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Paper #17

Superior Extension of Upper Instrumented Level in Distraction-Based Surgery: A Surrogate for Clinically Significant PJK

Ron El-Hawary, MD, MSc, FRCSC, John Heflin, MD, Nadim Joukhadar, BSc, Mohamad Yasin, MD, David Skaggs, MD, Children's Spine Study Group

Introduction: Proximal junctional kyphosis (PJK) is a reported complication of distraction-based treatment for early-onset scoliosis. Proximal junctional kyphosis can be detected radiographically; however, the clinical implications of PJK have not previously been studied for this population. A potential clinically significant consequence of PJK is revision surgery with superior extension of the upper instrumented level (UIL). The purpose of this study was to determine the rate of clinically significant PJK during rib-based distraction surgery. A secondary goal was to define the proximal junctional angle (PJA) at the time of revision surgery, with the hypothesis that PJA would be increased in this group of patients.

Methods: This was a retrospective review of a multicenter registry for children with early-onset scoliosis. All children treated with rib-based distraction surgery, with minimum 2-year follow-up, were evaluated to identify the rate of clinically significant PJK (ie, children who required a revision surgery that involved superior extension of the UIL). Two definitions of PJA were used: PJA-A was the angle between the caudal end plate of the upper instrumented vertebrae (UIV) to the cephalad end plate 2 vertebrae above UIV; and PJA-B was 2 levels below UIV to 2 levels above UIV.

Results: A total of 397 children were identified. At the time of implantation, these children had a mean age of 5.5 years, mean scoliosis of 69.9°, and mean kyphosis of 49.8°. Forty of these children required a revision surgery that involved superior extension of the UIL (10.1% rate of clinically significant PJK). Despite being younger (4.9 vs. 5.5 years; $p < .05$), the revision group had pre-implantation characteristics similar to the entire study population, with mean scoliosis of 70.0° and mean kyphosis of 50.0°. Average time to revision was 2.3 years with mean scoliosis of 66.7° and mean kyphosis of 54.7° at the time of revision. Proximal junctional angle A was 5.6° before surgery versus 11.8° at the time of revision ($p < .05$). Proximal junctional angle B was 13.1° before surgery versus 21.4° at the time of revision ($p = .07$).

Conclusions: A 10% rate of clinically significant PJK was found within this group of children who were treated with rib-based distraction surgery. At the time of revision surgery, PJA-A had increased significantly from preoperative values.

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Paper #18

Proximal Junctional Kyphosis in Surgically Treated Young Children With Scoliosis: Incidence, Risk Factors, and Management

Bangping Qian, MD, Xu Sun, MD, Yong Qiu, MD

Background: Proximal junctional kyphosis (PJK) is a well-recognized postoperative complication in adults and adolescents spinal deformity after surgery; however, no reports specifically addressed the incidence, risk factors, and behavior of PJK in young children after spinal correction surgery.

Methods: The authors performed a retrospective review of 61 consecutive young children with congenital scoliosis undergoing posterior instrumented spinal fusion (4 or more levels) from 2009 to 2011. Subjects were younger than age 10 years at surgery and the minimum follow-up was 2 years. Proximal junctional kyphosis was defined by a proximal junctional angle greater than 10° and at least 10° greater than the corresponding preoperative measurement. Radiographic measurements were performed preoperatively, immediate postoperatively, and at the final follow-up, including proximal junctional angle, thoracic kyphosis, lumbar lordosis, kyphosis angles at T1–UIV and UIV–T12, as well as the matching of rod contour. Matched rod contour referred to the difference between the proximal instrumented region angle and the proximal rod curvature less than 5°. Patients with and without PJK were compared.

Results: Average age at surgery was 5.4 years (range, 2–10 years) and average follow-up was 2.7 years (range, 2–4 years). Proximal junctional kyphosis developed in 11 of 61 patients (incidence, 18%). The PJK group had larger preoperative thoracic kyphosis (48.7° vs. 36.3°), longer fusion levels (9.2 vs. 6.8 levels) and a larger amount of kyphosis correction (20.8° vs. 11.7°). In addition, proximal instrumentation failure and mismatched rod contour were identified as risk factors significantly associated with the occurrence of PJK. In the PJK group, the average proximal junctional angle increased 14.4° after 3 to 6 months postoperatively and then increased 2.5° until the final follow-up. Six patients had proximal instrumentation failure but none reported pain or problems with appearance. They received brace treatment and the PJK resolved.

Conclusions: This study demonstrated that 18% of young children were observed with PJK after correction surgery for congenital scoliosis. Proximal junctional kyphosis mainly occurred within 6 months postoperatively; risk factors included preoperative hyperkyphosis and over-correction of kyphosis (greater than 40°), more than 5 fusion levels, proximal instrumentation failure, and mismatched rod contour. Brace treatment served as a salvage option for PJK in young children.

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Paper #19

The Etiology of Thoracic Insufficiency Syndrome in Neuromuscular Scoliosis Based on Quantitative Dynamic Lung MRI (QdMRI)

Robert M. Campbell, MD, Jay Udupa, PhD, Jack Flynn, MD, Hank Mayer, MD, Michael Nance, MD, Howard Panitch, MD, Wei-Hsun Wang, MD, Yubing Tong, PhD, Ivy Wu, PhD, Kieth Baldwin, MD, Joseph McDonough, MSBME, Andrew Mong, MD

Introduction: Neuromuscular (NM) scoliosis is associated with poor pulmonary function that is thought to result from muscle weakness. Thoracic insufficiency syndrome (TIS) caused by complex spine/chest deformity also contributes. It is addressable by surgery but it is difficult to assess with current technology. Standard dynamic lung magnetic resonance imaging (dMRI) can show only qualitatively TIS, but the authors have developed a new quantitative dMRI approach termed “QdMRI.” In this pilot study, the authors analyzed the dMRIs of 5 neuromuscular scoliosis patients. Based on these results, the researchers propose new physiologic mechanisms for TIS in NM scoliosis that appear to improve with treatment.

Methods: The authors retrospectively examined the records, X-rays, and preoperative dMRI scans of 5 NM scoliosis patients, as well as 3 postoperative scans. All patients were treated with bilateral rib to pelvis hybrid

Vertical Expandable Prosthetic Titanium Ribs. The QdMRI analysis was performed by constructing a consistent 4-dimensional image from free-breathing dMRI slice acquisitions, segmenting key anatomic structures in the 4-dimensional volume, and deriving quantitative dynamic parameters from the defined structures.

Results: Average age of patients was 7.9 years, with an average follow-up of 2.13 years. Scoliosis was 75° preoperatively and 45° at follow-up. SAL was 0.84 preoperatively and 0.96 at follow-up. Pelvic obliquity was 19.6° preoperatively and 13.6° at follow-up. One patient had substantial soft tissue remodeling of the kidney with increased excursion on dMRI. On QdMRI, preoperative concave change in rib cage volume (mL) was 46.5, change in diaphragm volume was 25.7, convex change in rib cage volume was 40.5, and change in diaphragm volume was 27.4. Kidney excursion (mm) concave was 2.58 and convex was 3.02. Two patients had concave change in diaphragm volume less than convex, 1 had concave greater than convex, and 2 had equal volumes. For 3 patients with postoperative scans, concave change in rib cage volume increased 57% (convex, 72%) and change in diaphragm volume increased 128% (convex, 109%). Concave kidney excursion increased 6.3 mm (320%), and convex 5.2 mm (192%). Complications included 2 patients with migration of devices proximally.

Conclusions: Quantitative dMRI can measure separate contributions of diaphragm and rib cage to lung expansion during respiration for each hemithorax, enabling accurate assessment of thoracic function for the first time. Vertical Expandable Prosthetic Titanium Rib treatment appears to increase rib cage expansion more on the convex side of the thorax than the concave, and conversely increases diaphragm excursion on the concave side. With treatment, renal excursion increases bilaterally, concave more than convex, probably reducing a blockade effect on the diaphragm. Quantitative dMRI has the potential to increase understanding of the anatomic mechanisms of spine/chest wall deformity that cause restrictive lung disease in NM scoliosis, and may enable the development of new treatments for it.

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Paper #20

Diminished PFTs in Growing Rod Graduates Despite Improved Thoracic Height and Curve Correction

Charles E. Johnston, MD, Dong-phuong Tran, MS, Anna McClung, RN

Introduction: Long-term outcomes of growing rod instrumentation (GRI) are becoming more available as adolescents achieving sufficient spine length or requiring final fusion complete the lengthening segment of the treatment. The authors report X-ray and pulmonary outcomes in 6 patients completing GRI treatment at a single institution.

Methods: Six patients with severe early-onset scoliosis (mean curve, 98°) completed surgical treatment as part of a program spanning 10 years from initial presentation. Outcomes measured included X-ray parameters, PFTs, and surgical data.

Results: Dxs included 2 IIS, 2 idiopathic-like, 1 congenital, and 1 ambulatory N-M. Mean preoperative age was 74 months (range, 58–89 months), with preliminary nonsurgical treatment (halo traction or cast/brace) delaying GRI 38 months (range, 21–56 months) in 4 cases. Most recent surgery at age 129 months (range, 121–157 months) included definitive fusion in 4 patients, with 1- to 2.4-years' follow-up after fusion, and lengthening only in 2, with 3- to 4-year further observation. There were 8.3 mean total procedures per patient, including initial implantation, 1.2 unplanned revision/I&D, and 6.2 lengthenings (range, 3–9 lengthenings). At last follow-up, mean age was 156 months (range, 133–180 months). T1–12 height increased from 13.9 cm (range, 9.9–17.7 cm) preoperatively to 22.8 cm (range, 18.6–29.5 cm) at last surgery, and to 23.9 cm (range, 20.3–29.6 cm) at last follow-up. Curve magnitude decreased from 98° (range, 75° to 123°) to 48° at last surgery and to 42° (range, 26° to 53°) at last follow-up. PFTs were first performed at a mean of 6+9 years (4+10-8+7), and by last follow-up, absolute forced expiratory

volume in 1 second had doubled from 0.71 L (range, 0.48–1.2 L) to 1.45 L (range, 0.64–2.4 L). However, percent predicted forced expiratory volume in 1 second decreased from 61% to 46.5%. Absolute forced vital capacity also increased from 0.75 L (range, 0.48–1.2) to 1.73 L (range, 1.1–3.0), but percent predicted forced vital capacity decreased from 62% to 49%. The only complications were 7 rod/anchor problems in 4 patients.

Conclusions: Despite what appears to be satisfactory thoracic length gain and curve correction during 7 years of surgical management with acceptable complication rates, pulmonary outcomes were diminished by percent predicted outcomes criteria. The significance of this finding is uncertain without other corroborating data, but room for improved techniques clearly exists.

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Paper #21

Serial Measures of Lung and Inspiratory Muscle Function in Children With Early-Onset Scoliosis

Gregory J. Redding, MD, Megan Roth, PhD, Walter Krengel, MD, Klane White, MD, Viviana Bompadre, PhD, John Waldhausen, MD, Ajeya Joshi, MD

Previous reports of serial lung function in children with early-onset scoliosis (EOS) used passive maneuvers to test lung function in the operating room that did not require use of respiratory muscles. The authors previously reported that respiratory muscle function in these children is impaired and that passive lung functions may underestimate loss of lung function over time. They hypothesized that active lung function measures (forced vital capacity [FVC]) while awake would demonstrate greater decline in FVC over time than previously reported. They also serially measured maximum inspiratory pressures (MIP) in children with EOS to determine whether respiratory muscle function was impaired chronically. In Group 1, the researchers measured FVC in 38 patients from Seattle and San Antonio over an average of 6 years (27–98 months) with EOS with and without fused ribs without known neuromuscular disease. In Group 2, they measured both FVC and MIP in 15 children aged over 1 year. The age of Group 1 children at first FVC measurement was 7.9 ± 1.6 years (range, 5.23–14.5.6 years). Thirteen had PFTs before initial implantation; 35 of 38 had multiple expansions ($x = 7.8$; range, 1–16) and rod revisions ($x = 1.6$; range, 0–7) during the follow-up period. Four had spine fusion during follow-up. Initial FVC values were 48% ± 18% in Seattle and 58.9% ± 24% in San Antonio. Final FVC values were 37.5% ± 16% and 43.6% predicted, respectively, using arm span, and a similar and significant loss of 16% ± 23% over 6 years ($p < .05$ paired t test). There was no difference in time interval of follow-up or initial age at first PFT between the 2 centers. In Group 2, 12 children with EOS, aged 8.9 ± 2.6 years at first test, who had serial measures with both FVC and MIP over 18 months (range, 7–41 months) showed persistently reduced but unchanged FVC and MIP (47.8% ± 14.6% vs. 47.0% ± 22.3%). The authors conclude that declines in lung function in older awake children with EOS over a mean of 6 years are twice the declines previously reported over shorter time intervals with passive assessment techniques, and that respiratory muscle weakness is sustained in children with EOS.

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Paper #22

Spine Growth Modulation Using Titanium Clip/Screw Device: Vertebral and Disc Height Changes at 1 Year

Donita I. Bylski-Austrow, PhD, Nana A. Entsuh, David L. Glos, Joseph E. Reynolds, Eric J. Wall, MD

Introduction: The purpose of this study was to determine changes to coronal plane symmetry in vertebral and disc heights from before surgery to 12 months afterward in a prospective clinical safety trial of a titanium clip/screw device in children with late juvenile and early adolescent