Aotearoa New Zealand

Guidelines for the Prevention, Diagnosis, and Management of Acute Rheumatic Fever and Rheumatic Heart Disease

2024 Update

Dedication

These updated Rheumatic Fever and Rheumatic Heart Disease Guidelines are dedicated to **Professor Diana Lennon**, whose unwavering commitment and lifelong contributions to the prevention, diagnosis, and management of rheumatic fever and rheumatic heart disease have shaped care and improved outcomes for countless individuals and their whānau in Aotearoa New Zealand and the Pacific region. Professor Diana Lennon strongly advocated for rheumatic fever registers and championed nurse-led community-based penicillin prophylaxis programmes.

She co-led the first two editions of the Aotearoa New Zealand guidelines and would be heartened to see a new generation of academics and clinicians contributing to this third iteration, alongside those who have dedicated many years to this important work.

Professor Diana Lennon's pioneering research, clinical expertise, and dedication to advancing equitable health outcomes have inspired generations of healthcare professionals in Aotearoa New Zealand and around the world.

Her understanding of the epidemiology and the social determinants of child health continue to influence our ongoing determination to prevent and control rheumatic fever and rheumatic heart disease, in Aotearoa New Zealand and globally.

Professor Diana Lennon

Acknowledgements

The coloured illustrations in the guidelines were designed by MITA Creative Ltd. The illustrations were inspired by Rehua, an Atua (celestial Māori deity), who is a powerful healer. The images represent kawakawa, a native plant of Aotearoa commonly used for healing in rongoā Māori (Māori healing systems). The colour variations used symbolise the range of colours of kawakawa plants.

Thank you to the Starship Foundation for helping to support the update of these guidelines.

Starship Foundation

Disclaimer

These guidelines were commissioned by Health New Zealand | Te Whatu Ora and produced by Te Kupenga Hauora Māori at the University of Auckland. Oversight was provided by the guidelines Steering Committee, informed by Māori and Pacific governance groups. These guidelines are designed to be used by healthcare professionals involved in acute rheumatic fever and rheumatic heart disease care and management. They are not a substitute for clinical advice and healthcare professionals using these guidelines should exercise their professional judgement in providing care and treatment to any particular patient.

Suggested citation

Anderson, A., Mills, C., Rentta, N., Devlin, G., Farrant, B., Kando, I., Kistanna, D., Middleton, F., Wheeler, M., Wilson, N., & Webb, R. 2025. *Aotearoa New Zealand Guidelines for the Prevention, Diagnosis, and Management of Acute Rheumatic Fever and Rheumatic Heart Disease: 2024 Update*. Wellington: Health New Zealand | Te Whatu Ora.

Published in May 2025 by Health New Zealand | Te Whatu Ora
PO Box 793, Wellington 6140, New Zealand

ISBN 978-1-991139-26-9 (online)

Endorsing organisations

Mihi

Manawa nei e, te huaki rangi	**Heartfulness in the subconscious**
Manawa nei e, te huaki papa	**Heartfulness in the conscious**
Hohou nuku te kokonga whare kia kitea	**To stimulate further what we understand**
Hohou rangi te kokonga ngākau kia rongohia	**To foster further what is yet to be understood**
Kauae rungatia, kauae rarotia	**Internalise it, externalise it**
Kia pūkawatia te mānehurangi	**So that it may imbue a new reality**
Mō Hine-ngākau, mō Tama-ngākau	**For her soul, for his soul**
Hei oranga tinana, hei oranga wairua	**For physical and spiritual wellbeing**
Tau te Mauri!	**Contentment in life anew**
Tau hā, tau ana!	**Bring forth vitality!**

Nā Te Tīma Māori, Rheumatic Fever Co-Design Initiative

Karakia gifted by Te Tīma Māori Rheumatic Fever Co-Design Initiative

Hūtia te rito o te harakeke	**If you pluck out the centre shoot of the flaxbush**
Kei hea te kōmako, e ko?	**Where will the bellbird sing?**
Kii mai koe ki ahau, he aha te mea nui o tēnei ao?	**What is the most important thing in the world?**
Māku e kii atu kia koe	**I will reply to you**
He tangata, he tangata, he tangata!	**It is people, it is people, it is people!**

Tēnā rā koutou katoa!

E rere haere ngā mihi maioha ki te hunga nei, ki te mana whenua o te rohe nei ko Ngāti Whātua ki Ōrākei, tēnā koutou.
Ka tuku mihi ki te rōpū whakahaere o te waka hauora ko te National Hauora Coalition.
E mihi ana ki a Pū Manawa Aotearoa, Te Whatu Ora, te mātanga rūmātiki me te katoa e tautoko ana i te kaupapa nei.
Nōku te waimārie ke te tuhi tēnei mihi ki a koutou.

Mauriora kia tātou katoa.

I am honoured to have the opportunity to write this mihi drawing from my lived experience of acute rheumatic fever (ARF) and rheumatic heart disease (RHD), Chairperson of the Māori governance group of Pū Manawa Aotearoa and a kaimahi with the National Hauora Coalition. At the National Hauora Coalition, we celebrate those who come before us, who believed in building a future where whānau Māori thrive. Our moemoeā, Mana whānau, whānau ora — prosperous families living well, reflect this commitment. Whānau voice is at the core of our being and steers our actions, from the insights we gather, to the way our programmes are designed, to how they are delivered.

As a person with lived experience of ARF and RHD, I have the great pleasure of sharing my journey in many spaces and contributing to various projects addressing these conditions. I was diagnosed with ARF at a very young age at St Mary's Hospital in Auckland and spent a considerable time in the Cardiology ward at Greenlane hospital. I found this experience quite scary. I was not able to attend my school. I was confined to a wheelchair or bedrest most of the time that I was in hospital. In 1995, I had a heart operation to replace the mitral valve of my heart, luckily, the valve was able to be repaired by Dr Allan Kerr and his team and did not need to be replaced.

In November 2023, I was asked by the clinical trials team at Middlemore Hospital to attend the World Congress of Rheumatic Heart Disease conference in Abu Dhabi as a lived experience Champion of Change for ARF and RHD. I was able to connect with other Champions for Change from Australia, Namibia, Nepal and Fiji.

These experiences are a reminder of how far we have come in addressing ARF and RHD and how much further we still need to go to eliminate these preventable diseases, especially for our Māori and Pacific communities. We are still on a journey, and these guidelines will serve as a critical tool to guide clinicians and health professionals in their work to support whānau affected by ARF and RHD.

The whakataukī I have used at the beginning asks what is the most important thing in this world? The reply is it is people, it is people, it is people! This is a favourite of mine as I reflect on this mahi which has bought us altogether in this space. It is you, it is me, it is us.

Ma Ihoa o ngā Mano hei manaaki, hei tiaki.

Tēnā koutou katoa.

Shannon Leilua
Expert Patient Advisor for ARF and RHD, Pū Manawa Māori Governance Group Chair and Pou Tikanga at the National Hauora Coalition

Contents

Introductory pages

Chapters

List of figures

List of tables

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Declaration

No conflicts of interest were apparent in the development of this guideline update.

Scope and purpose

These 2024 guidelines are an update of national acute rheumatic fever (ARF) and rheumatic heart disease (RHD) guidelines published in 2014 and 2019.

These ARF and RHD guidelines have been developed under a culturally responsive framework, guided by Māori and Pacific governance groups and people with lived experience of ARF and RHD.

The 2024 ARF and RHD guidelines include:

- Chapters which provide updated clinical guidance about the diagnosis and management of sore throat, ARF and RHD.
- Background information about the epidemiology, pathogenesis and risk factors for ARF and RHD in Aotearoa.
- Guidance for clinicians about culturally responsive care and working with rangatahi (young people).

Notable changes in this 2024 edition include:

- Inclusion of a sore throat chapter (previously this was a separate document).
- New chapters on cultural responsiveness, ARF pathogenesis, Strep A infection, ARF and RHD: risk factors, social determinants of health and primordial prevention, administration of intramuscular benzathine penicillin, diagnosis of RHD, RHD and pregnancy and developmentally appropriate care for rangatahi.

The 2024 ARF and RHD guidelines are designed to be used by healthcare professionals from a range of clinical backgrounds, as well as those involved in the design and delivery of ARF and RHD healthcare and prevention services.

Hyperlinks are included to enable users to navigate to relevant content quickly.

Additionally, a summary guide has been produced. Key information has been extracted from each chapter in order to provide quick access to key changes, clinical recommendations and tables. The summary guide can be accessed here.

Māori Governance Group — positioning statement

E ngā reo, e ngā mana, e ngā rau rangatira mā, o ngā waka, tēnā koutou, tēnā koutou, tēnā koutou katoa.

The Māori Governance Group was established to ensure that mana motuhake (sovereignty) and kāwanatanga (governance) by tangata whenua operate in the development and content of the Aotearoa New Zealand Guidelines for Prevention, Diagnosis and Management of Acute Rheumatic Fever and Rheumatic Heart Disease. The group honours and oversees the implementation of Te Tiriti o Waitangi across the guidelines and upholds a rights-based partnership between tangata whenua and tangata Tiriti, particularly for Pacific peoples in Aotearoa who are also inequitably affected by acute rheumatic fever and rheumatic heart disease. The group further oversees a strengths-based (non-deficit) framing of the guidelines, inclusion of high quality, evidence-based and culturally safe and responsive clinical practice, and inclusion and monitoring of ethnicity data to benefit whānau Māori.

Members

Dr Rachel Brown, Professor Ricci Harris, Donna Kielar, Jane Kelly, Dr Lisa Kremer, Shannon Leilua, Dr Rawiri Jansen, Dr Ranche Johnson, Professor Bridget Robson, Dr Karen Wright.

Pacific Governance Group — positioning statement

Addressing the inequitable burden of acute rheumatic fever borne by Pacific communities is a key priority for the Aotearoa New Zealand Guidelines for Prevention, Diagnosis and Management of Acute Rheumatic Fever and Rheumatic Heart Disease. The Pacific Governance Group was formed to ensure that Pacific peoples' perspectives, knowledge, clinical expertise, and lived experiences were available to support the development of the refreshed guidelines to achieve positive health outcomes in Pacific communities. The 2024 guidelines use the term 'Pacific peoples' to refer to diverse communities in Aotearoa with ancestral ties to the Pacific Islands. The Pacific Governance Group acknowledges Māori as tangata whenua of Aotearoa and strongly commits to upholding Te Tiriti o Waitangi. The Group endorses and supports the leadership role of the Māori Governance Group. The Group also endorses and supports the focus on top-quality practice that is strengths-based, culturally safe and responsive, and informed by high-quality evidence relevant to Pacific peoples' health.

Members

Dr Sainimere Boladuadua, Dr Florina Chan Mow, Dr Adam Dennison, Monleigh Ikiua, Vivien Pole, Dr Debbie Suitafa Ryan-Nicholson.

Grading methodology

Targeted literature reviews of new evidence (since 2012) were conducted, after identifying key questions with the Steering Committee and expert review groups. Evidence from published systematic reviews was privileged, where available. Search strategies were carried out with reference librarian support and are available on request. Additional searches were carried out as the need arose and included review of current international clinical practice guidelines on sore throat, ARF and RHD; and discussion with subject matter experts and chapter review groups. Recommendations in these guidelines are based on the available evidence of both benefits and harms of an intervention, and an assessment of the balance. However, clinical decision-making requires consideration of more than evidence alone and should be individualised in discussion with the person and/or their whānau.

The grading used in this 2024 update is adapted from that previously used in the '2019 Rheumatic Fever New Zealand Guidelines for Group A Streptococcal Sore Throat Management Guideline: 2019 Update'.[1, 2, 3]

Evidence levels		Recommendation Grading	
I	Evidence obtained from a systematic review of all relevant randomised controlled trials (RCT)	A	Rich body of high-quality RCT data
II	Evidence obtained from at least one properly designed randomised controlled trial	B	Limited body of RCT data or high-quality non-RCT data
III-1	Evidence obtained from well-designed pseudo-randomised controlled trials (alternate allocation or some other method)	C	Limited evidence
III-2	Evidence obtained from comparative studies with concurrent controls and allocation not randomised (cohort studies), case-control studies or interrupted time series with a control group	D	Inadequate evidence, expert consensus
III-3	Evidence obtained from comparative studies with historical control, 2 or more single-arm studies or interrupted time series with a parallel control group		
III-4	Evidence obtained from case series, either post-test or pre- and post-test.		

References

1 Owens JK. Systematic reviews: Brief overview of methods, limitations, and resources. Nurse Author & Editor. 2021;31:69–72. [Cited 2024 July 16]. Available from: https://onlinelibrary.wiley.com/doi/pdf/10.1111/nae2.28

2 Page MJ, McKenzie JE, Bossuyt PM, Boutron I, Hoffmann TC, Mulrow CD, et al. The PRISMA 2020 statement: an updated guideline for reporting systematic reviews. BMJ (Online) 2021;372:n71. https://doi.org/10.1136/bmj.n71

3 National Heart Foundation of New Zealand. Guidelines for Group A Streptococcal Sore Throat Management Guideline: 2019 Update. Auckland: National Heart Foundation of New Zealand; 2019. [Cited 2024 May 7]. Available from: https://www.heartfoundation.org.nz/resources/group-a-streptococcal-sore-throat-management

List of acronyms and initialisms

Acronym or initialism	Meaning
ACC	American College of Cardiology
ACEi	Angiotensin-converting enzyme inhibitor
ADB; anti-DNase B	Anti-deoxyribonuclease B
AF	Atrial fibrillation
AHA	American Heart Association
AI	Artificial intelligence
AMOSS	Australasian Maternity Outcomes Surveillance System
AMVL	Anterior mitral valve leaflet
ANA	Anti-nuclear antibody
AOR	Adjusted odds ratio
APTT	Activated partial thromboplastin clotting time
AR	Aortic regurgitation
ARB	Angiotensin receptor blocker
ARF	Acute rheumatic fever
ARNI	Angiotensin receptor-neprilysin inhibitor
AS	Aortic stenosis
ASO; antiSLO	Anti-streptolysin O
AV	Aortic valve
AV	Atrioventricular
BPG	Benzathine Benzylpenicillin G
BMI	Body mass index
BMV	Balloon Mitral Valvuloplasty
BNP	B-type natriuretic peptide
CARM	Centre for Adverse Reactions Monitoring
CHADSVASc	Congestive heart failure, hypertension, age, diabetes, previous stroke/ transient ischemic attack or thromboembolism, vascular disease
CHD	Congenital heart disease

Acronym or initialism	Meaning
CI	Confidence interval
CMR	Cardiac magnetic resonance
CRP	C-reactive protein
CT	Computed tomography
DHB	District health board
DOAC	Direct oral anticoagulant
EACTS	European Association for Cardio-Thoracic Surgery
ECG	Electrocardiogram
Echo	Echocardiography or echocardiogram
EF	Ejection fraction
EOA	Effective orifice area
ESC	European Society of Cardiology
ESR	Erythrocyte sedimentation rate
iGAS	Group A streptococcus infection
GLS	Global longitudinal strain
GP	General practitioner
HEAT	Health equity and assessment tool
HHI	Healthy Homes Initiative
HLA	Human leukocyte antigen
IDSA	Infectious Diseases Society of America
IM	Intramuscular
INR	International normalised ratio
IQR	Interquartile range
IV	Intravenous
IVIg	Intravenous immunoglobulin
LMWH	Low molecular weight heparin
LV	Left ventricle
LVEF	Left ventricular ejection fraction

Acronym or initialism	Meaning
LVESD	Left ventricular end-systolic diameter
LVOT	Left ventricular outflow tract
MET	Metabolic equivalents
MPHV	Mechanical prosthetic heart valve
MPVT	Mechanical prosthetic valve thrombosis
MR	Mitral regurgitation
MRI	Magnetic resonance imaging
MS	Mitral stenosis
MV	Mitral valve
MVA	Mitral valve area
NAAT	Nucleic acid amplification tests
NASC	Needs Assessment and Service Co-ordination
NOAC	Non-vitamin K antagonist oral anticoagulant
NSAID	Nonsteroidal anti-inflammatory drug
NYHA	New York Heart Association
NZF	New Zealand Formulary
PANDAS	Paediatric auto-immune neuropsychiatric disorder associated with streptococcal infection
PBMV	Percutaneous balloon mitral valvuloplasty
PCR	Polymerase chain reaction
PET	Positron emission tomography
PMVL	Posterior mitral valve leaflet
PO	Oral administration
PSGN	Post-streptococcal glomerulonephritis
PSRA	Post-streptococcal reactive arthritis
RADT	Rapid antigen detection test
RCT	Randomised controlled trial
RFCCS	Rheumatic Fever Care Coordination System
RFPP	Rheumatic Fever Prevention Programme

Acronym or initialism	Meaning
RHD	Rheumatic heart disease
RR	Rate ratio
RV	Right ventricle
SAP	Secondary antibiotic prophylaxis
SAVR	Surgical aortic valve replacement
SDOH	Social determinants of health
SGLT2	Sodium-glucose-cotransporter-2
SVD	Structural valve degeneration
TAVR	Transcatheter aortic valve replacement
TKHM	Te Kupenga Hauora Māori
TR	Tricuspid regurgitation
WHF	World Heart Federation
WHO	World Health Organization
mWHO classification	modified World Health Organization
UFH	Unfractionated heparin
USCRS	UFMG Sydenham's chorea rating scale
VKA	Vitamin K antagonist
VTI	Velocity time integral

Te reo glossary

Te Reo Māori	English
Aotearoa	New Zealand
Hapori	Community or society
Hapū	Sub-tribe or kinship group (can also mean to be pregnant)
Hauora	Health
Hauora Māori	Māori health
He wā pai	A good time
Hinengaro hauora	Mental health
Hui	Meeting
Iwi	Tribe or extended kinship group or nation or people
Kaiāwhina	Helper, assistant, contributor, counsel, advocate
Kaimahi	Worker or employee
Kaitiaki	Guardians/caretakers
Kaupapa	Principle or policy
Kaupapa Māori	A critical, decolonising framework that prioritises Māori worldviews and avoids cultural deficit perspectives
Kaupapa pai	A genuine kaupapa (activity, process) — emphasises rangatahi connections responding to their desires and aspirations
Kāwanatanga	Governorship
Kawa whakaruruhau	Cultural safety
Kōhanga	Nest or nursery
Kōhanga reo	Early childhood care based on immersion in kaupapa Māori
Ko wai	A reciprocal connection — emphasises the importance of connections with people and beyond
Kura kaupapa	Primary school operating under Māori custom and using Māori as the medium of instruction
Mahi	Work
Mana motuhake	Sovereignty or self-determination
Mana Ōrite	Equal standing
Mana whakamārama	Equal explanatory power

Te Reo Māori	English
Mātauranga Māori	Māori knowledge or Māori understanding
Mauri	Life essence
Moemoeā	Dream or vision
Ōritetanga	Equality or equal opportunity
Pākehā	New Zealander of European descent
Pēpi	Baby or infant
Rangatahi	Teenager or young adult
Tamaiti	Children of Pacific peoples or child
Tamariki	Children
Tangata whenua	Indigenous people of Aotearoa
Tangata tiriti	People of the treaty. Refers to Non-Māori living in Aotearoa who are committed to building a truly Tiriti-centric nation, grounded in the principles and articles of Te Tiriti o Waitangi.
Taonga	Treasure or goods or possession
Te ao Māori	Māori world view
Te Pāti Māori	Māori political party
Te taiao	World or earth or environment or natural world or nature.
Te Tiriti o Waitangi	Te reo Māori version of the Treaty of Waitangi
Tikanga Māori	Māori customs, practices and procedures
Tinana	Physical body
Tino Rangatiratanga	Self-determination or sovereignty or autonomy or self-government
Wairua/wairuatanga	Spirit/spirituality
Wakahourua	Double-hulled canoe
Whakapapa	Genealogy
Whakataukī	Proverb saying from a person known who first used the proverb
Whānau	Extended family or to be born
Whānau Ora	Healthy families
Whanaungatanga	A relationship through shared experiences or kinship or sense of a family connection
Whanonga Pono	Values
Whenua	Local peoples

1

Cultural Responsiveness

In this chapter

1

"I think whānau voice should be, you know, or consumers and communities should have their voice heard, because so often the people that are affected [by streptococcus A] don't get asked."

Whānau Māori participant, 2022

Key changes

This is a new chapter.

Key points

- This chapter outlines key concepts of cultural responsiveness and cultural safety that are critical to services providing care for Māori and Pacific communities at risk of acute rheumatic fever (ARF) and rheumatic heart disease (RHD).
- Cultural responsiveness is crucial for achieving safe, equitable healthcare outcomes. It involves understanding equity in patient and whānau engagement and ensuring inclusive practices at all levels of healthcare.
- Applying cultural models of health in sore throat management, ARF and RHD service delivery and design promotes culturally safe and responsive care.
- Cultural safety requires critical self-reflection by healthcare professionals to address power dynamics, biases, and stereotypes that impact patient care.
- Upholding the articles of Te Tiriti o Waitangi is crucial in emphasising equitable engagement, leadership, and resource allocation for Māori (and Pacific) communities.
- Indigenous data sovereignty is integral to Indigenous rights and self-determination, empowering communities to control the collection, use, and protection of their data for their benefit.
- Collaborating with Māori and Pacific communities in designing sore throat, ARF, and RHD interventions is essential for tailoring culturally responsive care, ensuring services meet community contexts effectively.

Introduction

This chapter provides an overview of cultural responsiveness and offers some approaches for key elements of its practice (**Figure 1.1**). This chapter is designed to guide services focused on sore throat, ARF, and RHD, supporting a more equitable path for Māori and Pacific peoples in Aotearoa.

Cultural responsiveness is how health practitioners deliver culturally safe, equitable, and effective healthcare to patients and whānau.[1-3] Cultural responsiveness requires both individual health practitioners and broader health services to engage in critical reflective practices, shifting power from themselves to the communities they work with.[1]

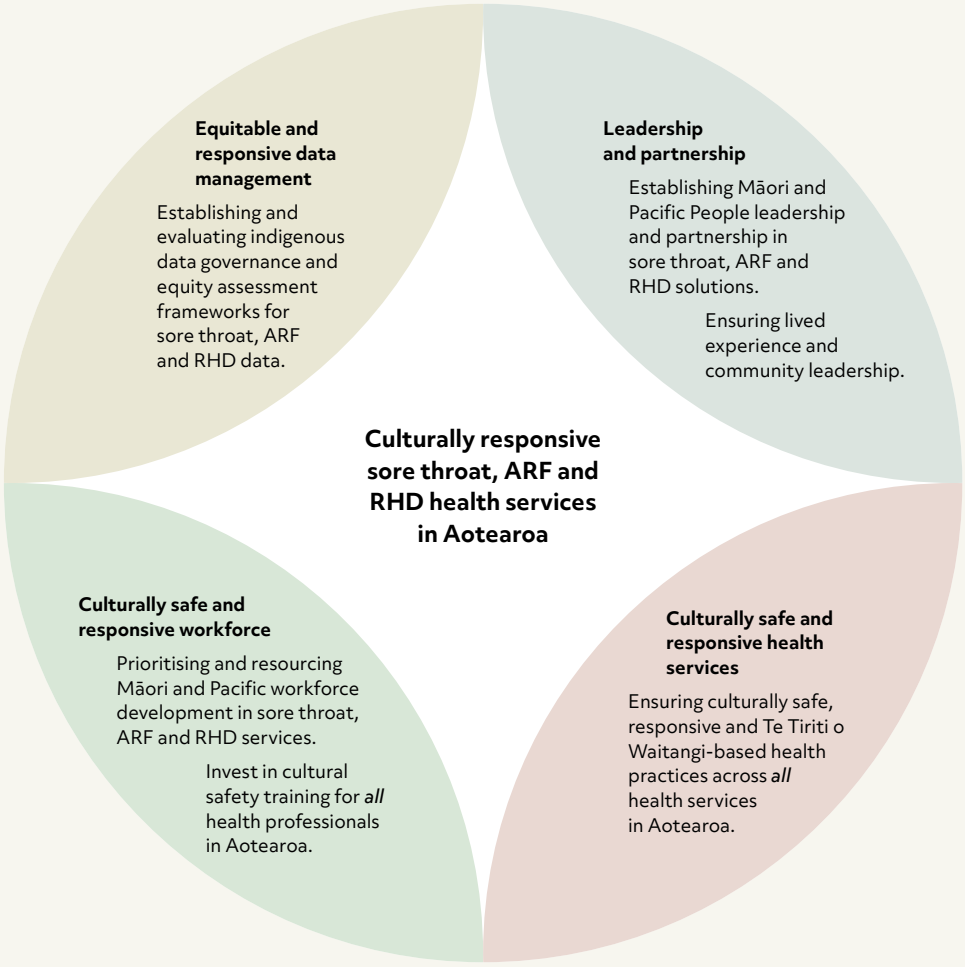

Figure 1.1. Key areas to focus on improving culturally responsive sore throat, acute rheumatic fever and rheumatic heart disease health services in Aotearoa

As described in Chapter 4: Epidemiology of Strep A Infections, Acute Rheumatic Fever and Rheumatic Heart Disease, Māori and Pacific peoples experience significant inequities in ARF and RHD in Aotearoa. Research with Māori and Pacific peoples in both conditions has revealed notable gaps in the practice of cultural responsiveness in healthcare.[4, 5] Key findings from these studies included experiences of differential and inequitable prescribing, diagnostic practices, racism, ageism, and misalignment of cultural values within health services.[6, 7]

Culturally unresponsive care can create barriers to engaging with health services, promote non-adherence to medication, and reduce access to and uptake of health information.[2, 5, 8] These barriers can cause delays in diagnosis and management of sore throat, ARF and RHD and reduce engagement with ARF and RHD care.[4, 5, 7]

Given the significant impact of culturally unresponsive care on people with ARF and RHD, there needs to be more focus and resourcing towards creating and maintaining cultural responsiveness in ARF and RHD services.

Kawa whakaruruhau/Cultural safety

Kawa whakaruruhau or cultural safety creates and maintains responsive, equitable, and safe healthcare and should be prioritised in health services for sore throat, ARF, and RHD management.[9-11] Cultural safety is a practice where healthcare professionals critically focus on themselves and the interpersonal power dynamics that occur with patients during engagement with health services.[9] Cultural safety is undertaken through ongoing critical self-reflection by health professionals, taking into consideration their values, beliefs, and associated biases and stereotypes. A key element of cultural safety is that health service users evaluate the 'safeness' of the health services.[10]

Health Navigator New Zealand and Health Literacy New Zealand have developed a useful resource for cultural safety training and support that could be used for health professionals working across sore throat, ARF, and RHD services: Cultural safety | Smstoolkit.[12] This is also available at: www.journeytowellbeing.nz.

The Manaaki Mana Strategy was developed to provide excellence in emergency care for Māori patients and whānau but could also be applied to sore throat, ARF, and RHD health services. This strategy focuses on cultural safety and provides a framework of actions to achieve culturally responsive care, including applying it to workplace and personal practice audits.[13]

Cultural safety is often used interchangeably with cultural competency. Despite both concepts prioritising culture, they are distinct practices:

- Cultural safety focuses on critical self-reflection to examine self-bias, culture, and practice.
- Cultural competency focuses on health professionals learning about the diverse cultures of those other than themselves.[11]

Cultural competency on its own has been critiqued as being a tick-box practice that can further perpetuate racialised stereotypes and bias through 'othering' diverse cultural groups.[9] Despite these concerns, cultural competency remains a dominant element of medical and health practice regulation and training programmes in Aotearoa.[9] Simmonds et al.[11] have developed an equity-based cultural safety training plan for medical colleges in Aotearoa. They consider cultural safety, cultural competency, and understandings of Hauora Māori (Māori health) as "separate but entwined pursuits, and all pertain to equitable outcomes and optimal health for Māori,"[11, p. 5] as noted in **Appendix 1**.

Given the importance of cultural safety to inform culturally responsive care for Māori and Pacific peoples, a key recommendation is to embed cultural safety training, practice, and evaluation into all services dealing with sore throat management, ARF, and RHD in Aotearoa.

Te Tiriti o Waitangi and health

Along with cultural safety, accountability is a crucial component of responsive health systems. The World Health Organization recommends that equitable health outcomes:

- Are grounded on the inclusion of diverse communities and stakeholders.
- Allow human rights to be actioned at national, regional, and local levels.[14]

Internationally and nationally, various declarations, legislative acts, and treaties recognise health as an essential right of people.[15] In Aotearoa, peoples' right to experience good health and freedom from discrimination in health services are protected by the New Zealand Bill of Rights Act 1990, Human Rights Act 1993, and Health and Disability Services Act 1994.[16]

Further, for Māori as tangata whenua (Indigenous people of Aotearoa), Te Tiriti o Waitangi is an essential framework to operationalise rights and responsive health services. Te Tiriti o Waitangi is a covenantal agreement between tangata whenua and the Crown. Te Tiriti o Waitangi recognises Māori as the original occupants of Aotearoa and is considered the founding document for rights and relationships between the Crown (including Pacific peoples and all other non-Māori representatives) and Māori.[15]

Various Te Tiriti o Waitangi frameworks are available to inform culturally responsive health services.[15, 17] Based on these frameworks, Table 1.1 proposes recommendations for health services managing sore throat, ARF, and RHD.

Table 1.1. Te Tiriti o Waitangi articles and recommendations for health services managing sore throat, acute rheumatic fever, and rheumatic heart disease

Te Tiriti o Waitangi article description	Recommendations for services dealing with sore throat, ARF and RHD
Preamble Outlines the purpose of Te Tiriti o Waitangi — with Crown intentions to protect Māori interests, provide British settlement, establish a government, and maintain peace and order.[15]	Practise these intentions by prioritising Te Tiriti o Waitangi and ensuring Māori are equal or lead parties in health policies, practices, and services.[17]
Article 1: Kāwanatanga This article provides the Crown the right to govern in Aotearoa.	Practise this article through fair, ethical, and just governance for Māori and Pacific peoples, ensuring equitable engagement and leadership of Māori in priority decision-making and health resourcing.[15, 17]
Article 2: Tino Rangatiratanga This article provides Māori the right to absolute sovereignty and autonomy over anything considered a taonga (treasure).	Practise this article by ensuring mana motuhake (sovereignty) of Māori through leadership, investment, professional development, resourcing, values, and worldviews.[15, 17]
Article 3: Ōritetanga This article guarantees Māori equal protection, rights, and opportunities to those of the Crown.	Practise this article by ensuring equitable access, experience, and outcomes for tangata whenua and Crown citizens — including Pacific peoples.[15]
Article 4: Wairuatanga This article upholds the rights of tangata whenua to practice spiritual and cultural beliefs, paradigms, and values.	Practise this article through inclusive processes, valuing diverse beliefs, and developing and investing in mātauranga Māori and Pacific paradigms.[17]

Indigenous data sovereignty

Indigenous data sovereignty is an important element of Indigenous peoples' rights. It refers to the collection, control, use, and protection of Indigenous data — including all forms of information and knowledge about and of Indigenous people, to benefit Indigenous people. Jansen describes Indigenous data sovereignty as Indigenous people having control of "knowing about ourselves. Knowing who we are, where we are, what we do, when we do it, how we do it or how much we do what we do".[18]

Kaupapa Māori epidemiology approaches to collecting and analysing data provide a methodology that ensures Indigenous rights, self-determination, and data sovereignty are upheld.[19] The approach draws on Kaupapa Māori theory — a critical, decolonising framework that prioritises Māori worldviews and places Māori at the centre of the enquiry. The theory also avoids explanations that have a cultural deficit perspective (a view that individuals from some cultural groups lack the ability to achieve only because of their cultural background).[18]

Paine et al.[19] provide four fundamental principles of Māori data sovereignty (see Table 1.2). These principles can uphold Indigenous data sovereignty when working with quantitative data, such as epidemiological data informing surveillance and services dealing with sore throat, ARF, or RHD.

Table 1.2. Four principles to upholding Māori data sovereignty with quantitative data[19]

1. Māori rights to monitor the Crown
This right includes the power to question and hold the Crown accountable for Māori wellbeing through their actions or inactions.
2. Māori rights to be counted
Māori have the right to be represented, not underrepresented, in datasets of importance using consistent and robust standards for collecting ethnicity data.
3. The right of Māori to have a powerful voice
Māori have the right to be represented as a people, not a sub-population or minority group. A key recommendation here is to avoid using approaches for sampling total populations that underrepresent Māori. The recommendation is to apply mana whakamārama or equal explanatory power instead to enable enough statistical power analyses for Māori.
4. The right to name racism and colonisation
Racism, stemming from colonisation, creates white privilege in Aotearoa and inequities for Māori. Naming these mechanisms allows for a transparent critique of power structures and for establishing equity-based interventions to address power imbalances.

As noted in **Table 1.2**, ethnicity data is important and should be collected using the standard ethnicity question in the Aotearoa Census (and most official national datasets), with processes following the ethnicity data protocols for the health sector.[20, 21] This will ensure high-quality ethnicity data for all groups, including Pacific peoples.

At a national level, Kukutai et al.[22] have developed a Māori data-governance model — Te Kāhui Raraunga, for public services and agencies in Aotearoa. Although Te Kāhui Raraunga has not yet been evaluated, it could help improve data governance and sovereignty for Māori. Te Kāhui Raraunga uses the metaphor of a wakahourua (double-hulled canoe) to embody Te Tiriti o Waitangi relationships. The model comprises three key phases (see **Figure 1.2**) to co-develop and implement data governance for Māori data by Māori.[22]

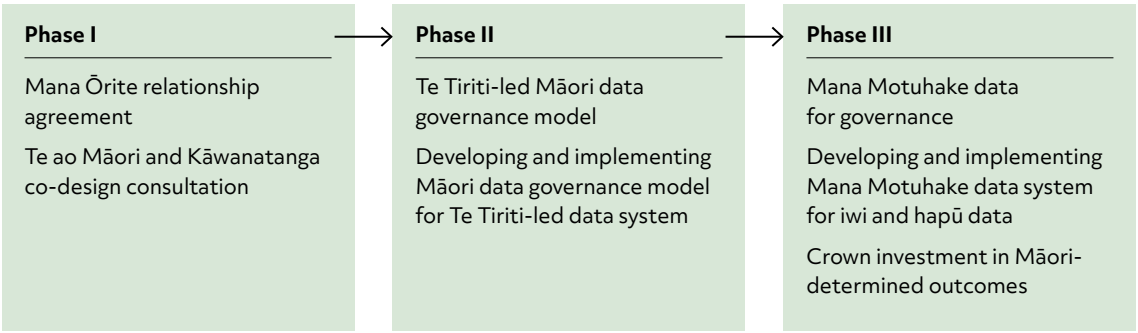

Figure 1.2. The three phases of Te Kāhui Raraunga data governance model[22]

Adapted from Kukutai et al. (2023).[22]

Resources to improve Pacific data sovereignty

Reviewing and improving data sovereignty processes for Pacific peoples has also seen some current changes. The Ministry for Pacific peoples[23] applied the Kakala model (a Tongan model based on a metaphor of garland making[24]) to compile a long-term briefing document as a resource to inform Pacific data equity across public services in Aotearoa. The document identifies three key areas that need improvement in the public sector and provides practical pathways to achieve these transformations (see **Figure 1.3**).

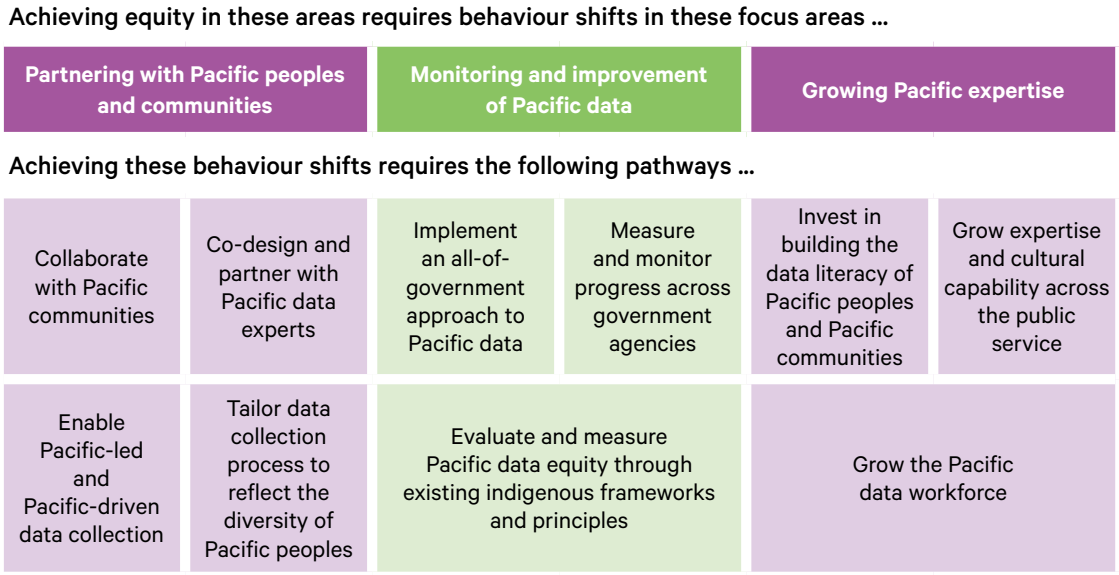

Figure 1.3. Framework to achieve data equity for Pacific peoples in Aotearoa[23]

Equity

Aiming to achieve equity is another foundational principle of cultural responsiveness.[1] Health equity is defined by Braveman and Gruskin (2003) as 'the absence of systematic disparities in health (or its social determinants) between more and less advantaged social groups'.[25]

The Equity of Health Care for Māori Framework was developed by the Ministry of Health in 2014 to guide health professionals and health services to achieve equitable healthcare for Māori. The framework proposed three key equity actions:

1. **Leadership:** Championing health services that deliver equity outcomes
2. **Knowledge:** Developing knowledge to deliver and monitor quality, equitable health services effectively
3. **Commitment:** Providing health services that meet the healthcare contexts and aspirations for Māori.

The Health Equity and Assessment Tool (HEAT) is another resource developed in Aotearoa to help health services, policies, and programmes become more equitable through evaluating 10 key questions (see **Table 1.3**).[26] HEAT can be used alongside other culturally responsive or rights-based frameworks, and Māori and other community stakeholders should be involved in applying the tool.[26] This tool or similar equity assessment processes should be included in all sore throat management, ARF, and RHD health service, policy, or programme planning.

Table 1.3. Key questions used for evaluative consideration in the Health Equity Assessment Tool

Key questions
1. What inequities exist in relation to the health issue under consideration?
2. Who is most advantaged and how?
3. How did the inequities occur? What are the mechanisms by which the inequities were created, maintained or increased?
4. Where/how will you intervene to tackle this issue?
5. How will you improve Māori and Pacific health outcomes and reduce health inequities experienced by Māori and Pacific peoples?
6. How could this intervention affect health inequities?
7. Who will benefit most?
8. What might the unintended consequences be?
9. What will you do to ensure the intervention reduces inequities?
10. How will you know if inequities have been reduced?

Adapted from the HEAT planning tool.[26]

Māori and Pacific leadership and workforce development

To fully achieve cultural responsiveness across all levels of primary and secondary ARF and RHD health services, Māori and Pacific leadership and partnership should be established, with representation from Māori and Pacific health professionals, health services and providers, iwi (tribe), hapū (sub-tribe), hapori (community) and people with lived experience of ARF or RHD.[27] Resourcing and prioritising the workforce development of Māori and Pacific peoples is critical to achieving successful and responsive health services for sore throat, ARF, and RHD in Aotearoa.[5] Only then can authentic representation of Māori and Pacific peoples in health services become a reality.

Ethnic representative workforces improve health outcomes by creating safe environments and allowing rapport, trust, and better engagement for service users. Such work environments can also buffer racism.[28-30] To recognise the importance of Māori and Pacific workforce development, several universities in Aotearoa have equity-based initiatives in place to increase the representation of Māori and Pacific health professionals in Aotearoa.[31, 32] Continued support for such initiatives should be prioritised in health education and sore throat, ARF, and RHD health delivery sectors in Aotearoa.

Māori and Pacific models of healthcare

In addition to Māori and Pacific leadership and partnership, the application of cultural models of health in sore throat management, ARF and RHD service delivery and design can also promote culturally safe and responsive care.[33] The hui process is an example of a cultural model that promotes engagement between Māori and health professionals by integrating cultural beliefs, values, and practices in clinical consultations.[33] This model can be applied to any clinical context, including services focused on sore throat management, ARF, and RHD.

At a service level, Mana Tū is a Māori-led diabetes model. Mana Tū was co-designed with diabetes patients, health professionals, and Māori providers and is underpinned by whānau ora and Kaupapa Māori principles.[34] The model could also be adapted to services focused on ARF and RHD.

As with Māori health models, several Pacific health models have been applied in Aotearoa. The Soālaupule framework is founded on a Samoan collective decision-making process and was designed by the Rheumatic Fever Co-Design Initiative Samoan Team. The framework aims to support health professionals in providing better health outcomes for Pacific peoples with a focus on ARF management, including secondary prophylaxis. The model aims to create "equity by sharing power and decision-making" and can be accessed as an online course at www.goodfellowunit.org/group/262.

The Yavu framework is a Pacific community engagement tool recommended by the Ministry for Pacific peoples. Yavu is based on the Fijian notion of Yavusa (the foundation of human essence, identity, and origin).[35] Similarly, the Kakala model can also be used as an engagement framework that draws on key cultural values and processes to promote engagement between the health sector and Pacific peoples to improve health outcomes.[35, 36] Seitapu is a framework (based on the Tongan metaphor of healing oil) developed within mental health and addiction services to improve engagement and health outcomes for Pacific peoples.[37] These models could be implemented within sore throat, ARF, and RHD services for specific Pacific communities to improve the cultural responsiveness of health services.

Many pan-Pacific models based on shared values of relationality, collectivism, and spirituality are used in health contexts and the 'by Pacific for Pacific' approach has been demonstrated to be of value for Pacific peoples.[36, 38] However, Naepi[39] critiques this approach and argues it can homogenise the diversity seen within and between Pacific ethnic groups that each have distinct histories, languages, and cultures.

Agnew et al.[36] particularly note the diversity within groups, emphasising generational and migrant contexts. They caution the use of such models that can exclude specific Pacific groups such as those that "adopt an island-born adult emphasis [that] can exclude Aotearoa-born youth".[36] Using cultural models along with diverse community consultation, governance, and the application of cultural safety could help address the homogenising effects of these models.[1, 9]

Summary of recommendations

- Establish Māori and Pacific leadership and partnership, e.g. including representation from Māori and Pacific health professionals, services and providers, iwi, hapū, hapori and people with lived experience of ARF or RHD.
- Implement cultural safety training and evaluation for all sore throat, ARF, and RHD health services.
- Resource and prioritise the workforce development of Māori and Pacific peoples across sore throat, ARF, and RHD health services.
- Apply Māori and Pacific health engagement and delivery models in sore throat, ARF, and RHD services, e.g. hui process and Soālapule.
- Prioritise Te Tiriti o Waitangi and implement its key articles in sore throat, ARF, and RHD health service planning and practice.
- Embed and evaluate Indigenous data sovereignty models when using or monitoring Māori and Pacific health data.
- Ensure ethnicity data is collected using the standard ethnicity question in the Aotearoa Census for all sore throat, ARF, and RHD data.
- Use HEAT or similar equity assessment tools in all sore throat, ARF, and RHD health service, policy and programme planning.

Appendix

Appendix 1. Definitions of cultural safety, cultural competency, and Hauora Māori

Cultural safety	Cultural competence	Hauora Māori[9]
Culturally safe medical practitioners engage in the ongoing development of critical consciousness. Developing such consciousness involves self-reflection on their own biases, attitudes, assumptions, stereotypes, prejudices, structures, and characteristics that may affect their practice. Medical practitioners who are culturally safe: • examine and redress power relationships in consultations, with colleagues, and within the healthcare ecosystem • commit to transformative action internally, horizontally, and vertically. Medical practitioners who are culturally safe ensure that cultural safety is defined by the patients, whānau, and communities they serve.	Culturally competent medical practitioners are committed to the ongoing development of the knowledge and skills to work effectively within cross-cultural contexts. They recognise that the definition of culture is wider than ethnic understandings and includes other social groups defined by their behaviours, beliefs, and values. Culturally competent medical practitioners: • accommodate for the cultural preferences of patients, whānau, and communities • know cultural protocols, beliefs, and language. They use these competencies to facilitate engagement with patients during clinical encounters. Culturally competent medical practitioners have the communication skills and confidence to ask about cultural expectations and traditional practices, including the correct pronunciation of names.	Medical practitioners: • have knowledge of the historical and contemporary Māori health situation • use Māori health models within clinical practice • engage appropriately with Māori patients, whānau, and communities • are familiar with te reo Māori and tikanga Māori • are familiar with the diversity of Māori beliefs, values, and experiences. Health is considered a priority of the collective rather than the individual. Health is viewed holistically, incorporating: • physical, mental, emotional, spiritual, and whānau dimensions • the relationship with whenua and environment.

References

1. Reid P, Paine SJ, Curtis E, Jones R, Anderson A, Willing E, et al. Achieving health equity in Aotearoa: strengthening responsiveness to Māori in health research. *New Zealand Medical Journal*. 2017;130(1465):96–103.
2. Wilson D, Heaslip V, Jackson D. Improving equity and cultural responsiveness with marginalised communities: understanding competing worldviews. *Journal of Clinical Nursing*. 2018;27(19-20):3810–3819. https://doi.org/10.1111/jocn.14546
3. Indigenous Allied Health Australia. Cultural responsiveness in action: an IAHA framework. 2015. https://iaha.com.au/workforce-support/training-and-development/cultural-responsiveness-in-action-training/ (Accessed February 18 2025).
4. Anderson A, Mills C, Eggleton K. Whānau perceptions and experiences of acute rheumatic fever diagnosis for Māori in Northland, New Zealand. *New Zealand Medical Journal*. 2017;130(1465):80–88.
5. Anderson A, Peat B, Ryland J, Ofanoa M, Burgess H, Malungahu G, et al. Mismatches between health service delivery and community expectations in the provision of secondary prophylaxis for rheumatic fever in New Zealand. *Australian and New Zealand Journal of Public Health*. 2019;43(3):294–299. https://doi.org/10.1111/1753-6405.12890
6. Shetty A, Mills C, Eggleton K. Primary care management of group A streptococcal pharyngitis in Northland. *Journal of Primary Health Care*. 2014;6(3). https://doi.org/10.1071/hc14189
7. Anderson A, Spray J. Beyond awareness: Towards a critically conscious health promotion for rheumatic fever in Aotearoa, New Zealand. *Social Science and Medicine*. 2020;247:112798. https://doi.org/10.1016/j.socscimed.2020.112798
8. Minnican C, O'Toole G. Exploring the incidence of culturally responsive communication in Australian healthcare: the first rapid review on this concept. *BMC Health Services Research*. 2020;20(1):20. https://doi.org/10.1186/s12913-019-4859-6
9. Curtis E, Jones R, Tipene-Leach D, Walker C, Loring B, Paine SJ, et al. Why cultural safety rather than cultural competency is required to achieve health equity: a literature review and recommended definition. *International Journal for Equity in Health*. 2019;18(1):174. https://doi.org/10.1186/s12939-019-1082-3
10. Ramsden I. Towards cultural safety. In: Wepa D, editor. Cultural safety in Aotearoa New Zealand. 2nd ed. Melbourne: Cambridge University Press; 2015. p. 5–25.
11. Simmonds S, Carter M, Haggie H, Mills V, Lyndon M, Tipene-Leach D. Cultural safety training plan for vocational medicine in Aotearoa. New Zealand: Te ORA and the Council of Medical Colleges; 2023. https://www.cmc.org.nz/media/4xmpx1dz/cultural-safety-training-plan-for-vocational-medicine-in-aotearoa.pdf (Accessed December 16 2024).
12. Health Navigator New Zealand. Cultural safety. 2019. https://www.smstoolkit.nz/cultural-safety (Accessed February 18 2025).
13. Australasian College for Emergency Care. Te rautaki manaaki mana: excellence in emergency care for Māori 2022–2025. 2022. https://acem.org.au/Content-Sources/Advancing-Emergency-Medicine/Cultural-safety/Aotearoa-Manaaki-Mana-Strategy/Te-Manaaki-Mana-Strategy (Accessed December 17 2025).

14. Hammonds R, Hanefeld J, Ooms G. Accountability as a driver of health equity. Copenhagen: WHO Regional Office for Europe; 2019.

15. Reid P. Good governance: the case of health equity. In: Tawhai V, Gray-Sharp K, editors. 'Always speaking': the Treaty of Waitangi and public policy. Wellington: Huia; 2011. p. 39–50.

16. Kukutai T, Cassim S, Clark V, Jones N, Mika J, Morar R. Māori data sovereignty and privacy. Hamilton: Te Ngira Institute for Population Research; 2023.

17. Came H, Kidd J, Heke D, McCreanor T. Te Tiriti o Waitangi compliance in regulated health practitioner competency documents in Aotearoa. *New Zealand Medical Journal*. 2021;134(1535):35–43.

18. Jansen R. Indigenous data sovereignty: a Māori health perspective. In: Kukutai T, Taylor J, editors. Indigenous data sovereignty: toward an agenda. Australia: ANU Press. p. 193–212.

19. Paine S, Cormack D, Reid P, Harris R, Robson B. Kaupapa Māori-informed approaches to support data rights and self-determination. In: Walter M, Kukutai T, Russo C, S, Rodriguez-Lonebear D, editors. Indigenous data sovereignty and policy. London: Routledge; 2021. p. 187–203.

20. Health New Zealand | Te Whatu Ora. HISO 10001:2017 ethnicity data protocols. Wellington: Health New Zealand | Te Whatu Ora. https://www.tewhatuora.govt.nz/health-services-and-programmes/digital-health/data-and-digital-standards/approved-standards/identity-standards (Accessed February 12 2025).

21. McLeod M, Harris R. Action plan for achieving high quality ethnicity data in the health and disability sector. A report for Te Aka Whai Ora: Māori Health Authority. Wellington: Te Rōpū Rangahau Hauora a Eru Pōmare; 2023.

22. Kukutai T, Campbell-Kamariera K, Mead A, Mikaere K, Moses C, Whitehead J, et al. Māori data governance model. Te Kāhui Raraunga. Tikanga in Technology discussion paper: Te Ngira Institute for Population Research; 2023.

23. Ministry for Pacific peoples. Improving Pacific data equity: opportunities to enhance Pacific wellbeing. Wellington: Ministry for Pacific peoples; 2022. https://www.mpp.govt.nz/assets/Reports/Long-Term-Insights-Briefing/MPP-LTIB-v3.pdf (Accessed February 15 2025).

24. Fua S. Kakala Research Framework: A garland in celebration of a decade of rethinking education. In: 'Otunuku M, Nabobo-Baba U, Johansson Fua S, editors. Of waves, winds and wonderful things: a decade of rethinking Pacific education. Fiji: USP Press; 2014. p. 50–72.

25. Braveman P, Gruskin S. Defining equity in health. *Journal of Epidemiology and Community Health*. 2003;57(4):254–258.

26. Signal L, Martin J, Cram F, Robson B. The Health Equity Assessment Tool: a user's guide. Wellington: Ministry of Health; 2008.

27. Sporle A, Koea J. Maori responsiveness in health and medical research: key issues for researchers (part 1). *New Zealand Medical Journal*. 2004;117(1199):U997.

28. Mullane T, Harwood M, Warbrick I, Tane T, Anderson A. Understanding the workforce that supports Maori and Pacific peoples with type 2 diabetes to achieve better health outcomes. *BMC Health Services Research*. 2022;22(1):672. https://doi.org/10.1186/s12913-022-08057-4

29. Robson B, Harris R, editors. Hauora: Māori Standards of Health IV. A study of the years 2000–2005. Wellington: Te Rōpū Rangahau Hauora a Eru Pōmare; 2007.

30. Becares L, Nazroo J, Stafford M. The buffering effects of ethnic density on experienced racism and health. *Health & Place*. 2009;15(3):670–678. https://doi.org/10.1016/j.healthplace.2008.10.008

31. Curtis E, Stokes K, Wikaire E, Reid P. Recruiting via Hui-ā-Rohe: how the Whakapiki Ake Project has increased engagement with Māori students, their whānau (families) and communities. In: Mazel O, Ryan C, editors. LIME good practice case studies. 3. Melbourne: University of Melbourne; 2015. p. 16–23.

32. Sopoaga F, Zaharic T, Kokaua J, Ekeroma AJ, Murray G, van der Meer J. Pacific students undertaking the first year of health sciences at the University of Otago, and factors associated with academic performance. *New Zealand Medical Journal*. 2013;126(1384):96–108.

33. Lacey C, Huria T, Beckert L, Gilles M, Pitama S. The Hui Process: a framework to enhance the doctor-patient relationship with Māori. *New Zealand Medical Journal*. 2011;124(1347):72–78.

34. Harwood M, Tane T, Broome L, Carswell P, Selak V, Reid J, et al. Mana Tū: a whānau ora approach to type 2 diabetes. *New Zealand Medical Journal*. 2018;131(1485):76–83.

35. Ministry for Pacific peoples. Yavu: foundations of Pacific engagement tool. Wellington: Ministry for Pacific peoples; 2022. https://www.mpp.govt.nz/publications-resources/resources/yavu/ (Accessed February 11 2025).

36. Agnew F, Pulotu-Endemann F, Suaalii-Sauni T, Warren H, Wheeler A, Erick M, et al. Pacific Models of Mental Health Service Delivery in New Zealand ("PMMHSD") Project. Auckland: Health Research Council of New Zealand; 2004. https://www.leva.co.nz/uploads/files/resources/Pacific-Models-of-Mental-Health-Service-Delivery-in-New-Zealand-PMMHSD-Project.pdf (Accessed February 18 2025).

37. Le Va, Te Pou o Te Whakaaro Nui. Real skills plus Seitapu: working with Pacific peoples. Auckland: The National Centre of Mental Health Research, Information and Workplace Development; 2009. https://www.leva.co.nz/wp-content/uploads/2024/08/Lets-Get-Real-Real-Skills-Plus-Seitapu-Working-with-Pacific-Peoples.pdf (Accessed February 8 2025).

38. Suaalii-Sauni T, Wheeler A, Saafi E, Robinson G, Agnew F, Warren H, et al. Exploration of Pacific perspectives of Pacific models of mental health service delivery in New Zealand. *Pacific Health Dialog*. 2009;15(1):18–27.

39. Naepi S. Navigating the currents of kaupapa Māori and pan-Pacific research methodologies in Aotearoa New Zealand. *MAI Journal*. 2015;4:71–84.

2

Acute Rheumatic Fever Pathogenesis

In this chapter

"My kids have had Strep a couple of times. I actually took my child to the doctor last Wednesday. They took a throat swab and gave her antibiotics... But, yeah, that's pretty scary when I look at it. I didn't actually realise, like I heard of rheumatic fever, but then I never really realised how serious the heart part of it was."

Whānau with lived experience

Key changes

This new chapter describes current understanding of the disease mechanisms of acute rheumatic fever and rheumatic heart disease.

Key points

Strep A infection in the throat acts as a trigger for acute rheumatic fever (ARF) in susceptible individuals, leading to an exaggerated immune response. The immune system makes antibodies to fight Strep A infections (**Figure 2.1**). When this response becomes over-active, the immune system starts producing antibodies that may be damaging to parts of the body. This causes ARF which may cause heart damage (carditis), joint damage (arthritis), or inflammation in the brain (chorea).

Why this dangerous immune system imbalance occurs is unknown, but possible reasons are:

- Parts of the bacteria look similar to parts of our heart — as the immune system cannot tell the difference, it attacks the heart while trying to attack the bacteria.
- The bacteria change the shape of heart tissue, making the immune system think the heart is foreign and needs to be attacked.

Over time, repeated Strep A infections may cause unbalanced immune responses that lead to permanent damage and scarring of the heart valves, known as rheumatic heart disease (RHD). In severe cases, this damage means the heart cannot work properly and may lead to congestive heart failure.

Finding effective treatments has been difficult because there is limited understanding of how infection causes the immune system to become unbalanced and attack the heart. Researchers are urgently working to find out how infections cause this ongoing injury and scarring so that effective treatments to limit heart damage can be found.

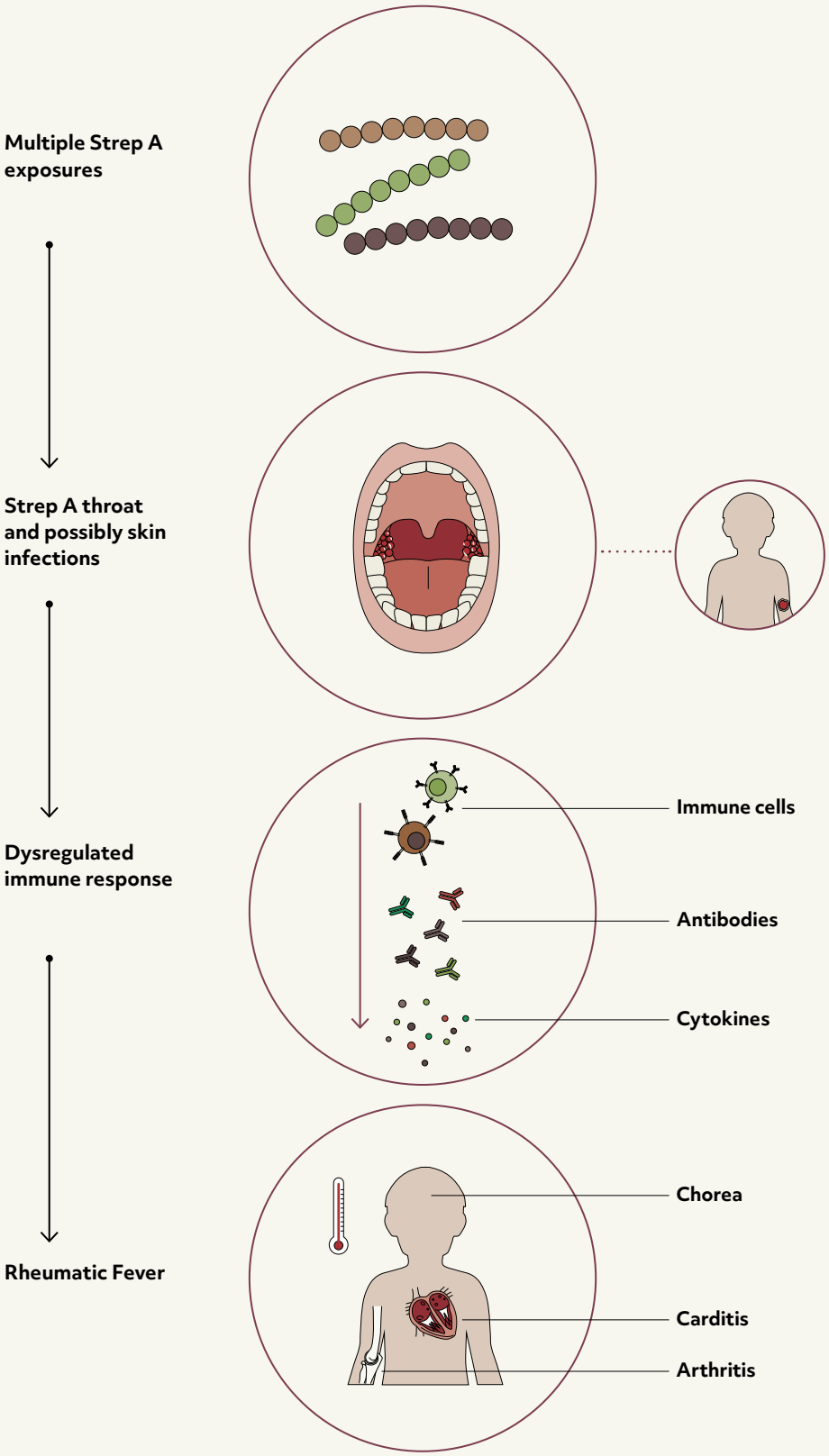

Figure 2.1. From infection to acute rheumatic fever

Adapted from an image created with BioRender.com.

Introduction

Strep A and acute rheumatic fever

Streptococcus pyogenes, also known as Strep A, Group A Strep, and GAS, is the bacterium that triggers ARF. It only infects humans. It spreads primarily by respiratory droplets but also via nasal secretions, skin-to-skin contact, and occasionally via food.[1, 2] Older studies[3] and a more recent study in the United Kingdom indicate that airborne transmission of Strep A is also possible.[2, 4]

Infection with Strep A can cause a spectrum of illnesses, from sore throat to invasive Strep A sepsis (also known as iGAS) to immune-mediated diseases such as ARF. Strep A can also be carried by people who are not ill but who can still pass Strep A on to others. This is known as 'carriage.'[5]

ARF is a multi-organ inflammatory (autoimmune) condition that may follow Strep A infection in susceptible individuals.

A strong epidemiological association and causal relationship exists between Strep A throat infections (pharyngitis or sore throat) and ARF.[6-10] However, the pathogenesis of ARF remains incompletely understood. For example, why are some people more likely than others to develop ARF? What causes ARF to progress to RHD?

An epidemiological association between Strep A skin infections and increased risk of ARF has been demonstrated in some Australian and Aotearoa studies, however, a causal relationship has not been proven.[11-16] A recent study in the Northern Territory of Australia highlighted the complexities of Strep A transmission dynamics and found asymptomatic throat infection to be an important reservoir of infection.[17]

Similarly, the contribution of asymptomatic Strep A infection[4, 18] and infections with other Streptococci (groups C and G) infections to ARF and RHD pathogenesis is uncertain.[1, 19-21]

ARF presents with various features, including carditis, arthritis, and chorea. Following ARF, progression to the chronic form of the disease, RHD occurs in approximately two-thirds and is characterised by persistent inflammatory damage to cardiac valves. RHD can cause heart failure and other complications.

The pathogenesis of ARF is characterised by an immune response to Strep A infection that becomes dysregulated and self-reactive.[22] The precise mechanisms leading to self-reactivity and how this causes disease manifestations and progression to chronic RHD are not well understood.

Infectious burden could be linked to the initiation of acute rheumatic fever

Understanding the pathogenesis of ARF requires knowing why only some Strep A infections in some tamariki (children) may cause disease.[23]

Historically, only a small number of rheumatogenic Strep A strains were considered capable of causing ARF.[24] However, contemporary analyses have shown a broad range of Strep A strains are associated with ARF, with classic rheumatogenic strains comprising only a small fraction.[25]

Another explanation focuses on the infectious burden. Tamariki may develop skin and throat infections from an early age, but ARF is extremely uncommon in preschool-aged tamariki. Two studies present compelling evidence that a high Strep A infectious burden (repeated infections) needs to prime the immune system before self-reactivity can occur. The number of priming infections required to 'tip' the immune system into ARF is currently unclear. Research in Aotearoa found serological evidence that ARF patients had experienced multiple Strep A infections before ARF diagnosis compared to demographically and ethnically matched healthy controls.[26] Another serological study compared the antibody response to Strep A in ARF patients with tamariki from the general Auckland population with superficial Strep A infections. ARF patients showed increased breadth (antibodies reacting with more Strep A antigens) and magnitude (higher levels of these antibodies) of antibody responses.[27]

Mimicry and collagen-driven damage might trigger self-reactivity

The traditional hypothesis for the initiation of self-reactivity is molecular mimicry. Proteins (M protein) and carbohydrates (group A carbohydrate) on the surface of Strep A share regions of similarity with self-molecules in the heart (myosin and laminin) and brain (dopamine receptors).[28] This molecular similarity is thought to allow cross-reactive antibodies and immune cells (T-cells) to infiltrate tissues.

Another hypothesis suggests that during infection, Strep A colonisation disrupts host connective tissue, exposing previously hidden regions of self-molecules such as collagen (cryptic epitopes). This exposure may trigger the production of anti-collagen auto-antibodies that can potentially initiate tissue damage.[29]

Mimicry and collagen-driven damage, as well as other mechanisms yet to be described, may contribute to self-reactivity in ARF.

Immune-mediated damage and symptoms

Epitope spreading occurs when the immune system targets molecules beyond those that initiate an immune response. Epitope spreading likely worsens the autoimmune response in ARF and contributes to the presence of immune molecules and cells that infiltrate cardiac tissue and cause immune-mediated tissue damage.[30, 31]

However, the exact mechanisms that worsen this damage and cause symptoms are not fully understood. This has hindered efforts to identify effective immunomodulatory therapy that could interrupt disease progression and improve clinical outcomes for people affected by ARF and RHD. Recent research points to the pathological involvement of a highly potent antibody subtype (IgG3),[32] migration of inflammatory T-cells into cardiac lesions,[33, 34] and proinflammatory cytokines (immune signalling molecules) such as IL-6 and TNF-a.[35]

Inflammation can affect heart valve function

Inflammatory cardiac lesions resulting from immune-mediated damage are characterised by excessive connective tissue production — the pathological development of blood vessels and fibrosis.[36] These lesions are associated with increased expression of the pro-fibrotic cytokine TGFβ and mainly occur in the mitral and aortic valves that experience the highest pressure gradients.[37, 38]

The inflammatory infiltrate in valvular lesions can cause fibrotic scars that impair valvular function, leading to heart failure associated with RHD. This outcome underscores the need to better understand the processes that cause immune-mediated tissue damage in ARF and RHD.

References

1. Avire NJ, Whiley H, Ross K. A review of streptococcus pyogenes: public health risk factors, prevention and control. *Pathogens*. 2021;10(2). https://doi.org/10.3390/pathogens10020248

2. Barth DD, Daw J, Xu R, Enkel S, Pickering J, McRae T, et al. Modes of transmission and attack rates of group A atreptococcal infection: a protocol for a systematic review and meta-analysis. *Systematic Reviews*. 2021;10(1):90. https://doi.org/10.1186/s13643-021-01641-5

3. Hamburger M, Jr., Robertson OH. Expulsion of group A hemolytic streptococci in droplets and droplet nuclei by sneezing, coughing and talking. *American Journal of Medicine*. 1948;4(5):690–701. https://doi.org/10.1016/s0002-9343(48)90392-1

4. Cordery R, Purba AK, Begum L, Mills E, Mosavie M, Vieira A, et al. Frequency of transmission, asymptomatic shedding, and airborne spread of streptococcus pyogenes in schoolchildren exposed to scarlet fever: a prospective, longitudinal, multicohort, molecular epidemiological, contact-tracing study in England, UK. *Lancet Microbe*. 2022;3(5):e366–e375. https://doi.org/10.1016/S2666-5247(21)00332-3

5. RHDAustralia, Menzies School of Health Research. The 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (3.2 edition). 2022. https://www.rhdaustralia.org.au/arf-rhd-guidelines (Accessed December 16 2024).

6. Spinks A, Glasziou PP, Del Mar CB. Antibiotics for treatment of sore throat in children and adults. *Cochrane Database of Systematic Reviews*. 2021;12(12):CD000023. https://doi.org/10.1002/14651858.CD000023.pub5

7. Wessels MR. Clinical practice. Streptococcal pharyngitis. *New England Journal of Medicine*. 2011;364(7):648–655. https://doi.org/10.1056/NEJMcp1009126

8. Denny FW, Wannamaker LW, Brink WR, Rammelkamp CH, Jr., Custer EA. Prevention of rheumatic fever; treatment of the preceding streptococcic infection. *Journal of the American Medical Association*. 1950;143(2):151–153. https://doi.org/10.1001/jama.1950.02910370001001

9. Kerdemelidis M, Lennon DR, Arroll B, Peat B, Jarman J. The primary prevention of rheumatic fever. *Journal of Paediatrics and Child Health*. 2010;46(9):534–548. https://doi.org/10.1111/j.1440-1754.2010.01854.x

10. Robertson KA, Volmink JA, Mayosi BM. Antibiotics for the primary prevention of acute rheumatic fever: a meta-analysis. *BMC Cardiovascular Disorders*. 2005;5(1):11. https://doi.org/10.1186/1471-2261-5-11

11. Bennett J, Moreland NJ, Zhang J, Crane J, Sika-Paotonu D, Carapetis J, et al. Risk factors for group A streptococcal pharyngitis and skin infections: a case control study. *The Lancet Regional Health — Western Pacific*. 2022;26:100507. https://doi.org/10.1016/j.lanwpc.2022.100507

12. Williamson DA, Smeesters PR, Steer AC, Steemson JD, Ng AC, Proft T, et al. M-protein analysis of streptococcus pyogenes isolates associated with acute rheumatic fever in New Zealand. *Journal of Clinical Microbiology*. 2015;53(11):3618–3620. https://doi.org/10.1128/JCM.02129-15

13. Thomas S, Bennett J, Jack S, Oliver J, Purdie G, Upton A, et al. Descriptive analysis of group A streptococcus in skin swabs and acute rheumatic fever, Auckland, New Zealand, 2010–2016. *The Lancet Regional Health — Western Pacific*. 2021;8:100101. https://doi.org/10.1016/j.lanwpc.2021.100101

14. Oliver J, Bennett J, Thomas S, Zhang J, Pierse N, Moreland NJ, et al. Preceding group A streptococcus skin and throat infections are individually associated with acute rheumatic fever: evidence from New Zealand. *BMJ Global Health*. 2021;6(12). https://doi.org/10.1136/bmjgh-2021-007038

15. Parks T, Smeesters PR, Steer AC. Streptococcal skin infection and rheumatic heart disease. *Current Opinion in Infectious Diseases*. 2012;25(2):145–153. https://doi.org/10.1097/QCO.0b013e3283511d27

16. McDonald MI, Towers RJ, Andrews RM, Benger N, Currie BJ, Carapetis JR. Low rates of streptococcal pharyngitis and high rates of pyoderma in Australian aboriginal communities where acute rheumatic fever is hyperendemic. *Clinical Infectious Diseases*. 2006;43(6):683–689. https://doi.org/10.1086/506938

17. Lacey JA, Marcato AJ, Chisholm RH, Campbell PT, Zachreson C, Price DJ, et al. Evaluating the role of asymptomatic throat carriage of streptococcus pyogenes in impetigo transmission in remote Aboriginal communities in Northern Territory, Australia: a retrospective genomic analysis. *Lancet Microbe*. 2023;4(7):e524-e533. https://doi.org/10.1016/s2666-5247(23)00068-x

18. Heart Foundation of New Zealand. Guidelines for group A streptococcal sore throat management guideline: 2019 update. National Heart Foundation of New Zealand; 2019. https://www.heartfoundation.org.nz/resources/group-a-streptococcal-sore-throat-management (Accessed February 18 2025).

19. Dougherty S, Carapetis J, Zühlke L, Wilson N. Acute rheumatic fever and rheumatic heart disease. Amsterdam: Elsevier; 2020.

20. Auala T, Zavale BG, Mbakwem AC, Mocumbi AO. Acute rheumatic fever and rheumatic heart disease: highlighting the role of group A streptococcus in the global burden of cardiovascular disease. *Pathogens*. 2022;11(5). https://doi.org/10.3390/pathogens11050496

21. Bright PD, Mayosi BM, Martin WJ. An immunological perspective on rheumatic heart disease pathogenesis: more questions than answers. *Heart*. 2016;102(19):1527–1532. https://doi.org/10.1136/heartjnl-2015-309188

22. Cunningham MW. Rheumatic fever, autoimmunity, and molecular mimicry: the streptococcal connection. *International Reviews of Immunology*. 2014;33(4):314–329. https://doi.org/10.3109/08830185.2014.917411

23. Bryant PA, Robins-Browne R, Carapetis JR, Curtis N. Some of the people, some of the time: susceptibility to acute rheumatic fever. *Circulation*. 2009;119(5):742–753. https://doi.org/10.1161/CIRCULATIONAHA.108.792135

24. Stollerman GH. Rheumatogenic and nephritogenic streptococci. *Circulation*. 1971;43(6):915–921. https://doi.org/10.1161/01.cir.43.6.915

25. de Crombrugghe G, Baroux N, Botteaux A, Moreland NJ, Williamson DA, Steer AC, et al. The limitations of the rheumatogenic concept for group A streptococcus: Systematic review and genetic analysis. *Clinical Infectious Diseases*. 2020;70(7):1453–1460. https://doi.org/10.1093/cid/ciz425

26. Lorenz N, Ho TKC, McGregor R, Davies MR, Williamson DA, Gurney JK, et al. Serological profiling of group A streptococcus infections in acute rheumatic fever. *Clinical Infectious Diseases*. 2021;73(12):2322–2325. https://doi.org/10.1093/cid/ciab180

27. Whitcombe AL, McGregor R, Bennett J, Gurney JK, Williamson DA, Baker MG, et al. Increased breadth of group A streptococcus antibody responses in children with acute rheumatic fever compared to precursor pharyngitis and skin infections. *Journal of Infectious Diseases*. 2022;226(1):167–176. https://doi.org/10.1093/infdis/jiac043

28. Carapetis JR, Beaton A, Cunningham MW, Guilherme L, Karthikeyan G, Mayosi BM, et al. Acute rheumatic fever and rheumatic heart disease. *Nature Reviews Disease Primers*. 2016;2:15084. https://doi.org/10.1038/nrdp.2015.84

29. Tandon R, Sharma M, Chandrashekhar Y, Kotb M, Yacoub MH, Narula J. Revisiting the pathogenesis of rheumatic fever and carditis. *Nature Reviews: Cardiology*. 2013;10(3):171–177. https://doi.org/10.1038/nrcardio.2012.197

30. Kaplan MH, Bolande R, Rakita L, Blair J. Presence of bound immunoglobulins and complement in the myocardium in acute rheumatic fever. Association with cardiac failure. *New England Journal of Medicine*. 1964;271:637–645. https://doi.org/10.1056/NEJM196409242711301

31. Kemeny E, Grieve T, Marcus R, Sareli P, Zabriskie JB. Identification of mononuclear cells and T cell subsets in rheumatic valvulitis. *Clinical Immunology and Immunopathology*. 1989;52(2):225–237. https://doi.org/10.1016/0090-1229(89)90174-8

32. Chung AW, Ho TK, Hanson-Manful P, Tritscheller S, Raynes JM, Whitcombe AL, et al. Systems immunology reveals a linked IgG3-C4 response in patients with acute rheumatic fever. *Immunology and Cell Biology*. 2020;98(1):12–21. https://doi.org/10.1111/imcb.12298

33. Guilherme L, Cury P, Demarchi LM, Coelho V, Abel L, Lopez AP, et al. Rheumatic heart disease: proinflammatory cytokines play a role in the progression and maintenance of valvular lesions. *American Journal of Pathology*. 2004;165(5):1583–1591. https://doi.org/10.1016/S0002-9440(10)63415-3

34. Roberts S, Kosanke S, Terrence Dunn S, Jankelow D, Duran CM, Cunningham MW. Pathogenic mechanisms in rheumatic carditis: focus on valvular endothelium. *Journal of Infectious Diseases*. 2001;183(3):507–511. https://doi.org/10.1086/318076

35. Middleton FM, McGregor R, Webb RH, Wilson NJ, Moreland NJ. Cytokine imbalance in acute rheumatic fever and rheumatic heart disease: mechanisms and therapeutic implications. *Autoimmunity Reviews*. 2022;21(12):103209. https://doi.org/10.1016/j.autrev.2022.103209

36. Fae KC, Palacios SA, Nogueira LG, Oshiro SE, Demarchi LM, Bilate AM, et al. CXCL9/Mig mediates T cells recruitment to valvular tissue lesions of chronic rheumatic heart disease patients. *Inflammation*. 2013;36(4):800–811. https://doi.org/10.1007/s10753-013-9606-2

37. Kim L, Kim DK, Yang WI, Shin DH, Jung IM, Park HK, et al. Overexpression of transforming growth factor-beta 1 in the valvular fibrosis of chronic rheumatic heart disease. *Journal of Korean Medical Science*. 2008;23(1):41–48. https://doi.org/10.3346/jkms.2008.23.1.41

38. Karthikeyan G, Fung E, Foo RS. Alternative hypothesis to explain disease progression in rheumatic heart disease. *Circulation*. 2020;142(22):2091–2094. https://doi.org/10.1161/CIRCULATIONAHA.120.050955

3

Strep A Infection, Acute Rheumatic Fever and Rheumatic Heart Disease: Risk Factors, Social Determinants of Health and Primordial Prevention

In this chapter

"You have all these other issues going on that make you barely able to function as a parent or a person … I was like 'dude, I just had a baby and he's only six weeks old, my oldest son has just come out of surgery and is still really sick with rheumatic fever and I've got two toddlers and a partner but we haven't had enough time to find a house, you try finding one … who can find a house in Auckland to rent'?"

Mother of a tamaiti with ARF

Key changes

This new chapter addresses the social determinants of health (SDOH) that drive inequities in acute rheumatic fever (ARF) and rheumatic heart disease (RHD) incidence and outcomes. Efforts in Aotearoa to address the upstream drivers of ARF and RHD are described.

Key points

- Although the pathogenesis of ARF and RHD is not well understood, the risk factors associated with Strep A infection, ARF, and RHD are well established, both globally and in Aotearoa.
- The risk of ARF and subsequent RHD is partly influenced by factors associated with Strep A and the individual and partly by upstream risk factors. These are also known as the social determinants of health.
- International evidence points to the importance of the SDOH — the socioeconomic and political factors that influence people's lives — in creating structural barriers and inequities in health outcomes.
- Household crowding, poverty, and lack of access to healthcare are key SDOH that are modifiable risk factors for Strep A, ARF, and RHD.
- In Aotearoa, these risk factors disproportionately impact Māori and Pacific peoples.
- The health system in Aotearoa performs poorly for Māori and Pacific peoples, who have the greatest need for care related to Strep A, ARF, and RHD, whilst privileging other population groups.
- Primordial prevention focuses on addressing the root causes of diseases at a population level, such as poverty and crowding, and aims to avoid the development of risk factors.
- In Aotearoa, the distribution of SDOH privileges Pākehā and is influenced by colonisation and racism. Primordial prevention of ARF requires addressing these complex issues.

Risk factors and social determinants of health

Family/whānau history

Familial association for ARF is seen globally. Both environmental and genetic factors likely contribute to elevated risk, but the interplay between these is uncertain. Genetic susceptibility alone does not adequately explain the inequities in ARF seen by socioeconomic status in endemic regions worldwide. Nor does it explain why high rates of ARF previously seen in Pākehā sharply declined in the 20th century.

Studies demonstrating elevated familial risk for ARF include twin studies, case-control studies, and epidemiological reports.

- A case-control study in Aotearoa found that the risk of ARF was five times higher for those with a self-reported family/whānau history of ARF (aOR 4.97; 95% CI: 2.53–9.77).[1]
- A meta-analysis of twin studies showed a pooled concordance risk for ARF of 44% in monozygotic and 12% in dizygotic twins.[2]
- Studies in the 1980s and 1990s identified genetic polymorphisms associated with ARF and RHD, including studies linking ARF to the human leukocyte antigen (HLA) locus on chromosome 6.
- Genome-wide association studies have expanded the search for susceptibility loci.[3]
- A study of 206 people from Auckland with Māori and Pacific ancestry demonstrated an association of the HLA class 11 IL6 promoter variant (rs1800797, -597G/A) with RHD and an association of an IL1RN variant (rs447713) with the severity of cardiac involvement.[4]
- Echocardiography screening studies in Aotearoa and Uganda have shown an increased prevalence of RHD amongst first-degree relatives of those with ARF and RHD.[5, 6]

Social determinants of health

ARF and RHD are associated with socioeconomic disadvantage across Africa, the Americas, Asia, Europe, and the Pacific.[7-9] This link can be viewed as one of cause and effect when scrutinised against the Bradford Hill criteria for assessing the evidence of a causal relationship.[9, 10]

Socioeconomic status likely contributes to the risk of Strep A infection, ARF, and RHD through multiple intermediary factors such as household crowding, income, and education. These factors combine to compound the adverse effect.[8, 9]

The SDOH are the "non-medical factors that influence health outcomes. They are the conditions in which people are born, grow, work, live, and age, and the wider set of forces and systems shaping the conditions of daily life". [11, 12] These factors are estimated to contribute directly to 30-55% of overall health outcomes.[11]

In Aotearoa, there is strong evidence that SDOH, strongly associated with Strep A, ARF, and RHD, are shaped by political factors, colonisation, and racism, resulting in structural barriers and causing differential exposures to these determinants.[13-16]

Strep A, ARF, and RHD are all associated with socioeconomic disadvantage in Aotearoa,[17-19] though the strength of the association is strongest for ARF. An analysis of ARF (2000–2018) showed that people hospitalised with ARF were over five times more likely to live in the least socioeconomically privileged areas of Aotearoa (NZ Index of Deprivation (NZDep Index) 9–10; aRR 5.2; 95% CI: 4.0–6.8). People hospitalised with RHD or dying from RHD are three times more likely to live in the least privileged quintile of communities (NZDep Index 9–10) in Aotearoa.[18]

Three SDOH have established well-evidenced links with exposure to Strep A and subsequent development of ARF and RHD:

- Poor housing quality and household crowding.
- Low incomes, material deprivation, and poverty.
- Lack of access to quality healthcare that is also affordable.

Other SDOH, including food security, education, and employment, are also acknowledged as being interrelated and important for overall whānau wellbeing.

Household crowding

Household crowding rates are associated with increased exposure to Strep A and risk of ARF. Figure 3.1 shows that crowding may be structural (for example, an inadequate number of rooms) or functional (for example, sleeping in one room because heating is unaffordable).[20] Living in large intergenerational households benefits wellbeing. However, housing in Aotearoa is often poorly designed for big households, resulting in structural or functional crowding.[21, 22]

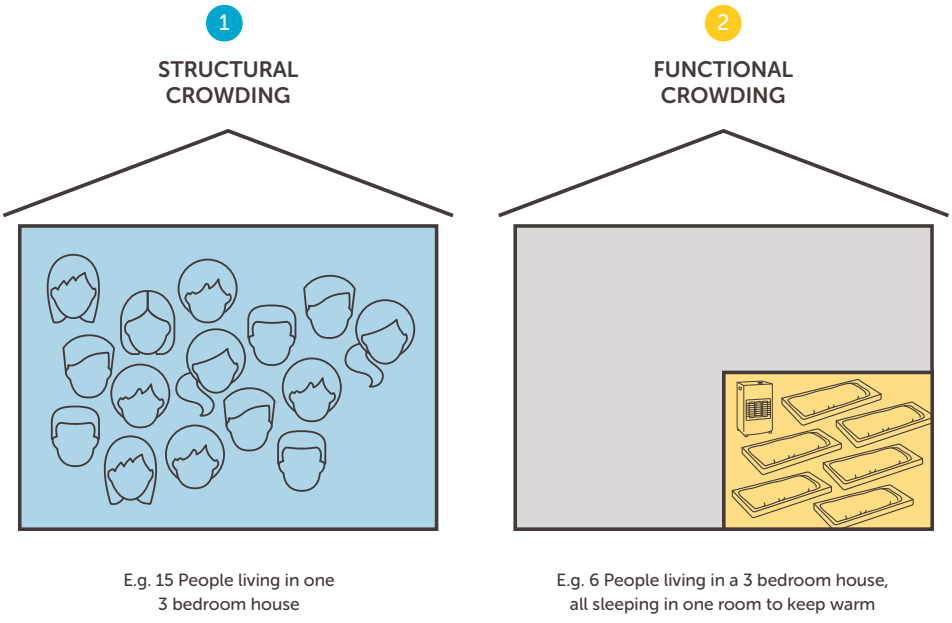

Figure 3.1. Crowding may be structural or functional (structural crowding as defined by the Canadian National Occupancy Standard)[23]

Māori and Pacific whānau are more likely to live in crowded households than other ethnic groups (40% of Pacific and 20% of Māori whānau, compared with 10% of the total population in 2013). Household crowding rates are higher in tamariki and rangatahi under 20 years of age.[23] Other housing factors include homelessness, transient or rental housing, and inadequate heating and insulation.[1, 9, 23-30]

A study of 55 whānau with experience of ARF in 2012 and 2013 found:

- 58% of people recently diagnosed with ARF reported household crowding.
- 35% lived in severe crowding (short of at least two bedrooms).
- 49% shared a bed.[27]

The Rheumatic Fever Risk Factors Study, a case-control study in Aotearoa, found strong associations between ARF and various forms of crowding (structural, functional, bed-sharing) and living in a rental home. Multi-variable analysis between ARF and housing tenure (living in a rental home) showed an adjusted Odds Ratio of 3.48 (95% CI: 1.81–6.70). The finding for household crowding showed an adjusted Odds Ratio of 3.88 (95% CI: 1.68–8.98).[1]

Differential access to quality healthcare

Poor access to healthcare increases the risk of ARF. Barriers to accessing primary healthcare were important modifiable risk factors in two case-control studies in Aotearoa. Barriers were associated with an increased risk of Strep A sore throat and ARF [1, 19] and included:

- Inability to book an appointment within 24 hours.
- The cost of the appointment and prescription.
- Lack of transport to attend the appointment.
- Lack of childcare for other tamariki.

Even when Māori and Pacific peoples can access care in Aotearoa, systemic racism and discrimination may be present, which can result in poorer quality of care for them than for people of other ethnicities.[31-35] Racism was found in audits of sore throat management in Te Tai Tokerau (Northland) general practices. There were differences in management for Māori and non-Māori patients. Young Māori were significantly less likely than non-Māori to receive antibiotics according to criteria in the national guidelines, suggesting that implicit bias in prescribing practices contributes to inequitable outcomes.[36, 37]

Poor nutrition

The evidence for an association between body weight, nutrition, sugar intake, oral health, and the risk of ARF is weak, although these factors may be proxies for other risk factors.

Some studies have shown malnutrition is associated with ARF and RHD.[7] Others have associated low vitamin D levels with ARF.[38, 39] An Israeli cohort study proposed high body weight (obesity) as a risk factor.[40]

An Auckland-based cohort study with Māori and Pacific tamariki, suggested an association with dental caries. After adjusting for potential confounders, tamariki with five or more primary teeth affected by caries were 57% more likely to develop ARF (95% CI: 20–106%) compared to tamariki whose primary teeth had no caries. However, a major

limitation of this study was the lack of data on Strep A infections in the cohort and the findings of this study were not replicated in the NZ case control study.[41]

Intake of sugar-sweetened beverages (1–9 drinks per day versus none) was associated with risk of ARF in the Aotearoa case-control study (aOR 2.00; 1.13–3.54), but increased caries were not.[1] Sugar intake may be a proxy for body mass or tooth decay. It is also postulated that sugar may enhance conditions in the throat that promote Strep A sore throat.[42]

Smoke exposure

Current evidence associating smoking exposure with ARF is insufficient, but there is a recognised association between household tobacco smoking with other infectious respiratory illnesses and meningococcal disease in tamariki.[43, 44]

Primordial prevention of acute rheumatic fever and rheumatic heart disease

Primordial prevention of ARF refers to modifying the inequitable distribution of the SDOH — which drives inequities in exposure to Strep A and the incidence of ARF and RHD.

Global primordial prevention experience

Improved living conditions (income and education, better housing, water, and sanitation) contributed to a decline in Strep A infections in many countries in the 20th century, with a sharper decline in rates of ARF and RHD seen in high and middle-income countries from the 1940s onwards. Evidence linking the decrease in ARF with improved living conditions, housing, and hygiene is mostly ecological and observational. Even so, many studies have confirmed a consistently strong epidemiological association globally with poverty, social inequity, and household crowding.[8, 9, 24, 45, 46]

A large proportion of the decrease in ARF occurred before the widespread availability of antibiotics, indicating the important role of SDOH in exposure to Strep A and the subsequent development of ARF and RHD.[8, 9, 47-49] However, high rates of ARF and RHD persist in many low and middle-income countries, as well as among Māori and Pacific communities in Aotearoa and Indigenous Peoples in Australia (see **Chapter 4: Epidemiology of Strep A Infections, Acute Rheumatic Fever and Rheumatic Heart Disease**).

Improvements in access to healthcare have also been associated with declines in ARF and RHD at a population level, with sharper declines observed since the widespread availability of penicillin. Gordis reported on studies in underserved parts of inner-city Baltimore, United States, where comprehensive primary care resulted in reduced ARF incidence, independent of ethnicity.[50] Large declines in ARF and RHD seen in Costa Rica, Cuba, and the Caribbean from the 1970s to the 1990s have been attributed to improved access to healthcare, with programmes targeting ARF prevention through treatment of Strep A pharyngitis, alongside socioeconomic development.[8, 25, 51-55]

Primordial prevention of acute rheumatic fever in Aotearoa

Māori and Pacific peoples are most impacted by ARF and RHD in Aotearoa.[56] As part of its Te Tiriti o Waitangi obligations, the Crown must address inequities in health for all populations. Māori have unique rights as tangata whenua of Aotearoa (see **Chapter 1: Cultural Responsiveness**). Reducing ARF and RHD in Aotearoa requires acknowledging these rights and addressing the inequitable distribution of power and resources that drive inequities in the key SDOH that increase exposure to Strep A, ARF, and RHD. In Aotearoa, colonisation exerts a detrimental impact on Māori health through mechanisms including land appropriation, social deprivation, experiences of racism, cultural subjugation, and loss of political power.[16, 57-59] As depicted in the William's model, adapted by Te Kupenga Hauora Māori (TKHM), these 'basic causes' drive social determinants and social status, and intervening mechanisms or 'surface causes' result in behavioural, physiological or psychological responses (see **Figure 3.2**).[16, 60]

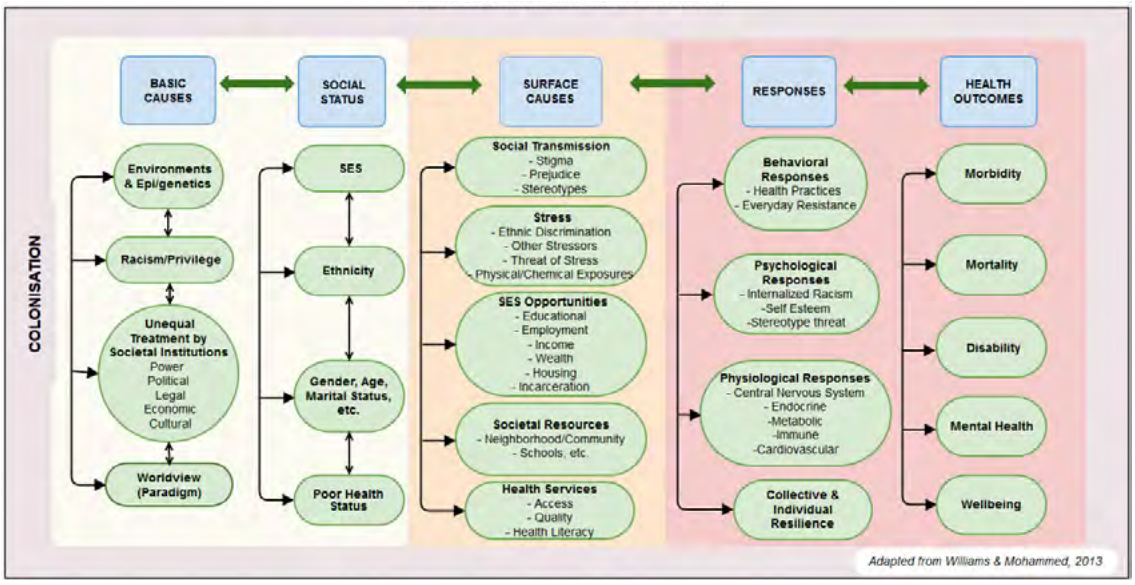

Figure 3.2. TKHM-modified Williams & Mohammed model for explaining Indigenous/ethnic determinants of health

Interventions to address poor housing quality and crowding

Decades of high-quality research in Aotearoa have reported the positive health benefits of living in warm, dry, and safe homes, as well as the importance of appropriate design to meet the intergenerational and cultural contexts of whānau.[61-65] Evidence from existing programmes demonstrates benefits from improved housing, including improved health outcomes. Scaling up housing interventions would likely deliver positive impacts to offset inequities in Strep A exposure and ARF and RHD (Evidence level III-2, Grade B).[61, 66]

Alleviation of child poverty

As of June 2023, 12.5% of all tamariki in Aotearoa lived in households experiencing material hardship.[67] Additionally, 17.% of tamariki lived in households with an equivalised disposable income below 50% of the median, after accounting for housing costs. This percentage was about 3% more than in 2021/2022.[67]

Household levels of material hardship are higher for tamariki Māori and Pacific tamaiti, with 21.5% of tamariki Māori and 28.9% of Pacific tamaiti living in households experiencing significant restrictions or deprivations of essential items compared with 9.4% of Pākehā.[67-69] These ethnic disparities are even more extreme for severe material hardship (defined as households going without nine or more basic needs). In 2022/23, 10.4% of tamariki Māori and 14.7% of Pacific tamaiti lived in households experiencing severe material hardship, compared to 3.5% of European tamariki.

In 2023, around one in seven tamariki in Aotearoa were living in material hardship, where fresh food was unaffordable, whānau put up with feeling cold, or visits to the doctor or dentist had to be postponed because of cost. For Pacific tamaiti this was the case for nearly one in three and for one in five tamariki Māori; for Pākehā tamariki, it was less than one in ten.[67]

Figure 3.3. Thresholds for tamariki in relation to material hardship and severe material hardship[69]

As a primordial intervention, poverty alleviation would have multiple benefits for ARF prevention by improving access to warm, dry housing, food security, and better access to primary healthcare.

Access to affordable, quality healthcare

Access to quality healthcare is an important determinant of health in Aotearoa. Barriers to accessing primary healthcare include:

- Upfront costs.
- Living remotely (rurally).
- Lack of transport.
- Caregivers do not have enough time.
- Previous negative experiences, including experiences of racism.

In addition, barriers created by the health system are well described, such as:

- Lack of available appointments.
- Lack of after-hours services.
- Scarcity of locations and locations that are hard to access.
- Structural racism.
- Unconscious bias.[70-73]

Evidence over decades shows that these healthcare inequities negatively impact health outcomes for Māori and Pacific peoples, which extends to Strep A and the development of ARF and RHD.[35, 59, 74-81]

Frameworks, key principles, and decades of experience in Hauora Māori and Pacific peoples' health and community services can guide future strategies and effective interventions to address these structural issues.[82-84] A recent review identified key features of interventions that improved health outcomes for rangatahi Māori:

- Evidence-based, sustained, comprehensive approaches, including both universal levers and Indigenous youth-specific policies.
- Indigenous and rangatahi leadership.
- The political will to address Indigenous youth rights, preferences, and priorities.
- A commitment to an anti-racist praxis and healthcare Indigenisation.[82]

Previous interventions have shown how to successfully improve access to primary prevention services, as reinforced by outcomes experienced during the response to the COVID-19 pandemic. Key learnings from previous successful interventions are the need to:

- Sustainably and equitably resource hauora and community services led by Māori and Pacific peoples.
- Increase access by Māori and Pacific peoples to quality, culturally safe, comprehensive (multidisciplinary) primary care in appropriate settings, such as early childhood services, kōhanga reo and a'oga, schools, kura kaupapa, marae, and pharmacies.[85-87]

Previous experiences of ensuring free primary healthcare services for tamariki and eliminating prescription charges suggest that reducing cost barriers can successfully increase access for whānau.[88, 89]

Interventions to address racism and privilege

To successfully address inequitable ARF and RHD rates impacting Māori and Pacific peoples, primordial prevention needs to target the complex impacts of colonisation, systemic racism, socioeconomic inequities, and health system deficits.

Māori and Pacific peoples experience racism as individuals, as healthcare practitioners, and structurally in the health sector.[57, 59, 79, 90] Therefore, racism and privilege at both institutional and personal levels should be included in primordial prevention efforts.

To date, strategies to address racism in Aotearoa healthcare settings have mostly been implemented at the level of a healthcare practitioner, in workforce training and education, centred around cultural safety, implicit bias, or Indigenous framing.[79, 91]

Other actions to embed in the health system are:

- Addressing the power imbalances and privilege reflected in decision-making, commissioning, and resourcing within the health system (structural racism).
- Devolving governance and resources to Pacific, iwi, hapū, and Māori community and Hauora services.
- Building a culturally safer workforce (see **Chapter 1: Cultural Responsiveness**).[80, 90]

References

1. Baker MG, Gurney J, Moreland NJ, Bennett J, Oliver J, Williamson DA, et al. Risk factors for acute rheumatic fever: a case-control study. *The Lancet Regional Health — Western Pacific*. 2022;26:100508. https://doi.org/10.1016/j.lanwpc.2022.100508

2. Engel ME, Stander R, Vogel J, Adeyemo AA, Mayosi BM. Genetic susceptibility to acute rheumatic fever: a systematic review and meta-analysis of twin studies. *PloS One*. 2011;6(9):e25326. https://doi.org/10.1371/journal.pone.0025326

3. Muhamed B, Parks T, Sliwa K. Genetics of rheumatic fever and rheumatic heart disease. *Nature Reviews: Cardiology*. 2020;17(3):145–154. https://doi.org/10.1038/s41569-019-0258-2

4. Azevedo PM, Merriman TR, Topless RK, Wilson NJ, Crengle S, Lennon DR. Association study involving polymorphisms in IL-6, IL-1RA, and CTLA4 genes and rheumatic heart disease in New Zealand population of Māori and Pacific ancestry. *Cytokine*. 2016;85:201–206. https://doi.org/10.1016/j.cyto.2016.06.029

5. Culliford-Semmens N, Tilton E, Wilson N, Stirling J, Doughty R, Gentles T, et al. Echocardiography for latent rheumatic heart disease in first degree relatives of children with acute rheumatic fever: implications for active case finding in family members. *EClinicalMedicine*. 2021;37:100935. https://doi.org/10.1016/j.eclinm.2021.100935

6. Aliku T, Sable C, Scheel A, Tompsett A, Lwabi P, Okello E, et al. Targeted echocardiographic screening for latent rheumatic heart disease in northern Uganda: evaluating familial risk following identification of an index case. *PLoS Neglected Tropical Diseases*. 2016;10(6):e0004727. https://doi.org/10.1371/journal.pntd.0004727

7. Watkins D, Baker M, Kumar R, Parks T. Epidemiology, risk factors, burden and cost of acute rheumatic fever and rheumatic heart disease. In: Dougherty S, Carapetis J, Zühlke L, Wilson N, editors. Acute rheumatic fever and rheumatic heart disease. Amsterdam: Elsevier; 2020. p. 1–18.

8. Baker MG, Masterson MY, Shung-King M, Beaton A, Bowen AC, Bansal GP, et al. Research priorities for the primordial prevention of acute rheumatic fever and rheumatic heart disease by modifying the social determinants of health. *BMJ Global Health*. 2023;8(Suppl 9). https://doi.org/10.1136/bmjgh-2023-012467

9. Coffey PM, Ralph AP, Krause VL. The role of social determinants of health in the risk and prevention of group A streptococcal infection, acute rheumatic fever and rheumatic heart disease: a systematic review. *PLoS Neglected Tropical Diseases*. 2018;12(6):e0006577. https://doi.org/10.1371/journal.pntd.0006577

10. Hill AB. The environment and disease: association or causation? 1965. *Journal of the Royal Society of Medicine*. 2015;108(1):32–37. https://doi.org/10.1177/0141076814562718

11. World Health Organization. Social determinants of health. World Health Organization; 2023. https://www.who.int/health-topics/social-determinants-of-health (Accessed February 21 2025).

12. World Health Organization. Closing the gap in a generation: health equity through action on the social determinants of health. World Health Organization; 2008. https://iris.who.int/handle/10665/69832.

13. Graham R, Masters-Awatere B. Experiences of Māori of Aotearoa New Zealand's public health system: a systematic review of two decades of published qualitative research. *Australian and New Zealand Journal of Public Health*. 2020;44(3):193–200. https://doi.org/10.1111/1753-6405.12971

14. Harris R, Tobias M, Jeffreys M, Waldegrave K, Karlsen S, Nazroo J. Racism and health: the relationship between experience of racial discrimination and health in New Zealand. *Social Science and Medicine*. 2006;63(6):1428–1441. https://doi.org/10.1016/j.socscimed.2006.04.009

15. Ministry of Health. Racial discrimination 2011/12, 2016/17 and 2020/21. New Zealand Health Survey. Wellington: Ministry of Health; 2023.

16. Curtis E, Jones R, Willing E, Anderson A, Paine S-J, Herbert S, et al. Indigenous adaptation of a model for understanding the determinants of ethnic health inequities. *Discover Social Science and Health*. 2023;3(1). https://doi.org/10.1007/s44155-023-00040-6

17. Lennon D, Stewart J, Farrell E, Palmer A, Mason H. School-based prevention of acute rheumatic fever: a group randomized trial in New Zealand. *Pediatric Infectious Disease Journal*. 2009;28(9):787–794. https://doi.org/10.1097/INF.0b013e3181a282be

18. Bennett J, Zhang J, Leung W, Jack S, Oliver J, Webb R, et al. Rising ethnic inequalities in acute rheumatic fever and rheumatic heart disease, New Zealand, 2000–2018. *Emerging Infectious Diseases*. 2021;27(1):36–46. https://doi.org/10.3201/eid2701.191791

19. Bennett J, Moreland NJ, Zhang J, Crane J, Sika-Paotonu D, Carapetis J, et al. Risk factors for group A streptococcal pharyngitis and skin infections: a case control study. *The Lancet Regional Health — Western Pacific*. 2022;26:100507. https://doi.org/10.1016/j.lanwpc.2022.100507

20. Goodyear R, Fabian A, Hay J. Finding the crowding index that works best for New Zealand. Working Paper No. 11–04. Wellington: Statistics New Zealand; 2011. https://www.stats.govt.nz/assets/Uploads/Retirement-of-archive-website-project-files/Research/Finding-the-crowding-index-that-works-best-for-New-Zealand-working-paper/finding-crowding-index-best-for-nz-working-paper-11-04.pdf (Accessed February 23 2025).

21. Boulton A, Allport T, Kaiwai H, Harker R, Potaka Osborne G. Māori perceptions of 'home': Māori housing needs, wellbeing and policy. *Kōtuitui: New Zealand Journal of Social Sciences Online*. 2021;17(1):44–55. https://doi.org/10.1080/1177083x.2021.1920984

22. Malungahu G. Too little space! Experiences and perspectives of housing and housing policy: Tongan families with rheumatic fever in South Auckland and key housing informants [Doctoral thesis]. Auckland: University of Auckland; 2020.

23. Southern Initiative. Healthy Homes Initiative — Auckland: co-design: making Auckland homes warmer and drier. Auckland: Southern Initiative; 2018. https://www.health.govt.nz/publications/healthy-homes-initiative-auckland-co-design-testing-ideas-to-make-homes-warmer-and-drier-report-on-0.

24. Baker MG, Gurney J, Oliver J, Moreland NJ, Williamson DA, Pierse N, et al. Risk factors for acute rheumatic fever: literature review and protocol for a case-control study in New Zealand. *International Journal of Environmental Research and Public Health*. 2019;16(22). https://doi.org/10.3390/ijerph16224515

25. Kerdemelidis M, Lennon DR, Arroll B, Peat B, Jarman J. The primary prevention of rheumatic fever. *Journal of Paediatrics and Child Health*. 2010;46(9):534–548. https://doi.org/10.1111/j.1440-1754.2010.01854.x

26. Jaine R, Baker M, Venugopal K. Acute rheumatic fever associated with household crowding in a developed country. *Pediatric Infectious Disease Journal*. 2011;30(4):315–319. https://doi.org/10.1097/INF.0b013e3181fbd85b

27. Oliver JR, Pierse N, Stefanogiannis N, Jackson C, Baker MG. Acute rheumatic fever and exposure to poor housing conditions in New Zealand: a descriptive study. *Journal of Paediatrics and Child Health*. 2017;53(4):358–364. https://doi.org/10.1111/jpc.13421

28. Cannon JW, Abouzeid M, de Klerk N, Dibben C, Carapetis JR, Katzenellenbogen JM. Environmental and social determinants of acute rheumatic fever: a longitudinal cohort study. *Epidemiology and Infection*. 2019;147:e79. https://doi.org/10.1017/s0950268818003527

29. Carapetis JR, Beaton A, Cunningham MW, Guilherme L, Karthikeyan G, Mayosi BM, et al. Acute rheumatic fever and rheumatic heart disease. *Nature Reviews Disease Primers*. 2016;2:15084. https://doi.org/10.1038/nrdp.2015.84

30. Avire NJ, Whiley H, Ross K. A review of streptococcus pyogenes: public health risk factors, prevention and control. *Pathogens*. 2021;10(2). https://doi.org/10.3390/pathogens10020248

31. Paine SJ, Li C, Wright K, Harris R, Loring B, Reid P. The economic cost of Indigenous child health inequities in Aotearoa New Zealand-an updated analysis for 2003-2014. *New Zealand Medical Journal*. 2022;136(1568):23–45. https://doi.org/10.26635/6965.5874

32. Robson B, Harris R, editors. Hauora: Māori Standards of Health IV. A study of the years 2000–2005. Wellington: Te Rōpū Rangahau Hauora a Eru Pōmare; 2007.

33. Tipene-Leach D, Walker R. Pervasive kidney health inequities for Maori require multi-level attention. *Nature Reviews Nephrology*. 2022;18(9):541–542. https://doi.org/10.1038/s41581-022-00606-8

34. Svensen G, Hikaka J, Cavadino A, Kool B. Ethnic variation in hospitalisation due to treatment injury and complications of healthcare in older adults residing in New Zealand. *New Zealand Medical Journal*. 2023;136(1579):70–85. https://doi.org/10.26635/6965.6065

35. Jeffreys M, Smiler K, Ellison-Loschmann L, Pledger M, Kennedy J, J. C. Prevalence and consequences of barriers to primary health care. Wellington: Ministry of Social Development. https://www.msd.govt.nz/documents/about-msd-and-our-work/publications-resources/research/barriers-to-primary-health-care/prevalence-and-consequences-of-barriers-to-primary-health-care.pdf (Accessed February 23 2025).

36. Shetty A, Mills C, Eggleton K. Primary care management of group A streptococcal pharyngitis in Northland. *Journal of Primary Health Care*. 2014;6(3). https://doi.org/10.1071/hc14189

37. Shetty A, Mills C, Eggleton K. A repeat audit of primary care management of group A streptococcal pharyngitis in Northland, New Zealand 2016. *Journal of Primary Health Care*. 2018;10(1):18–24. https://doi.org/10.1071/hc17056

38. Thorup L, Hamann SA, Tripathee A, Koirala B, Gyawali B, Neupane D, et al. Evaluating vitamin D levels in rheumatic heart disease patients and matched controls: a case-control study from Nepal. *PloS One*. 2020;15(8):e0237924. https://doi.org/10.1371/journal.pone.0237924

39. Onan SH, Demirbilek H, Aldudak B, Bilici M, Demir F, Yilmazer MM. Evaluation of vitamin D levels in patients with acute rheumatic fever. *The Anatolian Journal of Cardiology*. 2017;18(1):75–76. https://doi.org/10.14744/AnatolJCardiol.2017.7720

40. Machluf Y, Chaiter Y, Farkash R, Sebbag A, Fink DL. Rheumatic fever in large cohort of adolescents in Israel. *Frontiers of Medicine*. 2019;6:328. https://doi.org/10.3389/fmed.2019.00328

41. Thornley S, Marshall RJ, Bach K, Koopu P, Reynolds G, Sundborn G, et al. Sugar, dental caries and the incidence of acute rheumatic fever: a cohort study of Maori and Pacific children. *Journal of Epidemiology and Community Health*. 2017;71(4):364–370. https://doi.org/10.1136/jech-2016-208219

42. Office of the Prime Minister's Chief Science Advisor. Group A streptococcus and acute rheumatic fever in Aotearoa New Zealand: a summary of current knowledge in Aotearoa New Zealand. Office of the Prime Minister's Chief Science Advisor; 2021. https://www.dpmc.govt.nz/sites/default/files/2024-01/PMCSA-21-11-02-V1-OPMCSA-rheumatic-fever-to-upload-on-Nov-19-.pdf (Accessed February 18 2025).

43. Adenaiye O, Bueno de Mesquita PJ, Wu Q, Hong F, Lai J, Chen S, et al. The effect of COVID-19 stay-at-home order and campus closure on the prevalence of acute respiratory infection symptoms in college campus cohorts. *Influenza and Other Respiratory Viruses*. 2021;15(3):331–335. https://doi.org/10.1111/irv.12837

44. Baker M, McNicholas A, Garrett N, Jones N, Stewart J, Koberstein V, et al. Household crowding a major risk factor for epidemic meningococcal disease in Auckland children. *Pediatric Infectious Disease Journal*. 2000;19(10):983–990. https://doi.org/10.1097/00006454-200010000-00009

45. Steer AC. Historical aspects of rheumatic fever. *Journal of Paediatrics and Child Health*. 2015;51(1):21–27. https://doi.org/10.1111/jpc.12808

46. Dougherty S, Carapetis J, Zühlke L, Wilson N. Acute rheumatic fever and rheumatic heart disease. Amsterdam: Elsevier; 2020.

47. Gordis L. The virtual disappearance of rheumatic fever in the United States: lessons in the rise and fall of disease. T. Duckett Jones memorial lecture. *Circulation*. 1985;72(6):1155–1162. https://doi.org/10.1161/01.cir.72.6.1155

48. Bland EF. Rheumatic fever: the way it was. *Circulation*. 1987;76(6):1190–1195. https://doi.org/10.1161/01.cir.76.6.1190

49. Carapetis JR, McDonald M, Wilson NJ. Acute rheumatic fever. *Lancet*. 2005;366(9480):155–168. https://doi.org/10.1016/s0140-6736(05)66874-2

50. Gordis L. Effectiveness of comprehensive-care programs in preventing rheumatic fever. *New England Journal of Medicine*. 1973;289(7):331–335. https://doi.org/10.1056/nejm197308162890701

51. Nordet P, Lopez R, Dueñas A, Sarmiento L. Prevention and control of rheumatic fever and rheumatic heart disease: the Cuban experience (1986-1996-2002). *Cardiovascular Journal of Africa*. 2008;19(3):135–140.

52. Arguedas A, Mohs E. Prevention of rheumatic fever in Costa Rica. *Journal of Pediatrics*. 1992;121(4):569–572. https://doi.org/10.1016/s0022-3476(05)81146-1

53. Watkins DA, Mvundura M, Nordet P, Mayosi BM. A cost-effectiveness analysis of a program to control rheumatic fever and rheumatic heart disease in Pinar del Rio, Cuba. *PloS One*. 2015;10(3):e0121363. https://doi.org/10.1371/journal.pone.0121363

54. Bach JF, Chalons S, Forier E, Elana G, Jouanelle J, Kayemba S, et al. 10-year educational programme aimed at rheumatic fever in two French Caribbean islands. *Lancet*. 1996;347(9002):644–648. https://doi.org/10.1016/s0140-6736(96)91202-7

55. Yusuf S, Narula J, Gamra H. Can we eliminate rheumatic fever and premature deaths from RHD? *Global Heart*. 2017;12(1):3–4. https://doi.org/10.1016/j.gheart.2017.05.001

56. Reid P. Good governance: the case of health equity. In: Tawhai V, Gray-Sharp K, editors. 'Always speaking': the Treaty of Waitangi and public policy. Wellington: Huia; 2011. p. 39–50.

57. Moewaka Barnes A, Taiapa K, Borell B, McCreanor T. Māori experiences and responses to racism in New Zealand. *MAI Journal*. 2013;2(2):63–77.

58. Moewaka Barnes H, McCreanor T. Colonisation, hauora and whenua in Aotearoa. *Journal of the Royal Society of New Zealand*. 2019;49:19–33. https://doi.org/10.1080/03036758.2019.1668439

59. Harris RB, Stanley J, Cormack DM. Racism and health in New Zealand: prevalence over time and associations between recent experience of racism and health and wellbeing measures using national survey data. *PloS One*. 2018;13(5):e0196476. https://doi.org/10.1371/journal.pone.0196476

60. Williams DR, Mohammed SA. Racism and health I: Pathways and scientific evidence. *American Behavioral Scientist*. 2013;57(8). https://doi.org/10.1177/0002764213487340

61. Howden-Chapman P, Crane J, Keall M, Pierse N, Baker MG, Cunningham C, et al. He Kainga Oranga: reflections on 25 years of measuring the improved health, wellbeing and sustainability of healthier housing. *J R Soc N Z*. 2024;54(3):290–315. https://doi.org/10.1080/03036758.2023.2170427

62. Chapman R, Howden-Chapman P, Viggers H, O'Dea D, Kennedy M. Retrofitting houses with insulation: a cost-benefit analysis of a randomised community trial. *Journal of Epidemiology and Community Health*. 2009;63(4):271–277. https://doi.org/10.1136/jech.2007.070037

63. Telfar-Barnard L, Preval N, Howden-Chapman P, Arnold A, Young C, Grimes A, et al. The impact of retrofitted insulation and new heaters on health services utilisation and costs, pharmaceutical costs and mortality: evaluation of Warm Up New Zealand: Heat Smart. Department of Public Health, University of Otago; 2011. https://www.motu.nz/assets/Documents/our-work/urban-and-regional/housing/The-Impact-of-Retrofitted-Insulation-and-New-Heaters-on-Health-Services-Utilisation-and-Costs-Pharmaceutical-Costs-and-Mortality-Evaluation-of-Warm-Up-New-Zealand-Heat-Smart.pdf (Accessed February 23 2025).

64. Howden-Chapman P, Pierse N, Nicholls S, Gillespie-Bennett J, Viggers H, Cunningham M, et al. Effects of improved home heating on asthma in community dwelling children: randomised controlled trial. *BMJ*. 2008;337:a1411. https://doi.org/10.1136/bmj.a1411

65. Baker M, Zhang J, Howden-Chapman P. Health impacts of social housing: hospitalisations in Housing New Zealand applicants and tenants, 2003–2008. Wellington: He Kainga Oranga/Housing and Health Research Programme University of Otago; 2010. https://www.healthyhousing.org.nz/sites/default/files/2022-01/Microsoft-Word-Health-Impacts-of-Social-Housing-June-2010-FINAL1.pdf (Accessed February 24 2025).

66. Pierse N, Johnson E, Riggs L, Watson N. Healthy Homes Initiative: three years outcomes evaluation. Health New Zealand | Te Whatu Ora; 2022. https://www.tewhatuora.govt.nz/publications/heathy-homes-initiative-three-year-outcomes-evaluation/ (Accessed February 18 2025).

67. Stats NZ. Child poverty statistics: year ended June 2023. Stats NZ; 2024. https://www.stats.govt.nz/information-releases/child-poverty-statistics-year-ended-june-2023/ (Accessed February 23 2025).

68. Duncanson M, Roy M, van Asten H, Oben G, Wicken A, Tustin K, et al. Child Poverty Monitor 2022. Technical report. Dunedin: NZ Child and Youth Epidemiology Service, University of Otago; 2022. https://www.nzchildren.co.nz/ (Accessed February 23 2025).

69. Stats NZ. Child poverty statistics: year ended June 2022. Stats NZ; 2022. https://www.stats.govt.nz/information-releases/child-poverty-statistics-year-ended-june-2022 (Accessed February 23 2025).

70. Anderson A, Mills C, Eggleton K. Whānau perceptions and experiences of acute rheumatic fever diagnosis for Māori in Northland, New Zealand. *New Zealand Medical Journal*. 2017;130(1465):80–88.

71. Ministry of Health. Annual update of key results 2015/16: New Zealand Health Survey. Wellington: Ministry of Health; 2016. https://www.health.govt.nz/publications/annual-update-of-key-results-201516-new-zealand-health-survey (Accessed February 24 2025).

72. Ministry of Health. Barriers to visiting a GP. Ministry of Health; 2024. https://www.health.govt.nz/publications/annual-update-of-key-results-202324-new-zealand-health-survey (Accessed February 22 2025).

73. Jeffreys M, Ellison-Loschmann L, Irurzun-Lopez M, Cumming J, McKenzie F. Financial barriers to primary health care in Aotearoa New Zealand. *Family Practice*. 2024;41(6):995–1001. https://doi.org/10.1093/fampra/cmad096

74. Ryan D, Grey C, Mischewski B. Tofa Saili: a review of evidence about health equity for Pacific peoples in New Zealand. Pacific Perspectives Limited; 2019. https://www.nzdoctor.co.nz/sites/default/files/2019-09/Tofa%20Saili-%20A%20review%20of%20evidence%20about%20health%20equity%20for%20Pacific%20Peoples%20in%20New%20Zealand.pdf (Accessed February 22 2025).

75. Jeffreys M, Smiler K, Ellison Loschmann L, Pledger M, Kennedy J, Cumming J. Consequences of barriers to primary health care for children in Aotearoa New Zealand. *SSM — Population Health*. 2022;17:101044. https://doi.org/10.1016/j.ssmph.2022.101044

76. Espiner E, Paine SJ, Weston M, Curtis E. Barriers and facilitators for Māori in accessing hospital services in Aotearoa New Zealand. *New Zealand Medical Journal*. 2021;134(1546):47–58.

77. Goodyear-Smith F, Ashton T. New Zealand health system: universalism struggles with persisting inequities. *Lancet*. 2019;394(10196):432–442. https://doi.org/10.1016/s0140-6736(19)31238-3

78. Health New Zealand | Te Whatu Ora. Aotearoa New Zealand health status report 2023. Health New Zealand | Te Whatu Ora; 2024. https://www.tewhatuora.govt.nz/publications/health-status-report (Accessed February 22 2025).

79. Talamaivao N, Harris R, Cormack D, Paine SJ, King P. Racism and health in Aotearoa New Zealand: a systematic review of quantitative studies. *New Zealand Medical Journal*. 2020;133(1521):55–68.

80. Waitangi Tribunal. HAUORA report on stage one of the Health Services and Outcomes Kaupapa Inquiry. Waitangi Tribunal; 2023. https://forms.justice.govt.nz/search/Documents/WT/wt_DOC_195476216/Hauora%202023%20W.pdf (Accessed February 22 2025).

81. Malcolm L. New Zealand action to address major inequities in the distribution and ultilisation of primary health care services. *Australian Journal of Primary Health*. 2004;10(3). https://doi.org/10.1071/py04053

82. Clark TC, Ball J, Fenaughty J, Drayton B, Fleming TT, Rivera-Rodriguez C, et al. Indigenous adolescent health in Aotearoa New Zealand: trends, policy and advancing equity for rangatahi Maori, 2001–2019. *The Lancet Regional Health — Western Pacific*. 2022;28:100554. https://doi.org/10.1016/j.lanwpc.2022.100554

83. Gustafson P, Abdul Aziz Y, Lambert M, Bartholomew K, Rankin N, Fusheini A, et al. A scoping review of equity-focused implementation theories, models and frameworks in healthcare and their application in addressing ethnicity-related health inequities. *Implementation Science*. 2023;18(1):51. https://doi.org/10.1186/s13012-023-01304-0

84. Palmer SC, Gray H, Huria T, Lacey C, Beckert L, Pitama SG. Reported Māori consumer experiences of health systems and programs in qualitative research: a systematic review with meta-synthesis. *International Journal for Equity in Health*. 2019;18(1):163. https://doi.org/10.1186/s12939-019-1057-4

85. Sinclair O, Lyndon M. Pathways towards health equity for tamariki Māori. CPAG 2023 policy brief on Māori child health. Child Poverty Action Group; 2023. https://www.cpag.org.nz/policy-briefs/maori-child-health (Accessed February 23 2025).

86. Whānau Ora Review Independent Panel. Whānau Ora review Tipu Matoro ki te Ao — final report to the Minister for Whānau Ora. Wellington: Te Puni Kōkiri; 2018. https://www.tpk.govt.nz/docs/tpk-wo-review-2019.pdf (Accessed February 24 2025).

87. King J, Moss M, Spee KE. Evaluation of school-based health services in primary and intermediate schools (Mana Kidz). Auckland: Counties Manukau Health; 2022. https://www.nhc.maori.nz/wp-content/uploads/2022/08/220601-Mana-Kidz-evaluation-final-report.pdf (Accessed February 11 2025).

88. Crampton P. The ongoing evolution of capitation funding for primary care: the December 2018 PHO capitation funding changes for Community Services Card holders. *New Zealand Medical Journal*. 2019;132(1498):69–78.

89. Norris P, Horsburgh S, Cumming J, Tordoff J. Prescription charge increases in New Zealand penalise the poor and sick. *Journal of Primary Health Care*. 2014;6(1):4–5.

90. Came H, O'Sullivan D, Kidd J, McCreanor T. The Waitangi Tribunal's WAI 2575 report: implications for decolonizing health systems. *Health and Human Rights*. 2020;22(1):209–220.

91. Curtis E, Jones R, Tipene-Leach D, Walker C, Loring B, Paine SJ, et al. Why cultural safety rather than cultural competency is required to achieve health equity: a literature review and recommended definition. *International Journal for Equity in Health*. 2019;18(1):174. https://doi.org/10.1186/s12939-019-1082-3

4

Epidemiology of Strep A Infections, Acute Rheumatic Fever and Rheumatic Heart Disease

In this chapter

"And the fact they always say it's [ARF] Māori and Pacific islands [people] you know? It's like oh you know it's because they say that not a lot of us live in Remuera so it must be because we're in poverty. So now this is a poor person's disease isn't it, coz that's how you make us feel. Like it is our fault."

Parent of a rangatahi with ARF

Key changes

This chapter discusses the updated epidemiology of Strep A infections, acute rheumatic fever (ARF), and rheumatic heart disease (RHD) and the pattern of these conditions in Aotearoa.

Key points

- About 140 new ARF cases are diagnosed each year in Aotearoa. ARF is most common in tamariki aged 5–14 years. Of people diagnosed with ARF, 94% are under 35 years of age.
- Nearly 50% of people diagnosed with ARF are Māori, and over 40% are Pacific peoples.
- ARF rates for Māori are 10 times higher than for non-Māori, non-Pacific peoples after correcting for age, sex, socioeconomic deprivation, and location. Rates for Pacific peoples are 20 times higher.
- Rates of ARF are highest in the North Island, particularly in the northern region (South Auckland and Northland/Te Tai Tokerau) and the East Coast/Te Tai Rāwhiti.
- An estimated 1,800 people receive secondary antibiotic prophylaxis (SAP) with 4-weekly injections of long-acting penicillin and are enrolled on rheumatic fever registers in Aotearoa.
- The rate of recurrent ARF in Aotearoa is around 7% (the median age for recurrent ARF is 21 years).[1]
- In Aotearoa, around 40% of people with moderate to severe RHD have not had a confirmed ARF episode.[2]
- The Aotearoa New Zealand Rheumatic Heart Disease Registry has found about 3,500 people living with moderate to severe RHD in Aotearoa, and around two-thirds have had heart surgery. The average age at surgery was 40 years. Nearly one in five had two or more cardiac interventions.[2]
- RHD shows stark inequities in mortality. A recent study estimated the average age of death attributed to RHD at 59 years for Māori, 55 years for Pacific peoples, and 80 years for Pākehā.[3]

Introduction

Behind the numbers presented in this chapter are whānau who have experienced the impact of ARF and RHD on individual and collective wellbeing. We also acknowledge the loss and grief from RHD-related deaths and loss of health and wellbeing for impacted whānau.

Global epidemiology of Strep A, acute rheumatic fever and rheumatic heart disease

Historically, Strep A diseases such as scarlet fever, ARF and RHD were important causes of illness and death in Europe and the United States. In the late 19th century and the first half of the 20th century, rates of these conditions declined rapidly, thought to be due to improved sanitation and living conditions.[4-7]

However, ARF and RHD persist in many Indigenous and socioeconomically disadvantaged communities within high-income countries (see **Chapter 3: Strep A Infection, Acute Rheumatic Fever and Rheumatic Heart Disease: Risk Factors, Social Determinants of Health and Primordial Prevention**).[8] In low- and middle-income countries, Strep A infections, ARF, and RHD continue to represent a significant burden of disease, accounting for most of the global burden.[9-11] Strep A results in at least half a million deaths a year.[9] The global resurgence of invasive infections and regional outbreaks of diseases such as scarlet fever and invasive Group A Strep infections (iGAS) in the last decade have increased awareness about Strep A infections internationally.[8]

Global epidemiology of acute rheumatic fever

ARF is rare in tamariki under 4 years old. Incidence peaks around 9–14 years of age and declines after 20 years of age. Some studies report a higher risk of ARF and RHD for females, which may be at least partially associated with an increased chance of diagnosis when accessing healthcare during pregnancy.[12]

Globally, ARF annual incidence estimates among tamariki and rangatahi range from 8–51 per 100,000;[13, 14] however, robust ARF incidence data from endemic regions is limited.[15, 16] Higher rates have been reported among specific populations; for example, up to 194 per 100,000 person-years in the Indigenous populations in the Northern Territory of Australia. Higher rates may reflect better access to healthcare resources for diagnosis and better surveillance systems to record cases rather than truly higher incidence.

Global epidemiology of rheumatic heart disease

Around the world, 40%–80% of people diagnosed with RHD have no prior diagnosis of ARF.[2, 12, 17, 18]

RHD is most common in sub-Saharan Africa, Asia, and the Pacific, with a disproportionate impact on Indigenous peoples and marginalised communities. RHD prevalence is highest in tamariki aged 5–14 years in Sub-Saharan Africa (5.7 cases per 1000 tamariki aged 5–14 years old) and the Pacific (3.5 cases per 1000 tamariki aged 5–14 years old). High rates are also seen in Indigenous and Pacific peoples in Australia and Aotearoa.[19-21] Infective endocarditis, stroke, arrhythmia, and heart failure are common complications of RHD, contributing to the large numbers of premature deaths globally.[19]

RHD remains a significant contributor to heart failure and related mortality in low- and middle-income countries. Over 80% of global RHD deaths occur in endemic countries, mainly in young people between the ages of 20 and 40 years.[22] 2019 Global Burden of Disease data estimates:[23, 24]

- 2.79 million people are diagnosed with RHD globally each year.
- More than 40.5 million people live with RHD.
- More than 305,600 deaths from RHD globally.
- RHD disability-adjusted life years (DALY) exceeding 10.6 million person-years.[25, 26]

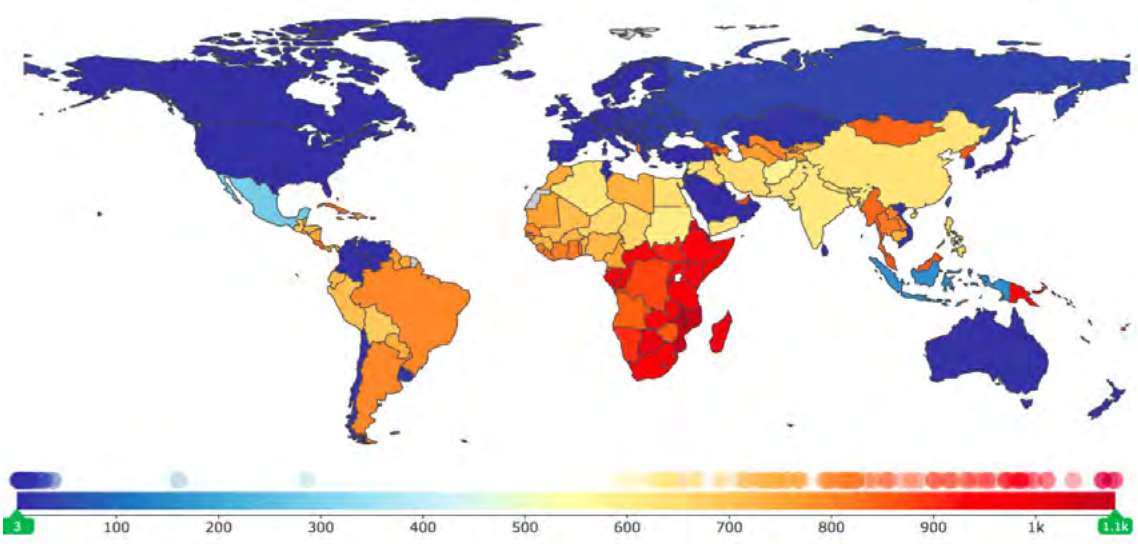

Figure 4.1. Prevalence of rheumatic heart disease globally[27]

Global map showing prevalence/100,000 population in 2017 by country.
Source: healthdata.org

Echocardiographic population-based studies provide a further source of prevalence data. A 2019 meta-analysis of 82 studies from 35 countries showed:

- A pooled prevalence of definite RHD of 1,140/100,000 population (95% CI: 670–1,720/100,000), using World Heart Federation (WHF) 2012 criteria.
- A prevalence of clinically manifested RHD at 470/100,000 population (95% CI: 190–880/100,000).

RHD prevalence is inversely related to country income, decreasing as income increases.[20] The lower a country's income category, the younger the median age, and the more advanced the disease was at presentation.[25, 28] A strong association between social inequality (as expressed by the Gini coefficient) and prevalence of RHD ($p = 0.0002$) has also been shown.[29] However, the true impact of RHD is probably underestimated and uncertain because the most impacted regions typically have the least robust data on RHD prevalence.

Surveillance in Aotearoa

Surveillance involves systematically collecting, analysing, interpreting, and disseminating disease information for defined populations to inform actions to improve health outcomes. Active surveillance involves an active process of collecting an agreed set of data; passive surveillance is the secondary use of data collected for another purpose, e.g. laboratory or hospitalisation data.

There is limited active systematic surveillance of Strep A infections in Aotearoa, and current approaches have limitations that should be considered when interpreting epidemiology.

Establishing national surveillance for Strep A disease is challenging for several reasons:

- Strep A related conditions have different diagnostic approaches:
  - Laboratory testing — Strep A sore throats and skin infection, invasive Strep A.
  - Diagnostic criteria, e.g. ARF, post-streptococcal glomerulonephritis (PSGN).
  - Echocardiogram (echo) — RHD.
- Diagnostic data are dispersed across numerous information systems.
- Barriers to accessing community and primary healthcare introduce bias as higher-risk populations are often under-represented in surveillance data for more minor Strep A diseases like sore throat and skin infections.
- Strep A diagnoses are not always certain e.g.
  - Positive throat and skin swabs can indicate carriage and not infection.
  - Criteria-based approaches are inherently more complex and subjective.

Table 4.1. Surveillance of Strep A diseases in Aotearoa

Strep A Disease	Current surveillance approach	Limitations
Sore throats	• Passive surveillance using laboratory data (throat swabs that are Strep A positive) • Ad hoc district or regional studies • Hospital laboratory data are often not included • Positive test rates vary depending on the context of swabbing, e.g. schools, pharmacies, primary care, public health contact tracing	• No national surveillance • No national laboratory data collection, which means data must be sourced from various community and hospital laboratories • No clinical information to distinguish between Strep A carriage and infection • Limited contextual information on swabbing context to allow adjustment for testing bias • Barriers to healthcare access introduce bias
Skin infections	• Passive surveillance using laboratory data (skin swabs that are Strep A positive) • Ad hoc studies • Hospital laboratory data are often not included • Passive surveillance using skin infection hospitalisation data* which are not limited to Strep A	• No national surveillance • No national laboratory data collection, which means data must be sourced from various community and hospital laboratories • No clinical information to distinguish between Strep A carriage and infection • Limited contextual information on swabbing context to allow adjustment for testing bias • Barriers to healthcare access introduce bias • Hospitalisation data don't reliably identify Strep A as the cause (vs. other common causes, e.g. Staph infection)
Scarlet fever	No current surveillance	• No current surveillance approach • Public health units are frequently contacted about community scarlet fever "outbreaks", but there is no system for recording these. Unless scarlet fever results in hospitalisation, it is unlikely that an increase would be noticed in NZ

Strep A Disease	Current surveillance approach	Limitations
Peri-tonsilar abscess (quinsy)	No current surveillance	• No current surveillance approach • ED and hospital data could likely be used for high-level surveillance
ARF	There are three sources used for ARF surveillance: • National notification data collected in EpiSurv** with regular surveillance reports • District and regional secondary prophylaxis rheumatic fever registers (n=15) used for ad hoc studies and reports • National hospitalisation data* used for national passive surveillance • During the National Rheumatic Fever Prevention Programme (2012–2017), efforts were made to improve EpiSurv data quality by regular triangulation of hospital discharge data, rheumatic fever register notifications data, and notifications to public health[30]	• Limitations exist with all current surveillance approaches as described • Notification data relies on clinicians notifying public health of suspected cases. Older data (pre-2013) is less complete, impacting its use for time-series analysis. Missing data and inconsistent use of diagnostic criteria limit accuracy • Rheumatic fever registers collect disparate data, limiting their use in national surveillance reporting. They are considered the gold standard for ARF surveillance • Hospitalisation data both over and undercounts ARF cases due to International Classification of Diseases (ICD) coding issues
RHD	• Ad hoc district or regional studies provide limited information on RHD prevalence • A retrospective RHD registry cohort study on moderate to severe RHD compiled a dataset from multiple clinical sources	• No national surveillance • No national echo data collection, and existing district IT systems are old with largely un-coded data, making reliable identification of RHD impractical • No prospective collection
Invasive GAS (iGAS)	• National surveillance via EpiSurv** started on 1st Oct 2024 with direct laboratory notification	• Prior to 1st Oct 2024, passive surveillance using laboratory data • Ad hoc studies using laboratory and discharge data provide some information
PSGN	• Ad hoc studies	• No national surveillance • Criteria-based diagnosis limits passive surveillance • ICD coding limits the use of hospitalisation data

* National Minimum Dataset — a national collection of ICD-10 coded hospital admissions

** EpiSurv — national notifiable disease data collection

Surveillance of Strep A-related diseases is complex. Multiple disease entities occur across the lifespan. A wide range of health services are involved in the diagnosis and management of Strep A diseases.

In Aotearoa, Strep A sore throat data comes from community laboratory data, with throat swabbing performed in general practice, emergency departments, school programmes in some regions, and in other regions, pharmacies. A potential bias is that these populations may not be representative of the total population of tamariki. Additionally, tamariki at the highest risk of ARF may not be able to access general practice.

ARF is nationally notifiable in Aotearoa, and ARF cases are recorded across a number of information systems, including:

- District rheumatic fever registers (15 in existence in 2024).
- The National Notifiable Disease Surveillance System (EpiSurv) — ARF only.
- Hospital discharge coding data (the national minimum dataset).

There are strengths and limitations of notification and admission data. Cases of ARF may not be notified to public health, and older EpiSurv data may be unreliable, with implications for time series reporting. ICD discharge coding data tends to mis-classify and overcount ARF and register data.[31, 32] During the National Rheumatic Fever Prevention Programme (2012–2017), efforts were made to improve ARF data quality by regular triangulation of hospital discharge data and register notifications.[30]

RHD is not nationally notifiable in Aotearoa. ICD coding of hospitalisations has also been shown to have poor predictive value for RHD in older and low-risk populations.[33, 34] The Aotearoa New Zealand RHD Registry Project aims to improve understanding of the true burden of RHD. This retrospective cohort study collated data on individuals with moderate and severe RHD from multiple clinical datasets.[2]

Epidemiology of Strep A throat and skin infections in Aotearoa

Sore throat and skin infection are the most common manifestations of Strep A in Aotearoa. Cellulitis, a form of skin infection, is estimated to be the most substantial contributor to the total economic and health burden of all Strep A-related diseases (42.0% of cases).[35] Pacific and Māori tamariki are disproportionately affected.[36, 37] (See **Table 4.2**).

As is the case internationally, tamariki under 15 years have the highest incidence of Strep A throat infection in Aotearoa. There is limited national data available, therefore, data presented here is limited to the Auckland region.

In a school-based study from 1998–2001, tamariki were swabbed if symptomatic. They were also screened monthly to identify undeclared clinical sore throat (an inflamed throat and/or tonsils without a declared sore throat). Seven percent of all throat swabs were positive for Strep A. Yearly rates of one or more positive throat swabs were 39% for Māori tamariki, 35% for Pacific tamaiti, and 20% for tamariki of other ethnicities. Ethnicity, school, and age were highly associated with Strep A (P <0.0001).[38]

An Auckland study analysing Strep A sore throat data from 2010–2016 (covering part of the Rheumatic Fever Prevention Programme period, 2012–2017),[39] found:

- Tamariki 5–9 years of age had the highest rates of Strep A sore throat (82.9 cases/1,000 person-years), followed by tamariki 10–14 years of age (44.3 cases/1,000 person-years).
- Among tamariki 5–14 years of age in primary healthcare (general practice) clinics, the incidence of Strep A sore throat was significantly higher among Pacific (99.6 cases/1,000 person-years) and Māori (79.0 cases/1,000 person-years) than among non-Māori, non-Pacific peoples (58.3 cases/1,000 person-years). Incidence increased with deprivation.[39]
- The incidence of Strep A sore throat was even higher in school-based clinics, which were mainly in South Auckland (121.8 Strep A positive swabs/1,000 person-years).[39]

In a further analysis of the results of throat swab cultures for the total Auckland population from 2010–2017, Strep A was detected in 14.3% of throat swabs. Māori and Pacific peoples had a slightly higher proportion of Strep A detection from throat swabs than non-Māori, non-Pacific peoples (Māori RR: 1.03, Pacific RR: 1.01, p < 0.01).[40]

Overall, incidence estimates from these studies suggest Pacific and Māori tamariki in the Auckland region are 1.3–4.8 times more likely than other ethnicities to suffer a sore throat from which Strep A is cultured. This incidence may partly reflect increased access to school-based throat swabbing and adherence to clinical guidelines in areas of high risk for ARF through the Mana Kidz programme in South Auckland schools since 2011.[41]

Pacific and Māori tamariki are also more likely than other tamariki to have a positive skin swab for Strep A (Pacific 5–7 times more likely and Māori 4–5 times more likely).[40, 42] This data must be interpreted cautiously due to variable access to primary healthcare and testing practices. Multivariate analysis found significant associations between Strep A skin infection and:[43]

- History of eczema.
- Household crowding.
- Access barriers to primary care.
- Having four Māori or Pacific grandparents.[43]

Invasive Strep A infections

Prior to 1 October 2024, invasive Strep A was not nationally notifiable in Aotearoa, although blood cultures were sent to the national reference laboratory, and some monitoring occurred via passive surveillance. From 2002–2012, the invasive Strep A incidence rate, based on national laboratory data, was approximately seven times higher for Pacific peoples than for non-Māori, non-Pacific peoples, while the rate among Māori was approximately three times higher.[44]

Invasive Strep A infections dropped markedly in 2021, possibly related to public health measures for the COVID-19 pandemic. Rates of invasive Strep A sharply increased in 2023, with 591 cases reported by passive surveillance (11.3 per 100,000).[45]

From 1 October 2024, invasive Strep A infections became nationally notifiable in Aotearoa. Details of surveillance and approaches to case and contact management can be found in the Communicable Disease Control Manual.

Post-streptococcal glomerulonephritis

PSGN is another autoimmune-mediated sequela of Strep A infection. It is not nationally notifiable, but cohort studies from the Auckland region show similar disparities in incidence according to ethnicity. Between 2007 and 2015, the average annual incidence in the Auckland region was 15.2 cases/100,000 population (95%, CI: 14.9–15.6). Rates among Pacific tamaiti were 17 times higher than in non-Māori, non-Pacific peoples, and rates among Māori were 7 times higher than in non-Māori, non-Pacific peoples.[46]

Epidemiology of acute rheumatic fever in Aotearoa

ARF mainly affects Māori and Pacific tamariki aged 5–14 years in socioeconomically deprived areas of the upper North Island. Around half of cases live in the Auckland region. ARF is rare under 4 years of age. Over 90% of notified cases (confirmed or probable) are under the age of 30 years.[47, 48]

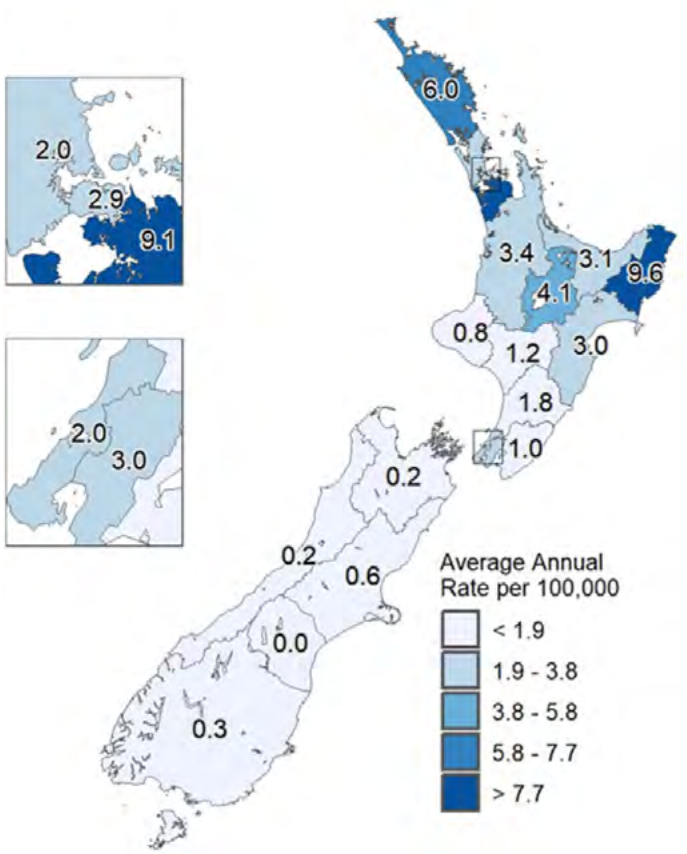

Figure 4.2. Map showing rates of acute rheumatic fever notifications by health district 2010–2022

Rates of ARF in Aotearoa, documented since early surveys in the 1900s, have always been high. In the 1920s, surveys of school records suggested an approximate incidence in school tamariki of 65 cases/100,000 population.[49] By the 1970s, total population rates had dropped significantly,[50] but evidence existed of much higher rates in rural and lower socioeconomic areas (for example, evidence from the Wairoa College Study[51]).

The overall rate of ARF in Aotearoa has not changed significantly since the mid-1990s, but ethnic inequities have increased. Between 1996 and 2005, Māori and Pacific peoples made up 83% of new ARF diagnoses — in 2024, they made up 91%. However, some of this change is likely due to improved data with the introduction of ethnicity data protocols for the health sector in 2004. In the decade from 1996–2005, ARF hospitalisations averaged 125 a year, an average annual rate of 3.4 per 100,000. However, rates for Māori and Pacific peoples increased, while those for non-Māori, non-Pacific peoples decreased by nearly two-thirds. Māori rates were 10 times higher than those of Pākehā. Pacific rates were 20.7 times higher.[52]

An analysis of initial ARF hospitalisations from 2000–2018 in Aotearoa[3] found an average rate of 3.4 hospitalisations per 100,000 (unchanged from the late 1990s). Between 2000–2009 and 2010–2018, annual rates of initial ARF hospitalisation increased significantly for rangatahi aged 15–19 and 20–29 years ($p < 0.05$) but remained steady in other age groups. This study reported persisting inequities in ARF incidence according to ethnicity:

- Māori accounted for 48.9% of cases, and Pacific peoples for 43.7%.[3]
- Initial ARF hospitalisation rates peaked at 35.9 cases/100,000 population among Māori 5–14 years of age and at 79.6 cases/100,000 population for Pacific tamaiti. By comparison, rates for non-Māori, non-Pacific tamariki were 1.6 cases/100,000 population.
- Tamariki Māori had nearly 20 times the risk of ARF (RR 19.6). Pacific tamaiti had nearly 45 times the risk of ARF compared to non-Māori, non-Pacific tamariki (RR 44.5).
- When corrected for age, sex, socioeconomic deprivation, and location, a large residual increased risk remained (RR 9.0 for Māori and RR 16.6 for Pacific peoples).
- There was no decrease in rates over the period for Māori. Rates increased significantly for Pacific peoples compared with all other ethnicities.

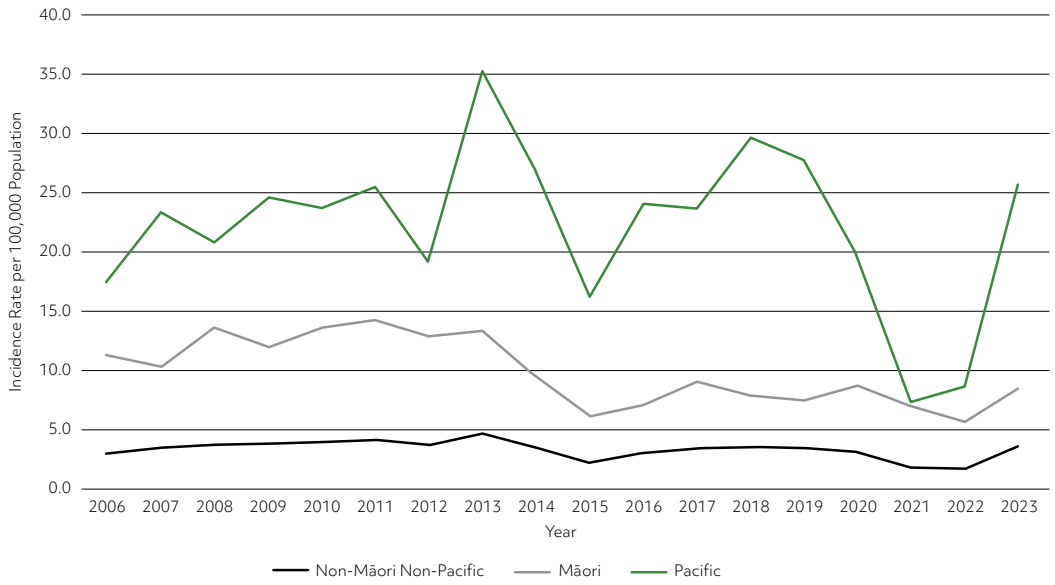

Figure 4.3. First episode acute rheumatic fever hospitalisations, annual rate per 100,000, by ethnic group, Aotearoa, 2006–2023[47]

The study also found inequities according to socioeconomic status and geographic location:

- Rates were 25 times higher for those living in the most socioeconomically disadvantaged neighbourhoods (NZDep Q5) compared with the least (NZDep Q1).[3] In addition, the most socioeconomically disadvantaged neighbourhoods had greater increases in ARF hospitalisations than more advantaged neighbourhoods.
- Large geographical inequities were also apparent. South Auckland had the highest rate for initial ARF hospitalisation among persons under 30 years of age (21.7 cases/100,000 population). But rates also were high in Northland/Te Tai Tokerau (17.4 cases/100,000 population) and the East Coast/Te Tai Rāwhiti (16.0 cases/100,000 population).[3]

In 2023, ARF hospitalisation rates returned to those of pre-pandemic levels. There were 158 confirmed and probable cases nationally for 2023 (compared with 75 notifications in 2022; 73 in 2021, and an average of 153 per year during 2018-2020, as per preliminary ESR data).[48, 53] In the Auckland region, ARF increased mainly in South Auckland.[54]

Epidemiology of recurrent acute rheumatic fever in Aotearoa

Recurrent ARF is associated with RHD progression. Between 1993 and 1999, an audit of the Auckland Regional Rheumatic Fever Register data identified 20 definite or probable recurrences among 360 episodes of ARF. The programme failure rate was 1.4/100 patient years and non-adherence accounted for 55% of recurrences.[55]

A retrospective national observational study of recurrent ARF covering the period 2010–2014, conducted using a detailed review of clinical records by clinical experts, found:

- An overall recurrence rate of 7.2% at a median age of 21.6 years.
- 83% of recurrences occurred in people over 15 years.
- Arthritis and carditis were the most common major manifestations.[1]

An analysis of discharge coding data from 2000–2018 found a slightly higher recurrence rate (9.5%) of ARF admissions.[3] As clinical records were not reviewed, it is likely that a number of re-admissions were misclassified as recurrences. Additionally, hospitalisation data overcounts ARF compared to registers, further compounding bias.[32]

Epidemiology of rheumatic heart disease in Aotearoa

Current trends in RHD prevalence in Aotearoa reflect patterns of ARF over the past few decades. They may also reflect changes in clinical awareness and diagnostic practices, notably the availability of echo. RHD may occur as a sequelae of ARF or may present years to decades later.[2, 3, 56] Aotearoa New Zealand RHD registry project data shows 40% of people with moderate to severe RHD had no prior history of ARF.[2] Echo screening in 2014–2016 in South Auckland suggests that up to 1 in 50 Pacific young adults may be living with RHD. This proportion is higher than suggested by current ARF rates.[17]

Noting the limitations of RHD hospitalisation data, Bennett et al. found that RHD hospitalisation rates increased from 2000–2018.[3] The average annual total rate of RHD hospitalisations as the principal diagnosis was 14.3 cases/100,000 population, including initial and repeat admissions.[3] Māori and Pacific peoples were most affected by RHD. They were also affected younger, accounting for 50.5% of all people affected under 70 years of age.[3]

An analysis of Aotearoa New Zealand RHD Registry project data suggests a doubling of cases of significant RHD in the last two decades. The registry consists of people identified with moderate or severe RHD by 2019. The increase may reflect rising disease increases, population increases, and improved recognition of RHD via wider application of echo. Improvements in secondary prophylaxis and healthcare for those with RHD may have also led to a cohort effect with greater numbers of people with moderate and severe RHD living longer.[2] Key findings of the RHD Registry project include:

- The average age of RHD presentation was 38.4 years.
- RHD prevalence fell among non-Māori, non-Pacific peoples but increased in Māori and Pacific peoples, both as a proportion of cases and in absolute numbers.[2]
- 64% of the cohort had at least one cardiac intervention at an average age of 40 years.
- 26.9% of the cohort had died at the latest follow-up.

Rheumatic heart disease in pregnancy

In those with RHD, pregnancy poses risks for both the pregnant person and their pēpi (baby). The increased cardiac demands of pregnancy can worsen clinical symptoms, unmask undiagnosed RHD, and lead to poorer pregnancy outcomes. (See **Chapter 12: Rheumatic Heart Disease and Pregnancy**.) Prevalence is around 0.18% for Māori and 0.48% for Pacific.[57-59] In a prospective cohort study of 122 pregnancies in 113 women with RHD in Aotearoa, serious RHD (severe regurgitation or stenosis, heart failure, or current or historical surgical repair) was present in 25% of the women.[59]

Rheumatic heart disease mortality

On average, 150 people die from RHD every year in Aotearoa.[3, 60] RHD mortality rates declined from 2000–2016, but Māori and Pacific peoples were disproportionately affected at younger ages, accounting for 73.8% of deaths among those under 70 years.[3]

Māori and Pacific peoples are around 11 times more likely to die from RHD than non-Māori, non-Pacific peoples.[3] They also die much younger, at an average of 55 years and 59 years, respectively, compared with 80 years in non-Māori, non-Pacific peoples.[2] As with ARF and RHD hospitalisations, the increased risk for Māori and Pacific peoples is associated with socioeconomic deprivation and location but not fully explained by those factors.[3]

Summary: inequitable rates

Table 4.2 presents estimates of rate ratios of Strep A infection, ARF, RHD hospitalisations, and deaths. It compares figures from studies between 1996 and 2021 for Māori and Pacific peoples with non-Māori, non-Pacific peoples (= 1). The table shows the extent and persistence of inequities in these conditions in Aotearoa. Although the methodology and time periods vary, the broad trend is clear: ARF, PSGN, and RHD deaths demonstrate the largest inequities.

Table 4.2. Inequities in Strep A disease, acute rheumatic fever, rheumatic heart disease hospitalisations and rheumatic heart disease deaths

Rate ratios for Māori and Pacific peoples (rate ratios compared to non-Māori, non-Pacific = 1)

Condition	Māori	Pacific peoples
Strep A sore throat in tamariki 5–14 years in primary care rate ratios Auckland region, 2010–2016[39]	1.3	1.7
Strep A sore throat in tamariki 5–14 years in primary care and school programmes rate ratios Auckland region, 2010–2016[39]	3.4	4.8
Strep A skin infections in community <20 years rate ratios Auckland region, 2010–2016[42] and 2010–2017[40]	4.0–4.9	5.4–6.8
All-cause skin infection hospitalisations in tamariki 0–14 years rate ratios (note Staphylococcus aureus accounts for many skin infections in childhood) 2004–2014 Aotearoa[36]	3.51	4.17
Invasive Strep A in total population rate ratios 2017–2022[61]	3.3	5.4
Post-streptococcal glomerulonephritis in tamariki <15 years rate ratios Auckland region, 2007–2015[46]	6.8	17.3
Acute rheumatic fever		
1996–2005 total population, age-standardised rates[52]	10	20.7
2010–2016 <20 years crude rate ratios[42]	30.3	69.7
2010–2013 total population*[62]	14.5	20.6
2000–2018 <30 years crude rate ratios[3]	19.6	44.5
2000–2018 <30 years adjusted rate ratios*[3]	11.8	23.6
2000–May 2021 <15 years crude rate ratios[63]	23.1	52.6
Rheumatic heart disease hospitalisation		
2000–2018 <70 years adjusted rate ratios*[3]	3.2	4.6
2000–2022 <75 years adjusted rate ratios** (WTM)[64]	4.3	9.2
Rheumatic heart disease deaths under 70 years		
2000–2018 <70 years adjusted rate ratios*[3]	12.3	11.2
2000–2022 <75 years adjusted rate ratios** (WTM)[64]	11.2	11.7

* aRR adjusted for age, sex and socioeconomic deprivation

** aRR adjusted for age, sex, district health board, socioeconomic deprivation and year

WTM: Whāia Te Māramatanga

References

1. Dennison A, Peat B, Wilson E, Leversha A, Wheeler M, Briggs S, et al. Rheumatic fever recurrences in New Zealand 2010-14. *New Zealand Medical Journal*. 2020;133(1516):47–57.

2. Tilton E, Mitchelson B, Anderson A, Peat B, Jack S, Lund M, et al. Cohort profile: methodology and cohort characteristics of the Aotearoa New Zealand Rheumatic Heart Disease Registry. *BMJ Open*. 2022;12(12):e066232. https://doi.org/10.1136/bmjopen-2022-066232

3. Bennett J, Zhang J, Leung W, Jack S, Oliver J, Webb R, et al. Rising ethnic inequalities in acute rheumatic fever and rheumatic heart disease, New Zealand, 2000–2018. *Emerging Infectious Diseases*. 2021;27(1):36–46. https://doi.org/10.3201/eid2701.191791

4. Gordis L. The virtual disappearance of rheumatic fever in the United States: lessons in the rise and fall of disease. T. Duckett Jones memorial lecture. *Circulation*. 1985;72(6):1155–1162. https://doi.org/10.1161/01.cir.72.6.1155

5. Bland EF. Rheumatic fever: the way it was. *Circulation*. 1987;76(6):1190–1195. https://doi.org/10.1161/01.cir.76.6.1190

6. Coffey PM, Ralph AP, Krause VL. The role of social determinants of health in the risk and prevention of group A streptococcal infection, acute rheumatic fever and rheumatic heart disease: a systematic review. *PLoS Neglected Tropical Diseases*. 2018;12(6):e0006577. https://doi.org/10.1371/journal.pntd.0006577

7. Baker MG, Masterson MY, Shung-King M, Beaton A, Bowen AC, Bansal GP, et al. Research priorities for the primordial prevention of acute rheumatic fever and rheumatic heart disease by modifying the social determinants of health. *BMJ Global Health*. 2023;8(Suppl 9). https://doi.org/10.1136/bmjgh-2023-012467

8. Barnett TC, Bowen AC, Carapetis JR. The fall and rise of group A streptococcus diseases. *Epidemiology and Infection*. 2018;147:e4. https://doi.org/10.1017/S0950268818002285

9. Auala T, Zavale BG, Mbakwem AC, Mocumbi AO. Acute rheumatic fever and rheumatic heart disease: highlighting the role of group A streptococcus in the global burden of cardiovascular disease. *Pathogens*. 2022;11(5). https://doi.org/10.3390/pathogens11050496

10. Miller KM, Carapetis JR, Van Beneden CA, Cadarette D, Daw JN, Moore HC, et al. The global burden of sore throat and group A streptococcus pharyngitis: a systematic review and meta-analysis. *EClinicalMedicine*. 2022;48:101458. https://doi.org/10.1016/j.eclinm.2022.101458

11. Sims Sanyahumbi A, Colquhoun S, Wyber R, Carapetis JR. Global disease burden of group a streptococcus. In: Ferretti JJ, Stevens DL, Fischetti VA, editors. Streptococcus pyogenes: basic biology to clinical manifestations. Oklahoma City: University of Oklahoma Health Sciences Center; 2016.

12. Dougherty S, Carapetis J, Zühlke L, Wilson N. Acute rheumatic fever and rheumatic heart disease. Amsterdam: Elsevier; 2020.

13. Tibazarwa KB, Volmink JA, Mayosi BM. Incidence of acute rheumatic fever in the world: a systematic review of population-based studies. *Heart*. 2008;94(12):1534–1540. https://doi.org/10.1136/hrt.2007.141309

14. Karthikeyan G, Guilherme L. Acute rheumatic fever. *Lancet*. 2018;392(10142):161–174. https://doi.org/10.1016/S0140-6736(18)30999-1

15. Jackson SJ, Steer AC, Campbell H. Systematic review: estimation of global burden of non-suppurative sequelae of upper respiratory tract infection: rheumatic fever and post-streptococcal glomerulonephritis. *Tropical Medicine and International Health*. 2011;16(1):2–11. https://doi.org/10.1111/j.1365-3156.2010.02670.x

16. Seckeler MD, Hoke TR. The worldwide epidemiology of acute rheumatic fever and rheumatic heart disease. *Clinical Epidemiology*. 2011;3:67–84. https://doi.org/10.2147/CLEP.S12977

17. Webb R, Culliford-Semmens N, ChanMow A, Doughty R, Tilton E, Peat B, et al. High burden of rheumatic heart disease confirmed by echocardiography among Pacific adults living in New Zealand. *Open Heart*. 2023;10(1):e002253. https://doi.org/10.1136/openhrt-2023-002253

18. Oliver J, Robertson O, Zhang J, Marsters BL, Sika-Paotonu D, Jack S, et al. Ethnically disparate disease progression and outcomes among acute rheumatic fever patients in New Zealand, 1989–2015. *Emerging Infectious Diseases*. 2021;27(7):1893–1902. https://doi.org/10.3201/eid2707.203045

19. Zühlke LJ, Beaton A, Engel ME, Hugo-Hamman CT, Karthikeyan G, Katzenellenbogen JM, et al. Group A streptococcus, acute rheumatic fever and rheumatic heart disease: epidemiology and clinical considerations. *Current Treatment Options in Cardiovascular Medicine*. 2017;19(2):15. https://doi.org/10.1007/s11936-017-0513-y

20. Noubiap JJ, Agbor VN, Bigna JJ, Kaze AD, Nyaga UF, Mayosi BM. Prevalence and progression of rheumatic heart disease: a global systematic review and meta-analysis of population-based echocardiographic studies. *Scientific Reports*. 2019;9(1):17022. https://doi.org/10.1038/s41598-019-53540-4

21. Carapetis JR, Steer AC, Mulholland EK, Weber M. The global burden of group A streptococcal diseases. *Lancet Infectious Diseases*. 2005;5(11):685–694. https://doi.org/10.1016/S1473-3099(05)70267-X

22. Rwebembera J, Beaton AZ, de Loizaga SR, Rocha RTL, Doreen N, Ssinabulya I, et al. The global impact of rheumatic heart disease. *Current Cardiology Reports*. 2021;23(11):160. https://doi.org/10.1007/s11886-021-01592-2

23. Watkins DA, Johnson CO, Colquhoun SM, Karthikeyan G, Beaton A, Bukhman G, et al. Global, regional, and national burden of rheumatic heart disease, 1990–2015. *New England Journal of Medicine*. 2017;377(8):713–722. https://doi.org/10.1056/NEJMoa1603693

24. Watkins DA, Beaton AZ, Carapetis JR, Karthikeyan G, Mayosi BM, Wyber R, et al. Rheumatic heart disease worldwide: JACC Scientific Expert Panel. *Journal of the American College of Cardiology*. 2018;72(12):1397–1416. https://doi.org/10.1016/j.jacc.2018.06.063

25. Zhang L, Tong Z, Han R, Li K, Zhang X, Yuan R. Spatiotemporal trends in global burden of rheumatic heart disease and associated risk factors from 1990 to 2019. *International Journal of Cardiology*. 2023;384:100–106. https://doi.org/10.1016/j.ijcard.2023.04.060

26. Yu G, Gong X, Xu Y, Sun H, Liu Y, Zhai C, et al. The global burden and trends of four major types of heart disease, 1990-2019: a systematic analysis for the Global Burden of Disease Study 2019. *Public Health*. 2023;220:1–9. https://doi.org/10.1016/j.puhe.2023.04.005

27. Kumar RK, Antunes MJ, Beaton A, Mirabel M, Nkomo VT, Okello E, et al. Contemporary diagnosis and management of rheumatic heart disease: implications for closing the gap: a scientific statement from the American Heart Association. *Circulation*. 2020;142(20):e337–e357. https://doi.org/10.1161/CIR.0000000000000921

28. French KA, Poppas A. Rheumatic heart disease in pregnancy: global challenges and clear opportunities. *Circulation*. 2018;137(8):817–819. https://doi.org/10.1161/CIRCULATIONAHA.118.033465

29. Rothenbuhler M, O'Sullivan CJ, Stortecky S, Stefanini GG, Spitzer E, Estill J, et al. Active surveillance for rheumatic heart disease in endemic regions: a systematic review and meta-analysis of prevalence among children and adolescents. *Lancet Global Health*. 2014;2(12):e717–726. https://doi.org/10.1016/S2214-109X(14)70310-9

30. Oliver J, Pierse N, Williamson DA, Baker MG. Estimating the likely true changes in rheumatic fever incidence using two data sources. *Epidemiology and Infection*. 2018;146(2):265–275. https://doi.org/10.1017/S0950268817002734

31. Atatoa-Carr P, Bell A, Lennon DR. Acute rheumatic fever in the Waikato District Health Board region of New Zealand: 1998–2004. *New Zealand Medical Journal*. 2008;121(1285):96–105.

32. Moxon TA, Reed P, Jelleyman T, Anderson P, Leversha A, Jackson C, et al. Is a rheumatic fever register the best surveillance tool to evaluate rheumatic fever control in the Auckland region? *New Zealand Medical Journal*. 2017;130(1460):48–62.

33. Fitz-Gerald JA, Ongzalima CO, Ng A, Greenland M, Sanfilippo FM, Hung J, et al. A validation study: how predictive is a diagnostic coding algorithm at identifying rheumatic heart disease in Western Australian hospital data? *Heart, Lung & Circulation*. 2020;29(8):e194–e199. https://doi.org/10.1016/j.hlc.2019.08.020

34. Katzenellenbogen JM, Nedkoff L, Cannon J, Kruger D, Pretty F, Carapetis JR, et al. Low positive predictive value of International Classification of Diseases, 10th Revision codes in relation to rheumatic heart disease: a challenge for global surveillance. *Internal Medicine Journal*. 2019;49(3):400–403. https://doi.org/10.1111/imj.14221

35. Cannon JW, Zhung J, Bennett J, Moreland NJ, Baker MG, Geelhoed E, et al. The economic and health burdens of diseases caused by group A streptococcus in New Zealand. *International Journal of Infectious Diseases*. 2021;103:176–181. https://doi.org/10.1016/j.ijid.2020.11.193

36. Lim A, Rumball-Smith J, Jones R, Kawachi I. The rise and fall of hospitalizations for skin infections in New Zealand, 2004–2014: trends by ethnicity and socioeconomic deprivation. *Epidemiology and Infection*. 2017;145(4):678–684. https://doi.org/10.1017/S0950268816002685

37. O'Sullivan CE, Baker MG, Zhang J. Increasing hospitalizations for serious skin infections in New Zealand children, 1990–2007. *Epidemiology and Infection*. 2011;139(11):1794–1804. https://doi.org/10.1017/S0950268810002761

38. Lennon D, Stewart J, Farrell E, Palmer A, Mason H. School-based prevention of acute rheumatic fever: a group randomized trial in New Zealand. *Pediatric Infectious Disease Journal*. 2009;28(9):787–794. https://doi.org/10.1097/INF.0b013e3181a282be

39. Oliver J, Upton A, Jack SJ, Pierse N, Williamson DA, Baker MG. Distribution of streptococcal pharyngitis and acute rheumatic fever, Auckland, New Zealand, 2010–2016. *Emerging Infectious Diseases*. 2020;26(6):1113–1121. https://doi.org/10.3201/eid2606.181462

40. Oliver J, Bennett J, Thomas S, Zhang J, Pierse N, Moreland NJ, et al. Preceding group A streptococcus skin and throat infections are individually associated with acute rheumatic fever: evidence from New Zealand. *BMJ Global Health*. 2021;6(12). https://doi.org/10.1136/bmjgh-2021-007038

41. King J, Moss M, Spee KE. Evaluation of school-based health services in primary and intermediate schools (Mana Kidz). Auckland: Counties Manukau Health; 2022. https://www.nhc.maori.nz/wp-content/uploads/2022/08/220601-Mana-Kidz-evaluation-final-report.pdf (Accessed February 11 2025).

42. Thomas S, Bennett J, Jack S, Oliver J, Purdie G, Upton A, et al. Descriptive analysis of group A streptococcus in skin swabs and acute rheumatic fever, Auckland, New Zealand, 2010–2016. *The Lancet Regional Health — Western Pacific*. 2021;8:100101. https://doi.org/10.1016/j.lanwpc.2021.100101

43. Bennett J, Moreland NJ, Zhang J, Crane J, Sika-Paotonu D, Carapetis J, et al. Risk factors for group A streptococcal pharyngitis and skin infections: a case control study. *The Lancet Regional Health — Western Pacific*. 2022;26:100507. https://doi.org/10.1016/j.lanwpc.2022.100507

44. Williamson DA, Morgan J, Hope V, Fraser JD, Moreland NJ, Proft T, et al. Increasing incidence of invasive group A streptococcus disease in New Zealand, 2002–2012: a national population-based study. *Journal of Infection*. 2015;70(2):127–134. https://doi.org/10.1016/j.jinf.2014.09.001

45. ESR. Invasive group A streptococcal infections in New Zealand — update 18 September 2023. Presentation to Public Health Association hui [meeting]. New Zealand: ESR; 2023.

46. Vogel AM, Lennon DR, van der Werf B, Diack M, Neutze JM, Horsfall M, et al. Post-streptococcal glomerulonephritis: some reduction in a disease of disparities. *Journal of Paediatrics and Child Health*. 2019;55(6):652–658. https://doi.org/10.1111/jpc.14263

47. Health New Zealand | Te Whatu Ora. Reducing rheumatic fever. Health New Zealand | Te Whatu Ora; 2024. https://www.tewhatuora.govt.nz/for-the-health-sector/health-sector-guidance/diseases-and-conditions/rheumatic-fever-guidance/reducing-rheumatic-fever/ (Accessed February 18 2025).

48. ESR. Summary of rheumatic fever notifications 1 January–30 June 2023. Porirua: ESR; 2023.

49. Stanhope JM. New Zealand trends in rheumatic fever: 1885–1971. *New Zealand Medical Journal*. 1975;82(551):297–299.

50. Wabitsch KR, Prior IA, Stanley DG, Pearce N. New Zealand trends in acute rheumatic fever and chronic rheumatic heart disease 1971–1981. *New Zealand Medical Journal*. 1984;97(763):594–597.

51. Frankish JD, Stanhope JM, Martin DR, Clarkson PM, Leslie PN, Langley RB. Rheumatic fever and streptococci: the Wairoa College study. *New Zealand Medical Journal*. 1978;87(604):33–38.

52. Jaine R, Baker M, Venugopal K. Epidemiology of acute rheumatic fever in New Zealand 199–2005. *Journal of Paediatrics and Child Health*. 2008;44(10):564–571. https://doi.org/10.1111/j.1440-1754.2008.01384.x

53. ESR. Monthly notifiable disease surveillance report Dec 2023. 2023. https://www.esr.cri.nz/digital-library/monthly-notifiable-disease-surveillance-report-dec-2023/ (Accessed February 18 2025).

54. National Public Health Service Te Whatu Ora. Acute rheumatic fever notifications to Auckland Regional Public Health Service 1 April 2023 to 30 June 2023. 2023. https://view.officeapps.live.com/op/view.aspx?src=https%3A%2F%2Fwww.tewhatuora.govt.nz%2Fassets%2FUploads%2F20230420-Rheumatic-Fever-Report-2022-Public.xlsx&wdOrigin=BROWSELINK (Accessed February 11 2025).

55. Spinetto H, Lennon D, Horsburgh M. Rheumatic fever recurrence prevention: a nurse-led programme of 28-day penicillin in an area of high endemnicity. *Journal of Paediatrics and Child Health*. 2011;47(4):228–234. https://doi.org/10.1111/j.1440-1754.2010.01942.x

56. McLintock C. Still casting its long shadow: rheumatic heart disease in Australia and New Zealand. *Internal Medicine Journal*. 2012;42(9):963–966. https://doi.org/10.1111/j.1445-5994.2012.02871.x

57. Sadler L, McCowan L, White H, Stewart A, Bracken M, North R. Pregnancy outcomes and cardiac complications in women with mechanical, bioprosthetic and homograft valves. *BJOG: An International Journal of Obstetrics and Gynaecology*. 2000;107(2):245–253. https://doi.org/10.1111/j.1471-0528.2000.tb11696.x

58. North RA, Sadler L, Stewart AW, McCowan LM, Kerr AR, White HD. Long-term survival and valve-related complications in young women with cardiac valve replacements. *Circulation*. 1999;99(20):2669–2676. https://doi.org/10.1161/01.cir.99.20.2669

59. Sullivan EA, Vaughan G, Li Z, Peek MJ, Carapetis JR, Walsh W, et al. The high prevalence and impact of rheumatic heart disease in pregnancy in First Nations populations in a high-income setting: a prospective cohort study. *BJOG: An International Journal of Obstetrics and Gynaecology*. 2020;127(1):47–56. https://doi.org/10.1111/1471-0528.15938

60. Wilson N. Rheumatic heart disease in indigenous populations--New Zealand experience. *Heart, Lung & Circulation*. 2010;19(5-6):282–288. https://doi.org/10.1016/j.hlc.2010.02.021

61. ESR. Invasive group A streptococcal infection in New Zealand, 2017–2022. Porirua: ESR; 2023. https://www.esr.cri.nz/media/4zwlsolx/final-invasive-gas-in-nz-2017-2022-report_feb2024-update.pdf (Accessed February 11 2025).

62. Gurney JK, Stanley J, Baker MG, Wilson NJ, Sarfati D. Estimating the risk of acute rheumatic fever in New Zealand by age, ethnicity and deprivation. *Epidemiology and Infection*. 2016;144(14):3058–3067. https://doi.org/10.1017/s0950268816001291

63. Oben G, Duncanson M, Adams J, Satyanand T. State of child health: acute rheumatic fever in Aotearoa New Zealand. *Journal of the Royal Society of New Zealand*. 2023;53(5):631–640. https://doi.org/10.1080/03036758.2022.2113102

64. Wright K, Dennison A, Mills C, van der werf B. Whāia te Māramatanga: incidence and trends in acute rheumatic fever and rheumatic heart disease, 2000–2022. Auckland: University of Auckland; 2024. (Unpublished).

5

Primary Prevention of Acute Rheumatic Fever: Sore Throat Diagnosis and Management

In this chapter

Key changes

Changes have been made in three main areas.

Assessing the risk of ARF

Modifications have been made to **Algorithm 1: Assessment and management of sore throats in Aotearoa**. These changes focus on high-risk age groups and align more accurately with current acute rheumatic fever (ARF) epidemiology in Aotearoa. Of people hospitalised with ARF, 93% are Māori and Pacific peoples under 35 years of age.[1, 2, 3]

Diagnosis of Strep A sore throat

- Rapid antigen diagnostic tests (RADT) are not recommended in Aotearoa.
- Culture remains the gold standard.
- Rapid molecular tests are recommended to support timely diagnosis of Strep A sore throat in high-incidence ARF populations as part of community 'test and treat' services.
- Nationally coordinated clinical governance and oversight of rapid testing for Strep A is urgently needed.

Antibiotic treatment of Strep A sore throat

- Phenoxymethylpenicillin dosing has been simplified to twice daily dosing.
- Recommendations have been added for the administration of intramuscular (IM) benzathine penicillin.
- Roxithromycin is no longer recommended for people with documented penicillin allergy, while erythromycin remains available for this indication.

Key points

- In Aotearoa, the primary reason for treating Strep A sore throat is to prevent ARF. Historical studies (pre-1960s) show that treating Strep A sore throat with antibiotics reduces the risk of ARF by up to two-thirds.[4]
- Assessing the risk of ARF in patients is key to appropriate clinical management.
- People at low risk of ARF usually only require their symptoms to be managed and neither swabbing nor antibiotics are usually indicated.
- Strep A sore throat should be treated with a 10-day course of antibiotics for people at higher risk of ARF. **Algorithm 1**, **Algorithm 2**, and **Algorithm 3** show the approach to assessment and diagnosis, and **Table 5.1** shows recommended antibiotics for treating Strep A sore throat.

Algorithm 1: Assessment and management of sore throats in Aotearoa

See Table 5.1 for recommended antibiotic treatment for Strep A sore throat (Grade A).

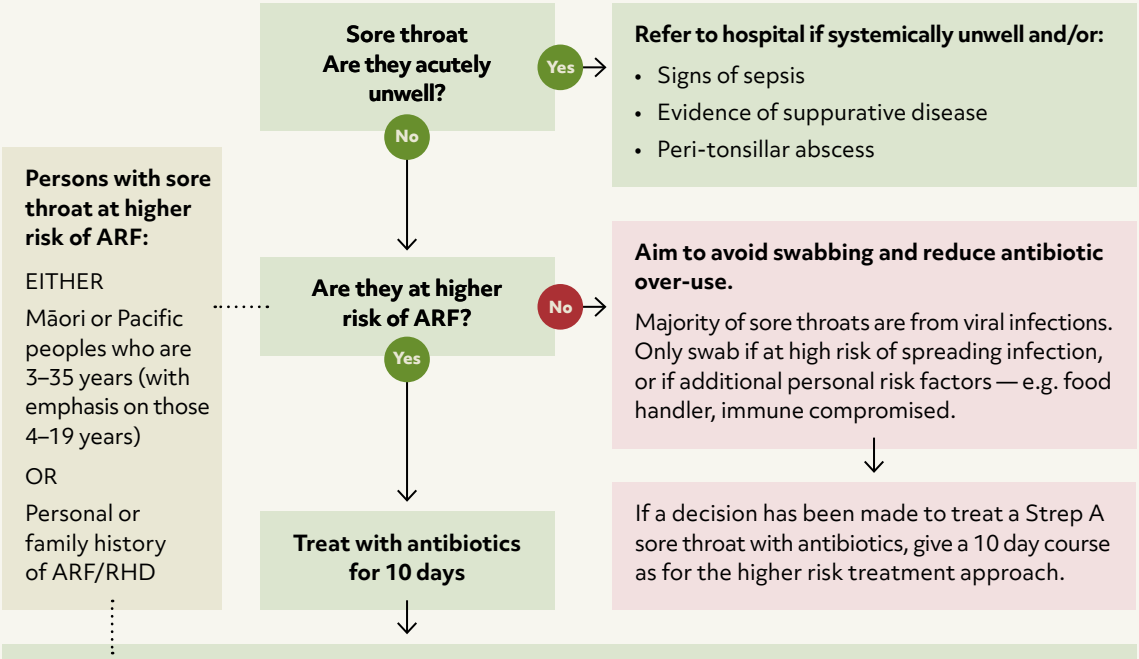

Table 5.1. Recommended antibiotic treatment for Strep A sore throat

Antibiotic and type of patient	Dose
Phenoxymethylpenicillin (Pen V)	15mg/kg (maximum 500mg/dose) two times daily PO
Amoxicillin	50mg/kg (max 1000mg/dose) once daily PO
Benzathine penicillin Single dose — tamariki <20kg	600,000 international units (450mg) IM Use with lignocaine and distraction techniques. See Chapter 9: Administration of Intramuscular Benzathine Penicillin (Bicillin®L-A)
Benzathine penicillin Single dose — tamariki ≥20kg and adults	1,200,000 international units (900mg) IM Use with lignocaine and distraction techniques. See Chapter 9: Administration of Intramuscular Benzathine Penicillin (Bicillin®L-A)
Erythromycin ethyl succinate — tamariki and adults, for documented penicillin anaphylaxis or suspected true penicillin allergy (see Table 5.3)	20mg/kg/dose two times daily for 10 days (Max 1.6 g daily)

Algorithm 2: Guide for whānau/household Strep A sore throat management in populations at high risk of ARF

Person has Strep A sore throat and is at higher risk of ARF.
Treat as per Algorithm 1.

AND

Does the person have a personal, whānau or household history of ARF?

OR

Has this whānau/household had 3 or more cases of Strep A pharyngitis within the last 3 months?

Persons with sore throat at higher risk of ARF:

EITHER

Māori or Pacific peoples who are 3–35 years (with emphasis on those 4–19 years)

OR

Personal or family history of ARF/RHD

Yes

Offer throat swabbing to all whānau/household, with an emphasis on those aged 4–19 years, whether symptomatic or not.

Treat those who are Strep A positive.*

No

Check for any symptomatic household members and offer throat swabbing.

Treat those who are Strep A positive.*

If one or more household members are Strep A positive:

- Use a health literacy approach:
  - Ask what the whānau understands about treating sore throats to prevent rheumatic fever.
  - Fill in any gaps in understanding.
  - Help them solve problems so they can complete their treatment.
  - Discuss the importance of basic hygiene practices (hand washing, sneeze/cough into elbow or a tissue).
- Assess and treat any skin infections and treat as per local HealthPathways.
- Swabbing at end of treatment is not recommended except for recurrent Strep A cases as per Algorithm 3.
- Discuss referral to Healthy Homes Initiative if relevant (see Healthy Homes Initiative — Health New Zealand | Te Whatu Ora for local contact information) and/or offer advice on keeping home warm and dry (see Healthy Homes — Health New Zealand | Te Whatu Ora).

* If any household contact has had three or more Strep A infections in three months, follow Algorithm 3 for treatment.

Algorithm 3: Managing recurrent Strep A sore throat in tamariki and rangatahi at high risk of ARF (Grade D)

Tamariki or rangatahi in **high-risk population** (see Algorithm 1) presenting with three or more symptomatic Strep A positive sore throats in a three-month period?

↓

Give a single dose of IM benzathine penicillin OR 10 days of amoxicillin with directly observed therapy.

↓

In addition, swab the whānau/household as per Algorithm 2 and treat if Strep A positive.

↓

Take a follow up post treatment swab 3–6 days after finishing oral amoxicillin treatment or one month after IM benzathine penicillin.

↓

Persisting Strep A positive

↓

Seek advice from specialist paediatrician, ID physician or clinical microbiologist on appropriate management which may include further antibiotic regimens — see Appendix 1.

↓

Use a health literacy approach:

- Ask what the whānau understands about the treatment of sore throats to prevent rheumatic fever.
- Fill in any gaps in understanding, explain Strep A carriage, potential risk in high-risk whānau, and treatment options available.
- Check what they've understood. Help them solve problems so they can complete their treatment.
- Discuss the importance of antibiotic adherence and basic hygiene practices (hand washing, sneeze/cough into elbow or a tissue).
- Assess and treat any skin infections as per local HealthPathways.
- Discuss referral to Healthy Homes Initiative if relevant (see Healthy Homes Initiative — Health New Zealand | Te Whatu Ora for local contact information) and/or offer advice on keeping home warm and dry (see Healthy Homes — Health New Zealand | Te Whatu Ora).

Strep A negative

↓

Strep A infection resolved

For any further sore throats, manage as per Algorithm 1.

Introduction

Most people presenting with a sore throat in primary care have a pharyngitis caused by a virus, but between 10% and 30% will have Strep A infection.[5-10]

Strep A only infects humans and is mainly transferred from person to person through respiratory droplets and close contact.

The incubation period for Strep A sore throat is typically two to five days.[5, 11]

"Yeah and um, sometimes they [doctors] are understanding, [sometimes] they're like "oh no it's viral, you don't need it." But then I have to like, stress to them how "I've got rheumatic fever, you know my history" and they look up like "oh yep, yeah" so they do it, you know..."

Person with lived experience of ARF

The rationale for treating Strep A sore throat

In Aotearoa, the main reason for diagnosing and treating Strep A sore throat is to prevent ARF.

The most effective way to prevent the development of ARF and subsequent rheumatic heart disease (RHD) in an individual is to treat Strep A sore throat with antibiotics. Randomised controlled trials and quasi-randomised studies from the 1950s–1970s demonstrated that penicillin reduced the risk of ARF by more than two-thirds compared to placebo.[4, 12-15]

Reduced incidence of ARF occurred in a number of regions around the globe where improved healthcare access and effective penicillin treatment of Strep A sore throat were implemented in parallel with improved socioeconomic conditions. This association has been demonstrated at a population level in Baltimore, United States,[16] Costa Rica,[17] Cuba,[18] Caribbean,[19] China,[20] and the Americas.[21] (See **Chapter 15: Primary Prevention**, and **Chapter 3: Strep A Infection, Acute Rheumatic Fever and Rheumatic Heart Disease: Risk Factors, Social Determinants of Health and Primordial Prevention**.)

Conversely, in populations with a low incidence of ARF, there is limited rationale to confirm a diagnosis of Strep A sore throat and treat it with antibiotics. More than 80% of people who are not treated also become symptom-free within one week. Pain and headache on day three are only slightly reduced by using antibiotics.[4] There is some rationale for treating people at higher risk of Strep A transmission, such as food handlers and healthcare workers (Grade D). However, antibiotics may still be beneficial in the following selected situations:

- **Treating or preventing suppurative complications** — Treating Strep A sore throat has been shown to reduce suppurative complications, such as peritonsillar cellulitis or abscess (quinsy) and otitis media,[22, 23] but not the risk of sinusitis. However, an estimated 4,000 people need to be treated with antibiotics to prevent one case of quinsy or mastoiditis.[4, 24]
- **Scarlet fever** — This complication of Strep A infection usually presents with a 'scarlatiniform' rash (like sandpaper), fever, and sore throat. It may occur in outbreaks with a high attack rate, especially in school and early childhood settings. Antibiotic treatment is recommended for a person with scarlet fever to minimise complications and reduce transmission.[25, 26]
- **Prevention of invasive Strep A (iGAS)** — Some studies have suggested a link between Strep A nasopharyngeal carriage or sore throat and iGAS. iGAS includes bacteraemia, with pneumonia or skin and soft tissue infection, meningitis, osteomyelitis, septic arthritis, and streptococcal toxic shock syndrome (STSS).[27, 28] iGAS was made a notifiable condition in Aotearoa on 1 Oct 2024 (Severe disease invasive Group A Strep now a 'notifiable' disease | Ministry of Health NZ).
- **Post-streptococcal glomerulonephritis (PSGN)** — PSGN is a rare complication of Strep A infection. Evidence is limited on whether antibiotic treatment of Strep A sore throat reduces PSGN.[4] A 2022 review suggests there may be a small treatment benefit.[29]

- **Strep A transmission** — Antibiotics reduce the infectivity period of Strep A sore throat to 24 hours in more than 90% of cases.[30] This may be an important consideration in specific settings:
  - In school tamariki in communities at high risk for ARF.
  - In school staff and early childhood education staff in communities at high risk for ARF.
  - In healthcare workers and food handlers.
  - During outbreaks, including in closed communities such as boarding hostels.[31-33]

Assessing the risk of acute rheumatic fever in a person with a sore throat

Managing patients with sore throats effectively, safely, and appropriately requires:

- Good clinical assessment.
- Consideration of the person's risk of ARF.
- Consideration of the wider environment of the individual and their whānau.

Algorithm 1 on assessing and managing sore throats is informed by the current epidemiology of ARF in Aotearoa. The algorithm helps identify patients who are most likely to be at risk of ARF: Māori and Pacific peoples under 35 years of age (particularly those 4–19 years). See **Chapter 4: Epidemiology of Strep A Infections, Acute Rheumatic Fever and Rheumatic Heart Disease**.

Take the following steps when assessing a person with a sore throat in discussion with the individual and/or their whānau:

- Ensure the patient (or their whānau) has the chance to say the ethnic groups they identify with.
- Ask **open questions** about their knowledge of Strep A and ARF: *"Can you tell me what you know about Strep A and rheumatic fever?"*
- Find out if anyone in the whānau has had ARF or RHD.
- Ask about who lives in the household, including whether there are any immune-compromised people or young infants who may be at increased risk of serious Strep A disease (other than ARF).
- Assess the housing situation. Assessing household crowding needs to be done sensitively. Ask, *"How many people normally sleep in the same bedroom/same room as (insert name of the person)?"* More than two per bedroom is a marker of crowding.[34]

For people assessed as low risk of ARF, discussing the following is important:

- The viral nature of most sore throats.
- The ineffectiveness and potential harms of over-testing and over-treating with antibiotics.
- The management of pain and fever.
- The plan for review (as needed).

See 'Managing patients' antibiotic expectations' — He Ako Hiringa.

Diagnosis and management of Strep A sore throat

Onset of Strep A sore throat is usually rapid, with throat pain and fever. However, tamariki may present in different ways. They may have less severe or transient symptoms, which may not come to their parent's/caregiver's attention. Classic signs and symptoms are outlined below.

However, as signs and symptoms may not be classical, take a swab in a high-risk patient for any reported sore throat (or alternatively treat empirically) as per **Algorithm 1**.[5]

Symptoms of a Strep A sore throat may include:

- Sore throat — typically with pain when swallowing.
- Fever.
- Headache.
- Nausea and vomiting.
- Abdominal pain.

Signs of a Strep A sore throat may include:

- Large red swollen tonsils, sometimes with exudate (pus).
- Fever >38°C.
- Tender enlarged anterior cervical (neck) lymph nodes.
- Petechiae on the soft palate and posterior pharynx.
- Strawberry tongue.

Diarrhoea, cough, coryza, ulcerations, conjunctivitis, and croup/laryngitis/hoarseness are not typically associated with Strep A sore throat. Such symptoms increase the likelihood of a viral aetiology.

Common differential diagnoses of Strep A sore throat

Viral infections are common. Bacterial infections are infrequent (except for Group C and G streptococci).[5, 35, 36]

Viral infections include:

- Adenovirus.
- SARS-CoV-2 (COVID-19).
- Influenza.
- Parainfluenza.
- Rhinoviruses.
- Respiratory syncytial virus (RSV).
- Epstein-Barr virus.
- HIV (seroconversion illness).
- Coxsackievirus.

Bacterial infections include:

- Group C and G β haemolytic streptococcus.
- Fusobacterium necrophorum.
- Sexually transmitted infections, including Treponema pallidum and Neisseria gonorrhoeae.
- Corynebacterium diphtheriae.
- Arcanobacterium haemolyticum.

Investigations for diagnosis of Strep A sore throat

Summary recommendations are as follows:

- Take a throat swab for culture in populations at high risk of ARF. In Aotearoa, throat culture remains the gold standard for diagnosing Strep A (Evidence level II, Grade B).
- If it is not possible to perform a throat swab or to ensure timely follow-up of swab results, empiric antibiotic treatment should be prescribed (Grade D).
- Do not use rapid antigen detection tests — studies in Aotearoa have shown they are less sensitive, and their positive predictive value is poor[37, 38] (Grade C).
- Rapid molecular tests are highly sensitive and specific and can be used for timely diagnosis and treatment of Strep A sore throat within organised community-based test-and-treat programmes. The coordinated roll-out of rapid molecular testing should be prioritised for communities with a high incidence of ARF (Grade B).

Comparison of different diagnostic methods

Clinical scoring systems

Differentiating between Strep A and viral sore throat solely on clinical grounds is not possible.[5, 6] Although clinical scoring tools are used in many settings, studies in Aotearoa have found that these tools have low predictive value (positive or negative) in most environments.[39, 40] A recent meta-analysis of the CENTOR and McIsaac scores in the United Kingdom confirmed the poor predictive value.[41] Accurately predicting, at an individual level (in people presenting with a Strep A sore throat), who will go on to develop ARF is also not possible. A recent meta-analysis found a higher risk only in those with a positive Strep A swab, a cardiac murmur, or previous history of ARF.[42]

Pharyngeal culture

Diagnosis of Strep A sore throat by culturing a throat swab remains the gold standard. Traditional culture techniques require access to a community or hospital microbiology laboratory. In rural areas, transport of swabs to the laboratory followed by specimen processing may lead to delays of several days before results are reported.

Rapid antigen detection tests (RADT)

RADT for Strep A mostly identify the Strep A-specific cell-wall antigen, Lancefield group A carbohydrate. Detection methods include latex agglutination assay, enzyme immunoassay, and optical enzyme immunoassay.[43] Globally, most published literature relating to the performance of RADTs comes from low-incidence ARF populations, with sensitivities between 50 and 92%, specificities generally greater than 95%, and varying predictive values. A 2014 meta-analysis of RADT studies on adults concluded that in low-incidence ARF settings, RADTs provided a sufficiently accurate diagnosis of Strep A sore throat in primary care and that positive RADTs may not require culture backup for negative tests.

A 2016 Cochrane review noted issues with the sensitivity of RADTs, with a pooled sensitivity of 85.6% and specificity of 95.4%. Based on these results, of 100 tamariki with strep throat, 86 would be correctly detected with the RADT, and 14 would be missed. The review concluded that whether or not a RADT could be used as a stand-alone test to rule out Strep A would depend on the epidemiological context.[43]

RADTs have been used in primary care and pharmacy 'test and treat' models in low-incidence ARF settings in the United Kingdom, the United States, France, Scandinavia, and Canada. (The tests in the United Kingdom use a clinical score plus point-of-care RADT).[44-46] In these populations where ARF is rare, they have generally reduced antibiotic use and performed better than clinical scoring alone.

The 2012 Infectious Diseases Society of America (IDSA) Group A Streptococcal Pharyngitis Guidelines (under review in 2024) contain recommendations for populations where ARF is rare and the priority is to reduce antibiotic use. IDSA recommendations are not appropriate for use in Aotearoa.

In Aotearoa, two validation studies of RADTs, performed in 2013 to inform the design of school-based primary prevention programmes, showed that RADTs performed poorly with variable and poor sensitivities (26–84%).[38, 47] The authors concluded that RADT should not be used for sore throat diagnosis in high-risk ARF population in Aotearoa. In the absence of contemporary data, RADT are not recommended in Aotearoa at present.

Rapid molecular tests

Rapid nucleic acid amplification tests (NAAT), including Polymerase chain reaction (PCR) tests, are more sensitive. Benchtop-sized machines suitable for community and primary care settings are now commercially available (Abbott IDNow®, Roche Cobas® Liat, and Cepheid Xpert Xpress® Strep A). They have considerable advantages over culture in terms of improved sensitivity, speed, and ease of performance. They offer definitive results within 5–30 minutes. Some are approved for use outside microbiology laboratories.[48-52, 53]

The Xpert Xpress® Strep A molecular test recently performed well at Middlemore Hospital, a region with a high-risk ARF population.[54] The test was significantly faster than culture, and its negative predictive value was excellent. The study's authors concluded that a negative Xpert Xpress® Strep A test allows clinicians to withhold antibiotics confidently. It is likely to result in a net antimicrobial stewardship benefit in populations with a high burden of Strep A-related disease, especially where the alternative is empiric antibiotics.

Experience with other infectious diseases shows that molecular testing can be successfully implemented in high-risk communities in non-laboratory and primary care settings with appropriate clinical governance, funding, and training. In Aotearoa, this experience includes:

- COVID-19 rapid molecular testing.
- Experience in PCR testing for respiratory viruses in emergency departments.
- Rapid molecular testing in Hepatitis C control programmes.

The high sensitivity of molecular tests (which may be more sensitive than culture) has raised concerns about detecting carriage rather than acute symptomatic infection. Similar concerns exist regarding the inability of traditional culture methods to differentiate recently acquired infection from carriage. Restricting testing to symptomatic people is important, except in specific circumstances with specialist guidance (for example, in localised outbreaks of ARF).[48]

Governance of rapid testing methods for Strep A sore throat is urgently required

Currently, there is a lack of national oversight and governance of point-of-care testing for infectious diseases, including Strep A. Such governance is needed to ensure that rapid and molecular point-of-care testing devices are appropriately used, maintained, and reported. Carefully developed oversight systems involving key stakeholders, including clinical microbiologists, primary care organisations, and other community end users, are needed prior to any roll-out of rapid testing for Strep A. Diagnostic stewardship principles need to be considered to ensure that access to testing (and treatment) is prioritised for populations at highest risk of ARF.

Table 5.2 compares the different characteristics of different testing options for Strep A throat infection.

Table 5.2. Comparing laboratory tests for Strep A sore throat

Key points	Throat culture	Rapid antigen diagnostic test (RADT)[55]	Molecular methods (nucleic acid amplification tests, or NAAT)[48]
Throat swab	Yes	Yes	Yes
Turn-around time	18–48 hours, but may be longer in rural areas	5–10 minutes	15 minutes–1 hour
Sensitivity	72– 95%[48]	**Overall 86%**[55] • Latex agglutination 53–92%[48] • Lateral flow immunoassay 85% • Optical immunoassay 86% **Aotearoa studies** • 42.9–86.4% in in-vitro lab studies • Lower in school programmes (26–42%)[38, 47]	**Overall** • Abbot Strep A and Strep A2® 98.5% • Cobas®Liat Strep A 95% • Xpert Xpress Strep A® 99.4% **Aotearoa studies** • Xpert Xpress Strep A® molecular test • 100% (95% CI: 87.6–100) • BioGX® 100%[54] • *Illumigene*® Strep A assay (LAMP) 87%[37]
Specificity	100%	**Overall 95%**[55] • Latex agglutination 90%[48] • Lateral flow immunoassay 97% • Optical immunoassay 94% **Aotearoa studies** • 80–89%, 100% in-vitro lab studies[38, 47]	**Overall** • Abbot Strep A and Strep A2® 93.4% • Cobas®Liat Strep A 94.2% • Xpert Xpress-Strep A® 94.1% **Aotearoa studies** • Xpert Xpress® Strep A 90.4% (95 CI: 85.1–94.3)[54] • *Illumigene*® Strep A assay 98%[37]

Key points	Throat culture	Rapid antigen diagnostic test (RADT)[55]	Molecular methods (nucleic acid amplification tests, or NAAT)[48]
Advantages	• Cheap • Facilitates *emm*-typing/whole genome sequencing and susceptibility testing • Identifies other bacterial causes of sore throat	• Cheap • Rapid • Simple — can be performed in the community	• Many NAATs are rapid • Highly sensitive and specific • Can be performed by non-lab healthcare workers
Disadvantages	• Time to result (18–48 hours, significantly higher turn-around time in rural areas) • Requires skilled laboratory staff and equipment	• Cannot sequence isolates or perform susceptibility testing • Many RADTs have low sensitivity and risk of false negatives in high-incidence ARF communities • Not validated in high-risk ARF settings • Unregulated market in Aotearoa: lack of quality control, potential for over-use or inequitable access in low-risk ARF populations in primary care settings	• Requires some training and benchtop lab equipment • Cannot sequence isolates or perform susceptibility testing • Costly (although it may be offset by reduced microbiology lab specimen processing and reporting costs)

Adapted from Taylor et al. Table 3.[54]

Blood tests are unhelpful for diagnosing Strep A throat infection

Serology: Anti-streptococcal antibody titres (anti-streptolysin O titre and anti-DNAse B) are not useful or appropriate for the timely diagnosis of acute Strep A sore throat in primary care.[35]

C-reactive protein: Although C-reactive protein (CRP) may be elevated in Strep A sore throat, CRP is insufficiently specific to differentiate between viral and Strep A sore throat, and its use in sore throat is not recommended.[56-58]

Procalcitonin (PCT): Has not been found to usefully distinguish Strep A sore throat from viral throat infections.[56]

Novel approaches to diagnosis are being trialled

A French study proposes to analyse the diagnostic accuracy of rapid nucleic acid tests for Strep A using saliva in a prospective multicentre study in primary care. Obtaining saliva samples could potentially be more convenient and advantageous for self-testing in the future.[59]

A very small study in the United States suggests gene expression profiles and host transcriptomic markers have the potential to differentiate Strep A throat infection from viral infections.[58]

Other new diagnostic approaches using artificial intelligence (AI) and smartphone technology are being evaluated but are not yet regulated for this indication in Aotearoa. A small study in Texas used throat colour analysis from smartphone photographs for Strep A sore throat diagnosis and found 88% specificity and 87.5% sensitivity in low-risk settings.[60]

Recommendations for antibiotic treatment of Strep A sore throat

Internationally, recommended treatments for Strep A sore throat include oral phenoxymethylpenicillin (Pen V) for 10 days or single dose intramuscular penicillin (benzathine penicillin, also known as BPG).[5, 6, 35, 36] Penicillin is narrow-spectrum and inexpensive. To date, no clinically relevant resistance to penicillin has been documented in Strep A.

One intra-muscular dose of benzathine penicillin may be a useful option when adherence to a 10-day course is challenging, but the injection is painful. Administration requires training, good patient preparation, and a trusting relationship with the patient and whānau. Administering benzathine penicillin with lignocaine and developmentally appropriate support is strongly recommended. See **Chapter 9: Administration of Intramuscular Benzathine Penicillin (Bicillin®L-A).**

In Aotearoa, once-daily amoxicillin is also recommended for antibiotic treatment for Strep A sore throat. Amoxicillin has been shown to be non-inferior to oral penicillin in an RCT conducted in Auckland.[61] It is also acid stable, has a highly palatable syrup formulation and is more reliably absorbed when taken with food than penicillin V (Grade C). Once-daily dosing is convenient and may help adherence, particularly in school-based primary prevention programmes where daily observed therapy is possible.

Antibiotic duration

A 10-day course of oral penicillin is recommended to treat Strep A sore throat in Aotearoa.[6, 32, 35, 62, 63] Recent international research indicates that three- to five-day courses may be equivalent to 10-days in reducing clinical symptoms and eradicating Strep A. However these studies were conducted in populations at low risk of ARF and there is no contemporary data from Aotearoa or globally, to indicate that short course treatment reduces the risk of subsequent ARF in a person with Strep A sore throat. It is also recognised that shorter courses may be less effective at preventing suppurative sequelae such as quinsy. Currently, 10 days remain the recommendation for antibiotic treatment of Strep A sore throat in Aotearoa (Grade D).

A 2021 Cochrane review of Strep A sore throat treatment found "low certainty for any findings of difference in effectiveness of cure or reduction of complications between any of the antibiotics and penicillin."[64] The quality of studies was noted to be generally poor, with significant variation.[64] The necessity and strength of evidence for a 10-day course has also been questioned by others[65] on the basis of poor-quality historical studies. They found no significant difference in the incidence of streptococcal sore throat or scarlet fever in the next three months between those given a five- to seven-day course and those given an eight- to ten-day course.[66] However, ARF was not an outcome measured in this study.

A recent meta-analysis suggests that antibiotic treatment clears pharyngeal Strep A in more than 90% of individuals 24 hours after the therapy started. However, as in previous meta-analyses, the evidence overall is moderate at best. Half of the included studies were at moderate risk of bias. Only six had a low risk of bias.[30] The authors concluded that studies had important methodological limitations and pooled estimates are imprecise.

However, Strep A was cultured from the pharynx of almost 10% of people on routine follow-up 10 or more days after finishing antibiotics of any class. Where typing was available, four of five were confirmed to be a relapse or were found to have reacquired the original Strep A strain.[30]

Antibiotic use in low-risk populations

In populations at low risk of ARF in Aotearoa, best care will rarely require either testing of a throat swab for Strep A or any antibiotic treatment, of whatever duration, regardless of whether Strep A infection is present or not. The exception is individuals at increased risk of transmission, such as healthcare workers, or individuals at risk of severe GAS disease for example those with underlying immune compromise.

Assessing suspected penicillin allergy when managing sore throat

Most people said to be allergic to penicillin are not truly allergic when re-challenged.

The 'label' of penicillin allergy is associated with suboptimal treatment and leads to worse clinical outcomes in many conditions. Penicillin is generally more effective, better tolerated, and less impacted by antibiotic resistance than many other antibiotics, including erythromycin, for treatment of Strep A sore throat. Clinicians must carefully consider whether a penicillin allergy label is likely to represent true penicillin allergy before prescribing second-line non-penicillin antibiotics such as erythromycin.[67-72]

Identifying true penicillin allergy can be problematic in community settings.

There is a low likelihood of an allergic reaction in the following circumstances:

- Delayed onset >72hours after first dose.
- Mild spotty childhood rash: transient morbilliform or maculopapular rash, may be mildly itchy but not associated with systemic symptoms.
- Gastro-intestinal symptoms (e.g. nausea, vomiting, and diarrhoea).
- Family/whānau history of penicillin allergy.

Table 5.3. Treating those who may have a penicillin allergy

Likelihood of penicillin allergy	Suggested treatment
Low likelihood of true penicillin allergy	Penicillin or amoxicillin
High likelihood of true penicillin allergy	Erythromycin

If the patient has a suspected true penicillin allergy and requires (or is likely to require) frequent courses of antibiotics, consider referring them to a paediatrician or an allergist/immunologist.

Non-antibiotic management of sore throat

Strong evidence from multiple good-quality studies shows that paracetamol and non-steroidal anti-inflammatory (NSAID) medication (such as ibuprofen) provide significant symptom relief for acute sore throat, compared to placebo (Grade B).[22, 24, 36] Evidence is inconclusive that probiotics and homeopathic treatments relieve symptoms and prevent Strep A sore throat. Probiotics are considered safe and well tolerated and may have a role in managing or preventing persistent Strep A infection.[73-75] However, further studies are needed (Grade C).

Monitoring antimicrobial resistance to Strep A

Strep A historically has shown 100% susceptibility to penicillin, such that microbiology laboratories do not consistently report antimicrobial susceptibility test results when they report positive Strep A cultures. Although clinically significant penicillin resistance amongst Strep A has never been reported, genetic resistance mutations affecting penicillin-binding protein genes were identified among Strep A isolates from a localised community outbreak in Washington, United States.[76] Further studies have confirmed more widespread clonal expansion of similar penicillin-binding gene mutations[77] underscoring the need for ongoing national laboratory surveillance of Strep A resistance.

Monitoring of macrolide susceptibility rates among Strep A isolates is also important. Macrolide resistance among Strep A isolates in Aotearoa appears to currently sit between 4 and 8% (based on unpublished data from hospital laboratories) but is not consistently monitored nationally.

Knowledge of macrolide susceptibility results is important for anyone at high risk of ARF with Strep A sore throat and penicillin allergy. If needed, susceptibility results can be provided by the microbiology laboratory on request to help guide treatment.

Carriage of Strep A: Definition, prevalence and persistence

Although there is no universally agreed consensus definition of 'carriage,' classically, 'carriage' is considered to mean detecting persistent Strep A in the throat in an asymptomatic person without evidence of an immune system (antibody) response.[35, 78]

Some people may 'carry' Strep A without becoming ill. Others with a sore throat can be carriers (have a positive Strep A culture without an antibody response) with sore throat symptoms caused by a viral illness. Studies have also shown that serological responses to Strep A are highly variable and may not always be correlated with presence or absence of sore throat symptoms.[79, 97] Differentiating between the following scenarios may be challenging or impossible in a clinical context:

- Between carriage and 'true' Strep A sore throat.
- Between carriage and recurrent Strep A (re-infection with the same or a new strain).
- In an asymptomatic infection in a person at high risk for ARF, the immune response differs between infections that trigger an immune reaction and those that do not.

In primary care, the term carriage should be applied only rarely and after very careful consideration in persons at higher risk of ARF. A pragmatic definition of Strep A carriage is persistent Strep A after adequate antibiotic therapy and resolution of symptoms on culture or PCR testing. This definition considers the difficulty of interpreting serology and that Strep A molecular testing is not routinely available.[78]

Table 5.4. Characteristics to help differentiate carriage from recurrence

True recurrence	Carriage
Classic Strep A symptoms (see '**Diagnosis and management of Strep A sore throat**' above)	Other respiratory symptoms (cough, nasal congestion)
Symptoms improve within 24–48 hours of starting antibiotics	Little improvement in symptoms after antibiotics; illness lasts >5 days
The culture between episodes is negative	The culture between episodes remains positive

Adapted from 'The carrier state of Streptococcus pyogenes'.

The prevalence of carriage has been reported to be between 7 and 12% in tamariki in OECD countries[8] and Aotearoa up to 35%.[80] Prevalence may vary by age group, season, and geographic location.[8, 81] Throat culture surveys of healthy asymptomatic tamariki may yield Strep A prevalence rates as high as 20–25%.[11, 78]

Strep A presence in the throat can persist for many months. The risk of transmission from "carriers" has traditionally been considered low because of the lack of respiratory symptoms and the lower density of bacteria in the pharynx.[11, 78] Recently, Cordery et al. showed evidence of transmission from tamariki with asymptomatic infection in a scarlet fever outbreak in school and nursery settings in the United Kingdom.[25] Lacey et al. also used whole genome sequencing to demonstrate that asymptomatic Strep A throat carriage acted as a 'reservoir' in the transmission of Strep A associated with impetigo in northern Australia.[82]

Carriage of Strep A: Treatment considerations

Factors to consider when treating persistent carriage in Aotearoa

In Aotearoa, antibiotic treatment is usually unnecessary for people at **low risk** of ARF.[11] However, antibiotic treatment is recommended for people at high risk of ARF, given the uncertainty about whether the presence of Strep A in the throat is due to an acute infection that might cause ARF, or due to long-term carriage that is less likely to trigger ARF (Grade C) — seek specialist advice if considering treating carriage. Thus consider:

- The individual's risk of ARF.
- Whether clinical findings are more suggestive of Strep A or viral illness.
- The response to antibiotic treatment (usually within 24 hours for Strep A sore throat).
- Whether Strep A testing between episodes (when asymptomatic) is positive.
- Whether a patient is likely to complete a course of oral antibiotics or to have other reasons for suboptimal absorption of oral antibiotics (note that around 10% of all fully adherent patients will remain Strep A positive after treatment).

Guidance in the United States on eradicating Strep A carriage

The Red Book 2021–2024[11] provides advice for predominantly low-risk populations in the United States and lists only a few indications for attempted eradication of Strep A carriage:

- Localised outbreaks of ARF or PSGN or outbreaks of Strep A in a closed community.
- A family/whānau history of ARF.
- Multiple 'ping-pong' episodes of documented symptomatic Strep A sore throat in a whānau despite appropriate therapy.
- As an alternative to tonsillectomy because of very frequent Strep A episodes.

Defining and managing recurrent symptomatic Strep A sore throat in Aotearoa

As noted above, differentiating new infections from carriage is challenging in the community setting without timely access to molecular sequencing. A new infection could be a re-infection with the same strain or a new one. In a 2008 study in a high-risk population in Auckland, 2.5% of all participants treated with penicillin and 5% of participants treated with amoxicillin had acquired a new serotype (as defined by serotyping) 26–36 days after treatment started. Another approximately 10% cultured the original serotype.[61]

In Aotearoa, treatment for recurrent Strep A throat infection is recommended for people:

- At high risk of ARF.
- **AND** presenting with three or more episodes of **symptomatic** sore throat within three months, and those episodes are positive for Strep A.

Manage recurrent infections as per Algorithm 3. Recommended treatment is as follows:

- Treat the third and subsequent Strep A episodes using IM benzathine penicillin or oral amoxicillin (see Algorithm 3) (Grade C/D).
- Collect a post-treatment swab three to six days after the person has completed 10 days of amoxicillin or one month after IM benzathine penicillin (Grade D).
- If the patient remains Strep A positive on follow-up swabbing, ask a paediatrician, infectious disease specialist, or clinical microbiologist for management advice, which may include further antibiotics (usually the addition of rifampicin — see Table 5.5).

Evidence regarding the effectiveness of particular antibiotic regimens for treating Strep A recurrence or clearing carriage is of low quality. In a review, Munck suggests the incidence of future sore throats can be effectively decreased with clindamycin, amoxicillin with clavulanate, or cefpodoxime, but a significant proportion of patients experience spontaneous clearance.[83] Ng et al. found no robust studies met the criteria for a Cochrane systematic review.[84]

Table 5.5 outlines the recommended antibiotic regimens for the treatment of a third or subsequent symptomatic episode of Strep A sore throat in a three-month period in a person at high risk of ARF.

These antibiotic regimens are for treating high-risk persons with recurrent Strep A sore throat who have already been treated for three or more symptomatic episodes within three months. They are only to be used on advice from a paediatrician, adult infectious disease specialist, or clinical microbiology specialist. Rifampicin and clindamycin require specialist approval. Rifampicin is relatively contraindicated in pregnancy and interacts with many drugs (in particular oral contraceptives, anticonvulsants and warfarin).

Table 5.5. Antibiotic regimens for recurrent Strep A sore throat

Antibiotic	Dose	Note
Amoxicillin (PO)	50mg/kg (maximum 1000mg/day) once daily for 10 days	**PLUS** on the final 4 days (days 7–10 of treatment) add **oral rifampicin** 20 mg/kg (max 600mg) once daily
Phenoxymethyl-penicillin (PO)	15mg/kg (maximum 500mg/dose) two times daily	**PLUS** on the final 4 days (days 7–10 of treatment) add **oral rifampicin** 20 mg/kg (max 600mg) once daily
Benzathine penicillin (IM) single dose	**Tamariki <20kg** 600,000 units (450mg) Recommend use with lignocaine and distraction techniques. Chapter 9: Administration of Intramuscular Benzathine Penicillin (Bicillin®L-A)	**PLUS oral rifampicin**, 20 mg/kg/day (max 600 mg/day), once daily for 4 days
	Children ≥20kg or adults 1,200,000 units (900mg) Recommend use with lignocaine and distraction techniques. Chapter 9: Administration of Intramuscular Benzathine Penicillin (Bicillin®L-A)	**PLUS oral rifampicin**, 20 mg/kg/day (max 600 mg/day), once daily for 4 days
Amoxicillin + clavulanic acid (PO)	15–30mg/kg (max 625mg/dose) three times daily for 10 days	
Treatment in documented/high risk of PENICILLIN ALLERGY		
Clindamycin	7–10mg/kg (max 300mg/dose) three times daily for 10 days Available in 150mg capsules, round to nearest capsule dose	

Managing household contacts of people with Strep A throat infection

Prioritise swabbing household contacts at high risk of ARF, especially where resources are limited (see **Algorithm 2**) (Evidence level III-1, Grade B).

The usual purpose of identifying and treating contacts of someone with an infectious disease is to prevent further disease. The secondary attack rate of Strep A within households is known to be relatively high. Historic estimates of onward transmission of pharyngeal Strep A after exposure to a symptomatic index case range between 13% and 28%. Transmission may occur over weeks to months.[85]

The rationale for treating contacts of people with Strep A throat infections is to eradicate Strep A from household members. Treating contacts temporarily reduces the risk of transmitting Strep A to others in the household and reduces their risk of developing a Strep A-related disease. However, treating contacts is resource intensive, and incomplete knowledge of ARF pathogenesis, along with the potential ineffectiveness of Strep A treatment for carriage or recurrence, all need to be balanced against potential benefits. Thus, indications for testing contacts for Strep A throat infection in the context of ARF prevention are limited.[11]

In Aotearoa, the 2019 Sore Throat Guidelines recommended swabbing household contacts:

- Of a person with Strep A throat infection and a family/whānau or personal history of ARF or RHD.
- Of a person with Strep A throat infection living in a household where three or more cases of Strep A infection occurred within the last three months.[86]

As of 2024, no new evidence supports or refutes these recommendations. For low-risk ARF populations, household swabbing is not generally recommended, with the exception of people employed in higher-risk professions such as education, healthcare, and food handling, or households with immune compromised individuals. A careful explanation of carriage, the limitations of our knowledge of Strep A transmission, and treatment options is required.

The role of tonsillectomy

No specific evidence exists to suggest that tonsillectomy reduces the risk of ARF or that different criteria need to be used for tamariki at high risk of ARF when assessing them for potential tonsillectomy. However, tonsillectomy appears to reduce the frequency of sore throat episodes in tamariki with recurrent tonsillitis.[87-91]

Treatment of Strep A sore throat in people already on secondary antibiotic prophylaxis (SAP)

Recommendations for patients on SAP for ARF or RHD who test Strep A positive on throat swab are as follows (Evidence level IV, Grade D):

- Treat with a 10-day course of oral phenoxymethylpenicillin or amoxicillin in addition to continuing SAP if the person has a sore throat and the last benzathine penicillin dose was more than seven days ago.
- Review adherence to SAP and discuss options with the patient/their whānau if there are any concerns (see **Chapter 9: Administration of Intramuscular Benzathine Penicillin (Bicillin®L-A)**).

By weeks three and four after IM benzathine penicillin, serum penicillin levels fall to a level lower than required for effective treatment of Strep A throat infection.[92] However, historic studies suggest a low risk of recurrence for those with good adherence to benzathine penicillin, even if they experience Strep A sore throat.[93, 94]

Diagnosing and treating Strep A skin infections

To date, no conclusive patient-level evidence shows a direct causal relationship between Strep A skin infections and ARF, nor does treating infected skin with particular treatments (topical or oral antibiotics) directly prevent ARF. However, skin infections caused by Strep A and *Staphylococcus aureus* are common in population groups at risk for ARF. Skin infections can have many adverse sequelae, including cellulitis, suppurative infections, eczema flare-ups, PGSN, and invasive Strep A disease. Treating skin infections effectively in these populations remains important.

Diagnosis of skin infections is usually clinical. For indications for a swab refer to Health Pathways.

Group G and C streptococcus

Other strains of beta-haemolytic streptococci, in particular, Group C and G streptococcus (also known as *Streptococcus dysgalactiae* subspecies *equisimilis*), can infect humans. An association between Group C and G streptococci and ARF is postulated as these bacteria may share virulence factors and genetic elements with Group A streptococci.[95] Further investigation into any potential role in ARF pathogenesis is needed.

Group C and G β-haemolytic streptococci can cause a symptomatic sore throat and occasionally invasive infections.[96] Currently, antibiotic treatment of people with Group C and G strep throat can be considered for those:

- At increased risk of transmission (food handlers and early childhood workers).
- With severe and suppurative manifestations.
- Who are immune compromised.

References

1. Anderson A, Peat B, Ryland J, Ofanoa M, Burgess H, Malungahu G, et al. Mismatches between health service delivery and community expectations in the provision of secondary prophylaxis for rheumatic fever in New Zealand. *Australian and New Zealand Journal of Public Health*. 2019;43(3):294–299. https://doi.org/10.1111/1753-6405.12890

2. Bennett J, Zhang J, Leung W, Jack S, Oliver J, Webb R, et al. Rising ethnic inequalities in acute rheumatic fever and rheumatic heart disease, New Zealand, 2000–2018. *Emerging Infectious Diseases*. 2021;27(1):36–46. https://doi.org/10.3201/eid2701.191791

3. Health New Zealand | Te Whatu Ora. Rheumatic fever report 2022. Health New Zealand | Te Whatu Ora; 2023. https://view.officeapps.live.com/op/view.aspx?src=https%3A%2F%2Fwww.tewhatuora.govt.nz%2Fassets%2FUploads%2F20230420-Rheumatic-Fever-Report-2022-Public.xlsx&wdOrigin=BROWSELINK (Accessed February 20 2025).

4. Spinks A, Glasziou PP, Del Mar CB. Antibiotics for treatment of sore throat in children and adults. *Cochrane Database of Systematic Reviews*. 2021;12(12):CD000023. https://doi.org/10.1002/14651858.CD000023.pub5

5. Tanz R. Acute pharyngitis. In: Kliegman R, St Geme J, Blum N, Shah S, Tasker R, Wilson K, editors. Nelson textbook of pediatrics. 21st ed. Philadelphia: Elsevier; 2020. p. 2192–2196.

6. Dougherty S, Carapetis J, Zühlke L, Wilson N. Acute rheumatic fever and rheumatic heart disease. Amsterdam: Elsevier; 2020.

7. Lennon D, Stewart J, Farrell E, Palmer A, Mason H. School-based prevention of acute rheumatic fever: a group randomized trial in New Zealand. *Pediatric Infectious Disease Journal*. 2009;28(9):787–794. https://doi.org/10.1097/INF.0b013e3181a282be

8. Oliver J, Malliya Wadu E, Pierse N, Moreland NJ, Williamson DA, Baker MG. Group A streptococcus pharyngitis and pharyngeal carriage: A meta-analysis. *PLoS Neglected Tropical Diseases*. 2018;12(3):e0006335. https://doi.org/10.1371/journal.pntd.0006335

9. Oliver J, Upton A, Jack SJ, Pierse N, Williamson DA, Baker MG. Distribution of streptococcal pharyngitis and acute rheumatic fever, Auckland, New Zealand, 2010–2016. *Emerging Infectious Diseases*. 2020;26(6):1113–1121. https://doi.org/10.3201/eid2606.181462

10. Wessels MR. Streptococcus pyogenes pharyngitis and scarlet fever. In: Ferretti JJ, Stevens DL, Fischetti VA, editors. Streptococcus pyogenes: basic biology to clinical manifestations. Oklahoma City: University of Oklahoma Health Sciences Center; 2022.

11. Committee on Infectious Diseases. American Academy of Pediatrics. In: Kimberlin D, Barnett E, Lynfield R, Sawer M, editors. Red book 2021–2024: report of the Committee on Infectious Diseases. 32nd ed: American Academy of Pediatrics; 2021.

12. Wessels MR. Clinical practice. Streptococcal pharyngitis. *New England Journal of Medicine*. 2011;364(7):648–655. https://doi.org/10.1056/NEJMcp1009126

13. Denny FW, Wannamaker LW, Brink WR, Rammelkamp CH, Jr., Custer EA. Prevention of rheumatic fever; treatment of the preceding streptococcic infection. *Journal of the American Medical Association*. 1950;143(2):151–153. https://doi.org/10.1001/jama.1950.02910370001001

14. Robertson KA, Volmink JA, Mayosi BM. Antibiotics for the primary prevention of acute rheumatic fever: a meta-analysis. *BMC Cardiovascular Disorders*. 2005;5(1):11. https://doi.org/10.1186/1471-2261-5-11

15. Jones TD, Mote JR. The clinical importance of infection of the respiratory tract in rheumatic fever. *Journal of the American Medical Association*. 1939;113(10). https://doi.org/10.1001/jama.1939.02800350008003

16. Gordis L. Effectiveness of comprehensive-care programs in preventing rheumatic fever. *New England Journal of Medicine*. 1973;289(7):331–335. https://doi.org/10.1056/nejm197308162890701

17. Arguedas A, Mohs E. Prevention of rheumatic fever in Costa Rica. *Journal of Pediatrics*. 1992;121(4):569–572. https://doi.org/10.1016/s0022-3476(05)81146-1

18. Nordet P, Lopez R, Dueñas A, Sarmiento L. Prevention and control of rheumatic fever and rheumatic heart disease: the Cuban experience (1986-1996-2002). *Cardiovascular Journal of Africa*. 2008;19(3):135–140.

19. Bach JF, Chalons S, Forier E, Elana G, Jouanelle J, Kayemba S, et al. 10-year educational programme aimed at rheumatic fever in two French Caribbean islands. *Lancet*. 1996;347(9002):644–648. https://doi.org/10.1016/s0140-6736(96)91202-7

20. Lin S, Kaplan EL, Rao X, Johnson DR, Deng M, Zhuo Q, et al. A school-based program for control of group a streptococcal upper respiratory tract infections: a controlled trial in Southern China. *Pediatric Infectious Disease Journal*. 2008;27(8):753–755. https://doi.org/10.1097/INF.0b013e31816be02f

21. Ordunez P, Martinez R, Soliz P, Giraldo G, Mujica OJ, Nordet P. Rheumatic heart disease burden, trends, and inequalities in the Americas, 1990-2017: a population-based study. *Lancet Global Health*. 2019;7(10):e1388–e1397. https://doi.org/10.1016/s2214-109x(19)30360-2

22. Kociolek LK, Shulman ST. In the clinic. Pharyngitis. *Annals of Internal Medicine*. 2012;157(5):Itc3–1 – itc3–16. https://doi.org/10.7326/0003-4819-157-5-201209040-01003

23. Miller KM, Carapetis JR, Van Beneden CA, Cadarette D, Daw JN, Moore HC, et al. The global burden of sore throat and group A streptococcus pharyngitis: a systematic review and meta-analysis. *EClinicalMedicine*. 2022;48:101458. https://doi.org/10.1016/j.eclinm.2022.101458

24. National Institute for Health and Care Excellence. Sore throat (acute): antimicrobial prescribing. NICE guideline [NG84]. National Institute for Health and Care Excellence. https://www.nice.org.uk/guidance/ng84 (Accessed February 24 2025).

25. Cordery R, Purba AK, Begum L, Mills E, Mosavie M, Vieira A, et al. Frequency of transmission, asymptomatic shedding, and airborne spread of streptococcus pyogenes in schoolchildren exposed to scarlet fever: a prospective, longitudinal, multicohort, molecular epidemiological, contact-tracing study in England, UK. *Lancet Microbe*. 2022;3(5):e366–e375. https://doi.org/10.1016/S2666-5247(21)00332-3

26. Herdman MT, Cordery R, Karo B, Purba AK, Begum L, Lamagni T, et al. Clinical management and impact of scarlet fever in the modern era: findings from a cross-sectional study of cases in London, 2018-2019. *BMJ Open*. 2021;11(12):e057772. https://doi.org/10.1136/bmjopen-2021-057772

27. Zachariadou L, Stathi A, Tassios PT, Pangalis A, Legakis NJ, Papaparaskevas J. Differences in the epidemiology between paediatric and adult invasive streptococcus pyogenes infections. *Epidemiology and Infection*. 2014;142(3):512–519. https://doi.org/10.1017/s0950268813001386

28. Kailankangas V, Vilhonen J, Gröndahl-Yli-Hannuksela K, Rantakokko-Jalava K, Seiskari T, Auranen K, et al. Presence of streptococcus pyogenes in the throat in invasive group A streptococcal disease: a prospective two-year study in two health districts, Finland. *Infect Dis (Lond)*. 2023;55(6):405–414. https://doi.org/10.1080/23744235.2023.2192287

29. Bateman E, Mansour S, Okafor E, Arrington K, Hong BY, Cervantes J. Examining the efficacy of antimicrobial therapy in preventing the development of postinfectious glomerulonephritis: a systematic review and meta-analysis. *Infectious Disease Reports*. 2022;14(2):176–183. https://doi.org/10.3390/idr14020022

30. McGuire E, Li A, Collin SM, Decraene V, Cook M, Padfield S, et al. Time to negative throat culture following initiation of antibiotics for pharyngeal group A streptococcus: a systematic review and meta-analysis up to October 2021 to inform public health control measures. *Euro Surveillance*. 2023;28(15). https://doi.org/10.2807/1560-7917.Es.2023.28.15.2200573

31. Mustafa Z, Ghaffari M. Diagnostic methods, clinical guidelines, and antibiotic treatment for group A streptococcal pharyngitis: a narrative review. *Frontiers in Cellular and Infection Microbiology*. 2020;10:563627. https://doi.org/10.3389/fcimb.2020.563627

32. Heart Foundation of New Zealand. New Zealand guidelines for rheumatic fever. 2. Group A streptococcal sore throat management guideline: 2014 update. Auckland: Heart Foundation of New Zealand; 2014. https://www.heartfoundation.org.nz/resources/group-a-streptococcal-sore-throat-management (Accessed February 24 2025).

33. Gatman K, Thompson B, Harrower J, Rajanaidu S. A foodborne outbreak of group A streptococcus: an under-recognised method of spread. *New Zealand Medical Journal*. 2024;137(1589):73–76. https://doi.org/10.26635/6965.6313

34. Goodyear R, Fabian A, Hay J. Finding the crowding index that works best for New Zealand. Working Paper No. 11–04. Wellington: Statistics New Zealand; 2011. https://www.stats.govt.nz/assets/Uploads/Retirement-of-archive-website-project-files/Research/Finding-the-crowding-index-that-works-best-for-New-Zealand-working-paper/finding-crowding-index-best-for-nz-working-paper-11-04.pdf (Accessed February 23 2025).

35. Shulman ST, Bisno AL, Clegg HW, Gerber MA, Kaplan EL, Lee G, et al. Clinical practice guideline for the diagnosis and management of group A streptococcal pharyngitis: 2012 update by the Infectious Diseases Society of America. *Clinical Infectious Diseases*. 2012;55(10):e86–102. https://doi.org/10.1093/cid/cis629

36. Taylor A, Webb R. Fifteen-minute consultation: group A streptococcal pharyngitis, diagnosis and treatment in children. *Archives of Disease in Childhood: Education and Practice Edition*. 2024;109(5):210–221. https://doi.org/10.1136/archdischild-2023-325755

37. Upton A, Bissessor L, Farrell E, Shulman ST, Zheng X, Lennon D. Comparison of illumigene group A streptococcus assay with culture of throat swabs from children with sore throats in the New Zealand school-based rheumatic fever prevention program. *Journal of Clinical Microbiology*. 2016;54(1):153–156. https://doi.org/10.1128/jcm.02440-15

38. Upton A, Farrell E, Stewart J, Lennon D. Disappointing performance of rapid antigen detection tests for group A streptococcus in the Auckland school-based sore throat programme. *New Zealand Medical Journal*. 2014;127(1389):103–105.

39. Jamiel Y. The validity of scorecard as a predictive of streptococcal pharyngitis by throat swab [Master's thesis]: University of Auckland; 2005.

40. Kerdemelidis M, Lennon DR, Stewart J. How should sore throats be managed? Performance of four group A streptococcal sore throat prediction rules in New Zealand children. *Open Forum Infectious Diseases*. 2015;2. https://doi.org/10.1093/ofid/ofv133.720

41. Willis BH, Coomar D, Baragilly M. Comparison of Centor and McIsaac scores in primary care: a meta-analysis over multiple thresholds. *British Journal of General Practice*. 2020;70(693):e245–e254. https://doi.org/10.3399/bjgp20X708833

42. Kulik E, Stuart B, Willcox M. Predictors of rheumatic fever in sore throat patients: a systematic review and meta-analysis. *Transactions of the Royal Society of Tropical Medicine and Hygiene*. 2022;116(4):286–297. https://doi.org/10.1093/trstmh/trab156

43. Cohen JF, Bertille N, Cohen R, Chalumeau M. Rapid antigen detection test for group A streptococcus in children with pharyngitis. *Cochrane Database of Systematic Reviews*. 2016;7(7):Cd010502. https://doi.org/10.1002/14651858.CD010502.pub2

44. Mantzourani E, Cannings-John R, Evans A, Ahmed H. To swab or not to swab? Using point-of-care tests to detect group A streptococcus infections as part of a sore throat test and treat service in community pharmacy. *Journal of Antimicrobial Chemotherapy*. 2022;77(3):803–806. https://doi.org/10.1093/jac/dkab470

45. Rystedt K, Hedin K, Tyrstrup M, Skoog-Ståhlgren G, Edlund C, Giske CG, et al. Agreement between rapid antigen detection test and culture for group A streptococcus in patients recently treated for pharyngotonsillitis — a prospective observational study in primary care. *Scandinavian Journal of Primary Health Care*. 2023;41(1):91–97. https://doi.org/10.1080/02813432.2023.2182631

46. Pallon J, Rööst M, Sundqvist M, Hedin K. The aetiology of pharyngotonsillitis in primary health care: a prospective observational study. *BMC Infectious Diseases*. 2021;21(1):971. https://doi.org/10.1186/s12879-021-06665-9

47. Upton A, Lowe C, Stewart J, Taylor S, Lennon D. In vitro comparison of four rapid antigen tests for group A streptococcus detection. *New Zealand Medical Journal*. 2014;127(1398):77–83.

48. Thompson TZ, McMullen AR. Group A streptococcus testing in pediatrics: The move to point-of-care molecular testing. *Journal of Clinical Microbiology*. 2020;58(6). https://doi.org/10.1128/jcm.01494-19

49. Shapiro DJ, Fine AM, Hersh AL, Bourgeois FT. Association between molecular streptococcal testing and antibiotic use for pharyngitis in children. *Journal of the Pediatric Infectious Diseases Society*. 2022;11(6):303–304. https://doi.org/10.1093/jpids/piac008

50. Weinzierl EP, Jerris RC, Gonzalez MD, Piccini JA, Rogers BB. Comparison of Alere istrep A rapid molecular assay with rapid antigen testing and culture in a pediatric outpatient setting. *American Journal of Clinical Pathology*. 2018;150(3):235–239. https://doi.org/10.1093/ajcp/aqy038

51. Klepser DG, Klepser ME, Murry JS, Borden H, Olsen KM. Evaluation of a community pharmacy-based influenza and group A streptococcal pharyngitis disease management program using polymerase chain reaction point-of-care testing. *Journal of the American Pharmacists Association*. 2019;59(6):872–879. https://doi.org/10.1016/j.japh.2019.07.011

52. Dubois C, Smeesters PR, Refes Y, Levy C, Bidet P, Cohen R, et al. Diagnostic accuracy of rapid nucleic acid tests for group A streptococcal pharyngitis: systematic review and meta-analysis. *Clinical Microbiology and Infection*. 2021;27(12):1736–1745. https://doi.org/10.1016/j.cmi.2021.04.021

53. Barth DD, Cinanni G, Carapetis JR, Wyber R, Causer L, Watts C, et al. Roadmap to incorporating group A streptococcus molecular point-of-care testing for remote Australia: a key activity to eliminate rheumatic heart disease. *Medical Journal of Australia*. 2022;217(6):279–282. https://doi.org/10.5694/mja2.51692

54. Taylor A, Morpeth S, Webb R, Taylor S. The utility of rapid group A streptococcus molecular testing compared with throat culture for the diagnosis of group A streptococcal pharyngitis in a high-incidence rheumatic fever population. *Journal of Clinical Microbiology*. 2021;59(12):e0097821. https://doi.org/10.1128/jcm.00978-21

55. Cohen JF, Pauchard JY, Hjelm N, Cohen R, Chalumeau M. Efficacy and safety of rapid tests to guide antibiotic prescriptions for sore throat. *Cochrane Database of Systematic Reviews*. 2020;6(6):Cd012431. https://doi.org/10.1002/14651858.CD012431.pub2

56. Christensen AM, Thomsen MK, Ovesen T, Klug TE. Are procalcitonin or other infection markers useful in the detection of group A streptococcal acute tonsillitis? *Scandinavian Journal of Infectious Diseases*. 2014;46(5):376–383. https://doi.org/10.3109/00365548.2014.885656

57. Calviño O, Llor C, Gómez F, González E, Sarvisé C, Hernández S. Association between C-reactive protein rapid test and group A streptococcus infection in acute pharyngitis. *Journal of the American Board of Family Medicine*. 2014;27(3):424–426. https://doi.org/10.3122/jabfm.2014.03.130315

58. Yu J, Tycksen E, Yang W, Mariani TJ, Bhattacharya S, Falsey AR, et al. Use of host response to refine the diagnosis of group A streptococcal pharyngitis. *Journal of the Pediatric Infectious Diseases Society*. 2022;11(11):482–491. https://doi.org/10.1093/jpids/piac072

59. Touitou R, Bidet P, Dubois C, Partouche H, Bonacorsi S, Jung C, et al. Diagnostic accuracy of a rapid nucleic acid test for group A streptococcal pharyngitis using saliva samples: protocol for a prospective multicenter study in primary care. *Diagnostic and Prognostic Research*. 2023;7(1):13. https://doi.org/10.1186/s41512-023-00150-4

60. Askarian B, Yoo SC, Chong JW. Novel image processing method for detecting strep throat (streptococcal pharyngitis) using smartphone. *Sensors*. 2019;19(15). https://doi.org/10.3390/s19153307

61. Lennon DR, Farrell E, Martin DR, Stewart JM. Once-daily amoxicillin versus twice-daily penicillin V in group A beta-haemolytic streptococcal pharyngitis. *Archives of Disease in Childhood*. 2008;93(6):474–478. https://doi.org/10.1136/adc.2006.113506

62. Skoog Ståhlgren G, Tyrstrup M, Edlund C, Giske CG, Mölstad S, Norman C, et al. Penicillin V four times daily for five days versus three times daily for 10 days in patients with pharyngotonsillitis caused by group A streptococci: randomised controlled, open label, non-inferiority study. *BMJ*. 2019;367:l5337. https://doi.org/10.1136/bmj.l5337

63. Holm AE, Llor C, Bjerrum L, Cordoba G. Short- vs. Long-course antibiotic treatment for acute streptococcal pharyngitis: Systematic review and meta-analysis of randomized controlled trials. *Antibiotics*. 2020;9(11). https://doi.org/10.3390/antibiotics9110733

64. van Driel ML, De Sutter AI, Thorning S, Christiaens T. Different antibiotic treatments for group A streptococcal pharyngitis. *Cochrane Database of Systematic Reviews*. 2021;3(3):Cd004406. https://doi.org/10.1002/14651858.CD004406.pub5

65. Radetsky M. Hostage to history: the duration of antimicrobial treatment for acute streptococcal pharyngitis. *Pediatric Infectious Disease Journal*. 2017;36(5):507–512. https://doi.org/10.1097/inf.0000000000001480

66. Salinas Salvador B, Moreno Sánchez A, Carmen Marcén G, Molina Herranz D, Arana Navarro T, García Vera C. Retrospective study on the effectiveness and safety of the shortened 5- to 7-day antibiotic regimen for acute streptococcal pharyngotonsillitis compared to the classic 10-day regimen. *Anales de Pediatría*. 2022;97(6):398–404. https://doi.org/10.1016/j.anpede.2022.07.005

67. International Rheumatic Fever Study Group. Allergic reactions to long-term benzathine penicillin prophylaxis for rheumatic fever. International Rheumatic Fever Study Group. *Lancet*. 1991;337(8753):1308–1310.

68. Blumenthal KG, Peter JG, Trubiano JA, Phillips EJ. Antibiotic allergy. *Lancet*. 2019;393(10167):183–198. https://doi.org/10.1016/s0140-6736(18)32218-9

69. Lagacé-Wiens P, Rubinstein E. Adverse reactions to β-lactam antimicrobials. *Expert Opinion on Drug Safety*. 2012;11(3):381–399. https://doi.org/10.1517/14740338.2012.643866

70. Krishna MT, Huissoon AP, Li M, Richter A, Pillay DG, Sambanthan D, et al. Enhancing antibiotic stewardship by tackling "spurious" penicillin allergy. *Clinical and Experimental Allergy*. 2017;47(11):1362–1373. https://doi.org/10.1111/cea.13044

71. Kaminsky LW, Ghahramani A, Hussein R, Al-Shaikhly T. Penicillin allergy label is associated with worse clinical outcomes in bacterial pneumonia. *The Journal of Allergy and Clinical Immunology: In Practice*. 2022;10(12):3262–3269. https://doi.org/10.1016/j.jaip.2022.08.027

72. Blumenthal KG, Lu N, Zhang Y, Walensky RP, Choi HK. Recorded penicillin allergy and risk of mortality: a population-based matched cohort study. *Journal of General Internal Medicine*. 2019;34(9):1685–1687. https://doi.org/10.1007/s11606-019-04991-y

73. Wilcox CR, Stuart B, Leaver H, Lown M, Willcox M, Moore M, et al. Effectiveness of the probiotic streptococcus salivarius K12 for the treatment and/or prevention of sore throat: a systematic review. *Clinical Microbiology and Infection*. 2019;25(6):673–680. https://doi.org/10.1016/j.cmi.2018.12.031

74. Wescombe PA, Heng NC, Burton JP, Chilcott CN, Tagg JR. Streptococcal bacteriocins and the case for streptococcus salivarius as model oral probiotics. *Future Microbiology*. 2009;4(7):819–835. https://doi.org/10.2217/fmb.09.61

75. Doyle H, Pierse N, Tiatia R, Williamson D, Baker M, Crane J. Effect of oral probiotic streptococcus salivarius K12 on group A streptococcus pharyngitis: a pragmatic trial in schools. *Pediatric Infectious Disease Journal*. 2018;37(7):619–623. https://doi.org/10.1097/inf.0000000000001847

76. Vannice KS, Ricaldi J, Nanduri S, Fang FC, Lynch JB, Bryson-Cahn C, et al. Streptococcus pyogenes pbp2x mutation confers reduced susceptibility to β-lactam antibiotics. *Clinical Infectious Diseases*. 2020;71(1):201–204. https://doi.org/10.1093/cid/ciz1000

77. Musser JM, Beres SB, Zhu L, Olsen RJ, Vuopio J, Hyyryläinen HL, et al. Reduced in vitro susceptibility of streptococcus pyogenes to β-lactam antibiotics associated with mutations in the pbp2x gene is geographically widespread. *Journal of Clinical Microbiology*. 2020;58(4). https://doi.org/10.1128/jcm.01993-19

78. Martin J. The carrier state of streptococcus pyogenes. In: Ferretti J, Stevens D, Fischetti V, editors. Streptococcus pyogenes: basic biology to clinical manifestations. 2nd ed. Oklahoma City: University of Oklahoma Health Sciences Center; 2022.

79. Johnson DR, Kurlan R, Leckman J, Kaplan EL. The human immune response to streptococcal extracellular antigens: clinical, diagnostic, and potential pathogenetic implications. *Clinical Infectious Diseases*. 2010;50(4):481–490. https://doi.org/10.1086/650167

80. Dierksen KP, Inglis M, Tagg JR. High pharyngeal carriage rates of Streptococcus pyogenes in Dunedin school children with a low incidence of rheumatic fever. *New Zealand Medical Journal*. 2000; 24(1122):496-469.

81. Pallon J, Sundqvist M, Rööst M, Danielsson P, Neumark T, Skovbjerg S, et al. Presence of microorganisms in children with pharyngotonsillitis and healthy controls: a prospective study in primary healthcare. *Infection*. 2021;49(4):715–724. https://doi.org/10.1007/s15010-021-01595-9

82. Lacey JA, Marcato AJ, Chisholm RH, Campbell PT, Zachreson C, Price DJ, et al. Evaluating the role of asymptomatic throat carriage of streptococcus pyogenes in impetigo transmission in remote Aboriginal communities in Northern Territory, Australia: a retrospective genomic analysis. *Lancet Microbe*. 2023;4(7):e524–e533. https://doi.org/10.1016/s2666-5247(23)00068-x

83. Munck H, Jørgensen AW, Klug TE. Antibiotics for recurrent acute pharyngo-tonsillitis: systematic review. *European Journal of Clinical Microbiology and Infectious Diseases*. 2018;37(7):1221–1230. https://doi.org/10.1007/s10096-018-3245-3

84. Ng GJ, Tan S, Vu AN, Del Mar CB, van Driel ML. Antibiotics for preventing recurrent sore throat. *Cochrane Database of Systematic Reviews*. 2015;2015(7):CD008911. https://doi.org/10.1002/14651858.CD008911.pub2

85. O'Brien B. Household contact tracing for acute rheumatic fever: a review of the literature and case series [Master's thesis]. Auckland: University of Auckland; 2010.

86. Heart Foundation of New Zealand. Guidelines for group A streptococcal sore throat management guideline: 2019 update. National Heart Foundation of New Zealand; 2019. https://www.heartfoundation.org.nz/resources/group-a-streptococcal-sore-throat-management (Accessed February 18 2025).

87. Paradise JL, Bluestone CD, Bachman RZ, Colborn DK, Bernard BS, Taylor FH, et al. Efficacy of tonsillectomy for recurrent throat infection in severely affected children. Results of parallel randomized and nonrandomized clinical trials. *New England Journal of Medicine*. 1984;310(11):674–683. https://doi.org/10.1056/nejm198403153101102

88. Paradise JL, Bluestone CD, Colborn DK, Bernard BS, Rockette HE, Kurs-Lasky M. Tonsillectomy and adenotonsillectomy for recurrent throat infection in moderately affected children. *Pediatrics*. 2002;110(1 Pt 1):7–15. https://doi.org/10.1542/peds.110.1.7

89. Lock C, Wilson J, Steen N, Eccles M, Mason H, Carrie S, et al. North of England and Scotland Study of Tonsillectomy and Adeno-tonsillectomy in Children(NESSTAC): a pragmatic randomised controlled trial with a parallel non-randomised preference study. *Health Technology Assessment*. 2010;14(13):1–164, iii–iv. https://doi.org/10.3310/hta14130

90. Burton MJ, Glasziou PP, Chong LY, Venekamp RP. Tonsillectomy or adenotonsillectomy versus non-surgical treatment for chronic/recurrent acute tonsillitis. *Cochrane Database of Systematic Reviews*. 2014;2014(11):Cd001802. https://doi.org/10.1002/14651858.CD001802.pub3

91. Guntinas-Lichius O, Geißler K, Mäkitie AA, Ronen O, Bradley PJ, Rinaldo A, et al. Treatment of recurrent acute tonsillitis-a systematic review and clinical practice recommendations. *Frontiers in Surgery*. 2023;10:1221932. https://doi.org/10.3389/fsurg.2023.1221932

92. Kassem AS, Zaher SR, Abou Shleib H, el-Kholy AG, Madkour AA, Kaplan EL. Rheumatic fever prophylaxis using benzathine penicillin G (BPG): two- week versus four-week regimens: comparison of two brands of BPG. *Pediatrics*. 1996;97(6 Pt 2):992–995.

93. Feinstein AR, Spagnuolo M, Jonas S, Kloth H, Tursky E, Levitt M. Prophylaxis of recurrent rheumatic fever. Therapeutic-continuous oral penicillin vs monthly injections. *JAMA*. 1968;206(3):565–568.

94. Tompkins DG, Boxerbaum B, Liebman J. Long-term prognosis of rheumatic fever patients receiving regular intramuscular benzathine penicillin. *Circulation*. 1972;45(3):543–551. https://doi.org/10.1161/01.cir.45.3.543

95. Xie O, Zachreson C, Tonkin-Hill G, Price DJ, Lacey JA, Morris JM, et al. Overlapping streptococcus pyogenes and streptococcus dysgalactiae subspecies equisimilis household transmission and mobile genetic element exchange. *Nature Communications*. 2024;15(1):3477. https://doi.org/10.1038/s41467-024-47816-1

96. Itzek A, Weißbach V, Meintrup D, Rieß B, van der Linden M, Borgmann S. Epidemiological and clinical features of streptococcus dysgalactiae ssp. Equisimilis stg62647 and other emm types in Germany. *Pathogens*. 2023;12(4). https://doi.org/10.3390/pathogens12040589

97. Hysmith ND, Kaplan EL, Cleary PP, Johnson DR, Penfound TA, Dale JB. Prospective Longitudinal Analysis of Immune Responses in Pediatric Subjects After Pharyngeal Acquisition of Group A Streptococci. *Journal of Pediatric Infectious Disease Society*. 2017;6(2):187-196. https://doi.org/10.1093/jpids/piw070

6

Diagnosis of Acute Rheumatic Fever

In this chapter

Key changes

- Transient advanced atrioventricular (AV) block is now included as a major manifestation.
- Streptococcal antibody titres to support the diagnosis of acute rheumatic fever (ARF) have been revised, and the upper limits of normal have been updated (see **Table 6.4**).
- For persons with carditis, either a Strep A throat culture, polymerase chain reaction (PCR), or serology is acceptable to confirm the diagnosis of Definite ARF.
- For persons without carditis, positive serology is required to confirm the diagnosis of Definite ARF.

Key points

- Accurate diagnosis of ARF is important:
  - Missing the diagnosis may lead to an individual experiencing further attacks of ARF, cardiac damage, and premature death.
  - Over-diagnosis will result in the individual receiving benzathine penicillin injections unnecessarily every four weeks for a minimum of ten years.
  - Misdiagnosis may result in another condition remaining undiagnosed and untreated.
- In Aotearoa, high-risk population groups for ARF and rheumatic heart disease (RHD) are Māori and Pacific tamariki and rangatahi, especially those living in low socioeconomic environments.
- In tamariki and rangatahi from high-risk population groups, ARF should always be considered when they present with arthritis or arthralgia.
- Persons with suspected ARF should be discussed with the appropriate hospital team (paediatrics or adult medical), and an acute admission should be arranged for a diagnostic workup.
- Māori tamariki and Pacific tamaiti, with suspected septic arthritis but no identified pathogen on joint aspirate, should be investigated for ARF.
- All persons with suspected ARF should undergo echocardiography (echo) to identify carditis and assess the severity of valvular regurgitation and left ventricular size and function.
- Categories of Definite, Probable, and Possible ARF can be determined by applying Aotearoa diagnostic criteria (adapted Jones Criteria) to each case.

Factors that influence the risk of acute rheumatic fever

Several factors influence the risk of a person experiencing ARF. These include:

- Family/whānau history of ARF or RHD.[1-3]
- Frequency of exposure to a Group A Streptococcus infection.[4]
- Age — most cases occur between the ages of 4 and 20 years[5], and the highest incidence of ARF is in tamariki aged 5 to 14 years.[6] Cases occasionally occur in people in their 20s and 30s.
- Poverty, material deprivation, and household crowding.[1, 6]
- Māori and Pacific peoples are more likely than others to have ARF.[1]
- Barriers to accessing healthcare services.[1]

Diagnostic criteria and challenges

No single gold-standard clinical or laboratory test exists for diagnosing ARF. The diagnosis is made based on the presence of a combination of clinical and laboratory features, along with evidence of a recent Strep A infection. In 1944, the original Jones Criteria were developed in the United States as a clinical classification system, encompassing the clinical and laboratory features of ARF (classified as major and minor criteria). The Jones Criteria have been modified and adapted many times since the 1940s to reflect disease patterns and epidemiology in affected populations, most recently in 2015.[7]

In Aotearoa, national criteria for diagnosing ARF was first published in 2006, reflecting local epidemiologic and clinical data and aiming to avoid both under-diagnosis and over-diagnosis. The 2006 Aotearoa criteria was among the first globally to incorporate carditis detected solely by echo (subclinical carditis) as a major manifestation.

Whilst the 2015 American Heart Association (AHA) Jones Criteria are appropriate for use in international settings, they should not be applied in Aotearoa.[7] The 2015 AHA criteria contain differentiated criteria for high-risk and low-risk population groups, and the Aotearoa guidelines are designed to apply across all population groups. Additionally, the 2015 AHA criteria include more permissive joint manifestations and a lower threshold for inflammatory markers — neither reflects disease patterns in Aotearoa.

Diagnosing ARF can be challenging as clinical features may be non-specific or atypical. Symptoms (particularly arthritis and arthralgia) may overlap with numerous other clinical syndromes. Additionally, the time course of ARF typically lasts for many weeks, and individuals can present at different stages of the illness.

Timely diagnosis of ARF requires four key elements:

1. Health professionals across the entire health system (primary care, emergency departments, paediatrics, orthopaedics, and cardiology) having a high level of awareness of the presenting features of ARF and how to manage a person with suspected ARF.
2. Clear referral pathways for diagnostic evaluation of people with suspected ARF.
3. Accessible, culturally responsive primary and acute care services.
4. Community awareness of the signs and symptoms of ARF.

The likelihood of a person having ARF varies according to the setting and a person's demographic profile. For example, in a region with a high incidence of ARF (such as the northern half of the North Island), a Samoan tamaiti with a fever and arthritis is more likely to have ARF than a non-Māori, non-Pacific adult living in the South Island. Māori and Pacific peoples are more likely than non-Māori, non-Pacific peoples to have ARF.

If the diagnosis is uncertain, then careful communication is required when explaining the complexities and time course of an ARF diagnosis. Make sure that whānau understand that confirming or ruling out ARF is often not possible when the person first presents for assessment and that follow-up and investigations may take weeks or months.

Practitioners should be acutely aware of the consequences of missing a true case of ARF, which puts a person at risk of recurrent disease and lifelong cardiac damage.

In the absence of carditis or an alternative cause for arthritis or involuntary movements, ARF may still be the diagnosis by exclusion. Actively consider alternative diagnoses and plan ongoing review. In instances where the person does not fulfil criteria for Definite ARF, yet the clinician believes the diagnosis may still be ARF, start the person on oral penicillin or give them a first dose of benzathine penicillin and then review in two to four weeks with a second echo to check for evolving carditis.[8, 9]

Triggers for referral — could this person have acute rheumatic fever?

People with the following features should be prioritised for **referral to hospital for assessment** of suspected ARF:

Either: A personal, whānau, or household history of ARF.

Or: Age 3–35 years (highest risk 4–20 years) AND one or more of the following:

- Identify as Māori or Pacific peoples and/or
- Live in crowded circumstances (>2 people sharing a bedroom) in lower socioeconomic areas of the North Island.

In either of these categories, consider ARF if **any** of the following are present:

- Painful or swollen joints (may or may not have a migratory pattern).
- A single painful or stiff joint or limping in tamariki or rangatahi (arthralgia).
- Unexplained breathlessness or concern for congestive heart failure.
- Abnormal involuntary jerky movements (chorea).

Table 6.1. 2024 Aotearoa criteria for the diagnosis of acute rheumatic fever

Manifestations of ARF	Criteria
Major manifestations**	• Carditis**§** (including evidence of subclinical rheumatic valve disease on echo or advanced AV block **§§**) • Polyarthritis or aseptic monoarthritis[a] • Sydenham's chorea**‡‡** • Erythema marginatum • Subcutaneous nodules
Minor manifestations^	• Fever ≥38°C • Raised ESR ≥50mm/hr or CRP ≥30mg/L • Polyarthralgia • Prolonged P-R interval on ECG (corrected for age)
Definite initial episode of ARF	2 major manifestations and evidence of preceding Strep A infection 1 major and 2 minor manifestations, and evidence of a preceding Strep A infection****
Probable initial episode of ARF	1 major and 2 minor, with the inclusion of evidence of a preceding Strep A infection**** as a minor manifestation (as per Jones, 1956)[10]
Possible initial episode of ARF	Strong clinical suspicion of ARF, but insufficient signs and symptoms to fulfil diagnosis of definite or probable ARF
Recurrent ARF in a person with a known history of ARF or RHD	• **Definite**: 2 major manifestations • **Probable**: 1 major and 2 minor manifestations • **Possible**: 3 or more minor manifestations or strong clinical suspicion but insufficient to fulfil diagnosis of Definite or Probable Recurrence Evidence of preceding Strep A infection must be present

Notes

CRP = C-reactive protein

ECG = electrocardiogram

ESR = erythrocyte sedimentation rate

All categories assume that other more likely diagnoses have been excluded.

***** Elevated or rising antistreptolysin O or other streptococcal antibody (Table 6.4) is sufficient support for diagnosing definite ARF. A positive throat culture, PCR, or rapid test alone is less secure, as 50% of people with a positive throat culture will be carriers only.

****** Modified from Jones (1992) and Jones (2015): See text for details about major manifestations.

^ See text for key points about minor manifestations.

‡‡ Chorea can be a stand-alone manifestation for ARF diagnosis, provided other causes are excluded.

§ When carditis is present as a major manifestation (clinical and/or echocardiographic), a prolonged P-R interval cannot be considered an additional minor manifestation.

§§ Advanced AV block (transient second or third-degree heart block or junctional rhythm) is now included in the definition of carditis and as a major manifestation of ARF. Among people with other manifestations of ARF, advanced AV block is highly specific for ARF.[11]

a History of any joint too sore to walk is considered to be arthritis. Other causes of arthritis/arthralgia should be carefully excluded (refer to the differential diagnosis section). If polyarthritis or monoarthritis is present as a major manifestation, then polyarthralgia cannot be considered an additional minor manifestation.

Table 6.2. Comparison of Aotearoa Criteria 2024, Aotearoa Criteria 2014[12], and the revised Jones Acute Rheumatic Fever Diagnostic Criteria[7]

Manifestation	Aotearoa Criteria 2024	Aotearoa Criteria 2014	AHA Revised Jones Criteria 2015 (Moderate and High Risk)
Carditis	Major	Major	Major
Subclinical carditis	Major	Major	Major
Transient advanced AV block	Major	N/A	N/A
Polyarthritis	Major	Major	Major
Aseptic monoarthritis	Major	Major	Major
Polyarthralgia	Minor	Minor	Major
Monoarthralgia	N/A	N/A	Major
Prolonged PR interval	Minor	Minor	Minor
Sydenham's chorea	Major	Major	Major
Erythema marginatum	Major	Major	Major
Subcutaneous nodules	Major	Major	Major
Fever	Minor	Minor ≥38^{0}C	Minor ≥38^{0}C
Elevated acute phase reactants	Minor Either of: • CRP of ≥30mg/L • ESR of ≥50mm/hr	Minor Either of: • CRP of ≥30mg/L • ESR of ≥50mm/hr	Minor Either of: • CRP of ≥30mg/L • ESR of ≥30mm/hr

Clinical features of acute rheumatic fever: Timing in relation to Strep A infection

A latent period usually follows the triggering Strep A infection. During this time, the person appears well before developing symptoms of ARF. This period is generally between 2–4 weeks but is occasionally longer. The latent period may be more extended (up to 8 months) in people with Sydenham's chorea or indolent carditis.[5, 13]

Major manifestations of acute rheumatic fever

Major manifestations of ARF are:

- Arthritis.
- Carditis.
- Sydenham's chorea.
- Subcutaneous nodules.
- Erythema marginatum.

Arthritis

Arthritis is the most common presenting symptom of ARF, occurring in 70% to 80% of first attacks.[5, 6, 14-16] Joint pain is a common childhood concern and can have many underlying causes. It is important that any tamariki with arthritis and suspected ARF are carefully evaluated for other diagnoses (e.g. septic arthritis, autoimmune disease), particularly in the absence of carditis.

Arthritis is defined as swelling of the joint (except for the hip joint) when two or more of the following are present:[17]

- Limitation of movement.
- Hotness of the joint.
- Pain or tenderness in the joint, or both.

Typically, the arthritis in ARF is extremely painful yet highly responsive to Non-Steroidal Anti-Inflammatory Drugs (NSAID).[5, 18] Large joints are most affected, especially the knees and ankles. Polyarthritis is usually asymmetrical and migratory (one joint becoming inflamed as another subsides) but can be additive (multiple joints progressively becoming inflamed without warning). Polyarthritis is non-suppurative. Since the arthritis of ARF is fleeting, a careful history may identify arthritis even in the absence of examination findings. A hip examination is challenging, and the diagnosis of arthritis of the hip is accepted by a history of pain with limping, inability to weight-bear, or limitation of movement on examination.

Polyarthritis is included as a manifestation if at least one joint has been observed in a clinical setting, accompanied by a history of arthritis in other joints (Grade D).

Monoarthritis is included as a major manifestation even if there is no history of NSAID use. In an Auckland case series of rheumatic monoarthritis, 85% had subclinical carditis. This is a similar proportion to cases of ARF with polyarthritis and supports the inclusion of monoarthritis as a major manifestation.[19]

In Aotearoa, NSAIDs are readily available over the counter and are often used before presentation to a GP or hospital.[12] Tamariki or rangatahi who are unable to walk due to severe pain (who do not have clinical arthritis of the knee or ankle) and who have received NSAIDs can also be considered as fulfilling the diagnosis for arthritis in Aotearoa.

Always consider ARF in persons from high-incidence population groups who present with arthritis. Persons with suspected septic arthritis who have sterile joint aspirates, particularly in the absence of prior antibiotics, should also be investigated for ARF.[20]

Carditis

Carditis usually affects the endocardium of the left-sided heart valves. Historically, the terms carditis and valvulitis have been used interchangeably. The incidence of carditis during an initial attack of ARF ranges from 40% to 90%, depending on the setting and the diagnostic criteria applied.[21-24] Rheumatic valvulitis is the active inflammation of the endocardium in ARF. The diagnosis of rheumatic valvulitis is based on the presence of mitral regurgitation (MR) or aortic regurgitation (AR). Isolated tricuspid regurgitation is not a feature of ARF.

Carditis is defined as at least one of the following:

- **Clinical carditis** occurs when abnormal signs are evident, such as a cardiac murmur or signs of congestive heart failure. Clinical mitral valvulitis is an apical holosystolic murmur (mitral regurgitation) with or without a mid-diastolic flow murmur (Carey-Coombs murmur). AR occurs less often, where an early diastolic murmur is heard at the base of the heart. If pericarditis is present, the friction rub may obscure valvular murmurs.
- **Subclinical carditis** is defined as carditis detected on an echo with a normal cardiac examination.
- **Transient advanced AV block** is an uncommon but recognised manifestation of carditis.
- **Indolent carditis** is rare and occurs without other clinical features, such as arthritis, over a protracted time.

Acute rheumatic carditis evolves to chronic RHD, making it the most important prognostic feature in ARF.[5] Rheumatic aetiology can usually be confirmed by a typical appearance on echo (see Table 6.5 and Table 6.6).

Pericarditis and myocarditis may also occur as part of acute carditis. It is rare for these to occur in the absence of valvulitis, and alternate diagnoses (e.g. viral infections) should be carefully considered if myocarditis or pericarditis occurs. Congestive heart failure can occur in ARF due to severe valvular dysfunction secondary to valvulitis and is not due to primary myocarditis.[25] Early disease is typically associated with valvular regurgitation, and ARF recurrences typically lead to worsening valvular regurgitation, valvular stenosis, or a combination of both. While carditis is present at diagnosis in most people with ARF, in some cases, carditis can evolve over several weeks (usually within two to six weeks). Re-assessment and repeat echo is recommended in 2–4 weeks if the initial echo is normal.[26, 27, 28] Regurgitation will improve in 25–50% of people with ARF within one year of the acute episode if secondary antibiotic prophylaxis (SAP) is well-delivered.[29, 30]

Transient advanced AV block

Extreme first-degree heart block (see **Figure 6.1**) is sometimes associated with junctional escape rhythm (see **Figure 6.2**) in ARF, usually with a heart rate similar to the sinus rate. Second-degree and even third-degree AV block (or complete heart block) can occur and, if associated with a slow ventricular rate, may give a false impression that carditis is not significant (see **Figure 6.3**).[31]

In the 1908s, during a resurgence of ARF in the United States, 32% of patients had abnormal AV conduction, usually a prolonged P-R interval. A small proportion had more severe conduction abnormalities, which were sometimes found in the absence of valvular regurgitation.[32]

A recent Aotearoa study found that 8.5% of ARF cases had either advanced AV block or junctional rhythms.[11] Valvulitis was present in 8 of 17 people with advanced AV block.[11] Updated expert consensus is that transient advanced AV block (second or third-degree AV block) accompanied by other clinical manifestations of ARF (typically arthritis or arthralgia) is considered a major manifestation of carditis (Evidence level III-4, Grade C), even in the absence of echocardiographic valvulitis. By definition, this must be transient, i.e. the advanced AV block must return to first-degree AV block or sinus rhythm.

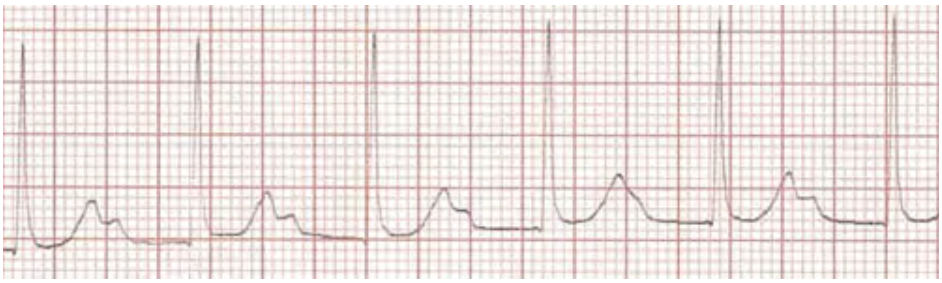

Figure 6.1. Example of extreme first-degree block: P waves merging with T waves[11]

Agnew J, Wilson N, Skinner J, Nicholson R. Beyond first-degree heart block in the diagnosis of acute rheumatic fever. *Cardiology in the Young*, 29, 6, 744–748, 2019 © Cambridge University Press, reproduced with permission.

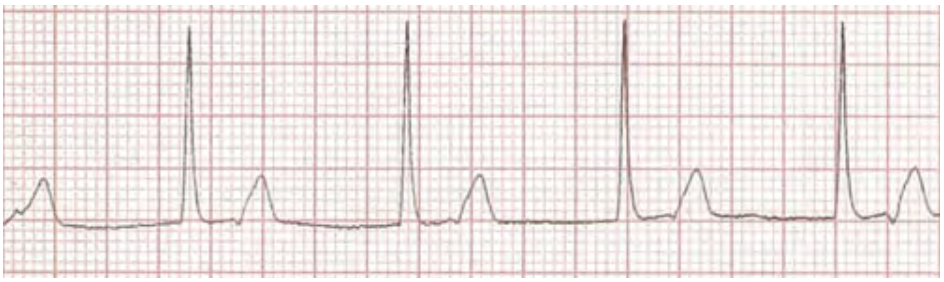

Figure 6.2. Example of junctional rhythm[11]

Agnew J, Wilson N, Skinner J, Nicholson R. Beyond first-degree heart block in the diagnosis of acute rheumatic fever. *Cardiology in the Young*, 29, 6, 744–748, 2019 © Cambridge University Press, reproduced with permission.

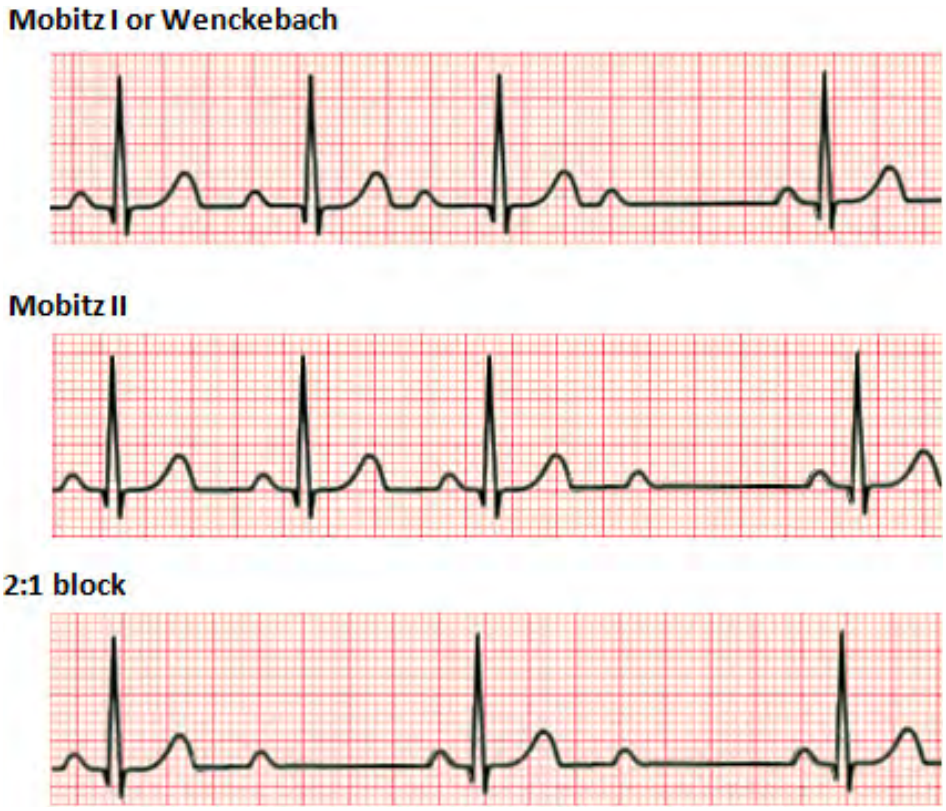

Figure 6.3. Examples of second-degree heart block[33]

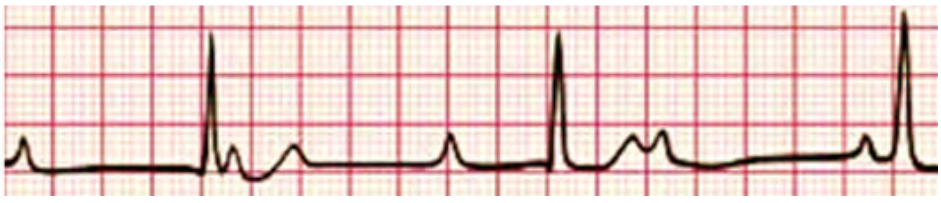

Figure 6.4. Example of third-degree heart block[33]

Indolent carditis

First described in the United States,[10, 34] and sometimes called 'insidious onset carditis.' Indolent carditis is characterised by a subacute illness of several weeks or months, with severe cardiac involvement, few or no joint symptoms, and only modest elevation of inflammatory markers. This rare scenario is recognised in Aotearoa in about two to three tamariki each year.[35] A recent report from the Aotearoa New Zealand Rheumatic Heart Disease Registry research project found that only 0.1% of cases presented with indolent carditis.[36] Evidence of a Strep A infection is not required.[37] Younger tamariki with indolent carditis may have cardiac cachexia and weight loss. However, older tamariki may present with ARF, severe chronic rheumatic valve disease, and significantly impaired ventricular function. This is sometimes described as an "acute on chronic" presentation.

Sydenham's chorea

Sydenham's chorea, also known as rheumatic chorea or St Vitus dance,[5] consists of jerky, involuntary, uncoordinated movements, especially affecting the hands, feet, tongue, and face. It is usually symmetric but sometimes affects one side only (hemichorea). The movements generally disappear during sleep.

Clinical signs of chorea include:[38]

- **"Milkmaid's grip":** Rhythmic squeezing when the individual grasps the examiner's fingers.
- **"Spooning":** Flexion of the wrists and finger extension when the hands are extended.
- **The "pronator sign":** The arms and palms turning outwards when held above the head.
- The person is unable to keep their tongue stuck out.

Chorea affects females predominantly, particularly in adolescence.[39, 40] Chorea typically occurs late in the inflammatory phase of ARF, after a prolonged latent period following Strep A infection.[41-43] Chorea can be a stand-alone manifestation for the diagnosis of ARF, and the ESR and streptococcal antibody titres may be low.

Chorea is strongly associated with carditis, and an echo is essential, regardless of cardiac murmurs (Evidence level IV, Grade C). During outbreaks of ARF in the United States, up to 71% of patients with chorea had carditis.[44]

Even in the absence of echocardiographic evidence of carditis, people with chorea should be considered at risk of subsequent cardiac damage.[45] Prior to the availability of echo, approximately 25% of patients with 'pure' chorea also eventually developed clinical RHD.[45, 46] Therefore, all persons with chorea should receive SAP and be carefully followed up for subsequent development of RHD.

Chorea usually resolves within six months (usually in 12 to 15 weeks),[47] but rare cases have lasted as long as three years.[38] Chorea can recur during times of stress, and intercurrent flares of chorea symptoms may be associated with pregnancy (chorea gravidarum) or oral contraceptive use.

Emotional lability and functional impairment (inability to hold utensils, pens, clumsiness, falls due to unsteady gait) are common manifestations of chorea, although they may be under-recognised. Getting a detailed history from the individual and their caregivers is important.

A study using the Work and Social Adjustment Scale (WSAS) showed significantly ($p = 0.021$) higher alterations in the Sydenham's chorea group than in the non-Sydenham's chorea group, indicating that chorea has a strong adverse psychological impact on individuals, even years after its onset.[47]

Subcutaneous nodules

Subcutaneous nodules (see **Figure 6.5**) are a rare (less than 2% of cases) yet highly specific manifestation of ARF.[48] The nodules are between 0.5cm and 2.0cm in diameter, round, firm, freely mobile, and painless. The nodules occur in crops of up to 12 on extensor surfaces and bony prominences, including occiput, elbows, wrists, knees, ankles, Achilles tendon, and posterior spinal processes of the vertebrae. They typically appear 1 to 2 weeks after the onset of other symptoms and last only 1 to 2 weeks (rarely more than 1 month). Nodules are strongly associated with carditis and must be accompanied by additional major manifestations to make the diagnosis.

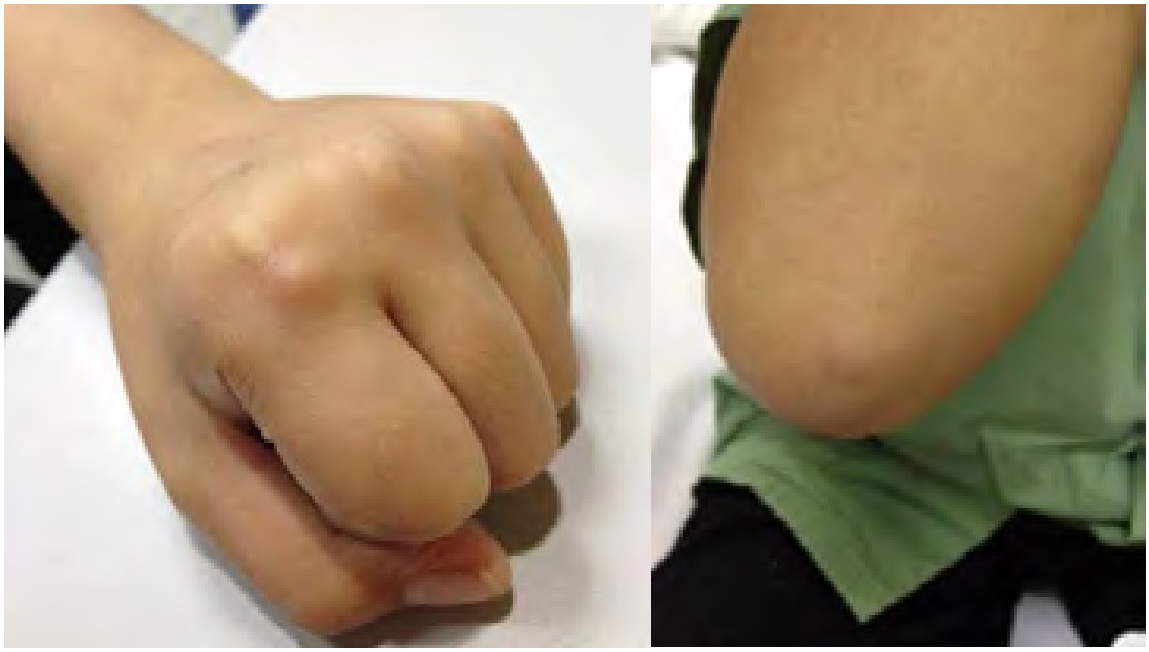

Figure 6.5. Subcutaneous nodules[49]

Erythema marginatum

Erythema marginatum occurs as circular patterns of bright pink macules or papules that blanch under pressure and spread outwards in a circular or serpiginous pattern on the trunk and proximal extremities (almost never on the face). See **Figure 6.6** for examples.

Erythema marginatum is rare, seen in <6% of individuals with ARF[7], and can be hard to detect in darker-skinned people. The rash can fade and reappear within hours; it may reappear in hot conditions and be more apparent after showering. The patient and their caregiver should be alerted to look for this. Erythema marginatum lesions are not itchy or painful and are not affected by anti-inflammatory medication. Erythema marginatum may recur for weeks or months, even after other features of ARF are resolved.

Erythema marginatum is rarely seen as the sole major manifestation of ARF. Additional major manifestations are needed to make the diagnosis.

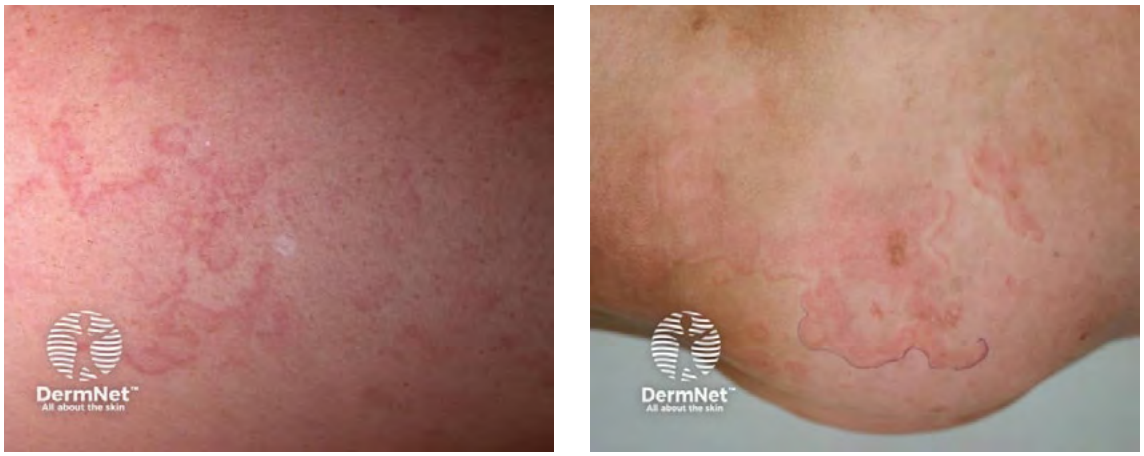

Figure 6.6. Erythema marginatum[50]

Minor manifestations of acute rheumatic fever

Minor manifestations of ARF are:

- Polyarthralgia.
- Fever.
- Elevated acute phase reactants.
- First-degree heart block (prolonged P-R interval).

Polyarthralgia

Arthralgia is pain in the joints without objective findings such as swelling or heat.[10] The pain is associated with movement and is usually out of proportion to clinical findings. Polyarthralgia is common in tamariki and may be non-specific, although in ARF, it tends to follow a similar pattern to polyarthritis (migratory, asymmetrical, and affecting large joints). In Aotearoa, polyarthralgia remains a minor manifestation (Grade D). Polyarthralgia cannot be counted as a minor manifestation if polyarthritis is present as a major manifestation.[10]

Polyarthralgia has been included as a major criteria in high-risk population groups in the 2015 Jones criteria and the 2020 Australian guidelines.[7, 51]

Consider alternative diagnoses (as suggested in a differential diagnosis list, see page 21) in an individual with arthralgia, particularly if they do not meet the criteria for Definite ARF.

In Aotearoa, monoarthralgia (single joint pain with retained ability to walk or freely move the joint) is not considered a minor manifestation.

Fever

Most individuals with ARF report fever (except for chorea). In Aotearoa, an oral, tympanic, or rectal temperature greater than or equal to 38°C on admission or documented during the current illness should be considered a fever (Evidence level IV, Grade C). In Australia, defining fever as a temperature of >38°C instead of 39°C improved the sensitivity for diagnosing ARF in Aboriginal and Torres Strait Islander People.[48] Fever, like joint manifestations, is usually quickly responsive to NSAIDs and may have resolved by the time of presentation.

Elevated acute phase reactants

ARF is a systemic inflammatory process. The serum C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) are elevated. In Aotearoa, serum CRP level of ≥30mg/L or ESR of ≥50mm/h meets the diagnostic threshold (Grade D). The peak ESR in ARF is typically >80mm/hr, usually remains elevated for around 4 weeks, and may remain elevated for 3 to 6 months even if the symptoms have resolved. The serum CRP concentration rises more rapidly than the ESR and falls more rapidly. It is also less consistently raised on admission in ARF compared to the ESR.[19] A retrospective audit from the Auckland Region Rheumatic Fever Register reviewed trends in inflammatory markers from ARF cases between 2000 and 2016. Among 361 ARF cases, 86% met the CRP cut-off and 95% met the ESR cut-off, with only a fair correlation between the two markers (87% observed agreement, Kappa 0.27 (unpublished).*

First-degree heart block (prolonged P-R interval)

Transient prolongation of the P-R interval can occur in ARF. A prolonged P-R interval can also occasionally occur in healthy people. Even so, a prolonged P-R interval that appears or resolves over the ensuing days to weeks may be a useful diagnostic feature of ARF, and serial ECGs may be useful, especially if carditis is not evident on echo.[32]

The P-R interval increases with age and needs to be age-adjusted. The upper limits of normal are:

- Age 3–12 years 0.16 seconds
- Age 12–16 years 0.18 seconds
- Age ≥17 years 0.20 seconds.

Adapted from Park's pediatric cardiology for practitioners.[52]

When carditis is present as a major manifestation, prolonged P-R interval is not considered an additional minor manifestation.

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* Stuart PM, Lennon D, Reed P. The Diagnostic Value of Erythrocyte Sedimentation Rate Versus C-Reactive Protein in Acute Rheumatic Fever. 2019.

Other disturbances of cardiac rhythm in acute rheumatic fever

(Also see the subsection **Transient advanced AV block** under 'Clinical features of acute rheumatic fever: Major manifestations'.)

In a 2019 study of tamariki in Aotearoa aged <15 years with a clinical diagnosis of ARF, QTc prolongation was noted. The mean (SD) QTc was 445 milliseconds, ranging from 370 to 545 milliseconds. Also, 4% of patients had a QTc over 500 milliseconds, while 18% of the cohort had a QTc >99th percentile of normal by age.[53]

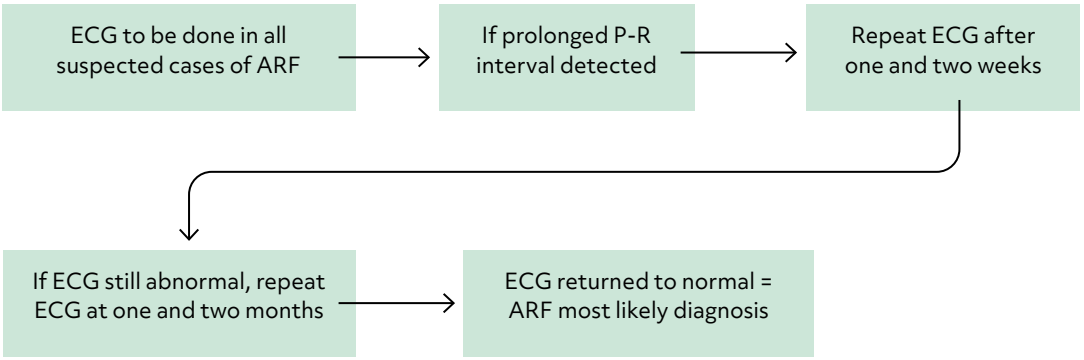


Figure 6.7. The recommended approach to performing an electrocardiogram in the diagnosis of acute rheumatic fever

Key messages regarding prolonged P-R interval and other rhythm abnormalities in ARF

- An ECG should be performed in all cases of suspected ARF (Evidence level IV, Grade B).
- Prolonged P-R interval is a minor manifestation but cannot be included when carditis is present as a major manifestation.
- In the absence of clinical or echocardiographic carditis, transient second or third-degree heart block accompanied by other manifestations of ARF is elevated to a major manifestation of carditis due to its high specificity for ARF in Aotearoa[11] (Grade B).

Other uncommon clinical features of acute rheumatic fever

Other uncommon clinical features of ARF include:

- Epistaxis (occurs commonly but is not specific for ARF).
- Abdominal pain (occurs commonly but is not specific for ARF).
- Rheumatic pneumonia.
- Mild elevations of plasma transaminase levels and microscopic haematuria.
- Pyuria or proteinuria.

Rheumatic pneumonia represents pulmonary infiltrates (pulmonary oedema) in individuals with acute carditis with ruptured chordae tendineae.[54] There are rare case reports of acute post-streptococcal glomerulonephritis (APSGN), another immune-mediated sequala of Strep A infection, occurring at the same time as ARF.[55]

Evidence of preceding Group A streptococcal infection

Evidence of a preceding Strep A pharyngitis (preferably with evidence of immunologic response to pharyngitis measured by streptococcal antibody titres) is critical to diagnosing ARF. The exceptions are chorea and indolent carditis, which are typically late presentations.

Table 6.3. Summary of Strep A testing for acute rheumatic fever diagnosis in Aotearoa

Test	Timing	Evidence to support diagnosis
Strep A serology	At presentation Repeat at 2–4 weeks if not initially raised	Can support a diagnosis of Definite or Probable ARF
Positive Strep A throat culture or PCR	At presentation or in the 2–4 weeks prior to symptom onset (occasional cases occur outside this timeframe)	Can support a diagnosis of Definite ARF if carditis is present Can support a diagnosis of Probable or Possible ARF in the absence of carditis
Positive Strep A rapid Antigen test of throat		Insufficient evidence and is not accepted as evidence for diagnosis in Aotearoa
Positive Strep A skin swab		Insufficient evidence and is not accepted as evidence for diagnosis in Aotearoa

Streptococcal serology

Elevated or rising streptococcal antibody titres are crucial to support the diagnosis of ARF.

The tests used in Aotearoa and most of the world are the antistreptolysin O (anti-SLO, ASO) and the anti-deoxyribonuclease B (anti-DNase B, ADB) titres. ASO titres typically reach a maximum at about 3 to 6 weeks after Strep A infection and ADB titres can take up to 6 to 8 weeks to reach a maximum.[56] The rate of decline of these antibodies varies considerably, with the ASO titre starting to fall 6 to 8 weeks after infection and the ADB titre starting to fall 3 months after infection.[57] In the absence of re-infection, the ASO titre usually approaches pre-infection levels between 6 and 12 months after infection, while the ADB titre tends to remain elevated for longer.[58]

Indications for streptococcal serology

Streptococcal serology should only be performed when there is a clinical concern for immune sequelae of Strep A infection (i.e. suspected ARF or PSGN), usually in a hospitalised patient. It has no role in the diagnosis of acute suppurative Strep A infections (Grade D).

Reference intervals for streptococcal serology

Upper limits of normal for streptococcal titres have been determined for several populations around the globe, usually set at the 80th percentile.

In Aotearoa, in the early 1980s, thresholds were derived from residual sera of a historic cohort of young hospitalised Auckland patients without ARF and reported in 1982. An ASO titre of greater than or equal to 480 international units and/or an ADB titre of greater than or equal to 680 international units was accepted as elevated, with lower levels acceptable in the very young or those over the age of 15 years.

These historic reference intervals have recently been updated using modern laboratory methods. The sample population tested was a contemporary cohort of 224 healthy tamariki from Auckland who participated in a rheumatic fever case-control study and a study comparing antibody titres of healthy tamariki to those with skin and throat infections.[1, 59]

Table 6.4. 2024 updated upper limits of normal for serum streptococcal antibody titres used in Aotearoa for acute rheumatic fever

Antibody titres	International units/mL for Aotearoa in 2024	International units/mL for Aotearoa in 2006 and 2014
Anti-Streptolysin-O (ASO)	≥450	≥480
Anti-DNase B (ADB)	≥400	≥680

Interpretation of streptococcal serology

Interpretation of streptococcal serology also has some limitations and caveats:

- The reference range for streptococcal antibody titres varies with age and geographical location. In settings with high exposure rates to streptococcal infections, background population titres in healthy tamariki are high. Younger (preschool) tamariki and adults typically have lower titres than school-aged tamariki and rangatahi.
- Rising titres are preferred to single-raised titres. Historically, the best evidence of an immune response to an antecedent Strep A infection was said to be a twofold or more increase in titre from acute to convalescence (usually 12 to 28 days apart).[7] However, people with ARF typically present with an established immunological response. Additionally, it is unclear whether a twofold increase and eventual fall in titres are always seen with modern laboratory methods (turbidimetry and nephelometry).
- Different laboratories may use different laboratory methods (assays) for ASO and ADB measurement, and results may not be comparable. Additionally, most laboratories acknowledge measurement of uncertainty of up to 10% when reporting results.
- Repeat (paired) serology should always be performed after 2–4 weeks if the initial titres are not elevated and the diagnosis of ARF is suspected but not certain.[5]
- At some laboratories, reference ranges accompanying test results reflect the test manufacturer's guidance and do not align with the Aotearoa New Zealand Guidelines for the Prevention, Diagnosis, and Management of Acute Rheumatic Fever and Rheumatic Heart Disease. Typically, manufacturers' cut-offs are lower than those of Aotearoa New Zealand Guidelines for the Prevention, Diagnosis, and Management of Acute Rheumatic Fever and Rheumatic Heart Disease. The recommendation is that guideline values are used in place of any manufacturer's cut-offs, for the diagnosis of ARF in Aotearoa.

Pharyngitis and pharyngeal culture for Strep A

Pharyngitis is a common problem in tamariki and rangatahi. Most pharyngitis has a viral aetiology, and Strep A accounts for around 10% of pharyngitis in high-incidence ARF populations globally.[60] A positive pharyngeal culture is the gold standard for diagnosing Strep A pharyngitis,[5] and has a sensitivity of 90% to 95% in detecting Strep A when performed correctly.[61] Recent data from South Auckland suggests that around 60% of ARF cases have Strep A cultured from the pharynx in the month before diagnosis. Rates of pharyngeal Strep A isolation are much lower in Northern Australia — in one study, Strep A was isolated in less than 5% of ARF cases.[48] A positive culture without supportive antibody elevation may reflect asymptomatic colonisation (carriage) and some practitioners consider this to be less secure evidence of antecedent Strep A infection than elevated antibody titres.[34] In Aotearoa, a positive throat swab in the 2–4 weeks prior to diagnosis or at presentation can support a diagnosis of Definite ARF if carditis is present. In the absence of carditis, positive serology is required to support the diagnosis of Definite ARF (Grade D).

Strep A skin cultures are not accepted for acute rheumatic fever diagnosis in Aotearoa

Although data linkage studies have observed an epidemiologic association between Strep A skin culture and ARF at a population level in Aotearoa,[4] the role of skin infection and ARF immunopathogenesis at the individual patient level remains uncertain. At present, bacteriologic cultures of Strep A from the skin are not accepted as evidence of antecedent Strep A infection for the diagnosis of ARF in Aotearoa.

Rapid molecular testing for Strep A

Rapid molecular tests for Strep A have become commercially available in recent years. These may be useful in rapidly assessing and managing sore throats in people at high risk of ARF. TheGeneXpert® Xpress A test has been shown to be highly sensitive, with a strong negative predictive value when compared to traditional culture at Middlemore Hospital, Auckland.[62] Molecular testing has also shown promise in Northern Australia.[63] The scale-up of rapid molecular testing for Strep A pharyngitis has the potential to:

- Improve access to timely sore throat diagnosis and treatment.
- Assist with the diagnosis of ARF.
- Assist with community antimicrobial stewardship.

Workforce training, equipment costs and ensuring reach into at risk populations require further consideration.[64] Further research is needed regarding the clinical significance of positive molecular results when pharyngeal culture is negative. At this time in Aotearoa, molecular detection of Strep A is considered equal to culture to support a diagnosis of ARF.

Rapid antigen tests for Strep A

Rapid antigen detection tests (RADT) are currently not recommended in Aotearoa. They are quick and easy to use however, their diagnostic accuracy varies. Two studies performed in the Auckland region showed suboptimal performance, with one study finding a sensitivity of only 26% compared to conventional culture.[65] RADT is not recommended for diagnosing Strep A pharyngitis or ARF in Aotearoa due to concerns about missing Strep A in high-incidence ARF populations.[66] Additionally, widespread use of RADT without appropriate clinical governance may lead to over-testing and over-treating low-incidence ARF populations, with adverse consequences for community antimicrobial stewardship.

Recurrent acute rheumatic fever and its diagnosis

After a first episode of ARF, a person remains susceptible to further episodes.

Recurrent ARF is classified as Definite, Probable, or Possible ARF using the approach listed in **Table 6.1**. To make a diagnosis of recurrent ARF, the person must have a previous formal diagnosis of ARF or RHD, for which SAP was recommended. Recurrences of ARF are an important indicator of the performance of SAP services.

In Aotearoa, ARF recurrence is uncommon compared to many regions of the world. A recent national audit found that 7.2% of people with ARF experienced an episode of recurrent ARF. However, rates were lower (4%) among tamariki <16 years receiving SAP compared to rangatahi.[67] Most recurrences occur due to suboptimal delivery of SAP because the person is not receiving benzathine penicillin on a 28-day cycle. True penicillin failure in persons strictly adherent with 28-day/four-weekly SAP can occur,[68] but this is very rare in Aotearoa, particularly for tamariki on SAP administered through community nursing programmes.

Recurrence may also occur many years after the initial ARF diagnosis, even following the planned cessation of SAP — the time since diagnosis influences the risk of recurrence. Recent data for Aotearoa found the median time from initial ARF to recurrent ARF hospitalisation was 3.2 years (IQR 1.9–8.4 years).[69] In Australia, recurrent ARF rates were highest (an incidence of 3.7 per 100 person-years) in the first year after the initial ARF episode, with a low level of risk persisting for >10 years.[70]

Before a diagnosis of recurrent ARF can be made, there should be a period of more than 180 days after the onset of symptoms from the previous ARF episode (Grade D). The exception is chorea, which can take a fluctuating course over many months. If there is evidence that the ESR has normalised or benzathine penicillin has been missed since the previous ARF episode, a recurrence may be considered within 180 days. Care must also be taken to consider and rule out other differential diagnoses that may cause prolonged joint or neurologic manifestations (particularly rheumatologic disorders such as juvenile arthritis).

In some situations, it can be difficult to clinically distinguish first-episode ARF from recurrent ARF (for example, in a person hospitalised for the first time with ARF, who has features of chronic RHD on echo, or who gives a history of a past illness suspicious for missed ARF). In these circumstances, the individual should be notified as a first ARF episode.

The diagnosis of ARF recurrence should only be made when a person with a **reliable** and **documented** history of previous ARF or RHD presents with clinical features of ARF **and** evidence of a preceding Strep A infection.[71]

Differential diagnosis of acute rheumatic fever

Many features of ARF are non-specific. Consider a wide range of differential diagnoses.

Polyarthritis or monoarthritis

The differential diagnosis of polyarthritis or monoarthritis includes:

- Bacterial infection, such as septic arthritis due to *Staphylococcus aureus*.

- Reactive arthritis: infectious triggers may include cytomegalovirus, Epstein-Barr Virus, Mycoplasma, rubella vaccination, hepatitis B, parvovirus, Yersinia, and other gastrointestinal pathogens.
- Autoimmune and connective tissue disorders include rheumatoid arthritis, juvenile chronic arthritis, inflammatory bowel disease, Still's disease, systemic lupus erythematosus, and systemic vasculitis.
- Sickle-cell anaemia.
- Infective endocarditis.
- Leukaemia or lymphoma.
- Gout and pseudogout.
- Post-streptococcal reactive arthritis (see **Poststreptococcal reactive arthritis** section).

Carditis

A detailed echo can differentiate carditis. Differential diagnoses include:

- Congenital mitral valve prolapse.
- Congenital heart disease.
- Innocent (physiologic) murmur.
- Infective endocarditis.
- Viral myocarditis or pericarditis.

Transient AV block

- Lyme disease (does not occur in Aotearoa, but consider if the person has travelled overseas to a Lyme-endemic region).
- Diphtheria (consider in an un-immunised tamariki with other clinical features).

Chorea

Differential diagnosis of chorea include:

- Systemic lupus erythematosus.
- Drug ingestion (includes anticonvulsants, antidepressants, lithium, scopolamine, calcium channel blockers, methylphenidate, theophylline, and antihistamines).
- Wilson's disease (also called Hepatolenticular degeneration).
- Tic disorder and Tourette syndrome.
- Choreoathetoid cerebral palsy.
- Familial chorea (including Huntington's disease).
- Intracranial tumour.
- Metabolic diseases such as Lesch-Nyhan, hyperalanaemia, ataxia, or telangiectasia.
- Antiphospholipid syndrome.
- Chorea gravidarum while pregnant.
- Hyperthyroidism and hypoparathyroidism.
- PANDAS.

Paediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcal infection (PANDAS)

The term PANDAS describes clinical features occurring in tamariki with tic or obsessive-compulsive disorders (OCD), whose symptoms are thought to develop or worsen following Strep A infection. Mild or atypical chorea may be confused with motor tics, Tourette syndrome, or PANDAS.

The five criteria used to define PANDAS are:[72, 73]

- The presence of a Tic disorder, OCD, or both.
- The pre-pubertal age of onset (usually between 3 and 12 years of age).
- Abrupt symptom onset, episodic course of symptom severity, or both.
- Temporal association between the symptom and the streptococcal infection (usually between 7 and 14 days).
- Presence of neurologic abnormalities during periods of symptom exacerbation (typically adventitious movements or motoric hyperactivity).

The evidence supporting PANDAS as a distinct disease entity is debated.[73] In Aotearoa, where there is a high incidence of ARF and chronic RHD, clinicians should rarely diagnose PANDAS. Instead, clinicians should consider Sydenham's chorea and commence presumptive SAP (Grade D). These cases should be carefully evaluated by a neurologist and followed up to ensure they do not develop movement disorders. An echo should be performed to check for RHD.

Poststreptococcal reactive arthritis (PSRA)

Rarely, people may present with atypical arthritis affecting the small joints (such as fingers), along with evidence of recent streptococcal infection. This entity is described in some settings as PSRA. PSRA has been noted to have a bimodal age distribution involving the age groups 8 to 14 and 21 to 37 years.[74] PSRA is considered less responsive to anti-inflammatory treatment than ARF. Historically, individuals with PSRA have been considered to be at no risk of carditis and therefore, do not require SAP. However, some individuals diagnosed with PSRA have developed later episodes of ARF, indicating that the initial diagnosis should have been ARF (Grade D).[75, 76]

It is recommended that the diagnosis of PSRA is rarely made in high-risk populations and with caution in low-risk populations (Evidence level IV, Grade C). In Aotearoa, these cases should usually be classified as Possible or Probable ARF (Grade D). Alternate rheumatologic diagnoses should also be carefully considered. Individuals with suspected PSRA should be followed up for at least one year.[77] SAP may be recommended for one year if in a low-risk population group for ARF (Grade D) and 5 years if high-risk (Grade D). Use an echo to confirm the absence of RHD before discontinuing SAP (Grade D).

Investigations in suspected acute rheumatic fever

The recommended investigations in ARF are listed below. Other investigations may be appropriate depending on the clinical picture and potential differential diagnoses.

Recommended investigations for all cases of suspected ARF

The recommended investigations in ARF are:

- ESR — repeat weekly after diagnosis is confirmed.
- CRP.
- Blood cultures if febrile.
- ECG — repeat as necessary, including if conduction abnormality is more than first degree or the initial ECG is normal and the individual has no other evidence of carditis.
- Echo — repeat as necessary in 2 to 4 weeks if the initial echo is normal or equivocal or if serious carditis.
- Throat swab culture for Strep A (preferably before giving antibiotics).
- Anti-streptococcal serology (both ASO and ADB titres) — repeat 10 to 14 days later if the first test does not provide confirmation.
- Chest X-ray if clinical or echo concern for heart failure.

Tests to consider for alternative diagnoses, depending on clinical features

A range of tests are available to confirm an alternative diagnosis, depending on the clinical features present. Examples include:

- Serology for arthritis: cytomegalovirus, Epstein-Barr Virus, Mycoplasma, rubella, hepatitis B, parvovirus, influenza, Yersinia and other gastrointestinal pathogens.
- Anti-nuclear antibody (ANA) and rheumatoid serology for autoimmune arthritis.
- Blood cultures for possible endocarditis or septic arthritis.
- Joint aspirate (microscopy and culture) for possible septic arthritis.
- Joint X-ray or MRI imaging (suspected septic arthritis).
- Copper, caeruloplasmin, drug screen, and consider MRI brain for atypical choreiform movements. Rheumatic chorea can usually be diagnosed based on history, physical examination, and laboratory evaluation. Neuroimaging is seldom necessary and should be reserved for cases with atypical features such as hemichorea.[78]

Echocardiography in acute rheumatic fever

In Aotearoa, echo is available for populations at high risk of ARF. All persons with suspected or Definite ARF should undergo echo to:

- Identify carditis (including subclinical carditis).
- Assess the severity of regurgitation and left ventricular size and function (Grade C).

Table 6.5. Uses of echocardiography in acute rheumatic fever

Diagnosis	Echocardiographic evidence of carditis
Valvulitis	• Identify subclinical evidence of valvular regurgitation. • Define the severity of mitral, aortic, or tricuspid regurgitation and any combination of these. • Define the mechanism of regurgitation (such as prolapse, flail leaflet, and annular dilatation). • Define the severity of mixed valve disease (mixed stenosis and regurgitation). • Visualise the anatomy of the valves, especially in MR. This is vital when making decisions about surgery.
Myocarditis and congestive heart failure	• Assess the left ventricular size and function. • Assess the severity of valvulitis (valvulitis is usually present in ARF with heart failure).
Pericarditis	• Confirm the presence of a pericardial effusion. • Reveal inaudible or subclinical valvular regurgitation in the presence of a friction rub.
Exclude other forms of cardiac murmur	• Identify congenital heart disease, such as bicuspid aortic valve and congenital mitral valve anomalies, as the cause of a pathological murmur. • Confirm normal valvular function and morphology if a flow murmur or innocent murmur is present.

Adapted from the 2014 New Zealand Guidelines for Rheumatic Fever[12] and the 2020 Australian guideline for prevention, diagnosis, and management of acute rheumatic fever and rheumatic heart disease.[51]

The use of colour-Doppler echo, permitting definitions of echo findings as a major manifestation of ARF diagnosis, has evolved over the past two decades. Wilson and Neutze summarised the original studies.[79]

The minimal colour-Doppler criteria for valvulitis of ARF are the same as those for diagnosing rheumatic MR and AR. The World Heart Federation (WHF) guidelines for echocardiographic diagnosis of rheumatic heart disease[80] were published in 2012 and updated in 2023.[81] Colour-Doppler findings of valvular regurgitation of acute carditis do not differ from those of chronic RHD valve regurgitation. The Aotearoa New Zealand Guidelines for the Prevention, Diagnosis, and Management of Acute Rheumatic Fever and Rheumatic Heart Disease endorse the same colour-Doppler criteria for defining the acute and chronic phases.[7, 51] Table 6.6 shows the colour-Doppler criteria for diagnosing pathological valvular regurgitation.

Table 6.6. Echocardiographic criteria for pathological regurgitation[80, 82]

Type	Criteria
Pathological mitral regurgitation (MR) (all criteria must be met)	• Observed in two views • In at least one view, MR length >2cm. In those weighing <30kg MR, jet length >1.5cm • Peak velocity >3.0m/s for one complete envelope • Pan-systolic jet in at least one envelope
Pathological aortic regurgitation (AR) (all criteria must be met)	• Observed in two views • Jet length >1cm • Peak velocity >3.0m/s in early diastole • Pan-diastolic jet in at least one envelope
Mitral stenosis (all criteria must be met)	• Restricted leaflet motion with reduced valve opening • Mean peak gradient >4mmHg

These criteria can usually readily distinguish a small colour jet of physiological regurgitation in normal tamariki from pathological regurgitation in a tamariki with ARF or RHD. The proportion of tamariki with physiological valve regurgitation in Aotearoa was 15%[83] and large international cohort studies show that this proportion rises with age.[84]

If the aetiology of AR or MR on the Doppler echo is not clear, the following features support (but are not individually specific for) a diagnosis of rheumatic valve damage:

- Both mitral and aortic valves have pathological regurgitation.
- The mitral regurgitant jet is directed posteriorly, as excessive leaflet motion of the tip of the anterior mitral valve leaflet (AMVL), often referred to as prolapse, is the most common mechanism of MR. Anterior leaflet prolapse is more common than posterior valve prolapse.
- Multiple jets of MR are evident.
- The presence of excessive leaflet motion of the tips /edges of the AMVL or posterior mitral valve leaflet (PMVL) is due to chordal lengthening, rupture, or both.

Morphological features of RHD take time to develop but may be present in ARF. Echo cannot date the duration of any of these changes. More advanced features that support an acute ARF on chronic RHD presentation are:

- Restrictive leaflet motion with gross subchordal thickening †,‡
- Gross thickening of AMVL at >5mm
- Unequivocal immobile PMVL †,‡
- Mitral stenosis with a mean valve gradient of >5mmHg (up to 5mmHg can be enhanced mitral filling in acute severe MR)

(Also see Chapter 10: Diagnosis of Rheumatic Heart Disease)

Symbols used

† Immobility of the subchordal apparatus and posterior leaflet is observed only after several months. Other findings have also been reported, including acute nodules, which show a beaded appearance of the mitral valve leaflets.[85]

‡ It is recommended to avoid using descriptive terms such as 'elbow,' 'dog leg' or 'hockey stick' when describing a deformity of the AMVL — such appearances are due to the combination of valve thickening and restrictive leaflet motion.[80]

Source: The content in this table is based on an original study by Wilson NJ and Neutze JM[79] as well as criteria that further evolved as part of both the Aotearoa and the Australian guidelines on rheumatic fever diagnosis, the WHO working groups on echocardiography and the 2023 WHF guidelines for echocardiographic diagnosis of rheumatic heart disease.[81]

Assessing the severity of acute rheumatic fever carditis

In Aotearoa, ARF carditis is classified as none, mild, moderate, or severe (as described in Table 6.7). These categories are used to inform clinical follow-up recommendations.

Table 6.7. Severity of acute rheumatic fever carditis

Severity	Presentation
Mild carditis	• Mild MR or AR clinically or on echo (fulfilling the minimal echocardiographic standards in Table 6.6 without heart failure, cardiac chamber enlargement on X-ray, ECG, or echo).
Moderate carditis	Any one of these: • Mitral or aortic valve lesion of moderate severity, as found on clinical examination • Cardiac chamber enlargement, as seen on an echo • Any valve lesion graded as moderate, as seen on echo. Regurgitation is considered moderate if: • A broad high-intensity proximal jet is filling half the left atrium — this means a mitral or a lesser volume high-intensity jet is producing a prominent blunting of the pulmonary venous inflow[9] • Diameter of the regurgitant jet is 15% to 30% of the diameter of the left ventricular outflow tract, flow reversal in the upper descending aorta.[9] When both MR and AR exist, one must be moderated by echo criteria for the carditis to be classified as being moderately severe.
Severe carditis	Any one of these: • Impending or previous cardiac surgery for RHD • Valve lesion associated with significant cardiomegaly or heart failure or graded as severe on clinical examination • Any valve lesion graded as severe on an echo. In tamariki: • An abnormal regurgitant colour and Doppler flow patterns in pulmonary veins is a prerequisite for severe MR[9] • Doppler reversal in the lower descending aorta is required to diagnose severe AR.[9] In adults: • Doppler flow reversal in the pulmonary veins (for severe MR) or abdominal aorta (for severe AR) is specific if present, but can be more difficult to detect — indeed, severe regurgitation may still be present if not detected.

Tricuspid and pulmonary regurgitation graded mild or greater may be seen in individuals with normal hearts with fever, volume overload, or pulmonary hypertension. For this reason, never base a diagnosis of carditis on right-side regurgitation alone. Tricuspid regurgitation is often seen in association with left-sided lesions in ARF, and pressure and volume overload need to be excluded before attributing even moderate tricuspid regurgitation to rheumatic valvulitis of the tricuspid valve. If both left and right-sided lesions coexist in ARF carditis, then the predominant influence for diagnosis is the severity of the left-sided lesion.

If valvulitis is not found at presentation, it may appear within two weeks,[8] or sometimes within one month[9] but not longer. If the initial echo is normal and the diagnosis of ARF is uncertain, then the individual should have a further follow-up in two to four weeks.[35] A series from Aotearoa during the late 1990s showed that clinical carditis occurred in 55% of cases and subclinical carditis in 30%, with only 14% of cases showing no carditis.[35]

Usually, it is not always possible to distinguish between acute carditis and pre-existing RHD by echo. In an individual with known previous RHD, the diagnosis of acute carditis during a recurrence of ARF relies on accurate documentation of the cardiac findings before the recurrence so that new clinical or echo features can be confirmed. In an individual with no history of ARF or RHD, whether echo changes are new or old makes little difference.

New developments in diagnosing acute rheumatic fever and carditis

No single definitive diagnostic test exists for ARF. Studies are ongoing to look for biomarkers and imaging to help diagnose ARF. A biomarker discovery study titled *Searching for a Technology-Driven Acute Rheumatic Fever Test* (START) aims to detect and test a biomarker signature that distinguishes ARF cases from non-ARF cases. Participants were recruited from three hospitals in Australia and Auckland between 2019 – 2021 and laboratory analyses are underway.[86]

Cardiac magnetic resonance (CMR) studies using the technique of myocardial T1 mapping have found that patients with ARF have elevated median native myocardial T1 times.[87] The studies showed that the increase in T1 time was significant in patients with ARF compared with both inflammatory control subjects (p= 0.002) and healthy control subjects (p= 0.003).[87] T1 elevation may be a mechanism for diagnosing carditis in ARF in the future.

"I was adamant that it was rheumatics [rheumatic fever] because I had dealt with the symptoms before with his brother but they kept putting it off and kept telling me it was rheumatism arthritis and then yeah, couple years later he's diagnosed with rheumatics."

Mother of tamaiti diagnosed with ARF

References

1. Baker MG, Gurney J, Moreland NJ, Bennett J, Oliver J, Williamson DA, et al. Risk factors for acute rheumatic fever: a case-control study. *The Lancet Regional Health — Western Pacific*. 2022;26:100508. https://doi.org/10.1016/j.lanwpc.2022.100508

2. Culliford-Semmens N, Tilton E, Wilson N, Stirling J, Doughty R, Gentles T, et al. Echocardiography for latent rheumatic heart disease in first degree relatives of children with acute rheumatic fever: implications for active case finding in family members. *EClinicalMedicine*. 2021;37:100935. https://doi.org/10.1016/j.eclinm.2021.100935

3. Vlajinac H, Adanja B, Marinković J, Jarebinski M. Influence of socioeconomic and other factors on rheumatic fever occurrence. *European Journal of Epidemiology*. 1991;7(6):702–704. https://doi.org/10.1007/bf00218687

4. Oliver J, Bennett J, Thomas S, Zhang J, Pierse N, Moreland NJ, et al. Preceding Group A streptococcus skin and throat infections are individually associated with acute rheumatic fever: evidence from New Zealand. *BMJ Global Health*. 2021;6(12). https://doi.org/10.1136/bmjgh-2021-007038

5. Dougherty S, Nascimento B, Carapetis J. Chapter 3 — Clinical evaluation and diagnosis of acute rheumatic fever. In: Dougherty S, Carapetis J, Zühlke L, Wilson N, editors. Acute rheumatic fever and rheumatic heart disease. San Diego: Elsevier; 2021. p. 31–54.

6. Carapetis JR, Beaton A, Cunningham MW, Guilherme L, Karthikeyan G, Mayosi BM, et al. Acute rheumatic fever and rheumatic heart disease. *Nature Reviews Disease Primers*. 2016;2:15084. https://doi.org/10.1038/nrdp.2015.84

7. Gewitz MH, Baltimore RS, Tani LY, Sable CA, Shulman ST, Carapetis J, et al. Revision of the Jones Criteria for the diagnosis of acute rheumatic fever in the era of Doppler echocardiography: a scientific statement from the American Heart Association. *Circulation*. 2015;131(20):1806–1818. https://doi.org/10.1161/cir.0000000000000205

8. Abernethy M, Bass N, Sharpe N, Grant C, Neutze J, Clarkson P, et al. Doppler echocardiography and the early diagnosis of carditis in acute rheumatic fever. *Australian and New Zealand Journal of Medicine*. 1994;24(5):530–535. https://doi.org/10.1111/j.1445-5994.1994.tb01753.x

9. Voss LM, Wilson NJ, Neutze JM, Whitlock RM, Ameratunga RV, Cairns LM, et al. Intravenous immunoglobulin in acute rheumatic fever: a randomized controlled trial. *Circulation*. 2001;103(3):401–406. https://doi.org/10.1161/01.cir.103.3.401

10. Rutstein DD, Bauer W, Dorfman A, Gross RE, Lichty JA, Taussig HB, et al. Jones criteria (modified) for guidance in the diagnosis of rheumatic fever; report of the Committee on Standards and Criteria for programs of care. *Circulation*. 1956;13(4):617–620. https://doi.org/10.1161/01.cir.13.4.617

11. Agnew J, Wilson N, Skinner J, Nicholson R. Beyond first-degree heart block in the diagnosis of acute rheumatic fever. *Cardiology in the Young*. 2019;29(6):744–748. https://doi.org/10.1017/s104795111900026x

12. New Zealand Heart Foundation. New Zealand guidelines for rheumatic fever: diagnosis, management and secondary prevention of acute rheumatic fever and rheumatic heart disease: 2014 update. Heart Foundation; 2014. https://www.heartfoundation.org.nz/resources/acute-rheumatic-fever-and-rheumatic-heart-disease-guideline (Accessed December 16 2024).

13. Lessof MH, Bywaters EG. The duration of chorea. *British Medical Journal*. 1956;1(4982):1520–1523. https://doi.org/10.1136/bmj.1.4982.1520

14. Feinstein AR, Wood HF, Spagnuolo M, Taranta A, Jonas S, Kleinberg E, et al. Rheumatic fever in children and adolescents. A long-term epidemiologic study of subsequent prophylaxis, streptococcal infections, and clinical sequelae. VII Cardiac changes and sequelae. *Annals of Internal Medicine*. 1964;60:87–123.

15. Steer AC, Kado J, Jenney AW, Batzloff M, Waqatakirewa L, Mulholland EK, et al. Acute rheumatic fever and rheumatic heart disease in Fiji: prospective surveillance, 2005-2007. *Medical Journal of Australia*. 2009;190(3):133–135. https://doi.org/10.5694/j.1326-5377.2009.tb02312.x

16. Güneş A, Akın A, Türe M, Balık H, Bilici M, Gül Ö. Evaluation of children with acute rheumatic fever: a single-center experience. *Turk Arch Pediatr*. 2022;57(1):26–31. https://doi.org/10.5152/TurkArchPediatr.2021.21064

17. Brewer EJ, Jr. New criteria for juvenile rheumatoid arthritis. *Texas Medicine*. 1973;69(2):84–92.

18. Carapetis JR, Brown A, Wilson NJ, Edwards KN. An Australian guideline for rheumatic fever and rheumatic heart disease: an abridged outline. *Medical Journal of Australia*. 2007;186(11):581–586. https://doi.org/10.5694/j.1326-5377.2007.tb01059.x

19. Mistry RM, Lennon D, Boyle MJ, Chivers K, Frampton C, Nicholson R, et al. Septic arthritis and acute rheumatic fever in children: the diagnostic value of serological inflammatory markers. *Journal of Pediatric Orthopedics*. 2015;35(3):318–322. https://doi.org/10.1097/bpo.0000000000000261

20. Mataika R, Carapetis JR, Kado J, Steer AC. Acute rheumatic fever: an important differential diagnosis of septic arthritis. *Journal of Tropical Pediatrics*. 2008;54(3):205–207. https://doi.org/10.1093/tropej/fmm116

21. Veasy LG. Myocardial dysfunction in active rheumatic carditis. *Journal of the American College of Cardiology*. 1994;24(2):581–582. https://doi.org/10.1016/0735-1097(94)90324-7

22. Caldas AM, Terreri MT, Moises VA, Silva CM, Len CA, Carvalho AC, et al. What is the true frequency of carditis in acute rheumatic fever? A prospective clinical and Doppler blind study of 56 children with up to 60 months of follow-up evaluation. *Pediatric Cardiology*. 2008;29(6):1048–1053. https://doi.org/10.1007/s00246-008-9242-z

23. Bhardwaj R, Sood A. Clinical profile of acute rheumatic fever patients in a tertiary care institute in present era. *Journal of the Association of Physicians of India*. 2015;63(4):22–24.

24. Mayosi B, Carapetis J. Part 11: Valvular heart disease. In: Fuster V, O'Rourke R, Walsh R, Poole-Wilson P, editors. Hurst's the heart. New York: McGraw-Hill Education; 2007.

25. Gentles TL, Colan SD, Wilson NJ, Biosa R, Neutze JM. Left ventricular mechanics during and after acute rheumatic fever: contractile dysfunction is closely related to valve regurgitation. *Journal of the American College of Cardiology*. 2001;37(1):201–207. https://doi.org/10.1016/s0735-1097(00)01058-5

26. Marcus RH, Sareli P, Pocock WA, Barlow JB. The spectrum of severe rheumatic mitral valve disease in a developing country. Correlations among clinical presentation, surgical pathologic findings, and hemodynamic sequelae. *Annals of Internal Medicine*. 1994;120(3):177–183. https://doi.org/10.7326/0003-4819-120-3-199402010-00001

27. Williams RV, Minich LL, Shaddy RE, Veasy LG, Tani LY. Evidence for lack of myocardial injury in children with acute rheumatic carditis. *Cardiology in the Young*. 2002;12(6):519–523. https://doi.org/10.1017/s104795110200094x

28. Alehan D, Ayabakan C, Hallioglu O. Role of serum cardiac troponin T in the diagnosis of acute rheumatic fever and rheumatic carditis. *Heart*. 2004;90(6):689–690. https://doi.org/10.1136/hrt.2003.026088

29. Wilson NJ VL, Neutze JM, Ameratunga RV, Lennon DR. The natural history of acute rheumatic fever to one year in the echocardiographic era. In: Imai Y, Momma K, editors. Proceedings of the Second World Congress of Paediatric Cardiology and Cardiac Surgery 1997. Oxford: Blackwell Publishing; 1998. p.971–972.

30. Grayson S, Horsburgh M, Lennon D. An Auckland regional audit of the nurse-led rheumatic fever secondary prophylaxis programme. *New Zealand Medical Journal*. 2006;119(1243):U2255.

31. Zalzstein E, Maor R, Zucker N, Katz A. Advanced atrioventricular conduction block in acute rheumatic fever. *Cardiology in the Young*. 2003;13(6):506–508.

32. Veasy LG, Tani LY, Hill HR. Persistence of acute rheumatic fever in the intermountain area of the United States. *Journal of Pediatrics*. 1994;124(1):9–16. https://doi.org/10.1016/s0022-3476(94)70247-0

33. Wikimedia. Image of second degree heart block. 2016. https://en.wikipedia.org/wiki/Atrioventricular_block (Accessed February 23 2025).

34. Dajani A, Ayoub E, Bierman F, Bisno A, Denny F, Durack D, et al. Guidelines for the diagnosis of rheumatic fever: Jones criteria, updated 1992: special writing group of the committee on rheumatic fever, endocarditis, and Kawasaki disease of the council on cardiovascular disease in the young, American Heart Association. *Circulation*. 1993;87(1):302.

35. Wilson NJ, Voss L, Morreau J, Stewart J, Lennon D. New Zealand guidelines for the diagnosis of acute rheumatic fever: small increase in the incidence of definite cases compared to the American Heart Association Jones criteria. *New Zealand Medical Journal*. 2013;126(1379):50–59.

36. Tilton E, Mitchelson B, Anderson A, Peat B, Jack S, Lund M, et al. Cohort profile: methodology and cohort characteristics of the Aotearoa New Zealand Rheumatic Heart Disease Registry. *BMJ Open*. 2022;12(12):e066232. https://doi.org/10.1136/bmjopen-2022-066232

37. Lennon D, Stewart J, Farrell E, Palmer A, Mason H. School-based prevention of acute rheumatic fever: a group randomized trial in New Zealand. *Pediatric Infectious Disease Journal*. 2009;28(9):787–794. https://doi.org/10.1097/INF.0b013e3181a282be

38. Markowitz M, Gordis L. Rheumatic fever. *Major Problems in Clinical Pediatrics*. 1972;11:1–309.

39. Lessof M. Sydenham's chorea. *Guy's Hospital Reports*. 1958;107(3):185–206.

40. Carapetis JR, Currie BJ. Mortality due to acute rheumatic fever and rheumatic heart disease in the Northern Territory: a preventable cause of death in aboriginal people. *Australian and New Zealand Journal of Public Health*. 1999;23(2):159–163. https://doi.org/10.1111/j.1467-842x.1999.tb01227.x

41. Taranta A, Stollerman GH. The relationship of Sydenham's chorea to infection with Group A streptococci. *American Journal of Medicine*. 1956;20(2):170–175. https://doi.org/10.1016/0002-9343(56)90186-3

42. Taranta A. Relation of isolated recurrences of Sydenham's chorea to preceding streptococcal infections. *New England Journal of Medicine*. 1959;260(24):1204–1210. https://doi.org/10.1056/nejm195906112602402

43. Ayoub EM, Wannamaker LW. Streptococcal antibody titers in Sydenham's chorea. *Pediatrics*. 1966;38(6):946–956.

44. Centers for Disease Control and Prevention. Acute rheumatic fever. *MMWR: Morbidity and Mortality Weekly Report*. 1986;36:108–110, 115.

45. Bland EF. Chorea as a manifestation of rheumatic fever: a long-term perspective. *Transactions of the American Clinical and Climatological Association*. 1961;73:209–213.

46. Sanyal SK, Berry AM, Duggal S, Hooja V, Ghosh S. Sequelae of the initial attack of acute rheumatic fever in children from north India. A prospective 5-year follow-up study. *Circulation*. 1982;65(2):375–379. https://doi.org/10.1161/01.cir.65.2.375

47. Orsini A, Foiadelli T, Sica A, Santangelo A, Carli N, Bonuccelli A, et al. Psychopathological impact in patients with history of rheumatic fever with or without Sydenham's chorea: a multicenter prospective study. *International Journal of Environmental Research and Public Health*. 2022;19(17). https://doi.org/10.3390/ijerph191710586

48. Carapetis JR, Currie BJ. Rheumatic fever in a high incidence population: the importance of monoarthritis and low grade fever. *Archives of Disease in Childhood*. 2001;85(3):223–227. https://doi.org/10.1136/adc.85.3.223

49. Sean TG, Sheshadri S, Saravu K. Emphysematous pyelonephritis. *Journal of the Association of Physicians of India*. 2002;50:1413–1415. https://doi.org/10.47739/2641-7766/1006

50. Hartley M. Image of erythema marginatum. DermNet; 2010. https://dermnetnz.org/topics/rheumatic-fever (Accessed February 22 2025).

51. RHDAustralia, Menzies School of Health Research. The 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (3.2 edition). 2022. https://www.rhdaustralia.org.au/arf-rhd-guidelines (Accessed December 16 2024).

52. Park M. Park's pediatric cardiology for practitioners. 6th ed. Philadelphia: Elsevier Saunders; 2014.

53. Perelini F, Agnew J, Skinner JR, Han DY, Nicholson R, Wilson N. Revisiting QT prolongation in acute rheumatic fever — relevance for hydroxychloroquine treatment. *International Journal of Cardiology*. 2022;362:93–96. https://doi.org/10.1016/j.ijcard.2022.05.053

54. Anderson Y, Wilson N, Nicholson R, Finucane K. Fulminant mitral regurgitation due to ruptured chordae tendinae in acute rheumatic fever. *Journal of Paediatrics and Child Health*. 2008;44(3):134–137. https://doi.org/10.1111/j.1440-1754.2007.01214.x

55. Nakauyaca AV, Ralph AP, Majoni WS, Kangaharan N. Case report: concurrent rheumatic fever and acute post-streptococcal glomerulonephritis in a high-burden setting. *American Journal of Tropical Medicine and Hygiene*. 2019;101(5):1054–1057. https://doi.org/10.4269/ajtmh.18-0954

56. Kaplan EL, Ferrieri P, Wannamaker LW. Comparison of the antibody response to streptococcal cellular and extracellular antigens in acute pharyngitis. *Journal of Pediatrics*. 1974;84(1):21–28. https://doi.org/10.1016/s0022-3476(74)80548-2

57. McCarthy M. The antibody response to streptococcal infections. In: McCarthy M, editor. Streptococcal infections. New York: Columbia University Press; 1954. p. 130–142.

58. Stollerman GH, Lewis AJ, Schultz I, Taranta A. Relationship of immune response to Group A streptococci to the course of acute, chronic and recurrent rheumatic fever. *American Journal of Medicine*. 1956;20(2):163–169. https://doi.org/10.1016/0002-9343(56)90185-1

59. Bennett J, Moreland NJ, Williamson DA, Carapetis J, Crane J, Whitcombe AL, et al. Comparison of Group A streptococcal titres in healthy children and those with pharyngitis and skin infections. *Journal of Infection*. 2022;84(1):24–30. https://doi.org/10.1016/j.jinf.2021.10.014

60. Pearce S, Bowen AC, Engel ME, de la Lande M, Barth DD. The incidence of sore throat and Group A streptococcal pharyngitis in children at high risk of developing acute rheumatic fever: a systematic review and meta-analysis. *PloS One*. 2020;15(11):e0242107. https://doi.org/10.1371/journal.pone.0242107

61. Gerber M. Pharyngitis: management in an era of declining rheumatic fever. In: Shulman S, editor. Diagnosis of pharyngitis: methodology of throat cultures. New York: Praeger; 1984. p. 61–72.

62. Taylor A, Morpeth S, Webb R, Taylor S. The utility of rapid Group A streptococcus molecular testing compared with throat culture for the diagnosis of Group A streptococcal pharyngitis in a high-incidence rheumatic fever population. *Journal of Clinical Microbiology*. 2021;59(12):e0097821. https://doi.org/10.1128/jcm.00978-21

63. Ralph A, Holt D, Islam S, Osowicki J, Carroll D, Tong S, et al. Potential for molecular testing for Group A streptococcus to improve diagnosis and management in a high-risk population: a prospective study. *Open Forum Infectious Diseases*. 2019;6(4):ofz097.

64. Barth DD, Cinanni G, Carapetis JR, Wyber R, Causer L, Watts C, et al. Roadmap to incorporating Group A streptococcus molecular point-of-care testing for remote Australia: a key activity to eliminate rheumatic heart disease. *Medical Journal of Australia*. 2022;217(6):279–282. https://doi.org/10.5694/mja2.51692

65. Upton A, Lowe C, Stewart J, Taylor S, Lennon D. In vitro comparison of four rapid antigen tests for Group A streptococcus detection. *New Zealand Medical Journal*. 2014;127(1398):77–83.

66. Upton A, Farrell E, Stewart J, Lennon D. Disappointing performance of rapid antigen detection tests for Group A streptococcus in the Auckland school-based sore throat programme. *New Zealand Medical Journal*. 2014;127(1389):103–105.

67. Dennison A, Peat B, Wilson E, Leversha A, Wheeler M, Briggs S, et al. Rheumatic fever recurrences in New Zealand 2010–14. *New Zealand Medical Journal*. 2020;133(1516):47–57.

68. Spinetto H, Lennon D, Horsburgh M. Rheumatic fever recurrence prevention: a nurse-led programme of 28-day penicillin in an area of high endemnicity. *Journal of Paediatrics and Child Health*. 2011;47(4):228–234. https://doi.org/10.1111/j.1440-1754.2010.01942.x

69. Oliver J, Robertson O, Zhang J, Marsters BL, Sika-Paotonu D, Jack S, et al. Ethnically disparate disease progression and outcomes among acute rheumatic fever patients in New Zealand, 1989–2015. *Emerging Infectious Diseases*. 2021;27(7):1893–1902. https://doi.org/10.3201/eid2707.203045

70. He VY, Condon JR, Ralph AP, Zhao Y, Roberts K, de Dassel JL, et al. Long-term outcomes from acute rheumatic fever and rheumatic heart disease: a data-linkage and survival analysis approach. *Circulation*. 2016;134(3):222–232. https://doi.org/10.1161/CIRCULATIONAHA.115.020966

71. World Health Organization. Rheumatic fever and rheumatic heart disease: report of WHO expert consultation. Geneva: World Health Organization; 2001. https://iris.who.int/bitstream/handle/10665/42898/WHO_TRS_923.pdf?sequence=1&isAllowed=y (Accessed February 22 2025).

72. Swedo SE, Leonard HL, Mittleman BB, Allen AJ, Rapoport JL, Dow SP, et al. Identification of children with pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections by a marker associated with rheumatic fever. *American Journal of Psychiatry*. 1997;154(1):110–112. https://doi.org/10.1176/ajp.154.1.110

73. Kurlan R, Kaplan EL. The pediatric autoimmune neuropsychiatric disorders associated with streptococcal infection (PANDAS) etiology for tics and obsessive-compulsive symptoms: hypothesis or entity? Practical considerations for the clinician. *Pediatrics*. 2004;113(4):883–886. https://doi.org/10.1542/peds.113.4.883

74. Bawazir Y, Towheed T, Anastassiades T. Post-streptococcal reactive arthritis. *Current Rheumatology Reviews*. 2020;16(1):2–8. https://doi.org/10.2174/1573397115666190808110337

75. De Cunto CL, Giannini EH, Fink CW, Brewer EJ, Person DA. Prognosis of children with poststreptococcal reactive arthritis. *Pediatric Infectious Disease Journal*. 1988;7(10):683–686. https://doi.org/10.1097/00006454-198810000-00002

76. Shulman ST, Ayoub EM. Poststreptococcal reactive arthritis. *Current Opinion in Rheumatology*. 2002;14(5):562–565. https://doi.org/10.1097/00002281-200209000-00014

77. Chun C, Kingsbury DJ. Poststreptococcal reactive arthritis: diagnostic challenges. *The Permanente Journal*. 2019;23. https://doi.org/10.7812/tpp/18.304

78. Zomorrodi A, Wald ER. Sydenham's chorea in western Pennsylvania. *Pediatrics*. 2006;117(4):e675–679. https://doi.org/10.1542/peds.2005-1573

79. Wilson NJ, Neutze JM. Echocardiographic diagnosis of subclinical carditis in acute rheumatic fever. *International Journal of Cardiology*. 1995;50(1):1–6. https://doi.org/10.1016/0167-5273(95)02325-q

80. Rémenyi B, Wilson N, Steer A, Ferreira B, Kado J, Kumar K, et al. World Heart Federation criteria for echocardiographic diagnosis of rheumatic heart disease--an evidence-based guideline. *Nature Reviews: Cardiology*. 2012;9(5):297–309. https://doi.org/10.1038/nrcardio.2012.7

81. Rwebembera J, Marangou J, Mwita JC, Mocumbi AO, Mota C, Okello E, et al. 2023 World Heart Federation guidelines for the echocardiographic diagnosis of rheumatic heart disease. *Nature Reviews: Cardiology*. 2024;21(4):250–263. https://doi.org/10.1038/s41569-023-00940-9

82. Pandian NG, Kim JK, Arias-Godinez JA, Marx GR, Michelena HI, Chander Mohan J, et al. Recommendations for the use of echocardiography in the evaluation of rheumatic heart disease: a report from the American Society of Echocardiography. *Journal of the American Society of Echocardiography*. 2023;36(1):3–28. https://doi.org/10.1016/j.echo.2022.10.009

83. Webb RH, Gentles TL, Stirling JW, Lee M, O'Donnell C, Wilson NJ. Valvular regurgitation using portable echocardiography in a healthy student population: implications for rheumatic heart disease screening. *Journal of the American Society of Echocardiography*. 2015;28(8):981–988. https://doi.org/10.1016/j.echo.2015.03.012

84. Reid CL, Anton-Culver H, Yunis C, Gardin JM. Prevalence and clinical correlates of isolated mitral, isolated aortic regurgitation, and both in adults aged 21 to 35 years (from the CARDIA study). *American Journal of Cardiology*. 2007;99(6):830–834. https://doi.org/10.1016/j.amjcard.2006.10.048

85. Vasan RS, Shrivastava S, Vijayakumar M, Narang R, Lister BC, Narula J. Echocardiographic evaluation of patients with acute rheumatic fever and rheumatic carditis. *Circulation*. 1996;94(1):73–82. https://doi.org/10.1161/01.cir.94.1.73

86. Ralph AP, Webb R, Moreland NJ, McGregor R, Bosco A, Broadhurst D, et al. Searching for a technology-driven acute rheumatic fever test: the START study protocol. *BMJ Open*. 2021;11(9):e053720. https://doi.org/10.1136/bmjopen-2021-053720

87. Gutman SJ, Costello BT, van Leeuwen MG, Wright LM, Varghese SE, O'Brien J, et al. Noninvasive identification of carditis in acute rheumatic fever. *JACC: Cardiovascular Imaging*. 2022;15(4):707–709. https://doi.org/10.1016/j.jcmg.2021.11.003

7

Initial Management of Acute Rheumatic Fever

In this chapter

Key changes

- Early discharge is now an endorsed option for medically stable patients with mild carditis, where close follow-up from community services and whānau support can be assured.
- Updated recommendations for clinical follow-up, including follow-up echocardiography (echo) after acute rheumatic fever (ARF), are outlined in **Table 7.5**.
- Updated recommendations about secondary antibiotic prophylaxis (SAP) duration are outlined in **Table 7.5**.
- Level of physical activity during initial ARF episode: Strict bed rest is no longer recommended for those with no or mild carditis. Mobilise as joint symptoms permit.
- Benzathine penicillin weight threshold is now 20kgs for the 1,200,000 unit dose.
- Household management: Routine swabbing of all household members is not recommended. Assess the household for those with symptomatic sore throats. Household members with a sore throat can be swabbed or treated empirically.

Key points

- The initial phase of managing ARF aims to alleviate symptoms — particularly joint pain, but also chorea or heart failure symptoms.
- Refer any person with suspected ARF to the hospital for initial assessment and investigation, including inpatient echo, regardless of clinical signs of carditis.
- Avoid giving nonsteroidal anti-inflammatory drugs (NSAIDs) until a diagnosis of ARF is confirmed. Give paracetamol instead.
- SAP should be started as part of initial management.
- Notify the regional Public Health Service of all suspected new cases — rheumatic fever is a notifiable condition in Aotearoa.
- Where the diagnosis of ARF is not clear initially, a person may require observation and investigation over several weeks, including a repeat echo.
- Early discharge is now an endorsed option for medically stable patients where close follow-up from community services and appropriate whānau support are available.
- Culturally safe care is essential.

Introduction

Hospital admission is recommended for all patients with suspected ARF. Initial hospital management has four main aims:

1. Timely assessment and investigation by clinicians experienced in ARF diagnosis, assuring access to investigations including ECG, serology, and echo (see **Chapter 6: Diagnosis of Acute Rheumatic Fever**).
2. Exclusion of alternative diagnoses.[1, 2]
3. Symptom management.
4. Care planning, including initiation of SAP, immunisations, dental care, and arranging follow-up.

Diagnosing ARF may be complex. Investigations over a period of several weeks may be required for people who do not initially meet the criteria for Definite ARF. A careful and detailed history is needed as symptoms may have already resolved before hospital admission (particularly joint symptoms, especially if NSAIDs have been given before hospital admission).

The initial clinical management of ARF focuses on symptom control and cardiac evaluation. Recommendations are largely based on expert clinical experience because very few randomised controlled trials have been carried out on managing ARF.

After diagnosis, the most important actions to reduce the risk of ARF recurrence and slow the progression of valvular disease are:

- Start the person on SAP.
- Refer them to a community-based Rheumatic Fever Secondary Prevention Service (benzathine penicillin programme) for ongoing care.

"..... 'Cause I think the paediatrician at one part kept going on to me about his weight and that and I kept trying to say to her, 'well, it's hard because, you know, he's not allowed to do anything.'"

Mother of tamaiti diagnosed with ARF

Table 7.1. Key priorities in the initial management of acute rheumatic fever

Step	Action
Admission to hospital	• Admit all patients with suspected ARF to facilitate investigations, particularly inpatient echo, clinical observation, and symptomatic management. • Ensure a multi-disciplinary team provides whānau-centred, culturally safe care in the hospital. The care should include cultural support, healthcare interpreters, play specialists, social workers, housing needs assessment, dietician and dental review, and other referrals as indicated. • Encourage early discharge for stable patients. 'Stable' means without progressive or severe carditis or severe chorea. Only consider discharge if whānau agrees and community services and supports are available.
Determine the diagnosis	• Take a careful history and physical examination. • Explain the investigations required to confirm a diagnosis of ARF to whānau. • Avoid NSAIDs until the diagnosis is confirmed — use paracetamol initially. • Exclude other diagnoses. If confirming an ARF diagnosis is not possible, discuss the need to re-evaluate over time. • Carry out the necessary investigations (see Table 7.3). • Consider whether the patient has Definite, Probable, or Possible ARF (note the initial classification may change with subsequent investigations). • Classify carditis as none, mild, moderate, or severe (according to echo). • Organise a whānau hui to discuss the diagnosis and management plan.
Notify the Public Health Service	• When the diagnosis is suspected, notify the appropriate regional or local Public Health Service using the local disease notification pathway.
Antibiotic treatment	• Give oral penicillin V or amoxicillin for 10 days or until the first dose of benzathine penicillin is given (see Chapter 9: Administration of Intramuscular Benzathine Penicillin (Bicillin®L-A)). • For those with penicillin anaphylaxis, give erythromycin instead. • Obtain consent for registration on the RFCCS from patient/caregiver. • Refer for SAP.
Arthritis	• Give paracetamol for analgesia until the diagnosis is confirmed. • When the diagnosis is confirmed, naproxen or ibuprofen should be given. • Mobilise the patient gently, as tolerated, unless they have severe carditis.
Sydenham's Chorea	• Explain the condition to the patient and whānau in detail. • Give medication (carbamazepine or valproate) if symptoms affect daily functioning (for example, eating, writing, walking). • Involve the play therapist, psychologist/psychiatrist, and occupational therapist in managing motor and/or neuropsychiatric manifestations. • Discuss severe and refractory cases with a paediatric neurologist.
Carditis	**Level of activity** — use severity of carditis as a guide. • No carditis, mild-moderate carditis — mobilise as joint symptoms permit. • Severe carditis — refer to a paediatrician with cardiology expertise. After initial bed rest, mobilise according to cardiac evolution. **Medication** — is usually only indicated for severe carditis. Be guided by a cardiologist or paediatrician/physician experienced in managing ARF/RHD. **Surgery for severe carditis** — the cardiosurgical team will consider whether surgery is needed and guide timing. The usual practice is to wait for inflammatory markers to normalise, but urgent surgery may be required for unstable patients.

First steps in managing acute rheumatic fever

This section outlines the first steps to take to manage ARF. Start by eradicating Strep A and treating joint pain. Carditis is common in ARF, and echo is important. Chorea is less common, but its symptoms can be debilitating.

Notify the Public Health Service of anyone with suspected acute rheumatic fever

ARF (whether Possible, Probable, or Definite) is a notifiable disease in Aotearoa and must be notified to the regional Public Health Service. Refer to the manual on communicable diseases and follow your local pathway for disease notification.

See 'Rheumatic fever' in the Communicable Disease Control Manual — Health New Zealand | Te Whatu Ora.

Eradicate Strep A and start secondary antibiotic prophylaxis

Penicillin is given in ARF to eradicate any persistent Strep A infection before starting SAP. Penicillin should be given regardless of pharyngeal culture status at doses recommended for treating Strep A pharyngitis (see **Table 7.4** on dosage).[3]

If the person has a history of rash or adverse reaction to penicillin, ask an allergy/immunology specialist if they may be suitable to trial oral penicillin in the hospital or require an alternative antibiotic (see **Table 7.4**). If IM benzathine penicillin is chosen for initial antibiotic therapy, this is considered the first dose of SAP (see **Chapter 9: Administration of Intramuscular Benzathine Penicillin (Bicillin®L-A)**). The first injection should be given before discharge. Prior to discharge, follow your local pathway to refer to the local Rheumatic Fever Secondary Prevention Service to support the delivery of SAP in the community. This referral must include the patient's diagnosis, diagnostic certainty (Definite, Probable, or Possible), echo findings, consent for registration on the Rheumatic Fever Care Coordination System (RFCCS), and the estimated duration of SAP. The secondary prophylaxis service will register the patient on the RFCCS.

See **Table 7.5** for guidance regarding clinical follow-up and anticipated duration of SAP at initial diagnosis. **Chapter 8: Secondary Prevention** provides guidance for ongoing SAP duration and conditions for ceasing SAP for use in non-acute and outpatient settings.

Treat symptoms of arthritis or polyarthralgia with NSAIDs

Joint pain in ARF is usually very sensitive to NSAIDs and may resolve symptoms rapidly. When the diagnosis is uncertain or not yet confirmed by an experienced ARF clinician, avoid NSAIDs, as these may mask the joint features. Use paracetamol instead as first-line therapy until a senior clinician has assessed the patient.[4]

NSAIDs are preferred to aspirin post-diagnosis based on their safety profile, and because they are as effective as aspirin.[5, 6] Aspirin is not recommended due to the risk of Reye's syndrome. Naproxen and ibuprofen are most commonly used in Aotearoa. Ibuprofen has not been extensively studied, specifically in ARF. However, a small retrospective non-randomised study in Turkey comparing aspirin and ibuprofen (30mg/kg/day) found that outcomes were equivalent, and ibuprofen had a superior safety profile.[7] Ibuprofen is available in suspension form, which many younger tamariki prefer.

In most patients, arthritis resolves within 10 days of regular NSAID therapy, but sometimes high and prolonged doses may be needed for up to 12 weeks in a minority of cases. Arthritis resolves by 12 weeks in 90% of cases.[2, 8]

As arthritis in ARF is usually very responsive to NSAIDS if joint symptoms do not reduce substantially within 72 hours of starting NSAIDs at the appropriate dosage, consider alternative diagnoses.

Joint symptoms may occasionally recur weeks later, but gradually reducing NSAIDs may prevent or reduce rebound. A rebound of joint symptoms does not indicate a recurrence of ARF.[9, 10] Occasionally, severe persisting arthritis in ARF may require increased analgesia and input from a rheumatologist, including consideration of alternate diagnoses.

Avoid Tramadol. It is contraindicated in tamariki under 12 years of age.[11]

Assess and manage any carditis

Perform echo in all patients with suspected ARF, regardless of clinical signs.[2, 12] In Aotearoa, around 70–80% of new ARF cases have carditis affecting the heart valves (also known as acute valvulitis). In mild and moderate cases, carditis is often subclinical (detected on echo, but the person does not have an audible cardiac murmur). Mitral regurgitation (MR) with or without aortic regurgitation (AR) is the most common form of valvulitis in ARF, followed by AR.[13, 14]

Level of activity

Most patients have mild carditis. These patients should rest only as needed to manage joint or other symptoms[3] (Grade D).

Managing conduction disturbances in acute rheumatic fever

First-degree heart block is common and needs no specific management. Less common are QT prolongation, advanced atrioventricular (AV) block, and junctional acceleration.[15, 16] A cardiologist should guide the management of advanced AV block/junctional acceleration, and cardiac monitoring/frequent ECGs may be required.[2, 17]

Managing pericardial effusion in acute carditis

Patients can occasionally present with pericardial effusion as part of acute carditis. Their management is individualised and should be guided by a paediatric or an adult cardiologist. A repeat echo will likely be required.

Medical management of heart failure associated with clinical carditis

Heart failure is uncommon and only occurs in severe carditis. Rest and diuretics usually improve heart failure. Although extended periods of bed rest were historically advised for such patients,[18] the current recommendation is to allow mobilisation as tolerated, guided more by echocardiographic evolution than the inflammatory markers.

Angiotensin-converting enzyme inhibitors (ACEi) can reduce afterload, mainly if AR is present (see **Table 7.4**). Digoxin is rarely used to manage atrial fibrillation (AF) under cardiology guidance.[19] Experience with beta-blockers in ARF is scarce. A recent Cochrane review of beta-blockers in tamariki with heart failure suggests they may be beneficial, but studies are small and heterogeneous.[20]

The role of corticosteroids in carditis has not been studied in large clinical trials. A recent meta-analysis found no benefit from corticosteroids or intravenous immunoglobulin (IVIg) over placebo at one year, but most studies were performed before echo was available.[21] Cardiologists experienced in ARF management sometimes use steroids in selected patients with severe carditis and intractably high inflammatory markers. IVIg does not appear to reduce carditis. Voss et al. conducted a randomised control trial in Aotearoa, which showed no impact of IVIg on echo parameters compared to placebo 12 months after diagnosis.[22] Clinical trials of novel therapies for ARF are urgently needed.

Valve surgery in severe carditis

Valve surgery is sometimes required for people with severe ARF and is usually delayed until after the acute inflammatory phase resolves[23] (see **Chapter 11: Management of Rheumatic Heart Disease**). However, in rare cases of fulminant carditis with severe regurgitation and mitral valve chordae tendineae rupture, urgent surgery may be needed.[24, 25]

Assess and manage chorea

Most people with rheumatic chorea have symptoms that resolve spontaneously within two to four months. In mild cases, supportive measures are enough — a calm environment, avoiding over-stimulation, rest, education, and reassurance about the condition.

However, some people with chorea have moderate or severe symptoms that impair day-to-day functioning, including the ability to eat, write, and walk steadily. Chorea may cause emotional lability and mood and behaviour disturbances. It can also fluctuate in severity and recur over two to three years, triggered by intercurrent illness or stress.[2]

Severely affected patients may require medication (see Table 7.2), but evidence to guide treatment is limited. In Aotearoa, carbamazepine is recommended as first-line treatment. It has a better safety profile than sodium valproate and appears as effective, but may be sedating.[26, 27] Sodium valproate is teratogenic and should be avoided in pregnancy and women of childbearing age. The response to anti-convulsants may take one to two weeks and may not fully eliminate all symptoms. Medication can be withdrawn gradually, two to four weeks after the chorea has subsided. A rating scale such as UFMG Sydenham's Chorea Rating Scale (USCRS) may help assess and measure response to therapy.[28]

Table 7.2. Managing chorea

Chorea severity	Management
Mild (no functional impairment)	Supportive.
Moderate (any functional impairment)	Carbamazepine as the first line treatment otherwise, sodium valproate.
Moderate to severe (significant functional impairment)	Prednisone or methylprednisolone; consider intravenous immunoglobulin if very severe, including chorea paralytica.

Neurologists occasionally use corticosteroids and other immunomodulatory agents (including IVIg) to treat severe chorea, but limited evidence supports this practice. A small placebo-controlled study showed significant reductions in symptom intensity and time to remission with prednisone but no change in relapse rate (the study failed to control for haloperidol use in both groups). A 2015 systematic review of IVIg[29] found only small non-randomised trials that showed possible short-term benefit.[30, 31] Robust clinical trials are needed. TREAT-SC is an Aotearoa-led randomised trial of short-course dexamethasone for chorea, recruiting from mid-2024.

Find out more about the trial on the Australian New Zealand Clinical Trials Registry.

Ask for neurology advice for severe or treatment-refractory chorea.

Monitoring clinical progress

Once the diagnosis is confirmed, monitor the erythrocyte sedimentation rate (ESR) and Creactive protein (CRP) weekly, then fortnightly until normal for one month.

Repeat echo may also be indicated in the following situations:

- The initial diagnosis is uncertain.
- Rheumatic pericardial effusion.
- Changes in clinical examination (murmurs, tachycardia/changing pulse pressure).
- Severe carditis, according to clinical course.

If the initial diagnosis is uncertain, repeat the echo in two to four weeks, as valvulitis can develop over several weeks. If individuals do not meet the criteria for Definite ARF and no clear alternate diagnosis exists, the categories of Possible ARF or Probable ARF can be used (see **Chapter 6: Diagnosis of Acute Rheumatic Fever**). Important considerations for these patients include:

- Whether they live in a high incidence area.
- Whether they are of Māori or Pacific ethnicity.
- Whether they have a family/whānau history of ARF or RHD.
- Whether they have been carefully evaluated for other possible diagnoses.

Dental assessment in acute rheumatic fever

Refer people with newly diagnosed ARF for a dental assessment, preferably while in hospital, and then for ongoing dental care by a community provider. Regular dental treatment may minimise the risk of infective endocarditis among people with RHD. ARF patients without carditis also require regular dental care as they are at risk of recurrent ARF. Those with RHD who require antibiotic prophylaxis for endocarditis should be given the New Zealand Heart Foundation's 'wallet card.' For people with ARF and RHD, ensure their dental service is aware of their RHD diagnosis. In Aotearoa, dental care is free for those under 18 years. Some districts also provide publicly funded dental services for adults with ARF or RHD.

Immunisations for people with acute rheumatic fever

- Check the person's current immunisation status on the Aotearoa Immunisation Register.
- Offer catch-up immunisations in the hospital.
- Recommend annual influenza immunisation. Note that influenza vaccine is only funded for patients with RHD. However, in some districts, Rheumatic Fever Secondary Prevention Services fund and offer annual influenza immunisation for all patients under their care.
- Refer to the Aotearoa Immunisation Handbook for guidance regarding other funded immunisations (including pneumococcal vaccine recommendations).

Holistic, culturally responsive care for acute rheumatic fever patients

Being hospitalised can be stressful, so providing the patient and whānau with the right support is essential. For whānau, the initial diagnosis and management of ARF is the start of a journey of living with ARF and potentially RHD. A multi-disciplinary team approach is recommended. Care should be tailored to the individual and their whānau and use available local services. Support should include:

- Cultural and spiritual support, using interpreters and Māori and Pacific health teams.
- Health information that is developmentally, linguistically, and culturally appropriate.
- Practical support for whānau during their hospital stay, including vouchers for food, parking, and petrol (via social work or cultural support teams).
- Social and housing needs assessments and referrals as relevant:
  - Healthy Homes Initiative (see Healthy Homes Initiative — Health New Zealand | Te Whatu Ora for local providers).
  - Whānau Ora services, MSD or WINZ Health and disability — Work and Income and Child Disability Allowance — Work and Income.
- Developmentally appropriate care, including play specialist or youth health assessment, and referral to health school if prolonged absence from school is anticipated.
- Facilitation of whānau hui and use of appropriate patient resources (see **Online resources for whānau with Strep A, acute rheumatic fever and rheumatic heart disease**).
- Peer or whānau support groups, if available. Offer to refer to Heart Kids Heart Kids New Zealand | Kids Charity.

Clinical management of household/whānau after acute rheumatic fever diagnosis

The diagnosis of ARF in tamariki and rangatahi impacts the entire whānau. Close relatives of those with ARF and RHD have an increased risk of chronic RHD (approximately 8–10%).[32-34]

ARF is an immune-mediated disorder and, as such, is not infectious. The Strep A throat infection that precedes ARF is infectious. It is therefore recommended to:

- Assess household members aged 3–35 years for sore throats (this may be undertaken by hospital teams, general practitioners (GP), or public health services, depending on local service arrangements).
- Household members with symptomatic sore throats should be managed per **Chapter 5: Primary Prevention of Acute Rheumatic Fever: Sore Throat Diagnosis and Management**. They should have a primary care assessment and throat swab, or if it is not possible to take a swab and follow-up the results, then empiric antibiotic treatment should be offered to household members with sore throats.
- Provide ongoing education for whānau and household members about the importance of timely recognition and treatment of sore throat, in particular Strep A sore throat.
- Support households in accessing free/low-cost local sore throat services, as available.

Previously, routine swabbing of all household members (including asymptomatic people, known as contact tracing) was undertaken in some regions of Aotearoa. In general, public health contact tracing aims to identify additional cases and prevent secondary cases via the provision of antibiotic prophylaxis.

The logic behind undertaking contact tracing for ARF, an immune-mediated, non-infectious condition, is uncertain, and secondary household cases remain extremely rare in Aotearoa. Any theoretical benefit needs to be considered against intensive public health resources required for contact tracing. Swabbing asymptomatic household members is therefore no longer recommended.

Unmet health needs may exist in whānau members. A holistic whānau health assessment is recommended. Healthcare providers should:

- Ask about whānau immunisation status (and link to immunisation providers as relevant).
- Ask about any other unmet health conditions and refer for treatment.
- Refer siblings of recently diagnosed ARF cases for echo and assessment (see **Chapter 14: Screening for Rheumatic Heart Disease**).[32, 33]

Summary: initial management of acute rheumatic fever

Table 7.3. Clinical observations and initial management of acute rheumatic fever

Observation	Initial management
Clinical observations	Check temperature, pulse, blood pressure, and respiratory rate. • If tachycardic, record an apical heart rate. • Perform an ECG if the pulse is rapid or irregular. Examine the patient daily for patterns of arthritis, presence of heart murmurs, skin rash, subcutaneous nodules, and choreiform movements. Consider the use of structured assessment for chorea, such as USCRS.[28]
Cardiac monitoring and management	Document cardiac symptoms and signs. **Fill out the weight and fluid balance chart** — initially daily, then reduce to twice weekly or as clinically indicated in moderate/severe carditis. **Normal echo at baseline** — repeat echo at 2–4 weeks. **Mild (subclinical) carditis** — repeat echo within 2 weeks–6 months of initial ARF diagnosis. The echo must be organised promptly if a changing murmur is identified during follow-up. Monitor ESR weekly. **Moderate carditis** — echo, pre-discharge, or as guided by cardiologist/experienced ARF physician/paediatrician. **Severe carditis** — provide individualised management guided by a cardiologist. May require an echo every one to two weeks while hospitalised if acute carditis is severe. Daily ECG or telemetry monitoring may be useful for conduction disturbances or arrhythmias. If the patient is in heart failure with severe carditis, avoid exertion until symptoms improve, as guided by a paediatrician, cardiologist, or other experienced ARF physician.
Diet and activity	Encourage a healthy diet. If the patient is overweight, give dietary advice and screen for obesity complications (including sleep apnoea and diabetes). Mobilise as tolerated, guided by cardiology advice in severe carditis.
Cultural support	Refer to cultural support teams (Pacific/Māori health) and local Pacific and Hauora Māori services as relevant.

Observation	Initial management
Age/ developmentally appropriate care	Get play specialist input to provide whānau support, procedural support (for injections and blood tests), and education on ARF (where they are trained and able to do this). Refer to hospital school and/or follow-up with the patient's usual school. Youth health services may be available in some regions.
Housing	Ask about housing concerns and refer to Healthy Homes Initiative, local iwi housing initiatives, MSD/Work and Income, or Kāinga Ora as needed.
Multi-disciplinary team (MDT)	Provide cultural support, social work, play specialist, income support, and child disability allowance.
Learning about ARF	See **Online resources for whānau with Strep A, acute rheumatic fever and rheumatic heart disease** for resources available.
Sibling/ whānau health assessment	Discuss sibling echo with whānau and refer according to local services as available. Ask if there is a family/whānau history of ARF or RHD.
Dental assessment	Carry out inpatient dental reviews. Explain the need for antibiotic prophylaxis for endocarditis for those with carditis/RHD to whānau and ensure the dentist is aware of the cardiac status. Tamariki with severe carditis who may need surgery should be assessed very promptly because they may need urgent dental treatment before valve surgery.
Immunisations	Check the immunisation register and offer catch-up immunisations in the hospital. The annual influenza vaccine is recommended for patients with ARF but is only funded for those with RHD and/or other eligible health conditions (e.g. asthma or diabetes). Proactively offer it to whānau/caregivers. The pneumococcal vaccine is also recommended for those with RHD. See the Immunisation Handbook for more details.

Table 7.4. Medication for acute rheumatic fever

Type of medication	Aotearoa Guidelines 2024	Tamariki (children)	Rangatahi (adolescents) and adults
Initial Strep A eradication	**Phenoxymethylpenicillin (Penicillin V)** **OR**	Weight ≤20kg 250mg two times daily for 10 days Weight ≥20kg 500mg two times daily for 10 days	500mg two times daily for 10 days
	Amoxicillin	50mg/kg once daily for 10 days (max 1000mg/day) OR 25mg/kg two times daily for 10 days (maximum 500mg per dose)	1000mg once daily for 10 days OR 500mg two times daily for 10 days
Secondary antibiotic prophylaxis (SAP)	**Benzathine penicillin** (see Chapter 9: Administration of Intramuscular Benzathine Penicillin (Bicillin®L-A))	Weight <20kg 600,000 units (450mg) IM Weight ≥20kg 1,200,000 units (900mg) IM	1,200,000 units (900mg) IM
Initial Strep A eradication or SAP, if allergy to beta-lactams	**Erythromycin**	20mg/kg two times daily for 10 days (maximum dose 1000mg/day)	
Initial analgesia	**Paracetamol**	Refer to New Zealand Formulary and New Zealand Formulary for Children for dosing	
Analgesia once a diagnosis is confirmed	**Naproxen (immediate release)** **OR**	10–20mg/kg/day PO two times daily (max of 1250mg/day) Taper dose as joint symptoms ease	
	Ibuprofen	Refer to New Zealand Formulary and New Zealand Formulary for Children for dosing	
Steroids	**Prednisone or prednisolone**	1–2mg/kg once daily (maximum 60mg/day) If used for more than 1 week, taper by 20–25% per week	
Cardiac drugs	Seek specialist cardiologist advice regarding diuretics and cardiac medications Refer to New Zealand Formulary and New Zealand Formulary for Children for dosing guidance		
	Digoxin, frusemide, spironolactone **ACEi for impaired left ventricular function (enalapril, lisinopril, captopril)** (choice depends on the clinical situation; monitor BP)		
Chorea medication	Seek specialist neurologist advice, especially for severe or refractory cases Refer to New Zealand Formulary and New Zealand Formulary for children for dosing		
	Carbamazepine or valproic acid		

Note: Weight-based dosing bands may be used in local guidelines.

The New Zealand Formulary (nzformulary.org) and New Zealand Formulary for Children — New Zealand Formulary for Children (nzfchildren.org.nz).

Managing recurrent acute rheumatic fever

Recurrent ARF usually occurs in the setting of late or missed SAP (suboptimal adherence or delivery) and occasionally occurs after the planned completion of SAP. In Aotearoa, recurrence is uncommon in tamariki and rangatahi, with 28% of recurrences occurring in those aged over 15 years. During 2010–2014, the recurrence rate was 4% in under 16 year-olds, 16% for 16–20 year-olds, and 25% for those over 20 years.

Reasons for recurrence may be due to health system errors, including:

- Discontinuation of SAP on medical recommendation (<10 years or stopped prior to 21 years of age) earlier than recommended in clinical guidelines.
- System errors in tracking benzathine penicillin delivery.
- Miscommunication between health service providers when a person moves region or to another provider.

People with recurrent ARF may not have received regular benzathine penicillin injections due to actively declining them. In this situation, a careful approach with cultural and multi-disciplinary input is needed to ensure the person understands their condition and the implications and risks of discontinuing SAP. Injection-related pain and barriers to accessing healthcare should be explored and addressed.

When a person is diagnosed with recurrent ARF, they are more likely to have worsening RHD. Additional measures are needed for individuals with recurrent ARF.

- Reduce SAP interval to 21 days/3 weeks (if the person was strictly adherent to 4-weekly) — seek specialist advice and extend the end date of SAP. The exact duration of SAP is to be decided by a specialist and informed by RHD severity.
- Talk to SAP providers about additional support to optimise adherence. Minimising injection-related pain and inconvenience to the person and whānau (see **Chapter 9: Administration of Intramuscular Benzathine Penicillin (Bicillin®L-A)**).
- Ensure the person is seen by a cardiologist in the hospital (for those with RHD).
- Provide education and culturally responsive care to ensure the person understands their condition and the rationale for treatment.

Patient, whānau and community factors affect the length of hospital stay

Hospital-based care for ARF diagnosis and management has been common practice since the mid-twentieth century, sometimes with prolonged stays for bed rest.

Most ARF patients without severe carditis can be discharged once the diagnosis is confirmed, management priorities are addressed, and discharge care planning is completed.

However, individuals and their whānau circumstances and available community healthcare services also influence the length of stay. If patients come from remote communities or experience other barriers in access to community healthcare or supervision at home for tamariki, prolonging the hospital stay may be advisable.

In selected medically stable patients, where resources are available, hospital-in-the-home (HITH) and comprehensive community nursing services may enable earlier discharge and whānau-centred, home-based care for ARF.

Involve the person, whānau, and local primary healthcare services early to ensure continuity of care. Include Māori or Pacific health providers where appropriate and possible.

Planning for discharge and follow-up

Planning ongoing care and follow-up with whānau is critical to optimise long-term outcomes. An agreed care plan (preferably one the patient owns/holds) should be provided before discharge, with a copy sent to the Rheumatic Fever Secondary Prophylaxis Service as part of the referral. Whānau should be given multiple, ongoing opportunities to ask questions and discuss concerns.

Most hospital services in high-incidence areas in Aotearoa have existing ARF inpatient or discharge checklists, or both, that outline the key steps in care and planning. Their key elements are discussed below.

Depending on local service configurations, the person should be reviewed within two weeks after discharge by a paediatrician/physician/specialist nurse. The purpose of the early post-discharge review is:

- To ensure the person remains clinically stable, for example, they have no new cardiac signs and joint symptoms remain controlled. Repeat blood tests may be indicated to check that the inflammatory markers are decreasing.
- To review understanding and check that community supports are in place.

Table 7.5 summarises recommendations for follow-up specialist, primary and dental care, echo, and SAP. The recommendations vary according to the severity of initial cardiac involvement and are informed by local experience and expert consensus. The recommendations in **Table 7.5** are intended to be used as a guide at the time of discharge from the hospital after an initial diagnosis of ARF.

Ongoing care recommendations (including SAP duration and conditions for ceasing SAP) can be found in **Chapter 8: Secondary Prevention**.

Table 7.5. Recommended clinical management and anticipated duration of secondary antibiotic prophylaxis following an initial episode of acute rheumatic fever: guidance at the time of discharge

Initial guidance regarding SAP duration and clinical management should always be reviewed depending on the person's situation and the progression of echo findings (see Chapter 8: Secondary Prevention). When inflammatory markers have resolved (typically within six months), any residual carditis is termed chronic RHD.

Severity	Clinical follow-up	Echo	Antibiotic prophylaxis for endocarditis	Penicillin prophylaxis (4-weekly)
Definite or Probable ARF with normal heart or mild carditis	Annual GP and dental review. Paediatric or physician follow-up 1–3 yearly (less often if stable).	2-yearly (less often if stable)	Recommended if any carditis	Minimum of 10 years or to age 21 years, whichever is longer. (Consider 5 years if >16 years at first episode and minimal carditis).
ARF with moderate carditis/RHD	Annual specialist, medical, and dental review. See Chapter 11: Management of Rheumatic Heart Disease.	Annual	Recommended — lifelong	Minimum of 10 years or until around age 21 years, whichever is longer, then reassess. If still moderate RHD or progresses to severe RHD, continue to age 30 years, then reassess.
ARF with severe carditis/RHD	6–12 monthly specialist, medical and dental review. See Chapter 11: Management of Rheumatic Heart Disease.	6–12 monthly	Recommended — lifelong	Reassess at 30 years. Beyond 30 years, individualised care is provided by the patient and physician. See Chapter 11: Management of Rheumatic Heart Disease.
Possible ARF	Specialist follow-up 6–12 months, then as needed if ARF/RHD is confirmed.	At 6 and 12 months	Not required	1 year and review. SAP may stop earlier if another diagnosis (e.g. juvenile arthritis) is confirmed. If a person from a high-risk population group has no other diagnosis confirmed, 5 years of SAP is recommended.

Checklist for planning discharge and ongoing care

Hold whānau hui before discharge, in the community, or at home.

Discuss the following with the whānau:

- Confirm Public Health notification is done (Possible, Probable, and Definite ARF are all notifiable).
- Consent for registration on the RFCCS and refer to Rheumatic Fever Secondary Prevention Services, if not done at diagnosis.
- Check and ensure the person is enrolled on the RFCCS. If not, contact your local RF Coordinator.
- Contact their GP and/or community hauora services to discuss the diagnosis and follow-up.

Provide a plain language discharge letter for the whānau, primary care team, and secondary prevention service, outlining the following:

- Diagnosis (Possible, Probable, or Definite ARF) and severity of carditis.
- Care plan based on severity of carditis.
- Date BPG given, the next due date and frequency (28 days or other), plus recommendations for administration based on experience of the first dose.
- Date of next medical or specialist follow-up and next echo.
- Anticipated duration of SAP.
- Information on any other care/advice provided, e.g. contraception, mental health, housing referrals, referral to community or private dental provider.
- Dental review date and whether endocarditis prophylaxis is indicated.
- Immunisations are given and indicate whether additional immunisations are indicated.

Engage and support the whānau by doing the following:

- Work with cultural liaison kaimahi and interpreters as required.
- Engage with whānau (and other caregivers as relevant) and introduce secondary prevention kaimahi (SAP provider) and other community kaimahi (community care worker) as required.
- Provide different ways of learning about ARF, RHD, sore throat management, and long-term care via a multi-disciplinary team (for example, peers, visual resources, learning kōrero) See **Online resources for whānau with Strep A, acute rheumatic fever and rheumatic heart disease**.
- Inform the person's school about their ARF/RHD and offer information to support their ongoing learning — only if the person and whānau agree.
- Ensure whānau knows who to contact for care and support and how (for example, contact numbers for secondary prevention service and primary care provider).
- Offer health assessment and echo for siblings (see **Chapter 14: Screening for Rheumatic Heart Disease**).

References

1. Ralph AP, Noonan S, Wade V, Currie BJ. The 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease. *Medical Journal of Australia*. 2021;214(5):220–227. https://doi.org/10.5694/mja2.50851

2. Dougherty S, Carapetis J, Zühlke L, Wilson N. Acute rheumatic fever and rheumatic heart disease. Amsterdam: Elsevier; 2020.

3. RHDAustralia, Menzies School of Health Research. The 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (3.2 edition). 2022. https://www.rhdaustralia.org.au/arf-rhd-guidelines (Accessed December 16 2024).

4. World Health Organization. Rheumatic fever and rheumatic heart disease: report of WHO expert consultation. Geneva: World Health Organization; 2001. https://iris.who.int/bitstream/handle/10665/42898/WHO_TRS_923.pdf?sequence=1&isAllowed=y (Accessed February 22 2025).

5. Hashkes PJ, Tauber T, Somekh E, Brik R, Barash J, Mukamel M, et al. Naproxen as an alternative to aspirin for the treatment of arthritis of rheumatic fever: a randomized trial. *Journal of Pediatrics*. 2003;143(3):399–401. https://doi.org/10.1067/s0022-3476(03)00388-3

6. Uziel Y, Hashkes PJ, Kassem E, Padeh S, Goldman R, Wolach B. The use of naproxen in the treatment of children with rheumatic fever. *Journal of Pediatrics*. 2000;137(2):269–271. https://doi.org/10.1067/mpd.2000.107158

7. Yılmaz M, Gürses D, Tükenmez G. The effectiveness and safety of ibuprofen and acetylsalicylic acid in acute rheumatic fever. *Pediatrics International*. 2022;64(1):e15133. https://doi.org/10.1111/ped.15133

8. New Zealand Heart Foundation. New Zealand guidelines for rheumatic fever: diagnosis, management and secondary prevention of acute rheumatic fever and rheumatic heart disease: 2014 update. Heart Foundation; 2014. https://www.heartfoundation.org.nz/resources/acute-rheumatic-fever-and-rheumatic-heart-disease-guideline (Accessed December 16 2024).

9. Osowicki J, Carr JP, Steer AC. Rheumatic fever: the rebound phenomenon returns. *Journal of Paediatrics and Child Health*. 2018;54(6):685–688. https://doi.org/10.1111/jpc.13848

10. Holt KS. The rebound phenomenon in acute rheumatic fever. *Archives of Disease in Childhood*. 1956;31(160):444–451. https://doi.org/10.1136/adc.31.160.444

11. MedSafe. Prescriber update ;41(2): 25–26. MedSafe; 2020. https://www.medsafe.govt.nz/profs/PUArticles/PDF/Prescriber_Update_Vol_%2041_%20No_2_June_2020.pdf (Accessed February 24 2025).

12. New Zealand Heart Foundation. New Zealand guidelines for rheumatic fever: diagnosis, management and secondary prevention of acute rheumatic fever and rheumatic heart disease: 2014 update. Heart Foundation; 2014. https://www.heartfoundation.org.nz/resources/acute-rheumatic-fever-and-rheumatic-heart-disease-guideline

13. Carapetis JR, Beaton A, Cunningham MW, Guilherme L, Karthikeyan G, Mayosi BM, et al. Acute rheumatic fever and rheumatic heart disease. *Nature Reviews Disease Primers*. 2016;2:15084. https://doi.org/10.1038/nrdp.2015.84

14. Reményi B, Carapetis J. Acute rheumatic fever and chronic rheumatic disease. In: Da Cruz E, Jaggers ID, editors. Pediatric and congenital cardiology, cardiac surgery and intensive care. London: Springer; 2014. p. 2329–2350.

15. Agnew J, Wilson N, Skinner J, Nicholson R. Beyond first-degree heart block in the diagnosis of acute rheumatic fever. *Cardiology in the Young*. 2019;29(6):744–748. https://doi.org/10.1017/s104795111900026x

16. Perelini F, Agnew J, Skinner JR, Han DY, Nicholson R, Wilson N. Revisiting QT prolongation in acute rheumatic fever - relevance for hydroxychloroquine treatment. *International Journal of Cardiology*. 2022;362:93–96. https://doi.org/10.1016/j.ijcard.2022.05.053

17. Agnew J, Nicholson R, Wilson N, Skinner J. Advanced AV conduction abnormality in acute rheumatic fever. *Heart, Lung and Circulation*. 2016;25. https://doi.org/10.1016/j.hlc.2016.06.718

18. Taran LM. The treatment of acute rheumatic fever and acute rheumatic heart disease in children. *Medical Clinics of North America*. 1947;31:557–580. https://doi.org/10.1016/s0025-7125(16)35812-6

19. Karthikeyan G, Devasenapathy N, Zühlke L, Engel ME, Rangarajan S, Teo KK, et al. Digoxin and clinical outcomes in the Global Rheumatic Heart Disease Registry. *Heart*. 2019;105(5):363–369. https://doi.org/10.1136/heartjnl-2018-313614

20. Alabed S, Sabouni A, Al Dakhoul S, Bdaiwi Y. Beta-blockers for congestive heart failure in children. *Cochrane Database of Systematic Reviews*. 2020;7(7):Cd007037. https://doi.org/10.1002/14651858.CD007037.pub4

21. Cilliers A, Adler AJ, Saloojee H. Anti-inflammatory treatment for carditis in acute rheumatic fever. *Cochrane Database of Systematic Reviews*. 2015(5):Cd003176. https://doi.org/10.1002/14651858.CD003176.pub3

22. Voss LM, Wilson NJ, Neutze JM, Whitlock RM, Ameratunga RV, Cairns LM, et al. Intravenous immunoglobulin in acute rheumatic fever: a randomized controlled trial. *Circulation*. 2001;103(3):401–406. https://doi.org/10.1161/01.cir.103.3.401

23. Finucane K, Wilson N. Priorities in cardiac surgery for rheumatic heart disease. *Global Heart*. 2013;8(3):213–220. https://doi.org/10.1016/j.gheart.2013.08.010

24. al Kasab S, al Fagih MR, Shahid M, Habbab M, al Zaibag M. Valve surgery in acute rheumatic heart disease. One- to four-year follow-up. *Chest*. 1988;94(4):830–833. https://doi.org/10.1378/chest.94.4.830

25. Strauss AW, Goldring D, Kissane J, Hernandez A, Hartmann AF, McKnight CR, et al. Valve replacement in acute rheumatic heart disease. *Journal of Thoracic and Cardiovascular Surgery*. 1974;67(4):659–670.

26. Genel F, Arslanoglu S, Uran N, Saylan B. Sydenham's chorea: clinical findings and comparison of the efficacies of sodium valproate and carbamazepine regimens. *Brain and Development*. 2002;24(2):73–76. https://doi.org/10.1016/s0387-7604(01)00404-1

27. Tariq S, Niaz F, Waseem S, Shaikh TG, Ahmed SH, Irfan M, et al. Managing and treating Sydenham chorea: a systematic review. *Brain and Behavior*. 2023;13(6):e3035. https://doi.org/10.1002/brb3.3035

28. Teixeira AL, Jr., Maia DP, Cardoso F. UFMG Sydenham's chorea rating scale (USCRS): reliability and consistency. *Movement Disorders*. 2005;20(5):585–591. https://doi.org/10.1002/mds.20377

29. Mohammad SS, Nosadini M, Grattan-Smith P, Dale RC. Intravenous immunoglobulin in acute Sydenham's chorea: a systematic review. *Journal of Paediatrics and Child Health*. 2015;51(12):1235–1238. https://doi.org/10.1111/jpc.12915

30. Walker K, Brink A, Lawrenson J, Mathiassen W, Wilmshurst JM. Treatment of Sydenham chorea with intravenous immunoglobulin. *Journal of Child Neurology*. 2012;27(2):147–155. https://doi.org/10.1177/0883073811414058

31. Garvey MA, Snider LA, Leitman SF, Werden R, Swedo SE. Treatment of Sydenham's chorea with intravenous immunoglobulin, plasma exchange, or prednisone. *Journal of Child Neurology*. 2005;20(5):424–429. https://doi.org/10.1177/08830738050200050601

32. Culliford-Semmens N, Tilton E, Wilson N, Stirling J, Doughty R, Gentles T, et al. Echocardiography for latent rheumatic heart disease in first degree relatives of children with acute rheumatic fever: implications for active case finding in family members. *EClinicalMedicine*. 2021;37:100935. https://doi.org/10.1016/j.eclinm.2021.100935

33. Aliku T, Sable C, Scheel A, Tompsett A, Lwabi P, Okello E, et al. Targeted echocardiographic screening for latent rheumatic heart disease in northern Uganda: evaluating familial risk following identification of an index case. *PLoS Neglected Tropical Diseases*. 2016;10(6):e0004727. https://doi.org/10.1371/journal.pntd.0004727

34. Baker MG, Gurney J, Moreland NJ, Bennett J, Oliver J, Williamson DA, et al. Risk factors for acute rheumatic fever: a case-control study. *The Lancet Regional Health — Western Pacific*. 2022;26:100508. https://doi.org/10.1016/j.lanwpc.2022.100508

8

Secondary Prevention

In this chapter

Key changes

- All people weighing ≥20kgs should receive the full dose of 1,200,000 units (900 mg) of benzathine penicillin.
- Revised recommendations regarding the duration of secondary antibiotic prophylaxis (SAP), including recommendations for people with rheumatic heart disease (RHD), are included.
- The National Rheumatic Fever Care Coordination System (RFCCS), which functions to coordinate secondary prevention and as a national register, is launched.
- Resources to support whānau and health kaimahi, co-designed by whānau with lived experience of ARF and RHD, are provided (see **Appendix 1** on managing ARF).

Key points

- Health services for secondary prevention of acute rheumatic fever (ARF) and RHD need to be culturally safe, provide a holistic approach and recognise the long-term nature of the condition.
- A comprehensive approach is needed, including clinical care, promoting health and wellbeing, ensuring warm, dry housing, and social and income support, as relevant.
- SAP in Aotearoa is best delivered in the community by trained nurses who are resourced to engage with whānau and coordinate care and support.
- Intramuscular (IM) benzathine penicillin is recommended every 28 days (4 weeks) unless the person has experienced a recurrent episode of ARF. In Aotearoa, recurrences are rare in those who fully adhere to a 28-day regimen.
- Following ARF, SAP is usually recommended for 10 years after ARF diagnosis or until age 21, whichever is longer. The duration of SAP should be reassessed when the person is around 21 years old according to RHD severity and is extended for people with persisting moderate or severe RHD.
- New methods of SAP, such as subcutaneous benzathine penicillin, may offer an alternative to the traditional IM route in the future. However, before implementation, further evaluation of safety, effectiveness, and health system readiness is needed.
- The development of the RFCCS and the completion of roll-out in 2025 supports secondary prevention services to coordinate the delivery and monitoring of care plans for those on SAP. It is a milestone towards improving secondary prevention.
- Clinical governance and oversight for the RFCCS will be developed and confirmed by early 2026.

Introduction

Secondary prevention is about promoting wellbeing and heart health as well as preventing the recurrence of ARF and the progression of RHD. A high-quality secondary prevention programme for ARF and RHD needs to:

- Provide culturally responsive clinical care and SAP.
- Respond to the strengths, contexts, and aspirations of whānau.

Aotearoa has some responsive secondary prevention models, but variations and inequities in resourcing and service models result in differing experiences and quality of service for people with ARF and RHD.

SAP is the cornerstone of secondary prevention: it prevents recurrences of ARF and slows heart valve damage. The most effective form of SAP is benzathine penicillin intra-muscularly every 28 days/4-weekly. Accepting painful injections every 28 days requires commitment. Frequent appointments pose considerable personal costs to people with ARF and RHD and their whānau (time off work, school, travel, discomfort). It is imperative that the benefits, risks, and rationale for the lengthy duration of SAP are well understood and supported as much as possible by SAP services.

Understanding the impact of adolescence, pregnancy, and other life course events is also critical in determining secondary prevention approaches and optimising adherence to SAP. As in managing other long-term/chronic conditions, a strong focus in ARF and RHD care should be to empower whānau to understand the condition and build the capacity to self-manage. (See the section **'Supporting adherence to SAP'**.)

Key elements of secondary prevention

A quality secondary prevention programme in Aotearoa should consist of the following:

- A robust national register and local coordinators to support the delivery and monitoring of SAP and related ARF and RHD care and epidemiological monitoring.
- Coordination and collaboration between hospital, community, and primary care services (particularly hauora Māori and Pacific health providers).
- Timely and equitable access to specialist follow-up and echocardiography (echo) services. Where possible, care should be provided in multidisciplinary teams, which include echo, cardiology, paediatric or adult medicine, and cultural support workers.
- Oral healthcare services, including provision of antibiotics for endocarditis prophylaxis.
- Pathways to support the transition from paediatric to rangatahi and adult services.[1, 2]
- Access to culturally responsive primary care services for timely management of Strep A throat infections, immunisations, and reproductive and mental healthcare.
- Dedicated community-based staff who deliver SAP and are adequately resourced to provide culturally safe care, benzathine penicillin injections at the location of the person's choice, and help whānau navigate other services as required.[3-5]
- Culturally and developmentally appropriate resources to support whānau understanding of ARF and RHD.
- Pathways and processes to support transfer or shared care arrangements for people who spend time in different districts or who move homes.
- Immunisations including influenza and pneumococcal conjugate vaccine.

Evidence for secondary antibiotic prophylaxis

The natural history of progression from ARF to RHD is not completely understood.[9] Disease regression related to prophylaxis cannot be easily differentiated from spontaneous regression of RHD, and few randomised controlled trials have studied this. Evidence to support using antibiotics to prevent recurrent ARF is drawn from:

- Studies of the natural history and progression of ARF in the pre-penicillin era.[10-13]
- Systematic reviews of small studies (primarily carried out in the 1950s–1960s).[14, 15]
- Contemporary evidence from SAP programme audits.[16-18]
- A World Health Organization report.[86]

Studies of penicillin prophylaxis after acute rheumatic fever

In a 1973 Indian study, penicillin (oral or IM benzathine penicillin) reduced the risk of ARF recurrence by 55% compared to placebo control (RR 0.45; 95%; CI: 0.22–0.92).[19] In two earlier studies of penicillin versus placebo from the 1950s–1960s, the trend was the same, but it was not statistically significant.[14] Other studies from the period support a positive effect.[20-22] Further studies from the 1960s showed IM benzathine penicillin was superior to oral penicillin in reducing recurrence risk, with four studies showing statistically significant reductions in ARF recurrences.[14] In a non-randomised study in Te Tai Rāwhiti (1984), IM benzathine penicillin was superior to oral Penicillin V. The benzathine penicillin group had no recurrences, compared to recurrences in 35% of the group on oral penicillin.[23]

Acute rheumatic fever recurrences in Aotearoa and secondary prophylaxis adherence

Most recurrences in Aotearoa are seen in rangatahi and young adults who do not strictly adhere to benzathine penicillin or have become disengaged.[8] ARF recurrence carries increased risk, especially for patients with pre-existing moderate or severe RHD. Historically, the risk of ARF recurrence was noted to be highest during the first year after the initial episode of ARF and to decrease as time passed.[8, 24, 67-69] A recent clinical audit of recurrences between 2010 and 2014 in Aotearoa showed very low recurrence rates in tamariki and rangatahi.

In an Auckland study of people diagnosed with ARF from 1993–1999[7], 360 cases met the case definition for ARF. Nineteen people (median age 21 years) had 20 recurrences. Most recurrences (72%) occurred within 5 years of the initial ARF diagnosis and 12% between 5 and 10 years. The four-weekly long-acting penicillin failure rate (n = 1) was 0.07/100 patient-years. The programme failure rate for Auckland residents was 1.4/100 patient-years (n = 20). Patient non-adherence accounted for 55% of recurrences. Dennison et al. [8] found that the ARF recurrence rate for 2010–2014 in Aotearoa was 7.2% using a detailed chart review to confirm the accuracy of cases coded as recurrences. Median age at recurrence was 21.6 years (8–42 years), and 83% of patients experiencing recurrence were over 15 years of age. At the time of recurrence, 65% were non-adherent to penicillin prophylaxis. Eleven patients (17%) had stopped SAP on medical recommendation. The median interval from ARF diagnosis until recurrence was 10.5 years (7 months to 29.1 years).

Contemporary evidence from audits

Internationally, programme audits and retrospective analyses of ARF and RHD cohorts consistently show a reduced risk of ARF recurrence in those with high adherence to a 28-day penicillin regimen and timely injection delivery compared with those with lower adherence.[12, 24-29] Regional programmes in Aotearoa have reported very good adherence,[7, 30-32] although local data on associated progression or regression to/from RHD is limited.[33, 34] More recent evidence on the natural history of RHD and the effectiveness of SAP is largely drawn from echo screening studies in asymptomatic tamariki with Stage A & B RHD (previously labelled echo-detected "borderline" and definite RHD).[35-37]

Secondary antibiotic prophylaxis: benzathine penicillin dose and frequency

This section discusses IM benzathine penicillin doses and administration, and oral penicillin. For more details, see **Chapter 9: Administration of Intramuscular Benzathine Penicillin (Bicillin®L-A)**.

Recommended dosing

The standard dose of benzathine penicillin for the secondary prevention of ARF in adults and tamariki weighing 20kgs or more is 1,200,000 international units, equivalent to 900 mg (see **Table 8.1**).[12, 16, 28] The cut-off weight for tamariki varies internationally. In Aotearoa, the recommended dose for those weighing less than 20kg is 600,000 international units (450 mg) (Grade D).

Table 8.1. Recommendations for benzathine penicillin and lignocaine dosing for secondary prophylaxis

Benzathine penicillin IM dose (units)	Lignocaine 2%	Interval*
<20kg 600,000 units (450 mg)	0.25 mL	28 days
≥20kg 1,200,000 units (900 mg)	0.25 mL	28 days

* 21 days (3-weekly) if ARF recurrence occurs despite full adherence to a 4-weekly regimen.

Frequency of benzathine penicillin injections

Benzathine penicillin is recommended every 28 days (4 weeks) in Aotearoa. Recurrence is rare in those who adhere fully to a 28-day regimen.[7, 30, 38] A 21-day regimen (3 weeks) may be recommended after specialist review for people with recurrent ARF episodes despite good adherence. Administration of benzathine penicillin several days (up to 2 weeks) early is reasonable and preferable to late delivery. The patient should not be changed back to a 28-day regimen unless explicitly advised by the patient's specialist physician/paediatrician.

Administering benzathine penicillin injections

In 2024, Bicillin®L-A (Pfizer) is the available benzathine penicillin formulation in Aotearoa. It comes as a pre-formulated thick white suspension in pre-filled syringes. Most other countries use dry powder formulations reconstituted with sterile water.

The first benzathine penicillin dose, especially in tamariki, should usually be given in the hospital, with appropriate play specialist support. However, for adults without a documented history of penicillin allergy, a first dose may be given in the community by a nurse experienced in administering benzathine penicillin.

Subsequently, people on SAP should be offered the option to receive injections at a location of their choice (for example, clinic, home, work, or school). Recalls and reminders to the patient (for example, by text messaging) minimise the risk of late delivery of injections.

Benzathine penicillin injections can cause pain and discomfort. Administration with 2% lignocaine and distraction techniques is recommended. Lignocaine reduces pain without affecting serum penicillin levels.[39] The Buzzy®, a vibrating device with a cold pack used in conjunction with lignocaine, also reduces injection-related pain.[40] See **Chapter 9: Administration of Intramuscular Benzathine Penicillin (Bicillin®L-A)** for more details.

Research on benzathine penicillin pharmacokinetics and formulation

Recent pharmacokinetic studies have raised questions about conventional benzathine penicillin dosing and the required penicillin level to prevent Strep A infections and ARF recurrences. These studies showed most people did not have concentrations of penicillin in plasma above the traditionally accepted pharmacological correlate of protection (>0.02 mg/L) beyond 10–14 days post-benzathine penicillin administration.[41-43] However, there was no increase in Strep A infections, suggesting that a lower target concentration may be effective.

Preliminary results from a human challenge model study (the CHIPS trial[44]) suggest the minimum protective penicillin concentration to prevent Strep A sore throat may be as low as 0.006–0.009 mg/L (less than half the current accepted cut-off).

Recent phase I and II studies assessing subcutaneous delivery of benzathine penicillin show promising tolerability and a good safety profile.[45-48] In a non-randomised phase II study, most participants preferred subcutaneous delivery to IM benzathine penicillin.[47] A robust prospective randomised control trial of the effectiveness of subcutaneous benzathine penicillin versus IM benzathine penicillin for preventing ARF recurrences is yet to be carried out. The benzathine penicillin dose used for subcutaneous administration is significantly higher than for IM benzathine penicillin, even once fewer doses are accounted for, which may have implications for managing benzathine penicillin supply within ARF control programmes. Additionally, pre-formulated Bicillin®L-A is not currently licensed to be given by the subcutaneous route and every dose must be individually prescribed under Section 29 Regulations — i.e. cannot be given under Standing Orders, which are used for most patients on SAP in Aotearoa New Zealand.

Oral penicillin for secondary prophylaxis is not generally recommended

Oral penicillin should only be used if benzathine penicillin injections cannot be given (e.g. severe needle phobia, anaphylaxis to penicillin or the components of Bicillin®L-A),

and after detailed counselling of the patient and whānau. Oral penicillin is less effective than benzathine penicillin at preventing recurrent ARF.[16, 49, 50] Oral penicillin results in less predictable serum penicillin concentrations, and food affects its absorption. Adherence to twice-daily medication is difficult for most people to maintain over many years.[51]

Those on oral SAP should be registered on the RFCCS. SAP services should closely support people with ARF and RHD on oral penicillin and all other services should be made available — e.g. immunisations and dental care. This may include support with repeat prescriptions, ongoing health education and ensuring that patients are regularly seen by their GP or hospital specialist. People on oral SAP without a history of anaphylaxis should be periodically reviewed by a hospital specialist to consider whether they could restart IM benzathine penicillin injections.

Table 8.2. Oral alternatives for secondary prophylaxis

Medication	Dosing	Notes
Penicillin V	<20kg 250 mg two times daily >20kg 500 mg two times daily	Oral penicillin is only recommended for short-term situations (for example, if IM benzathine penicillin is unavailable, on holiday or is out of stock).
Erythromycin ethyl succinate	20 mg/kg two times daily Max adult dose 1.6g daily	For documented penicillin allergy. Refer to allergy/immunology specialist to consider drug testing and penicillin challenge.

Managing a person on secondary prophylaxis who reacts to benzathine penicillin

Severe reactions following benzathine penicillin injections are very rare and are most likely to occur within 15 minutes of administration. For more details on how to recognise and respond to a possible benzathine penicillin reaction, see **Chapter 9: Administration of Intramuscular Benzathine Penicillin (Bicillin®L-A)**. If a person on SAP has an adverse reaction to benzathine penicillin:

- Verify the type and severity of the response.
- Notify the patient's GP and ARF team doctor.
- Refer the patient to a specialist allergist or immunologist.
- Start oral erythromycin until a specialist allergist or immunologist evaluates the person.
- Report adverse events to the Centre for Adverse Reactions Monitoring (CARM).
  Report a Problem (Medsafe.govt.nz)
  Adverse Reaction Reporting (Medsafe.govt.nz)

Skin tests or supervised penicillin challenge may be appropriate under the guidance of a specialist allergist or immunologist.[52]

Most people with labelled penicillin allergy tolerate penicillin antibiotics.[53, 54] Penicillin allergy labels are associated with suboptimal treatment of infections and worse clinical outcomes in a variety of conditions (including RHD and SAP with oral non-penicillin antibiotics).[53, 55, 56]

A very small number of people have reactions to excipients in Bicillin®L-A (Pfizer). Other dry powder formulations may be considered in these rare cases (for example, Tardocillin® and Lentocillin®). Seek specialist hospital pharmacist and clinical immunology advice if considering whether a patient should trial an alternative formulation of BPG.

Anaphylaxis has rarely been reported in patients who have previously tolerated the injection for months and years without incident. While hypersensitivity reactions to penicillin (such as rashes and hives) are relatively common,[53] anaphylaxis to benzathine penicillin is rare. The prevalence of severe (anaphylactic) type I reaction to penicillin is estimated at approximately 0.001% for parenteral exposures and 0.0005% for oral exposures.[53] In a 1991 prospective cohort study of people receiving benzathine penicillin for ARF and RHD:

- 57 of the 1,790 patients had an allergic reaction (3.2%).
- 4 had anaphylaxis (0.2% or 1.2 per 10 000 injections), with a fatality incidence of 0.05% (0.31/10 000 injections).[57]

Communication strategies for healthcare workers who deliver secondary prophylaxis

Culturally safe and respectful communication is essential for all healthcare workers involved in SAP services. Unless care is taken, the language used about ARF and RHD may perpetuate negative stereotypes and stigma. Healthcare workers may perceive low adherence as a patient deficit rather than a health system deficit. Service-wide strategies to improve communication, engagement, and whānau experience should be informed by those with lived experience of ARF and RHD and may include:

- Embedding tikanga and appropriate cultural processes into clinical services.[58-63]
- Changing service models to better meet the contexts of whānau (especially rangatahi).
- Provision of regular cultural safety and anti-racism training for staff.
- Co-design of resources for whānau.[4, 63]

Healthcare workers should take a strengths-based approach and use the hui process when communicating with whānau, see Chapter 1: Cultural Responsiveness.[5] The hui process has four key steps:

- **Mihi:** Initial greeting and engagement.
- **Whakawhanaungatanga:** Making a connection.
- **Kaupapa:** Attending to the main purpose of the encounter.
- **Poroporoaki:** Concluding the encounter.

Then, take these practical steps in an ongoing process:

- First, ask 'What is important to you?'
- Establish priorities and specific information needs for each whānau.
- Recognise that repeated opportunities are important for learning.

Several discussions over time and reassessing understanding are required. When discussing ARF and RHD, covering the following issues is important:

- Cause and complications of ARF and RHD.
- Reason for SAP.
- Sore throat management (for the person with ARF as well as whānau).
- Signs and symptoms of recurrent ARF.
- Importance of medical, echo, and oral health/dental follow-up.
- How to contact relevant people or agencies for further information or assistance.

Supporting adherence to secondary prophylaxis

In Aotearoa, there is a range of barriers to maintaining engagement with a SAP provider and, therefore, timely delivery of SAP:

- Complex living circumstances.
- Whānau mobility (due to unstable housing, employment, and financial pressures).
- Living rurally or remotely.
- Changes in schooling.
- Life events (for example, rangatahi transition to adult services).
- Culturally unsafe health services or negative experiences, including personal racism.
- Unmanaged pain and distress from injections.

The impact of adolescence, pregnancy, and other life events is critical in determining secondary prevention approaches and optimising adherence to SAP. Developmental and life changes over time need to be anticipated. For example, adolescence may be associated with rangatahi becoming more autonomous, facing new life challenges, and engaging in risk-taking activities. A strong focus in ARF and RHD care should be on building engagement with healthcare services. Enablers of engagement include:

- Whānau support.
- Postive relationship and continuity of care with the health worker/health service.
- Access to injections at home, school, or workplace.[3, 63-65]

At a programme level, the clinical governance measures support greater adherence:

- Coordinated services with dedicated teams delivering SAP in the community.
- Robust registers and recall systems.
- Strong linkages between health and school services.
- Use of appropriate health literacy tools.
- Tools such as text reminders.[3, 66]

To minimise days with sub-therapeutic levels of penicillin, providers should engage with patients on a 28-day regimen from day 21 onwards to schedule the next dose. SAP services should monitor the proportion of late injections and establish clinical pathways to enhance patient support and escalate concerns about late injections to the responsible clinician.

Planning for travel

If a person on SAP plans to travel away from their local region, organise in advance for them to receive continuous penicillin. Options include:

- Early injections (preferred).
- Referral to other services in Aotearoa (or internationally as needed).
- Bridging with oral penicillin (as a last resort if benzathine penicillin cannot be arranged).

Trust and engagement with the benzathine penicillin service is key, especially when whānau may have to move at short notice, live with housing insecurity, or are highly mobile. Whānau and staff should develop and agree on a contingency plan (including informing key people and sharing contact details) to ensure continuity of SAP.

Duration of secondary antibiotic prophylaxis

The guidance below on the duration of SAP in Aotearoa for people with ARF and RHD aims to balance the risks and benefits against the burden of regular benzathine penicillin injections. Estimated end dates for SAP should be reassessed periodically and adjusted if clinically indicated, with discussions with the patient and whānau and an update to the SAP service. The appropriate duration of SAP should be reassessed in early adulthood and revised if indicated depending on the severity of persisting disease and a person's risk of recurrence.

See **Appendix 1: Secondary antibiotic prophylaxis for people with acute rheumatic fever**.

See **Appendix 2: Secondary antibiotic prophylaxis for people with non-acute RHD**.

Discontinuing secondary antibiotic prophylaxis

Stopping SAP should be a joint decision between the person with ARF or RHD, their whānau, relevant specialists, and the community SAP provider. Reassessing the need for ongoing SAP is critical in early adulthood, as rangatahi enter higher education or employment. Reassess the severity of RHD rather than relying on a plan made when ARF was first diagnosed. The planned date of cessation may change if:

- There is no echocardiographic evidence of progression (cease on an earlier date).
- Clinical or echocardiographic severity progresses.
- Environmental exposure to Strep A changes.
- The person experiences a recurrence of ARF.
- A specialist recommends a change (Grade D).

Before discontinuing SAP, assess the risk of ongoing exposure to Strep A infections. Consider age, household, and occupation. People with a known history of acute carditis, RHD or intermittent adherence should be evaluated clinically and with an echo (Grade D). Document planned and actual dates of SAP cessation and clinical review in medical records.

Occasionally, secondary prevention services or clinicians may encounter older adults (>35 years) who have remained on SAP for many years without clinical review. These individuals should be referred to their local physician or cardiology service for reassessment, including echo and review, as to whether there is any reason to continue SAP.

Special considerations with secondary prophylaxis

Secondary prophylaxis and reproductive health

Most people with ARF and RHD can enjoy a healthy pregnancy and safe childbirth. However, pregnant people/women with moderate and severe RHD are at elevated risk of cardiac decompensation in pregnancy. Pre-conception counselling with a specialist obstetric physician is recommended for those with moderate or severe RHD, allowing in-depth discussion with the patient, their whānau and review with a cardiologist. For more details, see **Chapter 12: Rheumatic Heart Disease and Pregnancy**. Benzathine penicillin can also be safely given during pregnancy and while breastfeeding, as can erythromycin and oral penicillin.[70-72] Hormonal contraception can safely be offered for those on benzathine penicillin. A recent systematic review found no evidence of significant drug interactions between combined oral contraception and non-rifampicin antibiotics. The effectiveness of contraceptives should not be reduced for most women.[73] Earlier reviews also support this[74-76], while case-crossover studies have mixed results.[77, 78] Progesterone-only oral contraceptives do not interact with benzathine penicillin.

Low-dose lignocaine given with benzathine penicillin is considered safe in pregnancy and while breastfeeding. Lignocaine is excreted into breast milk in small amounts, but its oral bioavailability is very low (35%). Given the small amount of lignocaine used with benzathine penicillin, the amount excreted into breast milk that the infant is exposed to is minimal. More information can be found at:

Data Sheets and Consumer Medicine Information from Medsafe

Xylocaine data sheet from Medsafe (PDF, 236kB)

Secondary antibiotic prophylaxis in anticoagulated patients

Intramuscular bleeding after benzathine penicillin injections when used in conjunction with anticoagulation has rarely been reported in Aotearoa. A 2020 single-centre, retrospective review over five years in Australia found no complications of haematoma in 48 patients on benzathine penicillin with concurrent anticoagulation.[79] The risk of ARF recurrence is higher than the risk of haematoma, especially for patients with severe RHD and previous mechanical valve surgery. Thus, benzathine penicillin injections should be continued for those who are anticoagulated unless there is a history or evidence of uncontrolled bleeding.

The patient's anticoagulation service provider should monitor the international normalised ratio (INR) regularly (suggested monthly at a minimum, more often in complex situations). INR monitoring services vary and can include self-testing and monitoring at community pharmacies or via the GP. See **Chapter 9: Administration of Intramuscular Benzathine Penicillin (Bicillin®L-A)** and **Chapter 11: Management of Rheumatic Heart Disease** for detailed guidance regarding administration of benzathine penicillin in people on anticoagulation.

Secondary antibiotic prophylaxis after valve surgery

The duration of benzathine penicillin prophylaxis for people with RHD post-valve surgery should be individualised depending on age, severity, and social and environmental factors.

See **Appendix 2: Secondary antibiotic prophylaxis for people with non-acute RHD** and **Chapter 11: Management of Rheumatic Heart Disease** for further details. Quality evidence to guide the duration of SAP after mechanical mitral and aortic valve replacement for RHD is lacking. A 2022 retrospective cohort study assessing outcomes in those with and without SAP after valve surgery[80] found higher survival in the SAP group. However, patients in the SAP group were significantly younger. They had less left ventricular dysfunction, a lower percentage of isolated mitral mechanical valve replacement, and a higher percentage of combined aortic and pulmonary biological valve replacement. An adjusted analysis with propensity score matching showed:

- No statistically significant difference in survival at 10 years.
- No differences in survival without re-operation.
- No differences in valve-related hospitalisation at 10 years.[80]

Risk of sudden death after benzathine penicillin in severe rheumatic heart disease

Deaths after benzathine penicillin injections have been reported internationally. A recent review reported 10 deaths in people with RHD between 2005 and 2018.[81] They occurred primarily in people who had severe symptomatic valvular heart disease, heart failure, or both. It was hypothesised that they may have died from vasovagal syncope or cardiovascular compromise after their benzathine penicillin injections.[81] This hypothesis resulted in an advisory from the American Heart Association (AHA) in 2022, advising oral antibiotic prophylaxis, preferably oral penicillin, for patients with RHD requiring SAP at elevated risk of death.[82]

No sudden cardiac events (collapse or death) have been observed in Aotearoa after administration of benzathine penicillin (Pfizer pre-formulated Bicllin®L-A). Expert clinicians from Aotearoa have recently challenged the AHA advisory statement's applicability to Aotearoa, where benzathine penicillin has had an excellent safety record for decades.[83]

People in Aotearoa with severe RHD should continue to be offered benzathine penicillin, preferably delivered by nurses in a register-based programme and reviewed by cardiologists before any change in SAP. Any person with a suspected adverse reaction should be reviewed by a specialist paediatrician or physician to determine the most appropriate antibiotic choice for SAP. Adverse reactions should be notified to Medsafe's CARM.

Those with severe RHD who may be anxious about continuing benzathine penicillin following the AHA advisory can discuss their individual SAP recommendations with a cardiologist experienced in RHD management.[83]

Measuring secondary prophylaxis programme success

Both completeness and timeliness of benzathine penicillin injections are important for individual and programme monitoring. Once fully implemented, the national RFCCS will have the capacity to calculate both parameters. For injections delivered more than three days after the due date, a reason the dose was delivered late is required on the RFCCS (personal communication, 2024, C. Jackson, Clinical Advisor Rheumatic Fever Care Coordination System | Public Health Medicine Specialist, Health New Zealand | Te Whatu Ora).

"She (nurse) comes out if she knows he's up here in the holidays, she comes out to do his injections and if she can't... like if she's on holiday or something, she gets a hold of the nurses to come out... so the nurses are primo as."

Whānau with lived experience of benzathine penicillin injections

Completeness

Completeness for the individual can be measured as the number of doses given divided by the number recommended:

$$\frac{\text{number of doses given}}{\text{number of doses recommended}} \times 100$$

Timeliness

Measuring the number of days at risk is another way to monitor timeliness. Days at risk for a 28-day benzathine penicillin regimen equals the number of days after 28 days that the next benzathine penicillin dose was administered.[43] If the benzathine penicillin administration date is considered day 0, the next benzathine penicillin dose is due on day 28. If the next benzathine penicillin dose is given late, the first day at risk is day 29. All subsequent days before the next dose administration date are counted as days at risk.

Managing Bicillin®L-A supply

Globally, supply shortages of Bicillin®L-A occur periodically. Access in Aotearoa is managed nationally by PHARMAC, with a secure supply chain. A secured stock of alternative dry powder formulations is an option if there is a global or regional stockout. As a last resort during stockouts, oral penicillin could be considered to assure continuity of SAP.

Oral health

Oral health is an important part of holistic secondary prevention of ARF and RHD. Linkages with dental providers should be established for patients on SAP. For those with RHD at risk of bacterial endocarditis, encourage whānau to have dental checks at least once a year and to exercise daily oral care. Offer practical support to arrange these checks. Standard dental treatment is free up to the age of 18 years at community dental services.[84] Some schools have onsite services. Ongoing dental checks after 18 years are recommended, also at least yearly.

Antibiotic prophylaxis for dental procedures

Antibiotic prophylaxis for dental procedures is recommended for people with RHD. The Heart Foundation's bacterial endocarditis wallet card can be given to whānau to support communication about the need for antibiotic prophylaxis with dental services. Some SAP services have standing orders for endocarditis prophylaxis, so refer to local protocols. Documentation and follow-up of dental care should be included in the RFCCS where possible.

See **Chapter 11: Management of Rheumatic Heart Disease** for more information.

National Rheumatic Fever Care Coordination System

Prior to 2024, fifteen separate district or regional rheumatic fever (RF) registers were developed using different approaches and IT solutions. These varied in scope, data, clinical governance, and capacity to monitor the delivery of secondary prevention activities, including SAP. Despite these limitations, RF registers have contributed significantly to the timely delivery of SAP seen in Aotearoa,[6] and the comparatively low ARF recurrence rate, and have supported largely nurse-led models of benzathine penicillin delivery in most communities. The commitment and engagement of nurses with whānau have been key to successful SAP delivery.[7, 8]

In April 2022, a business case was approved for Health New Zealand | Te Whatu Ora to develop and implement a national RF management system. Work commenced on 1 July 2022. In December 2022, Salesforce was approved as the platform for the RFCCS, and a phased approach was taken to development. In November 2023, the phase I pilot was launched with the Te Tai Tokerau and Waikato RF Secondary Prevention Services, migrating approximately 300 patients from district RF registers onto the RFCCS.

Deployment continued in 2024 with migration of the following local RF registers onto the RFCCS: Te Tai Rāwhiti, Bay of Plenty, Lakes, Taranaki, Hawkes Bay, Whanganui, Mid Central, Nelson-Marlborough, Canterbury, and Southern. In 2025, the three remaining RF registers (Wellington, Counties Manukau, and Auckland) will be migrated to the RFCCS to complete the national rollout.

The RFCCS has the key aims of:

- Improving delivery of SAP and reducing loss of follow-up as patients move between health districts and transition from tamariki to adult services.
- Linking up parts of the health system and ARF healthcare pathway to ensure people receive the care they need, when and where they need it.
- Improving healthcare provider access to patient information that is critical to providing the highest quality care and follow-up.
- Including patient and whānau voice.
- Where possible, leveraging existing IT systems.

Referral for secondary prevention services will continue via existing local referral pathways to the RF Coordinator. Patient/whānau consent is required for the RFCCS and this should be included as part of the referral process. The national consent form is available from your local secondary prevention service. RF Secondary Prevention services will register the patient on the RFCCS, create a care plan, and support delivery and coordination of this care plan.

All patients requiring SAP for RF/RHD in Aotearoa should be referred and registered on the RFCCS, including those on oral prophylaxis.

The following patients are currently out of the scope of the RFCCS:

- Patients receiving benzathine penicillin for another indication, e.g. Post-Streptococcal Glomerulonephritis (PSGN), reactive arthritis, and syphilis.
- Patients with non-acute RHD who are not receiving benzathine penicillin, e.g.
  - Those with Stage A screening detected RHD having follow-up echo's only.
  - Patients with RHD who have completed SAP.

Appendices

Appendix 1: Secondary antibiotic prophylaxis for people with acute rheumatic fever

Table 8.3. Anticipated duration of secondary antibiotic prophylaxis — guidance according to diagnostic classification and conditions for ceasing prophylaxis[1]

Diagnosis	Definition	Duration of secondary antibiotic prophylaxis (SAP) (first episode or recurrence)	Conditions for ceasing prophylaxis[4]
Definite ARF, with or without cardiac involvement[1]	2 major, or 1 major and 2 minor manifestations. **PLUS** Evidence of a preceding Strep A infection.	Minimum of 10 years after the most recent episode of ARF, or until age 21 years (whichever is longer).[2, 3, 4] Reassess around age 21 years (clinical assessment and echo). If still moderate RHD or progressed to severe RHD, continue SAP to age 30 years and reassess.	No ARF recurrence within the previous 10 years.[4]
Probable ARF[1] **(without cardiac involvement)**	Highly suspected ARF (1 major and 2 minor manifestations — evidence of a preceding Strep A infection counts as a minor manifestation).	Minimum of 10 years after the most recent episode of probable ARF, or until age 21 years (whichever is longer).[3]	No ARF recurrence within the previous 10 years.[4]
Possible ARF[1]	Incomplete features of ARF but no other diagnosis confirmed.	12 months, then reassess with clinical review, echo and ECG. • SAP may stop earlier if another diagnosis (for example, juvenile idiopathic arthritis) is confirmed during this period. • No other diagnosis confirmed by 12 months, a minimum of five years of SAP is recommended.	No signs or symptoms of ARF within the previous 12 months. Normal echo and ECG.[4]

Notes:

1 Evolution and resolution of initial carditis in ARF may take some months. Initial guidance about SAP should always be reviewed depending on the progression of echocardiographic findings, and the situation of the individual and their whānau.

2 See definitions of mild, moderate and severe carditis in **Table 6.7** in **Chapter 6: Diagnosis of Acute Rheumatic Fever**.

3 Stopping SAP early may be possible in these lower-risk situations:

- In people aged ≥16 years when their ARF was diagnosed (initial episode), cessation after 5 years may be considered if the person had no or mild carditis initially AND a normal follow-up echo at the time of cessation.
- In people aged ≤16 years when ARF was diagnosed (initial episode), cessation may be considered from 18 years of age if the person had no or mild carditis initially AND a normal follow-up echo at the time of cessation AND has completed 10 years SAP at the time of cessation.

Overall, these guidelines are relatively conservative compared to Australian and WHO guidelines. Late recurrences of ARF continue to occur among rangatahi and young adults in Aotearoa.[8] No new local evidence supports further shortening of SAP duration.

4 Moderate carditis frequently improves to mild carditis or better, and occasionally severe carditis improves. However, people with initial severe carditis, including those who have early RHD surgery, are likely to need longer duration SAP. They are likely to have persisting severe RHD, although evidence for SAP in older adults with RHD is limited. Recurrent ARF is rare after 35 years. As well as assessing RHD severity, consider the risk of Strep A exposure or recurrence and the benefits of continuing SAP. Any regression or progression of RHD should be determined by echo. Future recommendations for SAP should be determined by shared decision-making with the person and their whānau, based on echo findings and personal/whānau circumstances.

5 Clinical review and echo of all patients with moderate–severe carditis at 12–24 months after cessation of SAP is recommended as part of ongoing RHD management. This ensures no deterioration of RHD and promotes ongoing awareness of appropriate management of Strep A infections, oral health and other health issues.

The diagnosing clinician should obtain consent for registration on the RFCCS and refer the patient to the local district or regional secondary prevention service. Patients will be registered on the RFCCS by each SAP service.

Appendix 2: Secondary antibiotic prophylaxis for people with non-acute RHD

Table 8.4. Recommendations for secondary antibiotic prophylaxis for persons with newly diagnosed and non-acute RHD (includes RHD detected by echocardiographic screening, as an incidental finding and those with prior ARF)

WHF 2023 Stage	Echocardiographic features	Clinical management	Duration of secondary antibiotic prophylaxis	Conditions for ceasing prophylaxis
Stage A RHD This diagnosis applies only to people ≤20 years of age with no prior history of ARF.	Pathological MR or AR **without** morphological features in high prevalence population.	**Screen-detected RHD** Counsel whānau that the echo findings may or may not prove to be RHD. Usual advice is not to start SAP unless there is family history of ARF or RHD. Shared-decision-making and consideration of whānau preferences is required. Experience from screening in Aotearoa is that whānau of persons with Stage A RHD (previously called borderline RHD) tend to choose SAP if another family member has had ARF/RHD. If starting SAP, consent to register on the RFCCS. If not starting SAP, place the person on the screening programme database as relevant. Clinical review with echo in 1–2 years.	**Screen-detected RHD** If SAP is started, give for 1–2 years and consider stopping SAP if echo findings have normalised. If Stage A RHD persists after 1–2 years, SAP is recommended for 5 years. If SAP was not started but the follow-up echo shows persisting Stage A RHD, recommend 5 years of SAP. If the follow-up echo shows progress to Stage B or C, recommend prophylaxis for 10 years as per Stage B and C recommendations below.	If Stage A echo findings are normal after 12–24 months. OR if Stage A findings have normalised or not progressed after five years' SAP.
Stage B RHD or any prior ARF episode	**In individuals aged ≤20 years** — mild pathological valvular regurgitation plus at least one morphological feature. **In individuals aged >20 years** — at least two morphological features, or mild pathological regurgitation in both mitral and aortic valves.	**Person with prior ARF diagnosis (with or without residual RHD) or screen-detected RHD** SAP is recommended for all persons with Stage B RHD or prior ARF, with clinical review and repeat echo 1–2 years after diagnosis. Ensure the person is consented and registered on the RFCCS.	**Clinically diagnosed ARF or RHD** Minimum of 10 years or age 21, whichever is longer. Reassess SAP around 21 years of age. **Screen-detected Stage B RHD** 10 years duration currently in Aotearoa. This is an evolving area. International research to determine whether people with screen-detected Stage B RHD can safely have a shortened duration of SAP (e.g. 5 years) is in progress.	No probable or definite ARF within the previous 10 years; no progression of RHD.
Stage C	Moderate or severe MR, moderate or severe AR, mitral stenosis of any severity. Persons with Stage C RHD are at risk of clinical complications.	**Any person with Stage C RHD** SAP is recommended for all persons with Stage C or D RHD. Specialised RHD care as per Chapter 11: Management of Rheumatic Heart Disease. Ensure the person is consented and registered on the RFCCS.	10 years or until age 21 years, whichever is longer. Reassess SAP around 21 years of age. Those with continued moderate or severe RHD should continue to age 30–35 years and reassess. Consider individual risk of Strep A exposure or recurrence when deciding about cessation vs continuation of SAP.	No probable or definite ARF (within the previous 10 years). No progression of RHD.
Stage D	Established moderate or severe RHD detected by echocardiography with overt clinical complications including need for cardiac surgery, heart failure, arrhythmia, stroke and infective endocarditis.	**Any person with Stage D RHD** SAP is recommended for all persons with Stage C or D RHD. Specialised RHD care as per Chapter 11: Management of Rheumatic Heart Disease. Ensure the person is consented and registered on the RFCCS.	10 years or until age 30–35 years, whichever is longer. Reassess SAP around 30–35 years. SAP beyond age 30 years is individualised. Assess RHD severity, risk of Strep A exposure, and benefits /risks. In people with decompensated severe RHD who are not candidates for cardiac surgery, or are under palliative care, consider daily oral penicillin (as per AHA 2022 Advisory).	Individualised as detailed in Chapter 11: Management of Rheumatic Heart Disease.

Adapted from the 2023 World Heart Federation guidelines.

References

1. Mackie AS, Islam S, Magill-Evans J, Rankin KN, Robert C, Schuh M, et al. Healthcare transition for youth with heart disease: a clinical trial. *Heart*. 2014;100(14):1113–1118. https://doi.org/10.1136/heartjnl-2014-305748

2. Sable C, Foster E, Uzark K, Bjornsen K, Canobbio MM, Connolly HM, et al. Best practices in managing transition to adulthood for adolescents with congenital heart disease: the transition process and medical and psychosocial issues: a scientific statement from the American Heart Association. *Circulation*. 2011;123(13):1454–1485. https://doi.org/10.1161/CIR.0b013e3182107c56

3. Garden E, Butler R, Cram A, Fale Z, al e. Rheumatic fever nursing pilot evaluation report. Auckland: Te Whatu Ora Counties Manukau; 2023.

4. Pitama S, Huria T, Lacey C. Improving Māori health through clinical assessment: waikare o te waka o Meihana. *New Zealand Medical Journal*. 2014;127(1393):107–119.

5. Lacey C, Huria T, Beckert L, Gilles M, Pitama S. The Hui Process: a framework to enhance the doctor-patient relationship with Māori. *New Zealand Medical Journal*. 2011;124(1347):72–78.

6. New Zealand Heart Foundation. New Zealand guidelines for rheumatic fever: diagnosis, management and secondary prevention of acute rheumatic fever and rheumatic heart disease: 2014 update. Heart Foundation; 2014. https://www.heartfoundation.org.nz/resources/acute-rheumatic-fever-and-rheumatic-heart-disease-guideline (Accessed December 16 2024).

7. Spinetto H, Lennon D, Horsburgh M. Rheumatic fever recurrence prevention: a nurse-led programme of 28-day penicillin in an area of high endemnicity. *Journal of Paediatrics and Child Health*. 2011;47(4):228–234. https://doi.org/10.1111/j.1440-1754.2010.01942.x

8. Dennison A, Peat B, Wilson E, Leversha A, Wheeler M, Briggs S, et al. Rheumatic fever recurrences in New Zealand 2010–14. *New Zealand Medical Journal*. 2020;133(1516):47–57.

9. Karthikeyan G, Watkins D, Bukhman G, Cunningham MW, Haller J, Masterson M, et al. Research priorities for the secondary prevention and management of acute rheumatic fever and rheumatic heart disease: a National Heart, Lung, and Blood Institute workshop report. *BMJ Global Health*. 2023;8(Suppl 9). https://doi.org/10.1136/bmjgh-2023-012468

10. Bland EF, Duckett Jones T. Rheumatic fever and rheumatic heart disease; a twenty year report on 1000 patients followed since childhood. *Circulation*. 1951;4(6):836–843. https://doi.org/10.1161/01.cir.4.6.836

11. Wood HF, Simpson R, Feinstein AR, Taranta A, Tursky E, Stollerman G. Rheumatic fever in children and adolescents. A long-term epidemiologic study of subsequent prophylaxis, streptococcal infections, and clinical sequelae. I. Description of the investigative techniques and of the population studied. *Annals of Internal Medicine*. 1964;60:6–17. https://doi.org/10.7326/0003-4819-60-2-6

12. de Dassel J, Lennon D, Dougherty S, Ralph A. Secondary prevention of acute rheumatic fever and rheumatic heart disease. In: Dougherty S, Carapetis J, Zuhlke L, Wilson N, editors. Acute rheumatic fever and rheumatic heart disease. Maryland Heights: Elsevier Science; 2021. p. 207–234.

13. Taranta A, Wood HF, Feinstein AR, Simpson R, Kleinberg E. Rheumatic fever in children and adolescents. *Annals of Internal Medicine*. 1964;60(2):47–57. https://doi.org/10.7326/0003-4819-60-2-47

14. Manyemba J, Mayosi BM. Penicillin for secondary prevention of rheumatic fever. *Cochrane Database of Systematic Reviews*. 2002;2002(3):Cd002227. https://doi.org/10.1002/14651858.Cd002227

15. Bray JJ, Thompson S, Seitler S, Ali SA, Yiu J, Salehi M, et al. Long-term antibiotic prophylaxis for prevention of rheumatic fever recurrence and progression to rheumatic heart disease. *Cochrane Database of Systematic Reviews*. 2024;9(9):Cd015779. https://doi.org/10.1002/14651858.Cd015779

16. World Health Organization. Rheumatic fever and rheumatic heart disease: report of WHO expert consultation. Geneva: World Health Organization; 2001. https://iris.who.int/bitstream/handle/10665/42898/WHO_TRS_923.pdf?sequence=1&isAllowed=y (Accessed February 22 2025).

17. Baumgartner H, Falk V, Bax JJ, De Bonis M, Hamm C, Holm PJ, et al. 2017 ESC/EACTS Guidelines for the management of valvular heart disease. *European Heart Journal*. 2017;38(36):2739–2791. https://doi.org/10.1093/eurheartj/ehx391

18. Remenyi B, Carapetis J, Wyber R, Taubert K, Mayosi BM. Position statement of the World Heart Federation on the prevention and control of rheumatic heart disease. *Nature Reviews: Cardiology*. 2013;10(5):284–292. https://doi.org/10.1038/nrcardio.2013.34

19. Padmavati S, Sharma KB, Jayaram O. Epidemiology and prophylaxis of rheumatic fever in Delhi--a five year follow-up. *Singapore Medical Journal*. 1973;14(3):457–461.

20. Brick M, Mc KH, Gourley M, Roy TE, Keith JD. Oral penicillin prophylaxis in rheumatic fever patients. *Canadian Medical Association Journal*. 1950;63(3):255–258.

21. Cope S, Sanderson G, St Hill CA, Chamberlain EN. Prophylactic use of oral penicillin in rheumatic fever, chorea, and carditis. *British Medical Journal*. 1960;1(5177):913–917. https://doi.org/10.1136/bmj.1.5177.913

22. Evans JA. Oral penicillin in the prophylaxis of streptococcal infection and rheumatic relapse. *Proceedings of the Royal Society of Medicine*. 1950;43(3):206–208.

23. Frankish JD. Rheumatic fever prophylaxis: Gisborne experience. *New Zealand Medical Journal*. 1984;97(765):674–675.

24. Camara EJN, Santos JMd, Alves-Silva LS, Latado AL. Rheumatic fever recurrence: risk factors and clinical characteristics. *Clinical Trials and Regulatory Science in Cardiology*. 2016;19:5–8. https://doi.org/10.1016/j.ctrsc.2016.05.007

25. Mirabel M, Noel B, Tafflet M, Parks T, Braunstein C, Hagege A, et al. Rheumatic heart disease: factors associated with outcomes in a high-income country. *European Heart Journal*. 2015;1(269).

26. de Dassel JL, de Klerk N, Carapetis JR, Ralph AP. How many doses make a difference? An analysis of secondary prevention of rheumatic fever and rheumatic heart disease. *Journal of the American Heart Association*. 2018;7(24):e010223. https://doi.org/10.1161/JAHA.118.010223

27. Tal R, Hamad Saied M, Zidani R, Levinsky Y, Straussberg R, Amir J, et al. Rheumatic fever in a developed country — is it still relevant? A retrospective, 25 years follow-up. *Pediatric Rheumatology Online Journal*. 2022;20(1):20. https://doi.org/10.1186/s12969-022-00678-7

28. Dougherty S, Carapetis J, Zühlke L, Wilson N. Acute rheumatic fever and rheumatic heart disease. Amsterdam: Elsevier; 2020.

29. Doran J, Kangaharan N, Remeny B, Cass A, Ilton M, Mc Donald M, et al. 699 impact of adherence to antibiotic prophylaxis on rheumatic heart valve surgical outcomes in the Northern Territory of Australia. *Heart, Lung and Circulation*. 2020;29. https://doi.org/10.1016/j.hlc.2020.09.706

30. Culliford-Semmens N, Tilton E, Webb R, Lennon D, Paku B, Malcolm J, et al. Adequate adherence to benzathine penicillin secondary prophylaxis following the diagnosis of rheumatic heart disease by echocardiographic screening. *New Zealand Medical Journal*. 2017;130(1457):50–57.

31. French S. The complexities of rheumatic fever secondary prophylaxis in Tairawhiti. *Heart, Lung and Circulation*. 2019;28. https://doi.org/10.1016/j.hlc.2019.05.162

32. Grayson S, Horsburgh M, Lennon D. An Auckland regional audit of the nurse-led rheumatic fever secondary prophylaxis programme. *New Zealand Medical Journal*. 2006;119(1243):U2255.

33. North A, Martis W, Sutton T, Lennon D, Peat B, Vignakumar V, et al. The natural and unnatural history of acute rheumatic fever in the modern era. *Heart, Lung and Circulation*. 2016;25:S11–S12. https://doi.org/10.1016/j.hlc.2016.05.027

34. Oliver J, Robertson O, Zhang J, Marsters BL, Sika-Paotonu D, Jack S, et al. Ethnically disparate disease progression and outcomes among acute rheumatic fever patients in New Zealand, 1989–2015. *Emerging Infectious Diseases*. 2021;27(7):1893–1902. https://doi.org/10.3201/eid2707.203045

35. Saxena A, Ramakrishnan S, Roy A, Seth S, Krishnan A, Misra P, et al. Prevalence and outcome of subclinical rheumatic heart disease in India: the RHEUMATIC (Rheumatic Heart Echo Utilisation and Monitoring Actuarial Trends in Indian Children) study. *Heart*. 2011;97(24):2018–2022. https://doi.org/10.1136/heartjnl-2011-300792

36. Karki P, Uranw S, Bastola S, Mahato R, Shrestha NR, Sherpa K, et al. Effectiveness of systematic echocardiographic screening for rheumatic heart disease in Nepalese schoolchildren: a cluster randomized clinical trial. *JAMA Cardiology*. 2021;6(4):420–426. https://doi.org/10.1001/jamacardio.2020.7050

37. Okello E, Longenecker CT, Beaton A, Kamya MR, Lwabi P. Rheumatic heart disease in Uganda: predictors of morbidity and mortality one year after presentation. *BMC Cardiovascular Disorders*. 2017;17(1):20. https://doi.org/10.1186/s12872-016-0451-8

38. Spinetto H. Recurrences of rheumatic fever in Auckland 1993–1999 [Dissertation]. Auckland: University of Auckland; 2003.

39. Amir J, Ginat S, Cohen YH, Marcus TE, Keller N, Varsano I. Lidocaine as a diluent for administration of benzathine penicillin G. *Pediatric Infectious Disease Journal*. 1998;17(10):890–893. https://doi.org/10.1097/00006454-199810000-00008

40. Russell K, Nicholson R, Naidu R. Reducing the pain of intramuscular benzathine penicillin injections in the rheumatic fever population of Counties Manukau District Health Board. *Journal of Paediatrics and Child Health*. 2014;50(2):112–117. https://doi.org/10.1111/jpc.12400

41. Ketema EB, Gishen NZ, Hailu A, Leul A, Hadgu A, Hagos K, et al. High risk of early sub-therapeutic penicillin concentrations after intramuscular benzathine penicillin G injections in Ethiopian children and adults with rheumatic heart disease. *PLoS Neglected Tropical Diseases*. 2021;15(6):e0009399. https://doi.org/10.1371/journal.pntd.0009399

42. Hand RM, Salman S, Newall N, Vine J, Page-Sharp M, Bowen AC, et al. A population pharmacokinetic study of benzathine benzylpenicillin G administration in children and adolescents with rheumatic heart disease: new insights for improved secondary prophylaxis strategies. *Journal of Antimicrobial Chemotherapy*. 2019;74(7):1984–1991. https://doi.org/10.1093/jac/dkz076

43. Kado J, Salman S, Hand R, O'Brien M, Ralph A, Bowen AC, et al. Population pharmacokinetic study of benzathine penicillin G administration in Indigenous children and young adults with rheumatic heart disease in the Northern Territory, Australia. *Journal of Antimicrobial Chemotherapy*. 2022;77(10):2679–2682. https://doi.org/10.1093/jac/dkac231

44. Hla TK, Osowicki J, Salman S, Batty KT, Marsh JA, Kado J, et al. Study protocol for controlled human infection for penicillin G against streptococcus pyogenes: a double-blinded, placebo-controlled, randomised trial to determine the minimum concentration required to prevent experimental pharyngitis (the CHIPS trial). *BMJ Open*. 2022;12(12):e064022. https://doi.org/10.1136/bmjopen-2022-064022

45. Kado JH, Salman S, Henderson R, Hand R, Wyber R, Page-Sharp M, et al. Subcutaneous administration of benzathine benzylpenicillin G has favourable pharmacokinetic characteristics for the prevention of rheumatic heart disease compared with intramuscular injection: a randomized, crossover, population pharmacokinetic study in healthy adult volunteers. *Journal of Antimicrobial Chemotherapy*. 2020;75(10):2951–2959. https://doi.org/10.1093/jac/dkaa282

46. Kado J, Salman S, Hla TK, Enkel S, Henderson R, Hand RM, et al. Subcutaneous infusion of high-dose benzathine penicillin G is safe, tolerable, and suitable for less-frequent dosing for rheumatic heart disease secondary prophylaxis: a phase 1 open-label population pharmacokinetic study. *Antimicrobial Agents and Chemotherapy*. 2023;67(12):e0096223. https://doi.org/10.1128/aac.00962-23

47. Cooper J, Enkel SL, Moodley D, Dobinson H, Andersen E, Kado JH, et al. "Hurts less, lasts longer"; a qualitative study on experiences of young people receiving high-dose subcutaneous injections of benzathine penicillin G to prevent rheumatic heart disease in New Zealand. *PloS One*. 2024;19(5):e0302493. https://doi.org/10.1371/journal.pone.0302493

48. Enkel SL, Kado J, Hla TK, Salman S, Bennett J, Anderson A, et al. Qualitative assessment of healthy volunteer experience receiving subcutaneous infusions of high-dose benzathine penicillin G (SCIP) provides insights into design of late phase clinical studies. *PloS One*. 2023;18(4):e0285037. https://doi.org/10.1371/journal.pone.0285037

49. Manyemba J, Mayosi BM. Intramuscular penicillin is more effective than oral penicillin in secondary prevention of rheumatic fever--a systematic review. *South African Medical Journal*. 2003;93(3):212–218.

50. Feinstein AR, Wood HF, Epstein JA, Taranta A, Simpson R, Tursky E. A controlled study of three methods of prophylaxis against streptococcal infection in a population of rheumatic children. II. Results of the first three years of the study, including methods for evaluating the maintenance of oral prophylaxis. *New England Journal of Medicine*. 1959;260(14):697–702. https://doi.org/10.1056/nejm195904022601405

51. Neutze JM, Clarkson PM. Rheumatic fever: an unsolved problem in New Zealand. *New Zealand Medical Journal*. 1984;97(763):591–593.

52. Lagacé-Wiens P, Rubinstein E. Adverse reactions to β-lactam antimicrobials. *Expert Opinion on Drug Safety*. 2012;11(3):381–399. https://doi.org/10.1517/14740338.2012.643866

53. Blumenthal KG, Peter JG, Trubiano JA, Phillips EJ. Antibiotic allergy. *Lancet*. 2019;393(10167):183–198. https://doi.org/10.1016/s0140-6736(18)32218-9

54. Markowitz M, Lue HC. Allergic reactions in rheumatic fever patients on long-term benzathine penicillin G: the role of skin testing for penicillin allergy. *Pediatrics*. 1996;97(6 Pt 2):981–983.

55. Blumenthal KG, Lu N, Zhang Y, Walensky RP, Choi HK. Recorded penicillin allergy and risk of mortality: a population-based matched cohort study. *Journal of General Internal Medicine*. 2019;34(9):1685–1687. https://doi.org/10.1007/s11606-019-04991-y

56. Kaminsky LW, Ghahramani A, Hussein R, Al-Shaikhly T. Penicillin allergy label is associated with worse clinical outcomes in bacterial pneumonia. *The Journal of Allergy and Clinical Immunology: In Practice*. 2022;10(12):3262–3269. https://doi.org/10.1016/j.jaip.2022.08.027

57. International Rheumatic Fever Study Group. Allergic reactions to long-term benzathine penicillin prophylaxis for rheumatic fever. International Rheumatic Fever Study Group. *Lancet*. 1991;337(8753):1308–1310.

58. Belton S, Kruske S, Jackson Pulver L, Sherwood J, Tune K, Carapetis J, et al. Rheumatic heart disease in pregnancy: how can health services adapt to the needs of Indigenous women? A qualitative study. *Australian and New Zealand Journal of Obstetrics and Gynaecology*. 2018;58(4):425–431. https://doi.org/10.1111/ajo.12744

59. Crengle S, Luke JN, Lambert M, Smylie JK, Reid S, Harré-Hindmarsh J, et al. Effect of a health literacy intervention trial on knowledge about cardiovascular disease medications among Indigenous peoples in Australia, Canada and New Zealand. *BMJ Open*. 2018;8(1):e018569. https://doi.org/10.1136/bmjopen-2017-018569

60. Anderson A, Spray J. Beyond awareness: towards a critically conscious health promotion for rheumatic fever in Aotearoa, New Zealand. *Social Science and Medicine*. 2020;247:112798. https://doi.org/10.1016/j.socscimed.2020.112798

61. Anderson A, Brown R, Wheeler J, Jansen RM. Pacific Fono: a community-based initiative to improve rheumatic fever service delivery for Pacific Peoples in South Auckland. *Journal of Primary Health Care*. 2020;12(4):384–390. https://doi.org/10.1071/hc20022

62. Carlson T, Moewaka Barnes H, McCreanor T. Health literacy in action: kaupapa Māori evaluation of a cardiovascular disease medications health literacy intervention. *AlterNative: An International Journal of Indigenous Peoples*. 2019;15(2):101–110. https://doi.org/10.1177/1177180119828050

63. Anderson A, Peat B, Ryland J, Ofanoa M, Burgess H, Malungahu G, et al. Mismatches between health service delivery and community expectations in the provision of secondary prophylaxis for rheumatic fever in New Zealand. *Australian and New Zealand Journal of Public Health*. 2019;43(3):294–299. https://doi.org/10.1111/1753-6405.12890

64. Anderson A, Leversha A, Ofanoa M, Malungahu G, Burgess H, Wade W. Māori and Pacific whānau experiences of recurrent rheumatic fever and unexpected rheumatic heart disease in New Zealand. Auckland: University of Auckland; 2017. https://www.fmhs.auckland.ac.nz/assets/fmhs/MAPAS/Recurrent%20Rheumatic%20Final%20document.pdf (Accessed February 24 2025).

65. Barker H, Oetzel JG, Scott N, Morley M, Carr PEA, Oetzel KB. Enablers and barriers to secondary prophylaxis for rheumatic fever among Māori aged 14-21 in New Zealand: a framework method study. *International Journal for Equity in Health*. 2017;16(1):201. https://doi.org/10.1186/s12939-017-0700-1

66. Remond MG, Coyle ME, Mills JE, Maguire GP. Approaches to improving adherence to secondary prophylaxis for rheumatic fever and rheumatic heart disease: A literature review with a global perspective. *Cardiology in Review*. 2016;24(2):94–98. https://doi.org/10.1097/CRD.0000000000000065

67. Gerber MA, Baltimore RS, Eaton CB, Gewitz M, Rowley AH, Shulman ST, et al. Prevention of rheumatic fever and diagnosis and treatment of acute Streptococcal pharyngitis: a scientific statement from the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee of the Council on Cardiovascular Disease in the Young, the Interdisciplinary Council on Functional Genomics and Translational Biology, and the Interdisciplinary Council on Quality of Care and Outcomes Research: endorsed by the American Academy of Pediatrics. *Circulation*. 2009;119(11):1541–1551. https://doi.org/10.1161/circulationaha.109.191959

68. Taranta A. Factors influencing recurrent rheumatic fever. *Annual Review of Medicine*. 1967;18:159–172. https://doi.org/10.1146/annurev.me.18.020167.001111

69. He VY, Condon JR, Ralph AP, Zhao Y, Roberts K, de Dassel JL, et al. Long-term outcomes from acute rheumatic fever and rheumatic heart disease: a data-linkage and survival analysis approach. *Circulation*. 2016;134(3):222–232. https://doi.org/10.1161/CIRCULATIONAHA.115.020966

70. Briggs G, Freeman R. Brigg's drugs in pregnancy and lactation: a reference guide to fetal and neonatal risk. 10th ed. Philadelphia: Wolters Kluwer Health; 2014.

71. National Institute of Child Health and Human Development. Drugs and Lactation Database (LactMed®) Benzathine Penicillin G. Bethesda: National Institute of Child Health and Human Development; 2006.

72. Gardiner S. Drug safety in lactation. MedSafe; 2001. https://www.medsafe.govt.nz/profs/puarticles/lactation.htm (Accessed February 24 2025).

73. Simmons KB, Haddad LB, Nanda K, Curtis KM. Drug interactions between non-rifamycin antibiotics and hormonal contraception: a systematic review. *American Journal of Obstetrics and Gynecology*. 2018;218(1):88–97.e14. https://doi.org/10.1016/j.ajog.2017.07.003

74. Dickinson BD, Altman RD, Nielsen NH, Sterling ML. Drug interactions between oral contraceptives and antibiotics. *Obstetrics and Gynecology*. 2001;98(5 Pt 1):853–860. https://doi.org/10.1016/s0029-7844(01)01532-0

75. DeRossi SS, Hersh EV. Antibiotics and oral contraceptives. *Dental Clinics of North America*. 2002;46(4):653–664. https://doi.org/10.1016/s0011-8532(02)00017-4

76. Archer JS, Archer DF. Oral contraceptive efficacy and antibiotic interaction: a myth debunked. *Journal of the American Academy of Dermatology*. 2002;46(6):917–923. https://doi.org/10.1067/mjd.2002.120448

77. Koopmans PC, Bos JH, de Jong van den Berg LT. Are antibiotics related to oral combination contraceptive failures in the Netherlands? A case-crossover study. *Pharmacoepidemiology and Drug Safety*. 2012;21(8):865–871. https://doi.org/10.1002/pds.3267

78. Toh S, Mitchell AA, Anderka M, de Jong-van den Berg LT, Hernández-Díaz S. Antibiotics and oral contraceptive failure — a case-crossover study. *Contraception*. 2011;83(5):418–425. https://doi.org/10.1016/j.contraception.2010.08.020

79. Fox E, Misko J, Rawlins M, Manning L. The risk of intramuscular haematoma is low following injection of benzathine penicillin G in patients receiving concomitant anticoagulant therapy. *Journal of Thrombosis and Thrombolysis*. 2020;50(1):237–238. https://doi.org/10.1007/s11239-019-02013-6

80. Al-Jazairi AS, Althobaiti AM, Marek J, Devol EB, Al Halees Z, Mohty D, et al. Does secondary antibiotic prophylaxis improve clinical outcomes in adult rheumatic heart disease patients post-valve replacement? *World Journal for Pediatric and Congenital Heart Surgery*. 2023;14(2):161–167. https://doi.org/10.1177/21501351221139834

81. Marantelli S, Hand R, Carapetis J, Beaton A, Wyber R. Severe adverse events following benzathine penicillin G injection for rheumatic heart disease prophylaxis: cardiac compromise more likely than anaphylaxis. *Heart Asia*. 2019;11(2):e011191. https://doi.org/10.1136/heartasia-2019-011191

82. Sanyahumbi A, Ali S, Benjamin IJ, Karthikeyan G, Okello E, Sable CA, et al. Penicillin reactions in patients with severe rheumatic heart disease: a presidential advisory from the American Heart Association. *J Am Heart Assoc*. 2022;11(5):e024517. https://doi.org/10.1161/JAHA.121.024517

83. Webb R, Wheeler M, Peat B, Lund M, Luey C, Briggs S, et al. Aotearoa New Zealand clinicians respond to the 2022 American Heart Association Presidential Advisory Statement regarding penicillin reactions in people with severe rheumatic heart disease. *New Zealand Medical Journal*. 2023;136(1586):9–11. https://doi.org/10.26635/6965.e1586

84. Health New Zealand | Te Whatu Ora. Publicly funded dental care. Health New Zealand | Te Whatu Ora. https://info.health.nz/keeping-healthy/teeth-and-gums/dental-care?msclkid=f1628517c69711ecbedf3b73be29d55f (Accessed February 23 2025).

85. Rwebembera J, Marangou J, Mwita JC, Mocumbi AO, Mota C, Okello E, et al. 2023 World Heart Federation guidelines for the echocardiographic diagnosis of rheumatic heart disease. *Nature Reviews: Cardiology*. 2024;21(4):250–263. https://doi.org/10.1038/s41569-023-00940-9

86. Strasser T, Dondog N, El Kholy A, Gharagozloo R, Kalbain VV, Ogunbi O, et al. The community control of rheumatic fever and rheumatic heart disease: report of a WHO international cooperative project Bulletin of World Health Organization. 1981;59(2):285–294.

9

Administration of Intramuscular Benzathine Penicillin (Bicillin®L-A)

In this chapter

"So my first experience with the injections that was the worst experience; I got it in my thigh, and I just screamed the ward down, oh it was so bad, so I dreaded the next month … but nowadays they have the buzzy bee. And they, I LOVE that, I'm going, like, I'm 30 years old and I still ask for that."

Whānau with lived experience

Key changes

This is a new chapter. In the 2024 guidelines, the weight-based cut-off for the 600,000 unit (450 mg) dose has been changed to 20kg (from 30kgs). This change was made to avoid the potential under-dosing of growing tamariki.

Key points

- Benzathine penicillin is used for secondary antibiotic prophylaxis (SAP) of acute rheumatic fever (ARF) and treatment of Strep A throat infections. Aotearoa uses a pre-formulated suspension preparation of benzathine penicillin known as Bicillin®L-A.
- Intramuscular injections of benzathine penicillin are painful. Developmentally appropriate psychological support is essential for managing and minimising pain and distress.
- Adding lignocaine to benzathine penicillin is recommended to significantly reduce pain unless contraindicated.
- The ventrogluteal site is recommended and safer for administering intramuscular injections of benzathine penicillin.

Introduction

Benzathine penicillin comes in a variety of formulations, including dry powder and pre-formulated suspension (liquid). In Aotearoa, the Pfizer pre-formulated suspension Bicillin®L-A (active ingredient: Benzathine benzylpenicillin tetrahydrate 442 mg/mL (1,200,000 units/2.3 mL) is used for ARF SAP. Guidance in this chapter applies to this product but does not necessarily apply to reconstituted dry powder formulations.

Drug administration guidance in this chapter uses Bicillin®L-A to refer to benzathine penicillin. This chapter provides general guidance based on the Health New Zealand | Te Whatu Ora Counties Manukau benzathine penicillin administration guideline (2023).[1] Please also refer to MedSafe's product sheet for Bicillin®L-A as well as to local administration protocols and guidelines.

Indications for Benzathine penicillin

Intramuscular benzathine penicillin is given in the context of ARF to prevent recurrences and to treat Strep A infections.

- Benzathine penicillin is given for SAP following a diagnosis of ARF or rheumatic heart disease (RHD).
- The initial dose should usually be given in the hospital.
- Injections are given every 28 days (occasionally every 21 days) in homes, schools, workplaces, and primary healthcare clinics.
- A "one-off" injection of benzathine penicillin may be given to treat Strep A sore throat, in people at high risk of developing ARF, usually in primary healthcare settings or school-based programmes.
- Benzathine penicillin is also used to treat syphilis.

Benzathine penicillin dosing and adjunctive lignocaine

Dosing of benzathine penicillin and the dose interval

Weight-based dosing is used for benzathine penicillin in Aotearoa. Table 9.1 shows the recommended doses for people with ARF, RHD, and Strep A pharyngitis. For SAP, most people are prescribed benzathine penicillin every 28 days. A small number of people require it every 21 days following recurrent ARF. Decisions about dose interval should be made by an experienced ARF physician or paediatrician. Doses can also be brought forward up to two weeks if required (for example, if the person is going on a holiday overseas or in a location where benzathine penicillin may not be accessible). It is better for doses to be brought forward and given early than for doses to be given more than 28 days apart. If a person is travelling, every effort should be made to deliver the injection in the district the person is visiting.

Adjunctive Lignocaine

Adding 0.25 mL of 2% lignocaine to Bicillin®L-A is recommended in Aotearoa, as it significantly reduces pain immediately and in the first 24 hours after the injection. Lignocaine does not adversely affect serum penicillin concentrations (Grade C).

Contraindications to Adjunctive Lignocaine

Lignocaine is contraindicated in people with known hypersensitivity to local anaesthetics of the amide type, as well as in people with persistent second or third-degree heart block. Lignocaine is relatively contraindicated in people with bradycardia or hypovolaemia.

Required training for healthcare workers

Only healthcare workers trained in intramuscular injections and identifying anatomical landmarks should administer an injection of benzathine penicillin. Appropriate training is important to minimise the risk of intravascular administration, pain, and other complications. Trained healthcare workers who can administer benzathine penicillin include:

- Registered and enrolled nurses who have completed appropriate training and certification.
- Any additional local training or credentialing relevant to Bicillin®L-A administration.
- Nurse practitioners.
- Medical practitioners.

Avoiding distress associated with injections is vital

Intramuscular injections are painful. Careful preparation, which includes spending time with the person and their whānau is always needed. Pain and distress associated with injections can adversely affect:

- How a person feels about subsequent injections.
- Whether a person will continue with SAP.
- How a person will interact with healthcare providers in the future.

Psychological and pharmaceutical interventions are essential to manage pain and distress, particularly for tamariki and rangatahi on long-term SAP.

Physical restraint should never be used when delivering an injection of Bicillin®L-A to a tamariki or rangatahi, because they, their whānau, and clinicians may experience adverse psychological effects. This may adversely impact their trust and engagement with healthcare services.[2-5]

Non-pharmacologic strategies to support patients

- Ensure the environment and support provided are developmentally appropriate, culturally safe, and low-stress. Ensure the person has whānau support if available.
- Administration by healthcare workers who have established a trusting relationship with the tamaiti or rangatahi and whānau is preferred.
- In the hospital, support from play specialists is invaluable, especially when preparing for the first injection and for any tamaiti who experiences high levels of distress or anxiety.

- Document person's preferences for injection delivery, analgesia, and support in a care plan.
- Regularly check the plan with the person.
- Offer people the opportunity to receive their injections at a location of their choice, such as at a clinic, at home, in a workplace, or at school. Where possible, offer weekend appointments to minimise disruption to work and or school.
- Offer other non-pharmacologic options, including the BUZZY® device, ice packs, distraction, and cold spray.

Other medications to help reduce pain

Paracetamol or ibuprofen

Offer oral paracetamol or ibuprofen before delivering the injection and at appropriate time intervals afterward, as required. Provide a prescription, if possible, for use at home.

Entonox® — pre-mixed nitrous oxide

In inpatient settings, where available, consider using pre-mixed nitrous oxide (Entonox®) to reduce procedural pain and anxiety in inpatient settings, where the use of other non-pharmacological and pharmacological strategies is insufficient. Entonox® is a gas mixture of 50% nitrous oxide and 50% oxygen, which a person self-administers using a mouthpiece or mask.

No published data exists to confirm how effective Entonox® is with injections of benzathine penicillin, but it has proved effective for tamariki and rangatahi undergoing other painful procedures and is widely used in hospital paediatric practice.[6-9] Follow local hospital protocols when using Entonox®.

Continuous flow nitrous oxide

Continuous flow nitrous oxide at concentrations of between 50% and 70% is another option for procedural sedation and analgesia, which may be available in some hospitals and emergency departments. This is a resource-intensive process that requires:

- Appropriate hospital facilities.
- Senior medical staff who are credentialed in paediatric sedation.
- At least two registered nurses — one trained to deliver the continuous nitrous oxide and the other trained to give the injection of benzathine penicillin with lignocaine and the BUZZY® device.

Early experience with continuous nitrous oxide at KidzFirst Hospital has been favourable. In combination with psychologist, play-specialist support, and with careful whānau preparation, tamariki with complex needs (such as severe needle phobia or autism) have successfully transitioned to receiving injections in the community once they were better able to manage the procedure.

Other medications

Other analgesic medications, such as midazolam and intranasal fentanyl, have been used in some settings, but local experience suggests these medications are less successful (Personal communication, 2024, K. Russell. Clinical Psychologist, KidzFirst Children's Hospital). Although clonidine is occasionally used in Australia, experience with its use in Aotearoa is minimal when used for analgesia when administering benzathine penicillin.

Recommended injection sites

The ventrogluteal site is recommended for intramuscular benzathine penicillin

The ventrogluteal site (outer part of the hip that sticks out, see Figure 9.1) is safer for the person.[10-12] Newly diagnosed patients should be offered a ventrogluteal site as best practice. Sides should be alternated to avoid lipohypertrophy.

The Ministry of Health has produced a video (YouTube; 9:18) that shows how to deliver a ventrogluteal injection correctly: see www.youtube.com/watch?v=lYqR-q2Lg4g.

Table 9.1. Recommended dosing of Benzathine Benzylpenicillin (Bicillin-LA) with 0.25 mL Lignocaine 2% for Strep A pharyngitis and acute rheumatic fever secondary prophylaxis

Benzathine penicillin intramuscular dose (units)*	Lignocaine 2%
<20kg 600,000 units (450 mg, 1.15 mL)	0.25 mL
≥20kg 1,200,000 units (900 mg, 2.3 mL)	0.25 mL

Ventrogluteal site

1. Place the patient in a side-lying position.
2. Using your right hand on the patient's left hip: or left hand on the patient's right hip:
   a) With the palm of your hand, locate the greater trochanter of the femur.
   b) Place your index finger towards the front or anterior superior iliac spine, and fan the middle finger as far along the iliac crest as you can reach. (The thumb should always be pointed towards the front of the leg)
3. The injection site is in the middle of the triangle between the middle and index fingers
4. Remove your fingers prior to inserting the needle.

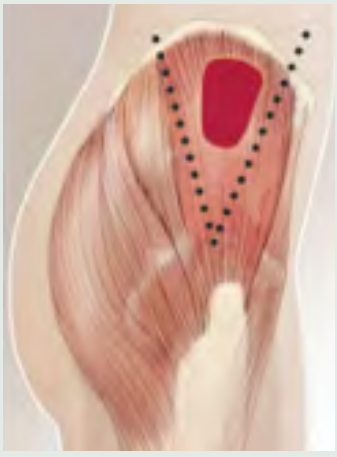


Figure 9.1. Steps to deliver an injection into the ventrogluteal site[13]

Image reproduced with permission from RHDAustralia.

The dorsogluteal (upper buttock) site can be used with caution for intramuscular benzathine penicillin injections

The dorsogluteal site (upper outer quadrant of the buttock) is another possible site. Use this site with caution: a related risk is injury to the sciatic nerve.

Dorsogluteal site

CAUTION — injections into the dorsogluteal muscle have been associated with sciatic nerve injury.

1. Place the patient in a prone (face down) position, or lying on the side. Some patients may prefer standing up. Patients with valve disease at risk of cardiac decompensation must lie down.
2. The site for injection can be identified by either:
   a) Dividing the buttock into four quadrants, selecting the upper outer quadrant;
   b) Drawing an imaginary diagonal line from the posterior superior liliac spine to the greater trochanter. From the middle of the line move up and out.

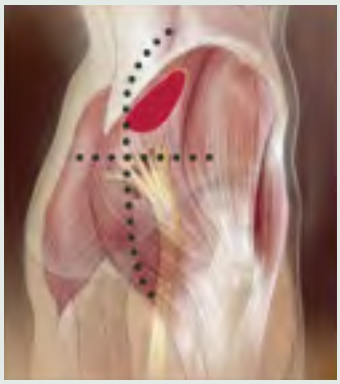


Figure 9.2. Steps to deliver an injection into the dorsogluteal site[13]

Image reproduced with permission from RHDAustralia.

The lateral thigh is acceptable for patients

The vastus lateralis (lateral thigh) is an acceptable site for an injection. Alternating sites (left and right thigh) help prevent lipohypertrophy.

Vastus Lateralis Injection site

1. Place the patient in a supine (on back) or sitting position. Patients with valve disease at risk of cardiac decompensation must lie down.
2. Place one hand on patient's thigh against greater trochanter, the other hand against the lateral femoral condyle near the knee.
3. Visualise a rectangle between the hands across the thigh.
4. The correct injection site is the middle third of the anterolateral thigh.

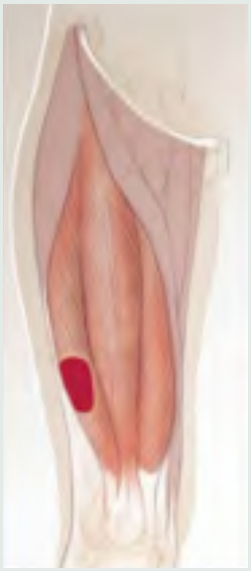


Figure 9.3. Positioning or preparing for an injection into the vastus lateralis site[13]

Image reproduced with permission from RHDAustralia.

Injection at the deltoid (upper arm) is not recommended.

Preparation before administering a Bicillin®L-A injection

Check the Standing Order, Medication Order, or Prescription is valid

- Check the medication, date, route, frequency, and dose.
- Check the medication expiry date and dose.
- Check the date Bicillin®L-A was last given.
- Check the most recent weight of the child if they were previously <20kgs.
- Check allergy status.
- Check for signed consent if under 16 years of age.

Make sure adrenaline and emergency equipment are available

- Ideally, already have the adrenaline in a syringe at the appropriate dose.

Ensure a safe environment, and ensure privacy

- **At-home injections:** Make sure another responsible person is in the home and they are capable of calling an ambulance should the person have an adverse reaction.
- **At-school injections:** Make sure an adult is present or nearby who knows what to do if the tamaiti or rangatahi has an adverse reaction.

Identify the person, gain consent, and check when any previous injection of benzathine penicillin was given

- Identify the person's full name and date of birth.
- Confirm that the person has given consent to receive Bicillin®L-A. A person under 16 years of age requires the written consent of a parent, guardian, or caregiver.
- Check the person's preferences for the injection site, distraction techniques, and analgesia.

Check the weight

For tamariki <20kgs, record the weight at each visit. If the weight has increased >20kgs, inform the specialist and ensure the higher dose is given, preferably at the same visit.

If the person is on warfarin, review their most recent international normalised ratio (INR) level

- **If the person is self-testing:** Ask them about and record their INR level.
- **If the person is under their General Practitioner (GP) or Community Pharmacy:** Check the last recorded INR level in the electronic laboratory records.

- **If the person has not had a recent INR or has stopped taking warfarin:** Administer the injection of Bicillin®L-A and notify their GP. Support the person to re-engage with their anticoagulation service provider.
- **If the person's INR is outside the recommended therapeutic range:** Follow local guidelines to notify the relevant clinician and their GP. The risk of ARF recurrence is likely to be higher than any risks they face associated with haematoma. Check with the person before administering the Bicillin®L-A injection about any history of bleeding or haematoma following injections.
- **If the INR is greater than 4.5 and still within the 28 day (or occasionally 21 day) window:** Defer the Bicillin®L-A injection, re-check the INR in 48 hours and if INR is below 4.5, then proceed with the injection. If the INR remains elevated >28 days and there are concerns for bleeding, seek specialist advice as per local guidelines.

Techniques to reduce pain when administering a Bicillin®L-A injection

Doing the initial preparation

1. Consider preparing the injection out of sight of the person.
2. Explain the injection process to the person. Allow time and do not rush the patient.
3. Check the preferred site with the person and position the person at the preferred angle for the injection. Wherever possible, positioning should accommodate the patient's preferences.
4. Check if the person wants to know when they will get the injection. Use appropriate distraction techniques (such as conversation, mobile phone, or picture book).

Using a BUZZY® device

1. If using a BUZZY® device for the first injection of Bicillin®L-A, explain how the device works. Let the person feel the vibrating device against their hand to experience the sensation. An icepack may be used with the BUZZY® device if the patient prefers.
2. Identify the site. Clean the site first with an alcohol swab, and then apply the clean BUZZY® device. Allow the site to dry before inserting the needle.
3. Leave the device on the injection site for at least **one minute** before delivering the injection.
4. When ready to deliver the injection, slide the BUZZY® device 2–5cm proximal to the injection site (along the dermatome towards the spine).
5. Leave the device in place, vibrating until the injection is complete.

Preparing and administering the injection

1. Ethyl chloride spray, an ice pack, or firm pressure at the site can be applied per local guidelines.
2. Warm the pre-filled syringe of Bicillin®L-A to room temperature by rolling it gently in the palms.
3. If using lignocaine, always use a filter needle to draw this up from a glass vial.
   - For the **1,200,000 units dose**, draw back the Bicillin®L-A plunger to allow for the addition of lignocaine. Then add the **0.25 mL of 2% lignocaine** to the syringe.
   - For the **600,000 units dose**, decant the required dose (1.15 mL) of Bicillin®L-A into a new 3 mL syringe. Then add the **0.25 mL of 2% lignocaine** into the new syringe using a 25 gauge needle.
4. Using the z-tracking technique, insert the needle and slowly deliver the Bicillin®L-A with lignocaine to the person. Give the first portion and then wait a few seconds. Then, slowly give the remaining Bicillin®L-A over 2–4 minutes. Keep checking and reassuring the person.
5. Remove the needle and activate the safety guard over the needle.
6. Dispose of the needle, syringe, and other sharp instruments safely in an appropriate bin.
7. Check the site for bleeding and apply gentle pressure if required. Cover the site with a sticking plaster if required.
8. Watch for any adverse reactions (see details in the next subsection).
9. Ask the person for a pain rating (such as using a face pain rating score) and document.
10. Advise the person about follow-up pain relief options such as taking paracetamol.

Preventing, recognising, and responding to adverse reactions

Never inject a person with a previous allergy or adverse reaction to penicillins (including Bicillin®L-A) or lignocaine unless medically cleared by a medical specialist.

All persons who administer Bicillin®L-A should be familiar with organisational recommendations around anaphylaxis management, including the administration of adrenaline.

Check for a history of adverse reactions prior to each injection.

For any person with a previous allergy or adverse reaction to Bicillin®L-A or penicillins:

- Document and report the nature of the reaction to the person's GP and ARF service for them to investigate.
- Refer to the local clinical guidelines for managing adverse reactions and anaphylaxis.
- Contact the patient's benzathine penicillin prescriber (this is usually the secondary prevention service but may be their GP or a hospital specialist) to arrange alternative SAP until the person is assessed.

Adverse reactions are uncommon; any that do occur will usually happen within 15 minutes of the person being injected. Observe the person for 15–20 minutes after delivering their first dose (the first dose they have ever had) and at least 10 minutes after subsequent doses.

Anaphylaxis signs and symptoms can include:

- Respiratory symptoms such as wheeze or stridor.
- Cardiovascular symptoms, such as tachycardia, dizziness, and prolonged hypotension.
- Skin symptoms, such as itching and welts.
- Gastrointestinal symptoms, such as abdominal cramps, nausea and vomiting.
- Neurological symptoms, such as severe anxiety, distress or confusion.

If a serious adverse reaction occurs, phone 111 and follow emergency procedures. Refer to your local protocol for managing anaphylaxis.

Rapid intramuscular administration of adrenaline is the first line for treating anaphylaxis.

Report any adverse reactions to the local ARF coordinator, the patient's GP, and medical specialist as appropriate.

Adverse reactions should also be reported to the Centre for Adverse Reactions Monitoring (CARM): **Report a Problem (Medsafe.govt.nz) Adverse Reaction Reporting (medsafe.govt.nz)**

Tasks after giving a Bicillin®L-A injection

- Follow local guidelines for documenting the administration of Bicillin®L-A /lignocaine in the patient record by adding the date, time, injection site, and other relevant health data (as set out in the local guidelines). Ensure the dose is recorded in the RFCCS.
- Give the person (or, their parent, or caregiver) follow-up information, including how to manage pain and the date of their next injection (if on SAP).
- Encourage whānau to record the next "due date" for the injection in their individualised plan or on their mobile phone.
- Re-check the safe disposal of all sharp instruments.

Storing and transporting Bicillin®L-A

Storing Bicillin®L-A

Bicillin®L-A has a shelf life of 48 months from the date of manufacture when stored at 2–8°C.

- Never freeze Bicillin®L-A. Store between 2° to 8°C.
- Store below 30°C for a single period of up to two months before its expiry date.

Transporting and returning Bicillin®L-A

- Bicillin®L-A may be stored out of the fridge in a room below 30°C for a single period of up to 2 months before its expiry date.
- Any un-used, uncontaminated pre-filled syringes (without added lignocaine) that are not used after being removed from the medication fridge (for example, if an injection is delayed or the person is not available) can be stored for up to two months in a temperate-controlled designated area below 30°C.
- Record the date on the Bicillin®L-A syringe the day it was removed from the refrigerated storage. Shelved Bicillin®L-A should be prioritised for use before refrigerated Bicillin®L-A.

Discarding Bicillin®L-A

Discard Bicillin®L-A if:

- Its temperature ever falls below 2°C or above 30°C.
- There are unused half-doses.
- There is uncertainty of its storage or efficacy.
- It has passed its expiration date.

Transporting Bicillin®L-A in a chilly bin

When transporting in a chilly bin Bicillin®L-A must stay within a temperate range of between 2°C and 30°C when being transported. A hard, chilly bin with ice packs and insulation mats is recommended to avoid freezing the Bicillin®L-A. Place icepack on the bottom, then the insulating mat, then Bicillin®L-A, then another insulating mat. The lidocaine and other equipment, such as needles and syringes can then be placed on top of the second insulating mat. Large items (such as a stethoscope or blood pressure cuff) should not be stored in the chilly bin.

References

1. Te Whatu Ora Counties Manukau. Procedure: administration of BicillinLA® for rheumatic fever prophylaxis in the community setting [30 August 2023 edition]. Auckland: Te Whatu Ora Counties Manukau; 2023.

2. Guideline statement: management of procedure-related pain in children and adolescents. *Journal of Paediatrics and Child Health*. 2006;42:S1–29. https://doi.org/10.1111/j.1440-1754.2006.00798_1.x

3. Mitchell A, Kelly J, Cook J, Atkinson N, Spain B, Remenyi B, et al. Clonidine for pain-related distress in Aboriginal children on a penicillin regimen to prevent recurrence of rheumatic fever. *Rural Remote Health*. 2020;20(4):5930. https://doi.org/10.22605/RRH5930

4. Mitchell AG, Belton S, Johnston V, Read C, Scrine C, Ralph AP. Aboriginal children and penicillin injections for rheumatic fever: how much of a problem is injection pain? *Australian and New Zealand Journal of Public Health*. 2018;42(1):46–51. https://doi.org/10.1111/1753-6405.12737

5. Russell K, Nicholson R, Naidu R. Reducing the pain of intramuscular benzathine penicillin injections in the rheumatic fever population of Counties Manukau District Health Board. *Journal of Paediatrics and Child Health*. 2014;50(2):112–117. https://doi.org/10.1111/jpc.12400

6. Cunico D, Rossi A, Verdesca M, Principi N, Esposito S. Pain Management in Children Admitted to the Emergency Room: A Narrative Review. *Pharmaceuticals (Basel, Switzerland)*. 2023;16(8). https://doi.org/10.3390/ph16081178

7. Annequin D, Carbajal R, Chauvin P, Gall O, Tourniaire B, Murat I. Fixed 50% nitrous oxide oxygen mixture for painful procedures: A French survey. *Pediatrics*. 2000;105(4):E47. https://doi.org/10.1542/peds.105.4.e47

8. Pedersen RS, Bayat A, Steen NP, Jacobsson ML. Nitrous oxide provides safe and effective analgesia for minor paediatric procedures--a systematic review. *Danish Medical Journal*. 2013;60(6):A4627.

9. Great Ormond Street Hospital for Children, NHS Foundation Trust. Pain relief using Entonox®. 2019. https://www.gosh.nhs.uk/conditions-and-treatments/procedures-and-treatments/pain-relief-using-entonox/ (Accessed December 14 2025).

10. Brown J, Gillespie M, Chard S. The dorso-ventro debate: in search of empirical evidence. *British Journal of Nursing*. 2015;24(22):1132, 1134, 1136–1139. https://doi.org/10.12968/bjon.2015.24.22.1132

11. Nicoll LH, Hesby A. Intramuscular injection: an integrative research review and guideline for evidence-based practice. *Applied Nursing Research*. 2002;15(3):149–162. https://doi.org/10.1053/apnr.2002.34142

12. Ogston-Tuck S. Intramuscular injection: exploring the evidence on effective administration. *Nursing Standard*. 2023;38(7):71–76. https://doi.org/10.7748/ns.2023.e12161

13. RHDAustralia, Menzies School of Health Research. The 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (3.2 edition). 2022. https://www.rhdaustralia.org.au/arf-rhd-guidelines (Accessed December 16 2024).

10

Diagnosis of Rheumatic Heart Disease

In this chapter

"Open heart surgery, penicillin shots until he's 30 years old, these are all things that could have been avoided if we had just taken him to the doctors at the time when he got a sore throat."

Parent of a tamaiti who was diagnosed with RHD

Key changes

This is a new chapter.

Key points

- Echocardiography (echo) is essential for diagnosing rheumatic heart disease (RHD).
- The 2023 World Heart Federation guidelines for the echocardiographic diagnosis of RHD use evidence-based standardised diagnostic criteria and staging.
- Over 40% of cases of diagnosed RHD have no previous reported history of acute rheumatic fever (ARF) in Aotearoa.
- The latent period between an episode of ARF and symptoms secondary to RHD may be long.
- RHD may be asymptomatic until cardiac decompensation occurs. Increased awareness of the spectrum of presentation and early referral for an echo are key to providing best-practice care for valvular heart disease.
- Reduced exercise tolerance or breathlessness in a pregnant person from a high-risk population should not be attributed solely to pregnancy or anaemia. Consider and investigate for RHD.

Introduction

RHD is defined as the persistent valvular changes occurring after acute or recurrent episodes of ARF, once all inflammation has subsided, with pathognomonic changes to valve leaflets and chordae resulting in pathological valve disease.[1, 2] Explaining RHD so that those living with RHD and their whānau can easily understand what it means for them is vital. Online educational resources are available, such as those by Pū Manawa.

RHD may be diagnosed:

- After the acute phase of ARF, any residual cardiac disease is then termed 'chronic RHD'.
- After clinical presentations of RHD, including those with complications of RHD.
- Following the investigation of a heart murmur.
- As an incdiental finding on echo for another reason.
- During screening programmes for RHD.

All patients with murmurs suggestive of possible valve disease or a possible history of ARF require an echo. A diagnosis of RHD is always confirmed with an echo. In Aotearoa, over 40% of cases of chronic RHD have not had a previously recognised episode of ARF.[3] The peak prevalence of RHD in Aotearoa occurs between the ages of 25 and 45 years due to the cumulative effects of episodes of ARF.[3, 4]

Acute changes of carditis are described in **Chapter 6: Diagnosis of Acute Rheumatic Fever**. In childhood, the most common RHD lesion is isolated mitral regurgitation. By adolescence and young adulthood, mixed disease involving both the aortic and mitral valves becomes the most common finding.[1, 5-7] Pacific peoples are 4.6 times and Māori 3.2 times more likely to be hospitalised for RHD than other ethnic groups in Aotearoa.[8]

For those with incidentally identified RHD, guidelines for the use of secondary prevention with benzathine penicillin are covered in **Chapter 8: Secondary Prevention** and **Chapter 11: Management of Rheumatic Heart Disease**.

Pathogenesis of rheumatic heart disease

The immunological response to ARF leads to distinctive valvular changes, which can help distinguish RHD from other valve pathologies (see **Table 10.3** and **Chapter 2: Acute Rheumatic Fever Pathogenesis**). In brief, through relative changes in collagen types and fibrosis, acute rheumatic changes can result in elongated chordae and leaflet prolapse or even acute chordal rupture, resulting in acute severe mitral regurgitation.[9-11] Annular dilatation also occurs, contributing to mitral regurgitation. Nodularity or beading of the free edges of the mitral leaflets can occur with or without chordal changes.[10] With repeated ARF episodes, progressive retraction of leaflet tips, thickening and shortening of mitral chordae occur, often affecting the posterior mitral leaflet, with secondary anterior leaflet override without true prolapse.[1, 2, 6, 12]

These changes due to an immunological response can take time to develop, highlighting the importance of follow-up and serial echo after episodes of ARF.[13] The combination of leaflet restriction and leaflet fusion leads to the commonly seen mixed valvular regurgitation and stenosis in chronic RHD.[1, 7, 14] In childhood, mitral stenosis is rare in Aotearoa. RHD can affect all four valves, but the aortic and mitral valves are almost always the most affected. The tricuspid and pulmonary valves are almost never affected without mitral involvement.[2, 15]

Clinical features of rheumatic heart disease

Features include symptoms, signs and complications. The clinical features of RHD are described in **Table 10.1**.

Table 10.1. Clinical features of rheumatic heart disease

Valve lesion	Symptoms associated with severe disease	Signs	Complications
Mitral regurgitation (MR)	May be asymptomatic despite severe mitral regurgitation Early symptoms: • Dyspnoea (breathlessness) or fatigue on exertion Advanced symptoms: • Orthopnoea (breathlessness when lying down), paroxysmal nocturnal dyspnoea (breathlessness at night) • Symptoms of right-sided heart failure if these co-exist with severe tricuspid regurgitation or pulmonary hypertension	Pan-systolic murmur, usually loudest at the apex Displaced apex beat Peripheral oedema or hepatomegaly	Left ventricular dilatation and systolic dysfunction Congestive cardiac failure Atrial arrhythmia Pulmonary hypertension
Mitral stenosis (MS)	Dyspnoea (breathlessness) or fatigue on exertion may occur with moderate or severe disease Advanced symptoms: • Orthopnoea (breathlessness when lying down), paroxysmal nocturnal dyspnoea (breathlessness at night) • Haemoptysis (coughing up blood) • Symptoms of right-sided heart failure if these co-exist with severe tricuspid regurgitation or pulmonary hypertension	Low-pitch, diastolic murmur at the apex with the patient in the left lateral position — often difficult to hear Peripheral oedema or hepatomegaly, if heart disease on the right side exists	Atrial arrhythmia Pulmonary hypertension Pulmonary haemorrhage Systemic embolism (stroke, peripheral arterial occlusion)
Mixed mitral valve disease	Symptoms as per mitral regurgitation and stenosis	Mixed systolic and diastolic murmurs Signs of right-sided heart failure in more advanced disease with pulmonary hypertension	As occurs with either mitral regurgitation or stenosis Atrial arrhythmias Pulmonary hypertension Cardiac failure
Aortic regurgitation (AR)	May be asymptomatic despite severe regurgitation Early symptoms: • Dyspnoea (breathlessness) or fatigue on exertion Advanced symptoms: • Orthopnoea (breathlessness when lying down), paroxysmal nocturnal dyspnoea (breathlessness at night) • Angina • Palpitations	Diastolic murmur is usually loudest at the left sternal edge. Diastolic flow murmur at the apex may be heard (Austin Flint murmur) Systolic murmur due to increased flow Wide pulse pressure in chronic severe regurgitation	Left ventricular dilatation and systolic dysfunction Congestive cardiac failure
Aortic stenosis (AS)	Rarely occurs in isolation, regurgitation is often present Can occur after homograft and bioprosthetic valve replacements Early symptoms: • Dyspnoea on exertion (breathlessness), pre-syncope Advanced symptoms: • Angina • Dyspnoea on exertion (breathlessness), • Orthopnoea (breathlessness when lying down), paroxysmal nocturnal dyspnoea (breathlessness at night)	Ejection systolic murmur over the aortic region, radiating to the neck Slow-rising pulse in very severe aortic stenosis	Congestive cardiac failure Left ventricular hypertrophy and preserved ejection fraction or reduced ejection fraction in advanced disease Arrhythmia
Tricuspid regurgitation (TR)	Peripheral oedema Abdominal distention and discomfort	Pan-systolic murmur at the left parasternal edge Elevated jugular venous pressure (JVP) Pulsatile enlarged liver and ascites	Right-sided heart failure Atrial arrhythmias
Tricuspid stenosis (TS)	Rarely severe and often occurs with regurgitation Fatigue Abdominal discomfort and bloating Anorexia	Soft, high-pitch diastolic murmur at the left parasternal edge, which may be difficult to hear Hepatomegaly and ascites	Right-sided heart failure Atrial arrhythmias
Multi-valve disease	Symptoms of severe valve disease can occur with two moderate lesions	Mixed systolic and diastolic murmurs Signs of cardiac failure if decompensation develops	Congestive cardiac failure Arrhythmias

Stages of valvular heart disease

Valvular heart disease of **any aetiology** may be associated with symptoms, or be asymptomatic, and can be categorised by stages A to D, as recommended in international valve management guidelines.[16]

- **Stage A** categorises individuals at risk of valvular heart disease where preventive strategies can be used.
- **Stage B** is classified as progressive disease (mild to moderate disease), where specific preventive therapies may be helpful to prevent or delay progression.
- **Stage C** is where the optimal timing of intervention is considered. The two sub-stages are asymptomatic severe valvular heart disease with compensated ventricles **(C1)** and asymptomatic severe valvular heart disease with decompensation of either ventricle **(C2)**.
- **Stage D** classification is severe symptomatic valvular disease, where intervention is generally indicated but balanced with options appropriate at an individual level.

Staging of RHD in the 2023 World Heart Federation guidelines for the echocardiographic diagnosis of RHD[14] follows a similar approach.

- **Stage A** echo features include the presence of mild MR or AR without morphological features, and the individual may or may not be at risk of valvular RHD progression.
- **Stage B** echo features include mild MR or AR and one morphological feature of RHD. If the person is over 20 years of age, two morphological features of RHD are required. Preventive therapies are recommended to prevent RHD progression.
- **Stage C** is defined by echo as moderate or severe RHD or any mitral stenosis (that is, established RHD).
- **Stage D** echo features include moderate or severe RHD, as well as clinical complications (such as heart failure, arrhythmia, embolic complications, or surgery).

These 2023 World Heart Federation (WHF) guidelines are discussed in more detail in Chapter 14: Screening for Rheumatic Heart Disease.

Many patients with RHD may report no symptoms. However, with chronic severe valvular disease, complications such as congestive cardiac failure, arrhythmias (particularly atrial fibrillation), stroke, and endocarditis may lead to an initial diagnosis of RHD.[5] Gradual symptoms of exercise intolerance (which may be mistaken for asthma), or increasing breathlessness during pregnancy may be the first signs of RHD.

Severe mitral stenosis, which often has a subtle murmur, creates stasis within the left atrium and may first present as a stroke. By the time heart failure is established and confirmed, RHD is more advanced.[16] The complications of chronic valvular heart disease contribute to the excess morbidity and mortality seen with RHD.[5]

A careful history from patients, and when appropriate with their whānau, is required to establish symptoms. Functional testing should support this history if there is uncertainty about symptom severity or if serial monitoring is needed. Multi-valve and mixed stenosis and regurgitation are a unique group, generally under-represented in valve management literature. Thresholds for follow-up and intervention are often lower for moderate lesions than for isolated valve disease. This is due to the combined impact of volume or pressure loading on the heart. Exercise testing can be helpful in providing an objective assessment of functional capacity.[16, 17]

The development of pulmonary hypertension is an important marker of reduced event-free survival.[18-20] Even in reportedly asymptomatic individuals, pulmonary hypertension is associated with a higher risk of cardiac events, and in some cases, reduced survival despite surgical correction of valvular disease. Assessment for pulmonary hypertension induced by exercise adds incremental information to help classify levels of risk.

Cultural responsiveness

Incorporating culturally responsive care at the time of diagnosis is important. Using health frameworks recommended in the hui process and Meihana Model[21] recognises the importance of whānau in assessing health. Whānau (or kaupapa whānau/non-related support people) play a key role in understanding an RHD diagnosis and supporting the diagnosed person to stay well. Including whānau is also important, as there may be concern about the health of others. This is particularly important to consider, given the higher incidence of RHD in first-degree relatives.[22] For more information about appropriate care, see **Chapter 1: Cultural Responsiveness**.

Consideration of hinengaro (thoughts and feelings) of whānau is critical when giving a diagnosis, including how whānau perceive and understand the diagnosis, as well as any potential impact on wellbeing, including stigmatisation.[23] Also, consider the impact of diagnosis on employment or sporting pursuits. Te taiao (the broader environment) also plays a key role in whānau wellbeing and is an important determinant of health. This includes opportunities to modify risk factors for ARF, such as improving housing and identifying barriers to achieving health goals, including financial or geographic barriers. Involve kaupapa Māori health providers, Pacific services, and other appropriate community services to address any gaps in care. The social determinants of health and risk for rheumatic fever are outlined in **Chapter 3: Strep A Infection, Acute Rheumatic Fever and Rheumatic Heart Disease: Risk Factors, Social Determinants of Health and Primordial Prevention**.

Understanding of health should be flexible, acknowledging the diversity of health perspectives within and between cultural groups. A culturally responsive approach can assist in identifying barriers to healthcare access, such as ensuring people can attend specialist appointments.

Echocardiographic diagnosis of rheumatic heart disease

Echo is the gold standard in establishing the presence of valvular disease.[7] (Also see **Chapter 14: Screening for Rheumatic Heart Disease**.)

Colour-Doppler criteria for mild pathological regurgitation

The colour-Doppler criteria for mild pathological regurgitation are shown in **Table 10.2**. These criteria from the 2023 World Heart Federation guidelines for the echocardiographic diagnosis of RHD,[14] are also used interchangeably as colour-Doppler criteria for acute carditis.

Table 10.2. Criteria for pathological regurgitation

Valve lesion	Criteria
Pathological mitral regurgitation (All criteria must be met)	• Observed in two views • In at least one view, MR length ≥2 cm (in individuals weighing >30 kg). In individuals weighing <30 kg MR jet length should measure ≥1.5 cm • Peak velocity ≥3.0 m/s for one complete envelope • Pan-systolic jet present in at least one envelope
Pathological aortic regurgitation (All criteria must be met)	• Observed in two views • Jet length ≥1 cm • Peak velocity ≥3.0 m/s in early diastole • Pan-diastolic jet present in at least one envelope
Mitral stenosis (All criteria must be met)	• Restricted leaflet motion with reduced valve opening • Mean peak gradient ≥4 mmHg

Rwebembera J, Marangou J, Mwita JC, Mocumbi AO, Mota C, Okello E, et al. 2023 World Heart Federation guidelines for the echocardiographic diagnosis of rheumatic heart disease. *Nature Reviews: Cardiology*. 21, 4, 250–263. 2020, reproduced with permission from SNCSC.

When assessing for mild pathological regurgitation using the WHF criteria,[2, 14] take care to align the imaging beam with the regurgitant jet, as detection of Doppler shift is directly related to the angle of incidence, with interrogation parallel to flow detecting the greatest Doppler shift.

An eccentric or wall-hugging jet may make obtaining a full Doppler envelope challenging, which may increase the risk of a severe eccentric jet producing an incomplete Doppler envelope, potentially leading to an underestimation of valvular disease.[24, 25] Operator experience in performing and reading echo can lead to variations in the inter-observer assessment.

The morphological features of rheumatic heart disease

The morphological features of RHD are shown in Table 10.3. The key features of chronic mitral RHD are:

- **Valve apparatus thickening** — thickening of leaflet edges, commissural fusion, and thickening of valve chordae.
- **Valve mobility abnormalities** — restricted leaflet motion in diastole and/or excessive leaflet motion in systole.

Both of these abnormalities lead to valve regurgitation, valve stenosis, or mixed stenosis and regurgitation.[7, 14]

The key morphological features of rheumatic aortic RHD are cusp thickening, cusp prolapse, restricted cusp motion, and a coaptation defect in diastole.[14]

A detailed description of echocardiographic interpretation for the morphological features of RHD is provided in the explanatory notes of Box 6 of the 2023 World Heart Federation guidelines for the echocardiographic diagnosis of RHD.[14]

Findings in RHD are contrasted with other valve pathologies in Table 10.3, with examples provided in Figures 10.1 to 10.6.

Table 10.3. Morphological features of rheumatic heart disease, with similarities and differences of other valve pathologies

Characteristic features of RHD	Differential valve lesion
Mitral valve morphological features Thickened leaflet tips Chordal thickening	
Leaflet prolapse (excessive leaflet motion in systole)	Floppy mitral valve prolapse Myxomatous mitral valve Ischaemic mitral regurgitation with posterior leaflet restriction from left ventricular remodelling
Restricted posterior leaflet Diastolic doming of anterior leaflet	Parachute mitral valve: • Commonly associated with additional congenital defects • Single papillary muscle (best seen in parasternal short axis)
Leaflet calcification (particularly at leaflet tips)	Mitral annular calcification extends from the annulus, usually sparing leaflet tips
Aortic valve morphological features Rolled cusp edges (central coaptation defect)	
Commissural fusion Cusp prolapse Cusp thickening	Bicuspid aortic valve (or congenital aortic valve): • Systolic doming of leaflets • Aortic regurgitation or stenosis • Bicuspid aortic valve confirmed in short axis view Aortic valve endocarditis Aortic root abscess may be associated with AV block on electrocardiogram (ECG)
Cusp calcification Dilated aortic root	Degenerative aortic valve disease Connective tissue disorders, bicuspid aortic valve
Tricuspid valve morphological features Leaflet thickening, calcification Leaflet restriction, retraction Chordal shortening	Carcinoid: • Diffuse thickening of tricuspid leaflets, with restriction and central coaptation defect Secondary tricuspid regurgitation: • Normal tricuspid leaflets, with defects in coaptation. These defects occur secondary to the annular dilatation or right ventricular remodelling in the presence of left heart disease

Secondary or functional mitral and tricuspid regurgitation occurs due to left or right ventricular remodelling, with displacement of the papillary muscles and with annular dilatation as the mechanism for regurgitation, but with normal valve leaflets. Primary rheumatic involvement of the tricuspid valve with the features shown in Table 10.3 can also occur. This distinction is important in planning valve surgery. Dedicated echo imaging to assess both valvular and sub-valvular involvement in mitral and tricuspid disease is important when considering valve repair. Additional off-axis imaging can help interrogate primary and secondary chordae and their role in valve dysfunction.

Diagnosing mild, moderate, or severe valvular heart disease

The quantification of valve lesion severity and important caveats with interpretation are well described in international valve guidelines and Table 10.4.[16, 17, 24, 25] Accurately defining severity is important for determining appropriate follow-up and management. This ensures timely medical and surgical intervention when needed, reducing the morbidity and mortality associated with RHD.

Table 10.4 lists the echo parameters used to diagnose mild, moderate, or severe RHD for six forms of valvular heart disease: mitral stenosis, mitral regurgitation, aortic stenosis, aortic regurgitation, tricuspid regurgitation, and tricuspid stenosis. The table is a synopsis of international guidelines for the quantification of valvular disease severity. Clinical events and mortality increase as disease severity increases from moderate to very severe.[26, 27]

Table 10.4. Echocardiographic parameters for diagnosing the severity of rheumatic heart disease

	Mild	Moderate	Severe
Mitral stenosis			
MV area (cm^2)	2.0–2.5	1.5–2.0	<1.5
MV mean gradient (mmHg)	4	5–10 *	>10
Mitral regurgitation			
MR EROA (cm^2)	<0.2	0.2–0.3	>0.4
MR vena contracta (cm)	<0.3	0.3–0.7	>0.7
MR regurgitant volume (mL)	<40	40–60	>60
Aortic stenosis			
AV area (cm^2)	1.5–2.0	1.0–1.5**	<1.0**
AV mean gradient (mmHg)	<20	20–39	>40
AV peak velocity (m/s)	2.0–2.9	3.0–3.9	>4
Aortic regurgitation			
AR ERO (cm^2)		0.1–0.29	>0.3
AR vena contracta (cm)	<0.3	0.3–0.6	>0.6
Aortic flow reversal			Holodiastolic flow reversal in descending aorta
Tricuspid regurgitation			
TR vena contracta (cm)	<0.3	<0.6	0.7
TR ERO (cm^2)	<20	20–39	>40
Hepatic vein flow	Systolic dominance	Systolic blunting	Systolic reversal
Tricuspid stenosis			
Mean gradient (mmHg)		>5 is considered significant	

* Low-gradient mitral stenosis may occur with low stroke volume, atrial fibrillation or decreased left ventricular compliance. High-gradient mitral stenosis may occur with co-existing mitral regurgitation or increased cardiac output.

** AV area may be increased with co-existing aortic regurgitation or decreased with low cardiac output.

Mixed and multi-valve rheumatic heart disease

Mixed valve disease occurs when a rheumatic valve has both regurgitation and stenosis. The presence of both can make echo assessment more complex. This complexity increases further when multi-valve disease is present. Table 10.5 describes the echo parameters for assessing mixed-valve and multi-valve disease.

Table 10.5. Echocardiographic assessment of mixed-valve and multi-valve disease

Echo parameter	Mixed valve	Mixed multi-valve
Area by pressure half-time	Unreliable in the presence of more than mild regurgitation	Unreliable in the presence of more than mild regurgitation
Area by planimetry	Consider 3D planimetry Independent of stroke volume	Reliable when performed by skilled operators Independent of stroke volume
Valve area by the continuity equation	Area increases with increasing stroke volume	Unreliable with mixed mitral valve disease and significant mitral regurgitation
Gradient *Important:* Consider low-gradient aortic and mitral stenosis in cases of multi-valve disease, low cardiac output, and atrial fibrillation	Peak velocity and gradient reflect the severity of mixed regurgitation and stenosis	Gradient increased in the setting of severe mitral regurgitation
Proximal isovelocity surface area (PISA) assessment of regurgitant lesion	Reliable	Reliable

Maintaining closer follow-up is vital in the presence of multi-valve or mixed-valve disease. The severity of valve disease determines the level of follow-up. See Chapter 11: Management of Rheumatic Heart Disease for the details of specific RHD valve lesions. There can be haemodynamic impacts of downstream valve disease. For example, co-existing aortic stenosis may result in hypertrophy and limit left ventricular dilatation of co-existing aortic or mitral regurgitation.

The usual cut-offs for severe aortic stenosis (peak velocity >4m/s and mean gradient >40mmHg) remain prognostically important, even if an aortic valve area falls into the moderate range due to co-existing regurgitation. Similarly, the impact of left atrial pressure on moderate mixed mitral stenosis and regurgitation may lead to earlier symptom onset and earlier progression to pulmonary hypertension thresholds than in isolated valve lesions.[28, 29]

Reduced peak left atrial strain on echo predicts clinical events of hospitalisations, atrial fibrillation, thromboembolic events, and the need for valve intervention at three years in asymptomatic severe mitral stenosis.[30]

Right ventricular (RV) pressure and volume overload are associated with greater myocardial fibrosis than volume overload alone from tricuspid regurgitation. Among the quantitative assessments of RV function, longitudinal strain demonstrated greater prognostic value compared with RV fractional area change and tricuspid annulus plane systolic excursion (TAPSE).[13]

Mitral valve area by planimetry is preferred in the setting of haemodynamically important aortic valve disease or mitral regurgitation, as these conditions limit the use of pressure half-time and continuity equation methods in valve area calculations. Assessment by planimetry is also independent of loading conditions and heart rate. Be aware that the criteria for clinically significant, or severe mitral stenosis in current valve management guidelines use a mitral valve area of <1.5cm^2. This area differs from some older echo texts, which defined the threshold for severe mitral stenosis as <1.0cm^2.

Investigating rheumatic heart disease

While echo is the definitive diagnostic test, additional investigations can assist in the clinical assessment of RHD, including chest X-rays and ECG. An ECG can show signs of cardiac remodelling due to severe RHD, such as left atrial or ventricular enlargement or right axis deviation and signs of right ventricular enlargement. A chest X-ray may show signs of cardiomegaly, left atrial dilatation, or signs of heart failure.

Multi-modality imaging in rheumatic heart disease

Given the complexities of multi-valve and mixed valve disease, additional imaging and supplementary assessment may be needed through:[16, 17]

- Echo global longitudinal strain.
- 3D left ventricular volume assessment.
- Functional testing with stress echo.
- Transoesophageal echo for 3D valve area assessment.
- Cardiac MRI.
- Biomarkers such as B-type natriuretic peptide (BNP).
- Right heart catheterisation.

B-type natriuretic peptide

BNP is a natriuretic peptide released from the heart in response to high ventricular filling pressures. Elevated BNP levels are predictive of symptom development and subclinical dysfunction in aortic valve disease and mitral regurgitation.[31, 32] BNP testing can be requested through community pathology services and may be helpful alongside chest X-ray and ECG while awaiting echo in the community setting.

BNP plays a role in diuresis and inhibits the renin-angiotensin system. It also has a role in endothelin secretion and sympathetic activity. These mechanisms contribute to increased urine output, dilatation of blood vessels, and reduced cardiac workload.[31] Elevated BNP levels have been described in isolated moderate to severe mitral stenosis, correlating with symptoms and cardiac remodelling.[32]

Echo for assessing suitability for percutaneous balloon mitral valvuloplasty

Percutaneous balloon mitral valvuloplasty (PBMV) is the preferred intervention for mitral stenosis if the anatomy is suitable. Comprehensive echo imaging of the anatomy and function of the mitral valve is required. PBMV may be considered if the mitral regurgitation is no more than mild.

The most widely recognised echo scoring system is the Wilkins Score,[33] with leaflet mobility and thickening, degree of subvalvular involvement and calcification, each contributing to the score. The Nunes and Sutaria score may also be used, focusing more on commissural calcification and the risk of procedure-related mitral regurgitation.[34, 35]

Left atrial thrombus is a contraindication to PBMV and may be present in the body of the left atrium or atrial appendage in cases of severe mitral stenosis.

Table 10.3 contrasts the findings of RHD with other valve pathologies. Figures 10.1 to 10.6 provide visual examples of RHD:

- Non-rheumatic mitral valve showing posterior leaflet prolapse.
- Rheumatic mitral valve showing pseudo-prolapse of the anterior mitral leaflet secondary to the posterior leaflet restriction.
- Rheumatic tricuspid valve showing thickening of leaflet tips and chordae.
- Carcinoid tricuspid valve disease showing diffuse thickening of leaflets.
- Rheumatic aortic valve showing rolled leaflet edges and a central coaptation.
- Rheumatic aortic valve showing cusp prolapse.

Recommendations for follow-up and management of rheumatic heart disease are detailed in Chapter 11: Management of Rheumatic Heart Disease and published international guidelines.[16, 17]

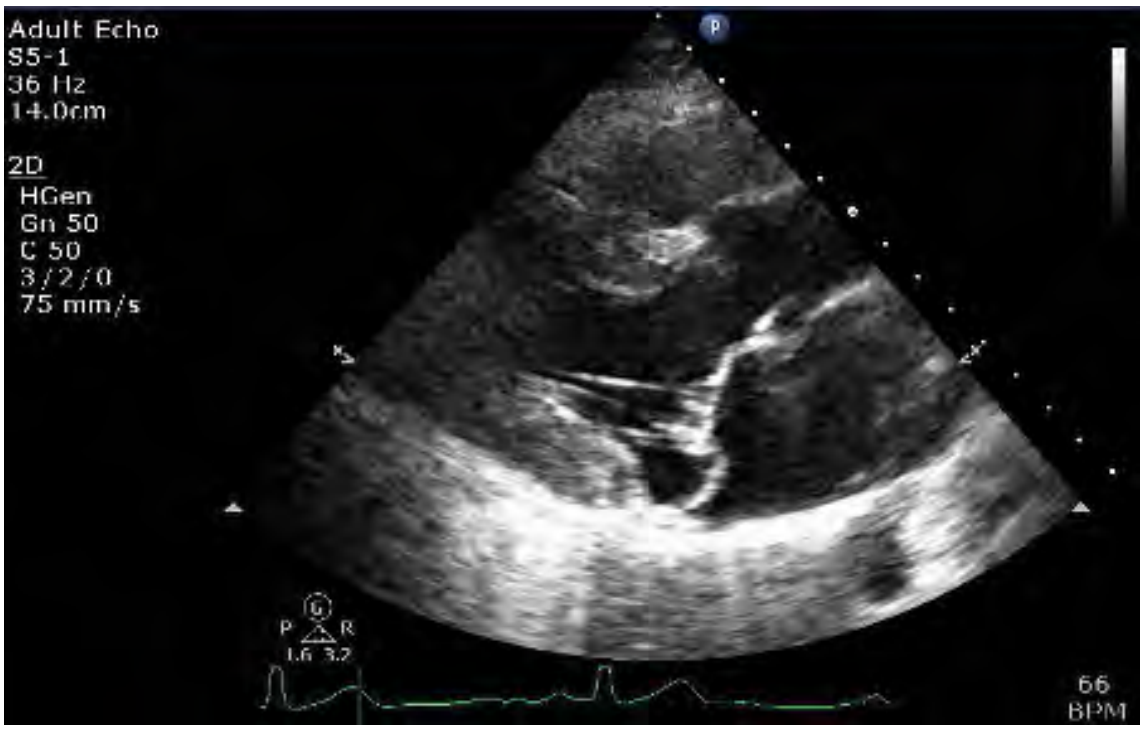


Figure 10.1. Non-rheumatic mitral valve showing posterior leaflet prolapse

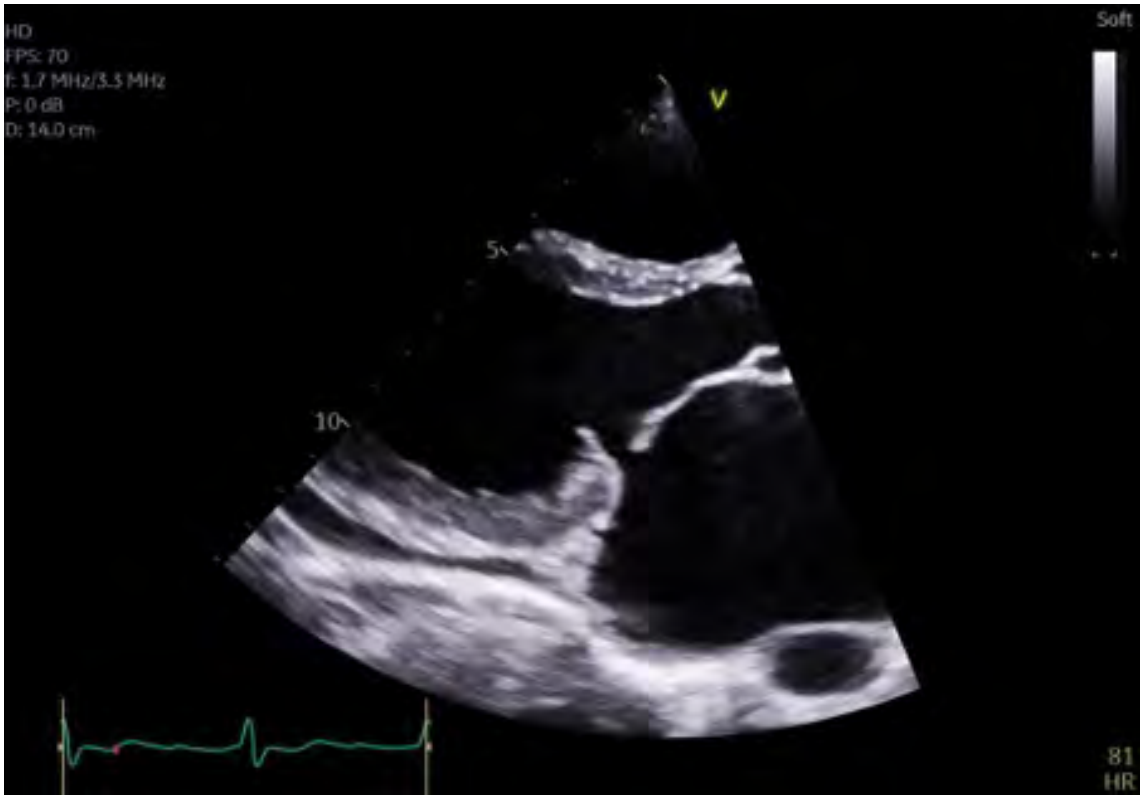


Figure 10.2. Rheumatic mitral valve showing pseudo-prolapse of the anterior mitral leaflet secondary to the posterior leaflet restriction

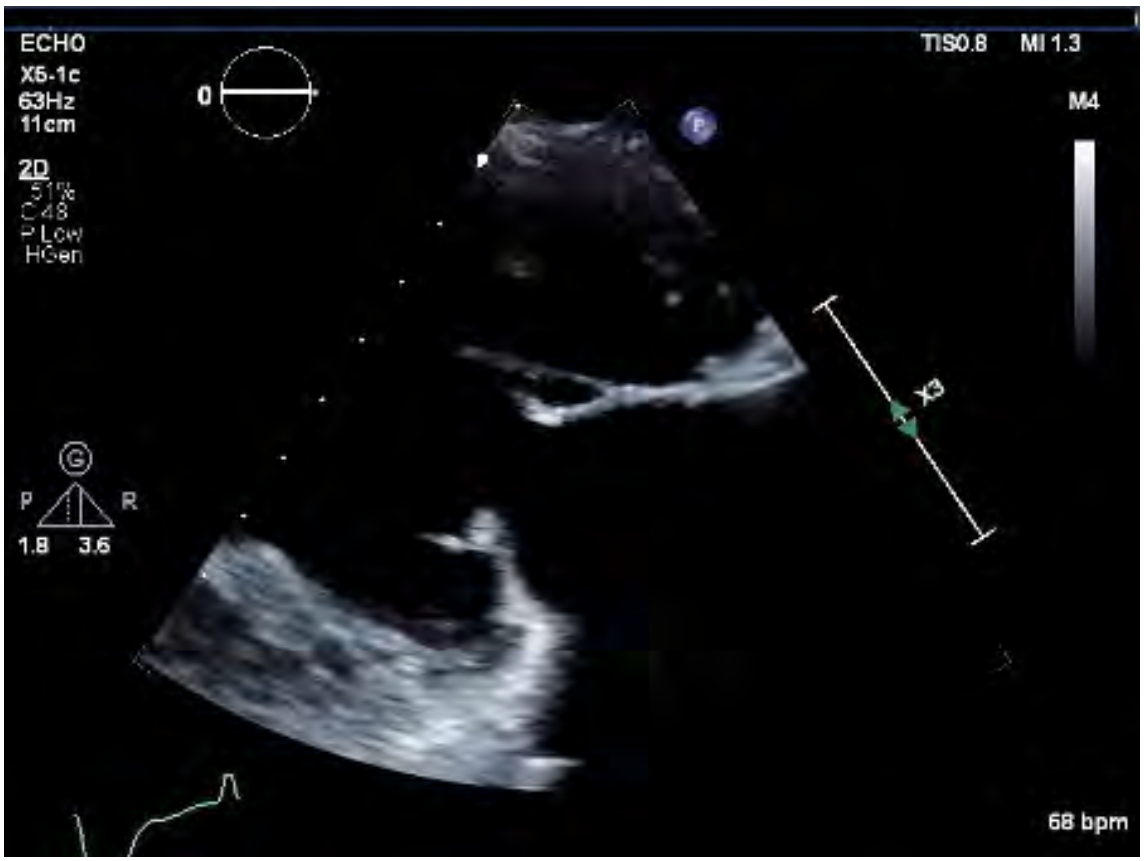


Figure 10.3. Rheumatic tricuspid valve showing thickening of leaflet tips and chordae

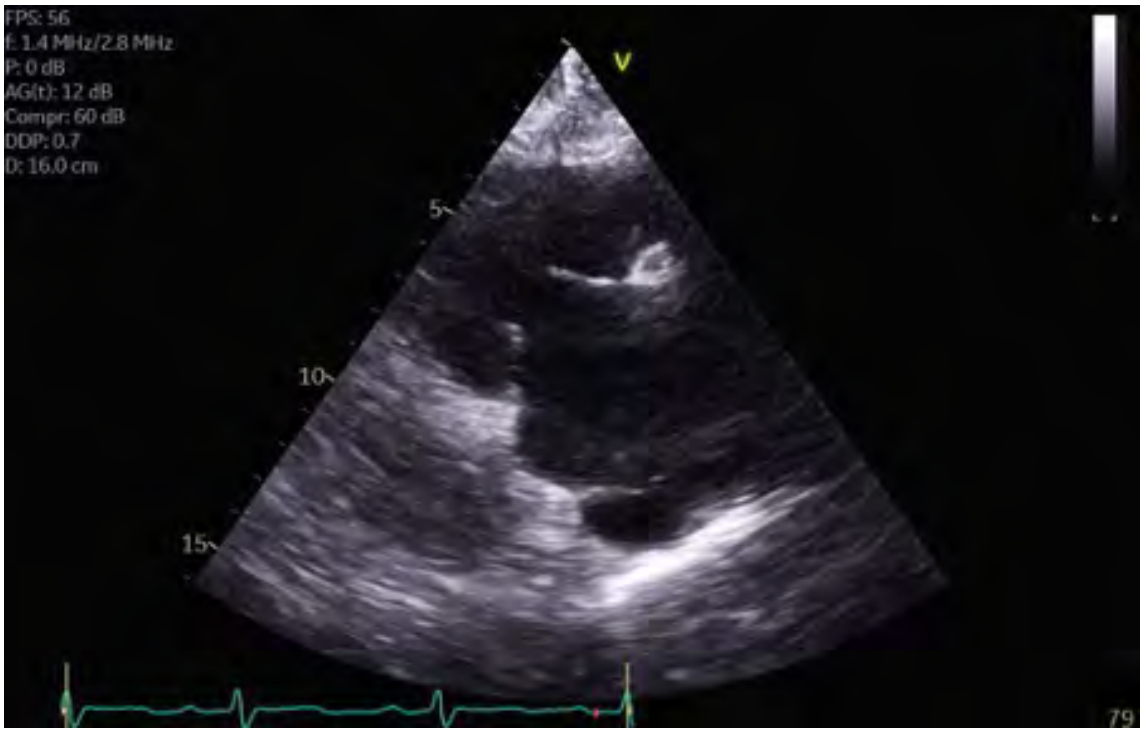


Figure 10.4. Carcinoid tricuspid valve disease showing diffuse thickening of leaflets

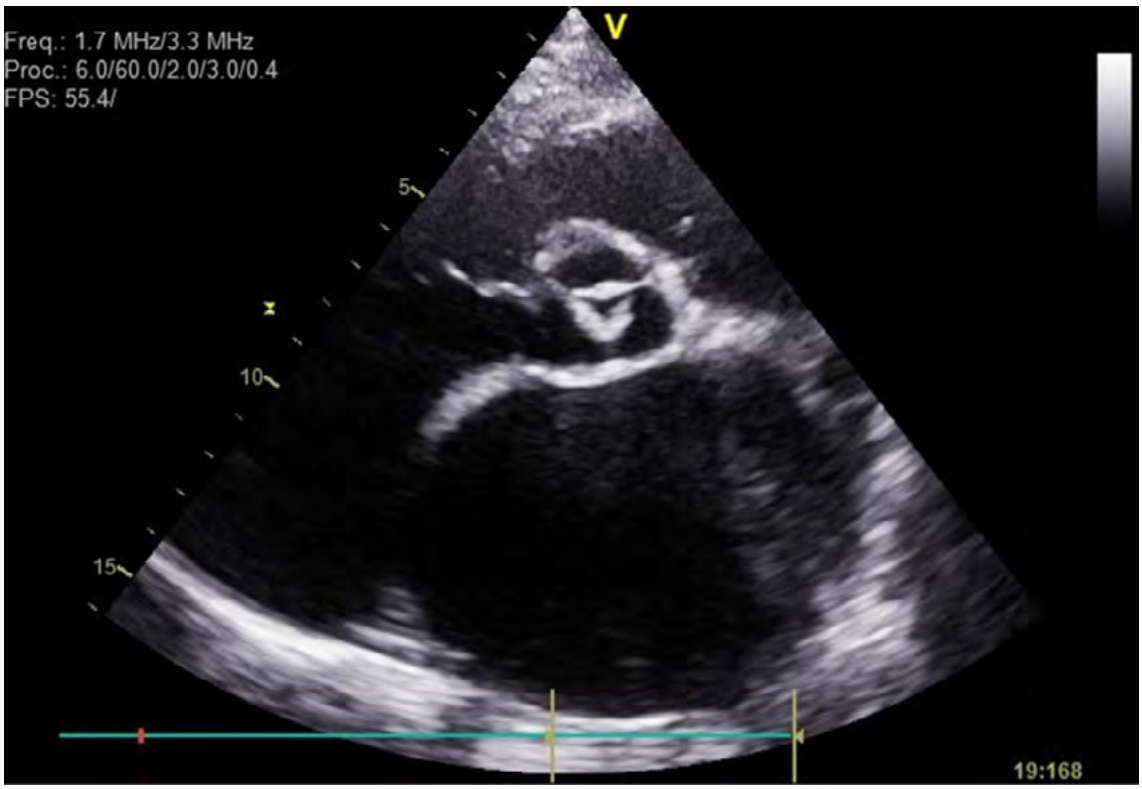


Figure 10.5. Rheumatic aortic valve showing rolled leaflet edges and a central coaptation defect

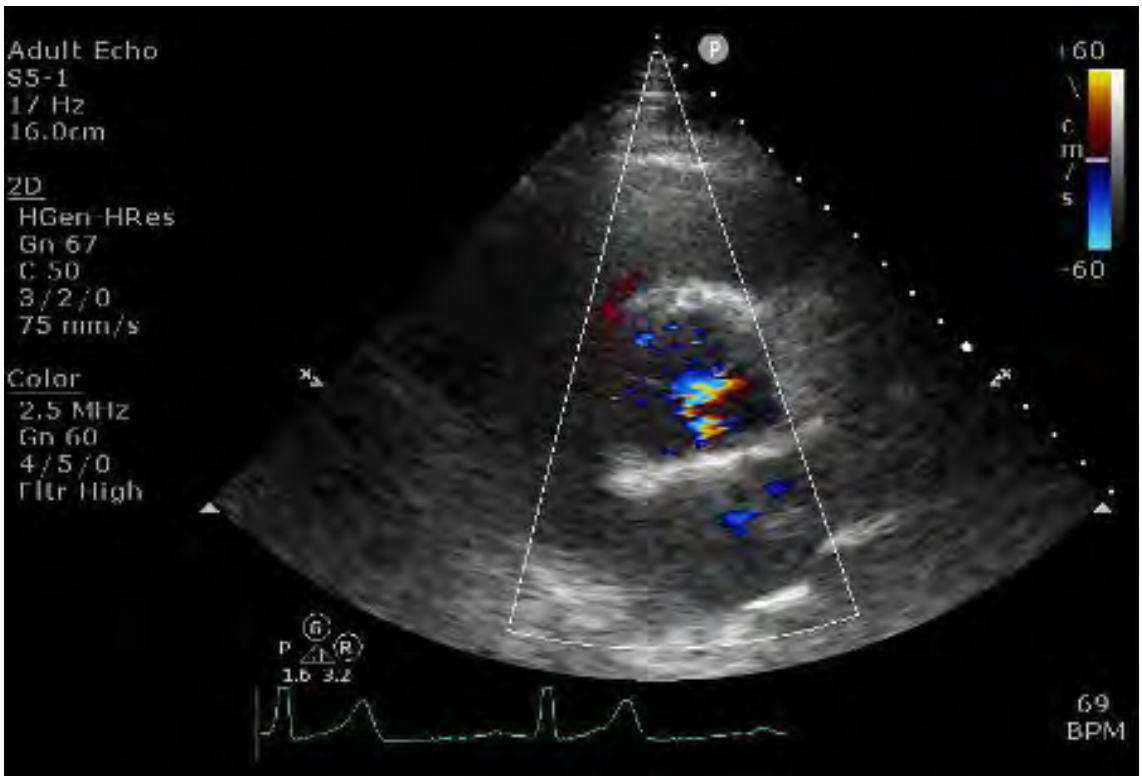


Figure 10.6. Rheumatic aortic valve showing cusp prolapse

Echocardiography after cardiac surgery

A baseline echo should be performed within three months of surgery, providing a reference for future follow-up. At this time, postoperative assessment of valve regurgitation may be more challenging due to acoustic shadowing. Indirect findings on haemodynamics from transthoracic echo may indicate the need for further transoesophageal echo.

Assessment of mechanical and bioprosthetic valve replacements

The previous operation report is useful for knowledge of the prosthesis type and size and will help with interpreting haemodynamics. If unavailable, the operation report should be sought from the surgical centre. Normal echo parameters for varying prostheses are available in published guidelines.[36] It is important to monitor for structural valve degeneration (SVD) in stented and stentless bioprostheses.

The presence of leaflet thickening or calcification may be:

- The first sign of SVD.
- An indication for early repeat imaging and follow-up.

Monitor for SVD more often in RHD than compared with monitoring in the elderly.[37]

As with native valve disease, gradients are influenced by flow, as well as the size and position of the valve. Low cardiac output will result in lower gradients. Mitral valve prosthesis assessment should always report the heart rate. Comparing the effective orifice area (EOA) to published normal values and using a dimensionless index will provide flow-independent assessments.

The dimensionless index for an aortic prosthesis is calculated using the ratio of the left ventricular outflow tract (LVOT) velocity time interval (VTI) to the aortic valve: LVOT VTI/AV VTI. A valve of <0.25 is suggestive of significant stenosis and >0.35 is considered normal. For a mitral prosthesis, the dimensionless index is calculated by LVOT VTI/MV VTI, with a value >2.5 suggestive of significant stenosis and <2.2 normal.[36] Paediatric echo labs primarily use peak and mean gradients when assessing prosthetic valves.

Specific guidelines exist for assessing percutaneous valves,[36] which differ from native valve disease in their mechanisms of paravalvular regurgitation.

Assessment of tricuspid valve replacements assessment

Tricuspid prostheses should also undergo a full assessment with peak velocity, mean gradient, EOA and dimensionless index. Freedom from bioprosthetic tricuspid valve dysfunction or mechanical valve thrombosis is significantly lower at five years compared to left-sided prostheses.[36]

Distinguishing patient-prosthesis mismatch from valve dysfunction

Using multiple haemodynamic parameters on transthoracic echo, with the addition of transoesophageal echo and cardiac CT, can help distinguish patient-prosthesis mismatch from valve dysfunction. Both transoesophageal echo and cardiac CT may help distinguish bioprosthetic or mechanical valve thrombus from pannus.

Additionally, cardiac PET plays a role in evaluating prosthetic valve endocarditis.

References

1. Horton A, Gentles T, Reményi B. Clinical evaluation and diagnosis of rheumatic heart disease. In: Dougherty JCS, Zühlke L, Wilson N, editors. Acute rheumatic fever and rheumatic heart disease: Elsevier; 2021. p. 69–106.

2. Rémenyi B, Wilson N, Steer A, Ferreira B, Kado J, Kumar K, et al. World Heart Federation criteria for echocardiographic diagnosis of rheumatic heart disease--an evidence-based guideline. *Nature Reviews: Cardiology*. 2012;9(5):297–309. https://doi.org/10.1038/nrcardio.2012.7

3. Tilton E, Mitchelson B, Anderson A, Peat B, Jack S, Lund M, et al. Cohort profile: methodology and cohort characteristics of the Aotearoa New Zealand Rheumatic Heart Disease Registry. *BMJ Open*. 2022;12(12):e066232. https://doi.org/10.1136/bmjopen-2022-066232

4. Dougherty S, Okello E, Mwangi J, Kumar RK. Rheumatic heart disease: JACC focus seminar 2/4. *Journal of the American College of Cardiology*. 2023;81(1):81–94. https://doi.org/10.1016/j.jacc.2022.09.050

5. Zuhlke L, Engel ME, Karthikeyan G, Rangarajan S, Mackie P, Cupido B, et al. Characteristics, complications, and gaps in evidence-based interventions in rheumatic heart disease: the Global Rheumatic Heart Disease Registry (the REMEDY study). *European Heart Journal*. 2015;36(18):1115–1122a. https://doi.org/10.1093/eurheartj/ehu449

6. RHDAustralia, Menzies School of Health Research. The 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (3.2 edition). 2022. https://www.rhdaustralia.org.au/arf-rhd-guidelines (Accessed December 16 2024).

7. Pandian NG, Kim JK, Arias-Godinez JA, Marx GR, Michelena HI, Chander Mohan J, et al. Recommendations for the use of echocardiography in the evaluation of rheumatic heart disease: a report from the American Society of Echocardiography. *Journal of the American Society of Echocardiography*. 2023;36(1):3–28. https://doi.org/10.1016/j.echo.2022.10.009

8. Bennett J, Zhang J, Leung W, Jack S, Oliver J, Webb R, et al. Rising ethnic inequalities in acute rheumatic fever and rheumatic heart disease, New Zealand, 2000-2018. *Emerging Infectious Diseases*. 2021;27(1):36–46. https://doi.org/10.3201/eid2701.191791

9. Lis Y, Burleigh MC, Parker DJ, Child AH, Hogg J, Davies MJ. Biochemical characterization of individual normal, floppy and rheumatic human mitral valves. *Biochemical Journal*. 1987;244(3):597–603. https://doi.org/10.1042/bj2440597

10. Vasan RS, Shrivastava S, Vijayakumar M, Narang R, Lister BC, Narula J. Echocardiographic evaluation of patients with acute rheumatic fever and rheumatic carditis. *Circulation*. 1996;94(1):73–82. https://doi.org/10.1161/01.cir.94.1.73

11. Marcus RH, Sareli P, Pocock WA, Meyer TE, Magalhaes MP, Grieve T, et al. Functional anatomy of severe mitral regurgitation in active rheumatic carditis. *American Journal of Cardiology*. 1989;63(9):577–584. https://doi.org/10.1016/0002-9149(89)90902-8

12. Câmara EJ, Neubauer C, Camara GF, Lopes AA. Mechanisms of mitral valvar insufficiency in children and adolescents with severe rheumatic heart disease: an echocardiographic study with clinical and epidemiological correlations. *Cardiology in the Young*. 2004;14(5):527–532. https://doi.org/10.1017/S1047951104005104

13. He VY, Condon JR, Ralph AP, Zhao Y, Roberts K, de Dassel JL, et al. Long-term outcomes from acute rheumatic fever and rheumatic heart disease: a data-linkage and survival analysis approach. *Circulation*. 2016;134(3):222–232. https://doi.org/10.1161/CIRCULATIONAHA.115.020966

14. Rwebembera J, Marangou J, Mwita JC, Mocumbi AO, Mota C, Okello E, et al. 2023 World Heart Federation guidelines for the echocardiographic diagnosis of rheumatic heart disease. *Nature Reviews: Cardiology*. 2024;21(4):250–263. https://doi.org/10.1038/s41569-023-00940-9

15. Sultan FA, Moustafa SE, Tajik J, Warsame T, Emani U, Alharthi M, et al. Rheumatic tricuspid valve disease: an evidence-based systematic overview. *Journal of Heart Valve Disease*. 2010;19(3):374–382.

16. Otto CM, Nishimura RA, Bonow RO, Carabello BA, Erwin JP, 3rd, Gentile F, et al. 2020 ACC/AHA guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Joint Committee on clinical practice guidelines. *Circulation*. 2021;143(5):e72–e227. https://pubmed.ncbi.nlm.nih.gov/33332149/

17. Vahanian A, Beyersdorf F, Praz F, Milojevic M, Baldus S, Bauersachs J, et al. 2021 ESC/EACTS Guidelines for the management of valvular heart disease: Developed by the Task Force for the management of valvular heart disease of the European Society of Cardiology (ESC) and the European Association for Cardio-Thoracic Surgery (EACTS). *European Heart Journal*. 2021;43(7):561–632. https://doi.org/10.1093/eurheartj/ehab395

18. Barbieri A, Bursi F, Grigioni F, Tribouilloy C, Avierinos JF, Michelena HI, et al. Prognostic and therapeutic implications of pulmonary hypertension complicating degenerative mitral regurgitation due to flail leaflet: a multicenter long-term international study. *European Heart Journal*. 2011;32(6):751–759. https://doi.org/10.1093/eurheartj/ehq294

19. Magne J, Pibarot P, Sengupta PP, Donal E, Rosenhek R, Lancellotti P. Pulmonary hypertension in valvular disease: a comprehensive review on pathophysiology to therapy from the HAVEC Group. *JACC Cardiovascular Imaging*. 2015;8(1):83–99. https://doi.org/10.1016/j.jcmg.2014.12.003

20. Bermejo J, Gonzalez-Mansilla A, Mombiela T, Fernandez AI, Martinez-Legazpi P, Yotti R, et al. Persistent pulmonary hypertension in corrected valvular heart disease: hemodynamic insights and long-term survival. *Journal of the American Heart Association*. 2021;10(2):e019949. https://doi.org/10.1161/JAHA.120.019949

21. Pitama S, Huria T, Lacey C. Improving Māori health through clinical assessment: waikare o te waka o Meihana. *New Zealand Medical Journal*. 2014;127(1393):107–119.

22. Culliford-Semmens N, Tilton E, Wilson N, Stirling J, Doughty R, Gentles T, et al. Echocardiography for latent rheumatic heart disease in first degree relatives of children with acute rheumatic fever: implications for active case finding in family members. *eClinicalMedicine*. 2021;37:100935. https://doi.org/10.1016/j.eclinm.2021.100935

23. Anderson A, Spray J. Beyond awareness: Towards a critically conscious health promotion for rheumatic fever in Aotearoa, New Zealand. *Social Science and Medicine*. 2020;247:112798. https://doi.org/10.1016/j.socscimed.2020.112798

24. Quinones MA, Otto CM, Stoddard M, Waggoner A, Zoghbi WA. Recommendations for quantification of Doppler echocardiography: a report from the Doppler Quantification Task Force of the Nomenclature and Standards Committee of the American Society of Echocardiography. *Journal of the American Society of Echocardiography*. 2002;15(2):167–184. https://doi.org/10.1067/mje.2002.120202

25. Bolger AF, Eidenvall L, Ask P, Loyd D, Wranne B. Understanding continuous-wave Doppler signal intensity as a measure of regurgitant severity. *Journal of the American Society of Echocardiography*. 1997;10(6):613–622. https://doi.org/10.1016/s0894-7317(97)70024-5

26. Antoine C, Benfari G, Michelena HI, Maalouf JF, Nkomo VT, Thapa P, et al. Clinical outcome of degenerative mitral regurgitation: critical importance of echocardiographic quantitative assessment in routine practice. *Circulation*. 2018;138(13):1317–1326. https://doi.org/10.1161/CIRCULATIONAHA.117.033173

27. Capoulade R, Le Ven F, Clavel MA, Dumesnil JG, Dahou A, Thebault C, et al. Echocardiographic predictors of outcomes in adults with aortic stenosis. *Heart*. 2016;102(12):934–942. https://doi.org/10.1136/heartjnl-2015-308742

28. Gentles TL, Colan SD, Wilson NJ, Biosa R, Neutze JM. Left ventricular mechanics during and after acute rheumatic fever: contractile dysfunction is closely related to valve regurgitation. *Journal of the American College of Cardiology*. 2001;37(1):201–207. https://doi.org/10.1016/s0735-1097(00)01058-5

29. Unger P, Pibarot P, Tribouilloy C, Lancellotti P, Maisano F, Iung B. Multiple and mixed valvular heart diseases: pathology, imaging, and management. *Circulation: Cardiovascular Imaging*. 2018;11(8). https://doi.org/10.1161/CIRCIMAGING.118.007862

30. Caso P, Ancona R, Di Salvo G, Comenale Pinto S, Macrino M, Di Palma V, et al. Atrial reservoir function by strain rate imaging in asymptomatic mitral stenosis: prognostic value at 3 year follow-up. *European Journal of Echocardiography*. 2009;10(6):753–759. https://doi.org/10.1093/ejechocard/jep058

31. Brunner-La Rocca HP, Kaye DM, Woods RL, Hastings J, Esler MD. Effects of intravenous brain natriuretic peptide on regional sympathetic activity in patients with chronic heart failure as compared with healthy control subjects. *Journal of the American College of Cardiology*. 2001;37(5):1221–1227. https://doi.org/10.1016/s0735-1097(01)01172-x

32. Sharma V, Stewart RA, Zeng I, Raffel C, Kerr AJ. Comparison of atrial and brain natriuretic peptide for the assessment of mitral stenosis. *Heart, Lung & Circulation*. 2011;20(8):517–524. https://doi.org/10.1016/j.hlc.2011.03.112

33. Wilkins GT, Weyman AE, Abascal VM, Block PC, Palacios IF. Percutaneous balloon dilatation of the mitral valve: an analysis of echocardiographic variables related to outcome and the mechanism of dilatation. *British Heart Journal*. 1988;60(4):299–308. https://doi.org/10.1136/hrt.60.4.299

34. Nobuyoshi M, Arita T, Shirai S, Hamasaki N, Yokoi H, Iwabuchi M, et al. Percutaneous balloon mitral valvuloplasty: a review. *Circulation*. 2009;119(8):e211–219. https://doi.org/10.1161/CIRCULATIONAHA.108.792952

35. Silbiger JJ. Advances in rheumatic mitral stenosis: echocardiographic, pathophysiologic, and hemodynamic considerations. *Journal of the American Society of Echocardiography*. 2021;34(7):709–722.e701. https://doi.org/10.1016/j.echo.2021.02.015

36. Zoghbi WA, Jone PN, Chamsi-Pasha MA, Chen T, Collins KA, Desai MY, et al. Guidelines for the evaluation of prosthetic valve function with cardiovascular imaging: a report from the American Society of Echocardiography developed in collaboration with the Society for Cardiovascular Magnetic Resonance and the Society of Cardiovascular Computed Tomography. *Journal of the American Society of Echocardiography*. 2024;37(1):2–63. https://doi.org/10.1016/j.echo.2023.10.004

37. Yang B, Malik A, Farhat L, Makkinejad A, Norton EL, Sareini MA, et al. Influence of age on longevity of a stentless aortic valve. *Annals of Thoracic Surgery*. 2020;110(2):500–507. https://doi.org/10.1016/j.athoracsur.2019.10.085

11

Management of Rheumatic Heart Disease

In this chapter

"My boy was so healthy six months ago, then one sore throat... and then he had to go through open heart surgery. I thought we were going to lose him. It's so unfair."

Parent of a tamaiti who underwent RHD surgery

Key changes

Group	Changes	Recommendation grading
High-risk category	The follow-up plan for bioprosthetic mitral valves has been separated from mechanical prosthetic valves. The recommended follow-up has been changed to every 6 months (previously every 6 to 12 months for bioprosthetic valves).	Grade C
Moderate rheumatic heart disease (RHD)	Follow-up for isolated, stable, moderate mitral or aortic regurgitation has been changed to every 2 years (previously every 2–3 years).	Grade D
Moderate mitral stenosis	Follow-up has been changed to annually if the mitral valve area (MVA) is <1.5cm^2 (previously every 2–3 years), as this is classified as severe in valve management guidelines.	Grade C
Mixed moderate RHD	Treat as severe RHD. Annual follow-up is recommended.	Grade D

The guidelines now include:

- A discussion on equity of care for RHD patients.
- A review and care plan based on the risk and severity of the individual cardiac diagnosis.
- A section on transitioning from paediatric to adult cardiac services.
- A section on best-practice discharge after cardiac surgery.
- A section on the complications of RHD.
- A section on managing RHD in primary care.

Key points

Table 11.1. Best practice rheumatic heart disease management principles

Best practice rheumatic heart disease management principles
1. The fundamental goals in the long-term management of RHD are: • To support patients and their whānau on their RHD journey and provide culturally safe care. • To recognise and understand the lived experience of individuals with RHD.
2. Provide access to a physician experienced in RHD for mild disease and a cardiologist for moderate to severe disease.
3. Ensure access to timely echocardiography (echo).
4. Manage secondary prevention with penicillin prophylaxis (see **Chapter 8: Secondary Prevention**) and ensure patient referral and registration to the national Rheumatic Fever Care Coordination System.
5. Refer patients for consideration of heart valve intervention in a timely manner, following best international practice guidelines.
6. Provide effective systems of care to support anticoagulation therapy in patients with atrial fibrillation (AF) and/or mechanical prosthetic valves.
7. Optimise oral health with regular dental reviews.
8. Offer annual influenza vaccination.
9. Implement strategies to prevent infective endocarditis by reducing the risk from oral microbes. Refer to a dentist with some urgency.
10. Provide individualised discussions with appropriate specialists to support informed decision making regarding pregnancy.
11. Increase awareness and encourage early recognition of RHD complications.
12. First-degree relatives of newly diagnosed RHD cases should undergo echocardiographic screening for RHD. See **Chapter 14: Screening for Rheumatic Heart Disease**.

Table adapted from Okello et al and page 108 of the New Zealand Guidelines for the Diagnosis, Management and Secondary Prevention of Acute Rheumatic Fever and Rheumatic Heart Disease: 2014 Update second edition.

Table 11.2 summarises the frequency of specialist reviews and echo. Rationale and explanations appear as footnotes.

Table 11.2. Review and care plan by severity of rheumatic heart disease diagnosis

Diagnosis/Criteria	Classification and rationale	Clinical specialist review	Echocardiography	Secondary antibiotic prophylaxis (SAP)
Priority 1a				
Bioprosthetic mitral valves	High risk of sudden valve degeneration **#**	6-monthly	6-monthly	4-weekly **§**
Recent valve repair within 1 year	To assess valve function stability		If stable at 6 months, move to yearly review	Until age 30 Beyond 30 years, individualised by patient and physician discussion Recurrences of are rare beyond ages 35-40 years Beyond 40 years, discontinue SAP for RHD of any severity
Priority 1b				
Pregnant people/women with RHD (any severity)	For the duration of the pregnancy	Individualised frequency of review in pregnancy	Individualised frequency of echo	Continue during pregnancy
Bioprosthetic aortic valves	Detect early degeneration	6-monthly initially	Frequency ≤ 6 months **#** if bioprosthetic valve gradient increases or onset of new regurgitation occurs	4-weekly **§**
Mechanical prosthetic valves	INR control & high risk of thrombosis	6-monthly initially	Initially, 6-monthly for baseline echo postoperatively, then yearly	See footnote ****
Priority 2				
Isolated severe valve lesion **Mitral stenosis with mitral valve area (MVA) <1.5cm²** **Double moderate valve lesion** **Mixed moderate valve lesion (stenosis and regurgitation)** **Prosthetic mitral valve** **Prosthetic aortic valve** **Bioprosthetic aortic valve** **Post-operative mitral valve repairs with stable gradients**	High risk of developing complications Severe by definition (previously 1.0cm²) Complex physiology/ outcomes equal to severe Mixed moderate RHD may be haemodynamically equivalent to severe RHD	Yearly Yearly, when stable gradients Yearly review even if gradients are stable	Yearly	4-weekly until aged 30 **§,** then review Beyond 30 years, individualised by patient and physician discussion After mitral valve repair, a recurrence of ARF may require mitral valve replacement. Continue to age >30, even with good mitral valve function
Priority 3				
Moderate RHD (for example, isolated stable MR or AR)	Medium risk	After ARF yearly until stable, then 2 yearly	After ARF yearly until stable, then 2 yearly	4-weekly until aged 30 **§**, then review

Diagnosis/Criteria	Classification and rationale	Clinical specialist review	Echocardiography	Secondary antibiotic prophylaxis (SAP)
Priority 4				
Mild RHD (including screen detected RHD)	Low risk for disease progression in the absence of recurrence of ARF	One review within the first year of diagnosis Then 3-yearly by a paediatrician or physician. No medical need for a cardiology review	One follow-up echo after the initial diagnosis If a new murmur starts or examination findings change, repeat echo If no new murmur or change, perform a 2–3-yearly echo, less often if stable Echo prior to recommendation of cessation of SAP	4-weekly until aged 21 or for 10 years after initial diagnosis, whichever is the longer **§ ###** Echo prior to recommendation of SAP cessation
ARF with no evidence of RHD	Low risk	2–3 yearly paediatrician review	Echo every 3–5 years even if clinical findings are unchanged until secondary prophylaxis is completed. **##**	

In addition, all RHD patients should have:

- Endocarditis prophylaxis
- 6 monthly to yearly dental review‡
- Annual influenza vaccination (funded for RHD patients)
- Polysaccharide pneumococcal vaccination (Pneumovax® 23), repeated once after 5 years

Post-surgical RHD is, by definition, still severe RHD. The priority category for post-surgical RHD varies as listed in this priority classification table and should be determined by an RHD specialist.

Review frequency should be adjusted based on individual needs. Most critically, prompt review should occur if:

- New symptoms develop
- Symptoms worsen
- Clinical findings change

Footnotes

Bioprosthetic mitral replacement is associated with higher rates of structural valve degeneration and requires closer follow-up due to unpredictable rapid onset of stenosis[3-5] and unpublished contemporary case series from Waikato RHD patients (N Patel FRACS 02/02/2024 written communication). Review 6-monthly (Evidence level III-4).

Feedback from whānau is that many expect intermittent echo reviews and guidance on exercise.

‡ Routine dental care is critically important for patients with a history of ARF and/or RHD. All patients should receive education about oral hygiene and be referred regularly for dental assessments (Grade D).

§ 28-day benzathine penicillin G is recommended in Aotearoa unless confirmed recurrent ARF has occurred despite full adherence to SAP. In that case, 21-day benzathine penicillin G is recommended (Grade C).

****** The main priority for an RHD patient with a double valve replacement is optimal anticoagulation management. Recurrence of ARF is very uncommon after age 30 and the risk of cardiac involvement is lower without a native mitral and aortic valve. It is reasonable to recommend oral penicillin as an alternative to stopping intramuscular SAP if the patient prefers to continue SAP (Grade D).

See Chapter 14: Screening for Rheumatic Heart Disease.

For the schedule on secondary prophylaxis with benzathine penicillin, see **Chapter 8: Secondary Prevention**.

A 21-day regimen (3 weeks) may be recommended after specialist review for people who have experienced recurrent ARF despite good adherence. There is no indication for SAP beyond age 40 for RHD of any severity.

For definitions of RHD severity, see **Chapter 10: Diagnosis of Rheumatic Heart Disease**.

Introduction

Knowledge about the epidemiology of RHD (as distinct from ARF) remains incomplete in Aotearoa. Some of these knowledge gaps are partly being addressed through data from the Aotearoa New Zealand Rheumatic Heart Disease Registry, which comprises 5,000 cases of moderate and severe RHD. However, further research is needed.[6] RHD is not a notifiable condition in Aotearoa, but registering new cases of RHD on the Rheumatic Fever Care Coordination System will improve epidemiological data for patients initiated on secondary prophylaxis.

It is pertinent that most countries, including Australia,[7] classify RHD as a notifiable condition, similar to ARF. Whānau and health professionals sometimes use the terms 'ARF' and 'RHD' interchangeably. Since ARF and RHD share the same risk factors, it is logical that a new diagnosis of RHD should trigger a holistic approach to care, which includes, for example, a housing assessment.

General management for those with rheumatic heart disease

Understanding the lived experience of individuals with RHD and their whānau should be at the forefront of all appointments: Table 11.1 summarises the fundamental goals for long-term RHD management. Longer clinic appointments should be available for initial consultations. Financial assistance for transport to the clinic may be required.

A multi-disciplinary approach

Access to cardiologists and echo for management and timely intervention before irreversible cardiac remodelling develops is important. A multi-disciplinary team (MDT) approach is often required to manage RHD, ideally offering a one-stop service for patients. Adequate primary, secondary, and tertiary services are often needed, including cardiology, cardiothoracic surgery, paediatrics, general medicine, cardiac obstetrics, general practice, dentistry and infectious diseases. Table 11.2 summarises the recommended frequency of specialist reviews and echo.

Due to the chronic nature of valvular disease, symptoms may not always be reported or recognised by the individual, but their functional capacity may be significantly less than expected for their age group. Stress testing and any supporting history from whānau are important for identifying functional impairment.

Moreover, irreversible myocardial damage, which is known to increase the risk of an adverse outcome, often occurs before the onset of symptoms. Multi-modality imaging and biomarkers can help reduce risk by providing important additional information to optimise the timing of valve intervention, especially in the asymptomatic patients.[8]

In cases of mild to moderate valvular regurgitation after ARF, RHD often improves over time, provided there is no recurrence of ARF.[9-11] In mild and moderate aortic and mitral valve regurgitation, the left ventricle (LV) is generally not at risk of failing.

In contrast, cardiac disease may worsen in individuals with severe RHD due to irreversible valve damage and secondary complications of left ventricular fibrosis, dysfunction, atrial dilatation, or pulmonary vascular remodelling. Deterioration can happen even if ARF does not recur. [9] Individuals with severe RHD are most at risk of developing complications described later in this chapter.

Best-practice care for rheumatic heart disease

Cardiologists have a key role in supporting best-practice RHD care. Table 11.1 is a useful checklist highlighting the importance of continued secondary prophylaxis.[12] RHD diagnosis is described in Chapter 10: Diagnosis of Rheumatic Heart Disease. The 2020 Australian Guideline for Acute Rheumatic Fever and Rheumatic Heart Disease (3rd edition)[7] also describes RHD and its management.

All patients with RHD or a history of ARF who develop new heart murmurs require an echo (Grade D). Table 11.2 outlines the recommended echo frequency. Echo is essential for grading the severity of valvular disease and serial echo assessments are critical in determining the optimal timing of any valve intervention.

Table 11.3 provides guidance on the timing of intervention in severe valve disease. Symptomatic severe valve disease is always an indication to consider valve intervention or surgery.

The role of medical therapy

Despite extensive guidance about the medical management of chronic heart failure[13, 14] (Grade A) in this context, evidence supporting medical (pharmacological) therapy for improving outcomes in severe but asymptomatic RHD (Grade B) is limited. Heart failure therapies are recommended in the presence of left ventricular dysfunction to optimise cardiac function before cardiac surgery. Standard goal-directed medical therapy remains a cornerstone of management in cases of residual cardiac dysfunction after valve replacement.[14, 15]

Improving equitable access to specialist services and surgery

Barriers to healthcare access are widely recognised for Indigenous and marginalised populations. They include geographic isolation, language, and financial barriers.[16] Barriers to healthcare for Māori and Pacific peoples in Aotearoa have been well-documented and must be systematically addressed.[17-19] Māori and Pacific peoples experience inequitable outcomes for ARF and RHD in Aotearoa.[20, 21] A study found that these population groups had:

- Higher initial RHD hospitalisation rates (Māori aRR 3.2; 95% CI: 2.9–3.5, Pacific peoples aRR 4.6; 95% CI: 4.2–5.1).
- Higher RHD mortality (Māori aRR 12.3, 95% CI: 10.3–14.6, Pacific peoples aRR 11.2, 95% CI: 9.1–13.8).[20, 21]

Ensuring equitable access to specialist cardiac services and surgical intervention for severe RHD is therefore important. Measures to achieve equitable healthcare outcomes for Māori and Pacific peoples include:

- Improving health literacy by allowing enough time in each consultation to ensure patients and whānau fully understand their RHD diagnosis and management. Care plans should be well articulated in correspondence.
- The RHD team should take a holistic approach to managing RHD and use a whānau/patient-focused approach.
- Establishing strong relationships between health practitioners and Māori communities and involving community health workers to bridge cultural gaps between Māori and Pacific patients and non-Māori/non-Pacific health practitioners.
- Offering clinic times and locations that align with patients' contexts. Providing social and financial support to facilitate clinic attendance if needed.

Leadership within health organisations must commit to reducing health inequities and supporting the health system to achieve these goals. Establishing universal health targets would further support a focus on equitable healthcare access for Māori.[17-19]

Historically, Aotearoa has lacked comprehensive data on the proportion of patients with severe RHD who attend specialist ARF and RHD clinics.[22] The Aotearoa New Zealand Rheumatic Heart Disease Registry[23] retrospectively records that approximately 85% of individuals with moderate or severe RHD enrolled on the registry were already attending specialist care at the time of enrolment.[24] This information led to an initiative for regional RHD experts to re-engage patients not linked to specialist care, and to reoffer assessment and review.

We recommend cardiothoracic units continue to develop RHD multi-disciplinary heart teams of cardiothoracic surgeons, cardiologists and specialist nurses with expertise in RHD (Grade D). Ongoing professional development would maintain a high volume of expertise, especially from surgeons. The RHD team should also develop additional quality improvement initiatives, such as auditing the unit's performance against these guidelines.

Transitioning from paediatric to adult cardiac services

The highest incidence of initial ARF occurs in tamariki aged 5–14 years. Initial cases are rare in individuals over 30 years of age.[9] Although RHD occurs in tamariki, the peak cumulative incidence occurs in adults, usually between 25 and 45 years of age.[9] Currently, two-thirds of individuals on the Rheumatic Fever Care Coordination System are aged 16 years and older (personal communication, 2024, C. Jackson, Clinical Advisor Rheumatic Fever Care Coordination System | Public Health Medicine Specialist, Health New Zealand | Te Whatu Ora).

The principles of rangatahi care and transition are covered in detail in **Chapter 13: Developmentally Appropriate Care for Rangatahi — Adolescents and Young People**. Cardiologists and adult physicians should be aware of the unique challenges rangatahi face, including the lack of age-appropriate services.

The transition to adult cardiology services usually starts from 15–16 years of age for those with moderate or severe RHD. For mild RHD or no RHD, continuing follow-up in paediatric services until 18–21 years of age, until their cardiac status is reassessed, is logical. Reassessment determines whether secondary prevention should continue (see **Chapter 7: Initial Management of Acute Rheumatic Fever** and **Chapter 8: Secondary Prevention**). Regional variations in transition models are determined by local expertise.

Clinicians and ARF secondary prevention services are important to ensure a smooth transition of care from paediatric to adult cardiac services. This includes ongoing SAP where needed.[2, 6, 25] The national Rheumatic Fever Care Coordination System can also support this transition, particularly in districts where paediatric and adult secondary prophylaxis services are provided separately.

Key aspects of managing rheumatic heart disease in primary care

Effective primary care management of RHD includes:

- Oral healthcare.
- Secondary prevention (where needed).
- Anticoagulation management and support.
- Influenza and pneumococcal vaccine.
- Reproductive health and pregnancy (see **Chapter 12: Rheumatic Heart Disease and Pregnancy**). Pregnant people/women with RHD who present with new symptoms of breathlessness should be referred back to specialist care.

Table 11.3. Summary of indications for medical and surgical management for severe rheumatic heart disease in adults and tamariki

Valve disease	Medical therapy and discussion with the heart team	Indications for considering intervention and surgery	Valve intervention[26]
Mitral regurgitation (MR)	Acute MR: Afterload reduction, such as nitrates and diuretics, to reduce filling pressure.[14] Chordal rupture/flail segment: Early surgery is required. ARF without chordal rupture: Diuresis, afterload reduction (if indicated), and ARF management. Chronic MR: Heart failure management as per guidelines[15] while awaiting surgery or after valve replacement. Timing of surgery.	**Tamariki** A. Severe MR with symptoms of breathlessness **or** B. Asymptomatic MR and one of the following: • Impaired LV function LVEF <60% • LVESV z-score >+2.5 • Pulmonary hypertension >50mmHg. **Adults** A. Severe MR with symptoms (NYHA class 2–4)* **or** B. Asymptomatic MR and one of the following: • LVESD ≥40 mm • Impaired LV function LVEF <60% • Pulmonary hypertension >50mmHg • New-onset AF • Left atrial volume >60 mL/m^2.[14]	Mitral valve repair is preferred. If repair is deemed too complex, unfavourable, or fails, replace the valve with a mechanical or bioprosthetic valve. Mitral valve repair is preferred. Prosthetic valve replacement is indicated in patients where repair is deemed too complex or unfavourable or if the repair fails.
Mitral stenosis (MS)	Symptom control: Use beta-blockers, diuretics, ivabradine, non-dihydropyridine, calcium channel blockers, or digoxin if in AF. Anticoagulation with vitamin K antagonist if patients have AF.	**Tamariki and adults** A. Severe MS with symptoms (NYHA class 2–4) **or** B. Asymptomatic severe MS and one of the following: • Paroxysmal AF • MVA <1.5cm^2 • Pulmonary hypertension >50mmHg • Thromboembolism.	In patients with isolated MS and suitable anatomy, perform BMV. If BMV is not feasible, surgery should be performed.
Aortic regurgitation (AR)	Chronic asymptomatic severe AR: Manage hypertension and heart failure as per guidelines[15] while awaiting surgery or after valve replacement.	**Tamariki** A. Severe AR with symptoms of breathlessness **or** B. Asymptomatic severe AR and one of the following: • LVESV z-score >+4 • Impaired LV function LVEF <55%[28] concordant with ESC[14] and ACC/AHA guidelines.[29] **Adults with severe AR** Symptomatic patients with severe AR regardless of LV systolic function.[29] **Adults with asymptomatic severe AR** Asymptomatic patient with severe AR with one of the following: • LVESD >50 mm (or LVESD >25 mm/m^2 BSA in patients with small body size) • Impaired LV function LVEF ≤55%.[29] Surgery may be considered in asymptomatic patients with one of the following, if surgery is low risk: • LVESD >20 mm/m^2 BSA (especially in patients with small body size) • Resting LVEF ≤55%.[14]	Aortic valve repair if surgical expertise is available. Replacement in tamariki — currently homograft preferred. There is minimal data to support Ross procedure for RHD in tamariki. Mechanical valve replacement. Aortic valve repair if surgical expertise is available. Bioprosthetic valve or mechanical valve replacement after discussion with the heart team.

Valve disease	Medical therapy and discussion with the heart team	Indications for considering intervention and surgery	Valve intervention[26]
Aortic stenosis (AS)	Heart failure management as per heart failure guidelines[15] in patients with reduced LVEF, while awaiting intervention. Referral for surgery.	**Symptomatic AS** (in adults and tamariki, but extremely uncommon in tamariki) Severe AS: • Mean gradient ≥40mmHg • Peak velocity ≥4.0 m/s • Valve area ≤1.0 cm^2 (or ≤0.6 cm^2/m^2). **Asymptomatic AS** Severe AS and systolic LV dysfunction (LVEF <55%) without another cause. [14]	Surgical valve replacement or transcatheter valve replacement. Decision based on: • Surgical risk • Age • Anatomical assessment • Heart team discussion.
Tricuspid regurgitation (TR)	• Limited data to define appropriate medical therapy for TR, with no evidence level I recommendations in current guidelines.[14, 29] • Diuretics if right heart failure is present. • Treatment of pulmonary hypertension in specific cases by addressing left heart disease. • Control rhythm in patients with paroxysmal or recent onset AF or atrial arrhythmias.[14]	**Tamariki** See adult indications. **Adults** Establish aetiology of TR — primary or secondary valve disease. • **Primary TR:** Surgical intervention for severe TR. • **Secondary TR:** Consider surgery for asymptomatic severe TR with progressive RV dilatation. • Greater than mild secondary TR with annular dilation (>40 mm or >21 mm/m^2) undergoing left-sided valve surgery (Evidence level IIa). • **Isolated TR:** Severe TR with right atrial dilatation and/or right ventricular dilatation with borderline or low function.	Valve repair/ annuloplasty is preferred. If repair is not possible, surgical valve replacement with: • Bioprosthetic valve. • Mechanical valve. Valve repair/ annuloplasty is preferred. Carry out one of the following valve replacements if repair is not possible: • Bioprosthetic valve. • Mechanical valve. **Secondary TR:** Surgically treat moderate or greater TR with annular dilatation when performing left-sided valve surgery.

Adapted from European Society of Cardiology (ESC),[14] American Heart Association guidelines,[29] New Zealand Diagnosis, Management and Secondary Prevention of Acute Rheumatic Fever and Rheumatic Heart Disease: (2014 Update second edition),[2] 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease,[7] RHD Surgery chapter Antunes et al.[26] (Grades A and B).

Abbreviations used in the table

AF	Atrial fibrillation
BMV	Balloon mitral valvuloplasty
BSA	Body surface area
LV	Left ventricular
LVEF	Left ventricular ejection fraction
LVESD	Left ventricular end-systolic diameter
LVESV	Left ventricular end-systolic volume
NYHA	New York Heart Association classification
RV	Right ventricle

Managing rheumatic heart disease in tamariki

Since most evidence-based management of heart failure is based on adult data, the management principles for tamariki and rangatahi with RHD and heart failure have been extrapolated from published guidelines for adult patients.[1] Medical therapy plays a role in both preventing and treating complications of RHD, such as AF, heart failure, thromboembolic events and endocarditis.[30]

Surgical management of heart valve disease

Surgical planning should consider the lifetime trajectory of valve disease and the future valve options following initial surgery. Previous recommendations for tamariki and rangatahi in Aotearoa were largely based on cohorts from Green Lane Hospital and Starship Hospital.[31-36] However, the small body of literature on cardiac surgery outcomes in tamariki is growing.[37-39] More recent data from Starship Hospital are being analysed and may further refine surgical thresholds for childhood RHD.[28]

Repair mitral valves where possible

Mitral valve repair has lower morbidity than replacement in tamariki.[1, 33] Aortic valve repair should also be considered in tamariki and rangatahi, when feasible; however, data on the longevity of repair is limited.[7, 40] Surgical repair of rheumatic valvular disease is technically more difficult than for non-rheumatic valvular pathology.[38, 41]

Mitral valve repair may require reoperation.[1] In an Australian cohort of 79 tamariki (median age 11.4 years) who underwent mitral valve repair, 85% survived at 15 years; however, only 28% (CI: 17–47%) remained free from reoperation.[38] The risk factors for valve deterioration included preoperative anterior leaflet immobility and surgical posterior patch extension. A cohort study of 336 tamariki undergoing mitral valve repair in India found 93.9% survival at both 10 and 20 years, which was superior to replacement ($P < 0.001$).[37] Importantly, freedom from reoperation at 10 and 20 years after mitral valve repair was 81.7% and 72.6%, respectively. This study offers strong support that the rheumatic mitral valve should be repaired in the young when technically feasible, to maximise survival and reduce valve-related morbidity of mechanical prosthetic valve replacements.[37]

In Aotearoa, the reoperation rates for mitral valve repair in tamariki under 20 years are similar to those for valve replacement.[33] Bioprosthetic valves in the mitral position have limited and unpredictable durability in tamariki and should not be used.[26, 33]

Indications for cardiac surgery

Indications for cardiac surgery in tamariki include assessing LV size in systole and diastole, normalising for body size, and, where possible, tabulating over time.[26] Most paediatric and adult studies demonstrate that systolic dimensions and volume thresholds are related to late outcomes of mortality and LV dysfunction in tamariki with MR.[31-36] A recent paediatric study found that left ventricular end-diastolic indexed volumes (LVEDVi) were predictive of post-operative LV dysfunction.[39]

See Table 11.3 for detailed indications for surgical referral.

Mitral regurgitation

The indications for considering intervention or surgery are listed in Table 11.3.

There is no proven indication for medical treatment of asymptomatic severe rheumatic MR with normal LV function. Pre-operatively, angiotensin-converting enzyme inhibitor (ACEi) should be used in cases of LV impairment, and diuretics should be used if there is evidence of heart failure.

Various options for surgical valve repair for MR are available.[26, 42] Mitral valve repair is the operation of choice if possible.[32, 33] Where valve repair is deemed too complex or unfavourable, or if repair has failed, valve replacement is recommended.[26]

Mitral stenosis

The indications for considering intervention or surgery are listed in Table 11.3.

Balloon mitral valvuloplasty (BMV) is currently the first option in young patients presenting with isolated mitral stenosis, with surgical mitral commissurotomy as a subsequent procedure.[26]

Surgical indications are similar for tamariki and adults (see Table 11.3). However, early referral should be considered in younger patients, as they have a higher likelihood of successful mitral valvuloplasty.

Aortic regurgitation

Medical therapy with afterload reduction is indicated for tamariki and rangatahi with severe AR and concurrent systemic hypertension. Treatment options include afterload-reducing agents such as ACEi and calcium channel blockers (see Table 11.3 for surgical referral indications).

Aortic valve replacement has been the mainstay of treatment in Aotearoa, with aortic homograft as the initial surgical approach. Homograft explanation in patients with a large body habitus is technically challenging, so prosthetic valve replacement is often performed as the initial surgery. Aortic valve repair is feasible for a smaller proportion of rangatahi with RHD, but follow-up data remains limited outside of a small case series.[28] The usefulness of the Ross procedure for rheumatic AR in rangatahi with RHD remains controversial.[26]

Mitral and aortic regurgitation

Data from Aotearoa shows that combined severe MR and severe AR are most harmful to long-term ventricular function. Consider patients with MR and AR for surgical interventions early,[36] based on the threshold size of the LV for isolated MR (rather than isolated AR).

Managing rheumatic heart disease in adults

Cardiologists and cardiac nurse specialists should discuss the importance of frequent clinic reviews with the patient.

Generally, only those with severe valve lesions or multi-valve moderate lesions need cardiac surgery. Medical management alone is insufficient for those with severe RHD.[21, 26]

Recommendations for cardiac surgery for adults are adapted from the AHA/American College of Cardiology (ACC) and ESC/European Association for Cardio-Thoracic Surgery (EACTS) guidelines based on internationally relevant data[14, 29] including their grading of evidence. Table 11.3 summarises these recommendations.

Delays in timely surgical referral can result from:

- Reduced access to care and healthcare systems.
- Competing health needs (for example, waiting for dental management).
- A lack of patient and whānau readiness for surgery.

Pre-conception counselling by specialist obstetric physicians or cardio-obstetric specialists should be provided before cardiac surgery where possible. Individualised discussions about future pregnancy risks and management strategies are required for individuals of childbearing age.

Mitral regurgitation

The indications for considering intervention or surgery are listed in Table 11.3.

Medical management

Preventing further episodes of rheumatic fever is the only proven medical therapy that alters the progression of chronic valve disease.

For patients with heart failure or ARF, heart failure management and rate control strategies should be optimised pre-operatively where required.[1]

The combination of moderate MR and AR should be considered the equivalent of single severe valve regurgitation, with similar thresholds for surgery. Where MR is thought to be moderate but left ventricular dilatation or symptoms exist, additional imaging methods — such as transoesophageal echo, cardiac MRI, and stress echo, should be used for a comprehensive valve assessment.

Surgical management of mitral valve regurgitation in adults

Surgical planning should consider the following:

- The lifetime trajectory of valve disease.
- The expected future valve options following initial surgery.

Mitral valve repair is the preferred operation for MR due to its lower mortality and morbidity.[43] A retrospective study in Aotearoa examined outcomes in rangatahi aged 15–24 years who underwent mitral valve surgery for RHD. Patients who underwent mitral repair had better survival, with a medium-term mortality rate of 6.9% compared to 15.9% in the valve replacement group (mean follow-up: 6 years).[44]

In adults, mitral valve repair has a higher reoperation rate than replacement.[45, 46] However, in the absence of other risk factors, successful mitral valve repair could allow a decade or more without warfarin. Contact sports, such as rugby, would then not pose a risk of cerebral bleeding. A well-functioning repair without stenosis is also safer in pregnancy.

If the mitral valve is not suitable for repair, the option is to replace the valve with either:

- A mechanical valve prosthesis.
- A bioprosthetic valve (in the absence of an indication for anticoagulation).

Valve replacement surgery should only take place after a thorough discussion about:

- The significant risks of structural valve degeneration.
- Timing for re-do surgery.
- Individualised risk assessment.
- Anticoagulation considerations.

For a more detailed discussion of thresholds for valve surgery, see the 2021 ESC guidelines[14] and 2020 ACC/AHA guidelines.[29]

Mitral stenosis

The indications for considering intervention or surgery are listed in Table 11.3.

Severe mitral stenosis is defined as an MVA of <1.5cm^2 (normal MVA in adults is 4–6cm^2). A mean gradient of 10mmHg is typically associated with severe mitral stenosis, however gradients can be influenced by flow and heart rate. Mitral valve obstruction increases left atrial pressure, leading to left atrial remodelling and elevated pulmonary pressures. A baseline pulmonary artery systolic pressure of >40mmHg is associated with rapid progression and worse outcomes.[23] Close follow-up and timely valve intervention are key.

Low-flow, low-gradient mitral stenosis (mean gradient <10mmHg, MVA <1.5cm^2) may be associated with worse survival outcomes than high-gradient severe mitral stenosis. Patients with low-gradient mitral stenosis tend to benefit less from balloon valvuloplasty.

Higher rates of AF, severe TR, and reduced atrial and ventricular compliance contribute to discrepancies between gradient and valve area.[47, 48]

Exercise stress echo is helpful in identifying asymptomatic individuals with clinically significant mitral stenosis who would benefit from early intervention.

Medical management

The medical management of symptomatic rheumatic mitral stenosis while awaiting valve intervention includes:

- Beta-blockers or non-dihydropyridine calcium channel blockers.
- Diuretics.
- Digoxin in the presence of AF.

Ivabradine (for patients in sinus rhythm) can also improve symptoms but remains unfunded in Aotearoa.[14]

Mitral stenosis and AF significantly increase thrombosis risk. Anticoagulation with a vitamin K antagonist is indicated irrespective of CHADSVASc score, with a target international normalised ratio (INR) of 2 to 3. In patients with a history of systemic embolism and mitral stenosis but no documented AF, oral anticoagulation is recommended.[14] No robust evidence exists on whether those with severe mitral stenosis who remain in sinus rhythm should be anticoagulated.

Surgical management

Referral for cardiac surgery or BMV is indicated with mitral stenosis, as detailed in Table 11.3. BMV is also known as percutaneous balloon mitral commissurotomy (PBMC).

Mitral valve replacement is recommended for patients whose anatomy is unsuitable for BMV (see Chapter 10: Diagnosis of Rheumatic Heart Disease).

Aortic regurgitation

Referral for cardiac surgery is indicated in adults with AR, as detailed in Table 11.3.

Timely referral for surgery for severe AR is important to prevent chronic left ventricular fibrosis, which is strongly linked to adverse events.[49] LV systolic function is a key factor in determining survival and post-surgical functional status. Cardiac MRI and functional testing should be considered if LV size or borderline systolic function approaches surgical thresholds (see Table 11.3). Patients should be reviewed within 6 months to assess whether they meet the surgical threshold.

In asymptomatic patients with chronic AR, medical management includes treating hypertension (systolic blood pressure >140mm Hg).

In symptomatic patients with severe AR, surgery is recommended regardless of LV systolic function.[29]

Optimising surgical timing

The ACC/AHA guidelines recommend early aortic valve surgery in asymptomatic, low-risk patients with EF>55% if serial assessments show progressive LV end-diastolic diameter >65mm or a decline in EF 55–60%. Using linear dimensions and EF may delay the early detection of cardiac dysfunction. Left ventricular volumes may better predict adverse outcomes for women and older patients.[50] This has led to calls for more data to guide optimal surgical timing for women, who often experience worse outcomes using linear dimensions of the guidelines.[49]

Standard heart failure therapy

Patients who continue to have reduced ejection fraction after surgery should receive standard heart failure therapy.

Global longitudinal strain for earlier detection and prognosis

Global longitudinal strain (GLS) in echocardiographic imaging is a well-established prognostic tool that detects cardiac dysfunction earlier than LVEF. While the use of GLS is discussed in valve management guidelines, it is not yet included in guideline recommendations on the timing of surgery. Studies have shown that:

- Worsening GLS is associated with higher long-term mortality.[51]
- Fibrosis on cardiac MRI is also linked with increased mortality.[8]

These studies highlight the importance of close monitoring and timely surgical referral to improve patient outcomes.

Mixed aortic valve disease

Mixed AR and stenosis are common in RHD. In the absence of co-existing mild regurgitation or stenosis, relying on thresholds for severe regurgitation or stenosis may lead to worse outcomes due to the underlying competing compensatory mechanisms of LV dilatation and hypertrophy.

Moderate mixed aortic valve disease is associated with a higher risk of mortality than isolated moderate regurgitation or stenosis, with outcomes comparable to severe aortic stenosis.[52]

Independent risk factors for adverse outcomes include increased relative wall thickness and LV mass, peak aortic valve velocity (>3m/s), and peak gradient (>45mmHg).[53, 54] Biomarker use of B-type natriuretic peptide (BNP) offers additional prognostic information.[55]

Aortic stenosis

The indications for considering intervention or surgery are listed in Table 11.3.

Native isolated pure aortic stenosis is rare in RHD, while mixed aortic valve disease is more common.

Due to the challenges of lifelong anticoagulation associated with mechanical valve replacement, a bioprosthetic valve is a potential alternative, allowing patients to delay the need for a mechanical valve and subsequent anticoagulation.

Aortic stenosis can develop after bioprosthetic valve implantation. For guidance on assessing bioprosthetic valve dysfunction, see Chapter 10: Diagnosis of Rheumatic Heart Disease.

Transcatheter valve implantation for rheumatic heart disease

The widespread use of transcatheter valve techniques has led to growing interest in transcatheter valve-in-valve procedures for treating structural degeneration of bioprosthetic valves in RHD.[56] However, longer-term data is lacking, and studies on re-do surgery after valve-in-valve procedures have primarily focused on older patients with comorbidities.[57, 58]

Data for explantation of a surgical aortic valve placement (SAVR) with a subsequent valve-in-valve procedure is much more limited than data on transcatheter aortic valve replacement (TAVR) explantation. TAVR is more commonly referred to as transcatheter aortic valve implantation (TAVI). TAVR explantation carries unique risks, including

complex valve removal and a higher incidence of root replacement. Studies on TAVR explantation remain limited to an older population at higher risk for re-do surgery. The limited data does not suggest a significantly increased risk of re-do surgery for patients who undergo SAVR followed by transcatheter valve-in-valve procedure compared to those who undergo re-do SAVR alone.[59]

Transcatheter mitral valve-in-valve (TMViV) replacement is an alternative to re-do surgery in individuals with a previous bioprosthetic mitral valve.[56] However, evidence in rangatahi remains limited due to technical challenges such as the risk of LVOT obstruction, valve migration, or thrombosis.[4]

Tricuspid regurgitation

The prevalence of moderate or severe TR increases with age. It affects about 4% of patients aged 75 years or more, compared to 0.55% of the general population.[60]

Primary TR can be a feature of RHD. However, in ≥90% of TR cases, the aetiology is secondary to pressure and/or volume overload leading to right ventricular dilatation or is due to enlargement of the right atrium and tricuspid annulus due to chronic AF.[14, 60]

For patients with AF, a rhythm control strategy may help to reduce TR severity and contain annular dilatation.[14, 61]

Severe TR is associated with poor survival, so appropriate intervention timing is crucial.[14]

Tricuspid valve surgery

The indications for considering intervention or surgery are outlined in Table 11.3.

The more extensive the tricuspid rheumatic changes (leaflet thickening and chordal involvement), the lower the likelihood of successful surgical valve repair. Isolated tricuspid valve surgery has been associated with greater morbidity and mortality than other valve surgeries.

A retrospective review of patients who underwent isolated tricuspid valve surgery at Auckland Hospital reported one- and ten-year survival rates of 80–85% and 60–65%, respectively, with tricuspid repair and replacement having reduced survival over time. RHD was the underlying aetiology in 44% of the cohort, and 69% of the participants had prior cardiac surgery.[62] International guidelines are moving to earlier referral for tricuspid valve surgery. A more recent audit demonstrated improved outcomes, but a significant mortality rate of 20% at four years continues to persist.[63]

Surgery should be considered in patients with severe secondary TR (with or without previous left-sided surgery) if:

- They are symptomatic or have RV dilatation.
- They do not have severe RV or LV dysfunction.
- Severe pulmonary vascular disease/hypertension.[14]
- They have mild or moderate secondary TR with a dilated annulus (>40 mm or >21 mm/m^2) and are undergoing left-sided valve surgery (Evidence level IIa).[26]

Tricuspid stenosis can occur in the presence of rheumatic involvement but is often associated with regurgitation in native disease. For information on bioprosthetic tricuspid stenosis, see Chapter 10: Diagnosis of Rheumatic Heart Disease.

Best-practice discharge after cardiac surgery

Before discharge, the cardiac surgical team should consider and address the following areas of care:

Table 11.4. Best practice discharge after cardiac surgery

Area of care	Recommendation
Routine review and structured care planning	Develop and document a structured care plan in agreement with the patient.
Baseline echo	Perform a baseline echo before discharge or within 3 months of valve surgery.
Cardiac rehabilitation	Refer the patient to local cardiac rehabilitation services. Also, see the Heart Foundation's page on cardiac rehabilitation.
Secondary antibiotic prophylaxis	The surgical team should discuss the need for continued prophylaxis. If uncertain, seek clarification from the patient's cardiologist or physician. For more details, see **Chapter 8: Secondary Prevention**.
Preventing infective endocarditis	See Prevention of Infective Endocarditis — Guideline by the Heart Foundation.
Anticoagulation	Refer individuals with mechanical prosthetic valves to their local General Practitioner (GP) or pharmacy as appropriate for ongoing INR monitoring. A Referral to the community Pharmacy Anticoagulation Management Service should be made via the GP.
Oral healthcare	Refer the patient to dental services for check-ups at least annually. For dental procedures requiring antibiotic prophylaxis, see Table 11.5.
Specialist advice about pregnancy	Provide pre-conception counselling before valve replacement surgery. Where this is not possible, offer contraception and consultation with an appropriate obstetric medicine specialist.
Immunisations	Encourage annual influenza vaccinations. Ensure polysaccharide pneumococcal vaccination (Pneumovax® 23) is repeated once after 5 years. Find out about Pneumovax23.

Secondary antibiotic prophylaxis after rheumatic heart disease valve surgery

Continuing SAP after valve repair is paramount. There is limited quality evidence on how long SAP should be continued after mechanical prosthetic mitral or aortic valve replacement.

There is also insufficient risk-benefit data supporting SAP beyond the third decade of life. A retrospective low-evidence study in RHD patients over 30 years of age found no improved clinical outcomes (Evidence level III-4) for:

- Overall survival.
- Valve-related hospitalisation-independent survival.
- Re-do valve surgery-independent survival for those on secondary prophylaxis.[64]

For further details, see **Chapter 8: Secondary Prevention**.

Anticoagulation

Warfarin, a vitamin K antagonist (VKA), remains the preferred anticoagulant for patients with mechanical prosthetic heart valves for both RHD and non-rheumatic valve disease.[29] Long-term VKA oral anticoagulation is also recommended for patients with rheumatic MS with AF.[29]

Direct oral anticoagulants (DOAC) are non-vitamin K antagonists, also known as novel oral anticoagulants (NOAC). DOACSs may be appropriate in some instances but not for all RHD patients with AF:

- DOACs are appropriate where the CHA2DS2-VASC score is elevated in non-mitral stenosis RHD lesions (see **Table 11.2**).
- DOACs are not appropriate for patients with moderate or more significant mitral stenosis and mechanical prosthetic valves.[7]
- DOACs can be used in patients with bioprosthetic valves.[14, 29]

The INVICTUS study, a randomized, non-inferiority trial, compared the DOAC rivaroxaban with VKA therapy in RHD-associated AF.[65] Of the 4,531 participants included in the final analysis, 81.9% had moderate to severe mitral stenosis (valve area ≤2.0cm^2). Patients with RHD-associated AF had a lower rate of cardiovascular events or death with VKA therapy compared to those on rivaroxaban. Restricted mean survival time in the VKA group was 1,680 days, compared to 1,608 days in the rivaroxaban group, a difference of -72 days (95% CI: -117 to -28). Both groups had similar rates of bleeding.[65]

Recommendations

VKA therapy is superior to DOACs in patients with AF and moderate or severe mitral stenosis (Grade B).[65]

VKA therapy should be used in patients with RHD and mechanical prosthetic heart valves (Grade C).

INR ranges and monitoring

Since warfarin absorption is affected by diet, regular INR monitoring is required, with dose adjustments as needed.

The cardiologist should specify the target INR range for individual patients. The usual recommended INR ranges are:

- Prosthetic mitral valves — target: 3 (Range: 2.5–3.5).
- Prosthetic aortic valves — target: 2.5 (Range: 2.0–3.0).
- Mitral and aortic prosthetic valves — target: 3 (Range: 2.5–3.5).[66]

Challenges in anticoagulation management

Managing anticoagulation with warfarin can be difficult, even with easy access to INR monitoring.

Low INRs → Risk of valve thrombosis, thromboembolism, and strokes.

High INRs → Risk of spontaneous bleeding and strokes.

Although self-testing and self-management of anticoagulation with warfarin have extensive evidence supporting their effectiveness, these resources are not currently funded in Aotearoa.[14, 67-71]

The patient should be encouraged to actively control their INR. Home INR testing kits are already used for tamariki with RHD and congenital heart disease in some centres around Aotearoa and more routinely overseas.[72]

A retrospective audit of 150 patients from Tāmaki Makaurau (Auckland region) randomly selected from the Aotearoa RHD registry showed that nearly 40% of those on warfarin were hospitalised due to anticoagulation complications. This highlights the need for improved coagulation services.[24] The Community Pharmacy Anticoagulant Management Service in Aotearoa has been shown to achieve good anticoagulation control by providing:

- Point-of-care testing.
- Immediate dosing advice at the time of testing.[22]

Understanding lived experiences will help improve systems

Understanding the lived experiences of those affected by RHD will help foster systems that cater to patients' contexts. Patients are often young people balancing work and whānau life.

The healthcare system must provide safe, unambiguous communication to ensure optimal patient care. Language barriers can create significant anxiety for patients, particularly for those with English as a second language. Immigrants may experience limited access to interpreters, further complicating their healthcare experiences.

Infective endocarditis

People with RHD have an increased risk of developing infective endocarditis, a condition that carries significant morbidity and mortality.[73] Infective endocarditis may occur on native, repaired valves, especially prosthetic heart valves. Prophylaxis is recommended for people with rheumatic valvular heart disease but not for those who have had previous rheumatic fever without cardiac involvement on an echo.

Infective endocarditis carries a high risk of mortality, particularly prosthetic valve endocarditis.[74] In Aotearoa, patients with RHD tend to develop endocarditis at a younger age and have higher rates of prosthetic valve endocarditis than those without RHD. A history of previous endocarditis is more common among RHD patients.[75] An episode of endocarditis can sometimes be the first presentation of RHD.[75]

Guidelines on diagnosing and managing infective endocarditis

Diagnosing and managing infective endocarditis is complex and beyond the scope of this guideline. The ESC and AHA Clinical Practice guidelines are commonly used to inform diagnosis and management in Aotearoa.[14, 29, 76] The guidelines recommend:

- Obtaining blood cultures before starting antibiotics in the community.
- Ensure access to echo and cardiac surgery when needed.
- Use a multi-disciplinary approach involving cardiology, infectious diseases, imaging, and cardiosurgical expertise.[77]

Prevention of infective endocarditis

The evidence supporting antibiotic prophylaxis for preventing infective endocarditis is limited.[22] Most reviews and studies have used bacteraemia as the endpoint. A meta-analysis of 11 trials found that antibiotic prophylaxis reduced the risk of bacteraemia (RR 0.53; 95% CI: 0.49–0.57, P <0.01).[78] A Cochrane review on prophylaxis for bacterial endocarditis in dentistry was inconclusive.[79]

The current recommendation is to give people with established RHD antibiotic prophylaxis before procedures that are expected to cause bacteraemia. Individuals with a history of ARF but no valvular damage do not require antibiotic prophylaxis. Those already receiving penicillin for SAP should be offered a different antibiotic for endocarditis prevention.[2]

Recommendations for procedures that require endocarditis prophylaxis and the appropriate antibiotics are on the Heart Foundation of New Zealand website. See Prevention of infective endocarditis — Guideline by the Heart Foundation.[80]

Oral healthcare and rheumatic heart disease

Regular oral healthcare, which includes assessment, treatment, and preventive education, should be a routine part of managing RHD. All patients with RHD (regardless of severity) should have an annual oral health check.[2] Dental recall intervals should be based on clinical risk.[81] People should be offered more frequent dental checks if they have:

- Moderate or severe RHD.
- Prosthetic cardiac valves.
- Higher dental risk factors (for example, poor oral hygiene, dry mouth, untreated dental caries, and inflammatory periodontal disease).

Access to dental care for patients with RHD should be prioritised based on risk.

Dental procedures requiring antibiotic prophylaxis

Certain dental procedures have an increased risk of causing oral bacteraemia and may increase the risk of infective endocarditis. The effectiveness of additional antibiotic prophylaxis before dental procedures is controversial, but antibiotic prophylaxis is recommended for at-risk patients undergoing high-risk dental procedures. People with RHD should be empowered to discuss the need for antibiotic prophylaxis for dental procedures with their healthcare providers.

Dental procedures that require antibiotic prophylaxis are listed in Table 11.5.

Table 11.5. Dental procedures requiring antibiotic prophylaxis

Antibiotic prophylaxis is recommended for the following dental procedures:
• Procedures that involve manipulation of gingival tissue or the periapical region of teeth, such as fillings that extend to or below the gum margin • Cleaning teeth at or below the gingival margin • Early stages of a root filling when the length of the canal is still being measured • Procedures performed in the presence of a dental abscess or infection

The following procedures and events do not need antibiotic prophylaxis:

- Routine anaesthetic injections through non-infected tissue.
- Taking dental radiographs.
- Placing removable prosthodontic or orthodontic appliances.
- Adjusting orthodontic appliances.
- Placing orthodontic brackets.
- Shedding of deciduous (baby) teeth.
- Bleeding from trauma to the lips or oral mucosa.

Antibiotic prophylaxis regimen for dental and medical procedures

People with RHD should be empowered to inform health professionals about the need for antibiotic prophylaxis before medical procedures.

Prophylaxis for dental procedures and tonsillectomy is directed against *Viridans streptococci*. *Viridans streptococci* are the organisms most likely to cause endocarditis after these procedures, but not the only ones.

For a full list of medical conditions requiring antibiotic prophylaxis, refer to the New Zealand guideline for preventing infective endocarditis associated with dental and other medical procedures 2008.[80]

Table 11.6. Recommended antibiotics for infective endocarditis prophylaxis for dental procedures

Adult or tamariki	Antibiotic and dosage	Administration
Adult	Amoxicillin 2g	PO
Tamariki	Amoxicillin 50mg/kg up to 2g	One of these: • PO 1 hour before the procedure • IV given just before the procedure • IM given 30 minutes before the procedure.
Administer the amoxicillin parenterally if the person cannot take medication orally. Administer through IV if IV access is readily available. For penicillin allergy, or if a penicillin or cephalosporin-group antibiotic is taken more than once in the previous month (including for those on long-term penicillin prophylaxis for acute rheumatic fever):		
Adult	Option 1: Clindamycin* 600mg	One of these: • PO 1 hour before the procedure • IV given over at least 20 minutes, just before the procedure • IM given 30 minutes before the procedure.
Adult	Option 2: Clarithromycin† 500mg	PO 1 hour before the procedure.
Tamariki	Option 1: Clindamycin* 15mg/kg up to 600mg)	One of these: • PO 1 hour before the procedure • IV, given over at least 20 minutes, just before the procedure • IM, given 30 minutes before the procedure.
Tamariki	Option 2: Clarithromycin† 15mg/kg up to 500mg	PO 1 hour before the procedure.

Adapted from the Heart Foundation of New Zealand. *New Zealand Guideline for the Prevention of Infective Endocarditis Associated with Dental and Other Medical Procedures 2008*.[80]

* Clindamycin is not available in syrup form in Aotearoa.

† Beware of potential interactions between clarithromycin and other medications.

If the antibacterial agent is inadvertently not administered before the procedure, it may be administered up to two hours after the procedure.

Patients may require more than one appointment to complete their care. If possible, schedule appointments two weeks apart and alternate Clindamycin and Clarithromycin, especially if the patient is already taking penicillin for secondary RHD prophylaxis or is allergic to penicillin. Dentist-prescribed antibiotics are not currently funded in Aotearoa, with a dispensing fee of $15 per medication, which may be a barrier for some whānau.

For more details, see these Heart Foundation resources:

- Antibacterial prophylaxis for dental procedures.
- New Zealand guideline for preventing infective endocarditis associated with dental and other medical procedures 2008.[80]

Complications of rheumatic heart disease

The complications of RHD include heart failure, AF, pulmonary hypertension, thromboembolic events, and infective endocarditis.[30]

Heart failure

Chronic mitral or aortic regurgitation can lead to reduced left ventricular ejection fraction, resulting in heart failure. International published guidelines summarise management and optimal medical therapy for patients with left ventricular systolic dysfunction.[15] Recommendations for initiating and achieving goal-directed medical therapy for patients with heart failure in Aotearoa are summarised below. [82]

- Renin-angiotensin system antagonist — use angiotensin receptor blocker (ARB) with a neprilysin inhibitor (ARNI) as first-line therapy. If ARNI is not tolerated, use ACEi or ARB.
- Beta-blocker.
- Mineralocorticoid receptor antagonist.
- Sodium-glucose co-transporter-2 (SGLT2) inhibitor.

Device-based therapies are discussed in international guidelines.[15, 83]

Pulmonary hypertension

Pulmonary hypertension is common in severe left heart valve disease and is associated with poorer outcomes. Pulmonary hypertension is associated with a higher risk of cardiac events and sometimes reduced survival despite correction of valvular disease.

Chronically elevated pulmonary pressures can cause irreversible pulmonary vascular remodelling and pulmonary arterial hypertension. 5-phosphodiesterase (PDE5) inhibitors have an established role in primary pulmonary artery hypertension. However, studies have not shown benefits in persistent pulmonary hypertension post-valve surgery.[84]

PDE5 inhibitors have also not shown clinically significant improvements in unoperated left-sided valve disease. Timely referral for valve surgery is key to prevent chronic pulmonary vascular changes.

Treatment for pulmonary hypertension is described in the 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension.[85]

End-stage rheumatic heart disease

Patients with advanced (Stage D) heart failure have severe ventricular dysfunction and chronic heart failure due to long-standing valvular disease. In the absence of any options for valvular intervention, referring patients for cardiac transplantation or other advanced therapies may be an option. 13 patients with end-stage RHD have received cardiac transplantation since the transplant programme started in Aotearoa in 1987.[6]

Markers of advanced heart failure and the need to consider referral to an advanced heart failure or transplant team can be summarised with the mnemonic 'I need help':

I Inotropic use
N NYHA class III or IV or elevated natriuretic peptides
E Ejection fraction <25%
E End-organ dysfunction (renal or liver impairment)
D Defibrillator shock
H Hospitalisation
E Edema
L Low blood pressure
P Prognostic medications stopped due to intolerance

Mechanical prosthetic aortic valve may preclude ventricular assist devices. The time required for assessment and the complicating factors in the presence of mechanical valves further increase the risk associated with late referrals.

In high-risk individuals in the appropriate clinical setting, clinicians should stress the importance of the following, which are key considerations should a transplant ever be needed:

- Medication adherence.
- Regular clinical review.
- A healthy weight with a BMI below 35.
- The importance of not smoking, vaping, or taking recreational drugs early in the clinical management and throughout follow-up.

Patients with end-stage RHD and prohibitive surgical risk should be offered referral to an appropriate and skilled palliative care team. Discontinuing secondary prophylaxis is reasonable. Symptom management should be tailored, including continued judicious use of diuretics and other cardiac medications.

Stroke

Stroke can be a complication of valve dysfunction due to RHD — 4% of RHD patients had a stroke within 10 years of RHD diagnosis.[7, 86]

Strokes in RHD can be due to mitral stenosis with or without AF. When young patients from a high-risk population present with arterial thromboembolism or stroke, consider if RHD could be a cause.[7]

Prosthetic valve dysfunction

Patients with prosthetic heart valves are at risk of various complications, which can significantly impact long-term outcomes.

Prosthetic valve complications include:

- Structural valve degeneration.
- Haemolysis.
- Endocarditis.
- Paravalvular leak.
- Thrombosis.

Bioprosthetic valves, including aortic homografts and tissue mitral valves, have limited durability and eventually fail. Clinical event rates can occur within the first year and is more common within the first five years.[87, 88] However, bioprosthetic or aortic homograft valves have been reported to last up to 10 years.[14, 89] Once echo imaging detects structural valve degeneration (valve thickening, valvular regurgitation, or stenosis), close follow-up is critical as bioprosthetic valve degeneration can occur rapidly.

Early investigation of breathlessness with echo imaging is essential in patients with bioprosthetic valves.

Managing thrombosis

Prosthetic valve thrombosis most commonly occurs within the first 3 months of implantation, but it can also happen years after the implantation, typically after 1–2 years but as much as 6.5 years.

In cases of acute mechanical valve thrombosis with symptoms of valve obstruction, urgent initial treatment with either slow infusion, low-dose fibrinolytic therapy, or emergency surgery has been recommended.[29]

Untreated prosthetic valve thrombosis that leads to left-sided prosthetic valve obstruction has high mortality and morbidity rates.[14, 77]

Paravalvular leaks may cause haemolysis and require assessment for intervention. The decision on transcatheter or surgical closure of clinically significant paravalvular leaks should depend on the patient's profile, paravalvular leak morphology, and local expertise.[14] Prosthetic valve infection should be excluded.

The ESC and the EACTS provide detailed guidelines on managing prosthetic valve dysfunction.[14]

References

1. Okello E, Mordi I, Lang C, Sable C, Dougherty S, Wilson N. Chapter 6 — Medical management of rheumatic heart disease. In: Dougherty S, Carapetis J, Zühlke L, Wilson N, editors. Acute rheumatic fever and rheumatic heart disease: Elsevier; 2020. p. 107–132.

2. New Zealand Heart Foundation. New Zealand guidelines for rheumatic fever: diagnosis, management and secondary prevention of acute rheumatic fever and rheumatic heart disease: 2014 update. Heart Foundation; 2014. https://www.heartfoundation.org.nz/resources/acute-rheumatic-fever-and-rheumatic-heart-disease-guideline (Accessed December 16 2024).

3. Chen CY, Chan YH, Wu VC, Liu KS, Cheng YT, Chu PH, et al. Bioprosthetic versus mechanical mitral valve replacements in patients with rheumatic heart disease. *Journal of Thoracic and Cardiovascular Surgery*. 2023;165(3):1050–1060. https://doi.org/10.1016/j.jtcvs.2021.03.033

4. Keenan NM, Newland RF, Baker RA, Rice GD, Bennetts JS. Outcomes of re-do valve surgery in indigenous Australians. *Heart, Lung & Circulation*. 2019;28(7):1102–1111. https://doi.org/10.1016/j.hlc.2018.05.198

5. Russell EA, Tran L, Baker RA, Bennetts JS, Brown A, Reid CM, et al. A review of outcome following valve surgery for rheumatic heart disease in Australia. *BMC Cardiovascular Disorders*. 2015;15:103. https://doi.org/10.1186/s12872-015-0094-1

6. Tilton E, Mitchelson B, Anderson A, Peat B, Jack S, Lund M, et al. Cohort profile: methodology and cohort characteristics of the Aotearoa New Zealand Rheumatic Heart Disease Registry. *BMJ Open*. 2022;12(12):e066232. https://doi.org/10.1136/bmjopen-2022-066232

7. RHDAustralia, Menzies School of Health Research. The 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (3.2 edition). 2022. https://www.rhdaustralia.org.au/arf-rhd-guidelines (Accessed December 16 2024).

8. Ajmone Marsan N, Delgado V, Shah DJ, Pellikka P, Bax JJ, Treibel T, et al. Valvular heart disease: shifting the focus to the myocardium. *European Heart Journal*. 2023;44(1):28–40. https://doi.org/10.1093/eurheartj/ehac504

9. Carapetis JR, Beaton A, Cunningham MW, Guilherme L, Karthikeyan G, Mayosi BM, et al. Acute rheumatic fever and rheumatic heart disease. *Nature Reviews Disease Primers*. 2016;2:15084. https://doi.org/10.1038/nrdp.2015.84

10. The Rheumatic Fever Working Party of the Medical Research Council of Great Britain, The Subcommittee of Principal Investigators of the American Council on Rheumatic Fever and Congenital Heart Disease. The natural history of rheumatic fever and rheumatic heart disease. Ten-year report of a cooperative clinical trial of ACTH, cortisone, and aspirin. *Circulation*. 1965;32(3):457–476. https://doi.org/10.1161/01.cir.32.3.457

11. Wilson NJ, Voss LM, Neutze JM, Ameratunga RV, Lennon DR. The natural history of acute rheumatic fever to one year in the echocardiographic era. In: Momma K, Imai Y, editors. Proceedings of the 2nd World Congress of Paediatric Cardiology and Cardiac Surgery: Futura Publishing; 1998. p. 971–972.

12. Wilson N. Secondary prophylaxis for rheumatic fever: simple concepts, difficult delivery. *World Journal for Pediatric and Congenital Heart Surgery*. 2013;4(4):380–384. https://doi.org/10.1177/2150135113497240

13. Nishimura RA, Otto CM, Bonow RO, Carabello BA, Erwin JP, 3rd, Guyton RA, et al. 2014 AHA/ACC guideline for the management of patients with valvular heart disease: executive summary: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. *Circulation*. 2014;129(23):2440–2492. https://doi.org/10.1161/CIR.0000000000000029

14. Vahanian A, Beyersdorf F, Praz F, Milojevic M, Baldus S, Bauersachs J, et al. 2021 ESC/EACTS Guidelines for the management of valvular heart disease: Developed by the Task Force for the management of valvular heart disease of the European Society of Cardiology (ESC) and the European Association for Cardio-Thoracic Surgery (EACTS). *European Heart Journal*. 2021;43(7):561–632. https://doi.org/10.1093/eurheartj/ehab395

15. Heidenreich PA, Bozkurt B, Aguilar D, Allen LA, Byun JJ, Colvin MM. 2022 AHA/ACC/HFSA guideline for the management of heart failure: a report of the American College of Cardiology/American Heart Association Joint Committee on clinical practice guidelines. *Journal of the American College of Cardiology*. 2022;79:e263–e421. https://doi.org/10.1161/CIR.0000000000001063

16. Jeremy R, Tonkin A, White H, Riddell T, Brieger D, Walsh W, et al. Improving cardiovascular care for indigenous populations. *Heart, Lung & Circulation*. 2010;19(5-6):344–350. https://doi.org/10.1016/j.hlc.2010.02.015

17. Ellison-Loschmann L, Pearce N. Improving access to health care among New Zealand's Maori population. *American Journal of Public Health*. 2006;96(4):612–617. https://doi.org/10.2105/AJPH.2005.070680

18. Cram F. Improving Māori access to health care: research report: Katoa Ltd; 2014.

19. Marrone S. Understanding barriers to health care: a review of disparities in health care services among indigenous populations. *International Journal of Circumpolar Health*. 2007;66(3):188–198. https://doi.org/10.3402/ijch.v66i3.18254

20. Oliver J, Robertson O, Zhang J, Marsters BL, Sika-Paotonu D, Jack S, et al. Ethnically disparate disease progression and outcomes among acute rheumatic fever patients in New Zealand, 1989-2015. *Emerging Infectious Diseases*. 2021;27(7):1893–1902. https://doi.org/10.3201/eid2707.203045

21. Bennett J, Zhang J, Leung W, Jack S, Oliver J, Webb R, et al. Rising ethnic inequalities in acute rheumatic fever and rheumatic heart disease, New Zealand, 2000-2018. *Emerging Infectious Diseases*. 2021;27(1):36–46. https://doi.org/10.3201/eid2701.191791

22. Rentta NN, Bennett J, Leung W, Webb R, Jack S, Harwood M, et al. Medical treatment for rheumatic heart disease: a narrative review. *Heart, Lung & Circulation*. 2022;31(11):1463–1470. https://doi.org/10.1016/j.hlc.2022.07.013

23. Ko KY, Cho I, Kim S, Seong Y, Kim DY, Seo JW, et al. Identification of distinct subgroups in moderately severe rheumatic mitral stenosis using data-driven phenotyping of longitudinal hemodynamic progression. *Journal of the American Heart Association*. 2022;11(15):e026375. https://doi.org/10.1161/JAHA.121.026375

24. Meng A, Wilson N, Webb R, Han DY, Tilton E. Contemporary specialist care for rheumatic heart disease meets New Zealand Heart Foundation recommendations for most patients in Tāmaki Makaurau. *Heart, Lung and Circulation*. 2023;32(2). https://doi.org/10.1016/j.hlc.2023.04.216

25. Wyber R, Kado J. Chapter 12 — Rheumatic heart disease control programs, registers, and access to care. In: Dougherty S, Carapetis J, Zühlke L, Wilson N, editors. Acute rheumatic fever and rheumatic heart disease: Elsevier; 2020. p. 235–259.

26. Antunes MJ, Finucane K, Kumar AS, Coutinho GF. Chapter 8 — Surgical management of rheumatic valvular heart disease. In: Dougherty S, Carapetis J, Zühlke L, Wilson N, editors. Acute rheumatic fever and rheumatic heart disease: Elsevier; 2020. p. 147–170.

27. Abdelnabi M, Benjanuwattra J, Ahmed A, Almaghraby A. Can warfarin be replaced by non-vitamin K anticoagulants in prosthetic valves? *Expert Review of Cardiovascular Therapy*. 2022;20(12):905–909. https://doi.org/10.1080/14779072.2022.2152329

28. Hardefeldt H, Kiyokawa S, Mitchelson B, Finucane K, Gentles T, Wilson N. Preoperative left ventricular dysfunction is a risk factor for late death in paediatric patients after rheumatic heart disease surgery for isolated aortic valve disease. World Congress on Rheumatic Heart Disease; 2023. https://world-heart-federation.org/world-congress-on-rhd/wp-content/uploads/sites/6/2023/10/WHF-Abstracts-90.pdf.

29. Otto CM, Nishimura RA, Bonow RO, Carabello BA, Erwin JP, 3rd, Gentile F, et al. 2020 ACC/AHA guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Joint Committee on clinical practice guidelines. *Circulation*. 2021;143(5):e72–e227. https://doi.org/10.1161/CIR.0000000000000923

30. Zuhlke L, Karthikeyan G, Engel ME, Rangarajan S, Mackie P, Cupido-Katya Mauff B, et al. Clinical outcomes in 3343 children and adults with rheumatic heart disease from 14 low- and middle-income countries: two-year follow-up of the Global Rheumatic Heart Disease Registry (the REMEDY study). *Circulation*. 2016;134(19):1456–1466. https://doi.org/10.1161/CIRCULATIONAHA.116.024769

31. Gentles TL, Colan SD, Wilson NJ, Biosa R, Neutze JM. Left ventricular mechanics during and after acute rheumatic fever: contractile dysfunction is closely related to valve regurgitation. *Journal of the American College of Cardiology*. 2001;37(1):201–207. https://doi.org/10.1016/s0735-1097(00)01058-5

32. Finucane K, Wilson N. Priorities in cardiac surgery for rheumatic heart disease. *Global Heart*. 2013;8(3):213–220. https://doi.org/10.1016/j.gheart.2013.08.010

33. Rémenyi B, Webb R, Gentles T, Russell P, Finucane K, Lee M, et al. Improved long-term survival for rheumatic mitral valve repair compared to replacement in the young. *World Journal for Pediatric and Congenital Heart Surgery*. 2013;4(2):155–164. https://doi.org/10.1177/2150135112474024

34. Gentles TL, French JK, Zeng I, Milsom PF, Finucane AK, Wilson NJ. Normalized end-systolic volume and pre-load reserve predict ventricular dysfunction following surgery for aortic regurgitation independent of body size. *JACC: Cardiovascular Imaging*. 2012;5(6):626–633. https://doi.org/10.1016/j.jcmg.2011.12.021

35. Cheung MM, Sullivan ID, de Leval MR, Tsang VT, Redington AN. Optimal timing of the Ross procedure in the management of chronic aortic incompetence in the young. *Cardiology in the Young*. 2003;13(3):253–257.

36. Gentles TL, Finucane AK, Remenyi B, Kerr AR, Wilson NJ. Ventricular function before and after surgery for isolated and combined regurgitation in the young. *Annals of Thoracic Surgery*. 2015;100(4):1383–1389. https://doi.org/10.1016/j.athoracsur.2015.06.009

37. Krishna Moorthy PS, Sivalingam S, Dillon J, Kong PK, Yakub MA. Is it worth repairing rheumatic mitral valve disease in children? Long-term outcomes of an aggressive approach to rheumatic mitral valve repair compared to replacement in young patients. *Interactive Cardiovascular and Thoracic Surgery*. 2019;28(2):191–198. https://doi.org/10.1093/icvts/ivy234

38. McGurty D, Remenyi B, Cheung M, Engelman D, Zannino D, Milne C, et al. Outcomes after rheumatic mitral valve repair in children. *Annals of Thoracic Surgery*. 2019;108(3):792–797. https://doi.org/10.1016/j.athoracsur.2019.03.085

39. Tarca AJ, Causer LE, Maslin KL, Ramsay JM, Andrews DR, MacDonald BR, et al. Impact of mitral regurgitation on left ventricular remodeling and function in children with rheumatic heart disease. *International Journal of Cardiovascular Imaging*. 2022;38(12):2667–2676. https://doi.org/10.1007/s10554-022-02678-w

40. Wu DM, Buratto E, Schulz A, Zhu MZL, Ivanov Y, Ishigami S, et al. Outcomes of mitral valve repair in children with infective endocarditis: a single-center experience. *Seminars in Thoracic and Cardiovascular Surgery*. 2023;35(2):339–347. https://doi.org/10.1053/j.semtcvs.2022.05.003

41. Bolling SF, Li S, O'Brien SM, Brennan JM, Prager RL, Gammie JS. Predictors of mitral valve repair: clinical and surgeon factors. *Annals of Thoracic Surgery*. 2010;90(6):1904–1911; discussion 1912. https://doi.org/10.1016/j.athoracsur.2010.07.062

42. Madesis A, Tsakiridis K, Zarogoulidis P, Katsikogiannis N, Machairiotis N, Kougioumtzi I, et al. Review of mitral valve insufficiency: repair or replacement. *Journal of Thoracic Disease*. 2014;6(Suppl 1):S39–51. https://doi.org/10.3978/j.issn.2072-1439.2013.10.20

43. Enriquez-Sarano M, Akins CW, Vahanian A. Mitral regurgitation. *Lancet*. 2009;373(9672):1382–1394. https://doi.org/10.1016/S0140-6736(09)60692-9

44. Singh N, Haydock DA, Goh SSC. Medium-term outcomes from mitral valve surgery for rheumatic heart disease in young adults in Aotearoa New Zealand: a cohort study. *ANZ Journal of Surgery*. 2022;92(5):1060–1065. https://doi.org/10.1111/ans.17685

45. DiBardino DJ, ElBardissi AW, McClure RS, Razo-Vasquez OA, Kelly NE, Cohn LH. Four decades of experience with mitral valve repair: analysis of differential indications, technical evolution, and long-term outcome. *Journal of Thoracic and Cardiovascular Surgery*. 2010;139(1):76–83; discussion 83–84. https://doi.org/10.1016/j.jtcvs.2009.08.058

46. Liao YB, Wang TKM, Wang MTM, Ramanathan T, Wheeler M. Meta-analysis of mitral valve repair versus replacement for rheumatic mitral valve disease. *Heart, Lung & Circulation*. 2022;31(5):705–710. https://doi.org/10.1016/j.hlc.2021.11.011

47. El Sabbagh A, Reddy YNV, Barros-Gomes S, Borlaug BA, Miranda WR, Pislaru SV, et al. Low-gradient severe mitral stenosis: Hemodynamic profiles, clinical characteristics, and outcomes. *Journal of the American Heart Association*. 2019;8(5):e010736. https://doi.org/10.1161/JAHA.118.010736

48. Soesanto AM, Roeswita D, Atmosudigdo IS, Adiarto S, Sahara E. Clinical and hemodynamic factors associated with low gradient severe rheumatic mitral stenosis. *International Journal of Angiology*. 2023;32(1):43–47. https://doi.org/10.1055/s-0042-1751231

49. Sannino A, Fortuni F. Timing for intervention in aortic regurgitation: When one does not fit all. *Journal of the American College of Cardiology*. 2023;81(15):1488–1490. https://doi.org/10.1016/j.jacc.2023.02.036

50. Akintoye E, Saijo Y, Braghieri L, Badwan O, Patel H, Dabbagh MM, et al. Impact of age and sex on left ventricular remodeling in patients with aortic regurgitation. *Journal of the American College of Cardiology*. 2023;81(15):1474–1487. https://doi.org/10.1016/j.jacc.2023.02.037

51. Alashi A, Mentias A, Abdallah A, Feng K, Gillinov AM, Rodriguez LL, et al. Incremental prognostic utility of left ventricular global longitudinal strain in asymptomatic patients with significant chronic aortic regurgitation and preserved left ventricular ejection fraction. *JACC: Cardiovascular Imaging*. 2018;11(5):673–682. https://doi.org/10.1016/j.jcmg.2017.02.016

52. Egbe AC, Poterucha JT, Warnes CA. Mixed aortic valve disease: midterm outcome and predictors of adverse events. *European Heart Journal*. 2016;37(34):2671–2678. https://doi.org/10.1093/eurheartj/ehw079

53. Isaza N, Desai MY, Kapadia SR, Krishnaswamy A, Rodriguez LL, Grimm RA, et al. Long-term outcomes in patients with mixed aortic valve disease and preserved left ventricular ejection fraction. *Journal of the American Heart Association*. 2020;9(7):e014591. https://doi.org/10.1161/JAHA.119.014591

54. Zilberszac R, Gabriel H, Schemper M, Zahler D, Czerny M, Maurer G, et al. Outcome of combined stenotic and regurgitant aortic valve disease. *Journal of the American College of Cardiology*. 2013;61(14):1489–1495. https://doi.org/10.1016/j.jacc.2012.11.070

55. Onishi H, Naganuma T, Izumo M, Ouchi T, Yuki H, Mitomo S, et al. Prognostic relevance of B-type natriuretic peptide in patients with moderate mixed aortic valve disease. *ESC Heart Failure*. 2022;9(4):2474–2483. https://doi.org/10.1002/ehf2.13946

56. Knox A, Bennetts JS, Gimpel D, Newland RF, Baker RA, Joseph MX, et al. Transcatheter mitral valve-in-valve: treatment of rheumatic heart disease in young patients. *ANZ Journal of Surgery*. 2022;92(12):3298–3303. https://doi.org/10.1111/ans.18076

57. Hahn RT, Webb J, Pibarot P, Ternacle J, Herrmann HC, Suri RM, et al. 5-Year follow-up from the PARTNER 2 Aortic Valve-in-Valve Registry for Degenerated Aortic Surgical Bioprostheses. *JACC: Cardiovascular Interventions*. 2022;15(7):698–708. https://doi.org/10.1016/j.jcin.2022.02.014

58. Gozdek M, Raffa GM, Suwalski P, Kolodziejczak M, Anisimowicz L, Kubica J, et al. Comparative performance of transcatheter aortic valve-in-valve implantation versus conventional surgical re-do aortic valve replacement in patients with degenerated aortic valve bioprostheses: systematic review and meta-analysis. *European Journal of Cardio-Thoracic Surgery*. 2018;53(3):495–504. https://doi.org/10.1093/ejcts/ezx347

59. Hawkins RB, Deeb GM, Sukul D, Patel HJ, Gualano SK, Chetcuti SJ, et al. Re-do surgical aortic valve replacement after prior transcatheter versus surgical aortic valve replacement. *JACC: Cardiovascular Interventions*. 2023;16(8):942–953. https://doi.org/10.1016/j.jcin.2023.03.015

60. Topilsky Y, Maltais S, Medina Inojosa J, Oguz D, Michelena H, Maalouf J, et al. Burden of tricuspid regurgitation in patients diagnosed in the community setting. *JACC: Cardiovascular Imaging*. 2019;12(3):433–442. https://doi.org/10.1016/j.jcmg.2018.06.014

61. Muraru D, Badano LP, Hahn RT, Lang RM, Delgado V, Wunderlich NC, et al. Atrial secondary tricuspid regurgitation: pathophysiology, definition, diagnosis, and treatment. *European Heart Journal*. 2024;45(11):895–911. https://doi.org/10.1093/eurheartj/ehae088

62. Oh TH, Wang TK, Sidhu K, Haydock DA. Isolated tricuspid valve surgery at a single centre: the 47-year Auckland experience, 1965–2011. *Interactive Cardiovascular and Thoracic Surgery*. 2014;18(1):27–32. https://doi.org/10.1093/icvts/ivt452

63. Singh N, Kim H, Haydock DA. Isolated tricuspid valve surgery: The Auckland experience 2011–2019. *Heart, Lung & Circulation*. 2022;31(4):582–589. https://doi.org/10.1016/j.hlc.2021.09.020

64. Al-Jazairi AS, Althobaiti AM, Marek J, Devol EB, Al Halees Z, Mohty D, et al. Does secondary antibiotic prophylaxis improve clinical outcomes in adult rheumatic heart disease patients post-valve replacement? *World Journal for Pediatric and Congenital Heart Surgery*. 2023;14(2):161–167. https://doi.org/10.1177/21501351221139834

65. Connolly SJ, Karthikeyan G, Ntsekhe M, Haileamlak A, El Sayed A, El Ghamrawy A, et al. Rivaroxaban in rheumatic heart disease-associated atrial fibrillation. *New England Journal of Medicine*. 2022;387(11):978–988. https://doi.org/10.1056/NEJMoa2209051

66. Kido K, Ball J. Optimal intensity of warfarin therapy in patients with mechanical aortic valves. *Journal of Pharmacy Practice*. 2019;32(1):93–98. https://doi.org/10.1177/0897190017734765

67. Thompson JL, Burkhart HM, Daly RC, Dearani JA, Joyce LD, Suri RM, et al. Anticoagulation early after mechanical valve replacement: improved management with patient self-testing. *Journal of Thoracic and Cardiovascular Surgery*. 2013;146(3):599–604. https://doi.org/10.1016/j.jtcvs.2012.03.088

68. Witt DM, Nieuwlaat R, Clark NP, Ansell J, Holbrook A, Skov J, et al. American Society of Hematology 2018 guidelines for management of venous thromboembolism: optimal management of anticoagulation therapy. *Blood Advances*. 2018;2(22):3257–3291. https://doi.org/10.1182/bloodadvances.2018024893

69. Regier DA, Sunderji R, Lynd LD, Gin K, Marra CA. Cost-effectiveness of self-managed versus physician-managed oral anticoagulation therapy. *CMAJ: Canadian Medical Association Journal*. 2006;174(13):1847–1852. https://doi.org/10.1503/cmaj.051104

70. Heneghan CJ, Garcia-Alamino JM, Spencer EA, Ward AM, Perera R, Bankhead C, et al. Self-monitoring and self-management of oral anticoagulation. *Cochrane Database of Systematic Reviews*. 2016;7:CD003839. https://doi.org/10.1002/14651858.CD003839.pub3

71. Matchar DB, Jacobson A, Dolor R, Edson R, Uyeda L, Phibbs CS, et al. Effect of home testing of international normalized ratio on clinical events. *New England Journal of Medicine*. 2010;363(17):1608–1620. https://doi.org/10.1056/NEJMoa1002617

72. Soper J, Chan GT, Skinner JR, Spinetto HD, Gentles TL. Management of oral anticoagulation in a population of children with cardiac disease using a computerised system to support decision-making. *Cardiology in the Young*. 2006;16(3):256–260. https://doi.org/10.1017/S1047951106000333

73. Webb R, Voss L, Roberts S, Hornung T, Rumball E, Lennon D. Infective endocarditis in New Zealand children 1994-2012. *Pediatric Infectious Disease Journal*. 2014;33(5):437–442. https://doi.org/10.1097/INF.0000000000000133

74. Birrell JM, Evans T, Fisher R, Davis A, Wilkinson L. The economic and health burden of infective endocarditis in Northland, New Zealand. *New Zealand Medical Journal*. 2022;135(1551):13–24.

75. Alsamarrai A, Saavedra C, Bryce A, Dimalapang E, Leversha A, Briggs S, et al. Infective endocarditis in patients with rheumatic heart disease: a single-centre retrospective comparative study. *New Zealand Medical Journal*. 2022;135(1550):62–73.

76. Habib G, Lancellotti P, Antunes MJ, Bongiorni MG, Casalta JP, Del Zotti F, et al. 2015 ESC Guidelines for the management of infective endocarditis: The Task Force for the Management of Infective Endocarditis of the European Society of Cardiology (ESC). Endorsed by: European Association for Cardio-Thoracic Surgery (EACTS), the European Association of Nuclear Medicine (EANM). *European Heart Journal*. 2015;36(44):3075–3128. https://doi.org/10.1093/eurheartj/ehv319

77. Dougherty S, Essop MR, Webb R, Price S, Wilson N. Chapter 16 — Complications of rheumatic heart disease and acute emergencies. In: Dougherty S, Carapetis J, Zühlke L, Wilson N, editors. Acute rheumatic fever and rheumatic heart disease: Elsevier; 2020. p. 301–336.

78. Cahill TJ, Harrison JL, Jewell P, Onakpoya I, Chambers JB, Dayer M, et al. Antibiotic prophylaxis for infective endocarditis: a systematic review and meta-analysis. *Heart*. 2017;103(12):937–944. https://doi.org/10.1136/heartjnl-2015-309102

79. Rutherford SJ, Glenny AM, Roberts G, Hooper L, Worthington HV. Antibiotic prophylaxis for preventing bacterial endocarditis following dental procedures. *Cochrane Database of Systematic Reviews*. 2022;5:CD003813. https://doi.org/10.1002/14651858.CD003813.pub5

80. Heart Foundation. New Zealand guideline for prevention of infective endocarditis associated with dental and other medical interventions. Heart Foundation; 2008. https://www.heartfoundation.org.nz/resources/prevention-of-infective-endocarditis (Accessed December 16 2024).

81. National Institute for Health and Care Excellence. Dental recall: recall interval between routine dental examinations. National Institute for Health and Care Excellence; 2004. https://www.nice.org.uk/guidance/cg19/evidence/full-guideline-pdf-193348909 (Accessed December 16 2024).

82. Doughty RN, Devlin G, Wong S, McGrinder H, Chirnside J, Sinclair L. 2023 position statement on improving management for patients with heart failure in Aotearoa New Zealand. *New Zealand Medical Journal*. 2023;137(1590):93–99.

83. McDonagh TA, Metra M, Adamo M, Gardner RS, Baumbach A, Bohm M, et al. 2021 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure. *European Heart Journal*. 2021;42(36):3599–3726. https://doi.org/10.1093/eurheartj/ehab368

84. Bermejo J, Yotti R, Garcia-Orta R, Sanchez-Fernandez PL, Castano M, Segovia-Cubero J, et al. Sildenafil for improving outcomes in patients with corrected valvular heart disease and persistent pulmonary hypertension: a multicenter, double-blind, randomized clinical trial. *European Heart Journal*. 2018;39(15):1255–1264. https://doi.org/10.1093/eurheartj/ehx700

85. Humbert M, Kovacs G, Hoeper MM, Badagliacca R, Berger RMF, Brida M, et al. 2022 ESC/ERS guidelines for the diagnosis and treatment of pulmonary hypertension. *European Respiratory Journal*. 2022;61(1):2200879. https://doi.org/10.1093/eurheartj/ehac237

86. He VY, Condon JR, Ralph AP, Zhao Y, Roberts K, de Dassel JL, et al. Long-term outcomes from acute rheumatic fever and rheumatic heart disease: a data-linkage and survival analysis approach. *Circulation*. 2016;134(3):222–232. https://doi.org/10.1161/CIRCULATIONAHA.115.020966

87. Cremer PC, Rodriguez LL, Griffin BP, Tan CD, Rodriguez ER, Johnston DR, et al. Early bioprosthetic valve failure: Mechanistic insights via correlation between echocardiographic and operative findings. *Journal of the American Society of Echocardiography*. 2015;28(10):1131–1148. https://doi.org/10.1016/j.echo.2015.07.003

88. Cremer PC, Rodriguez LL, Griffin BP, Tan C, Rodriguez R, Johnston DR, et al. Early Bioprosthetic Valve Failure: A Pictorial Review of Rare Causes. *JACC: Cardiovascular Imaging*. 2015;8(6):737–740. https://doi.org/10.1016/j.jcmg.2014.06.025

89. Johnston DR, Soltesz EG, Vakil N, Rajeswaran J, Roselli EE, Sabik JF, 3rd, et al. Long-term durability of bioprosthetic aortic valves: implications from 12,569 implants. *Annals of Thoracic Surgery*. 2015;99(4):1239–1247. https://doi.org/10.1016/j.athoracsur.2014.10.070

12

Rheumatic Heart Disease and Pregnancy

In this chapter

“Because when I was pregnant, I got really out of breath, and like I couldn’t really breathe properly like when I would lie down, I couldn’t really breathe unless I was sitting up. Then got it checked and that was it [RHD.]”

Pregnant person with RHD

Key changes

This is a new chapter and includes the following changes:

- Inclusion of the modified World Health Organization (mWHO) classification of maternal risk of valvular heart disease during pregnancy.
- Referral to a tertiary centre may be appropriate for managing pregnant people/women with moderate to severe rheumatic heart disease (RHD).
- A treadmill exercise stress echocardiogram (echo) before pregnancy can help identify lesions at risk of decompensation.
- Updated dosage and frequency recommendations for monitoring anticoagulation during pregnancy.
- Guidance on managing mechanical prosthetic valve thrombosis (MPVT) in pregnancy.
- Links to external resources for prescribing medication during pregnancy and breastfeeding.

Key points

- Pre-conception counselling is strongly recommended for people with RHD who can become pregnant.
- Collaborative care is essential, ensuring clear communication between specialists, midwives, and whānau.
- Planned pregnancies and pre-conception assessments are associated with better pregnancy outcomes in those with cardiac disease.
- This chapter includes the internationally endorsed WHO classification, detailing pregnancy risks for specific valvular heart disease lesions.
- Mitral stenosis (MS) of any severity can cause decompensation during pregnancy.
- Anticoagulation for mechanical prosthetic heart valves poses risks to the pregnant person/woman and pēpi. Anticoagulation regimens are detailed.

Recommendations

- Antenatal care for people with RHD should begin early from gestation to reassess risk factors and detect changes in cardiac status since the previous assessment (Grade D).
- Anticoagulation regimens for those with mechanical heart valves are best managed by a multi-disciplinary, high-risk maternity team. Birthing is recommended to be at a centre with expertise in bridging peri-birth and postpartum anticoagulation, with access to cardiothoracic intensive care in case of acute valve thrombosis (Evidence level III-3, Grade C).

Introduction

Cardiac disease, including RHD, remains the second leading cause of death during pregnancy and the postpartum period in Aotearoa after suicide. Between 2006 and 2020, it accounted for 10.7% of all pregnancy-related deaths, surpassing the combined deaths from hypertensive disorders of pregnancy, venous thromboembolism, and obstetric haemorrhage.[1, 2]

Several factors contribute to poor outcomes in pregnant people/women with cardiac disease, including:

- Delayed pregnancy booking, leading to missed opportunities for early intervention.
- Delayed access to healthcare due to rural locality, socioeconomic disadvantage, poor health literacy, and cultural or health belief differences.
- Symptoms mistakenly attributed to pregnancy rather than underlying cardiac disease, delaying diagnosis and treatment.
- Gaps in high-risk maternity care and variability in care between centres.

RHD can be a silent condition with minimal symptoms before pregnancy and may first be diagnosed during pregnancy. The diagnosis could be made:

- By chance, when a cardiac murmur is investigated.
- Following presentation with tachyarrhythmia or symptoms of decompensated heart failure.
- Incidentally, due to heightened clinical suspicion based on ethnicity or demographic risk factors.[2]

A single-centre registry of contemporary prospective cases examining women with heart disease in a low- and middle-income country found that 60.5% of the 1,005 women were diagnosed with heart disease for the first time during pregnancy.[3] RHD was the most common heart condition identified in this study.

The Australasian Maternity Outcomes Surveillance System (AMOSS) study found that 4.3 in 10,000 women giving birth had RHD.[4] The study, which surveyed the epidemiology of RHD in Australia and Aotearoa over 2 years from January 2013, also found that 90% of affected women in Aotearoa were Māori and Pacific peoples. RHD was associated with high rates of postpartum admissions to coronary or intensive care units (10%) and pre-term births (21%). Since then, anecdotally, many cardiologists and obstetric physicians have reported cases of previously unrecognised RHD leading to cardiac decompensation during pregnancy or in the immediate postpartum period.

When a pregnant person/woman with RHD presents acutely unwell with decompensated heart failure, the health of the pregnant person/woman takes priority. A multi-disciplinary team making decisions about their ongoing care will follow set principles and consider the following:

- The optimal treatment for the person, independent of pregnancy.
- The gestation of the pregnancy.
- The person's perspective on continuing the pregnancy.

Pre-conception assessment and counselling

People with known RHD should be given the opportunity to discuss their plans to have whānau at every routine clinical assessment, where appropriate. Starting these conversations early helps normalise this aspect of care for patients and whānau.

Counselling should ideally include:

- A discussion about effective contraception options, with referral to an appropriate service if the person agrees.
- Consider cardiac disease and other relevant medical history.[5]
- Unplanned pregnancies are common.[5] It is important to explain that RHD does not necessarily reduce fertility, but careful pregnancy planning is essential.
- Emphasising the role of healthcare providers in supporting pregnancy planning.

Managing lesions

The purpose of pre-conception assessment is to identify clinically important heart lesions that carry a significant risk of cardiac events during pregnancy. For those with severe RHD, maternity specialists, and cardiologists should consider whether interventional therapy such as balloon valvuloplasty or valve surgery (either valve repair or replacement) is needed before pregnancy. The choice of valve replacement in a young person should be informed by their pregnancy plans.[6] Ideally, pregnancy should only be attempted once RHD is optimally managed.

People with stable moderate to severe lesions may not require immediate surgical or catheter intervention but remain at risk of decompensation due to the cardiovascular changes of pregnancy. Making an informed choice about pregnancy while managing the potential health risks is often challenging, therefore, expert counselling by a multi-disciplinary team is highly recommended.

Discussing medication and other risk factors

Pre-pregnancy assessment by specialists provides an opportunity to review and advise on medications that:

- May be teratogenic (harmful to the baby/pēpi), such as warfarin used for mechanical prosthetic heart valves.
- Must be discontinued when pregnancy is detected, such as angiotensin-converting enzyme inhibitors (ACEi) like enalapril or angiotensin-receptor blockers (ARB) like candesartan used for cardiac remodelling.

For those on warfarin, appropriate contraception counselling is essential to avoid unplanned pregnancy. People need to be informed that they must not stop their warfarin without consulting their specialist, even if they think they might be pregnant or are pregnant. (Also see Chapter 1: Cultural Responsiveness and Chapter 11: Management of Rheumatic Heart Disease.)

Other important discussion points in the pre-conception assessment include modifiable risk factors such as smoking and excess weight. Prenatal supplementation with a standard dose of folic acid should be offered to all people planning a pregnancy.

Involving social support

Involving the whānau and wider support networks in the pre-conception counselling is important and should be offered when appropriate. Discussions should include how pregnancy-related physiological changes can impact heart function and, in turn, affect both maternal and fetal outcomes. For those with moderate or severe RHD, decompensation can occur when the fetus (baby) is peri-viable, leaving only two options: delivering an extremely premature pēpi or undergoing valve intervention during pregnancy. These discussions should be tailored to individual contexts and understanding, with whānau support and interpreters available as required.

Counselling by a multi-disciplinary team

All pregnant people/women with cardiac valve disease, including RHD, should be referred to an Obstetric and related medical service for assessment following standard maternity guidelines.[7] Pregnant people/women should be supported to receive counselling closer to home at their domicile hospital. Still, referral to a tertiary centre may be more appropriate for those with moderate to severe cardiac disease. Some larger centres have dedicated high-risk maternity services where a multi-disciplinary team provides pre-conception counselling. The team typically includes Maternal Fetal Medicine specialists, Obstetric Physicians, and Cardiologists.

Risk prediction with scoring systems

Several historical risk scoring tools (CARPREG,[8] CARPREG II[9] and ZAHARA[10]) are available to estimate the risk of cardiac events during pregnancy. However, these tools were primarily developed for pregnant patients with congenital heart disease, limiting their accuracy in predicting risk for RHD.

The widely accepted mWHO classification provides a 'simplified' approach based on lesions to estimate risk (see **Table 12.1**). International cardiac societies endorse this classification.[11-14] The mWHO classification also identifies conditions where the risk of pregnancy is considered prohibitive. However, because this classification system is based solely on the type of lesion to estimate risk, it does not consider other important contributing factors that may indirectly affect outcomes, such as geography, demographics, and access to healthcare.

Table 12.1. The modified World Health Organization classification of risk of valvular heart disease during pregnancy

	mWHO I	mWHO II	mWHO III	mWHO IV
Lesion	Mild asymptomatic RHD	Mild MS Moderate Aortic Stenosis (AS) LVEF >45%	Moderate MS Severe asymptomatic AS Severe asymptomatic mitral regurgitation (MR) or aortic regurgitation (AR) Left ventricular ejection fraction (LVEF) 30–45% Mechanical prosthetic heart valve	Severe MS Severe symptomatic AS Severe symptomatic MR or AR LVEF <30% or New York Heart Association (NYHA) class III–IV
Risk of cardiac events	Not increased	Small	Intermediate	Very high (pregnancy may be contraindicated, termination to be discussed)
Maternal cardiac event rate	2.5–5%	5–10%	10–19%	40–100%
Location of pregnancy care	Local	Local	Centre with expertise	Centre with expertise
Number of visits during pregnancy	1–2	Once per trimester	Monthly	Monthly or twice per month
Location of birth	Local	Local	Centre with expertise	Centre with expertise

The following five general and lesion-specific risk factors have been identified in various risk-scoring systems as highly associated with maternal cardiac events:[11-13]

1. Left-sided valve disease (mitral or aortic stenosis; severe mitral or aortic regurgitation).
2. Symptomatic RHD.
3. Impaired left ventricular systolic function.
4. Moderate or severe pulmonary hypertension.
5. Prior history of heart failure or arrhythmia.

Risk scores cannot capture all nuances of a case. Clinical judgement remains essential in risk stratification, highlighting the importance of expert assessment of heart disease during pregnancy.

Treadmill exercise stress echocardiogram

Risk assessment should also assess baseline functional status. A treadmill exercise stress echo before pregnancy can serve as a 'surrogate' for the cardiovascular changes in pregnancy, potentially identifying lesions at risk of decompensation during pregnancy.[11]

A functional stress echo could be part of pre-conception or early pregnancy risk stratification in tertiary centers with appropriate resources and expertise.[11] Pregnant people/women with known moderate to severe disease (mWHO III and mWHO IV) should be discussed with tertiary centres for consideration of a pre-conception treadmill exercise stress echo.

A retrospective cohort study of 16 women with known left-sided valve stenosis (nine with MS, seven with AS) used a screening treadmill stress echo as part of a pre-conception risk assessment.[15] Women who were screened were supported to proceed with pregnancy if they completed more than 7 METS on the standard Bruce protocol without developing pulmonary hypertension (pulmonary artery systolic pressure >60 mmHg). Maternal and pregnancy outcomes were compared with those who conceived without the stress echo. In those with MS, baseline NYHA results were documented (class I: 3 women, class II: 5 women, class III: 1 woman). None of the nine women with MS had a major cardiac event or required valve intervention during pregnancy, though two had documented functional decline in their pregnancy.

B-type natriuretic peptide

B-type natriuretic peptide (BNP), when used alongside other risk assessment tools, may help identify pregnancies at the highest risk of cardiac complications. Checking BNP at the beginning and then at intervals throughout pregnancy may be particularly helpful. In a Canadian cohort of 86 pregnant people/women with cardiac disease (only 4% with RHD), a BNP level ≤100 pg/ml was associated with a negative predictive value of 100% for adverse maternal cardiac events.[16] However, specific data linking BNP levels with rheumatic MS (the classic RHD lesion that causes decompensation with pregnancy) does not exist.

Table 12.2. Pre-conception counselling and assessment for pregnancies with rheumatic heart disease

Classification	mWHO I Low-risk maternal cardiac event (2.5–5%)	mWHO II Intermediate-risk maternal cardiac event (10–19%)	mWHO III High-risk maternal cardiac event (20–27%)	mWHO IV Extremely high-risk maternal cardiac event (40–100%)
Category description	• History of acute rheumatic fever (ARF) without carditis or mild MR or AR • Mild asymptomatic RHD	• Bioprosthetic valve or previous PBMV • Mild MS • Moderate AS • Moderate MR or AR • Mild LV impairment with EF>45% without severe regurgitation or stenosis and good functional capacity	• MPHV • Moderate MS with MVA 1.5–2.0 cm^2 • Severe asymptomatic AS • Severe asymptomatic MR or AR • Moderate LV impairment with EF 30–45%	• Mitral or aortic valve disease with pulmonary hypertension • Severe MS with MVA <1.5 cm^2 Pregnancy is prohibitive in the following. Discussing the termination of pregnancy may be required. • Severe symptomatic MS with MVA <1.0 cm^2 • Severe symptomatic AS • Severe LV impairment with EF <30% • NYHA class III/IV
Pre-conception counselling	• Enquire about plans to start a whānau. Supplement with prenatal folic acid if pregnancy is planned • Advise on effective contraception if pregnancy is not planned or if significant cardiac disease is identified (mWHO IV) • Review medications (ACEi, ARB, warfarin, heart failure therapy) • Optimise general health (for example, reducing weight, stopping smoking) • Involve partner and whānau if appropriate • Consider the location of counselling. A tertiary centre with multi-disciplinary specialties may be more appropriate for mWHO III and IV			
Care during pregnancy	• Consider an updated echo if cardiac symptoms are new • Counselling and care at the local hospital are appropriate • Lead maternity care with a midwife may be appropriate	• An updated echo is recommended • Counselling at the local hospital may be considered • A shared model of pregnancy care between lead maternity carer, local and tertiary hospitals may be appropriate • Giving birth at a secondary hospital is recommended	• An updated echo is recommended • Counselling at a tertiary hospital is recommended • A shared model of pregnancy care between local and tertiary hospitals is recommended • Giving birth at a tertiary hospital is recommended	• An updated echo is recommended • Counselling at a tertiary hospital is recommended • Shared model of pregnancy care between local and tertiary hospitals with a low threshold for transfer of care to a tertiary hospital
Frequency of visits	• When pregnant, the frequency of visits follows standard maternity guidelines • Review with specialists as indicated	• When pregnant, review with specialists once per trimester but consider more frequent visits from 28 weeks	• When pregnant, review with specialists every month but increase the frequency of visits from 24 weeks	• When pregnant, review with specialists every month but increase the frequency of visits from 20 weeks

Adapted from Regitz-Zagrosek (2011)[11] and Bonow (2008)[17].

Key:

EF	Ejection fraction	**MVA**	Mitral valve area	**NYHA**	New York Heart Association
LV	Left ventricular	**MVO**	Mitral valve orifice	**PBMV**	Percutaneous balloon mitral valvuloplasty
MPHV	Mechanical prosthetic heart valve	**mWHO**	Modified World Health Organization		

In low- and middle-income countries, echocardiographic screening for RHD is justified by the high maternal mortality associated with RHD.[2, 18, 19] The current recommendations for echo referral during pregnancy are detailed in **Chapter 14: Screening for Rheumatic Heart Disease**.

Health professionals need to be aware of unrecognised RHD among Māori and Pacific peoples. Some people may be unaware they have RHD, but clinicians should seek a history of previous episodes of arthritis, ARF, the need for monthly injections, previous echo's, or whānau members with ARF or RHD. Current evidence does not support routine antenatal screening for all Māori and Pacific peoples during pregnancy.

Cardiac referral for echo should have a low threshold, as breathlessness in pregnancy may be difficult to distinguish from early cardiac decompensation, and auscultation is unreliable.

How pregnancy changes the cardiovascular system

The cardiovascular system changes in several ways during pregnancy.[16]

- Maternal blood volume increases by an average of 50%. It rises rapidly from as early as six weeks gestation to the mid-trimester, then more slowly until term gestation.[20]
- Systemic vascular resistance drops correspondingly, helping to maintain the physiological low blood pressure until the end of the second trimester. Blood pressure then rises to its pre-pregnancy level.
- Cardiac output increases by about 50% during pregnancy, reaching its peak at 24–28 weeks' gestation.[20]

During labour, maternal cardiac output increases by a further 50%.[20] During labour and birth, the contracting uterus auto-transfuses blood into the maternal systemic circulation, increasing stroke volume. Heart rate can also increase during labour and birth due to pain and anxiety. The Valsalva manoeuvre in the second stage of labour increases intrathoracic pressure and reduces venous return. This activates the baroreceptor reflex, which causes heart rate and systemic vascular resistance to rise. Aortocaval compression from the gravid uterus in a supine maternal position during labour can reduce blood flow to the placenta.

After birth, blood returns to the maternal circulation as a result of the immediate relief of aorto-caval compression and involution of the uterus.[20] Fluid shifts between maternal compartments occur with the resolution of peripheral oedema, which typically happens over the days to weeks postpartum. These postpartum changes contribute to the high stroke volume and high cardiac output seen in the first few days after birth.

How pregnancy can affect rheumatic heart disease

The maternal cardiac workload peaks at three important times (see **Figure 12.1**):

- Between 24 to 28 weeks.
- During the second stage of labour.
- In the first 24 to 48 hours postpartum.[20]

Pregnant people/women with valvular disease who are unable to meet the challenge of the increased cardiac output at these critical times have a higher risk of decompensation with heart failure and may even face the risk of death.

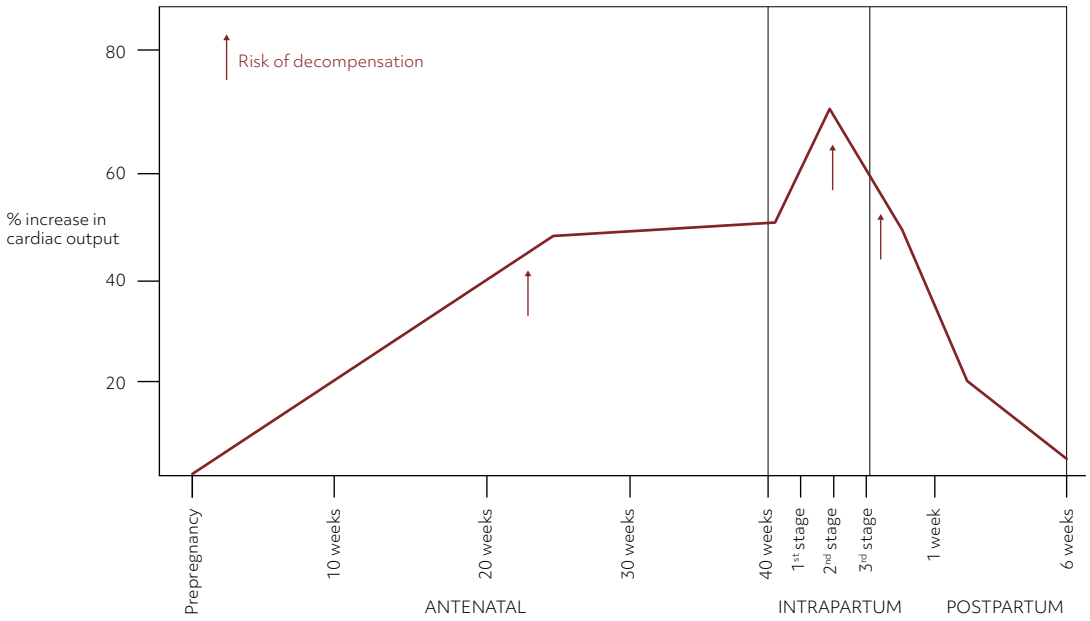


Figure 12.1. Times of peak cardiac output and decompensation risk[20]

This line graph shows the percentage increase in cardiac output from pre-pregnancy to six weeks postpartum. Cardiac output increases by about 50% around 24 weeks. It continues to rise, peaking at the second stage, intrapartum. It then falls steadily to nearly pre-pregnancy levels at six weeks postpartum.

Lesions and the risk of decompensation

Regurgitant valvular lesions generally tolerate a higher cardiac output state better than stenotic lesions. The decrease in systemic vascular resistance and the left ventricle's ability to stretch to accommodate the higher stroke volume provides some protection for regurgitant lesions. However, the overall context of the cardiac disease is equally important in determining the tolerability of the valve lesion during pregnancy. For example, severe MR combined with a failing left ventricle and pulmonary hypertension may pose a risk level comparable to an 'uncomplicated' severe MS.

Stenotic lesions are associated with fixed obstructions, which pose significant concerns during pregnancy (see **Figure 12.2**). With MS, the physiological tachycardia of pregnancy reduces the time available for blood to empty from the left atrium during diastole (1). This underfills the left ventricle (2) while simultaneously increasing the left atrial pre-load (3).[20] The increased pre-load, in turn, increases the pulmonary pressures (4). Coupled with the lower oncotic pressure in pregnancy, the increased pulmonary pressure can lead to acute pulmonary oedema (5).

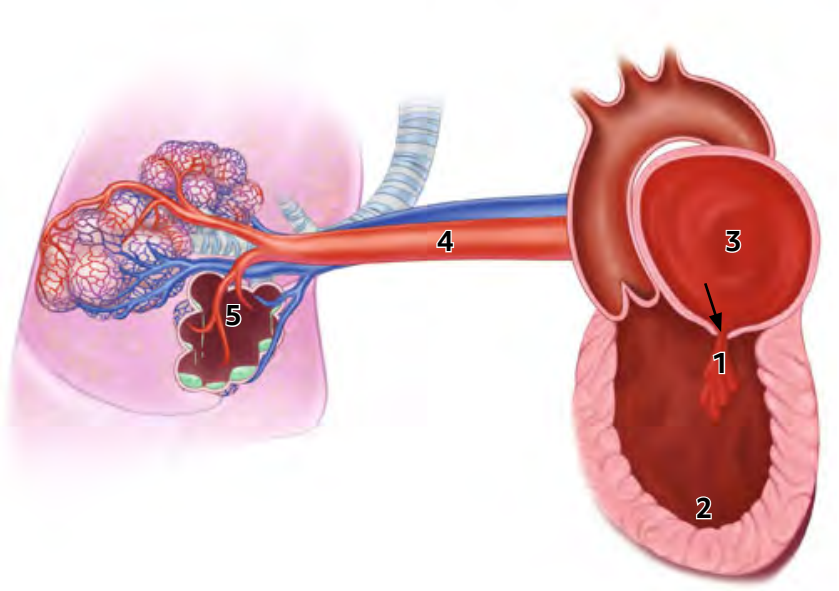


Figure 12.2. Pathogenesis of pulmonary oedema in pregnancy with mitral stenosis[21]

Johal K, Lau L, Card ME. Mitral stenosis. In: Taylor A, editor. Learning cardiac auscultation: Springer, 135–149, 2025, reproduced with permission from SNCSC.

The risk of decompensation is greatest in those with severe MS, but even mild MS should be considered a risk factor.[22] (see **Table 12.1**) Atrial fibrillation (AF) with an enlarged left atrium can further compromise the emptying of the left atrium, reducing cardiac output and worsening pulmonary hypertension. The need for anticoagulation with valvular AF adds another layer of complexity.

Impact of rheumatic heart disease on pregnancy outcomes

The three most common cardiac complications associated with valvular disease in pregnancy are arrhythmia, heart failure, and thromboembolism. Maternal deaths from RHD are still reported in some countries despite being preventable.[23]

In pregnant people/women experiencing cardiac decompensation, the first-line management involves admission for bed rest to:

- Reduce the additional cardiac workload imposed by physical activity.
- Initiate appropriate medical treatment.

Beta-blockers and digoxin, commonly prescribed for rate control of AF, are safe in pregnancy.[11] Diuretic treatment with furosemide for managing fluid overload and nitrates to reduce cardiac afterload are also compatible and efficacious with pregnancy.[11]

Episodes of maternal decompensation can compromise placental perfusion, leading to a higher incidence of fetal growth restriction. Regular fetal growth surveillance is recommended for all pregnant people/women with RHD, particularly those with moderate to severe disease.

Valve intervention (balloon valvuloplasty or valve surgery) should be considered for those with refractory symptoms despite medical therapy. Percutaneous balloon mitral valvuloplasty (PBMV), if the valve characteristics allow, is often preferred over valve surgery given the high maternal morbidity risk and the risk of fetal loss of 20 to 30% associated with cardiac surgery and bypass.[11, 24] Favourable outcomes have been reported with PBMV during pregnancy.[25] However, PBMV should only be performed by experienced operators in tertiary centres with cardiothoracic intensive care units available, as procedural complications like torrential MR may necessitate emergency birth to facilitate maternal resuscitation.

Essential components of counselling pregnant people/women with severe symptomatic RHD at significant risk of early decompensation should include:

- Termination of pregnancy, especially for those at risk of becoming critically unwell before reaching viability.
- Birth around the peri-viability of gestation, including the risk of operative morbidity for the pregnant person/woman and long-term fertility implications.
- Pre-term birth risks and complications of prematurity.

Cardiac complications can also be compounded by pregnancy-related disorders like preeclampsia, especially in severe AS, MS, or severe AR with left ventricular impairment. Acute decompensation may also occur following fluid resuscitation for severe blood loss from postpartum haemorrhage, potentially unmasking previously undiagnosed stable MS.

Pregnancy care for people with rheumatic heart disease

People with RHD who wish to become pregnant should have a pre-conception plan in place, ideally prior to the pregnancy, for the pregnancy, and after birth.

Antenatal care

People with heart disease who receive pre-conception counselling and have planned pregnancies generally have better pregnancy outcomes.[22, 23, 26] Unplanned pregnancies or lack of pre-pregnancy counselling are associated with poorer outcomes.

Early antenatal assessment

If more than one year has passed since the pre-conception review, antenatal care should start from early gestation (ideally in the first trimester or as soon as pregnancy is confirmed). Early referral is crucial for re-evaluating risk factors and assessing for changes in cardiac status. Serial cardiac surveillance should be tailored to RHD severity and clinical symptoms (see Table 12.2). An echo assessment at 24–28 weeks at the stage of peak cardiac output can guide the frequency of ongoing echo and clinical surveillance.

A study of 47 women with MS found that certain baseline and third-trimester echocardiographic characteristics predicted adverse cardiac events:

- Mitral valve gradient (MG) >10 mmHg.
- Right ventricular systolic pressure (RSPV) >40 mmHg.[22]

For those with MS, an MVA ≤1.2 cm^2 at any stage of pregnancy is a strong predictor of the risk of maternal pulmonary oedema, especially if symptoms are already present.[27] (Also see **Chapter 10: Diagnosis of Rheumatic Heart Disease**).

Pregnant people/women, their whānau, and support networks should receive accessible resources to assist their understanding of early warning signs of cardiac deterioration. Breathlessness, a common physiological symptom of pregnancy, can be challenging to distinguish from early cardiac decompensation.

In high-risk cases (see **Table 12.1**), baseline and serial BNP measurements may help identify important clinical changes.[16] BNP levels remain low in normal pregnancies.[28, 29]

Iron status should be monitored and optimised opportunistically, particularly for those at higher risk of bleeding complications.

Fetal growth should be monitored closely and updated when the clinical status changes or an acute cardiac event occurs.

Pregnant people/women should continue to receive routine midwifery or lead maternity care throughout pregnancy. Infant feeding discussions should occur early, with a breastfeeding support plan in place in case of complications after the birth.

Planning for birth

Given the unpredictability of labour, the birth location should be discussed early, with a contingency plan for out-of-hours emergencies. Pregnant people/women and their whānau should be offered the opportunity to meet with the multi-disciplinary team during pregnancy to establish rapport and clarify care plans.

Intrapartum care

Pregnant people/women with uncomplicated RHD can safely have a vaginal birth. Caesarean section should be reserved for those with an obstetric indication. The maternity early warning system (MEWS) should be used to record all maternal observations. For complex or severe lesions, the mode of birth should be decided on a case-by-case basis, together with the multi-disciplinary team, considering:

- Obstetric history.
- Future whānau planning, for example, whether they have completed their whānau and would plan tubal sterilisation.
- Other cardiac findings, such as left ventricular systolic impairment or pulmonary hypertension.
- Resources available at the local birth centre including cardiac monitoring, access to specialist cardiac anaesthesia, and cardiothoracic intensive care.
- Gestation at birth and availability of neonatal intensive care beds.

- Out-of-hours emergency management, for example, whether a planned induction of labour during working hours is preferable.
- The person's preference.

Pregnant people/women in the higher risk groups who wish to have a vaginal birth can be supported with careful planning, including:

- Early epidural placement to minimise the sympathetic drive on heart rate and blood pressure.
- Fluid restriction during labour and detailed fluid balance monitoring.
- Assisted instrumental birth (with or without maternal effort) to shorten the second stage.
- Diuretic administration in the second stage to reduce the impact of auto-transfusion from involution of the uterus after birth.
- Selective use of ecbolics, for example, avoiding ergotamine in hypertension or preeclampsia, or prostaglandin F 2-alpha (such as Carboprost) in pulmonary hypertension.
- Active third-stage management to prevent postpartum haemorrhage.

While there is no absolute indication for an elective caesarean section in the context of RHD, many clinicians would be concerned about the following lesions:

- Severe MS with severe pulmonary hypertension.
- Severe AR or MR with severe left ventricular impairment (LVEF <30%).
- Severe symptomatic AS.

Vaginal, assisted instrumental, and caesarean births are not associated with an increased risk of bacterial endocarditis. Therefore, routine endocarditis prophylaxis is not indicated. Prophylactic antibiotics should be used as per standard obstetric indications.

Postpartum care

Early discharge may not be suitable for those with moderate to severe RHD, even after an uncomplicated birth, due to the ongoing risk of postpartum decompensation. People with RHD who receive intravenous fluid as part of resuscitation for puerperal sepsis or postpartum haemorrhage must be monitored closely for symptoms of heart failure. Those with moderate to severe RHD should not be transferred early to primary birthing units for postnatal care.

Finalise the person's contraception plan if not yet finalised. All newly diagnosed cases of RHD during pregnancy should be followed up in a cardiology clinic and linked with a community provider for secondary penicillin prophylaxis (SAP), if appropriate. (Also see Chapter 8: Secondary Prevention).

Pregnancy after valve replacement: bioprosthetic versus mechanical prosthetic valve

Prophylactic valve replacement before pregnancy is only indicated in those who meet the criteria for intervention. For those qualifying for valve replacement, the choice between a bioprosthetic and a mechanical prosthetic valve requires careful consideration of multiple factors, including:

- Whether there is another indication for anticoagulation, like concurrent AF.
- Concerns about adherence to treatment.
- Risks associated with re-do valve surgery.
- Fertility history and planning.
- The person's preference.

Bioprosthetic valves

The major advantage of bioprosthetic valves in pregnancy is that anticoagulation is not required, and some may choose this option. However, bioprosthetic valves do not last as long as mechanical valves and structural degeneration increases the likelihood of re-operation. Pregnancy with a normal functioning bioprosthetic valve is well tolerated and does not appear to accelerate structural valve degeneration.[30, 31] However, before considering pregnancy, an up-to-date assessment of bioprosthetic valve function or prior valve repair should be undertaken.

Mechanical prosthetic valves

A mechanical prosthetic heart valve (MPHV) is the most durable option for adults but requires lifelong anticoagulation with a vitamin K antagonist (VKA).

People with MPHV represent one of the highest-risk groups in pregnancy due to the need for therapeutic anticoagulation. Anticoagulation is essential to prevent valve thrombosis and its sequelae of valve dysfunction or valve failure and systemic thromboembolism. Achieving optimal anticoagulation is challenging as it must balance preventing thromboembolic events and minimise the risk of major bleeding from excessive anticoagulation treatment.[32-36]

Anticoagulation in pregnancy with a mechanical prosthetic heart valve

The ideal anticoagulation in pregnant people/women with MPHV remains contentious.[11, 17, 32] No anticoagulation option is completely safe for both the mother and pēpi.[32, 37, 38] For this reason, some people with an MPHV will choose to avoid pregnancy altogether.

Oral VKA, like warfarin, is the treatment of choice for MPHV, but it can cross the placenta and affect the fetus at all stages of development. Warfarin-related embryopathy occurs with first-trimester exposure (between 6 to 12 weeks). Importantly, fetopathy and fetal anticoagulation also occur when warfarin is used after 12 weeks. The effect on fetal anticoagulation is dose-dependent, with a daily dose >5 mg associated with higher fetal risks[33, 38] (see Table 12.3).

The warfarin dose must be based on the international normalised ratio (INR) levels. Pregnant people/women should not opt to reduce the warfarin dose if it results in a subtherapeutic INR, as this increases the risk of prosthetic valve thrombosis.

Table 12.3. Fetal effects of vitamin K antagonist exposure after the first trimester

Warfarin fetopathy	Fetal anticoagulation
Central nervous system abnormalities: hydrocephalus, spasticity, hypotonia, intellectual disability **Ocular abnormalities:** microphthalmia, cataract, optic atrophy	Central nervous system bleeding Stillbirth

VKAs are associated with lower live birth rates. A systematic review and meta-analysis of 2,468 pregnancies in women with MPHV found:

- Live birth rates were lower when VKA was used as the main anticoagulation throughout pregnancy, compared to low molecular weight heparin (LMWH) (64.5% versus 92%).[35]
- The live birth rate improved to 79.97% with sequential treatment that avoids first-trimester VKA exposure.
- Live birth rates were 83.6% when warfarin was used at ≤5 mg (either as the predominant treatment or part of sequential treatment), but dropped to 43.9% with doses >5 mg.

The true incidence of fetopathy is unknown due to the heterogeneity of data and the underreporting of fetal outcomes with VKA use beyond the first trimester.[35, 36]

LMWH and maternal thromboembolism risk

Unfractionated heparin (UFH) and LMWH, like enoxaparin, do not cross the placenta, making them attractive alternatives to VKA. However, the potential advantage to the fetus is offset by an increased risk of maternal thromboembolism. Fatal valve thrombosis has been reported in pregnant people/women treated with UFH or LMWH for part of all of pregnancy.[35, 36, 39] Even with perfect LMWH use, breakthrough thromboembolism remains a concern.[39]

A meta-analysis of maternal outcomes with MPHV showed that:

- Thromboembolism rates were significantly lower with VKA use compared to LMWH (0.9% versus 8.7%).[35]
- Maternal mortality was lower with a VKA regimen than with LMWH (less than 1% versus 2.9%).

When sequential anticoagulation therapy is used , the highest thromboembolism risk occurs during the medication transitions:

- From VKA to LMWH in early pregnancy.
- From LMWH back to VKA after the first trimester.
- From VKA to LMWH from 34 weeks to allow 'wash out' of VKA in the fetus before a planned birth.

Rates of thromboembolism as high as 5.8% have been reported with sequential treatment.[35] Major bleeding complications were as high as 11.5% with LMWH compared to 1.3% in VKA regimen.

Despite the lack of a consensus on best practice anticoagulation in pregnancy for people with MPHV, LMWH continues to be offered in some centres. This is often due to patients' concerns about VKA use in pregnancy. Favourable maternal and fetal outcomes have been reported when twice-daily therapeutic LMWH dosing is used with rigorous monitoring of peak and trough anti-Xa levels.[33, 39, 40]

Comparing the risks of different anticoagulation options

Table 12.4. Maternal and fetal risks for the different anticoagulation options

	Risk to mother — % average (95% CI)	Risk to fetus — % average (95% CI)	Risk to both mother and fetus — % average
VKA predominant	5% (2–9%)	39% (27–52%)	44%
Low-dose VKA	5% (0–16%)	15% (7–27%)	20%
LMWH predominant	15% (8–25%)	14% (4–29%)	29%
Sequential treatment	16% (5–32%)	16% (1–41%)	32%

Adapted from Steinberg (2017).[36]

Antenatal care for people with mechanical prosthetic heart valves

The 2016 United Kingdom Obstetric Surveillance System (UKOSS) study found that the lack of a standardised approach for managing pregnant people/women with MPHV was a major contributor to poor maternal and fetal outcomes.[41] People with MPHV are at high risk during pregnancy and should be reviewed by experienced clinicians as early as possible. For those who have not had pre-conception counselling, clinicians should thoroughly discuss treatment options and their associated risks and benefits. Individualised, joint decision making is recommended, covering the following:

- The pros and cons of each anticoagulation option must be discussed with those who wish to continue their pregnancy.
- The most dangerous option is to take no anticoagulation, as this significantly increases the risk of fatal valve thrombosis.
- Patients must be informed that there are currently no randomised comparative studies of the different anticoagulation regimens.[11, 17, 32, 38]

Strict adherence to treatment and monitoring must be emphasised, irrespective of the anticoagulation chosen.[40]

Consider using culturally appropriate services for interpreting and support, particularly for Pacific peoples. Given the sensitive and complex nature of these discussions, pregnant people/women should be encouraged to identify who they would like to be present during the counselling sessions.

Deciding on the type of anticoagulation

When discussing anticoagulation options, key considerations include:

- Cardiac and valve status at baseline.
- Additional risk factors for thromboembolism.
- Warfarin maintenance dose.
- Concerns with adherence.
- Geographical barriers, for example, access to regular blood testing.
- The person's preference and views.

Additional risk factors for thromboembolism for MPHV:

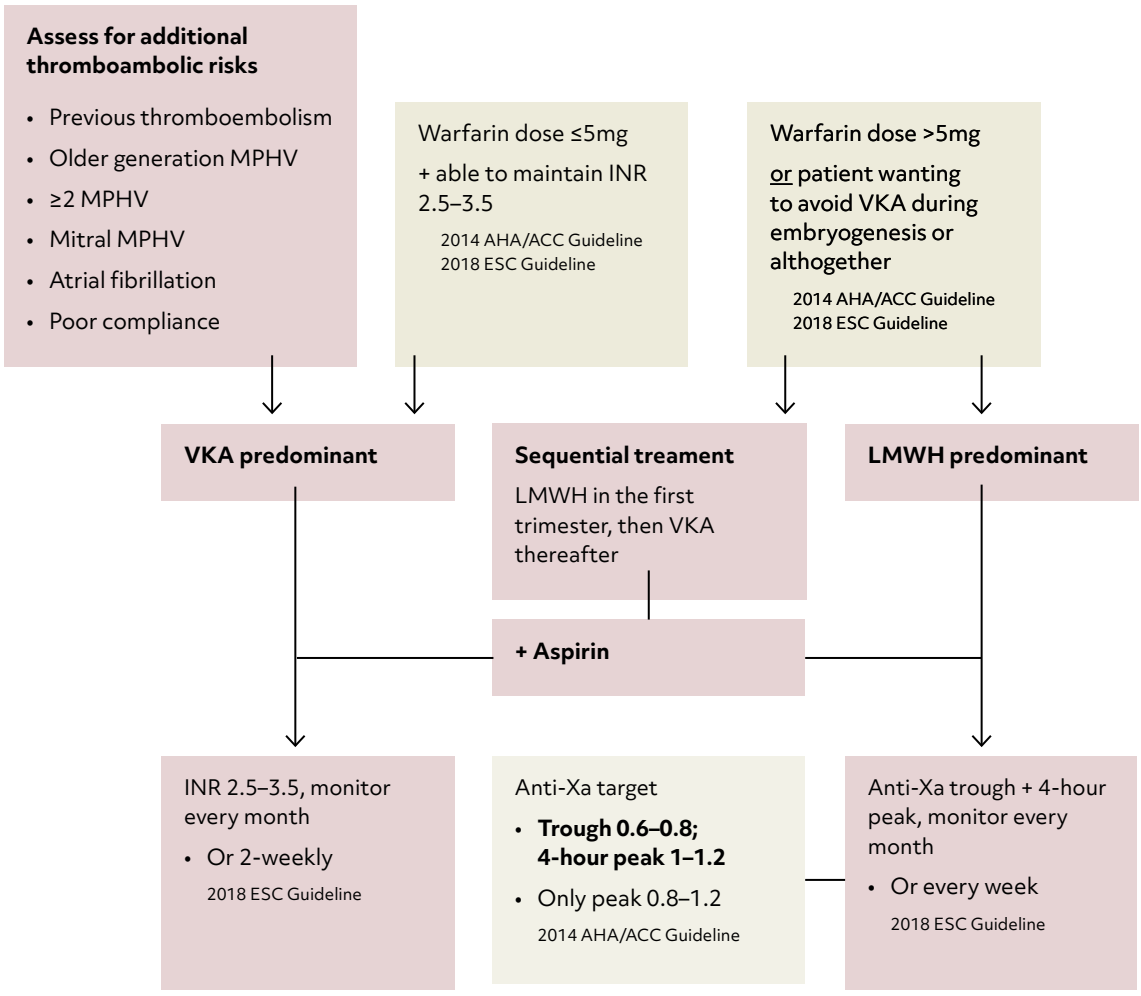
- Previous thromboembolism.
- Older generation MPHV, for example, Starr-Edwards valve.
- Mechanical valve in the mitral position.
- Two or more mechanical valves.
- Presence of concurrent AF.
- History of poor adherence to anticoagulation.

Patients should be fully informed about the three available anticoagulation regimens (see Figure 12.3):

- VKA predominant.
- Sequential (LMWH in the first trimester, then VKA).
- LMWH predominant.

Those at very high risk of thrombosis should be advised that using warfarin for some or all of pregnancy offers the greatest maternal safety.

All pregnant people/women with an MPHV should receive aspirin 100 mg daily, regardless of their anticoagulation regimen during pregnancy.



Key:

AHA/ACC	American Heart Association/American College of Cardiology
ESC	European Society of Cardiology

Figure 12.3. Anticoagulation options in pregnancy with mechanical prosthetic heart valve

Table 12.5. Dosage and frequency of monitoring of anticoagulation during pregnancy

Anticoagulation options	Regimen
Warfarin predominant	• Warfarin until 34 weeks + INR monitoring • Then, enoxaparin 1 mg/kg two times daily + anti-Xa monitoring until the planned birth
Enoxaparin throughout	• Enoxaparin 1 mg/kg two times daily + anti-Xa monitoring until planned birth
Sequential anticoagulation	• Enoxaparin 1 mg/kg two times daily between 6 to 12 weeks + anti-Xa monitoring • Then warfarin until 34 weeks + INR monitoring • Then enoxaparin 1 mg/kg two times daily + anti-Xa monitoring until the planned birth
	Recommendations for monitoring and target
Warfarin	• At least monthly, more often if dose changes or clinical concerns arise • Target INR: 2.5–3.5
Enoxaparin	• At least monthly, more often if dose changes or clinical concerns arise • Monitor trough anti-Xa (pre-dose) and peak anti-Xa (4 hours post-dose) • Target trough anti-Xa: 0.6–0.8 • Target peak anti-Xa: 1–1.12 Tough levels indicate the minimum anticoagulation required to prevent thromboembolism. Peak levels indicate the maximal anticoagulation effect and bleeding risk. Gaps remain in the evidence about the optimal anti-Xa level targets and the best frequency for anti-Xa monitoring.[85]

Deciding on where to give birth

For pregnant people/women with MPHV, the decision about where to give birth should be made early. The decision should include a contingency plan for out-of-hours emergencies because of the unpredictability of labour and the potential bleeding risks.

Key considerations:

- Pregnant people/women must be informed about the complex anticoagulation management around the time of birth.
- Bridging from maintenance anticoagulation to short-acting UFH infusion is required.

- Re-initiation of anticoagulation after birth will need very close monitoring, particularly in cases of postpartum bleeding.
- The period of anticoagulation bridging before and immediately after birth is high-risk for breakthrough thromboembolism.

Standard obstetric indications should determine the mode of birth. Pregnant people/women with MPHV should give birth at a centre that:

- Has expertise in managing peri-birth and postpartum anticoagulation bridging.
- Provides cardiothoracic intensive care in case of acute valve thrombosis.

While few people with advanced RHD and MPHV undergo pregnancy in Aotearoa, National Women's Health, in conjunction with the Auckland City Hospital Cardiology service, have the most experience in managing these pregnancies.[39]

Pregnant people/women and their whānau should be offered a meeting with the multi-disciplinary team during their pregnancy to establish rapport. Close fetal growth surveillance is reasonable and should be updated at the time of an acute maternal cardiac event.

Iron status should be reviewed at standard times and optimised opportunistically due to the risk of bleeding.

Peri-birth and postpartum care for people with mechanical prosthetic heart valves

The peri-birth and postpartum period are high-risk for cardiac, thromboembolic, or bleeding complications, even in those whose antenatal period was straightforward.

Key considerations for peri-birth and postpartum care:

- Timing of the last dose of LMWH to allow appropriate 'wash-out' before planned birth (see **Figure 12.4**). Wash-out is important to allow access to regional neuraxial analgesia.
- Strict monitoring of intravenous UFH infusion through local protocol to maintain activated partial thromboplastin clotting time (APTT) within the therapeutic range until the start of active labour, or until an epidural catheter for regional anaesthesia is placed for a planned caesarean section. National Women's Health is one of the few centres in Aotearoa with a dedicated local protocol for peri-birth management of those on therapeutic anticoagulation in pregnancy.
- Re-initiation of intravenous UFH infusion guided by mode of birth and postpartum bleeding status.
- Re-initiation of VKA with appropriate bridging anticoagulation cover until therapeutic INR is achieved. Warfarin is compatible with breastfeeding.
- Finalisation of contraception plan if not previously completed.

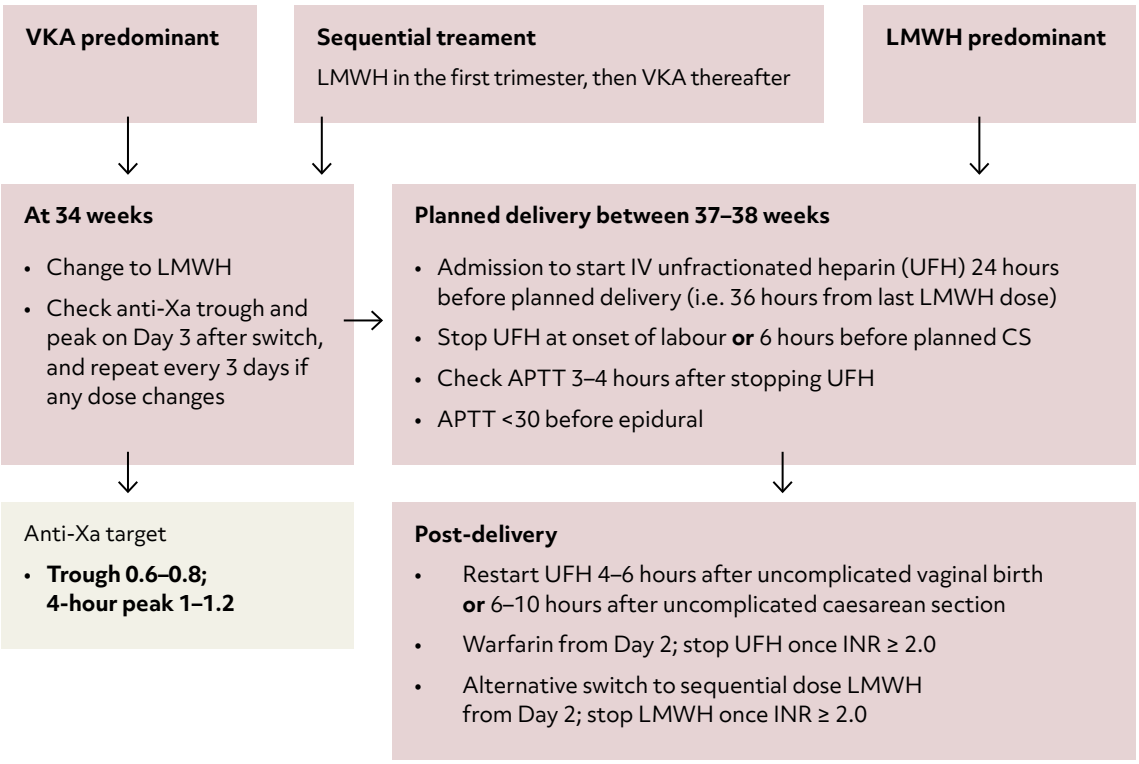


Figure 12.4. Peri-birth and postpartum management of anticoagulation in pregnancy with mechanical prosthetic heart valve

This is the local protocol developed at National Women's Health.

Mechanical prosthetic valve thrombosis in pregnancy

MPVT is a rare but potentially life-threatening complication in pregnancy.[35, 42] This serious complication must be considered for all pregnant people/women with MPHV who present with:

- New cardiac or thromboembolic symptoms, irrespective of their anticoagulation status.
- Therapeutic LMWH use and additional thromboembolism risk factors (see section on 'Deciding on the type of anticoagulation').
- Non-adherence or intermittent adherence to anticoagulation therapy.
- Minimal or sub-therapeutic anticoagulation monitoring.
- Recent transition between different anticoagulation treatments.

A routine surveillance echo that demonstrates raised gradients across the MPHV or abnormal leaflet movement should raise suspicion of MPVT. Immediate discussion with a view to transfer to a tertiary centre experienced in managing MPVT in pregnancy is recommended.

Managing the risk of mitral prosthetic valve thrombosis

Currently, there are no evidence-based guidelines specific to pregnancy, but management follows similar principles to non-pregnant cases. For stable and non-critical cases, medical management with transoesophageal echo monitoring can be offered. Medical management involves anticoagulation optimisation with intravenous UFH infusion, followed by a protocol of slow, low-dose infusion of tissue-type plasminogen activator (tPA) if required.[11, 43, 44] Complications with tPA use include significant maternal and placental bleeding and cardioembolism.

If treatment is successful, VKA (warfarin) for the remainder of pregnancy is recommended to offer the best maternal protection from re-thrombosis. People should receive counselling about the ongoing risks of continuing with the pregnancy.

Valve surgery should be reserved for critically unwell people with MPVT or those who fail medical therapy due to its high morbidity and mortality risk.[44]

Resources on prescribing medication in pregnancy and during breastfeeding

The New Zealand Formulary (NZF) replaced the United States Food and Drug Administration (US FDA) classification system in 2018. It provides up-to-date advice and recommendations on medication safety in pregnancy and breastfeeding.[45] Medication safety between cardiac medications of the same class can vary. Use medications with the most clinical experience in pregnancy and during breastfeeding when possible.

The US FDA classification system has several critical limitations and is no longer recommended as the sole reference for prescribing medication during pregnancy and breastfeeding.[11] Concerns about potential risk to the fetus must be weighed against the maternal benefits of treatment. Withholding necessary treatment can be harmful to the pregnant person/woman.

The 2018 European Society of Cardiology (ESC) provides a comprehensive table with a summary of each cardiac-related medication and its clinical safety data.[11] Other online resources are available.

New Zealand Formulary (NZF)

Best Use of Medicines in Pregnancy (BUMPS, UK Teratology Information Service)

Drugs and Lactation Database (Lactmed®, National Institutes of Health)

Breastfeeding has many benefits, and people should be supported to breastfeed where possible. Before prescribing medication to breastfeeding people, check guidelines and safety resources. Consult a lactation consultant if available for additional support.

References

1. Perinatal and Maternal Mortality Review Committee. Fifteenth annual report of the Perinatal and Maternal Mortality Review Committee: reporting mortality and morbidity 2020. Health Quality & Safety Commissions; 2022. https://www.hqsc.govt.nz/assets/Our-work/Mortality-review-committee/PMMRC/Publications-resources/15thPMMRC-report-final.pdf (Accessed December 16 2024).

2. Webb R, Culliford-Semmens N, ChanMow A, Doughty R, Tilton E, Peat B, et al. High burden of rheumatic heart disease confirmed by echocardiography among Pacific adults living in New Zealand. *Open Heart*. 2023;10(1):e002253. https://doi.org/10.1136/openhrt-2023-002253

3. Justin Paul G, Anne Princy S, Anju S, Anita S, Cecily Mary M, Gnanavelu G, et al. Pregnancy outcomes in women with heart disease: the Madras Medical College Pregnancy and Cardiac (M-PAC) Registry from India. *European Heart Journal*. 2023;44(17):1530-1540. https://doi.org/10.1093/eurheartj/ehad003

4. Sullivan EA, Vaughan G, Li Z, Peek MJ, Carapetis JR, Walsh W, et al. The high prevalence and impact of rheumatic heart disease in pregnancy in First Nations populations in a high-income setting: a prospective cohort study. *BJOG: An International Journal of Obstetrics and Gynaecology*. 2020;127(1):47–56. https://doi.org/10.1111/1471-0528.15938

5. Hohmann-Marriot BE. Unplanned pregnancies in New Zealand. *Australian and New Zealand Journal of Obstetrics and Gynaecology*. 2017;58(2):247–250. https://doi.org/10.1111/ajo.12732

6. Russell EA, Tran L, Baker RA, Bennetts JS, Brown A, Reid CM, et al. A review of valve surgery for rheumatic heart disease in Australia. *BMC Cardiovascular Disorders*. 2014;14:134. https://doi.org/10.1186/1471-2261-14-134

7. Health New Zealand | Te Whatu Ora. Guidelines for consultation with obstetric and related medical services (referral guidelines). 2023. https://www.tewhatuora.govt.nz/publications/guidelines-for-consultation-with-obstetric-and-related-medical-services-referral-guidelines (Accessed December 16 2024).

8. Siu SC, Sermer M, Colman JM, Alvarez AN, Mercier LA, Morton BC, et al. Prospective multicenter study of pregnancy outcomes in women with heart disease. *Circulation*. 2001;104(5):515–521. https://doi.org/10.1161/hc3001.093437

9. Silversides CK, Grewal J, Mason J, Sermer M, Kiess M, Rychel V, et al. Pregnancy outcomes in women with heart disease: the CARPREG II study. *Journal of the American College of Cardiology*. 2018;71(21):2419–2430. https://doi.org/10.1016/j.jacc.2018.02.076

10. Drenthen W, Boersma E, Balci A, Moons P, Roos-Hesselink JW, Mulder BJ, et al. Predictors of pregnancy complications in women with congenital heart disease. *European Heart Journal*. 2010;31(17):2124–2132. https://doi.org/10.1093/eurheartj/ehq200

11. European Society of Gynecology, Association for European Paediatric Cardiology, German Society for Gender Medicine, Regitz-Zagrosek V, Blomstrom Lundqvist C, Borghi C, et al. ESC guidelines on the management of cardiovascular diseases during pregnancy: the Task Force on the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology (ESC). *European Heart Journal*. 2011;32(24):3147297. https://doi.org/10.1093/eurheartj/ehr218

12. Mehta LS, Warnes CA, Bradley E, Burton T, Economy K, Mehran R, et al. Cardiovascular considerations in caring for pregnant patients: a scientific statement from the American Heart Association. *Circulation*. 2020;141(23):e884–e903. https://doi.org/10.1161/CIR.0000000000000772

13. RHDAustralia, Menzies School of Health Research. The 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (3.2 edition). 2022. https://www.rhdaustralia.org.au/arf-rhd-guidelines (Accessed December 16 2024).

14. Balci A, Sollie-Szarynska KM, Bijl AGL, Ruys TPE, Mulder BJM, Roos-Hesselink JW. Prospective validation and assessment of cardiovascular and offspring risk model for pregnant women with congenital heart disease. *Heart*. 2014;100(17):1373–1381.

15. Mohammadi N, Shojaeifard M, Kashfi F, Larti F, Chenaghlou M, Rezaei Y, et al. Pre-conception consultation using treadmill exercise stress echocardiography for pregnant women with the left-sided heart valve stenosis: a preliminary report. *Northern Clinics of İstanbul*. 2022;9(6):550–556. https://doi.org/10.14744/nci.2021.67809

16. Tanous D, Siu SC, Mason J, Greutmann M, Wald RM, Parker JD, et al. B-type natriuretic peptide in pregnant women with heart disease. *Journal of the American College of Cardiology*. 2010;56(15):1247–1253. https://doi.org/10.1016/j.jacc.2010.02.076

17. Bonow RO, Carabello BA, Chatterjee K, de Leon AC, Jr., Faxon DP, Freed MD, et al. 2008 Focused update incorporated into the ACC/AHA 2006 guidelines for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Revise the 1998 Guidelines for the Management of Patients With Valvular Heart Disease): endorsed by the Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. *Circulation*. 2008;118(15):e523–661. https://doi.org/10.1161/CIRCULATIONAHA.108.190748

18. Gutman SJ, Shemesh E, Marwick TH, Taylor AJ. Echocardiographic screening to determine progression of latent rheumatic heart disease in endemic areas: a systematic review and meta-analysis. *PloS One*. 2020;15(6):e0234196. https://doi.org/10.1371/journal.pone.0234196

19. Rwebembera J, Marangou J, Mwita JC, Mocumbi AO, Mota C, Okello E, et al. 2023 World Heart Federation guidelines for the echocardiographic diagnosis of rheumatic heart disease. *Nature Reviews: Cardiology*. 2024;21(4):250–263. https://doi.org/10.1038/s41569-023-00940-9

20. Sanghavi M, Rutherford JD. Cardiovascular physiology of pregnancy. *Circulation*. 2014;130(12):1003–1008. https://doi.org/10.1161/CIRCULATIONAHA.114.009029

21. Johal K, Lau L, Card ME. Mitral stenosis. In: Taylor A, editor. Learning cardiac auscultation: Springer; 2015. p. 135–149.

22. Wichert-Schmitt B, Steckham KE, Pfaller B, Colman JM, Wald RM, Sermer M, et al. Cardiac complications in pregnant women with isolated mitral stenosis and their association with echocardiographic changes during pregnancy. *American Journal of Cardiology*. 2021;158:81–89. https://doi.org/10.1016/j.amjcard.2021.07.037

23. Zuhlke L, Engel ME, Karthikeyan G, Rangarajan S, Mackie P, Cupido B, et al. Characteristics, complications, and gaps in evidence-based interventions in rheumatic heart disease: the Global Rheumatic Heart Disease Registry (the REMEDY study). *European Heart Journal*. 2015;36(18):1115–1122a. https://doi.org/10.1093/eurheartj/ehu449

24. Weiss BM, von Segesser LK, Alon E, Seifert B, Turina MI. Outcome of cardiovascular surgery and pregnancy: a systematic review of the period 1984-1996. *American Journal of Obstetrics and Gynecology*. 1998;179(6 Pt 1):1643–1653. https://doi.org/10.1016/s0002-9378(98)70039-0

25. Hussein A, Eid M, Mahmoud SED, Sabry M, Altaher A. The outcomes of PBMV in pregnancy, and when is the best time? *Vascular Health and Risk Management*. 2023;19:13–20. https://doi.org/10.2147/VHRM.S388754

26. Gnanaraj JP, Princy S A, Surendran S A. Counselling and pregnancy outcomes in women with congenital heart disease — current status and gap analysis from Madras Medical College Pregnancy And Cardiac disease (M-PAC) registry. *International Journal of Cardiology Congenital Heart Disease*. 2021;5:100207. https://doi.org/10.1016/j.ijcchd.2021.100207

27. Desai DK, Adanlawo M, Naidoo DP, Moodley J, Kleinschmidt I. Mitral stenosis in pregnancy: a four-year experience at King Edward VIII Hospital, Durban, South Africa. *BJOG: An International Journal of Obstetrics and Gynaecology*. 2000;107(8):953–958. https://doi.org/10.1111/j.1471-0528.2000.tb10395.x

28. Sarma AA, Aggarwal NR, Briller JE, Davis M, Economy KE, Hameed AB, et al. The Utilization and Interpretation of Cardiac Biomarkers During Pregnancy: *JACC: Advances* Expert Panel. *JACC Advances*. 2022;1(3):100064. https://doi.org/10.1016/j.jacadv.2022.100064

29. Balaceanu A. B-type natriuretic peptides in pregnant women with normal heart or cardiac disorders. *Medical Hypotheses*. 2018;121:149–151. https://doi.org/10.1016/j.mehy.2018.09.034

30. El SF, Hassan W, Latroche B, Helaly S, Hegazy H, Shahid M, et al. Pregnancy has no effect on the rate of structural deterioration of bioprosthetic valves: long-term 18-year follow up results. *Journal of Heart Valve Disease*. 2005;14(4):481–485.

31. North RA, Sadler L, Stewart AW, McCowan LM, Kerr AR, White HD. Long-term survival and valve-related complications in young women with cardiac valve replacements. *Circulation*. 1999;99(20):2669–2676. https://doi.org/10.1161/01.cir.99.20.2669

32. McLintock C. Anticoagulant therapy in pregnant women with mechanical prosthetic heart valves: no easy option. *Thrombosis Research*. 2011;127 (Suppl 3):S56–60. https://doi.org/10.1016/S0049-3848(11)70016-0

33. McLintock C. Anticoagulant choices in pregnant women with mechanical heart valves: balancing maternal and fetal risks--the difference the dose makes. *Thrombosis Research*. 2013;131(Suppl 1):S8–10. https://doi.org/10.1016/S0049-3848(13)70010-0

34. Castellano JM, Narayan RL, Vaishnava P, Fuster V. Anticoagulation during pregnancy in patients with a prosthetic heart valve. *Nature Reviews: Cardiology*. 2012;9(7):415–424. https://doi.org/10.1038/nrcardio.2012.69

35. D'Souza R, Ostro J, Shah PS, Silversides CK, Malinowski A, Murphy KE, et al. Anticoagulation for pregnant women with mechanical heart valves: a systematic review and meta-analysis. *European Heart Journal*. 2017;38(19):1509–1516. https://doi.org/10.1093/eurheartj/ehx032

36. Steinberg ZL, Dominguez-Islas CP, Otto CM, Stout KK, Krieger EV. Maternal and fetal outcomes of anticoagulation in pregnant women with mechanical heart valves. *Journal of the American College of Cardiology*. 2017;69(22):2681–2691. https://doi.org/10.1016/j.jacc.2017.03.605

37. Sadler L, McCowan L, White H, Stewart A, Bracken M, North R. Pregnancy outcomes and cardiac complications in women with mechanical, bioprosthetic and homograft valves. *BJOG: An International Journal of Obstetrics and Gynaecology*. 2000;107(2):245–253. https://doi.org/10.1111/j.1471-0528.2000.tb11696.x

38. Elkayam U, Singh H, Irani A, Akhter MW. Anticoagulation in pregnant women with prosthetic heart valves. *Journal of Cardiovascular Pharmacology and Therapeutics*. 2004;9(2):107–115. https://doi.org/10.1177/107424840400900206

39. McLintock C, McCowan LM, North RA. Maternal complications and pregnancy outcome in women with mechanical prosthetic heart valves treated with enoxaparin. *BJOG: An International Journal of Obstetrics and Gynaecology*. 2009;116(12):1585–1592. https://doi.org/10.1111/j.1471-0528.2009.02299.x

40. Goland S, Schwartzenberg S, Fan J, Kozak N, Khatri N, Elkayam U. Monitoring of anti-Xa in pregnant patients with mechanical prosthetic valves receiving low-molecular-weight heparin: peak or trough levels? *Journal of Cardiovascular Pharmacology and Therapeutics*. 2014;19(5):451–456. https://doi.org/10.1177/1074248414524302

41. Vause S, Clarke B, Tower CL, Hay C, Knight M. Pregnancy outcomes in women with mechanical prosthetic heart valves: a prospective descriptive population based study using the United Kingdom Obstetric Surveillance System (UKOSS) data collection system. *BJOG: An International Journal of Obstetrics and Gynaecology*. 2017;124(9):1411–1419. https://doi.org/10.1111/1471-0528.14478

42. van Hagen IM, Roos-Hesselink JW, Ruys TP, Merz WM, Goland S, Gabriel H, et al. Pregnancy in women with a mechanical heart valve: Data of the European Society of Cardiology Registry of Pregnancy and Cardiac Disease (ROPAC). *Circulation*. 2015;132(2):132–142. https://doi.org/10.1161/CIRCULATIONAHA.115.015242

43. Ozkan M, Cakal B, Karakoyun S, Gursoy OM, Cevik C, Kalcik M, et al. Thrombolytic therapy for the treatment of prosthetic heart valve thrombosis in pregnancy with low-dose, slow infusion of tissue-type plasminogen activator. *Circulation*. 2013;128(5):532–540. https://doi.org/10.1161/CIRCULATIONAHA.113.001145

44. Bigdelu L, Maadarani O, Yadolh A, Bitar Z, Azadi N. Successful thrombolytic therapy using ultraslow low-dose infusion during pregnancy for mitral mechanical valve thrombosis: a case series and review of the literature. *European Journal of Case Reports in Internal Medicine*. 2023;10(5):003856. https://doi.org/10.12890/2023_003856

45. Best Practice Advocacy Centre New Zealand. NZF is changing the way that pregnancy and breastfeeding advice for medicines is presented. 2018. https://bpac.org.nz/2018/nzf-pregnancy.aspx (Accessed December 16 2024).

13

Developmentally Appropriate Care for Rangatahi — Adolescents and Young People

In this chapter

"I had a [ARF] relapse a few years ago … it was because I wasn't taking my injections. I was just being silly. They [nurses] will give me a call and be like, 'okay you're finishing school this year, what's the plan for the future?' I was like, 'I don't know what I'm going to do'."

Person receiving secondary prophylaxis

Key changes

This is a new chapter.

Key points

- Healthcare must be developmentally, age, and culturally appropriate to support tamariki (children) and rangatahi (young people) in their development during illness and treatment.
- Cultural frameworks and models such as Te Tapatoru and Te Ūkaipō consider rangatahi perspectives and can help build trust and support the hauora (health) of rangatahi.
- Providing regular opportunities for healthcare engagement and integrating acute rheumatic fever (ARF) and rheumatic heart disease (RHD) treatment with other healthcare promotes overall hauora.
- Clear communication and coordination among healthcare providers support continuous care and smooth transitions between services for rangatahi.

Introduction

"Adolescents are not older children or younger adults: adolescence is a unique, formative stage of human development. Notwithstanding, adolescents are extremely diverse — in culture, nationality, wealth, education, family and many other ways. In all societies and settings, they have key developmental experiences as they transition from childhood to adulthood. This complex passage involves rapid physical growth, hormonal changes, sexual development, new and conflicting emotions, increased cognitive and intellectual capacities, moral development and evolving relationships with peers and families."[1, p.1]

This chapter starts by defining 'developmentally appropriate care' before discussing a few frameworks that underpin a range of strengths-based, cultural approaches to working with rangatahi (adolescents and young people between 15–24 years[2]). The rest of this chapter offers practical insights and tips on how to speak and engage with rangatahi about their hauora.

Some current approaches harness rangatahi potential by creating reciprocal and invigorating supportive environments based on the aspirations and insights of rangatahi. For instance, they associate a strong cultural identity with better wellbeing and mental health.[3] When asked what will help with positive development, rangatahi identified that "wellbeing means hauora, wairua [spirit], mauri [life essence] — an interwoven presence of wayfinding time, space and generations".[4, p.70]

Two well-known examples of te ao Māori models of hauora are Te Whare Tapa Whā and Te Wheke.[5] Healthcare workers who care for rangatahi with ARF or RHD may find these models useful as they allow better alignment with clinical health practice and cultural values of many rangatahi Māori including the importance of whānau, wairuatanga (spirituality) and hinengaro hauora (mental health). Adolescent development and working with Māori and Pacific rangatahi and their whānau need extra thought and effort due to the importance of collective values, cultural contexts, connection, and whanaungatanga (also see **Chapter 1: Cultural Responsiveness**). Specific models that consider youth perspectives of achieving good health and wellbeing have been developed. For instance, Te Tapatoru model (**Figure 13.2**) highlights the importance of whanaungatanga (nurturing of relationships) for wellbeing and building a relationship of trust.[6]

Te Ūkaipō is a values and vision framework that was developed as a model of care for School-based Health Services, in the way they work, the mahi they do and the outcomes for rangatahi. Te Ūkaipō was developed by Te Rōpū Mātanga o Rangatahi as an expression of te ao Māori (Māori world view), Mātauranga Māori (Māori knowledge and understanding) and Te Tiriti o Waitangi principles. Te Rōpū Mātanga o Rangatahi are the kaitiaki (guardians/caretakers) of Te Ūkaipō, ensuring its integrity. The vision and the values together form a strong and sturdy kōhanga (nest), a safe space for rangatahi to come into, ask questions, grow, and develop. Te Ūkaipō Vision and Values Framework is available, along with a compilation video and podcasts. Increasingly kaimahi are seeing Te Ūkaipō as providing a foundation in wider health and wellbeing care contexts when caring for young people. Ōritetanga is the importance of equity. "All rangatahi have accessible and equitable care". Rheumatic fever affects Māori and Pacific children and young people at far higher rates than non-Māori or non-Pacific. Ensuring all Māori and Pacific rangatahi have equitable access to throat swabs in our local communities demonstrates Ōritetanga.

Te hoki atu ki te wāhi i ahu mai koe, te wāhi i whāngaitia e koe hei oranga mōu mō te rerenga ki mua.

To return to the place of your origin, the place where you can be nourished to sustain you for the journey ahead.[7]

Whanonga Pono | Values

Tino Uaratanga — Potential — "I have potential"

We recognise the unique potential of each rangatahi.

Wairua — Spirituality — "I am essential"

We acknowledge wairua-based practices as a way to restore and enhance hauora.

Aroha — Love & Compassion — "I matter"

We lead with compassion and understanding and actively demonstrate this throughout our mahi.

Whanaungatanga — Connection to Others and Self — "I am connected"

We are passionate about our meaningful connection.

Connection to each other, to te taiao, to our whānau, schools and most of all, connection to ourselves.

Rangatiratanga — Autonomy — "I have self determination"

We listen to the individual needs of rangatahi and empower them to make choices for themselves.

Whakapapa — Identity — "I belong"

We respect all whakapapa and value the power of knowing where we come from.

Te Reo — Language — "I have mana"

We love Te Reo Māori. No matter how little or how much we understand, we speak and write it as often as possible, and we ensure pronunciation is correct — always.

Manaakitanga — Nurturing — "I am valued"

We value the exchange of supporting and caring for others and the inner reward that is brings.

Ōritetanga — Equality — "I am equal"

We believe that all people are of equal worth and are entitled to equal respect.

Figure 13.1. Te Ūkaipō Framework[7]

For more detail and examples of both how these values can be embedded and how they can be measured, see the Te Ūkaipō Framework (December 2023).[7]

What is developmentally appropriate care?

Rangatahi often experience adolescence as a complex time that involves many physical, hormonal, emotional, cognitive, and social changes. These experiences are diverse and intersect with cultural and social contexts.[8] An illness like ARF adds an additional layer of complexity to an often already complex period in a rangatahi's life.

A critical component of appropriate healthcare for rangatahi is delivering culturally and age-appropriate care that supports adolescent development throughout their illness and treatment. Developmentally appropriate care engages rangatahi. It recognises and addresses the broad, holistic health and developmental contexts of rangatahi and fosters skills in self-management and self-efficacy.[9-11] It requires ongoing engagement with support people — usually whānau — as relationships are renegotiated.

Engaging with rangatahi and their whānau requires a respectful understanding of their cultural perspectives.[7, 12] Such engagement is coordinated and uninterrupted, which requires sharing clinical and psychosocial information between teams and with rangatahi and whānau, as well as their other supports.[9]

What happens if developmentally appropriate care is not provided?

Rangatahi and whānau have reported that health services in Aotearoa are not adequately meeting the complex contexts of rangatahi. [13-15] Rangatahi value developmentally appropriate healthcare, yet healthcare systems are traditionally designed around tamariki or adults and often fail to meet the complex contexts of rangatahi.[16-19]

Rangatahi with long-term health conditions (such as RHD) suffer poorer emotional health and social outcomes, with higher rates of health-compromising behaviours and impairments in socialisation with peers than those without long-term health conditions.[20-22] Rangatahi with complex, specialised healthcare requirements also experience less educational and employment success.[23]

Emerging evidence indicates that delivering developmentally appropriate healthcare to rangatahi with long-term health and developmental conditions from early adolescence has benefits. Taking this approach can improve access to services and disease management and increase satisfaction with the healthcare provided.[18, 24-27] Improved care has the potential to reduce inequitable health outcomes.

Strengths-based approaches: what this means when working with rangatahi

A strengths-based approach prioritises the inherent strengths and connections of the individual and their whānau. Instead of focusing on weakness or risk, a strengths-based approach encourages resilience and self-efficacy.[7, 28]

Historical, social, and cultural contexts add a layer of complexity to how Indigenous adolescents come of age in terms of age-related roles, values, and expectations.[29] In contemporary colonial contexs Indigenous adolescents can become confused or feel out of place in such misaligned spaces. They can also encounter challenges when expected to take on more responsibility without first having the opportunity to develop self-management skills.[9-11]

Structural influences such as colonisation, poverty, and racism play a role in Māori and Pacific rangatahi health outcomes. Decolonisation, anti-racism, and equity interventions are critical to negate these influences.[16] Specific considerations for rangatahi Māori and Pacific should focus on:

- The impacts of colonisation and the continued discrimination and racism in accessing care within Aotearoa's health system.
- The lack of health funding and interventions specific to both Māori and Pacific rangatahi.[16]

Age-appropriate engagement is important when communicating with rangatahi in health settings. Rangatahi may want to discuss a range of health-related behaviours (including oral health, ARF, or RHD management) but may only feel comfortable responding to questions that are asked directly in a way that lets them easily understand and not feel overwhelmed or whakamā (embarrassed). Such discussions must be done in private settings and are confidential, particularly for conditions such as ARF or RHD, which carry stigma.[26, 27, 30-35] If discussions and questions are not framed in this way, rangatahi may decide to go without healthcare and miss opportunities for health promotion and ARF and RHD prevention.[35]

The importance of a rangatahi's connections to their peers and vocational or training goals is also highly valued, yet often poorly addressed in health contexts.[9-11, 14, 15, 34, 36] As tamariki develop into rangatahi and through their teen years, their aspirations, contexts, and challenges change. A regular comprehensive review of their haurora is needed, including these elements: tinana (physical body), hinengaro hauora, wairua, and whānau.[37] Inappropriate services can result in a range of negative consequences, including poorer health outcomes, fragmented care, increased psychological distress, and disrupted developmental progress. These inadequate services may contribute to reduced treatment adherence, strained relationships with healthcare providers, and financial burdens to whānau and healthcare systems.[13, 15]

Cultural frameworks for developmentally appropriate care in Aotearoa

Rangatahi in some cultures, including Māori and Pacific peoples, may consider adolescence as a time when whānau relationships remain important, and being part of a community to which the rangatahi identifies and belongs plays a vital role in hauora.[38]

Providing regular opportunities for engagement and healthcare delivery for rangatahi, along with integrating ARF and RHD treatment into their overall care, can promote a holistic approach to hauora.[13, 15] Receiving regular dental care (which currently in Aotearoa is free until their 18th birthday), is crucial for rangatahi with ARF or RHD as it decreases their risk of poor oral health and infective endocarditis. Similarly, rangatahi with ARF or RHD require injections of benzathine penicillin (Bicillin®L-A brand is used in Aotearoa) every 28 days and may require additional medical management, including anticoagulation (see **Chapter 8: Secondary Prevention**). Negotiation and flexibility in delivering benzathine penicillin, along with regular monitoring for ARF and RHD, are essential for health practitioners to manage with rangatahi and their whānau. This flexibility can assist in maximising treatment adherence and promoting rangatahi independence in managing their own health.

In addition to the experience of rangatahi and whānau, health professionals report many issues with providing developmentally appropriate care to rangatahi. These include existing systems that are inflexible and non-youth-focused, inappropriate facilities, lack of training, and time constraints.[13, 24, 39] It is the health system's responsibility to manage these issues and health practitioners should advocate and be willing to support flexible approaches for developmentally appropriate care for rangatahi.

Delivery of healthcare must be clear and coordinated

Many different models of healthcare for ARF exist across Aotearoa. These usually involve a range of different teams. Clear communication between different services involved is vital. Poor communication and care coordination among different specialties result in confusion and potentially reduced adherence and mistrust in health services.[13, 15] This may lead to ARF recurrences and other serious medical complications of RHD, such as stroke, pregnancy loss, or infective endocarditis.

High-risk times include transitioning between paediatric and adult services or returning to primary care; and when transitioning between or having care shared by several specialist services. Several models provide guidance for transition support within a framework of developmentally appropriate care. These models have been shown to improve health outcomes and self-reported engagement in care and are cost-effective.[9-11, 18] The key to many of these models is having rangatahi and their whānau involved with transition care plans.

Deep understanding of hauora rangatahi is required

SAP adherence decreases when tamariki reach adolescence. Given this, the health workforce requires a deep understanding of hauora rangatahi related to ARF, RHD, and SAP.[40-42] Data within Aotearoa shows that recurrences of ARF increase in adolescence and, proportionally, are highest in people over 21 years of age.[43] Health services that acknowledge hauora rangatahi and support the provision of developmentally appropriate care are likely to achieve better delivery of SAP.[44]

Warfarin management is a particular issue for many with severe RHD and a prosthetic heart valve. Self-management is key. Taking over the self-management of oral health, chronic health conditions, and disability is a critical task for rangatahi[45]. Where conditions have persisted since childhood, some rangatahi may not have received a good explanation about their condition. Learning self-management skills is a gradual task that may take some rangatahi many years and require significant support.[18, 31, 46, 47]

Along with knowledge, rangatahi, with enablers and support, will need to accept and be motivated to take a greater part in their care. People are more likely to adhere to health behaviours when they are motivated to follow them. Motivation is enhanced when people are involved in decisions about their care or treatment. This involvement must be developmentally and culturally appropriate, see **Chapter 1: Cultural Responsiveness**. For rangatahi, engagement in developmentally appropriate care is central to their ability to be involved in decision-making. The level of engagement will depend on their understanding, the appropriate information being made available, and the provision of a supportive environment where they can voice their views and feel valued.[9-11, 13-15, 18, 34, 36]

Ten tips when interacting with rangatahi

Here are ten helpful tips that are useful to apply when interacting with rangatahi.

Tip 1: Have appropriate skills and understanding

All staff working with tamariki, rangatahi, and their whānau should receive appropriate training to enhance their skills and understanding in working with rangatahi. This should include:

- Providing confidential care.
- Conducting hauora psychosocial assessments.
- Practicing cultural safety.
- Supporting rangatahi in developing self-management skills.
- Facilitating transitions through services.

See these resources for training and other materials:

Adolescent HEEADSSS — a cultural perspective on what's new with GenZ | Goodfellow Unit

Working with Youth: HEEADSSS Assessment | Goodfellow Unit

Te-Ukaipo-Framework

Overarching Te Ukaipo Podcast on Vimeo

Tip 2: Plan, monitor and evaluate with rangatahi and whānau

Engage with rangatahi and whānau for advice in service planning, monitoring, and evaluation. Have rangatahi and their whānau lead the service design. If this is not possible, co-design is an effective way to improve service quality.[13]

Think about what **developmental stage** the rangatahi you are working with is at, and with their leadership or partnership, plan care accordingly — see Table 13.1. As an example, an early adolescent is developmentally less likely to be able to have a future focus. Long-term implications of ARF recurrence will not mean much to them. Instead, a current focus, such as the thought of being unwell and missing out on an activity with friends or whānau next week, may be more meaningful.

Table 13.1. Early, middle, and late developmental stages

	Early **"Am I normal?"**	**Middle** **"Who am I?"**	**Late** **"Where am I going?"**
Developmental issues	Puberty Strive for autonomy Peer relationships	Experimentation and risk-taking Identity exploration Peer acceptance	Independence or renegotiated relationship with parents and whānau Educational and vocational goals Own value system
Psychological or cognitive	Concrete thinking The future is now Difficulty identifying how current behaviour impacts their future	Can think more conceptually Able to take more responsibility for their actions	Longer attention span Abstract thinking Future thinking
Practice points	**Confidentiality** Short and simple Reassure about normality	**Confidentiality** Support identity exploration	**Confidentiality** Base interventions on short- and long-term goals

Tip 3: Create an accessible, culturally safe and developmentally appropriate space

Healthcare environments should be accessible, culturally safe, and developmentally appropriate for rangatahi to help improve access to and engagement with healthcare. Consideration should be given to convenience, welcome, physical safety, and maintenance of privacy and confidentiality.[7] Māori and Pacific engagement frameworks such as the hui process and the Soālaupule model are described more in Chapter 1: Cultural Responsiveness.

1. Meet the rangatahi with a smile. Pronounce their name correctly. Before you start, check their preferred pronouns and ethnicity. Checking how they self-identify themselves is important.
2. Greet the rangatahi using their own language greetings or offer to bless the space and session with a karakia. Acknowledgement of culture is critical.
3. Be present in the moment. Listen to what they say even if you feel rushed. Start with an informal conversation. Listening can help decrease the anxiety a rangatahi may be feeling.
4. Create a caring, respectful environment that keeps wellbeing in focus.

Te Tapatoru model for nurturing relationships

Te Tapatoru is a model[6] that consists of three interconnected concepts: Ko wai, He wā pai and Kaupapa pai. The model highlights the importance of whanaungatanga for hauora. Harnessing rangatahi potential by creating reciprocal and invigorating supportive environments based on their aspirations and insights is crucial.

Figure 13.2. Te Tapatoru model[6]

Ko wai: A reciprocal connection — emphasises the importance of reciprocal connections with people (or more than only people).

He wā pai: When directly translated this means 'a good time' in the context of the model refers to a genuine time and place — emphasises that contexts, time, and place provide space for meaningful connections to take root and flourish.

Kaupapa pai: A good foundation or in the context of this model a genuine kaupapa (activity, process) — emphasises that rangatahi desire connections which respond to their desires and aspirations.

Tip 4: Ensure care is private and confidential

Confidentiality can be a barrier to tamariki and rangatahi accessing healthcare, particularly with ARF and RHD, which may have stigma attached.[48] Many rangatahi will choose to go without care around sensitive issues that lack any guarantee of confidentiality.[30, 32, 33, 35]

Discuss confidentiality and its limits with all rangatahi and their whānau. Offer to see any rangatahi alone for part of their consultation.

Like adults, rangatahi have the right to be treated with respect and the right to access care confidentially. However, sharing information is required where there is a risk of harm — to the rangatahi or to others — or where others are harming the rangatahi. If confidentiality must be broken, talk with the rangatahi and create a plan for how you will do this.

Tip 5: Carry out a broad hauora assessment

All rangatahi should have a broad hauora assessment taken and updated regularly as part of their care. Using a good hauora or psychosocial assessment framework is at least as important as the physical exam when working alongside rangatahi.

Ensure interventions and follow-up are grounded in a strong understanding of rangatahi's social circumstances, cultural context, self-defined identity, and overall wellbeing. Doing this improves engagement and is more likely to deliver a successful plan.

Framework for psychosocial assessment

Get to know the rangatahi who you are working with and what they consider to be important. A popular framework for psychosocial assessment used in Aotearoa is HEEADSSS:

Home/Whakapapa; **E**ducation/Vocation; **E**ating; **A**ctivity; **D**rugs and other substances; **S**exual health; **S**uicide, mood, and mental health; **S**afety

The HEEADSSS framework helps health services explore different areas of a rangatahi's world.

Useful resource about the HEEADSSS framework:

Goldenring JM, et al.: HEEADSSS 3.0: The psychosocial interview for adolescents updated for a new century fuelled by media

Useful videos about the HEEADSSS framework:

Adolescent HEEADSSS — a cultural perspective on what's new with GenZ | Goodfellow Unit

HEEADSSS Assessment learning video resource — Youth health and wellbeing

Working with Youth: HEEADSSS Assessment | Goodfellow Unit

Other useful resources:

Te-Ukaipo-Framework-December

Overarching Te Ukaipo Podcast on Vimeo

A flexible approach allows a rapport to develop and secures engagement

Allow the conversation to move from typically less sensitive to more sensitive areas. This approach allows a rapport to develop with the rangatahi in a way that will maintain engagement.

Always invest time in whanaungatanga (nurturing of relationships) throughout. Remember the Te Tapatoru model[6] with its three interconnected concepts: Ko wai, he wā pai, and kaupapa pai. Do not use the model as a tick box but as a guide to conversation that will occur as part of a broader consultation. Establishing a conversation pattern with the rangatahi and their whānau as standard practice from the start is vital. For example, "we value getting to know all tamariki and rangatahi and whānau so we can work with you to support the best care." Normalise these types of conversations.

Be flexible with assessments and engagement. Depending on the circumstances, you could complete it over a few visits. For example:

- **If on a shift in a ward** — consider visiting at different times over two days.
- **If visiting monthly for BPG** — complete over a couple of visits and refresh every six months.

Incorporating elements of Te Ūkaipō into your approach will improve the value and quality of your time spent with the rangatahi and their whānau. Across the nine kaupapa Māori values and corresponding whakataukī, the full framework contains measures and outcomes for both rangatahi and kaimahi.[7]

Helping rangatahi to self-manage starts with a management plan

Management plans should always be made with rangatahi and their whānau. As rangatahi develop, they transition to the appropriate level of self-management of their hauora, with their whānau or caregivers walking alongside.

A particular time of risk is the transition between services. Providers must be respectful at this time. Clarify the differences in how services operate and deliver care as the rangatahi moves between services. Offer support services that might be available.

Guides that focus on transition may be helpful, such as Starship's Transition Tools: see https://starship.org.nz/transition-tools/

Self-management is a skill that requires support to develop. Do not allow it to simply happen. Rangatahi need to be enabled and supported to develop their skills in self-management.

Tip 6: Respect the rights of tamariki and rangatahi

Respect the rights of tamariki and rangatahi. This will facilitate the development of trust in the service, the care provided, and the team.

We encourage all tamariki and rangatahi to have whānau involved in their care, but rangatahi can access care independently and consent to treatment if they are competent to consent.[49]

Tip 7: Explore broader aspects of hauora

Move beyond the presenting issue (the initial reason) for the contact to explore broader aspects of hauora. Concerns other than RHD may impact adversely on, and be more important to, the rangatahi.

Ask them, "What's on top? What is important for you to get support with today?"

Tip 8: Empower rangatahi through health literacy

Health literacy is critical for empowerment. Rangatahi and whānau should be able to understand their health-related behaviours and contexts. They need to be able to describe their condition, medications and treatments, follow-up schedules, and how to access help if needed.

A rangatahi's positive experience of care is enhanced by the availability of developmentally appropriate health information in various formats.[34, 46] Check the rangatahi understands what you are saying. Also, address any knowledge gaps. If a rangatahi was diagnosed when they were a tamariki, much of the information presented to them was likely given to their whānau or caregivers. This may have led to the rangatahi not understanding much about their health. See **Online resources for whānau with Strep A, acute rheumatic fever and rheumatic heart disease**.

Steps to finding out what the rangatahi knows

Table 13.2 sets out steps to help understand the rangatahi's current level of knowledge and what you can do to assist.

Table 13.2. Actions to understand a rangatahi's current knowledge

Step	Action
Assess what the rangatahi already knows and understands.	**Assess** what the rangatahi already knows and understands about their disease and what's required to manage it. This includes them answering some questions. Remember to word the questions below in a way they can understand: • Do they understand the cause and complications of ARF? • Do they understand the reason for SAP? • Do they understand the signs and symptoms of recurrent ARF? • Do they know the difference between endocarditis and prophylaxis of ARF? • Do they know how to manage a sore throat (for themselves as well as their whānau/household members)? • Do they understand why medical follow-up is important? • Do they understand why dental follow-up is important? (to prevent endocarditis). • Are they pregnant or considering pregnancy? If they are, do they understand the importance of pregnancy planning and monitoring — including ensuring they are sharing their medical history and diagnosis with their pregnancy provider? • Do they know which people or agencies to contact for further information or help? • Do they know how to contact those relevant people or agencies?
Build on the rangatahi's health literacy skills and knowledge.	**Explain** concepts in a manner that: • Is culturally and developmentally appropriate. • Aligns with the rangatahi's literacy and cultural contexts (such as using simple visual aids, diagrams, or videos). **Ask** the rangatahi how they want the information (and in what format).
Take care with the language you use. **Check** their understanding of any steps frequently. **Explain** technical words simply to help the rangatahi understand.	**Give** information in an order of logical, simple steps, such as: • **Condition** — prevent Strep A infection (the name of the prevention measure). • **Action** — have regular penicillin injections as prescribed by the healthcare team (what action needs to happen). • **Rationale** — to prevent a further episode of ARF (why the action needs to happen).
Give small amounts of information that are important at the time.	Avoid overwhelming them by giving too much information at the same time. Keep it simple. Where possible, working with the same health professional at each visit may help achieve this.
Prioritise the information you provide.	Work with the rangatahi and their whānau to prioritise the information you provide: "When can you return to school and sport?" instead of "What happens when you are ready to have a tamariki?"
Give practical information.	Use practical application — put teaching into practice: "How do you contact the team who delivers benzathine penicillin?"
Reinforce and **emphasise** key points.	Repeat the main point and check they understand it. Anticipatory guidance related to their situation. For example: "If you start a job, how do we need to change when and where we deliver your medication to suit your needs?"
Check for understanding by using open questions.	Ask open questions for a more open conversation. Give scenarios to check what they would do in certain situations and if they've taken on board the information you have shared with them. For example: "Who would you contact if you ran out of warfarin?" "How often do you need to see the rheumatic fever clinic?" "What is a good way for you to know when your appointments are?" Asking yes/no questions will not help you find out how much the rangatahi or their whānau understand. Effective communication relies on the person giving the information — it is not a test for the rangatahi on how well they understood you. Here are some more examples: "Can you tell me key points around oral healthcare? I want to make sure I have explained everything so that you understand it. I also want to check that I haven't missed anything." "Why is oral healthcare important?" "What information must you remember to give the dentist?"

Tip 9: Consider co-locating rangatahi in hospital

For inpatient care — wherever possible, consider co-locating rangatahi in the hospital. Doing this will enhance developmental care, experience, and social support. Rangatahi identify cohorting as an important factor in their level of satisfaction.[34]

Tip 10: Offer opportunities for peer support

Offer opportunities for peer support for rangatahi and their whānau, where they can connect, share experiences and practical tips with others going through the same experience. Rangatahi and whānau value this connection.

"I know he needs to talk to somebody because half of the time he doesn't even know what he's got."

Aiga member of a rangatahi aged 14 who has RHD, discussing secondary antibiotic prophylaxis (SAP)

"I don't know why I get the injection — they just said I need it. I don't know how long for."

Rangatahi aged 13, receiving SAP for ARF

References

1. World Health Organization. Working for a brighter, healthier future. How WHO improves health and promotes wellbeing for the world's adolescents. Geneva: World Health Organization; 2022. https://www.paho.org/en/documents/working-brighter-healthier-future-how-who-improves-health-and-promotes-well-being-worlds (Accessed February 18 2025).

2. Ministry of Business, Innovation and Employment. Rangatahi — helping our young people thrive in the workforce. n.d. https://www.mbie.govt.nz/business-and-employment/employment-and-skills/regional-skills-leadership-groups/nelson-tasman/regional-workforce-plans/regional-workforce-plan/new-content-page/rangatahi-helping-our-young-people-thrive-in-the-workforce (Accessed February 18 2025).

3. Williams AD, Clark TC, Lewycka S. The associations between cultural identity and mental health outcomes for Indigenous Māori youth in New Zealand. *Frontiers in Public Health*. 2018;6:319. https://doi.org/10.3389/fpubh.2018.00319

4. Calder-Dawe O, Carlson T, Mulholland J, Squire D. Rangatahi perspectives on hauora and wellbeing: a qualitative report from Aotearoa. Victoria University of Wellington; 2023. https://openaccess.wgtn.ac.nz/articles/report/Rangatahi_perspectives_on_hauora_and_wellbeing_A_qualitative_report_from_Aotearoa/23904453?file=41908929 (Accessed February 18 2025).

5. Ministry of Health. Māori health models. Wellington: Ministry of Health; 2015. https://www.health.govt.nz/our-work/populations/maori-health/maori-health-models (Accessed February 17 2025).

6. Hamley L, Le Grice J, Greaves L, Groot S, Lindsay Latimer C, Renfrew L, et al. Te Tapatoru: a model of whanaungatanga to support rangatahi wellbeing. *Kōtuitui: New Zealand Journal of Social Sciences Online*. 2022;18(2):171–194. https://doi.org/10.1080/1177083x.2022.2109492

7. Health New Zealand | Te Whatu Ora. Te Ūkaipō Framework. 2023. https://www.tewhatuora.govt.nz/assets/For-the-health-sector/Specific-life-stage/Youth/SBHS/Te-Ukaipo-Framework-December-2023.pdf (Accessed February 18 2025).

8. World Health Organization. The second decade: improving adolescent health and development. Geneva: World Health Organization; 2001. https://iris.who.int/bitstream/handle/10665/64320/WHO_FRH_?sequence=1 (Accessed February 18 2025).

9. White PH, Cooley WC, Transitions Clinical Report Authoring Group, American Academy of Pediatrics, American Academy of Family Physicians, American College of Physicians. Supporting the health care transition from adolescence to adulthood in the medical home. *Pediatrics*. 2018;142(5). https://doi.org/10.1542/peds.2018-2587

10. Gray WN, Schaefer MR, Resmini-Rawlinson A, Wagoner ST. Barriers to Transition From Pediatric to Adult Care: A Systematic Review. *Journal of Pediatric Psychology*. 2018;43(5):488–502. https://doi.org/10.1093/jpepsy/jsx142

11. Colver A, Rapley T, Parr JR, McConachie H, Dovey-Pearce G, Couteur AL, et al. Facilitating transition of young people with long-term health conditions from children's to adults' healthcare services — implications of a 5-year research programme. *Clin Med (Lond)*. 2020;20(1):74–80. https://doi.org/10.7861/clinmed.2019-0077

12. Medical Council of New Zealand. Cultural safety Wellington: Medical Council of New Zealand; 2019. https://www.mcnz.org.nz/our-standards/current-standards/cultural-safety/ (Accessed February 18 2025).

13. Wong L, Wong A, Maher L, Farrant B, Palmer-Neels K, Pio F, et al. Co-design of youth appropriate services for young people with rheumatic fever/rheumatic heart disease in Counties Manukau District. *New Zealand Medical Journal*. 2023;136(1585):63–73. https://doi.org/10.26635/6965.6123

14. Engelman D, Ah Kee M, Mataika RL, Kado JH, Colquhoun SM, Tulloch J, et al. Secondary prevention for screening detected rheumatic heart disease: opportunities to improve adherence. *Transactions of the Royal Society of Tropical Medicine and Hygiene*. 2017;111(4):154–162. https://doi.org/10.1093/trstmh/trx035

15. Anderson A, Peat B, Ryland J, Ofanoa M, Burgess H, Malungahu G, et al. Mismatches between health service delivery and community expectations in the provision of secondary prophylaxis for rheumatic fever in New Zealand. *Australian and New Zealand Journal of Public Health*. 2019;43(3):294–299. https://doi.org/10.1111/1753-6405.12890

16. Clark TC, Ball J, Fenaughty J, Drayton B, Fleming TT, Rivera-Rodriguez C, et al. Indigenous adolescent health in Aotearoa New Zealand: trends, policy and advancing equity for rangatahi Maori, 2001–2019. *The Lancet Regional Health — Western Pacific*. 2022;28:100554. https://doi.org/10.1016/j.lanwpc.2022.100554

17. Nair M, Baltag V, Bose K, Boschi-Pinto C, Lambrechts T, Mathai M. Improving the quality of health care services for adolescents, globally: a standards-driven approach. *Journal of Adolescent Health*. 2015;57(3):288–298. https://doi.org/10.1016/j.jadohealth.2015.05.011

18. Campbell F, Biggs K, Aldiss SK, O'Neill PM, Clowes M, McDonagh J, et al. Transition of care for adolescents from paediatric services to adult health services. *Cochrane Database of Systematic Reviews*. 2016;4(4):CD009794. https://doi.org/10.1002/14651858.CD009794.pub2

19. Sawyer SM, Azzopardi PS, Wickremarathne D, Patton GC. The age of adolescence. *The Lancet Child & Adolescent Health*. 2018;2(3):223–228. https://doi.org/10.1016/s2352-4642(18)30022-1

20. Denny S, de Silva M, Fleming T, Clark T, Merry S, Ameratunga S, et al. The prevalence of chronic health conditions impacting on daily functioning and the association with emotional wellbeing among a national sample of high school students. *Journal of Adolescent Health*. 2014;54(4):410–415. https://doi.org/10.1016/j.jadohealth.2013.09.010

21. DiFusco LA, Schell KA, Saylor JL. Risk-taking behaviors in adolescents with chronic cardiac conditions: a scoping review. *Journal of Pediatric Nursing*. 2019;48:98–105. https://doi.org/10.1016/j.pedn.2019.07.011

22. Sullivan M, Ballantine K. The incidence of childhood cancer in New Zealand 2000–2009: the first outcome analysis of the New Zealand Children's Cancer Registry. Auckland: National Child Cancer Network; 2014. https://childcancernetwork.org.nz/wp-content/uploads/2015/10/Childhood-Cancer-Incidence-in-New-Zealand-2000-2009-1.pdf (Accessed February 18 2025).

23. Bloom SR, Kuhlthau K, Van Cleave J, Knapp AA, Newacheck P, Perrin JM. Health care transition for youth with special health care needs. *Journal of Adolescent Health*. 2012;51(3):213–219. https://doi.org/10.1016/j.jadohealth.2012.01.007

24. Denny S, Farrant B, Utter J, Fleming T, Bullen P, Peiris-John R, et al. The Prevalence of Postgraduate Education in Youth Health Among High School Clinicians and Associated Student Health Outcomes. *Journal of Adolescent Health*. 2016;59(5):555–561. https://doi.org/10.1016/j.jadohealth.2016.07.012

25. Wilson H, Bostock N, Phillip N, Shannon P, Payne D, Kennedy A. Opportunistic adolescent health screening of surgical inpatients. *Archives of Disease in Childhood*. 2012;97(10):919–921. https://doi.org/10.1136/archdischild-2012-301835

26. Payne D. Meeting the needs of young people in hospital. *Archives of Disease in Childhood*. 2013;98(12):930–932. https://doi.org/10.1136/archdischild-2013-304294

27. Hawkrigg S, Smith L, Johnson A, Kennedy A, Payne D. Opportunistic adolescent health assessment in the child protection unit. *Journal of Paediatrics and Child Health*. 2016;52(6):656–661. https://doi.org/10.1111/jpc.13203

28. Ara Taiohi. Mana taiohi. Wellington: Ara Taiohi; 2019. https://arataiohi.org.nz/mana-taiohi/ (Accessed February 17 2025).

29. Whitbeck L, Walls M, Hartshorn K. Indigenous adolescent development: psychological, social and historical contexts. London: Routledge, Taylor & Francis Group; 2014.

30. Denny S, Farrant B, Cosgriff J, Hart M, Cameron T, Johnson R, et al. Access to private and confidential health care among secondary school students in New Zealand. *Journal of Adolescent Health*. 2012;51(3):285–291. https://doi.org/10.1016/j.jadohealth.2011.12.020

31. Royal Australasian College of Physicians. RACP Joint Adolescent Health Committee. Confidential health care for adolescents and young people (12–24 years). Position statement: July 2021. 2021. https://www.racp.edu.au/docs/default-source/advocacy-library/racp-position-statement-confidential-health-care-for-adolescents-and-young-adults-12-24-years.pdf?sfvrsn=e77bc31a_6 (Accessed February 18 2025).

32. Ford CA. Influence of physician confidentiality assurances on adolescents' willingness to disclose information and seek future health care. A randomized controlled trial. *JAMA*. 1997;278(12):1029–1034. https://doi.org/10.1001/jama.1997.03550120089044

33. English A, Ford CA. More evidence supports the need to protect confidentiality in adolescent health care. *Journal of Adolescent Health*. 2007;40(3):199–200. https://doi.org/10.1016/j.jadohealth.2006.12.016

34. Ambresin AE, Bennett K, Patton GC, Sanci LA, Sawyer SM. Assessment of youth-friendly health care: a systematic review of indicators drawn from young people's perspectives. *Journal of Adolescent Health*. 2013;52(6):670–681. https://doi.org/10.1016/j.jadohealth.2012.12.014

35. Denny S, Farrant B, Cosgriff J, Harte M, Cameron T, Johnson R, et al. Forgone health care among secondary school students in New Zealand. *Journal of Primary Health Care*. 2013;5(1). https://doi.org/10.1071/hc13011

36. Sawyer S, Ambresin A-E, Bennett K, Hearps S, Romaniuk H, Patton G. Towards an Adolescent Friendly Children's Hospital. The RCH Adolescent Friendly Hospital Survey (2011 Baseline Assessment). Melbourne: Royal Children's Hospital. https://www.rch.org.au/uploadedFiles/Main/Content/cah/2012%20Adolscent%20Report.pdf (Accessed February 19 2025).

37. Sawyer SM, Ambresin AE, Bennett KE, Patton GC. A measurement framework for quality health care for adolescents in hospital. *Journal of Adolescent Health*. 2014;55(4):484–490. https://doi.org/10.1016/j.jadohealth.2014.01.023

38. Bécares L, Cormack D, Harris R. Ethnic density and area deprivation: neighbourhood effects on Māori health and racial discrimination in Aotearoa/New Zealand. *Social Science and Medicine*. 2013;88:76–82. https://doi.org/10.1016/j.socscimed.2013.04.007

39. Albertella L, Farrant B, Denny S. Improving the quality of care for adolescents and young adults on an adult medical ward. *Internal Medicine Journal*. 2022;52(9):1519–1524. https://doi.org/10.1111/imj.15463

40. de Dassel JL, de Klerk N, Carapetis JR, Ralph AP. How many doses make a difference? An analysis of secondary prevention of rheumatic fever and rheumatic heart disease. *Journal of the American Heart Association*. 2018;7(24):e010223. https://doi.org/10.1161/JAHA.118.010223

41. Kevat PM, Reeves BM, Ruben AR, Gunnarsson R. Adherence to secondary prophylaxis for acute rheumatic fever and rheumatic heart disease: a systematic review. *Current Cardiology Reviews*. 2017;13(2):155–166. https://doi.org/10.2174/1573403X13666170116120828

42. Ralph AP, de Dassel JL, Kirby A, Read C, Mitchell AG, Maguire GP, et al. Improving delivery of secondary prophylaxis for rheumatic heart disease in a high-burden setting: outcome of a stepped-wedge, community, randomized trial. *Journal of the American Heart Association*. 2018;7(14). https://doi.org/10.1161/JAHA.118.009308

43. Dennison A, Peat B, Wilson E, Leversha A, Wheeler M, Briggs S, et al. Rheumatic fever recurrences in New Zealand 2010–14. *New Zealand Medical Journal*. 2020;133(1516):47–57.

44. Mitchell AG, Belton S, Johnston V, Ralph AP. Transition to adult care for Aboriginal children with rheumatic fever: a review informed by a focussed ethnography in northern Australia. *Australian Journal of Primary Health*. 2018;24(1):9–13. https://doi.org/10.1071/PY17069

45. World Health Organization. Adolescent health. n.d. https://www.who.int/health-topics/adolescent-health#tab=tab_1 (Accessed February 18 2025).

46. World Health Organization. Making health services adolescent friendly: developing national quality standards for adolescent friendly health services. Switzerland: World Health Organization; 2012. https://www.who.int/publications/i/item/9789241503594.

47. Royal Australasian College of Physicians. Transition of young people with complex and chronic disability needs from paediatric to adult health services — position statement. Sydney: Royal Australasian College of Physicians; 2014. https://www.racp.edu.au/docs/default-source/advocacy-library/transition-of-young-people-with-complex-and-chronic-disability-needs.pdf (Accessed February 18 2025).

48. Anderson A, Spray J. Beyond awareness: towards a critically conscious health promotion for rheumatic fever in Aotearoa, New Zealand. *Social Science and Medicine*. 2020;247:112798. https://doi.org/10.1016/j.socscimed.2020.112798

49. van Rooyen A, Water T, Rasmussen S, Diesfeld K. What makes a child a 'competent' child? *New Zealand Medical Journal*. 2015;128(1426):88–95.

14

Screening for Rheumatic Heart Disease

In this chapter

Key changes

- A randomised control trial conducted in Uganda (the 'GOAL' study) published in 2022 has shown that secondary antibiotic prophylaxis (SAP) reduces disease progression of mild rheumatic heart disease (RHD) detected by echocardiography (echo). Echo screening for RHD broadly meets the criteria for a suitable screening test.
- The 2023 World Heart Federation guidelines describe four stages for the echo diagnosis of RHD. Stage A is equivalent to the 2012 WHF 'borderline' category, and Stage B is similar (but not identical) to the previous 'definite' RHD category. Stage C describes moderate or severe RHD. Stage D describes moderate or severe RHD with cardiac complications. See **Table 14.2**.

Key points

- Siblings of a person **recently diagnosed** with acute rheumatic fever (ARF) or new RHD should be offered echo screening (Grade C).
- Parents or guardians of a person under 30 years of age **recently diagnosed** with ARF or RHD should be offered echo screening (Grade D).
- Any pregnant person with RHD should be referred to their local high-risk maternity service for cardiac reassessment (Grade C).
- Current evidence does not support routine antenatal RHD screening for all Māori and Pacific peoples during pregnancy. However, lead maternity carers need to be aware that undetected RHD may be present in pregnant people/women from high-risk populations. Referral for suspected RHD is recommended on clinical grounds (Grade C).

Introduction

Prior to the mid-2000s, the World Health Organization (WHO) recommended school-based screening for RHD using auscultation with a stethoscope as a tool for estimating disease prevalence burden.[1] The WHO Global Programme on RHD undertook auscultatory screening of over one million tamariki.[2] In some regions of the world, this was augmented by echocardiography (cardiac ultrasound) to confirm the diagnosis of RHD. The field of echo screening for RHD rapidly expanded after the landmark publication from Cambodia and Mozambique in 2007[3] in which echo revealed a tenfold higher rate of RHD. Researchers used echo screening to define the true disease prevalence of RHD for their region and established that:

- Echo is more accurate than auscultation for screening.[4, 5]
- Portable echo can feasibly be conducted in school and community settings.[5, 6]
- Echo is acceptable to populations at risk.[7]

In 2012, a group of RHD researchers, led by Aotearoa and Australia, standardised the echo diagnosis of RHD using echocardiographic, surgical, and pathological descriptions of RHD. These diagnostic criteria received consensus agreement and endorsement from the World Heart Federation (WHF), ultimately becoming known as the WHF RHD Criteria.[8] The 2012 WHF Criteria became the gold standard for the echo diagnosis of mild RHD and have been widely adopted. Good inter-observer agreement has been found using the WHF criteria internationally[9] and locally.[10] In 2023, an expanded group of global RHD researchers and clinical experts published the second edition of the WHF criteria.[11]

Screening for rheumatic heart disease in Aotearoa

The first echo school screening study in Aotearoa was undertaken in South Auckland in 2007 and 2008.[5] Other regions followed, with collaborations involving public health, community paediatricians, nurses, research teams, and local school communities. Screening was undertaken in Te Tai Rāwhiti, [6] Porirua,[7] Bay of Plenty,[12] and Northland (personal communication, 2024, R. Tuck, Paediatrician, Health New Zealand | Te Whatu Ora). As expected, higher rates of RHD are found in adults than in tamariki in Aotearoa.[13, 14] A wider discussion of research from Aotearoa was published in 2024.[15]

In the early years of echo screening, there was no evidence individuals who had echo-detected RHD and began SAP had better clinical outcomes than those who did not receive penicillin.[16] However, since the publication of the New Zealand 2014 Rheumatic Fever Guidelines,[17] evidence has accumulated regarding the trajectory and evolution of mild RHD detected by echo.

In the 'GOAL' study, a randomised control trial conducted in Uganda,[18] 799 tamariki with mild RHD were randomised to benzathine benzylpenicillin G (BPG) or no intervention. After two years, RHD progression was observed in 8.3% of the non-treated group compared to 0.8% of those receiving BPG. Almost half of those with disease progression of RHD progressed to moderate-severe RHD. These findings from the GOAL study provide evidence that penicillin significantly reduces disease progression for an individual with echocardiographic mild RHD. However, 48% of those in both the treated and untreated groups had regression to a normal echo after two years, highlighting the need for surveillance with echo rather than treating all with BPG. While some adverse effects were reported in the prophylaxis group, they were mostly mild.[18]

Global perspective

After more than 15 years of research, bookmarked by two RHD papers published in the *New England Journal of Medicine*,[3, 18] echo screening for RHD now broadly meets the following criteria for a suitable screening test:[19]

- It is an important health issue.
- It has a recognisable latent or asymptomatic phase.
- It has an understood natural history.
- It has a suitable screening test.
- It has an accepted treatment that improves clinical outcomes.

Additional screening considerations include social and ethical issues, understanding the potential harms of screening, management of incidental findings, health system capacity and capability, cost-benefit analysis,[20] and in the Aotearoa setting, cultural safety, and responsiveness to Māori (and Pacific peoples).[19]

The number of RHD screening programmes is increasing in many regions and countries, as reflected by numerous presentations and discussions at the RHD World Congress in Abu Dhabi in late 2023. Experts and health officials in high-prevalence countries now recognise that scaled-up echo screening has the potential to play a key role in global RHD control. In other words, RHD echo screening has transitioned from the research phase to the implementation phase.

2023 World Heart Federation guidelines for the echocardiographic diagnosis of rheumatic heart disease

The 2023 World Heart Federation guidelines for the echocardiographic diagnosis of rheumatic heart disease introduce a new stage-based classification for RHD. This classification acknowledges the spectrum of disease and describes the risk of disease progression.[11] The 2023 WHF Criteria can be applied in both screening and clinical settings for individuals with newly diagnosed RHD,[11] but they are not intended to categorise carditis of ARF.

The 2023 WHF guidelines have two sets of echocardiographic criteria for RHD:[11]

- 'Screening criteria' for non-experts to use in appropriate settings, often resource-limited to detect suspected cases of RHD.
- 'Confirmatory criteria' are designed for experts to use to confirm an RHD diagnosis.

Morphological features for rheumatic heart disease

The colour-Doppler features for pathological mitral regurgitation (MR) and pathological aortic regurgitation (AR) are listed in Table 10.2 of Chapter 10: Diagnosis of Rheumatic Heart Disease. The morphological features of RHD, as listed in the 2023 WHF guidelines for echo diagnosis of RHD,[11] are shown in Table 14.1.

Table 14.1. Confirmatory morphological features of rheumatic heart disease

Valve	Morphological characteristics
Mitral valve	• Valve apparatus thickening (defined by the presence of either or both): – Anterior leaflet thickening[a, b] – Chordal thickening[c] • Valve mobility abnormalities (defined by the presence of either or both): – Restricted anterior or posterior leaflet motion in diastole[d] – Excessive anterior leaflet tip motion during systole[e]
Aortic valve	• Cusp thickening[f] • Cusp prolapse • Restricted cusp motion • Coaptation defect in diastole

Explanations and technical considerations

[a] Anterior mitral valve leaflet (AMVL) thickness should be measured during diastole at the full excursion. Measurement should be taken at the thickest portion of the leaflet, including focal thickening, beading and nodularity. Ideally, the measurement should be performed on a frame with maximal separation of chordae from the leaflet tissue.

[b] Abnormal thickening of the AMVL is age-specific and defined as follows: ≥3.0 mm for individuals aged ≤20 years, ≥4.0 mm for individuals aged 21–40 years and ≥5.0 mm for individuals aged >40 years.

[c] Chordal thickening might range from individual chordae tendineae to multiple chordae tendineae fusion and calcification. The structures of papillary muscles, chordae tendineae and margins of leaflets might not be clearly distinguishable. Assessment is subjective; however, in most cases, chordal thickening is attributable to fusion of two or more chordae and is seen as an echo-bright structure near to the leaflet tip insertion. Chordal thickening is typically associated with other morphological features.

[d] Restricted leaflet motion of either the AMVL or the posterior mitral valve leaflet is usually the result of chordal shortening or fusion, commissural fusion or leaflet thickening.

[e] Excessive leaflet tip motion results from elongation of the primary chords and is defined as displacement of the tip or edge of an involved leaflet towards the left atrium, resulting in abnormal coaptation and regurgitation. Excessive leaflet tip motion does not need to meet the standard echocardiographic definition of mitral valve prolapse disease,[8] given that they refer to different disease processes. This characteristic applies only to those aged <35 years.

[f] In the parasternal short-axis view, the right and non-coronary aortic cusp closure line often presents as echogenic (thickened) in healthy individuals, which should be considered normal. Image optimisation for valve thickness and morphology, including harmonic imaging, should be individualised on the basis of echocardiography devices and those performing echocardiograms.

Rwebembera J, Marangou J, Mwita JC, Mocumbi AO, Mota C, Okello E, et al. 2023 World Heart Federation guidelines for the echocardiographic diagnosis of rheumatic heart disease. *Nature Reviews: Cardiology*. 21, 4, 250–263. 2020, reproduced with permission from SNCSC. References in the footnotes are those of the original publication.

Rheumatic heart disease Stages A to D

The 2023 WHF guidelines classify RHD into stages A, B, C, and D based on the risk of progression to more advanced valvular heart disease. The terms 'borderline' and 'definite' RHD in the 2012 guidelines are now Stage A and Stage B respectively, with minor changes. The guidelines also recommend management strategies for each stage of RHD.[11]

Figure 14.1 shows stages A to D of RHD, as described in the 2023 World Heart Federation guidelines for the echocardiographic diagnosis of rheumatic heart disease.

Staging of rheumatic heart disease detected by echocardiography[a,b]

Stage A: minimal echocardiographic criteria for RHD
- Applies only to individuals aged ≤20 years old
- Clinical risk: might be at risk of valvular heart disease progression
- Echocardiographic features: the presence of mild[c] MR or AR without morphological features

Stage B: mild RHD
- Can apply to any age
- Clinical risk: at moderate or high risk of progression and at risk of developing symptoms of valvular heart disease
- Echocardiographic features: evidence of mild[c] valvular regurgitation plus at least one morphological feature in individuals aged ≤20 years and at least two morphological features in individuals aged >20 years[d]; or mild regurgitation in both mitral and aortic valves

Stage C: advanced RHD at risk of clinical complications
- Can apply to any age
- Clinical risk: at high risk of developing clinical complications that require medical or surgical intervention
- Echocardiographic features: moderate or severe MR, moderate or severe AR, any MS or AS[e], pulmonary hypertension and decreased LV systolic function

Stage D: advanced RHD with clinical complications
- Can apply to any age
- Clinical risk: established clinical complications include cardiac surgery, heart failure, arrhythmia, stroke and infective endocarditis
- Echocardiographic features: moderate or severe MR, moderate or severe AR, any MS or AS[e], pulmonary hypertension and decreased LV systolic function

AR, aortic regurgitation; AS, aortic stenosis; LV, left ventricular; MR, mitral regurgitation; MS, mitral stenosis, RHD, rheumatic heart disease. [a]To be applied in high-risk settings[3,51] and requires other causes of valvular heart disease to have been excluded. [b]After the application of the confirmatory echocardiographic criteria, diagnostic categories might include 'normal' and 'other', which encompasses other diseases such as congenital heart disease, cardiomyopathies and pericardial effusion. [c]Fulfilling the confirmatory criteria for pathological regurgitation (Box 4). [d]This cut-off value is derived from expert consensus. [e]Aortic stenosis is defined in accordance with international guidelines on valvular heart disease. A diagnosis of rheumatic aortic stenosis requires the exclusion of other causes, including bicuspid aortic valve and degenerative calcific AS.

Figure 14.1. Staging of rheumatic heart disease detected by echocardiography

Rwebembera J, Marangou J, Mwita JC, Mocumbi AO, Mota C, Okello E, et al. 2023 World Heart Federation guidelines for the echocardiographic diagnosis of rheumatic heart disease. *Nature Reviews: Cardiology*. 21, 4, 250–263. 2020, reproduced with permission from SNCSC.

Stage A: Minimal echocardiographic criteria for RHD

Stage A indicates the presence of early valvular changes that meet the minimum diagnostic criteria for RHD in a high-prevalence population. Individuals with Stage A disease may be at risk of developing valvular heart disease but are considered to have a low risk of disease progression. Note that a proportion of these individuals have the upper limit of physiological valvular regurgitation.[21-23]

This Stage A category corresponds to the disease stage previously known as 'borderline RHD'[8] and acknowledges the continuum of echo features from normal variant to mild RHD.

Note that Stage A RHD should not be diagnosed in individuals over 20 years of age.

A previously used subset of borderline RHD,[8] which involved the presence of morphological features alone without valvular regurgitation, no longer qualifies individuals for Stage A disease. The presence of morphological features alone is observed only in a very small number of individuals and is considered unlikely to be associated with the development of RHD.[11]

Stage B: Mild RHD

Stage B indicates the presence of mild pathological regurgitation (MR or AR) and one morphological feature of RHD. According to the 2012 guidelines, definite RHD required two morphological features.

The presence of both aortic and mitral pathological regurgitation, but without morphological features, is also classified as Stage B, given that the concurrence of pathological mitral and aortic regurgitation is probably indicative of true pathology.

Patients with Stage B disease are considered to have a moderate or high risk of disease progression based on the risk score.

Stage C: Moderate or severe RHD detected by echocardiography

Stage C indicates moderate or severe MR, moderate or severe AR, any mitral stenosis (MS), or any aortic stenosis (AS).

Stage D: Advanced RHD

Stage D indicates moderate or severe RHD (that is, the same as Stage C) **and** clinical complications of RHD, as described in **Figure 14.1**. Stages B, C, and D can apply to individuals of any age. Stage A only applies to individuals under 20 years of age.

Table 14.2 compares the echo features between the 2023 World Heart Federation Guidelines for the echocardiographic diagnosis of rheumatic heart disease[11] and the original 2012 WHF guidelines.[8]

"The lady [sonographer] made him [son with RHD] feel comfortable. She put a blanket on him. I think I had been to another one [echocardiogram], he was very uncomfortable and was cold, he didn't seem comfortable at all, and it was rushed."

Mother of tamaiti with RHD

Table 14.2. Comparison of World Health Federation (WHF) 2023 and 2012 categories and criteria/definitions

WHF 2023	WHF 2012
Stage A • Mild pathological MR • Mild pathological AR	**Borderline RHD** • Pathological MR • Pathological AR At least two morphological features of RHD of the MV without pathological MR or MS*
Stage B • Mild MR or mild AR + 1 morphological feature (<20 years) • Mild MR or AR + 2 morphological features (>20 years) • Both pathological mild MR and AR	**Definite RHD** • Pathological MR and at least two morphological features of RHD of the MV • MS mean gradient ≥4 mmHg* • Pathological AR and at least two morphological features of RHD of the AV • Borderline disease of both the AV and MV
Stages C and D Stage C: moderate or severe RHD Stage D: moderate or severe RHD plus clinical complications of RHD	**No equivalent**

* No longer used in the 2023 criteria.

Recommendations for care pathways for echocardiographic detected rheumatic heart disease

The New Zealand 2014 guidelines[17] previously recommended that:

- A person with 'borderline RHD' be followed up.
- A person with 'definite RHD' be started on SAP (Grade D).

The 2023 WHF guidelines are more prescriptive.[11]

Table 14.3 outlines the initial care and SAP for each stage of RHD, including the class of recommendation and level of evidence.

Table 14.3. Recommendations for care pathways for echocardiographic detected rheumatic heart disease by stage

Stage	Initial care	Secondary antibiotic prophylaxis (SAP) duration	Class of recommendation	Level of evidence
A	Counselling whānau the echo finding may or may not prove to be RHD.# Register on the Rheumatic Fever Care Coordination Systsem (RFCCS)** only if starting on SAP. Review with a comprehensive echo in 1–2 years.	The usual recommendation is **not** to start SAP (Grade D) as the risk-benefit of SAP is uncertain.# SAP may be commenced after a discussion with the whānau. If SAP is started, review with an echo in 1–2 years. Consider stopping SAP if the echo findings warrant this action. If SAP has not been started, but the follow-up echo shows persisting Stage A (mild) RHD, 5 years of SAP is recommended. If the echo features progress to Stage B, SAP for 10 years (total duration) is recommended.	2B*	B–R
B	Register on the RFCCS.** Recommend commencing SAP. Clinical follow-up as for mild RHD with repeat echo within 1–2 years.	10 year duration.##	1	A
C	Register on the RFCCS. Start SAP. Start specialised RHD care according to the Aotearoa guidelines. (See **Chapter 11: Management of Rheumatic Heart Disease**)	Until age 21 or for 10 years, whichever is longer, and then reassess. (See **Chapter 8: Secondary Prevention**) Those with continued moderate or severe RHD should continue SAP until age 30–35 years. Recurrences of ARF are rare after 35 years.	1	B–NR
D	Register on the RFCCS. Start SAP. Implement specialised RHD care according to the Aotearoa guidelines. (See **Chapter 11: Management of Rheumatic Heart Disease**)	For 10 years or until age 30–35 years, whichever is longer, and then reassess. Those with continued moderate or severe RHD should continue SAP until age 30 years. Recurrences of ARF are rare after 35 years. (See **Chapter 8: Secondary Prevention**)	1	B–NR

* The classification of recommendations and levels of evidence used in the 2023 WHF guidelines were in accordance with the 2013 American College of Cardiology/American Heart Association grading system.[24]

** Referral to an ARF Secondary Prevention Service will result in registration on the RFCCS.

Adapted from the WHF 2023 guidelines[11] for the Aotearoa setting.

Notes:

The risk-benefit for SAP at Stage A is unclear. Shared decision-making regarding individual patient preferences about SAP is required.

'Active surveillance' is a term used in public health for patients where follow-up without treatment is recommended.[25]

In Aotearoa, counselling with the whānau should include a discussion that their tamariki may or may not be at risk of progression of RHD.

The GOAL study[18] found that 13 (95% CI, 10 to 21) was the number needed to receive SAP to prevent one tamariki from having progression.

Stage A may also be the upper limit of physiological valvular regurgitation.[21-23] In addition, in the GOAL study, 48% of those in the control group (not receiving SAP) had regression of RHD at the two-year follow-up.

For these reasons, the usual recommendation is to **only start** SAP (Grade C) in Aotearoa when follow-up echo shows persistence or progression of RHD. This recommendation is unchanged from the 2014 New Zealand guidelines.[17]

Therefore, all tamariki with Stage A disease must have access to follow-up echo and longitudinal clinical evaluation to monitor for disease progression or regression. In the interim, all primary prevention measures are paramount to treat Strep A throat and skin infections. The tamaiti should be enrolled on to the register of the echo screening programme to ensure follow-up occurs in conjunction with the regional ARF and paediatric or cardiology services. To minimise harm from the diagnosis of RHD, counselling should emphasise that exercise poses no cardiac risks.

The experience from previous echo screening in Aotearoa is that whānau of tamariki with Stage A (previously termed 'borderline RHD') tend to choose SAP if another whānau member has had ARF or RHD.

The recommendation for 10 years (rather than until 21 years, whichever is longer) is influenced by international practices,[11] with a growing trend towards shorter SAP duration following RHD screening compared to post-ARF management.

Potential harms of screening

Harm will occur by a false positive result causing over-diagnosis of RHD or by a false negative causing under-diagnosis of RHD. Programmes may also result in harm by causing anxiety and decreasing quality of life after the abnormal test for some individuals. Studies of the impact of RHD screening have found that a negative screening test result has no negative impact.[26] However, a positive result can lead to increased anxiety, decreased physical activity, and decreased quality of life for the tamariki, parents, and whānau.[27, 28]

Healthcare workers counselling whānau about screening results should be aware of these issues and stress the importance of continued full participation in sports and physical activities for people with Stage A and B RHD. A study from Uganda showed that peer support groups have a positive impact on quality of life.[29]

Overall, the international consensus is that the benefits of preventing advanced RHD outweigh the harms of screening. As of 2024, a national programme of RHD screening has yet to be endorsed by the National Screening Advisory Committee in Aotearoa.

Barriers and enablers to perform screening

Addressing the enablers and barriers to carrying out screening is important to ensure equitable outcomes among communities, such as for Māori and Pacific peoples.

The 'Te Maatai Manawa a Whaanau: whānau heart screening study' demonstrated a whānau-informed model of care for ARF and RHD and identified enablers such as warmth and kindness, clear communication, and the involvement of kaiāwhina (support workers).[30] Conversely, barriers identified included a lack of support workers and an unwelcoming or indifferent approach.[30]

Considerations for developing a school-based echocardiographic screening programme

When developing a school-based echocardiographic screening programme, several factors should be taken into account, including but not limited to:

1. Endorsement by the Ministry of Health NZ.
2. Community engagement — establishing strong connections with local community.
3. Partnership with schools and whānau — ensuring collaboration with schools and whānau, along with the capacity to undertake school visits to educate teachers, students, and whānau.
4. Informed consent — providing clear information to whānau and obtaining informed consent for participation.

5. Programme logistics, such as:
    - Echo technology — hand-held versus portable devices, power sources, batteries.
    - Location and flow — appropriate scanning room, plinths, curtains, changing rooms, waiting areas, and student flow.
    - Echocardiographer workforce — ensuring adequate staffing with appropriate levels of training.
    - Reporting clinicians — echocardiographers, cardiologists, and physicians experienced in RHD echo interpretation.
6. Echo protocols — establishing criteria for abbreviated screening and full confirmatory echo.
7. Reporting systems — ensuring timely reporting and communication, including rapid release of results to whānau and copies sent to primary care providers.
8. Healthcare system capacity — providing clinical consultation and counselling for individuals with abnormal scans, including RHD, congenital heart disease (CHD) and other detected abnormalities.* Relevant to this, ARF (or new RHD) sibling screening is expected to be discussed at the National Screening Advisory Committee (NSAC) in 2025. NSAC's criteria will be used to assess sibling screening including health system requirements and readiness for a national screening programme.
9. Agreed treatment thresholds, including active surveillance of Stage A RHD.
10. Care pathways — establishing links to ongoing care for RHD and CHD patients, including access to SAP where appropriate.
11. Monitoring and evaluation of the programme.

* CHD and other detected abnormalities can represent a significant proportion of all abnormal scans identified through an RHD screening programme.[5]

Rheumatic heart disease echo school screening pilot in Aotearoa

As part of the Rheumatic Fever Roadmap 2023–2028,[31] in 2023 Health New Zealand | Te Whatu Ora announced funding for a feasibility study on RHD echo screening to detect undiagnosed RHD among tamariki and rangatahi in high-prevalence regions. The design is a cluster randomised control trial comparing the effectiveness and cost-effectiveness of school versus school and community-based settings of echo screening for undetected RHD for year 7 and 8 students. Planning began in 2024.

An implementation science approach will be used to assess community acceptability, workforce logistics, and delivery models (such as school-based screening and mobile bus services) to inform the case for a targeted national screening programme for high-risk populations.[31] It is currently unknown whether there is workforce capacity to develop a sustained programme of RHD screening in Aotearoa, particularly sonographers and an expert reporting workforce. The pilot will also analyse healthcare system capacity to deliver follow-up clinical care as well as SAP.

Globally, 'task shifting' is a strategy endorsed by the WHO to improve efficiency in healthcare systems with shortages in skilled healthcare workers.[32] The concept, when applied to RHD echo screening, is for the initial screening echo to be performed by 'briefly trained' healthcare workers who have undergone shorter, focused training programmes. This model has been adapted for RHD screening in low- or middle-income countries to address the shortage of skilled RHD echo screening staff.[33]

The study will also explore the utility of briefly trained healthcare workers to perform the initial screening echo building on the 'Te Maatai Manawa a Whaanau: Family heart screening study' which started in 2023. Led by nurses performing the screening echo, the study focuses on detecting RHD in both tamariki and rangatahi of whānau members who have recently had ARF or RHD. Co-designed with the community, the study aims to enrol siblings of tamariki diagnosed with ARF, providing holistic culturally responsive care and comprehensive health assessments.[30]

Echo screening using a trained echocardiographic sonographer for the initial screening has the logistic advantage that the sonographer can immediately move to the comprehensive protocol when either an RHD abnormality or non-RHD abnormality (such as CHD) is suspected. The utility of the sonographer workforce is critical for any national roll out of this programme of RHD screening in Aotearoa.

Artificial intelligence (AI) analysis has the potential for reporting RHD echo screening studies.[34] Currently, the use of AI for diagnosing RHD is in the research domain.

Table 14.4. Initial screening criteria for rheumatic heart disease

Initial screening criteria
Non-experts, such as briefly trained healthcare workers, or trained echocardiographers can do this initial screening.
Mitral regurgitation (requires all the following) • In individuals weighing <30 kg: MR jet length ≥1.5 cm* • In individuals weighing ≥30 kg: MR jet length ≥2.0 cm • MR jet is observed in at least one view • MR jet is observed in at least two consecutive frames
Aortic regurgitation (requires all the following) • Any AR • Observed in at least one view • Observed in at least two consecutive frames
Mitral stenosis • Restricted leaflet motion, with reduced valve opening
Positive screen: proceed to a confirmatory/diagnostic echo • Presence of any of the defined MR, AR or MS
Negative screen • Absence of any of the defined MR, AR or MS

AR — aortic regurgitation
MR — mitral regurgitation
MS — mitral stenosis

* This cut-off value comes from expert consensus. If the weight is not available, apply an age cut-off of <10 years or ≥10 years.

Adapted from the 2023 World Heart Federation guidelines for the echocardiographic diagnosis of rheumatic heart disease.[11]

Targeted screening

This brief section focuses on screening for RHD in two groups: first-degree relatives of people diagnosed with ARF or RHD and people who are pregnant.

First-degree relatives

A small but significant increased familial risk of RHD (2–3%) has been found in first-degree relatives (siblings or parents) compared to the general population in Aotearoa, though it remains unclear whether this is driven by genetic, environmental factors, or both.[35] The risk was highest when the index case had severe RHD.[35] Similar familial risk patterns have also been shown previously in Uganda.[36]

The above data for RHD is concordant with the marked five-fold higher ARF risk for those with a whānau history of ARF (OR 4·97; 95% CI 2·53–9·77) using the whānau reported history in the ARF risk factor study in Aotearoa.[37]

Recommendations

1. Siblings of an individual with a recent diagnosis of ARF (or new RHD) should be offered echo screening (Grade C).
2. Parents or guardians of an individual with a recent diagnosis of ARF or RHD in a person under 30 years of age should be offered echo screening (Grade D).#
3. First-degree relatives of people with established RHD+ if two or more whānau affected (Grade D recommendation).

It is reasonable to discuss the familial risk of RHD (2%–3%) for parents or guardians of individuals with newly diagnosed ARF or RHD, especially if the individual has moderate or severe carditis or moderate or severe RHD. It can be argued that parents have less benefit from preventing the progression of RHD as the chances of recurrence of ARF will be lower. This cut-off age is determined by consensus.

+ Currently, offering echo screening on a retrospective basis to all whānau with an individual who has established RHD is not being recommended. This is due in part to the expected low uptake and in part due to recommendations 1,2,3 above resulting in overburden of the current echo services in some regions of Aotearoa. However, if requested, it is reasonable to offer screening for first-degree relatives under 30 years of age.

Screening for rheumatic heart disease in people who are pregnant

There is a sound rationale for targeted screening for RHD in pregnancy[38, 39] in countries that have both:

- A high prevalence of RHD.
- Undiagnosed RHD (especially mitral stenosis) that carries a risk of maternal morbidity and mortality.[18, 40]

However, as of 2024, the evidence about pregnancy outcomes for those who have had echo screening compared to standard antenatal care is lacking.[41] Regional maternal outcomes for RHD were shown in the Australasian Maternity Outcomes Surveillance System (AMOSS) study in 2012.[42] Since then, anecdotally in the hui and other discussions for these guidelines, several cardiologists and obstetricians have reported examples of individuals with RHD presenting with decompensation in mid-to-late pregnancy or post-partum. An awareness that undetected RHD may be present in pregnant people/women from high-risk population groups is important for lead maternity carers.

An echo to screen for RHD in pregnancy should be requested if the pregnant person is Māori or Pacific or a recent migrant from high-risk endemic RHD regions and there is a clinical suspicion of RHD. Clinical suspicion includes excessive breathlessness on exertion or a whānau history of RHD.[35-37] (Also see **Chapter 12: Rheumatic Heart Disease and Pregnancy**).

Recommendations

Any pregnant person with RHD should be referred to their local high-risk maternity service for cardiac reassessment (Grade C).

Current evidence does not support routine screening. However, referral for suspected RHD is recommended on clinical grounds (Grade C).

References

1. World Health Organization. Rheumatic fever and rheumatic heart disease: report of WHO expert consultation. WHO Technical Report Series. Geneva: World Health Organization; 2001. https://iris.who.int/bitstream/handle/10665/42898/WHO_TRS_923.pdf?sequence=1&isAllowed=y (Accessed February 18 2025).

2. World Health Organization. The WHO global programme for the prevention of rheumatic fever and rheumatic heart disease: report of a consultation to review progress and develop future activities: Geneva, 29 November–1 December 1999. Geneva: World Health Organization; 2000. http://apps.who.int/iris/bitstream/10665/66273/1/WHO_CVD_00.1.pdf (Accessed February 18 2025).

3. Marijon E, Ou P, Celermajer DS, Ferreira B, Mocumbi AO, Jani D, et al. Prevalence of rheumatic heart disease detected by echocardiographic screening. *New England Journal of Medicine*. 2007;357(5):470–476. https://doi.org/10.1056/NEJMoa065085

4. Carapetis JR, Hardy M, Fakakovikaetau T, Taib R, Wilkinson L, Penny DJ, et al. Evaluation of a screening protocol using auscultation and portable echocardiography to detect asymptomatic rheumatic heart disease in Tongan schoolchildren. *Nature Clinical Practice: Cardiovascular Medicine*. 2008;5(7):411–417. https://doi.org/10.1038/ncpcardio1185

5. Webb RH, Wilson NJ, Lennon DR, Wilson EM, Nicholson RW, Gentles TL, et al. Optimising echocardiographic screening for rheumatic heart disease in New Zealand: not all valve disease is rheumatic. *Cardiology in the Young*. 2011;21(4):436–443. https://doi.org/10.1017/S1047951111000266

6. Cramp G, Stonehouse M, Webb R, Webb R, Chaffey-Aupouri G, Wilson N. Undetected rheumatic heart disease revealed using portable echocardiography in a population of school students in Tairawhiti, New Zealand. *New Zealand Medical Journal*. 2012;125(1363):53–64.

7. Perelini F, Blair N, Wilson N, Farrell A, Aitken A. Family acceptability of school-based echocardiographic screening for rheumatic heart disease in a high-risk population in New Zealand. *Journal of Paediatrics and Child Health*. 2015;51(7):682–688. https://doi.org/10.1111/jpc.12829

8. Rémenyi B, Wilson N, Steer A, Ferreira B, Kado J, Kumar K, et al. World Heart Federation criteria for echocardiographic diagnosis of rheumatic heart disease--an evidence-based guideline. *Nature Reviews: Cardiology*. 2012;9(5):297–309. https://doi.org/10.1038/nrcardio.2012.7

9. Remenyi B, Carapetis J, Stirling JW, Ferreira B, Kumar K, Lawrenson J, et al. Inter-rater and intra-rater reliability and agreement of echocardiographic diagnosis of rheumatic heart disease using the World Heart Federation evidence-based criteria. *Heart Asia*. 2019;11(2):e011233. https://doi.org/10.1136/heartasia-2019-011233

10. Culliford-Semmens N, Nicholson R, Tilton E, Stirling J, Sidhu K, Webb R, et al. The World Heart Federation criteria raise the threshold of diagnosis for mild rheumatic heart disease: three reviewers are better than one. *International Journal of Cardiology*. 2019;291:112–118. https://doi.org/10.1016/j.ijcard.2019.02.058

11. Rwebembera J, Marangou J, Mwita JC, Mocumbi AO, Mota C, Okello E, et al. 2023 World Heart Federation guidelines for the echocardiographic diagnosis of rheumatic heart disease. *Nature Reviews: Cardiology*. 2024;21(4):250–263. https://doi.org/10.1038/s41569-023-00940-9

12. Malcolm J, Ball S, Beharry J, Seal R, Lowe L, Biddle S, et al. Tautoko rheumatic hearts: to support those with rheumatic hearts, public health needs innovation, collaboration and evaluation. In: Zander A, Came H, editors. Proceedings of the 2013 Public Health Association Conference. Auckland: Public Health Association; 2013. p. 107–112.

13. Webb R. Rheumatic heart disease in New Zealand children: echocardiographic disease burden and clinical outcomes. [Master's thesis]. Auckland: University of Auckland; 2019. https://researchspace.auckland.ac.nz/items/641ebeb5-ade3-45e3-852d-927a423ac84e

14. Webb R, Culliford-Semmens N, ChanMow A, Doughty R, Tilton E, Peat B, et al. High burden of rheumatic heart disease confirmed by echocardiography among Pacific adults living in New Zealand. *Open Heart*. 2023;10(1):e002253. https://doi.org/10.1136/openhrt-2023-002253

15. Wilson N, Anderson A, Baker MG, Bennett J, Dennison A, McGregor R, et al. The roles of immunomodulator treatment and echocardiographic screening in rheumatic fever and rheumatic heart disease control: research from Aotearoa, New Zealand. *Journal of the Royal Society of New Zealand*. 2025;55(2):241–266. https://doi.org/10.1080/03036758.2024.2306981

16. Karthikeyan G. Measuring and reporting disease progression in subclinical rheumatic heart disease. *Heart Asia*. 2016;8(2):74–75. https://doi.org/10.1136/heartasia-2016-010857

17. New Zealand Heart Foundation. New Zealand guidelines for rheumatic fever: diagnosis, management and secondary prevention of acute rheumatic fever and rheumatic heart disease: 2014 update. Heart Foundation; 2014. https://www.heartfoundation.org.nz/resources/acute-rheumatic-fever-and-rheumatic-heart-disease-guideline (Accessed December 16 2024).

18. Beaton A, Okello E, Rwebembera J, Grobler A, Engelman D, Alepere J, et al. Secondary antibiotic prophylaxis for latent rheumatic heart disease. *New England Journal of Medicine*. 2022;386(3):230–240. https://doi.org/10.1056/NEJMoa2102074

19. National Advisory Committee on Health and Disability. Screening to improve health in New Zealand: criteria to assess screening programmes, April 2003. Wellington: National Health Committee; 2003. https://www.tewhatuora.govt.nz/assets/For-the-health-sector/NSU/Publications/Screening-to-Improve-Health-in-New-Zealand-Criteria-to-assess-screening-programmes-pdf-325-KB.pdf (Accessed February 17 2025).

20. National Advisory Committee on Health and Disability. Screening to improve health in New Zealand: criteria to assess screening programmes, April 2003. Wellington: National Health Committee; 2003. https://www.tewhatuora.govt.nz/assets/For-the-health-sector/NSU/Publications/Screening-to-Improve-Health-in-New-Zealand-Criteria-to-assess-screening-programmes-pdf-325-KB.pdf (Accessed February 18 2025).

21. Clark BC, Krishnan A, McCarter R, Scheel J, Sable C, Beaton A. Using a low-risk population to estimate the specificity of the World Heart Federation criteria for the diagnosis of rheumatic heart disease. *Journal of the American Society of Echocardiography*. 2016;29(3):253–258. https://doi.org/10.1016/j.echo.2015.11.013

22. Roberts K, Maguire G, Brown A, Atkinson D, Remény B, Wheaton G, et al. Echocardiographic screening for rheumatic heart disease in high and low risk Australian children. *Circulation*. 2014;129(19):1953–1961. https://doi.org/10.1161/circulationaha.113.003495

23. Webb RH, Gentles TL, Stirling JW, Lee M, O'Donnell C, Wilson NJ. Valvular regurgitation using portable echocardiography in a healthy student population: implications for rheumatic heart disease screening. *Journal of the American Society of Echocardiography*. 2015;28(8):981–988. https://doi.org/10.1016/j.echo.2015.03.012

24. Jacobs AK, Kushner FG, Ettinger SM, Guyton RA, Anderson JL, Ohman EM, et al. ACCF/AHA clinical practice guideline methodology summit report: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. *Journal of the American College of Cardiology*. 2013;61(2):213–265. https://doi.org/10.1016/j.jacc.2012.09.025

25. Saxena A, Zuhlke L, Wilson N. Echocardiographic screening for rheumatic heart disease: issues for the cardiology community. *Global Heart*. 2013;8(3):197–202. https://doi.org/10.1016/j.gheart.2013.08.004

26. Bradley-Hewitt T, Dantin A, Ploutz M, Aliku T, Lwabi P, Sable C, et al. The impact of echocardiographic screening for rheumatic heart disease on patient quality of life. *Journal of Pediatrics*. 2016;175:123–129. https://doi.org/10.1016/j.jpeds.2016.04.087

27. Gurney J, Chong A, Culliford-Semmens N, Tilton E, Wilson NJ, Sarfati D. The benefits and harms of rheumatic heart disease screening from the perspective of the screened population. *International Journal of Cardiology*. 2016;221:734–740. https://doi.org/10.1016/j.ijcard.2016.07.025

28. Wark EK, Hodder YC, Woods CE, Maguire GP. Patient and healthcare impact of a pilot rheumatic heart disease screening program. *Journal of Paediatrics and Child Health*. 2013;49(4):297–302. https://doi.org/10.1111/jpc.12154

29. Scheel A, Beaton A, Okello E, Longenecker CT, Otim IO, Lwabi P, et al. The impact of a peer support group for children with rheumatic heart disease in Uganda. *Patient Education and Counseling*. 2018;101(1):119–123. https://doi.org/10.1016/j.pec.2017.07.006

30. Dennison A, Anderson A, Brown R, Ikiua M, Philipson-Puna T, Chan Mow F, et al., editors. Te Maatai manawa a whānau: family health screening study. World Congress on Rheumatic Heart Disease: Abu Dhabi; 2023.

31. Health New Zealand | Te Whatu Ora. HISO 10001:2017 ethnicity data protocols. Wellington: Health New Zealand | Te Whatu Ora; 2024. https://www.tewhatuora.govt.nz/health-services-and-programmes/digital-health/data-and-digital-standards/approved-standards/identity-standards (Accessed February 12 2025).

32. World Health Organization. Task shifting: global recommendations and guidelines. Geneva: World Health Organization; 2007. https://www.unaids.org/sites/default/files/media_asset/ttr_taskshifting_en_0.pdf (Accessed February 18 2025).

33. Francis JR, Fairhurst H, Yan J, Fernandes Monteiro A, Lee A-M, Maurays J, et al. Abbreviated echocardiographic screening for rheumatic heart disease by non-experts with and without offsite expert review: a diagnostic accuracy study. *Journal of the American Society of Echocardiography*. 2023;36(7):733–745. https://doi.org/10.1016/j.echo.2023.02.007

34. Brown K, Roshanitabrizi P, Rwebembera J, Okello E, Beaton A, Linguraru MG, et al. Using artificial intelligence for rheumatic heart disease detection by echocardiography: focus on mitral regurgitation. *Journal of the American Heart Association*. 2024;13(2):e031257. https://doi.org/10.1161/jaha.123.031257

35. Culliford-Semmens N, Tilton E, Wilson N, Stirling J, Doughty R, Gentles T, et al. Echocardiography for latent rheumatic heart disease in first degree relatives of children with acute rheumatic fever: implications for active case finding in family members. *EClinicalMedicine*. 2021;37:100935. https://doi.org/10.1016/j.eclinm.2021.100935

36. Aliku T, Sable C, Scheel A, Tompsett A, Lwabi P, Okello E, et al. Targeted echocardiographic screening for latent rheumatic heart disease in northern Uganda: evaluating familial risk following identification of an index case. *PLoS Neglected Tropical Diseases*. 2016;10(6):e0004727. https://doi.org/10.1371/journal.pntd.0004727

37. Baker MG, Gurney J, Moreland NJ, Bennett J, Oliver J, Williamson DA, et al. Risk factors for acute rheumatic fever: a case-control study. *The Lancet Regional Health — Western Pacific*. 2022;26:100508. https://doi.org/10.1016/j.lanwpc.2022.100508

38. Nascimento BR, Sable C, Nunes MCP, Oliveira KKB, Franco J, Barbosa MM, et al. Echocardiographic screening of pregnant women by non-physicians with remote interpretation in primary care. *Family Practice*. 2021;38(3):225–230. https://doi.org/10.1093/fampra/cmaa115

39. Otto H, Saether SG, Banteyrga L, Haugen BO, Skjaerpe T. High prevalence of subclinical rheumatic heart disease in pregnant women in a developing country: an echocardiographic study. *Echocardiography*. 2011;28(10):1049–1053. https://doi.org/10.1111/j.1540-8175.2011.01520.x

40. Rokovunisei M, Sakumeni K, Wheeler M, Matanaicake S, Lee R, Kailawadoko J, editors. Fiji antenatal echocardiography screening (FANS) for rheumatic heart disease by trained non-expert health personnel. Abu Dhabi: World Congress on Rheumatic Heart Disease; 2023.

41. Seitler S, Ahmad M, Ahuja SAC, Ahmed MT, Stevenson A, Schreiber TR, et al. Routine antenatal echocardiography in high-prevalence areas of rheumatic heart disease: a WHO-guideline systematic review. *Global Heart*. 2024;19(1):39. https://doi.org/10.5334/gh.1318

42. Sullivan EA, Vaughan G, Li Z, Peek MJ, Carapetis JR, Walsh W, et al. The high prevalence and impact of rheumatic heart disease in pregnancy in First Nations populations in a high-income setting: a prospective cohort study. *BJOG: An International Journal of Obstetrics and Gynaecology*. 2020;127(1):47–56. https://doi.org/10.1111/1471-0528.15938

15

Primary Prevention

In this chapter

"Yeah [the doctor] just said I had the flu, sent me home on some different drugs … But he [doctor] never took a throat swab".

Mother of rangatahi

Key changes

This is a new chapter in the guidelines. In 2009, the primary prevention of acute rheumatic fever was a stand-alone guideline.[1]

Key points

Key messages for primary prevention in Aotearoa:

- Primary prevention of acute rheumatic fever (ARF) is currently based on the effective treatment of Strep A sore throats in susceptible individuals and populations (see **Chapter 5: Primary Prevention of Acute Rheumatic Fever: Sore Throat Diagnosis and Management**).
- Primary prevention also aims to raise awareness of getting sore throats checked and supports people to access treatment for them.
- Multiple barriers currently prevent access to sore throat diagnosis and management, especially for whānau in underserved urban and rural areas. Diagnosing and managing sore throats in schools, pharmacies, and other community settings makes ARF prevention more accessible to whānau.
- Communication and health promotion messages about ARF prevention should be informed by whānau to ensure they are culturally responsive and not stigmatising.
- Healthcare workers should implement best clinical practices (see **Chapter 5: Primary Prevention of Acute Rheumatic Fever: Sore Throat Diagnosis and Management**), along with cultural safety and anti-racism practices (see **Chapter 1: Cultural Responsiveness**).
- The development of a Strep A vaccine has the potential to deliver the most effective primary prevention of ARF.

Introduction

Primary prevention for ARF aims to diagnose and treat people with Strep A infection to prevent the immune response to Strep A that causes ARF in some people. For people living in communities with a high incidence of ARF, antibiotic treatment of Strep A sore throat is recommended to prevent ARF and subsequent rheumatic heart disease (RHD). Primary prevention is different from primordial prevention, which focuses on higher-level influences of the risk factors themselves, such as the effects of poverty and crowding on Strep A transmission within populations.

Between the 1950s and 1970s, randomised controlled trials (RCT) and quasi-randomised studies showed that intramuscular (IM) penicillin treatment for Strep A sore throat reduced the risk of ARF by more than two-thirds compared to placebo.[2-7] However, a landmark RCT of school-based primary prevention of ARF in Aotearoa using oral penicillin did not prove to be statistically effective in reducing ARF.[8]

Studies in Australia and Aotearoa demonstrate an epidemiological association between Strep A skin infections and an increased risk of ARF, but a causal link remains unproven.[9-14] It is also not known whether treating Strep A skin infections with antibiotics will reduce an individual's risk of ARF.

Primary prevention of acute rheumatic fever internationally

As noted in **Chapter 3: Strep A Infection, Acute Rheumatic Fever and Rheumatic Heart Disease: Risk Factors, Social Determinants of Health and Primordial Prevention**, several countries between the 1950s and 1990s, have reported a link between significantly improved access to healthcare, effective penicillin treatment for sore throats and reduced incidence of ARF at a population level. Such associations have been found in Baltimore in the United States,[15] Costa Rica,[16] Cuba,[17, 18] the Caribbean[16] and the Americas.[19] Significant socioeconomic development also occurred in these settings over the same period, with improvements in income, employment, housing, and education.[19, 20]

School programmes to reduce the incidence of Strep A sore throat and subsequent ARF in tamariki were implemented between the late 1950s and 1970s in some high-incidence areas within the United States. Observational studies ('before and after' interventions of programmes offering throat swabbing, culture, and treatment) were conducted in Colorado, Wyoming, Hawai'i, Arizona, and Alaska, mostly in rural areas or on reservations for Indigenous peoples. Some showed a positive impact in reducing Strep A prevalence, incidence, and ARF rates.[1, 4, 21-26]

Overall, international historical experience suggests that a comprehensive approach to primary prevention can reduce the incidence of ARF. Rates of ARF were falling substantially in Europe and the United States before the advent of penicillin. The long-term impact of positive changes in the broader social determinants of health (especially poverty reduction and reductions in household crowding)[20-25] on Strep A incidence and ARF may have been underestimated in the biomedical literature. Conversely, the ongoing influence of important factors such as colonisation and racism that continue to drive inequities in these determinants and outcomes may not have been fully recognised.[27] (See **Figure 3.2** TKHM-modified Williams & Mohammed model for explaining Indigenous/ethnic determinants of health about impacts of these determinants).

Primary prevention of acute rheumatic fever in Aotearoa

Prevention prior to 2000

In Aotearoa, rates of ARF declined nationally from the 1950s.[28] High rates persisted in mainly rural Māori communities between the 1970s and 2010, for example, in Wairoa and Te Tai Rāwhiti/East Coast,[29, 30] Te Tai Tokerau/Northland and Bay of Plenty.[31] At this time, primary prevention of ARF was not a political or health sector priority.

Since the 1980s, the focus on antimicrobial stewardship has increased in high-income countries, including Aotearoa. The use of antibiotics for sore throats was generally discouraged. A 1997 review for the Ministry of Health suggested there was no evidence that treating Strep A sore throat in general practice impacted the incidence of ARF in Aotearoa.[32] However, this review failed to recognise the increasing rates in ARF for Māori and Pacific peoples during the 1980s and 1990s, while the rates for non-Māori, non-Pacific groups decreased.[9, 33-35] Rates of ARF continued to rise for Māori and Pacific peoples during the 1990s and 2000s. General practitioners (GP) have a key role in the management of sore throats to prevent ARF in Aotearoa.[32] It has been recognised that systemic racism, inequitable GP access, and often the inadequate treatment of Strep A sore throat in general practice contributed to these increasing ARF inequities.[31-33]

Establishment of school-based primary prevention in Aotearoa

In the early 1990s, Professor Diana (Dinny) Lennon, Henare Mason, and colleagues hypothesised that a school-based study to treat Strep A sore throat could reduce ARF in South Auckland, which at the time had the highest rates of ARF nationally.[36]

Between 1998 and 2001, they conducted a large clinical trial of 22,000 school-aged tamariki in South Auckland. The trial, randomised by school, offered nurse-led, school-based sore throat services.[8] However, a subsequent meta-analysis by Lennon and colleagues suggested that implementing sore throat programmes in schools might reduce the incidence of ARF.[37]

The South Auckland school-based trial inspired other smaller primary prevention initiatives in high ARF incidence areas of the North Island of Aotearoa, for example, in the Whangaroa community in Te Tai Tokerau in 2002,[38] and in Ōpōtiki, Kawerau, and Murupara in Eastern Bay of Plenty from 2009–2011.[39]

The Rheumatic Fever Prevention Programme (RFPP) 2012–2017 in Aotearoa

Following successful advocacy by Hauora Māori and health advocates, and with the leadership of Te Pāti Māori and co-leader Dame Tariana Turia, the coalition government

(elected in 2011) invested approximately $65 million in the Rheumatic Fever Prevention Programme (RFPP) over five years (2012–2017). Additional investment and rollout beyond 2017 were provided at the district health board (DHB) level.

In 2012, the government set a Better Public Services target to reduce ARF by two-thirds by 2017 (from 4.0 to 1.4 cases per 100,000 population).[40] The three main strategies of the RFPP were to:

- Increase awareness of ARF and its prevention.
- Improve access to timely treatment for Strep A sore throat in priority communities.
- Reduce household crowding to reduce transmission of Strep A in households.

The RFPP initially funded eight high-incidence DHBs in the North Island. They later expanded funding to 11 DHBs, 10 of which implemented school-based programmes. 'High incidence' meant DHBs had:

- More than 1.5 first-episode ARF hospitalisations per 100,000 population over a three-year average baseline (2009/10–2011/12).
- A three-year average of four or more cases in a year.

The health districts (formerly DHB areas) with high incidences of ARF involved were:

- **Northern region** — Northland, Counties Manukau, Waitematā and Auckland.
- **Midland region** — Waikato, Bay of Plenty, Lakes, Hawkes Bay and Te Tai Rāwhiti.
- **Central region** — Capital and Coast, Hutt Valley.

School programmes were implemented incrementally between 2012 and 2014. Nearly all programmes were in decile 1–3 schools with high proportions of Māori tamariki, Pacific tamaiti, or both.[41] By 2014, over 250 schools and about 54,000 tamariki in high-risk communities were covered by a sore throat service. Consent rates and participation were high.[42, 43] Qualitative evaluations found positive whānau support.[44, 45]

Throat swabbing rates increased approximately 19-fold after the RFPP was implemented. They rose from 62 swabs per 1,000 tamariki aged 5–14 years in 2010, to 1,177 swabs per 1,000 tamariki at peak coverage in 2014. About 60% of this is related to swabs taken at schools.[41] Other methods to improve access to throat swabbing were introduced later (2014–2015), including walk-in and rapid-response clinics in primary care, as well as pharmacies.[46]

The RFPP provided resources for national and local communication and health promotion.[47] It also strengthened referral pathways, first to existing housing insulation programmes, and subsequently to the Healthy Homes Initiative (HHI),[48] which was implemented from 2013–2023[41, 49, 50] The HHI aimed to provide warm, dry and healthy housing to low-income whānau with tamariki at risk of ARF. Interventions included insulation, repairs, ventilation, bedding, referral for social housing, and housing support.

Crude rates of ARF decreased nationally from 2011 to 2016. There have been several evaluations of the RFPP's initial years. In 2016, the Centre for Public Impact assessed the RFPP in a public policy case study. The Centre rated the programme as 'strong' in nearly all aspects, including:

- Legitimacy (political commitment and public engagement).
- Clarity of objectives.
- Implementation management.[51]

After two years of the school programme implementation in South Auckland (2012–2014), Lennon's evaluation of this initiative reported that ARF cases had decreased by 58%, further supporting the RFPP's actions.[50]

In contrast, the 2015 interim quantitative evaluation of the sore throat management component of the RFPP, which assessed the effectiveness of school-based services, found no statistically significant difference.[42] Overall, the 10 DHBs implementing school-based services experienced a 17% reduction in ARF cases (95%; CI: -17– 42%; not statistically significant). Counties Manukau DHB saw a 31% reduction in ARF cases (95%; CI: 13– 58%; not statistically significant).

The interim RFPP evaluation report involved a 'before and after analysis' of ARF notifications for tamariki and rangatahi aged 5–12 years attending decile 1–3 schools with a school-based sore throat management service by the end of 2014. It compared ARF cases not exposed to the school-based service to ARF cases exposed.[42] The evaluators noted that variable implementation prevented a full assessment:

- Variations in the frequency of swabbing.
- Variable coverage (20–100%).
- Variable inclusion of case-finding.
- Variable management of skin infections.
- Lack of an evaluation component in programme design.

The intensive model implemented in Counties Manukau DHB was not affordable in some districts due to the higher number of schools required to cover the same proportion of high-risk children.

The final 2017 evaluation of the school-based sore throat programmes (funded as part of the RFPP) used a retrospective cohort design. The evaluation estimated that the national ARF incidence rate declined by 28%. The baseline (2009–2011) of 4.0 per 100,000 (95%; CI: 3.5–4.6) had decreased to 2.9 per 100,000 by 2016 (95%; CI: 2.4–3.4, $p<0.01$).

The school-based programme's overall effectiveness was estimated at 23% (95%; CI: -6%–44%; rate ratio (RR) 0.77, 95%; CI: 0.56–1.06).[41] Results were more compelling in Counties Manukau, where about one-third of Aotearoa's newly diagnosed ARF patients lived. High coverage of high-risk tamariki (84%), active case-finding and management of skin infections, and Strep A sore throat were associated with a 46% reduction in ARF (95%; CI: 16%–66%). The baseline ARF rate was 87.1 per 100,000 for tamariki aged 5–12 years.[41]

In contrast, positive results were reported from school-based programmes outside Auckland. For instance, an earlier review in Hawkes Bay of the 2015 'Say Ahh' school programme found important benefits in improving equity of access to throat swabbing. The review noted a significant decline in ARF, with average annual incidence rates 66% lower in the four years after the programme began than in the four years before.[52]

Walsh et al.[53] argued that in high-incidence areas of Bay of Plenty, the rate of ARF per student year among Māori tamariki in school-based programmes declined by up to 60% for cohorts between 2011 and 2018 compared to those from 2000 to 2010. In the same period, ARF rates increased in cohorts with only access to sore throat management in general practice.[53, 54] Ascertainment bias and more complete ARF notification data and hospital coding between 2011 and 2018,[55, 56] were potential reasons for the lack of effect estimated in the RFPP final evaluation. Using more appropriate denominator populations was also proposed (using Māori and Pacific school-year cohorts rather than all 5–15-year-olds or decile 1–3 school rolls).[53, 56]

A formative qualitative review of RFPP-funded 'rapid-response' sore throat services in primary care (2014–2015) described both positive and negative experiences of whānau. Experiences were positive in pharmacy, nurse-led, and Māori and Pacific health services. However, whānau experienced poor communication about the existence of sore throat clinics in high-risk communities, long wait times in general practice, and judgemental, culturally unsafe services.[46] Some areas experienced apparent 'capture' by low-risk populations (personal communication, 2023, N. Springford, Nurse Coordinator Rheumatic Fever, Northland DHB; and Personal communication, 2023, L. Hall, Pharmacist, Bay of Plenty). Pharmacy models were not quantitatively evaluated at a national level.

The Healthy Homes Initiative

Advocating for housing improvements for whānau at risk of ARF or affected by it and referring whānau to housing initiatives were integral to the RFPP. These actions were implemented variably. In some places, the HHI built on existing housing or subsidised insulation programmes. The programme's scope was expanded in 2016 to reach low-income whānau with tamariki under the age of 5 and pregnant people/women. It was expanded again in July 2022 to become nationwide.[48]

The 2024 five-year outcomes evaluation of the HHI demonstrated positive impacts for whānau[57], showing a 18.6% reduction in general hospitalisations for referred tamariki and their wider whānau. Additionally, when people were hospitalised, hospitalisations were shorter. A small but statistically significant reduction in days off school for medical reasons also occurred.[57]

"So if you turn the heaters off, go back to frost. If I don't turn the heaters off, I can't afford the power. I have to turn it off so ultimately we had all the kids in one room you know because it's easier to heat and then I'm worried, coz I'm worried about how I'm going to pay it. That's the biggest factor is [sic] these homes aren't insulated."

Whānau with lived experience

Community awareness of ARF

Increasing community and health worker awareness of the links between Strep A sore throat and ARF and treating high-risk people is important in primary prevention. The RFPP's communication campaigns reached a high proportion of the target audience.[47, 58] However, the messaging has been criticised by some groups as 'responsibilising' individuals and communities with the highest rates of ARF (blaming people for their illness), potentially creating further 'stigma, internalised blame, and hypervigilance'.[59]

In summary, the RFPP implementation from 2011–2017 was associated with a significant decline in ARF in Counties Manukau, where a large proportion of whānau with ARF live. Multiple factors were likely responsible for the decline during the RFPP.[41] School-based programmes offer tamariki access to primary healthcare without the barriers of cost and transport and parents having to take time off work. School programmes increased throat swabbing significantly and the tamariki most at risk were appropriately included.[42] It is important to record that throat swabbing also increased in low-risk tamariki at a significant cost to the RFPP. For example, in the Northland school-based and pharmacy sore throat programme, almost half of those swabbed were at low risk of developing ARF (personal communication, 2023, C. Jackson, Clinical Advisor Rheumatic Fever Care Coordination System | Public Health Medicine Specialist, Health New Zealand | Te Whatu Ora).

Overall, the RFPP achieved real progress in political, health worker, community, and whānau awareness of ARF. It improved access to sore throat and skin management for tamariki at high risk of ARF and developed kaimahi capacity in school programmes.[41, 58, 60] The RFPP contributed to advocacy on housing, improved housing quality, and reduced household crowding — key risk factors strongly associated with Strep A and ARF.[9, 61]

Several complexities surrounded the evaluation and impact of the RFPP. A standardised approach was not feasible, resulting in variable implementation across Aotearoa. Sore throat management was not standardised, with different frequencies of swabbing, coverage, and levels of access in different districts. Tight timeframes limited the ability of Māori and Pacific service providers to build kaimahi capacity and expand programmes.[44]

Primary prevention of ARF between 2018 and 2024 in Aotearoa

At the end of the RFPP in 2017, funding for ARF primary prevention was scaled back. School programmes in many areas reduced coverage, and some school programmes stopped as these services were costly and there was a lack of evidence of sustained effectiveness. The exceptions were in South Auckland and selected schools and kura in Bay of Plenty, Lakes,[62] and Hawkes Bay DHBs. Other high-incidence areas chose general practice and pharmacies as ways through which to fund sore throat management.[63] The cost-effectiveness evaluation of the RFPP concluded that primary prevention was unlikely to be cost-effective outside of Counties Manukau, where the high-risk population is less dispersed, allowing easier targeting of those at the highest risk.

In a series of workshops in 2018, stakeholders from across the health system identified the need for community-led solutions. As a result, the 2019 government budget allocated a further $12 million over four years for DHBs in the Auckland region to pilot 'high impact, short-term' approaches. These included co-design initiatives with Māori, Samoan, and Tongan communities in Auckland. In 2020, the government announced further funding for the HHI,[64] which was designed to expand the HHI.[65] From 2022, the expectation was that ARF prevention services would be 'embedded into ongoing delivery of services.'[40]

The Vote Health budget in 2023/24 was allocated to:

- The Rheumatic Fever Care Coordination System RFCCS (national ARF register).
- Support for culturally informed approaches to health and wellbeing.
- The updating of the national clinical RF and RHD guidelines.
- Raising awareness of ARF with Māori and Pacific whānau and communities.
- The South Auckland school-based programme to continue.
- Pilot projects (echo screening and oral health support).

Following declines between 2011 and 2015, annual crude rates of ARF initial hospitalisations slightly increased between 2016 and 2018.

The COVID-19 pandemic and lockdowns significantly reduced throat swabbing, particularly in school programmes. Crude rates of ARF fell during the pandemic, reaching a low of 1.6 per 100,000 in 2022, mainly associated with a large decrease in cases (over two-thirds) in Pacific peoples in Auckland in 2021–2022 (see **Chapter 4: Epidemiology of Strep A Infections, Acute Rheumatic Fever and Rheumatic Heart Disease**).[66] Pandemic public health measures, particularly prolonged school closures and restrictions on community gatherings in Auckland between 2020 and 2022, are likely to have reduced Strep A transmission and therefore ARF incidence. Reduced access to healthcare (including the temporary halt of all throat swabbing at schools during lockdowns) and a decline in funding and ascertaining cases of ARF may have also impacted ARF rates during and after the pandemic.[67-72]

Current status of primary prevention in Aotearoa

By 2023, ARF rates returned to pre-pandemic levels in the Auckland region and nationally.[66, 71, 73] Overall, a sustained reduction in ARF has not occurred with the introduction of primary prevention measures, although the impacts of COVID-19 have made assessment difficult from 2019–2023. Many of the structural inequities driving ARF in Aotearoa have worsened in recent years, including:

- Material hardship and child poverty
- Continued pressures on housing and household crowding
- High demands on the health sector, with worsening access to primary healthcare that, disproportionately impacts people in underserved urban and rural areas.

School-based programmes continue in Counties Manukau (Mana Kidz) and a smaller number of schools in Bay of Plenty, Lakes, and Hawke's Bay health districts. Pharmacy access to throat-swabbing is also available in Bay of Plenty, Lakes, Te Tai Tokerau, Waikato, Auckland and Hutt Valley.

A report by the Office of the Prime Minister's Chief Science Advisor (OPMCSA) reflects how primary prevention alone cannot control ARF:

> ARF remains a complex disease that will not be eliminated until poverty, household crowding, racism, and barriers to accessing health services are tackled. Inequities need to be addressed....[74]

The ultimate goal of primary prevention is the development and equitable uptake of an effective Strep A vaccine. In the interim, Māori and Pacific peoples (with a focus on those aged 3–35 years) presenting with symptomatic Strep A sore throats should receive antibiotic treatment.

Strep A vaccine development

Developing a Strep A vaccination is the ultimate goal for primary prevention. Developing a Strep A vaccine has been a challenge for scientists for decades, and to date, no Strep A vaccine has been available internationally. Currently, there are multiple vaccine candidates in both pre-clinical and clinical development based on M proteins and those involving Strep A toxins. Vaccine candidates with minimal likelihood of inducing autoreactive antibodies are essential to ensure safety and lasting immunity against Strep A. Internationally, a dedicated global network has been developed to support Strep A surveillance and vaccine trials. There has been considerable engagement with industry, governments, scientists, and funders to coordinate vaccine development.[75-77]

In 2021, the Government of Aotearoa invested funding to support the development of a Strep A vaccine to address ARF and RHD locally. The initiative — Rapua te mea ngaro ka tau — has a number of stages to complete before the vaccine will be ready for clinical trials.

References

1. Rheumatic Fever Guidelines Writing Group, Heart Foundation. New Zealand guidelines for rheumatic fever: 3. Proposed rheumatic fever primary prevention programme. Auckland: National Heart Foundation of New Zealand; 2009. https://assets.heartfoundation.org.nz/documents/shop/heart-healthcare/non-stock-resources/primary-prevention-rheumatic-fever-guideline.pdf?mtime=1667526707?1740182280&_gl=1*ni1pim*_gcl_au*OTYxMzcyMjQ2LjE3NDAxODIyODQ. (Accessed February 17 2025).

2. Spinks A, Glasziou PP, Del Mar CB. Antibiotics for treatment of sore throat in children and adults. *Cochrane Database of Systematic Reviews*. 2021;12(12):CD000023. https://doi.org/10.1002/14651858.CD000023.pub5

3. Denny FW, Wannamaker LW, Brink WR, Rammelkamp CH, Jr., Custer EA. Prevention of rheumatic fever; treatment of the preceding streptococcic infection. *Journal of the American Medical Association*. 1950;143(2):151–153. https://doi.org/10.1001/jama.1950.02910370001001

4. Kerdemelidis M, Lennon DR, Arroll B, Peat B, Jarman J. The primary prevention of rheumatic fever. *Journal of Paediatrics and Child Health*. 2010;46(9):534–548. https://doi.org/10.1111/j.1440-1754.2010.01854.x

5. Wessels MR. Clinical practice. Streptococcal pharyngitis. *New England Journal of Medicine*. 2011;364(7):648–655. https://doi.org/10.1056/NEJMcp1009126

6. Robertson KA, Volmink JA, Mayosi BM. Antibiotics for the primary prevention of acute rheumatic fever: a meta-analysis. *BMC Cardiovascular Disorders*. 2005;5(1):11. https://doi.org/10.1186/1471-2261-5-11

7. Jones TD, Mote JR. The clinical importance of infection of the respiratory tract in rheumatic fever. *Journal of the American Medical Association*. 1939;113(10). https://doi.org/10.1001/jama.1939.02800350008003

8. Lennon D, Stewart J, Farrell E, Palmer A, Mason H. School-based prevention of acute rheumatic fever: a group randomized trial in New Zealand. *Pediatric Infectious Disease Journal*. 2009;28(9):787–794. https://doi.org/10.1097/INF.0b013e3181a282be

9. Bennett J, Moreland NJ, Zhang J, Crane J, Sika-Paotonu D, Carapetis J, et al. Risk factors for group A streptococcal pharyngitis and skin infections: a case control study. *The Lancet Regional Health — Western Pacific*. 2022;26:100507. https://doi.org/10.1016/j.lanwpc.2022.100507

10. Williamson DA, Smeesters PR, Steer AC, Steemson JD, Ng AC, Proft T, et al. M-protein analysis of streptococcus pyogenes isolates associated with acute rheumatic fever in New Zealand. *Journal of Clinical Microbiology*. 2015;53(11):3618–3620. https://doi.org/10.1128/JCM.02129-15

11. Thomas S, Bennett J, Jack S, Oliver J, Purdie G, Upton A, et al. Descriptive analysis of group A streptococcus in skin swabs and acute rheumatic fever, Auckland, New Zealand, 2010–2016. *The Lancet Regional Health — Western Pacific*. 2021;8:100101. https://doi.org/10.1016/j.lanwpc.2021.100101

12. Oliver J, Bennett J, Thomas S, Zhang J, Pierse N, Moreland NJ, et al. Preceding group A streptococcus skin and throat infections are individually associated with acute rheumatic fever: evidence from New Zealand. *BMJ Global Health*. 2021;6(12). https://doi.org/10.1136/bmjgh-2021-007038

13. Parks T, Smeesters PR, Steer AC. Streptococcal skin infection and rheumatic heart disease. *Current Opinion in Infectious Diseases*. 2012;25(2):145–153. https://doi.org/10.1097/QCO.0b013e3283511d27

14. McDonald MI, Towers RJ, Andrews RM, Benger N, Currie BJ, Carapetis JR. Low rates of streptococcal pharyngitis and high rates of pyoderma in Australian aboriginal communities where acute rheumatic fever is hyperendemic. *Clinical Infectious Diseases*. 2006;43(6):683–689. https://doi.org/10.1086/506938

15. Gordis L. Effectiveness of comprehensive-care programs in preventing rheumatic fever. *New England Journal of Medicine*. 1973;289(7):331–335. https://doi.org/10.1056/nejm197308162890701

16. Bach JF, Chalons S, Forier E, Elana G, Jouanelle J, Kayemba S, et al. 10-year educational programme aimed at rheumatic fever in two French Caribbean islands. *Lancet*. 1996;347(9002):644–648. https://doi.org/10.1016/s0140-6736(96)91202-7

17. Nordet P, Lopez R, Dueñas A, Sarmiento L. Prevention and control of rheumatic fever and rheumatic heart disease: the Cuban experience (1986-1996-2002). *Cardiovascular Journal of Africa*. 2008;19(3):135–140.

18. Watkins DA, Mvundura M, Nordet P, Mayosi BM. A cost-effectiveness analysis of a program to control rheumatic fever and rheumatic heart disease in Pinar del Rio, Cuba. *PloS One*. 2015;10(3):e0121363. https://doi.org/10.1371/journal.pone.0121363

19. Ordunez P, Martinez R, Soliz P, Giraldo G, Mujica OJ, Nordet P. Rheumatic heart disease burden, trends, and inequalities in the Americas, 1990-2017: a population-based study. *Lancet Global Health*. 2019;7(10):e1388–e1397. https://doi.org/10.1016/s2214-109x(19)30360-2

20. Gordis L. The virtual disappearance of rheumatic fever in the United States: lessons in the rise and fall of disease. T. Duckett Jones memorial lecture. *Circulation*. 1985;72(6):1155–1162. https://doi.org/10.1161/01.cir.72.6.1155

21. Zimmerman RA, Biggs BA, Bolin RA, Wilson E, Mathews JH, Cropp CB, et al. An effective program for reducing group A streptococcal prevalence. *Pediatrics*. 1971;48(4):566–572.

22. Phibbs B, Lundin SR, Watson WB, Corbett JJ. Experience of a Wyoming county streptococcal control project. *Western Journal of Medicine*. 1988;148(5):546–550.

23. Phibbs B, Atha M, Enos E, Frank C, Murphy M, Ortega M, et al., editors. How an American Indian tribe controlled the streptococcus. World Health Forum; 1982.

24. Phibbs B, Becker D, Lowe CR, Holmes R, Fowler R, Scott OK, et al. The Casper project-an enforced mass-culture streptococcic control program. *Journal of the American Medical Association*. 1958;166(10):1113–1119. https://doi.org/10.1001/jama.1958.02990100001001

25. Phibbs B, Taylor J, Zimmerman RA. A community-wide streptococcal control project. The Natrona County Primary Prevention Program, Casper, Wyo. *JAMA*. 1970;214(11):2018–2024. https://doi.org/10.1001/jama.214.11.2018

26. Brant LJ, Bender TR, Bross DS. Evaluation of an Alaskan streptococcal control program: importance of the program's intensity and duration. *Preventive Medicine*. 1986;15(6):632–642. https://doi.org/10.1016/0091-7435(86)90068-x

27. Curtis E, Jones R, Willing E, Anderson A, Paine S-J, Herbert S, et al. Indigenous adaptation of a model for understanding the determinants of ethnic health inequities. *Discover Social Science and Health*. 2023;3(1). https://doi.org/10.1007/s44155-023-00040-6

28. Wabitsch KR, Prior IA, Stanley DG, Pearce N. New Zealand trends in acute rheumatic fever and chronic rheumatic heart disease 1971–1981. *New Zealand Medical Journal*. 1984;97(763):594–597.

29. Frankish JD. Rheumatic fever on the East Coast, North Island. *New Zealand Medical Journal*. 1974;80(520):48–51.

30. Frankish JD, Stanhope JM, Martin DR, Clarkson PM, Leslie PN, Langley RB. Rheumatic fever and streptococci: the Wairoa College study. *New Zealand Medical Journal*. 1978;87(604):33–38.

31. Prior I. A health survey in a rural Maori community, with particular emphasis on the cardiovascular, nutritional and metabolic findings. *New Zealand Medical Journal*. 1962;61:333–348.

32. Durham J, Kljakovic M. Primary prevention of rheumatic fever: is there a role for general practice. Wellington: Ministry of Health; 1997.

33. Norris P, Horsburgh S, Keown S, Arroll B, Lovelock K, Cumming J, et al. Too much and too little? Prevalence and extent of antibiotic use in a New Zealand region. *Journal of Antimicrobial Chemotherapy*. 2011;66(8):1921–1926. https://doi.org/10.1093/jac/dkr194

34. Harris R, Tobias M, Jeffreys M, Waldegrave K, Karlsen S, Nazroo J. Racism and health: the relationship between experience of racial discrimination and health in New Zealand. *Social Science and Medicine*. 2006;63(6):1428–1441. https://doi.org/10.1016/j.socscimed.2006.04.009

35. Shetty A, Mills C, Eggleton K. Primary care management of group A streptococcal pharyngitis in Northland. *Journal of Primary Health Care*. 2014;6(3). https://doi.org/10.1071/hc14189

36. Counties Manukau Health. Rheumatic fever prevention plan 2013–2017. 2013.

37. Lennon D, Kerdemelidis M, Arroll B. Meta-analysis of trials of streptococcal throat treatment programs to prevent rheumatic fever. *Pediatric Infectious Disease Journal*. 2009;28(7):e259–264. https://doi.org/10.1097/INF.0b013e3181a8e12a

38. Jarman J. How a community controlled the streptococcus: school-based rheumatic fever primary prevention in New Zealand. Australasian Faculty of Public Health Annual Conference. Cairns; 2008.

39. Malcolm J, Ball S, Beharry J, Seal R, Lowe L, Biddle S, et al. Tautoko rheumatic hearts: to support those with rheumatic hearts, public health needs innovation, collaboration and evaluation. In: Zander A, Came H, editors. Proceedings of the 2013 Public Health Association Conference. Auckland: Public Health Association; 2013. p. 107–112.

40. Health New Zealand | Te Whatu Ora. Reducing rheumatic fever. Health New Zealand | Te Whatu Ora; 2024. https://www.tewhatuora.govt.nz/for-the-health-sector/health-sector-guidance/diseases-and-conditions/rheumatic-fever-guidance/reducing-rheumatic-fever/ (Accessed February 18 2025).

41. Jack SJ, Williamson DA, Galloway Y, Pierse N, Zhang J, Oliver J, et al. Primary prevention of rheumatic fever in the 21st century: evaluation of a national programme. *International Journal of Epidemiology*. 2018;47(5):1585–1593. https://doi.org/10.1093/ije/dyy150

42. Jack S, Williamson D, Galloway Y, Pierse N, Milne R, Mackereth G. Interim evaluation of the sore throat component of the rheumatic fever prevention programme — quantitative findings. Porirua: Institute of Environmental Science and Research Limited; 2015.

43. King J, Moss M, McKegg K. School based primary health care programme evaluation: final report. Auckland: Kinnect Group; 2014.

44. Litmus Limited. Implementation and formative evaluation of the rheumatic fever prevention programme: final report. Wellington: Ministry of Health; 2014.

45. Anderson P, King J, Moss M, Light P, McKee T, Farrell E, et al. Nurse-led school-based clinics for rheumatic fever prevention and skin infection management: evaluation of Mana Kidz programme in Counties Manukau. *New Zealand Medical Journal*. 2016;129(1428):37–46.

46. Litmus Limited. Formative evaluation of sore throat clinics. Wellington: Ministry of Health; 2016.

47. Tu'akoi S, Ofanoa M, Ofanoa S, Lutui H, Heather M, Jansen RM, et al. Addressing rheumatic fever inequities in Aotearoa New Zealand: a scoping review of prevention interventions. *Journal of Primary Health Care*. 2023;15(1):59–66. https://doi.org/10.1071/HC22093

48. Health New Zealand | Te Whatu Ora. Healthy Homes. Health New Zealand | Te Whatu Ora; 2022. https://www.tewhatuora.govt.nz/keeping-well/for-families-and-children/healthy-homes-initiative (Accessed February 18 2025).

49. Dougherty S, Carapetis J, Zühlke L, Wilson N. Acute rheumatic fever and rheumatic heart disease. Amsterdam: Elsevier; 2020.

50. Lennon D, Anderson P, Kerdemilidis M, Farrell E, Crengle Mahi S, Percival T, et al. First presentation acute rheumatic fever is preventable in a community setting: a school-based intervention. *Pediatric Infectious Disease Journal*. 2017;36(12):1113–1118. https://doi.org/10.1097/inf.0000000000001581

51. Centre for Public Impact. New Zealand's rheumatic fever prevention programme (RFPP). New Zealand: Centre for Public Impact; 2016. https://www.centreforpublicimpact.org/case-study/rheumatic-fever-prevention-programme (Accessed February 18 2025).

52. Stevens J. Say ahh rheumatic fever prevention programme evaluation report. Hawke's Bay: Hawke's Bay District Health Board; 2015.

53. Walsh L, Innes-Smith S, Wright J, Michniewicz T, Tozer M, Humby J, et al. School-based streptococcal a sore-throat treatment programs and acute rheumatic fever amongst indigenous Maori: a retrospective cohort study. *Pediatric Infectious Disease Journal*. 2020;39(11):995–1001. https://doi.org/10.1097/INF.0000000000002770

54. Lennon D, Stewart J, Percival T, Anderson P, Crengle S, Malcolm J. Which school primary care model best reduces group A streptococcal burden as rheumatic fever precursor? Has the clinic model in CMDHB and ADHB reduced acute rheumatic fever in clinic schools? Auckland: Health Research Council; 2017.

55. Galloway Y, Jack S, Hambling T. Rheumatic fever in New Zealand: annual report July 2014 to June 2015. Porirua: ESR; 2017.

56. Oliver J, Pierse N, Williamson DA, Baker MG. Estimating the likely true changes in rheumatic fever incidence using two data sources. *Epidemiology and Infection*. 2018;146(2):265–275. https://doi.org/10.1017/S0950268817002734

57. Pierse N, Johnson E, Riggs L, Watson N. Healthy Homes Initiative: five-year outcomes evaluation. Health New Zealand | Te Whatu Ora; 2024. https://www.tewhatuora.govt.nz/publications/healthy-homes-initiative-five-year-outcomes-evaluation (Accessed February 18 2025).

58. Vermillion Peirce P, Akroyd S, Tavila A, Magill K, Goodwin D. Evaluation of the 2015 Rheumatic Fever Awareness Campaign. Wellington: Allen+Clarke; 2015. https://thehub.sia.govt.nz/resources/evaluation-of-the-2015-rheumatic-fever-awareness-campaign.

59. Anderson A, Spray J. Beyond awareness: towards a critically conscious health promotion for rheumatic fever in Aotearoa, New Zealand. *Social Science and Medicine*. 2020;247:112798. https://doi.org/10.1016/j.socscimed.2020.112798

60. Lennon D, Stewart J. An important investment to control acute rheumatic fever needs to run its course. *New Zealand Medical Journal*. 2015;128(1416):6–9.

61. Baker MG, Gurney J, Moreland NJ, Bennett J, Oliver J, Williamson DA, et al. Risk factors for acute rheumatic fever: a case-control study. *The Lancet Regional Health — Western Pacific*. 2022;26:100508. https://doi.org/10.1016/j.lanwpc.2022.100508

62. Toi Te Ora Public Health. Rheumatic fever. Toi Te Ora Public Health; 2023.

63. Northland District Health Board. Rheumatic fever prevention plan: July 2017–June 2019. 2017. https://www.northlanddhb.org.nz/assets/Your-Health/Northland-DHB-Rheumatic-Fever-Plan-2017-2019.pdf (Accessed February 18 2025).

64. Ministry of Health. Reducing the incidence and improving the management of rheumatic fever and rheumatic heart disease amongst Māori and Pacific. Budget 2019: guidance for agencies. https://www.health.govt.nz/system/files/2020-05/10806_-_reducing_the_incidence_improving_the_management_of_rheumatic_fever_redacted.pdf (Accessed February 18 2025).

65. New Zealand Labour Party. Healthy homes and hearts in rheumatic fever initiative. n.d. https://www.labour.org.nz/release-healthy-homes-and-hearts-in-rheumatic-fever-initiative (Accessed February 16 2025).

66. National Public Health Service Te Whatu Ora. Acute rheumatic fever notifications to Auckland Regional Public Health Service 1 April 2023 to 30 June 2023. 2023. https://view.officeapps.live.com/op/view.aspx?src=https%3A%2F%2Fwww.tewhatuora.govt.nz%2Fassets%2FUploads%2F20230420-Rheumatic-Fever-Report-2022-Public.xlsx&wdOrigin=BROWSELINK (Accessed February 11 2025).

67. Hills T, Ritchie S, Thomas M, Duffy E. Non-pharmaceutical interventions targeting COVID-19 were associated with large reductions in community antibiotic dispensing but no increase in severe morbidity from severe common bacterial infections. *New Zealand Medical Journal*. 2021;134(1544):179–182.

68. Duffy E, Thomas M, Hills T, Ritchie S. The impacts of New Zealand's COVID-19 epidemic response on community antibiotic use and hospitalisation for pneumonia, peritonsillar abscess and rheumatic fever. *The Lancet Regional Health — Western Pacific*. 2021;12:100162. https://doi.org/10.1016/j.lanwpc.2021.100162

69. Duncanson M, Wheeler BJ, Jelleyman T, Dalziel SR, McIntyre P. Delayed access to care and late presentations in children during the COVID-19 pandemic New Zealand-wide lockdown: A New Zealand Paediatric Surveillance Unit study. *Journal of Paediatrics and Child Health*. 2021;57(10):1600–1604. https://doi.org/10.1111/jpc.15551

70. Oben G, Duncanson M, Adams J, Satyanand T. State of child health: acute rheumatic fever in Aotearoa New Zealand. *Journal of the Royal Society of New Zealand*. 2023;53(5):631–640. https://doi.org/10.1080/03036758.2022.2113102

71. ESR. Summary of rheumatic fever notifications 1 January–30 June 2023. Porirua: ESR; 2023.

72. Wright K, Dennison A, Mills C, van der werf B. Whāia te Māramatanga: incidence and trends in acute rheumatic fever and rheumatic heart disease, 2000–2022. Auckland: University of Auckland; 2024. (Unpublished).

73. ESR. Monthly notifiable disease surveillance report Dec 2023. 2023. https://www.esr.cri.nz/digital-library/monthly-notifiable-disease-surveillance-report-dec-2023/ (Accessed February 18 2025).

74. Office of the Prime Minister's Chief Science Advisor. Group A streptococcus and acute rheumatic fever in Aotearoa New Zealand: a summary of current knowledge in Aotearoa New Zealand. 2021. https://www.dpmc.govt.nz/sites/default/files/2024-01/PMCSA-21-11-02-V1-OPMCSA-rheumatic-fever-to-upload-on-Nov-19-.pdf (Accessed February 18 2025).

75. Steer AC, Law I, Matatolu L, Beall BW, Carapetis JR. Global emm type distribution of group A streptococci: systematic review and implications for vaccine development. *Lancet Infectious Diseases*. 2009;9(10):611–616. https://doi.org/10.1016/S1473-3099(09)70178-1

76. Fulurija A, Cunningham MW, Korotkova N, Masterson MY, Bansal GP, Baker MG, et al. Research opportunities for the primordial prevention of rheumatic fever and rheumatic heart disease-streptococcal vaccine development: a national heart, lung and blood institute workshop report. *BMJ Global Health*. 2023;8(Suppl 9). https://doi.org/10.1136/bmjgh-2023-013534

77. Dale JB, Walker MJ. Update on group A streptococcal vaccine development. *Current Opinion in Infectious Diseases*. 2020;33(3):244–250. https://doi.org/10.1097/QCO.0000000000000644

Online resources for whānau with Strep A, acute rheumatic fever and rheumatic heart disease

These resources offer a wealth of information in different formats, including text, videos and apps to support whānau in Aotearoa.

Healthify

Strep throat

Rheumatic fever in children

Heart Foundation

Rheumatic fever and rheumatic heart disease — booklet

Rheumatic fever and rheumatic heart disease

We need to talk about heart disease

Heart Kids New Zealand

Takikava Joseph — Heart teen

Acquired heart conditions

Pū Manawa — Rheumatic Fever Network

Resources for whānau

Stop Sore Throats Hurting Hearts

Stop sore throats

Health New Zealand | Te Whatu Ora

Rheumatic fever

Rheumatic fever co-design initiative

KidsHealth New Zealand

For tamariki (children) and whānau:
My Rheumatic Fever & Rheumatic Heart Disease Journey — In Pictures

For rangatahi (youth) and whānau:
The Rheumatic Fever & Rheumatic Heart Disease Journey

Sore throat videos

Rheumatic fever — women and pregnancy