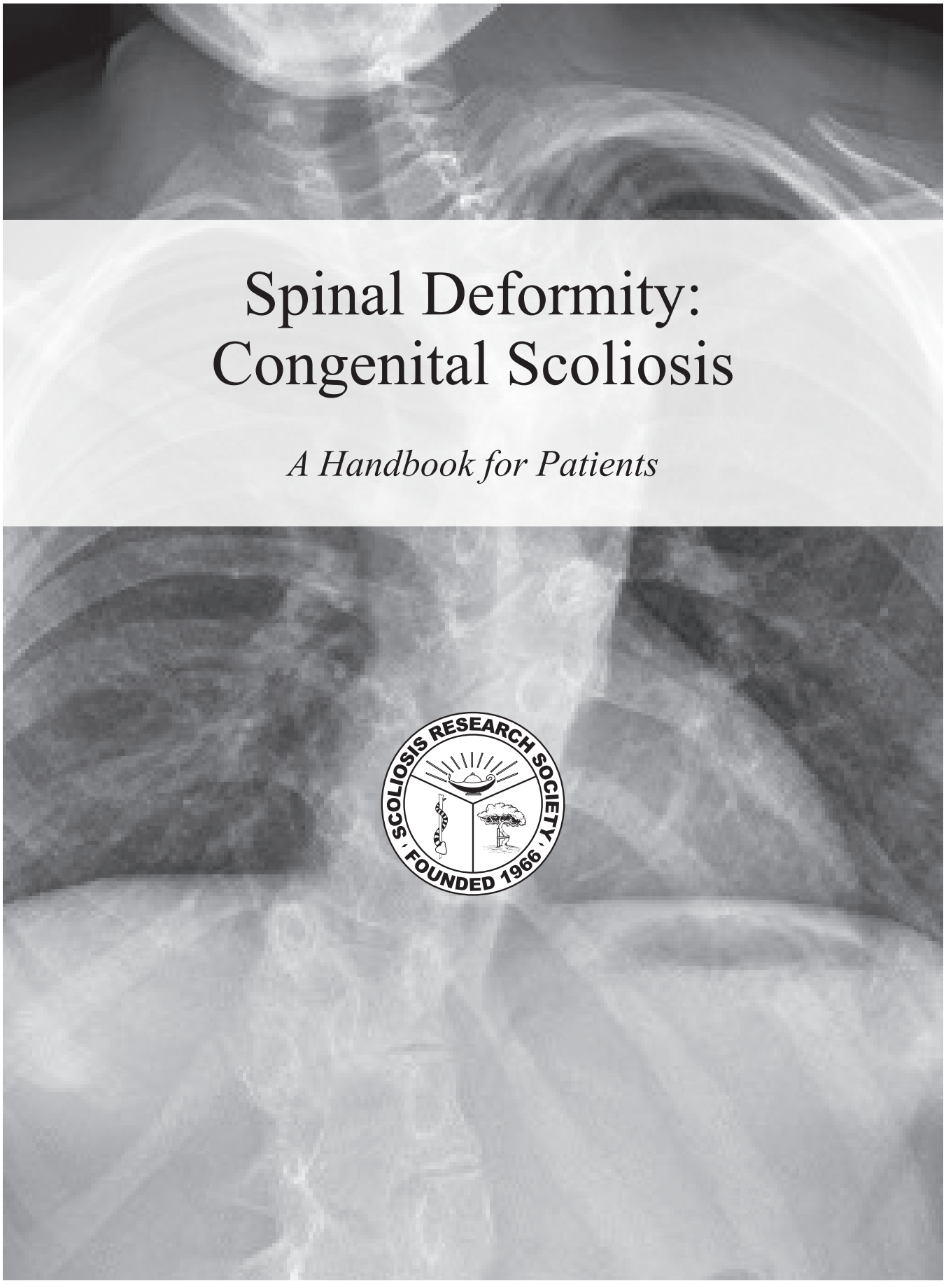
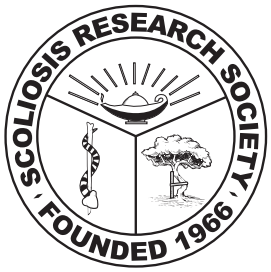


Spinal Deformity: Congenital Scoliosis

A Handbook for Patients

SCOLIOSIS RESEARCH SOCIETY · FOUNDED 1966 ·



Scoliosis Research Society
Dedicated to Education, Research and Treatment of Spinal Deformity
555 East Wells Street, Suite 1100
Milwaukee, WI 53202
Phone: (414) 289-9107 | Fax: (414) 276-3349
Email: info@srs.org

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Preface

The Scoliosis Research Society's Patient Education Committee has prepared this booklet to provide patients and their families with a better understanding of congenital scoliosis. This information is intended as a supplement to the information your physician will provide. The behavior of congenital spinal deformities may be very different from one individual to another, although a number of general statements can be made. However, your spinal deformity surgeon will be the most important source of information for you or your child's particular case.

It is beyond the scope of this booklet to discuss in detail the technical aspects of all the various surgical procedures that may be needed, but general concepts are discussed. Additionally, it must be stressed that congenital spinal deformities are much different than the more common idiopathic type of deformity, requiring different evaluation and treatment.

It is not intended that the contents of this manual be interpreted as standards or guidelines proposed by the Scoliosis Research Society.

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Why is it called congenital scoliosis?

Congenital scoliosis is an abnormal curve or twist of the spine that is caused by the way the spine bones (vertebrae) are formed. The term "congenital" means that patients were born with the abnormal vertebrae. This is different from the more common "idiopathic" scoliosis, which occurs in children with vertebrae that have a normal shape.

What causes congenital scoliosis?

The spine forms at a very early stage of pregnancy. In about the 8th week of pregnancy, the group of cells that form the spine start to become the individual bones of the spine. At this stage, errors can occur. When these cells don't form a complete vertebra, the spine will have partly formed bones called hemivertebra (half verebra) or even wedges. *(Figure 1)*

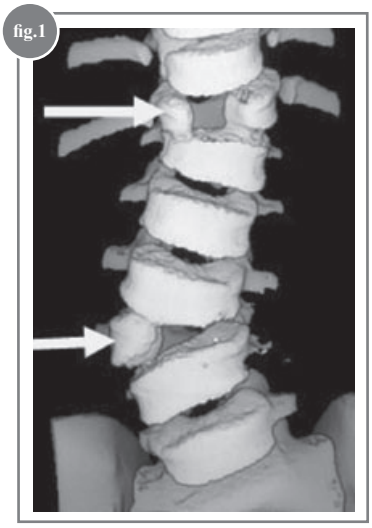

Figure 1:
3D image of abnormally-formed hemivertebra.

Sometimes, these cells don't leave space between the spinal bones and they end up being stuck together (sometimes called a fusion or bar). Fusions can also occur in the ribs, which connect to the vertebrae and form at the same time. The causes of these errors as the vertebrae are forming are unknown. Several factors may increase the risk of congenital scoliosis like a lack of oxygen as the baby develops, a lack of folic acid during pregnancy, cigarette smoking, alcohol or drug use during pregnancy, and if the mother has diabetes. Some cases of congenital scoliosis are inherited or passed from parents to children, but most are not.

How common is congenital scoliosis?

The estimate is that 1 in 1,000 children have congenital scoliosis. However, this could be underestimated because many people have no symptoms and are not aware that they have an abnormality in their spine. Ultrasound tests during pregnancy have improved our ability to diagnose more cases of congenital scoliosis.

How many different types of congenital scoliosis are there?

A vertebra may be partially formed or may be fully formed but not fully separated from surrounding bones. It is also possible to have combinations of both types. *(Figure 2)*

fig.2

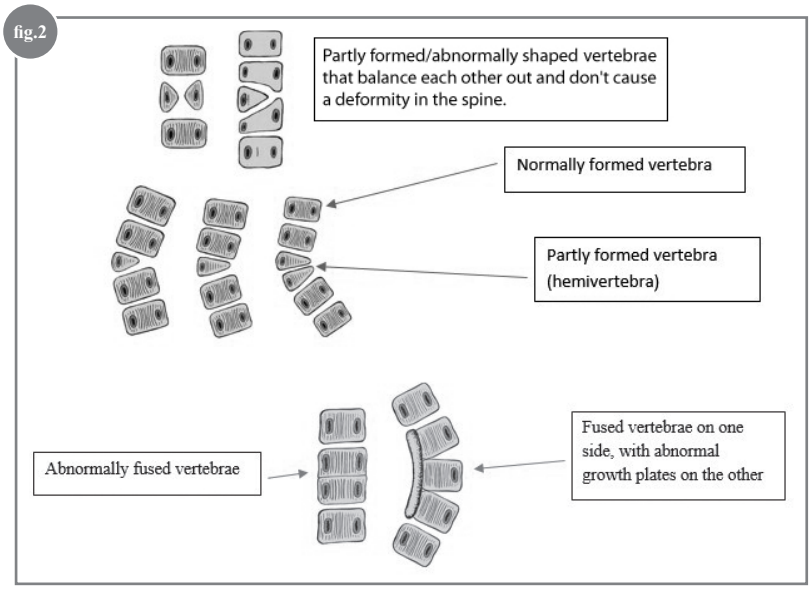

Figure 2:
Types of Congenital Scoliosis

Severe congenital scoliosis that effects the entire rib cage can make breathing difficult, and is called thoracic insufficiency syndrome. *(Figure 3)*

fig.3

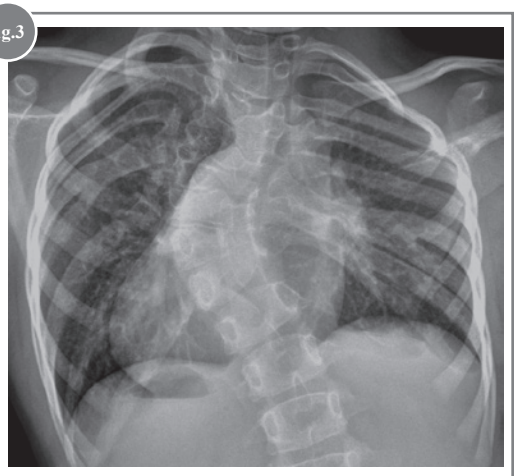

Figure 3:
A patient with multiple abnormal vertebrae and multiple fused ribs, characteristic of thoracic insufficiency syndrome.

What happens to congenital scoliosis as children grow?

The severity of congenital scoliosis can vary greatly. Some children will never notice anything at all: no pain, no change in shape, and no loss of function. In these cases, parents and children may be unaware they have the condition, and congenital scoliosis may only be discovered if X-rays are done for other reasons.

Others children will develop severe curves of the spine at an early age. Deformities can progress (get bigger) around the abnormally shaped vertebrae as one side grows more than the other. These patients may also form curves in the normal parts of the spine to maintain overall balance. These balancing curves are called *compensatory curves*.

Each developing vertebra normally has two growth plates, one on the top and one at the bottom. These are made of cartilage, which grows and then turns into bone. Abnormally shaped vertebra may have two growth plates, one or none. Normal vertebrae are shaped like rectangles, and the growth plates are even with each other, which causes them to grow up straight. Hemivertebrae are often shaped like triangles, which causes the growth plates to grow in the wrong direction, leading to spine curvatures. Think of your spine bones as building blocks on top of each other. If the ends are all flat, the spine will grow evenly. If you put a triangle between two rectangles, the 'tower' will curve. Some malformations can have no growth on one side of the curve and full growth on the other, causing scoliosis that gets worse quickly.

fig.4

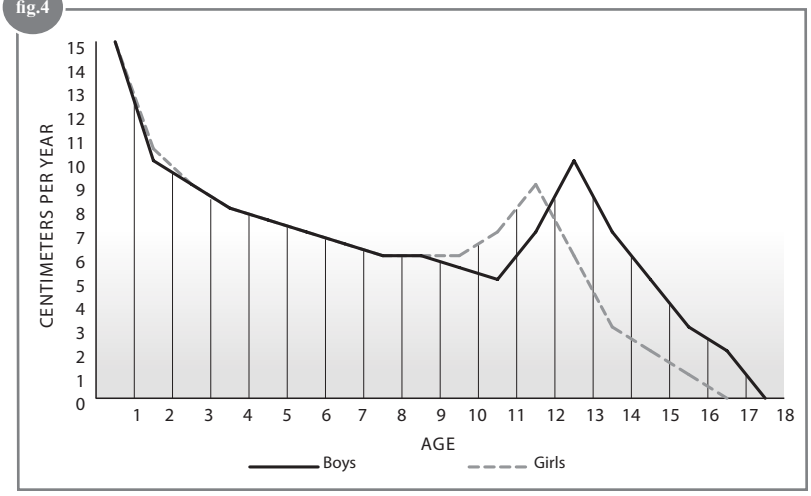

Figure 4:
Peak Height Velocity Curve for girls and boys showing the increase in height

Children grow most quickly as babies and toddlers (between the ages of 0 and 3 years), and during the adolescent growth spurt. Congenital scoliosis can get worse quickly during periods of rapid growth, so these are the times when monitoring needs to be more frequent.

The location of abnormalities in the spine can affect how bad the problem will be. Abnormalities at the junctions between different areas of the spine, such as those between the neck and upper back, between the upper and lower back, or between the lower back and the pelvis, may cause worse problems.

In severe cases, curves can be large enough to cause problems with the spinal cord, causeing weakness in the legs, partial, or even total paralysis. Injury to the spinal cord occurs most frequently in cases where there is a large, sharp forward curvature to the spine (congenital kyphosis).

What other problems can occur in patients with congenital scoliosis?

A number of other organs in the body are formed at the same time as the spine, including the brain, spinal cord, heart, and kidneys. Children with congenital scoliosis should be checked for abnormalities in these other organs. Approximately 30% of children with congenital scoliosis will have at least one other abnormality.

How are congenital spine abnormalities evaluated?

Ultrasound during pregnancy can show the location of some spinal abnormalities. However, not all abnormalities are visible on ultrasound, and the severity of curves cannot be properly assessed as babies rest in a curved position inside the uterus. Ultrasounds of the spine are less useful after birth for showing bony details, but may be used during the first few months of life to show the spinal cord. Ultrasounds are especially helpful in children of any age to check kidney development.

X-rays are the main method of assessing congenital scoliosis and tracking changes over time. Whenever possible, X-rays are performed in an upright (sitting or standing) position, because the way gravity affects the spine is important. The severity of curves is assessed by measuring the angle of the curve in the spine (called a Cobb angle). X-rays need to be repeated over many years to assess whether curves are progressing. Some changes will be very slow to develop, and may only be possible to spot by comparing X-rays over many years. Although all X-rays use radiation, modern techniques minimize the amount needed to get good pictures of the spine, and doctors routinely order only the number of X-rays needed to properly monitor the congenital scoliosis.

MRI scans may be used to see the brain, spinal cord, and nerves. Occasionally, abnormalities are detected in the spinal cord that need to be evaluated by a neurosurgeon. These abnormalities can include parts of the brain descending into the space normally reserved for the spinal cord (called a Chiari malformation), abnormal flow of spinal fluid creating pockets in the spinal cord (called a syrinx or syringomyelia), split spinal cords (called diastematomyelia), or an abnormal attachment at the base of the spinal cord (called a tethered cord).

What are the treatment options for congenital scoliosis?

If a curve has developed in the spine due to abnormal growth of vertebrae it will almost always require surgery to prevent it from getting worse. There is generally no role for non-surgical treatments such as exercise, stretching, physical therapy, chiropractic manipulation, or bracing. Occasionally, casts or braces may be used to control compensatory curves (curves that have formed in the normal parts of the spine to balance the congenital curves). In congenital scoliosis the vertebrae are abnormal, the way the spine grows is abnormal, and most will require surgical treatment.

What types of operations are used in the treatment of congenital scoliosis?

1. *In Situ Fusion:*

This is an operation generally performed at a young age, on curves that have not yet become too big or problematic. The goal of an *in situ* fusion is to eliminate the growth of the abnormal vertebrae to prevent further worsening of the curve. It is typically not used for larger curves, as there is no correction of the shape of the spine with this operation. It is also not suitable to perform this operation over a very long section of the spine in a young child because a successful fusion eliminates further growth. However, it may be used in situations where more extensive corrective surgery is too difficult or dangerous.

Metal rods anchored to the vertebrae by screws or hooks may be used in this operation to hold the bones still and increase the speed and quality of bone fusion. But unlike other scoliosis operations, the rods are not used to change the shape of the spine.

2. Hemivertebra Excision with or without Fusion and Instrumentation

This operation is used in situations where a partly formed vertebra is growing asymmetrically/unevenly and making the scoliosis worse. The aim of this operation is to remove the abnormal vertebra and its growing parts, and then connect some of the remaining vertebra above and below the malformed vertebra with metal implants. *(Figure 5)*

This operation can be performed in young children, although most surgeons choose to wait as long as possible because the fusion prevents further growth of the involved part of the spine. Since each case of congenital scoliosis is unique, the timing of an operation is best determined by your surgeon and takes into account remaining growth, current curve size, recent changes in the curve, the likelihood of future progression, and the risks of surgery.

fig.5

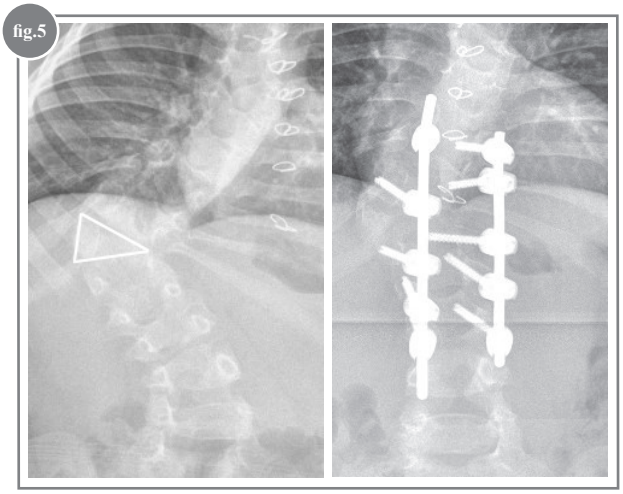

Figure 5:
Front view of a patient with a hemivertebra causing severe scoliosis, before and after hemivertebra resection.

3. Spine Fusion with Instrumentation

This operation is used closer to the end of growth. The goal of this operation is to fuse the abnormally curved part of the spine, decrease the size of the curve, and maintain overall spinal balance. Although most curves are improved, they rarely are made completely straight.

a. **Posterior Spine Fusion:** In this operation, an incision is made directly on the back over the curved part of the spine. Loosening of the spine is achieved by removing parts of the small joints (facets) and ligaments between the vertebrae. More extensive loosening may be added by performing spine Osteotomies (see below: Spine Osteotomies). Metal rods are anchored to the spine by screws, hooks, cables, or wires. The rods are used to move the spine into a straighter position. The exposed parts of the vertebrae are scraped, which helps the bones to fuse to each other. Bone graft may be added to speed the formation of the new bone. *(Figure 6)*

fig.6

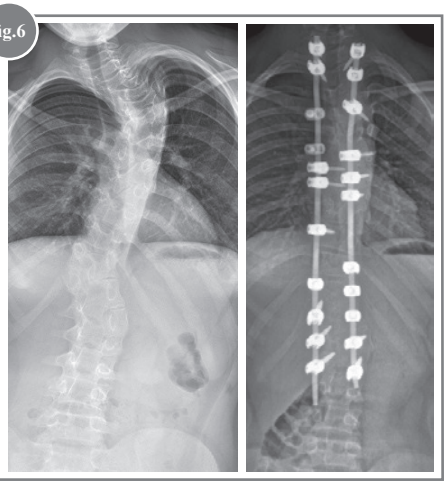

Figure 6:
A teenage patient before and after posterior spinal fusion for congenital scoliosis.

b. **Anterior Spinal Fusion:** The spine is approached through the side of the chest or abdomen (or both) to reach the front (anterior) part of the spine. Loosening of the spine is achieved by removal of the discs and growth plates connecting the vertebrae. Cages, bone graft, or both, may be inserted in the spaces where the discs were removed, and rods are passed to the front of the vertebrae.

c. In special situations, patients may require both anterior and posterior spine fusions. They might be performed on the same day, or separate days in two stages.

4. Growing Rods

This type of operation is performed in situations where the curve needs to be controlled, but there is too much growth remaining to do an extensive spinal fusion.

The goals of growing rod surgery are to prevent worsening of the curve, possibly improve the shape of the spine and to allow as much normal growth as possible.

These goals are achieved by inserting rods that are anchored to the spine and/or ribs by hooks or screws only at the top and the bottom of the curve. In between, the spine is not exposed to prevent fusion. The rods are tunneled beneath the skin and back muscles from one end to the other.

Growth is achieved by extending the rods. There are several ways this can be done:

1. Repeat surgery can be performed (usually every 6 months) to loosen the connectors between the rods and stretch them further apart.

2. Some rods can be made longer using a small magnet that is built into them. Lengthening is achieved by applying an external magnet to activate the device. This can typically be done in an office setting, and decreases the need for multiple surgeries. *(Figure 7)*

3. Some rods are designed to lengthen using a ratchet mechanism on a threaded rod. Lengthening is achieved by special movements or stretches, which may also decrease the need for multiple surgeries.
4. This construct is used frequently when there are abnormally fused ribs along with the congenital scoliosis. *(Figure 7)*

Growing rods in congenital scoliosis are not able to create as much spine length as they are in other types of scoliosis. This is because the abnormal vertebrae in congenital scoliosis cannot grow as much as normal vertebrae.

fig.7

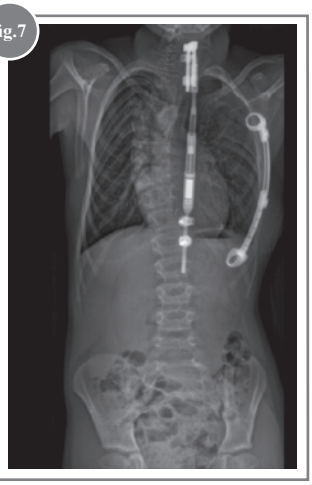

Figure 7:
X-ray of a patient with a magnetic lengthening rod adjacent to the spine and a growing rod construct attached to the ribs.

5. Spine Osteotomies

"Osteotomy" means to create a cut in a bone. Spine osteotomies are typically used when deformities are severe and adequate correction cannot be achieved with the rods and simpler methods to loosen the spine.

In these operations, parts of the vertebrae are removed. This creates more flexibility in the spine that is then controlled by the inserted rods and anchors. In some cases, osteotomies may include removal of parts of ribs. Osteotomies often involve a greater risk of injury to the spinal cord, blood vessels and nerves, and so are typically done only when needed to achieve correction that cannot be accomplished by other methods.

What factors do surgeons consider to pick the best operation?

1. Growth

Your surgeon will consider the positive and negative aspects of growth before surgery, as well as what effect the surgery will have on the remaining growth of the spine.

Positive aspects of allowing growth to continue before surgery include allowing children to become bigger and healthier, and to allow more time to optimize nutrition and increase their lung sizes. In general, larger children have better lung function, can tolerate more blood loss, and have bigger, stronger bones to hold the anchors for the metal rods.

The main negative aspect of growth is worsening of the spinal curvature, which may cause worsening lung function, pain, and imbalance of the spine. Some congenital scoliosis or kyphosis curves can even damage the spinal cord if they get too large. Also, as the curve gets worse, a bigger surgery is needed that increases the risks for the patient.

2. Size of the Current Curve

A number of factors determine how acceptable the current curve is. From the patient's and family's point of view, there may be concerns about pain, or the effects on walking, breathing, or spinal cord function. There may also be concerns about the appearance of the spine, which may not look like it does on the X-rays. All those involved need to consider how the curve may appear in the future, and what other risks are involved if the curve progresses. There may be a trade-off between the risks and benefits of simpler surgeries that keep the curve from getting larger instead of allowing the spine to grow and having to do a more difficult surgery in the future.

3. Location and Accessibility of the Scoliosis

Some parts of the spine are more difficult to reach than others and pose unique risks and challenges. These areas include the cervical spine (in your neck) and where it attaches to the skull, the upper thoracic spine (at the top of your rib cage) and the lower lumbar spine (low back) and its connection with the pelvis. Some of these areas may be treated more easily with an in situ fusion early, rather than a more difficult operation, such as a spine osteotomy, later.

4. Likelihood of Achieving the Surgical Goals

Your surgeon will consider which of the surgical techniques described above has the best chance of stopping progression of the congenital scoliosis, achieving correction of the curve, and maintaining as much normal spine growth as possible.

5. Risks of Surgical Treatment

All surgeries include some risk of complications. Spine surgeons are constantly working to prevent and minimize complications.

As a general rule, operations that carry greater risks are those that take longer, that involve more spine levels, that extend to the more difficult areas mentioned above, or that create greater instability of the spine (such as spine osteotomies). Complications known to occur during spine surgery include: blood loss that requires a blood transfusion; surgical site infections; damage to the nerves, spinal cord, or blood vessels; failure of the bones to fuse as intended; failure, loosening, or breaking of the metal implants; and unexpected worsening of the scoliosis despite a fusion operation. Although all are relatively uncommon, your surgeon can discuss how often these complications, or any others, occur in their practice, or at their hospital.

Common post operative problems that are not classified as complications include difficulty controlling pain, constipation, and limited mobility.

Summary

Congenital scoliosis is a unique type of scoliosis that happens because the spine bones (vertebrae) did not grow normally before the baby was born. People with congenital scoliosis may require additional tests to look for possible problems in other parts of the body like the heart, kidneys and spinal cord. Treatment for congenital scoliosis must take into account how large the curves are, whether they are progressing, in which part of the spine they are located, if there are other associated abnormalities, and how much growth the child has left. There are many different treatments for congenital scoliosis, most of which require specialized spine surgery. Your surgeon will work with you to identify which treatment is the most appropriate for you or your child.

Where can I get more information?

The best information about your specific condition typically comes directly from your surgeon. You can check to see if your surgeon is a member of the Scoliosis Research Society by going to http://www.srs.org/find/. Membership in SRS indicates that at least 20% of the doctor's practice is in spinal deformity, that they attend annual meetings, and stay abreast of new information and new research.

In addition to the Scoliosis Research Society website (www.srs.org), there are other reputable organizations that may offer information about Congenital Scoliosis.

Here is a list of some patient resource websites that may be of assistance:

- www.srs.org – Scoliosis Research Society
- www.posna.org – Pediatric Orthopaedic Society of North America
- www.aaos.org – American Academy of Orthopaedic Surgeons
- www.nih.gov – National Institutes of Health
- http://etext.srs.org/ - SRS provides information through the E-Text as an educational service. E-Text material is not intended to represent the only, nor necessarily best, methods or procedures appropriate for the medical situations discussed, but rather is intended to present an approach, view, statement or opinion of the chapter author(s) that may be helpful to others who face similar situations. SRS disclaims liability for all claims that may arise out of the use of techniques demonstrated therein by such individuals.

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100 percent of all contributions and donations to the Scoliosis Research Society's (SRS) Research, Education Outreach (REO) Fund are used entirely for research, outreach programs, and educational scholarships and fellowships seeking improved treatments, the causes and possible prevention of spinal deformities. Operating funds for SRS come from membership dues, educational meetings and courses, publication sales and other sources.

With your support, SRS can continue to support and offer necessary educational opportunities, beneficial research grants and maintain effective advocacy efforts that will change the lives of those living with spinal deformities.

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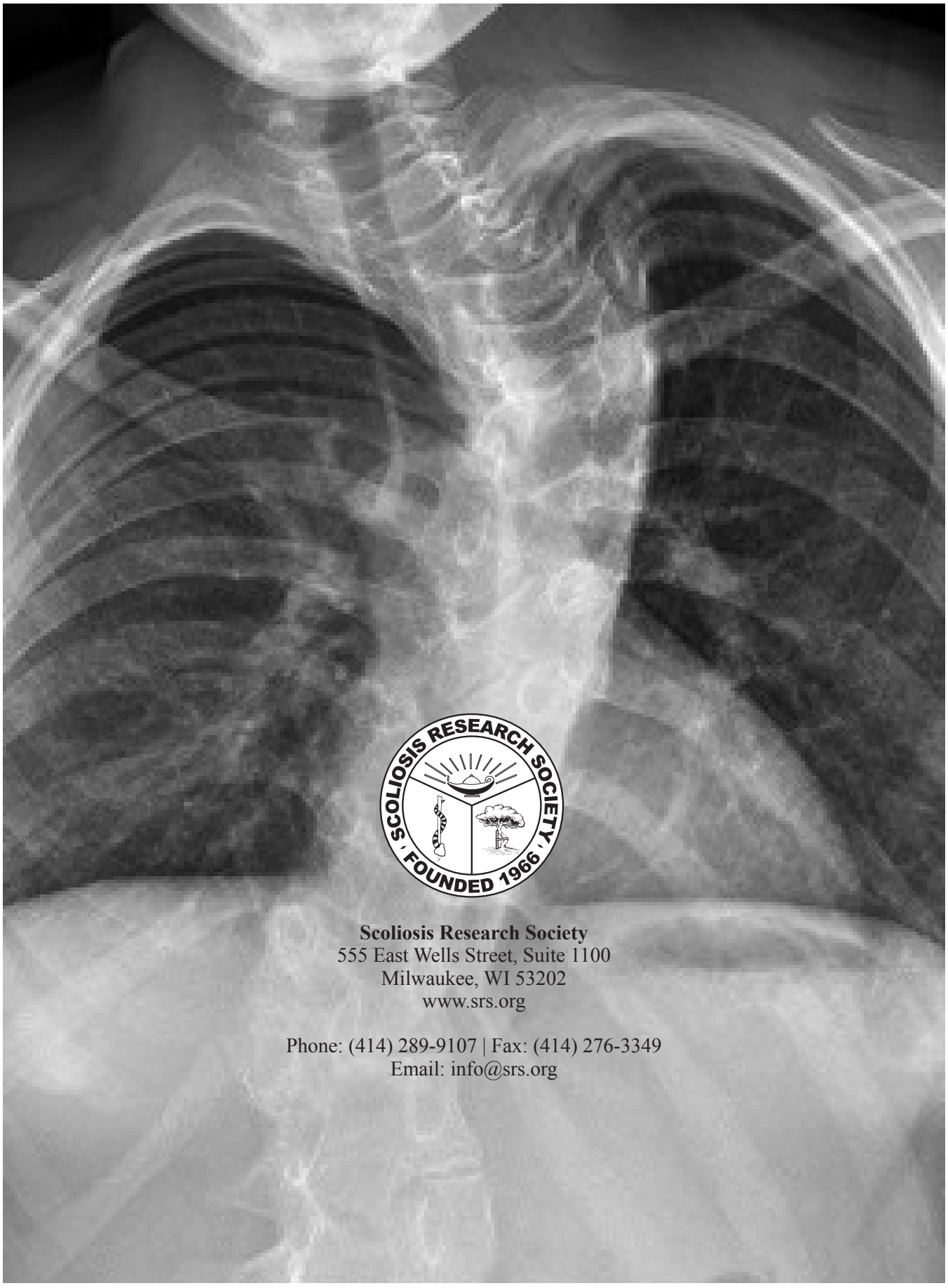

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