Scoliosis and spinal surgery

The spine (backbone) is made up of a line of small bones called vertebra stacked on top of each other and running from the base of the skull to the coccyx (tailbone). The vertebra range in size from 2-3 inches in the neck to 5-6 inches in the lumbar region of the lower back and are numbered according to what position they are in (cervical, thoracic, lumbar, sacral or coccygeal). The spine supports our skull and other bones and allows movement, as well as housing the spinal cord which supplies nerves to our bodies.

Scoliosis is a deformity of the spine where the vertebra twist on each other, causing the spine to curve to the side instead of staying in a straight line. Signs of scoliosis may be uneven shoulders or having one hip higher than the other, and slumping to one side. Scoliosis can occur in otherwise healthy people but is relatively much more common in people with neuromuscular conditions, and progression of the deformity is more common and more severe when there is a neurological condition associated with it. 60% of people with Friedreich's ataxia are affected by scoliosis to varying degrees.

The deformity is thought to occur because of weakness in the muscles around the spine. It is not caused by bad posture or diet or by wearing backpacks.

Whilst mild scoliosis may cause no problems and require no treatment beyond monitoring, severe scoliosis causes long term disablement as it impairs the ability to sit and affects mobility, and may lead to back pain. If the spine curvature becomes more extreme, it may also constrict the lungs, causing breathing difficulties, and put pressure on other organs including the heart.

Braces can be used to help support the spine and to minimise progression of the curve during growth of the spine but braces do not reverse or cure the deformity. If correct supportive treatment is started early in children with scoliosis bracing can delay the need for surgery. Modern braces are available which are less visible under clothes and made of flexible materials which conform to the body, although they still feel awkward and uncomfortable at first.

Surgical stabilisation is the only way to correct severe deformity. The decision to carry out surgery may be discussed when there is pain or the curvature is causing problems with breathing. It is a long operation (5-6 hours) that requires a full general anaesthetic so can have serious risks, as with any serious operation, including infection, blood loss and nerve damage, and risks associated with anaesthesia.

During the operation, the vertebrae are fused together to straighten them and rods may be inserted along the side of the spine to reinforce the position and prevent curving. Usually the surgery will be carried out through an incision made in the back, which will produce a long scar. The surgeon will insert rods, and use screws, hooks or wire to fuse the vertebrae together and this metalwork will remain in the spine permanently. After the bones have been fused together their growth will be limited so the operation cannot be carried out too early or uneven growth may occur. Children who undergo the operation may find themselves suddenly several inches taller because their spine is straightened out.

Recovery may take 6-7 months to 1 year. Having undergone a long operation, doctors know that people may feel worse afterwards, and may take some time to recover mobility that they had before the operation. As Friedreich's ataxia is a progressive condition, the long recovery time with impaired mobility can mean that people never regain the mobility they used to have. However back pain is usually reduced and being able to sit straight in a wheelchair improves comfort and control and reduces complications such as pressure sores from uneven weight distribution.

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Katie Henderson (24) was diagnosed with Friedreich's ataxia when she was eight years old and a year or so later discovered that she also had scoliosis. Here, she tells her story.

My scoliosis was identified after I fell and hurt myself whilst playing at a friend's house. I was in a lot of pain so my mum looked at my back to check for injuries and noticed my spine was curved. She assumed this had happened when I fell so she rushed me to casualty where I had x-rays taken which revealed the scoliosis. The doctors were astounded it hadn't been picked up before. I was referred to an Orthopaedic Consultant at the Royal Orthopaedic Hospital in Birmingham.

I had two curves which pushed my left hip and right shoulder blade out. I was fitted with a back brace and was told to wear it all day, every day. The brace was a very heavy and rigid plastic full-length corset, held tight with straps at the back. It was like stuff nightmares are made of, and I can still remember how tight and smothering it felt. And so I avoided wearing it and over the three years it spent far more time in the cupboard collecting dust than fulfilling its purpose.

Maybe as a consequence my scoliosis progressed quickly and it got to the point where the tops curve was pressing on my lung which was making breathing difficult. When I was 12 I had surgery to stabilise my condition. There was no option to choose who performed the surgery or where it was done, but it turned out that my consultant at the Royal Orthopaedic Hospital was an expert in the scoliosis procedure and had many years experience.

The surgery involved two different procedures, which took place two weeks apart. In the first procedure some of my rib bones were removed and used to replace the cartilage in my spine, making it solid. Originally, the second procedure was planned to happen a week after the first, but two days into my hospital stay I came down with German Measles and this slowed things down a bit as I needed extra time to recover from that. But two weeks later I was good to go and in the second procedure two metal rods (called Harrington rods) were fixed to both sides of my spine to hold it straight.

Throughout my time in hospital and in the weeks and months after leaving I did really struggle with the pain I was in. A family friend was a trained Bowen practitioner and this worked miracles in helping me to recover enough to start back to school, which I was away from for four months in the end.

All in all the surgery was successful and although it was four weeks of hospital and not-very-niceness, it was something that had to be done as it would have been dangerous not to intervene. My scoliosis has stayed stable since the surgery and I'm a lot straighter than I once was (I grew two inches in height immediately after the surgery).

If I had my time again I'd learn to put up with the brace because I don't think, if I had stuck with it, my Scoliosis would have progressed as much as it did. The curves in my spine, and the unevenness of my hips and shoulder blades, are still noticeable.

The rods themselves cause no problems, although they do greatly restrict movement in my trunk and I cannot bend or twist my back. The surgery lead to me using a wheelchair more than I had up until then.

More information

A summary of the whole experience from deciding to have the surgery to longer-term recovery, written from the point of view of two parents can be found on the website of the Friedreich's Ataxia Parents Group (http://www.fortnet.org/fapg/scoliosi.htm). This American site takes a no-holds-barred approach with very honest accounts of procedures and the progression of ataxia which some people may find distressing. They also provide email support between parents of children with Friedreich's ataxia.

Katie Henderson's own website is an inspiring and informative account of her personal journey and experiences whilst living with Friedreich's ataxia. Her website can be found at: http://www.onbother.co.uk/index.html