MICHIGAN MEDICINE
UNIVERSITY OF MICHIGAN

Restrictive Lung Disease

What is restrictive lung disease?

Restrictive lung disease occurs when the chest cannot expand fully to allow the lungs to completely fill with air. Conditions that can cause this include:

- Neuromuscular weakness (muscular dystrophy)
- Syndromes in which the chest does not grow (Jeune Syndrome)
- Conditions in which the spine or rib cage is abnormal (severe scoliosis)

Some children have a combination of these conditions.

What are the symptoms of restrictive lung disease?

- Fast and shallow breathing
- High carbon dioxide levels which can result in headache and fatigue
- Feeling short of breath
- Sounding "out of breath" when talking

How is restrictive lung disease diagnosed?

- The best available tool is pulmonary function testing. For this test, children must be old enough to follow instructions and be able to seal their lips on a mouthpiece.
- Carbon dioxide measurements
- Chest X-ray or chest Computed Tomography (CT) scan

How is restrictive lung disease treated?

- Ventilator support either:
    - At night with a mask (noninvasive machine called a BiPAP that is similar to a CPAP machine).

    - Tracheostomy 24 hours a day (requires invasive surgery to create an opening in the windpipe and insert a tube for oxygen)
- Surgery to correct the underlying problem that is restricting the chest (such as scoliosis repair)
- Treatments to prevent accumulation of secretions in the lungs which may lead to pneumonia

Where can I learn more?

- Scoliosis Research Society: www.srs.org/patients-and-families/conditions-and-treatments/parents/scoliosis/thoracic-insufficiency-syndrome
- Pulmonary function testing American Thoracic Society www.thoracic.org/patients/patient-resources/resources/pulmonary-function-testing-children.pdf