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## Scoliosis

The spine normally forms a straight vertical line when viewed from the back.



Scoliosis is an abnormal curve of the spine, most commonly affecting the spine in the chest area and lower back region.

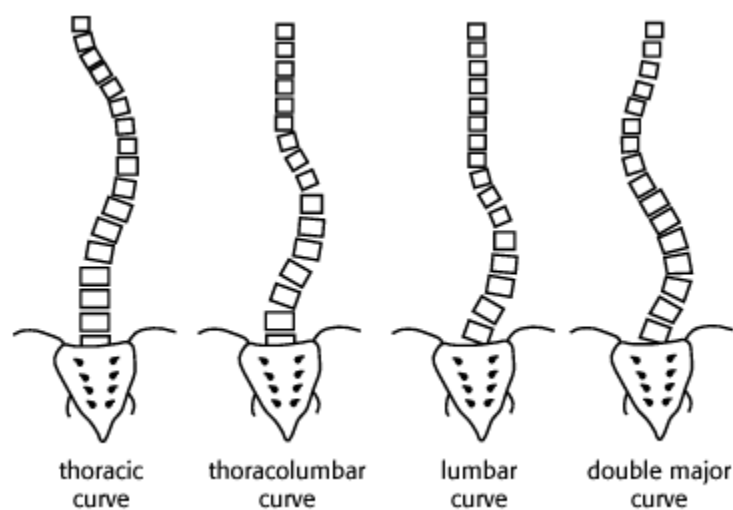
### What are the symptoms?

Scoliosis is usually asymptomatic, it gradually develops during childhood or adolescence.

However some signs may become notable:

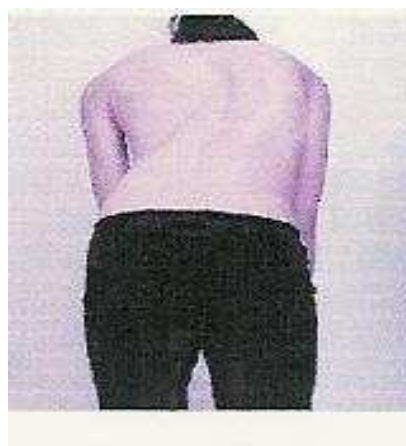
- 1) Visible curvature of the spine to one side, which is more obvious when bending forward.
- 2) One shoulder blade may appear more prominent than the other
- 3) Uneven waist causing clothing not to fit properly—uneven hemline or one trousers leg appearing longer than the other
- 4) One hip is higher than the other,
- 5) Body appears to lean to one side

Scoliosis affects 2 of every 1000 children. Most scoliosis is called idiopathic scoliosis because the cause of it is unknown. This type of scoliosis occurs most commonly in adolescents, especially girls, but may present at any time before the end of growth. Most adolescent idiopathic scoliosis do not progress and can be followed with observation until the person has finished their growing. It is unusual for these curves to progress after the growth spurt. However, scoliosis occurring in younger children, or complicated by another medical condition, has a higher probability of progression.



Patterns of scoliosis

The curves above show the 2 dimensional movement of the spine. Most scoliosis is also associated with rotation of the vertebra (bones in back) and this causes a rib hump see picture below.



## **Scoliosis and Friedreich's Ataxia (FA)**

Scoliosis in FA generally behaves like idiopathic scoliosis mentioned above but is more **progressive**. When a FA sufferer has finished growing, the curve will continue to grow. This is different from idiopathic scoliosis. In FA muscles are affected. The muscles in the back have a lot of influence on the position of spine. When muscles on one side of the spine become weaker than on the other side of spine, the stronger muscles "pull" the curve. The direction of the curve will indicate where the muscles are weakening.

Scoliosis can be the presenting sign of FA but this is unusual. FA usually presents over a long period with clumsiness of feet and hands. FA usually affects a sufferer's ability to run and balance initially. Later handwriting and walking are affected. Some people might present with shaking in their arms, cramps or spasms or numbness in their legs.

Symptoms of scoliosis in FA **include those mentioned earlier and** the following:

backache after physical activity or pain in his/her side and  
starting to lose balance in the direction of the curve.

### **What happens to the scoliosis curve at puberty?**

The curve of scoliosis worsens in the 12 months before girls have their 1<sup>st</sup> menstrual period and in the 3 months after the 1<sup>st</sup> menstrual period. Recording of height will help identify this period. During this time there is a significant risk of progression. Girls who have scoliosis and FA should be followed more closely during this time of their development. Otherwise a 6 monthly review is adequate but your doctor may decide to review you more often at puberty. Puberty in boys usually occurs between 12-15 years and last 3-4 years.

### **What can be done to prevent scoliosis from progressing?**

**Regular exercise:** While regular exercise can only be good for the child, there is no evidence to suggest that any form of exercises have any effect on scoliosis.

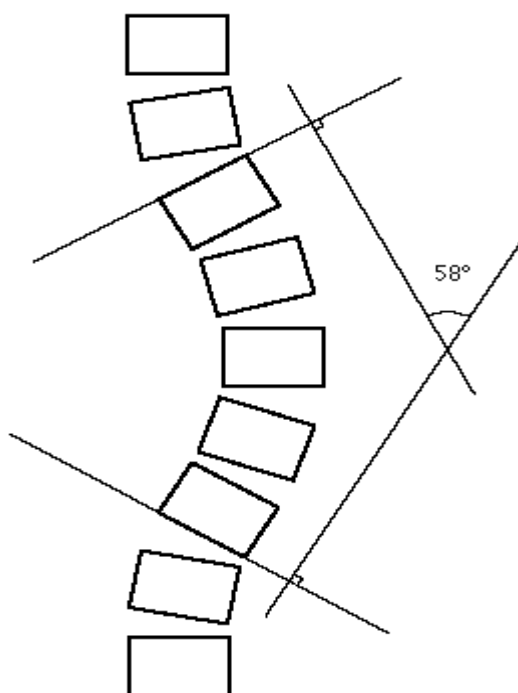
## What can be done to prevent scoliosis from progressing continued:

**Bracing:** The value of Bracing in the treatment of scoliosis is also hotly disputed. Some authorities believe that the brace can keep the curve from worsening if worn up to 23 hours a day. It is generally accepted that bracing will not reduce the curve and on stopping wearing the brace the behaviour of the curve is unpredictable. There is a further difficulty with bracing scoliosis in FA, in that it further restricts movement, which is not desirable for FA sufferer. In the long term, it does not prevent progression. However, not all orthopaedic surgeons agree on the treatment of scoliosis and some FA sufferers are still recommended to wear braces.

**Chiropractors:** These alternative practitioners are sometimes consulted to help back problems. Chiropractors themselves would say that their success in treating curves greater than 30 degrees is poor and no medical studies are available to support that any benefit can be gained from attending chiropractor with scoliosis.

**Surgery:** Surgery can help prevent scoliosis from progressing. It is recommended for

- a) Those whose curve is 45-50 degrees and continuing to progress
- b) Those whose curve is greater than 50 degrees



Cobb method of calculating the angle of scoliosis

### **The purpose of surgery is primarily:**

1. To fuse the spine that is balanced in the front to back and side to sides planes;
2. To prevent further progression of the curve. If a curve is allowed to progress to 70-90 degrees, it will not only result in a very disfiguring deformity, but will start to result in cardiopulmonary compromise. As mentioned earlier the spine also rotates in scoliosis and this further closes down the space available for the lungs and heart
3. To improve an undesired cosmetic appearance. The effect of straightening the spine, reducing the rib hump and balancing the head and trunk over the feet, is not a trivial affair. Un-operated scoliosis is a significant deformity which can make the child very unhappy
4. To aid respiratory muscles by giving a firmer structure - the fused spine to work with, which in turn will reduce the child's oxygen requirements (less work required to sit, stand or walk)
5. To relieve discomfort and postural fatigue. The scoliosis associated with FA can give rise to significant pain in the long-term. As the vertebrae press against each other and the nerves get pinched, correction of scoliosis ensures proper seating in the wheelchairs

### **Differences in surgical technique**

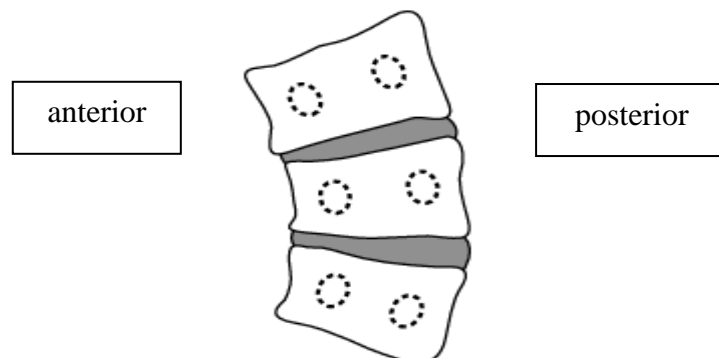
There are significant differences in surgical technique for the different spinal deformity, but the basic principles can be applied to almost any case:

1. mobilisation of the rigid scoliosis curve and rib hump deformity,
2. correction to as near as possible, normal anatomical position,
3. holding this new position with 2 long rods and inserting screws to facilitate bony fusion between the vertebrae. This maintains this position for life.

### **Mobilising the spine:**

As the spine and ribs have grown in this position, they fit together extremely well and are not readily changed into a more cosmetically acceptable alignment. Each vertebra within the structure of the curve is abnormally shaped both in its anterior and posterior elements.

Thus, part of the surgical technique must be to reshape these vertebrae individually, and to release the intra-vertebral structures to allow realignment.



The anterior element requires osteotomy (removal of bone), most commonly performed posteriorly, to shorten the elongated convex laminae (tiny layer of bone) and release the vertebrae. This bone can be placed in between the vertebra to help fusion of the spine. The posterior element is usually done by anterior discectomy (removal of disc), and a posterior release by excision of the posterior facet joints and inter-spinous ligaments. Having thus reshaped and loosened the spine, internal fixation devices are attached to the vertebrae.

**Details of fixation:** The principle is to use some form of fixation in the proximal part of the curve, either pedicle hooks or screws or claws, and a combination of either pedicle screws, +/- laminar hooks in the distal part of the spine, which have been attached to rods which are inserted, one on each side of the spine. Many different instrumentation systems are available for achieving this and there seems to be little difference between the different types available. Some are slightly easier to use than others.

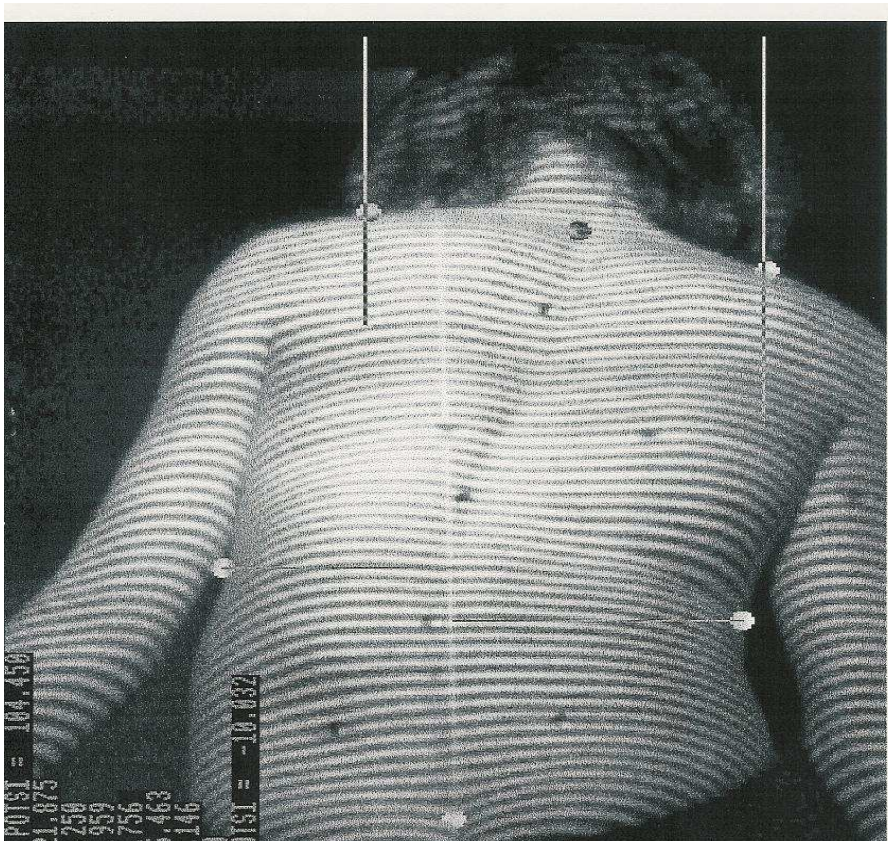
This x-ray shows the rods in situ over the spine.

The pedicle screws are clearly visible



**New position:** The patient is encouraged to stand 2 days after surgery and generally leaves hospital 1 week after surgery. It may be possible to get back to school after 6-8 weeks. Over the subsequent 6 months this bone graft becomes incorporated and the corrected area of the spine is fused into one segment. The patient can then resume contact sporting activities and horse-riding. Subsequent removal of implants can be carried out, generally, after 1 year and a half, if this is deemed necessary. However in most situations this is not required, as the high grade titanium or stainless steel implants that are used do not appear to cause any long term problems.

### What can go wrong?



This picture shows problems associated with just putting in rods and not reshaping and loosening the spine.

If surgery is done without releasing the anterior part of the vertebra, the front of the spine will continue growing while the rear cannot, causing an arched back and lead to further difficulty maintaining balance.

## **Risks of surgery:**

Scoliosis surgery is major surgery. It is often carried out in 2 stages, i.e. 2 operations with a 2 week interval between them. It involves intensive care nursing after both operations. The risks as with any surgery are significant (death, paraplegia or lesser nerve damage,) and must be carefully balanced against the perceived advantages. Despite all of the dangers, it is better done early, before deformity and respiratory compromise is severe, if it is to be done at all.

## **Who makes the decision for surgery?**

This is ideally made by the child who is to have the surgery in conjunction with her surgeon and family. However this may not always be possible.

Factors to be taken into consideration:

- 1) A child's underlying medical and psychological condition,
- 2) What benefit the child can expect from surgery, which is a major undertaking- e.g., a child with severe cerebral palsy who has no awareness of her appearance will not benefit, although her carers may push quite hard. On the other hand, a child who is less affected may benefit psychologically,
- 3) Spinal surgery may interfere with her ability to walk, so in FA, you have to balance any deterioration that a child may suffer in their ataxia against possible gains from straightening the spine,
- 4) Straightening the spine may limit full back movements. Usually the patient does not find this reduction of back movement interferes with the activities of daily living. Is it better to achieve trunk balance, which can improve mobility, and a more normal appearance which is a cosmetic effect and sacrifice some spinal movements?
- 5) There is no evidence to support the commonly held belief that spinal surgery will improve respiratory function from causes where the lungs did not grow properly but (as said earlier) it will help in the long term by giving the respiratory muscles a firmer structure to work with thus the child's requirements for sitting, standing and walking are reduced.

**This article was prepared by Dr Mary Kearney and reviewed by Dr Caroline Goldberg, Scoliosis Research unit, Our Lady Hospital for Sick Children, Crumlin, a major teaching and University hospital in Ireland. euro-ATAXIA is very grateful for her assistance.**

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Websites on Scoliosis –

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<http://www.scoliosis.org> ,

<http://www.srs.org>

[www.pediatric-orthopaedics.com/Treatments/Scoliosis/scoliosis.html](http://www.pediatric-orthopaedics.com/Treatments/Scoliosis/scoliosis.html)

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