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# Understanding Thoracic Insufficiency Syndrome

## Scoliosis can cause this disease of abnormal lung function and growth.

Written by [Isador H. Lieberman, MD, MBA, FRCSC](#) [1]

Thoracic insufficiency syndrome (TIS) occurs when the chest cavity prevents the lungs from growing and functioning normally. The chest cavity also known as the thorax is comprised of the spine, rib cage, and sternum—and it is the body's breathing machine. TIS develops in young children and is often caused by congenital scoliosis, which is when a baby is born with an abnormal spinal curve.

TIS is a rare condition and affects fewer than 4,000 children in the United States, according to the Children's Hospital of Philadelphia.

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### Causes of Thoracic Insufficiency Syndrome

Numerous syndromes and structural deformities can cause TIS, including kyphosis, lung disorders, or

genetic factors, but the most common cause is congenital scoliosis with rib fusion or absence of ribs. Congenital scoliosis occurs when the spine does not form properly while a baby is in the womb.

### How Scoliosis Affects the Lungs

When a child develops normally, the lungs grow at the same rate as the chest and spine. But in children with TIS, their lungs aren't able to develop properly because deformities, such as scoliosis, prevent the normal proportioned healthy growth of the rib cage and spine.

In short, the lungs need space to develop—and spinal and rib deformities won't allow that space.

Scoliosis and rib deformities may work together to constrict the lungs, so children with TIS experience breathing challenges. Some children with TIS may need supplemental oxygen or a ventilator to breathe.

### Thoracic Insufficiency Syndrome Signs and Symptoms

Many of the hallmark symptoms of TIS are related to breathing: Your child may struggle to breathe, have a spiked breathing rate, or seem breathless and tired.

From a body structure standpoint, signs of TIS include:

- An abnormal spinal curve (scoliosis or [kyphosis](#) [3])
- Shortened or narrow chest or rib cage
- Different shoulder heights
- A short neck

### Diagnosing Thoracic Insufficiency Syndrome

Most TIS diagnoses are in patients under the age of 5 years and always under the age of 10 years.

At the initial appointment, your doctor will first become familiar with your child's family and medical history, followed by a physical exam. After that, your doctor may conduct a gamut of tests to fully understand and confirm your child's TIS. These tests may include:

- **Pulmonary function studies:** These tests will reveal how well your child's lungs function.
- **X-rays:** X-rays are the test of choice when diagnosing and monitoring scoliosis, and your doctor may use x-ray to view the spinal curve and any rib cage problems.
- **Dynamic lung MRI:** This imaging test gives the doctor a clear view of your child's lung movement and breathing muscles. Your doctor may also order a separate MRI to see the spinal deformity.
- **EOS imaging:** This 3D imaging technology scans your child while he or she is standing up. It produces significantly less radiation than a standard x-ray.
- **Genetic testing:** Your doctor may use your child's saliva to understand if there's a genetic tie with the TIS or other associated syndromes.

### Treating Thoracic Insufficiency Syndrome

Three nonsurgical TIS treatment approaches are available to support healthy breathing:

- Nasal oxygen delivery
- Noninvasive positive-pressure ventilation (eg, CPAP)
- Ventilator support

When surgery is appropriate, the gold standard treatment is the vertical expandable [prosthetic titanium rib](#) [4] (VEPTR) device. This approach is the first FDA-approved treatment for TIS.

VEPTR uses titanium ribs to expand your child's chest and stabilize the scoliosis. The ribs are implanted into your child's back and chest. After the initial surgery, your child will have follow-up outpatient visits with your orthopaedic surgeon to have the ribs lengthened to facilitate healthy growth.

When the titanium ribs have reached their maximum length, and your child's spine and chest are developed (this typically occurs between 10 and 16 years old), your surgeon will remove the VEPTR device and perform a spinal fusion. The goals of the fusion are to straighten the scoliosis and stabilize the spine.

If your child has missing ribs, your surgeon may choose to leave the VEPTR device implanted permanently.

While VEPTR is still a relatively new treatment, outcomes have been good for children who've had the operation. The device allows the child to grow and develop, and the fusion completing treatment corrects the scoliosis.

### **Healthy Growth: Thoracic Insufficiency Syndrome Outcomes**

Receiving the diagnosis that your child has thoracic insufficiency syndrome as a result of his or her scoliosis can be unsettling, but treatments have advanced dramatically in recent years. The TIS treatments available today have enabled children to experience healthy growth, allowing both kids and parents to breathe easy.

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