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

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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

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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

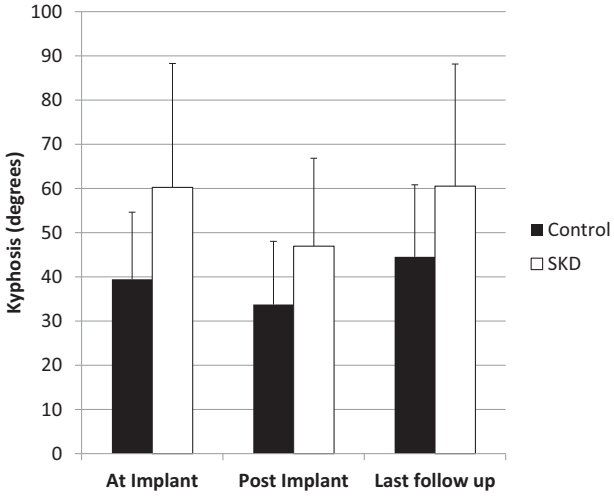

Fig. 1. Kyphosis is significantly greater in patients with skeletal dysplasias ($p = .0007$), but was stable over time after instrumentation in both groups.

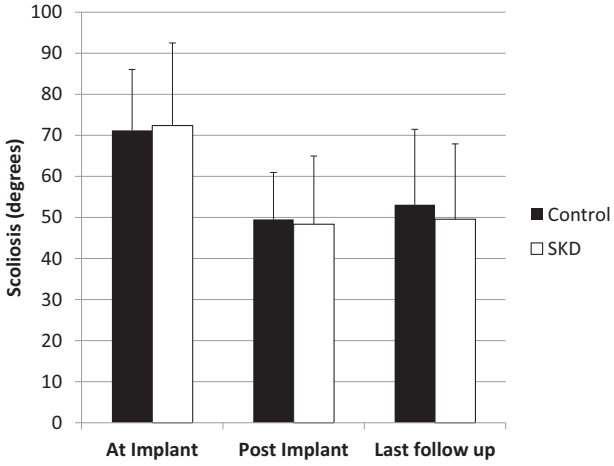

Fig. 2. Scoliosis was responsive to instrumentation in both groups ($p < .001$), and maintained at most recent follow-up.

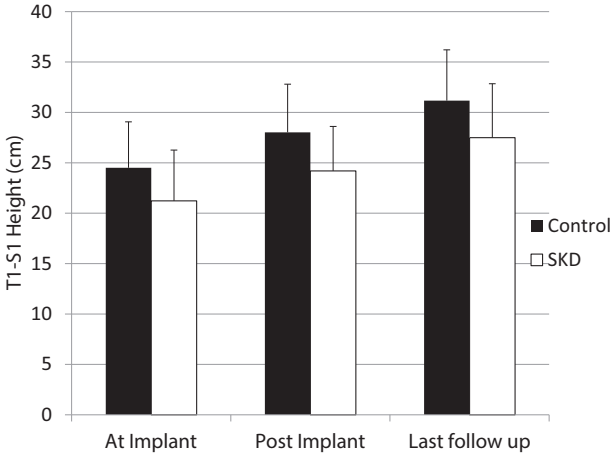

Fig. 3. Absolute T1–S1 height was greater for the CON group than the SKD group, but increased significantly in both groups ($p < .01$). CON, control; SKD, skeletal dysplasia.

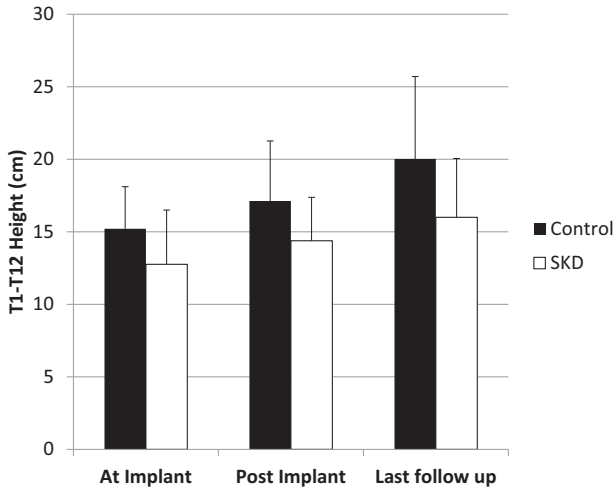

Fig. 4. Absolute T1–T12 height was greater for the CON group than the SKD group, but increased significantly in both groups ($p < .01$). CON, control; SKD, skeletal dysplasia.

Table 2
Average T1–S1 growth and percentage growth between preoperation and first follow-up after 3 months (FU1) and preoperation and last follow-up after 5 years (FU2).

Group	T1–S1 growth (cm)		p value	T1–S1 growth		p value
	Preoperation to FU1, Mean (SD)	Growth, %		Preoperation to FU2, Mean 2(SD)	Growth, %	
Skeletal dysplasia	2.92 (2.47)	12.25	.8097	6.71 (4.65)	23.27	.2388
Control	3.27 (3.70)	10.94		8.82 (4.14)	27.06	

SD, standard deviation.

Table 3
Average T1–T12 growth and percentage growth between preoperation and first follow-up after 3 months (FU1) and preoperation and last follow-up after 5 years (FU2).

Group	T1–T12 growth (cm)		p value	T1–T12 growth		p value
	Preoperation to FU1, Mean (SD)	Growth, %		Preoperation to FU2, Mean (SD)	Growth, %	
Skeletal dysplasia	1.73 (2.13)	17.45	.6899	4.27 (4.71)	29.98	.1899
Control	1.25 (1.74)	8.62		5.77 (4.60)	40.35	

significantly more pulmonary complications whereas the SKD group had more intraoperative monitoring changes ($p = .017$) and hardware failures ($p = .05$). There was greater variability in the number of complications observed in the control group, but the SKD group had a greater number of complications per patient (SKD: 2.43 complications/patient; Control: 1.73 complications/patient; Tables 5 and 6).

Table 4
Number of complications by group.

Group	Pneumonia/ pulmonary*	Infection	Hardware failure/ migration*	Fracture rib	Neurologic injury	IOM changes*	Other
Skeletal dysplasia	0	11	27	2	3	6	7
Control	11	9	11	2	1	0	6
Total	11	20	38	4	4	6	13

IOM, intraoperative monitoring.
* $p < .005$.

Table 5
Number of patients with complications.

Complications?	Skeletal dysplasia	Control	Total
No	6	8	14
Yes	17	15	32

No difference between groups ($\chi^2 = 0.4107$; $p = .522$).

Table 6
The range of number of complications.

Skeletal dysplasia		Control	
No. of complications	No. of patients	No. of complications	No. of patients
1	3	1	3
2	6	2	4
3	2	3	6
4	2	5	1
5	2	6	1
6	1		
9	1		

Discussion

Treatment of spinal deformity in skeletal dysplasia has been traditionally comprised of bracing followed by definitive fusion [1,2]. Because of the aggressive progression of deformity in many of these disorders, fusion has been historically recommended at a relatively young age. The rationale for recommending definitive fusion in young patients with skeletal dysplasia is based in the belief that these patients do not grow enough to be concerned about loss of potential lung growth.

With the increased awareness of potential pulmonary function compromise with early fusion, growth-friendly approaches to the treatment of spinal deformity in patients with skeletal dysplasia has been considered [9]. Good correction of coronal plane deformity and improvements in space available for the lung has been demonstrated with a predictable, but acceptable rate of complications in patients with skeletal dysplasia; however, it is not known whether these improvements are commensurate with those seen in other forms of early-onset scoliosis.

In this case-controlled study, we evaluate the response of early-onset spinal deformity associated with skeletal dysplasia as compared to patients with other etiologies of early-onset scoliosis. In this comparison, we have found that scoliosis can be corrected and maintained, whereas kyphosis tends to be more prevalent in patients with skeletal dysplasia but does not improve with treatment. Spine growth appears to be comparable between skeletal dysplasia patients and other forms of early-onset scoliosis over an average of five years' follow-up. The caveat here is that patients with skeletal dysplasias start with a shorter trunk and end with a shorter trunk.

There is considerable variability in chest growth to be expected among the skeletal dysplasia diagnoses, and for most there is no literature on norms for pulmonary function in affected adults. For some of these disorders (eg, camptomelic dysplasia, thanatophoric dysplasias, Conradi-Heunermann syndrome), compromised pulmonary function is often life-threatening and leads to early death. Generalizations about growth and pulmonary outcomes vary by diagnosis. Short trunk disorders such as the spondyloepiphyseal dysplasias, the spondyloepimetaphyseal dysplasias and the mucopolysaccharidoses should not be expected to grow in a fashion similar to short-limbed diagnoses such as achondroplasia and multiple epiphyseal dysplasia.

Nonetheless, this study clearly demonstrates the efficacy of growth-friendly spinal systems in this population. Much of the observed growth occurred with initial placement of the instrumentation in the skeletal dysplasia group, particularly when comparing T1–T12 spine lengths (about half the total growth, compared with one-quarter after initial implantation). When comparing T1–S1 lengths, however, this disparity was less (about one-half, compared to one-third of total growth after initial implantation). Statistically the percentage of spinal growth in both groups was similar for T1–T12 and T1–S1 measurements, whereas the absolute growth in T1–T12 length was less for the skeletal dysplasia group. Why a discrepancy exists between total spine growth and thoracic spine growth between these two groups is not a question these data can address.

Finally, we found that the overall rate of complications was not greater than the control group; however, there tended to be more implant failures and IOM changes in this group. This is likely attributable to the increased kyphotic deformity found in this patient population.

In conclusion, growth-friendly techniques promote growth of the spine in patients with skeletal dysplasia. They are therefore effective and appropriate in this patient population, and the risks are no greater than for other patients with early-onset scoliosis.

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