



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

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Edited by: Karelyn Munro BA

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