

## Facts about OI for Medical Professionals

### Take Charge of your Breathing

#### Respiratory Recommendations for Adults who have OI

Respiratory problems can seriously diminish a person's quality of life and are a leading cause of OI related death. Lung connective tissue is altered in all types of OI but the severity of symptoms and the risk for pulmonary problems is increased in people with short stature, abnormal chest shape, kyphoscoliosis, and vertebral or rib fractures. Breathing problems can develop slowly over time. So don't wait until you're seriously short of breath; start paying attention to your breathing today.

- Promptly seek medical care for all respiratory infections.
- Do not smoke and avoid secondhand smoke. Seek a smoking cessation program ASAP if needed.
- Pneumonia vaccine – follow CDC protocols for children and adults.\*
- Flu vaccine (annual).
- Maintain a healthy weight. Avoid obesity.
- Regular cardiopulmonary exercise such as swimming, bicycling, walking.
- Exercise to keep upper body strong.
- Maintain good hydration.
- Pulmonary consultation and pulmonary function tests (PFTs) are needed before any procedure requiring any form of anesthesia -- including spinal anesthesia and conscious sedation.
  - Track PFT changes over time by comparing the results to yourself rather than calculated predicted norms.
    - Get first between age 8 and 20
    - Biannually (or annually if you have symptoms) and after age 20
  - Routine annual oximetry at rest and with exertion.
    - If abnormal, consider arterial blood gas (ABG).
- Echocardiogram and CT scan of the chest as recommended by your doctor.
- Medications can include bronchodilators, inhaled corticosteroids for those with asthma or chronic obstructive pulmonary disease (COPD).

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*This fact sheet was prepared with assistance from Dr. Robert Sandhaus, University of Colorado School of Medicine, Denver, CO, May 2016.*

## FAQs

### **Are people with OI prone to more bronchial infections and pneumonia?**

As far as we can tell, the answer is yes. It's important to seek medical treatment at the first sign of an infection.

### **Do people with OI have a higher incidence of COPD or lung cancer?**

As far as we can tell the incidence of COPD and lung cancer are both more related to smoking than to OI. It is likely however, that a person with OI and short stature may have worse symptoms for a given amount of lung disease than a person in the general population.

### **What should anyone with OI and lung capacity issues do to help improve their situation?**

1. Follow a healthy lifestyle. Maintain a healthy weight, exercise regularly, don't smoke and be moderate when it comes to alcoholic beverages.
2. Follow a schedule of regular pulmonary function testing. The frequency will depend on each person's symptoms.

### **Will exercise increase lung capacity?**

Exercise that encourages a person to take deep breaths and to gradually increase their endurance is good for lung capacity. This can include swimming, walking, and bicycling. Swimming is a reasonable exercise for people with OI, but, as with any exercise, it must be done regularly.

### **Can singing help?**

Maybe -- there are no specific studies about singing and OI, but many therapy programs include activities such as blowing ping pong balls across a table, blowing bubbles in a liquid, playing a harmonica, and singing. The challenge is to find an exercise, keep doing it, and gradually increase effort and endurance.

### **Are there any new drugs that would help lung capacity?**

Drugs are available to treat specific diagnoses such as asthma and chronic obstructive pulmonary disease (COPD). Newer inhaled medications have longer duration of action with many now available as once daily inhalation. There is nothing that is universally recommended for all people who have OI.

### **Are there foods that can decrease inflammation in the lungs?**

The best recommendation is to eat a healthy diet. This can be described as – don't eat too much; eat more plants; and eat a variety of foods. In general, research suggests that being overweight is linked to breathing problems.

### **\*Which type of pneumonia vaccine should people with OI get?**

People with OI are considered to be in the "high risk" group for lung infections as defined by the Centers for Disease Control and Prevention (CDC).

For a summary of the recommendations see the OIF fact sheet **Pneumonia Vaccine Update**.