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Superior Extension of Upper Instrumented Vertebrae in Distraction-based Surgery: A Surrogate for Clinically Significant Proximal Junctional Kyphosis

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Abstract

Background: Proximal junctional kyphosis (PJK) is a reported complication of distraction-based growth-friendly surgery for early-onset scoliosis (EOS). A potential consequence of PJK is revision surgery with superior extension of the upper instrument vertebrae (UIV). The purpose of this study was to determine the risk of radiographic and clinically significant PJK during growth-friendly surgery.
Methods: This is a retrospective review of children treated with distraction-based growth-friendly surgeries from two EOS registries with minimum two-year follow-up. PJK is defined as clinically significant in this study if surgery with superior extension of the UIV was performed.
Results: Of 419 total patients, there was a 20% risk of developing clinically significant PJK (24% rib vs. 15% spine-based anchors, $p = .03$). These patients had a mean preoperative age of 5.6 years (5.2-year rib vs. 6.0-year spine, $p < .001$), scoliosis of 73° (69° rib vs. 77° spine, $p < .001$), and kyphosis of 51° (47° rib vs. 56° spine, $p < .01$). Regression analysis demonstrated that these differences in age, scoliosis, and kyphosis between anchor type did not account for a significant proportion of the measured variance.
Conclusions: There was a 20% risk of developing clinically significant PJK, with a slightly higher risk for patients treated with rib-based proximal anchors (24%) than for those patients treated with spine-based proximal anchors (15%).
Level of Evidence: Level III.

Keywords: Scoliosis; Early-onset scoliosis; Proximal junctional kyphosis; PJK; VEPTR

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Introduction

An estimated six million individuals in the United States are affected by scoliosis [1] and 42 per 1,000 patients between the ages of 8 and 15 are affected by scoliosis in Quebec alone [2]. Early-onset scoliosis (EOS) is defined as scoliosis with onset less than the age of 10 years, regardless of etiology [3]. A common form of treatment for EOS is distraction-based growth-friendly surgery, which involves insertion of expandable rods, with proximal rib-based or proximal spine-based attachments. Minor procedures are then performed approximately every six months with these patients to lengthen the rods so as to parallel the normal spine growth of the child [4]. Ideally, once the child passes her or his peak growth velocity, a spinal fusion procedure is then performed to halt scoliosis advancement.

Proximal junctional kyphosis (PJK) is a nonphysiologic, sagittal plane angulation that occurs cephalad to an instrumented spine and is a reported complication of surgical treatment of spinal deformity [5,6]. Previous radiographic work has demonstrated that the risk of developing postoperative PJK during distraction-based growth-friendly surgery is 20% immediately after implantation and 28% at minimum two-year follow-up. It also demonstrated that older age, greater thoracic kyphosis, preoperative lumbar lordosis, and pelvic tilt are all patient factors that may increase the risk of developing postoperative PJK. In that study, the risk of developing postoperative PJK was not significantly different between rib-based and spine-based growing systems [7].

Recent studies have demonstrated that the current radiographic definitions of PJK that are used in the evaluation of EOS are varied and potentially unreliable. The risk of developing PJK within the same EOS patient population was found to vary between 6% and 42% depending upon which of three common definitions of PJK were used [8]. For each definition, either ‘‘good’’ or ‘‘excellent’’ intra- and intra-rater reliability was noted [8]. In contrast, a separate study found a variability of approximately 18° and ‘‘poor’’ to ‘‘moderate’’ intra-/inter-rater reliability [9].

Although PJK can be detected radiographically in patients with EOS, its clinical implications have not been studied for this population. An unwanted clinical effect of radiographic PJK may include implant failure, which requires superior extension of the upper instrumented vertebrae (UIV) during growth-friendly treatment or during graduate surgery for EOS. These unplanned surgeries and revision to correct the PJK can place additional emotional and physical stress on young patients and their families during a period already laden with anxiety. Children who undergo repetitive surgery for EOS have been demonstrated to develop neurobehavioral dysfunction, with the majority of the children within the clinically significant or at-risk range [10]. Quality of life of both the children and their families could thus be seriously affected as a result. It is believed that efforts should be made to identify clinically significant PJK and that efforts to minimize this complication should be studied.

Although not perfect, an accepted definition of clinically significant PJK has been proposed to be ‘‘the risk of patients treated with distraction-based surgery who required superior extension of their upper instrumented vertebrae’’ (Fig. 1). The primary purpose of this study was to determine the risk of clinically significant PJK during distraction-based growth-friendly surgery. The secondary goal was to compare the risk of developing clinical PJK

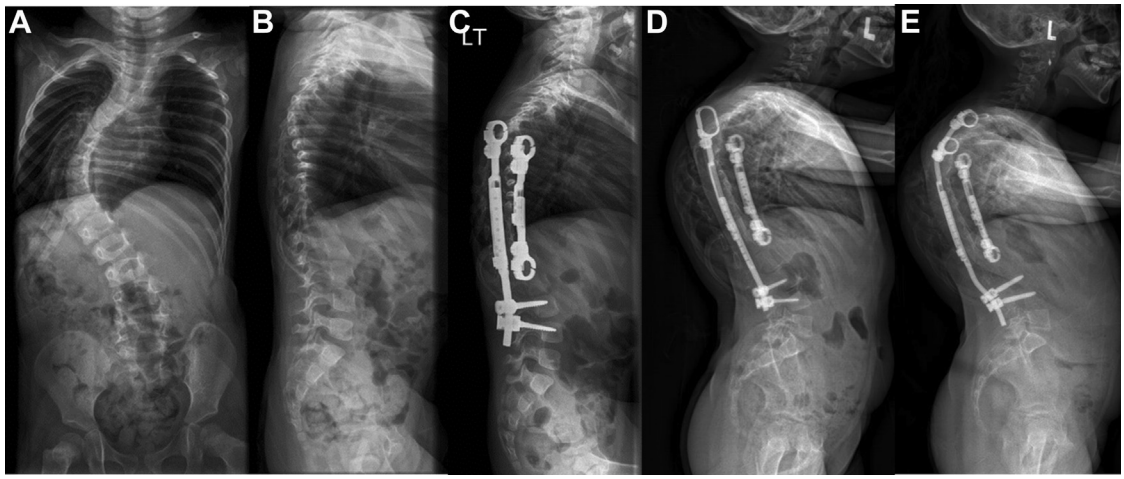

Fig. 1. Two-year-old patient with infantile idiopathic scoliosis treated with rib-based distraction surgery that developed clinically significant PJK. (A) Preimplantation supine posteroanterior scoliosis radiograph. (B) Preimplantation lateral scoliosis radiograph. (C) Initial postimplantation lateral scoliosis radiograph. (D) Five years postimplantation (eight lengthening surgeries and two exchange surgeries) with lateral radiograph demonstrating radiographic PJK. (E) Seven years postimplantation lateral radiograph demonstrating treatment of PJK with superior extension of the upper instrumented vertebrae. PJK, proximal junctional kyphosis.

between proximal rib-based and proximal spine-based distraction surgery.

Methods

This was a retrospective, comparative, clinical, and radiographic review of patients treated with distraction-based surgery. These patients were identified from two large EOS databases. Inclusion criteria included diagnosis of EOS, treatment with traditional surgically lengthened rib-based or spine-based distraction surgery with minimum two-year follow-up, and a minimum of three lengthening procedures. Patients with incomplete radiographic data, or those with proximal extension specifically due to the development of proximal scoliosis (proximal adding-on) were excluded.

The primary clinical outcome variable was the requirement for superior extension of the UIV during distraction phase or at graduation from distraction-based surgery.

Surgimap Spine V1.2.1.66 (Nemaris Inc., NY) software tool was used for the radiographic portion of the study. These measurements included scoliosis (thoracic and lumbar), cervical lordosis, thoracic kyphosis (high thoracic kyphosis was defined as $>50°$), lumbar lordosis, and proximal junctional angle (PJA). Two definitions of PJA were utilized: PJA-1 was measured as the angle between the caudal; end plate of the UIV to the cephalad end plate 2 vertebrae above the UIV (it was chosen as it is a very commonly used definition [5]); PJA-2 is measured as the angle from two levels below the UIV to two levels above the UIV (it has been found to be the most reliable measure for EOS patients [8]) (Fig. 2).

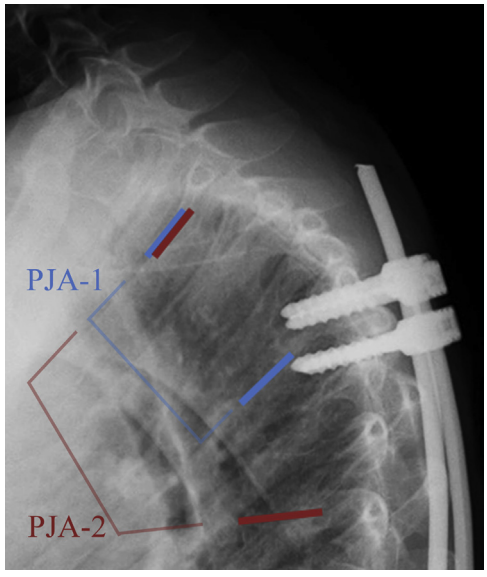

Fig. 2. Two definitions of proximal junctional angle (PJA). PJA-1 is measured as the angle between the caudal end plate of the UIV to the cephalad end plate 2 vertebrae above the UIV; PJA-2 is measured as the angle from 2 levels below the UIV to 2 levels above the UIV. UIV, upper instrumented vertebrae.

Continuous data were analyzed using paired *t* tests, and binary logistic and linear regression analyses were performed using Statistical Package for the Social Sciences, version 19 (IBM Corp, Armonk, NY). OpenEpi (OpenEpi 3.03; Atlanta, GA) with chi-square measure of association was used to determine risk between different groups of subjects. A p value $<.05$ was considered statistically significant.

Results

A total of 496 patients were identified from the two EOS registries. Seventy-seven patients with superior extension of implants specifically due to the development of proximal scoliosis, those with incomplete radiographic series, and those with a combination of rib and spine proximal anchors were excluded from analysis, resulting in a total of 419 patients included in this study. These patients had the following etiologies: 37 syndromic, 114 neuromuscular, 108 congenital, 73 idiopathic, and 87 with unknown etiologies.

Of these patients, 219 patients had been treated with rib-based proximal anchors [218 of the rib-based patients underwent VEPTR (DePuy-Synthes, Raynham, MA)–based treatment] and 200 patients had been treated with spine-based proximal anchors. These 419 patients had a mean preoperative age of 5.6 years (5.2 years rib-based vs. 6.0 years spine-based, $p<.001$), preoperative major scoliosis of 73° (69° rib-based vs. 77° spine-based, $p<.001$), and preoperative kyphosis of 51° (47° rib vs. 56° spine, $p<.01$).

With a minimum of two-year follow-up and a minimum of three lengthening procedures, 52 of 219 rib-based and 31 of 200 spine-based patients required proximal extension of the UIV for an overall risk of clinically significant PJK of 20% (24% rib-based vs. 15% spine-based, $p=.03$) (Table). Regression analysis demonstrated that these differences in age, scoliosis, and kyphosis between anchor type did not account for a significant proportion of the measured variance ($p=.06$). There were not any statistically significant differences in the risk of developing clinically significant PJK between the different etiologies of EOS (Fig. 3).

The mean final PJA-1 was 9.0° for the proximally extended group and was 7.1° for the comparison group ($p=.04$). The mean final PJA-2 was 20.0° for the proximally extended group and was 16.3° for the comparison group

Table
Comparison between rib-based and spine-based distraction.

	Rib-based	Spine-based	p	Total
Number of subjects	219	200		419
Preoperative age (years)	5.2	6.0	<.001	5.6
Preoperative scoliosis (°)	69	77	<.001	73
Preoperative kyphosis (°)	47	56	<.01	51
Clinical risk of PJK (%)	24	15	.03	20

PJK, proximal junctional kyphosis.

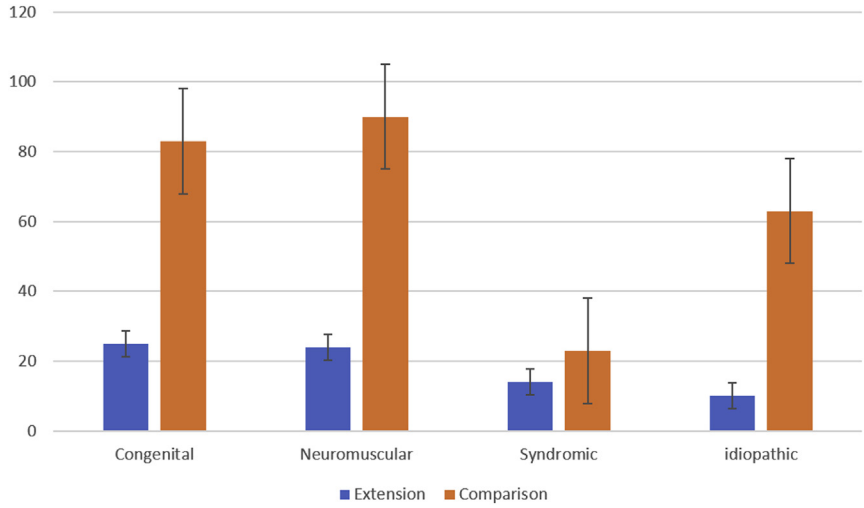

Fig. 3. Mean and standard error for percentage of patients within each etiologic category with clinically significant PJK (extended) versus those without clinically significant PJK. There were not any statistically significant differences between etiologies. PJK, proximal junctional kyphosis.

($p = .01$). Using a threshold of $>20°$ to represent high PJA, the risk of proximal extension was 42% for high PJA-1 and was 18% for low PJA-1 ($p = .02$). The risk of proximal extension was 25% for high PJA-2 and was 17% for low PJA-2 ($p = .02$). The risk ratio for requiring proximal extension of UIV with high PJA-1 was 2.3 ($p < .001$) and with high PJA-2 was 1.47 ($p = .08$).

Discussion

The primary purpose of this study was to determine the risk of clinically significant PJK during posterior distraction-based growth-friendly surgery. The secondary goal was to compare the risk of developing clinical PJK between proximal rib-based and proximal spine-based distraction surgery. We determined that there was a 20% risk of developing clinically significant PJK, with a slightly higher risk for patients treated with rib-based proximal anchors (24%) than for those patients treated with spine-based proximal anchors (15%).

Limitations of this study are that it analyzes a retrospective series of subjects with EOS that were identified from multicenter databases. Although a prospective study may have more easily identified the reasons for revision surgery, we do not think that the retrospective nature of our study had a significant impact on this variable. If the reason for revision surgery was not clearly identified directly from the databases, audits to the individual sites were performed in order to determine this information. In addition, the radiographic measurements for PJA would not be expected to be influenced by the retrospective nature of this study. As EOS is a rare condition with a heterogeneous etiology, its study often necessitates use of multicenter databases. This is an inherent weakness in the majority of studies on EOS.

One major strength of this study is that this is the first study in the EOS population examining the risk of developing PJK through a clinical surrogate. Despite the published results for the development of radiographic PJK for this population, there is still enough concern about the radiographic variability of this measurement that some feel it may not be a clinically useful tool. Barrett et al. published an interobserver variability of PJK greater than 20° [11]. As PJK is commonly defined as $>10°$ of kyphosis above the UIV, that group felt the measurement of PJK in patients with distraction-based growing rods on lateral radiographs has too much variability to be useful. Conversely, the study by Al Khudairy et al., evaluating the reliability of the measurements of radiographic PJK for EOS, determined that the reliability of the measurements was dependent on the definition of PJK used. From his reliability study, the definition for PJA-1 from our current study was found to have fair interobserver agreement [intraclass correlation coefficient (ICC) 0.48] and good intraobserver agreement (ICC 0.61) [8], whereas PJA-2 from our current study was found to have good interobserver agreement (ICC 0.71) and excellent intraobserver agreement (ICC 0.82). Although the radiographic variability of these measurements may be better than initially published, both groups of authors are in agreement that a clinical surrogate for significant PJK should be established.

Our definition for clinically significant PJK resulted in a 20% risk for patients treated with posterior distraction-based growth-friendly surgery. Results in the literature for radiographic PJK for EOS patients range from 6% to 54% depending upon the definition used [7,9,10]. Using the studies with the most reliable radiographic definitions, our 20% risk of developing clinically significant PJK is certainly less than the published risks of developing radiographic PJK. This supports our hypothesis that all

patients with radiographic PJK do not have clinically significant PJK.

Recent literature has observed a 28% risk of developing radiographic PJK and also compared patients treated with rib-based versus spine-based proximal anchors. A 2017 study found a significant difference between the treatment groups, with a 25% radiographic risk for patients treated with rib-based anchors versus a 31% radiographic risk for patients treated with spine-based anchors [7]. Our current study is only the second study to compare PJK between these two different treatment groups and found a slightly higher risk for patients treated with rib-based proximal anchors (24%) versus those patients treated with spine-based proximal anchors (15%). This is likely related to the location of UIV between treatment groups as the spine-based group tended to have a more cephalad UIV as compared with the rib-based group (T2 vs. Rib 3). As spine-based patients tended to already have a UIV in the upper thoracic spine, it is plausible to think that surgeons would be less likely to revise these patients with upper extension of the UIV into the cervical spine.

Another strength of our current study is the large number of patients. Our study of 419 patients is much larger than the 68 patients, 32 patients, and 40 patients evaluated in the previous studies on this subject [7,9,10]. We strive for large population studies as we continue to study EOS.

Overall, we were able to identify a relatively large number of patients in order to determine the risk for a clinically significant PJK. In addition, we were able to correlate radiographic definitions of PJA with our definition of clinically significant PJK. Both definitions of PJA were significantly higher for patients who had clinically significant PJK, and high PJA-1 had a statistically significant risk ratio of 2.3 for clinically significant PJK. By establishing that PJA can be predictive of clinically significant PJK, we have demonstrated its usefulness despite its potential high variability. Given the predictive nature of these radiographic measurements of PJA, we should continue to study these measures in an effort to try to improve upon the variability. Potentially the use of other diagnostic imaging modalities such as biplanar radiographs (EOS Imaging, Paris, France) or radiostereophotogrammetric analysis (RSA) (RSAcore, Leiden, the Netherlands) may be able to improve upon the variability of these radiographic measurements.

For patients treated with posterior distraction-based growth-friendly surgery, we identified a 20% risk of developing clinically significant PJK, with a slightly higher risk for patients treated with rib-based proximal anchors (24%) than for those patients treated with spine-based proximal anchors (15%).

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