Neuromuscular Scoliosis: Current Concepts

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abstract

Scoliosis is a common deformity in neuromuscular disorders. This spinal deformity usually presents at an early age, rapidly progresses during growth, and continues to progress even after skeletal maturity. Neuromuscular scoliosis classically involves the entire thoracolumbar spine, often extending to the pelvis and causing pelvic obliquity. Physical examination can be challenging, but it is important to assess the flexibility of the spinal curve and associated joint contractures. Upright anteroposterior and lateral radiographs are the preferred imaging. When formulating a treatment plan, it is important to take into account not only the degree of curvature, but also each patient’s needs and quality of life, the potentially high perioperative complication rates, and the natural history of the underlying neuromuscular disorder. Different neuromuscular conditions behave differently and should therefore be treated differently. With the exception of steroids for Duchenne muscular dystrophy, bracing remains the only reliable nonoperative intervention available. Preoperative optimization of medical comorbidities is crucial given the relatively high complication rate. Posterior segmental instrumentation has revolutionized the surgical management of neuromuscular scoliosis and is the most commonly used technique today. Despite reported improvement in postoperative quality of life, there are several limitations with currently used outcome measures that prevent a well-informed discussion on the outcomes after surgery. [Orthopedics. 2015; 38(6):e452-e456.]

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The incidence of scoliosis associated with neuromuscular disease can be as high as 90%, compared with up to 4% for idiopathic scoliosis (IS), which is the most common type of pediatric scoliosis. Unlike the idiopathic type, neuromuscular scoliosis (NMS) tends to progress even after skeletal maturity and is associated with significantly higher complication rates and economic burden. In a large retrospective study comparing children with NMS and IS after spinal surgery, those with NMS had longer lengths of hospital stay, higher total charges, higher complication rates, higher number of secondary procedures, and higher rates of disposition to other facilities or to home health care services. Neuromuscular scoliosis often presents at an early onset, rapidly progresses during growth, and continues to progress even after skeletal maturity. Among the various neuromuscular disorders, patients with spastic quadriplegic cerebral palsy have the highest incidence of scoliosis and the greatest risk of progression beyond skeletal maturity.

ETIOLOGY

Although NMS can result from a variety of neuropathic or muscular disorders (eg, cerebral palsy, myelodysplasia, muscular dystrophy), the central underlying etiology is the impaired function of the muscle forces acting on the spine leading to progressive trunk imbalance. The paraspinal muscles can be flaccid, spastic, or dyskinetic. As a result of the gross muscle imbalance, NMS classically involves the entire thoracolumbar spine, often extending to the pelvis and inducing pelvic obliquity (Figure 1A). Thoracic hyperkyphosis is another common feature (Figure 1B).

PHYSICAL EXAMINATION

Examining patients with NMS can be challenging because a sizeable proportion are nonambulatory or are unable to cooperate with the examination. Although it is important to observe the gait in an ambulatory patient, careful inspection of the seated position in a nonambulatory patient is equally important. The examiner may find truncal shift, kyphotic collapse, or pelvic obliquity (Figure 2A). The seated position may also reveal postural abnormalities in the lower extremities that can lead to pelvic obliquity. Typically, pelvic obliquity secondary to hip contracture, known as pelvic obliquity of the lower origin, is characterized by flexion and internal rotation contracture of 1 hip with abduction and external rotation contracture of the contralateral hip.

With the patient on the examination table, the ability to level the pelvis by asymmetric traction on the elevated (contracted) hip implies obliquity of a lower origin. However, if the pelvic obliquity persists, the scoliotic deformity is the underlying cause of the pelvic imbalance, which is known as pelvic obliquity of the upper origin (Figures 2A-D). The examiner can then perform a dynamic examination of the spine using the 3-point reduction technique to assess the flexibility of the deformity (Figure 2B). This is important to assess because rigid deformities cannot be corrected by bracing and make surgical correction more complex. Rigid deformities are also associated with higher complications, longer operative times, and a smaller percentage of age of final correction.

RADIOGRAPHIC EVALUATION

Although standing posteroanterior (PA) and lateral radiographs of the thoracolumbar spine provide the best information on the extent of spinal imbalance in an ambulatory patient, PA and lateral radiographs with the patient in a wheelchair can be alternatively obtained in a nonambulatory patient. However, plain radiographs with the patient in a wheelchair are often cumbersome to obtain and reproduce. As a result, supine PA and lateral radiographs are frequently obtained. Supine bending or traction radiographs should also be obtained because they provide information on the flexibility of the deformity. Magnetic resonance imaging is not routinely obtained, except in cases of suspected medullary pathology.

MEDICAL CONSIDERATIONS

Although the spinal deformity may be impressive and the most concerning complaint of some parents, scoliosis is often only 1 of the problems facing patients with neuromuscular disease. Nutritional depletion is common among patients with NMS, especially those who are severely disabled. Poor nutrition has been linked to increased postoperative complications, including infections, prolonged intubation, and longer length of hospital stay. Supplemental nutrition is advocated for children below the fifth percentile in weight and should be instituted several weeks to months preoperatively.

Impaired respiratory function is common in children with NMS, resulting in numerous perioperative complications. In addition to the restrictive lung disease caused by the progressive spinal deformity, pulmonary function may also be diminished by the underlying respiratory muscle weakness, as in patients with muscular dystrophy. Preoperative predictors of pulmonary complications include a
Cobb angle greater than 69°, a forced vital capacity less than 39.5%, a FEV1 less than 40%, and age older than 16.5 years.

Cardiomyopathy is another well-known comorbidity in patients with neuromuscular disease, particularly those with myopathies. Gastroesophageal reflux, chronic constipation, bladder dysfunction, and endocrine aberrancies are also common. Gastroesophageal reflex may place the patient at risk for aspiration in the perioperative period. Chronic constipation, which is especially prevalent in wheelchair-bound patients, can predispose patients to postoperative paralytic ileus and accentuate the pulmonary compromise.

**Decision Making**

When formulating a treatment plan, it is mandatory to take into account not only the degree of curvature, but also the patient’s needs and quality of life, the potentially high perioperative complication rates, and the natural history of the underlying neuromuscular disorder. Although different neuromuscular conditions tend to be grouped together, each condition behaves differently and should, therefore, be treated differently.

**Nonoperative Management**

With the exception of steroids for patients with Duchenne muscular dystrophy, bracing remains the only available nonoperative intervention for children with NMS. Bracing is primarily used for positional support and to help provide temporizing control until surgical timing is more optimal. A custom total contact thoracic-lumbar-sacral orthosis is the most effective in providing truncal support (Figure 3).

Among the different neuromuscular conditions, Duchenne muscular dystrophy has a positive response to glucocorticoids, with significant improvement in pulmonary function and a prolonged ability to ambulate independently. Patients treated with glucocorticoids also had significantly lower Cobb angles and were less likely to require surgical correction. More recently, a prospective study reporting on the longest follow-up on patients treated with high-dose steroids showed that steroids stabilized the spine and eliminated the need for surgery and showed that a protective benefit against scoliotic progression persisted beyond skeletal maturity.

**Operative Management**

The surgical techniques for NMS correction have evolved over the past decade. Traditionally, a combined anterior/posterior approach has been advocated. Ante-
rerior diskectomy and release of the anterior longitudinal ligament together with post-er instrumentation provide excellent ar-throdesis and flexibility for a rigid spinal deformity,15 while preventing the crank-shaft phenomenon in skeletally immature patients1 and allowing for correction and maintenance of pelvic obliquity.17 Staging the anterior/posterior approach is contro-versial and a review of literature shows con-flicting results with respect to complication rates.18,19 In general, although experienced surgeons may advocate 1-stage procedures, staging the anterior/posterior approach is likely more important in patients with large curves with concomitant medical morbidities.

Anterior surgery alone is rarely needed currently and is limited to patients with suf-ficient vital capacity, short isolated lumbar or thoracolumbar curves, minimal pelvic obliquity, and slow or nonprogressive pathology.16

Posterior segmental instrumentation with hooks, pedicle screws, or sublami-nar wires have revolutionized the surgical management of NMS. Posterior arthrodesis and instrumentation is by far the most com-monly used technique currently, especially in cases of skeletally mature patients with good curve flexibility, concomitant pel-vic obliquity, and limited lung function.20 There is some controversy as to whether it is necessary to extend the arthrodesis to the pelvis in all patients with NMS. Broom et al21 considered it mandatory to do so in cases of fixed pelvic obliquity exceeding 15°, when the sacrum is part of the curve, or when truncal decompensation occurs such that the vertical plumb line falls lateral to the sacroiliac joint.

INTRAOPERATIVE SPINAL CORD MONITORING

Somatosensory spinal evoked potentials are more useful than somatosensory cortical evoked potentials in providing real-time intraoperative detection of neu-rologic injury. Somatosensory cortical evoked potentials are unreliable and non-specific in neuromuscular scoliosis.22 To optimize the sensitivity and specificity of somatosensory spinal evoked potentials, an amplitude loss of 50% from baseline is considered clinically significant. Somato-sensory spinal evoked potentials traces remaining above 50% are unlikely to have neurologic sequelae.23

POSTOPERATIVE COMPLICATIONS

Patients with NMS have high complica-tion rates following surgical correction compared with those with IS. A meta-analysis of 15,218 patients with NMS showed that pulmonary complications are the most prevalent (22.71%), followed by implant-related complications (12.51%), infections (10.91%), neurological complica-tions (3.01%), and pseudoarthrosis (1.88%). There is a more than 10-fold increase in the risk of wound infections postoperatively for NMS compared with IS.24 Staphylococcus aureus is the most commonly cultured organism, followed by coagulase-negative staphylococci and Pseudomonas aeruginosa.

Compared to IS, patients with NMS also have greater blood loss, which is at-tributed to the complexity of correction with more instrumented levels and the frequent need to correct pelvic obliquity, poor nutritional status, impaired con-nective tissue function, and use of anti-seizure medications, such as valproic acid, which decreases platelet count, and factor VIII levels.2 In a retrospective study of risk factors for complications after NMS surgery, nonambulatory status and a Cobb angle equal to or greater than 60° were the greatest predictors of major complica-tions.5

POSTOPERATIVE QUALITY OF LIFE

Despite high complication rates, sur-veys of patients and their caregivers show high satisfaction rates and better quality of life (QOL) regardless of the amount of deformity correction.25 Specifically, the areas of most improvement are sitting balance, self-esteem, and reduction in caregiver time. However, a systemic lit-erature review cautiously pointed out that measurement of postoperative QOL in children with NMS is more complex and dynamic than simply surveying patients’ and caregivers’ perceptions.26 Children with NMS do not all share the same needs and expectations and there are multiple confounding factors when assessing QOL (eg, use of steroids in Duchenne muscular dystrophy, different sources for pain, cognitive difficulties). When accounting for the high complication rates and level of evidence for the different QOL studies, the evidence in support of QOL improve-ment in patients with cerebral palsy and Duchenne muscular dystrophy becomes weak, with no evidence for QOL improve-ment in patients with spina bifida.14

Conclusion

Scoliosis is a common deformity in children with neuromuscular disease. Compared with IS, management of NMS is more complex and is associated with higher complication rates. Unlike IS, NMS is often only 1 problem among patients with neuromuscular disease. A detailed medical history is important and multiple
consultations are often indicated preoperatively. Preoperative planning and decision making should take in account the underlying disease process and its associated comorbidities, including assessment of the cardiopulmonary system and nutritional status. Bracing is used to help with positioning and to provide temporary control until surgical timing is more optimal.

Posterior instrumentation and arthrodiasis is currently the most commonly used surgical technique. Although the different neuromuscular conditions tend to be grouped together, it is important to remember that each condition behaves differently and should, therefore, be considered separately. In the absence of high-quality evidence to guide the management of NMS, common sense guided by the patient’s needs, medical morbidities, and potential complications remains the first line of treatment.

REFERENCES