Scoliosis in Osteogenesis Imperfecta

What do you Need to Know?

When you look at someone from front to back, the spine should be straight with the head centred over the pelvis. Scoliosis is a bend in the spine which is seen when looking at someone front to back when the spine is no longer straight. This factsheet describes the different types and causes of scoliosis, the problems a scoliosis can cause, how it is assessed and can be treated.

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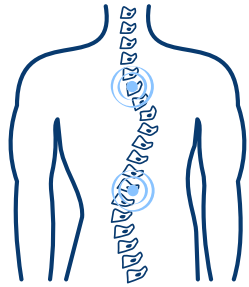

There are many different types and causes of scoliosis. Scoliosis is also associated with the diagnosis of Osteogenesis Imperfecta (OI). The reasons why scoliosis occurs in OI are not clear but will be related to:

- Fractures of the vertebral bodies (the blocks of bone that make up the spine).
- Weakness in the muscles around the spine, allowing the spine to bend as it is not being held upright as it should be as the muscles are not holding the spine upright.
- Periods of significant growth (such as the adolescent growth spurt) where the scoliosis can increase in size.

Why Does Scoliosis Matter?

Scoliosis matters for several reasons, which are related to the age that the scoliosis develops. When a scoliosis develops in the early years of life, then it can have implications for the development of the chest and the underlying heart and lungs. Once the heart and lungs have developed fully (and children are older than 10) then scoliosis is more around the difficulties associated with a shape that is not symmetrical, which can lead to some restrictions in mobility, be a cause of some pain and cause issues in comfort when sitting, particularly if the individual concerned is a wheelchair user. Whilst there is truth in the old wives' tale that scoliosis can prevent the heart and lungs from working properly, this is only for large curves in young children and is an uncommon presentation of scoliosis.

Scoliosis is assessed by using x-rays of the spine. For preference these x-rays will be taken in a standing or sitting position as this is the position where gravity will make the spinal curves as bad as they can be, such that an assessment is accurate.If the individual is unable to stand or sit then a lying down x-ray will be sufficient. It is worth noting that a good assessment of the shape of the spine can be obtained from the bone density measures that will be being undertaken on a regular basis in a similar fashion to a lying x-ray. Some centres are using these interchangeably.

The management of scoliosis is around the identification of a curve and then monitoring of that curve. The presence of a scoliosis does not mean that something must be done. A referral to a spinal unit will be made by an Osteogenesis Imperfecta doctor when there is a large curve, a curve that is getting bigger with time or the curve is causing the child symptoms that might benefit from some intervention. A scoliosis surgeon will make an assessment of the child in their totality including their diagnosis of Osteogenesis Imperfecta, their treatment with medications, their bone strength and the trend of development of the scoliosis curve.The reasons to do something are to prevent things from getting worse, get a more symmetrical torso and trunk for the comfort of the individual and in the small child guide growth to maximise heart and lung development.

Whilst in some types of scoliosis there is a role for the use of a brace or a plaster cast round the torso this is not something that is routinely done in OI due to the effects of pressure on the chest and possible rib fractures.Surgical intervention is not particularly common. In the early years growing spine technology can be used, and the use of growing rods and MAGEC rods have been reported with some success. More commonly it is a correction and fusion of the spine to improve shape and prevent progression and this is done with implantation of screws and rods.

How Is Surgery Performed?

All procedures are performed under general anaesthetic and will be performed through an incision that runs down the length of the back. In particularly fragile bone there are techniques that can improve the hold of the screws in the bone such as the use of bone cement.Invariably young people are in hospital for between 7 and 10 days following this procedure. The spine will be well fixed and the time in hospital is spent mobilising and getting over the operation before the person can go home.There is certainly a role for getting ready for the operation through the use of pre-habilitation with physiotherapy and certainly physiotherapy and rehabilitation after the surgery is very important. Once the surgery is performed then follow up with the spinal surgeon will occur on a reasonably regular basis for 2-3 years to make sure that all has healed.

It is of note that surgery does come with some risks and these include:

- The fusion not healing.
- The need for further surgery at some point.
- Ongoing symptoms because of pain or further spinal deformity.
- The risk of infection, because metalwork is being implanted into the body.
- The risk of neurological injury.

Neurological injury is damage to the nerves and/or spinal cord during surgery which would result in paralysis in worse case. It is fair to note that this is a very uncommon complication because surgeons are very careful in making sure that the spinal cord is looked after during the surgery, and because there are techniques to monitor how the spinal cord is working during the surgery. It has also been reported that there is a risk of blindness associated with surgery to do with being face down for a long period of time. This is incredibly rare but again, the surgical teams are very conscious of this and make every effort to make things as safe as possible.

It is worth noting that spinal surgery with rods and screws does lead to loss of movement and some stiffness. It may be necessary to extend the rods and screws down into the pelvis to get the correct fixation and position following the treatment of scoliosis.This results in a reasonable amount of stiffness and loss of flexibility however, this does not seem to lead to a loss of function.

The National Charity for Scoliosis in the United Kingdom is Scoliosis Association UK (SAUK). They are a charity that deals with support of individuals and their families who have a diagnosis of scoliosis from all causes. Whilst this guide has hopefully been of some use in explaining the scoliosis from an OI point of view, SAUK has lots more information along with the point of view of those that have scoliosis and those that have been treated for scoliosis. Further information on this can be found on their website at www.sauk.org.uk

Notes

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Compiled by the Brittle Bone Society in collaboration with BBS Medical Advisory Board and POINT (Paediatric Osteogenesis Imperfecta National Team). The information in this leaflet is correct as at 31st July 2022 but we cannot guarantee that it will be accurate and current at any given time. This leaflet is not intended in any way to replace the advice of your doctor or other medical professional. Leaflets are available online at www.brittlebone.org. This information is available in accessible formats on request.

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