Neuromuscular Scoliosis: Diagnostic and Therapeutic Considerations

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Neuromuscular scoliosis in a child presents as a diagnostic and therapeutic dilemma to the pediatric neurologist, the pediatric neurological surgeon, and the pediatric orthopaedist. Because of its progressive nature, it tends to be refractory to conservative management and requires active intervention. Neuromuscular scoliosis is a symptom, and the diseases leading to the deformity must be addressed together with the spinal abnormality. The framework of the diagnosis and management is addressed in this article.

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ABNORMALITIES OF spinal curvature have long been recognized as infirmities that demand medical vigilance. In ancient Greece, Hippocrates (b. 460 BC) wrote extensively of spinal deformity, noting that “There are many varieties of curvatures of the spine even in persons who are in good health, for it takes place from natural conformation and from habit, and the spine is liable to be bent from old age and from pains.” He appreciated the relationship between age of onset and curve severity, and developed the concept of longitudinal traction for treatment of the disorder. Subsequently, the prominent Greek physician and anatomist Galen (b. 129 AD) categorized spinal deformity based on curve morphology, introducing the terms kyphosis, lordosis, and scoliosis into medical nomenclature. Like Hippocrates, Galen used elaborate traction mechanisms to effect curve correction. Although largely ineffectual, this mode of treatment persisted well into the 19th century AD.

As an alternative to traction, thoracolumbar orthoses were introduced by Ambrose Pare in the mid-16th century. He used metal corsets fashioned by an armorer to treat children and adolescents with spinal deformity, recognizing that bracing was ineffectual after skeletal maturity had been achieved. The art of bracing and casting was refined over the ensuing centuries, culminating in the advent of the Milwaukee brace in 1945. Intended initially as a means of postoperative immobilization, this brace is now used widely as a primary means of nonoperative treatment of kyphotic and scoliotic spinal deformities.

Early efforts at surgical correction of scoliosis were reported in the mid-19th century by the French surgeons Guerin and Malgigne. These procedures consisted of paraspinous myotomies that were performed to overcome the “muscle imbalance” perceived to be responsible for the condition. The modern era of surgical treatment for spinal deformity was heralded by Hibbs, who performed the first successful posterior spinal fusion procedure for kyphosis in a patient with tuberculous spondylitis in 1911. Initially, operative intervention was limited to in situ fusion, intended to halt curve progression but not capable of correcting an existing deformity. However, with the advent of the Harrington distraction rod system in 1962, correction of even extensive deformities was made possible by applying reductive forces to the malaligned spine via dorsally applied internal fixators. Subsequently, a similar posterior instrumentation system developed by Cotrel and Dubousset permitted the application of both corrective and compressive forces to the same construct to address more complex deformities.

Segmental spinal fixation using rods and sublaminar wires was introduced by Resina and Alves in 1977. This technique, further refined by Luque, empowered the surgeon with the capacity to correct deformities in both the sagittal and the coronal planes, a concept germane to modern spinal surgery. In the late 1960s, Dwyer et al developed a ventrally applied thoracolumbar instrumentation system that incorporated multilevel anterior disectomy and segmental vertebral body screw and rod fixation to achieve superior reduction of rotational and coronal deformities. These major innovations in surgical technique and internal fixation instrumentation, coupled with advances in anesthesiology, transfusion medicine, and critical care, have increased the safety and the efficacy of corrective surgical procedures for children with spinal deformity.
GENERAL CONSIDERATIONS

Nomenclature

The Scoliosis Research Society defines scoliosis as a lateral curvature of the spine that exceeds 10° with consequent rotation of a series of vertebrae away from the midline spinal axis. Although most obvious in the coronal plane, true scoliotic deformity is evident in the sagittal and axial projections as well. Deformities of this nature are common in the pediatric population and range widely in severity from the barely detectable to the grotesquely disfiguring. When mild, a scoliotic curvature may be purely a cosmetic debility but may carry with it profound psychological ramifications, both in childhood (poor self-image, social isolation) and as an adult (lower marriage rate, higher rates of unemployment and disability). When severe, spinal deformity may incite potentially life-threatening physiological alterations, including restrictive pulmonary insufficiency and cor pulmonale. Irrespective of the extent of deformity, it must be remembered that scoliosis is only a physical finding and not a disease process. Successful treatment is contingent on the accurate and timely diagnosis of the underlying disorder(s).

Before proceeding, some clarification of the nomenclature used to describe scoliotic deformities is warranted. Scoliosis is deemed structural or nonstructural based on two fundamental elements: the extent of spinal flexibility in the region of the curve and the influence of posture on curve morphology. Structural scoliosis is a fixed lateral curvature of the spine that cannot be corrected or maintained by volitional effort, persists in supine and bending postures, and demonstrates restricted spinal mobility. Irrespective of the extent of deformity, it must be remembered that scoliosis is only a physical finding and not a disease process. Successful treatment is contingent on the accurate and timely diagnosis of the underlying disorder(s).

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Etiology and Epidemiology

An estimated 500,000 individuals in the United States are afflicted with scoliosis, and in most, this type of spinal deformity develops during childhood or adolescence. A lateral spinal curve measuring greater than 10° may be identified in roughly 2% to 3% of adolescents at the terminus of skeletal growth; however, fewer than 0.5% exhibit curves exceeding 20°. The etiology of scoliosis in the pediatric population is varied. Many disparate conditions may present with morphologically similar spinal curvatures, and thus the magnitude, location, or direction of the curve have poor predictive value in identifying the underlying disease process. In many cases, the cause of deformity may be multifactorial. Table 1 presents a general classification scheme proposed by the Scoliosis Research Society that subdivides scoliotic deformities based on origin.

Idiopathic scoliosis is the most common of all forms of pediatric spinal deformity and accounts for roughly 65% of all cases of structural scoliosis.
Table 1. Classification of Scoliotic Deformities by Etiology

<table>
<thead>
<tr>
<th>Structural scoliosis</th>
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<tr>
<td>Idiopathic</td>
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<tr>
<td>Infantile (0 to 3 years)</td>
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<tr>
<td>Juvenile (3 to 10 years)</td>
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<tr>
<td>Adolescent (10+ years)</td>
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<tr>
<td>Neuromuscular</td>
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<tr>
<td>Neuropathic</td>
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<tr>
<td>Myopathic</td>
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<tr>
<td>Congenital</td>
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<tr>
<td>Failure of formation</td>
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<tr>
<td>Failure of segmentation</td>
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<tr>
<td>Neurofibromatosis</td>
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<tr>
<td>Mesenchymal disorders</td>
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<td>Marfan's syndrome</td>
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<td>Ehlers-Danlos syndrome</td>
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<tr>
<td>Rheumatoid disease</td>
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<tr>
<td>Spinal trauma</td>
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<tr>
<td>Fracture</td>
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<tr>
<td>Postoperative</td>
<td></td>
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<tr>
<td>Osteochondrodystrophies</td>
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<tr>
<td>Achondroplasia</td>
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<tr>
<td>Spondyloepiphyseal dysplasia</td>
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<td>Mucopolysaccharidoses</td>
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<tr>
<td>Infection</td>
<td></td>
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<tr>
<td>Metabolic disorders</td>
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<tr>
<td>Rickets</td>
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<tr>
<td>Osteogenesis imperfecta</td>
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<td>Homocystinuria</td>
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<td>Spinal neoplasia (tumors of the vertebral body)</td>
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<thead>
<tr>
<th>Nonstructural scoliosis</th>
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<tr>
<td>Postural</td>
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<tr>
<td>Hysterical</td>
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<tr>
<td>Irritative lesions</td>
<td></td>
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<tr>
<td>Limb length inequality</td>
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Although not the focus of this article per se, a brief discussion of this condition is warranted as a prelude to addressing the neuromuscular scolioses, owing to the former’s relative prevalence and comparable demographics. By definition, idiopathic scoliosis arises in otherwise healthy children and represents a process for which no identifiable cause has yet been established. Three categories of idiopathic scoliosis have been proposed, based on age at the time of curve detection: (1) infantile (0 to 3 years of age), (2) juvenile (3 to 10 years of age), and (3) adolescent (greater than 10 years of age).

Infantile scoliosis accounts for fewer than 1% of all cases of idiopathic scoliosis, is more common in males, and tends to involve the thoracic spine preferentially, with most curves projecting leftward.17,18 Juvenile scoliosis is relatively more frequent, comprising roughly 10% of idiopathic cases.17,18 Within this patient subgroup, no gender predilection has been identified and the curves are equally distributed to the right and the left.

Most idiopathic scoliotic curvatures (nearly 90%) appear after the onset of the pubertal growth spurt and thus are classified as “adolescent.” Although school screening studies indicate that the gender ratio of young patients (ie, 10 years) with mild curvatures approaches 1:1, it is the female patient that is most likely to have a progressive curve and thus require treatment consisting of orthotic bracing, surgery, or both. In addition to gender, factors that predispose to curve progression include the patient’s age, extent of skeletal maturity, curve pattern, and magnitude of curvature.12,19 Progression is most likely in younger patients who exhibit relative skeletal immaturity.12 Large curves and curves that display a double major configuration are also more prone to progression; conversely, smaller curves and single lumbar and single thoracolumbar curve patterns are less likely to progress.19 If left untreated, progressive scoliotic curvatures may result in severe cosmetic deformity, chronic back pain, impairments of posture and integument, restrictive pulmonary insufficiency, and cor pulmonale.20

Spinal deformity that arises as a consequence of neuromuscular disease is far less prevalent than that associated with idiopathic scoliosis. As a rule, neuromuscular scoliosis is a disease of the very young, presenting much earlier than the idiopathic form. Indeed, most of these conditions are manifest in early childhood, even in infancy, and thus are often associated with an early onset of spinal deformity. Furthermore, the neurological dysfunction inherent in such conditions frequently produces varying degrees of paralysis and axial muscle imbalance, both permissive factors that favor early and profound curve progression. Unlike idiopathic scoliosis, in which curve progression usually ceases when skeletal maturity is achieved, neuromuscular scoliotic curvatures may continue to increase throughout the patient’s lifetime, irrespective of skeletal maturation.

As many of these patients have generalized neurological dysfunction, development of a spinal curvature may have a disastrous effect on an individual’s overall functional capacity. Spinal deformity may induce pelvic obliquity or trunk imbalance, postural factors that may alter a patient’s ability to stand, ambulate, or sit. Previously independent patients may require additional orthoses to
ambulate or may become wheelchair bound; those
confined to a wheelchair may lose the ability to
operate assist devices or may develop decubitus
ulcers, further restricting their independence.21 Fur-
thermore, the management of spinal deformity in
such patients is confounded by the often progress-
vie nature of the underlying neurological condi-
tion, which may alter cognition and autonomic
function as well.

Neuromuscular causes of scoliosis may be subdi-
vided into neuropathic and myopathic conditions;
the former may be further differentiated as disor-
ders affecting the upper motor neuron and those
involving the lower motor neuron, respectively.
Table 2 presents a classification scheme of neu-
romuscular scoliosis proposed by the Scoliosis Re-
search Society.16

PATIENT EVALUATION

A complete evaluation of the child with neuro-
muscular scoliosis requires an appreciation of both
the neurological and orthopaedic manifestations of
the condition. Emphasis should be placed on deter-
mining the patient’s overall level of function and
independence, and restrictions created by the under-
lying neurological dysfunction and those imposed
by the progressive spinal deformity quantified.
Intellectual performance and developmental mile-
stones should be carefully scrutinized. Associated
general medical deficiencies, such as pulmonary
insufficiency and cardiomyopathy, must also be
identified. A complete history, general physical
examination, and neurological examination, supple-
mented by plain spinal radiographs and other
complementary diagnostic tests, such as computed
tomography (CT), magnetic resonance imaging
(MRI), electromyography, and muscle biopsy,
should establish the correct diagnosis as to the
nature of the spinal deformity and its underlying
neuropathic or myopathic condition.

**History and Physical Examination**

Historical data should be collected as in the
assessment of any putative neuromuscular disorder.
All interviews should include a comprehensive
prenatal and perinatal history. Developmental mile-
stones during infancy and early childhood should
be documented, and any deviation from their
timely achievement should be noted. In older
children, an accurate assessment of physiological
maturity should be undertaken, including age of
pubertal onset and menarche.22,23 A detailed family
history is crucial in establishing a diagnosis in
conditions, such as spinal muscular atrophy (Wer-
ding-Hoffman, Kugelberg-Welander’s disease), Frie-
dreich’s ataxia, Charcot-Marie-Tooth disease, famil-
ial dysautonomia, and Duchenne’s muscular
dystrophy.

In addition to routine historical data, specific
queries should be made with reference to the spinal
deformity itself. Attempts should be made to estab-
lish age of onset, rate of progression, and functional
significance of the spinal curvature. In particular,
the impact on the patient’s mobility and perfor-
mance of basic activities should be determined. The
presence of axial or radicular pain should be noted.
As a general rule, idiopathic scoliosis in children
and adolescents is not associated with pain; its
presence therefore suggests an alternate, often
neurological, etiology. Finally, symptoms suggest-
ing cardiac and pulmonary decompensation must
be investigated.

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**Table 2. Classification of Neuromuscular Scoliosis by Etiology**

<table>
<thead>
<tr>
<th>Neuropathic</th>
<th>Myopathic</th>
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<tbody>
<tr>
<td>Upper motor neuron</td>
<td>Arthrogryposis</td>
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<tr>
<td>Cerebral palsy</td>
<td>Muscular dystrophy</td>
</tr>
<tr>
<td>Spinocerebellar degeneration</td>
<td>Duchenne</td>
</tr>
<tr>
<td>Friedreich’s ataxia</td>
<td>Limb-girdle</td>
</tr>
<tr>
<td>Charcot-Marie-Tooth</td>
<td>Facioscapulohumeral</td>
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<tr>
<td>Roussy-Levy</td>
<td>Fiber-type disproportion</td>
</tr>
<tr>
<td>Syringomyelia</td>
<td>Congenital hypotonia</td>
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<tr>
<td>Spinal cord tumor</td>
<td>Myotonia dystrophica</td>
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<tr>
<td>Spinal cord trauma</td>
<td>Other</td>
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<td>Other</td>
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The physical examination of patients with scoliosis consists of three components: neurological, general, and orthopedic. A comprehensive neurological examination is performed, with particular emphasis placed on sensorimotor function. Alterations in muscular tone are noted, and the range of extremity motion is assessed. Gait and station are observed. Transfers between seated and standing postures are scrutinized.

The general examination endeavors to identify sequelae of the underlying neurological condition and/or of the spinal deformity itself. Severe curvatures, particularly those involving the thoracic spine deform the chest wall and may induce restrictive pulmonary insufficiency and cor pulmonale. In such instances, formal pulmonary function testing may be indicated, particularly if surgical intervention is proposed. Cardiomyopathy may accompany conditions, such as Friedreich’s ataxia and Duchenne’s muscular dystrophy. Autonomic instability, such as that observed with Riley-Day syndrome (a.k.a. familial dysautonomia) may be manifest as labile hypertension or hypotension. Sensory dysfunction may complicate many neuropathic conditions, including syringomyelia, spinal cord trauma, cord tumors, and dysautonomia, and violations of skin integrity and decubitus ulcerations are common. Other general examination findings, such as corneal opacification (mucopolysaccharidoses), high arching palate (Marfan’s syndrome), low-set hairline (Klippel-Feil syndrome), neck webbing (Turner’s syndrome), cafe-au-lait spots (neurofibromatosis), and skin dimpling/hyperpigmentation over the back (spinal dysraphism), suggest specific pathological entities.

The orthopedic examination emphasizes detection and quantification of spinal deformity and pelvic obliquity. The patient is observed undraped in both the standing and seated positions for obvious asymmetries and spinal deformities. Balance of the thorax over the pelvis may be assessed using a plumbline held at the C7 spinous process (vertebra prominens) or external occipital protuberance. Lateral deviation of the line from the gluteal cleft suggests a lateral spinal curvature. Relative shoulder height is compared from side to side using a carpenter’s level. Pelvic obliquity, a common deformity accompanying neuromuscular scoliosis, is measured in similar fashion by comparing the levels of the iliac crests. Spinal range of motion is calculated in flexion, extension, and lateral bending, and these data are used to discern the flexibility of the deformity. Axial traction may be applied to assist in this determination.

The forward-bending test is a simple yet highly sensitive physical examination maneuver that elucidates the presence of even mild scoliotic deformities. The patient stands with feet together, knees straight, and flexes approximately 90° at the waist. The arms are dependent and the hands are held with palms and fingers opposed. The back is observed from multiple vantage points for the presence of prominence or asymmetry. The site of maximal prominence (thoracic, thoracolumbar, or lumbar) is noted and its magnitude calculated using a level.

Diagnostic Studies

Radiological studies are nearly always indicated to supplement the clinical impression of spinal deformity. These studies confirm the presence and site of scoliotic curvature, permit quantification of its magnitude and flexibility, and document curve progression over time. An estimation of skeletal maturity may also be made using radiographic techniques. The radiological examination is particularly germane in assessing the patient with neuromuscular scoliosis as such studies may reveal the etiology underlying the spinal deformity.

Plain Roentgenograms. Plain roentgenograms constitute the initial diagnostic survey in evaluating the patient with scoliosis. These studies are inexpensive, readily obtainable, and do not require special expertise to interpret. So-called “scoliosis views” are obtained in the posteroanterior projection with the patient in the standing or upright sitting position. Standing radiographs are preferable; however, in those unable to stand, sitting views will suffice. Patients unable to sit (the very young or the severely paralyzed) are evaluated with supine radiographs. By convention, these studies are read as if viewing the patient from the back; thus, the right side of the radiograph represents the right side of the patient. Lateral projections are used to assess associated kyphosis or lordosis. Flexibility of the scoliotic curvature is quantified with supine side-bending radiographs in which the patient actively bends towards the side of the deformity’s apex.

Once the site and pattern of the scoliotic curvature have been established with upright plain radiography, the magnitude of the curve is calculated. To measure the curve accurately, one must first identify the vertebrae on the rostral and caudal
ends of the curve on the posteroanterior radiograph. These so-called "end vertebrae" are the final vertebrae maximally canted into the concavity of the curve being measured (Fig 1). The extent of the curve is then defined by these vertebrae; for example, a curve beginning at T3 and terminating at T11 with its apex projecting to the left is referred to as a T3 to T11 left thoracic curve. To assist in identifying the end vertebrae, the orientation of the intervertebral disc spaces are examined. Within the curve, the disc spaces are wider on the convexity of the curve and narrower on the concavity. The disc space above and below the end vertebrae exhibits either a parallel or reversed orientation when compared to the disc space immediately adjacent within the curve (Fig 1).

Once the upper and lower end vertebrae are identified, the curve is measured according to the method described by Cobb.26 Briefly, a line is drawn along (and parallel to) the superior endplate of the rostral end vertebra of the curve; a similar line is drawn along the inferior endplate of the caudal end vertebra (Fig 1). A line is then drawn perpendicular to each endplate line. The angle created by the intersection of these two perpendiculars is measured, yielding the magnitude or "Cobb angle" of the curvature. Using this technique, scoliotic curves may be measured with a high degree of accuracy and minimal intraobserver and interobserver variability.27,28

Similar methods are used to calculate curve magnitude on supine side-bending views. Such values are then compared with corresponding measurements obtained from upright neutral studies to determine the degree of curvature flexibility. Flexible curves will demonstrate smaller Cobb angles with side-bending toward the convexity of the curve; the magnitude of nonflexible curves is not altered significantly with lateral bending. Several investigators have proposed schemes to quantify the rotational component of vertebral displacement in scoliotic curvatures.29-31 In general, these techniques are complex and possess lesser clinical utility than that of Cobb angles, and thus their discussion lies outside the scope of this article.

Computed Tomography. In selected cases, CT scanning is a useful adjunct to plain radiography in evaluating the patient with neuromuscular scoliosis. CT provides unparalleled detail when imaging the osseous structures of the spine and presents calcific, osteoblastic, or osteolytic lesions to great advantage. When scoliosis is accompanied by myelopathy or radiculopathy, this modality permits a precise assessment of spinal canal and neural foraminal diameter at multiple levels along the length of a curvature to identify regions of stenosis. This function is enhanced by the capability of CT to reconstruct images in the sagittal and coronal planes. CT excels in localizing bone neoplasia and demonstrates the extent of bony destruction by tumor or infection more accurately than other modalities. Finally, operative planning is frequently facilitated by preoperative CT imaging, particularly when the use of internal spinal fixation devices, such as pedicle screws, is contemplated.

The use of CT scanning in the setting of spinal deformity is enhanced substantially by the addition of intrathecaly administered iodinated contrast medium. In addition to demonstrating bony anatomy, the CT-myelogram affords indirect visualization of the spinal cord and nerve roots, revealing thecal sac and neural element compression in a manner not possible with conventional noncontrast CT studies. CT-myelography is a relatively inexpensive study that is readily available at most medical centers on an outpatient basis.

Several disadvantages of CT and CT-myelography have relegated these modalities to a secondary role in the evaluation of the patient with neuromuscular scoliosis. Noncontrast CT images intraspinal soft tissues poorly and thus does not provide satisfactory visualization of the spinal cord and nerve roots. This modality is not sufficiently sensitive to reliably disclose noncalcific intraspinal mass lesions, such as intervertebral disc herniations, intradural or extradural neoplasms, or vascular anomalies. Furthermore, unlike MRI, CT imaging is limited to the axial plane, which limits its use in evaluating complex spinal curvatures.

CT-myelography is an invasive procedure necessitating percutaneous access of the subarachnoid space via the lumbar cistern or cisterna magna, and as such is less tolerated by the pediatric patient and more prone to morbidity than noninvasive studies, such as MRI. Serious complications of the procedure are rare, but include intracranial and intraspinal hemorrhage, meningitis, and contrast-induced anaphylaxis. In general, adverse reactions associated with myelography are mild; headache, nausea, vomiting, dizziness, and neck pain are those most commonly reported.32 Adverse reactions are less frequent when using nonionic contrast agents, such
as iohexal (Omnipaque) and iopamidal (Isovue).  

The incidence of postmyelography headache is proportional to the caliber of the spinal needle used and has been reported to be less than 4% when a 26-gauge needle is used.  

**Magnetic Resonance Imaging.** MRI has revolutionized the evaluation of spinal disorders and has supplanted CT-myelography as the procedure of choice when imaging the spinal canal in patients with neuromuscular scoliosis and other complex spinal pathology. This modality is noninvasive, devoid of ionizing radiation, and affords direct visualization of the neural elements and other soft-tissue structures within the craniospinal axis. In addition, images may be formatted in an infinite array of planes, both orthogonal and nonorthogonal. This multiplanar imaging capability renders MRI indispensable in evaluating the deformed spine; the imaging plane used can be tailored to conform to a given patient’s curvature. 

When a neuromuscular disorder is suspected to underlie a spinal deformity, MRI of the entire craniospinal axis is indicated to elucidate the causative lesion. Clinical settings that clearly suggest the need for MRI include the presence of congenital spinal deformity with or without myelodysplasia, juvenile scoliosis, painful curvatures, rapidly progressive curves, and leftward-projecting thoracic curvatures in adolescence. Any patient with scoliosis and an accompanying neurological deficit should undergo MRI as scoliotic curvatures, even when severe, rarely cause neurological dysfunction. Findings on plain radiography may also provide indication for MRI. Bony segmentation anomalies, spina bifida occulta, interpediculate widening, and pedicular erosion all mandate further investigation with this imaging modality. 

MRI possesses unsurpassed sensitivity and specificity in delineating intramedullary, intradural/extradural, and extradural spinal anomalies. Lesions involving the substance of the spinal cord, such as intrinsic cord tumors, syringomyelia, dys-

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Fig 1. Graphic rendition of a prototypical thoracic curvature. The magnitude of a scoliotic curve is determined by its Cobb angle, calculated by (1) identifying the end vertebrae of the curve (the last vertebra on each end of the curve that tilts into the curve’s concavity), (2) drawing a line parallel to the superior and inferior endplate of the rostral and caudal end vertebrae, respectively (solid lines), (3) drawing a line perpendicular to each endplate line (dotted lines), and (4) measuring the angle created by the intersection of these perpendiculars. Note that the disc spaces are narrower on the concave aspect of the curve than on its convex side.
raphic anomalies, diastematomyelia, and tethered cord, may be visualized directly. Intravenous administration of gadolinium is often helpful to define the extent of spinal cord neoplasms and to differentiate between syrinx cavities and tumor-associated intramedullary cysts (Fig 2). Imaging of the craniovertebral junction may disclose Chiari malformation, syringobulbia, or basilar invagination. The nature and extent of spinal cord or nerve root compression from pathology within the epidural space, including intervertebral disc herniations, spinal tumors, and infectious/inflammatory lesions is readily apparent, owing to the exquisite soft-tissue imaging characteristics of MRI.

**Pulmonary Function Testing.** Assessment of pulmonary function is frequently indicated in patients with spinal deformities to identify and quantify respiratory compromise, particularly when surgical intervention is contemplated. The primary deficiency observed in scoliotic patients consists of restrictive pulmonary dysfunction with minimal or no obstructive component. Spirometric analysis of lung volume in patients with thoracic or thoracolumbar scoliosis indicates a preferential reduction in the vital capacity with relative maintenance of residual volume, contributing to an overall decrease in total lung capacity.\(^{38}\) Thus, the single most representative pulmonary function test to assess the degree of respiratory impairment is the vital capacity.

In general, the severity of pulmonary dysfunction is directly proportional to the magnitude of the thoracic curvature. Patients with thoracic curves measuring 80° or less are typically asymptomatic.\(^ {38}\) Dyspnea on exertion is often seen when the curve approaches 100°. Curvatures exceeding 120° are associated with alveolar hypoventilation, carbon dioxide retention, pulmonary hypertension, and eventually, cor pulmonale.\(^ {39}\) The cause of scoliosis is also an important factor in predicting the severity of respiratory compromise. For a given degree of curvature, patients with paralytic scoliosis have lower total lung capacities, functional reserve capacities, and vital capacities than do those with nonparalytic scoliosis due to weakened respiratory musculature.\(^ {39}\) Preoperative pulmonary function testing permits stratification of potential surgical patients by operative risk as the severity of restrictive pulmonary disease correlates well with surgical outcome and overall prognosis.

**Other Diagnostic Studies.** Depending on the clinical setting, other diagnostic studies may be required in the course of evaluating the patient with neuromuscular scoliosis. Electrocardiography should be performed when entertaining diagnoses, such as Friedreich's ataxia, Duchenne's muscular dystrophy, and myotonic dystrophy, due to concurrent cardiomyopathy and conduction abnormalities. Electromyography, nerve conduction studies, serum creatine phosphokinase levels, and muscle biopsy are standard when assessing scoliotic patients presenting with myopathy or peripheral neuropathy. Disorders of metabolism leading to conditions, such as homocystinuria and the mucopolysaccharidoses, may be confirmed with appropriate laboratory investigations.

**THERAPEUTIC STRATEGIES**

Although careful observation may be reasonable under certain clinical conditions for children with neuromuscular scoliosis, most require treatment to stabilize or reverse curve progression and its sequelae. Observation is appropriate only for those patients with mild curvatures, or those in whom the neuromuscular condition is nonprogressive. For patients who meet these criteria, frequent clinical and radiographic follow-up at regular intervals is mandatory. For those who do not, active treatment is indicated. Therapeutic strategies may be categorized as operative or nonoperative; the latter consists mainly of seating supports and external bracing. Surgical treatment may be further subdivided into that which addresses the primary neuromuscular disorder and that which endeavors only to correct the secondary spinal deformity. However, in practice, these subdivisions prove to be artificial, as correcting the underlying neurological condition (if feasible) frequently leads to improvement or resolution of the scoliotic curvature.\(^ {40}\)

**Orthotics**

Selected patients with curves measuring less than 30° and intact somatosensory function may be candidates for orthotic bracing as a first-line therapeutic option. A variety of custom-contoured orthoses are available, including the modified Milwaukee brace and the thoracolumbosacral orthosis (TLSO). In infants, a Kallabis brace may be used. In contrast to idiopathic scoliosis, in which braces are used to apply reductive forces to lessen the magnitude of a curvature, bracing in neuromuscular scoliosis assumes a more passive role, supporting the trunk and allowing for greater use of the upper extremities in daily activities. Only patients
with flaccid or mildly spastic forms of paralysis are considered; severe spasticity precludes orthotic treatment. In general, the goal of bracing is to retard curve progression in the young child while allowing for spinal growth and awaiting a more optimal age for spinal surgery, approximately age 10 years in females and age 12 years in males. However, caution must be exercised because improperly fitted braces may create pressure ulcerations or may restrict chest wall expansion, further exacerbating restrictive pulmonary insufficiency.

**Surgery**

Although nonoperative treatment may be undertaken as a palliative or temporizing measure, most patients with neuromuscular scoliosis will require surgical intervention at some time during their course of treatment. The rationale for operative treatment in this setting embraces two often complementary objectives: (1) to remedy the primary neurological condition inducing the spinal deformity, and (2) to correct the spinal curvature itself. Occasionally, both objectives may be addressed satisfactorily with a single operation; however, two or even a series of surgical procedures are frequently required.

Whenever feasible, the patient’s underlying neurological disorder should be addressed surgically before entertaining spinal stabilization. In simplistic terms, reversible conditions should be promptly reversed: the tethered cord should be released, the syrinx obliterated, the herniated hindbrain (Chiari’s malformations) reduced, the compressed spinal cord decompressed, and the spinal neoplasm resected. If the primary neurological abnormality is expeditiously and satisfactorily corrected and the degree of spinal deformity is relatively mild, no further surgical intervention may be required as the scoliotic curvature may spontaneously regress. This is particularly relevant in the very young patient, in whom the spinal column is immature and significant growth potential remains. Often, however, the neuromuscular abnormality is progressive and not amenable to surgical cure. In such cases and when the existing spinal deformity is severe, operative intervention may be indicated to stabilize or reduce the scoliotic curve.

Indications for operative reduction and internal fixation of the scoliotic spine include the following: (1) a curve measuring 30° or greater; (2) progressive curvatures; (3) curvatures that impair sitting or ambulatory capability; (4) thoracic deformity that compromises cardiopulmonary function; or (5) intractable pain. Although a comprehensive discussion of the various operative methods used for curve correction lies outside the scope of this text, three basic surgical principles are briefly reviewed.

First, if operative stabilization is to be undertaken, what type of instrumentation should be incorporated into the construct? Patients with neuromuscular scoliosis frequently suffer from severe osteopenia. Consequently, segmental fixation techniques using sublaminar wires or cables are, in general, superior to distraction/compression hook-rod constructs or constructs that rely on bone screw fixation at the bone-implant interface. The sublaminar wire and rod system introduced by Luque possesses superb biomechanical strength and boasts favorable clinical outcomes when used to correct scoliotic deformities in this setting. Such constructs achieve 40% to 60% curve correction; the incidence of pseudarthrosis ranges from 6.5% to 13%. Neurological complications resulting from the passage of sublaminar wires are encountered in 1.4% to 13.2% of patients, but are characteristically mild and transient when they do occur. Concern for the latter has prompted some to use interspinous process rather than sublaminar wires to reduce the risk of iatrogenic neurological compromise.

Second, which levels should be incorporated into the fusion? In general, fixation should extend at least as high as the T3 level, owing to the relative frequency of rostral curve progression when the construct ends at T4 or below. Neurological complications resulting from the passage of sublaminar wires are encountered in 1.4% to 13.2% of patients, but are characteristically mild and transient when they do occur. Concern for the latter has prompted some to use interspinous process rather than sublaminar wires to reduce the risk of iatrogenic neurological compromise.

Second, which levels should be incorporated into the fusion? In general, fixation should extend at least as high as the T3 level, owing to the relative frequency of rostral curve progression when the construct ends at T4 or below. The sacrum is incorporated into the fusion if the curve itself involves the sacrum (the so-called Galveston technique). Sacral extension may also be considered in the face of severe pelvic obliquity.
Third, which operative approach (anterior, posterior, or combined) should be used to achieve spinal fixation? Traditionally, all spinal fusions for correction of scoliosis were applied posteriorly; at present, most still are accomplished via a posterior approach. Anterior procedures with or without instrumentation may be considered in the following settings: (1) in the presence of severe coexistent kyphosis; (2) when the incidence of successful fusion would be increased substantially over that obtainable with the posterior approach alone; or (3) if progressive spinal deformity is anticipated to result from continued anterior vertebral growth in the presence of a solid posterior fusion (the so-called “crankshaft phenomenon”). However, regardless of approach, several authors have reported increased surgical morbidity in the subgroup of patients with neuromuscular scoliosis, particularly in terms of infection at the operative site.

**SPECIFIC CONDITIONS**

Several neuromuscular conditions warrant specific mention owing to their frequent association with scoliosis and other spinal deformities. For most clinical scenarios, the general diagnostic and treatment principles outlined above may be applied: (1) prompt identification of the underlying neuromuscular condition; (2) early surgical correction of the primary condition, if feasible; (3) palliative orthotic bracing to improve functional capacity and postpone surgical intervention for curve correction; and (4) operative spinal fixation for persistent or progressive curvatures as required. The following does not represent a comprehensive summary of neuromuscular disorders associated with scoliosis; rather, conditions encountered with relative frequency, or those with a notable propensity for spinal deformity are presented.

**Cerebral Palsy**

The prevalence of scoliosis in patients with cerebral palsy varies substantially with the population studied. Scoliosis is encountered in at least 20% of this cohort; in institutionalized study groups this prevalence is much higher, approaching 70%. Patients with profound spastic quadriplegia or quadriplegia are at greatest risk of developing severe, progressive spinal deformity. Those who ambulate independently appear to be less prone to develop this complication. When present, scoliosis may have devastating functional ramifications in this population, already challenged by physical debility. Severe curvatures may impair ambulation in those so able, or may create pelvic obliquity, sitting intolerance, and pain among nonambulators.

The severity of spinal deformity in this population is also quite variable, ranging from very mild to extensive. Most major curves involve the thoracic or thoracolumbar spine, although a significant minority are purely lumbar. The latter is associated with a high incidence of pelvic obliquity. Like many of the neuromuscular scolioses, the risk of curve progression persists beyond skeletal maturity. Such progression is more likely with curves exceeding 50° at the end of adolescence.

Operative spinal fixation is considered for those patients in whom progressive scoliosis is responsible for loss of ambulatory capacity, decreased sitting tolerance, or intractable pain. Segmental fixation methods incorporating sublaminar wires are generally used; hook-rod constructs (ie, Harrington rods) are associated with a higher incidence of failure in this population. With segmental fixation, curve correction of roughly 50% to 75% may be anticipated, with pseudarthrosis rates of less than 10%. Surgical complications are most prevalent in patients with severe spasticity and mental retardation, and those with athetoid cerebral palsy.

**Spinal Dysraphism**

Scoliosis may be identified in more than 80% of patients with myelodysplasia by the tenth year of life. The cause of spinal deformity in this population is variable. In some, the scoliosis is congenital, a consequence of anomalous vertebral development (segmentation failure, defects of formation). In others, spinal curvature is developmental (paralytic), arising secondary to concomitant abnormalities of the neural axis including hindbrain malformation, hydrocephalus, syringohydromyelia, diastematomyelia, and spinal cord tethering (Figs 3 and 4).

Similar to the child with cerebral palsy, the myelodysplastic child who develops progressive spinal deformity is often faced with inexorable functional decline. Ambulatory patients may lose the capacity to walk independently or may require cumbersome orthotic supports. Nonambulators may suffer from truncal collapse, pelvic obliquity, diminished sitting tolerance, integument breakdown, pain, and cardiopulmonary compromise.
Spinal deformity in this patient population may reach impressive proportions, with lateral curvatures often exceeding 100°. The curve typically involves the thoracic and upper lumbar levels, but may extend to the cervical regions and sacrum as well. The location of the myelodysplastic defect has strong predictive value in identifying those children with a propensity to develop spinal deformity. As a rule, the more rostral the defect, the higher the incidence of scoliosis. Children with thoracic or high lumbar motor levels manifest scoliosis in nearly 90% of cases, whereas such deformity is observed in only 50% at L3-L4, 20% at L5, and 5% at S1-S2.
Nonoperative treatment for scoliosis in the myelodysplastic patient is controversial, as most curves managed in this manner will progress. Curves less than 20° may be observed expectantly with serial radiographs obtained every 4 to 6 months. Bracing with a thoracolumbosacral orthosis should be instituted when the curve reaches 20°. However, orthoses typically serve only to slow curve progression and postpone surgery in an attempt to afford greater skeletal maturity at the time of spinal fixation. Indications for operative fixation include curve progression despite bracing, intolerance of orthotics, curvatures exceeding 40°, and progressive functional decline attributable to spinal deformity. As with most neuromuscular curves, stabilization is best accomplished by posterior fusion with segmental instrumentation. The addition of an anterior construct as well may diminish the risk of pseudarthrosis. Operative stabilization of a scolicotic curvature should be undertaken only after a meticulous clinical and radiological investigation has excluded the presence of an associated neural axis anomaly. Frequently, correction of such pathology (ie, hydrocephalus, Chiari malformation, syringomyelia, tethered cord) is sufficient to stabilize or reverse progressive spinal deformity and obviate the need for spinal fixation.

**Syringomyelia**

Syringomyelia may be encountered in association with a variety of neurological disorders, including spinal dysraphism, Chiari malformation, traumatic spinal cord injury, myelitis, meningitis, and arachnoiditis. Irrespective of cause, progressive scoliosis may accompany syringomyelia in up to 85% of cases. Curvatures are typically mild, but occasionally will exceed 50° and almost invariably involve the thoracic or thoracolumbar spine. Of note, leftward-projecting thoracic curves are far more common in this population than in patients with idiopathic scoliosis, in whom this configuration is decidedly rare.

In patients presenting with syringomyelia and progressive scoliosis, initial treatment consists of obliteration of the syrinx. If the syrinx is primary, simple surgical drainage usually suffices. When secondary, the underlying condition should be sought and corrected. Resolution of syringomyelia...
is associated with stabilization or, in some cases, improvement in the degree of scoliotic curvature.\textsuperscript{78} Spinal fixation procedures should be reserved for progressive spinal deformities refractory to syrinx obliteration, as paralysis may occur as a consequence of attempted operative curvature reduction in the face of persistent syringomyelia.\textsuperscript{74,79}

\textit{Chiari Malformation}

The association between scoliosis and Chiari malformation is well established in patients with spinal dysraphism. However, progressive scoliosis may be identified in conjunction with Chiari malformations in the absence of myelodysplasia as well (Fig 5). In one series, the prevalence of scoliosis exceeded 70\% in patients with symptomatic hindbrain herniation presenting before skeletal maturity.\textsuperscript{40} Spinal deformity in this population is typically mild, averaging approximately 30\(^\circ\), and characteristically involves the thoracic or thoraco-lumbar spine.\textsuperscript{40} Syringomyelia may or may not be present.

Successful treatment requires prompt surgical correction of the underlying abnormality, the Chiari malformation, with a dorsal posterior fossa decompressive procedure. Cerebrospinal fluid diversion via a fourth ventricle to cervical subarachnoid space shunt is required when syringomyelia coexists. With this approach, resolution of spinal deformity may be anticipated in nearly 80\% of patients.\textsuperscript{40} Children less than 10 years of age enjoy superior results in terms of the rate and extent of curve correction. Operative spinal fixation may be considered for the rare patient whose curve fails to respond following posterior fossa decompression, but should be avoided as a primary procedure.\textsuperscript{40}

\textit{Spinocerebellar Degeneration}

The prototypical spinocerebellar degenerative disease in the pediatric population is Friedreich's ataxia. This heritable disorder is manifest in patients aged 5 to 20 years with progressive gait instability, cavus foot deformities, areflexia, and impaired dorsal column function.\textsuperscript{80} As a result,
most children with this condition are wheelchair-bound within the first or second decades of life. Death generally occurs in the third or fourth decades, usually the consequence of progressive cardiomyopathy.81

Scoliosis is present in more than 75% of children with Friedreich’s ataxia. Most curvatures involve the thoracic or thoracolumbar spine and project towards the right, often in association with severe kyphosis. When scoliosis arises before adolescence, the curve tends to be rapidly progressive. Generally, orthotic bracing is ineffectual in halting curve progression in these patients and may actually be detrimental, as delays in surgical intervention are accompanied by further impairment of myocardial function. Surgical intervention should be considered once the magnitude of curvature exceeds 40° or in patients with rapidly progressive deformities. Segmental fixation techniques using sublaminar wire-and-rod constructs are preferable.82,83

**Spinal Cord Trauma**

Children who have sustained partial or complete paralysis as a consequence of spinal cord trauma frequently develop progressive spinal deformity as a late sequela of their injury. Although a multifactorial etiology is probable, paralysis of the postural muscles of the spine caudal to the level of injury and direct trauma to the spinal epiphyseal growth plates are factors putatively implicated in the pathogenesis of this condition. The most significant predictor of progressive post-traumatic spinal deformity is the child’s age at the time of cord trauma. If injured before the onset of rapid adolescent growth, the risk of spinal curvature is far greater than that experienced by older children, exceeding 90% in this younger population.84-87 Additional independent risk factors predisposing to progressive deformity include previous thoracic or thoracolumbar laminectomy, spasticity (even in older patients), and severe or complete motor deficit.84-87 Interestingly, the anatomical level of neurological injury appears to have little influence on the subsequent development of scoliosis.85,86

Prompt medical and surgical treatment must be instituted to prevent substantial functional decline in patients with progressive spinal deformity following spinal cord injury. Prophylactic bracing with a suitable thoracolumbosacral orthosis may retard curve progression and improve sitting tolerance. Generally, bracing should be instituted within 6 months of injury to yield maximal efficacy.84 However, despite aggressive bracing regimens, operative spinal fixation is required in most patients to stabilize or reverse progressive spinal deformity and preserve functional capacity. In such cases, surgical constructs that incorporate segmental spinal instrumentation and bony fusion are advocated.84,86

**Spinal Muscular Atrophy**

Spinal muscular atrophy is a group of heritable (autosomal recessive) disorders of the spinal cord ventral horn cells that result in hypotonia, areflexia, and progressive weakness, preferentially involving the proximal and axial musculature. At least three variants are described, with distinctions based on age of presentation and rapidity of disease progression. Werdnig-Hoffman disease is manifest within the first few months of life and is relentlessly progressive, leading to death by age 2 years in most cases. An intermediate form, known as “arrested” or “chronic” Werdnig-Hoffman disease, also presents in infancy but pursues a less aggressive course. Juvenile spinal muscular atrophy, or Kugelberg-Welander disease, generally presents between the ages of 2 and 12 years, yielding progressive disability and death by early adulthood. Children with Kugelberg-Welander disease may be ambulatory, at least for a few years; those with infantile or arrested Werdnig-Hoffman disease never walk.88

Scoliosis is prevalent in children with spinal muscular atrophy; estimates range from 30% to 90%.83,89-91 Long thoracic and thoracolumbar curvatures are typical, although other curve types may be observed. Curve progression is most rapid during the preadolescent growth spurt and may impair ambulation in those so able. Severe pelvic obliquity may result in decreased sitting tolerance and decubitus ulcers in nonambulators.

The role of bracing in the management of spinal muscular atrophy-associated scoliosis is controversial; generally, bracing serves only to retard curve progression in the young child and is not effective as a long-term therapy.89-92 Surgical stabilization is recommended once the curvature reaches 40°. Long fusions incorporating the rostral thoracic spine and the pelvis prevent curve progression above and below the level of fusion and improve
pelvic obliquity. Segmental instrumentation incorporated with sublaminar wires is most effective in these severely osteopenic children. With these techniques, curve correction averages approximately 50%.

Duchenne's Muscular Dystrophy

Duchenne-type muscular dystrophy is an X-linked recessive myopathic disorder that characteristically affects boys younger than 5 years old, presenting with muscle weakness and wasting in the proximal lower extremities and pseudohypertrophy of the calves. Progressive weakness leads to ambulatory impairment and eventual wheelchair dependence by (on average) age 10 years. Interestingly, the onset of wheelchair dependence usually heralds the beginning of progressive scoliosis, a condition encountered in up to 95% of patients with the disease.

Spinal curvatures in children with Duchenne's muscular dystrophy typically involve the thoracic or thoracolumbar regions. Curves frequently extend to the lumbosacral spine as well, resulting in pelvic obliquity and poor sitting tolerance. Invariably, spinal deformity is progressive, increasing roughly 20° per year with the greatest rate of progression observed during puberty. The treatment of scoliosis in these children is primarily surgical; bracing affords no lasting benefit. Owing to the inexorable progression of deformity in this population, surgery is entertained once the curve reaches 30°. The Luque-Galveston fixation technique using sublaminar wires and rods that extend from the rostral thoracic spine to the pelvis is the preferred method of stabilization in most cases; failure to extend the fusion rostrally and caudally results in postfusion curve progression and progressive pelvic obliquity, respectively.

Surgical intervention is not without morbidity in this population. Blood volume loss exceeds 2.5 L in most cases as hemostasis is impaired by the dystrophic smooth muscle that comprises blood vessel walls. Rhabdomyolysis and hyperkalemia may result from the use of depolarizing muscle relaxants as a component of general anesthesia, and thus should be avoided. The prevalence of concurrent cardiomyopathy in this population is high, and its severity may not correlate with the degree of skeletal muscle involvement. Preoperative cardiac evaluation and perioperative electrocardiographic monitoring are obligatory. Pulmonary insufficiency may be profound, with vital capacities of less than 30% of predicted values not uncommon. Some authors have recommended prophylactic tracheostomy to facilitate weaning from ventilatory support in the postoperative period. Although this is not routine in most centers, preoperative pulmonary function testing is mandatory to assess the extent of respiratory compromise and high-risk patients and their families should be counseled about the possibility of postoperative tracheostomy. Ideally, these patients should be managed by a coordinated, multidisciplinary team incorporating pediatric intensivists, spinal surgeons, neurologists, cardiologists, and pulmonologists to minimize perioperative morbidity and optimize outcome.

SUMMARY

The child with neuromuscular scoliosis presents a complex array of diagnostic and therapeutic challenges to the pediatric neurologist and neurological surgeon. The following points are germane and deserve reiteration: (1) scoliosis is a physical finding, not a disease process, and as such an underlying etiology should be sought; (2) spinal deformity arising as a consequence of neurological or neuromuscular disorders is characterized by early onset, rapid progression, and refractory course; (3) plain spinal radiographs demonstrate the nature and extent of deformity and may reveal clues to the primary diagnosis; (4) when a neuromuscular etiology is suspected, MRI of the neural axis is the diagnostic procedure of choice; (5) initial therapeutic interventions should target the underlying neurological or neuromuscular disorder as characterized by early onset, rapid progression, and refractory course; (6) orthotic bracing may slow curve progression but rarely is effective as long-term treatment; (7) surgical fixation is undertaken to correct curvatures that are large, progressive, or functionally debilitating; (8) segmental instrumentation using Luque rods and sublaminar wires provides superior fixation to other constructs and should incorporate the rostral thoracic spine and, in most cases, the pelvis for optimal stabilization; and (9) successful spinal stabilization corrects deformity, preserves function, facilitates care, and enhances the quality of life for patients suffering from neuromuscular disorders. Early diagnosis and prompt treatment are crucial to realize these goals.


88. Shapiro F, Bresnan MJ: Orthopaedic management of...