Spine Deformity

www.spine-deformity.org

Spine Deformity 6 (2018) 607−613

Achievement of Guided Growth in Children With Low-Tone Neuromuscular Early-Onset Scoliosis Using a Segmental Sublaminar Instrumentation Technique

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Received 28 August 2015; revised 19 February 2018; accepted 24 February 2018

Abstract

Objectives/Summary of Background Data: Segmental sublaminar spinal instrumentation without fusion results in guided growth and correction of deformity in early-onset scoliosis (EOS). The purpose of this study is to report the outcomes of a series of children with low-tone EOS that were treated surgically with a modification of a Luque trolley technique.

Methods: This study is a retrospective chart and radiographic review of a single-center series of 13 consecutive children who met inclusion criteria with documented progression of scoliosis greater than 25°. All children received surgical treatment with guided growth without fusion using a modified Luque trolley technique. The children's preoperative, postoperative, and most recent radiographs were assessed for Cobb angle, T1−T12 and T1−S1 height, and sagittal alignment including proximal junctional kyphosis. Surgimap spine software was used for calibration and measurement purposes. Complications and need for repeat and/or secondary surgical procedures were recorded.

Results: The mean age at surgery was 7.4 years (4.6−10.5). On average, 15 segments (13−16) were instrumented. None of the children went on to a spontaneous fusion, and the average growth rate per year from T1−T12 and T1−S1 was 0.9 cm/y and 1.5 cm/y, respectively. The mean total growth from T1−T12 and T1−S1 was 22.3 cm (16.6−30.2) and 37.5 cm (30.1−46.4). A total of three additional surgeries were needed in two children to address complications. There were no mortalities.

Conclusions: Sublaminar guided growth is a safe and effective treatment in the Low Tone Neuromuscular subset of EOS. Follow-up studies failed to show signs of auto fusion, and implant failure was not observed in our cohort. All children displayed growth post-operatively without the need for multiple distraction-based surgeries. Guided growth minimizes the risks associated with multiple surgical procedures while maintaining correction and allowing for near-normal rates of spinal growth.

Keywords: Early-onset scoliosis; Neuromuscular scoliosis; Segmental Sublaminar Spinal Instrumentation

Introduction

Early-onset scoliosis (EOS) is defined as a coronal Cobb angle greater than 20° in a child less than 10 years of age. The EOS classification divides children with spinal curvatures into Structural/Congenital, Low/High Tone Neuromuscular, Syndromic, and Idiopathic types [1]. Conditions that constitute the Low/High Tone Neuromuscular division of the classification often include myopathies and muscular dystrophies, cerebral palsy, and paralytic conditions.

One such low tone neuromuscular condition is spinal muscular atrophy (SMA). Clinical manifestations of SMA occur secondary to degeneration of the anterior horn cells of the spinal cord leading to symmetrical muscle weakness and atrophy of the trunk and proximal musculature of the shoulder and hip girdle [2]. One of the greatest problems faced by the orthopedic surgeon caring for patients with

Author disclosures: SR (consultant from Medicrea Spine and Ortho-Pediatrics Spine), JS (other from Arthrex, Inc., Naples, FL, outside the submitted work), BS (none).

IRB approval granted.

Sources of support and funding: None.

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2212-134X/$ - see front matter
https://doi.org/10.1016/j.jspd.2018.02.012

SMA is addressing the spinal deformity associated with this condition [3]. The scoliosis associated with SMA is of the neuromuscular type [4], in which the pelvic obliquity is proportional to the severity of the curve. Scoliosis in SMA has an early onset and progresses rapidly prior to the onset of puberty. Respiratory impairment in the form of restrictive lung disease advances simultaneously with the progression of the spinal curvature. In SMA, survival typically depends on the degree of respiratory involvement with atelectasis and pneumonia as the usual causes of death [5]. The outcomes of orthotic management to slow or halt curve progression have been poor and further contribute to restrictive pulmonary disease [6,7].

Surgical treatment of the spinal deformity associated with SMA and other low/high-tone neuromuscular EOS conditions has become the treatment of choice. For children with EOS, Karol et al. suggested that spinal arthrodesis be performed once thoracic vertebrae 1–12 height is greater than 22 cm to allow for optimal lung volume, otherwise thoracic insufficiency syndrome will likely develop [8]. DiMeglio showed that the spine grows in a bimodal distribution with rapid growth from zero to 5 years and again after 10 years of age and is completed in girls by 13 and boys by 17 [9,10]. Surgical techniques that have been utilized in the EOS population are distraction-based techniques (vertical expandable prosthetic titanium ribs and growing rods), guided growth (Luque trolley and Shilla), compression-based (tether and staples), and early limited arthrodesis [11]. Many of these techniques allow for growth and usually require multiple surgical interventions.

Sublaminar guided growth without fusion has previously been reported as an option to correct spinal deformity while maintaining spinal growth with varied results [12-15]. Recently, efforts to address the scoliotic deformity associated with EOS have focused on various nonfusion and growing techniques [16]. Over the past 15 years a modification of the Luque trolley technique has been performed on a subset of children with EOS at our institution. The purpose of this study is to report the outcomes of a series of children with neuromuscular and syndromic EOS that were treated surgically with sublaminar guided growth without fusion using a modified Luque technique during their years of peak growth.

Material and Methods

Our Institutional Review Board approved a retrospective chart review from 1998 to 2014. Children aged 10 years or younger who carried a diagnosis of scoliosis with progression of their curve greater than 25° and no prior surgery about the spine were included. Only patients who underwent sublaminar guided growth without fusion and had at least two years' follow-up with preoperative, intraoperative, and postoperative radiographs were included. The senior author performed all surgeries.

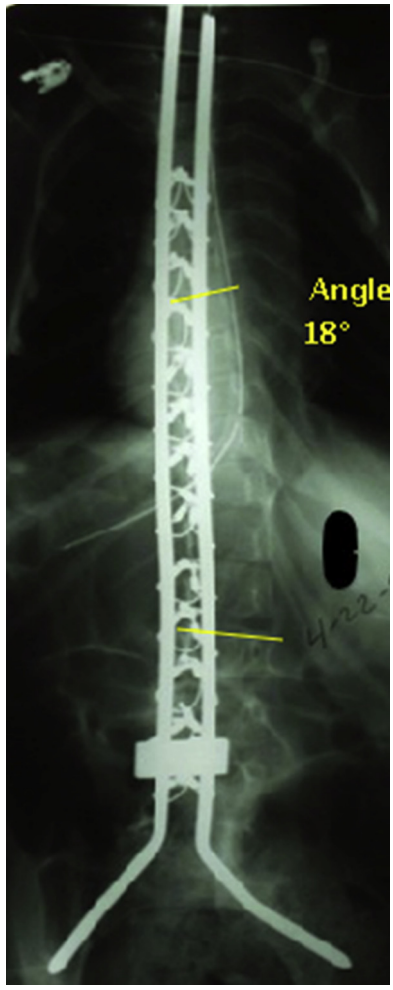

Fig. 1. Child #1 – Intraoperative PA Radiograph, yellow lines represent Cobb angle. Surgimap Spine Software (Nemeric Inc, 306 E 15th Street, New York, NY 10003).

A chart review was conducted and data were collected that included age, sex, length of follow-up, neuromuscular condition, complications, and decline in their pulmonary function studies defined as the need for tracheostomy and/or ventilator dependence post spinal surgery. Pulmonary functions studies were performed every six months for monitoring purposes.

All preoperative, intraoperative, initial postoperative, and most recent follow-up biplanar radiographs were reviewed. Surgimap spine software (Nemeric Inc, New York, NY) was used to measure radiographs. Images were uploaded and the known rod sizes (4.76 mm vs. 5.5 mm) were used for calibration purposes. Radiographic evaluation included coronal plane analysis using the Cobb method and monitoring the cranial end of the construct for sagittal balance. Growth was measured from the anterosuperior endplate of T1 to anteroinferior endplate of T12 and anterosuperior endplate of S1 accordingly via lateral radiographs intraoperatively after instrumentation and then at each visit. The radiographs were assessed for the initial