

Neuromuscular Scoliosis: Current Concepts on Surgical Care



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Submission: September 26, 2017; **Published:** October 03, 2017

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Background

Scoliosis is a common deformity in many types of neuromuscular disease. The orthopaedic surgeon is often the first physician who is required to diagnose and treat a young patient with spinal deformity and neuromuscular disorders. These diseases make to lose the ability to ambulate and determine the progression of scoliosis with collapse of the column. Severe spinal curvature can cause difficulty also in sitting. Conservative and surgical treatment of neuromuscular scoliosis differs from idiopathic scoliosis, being more complex and with a higher complications rate [1]. Non-surgical treatment rarely fully controls progressive deformity of the spine. Corrective surgery requires multidisciplinary management and perioperative difficult screening. Pelvic obliquity is commonly associated with neuromuscular scoliosis, making sitting difficult: correction needs to be considered during surgical planning.

The goal of surgical correction is to obtain and maintain a well-balanced spine above a well-positioned pelvis. Preoperative multidisciplinary study enables potential problems or complications. Respiratory function investigation will guide possible non-invasive perioperative ventilation. Nutritional and psychosocial assessment should also be incorporated in this preparation, as should overall postoperative care. Implementing this overall strategic planning can achieve a good surgical and functional result in the vast majority of cases. Pedicle screws have shown to be a safe and effective method of spinal fixation, offering superior multiplanar correction compared with hooks or sub laminar wires in selected situations [2]. We started over a decade, using only pedicle screw instrumentation for the surgical treatment of scoliosis. The positive results have led us to extend this technique with some certainty to the treatment of the most severe deformities such as neuromuscular scoliosis.

Methods and Results

We studied a consecutive series of 48 patients with neuromuscular scoliosis operated in our center only through a posterior instrumented fusion. They were 25 females and 23 males, mean age 15.8 years (min 10 - max 38 years) [3]. Cerebroathic spasticity in 17 cases, Duchenne muscular dystrophy in 8 cases, myelomeningocele in 6 cases, spinal muscular atrophy (SMA) in 8 cases, poliomyelitis in 3 cases, Friedreich's ataxia in 2 cases, Escobar syndrome in 2 cases, Steinert's myotonic dystrophy in 1 case and Charcot Marie Tooth's neuropathy in 1 case. The average angle value of the main scoliotic curve was 94.05° (34° min - max 165°), the headquarters of the curve was thoracic in 23 cases, thoracolumbar or lumbar in 25 cases. The angular value of the average thoracic kyphosis was 42.86° (7° min - max 90°), while that of the lumbar lordosis was 33.57 [4].

The posterior instrumented fusion, using only pedicle screw instrumentation (Figures 1A & 1B) was extended to the lumbar spine in 25 patients, up to the sacro-ileum with iliac screw in the other 23 patients (Table 1). All the instruments were made of titanium. The mean surgery time in 25 patients with lumbar fusion up was 4, 5 hours, while in 23 patients with fusion until ileum was 5 hours [5]. IOM was performed in 7 patients (3 male, 4 females) who presented mild to moderate motor deficits before surgical treatment [6]. Preoperative SEPs were pathologic in 1 patient and tce-MEPs amplitudes were reduced in 1 patient. Only one patient demonstrated reversible SEPs and tce-MEPs amplitude changes at left lower limb during surgery, with no postoperative sensory-motor or sphincteric deficits [7]. The complications that have been noted are minor type (Table 1) and led to a reoperation in 7 cases (14.5%).

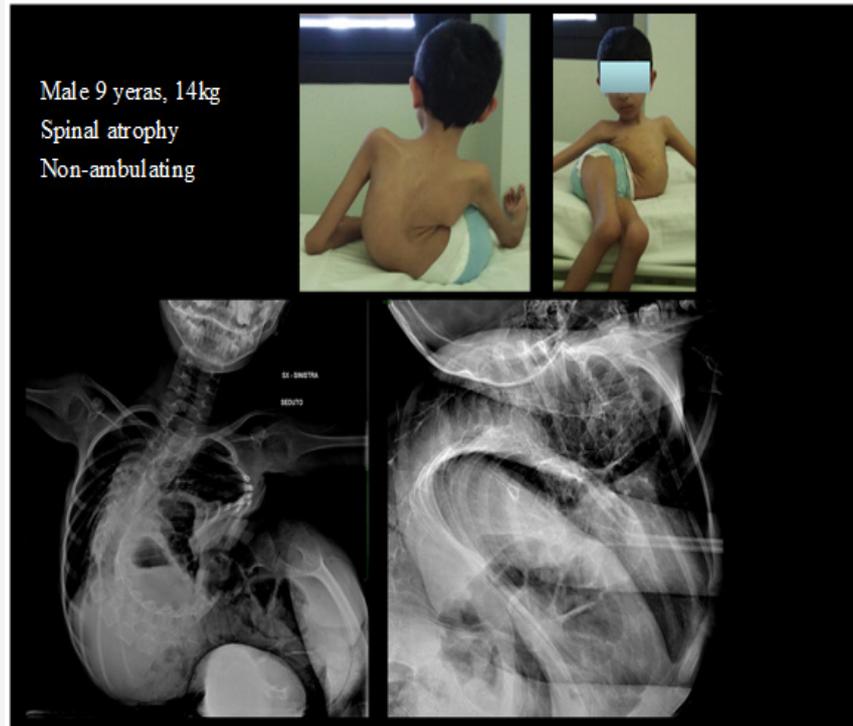


Figure 1A: Spinal atrophy Non-ambulating.



Figure 1B: Posterior instrumented fusion T4-Sacrum-ileum.

Table 1: Preoperative multidisciplinary study enables potential problems or complications.

COMPLICATIONS

Fusion up to the lumbar spine 25 cases	Fusion to the sacro-ileum 23 cases
6 Complications (24%)	2 Complications (8,6%)
3 mobilization of hooks	1 dural lesion

1 breakage of the instrumentation	1 deep infection
1 loss of correction	
2 proximal junctional kyphosis (PJK)	
5 Revision Surgery	2 Revision Surgery
2 replacements hooks mobilized	1 repair of the dura
1 replacement of instrumentation and fusion revision	1 debridement and removal of the hardware
2 proximal extension of arthrodesis	

Conclusion

Surgical treatment of neuromuscular scoliosis require a careful study of social and medical preoperatively aspects. The positive results with improvement of quality of life in patients with neuromuscular scoliosis, can be obtained when the decision takes into account for the treatment of multi-disciplinary assessments, this means where the spinal surgeon, cardiologist, neurologist or neuro-psychiatrist and a physiatrist participate all the decisions together. The surgery should be performed as early as possible; however, the sacrum and pelvis fixation should be avoided in patients with residual walking ability [8]. Surgical management of spinal deformity in neuromuscular diseases (NMDs) often requires a multidisciplinary approach beginning in the preoperative surgical planning period, owing to concomitant restrictive lung disease and cardiomyopathy in selected NMD conditions [9].

The need for thorough and thoughtful discussions must occur with the family and other caregivers before any scheduled surgery. The decision to proceed with spinal instrumentation may alter functional abilities in weak and marginally ambulatory NMD patients. With care and treatment involving a multidisciplinary team, proper planning, and support, patients will likely experience rewarding outcomes and improved quality of life. The instrumented posterior fusion alone proved to be an effective technique in neuromuscular scoliosis, and is seems the treatment of choice for patients with limited lung function.

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DOI: [10.19080/OROAJ.2017.08.555746](https://doi.org/10.19080/OROAJ.2017.08.555746)

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