Proximal Junctional Kyphosis in Early Onset Scoliosis

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Definition of Proximal Junctional Kyphosis (Adults / Adolescents)

Proximal junctional kyphosis (PJK) is a non-physiologic, sagittal plane angulation that occurs cephalad to an instrumented spine.

In adults and adolescents, proximal junctional failures above deformity constructs have been well established with different risk factors identified; however, the incidence, nature and clinical significance of this kyphosis remains unclear. Several reports in the literature have shed light on PJK in adults and adolescents with variable definition, incidence, prevalence, acuteness, risk factors, clinical significance and the need for revision surgery. Moreover, it followed diverse primary pathologies treated with different surgical techniques using variable types of fusion instrumentation.

The most consistent proximal junctional angle measurement in the literature as outlined by Glattes et al. has been between the caudal endplate of the upper instrumented vertebra (UIV) and the cephalad endplate of 2 levels above the UIV, with abnormal PJK defined as having proximal junctional cobb angle $>10^{\circ}$ and at least 10$^{\circ}$ greater than the pre-operative measurement. Despite this definition, there is difficulty in interpreting radiographic studies as the position of the upper extremity and head on lateral radiographs has an effect on sagittal plane alignment and subsequent radiographic measurements. Carmen et al. considered that a change in a kyphosis measurement should be $>10^{\circ}$ to be 95% confident that the change is true.

Kim et al. conducted a systematic review on PJK after spinal deformity fusion surgery in which they were able to include 7 studies according to their selection criteria, all sharing the aforementioned definition. He reported a PJK incidence of 17%-39%, mostly noted by 2 years postoperative with a moderate level of evidence. Risk factors included increased age, fusion to the sacrum, combined anterior and posterior spinal fusion, thoracoplasty, UIV at T1–T3, and non-anatomic restoration of thoracic kyphosis. The type of implants used at the proximal level (hooks, wires, or pedicle screws ) did not have a consistent statistically significant association with PJK across studies. He also concluded that PJK does not seem to have a detrimental effect on health related quality of life (HRQOL) outcomes, at least in milder/non-revision forms. Glattes et al. and Kim et al. also showed that there were no significant differences in Scoliosis Research Society Patient Questionnaire-24 outcome-scores between the PJK and non-PJK group in their series.
**Definition of Proximal Junctional Kyphosis (Early Onset Scoliosis)**

Currently, two common methods used to treat early onset scoliosis include rib-based and spine-based posterior distraction systems. These methods require repeated lengthening procedures that are believed to be kyphogenic and might predispose to PJK.

PJK in patients treated with growth friendly techniques has only been recently described, and thus there is no consensus on the definition, angles and measurement used. PJK Rates from 7% to 56% were reported in three separate studies presented at ICEOS 2011. As those studies were performed on different samples of young patients with scoliosis and used three different definitions of PJK, it is unclear as to whether the different rates were related to the different patient groups examined in those studies or to the choice of PJK definition that was used. These definitions were:

1. Proximal Junctional angle (PJA) $\geq 10^\circ$ and at least $10^\circ$ more than pre-implantation, with PJA defined as the angle between the caudal endplate of the upper instrumented level to the cephalad endplate 2 vertebrae above the upper instrumented level.

2. Proximal Junctional angle (PJA) $>10^\circ$ and at least $10^\circ$ more than pre-implantation, with PJA defined as the angle between the caudal endplate of 2 vertebrae below the upper instrumented level to the cephalad endplate 2 vertebrae the above upper instrumented level.

3. Proximal Junctional angle (PJA) $>20^\circ$, with PJA defined as the angle between the caudal endplate of the upper instrumented level to the cephalad endplate one vertebrae above the upper instrumented level.

In order to determine if the different rates of PJK were related to the patient populations that were studied or to the definitions that were employed, we conducted a review of a group of 36 children with EOS from the Children's Spine Study Group who were treated with distraction based systems between 2004 and 2008. Our main goal was to determine if the different PJK definitions will affect the rates of PJK in the same group of children, and secondarily to evaluate the reliability of these three different definitions of radiographic PJK. Five observers independently assessed children radiographs for PJA and PJK using all three definitions at two separate occasions (at least two weeks apart). Rates of PJK varied between 6% and 42% depending on the definition used. Highest rates were recorded with Definition 2 of around 39%, modest using Definition 1 $\approx 21\%$ and the lowest rates were with Definition 3 of about 7%. The variation we found in PJK rates with different definitions used was similar to what was observed in the 2011 studies. The segment length (number of vertebral levels) plus the angular threshold ($10^\circ$ vs. $20^\circ$) used to define PJK were reflected in the different rates reported. It was
found that by including more levels with a lower angular threshold (10°), higher PJK rates were observed (Definition 2).

Despite the difficulties in analyzing conventional radiographs taken at different institutions and the presence of anatomical / positional variability, we observed better inter and intra-observer agreement with Definition 2 than with the other two definitions. The use of a unified definition of PJK should improve the consistency of reporting this complication in future studies.

Notwithstanding these limitations, risk factors for PJK in early onset scoliosis have been identified. Li et al. studied 68 patients treated with VEPTR and determined that PJK developed in 6% of patients (generally developed within the first year of treatment). Risk factors included pre-operative thoracic hyper-kyphosis and the presence of weak paraspinal muscular support in neuromuscular patients.

Reinker et al. found that upper cradle placement below the third rib, distal anchor placement at too proximal a level, and recurrent erosion and loss of fixation on the proximal ribs contributed to progression of thoracic kyphosis. Thus, they recommended placing the upper cradle at the second rib, distal extension of the construct to the pelvis, and using bilateral implants. In contrast, Li et al. demonstrated that bilateral rib-to-pelvis constructs do not necessarily prevent progression of thoracic hyper-kyphosis. They identified indications for revision surgery to be a deformity sufficiently severe to endanger the neural elements, or the concern that a further increase in the deformity would necessitate a far more difficult and dangerous revision technique. In their small case series, patients who developed PJK were revised to growing rods.

In the Children's Spine Study Group review of 40 children treated with either spine-based or rib-based surgery, a PJK rate of 27.5% was identified. The type of growth friendly implant did not affect the rate of PJK; however, two risk factors for the development of PJK were observed: Pre-operative hyper-kyphosis (45° vs. 29°) and a higher age at implantation (7.1 years vs. 5.0 years). In this group of children, their hyper-kyphosis was corrected during implantation surgery; however, they experienced an increase in cervical lordosis in the initial post-operative period. With minimum two years follow up, their cervical lordosis continued to increase as did their proximal junctional angle.
**Future Directions**

Currently PJK is a radiographic parameter that has been demonstrated to occur after some growth friendly surgeries. Different definitions have been used and the reliability of these definitions has been variable. Ultimately, efforts should be made to try to correlate a unified definition of PJK with a clinically significant event (i.e. proximal implant failure and/or superior extension of the upper instrumented level during revision or graduate surgery). If these measures are found to be predictive of a clinically significant event, then further efforts should be made to improve the reliability of these measures.

**References**


