



## Syndromic scoliosis

### Cause

Syndromic scoliosis is diagnosed when the curvature is part of a recognised disease pattern, such as is seen in Marfan's syndrome.

### Diagnosis

A patient will be diagnosed with a syndrome in their early years. The medical staff monitoring their care will be aware that scoliosis is likely to occur. They will therefore be checked regularly for signs of curvature in their spine.

### Treatment

Dependent on the child's age, the treatment is likely to be similar to that of idiopathic scoliosis. In addition, however, because some patients will not be mobile, appropriate seating may also be used in an attempt to slow the progression of the curvature.

- Monitoring

In some cases no intervention will be necessary and the child will simply be monitored by a scoliosis specialist.

- Casting

In many cases of infantile idiopathic scoliosis the child's spine may need to be guided into its normal position as the child grows. This positioning is currently done by applying an external brace to the torso made out of a combination of plaster-of-Paris and modern casting materials. The cast is worn permanently and cannot be removed. Casts need to be made in a very specific way with a hole in the area of the chest to allow for the lungs to expand, so ensuring that the child sees a specialist who is knowledgeable about infantile scoliosis is essential.

In children less than age 2 years with the more benign type of curvature for whom the goal is curing the scoliosis, the cast will be changed under anaesthesia every 2-3 months with the aim of achieving a straight spine. Despite extensive casting a removable brace may still be needed after this treatment.

- Bracing

If the curve is progressive and the child is still growing the specialist may want to place the child in a cast or brace. Rarely does a brace permanently correct scoliosis; instead the goal of bracing is to allow the child to grow before a more definitive procedure is done. On average braces need to be worn 23 hours per day and are generally removed only for bathing and special occasions. As the child grows, new braces will need to be made. A permanent brace can sometimes be fitted. This is known as a plaster cast. Many parents find it preferable to braces, eliminating the problems of compliance and the difficulties of donning braces in uncooperative young children.

- Surgery

If the child's curve worsens despite bracing or casting, an operation may be necessary. The operation would probably involve the scoliosis specialist inserting growing rods. This growing-rod operation allows for continued controlled growth of the spine. In general, the rods are attached to the spine above and below the curve, usually correcting the curvature by 50% at the time of the first operation. The child then returns every 6 months to have the rods

lengthened by about 1 centimeter to keep up with the child's growth. This procedure is usually done through a small incision, and takes place in an outpatient clinic. Most children will have to wear a brace to protect the instrumentation. When the child becomes older and the spine has grown, the doctor will remove the instrumentation and undertake a final spinal fusion operation.

In some cases surgery will not be possible because of other health conditions relating to the syndrome.

### **Prognosis**

For the individual with scoliosis secondary to a syndrome, the prognosis after surgery should be permanent control of the curve in whatever safe position of balance the curve could be placed in relation to the individual's other health needs. In conditions with progressive neurological changes the scoliotic curve might change or a further curvature could develop, despite successful surgical control of the original curve.