

Congenital Scoliosis of the Pediatric Cervical Spine

Brandon Toll, Amer Samdani, Joshua Pahys
and Steven Hwang
Shriners Hospitals for Children-Philadelphia

ICEOS Portugal 2018

Introduction

- Literature on congenital scoliosis predominantly focuses on thoracic and thoracolumbar curves
- Papers to date include a handful of 1-2 patients and one series of 18 patients of cervicothoracic hemi vertebra (Chen et al, Spine 2018)
- Purpose: to better define surgical outcomes of patients having undergone spinal fusions for cervical scoliosis with osseous anomalies

Methods

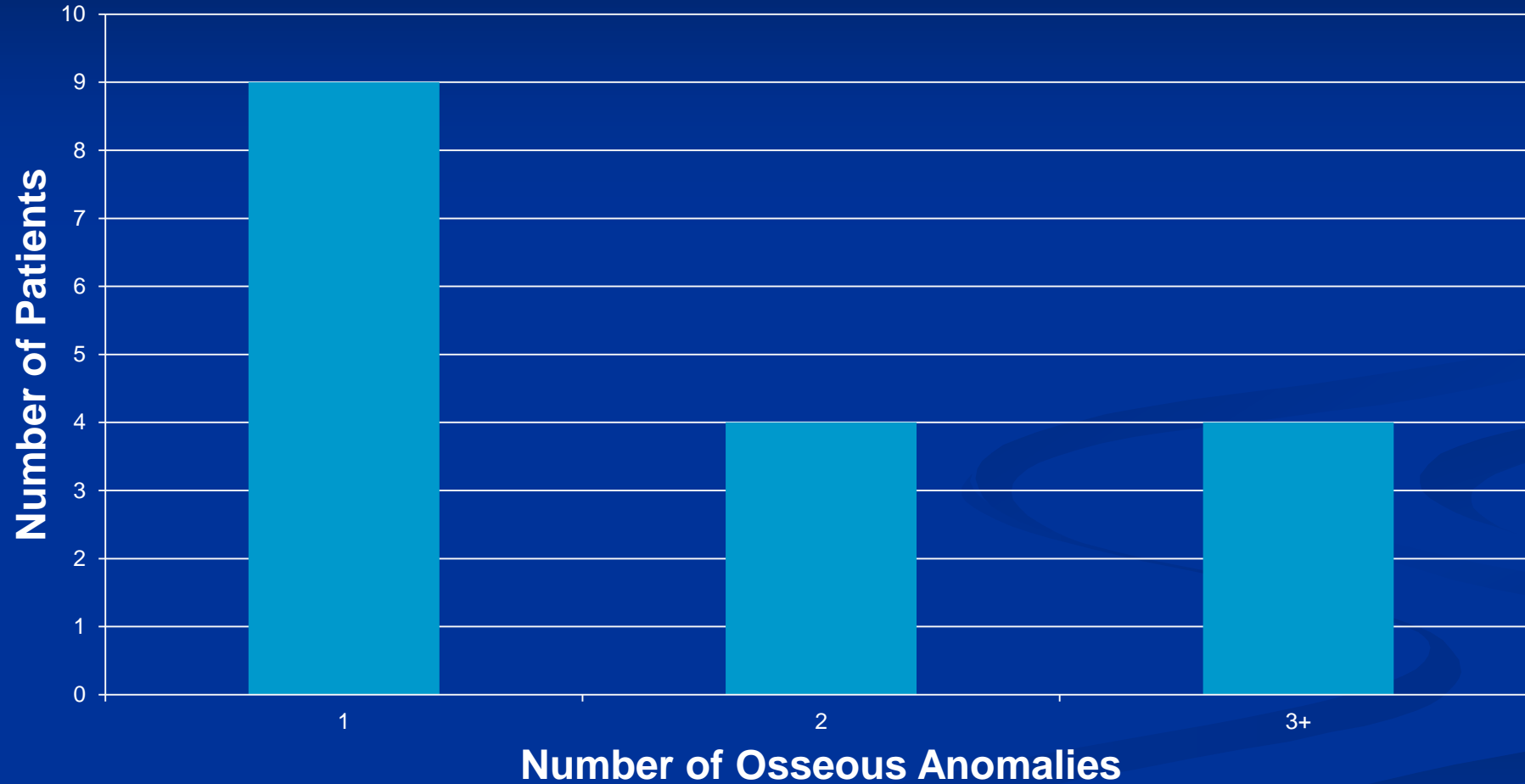
- Retrospective chart and radiographic review of 17 patients was conducted (2009-2016) from a single center (Shriners Hospitals for Children-Philadelphia)
- All patients treated surgically for cervical congenital scoliosis with adequate clinical and radiographic data available for analysis
- Analysis was performed with t-tests (paired and independent samples) and binary correlation

Cohort

- 9 boys/8 girls
- The most prevalent presentations were torticollis (29%), and Klippel-Feil syndrome (17.6%)
- Mean age at surgery was 7.07 ± 3.38 years
- Average follow-up of 3.06 ± 1.78 years
- 4 cognitively delayed (23.5%) and 3 non-ambulatory (17.6%)

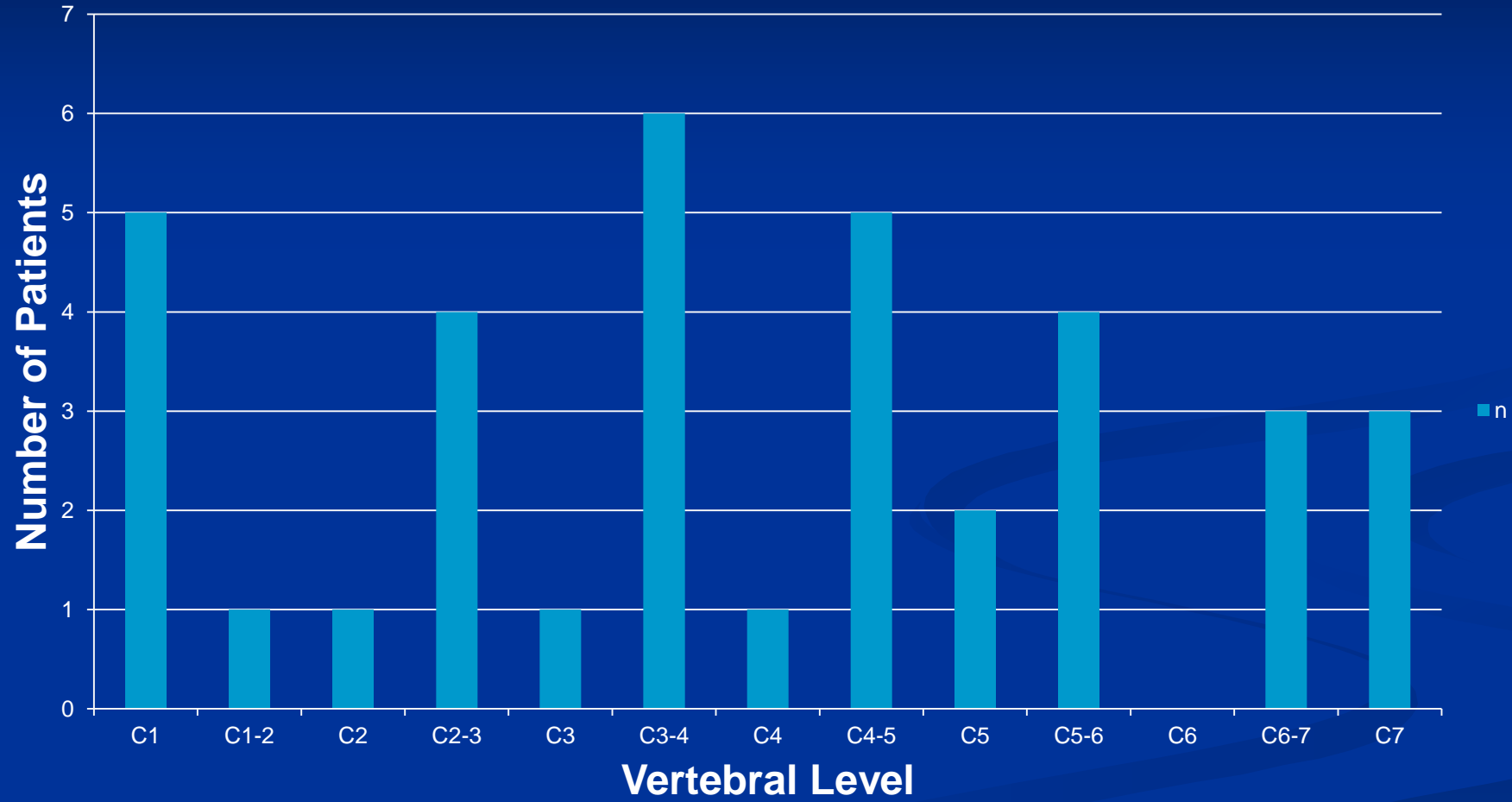
Concurrent Findings	
	Total Number (%)
VACTERL Syndrome	2 (11.8)
Tethered Cord	2 (11.8)
Sprengel's Deformity	2 (11.8)
Torticollis	5 (29.4)
Klippel-Feil Syndrome	3 (17.6)
Pierre Robin Syndrome	1 (5.9)
Brown Syndrome	1 (5.9)
Bruck Syndrome	1 (5.9)
Poland's Syndrome	1 (5.9)
Goldenhar Syndrome	1 (5.9)
Larsen Syndrome	2 (11.8)
Cognitively Delayed	4 (23.5)
Non-ambulatory	3 (17.6)
Total	17 (100.0)

Number of Osseous Anomalies per Patient



Results

Osseous Deformity by Vertebral Level



Summary of Clinical and Radiographic Metrics

	Pre-op Mean (SD)	Post-op Mean (SD)	FU Mean (SD)
Weight (kg)	23.39 (13.83)	23.11 (13.33)	25.95 (12.50)
Height (cm)	108.93 (21.58)	107.48 (25.28)	120.03 (17.49)
Coronal Cobb (°)	35.97 (19.75)	23.59 (14.15)	21.56 (12.39)
Cervical Lordosis (°)	22.81 (13.66)	21.17 (12.94)	20.76 (9.30)
Sagittal Balance (mm)	28.07 (38.19)	41.90 (36.73)	24.96 (41.97)
Coronal Balance (mm)	26.32 (11.31)	29.56 (17.52)	31.98 (21.85)
Occiput-C2 Angle (°)	11.52 (6.58)	9.79 (9.50)	9.74 (5.24)
T1 Slope (°)	20.73 (11.62)	18.03 (8.55)	14.63 (6.18)

Results

- Average total correction of $12.90 \pm 27.52^\circ$ in coronal Cobb angle
- Significant both at initial correction ($p = 0.020$) and last follow-up ($p = 0.004$)
- 3 complications: pressure ulcer (halo vest), vertebral artery injury, broken rod

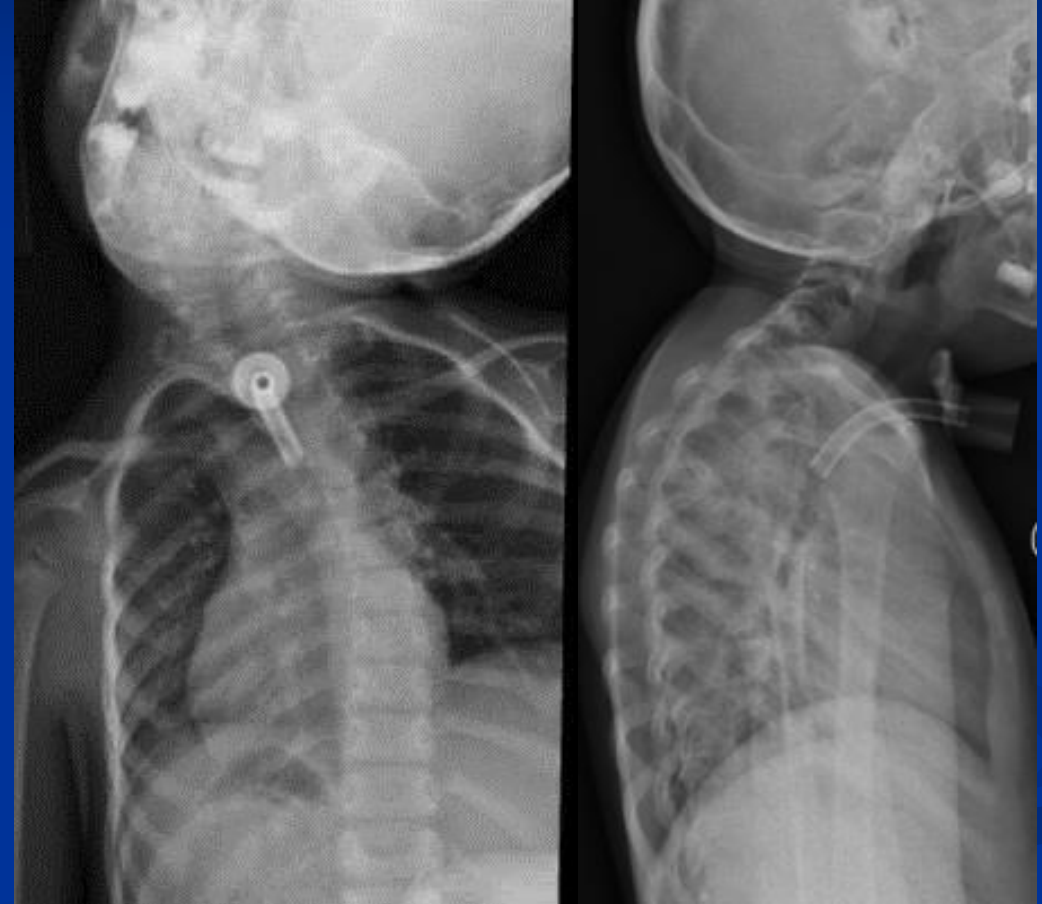
Correction vs. Outcome

- Patients stratified by degree of correction achieved in either coronal or sagittal planes
 - $\geq 50\%$ correction coronal vs. $< 50\%$
 - $\geq 50\%$ correction sagittal vs. $< 50\%$

Operative Characteristics of Overall Cohort			
	Total Number (%)	Total Number $\geq 50\%$ Coronal Correction (%)	Total Number $\geq 50\%$ Sagittal Correction (%)
Follow-up Time (yrs)	3.06 \pm 1.78	2.13 \pm 1.15	4.00 \pm 1.62
Age at Surgery (yrs)	7.07 \pm 3.38	5.8 \pm 3.78	6.94 \pm 3.19
Total	17	9	9

Patient LS

- 3 year old with Pierre Robin, multiple congenital anomalies
 - C1 hypoplasia
 - Severe torticollis
 - Surgery
 - Halo and SCM release
 - Occiput - C3 fusion



Pre-op CT scan at age 22 months

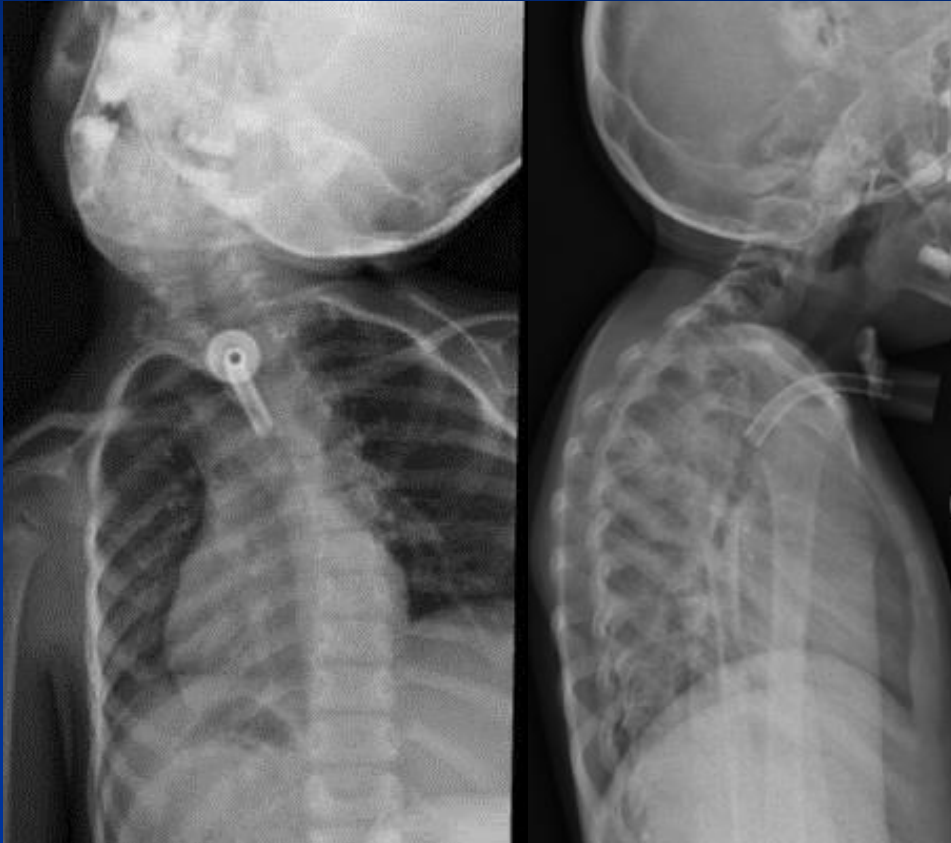


C1 left lateral mass

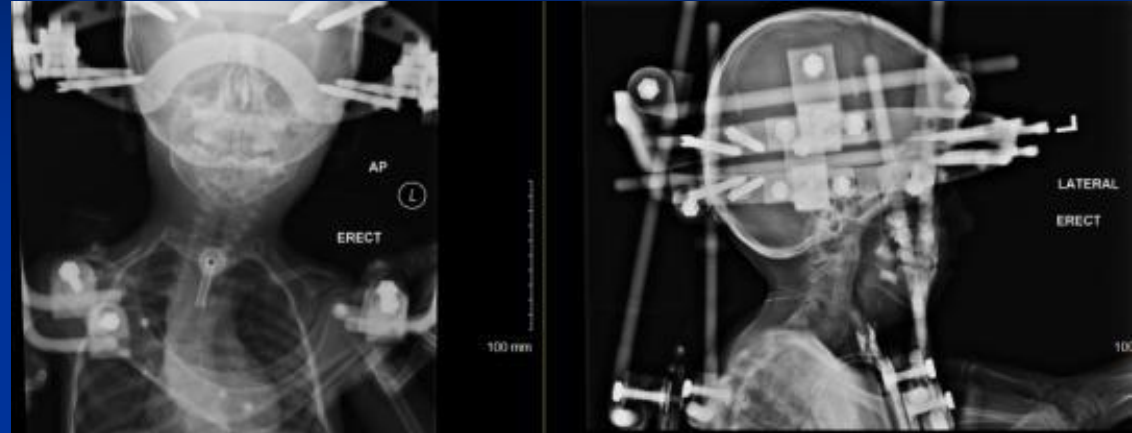
C1 right lateral mass

Plagiocephaly

Surgery



Pre-op AP, lateral



Halo – SCM Release



Post-op AP, lateral

Patient LS Post-op



Conclusion

- Broad group with variable goals with respect to correction
 - Stabilization versus correction
- Early surgery, prior to development of larger curves, likely provides better outcomes
- Halo vest immobilization often utilized after surgery

References

- Chen et al. Posterior-only hemivertebra resection for congenital cervicothoracic scoliosis: correcting neck tilt and balancing the shoulders. *Spine* 43(6):394-401, 2018
- Gruber et al. The prevalence of Klippel-Feil syndrome: A computed tomography-based analysis of 2917 patients. *Spine Deform* 6(4):448-453, 2018
- McMaster MJ, Ohtsuka K. The natural history of congenital scoliosis. A study of two hundred and fifty-one patients. *JBJS* 64(8):1128-47, 1982