

What is Scoliosis?

Scoliosis, a curvature of the spine, is common in children and adults who have OI. Many people with mild OI and almost everyone who have the more severe forms of OI develop some degree of scoliosis. Kyphosis is another type of spinal deformity seen in people with OI. Compression fractures of the spine can also lead to deformities of the individual vertebrae.

What is scoliosis?

It is a spinal curvature as seen from the front or back of the person who has it. It usually causes the spine to assume an "s" or "c" shape instead of being straight. Some curves get progressively worse. The curve is measured in degrees, and curves over 30 degrees need to be carefully watched because they can worsen rapidly. Once the spine has developed a significant curve, even surgery will not be able to make it perfectly straight again. Scoliosis occurs in many people, but it is seen more often in people with OI than in the general population.

What causes Scoliosis?

Scoliosis is probably caused by two aspects of OI – the associated laxity of the ligaments, and deformation of the vertebral bones themselves. Long periods of sitting can aggravate the problem of lax (overly relaxed or loose) ligaments and weak muscles in the back and chest. Many children and adults also experience painful compression fractures of the vertebrae (spine). These fractures can be caused by any activity that jars the spine. Over time, these fractures deform the bones of the spine and contribute to a worsening of scoliosis.

Can scoliosis be detrimental to an OI person's overall health?

Scoliosis increases pain, and collapse of the trunk. It can cause rotation of the ribs. The effects of scoliosis can make the child or adult significantly less mobile. Less mobility will limit the child's ability to develop bone mass and contributes to a loss of bone density in the adult. Progressively deforming scoliosis can also lead to life threatening heart and lung problems.

Why is scoliosis a problem for many children and adults who have OI?

Severe Scoliosis decreases lung function and the ability to breathe normally. This happens only in severe cases, and is not a worry for people with mild scoliosis. But for people who have more severe curves, lung capacity can become so reduced that the person takes very shallow breaths and does not get enough oxygen into their body to stay healthy. Often supplemental oxygen is needed. Individuals with severe curves should consult with a doctor who is a respiratory specialist to have their lung capacity tested.

How is scoliosis treated?

Once scoliosis is identified it should be watched. The degree of curve should be regularly checked to see if there are any changes. In mild to moderate cases, it does not need any treatment beyond maximizing overall fitness and bone density. In growing children with Type I OI, there may be some benefit to bracing for mild curves. This is decided on a case-by-case basis because the pressure exerted by a back brace can injure the fragile rib cage and chest deformities can get worse.

For more severe curves (over about 45 degrees), surgery is recommended to straighten and fuse the curved part of the spine using rods and bone graft. Both adults and children can be candidates for this surgery, but children undergo the procedure more readily and recover faster.

What can a person with OI and scoliosis do to protect their spine?

Everyone who has OI should make it a habit to always protect his or her spine by learning safe ways to bend, walk and sit. Twisting motions and anything that jars the spine should be avoided. Children and adults with scoliosis, especially serious curves, should also pay close attention to their respiratory health. Consulting with a

This fact sheet was prepared with assistance from Dr. Paul Sponseller, Orthopedic Surgeon, Kennedy Krieger Institute and co-director of their OI Clinic. Reviewed 2005 and 2016.

respiratory specialist is one way to monitor how well one's lungs are working. In addition, don't smoke, avoid second hand smoke and work with a doctor or physical therapist to develop a safe exercise program. These are things that can be done to promote respiratory health. **Of course it's especially important to check with an orthopedic surgeon knowledgeable about scoliosis to get the best recommendation appropriate for each individual's situation.**