Neuromuscular scoliosis: clinical presentation, types of deformity, assessment and principles of treatment

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Abstract

Scoliosis affects very commonly children with neurological or myopathic conditions. It can be associated with increased kyphosis or lordosis. The coronal curve extends to the sacrum producing pelvic obliquity. The deformity develops due to poor muscle control and spasticity as the spine cannot resist forces against gravity. It can produce severe problems including spino-pelvic imbalance causing back pain, costo-pelvic impingement pain on the concave side of the curve which can both affect sitting ability and posture, as well as respiratory complications and difficulties in provision of patient care. There is no conservative measure that can stop deformity progression. Surgical intervention is indicated in the presence of a progressive and symptomatic deformity which affects the patient's quality of life. The degree of neurological disability and associated co-morbidities must be taken into account during decision-making as these are directly correlated with the risk of perioperative complications. Thorough preoperative assessment should be undertaken by a multidisciplinary medical, surgical and allied health professional team in a centre with experience in the global management of such patients. This will reduce the risk of complications and improve patient outcomes. This review gives an overview of neuromuscular spinal deformity focussing on assessment and treatment.

Keywords complications; myopathic; neuromuscular; outcomes; scoliosis; spinal deformity; surgical treatment

Introduction

Neuromuscular scoliosis is the second most common spinal deformity after idiopathic. Contrary to idiopathic scoliosis, patients with neuromuscular deformities have a degree of physical and mental disability. Impaired control of trunk musculature can be due to either spasticity or hypotonia, resulting in the development of spinal deformity in most patients (Table 1). The deformity affects more often children who have no ambulatory function and those with poor head and neck control.[1]

Treatment of scoliosis is aimed largely at prevention of complications which arise from curve progression. The primary problems associated with a severe spinal curvature involve back or rib pain, difficulty looking after the patient and respiratory compromise. Curve prognosis depends on the underlying neurological or myopathic condition. This can dictate the timing and appropriateness of surgical treatment. Opposite to adolescent idiopathic scoliosis, neuromuscular curves occur in patients who can be markedly unwell with multiple medical problems affecting life expectancy, poor ability to cooperate and underlying osteoporosis.

Types of deformity

Scoliosis

Neuromuscular scoliosis involves a long, collapsing C-shaped curve which extends distally to include the pelvis, resulting in marked pelvic obliquity with the concave side being elevated (Figure 1). Pelvic imbalance can also be affected by hip subluxation or dislocation, as well as contractures of the hip flexors and adductors producing a typical 'wind-swept' deformity. When considering the order of correction, it is generally preferable to address the spino-pelvic deformity first in order to level the pelvis and allow relocation of the hips.

In patients with less severe neuromuscular involvement, such as hemiplegia, Friedrich's ataxia and Charcot–Marie–Tooth disease an 'idiopathic-like' curve is observed. This tends to be less severe with little or no pelvic obliquity and follows the principles of treatment that apply in idiopathic scoliosis.[2]

Thoracic or thoracolumbar kyphosis

This is the most common type of sagittal deformity seen in quadriplegic cerebral palsy, conditions characterized by low muscle tone and in children with fixed hip flexion contractures which eliminate lumbar lordosis. Its apex is at the junction between the mobile lumbar spine and the fixed thoracic cage due to the inability of trunk muscles to maintain balance. The deformity has a twofold effect: 1) due to the vertical space reduction it produces a restrictive lung defect; 2) as the child often has insufficient head control to maintain a horizontal gaze, interaction with the environment is reduced. It gives rise to pain from muscle fatigue but also due to pressure areas which occur across the apex of the curve. Thoracolumbar kyphosis can also develop as an iatrogenic form in children undergoing multilevel laminectomy for dorsal rhizotomy.

Lumbar hyperlordosis

This type of deformity is uncommon but its effects are more pronounced on sitting balance as the centre of gravity is shifted posteriorly and the anterior trunk muscles are too weak to accommodate. It develops in children with hip flexion contractures as compensation for their pelvic position and in those with ambulatory diplegia in whom it can predispose to lumbosacral

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Prevalence of scoliosis in common neuromuscular conditions

Upper motor neurone	
Cerebral palsy	25–74%
Friedreich's ataxia	80%
Brain or spinal cord tumour or trauma (prior to age 10)	100%
Lower motor neurone	
Myelodysplasia	60%
Spinal muscular atrophy	67%
Poliomyelitis	Decreasing (vaccinations)
Myopathic	
Duchenne muscular dystrophy	90%
Congenital myopathies	Dependent on diagnosis
Arthrogryposis multiplex congenital	30–67%

Table 1

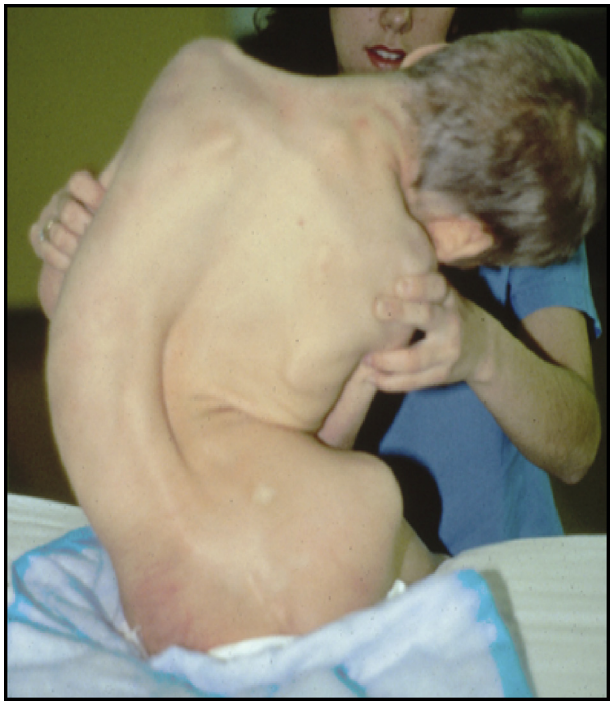

Figure 1 Typical C-shaped collapsing left thoracolumbar scoliosis with associated pelvic obliquity producing rib impingement against the elevated right iliac crest and asymmetrical loading of the left buttock (risk for pressure sore). The patient has complaints of severe pain and has lost any ability to sit independently.

spondylolysis/spondylolisthesis. Some patients can tolerate lumbar hyperlordosis; however, over time this is likely to become painful and is very difficult to manage with surgery which is the only predictable treatment.

Curve prognosis

Children with neuromuscular conditions generally present at a young age with a flexible postural curve which corrects on traction or suspension. No treatment is required at this stage other than observation on a 9-12-monthly basis. As the child grows their body mass increases producing greater workload upon the trunk muscles leading to fatigue and pain. As the vertebral column grows in height the lever arms through which the forces act increase. At this point the developing deformity requires close monitoring to gauge progression and guide management. In myopathic conditions progression of muscle weakness is accelerated. With growth the flexible postural curve evolves into a rotated structural scoliosis. Associated with the spinal curvature are changes to the chest wall and lung function the extent of which also depends on the underlying neuromuscular diagnosis.

As the child reaches puberty growth is rapid; however, rather than producing an increase in body height this can translate to a deteriorating spinal deformity which gradually becomes stiff. At this age, the problems associated with the curve become more problematic. With increasing pelvic involvement the patient may be unable to tolerate sitting and become effectively bed bound. Pain can arise from chronic muscle fatigue, pressure areas and costo-iliac impingement. Chest wall compromise can result in respiratory dysfunction.[3] Caring for children can become challenging with hygiene and feeding being compromised due to the deformity.

Common neuromuscular conditions

Cerebral palsy (CP)

This is the most common neuromuscular condition seen in orthopaedics. It is characterized by a static neurological deficit as the result of brain injury occurring in the perinatal period or early life. It is classified according to the Gross Motor Function Classification System (GMFCS-Table 2).[4] Observations of neuromuscular scoliosis' natural history are usually based on CP patients, largely as they are the most common. Patients with quadriplegia present with the typical collapsing C-shaped curve. Hypotonic patients tend to be more at risk of kyphosis or kyphoscoliosis, whilst those with athetosis or dystonia are less likely to develop scoliosis. Madigan and Wallace[1] reported an incidence of scoliosis up to 90% in severely affected children which is inversely related to ambulatory ability. Scoliosis in CP develops between 6 years and 10 years of age with a flexible curve. As the child grows, the deformity patterns described above are followed until the curve reaches 40–60 degrees. At this point the curve can progress up to 2 degrees per month with bracing doing little to prevent deterioration.[5]

An important caveat is that CP is sometimes a diagnosis of exclusion in the absence of clinical features of other neurological conditions. These children may have different curve progression and it is important to monitor them closely in the event of atypical findings.

Myelomeningocele

Spina bifida encompasses a group of congenital spinal anomalies in addition to failures of posterior arch formation. Congenital vertebral abnormalities can co-exist and the deformity can be a mixture of a congenital curve with a superimposed paralytic component due to the inherent muscle weakness. All patients radiologically have a tethered cord but this does not affect scoliosis surgery in the absence of clinical signs. They may also have hydrocephalus with ventriculoperitoneal shunts and Chiari malformation with invagination of the cerebellar tonsils in the spinal canal.

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