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Early-Onset Scoliosis: Updated Treatment Techniques and Results

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Abstract

Study Design: This is a review of the current literature on early-onset scoliosis (EOS) techniques and treatment written by the Growing Spine Committee of the Scoliosis Research Society.
Objectives: The Growing Spine Committee of the Scoliosis Research Society sought to update the information available on the definition and treatment of EOS, including new information about existing techniques.
Summary of Background Data: EOS represents a diverse, heterogeneous, and clinically challenging group of spinal disorders occurring in children under the age of 10. Our understanding of EOS has changed dramatically in the last 15 years, and management of EOS has changed even more rapidly in the last five years.
Methods: The Growing Spine Committee of the Scoliosis Research Society has embarked upon a review of the most current literature on EOS techniques and treatment.
Results: This white paper provides recent updates on current techniques, including a summary of new modalities, indications, contra-indications, and clinical results.
Conclusions: Although treatment of EOS is still challenging and complicated, the evolution of options and knowledge presents hope for better understanding and management in the future.
Level of Evidence: Level V.

Keywords: Early-onset scoliosis; Pediatric scoliosis; Spinal deformity; Techniques; Complications

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Introduction

Over the last 10 years, there has been a significant evolution in the knowledge and treatment of early-onset scoliosis (EOS), now defined as spinal deformity in patients less than 10 years of age. In 2012, the rapidly expanding volume of medical knowledge on EOS led the Growing Spine Committee of the Scoliosis Research Society to update the EOS white paper, which has already become outdated [1].

As we have come to better understand the relationship of the chest wall, the lungs, and the heart to the spine, as well as the three-dimensional changes the spine makes over time, treatment has secondarily evolved. In addition to a well-aligned spine treatment care now additionally focuses on creating a well-developed thoracic cavity, and volume, with good overall pulmonary function. The basic tenet in the treatment of EOS centers on maximizing T1–T12 length, which is important in optimizing pulmonary volume. The minimum T1–T12 length needed to assure relatively normal pulmonary and physical function is unknown, however a lower limit of 18 to 22 centimeters has been proposed [2-4]. The purpose of this paper is to review the changing landscape of EOS and provide the reader with a concise overview of current literature.

Materials and Methods

The Growing Spine Committee of the Scoliosis Research Society reviewed the current literature and summarized the most pertinent information related to current treatment of and information about EOS.

Results

Classification system

EOS is a heterogeneous group of varying pathologies, which until recently had no standardized classification system. However, there is now a new classification, C-EOS, designed by consensus of 15 experienced pediatric spine surgeons, which is based on 4 aspects of the deformity: etiology, major curve magnitude, kyphosis, and the annual rate of progression [5]. Two studies have demonstrated overall high reliability across the major categories of etiology (0.84), major curve (0.93), and kyphosis (0.98). The high level of agreement and consistency demonstrated by the C-EOS system shows it can be used as a reliable tool in clinical practice and research [6,7].

Nonoperative treatment options: Derotational casting

Serial casting is an effective growth-preserving technique, delaying or eliminating the need for operative treatment, without compromising the thoracic spine growth or pulmonary development. Although casted, the thoracic spine has been shown to grow at a similar rate as a typical child's [8], permitting pulmonary development to occur. Generally accepted guidelines for casting are curves $>25^{\circ}$, with $>10^{\circ}$ of documented progression, or rib-vertebral angle difference (RVAD) $>20^{\circ}$ [9,10]. Cast treatment, if indicated, should be initiated as early as possible as improved outcomes from casting have been demonstrated in younger patients with lower magnitude deformities. Casts are commonly changed every one to four months, depending on the age of the child, curve magnitude, and diagnosis [10,11]. If complete correction of the curve is achieved, the cast may be discontinued when radiographs demonstrate restoration of rib-cage symmetry and derotation of apical vertebra, followed by observation or bracing [11]. If casting can no longer control the curve, the treatment is typically converted to surgery [9].

Operative treatment options

Distraction-based techniques

Traditional dual growing rods (TGRs). TGRs are distraction-based constructs that allow for growth of the spine while providing stability and deformity correction. The spine fixation, or foundations, include pedicle screws or hooks, with or without pelvic fixation, with lengthening of the constructs at intervals averaging 6 to 8 months [12]. Akbarnia et al. published the largest clinical series of dual growing rods, reporting improvement in the mean curve magnitude from 82° to 38° after the first surgery with maintenance of the correction through final follow-up [12]. However, complications were frequent, with 13 occurring (56%) in 11 patients. Bess et al. reviewed 140 patients undergoing single- or dual-rod distraction and reported 58% of patients had at least 1 complication [13]. The complication rate increased by 24% for each additional procedure performed and decreased by 13% for each year of increased patient age at treatment initiation. TGRs are considered the gold standard to which other growth-friendly surgeries are compared. With all methods of distraction-based surgery, there has been a well-described decrease in the length achieved during distractions later in the process compared to the early post-insertion period.

Vertical expandable prosthetic titanium rib (VEPTR). The VEPTR system (DePuy Synthes Spine, West Chester, PA), a titanium alloy longitudinal rib distraction device, is approved for treatment of skeletally immature patients with primary thoracic insufficiency syndrome (TIS) or EOS that is at risk for secondary TIS [14]. In primary TIS, the goal of surgery is to maximize thoracic volume and symmetry of the deformed thorax by lengthening the constricted hemithorax through a transverse opening wedge thoracostomy of the concave side [15]. The thoracic correction and reconstruction is stabilized by the VEPTR construct, which is lengthened every 4 to 6 months. The VEPTR construct has also been used in other conditions in which secondary TIS may develop, but without thoracoplasty.

Campbell et al. studied 27 children with congenital scoliosis associated with fused ribs and found a mean