Spine Deformity
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Spine Deformity 6 (2018) 478–482

Early-Onset Spinal Deformity in Skeletal Dysplasias: A Multicenter Study of Growth-Friendly Systems

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Received 5 January 2018; accepted 7 January 2018

Abstract

Purpose: Severe, early-onset spinal deformity is common in patients with skeletal dysplasia. These deformities often present at young ages and are associated with significant pulmonary dysfunction. The objective of this study is to verify the effectiveness of growth-friendly spinal instrumentation systems in promoting growth in patients with skeletal dysplasia and early-onset kyphoscoliosis.

Methods: A retrospective, multicenter comparative cohort study was performed. Twenty-three patients identified to have a skeletal dysplasia (SKD) were evaluated for diagnosis, age at treatment, gender, and type of growing rod construct (spine vs. rib constructs). Patients were matched by age and construct type with similarly treated patients with early-onset scoliosis (CON) without skeletal dysplasia. Radiographic parameters including maximum coronal and sagittal Cobb angle with levels, T1–S1 height, and T1–T12 height were measured.

Results: T1–T12 (12.8 vs. 15.2 cm, $p = .01$) and T1–S1 (21.2 vs. 24.5 cm, $p = .05$) heights were significantly shorter for the SKD group at implantation, and kyphosis tended to be more severe in children with SKD ($p = .80$ and .07, respectively). Kyphosis did not improve with treatment. Scoliosis improved ($p < .01$), and ΔT1–T12 and ΔT1–S1 significantly increased in both groups ($p < .01$). Complication rates were similar between the two groups; however, patients with SKD had more intraoperative monitoring changes and hardware failures ($p < .005$).

Conclusion: Although patients with SKD start with shorter spine lengths, gains in spine length appear to be comparable to other forms of EOS. Neuromonitoring changes and implant failures are more common in the SKD group.

Significance: The effectiveness of growth-friendly techniques in promoting growth in early-onset spinal deformities in patients with skeletal dysplasia has not been previously studied. We report the first comprehensive review of this topic. Growth-friendly techniques are an appropriate treatment option in this patient population.

Keywords: Skeletal dysplasia; Early-onset scoliosis; Kyphosis; Growing rods

Introduction

Severe spinal deformity is common in patients with skeletal dysplasia [1-8]. These deformities often present at a young age and are associated with significant pulmonary dysfunction [1,2]. Treatment with growth-friendly systems for the complex deformities seen in these patients has been performed at many centers, yet only a few centers have adequate experience to report

IRB approval: This study was reviewed and approved by Seattle Children's Research Institute IRB ID# PIROSTUDY14864.

Author disclosures: KKW (none), VB (none), SAS (none), GJR (none), WFK III (none), WGM (none), Children's Spine Study Group, Growing Spine Study Group.

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2212-134X/$ - see front matter © 2018 Scoliosis Research Society. All rights reserved.
https://doi.org/10.1016/j.jspd.2018.01.002

meaningful outcomes in patients with a skeletal dysplasia [9]. Given the limited growth potential in late childhood relative to other children with early-onset spinal deformities, the appropriateness of treatment at relatively older ages (age > 5 years) is not clear. Additionally, given the severity of deformity in many of these patients, and the high incidence of medical comorbidities, the risk for complications is potentially higher in this group compared to other children who undergo similar treatment modalities. The objective of this study is to define the population of patients with skeletal dysplasia that has been treated with growth-friendly systems, to verify the effectiveness in promoting spine growth in this patient population, and to define the risks associated with this therapy.

Methods

A retrospective, multicenter comparative cohort study was performed. Using two multicenter databases, 23 patients identified to have a skeletal dysplasia (SKD) were evaluated for underlying diagnosis, age at treatment, gender, and type of growing rod construct (spine vs. rib constructs). Radiographic parameters including maximum coronal and sagittal Cobb angle with levels, T1–S1 height, and T1–T12 height were measured preimplantation, immediate postimplantation, and at most recent follow-up. These patients were matched by age and construct type with a control cohort (CON) of similarly treated patients with early-onset scoliosis (EOS) without skeletal dysplasia. To compare the groups, we used the Wilcoxon matched-pairs signed-rank test. We compared changes in T1–S1 height, T1–T12 height, Cobb angle, and kyphosis from time to implant, first postoperative follow-up (3 months), and last follow-up using repeated measures analysis of variance. Postoperative complications were tabulated and compared between groups. Complications were analyzed using chi-square tests and Spearman correlations.

Results

Twenty-three patients with SKD and 23 controls were included in this analysis (Table 1). In the SKD group, mean age at first implant was 5.4 years (range 2.08-12.33). Sixteen girls and 7 boys with diagnoses of spondyloepiphyseal dysplasia (SED; 6), multiple epiphyseal dysplasia (MED; 2), diastrophic dysplasia (3), achondroplasia (1), cleidocranial dysostosis (1), camptomelic dysplasia (2), Conradi-Heunermann syndrome (2), and other types of SKD (6) were included. The length of follow-up in this group was 5.05 years (range 1.3-10.4). In the control group, the mean age at first implant was 5.6 years (range 2.49-10.65). Nine girls and 14 boys with diagnosis of neuromuscular scoliosis (4), congenital scoliosis (8), idiopathic infantile scoliosis (3), and other syndromic related EOS (9) were included in this group. Length of follow-up was 4.76 years (range 1.9-9.5). Twelve patients were treated with spine-to-rib constructs and 11 with spine-to-spine constructs in each group.

The SKD and CON groups were significantly different in T1–T12 length (12.8 vs. 15.2 cm, $p = .011$) and T1–S1 (21.2 vs. 24.5 cm, $p = .05$) at implantation. The groups did not significantly differ in major Cobb angle (71° vs. 72°; $p = .819$), whereas kyphosis tended to be more severe for the SKD group (60° vs. 39°; $p = .066$). From the time of implant, 3 months postoperative follow-up, to the last follow-up, no significant difference in kyphosis (Fig. 1) was observed in either group. Cobb angle (Fig. 2) significantly decreased in both groups during this time period ($p = .001$ in SKD and $p = .012$ in CON), Both T1–T12 and T1–S1 growth (Figs. 3 and 4) significantly increased in the SKD group ($p = .002$ and $p = .001$, respectively) and in the control group ($p = .003$ and $p = .001$). The normalization of growth demonstrated as a percentage of spine growth, both with initial implantation and at final follow-up, was not statistically different between groups (Tables 2 and 3).

Total complications were similar between the two groups (Table 4); however, patients in the control group had

Table 1
Distribution of patients included in study.

	Average age at implant (years)	Sex	Proximal anchor construct	Diagnosis	Length of follow-up (years)
Skeletal dysplasia	5.4	16 (F) 7 (M)	12 rib 11 spine	Spondyloepiphyseal dysplasia (6) Multiple epiphyseal dysplasia (2) Diastrophic dysplasia (3) Achondroplasia (1) Cleidocranial dysostosis (1) Camptomelic dysplasia (2) Conradi-Heunermann syndrome (2) Other (6)	5.05
Control	5.6	9 (F) 14 (M)	12 rib 11 spine	Neuromuscular scoliosis (4) Congenital scoliosis (8) Idiopathic infantile scoliosis (3) Other (4)	5.6