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The Classification for Early-onset Scoliosis (C-EOS) Correlates With the Speed of Vertical Expandable Prosthetic Titanium Rib (VEPTR) Proximal Anchor Failure

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Abstract

Background—The Classification for Early-onset Scoliosis (C-EOS) was developed by a consortium of early-onset scoliosis (EOS) surgeons. This study aims to examine if the C-EOS classification correlates with the speed (failure/unit time) of proximal anchor failure in EOS surgery patients.

Methods—A total of 106 EOS patients were retrospectively queried from an EOS database. All patients were treated with vertical expandable prosthetic titanium rib and experienced proximal anchor failure. Patients were classified by the C-EOS, which includes a term for etiology [C: Congenital (54.2%), M: Neuromuscular (32.3%), S: Syndromic (8.3%), I: Idiopathic (5.2%)], major curve angle [1: 20 degrees (0%), 2: 21 to 50 degrees (15.6%), 3: 51 to 90 degrees (66.7%),

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4: >90 degrees (17.7%)), and kyphosis [“–”: 20 (13.5%), “N”: 21 to 50 (42.7%), “+”: >50 (43.8%)]. Outcome was measured by time and number of lengthenings to failure.

Results—Analyzing C-EOS classes with >3 subjects, survival analysis demonstrates that the C-EOS discriminates low, medium, and high speed of failure. The low speed of failure group consisted of congenital/51-90/hypokyphosis (C3–) class. The medium-speed group consisted of congenital/51-90/normal and hyperkyphosis (C3N, C3+), and neuromuscular/51-90/hyperkyphosis (M3+) classes. The high-speed group consisted of neuromuscular/51-90/normal kyphosis (M3N), and neuromuscular/>90/normal and hyperkyphosis (M4N, M4+) classes. Significant differences were found in time ($P < 0.05$) and number of expansions ($P < 0.05$) before failure between congenital and neuromuscular classes. As isolated variables, neuromuscular etiology experienced a significantly faster time to failure compared with patients with idiopathic ($P < 0.001$) and congenital ($P = 0.026$) etiology. Patients with a major curve angle >90 degrees demonstrated significantly faster speed of failure compared with patients with major curve angle 21 to 50 degrees ($P = 0.011$).

Conclusions—The ability of the C-EOS to discriminate the speeds of failure of the various classification subgroups supports its validity and demonstrates its potential use in guiding decision making. Further experience with the C-EOS may allow more tailored treatment, and perhaps better outcomes of patients with EOS.

Level of Evidence—Level III.

Keywords

early-onset scoliosis; scoliosis; vertical expandable prosthetic titanium rib; VEPTR; growing rod; growth friendly surgery; Classification for Early-onset Scoliosis

Early-onset scoliosis (EOS) is a heterogeneous population including congenital, neuromuscular, syndromic, and idiopathic etiologies. These etiologies have variable curve severities and differing natural histories.^{1–4} Communication and research with respect to EOS patients is a challenge due to this heterogeneity.^{5,6} In fact, numerous studies regarding the treatment of EOS have used mixed populations of patients, complicating the interpretation of such studies.^{7–10} EOS providers lack a common language to simplify communication regarding these patients, and in research, investigators are unable to compare management practices and outcomes.¹¹

To address this issue, classification systems are essential tools that define pathology and provide the language to describe it.¹² The Classification for Early-onset Scoliosis (C-EOS) (Fig. 1) was developed following a 3-step model proposed by Audigé et al.¹³ C-EOS facilitates the subgrouping of patients who may share natural histories and may benefit from similar treatments.¹² Establishing the validity and utility of the C-EOS is crucial for the utilization of this instrument within research and clinical contexts. The purpose of this study is to establish the validity and utility of the C-EOS by evaluating its association with the speed of a common complication of EOS surgery: vertical expandable prosthetic titanium rib (VEPTR; Synthes Spine Co., West Chester, PA) proximal fixation failure.

VEPTR relies on proximal fixation to the rib with circumferential rib “cradles” in order to support the thoracic cage. These fixation points migrate or fail with high frequency depending on the stresses on the construct, bone quality, and duration of attachment.¹⁴ This study aims to compare speed of proximal fixation failure in VEPTR constructs between classes of the C-EOS. We hypothesize that neuromuscular etiology, higher major curve angles, and higher kyphosis angles will correlate with higher speeds of VEPTR anchor failure.

METHODS

Design

This is a retrospective, multicenter study with data collected through an EOS study group registry from 2005 to 2012. Eight academic institutions contributed data: [Boston Children’s Hospital, University of Utah, Shriner’s Hospital for Children Los Angeles & Philadelphia, Children’s Hospital of Philadelphia, University of Colorado, Le Boenheur Children’s Hospital, and Children’s Hospital of New York].

Participants

Patients with a diagnosis of EOS were prospectively enrolled into an EOS study group registry. This database was queried to retrospectively gather data pertaining to patients with the following characteristics: age less than 8 years of age, underwent VEPTR implant surgery, and suffered subsequent proximal fixation failure.

Outcomes

The outcome analyzed was VEPTR proximal fixation failure speed. Failure was defined as a radiographic evidence of proximal fixation failure whether or not the patient necessitated emergent revision surgery. The speed of a failure was calculated using the formula:

$$\text{Speed} = \frac{\text{Total failures}}{\text{Months to failure}}.$$

For example, the speed of failure for the C3– C-EOS group was calculated by dividing the total observed failures (5) into the total months of observation for all 5 failures (250). This yielded a speed of 1 failure/50 months.

Procedures

The EOS study group registry was queried with the inclusion criteria listed above. Once study subjects were identified, data managers at contributing sites ensured data integrity and provide missing data fields. Study subject data was sent to [Children’s Hospital of New York] in order to classify patients by the C-EOS scheme and for subsequent analysis.

C-EOS classification groups with $n > 3$ were subjected to survivorship analysis. Speed stratification was accomplished by looking at months to <50% survival. If <50% survival occurred within 1 year of VEPTR implant, this was defined as high speed. If <50% survival

occurred within 1 to 3 years of VEPTR implant, this was defined as medium speed. If <50% survival occurred beyond 3 years of VEPTR implant, this was defined as low speed.

Statistical Analysis

To analyze differences within each classification component, analysis of variance was used. Statistical significance was defined as $P < 0.05$. To analyze differences among classification groups, Kaplan-Meier survivorship analysis was used.

RESULTS

The registry query resulted in 106 subjects meeting inclusion with 96 having etiology, major curve angle, and kyphosis for complete classification. Of the 105 subjects, there were 51 (48.1%) males, 44 (41.5%) females, and 11 (10.4%) unknown. Mean age at index surgery was 4.7 ± 2.6 years with a mean follow-up of 6.2 ± 2.7 years. All etiology groups of the C-EOS, major curve angle classification groups 2, 3, and 4, and all kyphosis classification groups were represented by the study population (Table 1). Twenty-six classification groups were represented, 7 of which had >3 subjects (Table 2).

When subjected to Kaplan-Meier Survivorship analysis, the 7 classification groups with >3 subjects were stratified into low-speed, medium-speed, and high-speed groups. The low speed of failure group consisted of congenital/51-90 degrees/hypokyphosis (C3-) class. The medium-speed group consisted of congenital/51-90 degrees/normal and hyperkyphosis (C3N, C3+), and neuromuscular/51-90 degrees/hyperkyphosis (M3+) classes. The high-speed group consisted of neuromuscular/51-90 degrees/normal kyphosis (M3N) and neuromuscular/>90 degrees/normal and hyperkyphosis (M4N, M4+) classes (Fig. 2) and (Table 3). Time to failure was calculated as an average of months or years within classes with $n > 3$. Congenital curves were slower to failure than neuromuscular, and within the neuromuscular etiology, higher major curve angle classes were faster to failure than lower major curve angle classes (Table 4).

Analysis of variance for speed of proximal fixation failure for individual C-EOS variables resulted in significant findings in the etiology and major curve angle variables. Subjects with neuromuscular scoliosis exhibited significantly faster speed of proximal fixation failure than idiopathic ($P = 0.026$) and congenital scoliosis ($P < 0.001$) subjects (Fig. 3). Subjects with curves >90 degrees exhibited significantly faster speed of proximal fixation failure in comparison to curves of 25 to 50 degrees ($P = 0.011$) and 51 to 90 degrees ($P = 0.031$) (Fig. 4). More failures occurred within the higher kyphosis classes (N, or +), but there were no significant differences in the speed of failure among kyphosis groups.

DISCUSSION

Our study demonstrates that the C-EOS with combined elements of etiology, major curve angle, and kyphosis has the capability to stratify the speed of rapid VEPTR proximal anchor failure. Furthermore, the manner in which the classification groups progress toward higher speed of VEPTR anchor failure is in concert with the conceptual understanding of mechanical stress upon the surgical construct.^{15,16} When examining the results,

neuromuscular etiology, increased major curve angle, and normokypnosis or hyperkypnosis were correlated with rapid VEPTR anchor failure. In fact, the order of classifications progressed from congenital to neuromuscular, major curve angle 51 to 90 degrees to >90 degrees, hypokypnosis to hyperkypnosis, which is in line with expectation. In terms of etiology, neuromuscular scoliosis can confer high stress to VEPTR devices as these patients often have rigid curves with associated spasticity or paralysis, factors implicated in implant-related complications of scoliosis surgery.^{17,18} It also follows that increased coronal and sagittal curves would also confer high stress upon VEPTR devices, factors that are also implicated in complications of growing spine surgery.¹⁹ Therefore, the progression of speed by classification group found in this study supports the validity and utility of the C-EOS.

When analyzed by individual variables, neuromuscular curves exhibited a significantly faster speed of failure than idiopathic or congenital curves. Studies have reported mixed results when examining the correlation between complications and etiology. However, there is evidence to suggest that neuromuscular scoliosis confers a high rate of complications. In a study of the Scoliosis Research Society Morbidity and Mortality database of 19,360 subjects, 4657 were neuromuscular, which was found to have a significantly greater complication rate in pediatric scoliosis surgery.²⁰ A common complication of these patients was implant related.²⁰ Bess et al⁹ found that neuromuscular patients treated with growing rods experienced the highest number of complications. However, McElroy et al²¹ found that the complication rate in spinal muscular atrophy subjects, a neuromuscular etiology of scoliosis, was less than that of idiopathic patients, providing evidence contrary to the notion of high rates of complications in neuromuscular scoliosis.

This study's findings suggest that higher degree curvatures are associated with a higher speed of proximal fixation failure in VEPTR surgery. In terms of major curve angle and kyphosis, major curve angle >90 degrees and higher degrees of kyphosis correlated with speed of anchor failure. More severe preoperative major curve angles have been shown to increase the speed of general complications in scoliosis surgery.^{22,23} Schroerlucke et al²⁴ found that hyperkypnosis subjects undergoing growing rods surgery had significantly more complications than normal kyphosis subjects. These findings lend support to the validity of the individual variables of the C-EOS.

The study's strengths include its multicenter and multisurgeon experience of a known complication of VEPTR surgery. However, the retrospective nature of this study with its low study size prevents the inclusion of more C-EOS classes that have >3 subjects. Furthermore, the study size does not adequately power statistical analysis between specific C-EOS classes. Therefore, future steps for the C-EOS include validation studies that follow the outcomes of EOS patients over medium to long term with larger sample sizes.

The ability of the C-EOS to discriminate among various classification subgroups supports its validity and demonstrates its potential use in guiding decision making. Further experience with the C-EOS may allow more tailored treatment, better advice to patients, and improved outcomes for patients with EOS.

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FIGURE 1. The Classification for Early-onset Scoliosis (C-EOS). Etiology: (1) congenital/structural: curves developing due to a structural abnormality/asymmetry of the spine and/or thoracic cavity; includes hemivertebrae, fused ribs, postthoracotomy, or congenital diaphragmatic hernia; (2) neuromuscular: patients with neuromuscular disease (ie, spinal muscular atrophy, cerebral palsy, muscular dystrophies, etc.); (3) syndromic: syndromes with known or possible association with scoliosis (including spinal dysraphism). Idiopathic: no clear causal agent (can include children with a significant comorbidity that has no defined association with scoliosis). Major curve angle: measurement of major spinal curve in position of most gravity; maximum measurable kyphosis: between any 2 levels; annual progression ratio modifier (optional): progression per year; minimum = 6 months between observation:

$$[(\text{Cobb at } t_2) - (\text{Cobb at } t_1)] \times \frac{12\text{months}}{[t_2 - t_1]}.$$

FIGURE 2.

Time to vertical expandable prosthetic titanium rib proximal fixation failure.

FIGURE 3.

Mean time to failure by diagnosis. Neuromuscular versus idiopathic ($P = 0.026$).

Neuromuscular versus congenital ($P < 0.001$).

FIGURE 4.

Mean time to failure by preoperative major curve angle. >90 degrees versus 21 to 50 degrees ($P = 0.011$). >90 degrees versus 51 to 90 degrees ($P = 0.031$).

TABLE 1

Demographics of Participants

C-EOS Variables	Classification	N (%)
Etiology	Congenital	52 (54.2)
	Neuromuscular	31 (32.3)
	Syndromic	8 (8.3)
	Idiopathic	5 (5.2)
Cobb (deg.)	20	0 (0)
	21–50	15 (15.6)
	51–90	64 (66.7)
	> 90	17 (17.7)
Maximum kyphosis (deg.)	20	13 (13.5)
	21–50	41 (42.7)
	> 50	42 (43.8)

C-EOS indicates Classification for Early-onset Scoliosis.

TABLE 2

Frequencies of Classes

Classification	N (%)
C2-	2 (2.1)
C2N	3 (3.1)
C2+	3 (3.1)
C3- *	5 (5.2)
C3N *	18 (18.8)
C3+ *	14 (14.6)
C4-	1 (1.0)
C4N	2 (2.1)
C4+	3 (3.1)
M2N	1 (1.0)
M3-	3 (3.1)
M3N *	7 (7.3)
M3+ *	9 (9.4)
M4-	1 (1.0)
M4N *	4 (4.2)
M4+ *	5 (5.2)
S2N	2 (2.1)
S2+	1 (1.0)
S3-	1 (1.0)
S3N	1 (1.0)
S3+	2 (2.1)
S4+	1 (1.0)
I2N	2 (2.1)
I2+	1 (1.0)
I3N	2 (2.1)
I3+	2 (2.1)

*
> 3 patients present.

TABLE 3**C-EOS Classes by Speed Group**

Speed	Classification
Low speed	C3–
Medium speed	C3N
	C3+
	M3+
High speed	M3N
	M4N
	M4+

C-EOS indicates Classification for Early-onset Scoliosis.

TABLE 4Average Months and Years to Failure in Classes With $n > 3$

C-EOS	Average Months to Failure	Average Years to Failure
C3–	50	4.17
C3N	26.28	2.19
C3+	28.43	2.37
M3N	3.93	0.33
M3+	19.44	1.62
M4N	3.75	0.31
M4+	10.6	0.88

C-EOS indicates Classification for Early-onset Scoliosis.