



Real Congenital Scoliosis (Rib Fusions/Jumbled Spine)

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Disclosures

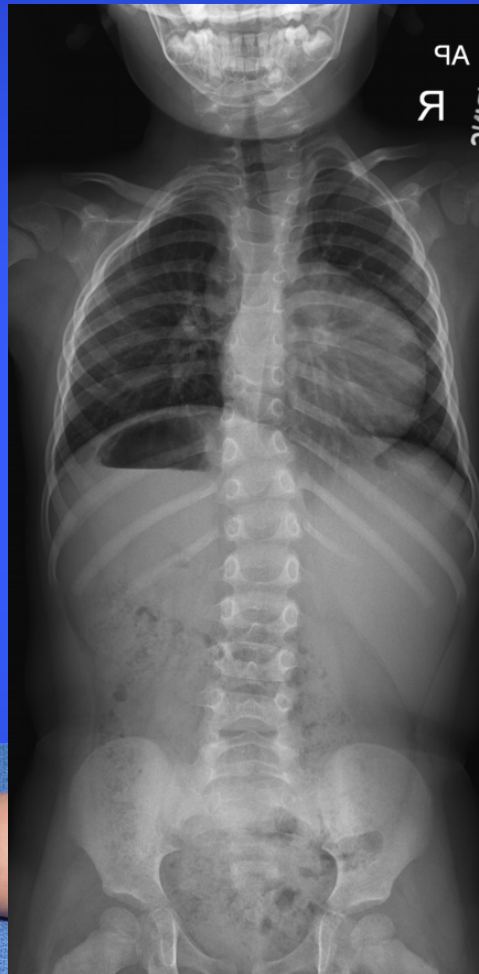
- Research funding SRS, OREF
- Honorarium/consulting funds directed to research from Medtronic, Zimmer

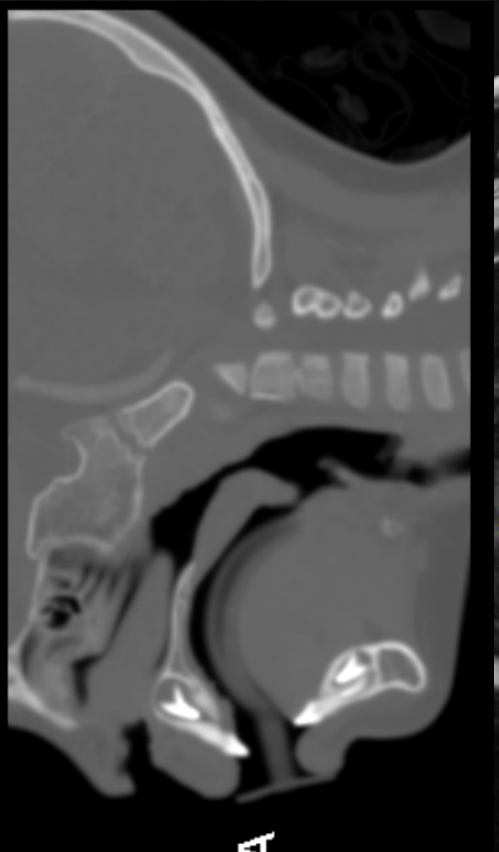
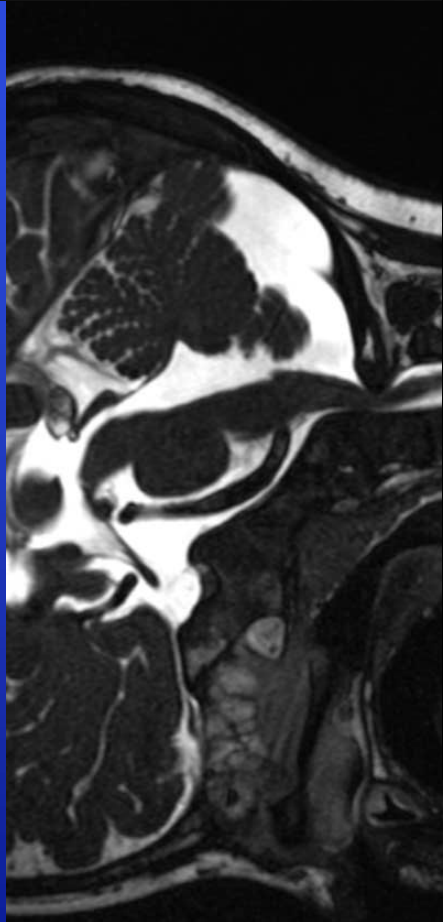
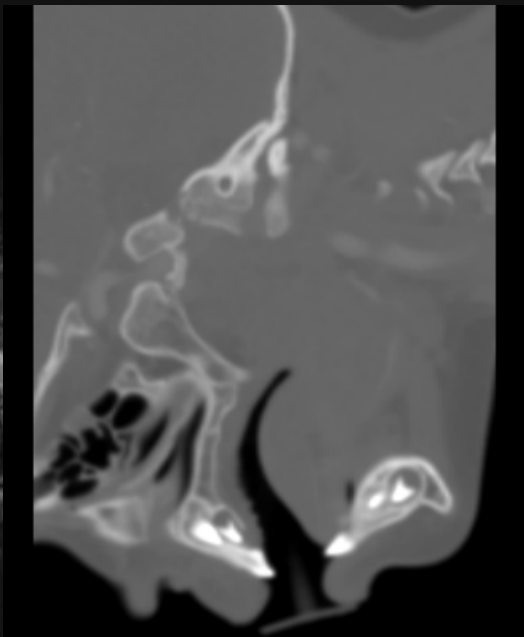
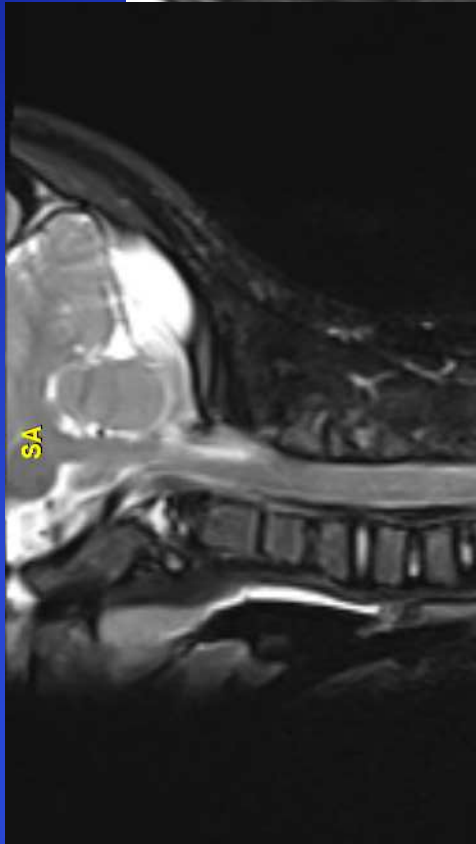
Congenital Scoliosis

- Associated Conditions
 - Renal
 - Spinal cord
 - Cardiac



- 3 yo M, evaluation for congenital scoliosis
- Family complains of gait abnormality
 - Left thumb hypoplasia
 - Imperforate anus
 - Dextrocardia
 - Fused kidneys





THE EFFECT OF OPENING WEDGE THORACOSTOMY ON THORACIC INSUFFICIENCY SYNDROME ASSOCIATED WITH FUSED RIBS AND CONGENITAL SCOLIOSIS

BY ROBERT M. CAMPBELL JR., MD, MELVIN D. SMITH, MD, THOMAS C. MAYES, MD,
JOHN A. MANGOS, MD, DONNA B. WILLEY-COURAND, MD, NUSRET KOSE, MD, RICARDO F. PINERO, MD,
MARDEN E. ALDER, DDS, HOA L. DUONG, MD, AND JENNIFER L. SURBER, BS

Investigation performed at the Thoracic Institute, Christus Santa Rosa Children's Hospital, San Antonio, Texas

Congenital Scoliosis with Rib Fusions

J Bone Joint Surg Am. 2014 Nov 5;96(21):e181. doi: 10.2106/JBJS.M.00185.

Management of thoracic insufficiency syndrome in patients with Jarcho-Levin syndrome using VEPTRs (vertical expandable prosthetic titanium ribs).

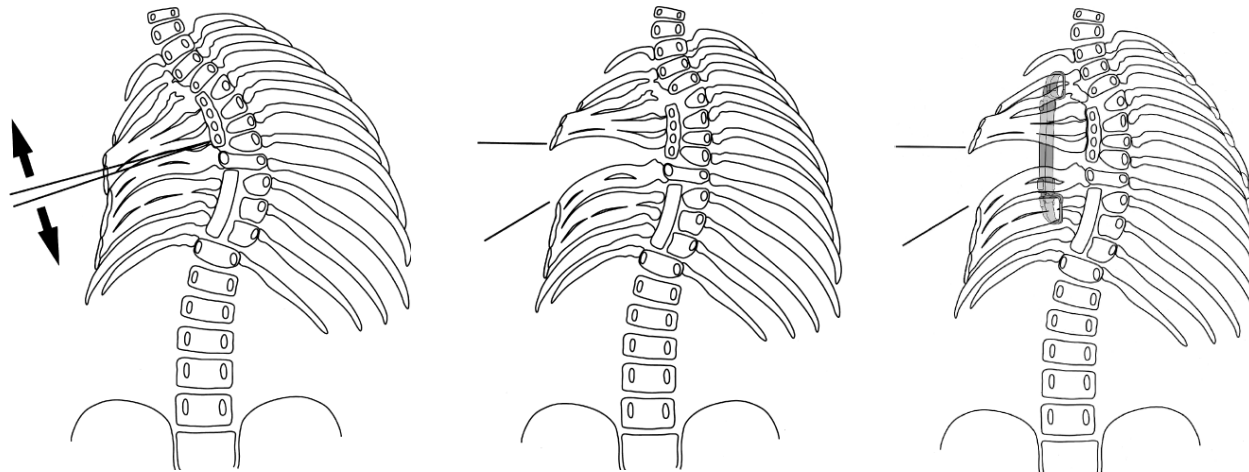
Karlin JG¹, Roth MK², Patil V¹, Cordell D¹, Trevino H², Simmons J², Campbell RM³, Joshi AP².

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THE EFFECT OF THORACOSTOMY ON THORACIC INSUFFICIENCY
SYNDROME WITH FUSED RIBS AND SCOLIOSIS



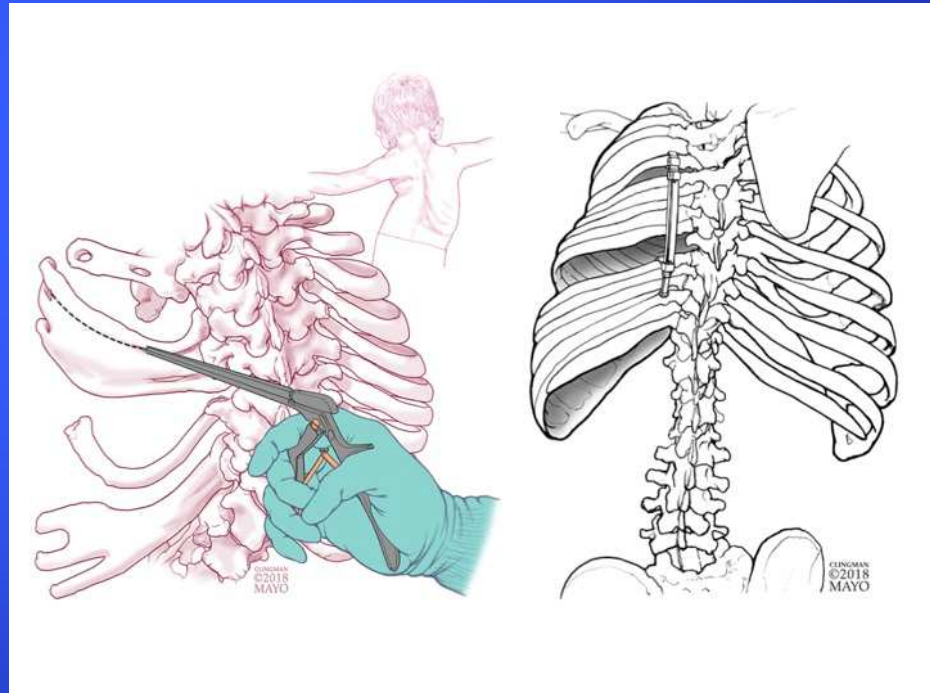
The Effect of Expansion Thoracostomy on Spine Growth in Patients with Spinal Deformity and Fused Ribs Treated with Rib-Based Growing Constructs

Fady J. Baky, MD^a, A. Noelle Larson, MD^{a,*}, Tricia St. Hilaire, MPH^b, Jeff Pawelek, MD^c, David L. Skaggs, MD^d, John B. Emans, MD^e, Joshua M. Pahys, MD^f, Children's Spine Study Group^b, Growing Spine Study Group^c

Spine Deform. 2019 Sep;7(5):836-841. doi: 10.1016/j.jspd.2019.01.004.



Does expansion thoracostomy with fused ribs result in improved outcomes?

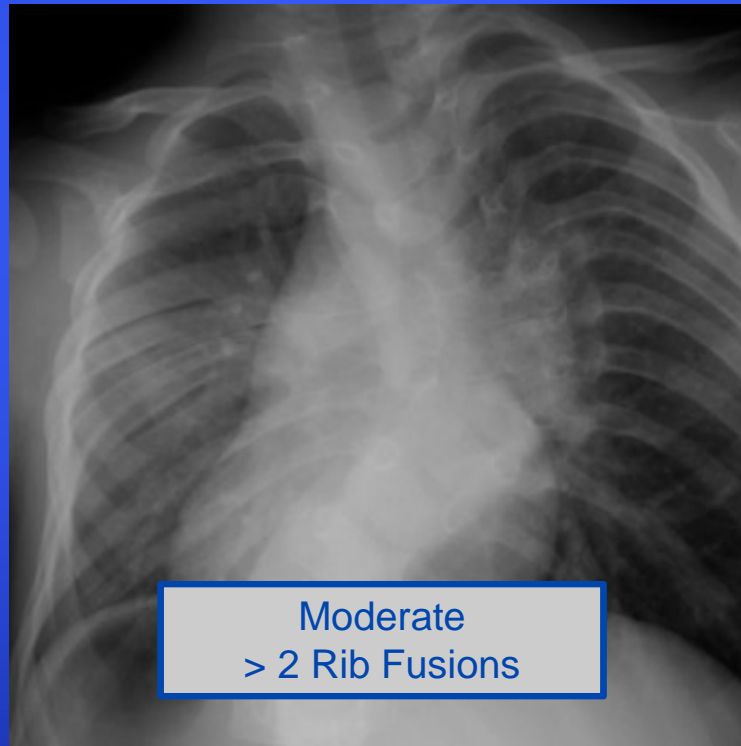


Rib Fusion Severity

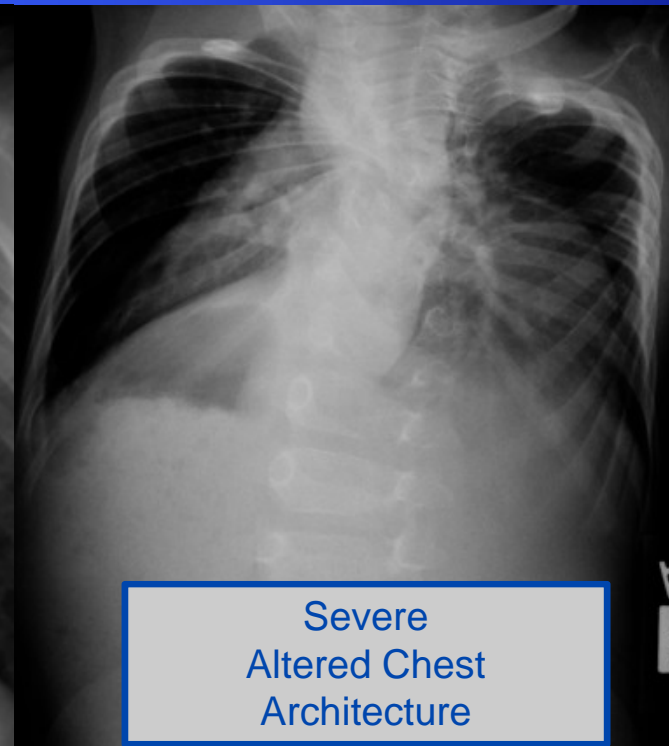
- Rated rib fusions as: mild/moderate/severe



Mild
1-2 Rib Fusions



Moderate
> 2 Rib Fusions



Severe
Altered Chest
Architecture

	Thoracostomy (N=103)	No Thoracostomy (N=48)	P-value
Follow-Up (Years)	6.8	5.8	0.08
# of Lengthenings	8.3	6.6	0.017
Total # of Surgeries	11.5	9.6	0.031
# of Revision Surgeries	2.3	2	0.48
Underwent Final Fusion	19/103	10/48	0.83
Change in T1-S1 height at initial procedure (cm)	2.01	1.24	0.20
Change in T1-S1 height with subsequent distractions (cm)	6.7	4.7	0.05
Change in T1-S1 height at Final Fusion (cm)	2.2	1.6	0.57
Total Change in T1-S1 (cm)	7.2	4.8	0.0043

What Proximal Anchors are Best?

- To determine outcomes (thoracic height and Cobb angle) in patients with congenital fused ribs treated with proximal spine anchors (spine-based growing devices) compared to constructs with proximal rib anchors (rib-based devices).

[Spine Deform.](#) 2019 Jan;7(1):152-157. doi: 10.1016/j.jspsd.2018.05.011.

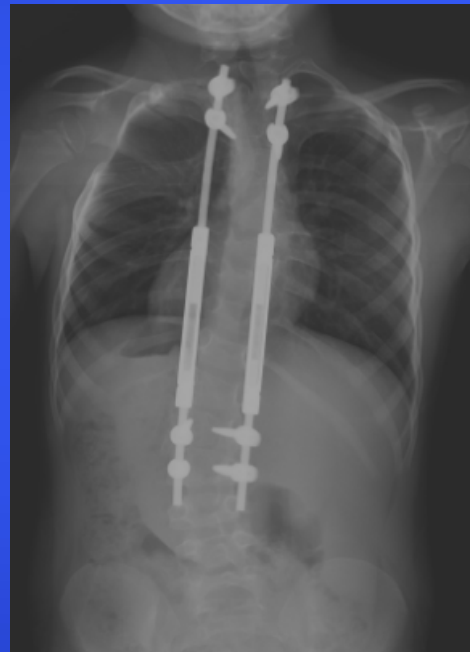
Spine Deformity With Fused Ribs Treated With Proximal Rib- Versus Spine-Based Growing Constructs.

[Larson AN](#)¹, [Baky FJ](#)², [St Hilaire T](#)³, [Pawelek J](#)⁴, [Skaggs DL](#)⁵, [Emans JB](#)⁶, [Pahys JM](#)⁷; [Children's Spine Study Group](#)³; [Growing Spine Study Group](#)⁴.

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- 2 Mayo Clinic, 200 First St SW, Rochester, MN 55905, USA.
- 3 Children's Spine Foundation, P.O. Box 397, Valley Forge, PA 19481, USA.
- 4 Growing Spine Foundation, 555 East Wells Street, Suite 1100, Milwaukee, WI 53202, USA.
- 5 Children's Hospital Los Angeles, 4650 Sunset Blvd, Los Angeles, CA 90027, USA.
- 6 Children's Hospital, Harvard Medical School, 300 Longwood Ave, Boston, MA 02115, USA.
- 7 Shriners Hospital for Children, 3551 N Broad St, Philadelphia, PA 19140, USA.

Proximal fixation assessed as spine-based or rib-based
(excluded those with both types of anchors)



Spine-based
Proximal Anchors



Rib-based
Proximal Anchors

Study Design

- Retrospective review of prospectively collected data from multicenter databases (GSSG & CSSG).
- Minimum 2 year follow-up
- Early onset scoliosis and rib fusions
- 176 patients identified
 - 16 proximal spine anchors
 - 160 proximal rib anchors
 - 154 VEPTR, 6 other
 - 106 had thoracoplasty at implantation
 - 90 had rib-to-rib construct

Results: Severity of Rib Fusions

Rib Deformity	Spine-Based Devices (N=16)	Rib-Based Devices (N=160)
Mild (%)	6 (38%)	60 (38%)
Moderate (%)	6 (38%)	66 (41%)
Severe (%)	2 (13%)	26 (16%)
Indeterminate	2 (13%)	8 (5%)

Results: # of Surgeries

	Spine-Based Devices (N=16)	Rib-Based Devices (N=160)	P-value
# Lengthening Surgeries	6.3 (1-14)	7.9 (1-21)	0.12
# All Surgeries	8.0 (2-18)	11.2 (2-30)	0.007
# Revision Surgeries	1.6 (0-7)	2.3 (0-12)	0.17

Results: Spinal Height

	Spine-Based Devices (N=16)	Rib-Based Devices (N=160)	P-value
Total Change in T1-T12	6.0 (-4.5-22.4)	3.4 (-3.01-11.7)	0.26
Total Change in T1-S1	9.1 (3.0-13.1)	6.3 (-4.1-18.2)	0.06
Distraction Change T1-T12	5.7 (-4.5-22.4)	3.3 (-3.3-11.7)	0.35
Distraction Change T1-S1	8.1 (-1.3-7)	5.9 (-5-10.3)	0.04
Length/Distraction	0.29	0.3	> 0.05
Final Fusion Change* T1T12	0.75 (-0.7-2.4)	1.4 (-2.1-6.2)	0.36
Final Fusion Change T1S1*	0.58 (-2.2-2.7)	2.1 (-5.5-8.2)	0.16

**Only 46 patients (40 rib-construct and 6 spine-construct) had final fusion.

Results: Coronal and Sagittal Plane

	Spine-Based Devices (N=16)	Rib-Based Devices (N=160)	P-value
Post-Treatment Cobb	36.7 (0-62)	57.8 (11-117)	<0.001
Post-Treatment Kyphosis	34.7 (0-72)	52.4 (0-108)	0.003
Δ in Cobb Angle	24.4 (-18-66)	11.3 (-31-88)	0.049
Δ in Kyphosis	20.3 (-10-62)	-7.3 (-63 - 74)	0.002

Discussion

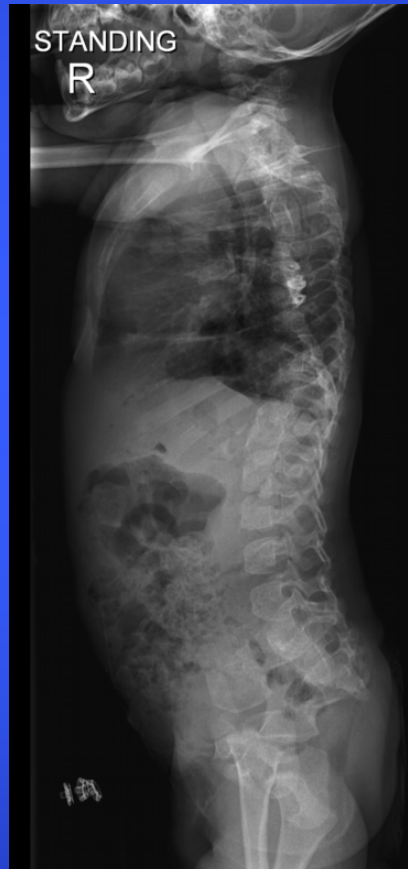
- No difference in T1-T12/T1-S1 growth achieved
- More surgeries in the rib-based group
- Increased thoracic Cobb angle and kyphosis at latest follow-up in rib-based group
 - Previously rib-based devices have been found to be associated with increasing kyphosis
 - Murphy RF et al. *JPO*. 2016 Jun; 36(4):329-35.
 - Waldhausen JH et al. *JPO*. 2016. 51:1747-1750.

Case: 6 month old



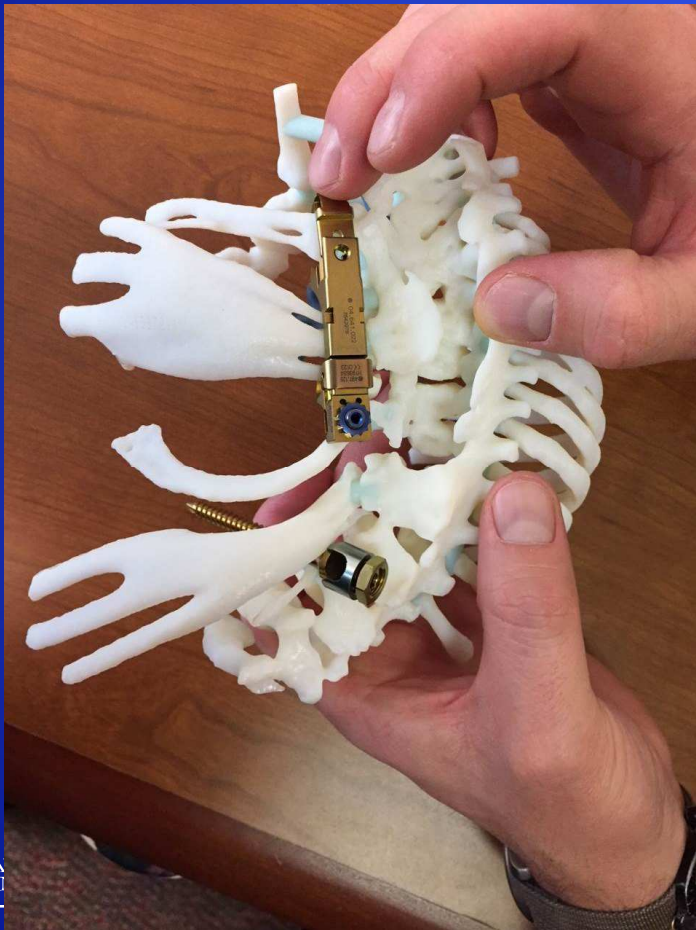
- Thoracic meningocele
- Solitary right kidney
- Rectal urethral fistula status post colon diversion takedown
- Left upper extremity abnormalities

Early Onset Scoliosis – Congenital 3 yo

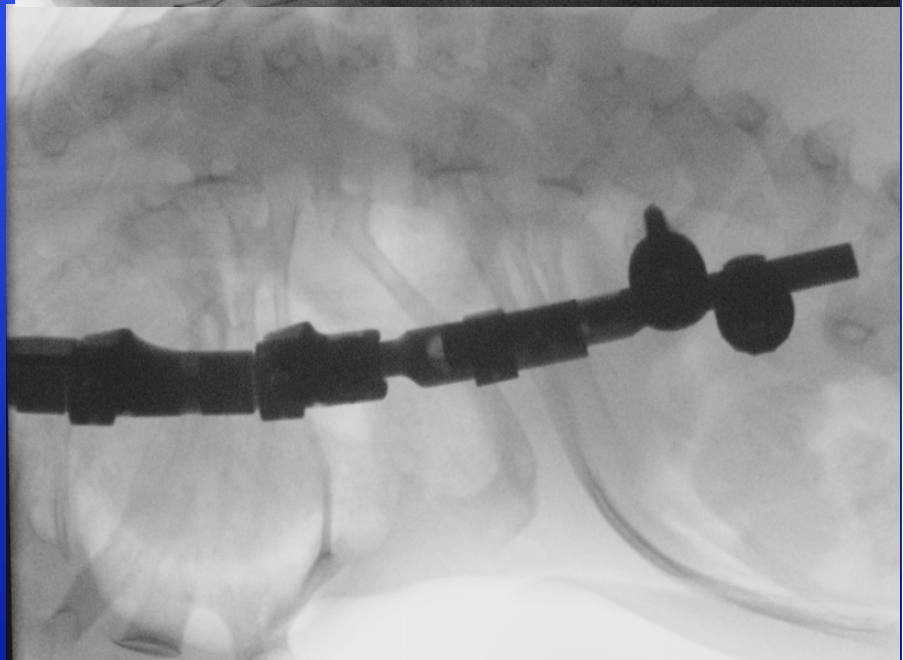
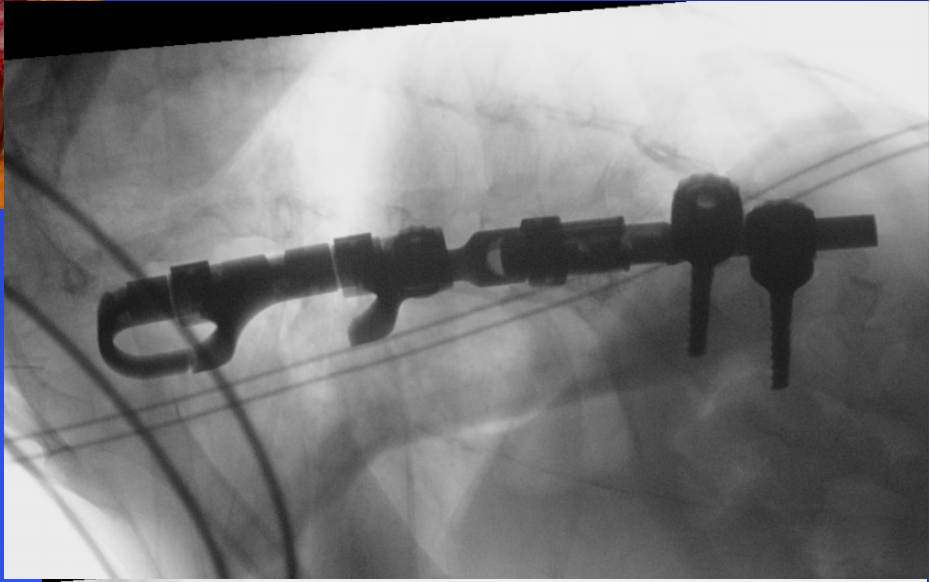
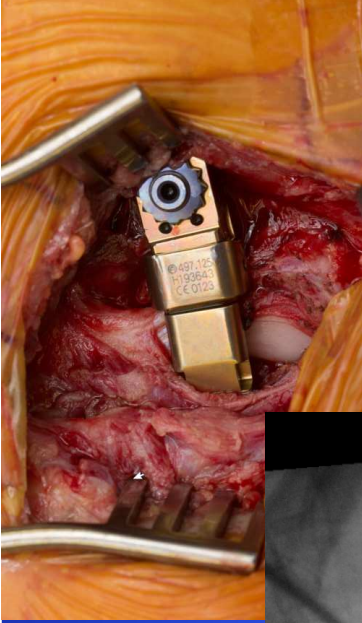


- 10.6 kg
- 83.7 cm
- T1-T12
8 cm

Congenital Deformities/Rib Fusions



- Three rib blocks
 - Planned two rib cradles
 - Two screws distally
- No thoracostomy (ribs somewhat mobile)
- Lack of soft tissue coverage



POD2





- Lower lesion
- Poor intrinsic function

Neuromonitoring During Index Procedure

- 3 lower extremity
- 1 upper lead (not ulnar n.)



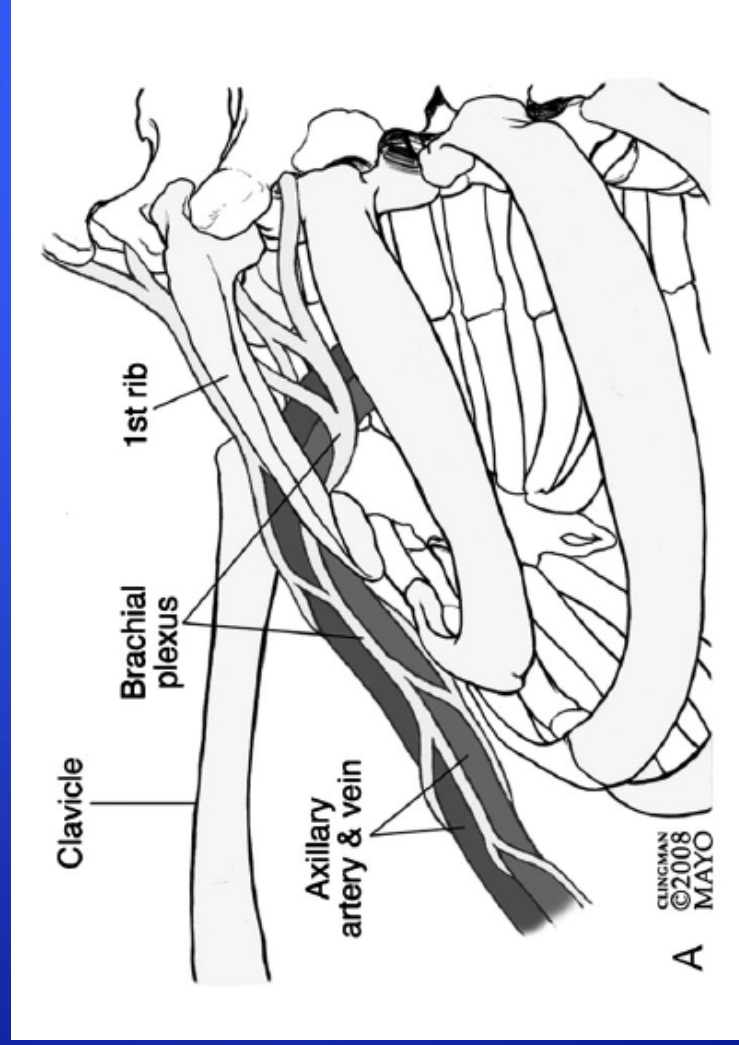
J.Pediatr.Orthop., 2009 Jan-Feb;29(1):31-4. doi: 10.1097/BPO.0b013e318192198a.

latrogenic thoracic outlet syndrome secondary to vertical expandable prosthetic titanium rib expansion thoracoplasty: pathogenesis and strategies for prevention/treatment.

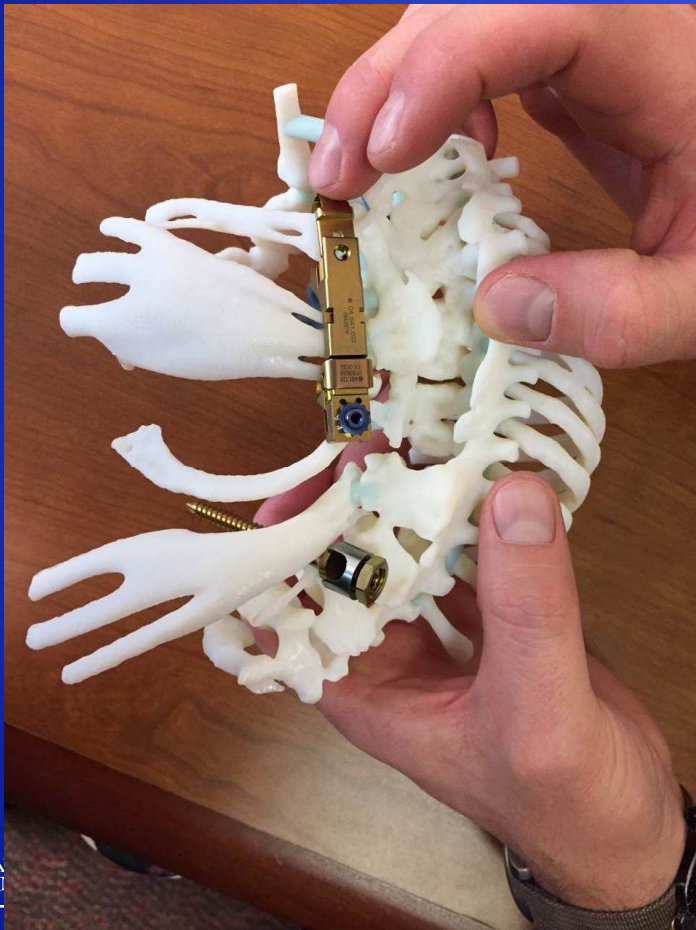
Nassr A¹, Larson AN, Crane B, Hammerberg KW, Sturm PF, Mardjetko SM.

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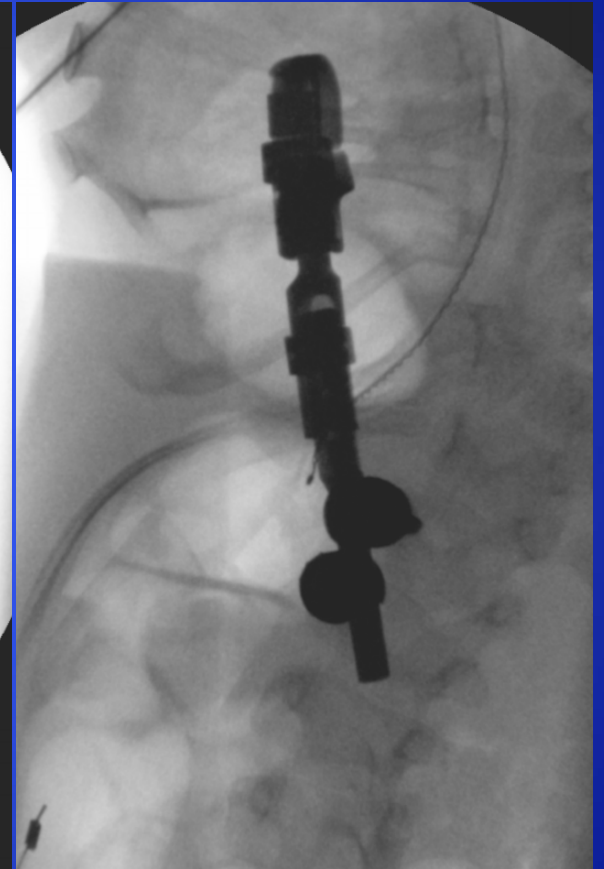
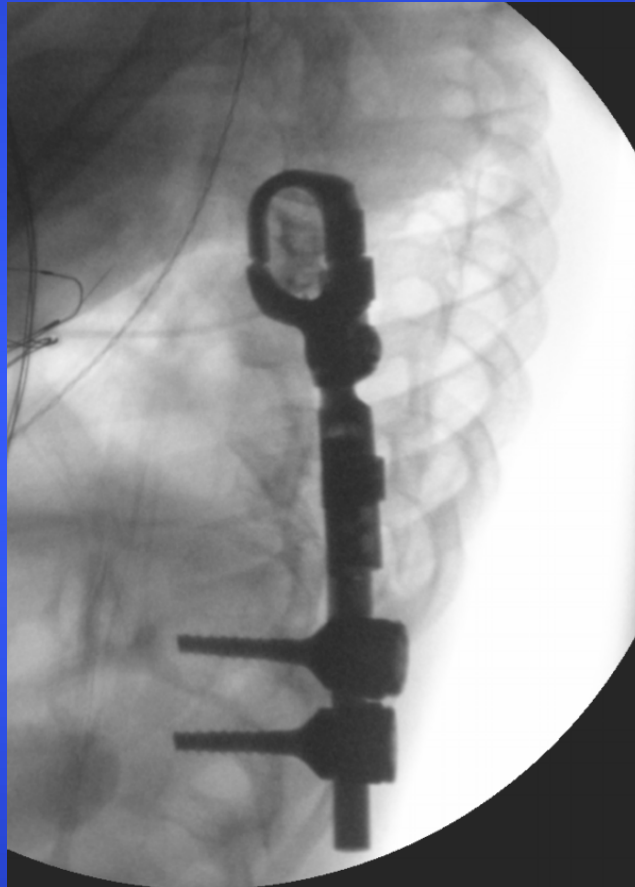
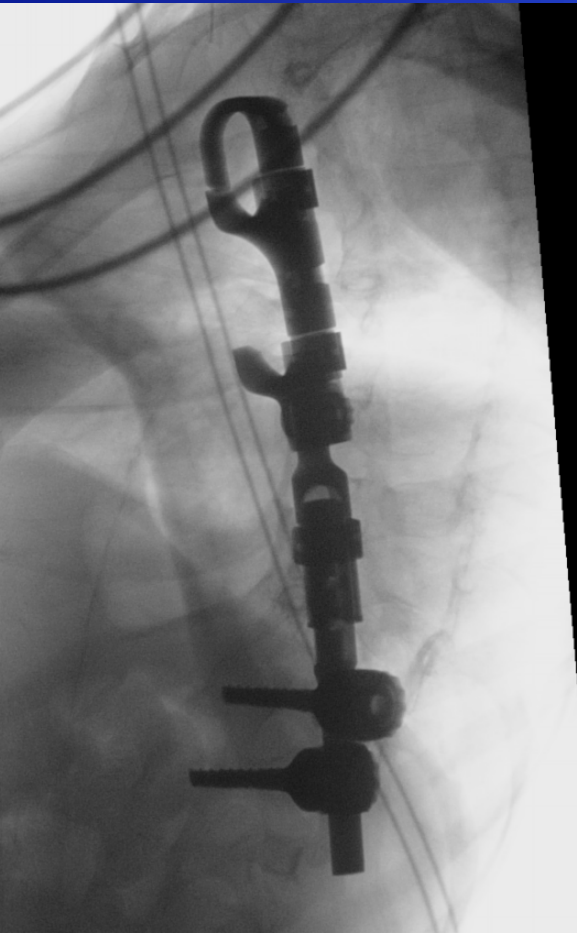


Congenital Deformities/Rib Fusions

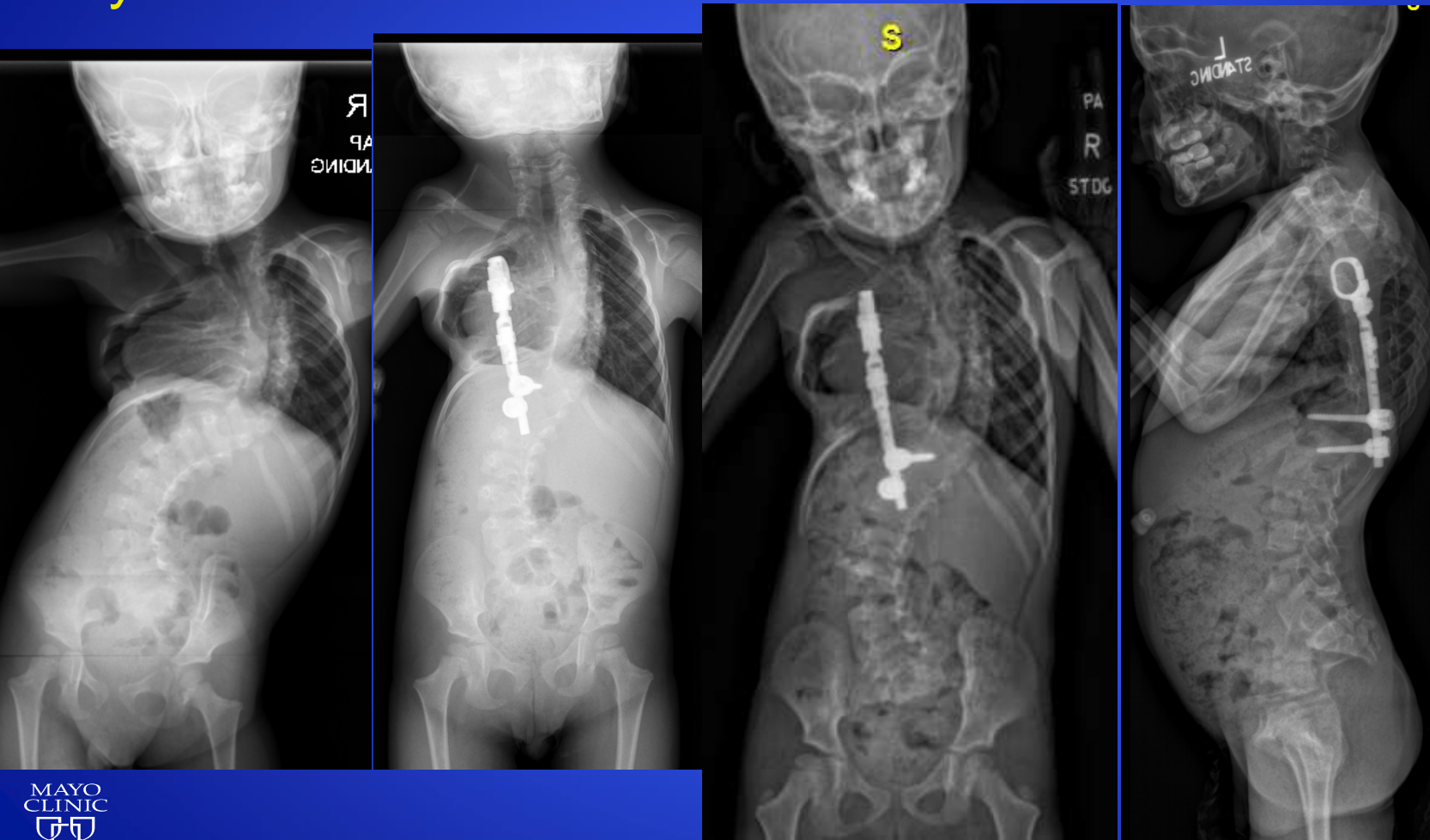


- Can sustain brachial plexus injuries from rib-based devices:
 - Compression from 1st rib
 - Retracted Superior pole of scapula
 - Secondary to Sprengel deformity reconstruction by pulling scapula down (Joiner et al., JBJS 2013)

Revised Construct POD2



2 Years Follow-Up → Upsized Rod and Screws 6 yo M



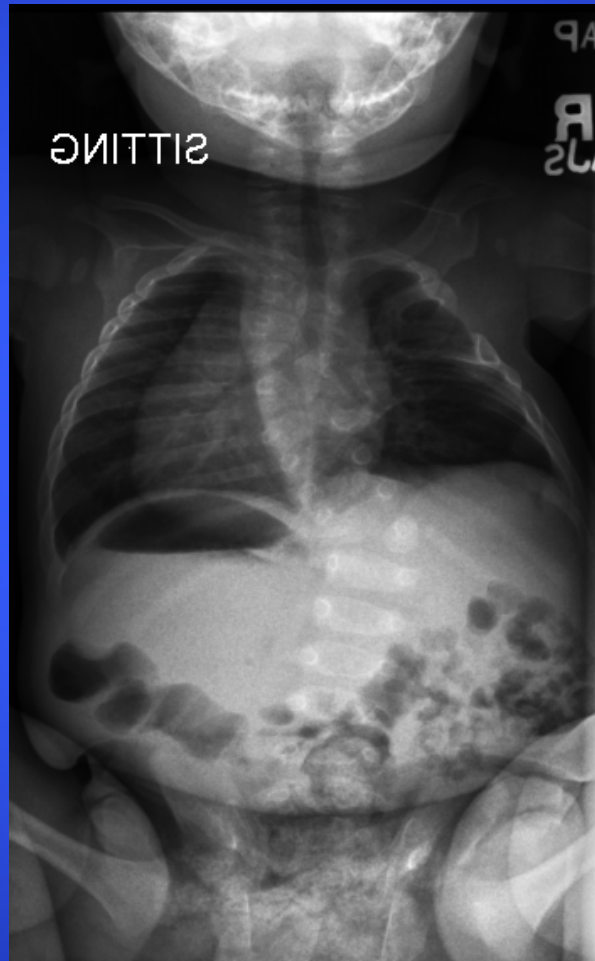
10.6 → 14 kg

83.7 → 98.7 cm

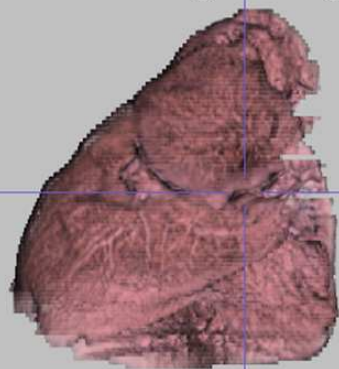
T1-T12

8 → 13.7 cm

- 1 yo M

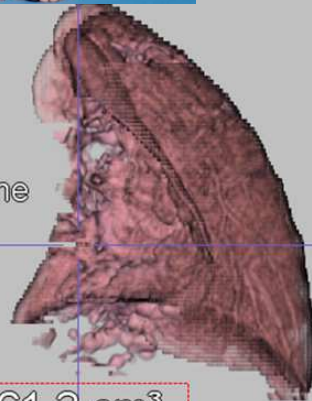


4 yo



11/18/2009 05:50:20
Karnis240

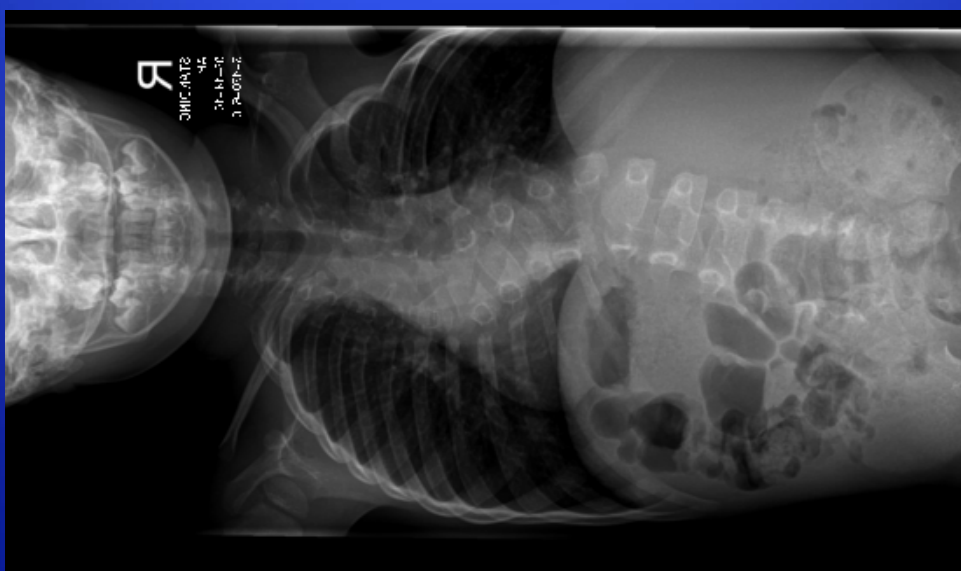
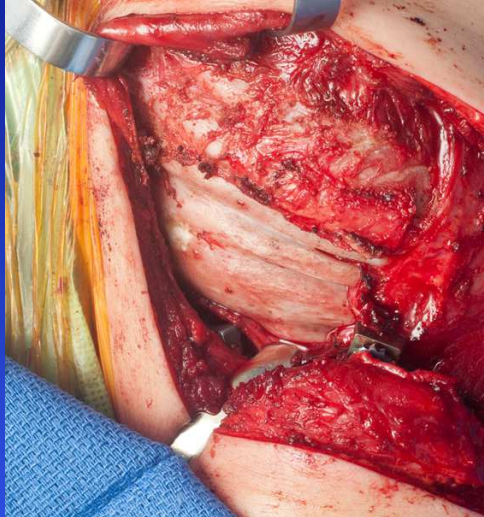
Left Lung Volume



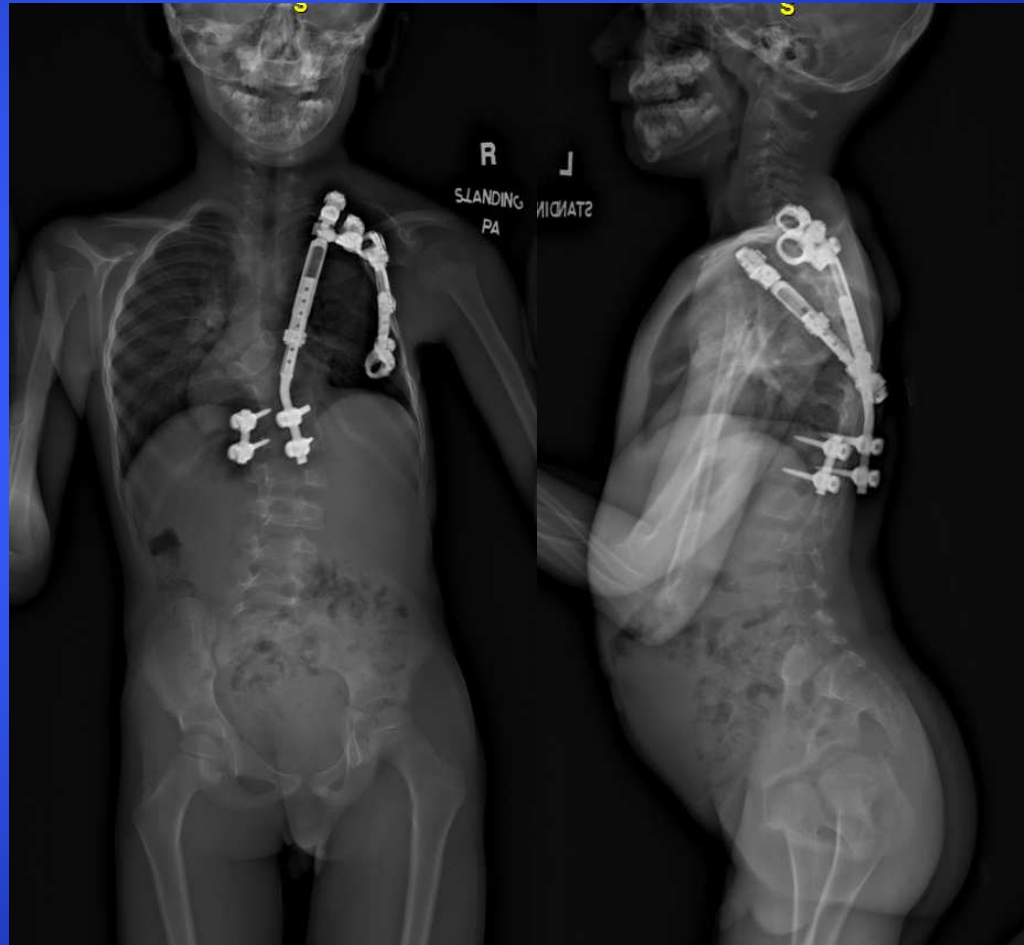
Volume = 174.3 cm³

Volume = 161.3 cm³

FDY:220.00
120.14

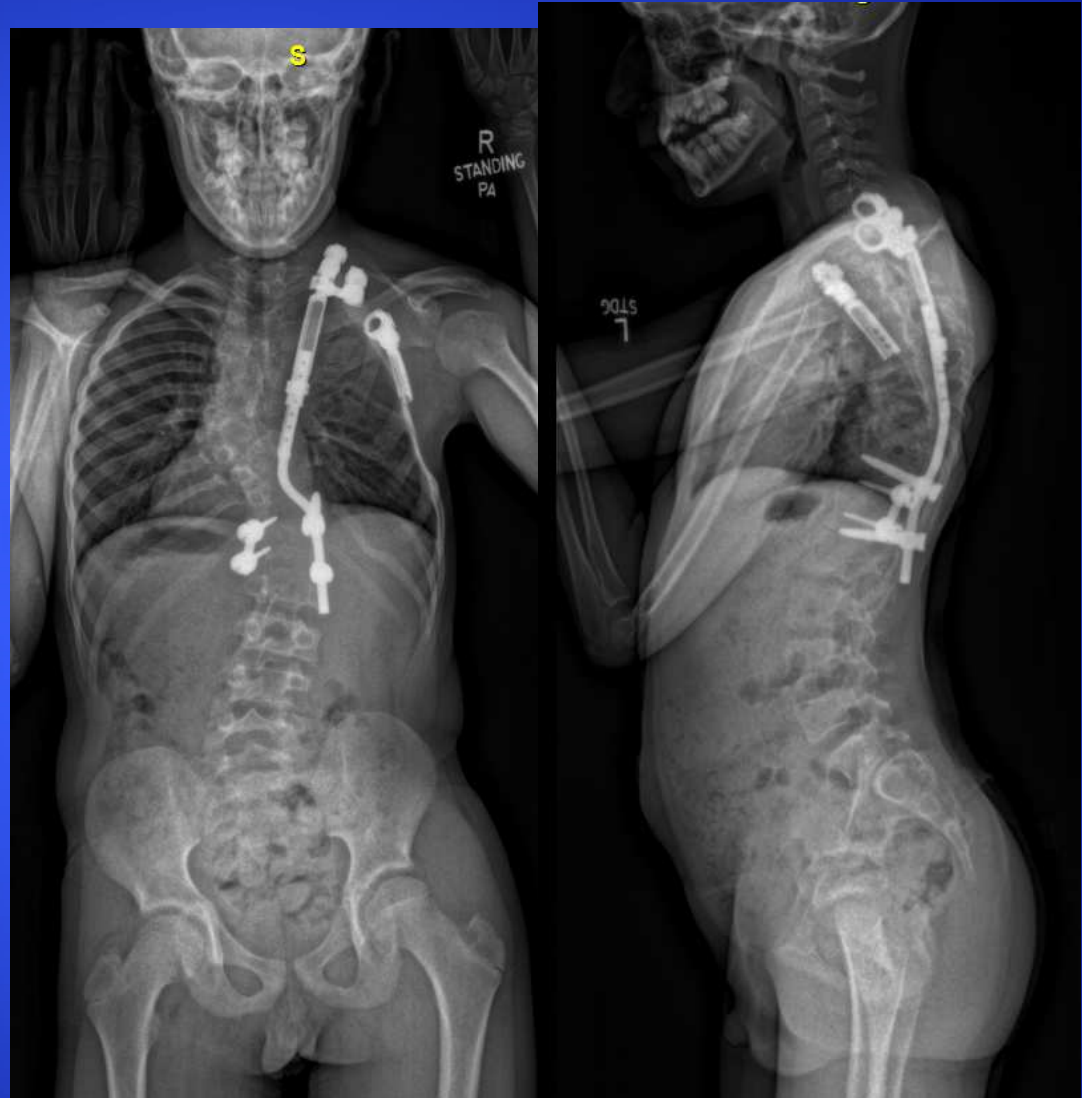


Age 8



12 yo M

- 16 cm thoracic ht



Summary: Congenital Scoliosis with Rib Fusions

- Always look for associated conditions (MRI, etc)
- Monitor arms and legs during surgical procedures
- PSSG database
 - Better deformity control with spine-based proximal rib anchors?
 - Improved overall spine length with thoracostomy
 - No data on PFTs, chest wall flexibility/function
- Area for further research
 - Use of MCGR
 - PFTs, more functional outcome measures



Thank you!

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