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ORIGINAL STUDY

The effect of distraction-based growth-friendly spinal instrumentation on growth in early-onset scoliosis

Mehmet Bülent Balioğlu, Yunus Atici, Akif Albayrak, Deniz Kargin, Yunus Emre Akman, Mehmet Akif Kaygusuz

The present study assessed the advantages and disadvantages of growth-friendly spinal instrumentation surgery for early-onset scoliosis in 17 patients who underwent this surgery with a minimum 2-year follow-up. The mean number of lengthening procedures was three, initial age at which surgery was performed was 108.1 ± 30.2 months, and follow-up duration was 40.6 ± 16.6 months. Spinal height (T1-S1 and T1-T12), lung space available, major Cobb angle for scoliosis, maximum thoracic kyphosis, lumbar lordosis, shoulder and pelvic balance, and coronal and sagittal balance were assessed preoperatively and at the last follow-up. Treatment with growth-friendly spinal instrumentation showed evident increases in the spinal height and space available for the lungs, and significant improvement in scoliosis and thoracic kyphosis. The most commonly observed complications were proximal anchor problems and proximal junctional kyphosis. To avoid proximal junctional kyphosis in treatments with growing rods, excessive thoracic kyphosis correction should not be performed.

Keywords : early-onset scoliosis ; growing rods ; growth-friendly spinal instrumentation ; proximal junctional kyphosis ; space available for lungs ; thoracic height ; thoracic kyphosis.

INTRODUCTION

Early-onset scoliosis (EOS) refers to progressive spinal deformities that occur in the early years of life (<10 years old) due to congenital, neuromuscular, syndromic, and idiopathic causes. Its prognosis and progression vary considerably depending on the particular aetiology *(5)*. In EOS, the speed of growth and ratio of the spine increase bimodally with age; initial growth occurs during the first 5 years of life, and then again during adolescence, from 10 years of age to skeletal maturity *(14,23)*. Most of the lung capacity, bronchial tree, and development of the alveolar structure are shaped by 8 years of age. During this period, progression of the spinal curvature and early spinal fusion restrict thoracic volume *(8,32)* and cause undesirable effects on pulmonary function *(13,21)*. EOS treatment aims to achieve normal physiological spinal growth and sufficient pulmonary function.

■ Mehmet Bülent Balioğlu, M.D.
■ Yunus Atıcı, M.D.
■ Akif Albayrak, M.D.
■ Deniz Kargın, M.D.
■ Yunus Emre Akman, M.D.
■ Mehmet Akif Kaygusuz, M.D.
Department of Orthopaedics and Traumatology, M.H. Metin Sabanci Baltalimani Bone Diseases Training and Research Hospital, Baltalimani, Istanbul, Turkey
Correspondence : Mehmet Bülent Balioğlu, M.H. Metin Sabanci Baltalimani Bone Diseases Training and Research Hospital, Rumeli Hisari Caddesi No. 62 Baltalimani, Istanbul, 34470, Turkey
E-mail : mbbalibey@gmail.com

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EOS treatment has rapidly developed over the last two decades, and many methods have been proposed, such as serial plastering *(17,24,29,31)*, the use of braces *(2,9)*, and surgery with growth-friendly implantation systems using non-fusion techniques *(7,18,19,22)*. The goals of surgical treatment include stabilization of the spinal deformity and optimization of the space available for the lungs (SAL) until skeletal maturity and definitive spinal fusion *(18,27,28)*. Various distraction and growth-guiding techniques, including the use of growing rods (GRs), are often used to treat severe progressive EOS *(35)*. However, high complication rates following the use of GR have been reported *(4)*; attention has been particularly focused on sagittal plane deformities and the increasing risk of junctional problems *(27)*. Our study's objectives were to 1) assess the effects of distraction-based, growth-friendly spinal instrumentation (GFSI) on spinal and thoracic growth, and 2) assess coronal and sagittal plane complications encountered during EOS deformity correction.

MATERIALS AND METHODS

We retrospectively examined data from patients with EOS treated with single, dual, or hybrid GRs between June 2007 and January 2013. Inclusion criteria were patients who had undergone hybrid-, single-, or dual-GR surgery, those who had open triradiate cartilage and a Risser grade of 0 at the time of the first surgery, and patients who had a follow-up duration of 2 years or more after index surgery. Anteroposterior (AP) and lateral-view standing radiographs taken preoperatively and at the last follow-up were assessed for each patient. Seventeen patients with complete sets of radiographs were included in the final analysis.

Radiographic parameters from preoperative and final follow-up AP and lateral standing whole-spine radiographs were measured to determine the magnitude of coronal and sagittal deformity, amount of spinal growth, proximal junctional kyphosis (PJK), and any signs of implant failure. On AP radiographs, the T1-S1 spinal height and T1-T12 height were measured. The SAL was measured on both the concave and convex sides of the thoracic cage on AP radiographs. The Cobb angle, coronal balance (C7 to central sacral vertical plumb line), shoulder, and pelvic balance were measured for scoliosis (Fig 1).

In the sagittal plane, the maximum thoracic kyphosis (TK), lumbar lordosis (LL) angle, and sagittal balance (posterior-superior corner of the S1-C7 vertical plumb line) were measured (Fig. 2). PJK was measured from the superior endplate two vertebral levels above the upper instrumented vertebra (UIV) to the inferior endplate of the UIV (Fig. 3) *(27)*. Patients were divided into subgroups on the basis of the PJK angle; ≤10° (n = 7, group 1) and >10° (n = 10, group 2).

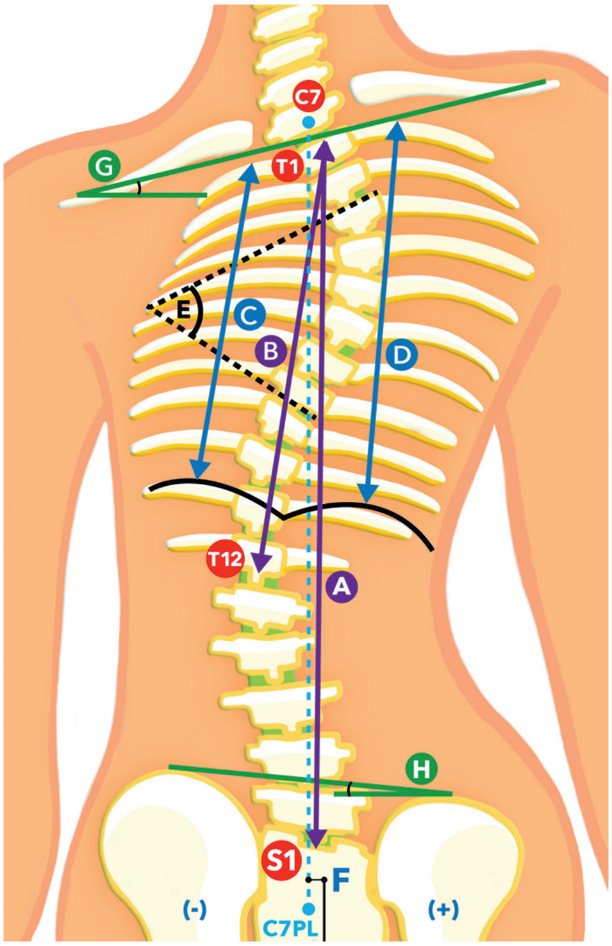

Fig. 1.— Radiographic parameters on the AP standing in whole-spine radiographs. Measured on AP: **(A)** T1–S1 spinal height, **(B)** T1–T12 height, **(C)** the space available for the lung (SAL) on both the concave, and **(D)** convex sides of the thoracic cage, **(E)** Cobb angle, **(F)** coronal balance (C7–central sacral vertical plumb line), **(G)** shoulder, and **(H)** pelvic balance.

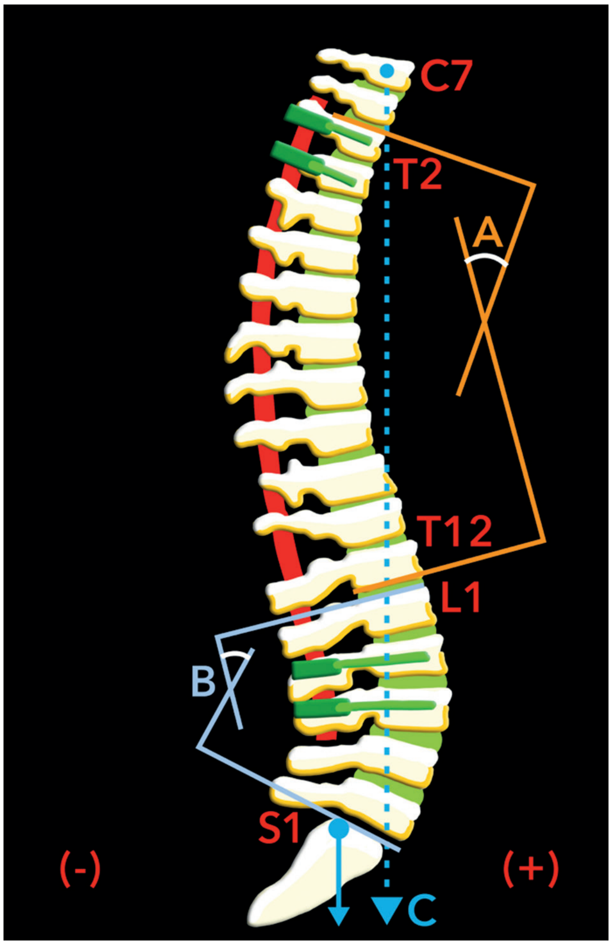

Fig. 2.— In the sagittal plane: **(A)** maximum thoracic kyphosis (TK), **(B)** lumbar lordosis (LL) angle, and **(C)** sagittal balance (posterior-superior corner of S1–C7 vertical plumb line) measured.

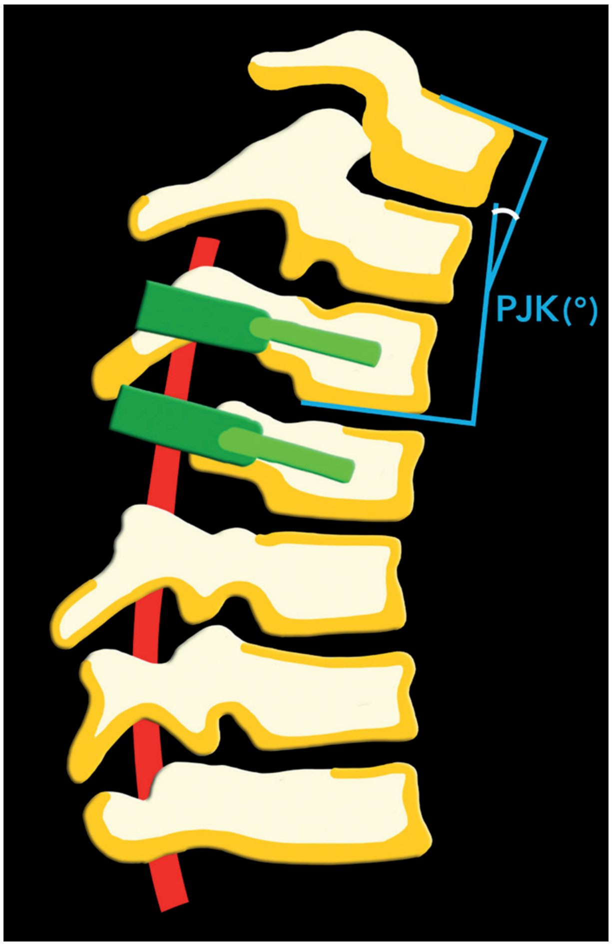

Fig. 3.— PJK measured from the superior endplate two vertebral levels above the upper instrumented vertebra to the inferior endplate of the upper instrumented vertebra *(32)*.

Eleven of 17 patients were girls and 6 were boys. The mean age at operation was 108.1 ± 30.2 months (median, 109; range, 32-144 months). The mean interval between lengthening procedures was 9.3 ± 2.8 months (median, 8.7; range, 6-18 months). The mean follow-up duration was 40.6 ± 16.6 months (median, 34; range, 24-72 months). The average number of lengthening procedures was 3 (range, 1-6). The number of patients discharged with post-lengthening fusion was 9, and we are currently following up with the remaining 8 with GRs.

The GFSI distributions were as follows: 14 double GRs (DGR), 2 hybrid GRs (HGR), and one single GR (SGR). Diagnoses were idiopathic scoliosis in 6 patients, syndromic scoliosis in 6 patients (1 cleidocranial dysplasia, 1 neurofibromatosis, 1 osteogenesis imperfecta, and 3 unknown), neuromuscular scoliosis in 3, and congenital scoliosis in 2 (1 segmentation and 1 formation anomaly). The patients were also grouped according to the newly defined classification of EOS *(34)*.

RESULTS

GFSI improved the coronal plane deformity in all patients. The mean concave and convex SAL also increased significantly from before surgery to the last follow-up. The overall mean SAL (convex/concave) was 1.02 cm preoperatively and 0.9 cm at the latest follow-up.

The maximum TK decreased significantly after surgery, but the mean LL did not change. Additionally, the mean T1-S1 distance and T1-T12 distance also significantly changed from the preoperative values to those obtained at the last follow-up.

Neither shoulder balance nor pelvic balance changed significantly by the last follow-up, and the coronal and sagittal balance did not change.

Twenty complications occurred in 13 patients due to the use of GRs, with the most frequent being PJK, which was corrected in 3 patients treated with fusion, and it was observed in 10 patients (58.8%) at the last follow-up. Other complications caused by the implant or related to the wound site were identified in 7 patients. Problems associated with implant migration occurred with the proximal anchor of 3 patients (implant loosening) and with the distal anchor of 3 patients (screw and rod breakage); wound-site complications were observed in 1 patient. PJK was corrected in 3 patients treated with fusion during post-GR treatment (PJK <10°). In 7 patients with an original PJK angle of <10°, PJK did not develop. Four of them underwent fusion, and in 3, GR treatment was on-going. For proximal or distal implant failures, implants were replaced and lengthened during the same session. Patients who developed infection were treated with wound-site debridement and antibiotics. None of the patients developed neurological complications. In 1 patient, HGR was replaced with GR, and in another, the GR was replaced with a magnetically controlled GR (MCGR; MAGEC Ellipse Technology, Irvine, CA).

At the final follow-up, there were no significant between-group differences in the preoperative and postoperative AP Cobb angles for patients in groups 1 and 2, and the AP Cobb angle was significantly decreased in both groups postoperatively. There were no significant differences between the two groups' preoperative and postoperative TK values. For group 1, the TK did not change after surgery, but for group 2, the TK value decreased significantly postoperatively. LL was not different between groups 1 and 2, and it did not change significantly in either group postoperatively. The spinal height (T1-S1 and T1-T12) did not differ between the groups, and it increased in both groups postoperatively.

A paediatrician assessed respiratory function preoperatively in all patients, and patients with no respiratory problems underwent surgery. No patients were treated with traction before or during surgery. Patients wore a protective corset for a minimum of 3 months after surgery.

DISCUSSION

EOS is a heterogeneous condition; its prognosis and progression vary according to its aetiology *(5)*. EOS refers to spinal curvature that occurs in the early years of life due to congenital, neuromuscular, syndromic, and idiopathic causes. The objective of EOS treatment is to increase spinal and thoracic growth while stopping or correcting deformity progression, to prevent thoracic insufficiency development, to avoid or restrict early fusion, to reduce negative impacts and surgical complications of the treatment, and to increase well-being *(12,13,28)*. Various surgical techniques were developed in the treatment of uncontrolled wide curves with serial plastering and brace treatment. These are growth-friendly methods and non-fusion surgical treatments *(2,7,12,18,19,22,28,29)*. According to the newly defined classification of EOS, the patients' aetiologies, major Cobb angles, kyphosis angles, and degrees of progression were used to classify patients with EOS *(34)*.

Growth-friendly implants are classified according to the direction of the corrective force they apply *(18)*. Among distraction-based, growth-friendly systems, GRs are recommended for all patients regardless of aetiologies with indications of flexible deformities and evident axial growth potential, with a Cobb angle of >60° (idiopathic scoliosis) and <10 years of age *(5,28,30,35)*. With distraction-based systems, lengthening is recommended at certain intervals in accordance with the annually expected growth; this is generally semi-annually *(1)*. In our study, the height of the spine (T1-T12 and T1-S1) and SAL (concave and convex) significantly increased. Moreover, a significant improvement in the scoliosis deformity and TK was found.

The DGR technique has been commonly used for treating EOS since it was popularized by Akbarnia *et al*. (Fig. 4). It was reported that greater correction was obtained with DGR than with SGR, and DGR was more stable, which resulted in an increase in autofusion *(5)*. The rate of complications with GR was found to be as high as 40-60%, and a 24% increase was identified in complications associated with each surgical application *(6)*. It has been reported that repeated distractions result in less lengthening due to the law of diminishing returns *(18,25,26,28)*. Sankar *et al.* showed that there is a decrease in the T1-S1 length gained with each subsequent DGR lengthening *(26)*. This decrease is attributed to stiffening of the spine, possibly due to autofusion. Additional surgical lengthening increases distraction forces, and these forces are doubled after the fifth lengthening *(20)*. Rigid distraction systems cause a lack of movement of facet joints and autofusion of the spine *(18)*. MCGR, which has recently become popular, has an effect similar to that of DGR, because it causes fewer lengthening-related complications and soft-tissue problems, as it is applied using a closed method *(3,9)*. With MCGR, wound infection, rod breaking, autofusion, and anchor insufficiency were reported to be very low in 34 patients with EOS with a mean age of 8 years due to a mean number of lengthening procedures of 4.8 *(10)*. Similar to vertical expandable prosthetic titanium rib, hybrid GRs can be implemented in cases with insufficient bone stock for upper anchors, spinal hooks to the ribs, lower anchors, pedicle screws (limited fusion) and/or supra-laminar hooks (non-fusion), and proximal pedicle fixation of vertebrae *(18,25)*. We also implemented HGR with hook fixation for the initial treatment of two patients who were ineligible for pedicle screw fixation: SGR in one case and DGR in the other case. Two patients (one treated with HGR) underwent replacement DGR, and in another patient (treated with DGR), the implants were replaced with MCGR to prevent repeated surgical intervention.

One possible complication during treatment is the development of a kyphotic deformity in the spinal segments immediately superior to the upper

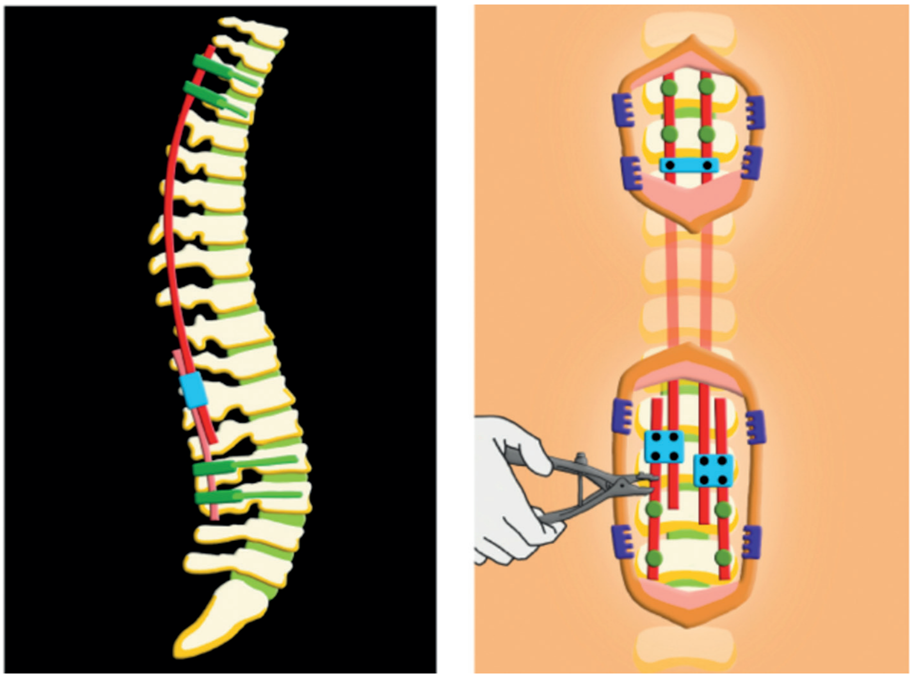

Fig. 4.— The GR technique commonly used in EOS treatment: **(A)** lateral and **(B)** anteroposterior view of cranial and caudal instrumentations using screws or hooks according to anatomic specialty. In both ends of the instrumentations, a stable foundation should be carefully established.

level of instrumentation (PJK) *(4,11,27,33)*. The incidence of PJK in patients with EOS with GRs has been reported to be 26-46% *(15,33)*. With the use of any rigid spinal instrumentation, PJK is a concern, and it has previously been noted as a potential complication *(4)*. Various methods have been defined to measure PJK; it is commonly defined as >10° of kyphosis above the UIV *(15,16)*. However, we preferred the method used by Shah *et al*. in which PJK was measured from the superior endplate two vertebral levels above the UIV to the inferior endplate of the UIV *(27)*. Shah *et al*. found a mean 10° increase in PJK at the latest follow-up. Their results showed that PJK increases over time, but the correlation between the number of lengthening procedures and PJK was not significant. According to Shah *et al*., PJK is a potential complication of the procedure, thus caution must be exercised when using rigid spinal instrumentation *(27)*. Recently, Watanabe *et al*. evaluated clinical and radiographic results from 88 patients with EOS who underwent DGR surgery *(33)*. PJK developed in 23 patients (26%); in 19 of these patients, the proximal foundation became dislodged following PJK *(33)*. In our follow-up, PJK (58.8%; mean 25.6°, range 13-40°) was higher than that reported in the literature *(27,33)* (Fig. 5). PJK developed in patients in whom TK was corrected excessively compared to the initial degree of TK (Fig. 6).

The weaknesses of our study were the small number of patients, the short follow-up period, the advanced age at first GR treatment, medical problems associated with the heterogeneity of the aetiologies, and failure to realise the lengthening at planned times due to various factors.

In conclusion, GR treatment of EOS may result in significant correction of scoliotic and kyphotic deformities, and a significant increase in spinal growth; it may also contribute to the long-term well-being and function of patients. However, attention should be focused on PJK. Nevertheless, long-term results of a larger number of patients with EOS

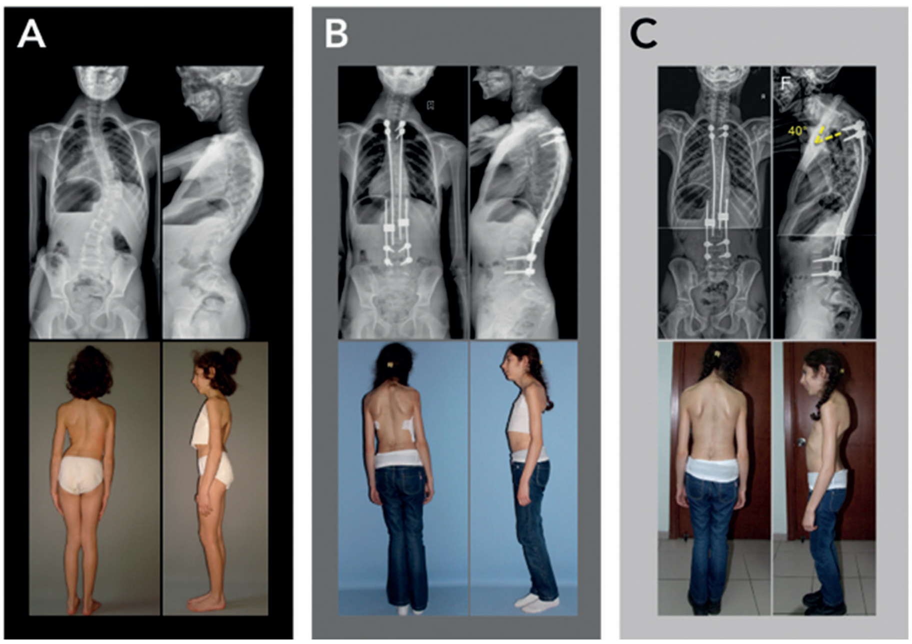

Fig. 5.— EOS with unidentified syndromic reason (in a 9-year-old female). The follow-up period was 24 months, three lengthenings perferformed during this period: **(A)** preoperative AP-lateral x-ray (50° major scoliosis and 40° kyphosis) and clinical pictures, **(B)** 12 month-follow-up postoperative GR, and **(C)** the latest follow-up PJK (40°) seen after GR distraction (29° major scoliosis and 39° kyphosis)**.**

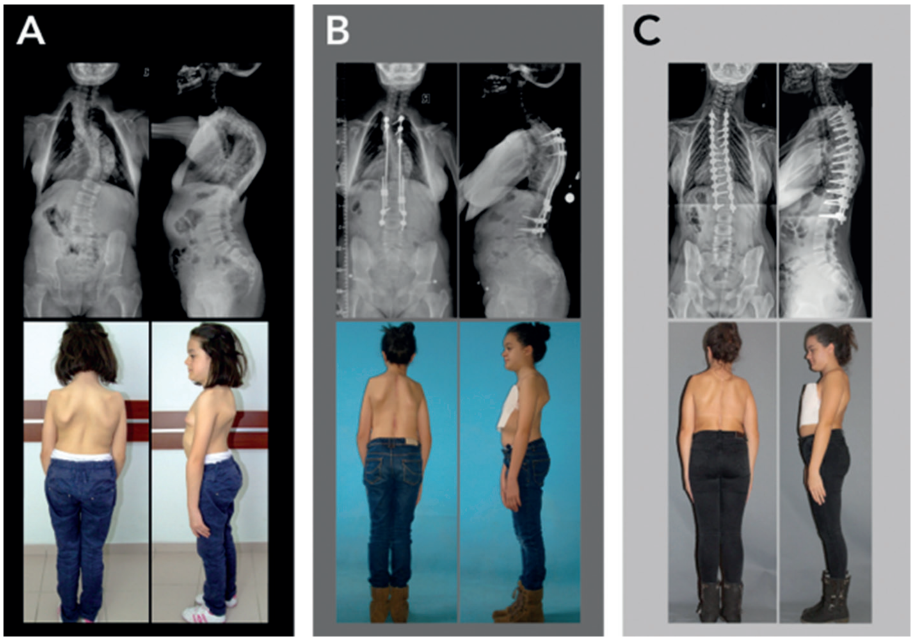

Fig. 6.— EOS with cleidocranial dysplasia (in a 12-year-old female). Treatment completed with fusion as a result of three lengthenings. The follow-up period was 33 months. **(A)** Preoperative AP-lateral x-rays (74° thoracolumbar scoliosis and 70° thoracic kyphosis), **(B)** PJK developed (25°) during GR, **(C)** the latest follow-up after posterior fusion with PJK regressed during the early postoperative-fusion period (9°). The major curve reduced to 19° scoliosis and 34° kyphosis.

are required to better assess the effectiveness of lengthening treatment using GFSI, and prevent and resolve the problems associated with this treatment.

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REFERENCES

1. **Akbarnia BA, Breakwell LM, Marks DS**, ***et al***. Dual growing rod technique followed for three to eleven years until final fusion: the effect of frequency of lengthening. *Spine (Phila Pa 1976)* 2008; 33 (9): 984-990.
2. **Akbarnia B, Campbell Jr R, McCarthy RE.** Optimizing Safety and Outcomes in Spinal Deformity Surgery: Early-Onset Scoliosis. *Spine Deformity (Preview Issue)* 2012: 53-63.
3. **Akbarnia BA, Cheung K, Noordeen H,** ***et al.*** Next generation of growth-sparing techniques: preliminary clinical results of a magnetically controlled growing rod in 14 patients with early-onset scoliosis. *Spine (Phila Pa 1976)* 2013; 38: 665-670.
4. **Akbarnia BA, Emans JB.** Complications of growth-sparing surgery in early-onset scoliosis . *Spine (Phila Pa 1976)* 2010; 35 (25): 2193-2204.
5. **Akbarnia BA, Marks DS, Boachie-Adjei O,** ***et al.*** Dual growing rod technique for the treatment of progressive early-onset scoliosis: A multicenter study. *Spine (Phila Pa 1976)* 2005; 30 (17 suppl): S46-S57.
6. **Bess S, Akbarnia BA, Thompson GH,** ***et al.*** Complications of growing-rod treatment for early-onset scoliosis: analysis of one hundred and forty patients. *J Bone Joint Surg Am* 2010; 92 (15): 2533-2543.
7. **Campbell RM Jr, Hell-Vocke AK.** Growth of the thoracic spine in congenital scoliosis after expansion thoracoplasty. *J Bone Joint Surg Am* 2003; 85-A: 409-420.
8. **Campbell RM Jr, Smith MD.** Thoracic insufficiency syndrome and exotic scoliosis. *J Bone Joint Surg Am* 2007; 89 (suppl 1): 108-122.
9. **Cheung KM, Cheung JP, Samartzis D,** ***et al.*** Magnetically controlled growing rods for severe spinal curvature in young children: a prospective case series. *Lancet* 2012; 379: 1967-1974.

10. **Dannai Z, Altaf F, Harshavardhana NS, *et al.*** Early results of a remotely-operated magnetic growth rod in early-onset scoliosis. *Bone Joint J* 2013; 95-B: 75-80.
11. **Glattes RC, Bridwell KH, Lenke LG, *et al.*** Proximal junctional kyphosis in adult spinal deformity following long instrumented posterior spinal fusion: incidence, outcomes, and risk factor analysis. *Spine (Phila Pa 1976)* 2005; 30: 1643-1649.
12. **Gomez JA, Lee JK, Kim PD, *et al.*** "Growth Friendly" Spine Surgery: Management Options for the Young Child With Scoliosis. *J Am Acad Orthop Surg* 2011; 19: 722-727.
13. **Karol LA**. Early definitive spinal fusion in young children: What we have learned. *Clin Orthop Relat Res* 2011; 469 (5): 1323-1329.
14. **Karol LA, Johnston C, Mladenov K, *et al*.** Pulmonary function following early thoracic fusion in non-neuromuscular scoliosis. *J Bone Joint Surg Am* 2008; 90 (6): 1272-1281.
15. **Lee C.** *Proximal junctional kyphosis in distraction-based growing rods*. In: Skaggs DL, ed. Scoliosis Research Society Meeting. Louisville, KY; 2011.
16. **Lee GA, Betz RR, Clements DH*, et al***. Proximal kyphosis after posterior spinal fusion in patients with idiopathic scoliosis. *Spine (Phila Pa 1976)* 1999; 24: 795-799.
17. **Mehta MH.** The rib-vertebra angle in the early diagnosis between resolving and progressive infantile scoliosis. *J Bone Joint Surg Br* 1972; 54 (2): 230-243.
18. **Mehta MH.** Growth as a corrective force in the early treatment of progressive infantile scoliosis. *J Bone Joint Surg Br* 2005; 87 (9): 1237-1247.
19. **Moe JH, Kharrat K, Winter RB, *et al.*** Harrington instrumentation without fusion plus external orthotic support for the treatment of difficult curvature problems in young children. *Clin Orthop Relat Res* 1984; 185: 35-45.
20. **Noordeen HM, Shah SA, Elsebaie HB, *et al.*** In vivo distraction force and length measurements of growing rods: which factors influence the ability to lengthen? *Spine (Phila Pa 1976)* 2011; 36 (26): 2299-2303.
21. **Pehrsson K, Larsson S, Oden A, *et al.*** Long-term follow-up of patients with untreated scoliosis: A study of mortality, causes of death, and symptoms. *Spine (Phila Pa 1976)* 1992; 17 (9): 1091-1096.
22. **Pratt RK, Webb JK, Burwell RG, *et al.*** Luque trolley and convex epiphysiodesis in the management of infantile and juvenile idiopathic scoliosis. *Spine (Phila Pa 1976)* 1999; 24: 1538-1547.
23. **Reid LM.** Lung growth in health and disease. *Br J Dis Chest* 1984; 78 (2) :113-134.
24. **Sanders JO, D'Astous J, Fitzgerald M*, et al.*** Derotational casting for progressive infantile scoliosis. *J Pediatr Orthop* 2009; 29 (6): 581-587.
25. **Sankar WN, Acevedo DC, Skaggs DL.** Comparison of complications among growing spinal implants. *Spine (Phila Pa 1976)* 2010; 35 (23): 2091-2096.
26. **Sankar WN, Skaggs DL, Yazici M, *et al.*** Lengthening of dual growing rods and the Law of Diminishing Returns. *Spine (Phila Pa 1976)* 2010; 36: 806-809.
27. **Shah SA, Karatas AF, Dhawale AA, *et al.*** The Effect of Serial Growing Rod Lengthening on the Sagittal Profile and Pelvic Parameters in Early-Onset Scoliosis. *Spine (Phila Pa 1976)* 2014; 39: 1311-1317.
28. **Skaggs DL, Akbarnia BA, Flynn JM**, ***et al***. A classification of growth friendly spine implants. *J Pediatr Orthop* 2014; 34 (3): 260-274.
29. **Sturm PF, Anadio JM, Dede O.** Recent advances in the management of early onset scoliosis. *Orthop Clin North Am* 2014; 45 (4): 501-514.
30. **Thompson GH, Akbarnia BA, Campbell RM Jr**. Growing rod techniques in early-onset scoliosis. *J Pediatr Orthop*. 2007; 27: 354-361.
31. **Tis JE, Karlin LI, Akbarnia BA *et al.*** Early onset scoliosis: modern treatment and results. *J Pediatr Orthop* 2012; 32 (7): 647-657.
32. **Vitale MG, Matsumoto H, Bye MR, *et al.*** A retrospective cohort study of pulmonary function, radiographic measures, and quality of life in children with congenital scoliosis: An evaluation of patient outcomes after early spinal fusion. *Spine (Phila Pa 1976)* 2008; 33 (11): 1242-1249.
33. **Watanabe K, Uno K, Suzuki T, *et al*.** Risk Factors for Proximal Junctional Kyphosis Associated with Dual-Rod Growing-Rod Surgery for Early-Onset Scoliosis. *J Spinal Disord Tech*. 2014 Jul 11.
34. **Williams BA, Matsumoto H, McCalla DJ, *et al.*** Development and Initial Validation of the Classification of Early-Onset Scoliosis (C-EOS). *Bone Joint Surg Am* 2014; 96 (16): 1359-1367.
35. **Yang JS, McElroy MJ, Akbarnia BA*, et al***. Growing rods for spinal deformity: characterizing consensus and variation in current use. *J Pediatr Orthop*. 2010; 30: 264-270.