Starting at the age of four years old individuals with FOP should begin seeing a respiratory therapist for pulmonary (or lung) function testing (PFTs). This often begins with a referral from your primary care provider. The results of your initial testing will be critical to serve as a baseline for future test to help identify decreased lung capacity.

Your healthcare provider may also consider information from up-to-date chest x-rays, echocardiograms, and pulse oximetry to understand your current lung health.

PFTs or other specialized testing should be done regularly, in discussion with your doctor. Frequency of re-testing will vary depending on baseline results.

Healthy lungs are free of secretions, able to inhale deeply, and have a strong cough, all traits which are key to recovery from respiratory illnesses.

**AN INTRODUCTION TO FOP LUNG HEALTH**

Having strong respiratory health is crucial for everyone, especially those with FOP. As FOP restrictions increase, the buildup of extra bone and changes in body positioning can restrict the movement of muscles around the lungs and heart. This limits chest expansion and causes additional breathing difficulties. To promote continued lung health and improve the chances of overcoming respiratory illness as FOP progresses, individuals with FOP should review the provided guidelines with their local health care provider. These guidelines offer general recommendations and can be implemented at varying degrees of FOP progression. Especially regarding lung health, being proactive in healthcare management is essential for overall well-being.

**FOP LUNG HEALTH Guidelines**

**START BY ASSESSING YOUR LUNG HEALTH**

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» Your healthcare provider may also consider information from up-to-date chest x-rays, echocardiograms, and pulse oximetry to understand your current lung health.

» PFTs or other specialized testing should be done regularly, in discussion with your doctor. Frequency of re-testing will vary depending on baseline results.

» Healthy lungs are free of secretions, able to inhale deeply, and have a strong cough, all traits which are key to recovery from respiratory illnesses.

**MAINTAINING RESPIRATORY CAPACITY**

» FOP medical experts recommend 15-30 minutes per day of active respiratory activity to maintain lung function. The goal of these activities is to promote deep breathing which strengthens the diaphragm (below the lungs) and the intercostal muscles (between the ribs) to improve lung expansion.

Examples of respiratory activity include:
  • Walking or active mobility*
  • Vigorous singing

  • Swimming/Hydrotherapy: A water safety assessment can be undertaken by a hospital physiotherapist or physical therapist who takes the child into a pool and makes recommendations for safe swimming.

  • Deep breathing/relaxation exercises*

  • Playing woodwind instruments (any musical instrument that relies on deep breathing)

  •Blowing bubbles

  • Using an incentive spirometer* (adults) or a peak flow whistle* (children)

*Make sure to consult with your primary care provider before incorporating new practices into your daily routine.
BEING PROACTIVE ABOUT DECREASED LUNG CAPACITY

Infection precaution and vaccination

» Discuss with your doctor if key vaccines for illnesses such as influenza, covid, and pneumonia are appropriate or available subcutaneously.
   • Refer to the Treatment Guidelines or consult with an FOP medical expert if you decide to get a vaccine.

» Practice infection precautions, especially during flu season such as: handwashing, masking, avoiding high-risk environments, social distancing, close family members getting vaccinated

Collaborating on Care

» It’s important to make sure primary care provider and any additional specialist you see are familiar with FOP and consult with FOP experts and the FOP treatment guidelines for optimal care.

» Discuss medication to thin the mucus, saline nebulizers and inhaled medications with your physician

» Work with a speech language pathologist to learn swallowing techniques which can help prevent aspiration pneumonia.

» Speak with a respiratory therapist about chest physiotherapy and whether it would be helpful for you.

» Explore pulmonary consultation for sleep studies which can be helpful in directing specific respiratory therapies.

Tools to Be Aware of

» Pill cutter/pill crusher for safe swallowing of medications

» Suction device to help clear airway

» Positive Expiratory Pressure (PEP) Device to loosen mucus and improve airway clearance

Consult with your clinician before using any of the above-mentioned tools or interventions before implementing to ensure that it is the right fit for you and that you are educated on how to safely use it.

ADDITIONAL LUNG HEALTH RESOURCES

Access the FOP Treatment Guidelines, watch the Introduction to Lung Health webinar, additional resource videos and access the Ability Toolbox Guidebook to learn more about tools to support pulmonary health at: ifopa.org/lung_health