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PEDIATRIC SCOLIOSIS

WHAT IS SCOLIOSIS?

Scoliosis is an abnormal curvature of the spine. The normal spine, when viewed from behind has no visible curve. When viewed from the side there will normally be a slight round back (kyphosis) in the upper back and a mild swayback (lordosis) in the lower back. When scoliosis develops (viewed from the behind) a curvature to the right or left will be seen (Figure 1). Associated with the abnormal curvature, there is also twisting of the spine. This twisting results in asymmetry of the rib cage. When viewed from the back a prominence of the ribs will be visible on the side of the curve.

Scoliosis may develop in one or more areas of the spine. Additionally, the curve patterns in scoliosis vary from short single curves to large double or triple curves (Figure 2).


Figure 1


Figure 2

WHAT CAUSES SCOLIOSIS?

There are several causes of scoliosis. In some cases it results from an abnormal development of the bones in the spine. This is called **congenital** scoliosis and is present from the time of birth, although it may not be recognized until the deformity increases with growth. The primary sources of the deformity in this case are the abnormally shaped bones or asymmetrically fused bones in the spine that do not grow properly (Figure 3a). Another cause for scoliosis is poor neurologic or muscular control of the spine. In these circumstances, support from the muscles of the back is insufficient to maintain an erect spinal position. This type of **neuromuscular** scoliosis is seen in conditions such as cerebral palsy, spina bifida, muscular dystrophy, etc (Figure 3b). The most common cause for scoliosis in children and adolescents, however, results from unknown causes. When there is no underlying abnormality of the spine or of the neuromuscular system, the scoliosis is considered to


Figure 3a


Figure 3b

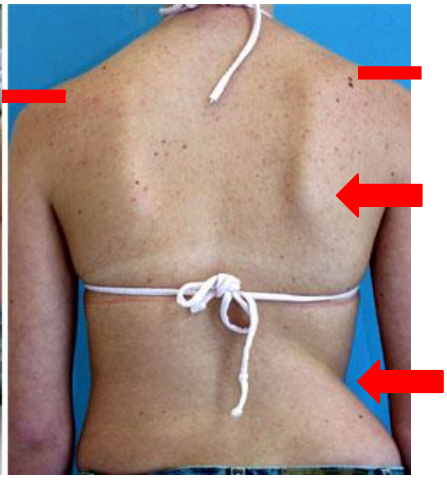


Figure 3c

be **idiopathic** (Figure 3c). Much investigation has gone into looking for the cause of scoliosis in these otherwise normal children, and to date no clear cause has been identified. There does appear to be an increased incidence of scoliosis within a family, suggesting some genetic influence. Thus, scoliosis may develop due to congenital, neuromuscular, or idiopathic causes.

WHAT ARE THE CONSEQUENCES OF SCOLIOSIS?

The deformity of the spine, in cases of scoliosis, tends to increase with increasing growth of the spine. There are cases, however, in which no progression of the deformity occurs or the increasing size of the deformity is minimal. The shape of the body is altered depending on the scoliosis curve pattern. Concern develops in cases when the deformity is progressive. With large curves, trunk and chest deformity develop to a degree that ultimately inhibits normal lung and heart function (Figure 4a, b). For these reasons, treatment of scoliosis is recommended when a progressive curvature is detected.

Figure 4a

Figure 4b

HOW IS SCOLIOSIS ASSESSED?

The diagnosis of scoliosis is made by examining the shape of the back and trunk. As scoliosis progresses, a difference in the height of the shoulders may develop, as well as an asymmetry of the waistline and contour of the back (Figure 5a). Because of the rotational changes in the spine with the development of scoliosis, prominence of the ribs occurs. This can be seen most readily by examining the patient from behind when bent forward at the hips (Figure 5b). This forward bending test (Adam's test) is used in screening patients for scoliosis. In many states a scoliosis school screening program exists in which all children are examined for back and rib asymmetry. When a rib prominence is noted, a standing radiograph of the spine is recommended. The x-ray evaluation of the spine is made to determine the magnitude and pattern of spinal curvature. The angulation (measured in degrees) between the most tilted vertebrae is determined using the Cobb angle method (Figure 5c). It is this measure of scoliosis on which treatment recommendations are based.

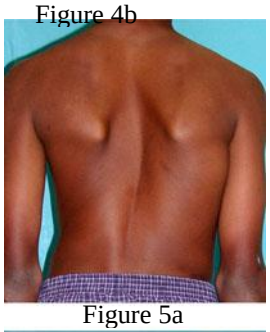


Figure 5a

Figure 5b

Figure 5c

RISK OF PROGRESSION:

In addition to the degree of scoliosis, it is important to assess the skeletal maturity of the patient. It is known that the risk of the curve enlarging is related to the amount of growth remaining in the skeleton and spine. This can be assessed in several ways. The first is a radiographic measure of the maturity of the skeleton, done by analyzing the growth centers of the pelvic bone. The Risser sign is a marker for skeletal maturity, which can be followed on the pelvic x-ray to help predict whether or not growth is expected to continue. Other measures of remaining growth relate to the changes of puberty that can be assessed in both boys and girls. An additional helpful time

Figure 5c

point in girls relates to the onset of menstrual periods. The beginning of menstrual period suggests 1-1/2 to 2 years of remaining spinal growth. It is largely these two facts, an estimate of the remaining spinal growth and the magnitude of the spinal curvature, which are used to predict the potential for scoliosis progression. The underlying cause of the scoliosis also plays a significant role in determining the likelihood of progression and need for treatment. It is recommended that a physician with expertise be involved in the care of a patient with scoliosis when the Cobb angle is greater than 15°.

HOW IS SCOLIOSIS TREATED?

The options for treating scoliosis include observation, the use of a brace (orthosis), or surgery. Currently, there is no evidence that exercise or spinal manipulation have a positive effect on preventing the progression of scoliosis.

The treatment of congenital scoliosis initially is observation by checking x-rays of the spine periodically. If increasing curvature is appreciated on serial examinations, surgery may be required to prevent the curve from increasing in size. Bracing is indicated in very few patients with congenital scoliosis. The surgical treatment for such cases is a spinal fusion, which limits the growth of the spine and prevents progression of the curve.

The indications for brace treatment in neuromuscular causes of scoliosis are variable. In many cases the brace is helpful to maintain sitting posture in patients who are unable to walk. The ultimate progression of the curvature may be delayed only slightly. In many neuromuscular conditions curve progression develops despite brace wear. The benefit of bracing in these circumstances is primarily in helping sitting balance. Surgical indications in neuromuscular scoliosis vary depending on the underlying diagnosis, pulmonary status, curve pattern, and magnitude.

Idiopathic scoliosis is treated with a brace in those patients who have curves that are progressing to greater than 25-30° and are thought to have substantial growth remaining.

HOW DOES BRACING HELP?

A brace used to control scoliosis is designed to apply external pressures to the trunk and pelvis, improving the alignment of the spine and allowing straighter growth of the spine (Figure 6a, b). Bracing of scoliosis is not helpful in patients who have completed or nearly completed their spinal growth. Bracing is, therefore, indicated for patients with idiopathic scoliosis with curves between 25-45° with greater than six months of skeletal growth remaining.

There are several types of orthoses or braces available (Figure 7a, b, c). All of the braces attempt to straighten the spine and are most effective if worn full time. The brace is made to fit intimately the patient's trunk, providing pressure against the pelvis and rib cage to reduce the deformity. A brace maker (orthotist) is used to

fit and adjust the brace accordingly. Initial brace wear may be uncomfortable, and a period of time is required to become accustomed to wearing the brace. X-rays are used to assess the correction of the spine in the brace and are taken periodically (every 3-4 months) to monitor the curve. The total length of time the brace will need to be worn is dependent on the remaining spinal growth. In general, the brace is worn until spinal growth has been completed. As this point is reached, the hours spent wearing the brace will be reduced and the brace ultimately discontinued.

WHEN IS SURGERY NEEDED?

The indication for proceeding with surgical treatment of scoliosis is dependent on multiple factors. The diagnosis and magnitude of the curve are the primary factors used in determining whether surgery is required to treat the deformity. Patients with idiopathic scoliosis, whose curves have progressed to greater than 40-50°, despite brace wear, are considered candidates for surgical treatment. Other important factors include the curve pattern, trunk imbalance, and skeletal maturity. The goal of surgical treatment is to provide a method for correcting the deformity, as well as preventing further progression of the curvature. Surgery is, therefore, useful in patients who have progressive curves that have not been controlled by brace treatment or who have curves that are too large to be amenable to brace treatment.

WHAT ARE THE SURGICAL OPTIONS?

The surgical treatment of scoliosis involves a spinal fusion. This procedure removes the flexible elements of the spine, creating a solid block of bone throughout the fused area of the curve. This, in most cases, is performed with the addition of an instrumentation system of rods, hooks, wires, and/or screws. The instrumentation system is used to correct the curvature of the spine as much as possible, as well as to maintain stability of the spine in the postoperative period while solidification of the fusion occurs. The surgical approach to the spine can be either through an incision on the back (posterior) or through an incision in the side (anterior).

Posterior Approach:

When doing a posterior spinal fusion the spinous processes, lamina, and transverse processes of the spine are exposed. It is to these elements of the vertebra that the hooks, wires, or screws are placed to provide fixation of the rod to the spine (Figure 8a, b, c). Bone graft is applied to the exposed bony surfaces of the spine, which allow these vertebrae to subsequently grow together or fuse with bone connecting each of the vertebra to one another.

Figure 8a Figure 8b Figure 8c

Anterior Approach:

In some curve patterns a similar correction and fusion can be performed via an anterior approach. This involves an incision on the side of the chest or abdomen to expose the front or anterior part of the spine. It is in this area where the vertebral bodies and discs are located. Fusion in this area involves removal of the soft disc material between the vertebrae and replacement of this tissue with bone graft, which will allow bony union or fusion between the adjacent vertebral bodies. Instrumentation systems that are applied anteriorly generally involve fixation to the vertebral body with a screw and connection between the screws with a rod system (Figure 9a, b). This anterior approach can be done in the thoracic, thoracolumbar, or lumbar regions, depending on the curve pattern.

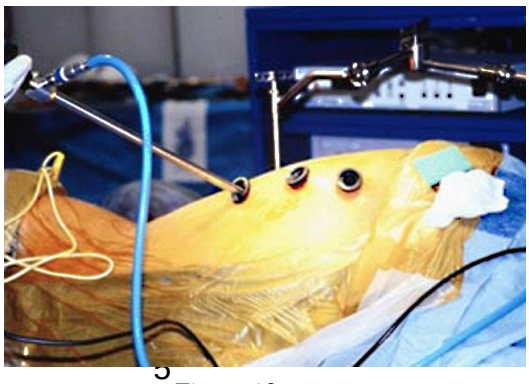
Anterior and Posterior Approach:

Figure 9a Figure 9b

There are times when both an anterior and posterior approach to the spine is required. In circumstances when the curve is extremely rigid or the patient quite young (less than 10-12 years), approaches to both the front and back of the spine may be required. Much of the growth potential in the spine is located in the vertebral body adjacent to the disc. This growth center is responsible for vertebral body growth and causes much of the progression of the curvature that occurs during spinal growth. Therefore, anterior fusion is useful at decreasing or stopping the anterior vertebral body growth. Therefore, in young patients, anterior fusion is recommended. The anterior fusion may be required in combination with a posterior fusion and instrumentation if the curve cannot be instrumented anteriorly. When a curve is particularly large and rigid, correction may be inadequate if posterior instrumentation is done without removal of the anterior discs. Removal of the anterior discs in such a case increases the flexibility of the spine and allows for better correction of the deformity by the instrumentation system.

Thoracoscopic Approach:

Minimally invasive methods are also available for approach to the anterior aspects of the spine. In the chest this is done with what is known as a thoracoscopic approach. The endoscopic approach utilizes small incisions and video technology to view the spine. Specialized instruments are used to perform disc removal, instrumentation, and fusion (Figure 10a, b). In some cases a posterior fusion is also required to perform the instrumentation portion of the procedure, although some curves can now be corrected with the endoscopic approach alone.



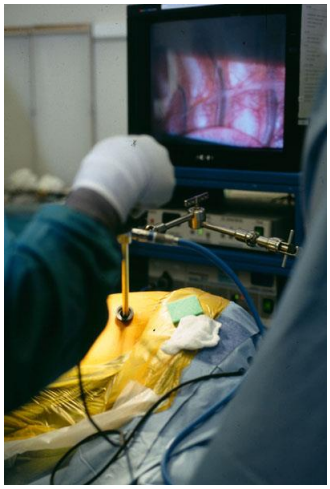


Figure 10a Figure 10b

WHAT ARE THE RISKS OF SURGERY?

With any surgical procedure, the risks and benefits of the procedure must be carefully assessed for each individual patient. The risk to the patient of not having surgery must also be addressed. In the case of progressive scoliosis, the risks of not having surgery if brace treatment has failed or is not indicated have been suggested above. These involve progressive deformity of the trunk with potential imbalance and offset between the trunk and pelvis when in an upright position. In addition, with thoracic curves, risk to the heart and lung function develops when curves reach 80-90°. This results from compression of the lung within the deformed chest cavity, which makes it difficult for the heart to pump blood through the compressed lung. Most cases of scoliosis are treated prior to curves reaching such a magnitude.

Blood Transfusion:

The specific surgical risks are dependent on multiple factors, including the general health of the patient, any underlying medical disorders, as well as the type of surgery that is required. In general, with all operations around the spine there are potential complications relating to the anesthesia, as well as risks of bleeding and wound infection. With anticipation preoperatively, blood loss can be managed by one of several means, depending on the anticipated magnitude of the blood loss. These include pre-donation of the patient's blood (autologous) or of blood from family members or friends (donor specific). An additional option involves the use of banked blood components. Intraoperative blood conservation techniques can be utilized with either use of hemodilution or blood salvage with recirculation methods to minimize need for other forms of blood replacement.

Neurologic Risk:

Additional potential intraoperative complications relate to neurologic injury. Surgery involving the spine places the neural elements of the spinal cord and nerve roots at some risk for injury. Injury to these structures may result in a spectrum of disability that spans from mild numbness to loss of bowel or bladder function to complete paralysis below the area of injury. The risk of spinal cord injury can be minimized, although not completely eliminated, by use of sophisticated spinal cord monitoring systems. This spinal cord monitoring is performed during the exposure and correction of the spinal deformity and monitors electrical function of the spinal cord throughout the surgical procedure. When indicated a "wake-up" test may be utilized to ensure normal neurologic function. This involves decreasing the level of anesthesia, after completion of the deformity correction, to the point that the patient is able to hear and follow commands. The patient is instructed to move the lower extremities to confirm that voluntary motor function remains present. If normal function is observed, the procedure is completed with wound closure. If spinal cord functional problems are noted with spinal cord monitoring and confirmed by abnormality appreciated on a wake-up test, removal or adjustment of the instrumentation may be required. This decreases the stress applied to the spinal cord by the instrumentation system, improving the likelihood of neurological recovery.

Postoperative Complications:

Postoperatively, complications may also arise. These include development of a wound infection, pulmonary difficulties, as well as problems associated with the hardware system and fusion. Postoperative wound infections occur in roughly 1% of patients. Measures taken to minimize this risk include the use of antibiotics before, during, and after the operation. There are risk factors that increase the likelihood of wound infection, which include poor nutritional status, history of urinary tract infections and revision surgery. In most cases

wound infection can be managed by a return to the Operating Room with cleaning of the wound and closure of the wound over drains, along with prolonged antibiotic use. In rare cases there is a delayed presentation of infection, which may come as late as 1-2 years postoperatively. In these cases, generally the treatment is cleaning of the wound and removal of the hardware. These delayed infections can be successfully managed with hardware removal and antibiotics.

Complications related to the instrumentation system and fusion are possible. In occasional instances, there is loss of fixation of the rod system to the spine, resulting in some loss of correction. This may require revision of the instrumentation system with replacement of hardware and reattachment to the spine. Additionally, there may be difficulties in obtaining fusion at each of the involved levels of the spine. If an area of nonunion (lack of fusion) or pseudarthrosis develops, there may eventually be breakage of the rod system. In many circumstances this does not cause further problems. However, in some cases it may require revision of the instrumentation system if there is pain or evidence of curve progression.

WHAT IS THE RECOVERY PERIOD?

The recovery after scoliosis surgery is variable, depending on the nature of the surgery performed. In general, after having 2-8 or more hours of surgery, time is spent initially in the Recovery Room. In this area the patient is allowed to wake up from the anesthesia and regain voluntary breathing. There are times when it is necessary to maintain respiratory support on a ventilator for one or more days following this surgery. If this is required or there are other indications to do so, the initial postoperative period may be spent in an Intensive Care Unit. This allows for ventilatory support of the respiratory system, as well as close monitoring of the cardiovascular system.

Pain Management:

There are several options for managing pain in the initial postoperative period. These include intravenous narcotic medicines, such as morphine or Demerol. Once the patient is drinking/eating, the transition from IV to oral pain medication will be initiated. The intravenous route is convenient and can be delivered either by the nurse or with a patient-controlled anesthesia (PCA) system. This allows the patient some control over the timing of administration of pain medication.

The 1st Day After Surgery:

In the initial postoperative period, there may be several tubes in place. The tubes will be placed while the patient is under anesthesia. These may include:

1) A nasogastric tube to drain the stomach contents, the tube will be in place while the intestines "wake up" or recover from the effects of anesthesia usually 1-3 days. After the intestines start to work again (bowel sounds and flatus present), the patient may then start to drink and eat again. Generally the diet is advanced gradually, starting with liquids and progressing to solid food.
2) There will be a bladder (urinary) catheter inserted during surgery.
3) Surgical drains may be placed in the back to allow for drainage of fluids.
4) If surgery in the chest area is performed, a chest tube will be required to drain fluid from the chest (thoracic cavity).

5) Intravenous lines will be in place to administer both medications and fluids. There may also be an arterial line for the same purpose, as well as to assist with blood pressure monitoring.

These monitoring lines and drainage tubes will be removed sequentially in the days following surgery when they are no longer necessary.

Getting Out of Bed:

In the first day or two following surgery, there is little physical activity required of the patient, though the nurses will be repositioning the patient in bed every 1-4 hours. There will then be a gradual increase in activity/mobilization, generally starting with sitting, then standing, then resuming walking. The timing and rate of the increase in activity will vary, depending on the type of surgery done. Depending on the patient and/or the type of surgery performed, some patients may need to wear a brace for 3-4 months after surgery. The brace provides external support to the trunk while the fusion is healing. This brace comes in various forms and the form will depend on the patient and type of surgery performed. This brace is to be worn at all times except when in bed. This brace will be fit and provided to you prior to discharge from the hospital.

Going Home:

In general, requirements for being discharged from the hospital following surgery include:

1) Tolerating oral liquids and food.
2) Comfortable on oral pain medications.
3) No fevers.
4) Ability to ambulate (if ambulating prior to surgery)
5) Able to urinate and have bowel movement.

After discharge from the hospital to home it usually takes several weeks for pain to completely resolve and in general, a gradual weaning from narcotics to Tylenol takes place naturally. It is not uncommon for patients to be comfortable on Tylenol during the day and need narcotics at night for comfort during this weaning process.

In general, school age children/teens are ready to return to school 3-4 weeks after surgery, depending on the child and the type of surgery. Some children tolerate a return to school for just half days initially, but we encourage returning as soon as possible to a "normal" schedule.

Limitations in Activity After Surgery:

Physical activity and sports are restricted after surgery for 6-12 months, depending on the type of surgery. We encourage children to begin walking for exercise (if child is ambulating prior to surgery) at the time of discharge. Bending, twisting, and lifting activities are to be avoided. At each follow-up visit, x-rays will be done to access healing of the fusion. This will allow your surgeon to make decisions regarding easing off on both bracing and activity restrictions. As healing occurs, brace wear will be discontinued, and permitted activities may be increased. Generally at 1 year following surgery, children may participate in all activities without restriction.

SUMMARY

Scoliosis is a curvature of the spine that most commonly affects adolescent girls. In some patients the deformity is progressive, necessitating treatment. Initial forms of treatment in the patient with remaining growth involve the use of a brace or orthosis. In some cases this is unsuccessful or the curve magnitude too large for this to be effective, in which case surgical treatment is required. In either case, the goal of treatment is to limit progression of the deformity of the spine and trunk and obtain some correction of the deformity.