Neuromuscular spine deformity

Neuromuscular spine deformity (NSD) refers to spinal deformity that is associated with a myriad of neuromuscular diseases (NMD) ranging from Chiari I malformations to cerebral palsy (CP), muscular dystrophy (DMD), and myelomeningocele (MM).

The incidence and severity of the NSD usually parallels the severity of the underlying NMD – but not always. NMD causes the NSD generally by three mechanisms: trunk muscle weakness, spasticity, or imbalance. These factors are influenced by growth especially during the adolescent growth spurt at which time rapid progression can occur.

Treatment of the underlying NMD (e.g., steroids in DMD) may prevent or slow the progression of NSD, but, on the other hand, some treatments may precipitate or cause rapid increase in NSD (e.g., baclofen pump in CP patients). The NMD can also be associated with abnormal anatomy (MM) that can affect the natural history of the NSD.

Nonoperative treatment (i.e., bracing) does not affect the natural history of a particular curve and therefore is not definitive treatment for progressive deformity. Bracing is used to provide improved sitting posture in slim patients with flexible NSDs and delay definitive surgery to allow for spinal growth. Progressive NSD in young children that are not candidates for bracing can be managed with posterior spinal growing rods. When NSDs reach a magnitude which indicates the need for definitive surgery (i.e., fusion), various options are available as dictated by the individual case. The goal of the surgery is to prevent further progression and to achieve a corrected spine that is balanced in the coronal and sagittal planes over a level pelvis. PSF/PSSI +/− to the pelvis is the standard surgical strategy though other options may be necessary. Special consideration of the overall medical condition (comorbidities) and ambulatory status in these often frail patients is essential.

Complication rates for NSD are significantly higher than those for AIS, especially infection, prolonged hospital stay, instrumentation failure, etc., and are influenced by the comorbidities present at the time of surgery. These events can be minimized by the control of preexisting conditions such as seizures, respiratory issues, and nutrition among others. Though the surgical treatment of NSD may be difficult for all concerned, there is generally a high degree of satisfaction among the caregivers of these patients.

Olafsson et al. reviewed 90 consecutive patients with various neuromuscular diseases and a progressive spine deformity treated with a prefabricated Boston-type underarm corrective brace. Of these, 38 patients had spastic tetraplegia; seven, syndrome-related muscular hypertonia; 24, muscular hypotonia; and 21, myelomeningocele. The mean age at the treatment start was 9.2 years (range, 1.4-17.7 years). Twenty-four were ambulating and 66 wheelchair-bound. Hypotonia was the dominant type of muscle involvement in 49, spasticity in 28, and athetosis in 13 patients. The mean pretreatment Cobb angle was 47 degrees, with a range from 23 to 95 degrees. The mean brace-induced Cobb-angle correction was 60%, thus well comparable to that in idiopathic scoliosis. However, this did not predict favorable treatment results. At the follow-up, on average 3.1 years (range, 1-5.5 years) after weaning from the brace, the brace treatment was successful in 23 patients. Successful was defined as <10 degrees curve progression during the observation time and a good brace compliance. Forty-one patients discontinued the brace treatment, and 19 progressed despite adequate brace wear. Five patients are still in treatment, and two have died. Successful treatment was seen in ambulating patients with muscle hypotonia and short thoracolumbar/lumbar curves

measuring <40 degrees as well as in nonambulating patients with spastic short lumbar curves. These types of neuromuscular scoliosis may be the only ones to respond to brace treatment. In other cases, the brace treatment cannot be expected to have a lasting corrective effect although it can be used as sitting support [1)].

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Olafsson Y, Saraste H, Al-Dabbagh Z. Brace treatment in neuromuscular spine deformity. J Pediatr Orthop. 1999 May-Jun;19(3):376-9. PubMed PMID: 10344323.