Instrumentation Strategies for Early Onset Scoliosis

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Abstract:
Nonoperative and operative management for early onset scoliosis has changed significantly since the Harrington era 50 plus years ago. Surgeons learned quickly that a spine fusion in the growing child can result in a short thorax and the development of thoracic insufficiency syndrome (TIS). Techniques were developed and refined over the subsequent decades to allow for spinal growth, control spine and chest wall deformity, and limit pulmonary demise. This "growth-friendly" concept is the modern-day approach to management of early onset scoliosis (EOS). In this article, we review the history of growth-friendly instrumentation following the Harrington era and present the authors preferred techniques for both growth guidance and posterior distraction-based management of EOS.

Key Concepts:
- Multiple techniques and various type of instrumentation exist to operatively manage EOS in 2021.
- The evolution and innovation of instrumentation for EOS resulted in the development of remote lengthening technology.
- Heterogeneity in the EOS population limits the ability to identify a single best practice or approach.
- Classic (infection, rod fracture, anchor failure, auto-fusion) and new (failure to lengthen) complications remain unsolved in the EOS population.

Introduction

In the latter part of the 20th century, it became evident that spinal fusions for early onset scoliosis (EOS) were detrimental to the growth of the thorax, often leading to significant pulmonary compromise. In addition, continued anterior growth after posterior spinal fusion often led to worsening spinal and thoracic deformity due to the crankshaft phenomenon. As a result, it was clear that "growth-friendly" techniques were needed for the optimal treatment of children with progressive EOS. However, the first generation of growth-friendly instrumentation for EOS, which included modified Harrington distraction rods inserted without fusion and "Luque trolleys," was marked by sparse long-term outcomes and limited overall success.[1]

Moe[2] reported on 20 EOS patients (average age 8.9 years) treated with a single modified Harrington rod placed subcutaneously on the concavity of the curve. Only the lamina of the upper and lower instrumented vertebra were subperiosteally exposed in order to prevent fusion in the intervening segments. All patients were treated with Milwaukee brace after growing rod insertion. Average gain in T1-S1 height was 2.9 cm, or 1.1 cm/year. At the time of publication, nine of these patients had been converted to final fusion at an average

age of 12.5 years. Ten patients experienced 22 complications, including six hook displacements and five rod breakages.

Luque[3] reported on 47 consecutive patients younger than eight with neuromuscular scoliosis who were treated with segmental fixation (hooks and wires) and a single rod placed either on the convex or concave side of the curve. No attempt at fusion was made. He reported average growth of 4.6 cm over the instrumented segments during an average of 4.7 years of follow-up. However, growth of the entire T1-S1 segment, which was 8.3 cm, was less than that predicted during the follow-up period, and subsequent authors[4] reporting on results of the Luque trolley technique found limited growth and extensive auto-fusion in the segments instrumented when these patients were converted to definitive fusion.

Given the suboptimal results associated with early fusion in early onset scoliosis and the first generation "growth-friendly" solutions, refinement of surgical techniques was an important development in the evolution of the surgical treatment of EOS beginning in the late 1980s and early 1990s.

Growing Rods

The next generation of innovation in pediatric spine deformity arose when surgeons moved away from the Harrington rod and Luque instrumentation and transitioned to a single submuscular rod with segmental instrumentation.[5] In an attempt to correct the deformity and maintain growth by minimizing auto-fusion, Blakemore et al. described placing this rod in a submuscular fashion without subperiosteal dissection with or without an apical fusion or convex hemiepiphysiodesis. In the first report on this technique, 29 patients received a single submuscular growing rod; 11 children simultaneously underwent apical intervention, and every patient was immobilized in an orthosis postoperatively for rod protection. Blakemore reported nine complications in seven patients (24%), comparable to the Harrington rod series complication profile reported by Klemme et al. with 33 complications in 25 children (37%).[6] The foundations for the anchors in the new growing rod consisted of the "claw" construct. Proximally, this meant a transverse process hook on the cephalad vertebrae and a sublaminar or pedicle hook 1-2 levels distally. The distal anchors consisted of an over-the-top laminar hook at the cephalad level and a sublaminar hook 1-2 levels distally. These rods were left long for periodic lengthening, which spinal growth suggests should occur every 5-9 months.[7] Farooq et al. published another single-center experience with a single submuscular growing rod with findings similar to the study by Blakemore et. al., indicating that the technique has acceptable levels of complications and is successful at managing early onset scoliosis.[8]

The combined experiences of Drs. Thompson and Akbarnia yielded the largest report to date on growing rods in 2005. This revealed dual submuscular growing rods without apical intervention as the most effective construct at initial correction, maintenance of correction, and amount of growth per year via T1-S1 height.[9] Group 1 in the study by Thompson et al. underwent an apical fusion in addition to the insertion of growing rods, which resulted in less spinal growth. This suggested no benefit from the hypothetical improved mobility created when performing a short apical fusion, leading the authors to ultimately recommend against it.

Over the next decade, many surgeons treating early onset scoliosis transitioned to dual growing rods for the management of pediatric spinal deformity (Figure 1), resulting in a new breadth of literature surrounding the technique. Mahar et al. identified the most biomechanically sound foundations when using growing rods attached to the spine, reporting that adjacent segment pedicle screws without a crosslink provided the greatest pull-out strength when tested in the laboratory.[10] This was a transformation in this patient population as hooks and sublaminar wires had predominated in the preceding generation of instrumentation systems.

Animal models confirmed that posterior-based distraction permits axial skeletal growth and may even stimulate it.[11] Knowing that growth was now possible, the

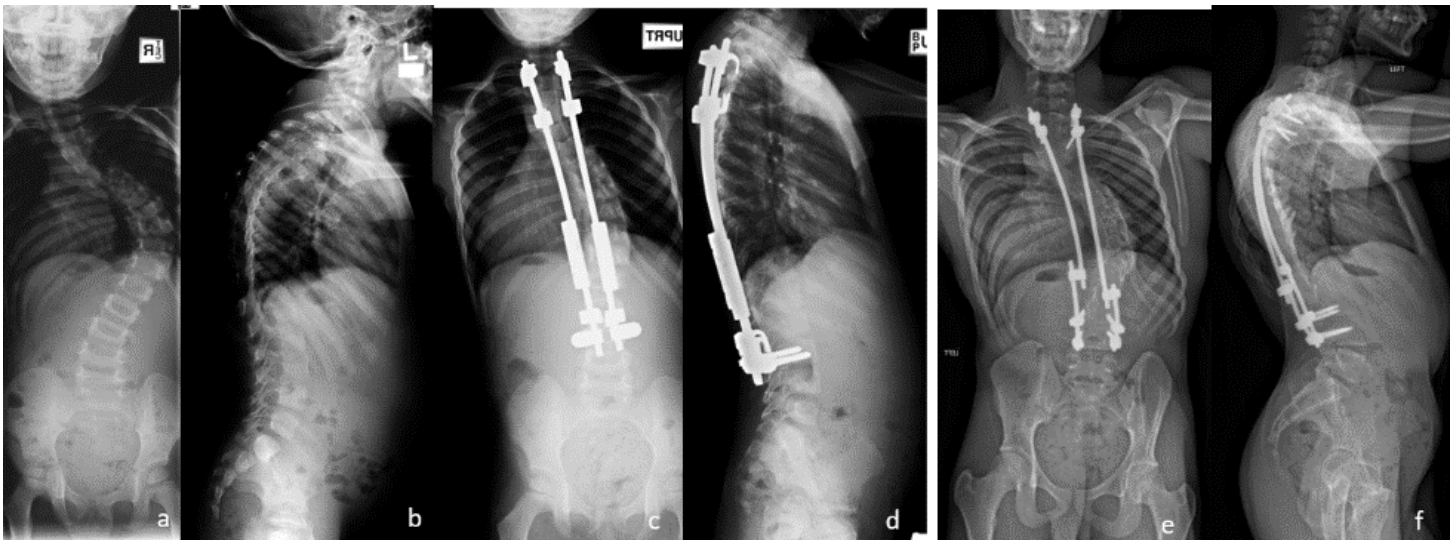

Figure 1. *Three-year-old male with infantile idiopathic scoliosis preop AP and lateral XR (a and b), 6 weeks (c and d) and 15 years (e and f) postop XR after dual-rod spine to spine growing rod insertion.*

timing of lengthening intervals became a popular topic of interest with studies documenting a lengthening interval of less than 6 months resulting in greater overall spine height and superior deformity correction.[12] In an ideal situation, surgeons could continue lengthening the growing rods until every patient reached a thoracic spinal height > 18 cm to minimize the likelihood of developing restrictive lung disease; however, numerous obstacles remain such as the auto-fusion problem described by Cahill et al. and the "law of diminishing returns" by Sankar et al.[13-14] Cahill reported 89% of the patients initially treated with growing rods had auto-fusion at the time of final fusion while Sankar identified decreased lengthening achieved with each subsequent lengthening surgery.

Auto-fusion and failure-to-lengthen were two of the many complications encountered in managing EOS with growing rods. Wound complications (superficial and deep), implant complications (rod fracture, anchor failure), neurologic complications, alignment complications, and medical complications were compiled in the study published by Bess et al. in *JBJS* in 2010.[15] Eighty-one patients experienced 177 complications out of the 140 patients in the study. Authors concluded that the younger the patient is at the time of insertion results in more lengthening surgery, thus increasing the likelihood of a complication. Older age at insertion surgery implies greater soft tissue envelope, less lengthening surgery, and improved bone stock for load sharing of dual rods which when combined, diminishes the overall complication profile. With each subsequent surgery, the risk of complications increases by 24%, according to Bess et al.[15] Additional reports identified other patient and surgeon-specific factors that play a role in complications including, but not limited to, small diameter rods, single rods, history of rod fracture, thoracic hyperkyphosis, and the magnitude of proximal thoracic deformity.[16-19]

In a single-center study, Phillips[20] found that the complication rate in a series of 28 EOS patients who underwent 165 procedures was 89% and the mortality rate was 18%. Four of the deaths were from respiratory failure and one occurred following infection. The authors eloquently highlighted the problems inherent to fusionless surgery, noting that without fusion, implants become load bearing rather than load sharing devices, making them susceptible to breakage. Improving bending resistance by doubling the implants (dual vs. single rods) increases the load to fatigue failure but at the potential cost of increasing the chance of skin breakdown and infection.

Heterogeneity in patient population and variations in practice patterns made consensus difficult to achieve when it came to growing rods and EOS. Yang et al.

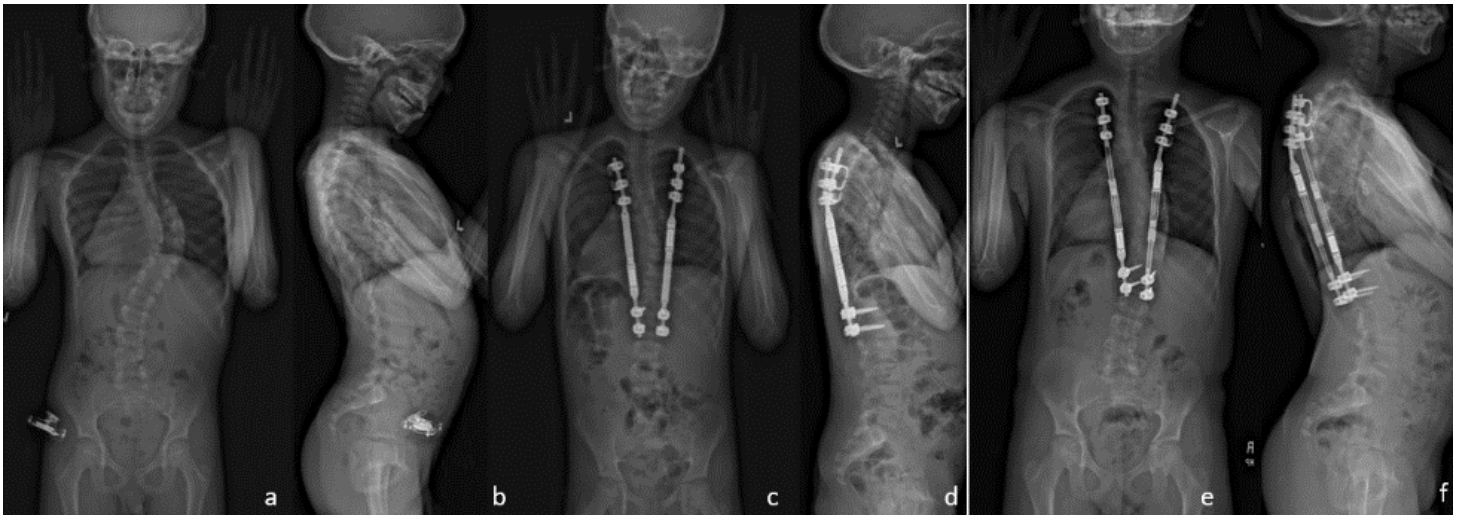

Figure 2. *Eight-year-old male with juvenile idiopathic scoliosis preop AP and lateral XR (a and b), 6 weeks (c and d) & 3 years (e and f) postop XR after rib to spine MCGR insertion.*

reported survey results in 2010 from a large group of surgeons on indications for growing rod treatment. While consensus was reached for indications in curve magnitude (over 60 degrees) and age of treatment (8-10 years old), a similar project repeated a decade later found no consensus on any of the six EOS cases presented.[21-22] The body of literature for growing rods continued to expand when authors began investigating construct success by disease type including but not limited to Marfan's syndrome, spinal muscular atrophy, Loeys-Dietz, and neurofibromatosis type 1.[23-27]

Magnetically Controlled Growing Rods (MCGR)

Repeat surgery, subcutaneous implants, and thin patients led surgeon innovators to realize that a remote-controlled lengthening rod could potentially be a landmark innovation for patients with early onset scoliosis. This concept first appeared in the literature in 1998 when Takaso et al. reported their experience with five beagles who had coronal cobb angles corrected, on average, from 25 degrees to 3 degrees over the course of 12 weeks via a remote-controlled system.[28] More than a decade later, Akbarnia et al. detailed their experience with nine Yucatan pigs, six of whom underwent weekly lengthening while three animals functioned as a control.[29] The authors identified the vertebral unit height (VUH), the distance between the center of the superior disc and the center of the inferior disc for each segment spanned by the rods. At the conclusion of the study (10 weeks), the VUH in the MCGR group was significantly greater ($p<0.05$) in the experimental group (32.2%) compared to the control (11.7%).[29] Less than a year later in the medical journal *Lancet*, Cheung et al. detailed the 24-month outcomes in two EOS patients treated with MCGR.[30] In their two patients, the T1-S1 and T1-T12 spinal length matched or exceeded the predicted growth of a healthy child aged 5–10 years.[31] This led the authors to conclude that the remote lengthening technology may be the solution to allow for continued spinal growth via posterior distraction while minimizing auto-fusion (Figure 2).

Over the next few years, the literature exploded with early reports of outcomes, which seemed promising, for the use of MCGR in management of pediatric spinal deformity. The U.S., British, and French experiences with the MCGR between 2013 and 2016 were mostly positive.[32-35] Consensus was that the new technology generally eliminated the need for planned repeat surgical lengthening, but MCGR did experience a similar complication profile to the traditional growing rod and unplanned return to the OR for anchor pull out, proximal junctional kyphosis, rod fracture, and the "law of diminishing returns." Lebon et al. were the first group to re-

port a significant discrepancy between the attempting lengthening measurement and the actual total spine length gained (45.5% discrepancy).

As with every new technology, longer follow-up produced new questions and unforeseen complications. First, surgeons questioned the unplanned return to the operating room (UPROR) rate for the MCGR. Early reports indicated that 46.7% of patients (14/30) returned to the operating room at an unplanned time for typical EOS complications such as rod fracture, infection, and proximal anchor failure, but additional new issues emerged such as failure of the MCGR to lengthen.[36] A 2-year analysis of all instrumentation strategies across all EOS diagnoses revealed an UPROR rate of 23%, with the hyperkyphotic neuromuscular patient population being the highest risk group for complication and the MCGR having a slightly higher risk of UPROR (p=0.009) compared to the traditional instrumentation.[37] This result surprised the authors; however, further investigation revealed many TGR/VEPTR complications are able to be put off and dealt with at the regularly scheduled surgery, thus not technically resulting in an UPROR. That approach is not possible with the MCGR. Many families currently being treated with TGR and VEPTR requested their child be converted to an MCGR to minimize anesthetic exposure with subsequent surgeries. The Pediatric Spine Study Group reported in 2018 on 383 patients comparing primary and conversion MCGR cases. The authors concluded that primary patients have improved radiographic correction and spine height gained, and the patients converted to MCGR had a greater rate of complications.[38]

Issues with remote lengthening have plagued the MCGR since its inception into the EOS community. Risk factors for rod slippage in the MCGR include increased distance between the magnet and the external remote control on the skin and a reduced distance between the magnets in a dual rod construct.[39] Cheung et al. concluded that desired lengthening returns to baseline following a rod exchange, suggesting that the "law of diminishing returns" may not be as clear cut in the MCGR as in the TGR. In contrast, Ahmad et al. reported that unlike TGR, the MCGR experiences a more linear decline in distraction achieved over a 51-month period.[40] Ahmad et al. did confirm the Cheung et al. conclusion that weight, age, and BMI of the patient negatively correlated with the ability of the MCGR to achieve the intended distraction length.[38,39] The literature on this topic continued to proliferate when Gilday et al. calculated the differences in rod length compared to actual distraction length, which came out to be 14% less than expected.[41] Similar to Dr. Robert M. Campbell's approach with the VEPTR, Lorenx et al. described the rib-to-pelvis MCGR construct for EOS deformity management and did not find as high of a rate of MCGR failure to achieve expected distraction length.[42] As follow-up increased, it became apparent that the rod slippage rate, failure to lengthen, and amount of force created by the MCGR decreases.[43] In two reports by Rushton et al., independent evaluation of explanted MCGR showed that rods implanted over 38 months previously produced no force (0/12) and 62% (34/55) of the constructs were no longer functional at the time of explant.[44,45] This is problematic for EOS patients who are more than 3 years away from implant removal or a final fusion surgery. It also begs the question, Is the MCGR as cost effective as original described? Cost-effectiveness studies indicate the MCGR must function correctly for 3–6 years to warrant the high price of the rod, and basic science studies now indicate that the rods may fail prior to 38 months, leading some authors to believe that the MCGR may not necessarily be the "game-changing" technology it was once thought to be.[46-48]

Authors' Preferred Technique: MCGR Insertion

The technique involves correction of the deformity with preservation of spinal soft tissues to minimize the development of auto-fusion. In the nonambulatory patient population, rib anchors proximally and pelvic hooks distally are preferred (Technique Video 1). The cephalad anchors are chosen by assessing the end vertebrae in the coronal plane and the apex of deformity in the sagittal plane placed through a midline skin incision. A minimum of five anchor sites for each rod is preferred to load share initial deformity correction and subsequent lengthening. Caudal anchors are placed through a longitudinal

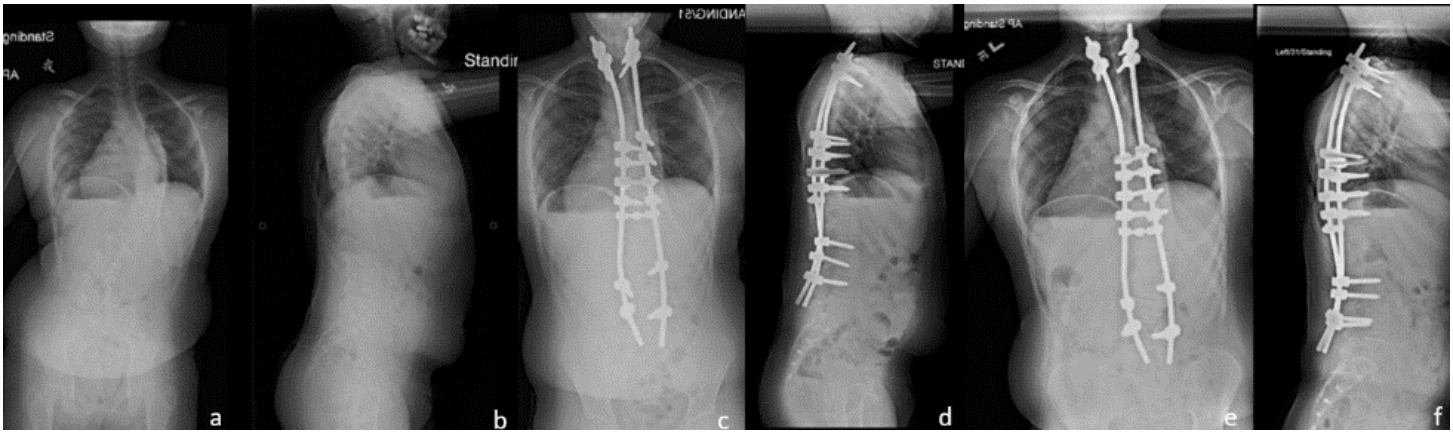

Figure 3. *Nine-year-old male with early onset scoliosis pre-op AP and lateral XR (a and b), immediate postop (c and d), and 9 months postop (e and f) XR after Shilla growth guidance with apical control.*

incision centered over the most cephalad aspect of the iliac crest. Ideal hook placement is a few millimeters medial to the most cephalad aspect of the iliac crest. Minimizing the soft tissue window decreases migration risk and the inner table of the pelvis must be gently dissected to facilitate hook placement. A temporary rod on the concavity of the deformity is placed and a combination of translation and distraction correct deformity. A definitive MCGR can then be cut, contoured, and tunneled from proximal to distal on the convexity, followed by removal of the temporary rod on the concavity and replacement with a definitive MCGR.

In the ambulatory EOS patient, the authors prefer the MCGR construct anchor to the spine distally and the ribs proximally. The rib anchors are selected as above while the distal anchors are the end vertebrae of the coronal plane deformity consisting of bilateral pedicle screws at two adjacent levels. No cross-link is applied when the rods are in reverse orientation on one side and standard orientation on the other. Implant prominence is minimized when the rod is cut and contoured to allow the actuator to lay over a flat part of the spine.

Growth Guidance

The second generation of growth guidance implants built upon the concept of a "Luque trolley." Whereas the Luque trolley attempted to guide growth using sublaminar wires, which ultimately led to interlaminar ankylosis and auto-fusion, the developers of the second generation of growth guidance constructs utilized pedicle screws placed in an extraperiosteal fashion to guide growth. Also known as the "Shilla growth guidance system," the construct was designed to guide spinal growth along a pair of parallel rods without any intentional manual distraction.[49]

Results of the second-generation growth guidance systems have been mixed. Luhmann and McCarthy,[50] who developed the technique, reviewed EOS patients treated with Shilla and those treated with dual growing rods. The average T1-S1 growth following Shilla instrumentation was 1.68 cm/yr, which compared favorably with dual growing rods (1.32 cm/yr). However, Nazareth and coauthors[51] reviewed a different series of 20 patients who underwent Shilla growth guidance instrumentation and found that the T1-S1 growth in these patients was only 4.2 mm per year or 36% of predicted growth. Likewise, in a comparison of growing rod patients and growth guidance patients, Andras and coauthors[52] found that patients who underwent dual growing rods had better scoliosis correction, superior T1-S1 growth, and fewer unplanned reoperations than those who underwent treatment with growth guidance.

Authors' Preferred Technique: Shilla Growth Guidance
The technique first involves correction of the apical deformity into a neutral three-dimensional alignment. This requires standard subperiosteal dissection of the apical

three or four levels, placement of fixed head pedicle screws, and periapical posterior column osteotomies to achieve optimal correction of the apex. The cephalad and caudal Shilla polyaxial growth guidance pedicle screws are then typically placed at the end vertebrae of the deformity. They are placed bilaterally at two cranial levels and two caudal levels using a minimally invasive technique through the muscle that avoids subperiosteal exposure with the bone only visualized fluoroscopically or with the aid of intraoperative 3D navigation. The use of a Jamshidi trocar system can be helpful for placement of the screws using a cannulated technique (Technique Video 2).

Once the anchors have been placed, a temporary contoured rod is placed on the convex side of the curvature and loosely attached at the apex and to the growth guidance screws. This provisional rod is rotated into a neutral position, then it is translated using coronal rod benders toward the midline. This is held by tightening the convex apical set plugs. The permanent concave rod is then attached to anchors and the temporary convex rod replaced by a permanent convex rod. Derotation tubes are used to perform direct vertebral rotation at the apex. The fixed head screws lock the rods at the apical screws through standard locking set screws that fix to the rods. The guidance screw caps capture the rods in the guidance screw head, leaving room for movement of the rod within the screw head. A crosslink is placed just below the apical fixation in order to help maintain rod rotation (Figure 3). Bone graft is placed at the apical levels only.

Summary

The treatment for patients with EOS has evolved dramatically over the past 50 years. Harrington and Moe introduced the concept of instrumenting the pediatric spine while Akbarnia, Thompson, McCarthy, and Campbell revolutionized the instrumentation and approach. One of the more recent innovations, the MCGR, has its limitations, and over time the literature will identify what patient factors increase the likelihood of early rod failure.[53] Likewise, the Shilla procedure, which represents the second generation of growth guidance implants, has shown mixed results. In comparison to growing rods, Shilla appears be less likely to facilitate the desired growth of the T1-S1 segment following implantation while having a complication profile and unplanned reoperation rate similar to that of growing rods. Future technology advancement that preserve pulmonary function, maintains spinal growth, and improves on the modern-day complication profile in EOS will have the health-related quality of life improvement (HRQOL) surgeons desire and will truly be the long sought after EOS "game-changing" technology.

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