



Strong respiratory health is important for everyone. This is particularly true for our FOP community since FOP can severely decrease respiratory capacity from the chest deformities and scoliosis.

Maintaining strong respiratory health involves a few things:

1. Infection precautions, particularly during the flu season—Make sure that all FOP patients and their family members wash their hands regularly and use alcohol gel. Avoid places where infection can be easily transmitted. In the event that exposures need to occur, we recommend that you wear a simple surgical mask to decrease the risk of breathing in infected droplets (such as from a sneeze). These masks are not meant to filter out all viruses and infectious bacteria, but will balance the need to decrease exposure to larger droplets with comfortable breathing.

FOP patients who decide to do yearly **flu immunizations** should use the subcutaneous immunization, given by an experienced provider. Even if the FOP patient decides not to get a flu vaccine, all immediate family members should be immunized. In the case where the flu vaccine is not obtained, for whatever reason, having a ready supply of Tamiflu on hand during flu season is a reasonable precaution. At the first signs of the flu – fever or feeling feverish/chills, cough, sore throat, runny or stuffy nose, muscle or body aches, headaches, fatigue (tiredness), vomiting and diarrhea – one should take the first dose of Tamiflu and then immediately call their doctor. **Please discuss with your physician whether flu immunizations and other immunizations such as pneumovax are appropriate for you.**

2. Maintaining respiratory capacity—We recommend 15-30 minutes per day of active respiratory activity. This should not be uncomfortable or cause pain, but is meant to help keep the diaphragm and other respiratory muscles strong and healthy. Activities that we recommend include vigorous vocalizations (for example, singing in a choir or loud continuous vocal activity like singing in the shower).

For some, an **incentive spirometer** can complement or serve as a substitute for vigorous vocalizations. These devices can be used to measure lung capacity; however, this is not the goal right now. Rather, we recommend using them as a way to maintain lung capacity and make sure your lungs are well ventilated.

There are many types of incentive spirometers available and many different strategies for maintaining lung function. The IFOPA is providing two types depending on your age and jaw needs. These are described on page two. Make sure to talk with your physician if you feel you want a different strategy.

To good health,
Your FOP clinicians and the IFOPA



**Whether using the Peak Flow Whistle or Incentive Spirometer,
you should complete these breathing exercises each day.**

PEAK FLOW WHISTLE (for young kids)

For young kids, the IFOPA is providing a Peak Flow Whistle. These whistles will make a sound when air is exhaled quickly through the whistle. **The most important part of using this device is taking the deep breath beforehand – not the actual ability to generate the whistle sound!**

The goal is to improve inspiration (i.e. inhalation), even though a peak flow meter measures expiration (i.e. exhalation).

User Instructions for the Peak Flow Whistle:

For Peak Flow Whistles, please discuss with your pediatrician how to set them (instructions for your clinician are below). Make sure to breathe through your mouth. You will need to have a good seal around the whistle for this to work. Stop if you feel dizzy at any time, and rest.

1. Sit upright or stand
2. Place the whistle in your mouth, and make sure your lips are tightly sealed
3. Slowly inhale as much as you can (this is actually the most important part); hold your breath for about 10 seconds
4. Exhale as quickly as you can through the whistle to generate the sound

Repeat 10 times, with short rest breaks in between. Stop if you feel dizzy at any time, or if you have any tenderness or chest discomfort. Make sure to rest in-between breaths.

Instructions for your clinician on how to set the whistle: The whistle should be set based on the patient's estimated peak flow on a regular day (for example, should just be able to whistle). Standard tables with FEV1 values are not useful in FOP due to the presence of chest deformities; however, prior PFT values can serve as a guide. The goal is to encourage deep breaths to minimize atelectasis (collapse of lung tissue with loss of volume), rather than increasing peak flow. We have found that the whistle serves as an incentive for kids.

INCENTIVE SPIROMETER

The incentive spirometer is being provided by the IFOPA for older children, teens and adults. The goal is to take slow, deep breaths to expand your lungs. The incentive spirometer is meant to improve inspiration or your ability to take deep breaths.

User Instructions for the Incentive Spirometer:

Make sure to breathe through your mouth. You will need to have a good seal around the intake tube of the spirometer. Stop if you feel dizzy at any time, or if you have chest pain or discomfort, and rest.

1. Sit upright in a chair or bed, or stand upright. Position the spirometer close enough to use by holding the spirometer in front of your face at eye level or having someone assist with holding the spirometer in place.
2. When you start using the spirometer you'll set the yellow bar at a lower level (e.g. 500mL). Put the mouthpiece in your mouth and close your lips around it to make a seal. Slowly exhale completely.
3. Then, **slowly inhale** through your mouth as deeply as you can. Try to get as deep of a breath as possible. The yellow meter will rise as you inhale. Hold your breath for 10 seconds; your goal is to keep the meter in the "Best" range for as long as possible; then exhale. The meter may fall during this time, but over time you'll get better at keeping it steadily in the Best range.
4. After you've mastered "Best," then you can raise the bar on the right to 1,000 mL and beyond. When you can keep it steadily in the "Best" range for 10 seconds, then raise the bar to 1,500 mL and so on.

Repeat 10 times, with short rest breaks in between. You should do this twice per day. Make sure to talk with your physician if you feel you need help in setting a goal.

A video with instructions for can also be viewed here:

<https://www.mskcc.org/cancer-care/patient-education/how-use-your-incentive-spirometer>