Early Onset Neuromuscular Scoliosis

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15.1 Definition

Early-onset scoliosis is associated with a variety of neuromuscular disorders. In comparison to other forms of scoliosis, neuromuscular scoliosis often has an early onset with rapid progression during growth, which may continue to progress following skeletal maturity. It is more likely to present with severe spinal deformities that frequently involve the sacrum, and may have associated pelvic obliquity; respiratory compromise is frequent. The associated disorders are broadly classified as either neuropathic or myopathic (Chap. 20).

15.2 Natural History

Progression throughout life is common as well as a respiratory compromise; depending on the underlying disorder, progression can be extremely rapid.

Supplementary Information The online version contains supplementary material available at [https://doi.org/10.1007/978-3-030-80356-8_15].

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A. Şenköylü, F. Canavese (eds.), *Essentials of Spine Surgery*,
https://doi.org/10.1007/978-3-030-80356-8_15

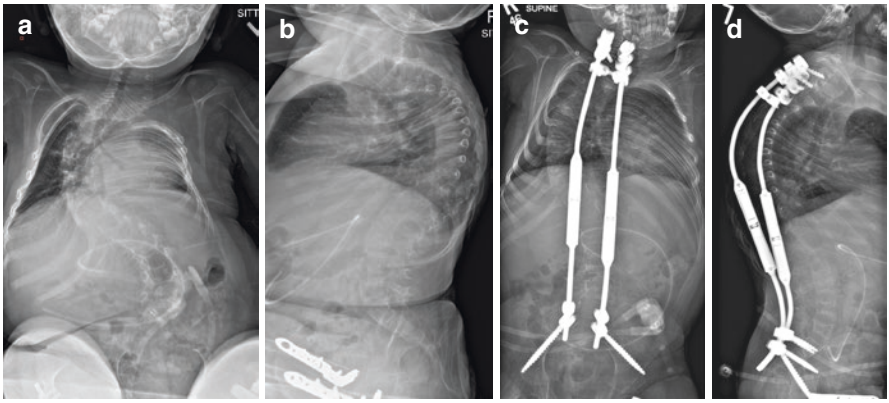

Fig. 15.1 A four-year-old patient with spinal muscular atrophy Type 1 with severe progressive kyphoscoliosis (**a** and **b**), treated with MCGR (**c** and **d**)

15.3 Physical Examination

Neuropathic disorders, including cerebral palsy (CP), spinal muscular atrophy (SMA), myelomeningocele, and Rett syndrome, are more commonly associated with early onset (Videos 15.4 and 15.5). In contrast, some patients with myopathic disorders such as Duchenne's muscular dystrophy are most likely to develop scoliosis after the age of 10 years old. Curve progression in all may continue after skeletal maturity, and a respiratory compromise is frequent. Patients with SMA are notable in that age of scoliosis onset may occur as early as less than 2 years of age (Fig. 15.1). Earlier onset is predictive of more severe development of scoliosis. Patients with myelomeningocele may develop tethered cord syndrome following surgical repair, which may be a cause of progressive scoliosis. Management of these patients is complicated by associated disorders including renal anomalies, and neurogenic bowel and bladder. Successful management of patients with myelomeningoceles requires a multi-disciplinary approach. Patients with Rett syndrome who present for management will be female, as the X-linked mutation leads to death in males within the first 2 years of life. As with other causes of neuromuscular scoliosis, patients with Rett syndrome can develop rapidly progressive scoliosis, which may continue after skeletal maturity.

15.4 Imaging

Radiographic evaluation of scoliosis involves determining the severity of the curve (Video 15.6), evaluating for abnormal vertebrae morphology or number, and obtaining additional imaging to assess complications of the associated syndrome. Spinal magnetic resonance imaging is also indicated in young patients under 10 years of age, those with neurologic deficits, and in patients who present with spinal dysraphism.

15.5 Treatment Options

Nonoperative management. Several goals of care, including maintenance of function and delay of surgical intervention (e.g., casting; Video 15.5), can be assisted by non-operative interventions. Bracing can be used to control the progression of curves during growth periods and delay the need for surgery until it can be performed safely. Patients with pulmonary conditions should be closely monitored, as bracing can cause decreased chest wall expansion. Wheelchair modifications, such as trunk support and posterior contouring, allow patients to sit in a functional position and maintain more independence. Frequent skin evaluations in patients treated with bracing or wheelchair modification are important in preventing the development of skin breakdown and pressure ulcers.

Intrathecal Nusinersen injections have improved the prognosis of SMA Type 1 and 2 patients. The medication is delivered directly to the central nervous system, which allows it to be distributed to the spinal motor neurons located in the spinal cord.

Surgery. Surgical intervention may be considered in patients with rigid curves greater than ~60° to 70°. A curve may be considered rigid when it has <50% of flexibility. Similar to conservative management, the goals of surgical intervention are to control curve progression and provide stability in the coronal and sagittal plane. Benefits include decreased pain, improved pulmonary function, improved seat positioning, and increased functional independence. Surgical options can be broadly categorized into definitive fusion and growth-friendly non-fusion techniques. In DMD patients cardiac function is an important parameter to consider when surgery is planned; anticipation is the best strategy.

Compared to idiopathic and congenital scoliosis, risks of surgery are greater in patients with neuromuscular scoliosis as a result of their underlying disorders. A preoperative assessment including a detailed history and evaluation of cardiac, respiratory, neurological, and urologic function should be performed. Furthermore, nutritional status and possible concomitant metabolic bone disease should be assessed.

The use of non-fusion surgical techniques has become favored in the treatment of early-onset scoliosis before a skeletal age of about 9 years as, compared to fusion, non-fusion techniques allow for the continued spine and chest growth. In addition to traditional growing rods, many non-fusion options are available, including magnetically controlled growing rods (MCGR), vertical expandable prosthetic titanium rib (VEPTR), Shilla growth guidance, self-expandable domino mechanical growing rods, and localized fusion in myelomeningocele (Videos 15.3 and 15.7). Compared to traditional growing rods, MCGR and the Shilla technique reduce the need for multiple operations to expand the construct as the patient grows. VEPTR constructs may be helpful in improving pulmonary function but require surgical adjustment every 6–8 months, increasing the risk of complications.

The extent of pelvic obliquity should be assessed in patients with neuromuscular scoliosis. Pelvic obliquity can cause uneven weight distribution while patients are sitting, leading to pressure ulcers and decreased sitting tolerance due to pain. Pelvic

fixation may be performed with fusion or as a part of a growing rod construct. The typical criterion for pelvic fixation is pelvic obliquity >10° to 15°, as measured on the anteroposterior radiograph.

Definitive surgical fusion is commonly viewed as the endpoint of treatment for neuromuscular scoliosis. Nevertheless, with the increased use of growth-friendly techniques, definitive instrumentation and fusion may not be necessary for all patients (Videos 15.1 and 15.5). Many patients treated with growth-friendly techniques undergo varying degrees of autofusion. Thus, patients with evidence of autofusion, minimal gain in length during their final distraction, and no implant-related complications can potentially be observed rather than undergo definitive fusion.

15.6 Expected Outcomes

Neuromuscular scoliosis, compared to idiopathic scoliosis, is less likely to respond to bracing and casting measures. Surgical intervention in these patients is associated with a greater risk of deep surgical infections, a longer length of hospital admission, and increased risk of pulmonary embolism and neurologic injury.

In comparison to idiopathic and congenital scoliosis patients, patients with neuromuscular scoliosis have high rates of complications associated with both treatment and their underlying disease state. Neuromuscular scoliosis patients, especially those with CP, have high rates of infection. Furthermore, patients may experience pulmonary insufficiency due to severe scoliosis curvature or associated deformities such as kyphosis. Uneven sitting posture and treatment with bracing increase risks of skin breakdown and pressure ulcers. Poor nutritional status in these patients is associated with decreased immune function, increasing the risk of surgical complications such as wound infections and sepsis. A goal serum albumin of >3.5 mg/dL should be reached prior to surgical intervention. Patients treated with growing rods that necessitate repeated distraction may experience progressive stiffness. In addition, instrumentation failure such as rod breakage or loosening of implants may occur. Finally, some curves may rapidly progress despite attempts to control them with growth-friendly techniques, leading to a more severe deformity that necessitates definitive surgical correction and fusion.

Further Readings

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