About Neuromuscular Scoliosis

Neuromuscular Scoliosis represents an abnormal lateral curvature of the spine that develops due to an abnormality of the neuromuscular axis (brain, spinal cord, skeletal muscle). There are a wide variety of conditions that affect the neuromuscular axis that can lead to scoliosis, which makes this a very diverse population. In general, the more severe the neuromuscular condition, the more likely the child is to develop a significant progressive scoliosis that may require surgical intervention. Children who are able to walk under their own power, or maintain their truncal strength, have a much better prognosis than children who spend most of their time in a wheelchair.

The most common neuromuscular disability of childhood is Cerebral Palsy (CP). In CP, there is an insult to the brain at an early stage of development that leads to progressive musculoskeletal impairment as the child grows and develops. Some children experience very mild impairment and are able to walk independently, while other children experience severe impairment and need a wheelchair for transport. This impairment is classified based on the Gross Motor Function Classification System (GMFCS) from 1 to 5. Children at GMFCS 1 are very unlikely to develop scoliosis, while children at GMFCS 5 have a greater than 90% chance of developing scoliosis.

At the level of the spinal cord there are several classical conditions that result in neuromuscular scoliosis: traumatic spinal cord injury, spina bifida/myelodysplasia, and spinal muscular atrophy (SMA).

In the setting of spinal cord injury, the younger the patient and the higher the level of the cord injured increases the risk of scoliosis. Patients under the age of ten with a complete cervical spinal cord injury have almost 100% risk of developing a progressive scoliosis.

Similarly, the risk of scoliosis in patients with spina bifida is dependent on the level of their neurologic impairment. It is rare to see a significant scoliosis in patients with a sacral level to their spina bifida, unless it is combined with a component of sacral agenesis. Scoliosis is very common in patients with a thoracic level spina bifida. Treatment of scoliosis in the setting of spina bifida can be extremely challenging because of the lack of bony elements at the back of the spinal column. There is a high risk of infection due to the poor soft tissues overlying this neural tube defect.

SMA is a disease that results in the progressive destruction of nerve cells in the spine that are responsible for controlling muscle function. The infants and children are floppy or low tone, and may regress in their motor milestones. The traditional classification of SMA was based on the age of onset and diagnosis, which correlated with the severity of the disease. In the past five years, novel gene therapies (i.e Nusinersen) have significantly improved the prognosis, such that neonatal screening for SMA is being introduced in Australia. Similar to other conditions, when a patient ends up in a wheelchair early in life, the risk of progressive scoliosis increases.

The most common condition affecting skeletal muscle leading to the development of scoliosis would be muscular dystrophy. There are several types of muscular dystrophies and myopathies, but the classic is Duchenne Muscular Dystrophy (DMD). DMD is inherited in an X-linked recessive pattern resulting in a condition that is almost exclusively seen in young males. It results in progressive global weakness through childhood that eventually impair mobility such that patients require a wheelchair. With the introduction of steroid therapy, DMD patients now remain ambulant for much later into life and thus the development of severe scoliosis has been significantly reduced. Again, the earlier in life a patient becomes non-ambulant, the more likely they are to develop a progressive scoliosis.

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There is a very common theme in neuromuscular scoliosis. The ability to walk independently provides a good prognosis. In order to walk, the brain, spinal cord, and muscles must coordinate to keep our head balanced over our pelvis, allowing us to stand up without falling over. If one, or more, of those components stops working properly during childhood, then the normal forces that guide the growth of a straight spine don't function correctly. This is exacerbated by forces that cause contractures of the hips and lower extremities resulting in

tilting of the pelvis and further tilting of the trunk. Utilising a wheelchair early in life provides a poor prognosis for both the development of scoliosis and progressive hip subluxation and dysplasia.

The treatment of neuromuscular scoliosis is based on one basic principle; getting the head and shoulders balanced over a level pelvis. In wheelchair patients with a tilted pelvis, instrumentation often is required down to the pelvis to create a level foundation upon which to build the rest of the spine. There are many techniques of fixation to the pelvis in this scenario. Long spinal instrumentation from the top of the chest/ thoracic spine down to the pelvis requires long procedure time, with a significantly increased risk for large blood loss requiring transfusion, and increased risk of wound infection. A deep infection/ metal infection may require surgical washouts and extended antibiotics.

Another factor that is always challenging to manage is growth. Spinal column height/ growth is necessary to allow a child to develop normal adult organ function, in particular development of the chest cavity and lungs. Restricting growth of the chest cavity has been shown to have permanent effects on lung function. Unfortunately, progressive severe spinal deformity can also severely restrict lung capacity. Non-operative options such as casting and bracing tend not to be effective in the neuromuscular population, and have their own set of complications because of the rigidity of the brace required to control the curve. There are several growing rod/ surgical options, but the rate of complications of growing rods in neuromuscular scoliosis approaches 100%. Newer technologies are emerging that will hopefully reduce this complication rate, but this group of patients with severe neuromuscular conditions and early onset scoliosis (<10yo) will probably remain one of the most challenging patient populations to manage.

Regardless of the surgery performed for the scoliosis, patients with neuromuscular disease require a strong multidisciplinary team to optimize their care and assist with decision making. Along with the spine surgeon, a paediatric orthopaedic surgeon with an interest in neuromuscular disease will be critical to help manage the hips and other limb deformities that will likely accompany the scoliosis. Medical teams involving Rehabilitation Medicine, Respiratory/ Sleep Medicine, Cardiology, Intensive Care and Anaesthesia will be important to help optimize perioperative decision making. Many allied healthcare providers including physiotherapy, occupational therapy, and orthotics will play important roles. It cannot be overstated the importance of the whole team that will need to be involved to help provide the best care to these patients with neuromuscular conditions and their families.

With all of these risk factors, many have questioned the benefits of surgery in patients with neuromuscular scoliosis. Several studies have shown that correction of the scoliosis in patients with neuromuscular disease provides improved quality of life and significant health benefits (improved lung function and gastric emptying) that lasts for years following surgery.

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