Similar Deformity Correction but Limited Spinal Growth of Growth-Friendly Management in Skeletal Dysplasia Associated Early Onset Scoliosis

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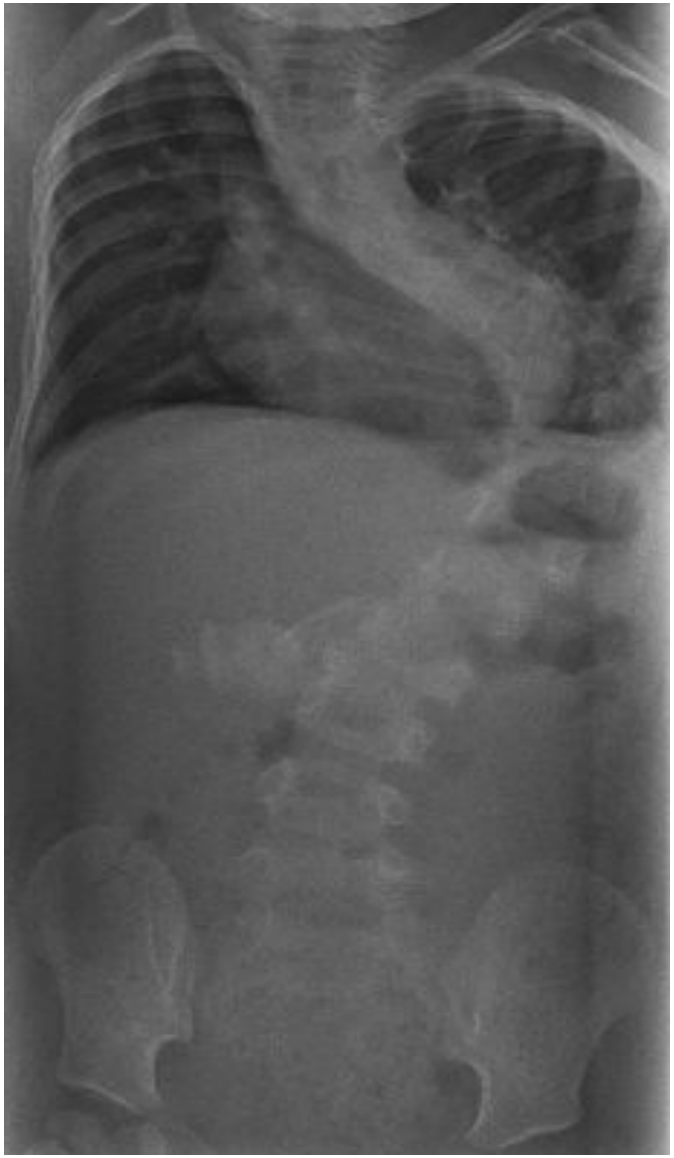
Skeletal Dysplasia Associated EOS



Skeletal dysplasias characterized by disturbances in the formation and growth of bone (Hall. Am J Med Gen 2002)

Spondyloepiphyseal dysplasia, osteogenesis imperfecta,diastrophic dysplasia have relatively high incidence of EOS (Sato et al. Bone 2016; Remes et al. Spine 2001)

EOS in these patients can be rapidly progressive resulting into severe, often short & angular deformity.

Traditional treatment: Delay with cast & brace, early anteroposterior spinal fusion (Bethem JBJS 1981; Jalanko et al. Spine 2009)

Effect of growth-friendly management remains unclear, failed previously in diastrophic dysplasia (Jalanko et al. Spine 2009; Kataras et al. Spine 2013; White et al. Spine Def 2018)

Skeletal dysplasia and Idiopathic EOS cohorts

A retrospective review of prospectively collected Growing and Children's Spine Study Group database for growth-friendly management in EOS with min 2-yr FU (n = 569)

Skeletal dysplasia associated SKD EOS:
33 children aged 10 years or less, EOS (major curve ≥30°) operated using growing rods or rib-based instrumentation, with minimum 2-yr FU after last lengthening or final fusion
- 7 with OI, 6 diastrophic, 4 camptomelic, 3 spondyloepiphyseal dysplasia, 3 achondroplasia, 2 each cleidocranial and atelosteognesis type III, 1 each chondrodysplasia punctata and bent bone dysplasia; 4 unknown SKD

Matched children with idiopathic EOS from the same database:
33 age (±1 year), gender, type of index surgery, and number of lengthening matched (±2) children with idiopathic EOS.

Data Collection

Time points of interest: Preop, Index surgery, Distraction period, Pre-definitive, Final follow-up

Clinical data collected: Age at surgery, Height, Weight, Etiology of EOS, Preop Halo traction, FU time, Number of lengthenings,

Surgical data: OR time; Blood loss; Type and levels of instrumentation; Revisions (Planned, unplanned)

Complications: Wound related (Deep surgical site infection); Implant (misplacement, pull-out, rod fracture); Alignment (PJK); Neurologic (New deficit, loss of MEPs); Other

Health-related quality of life using the 24-item Early-Onset Scoliosis Questionnaire (EOSQ-24) preoperatively and at final follow-up

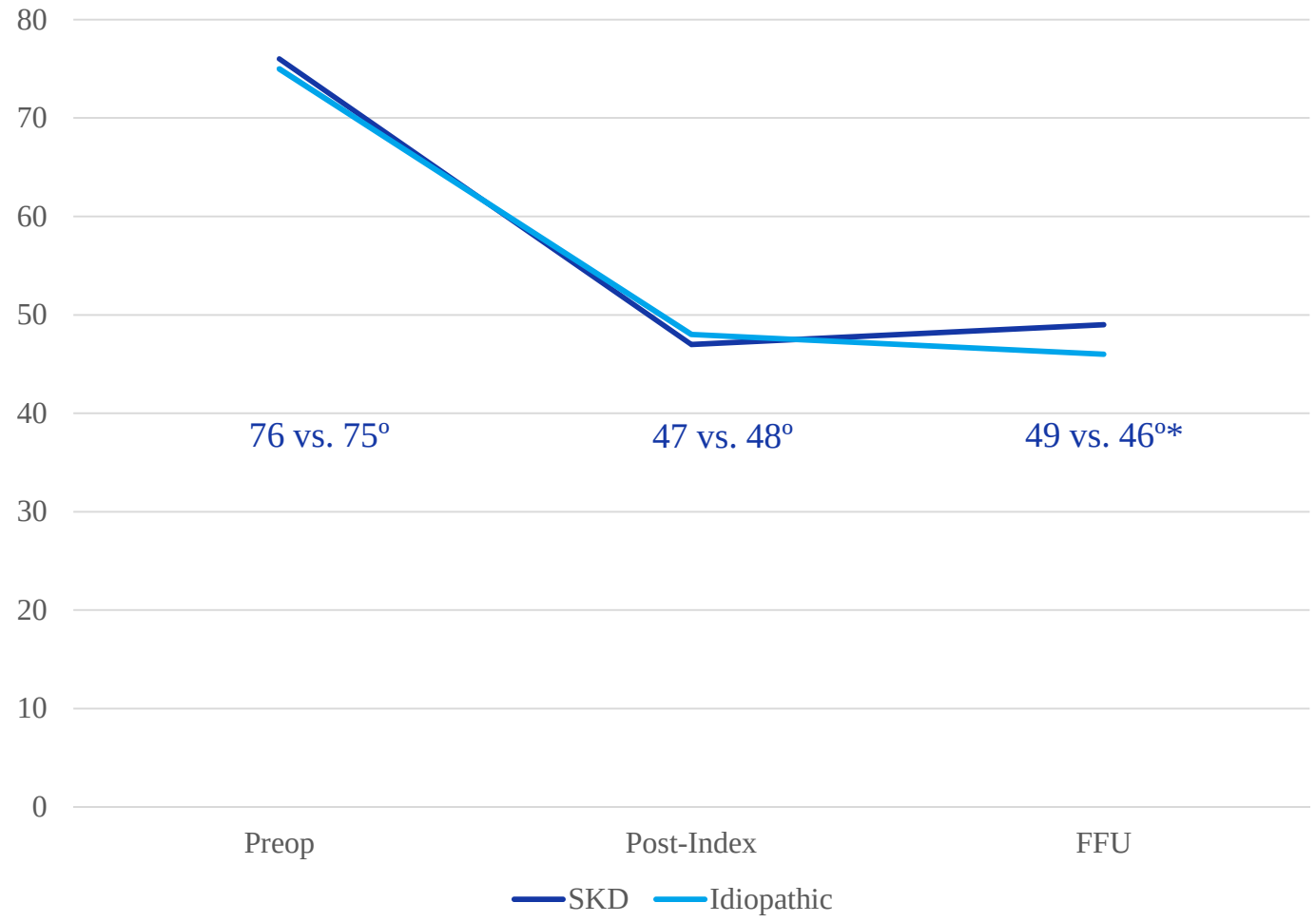
Clinical Characteristics

Characteristics	SKD (n=33)	Idiopathic (n=33)	P value
Age, yrs	5.3 (1.5-9.7)	5.4 (1.8-9.6)	0.41
Follow-up, yrs	5.6 (1.5-13)	7.1 (2.1-16)	0.046
Instrumentation Traditional GR VEPTR MCGR	 13 13 7	 13 13 7	1.0
No. of lengthenings	7.2 (3-19)	8.2 (3-22)	0.19
No. of surgeries	8.5 (2-21)	9.5 (4-23)	0.18
Final fusion	9	16	0.076

Radiographic Outcomes

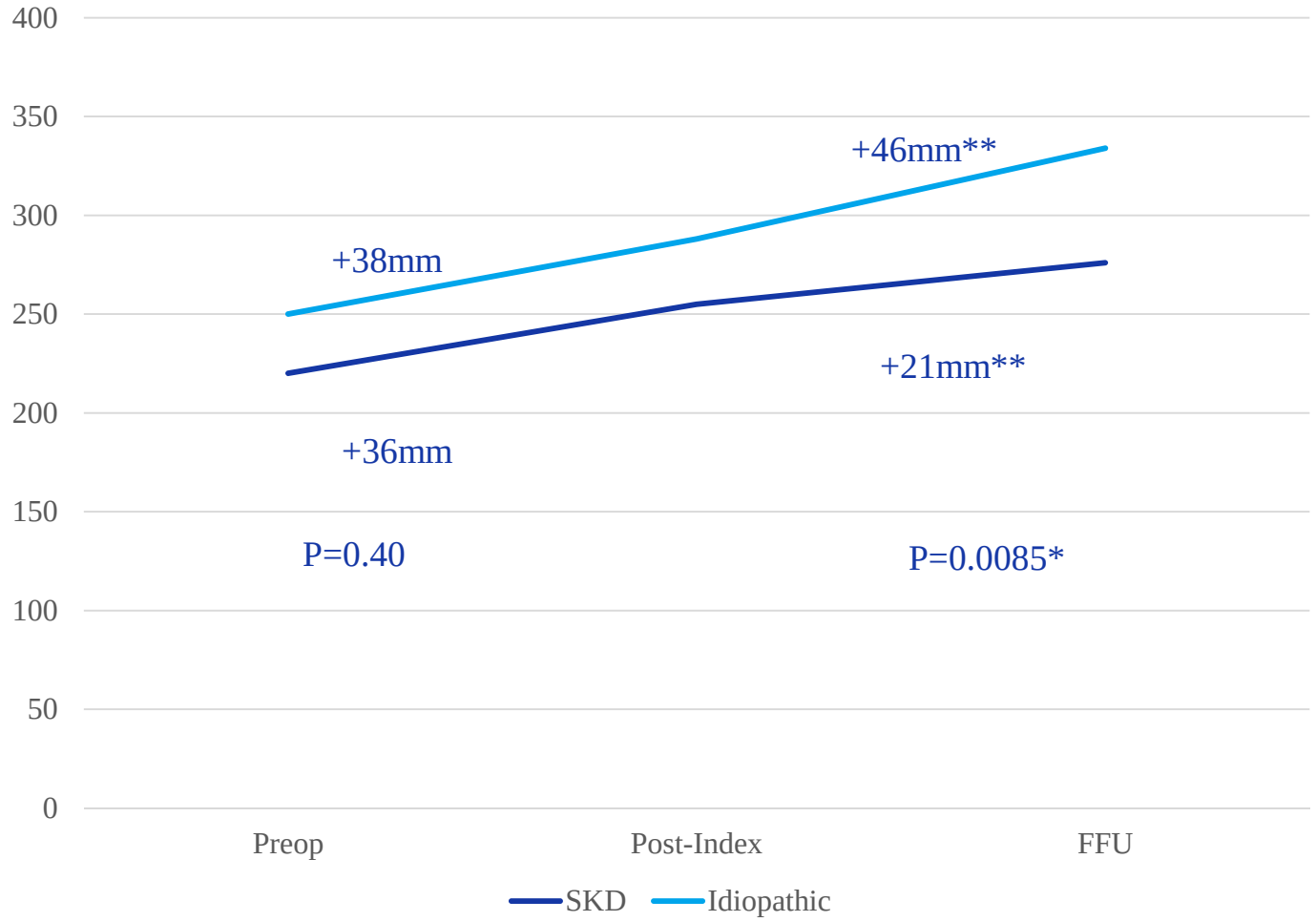
Characteristics	Skeletal dysplasia (n=33)	Idiopathic (n=33)	P value
Major curve (°)			
Preop	76 (34-115)	75 (51-113)	0.55
After Index	47 (19-82)	48 (18-95)	0.44
FFU	49 (13-113)	46 (12-112)	0.68
T1-S1 height (mm)			
Preop	220 (140-340)	250 (164-390)	0.0060
After Index	255 (160-337)	288 (173-430)	0.016
FFU	276 (182-385)	334 (205-472)	<0.001
T1-T12 height (mm)			
Preop	132 (72-207)	157 (115-242)	0.001
After Index	154 (93-211)	177 (115-257)	0.005
FFU	168 (93-229)	201 (114-282)	0.001

Major Curve: SKD vs. Idiopathic



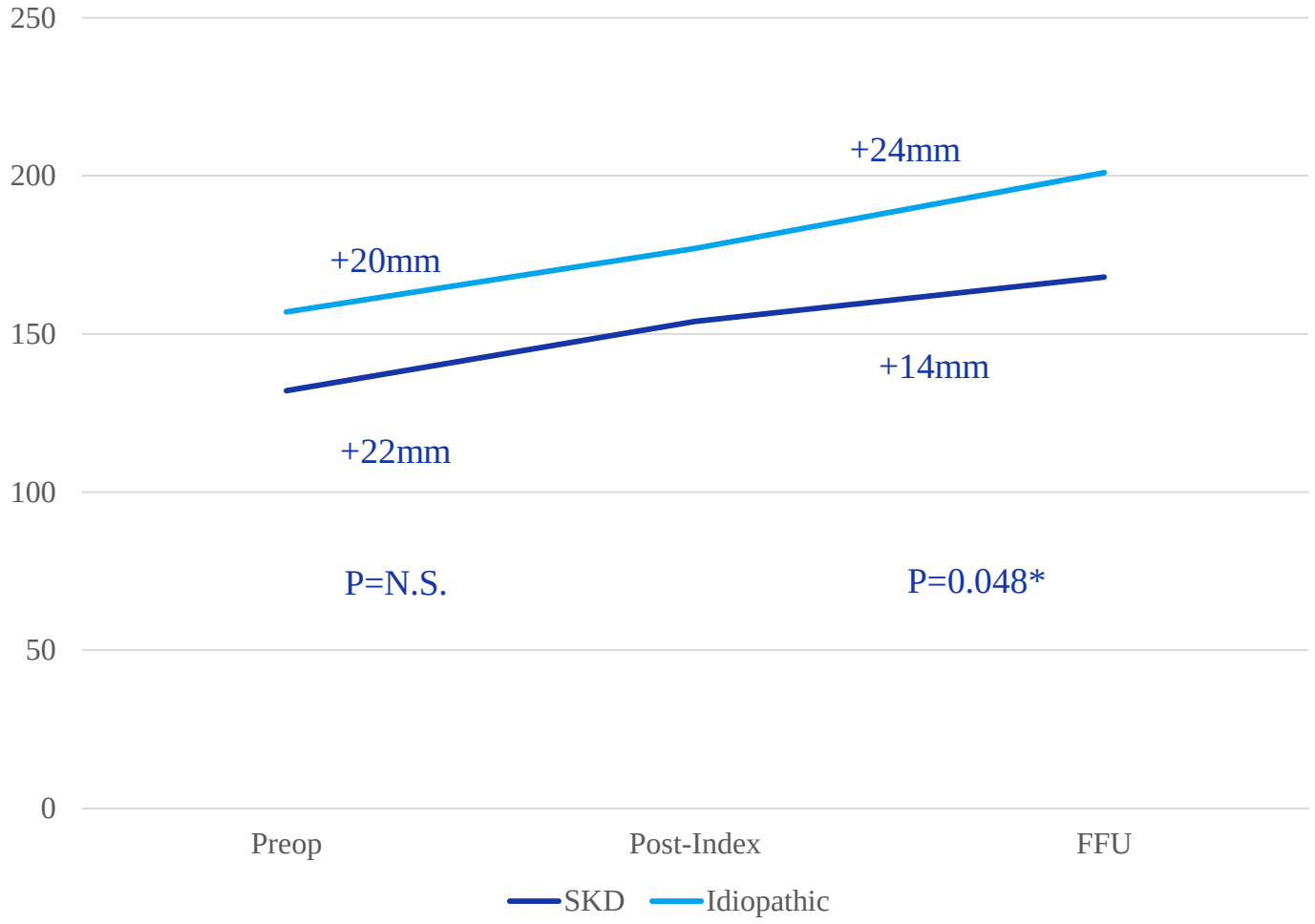
*P values not significant between groups.

T1-S1 Height: SKD vs. Idiopathic



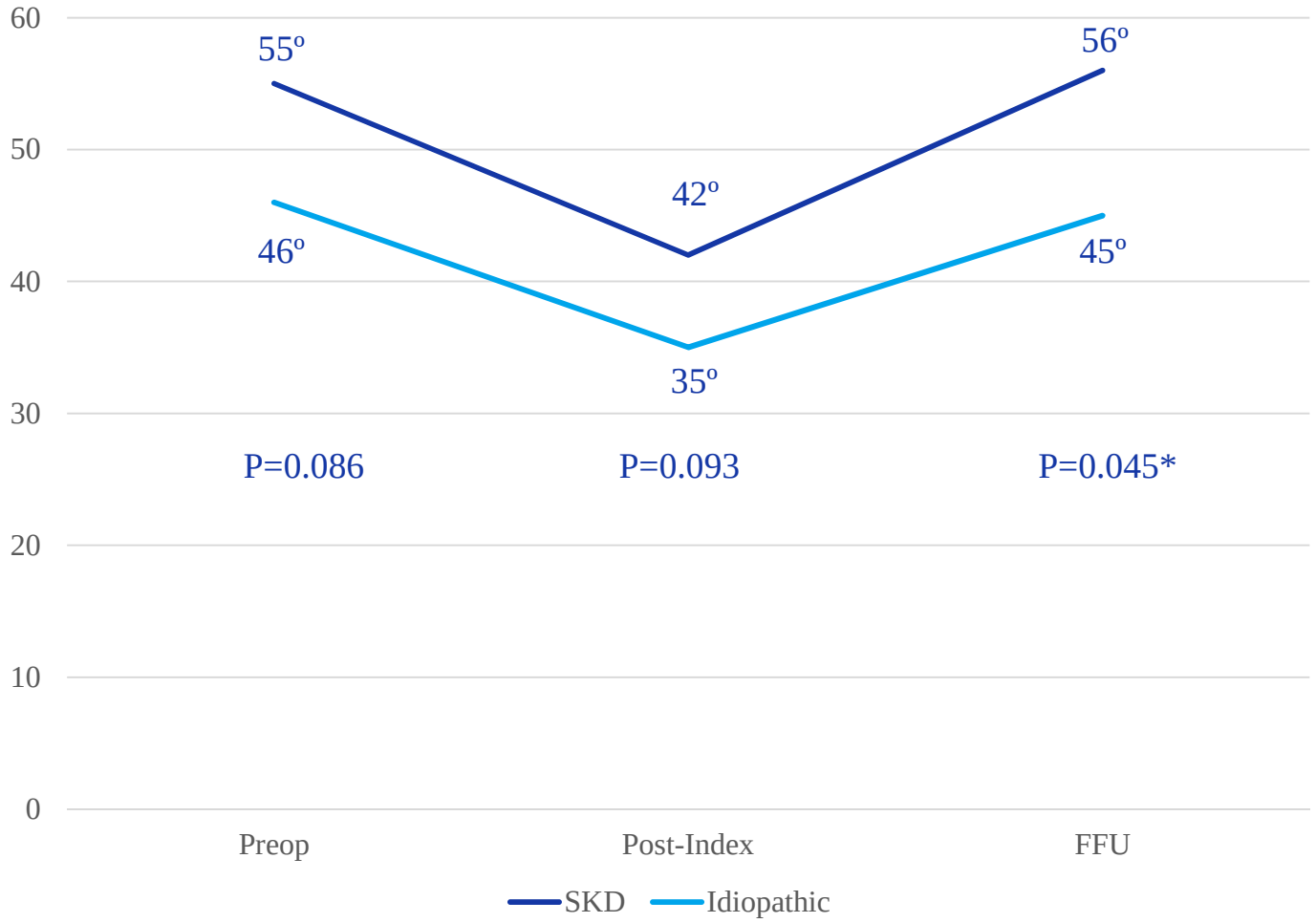
*P values between the study groups; **Annual T1-S1 growth: 3.8mm vs. 6.5mm, p=0.040

Thoracic height (T1-T12)



*P values between the groups

Thoracic kyphosis (T1-T12)



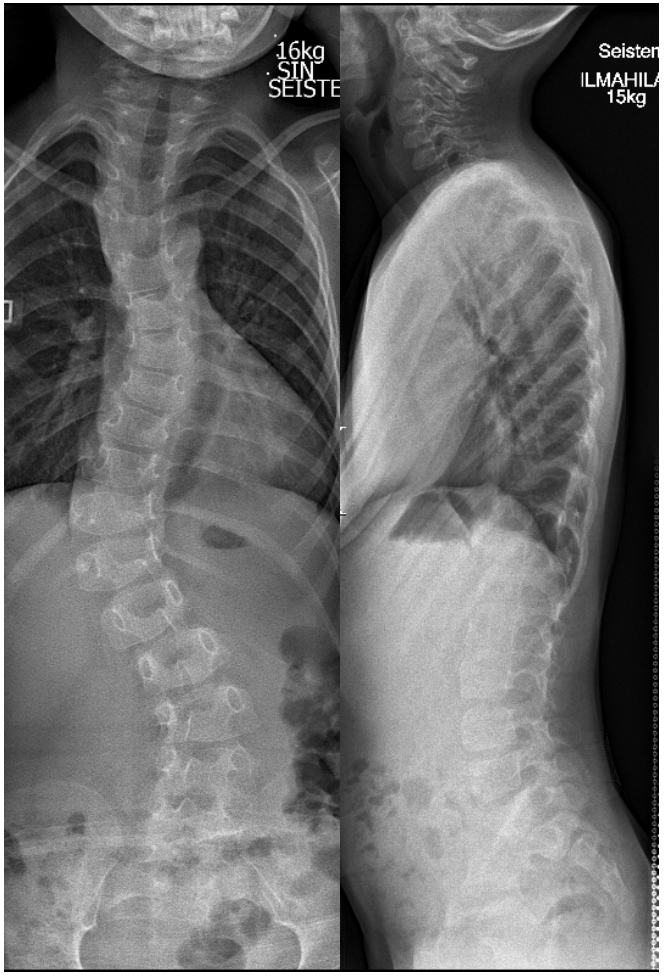
*P values between the groups

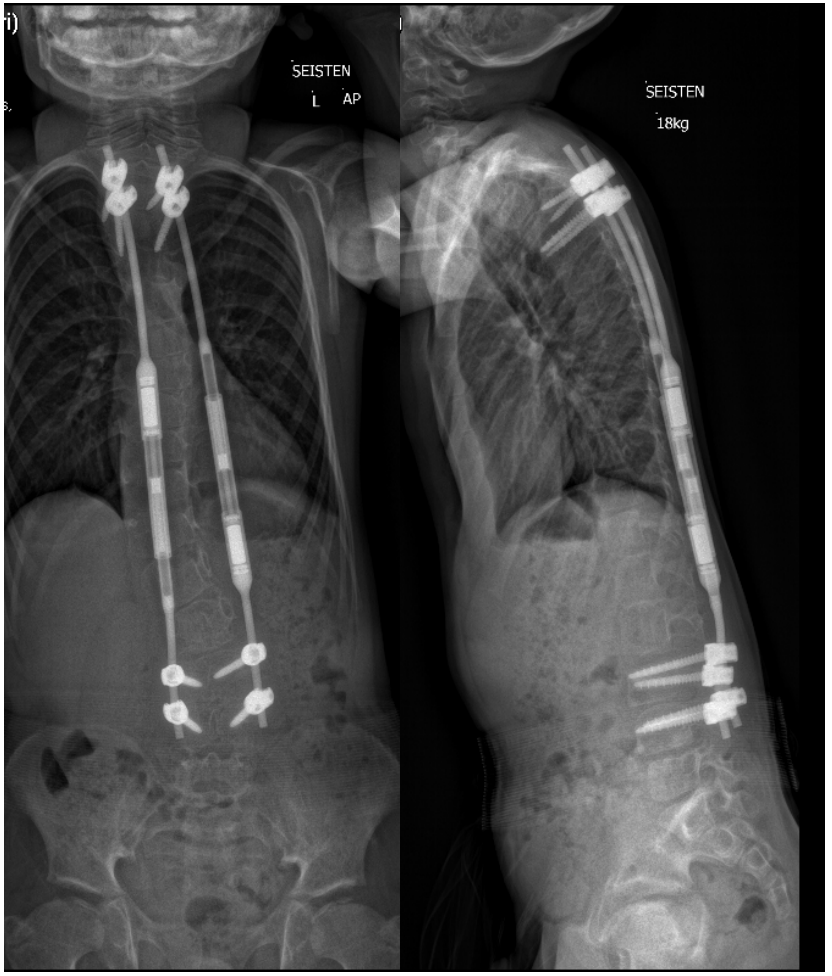
Complications

	SKD (n=33)	Idiopathic (n=40)	P value
Complication, n (%)	25 (76%)	22 (67%)	0.45
Surgery for complication, n (%)	18 (55%)	20 (61%)	0.62
Neurologic complication, n (%)*	6 (18%)	1 (3%)	0.045

*Included neurologic deficit and neuromontoring change.

Diastrophic Dysplasia & MCGR





5-year-old boy with rigid 50° early onset scoliosis. 2-yr FU. Treated at 1-yr of age with cast now progressed. MCGR after 8 outpatient lengthenings. T1-S1 growth (post index – FFU) 22 mm. Positive sagittal balance (hip extension deficit).

24-Item EOS Questionnaire

Domain	SKD (n=29/24) Preop vs. FFU	Idiop (n=31/27) Preop vs. FFU	P value (Groups)
Daily living	**46** vs. 56	**83** vs. 72	0.01/0.056
Emotion	71 vs. 68	79 vs. 70	0.13/0.60
Fatigue/energy level	61 vs. 67	73 vs. 68	0.09/0.99
Financial impact	**63** vs. **65**	**85** vs. **82**	0.01/0.018
General health	71 vs. 72	75 vs. 74	0.33/0.88
Child satisfaction	57 vs. **61**	65 vs. **75**	0.20/0.038
Parent satisfaction	**60** vs. 67	**73** vs. 68	0.044/0.75
Pain/Discomfort	69 vs. 69	75 vs. 69	0.14/0.96
Parental impact	65 vs. 64	72 vs. 74	0.37/0.14
Physical function	**61** vs. 70	**90** vs.77*	<0.01/0.29
Pulmonary function	76 vs. 85	81 vs. 80	0.83/0.23
Transfer	65 vs. 65	79 vs. 78	0.29/0.20

*P=0.010 within group

Conclusions

Growth-friendly management of early onset scoliosis in children with skeletal dysplasia provided limited additional spinal growth during the distraction phase: mean 21mm over 5.6 years (63% of spinal length gain obtained at index surgery).

Health-related quality of life is significantly reduced in children with skeletal dysplasias as compared with children having an idiopathic type of early onset scoliosis.

The benefits of growth-friendly management remains unclear in children with skeletal dysplasias as compared with the historical data on delaying with cast/brace and then early definitive spinal fusion (maximizing deformity correction and spinal length gain at index surgery).

■ CHILDREN'S ORTHOPAEDICS

Results of growth-friendly management of early-onset scoliosis in children with and without skeletal dysplasias

A MATCHED COMPARISON

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Aims

The aim of this study was to compare the surgical and quality-of-life outcomes of children with skeletal dysplasia to those in children with idiopathic early-onset scoliosis (EOS) undergoing growth-friendly management.

Patients and Methods

A retrospective review of two prospective multicentre EOS databases identified 33 children with skeletal dysplasia and EOS (major curve ≥ 30°) who were treated with growth-friendly instrumentation at younger than ten years of age, had a minimum two years of postoperative follow-up, and had undergone three or more lengthening procedures. From the same registries, 33 matched controls with idiopathic EOS were identified. A total of 20

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