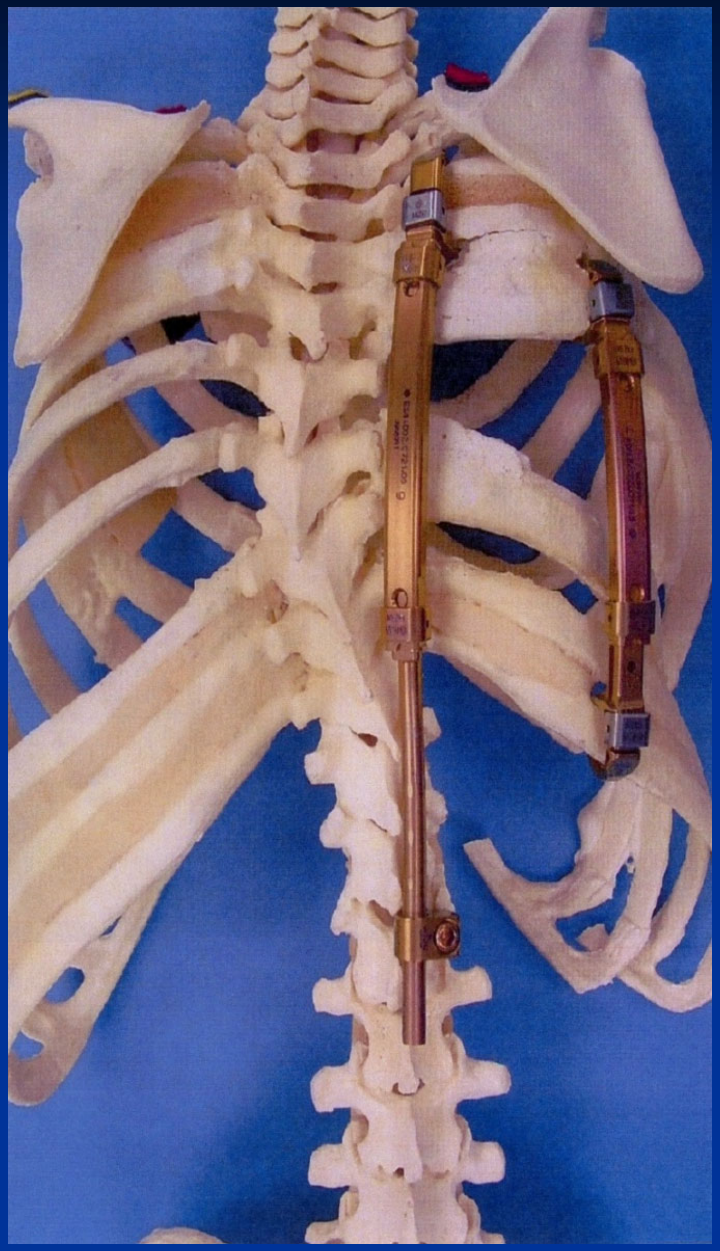


Non-Fusion Distraction Procedures: Thoracic Insufficiency Based

Robert M Campbell, Jr., MD
Division of Orthopaedics
CHOP
Philadelphia, PA

Thoracic Institute
Christus Santa Rosa Children's Hospital

Disclosures

- Grant Support
  - NORD
  - FDA office of Orphan Product Development
- Royalties, Honorarium
  - Synthes Spine Co.

Thoracic Institute
Christus Santa Rosa Children's Hospital

Thoracic Insufficiency Syndrome

The Inability of the Thorax to Support

Normal Respiration

or

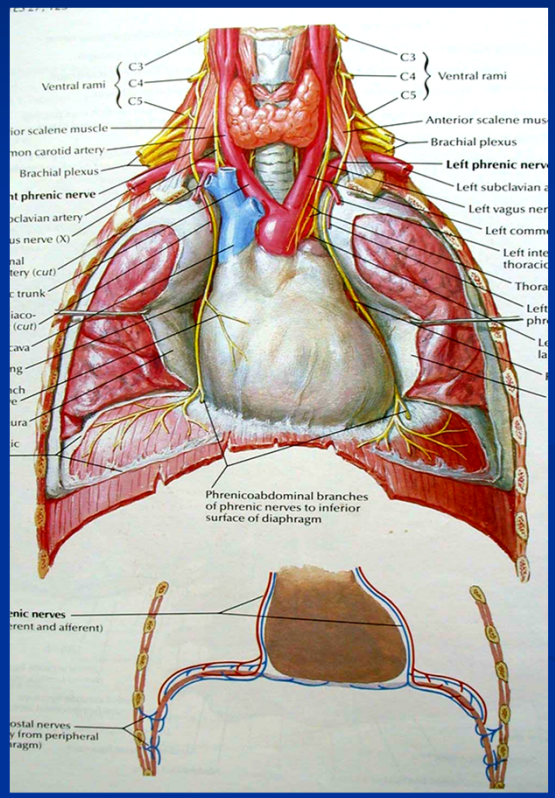
Lung Growth

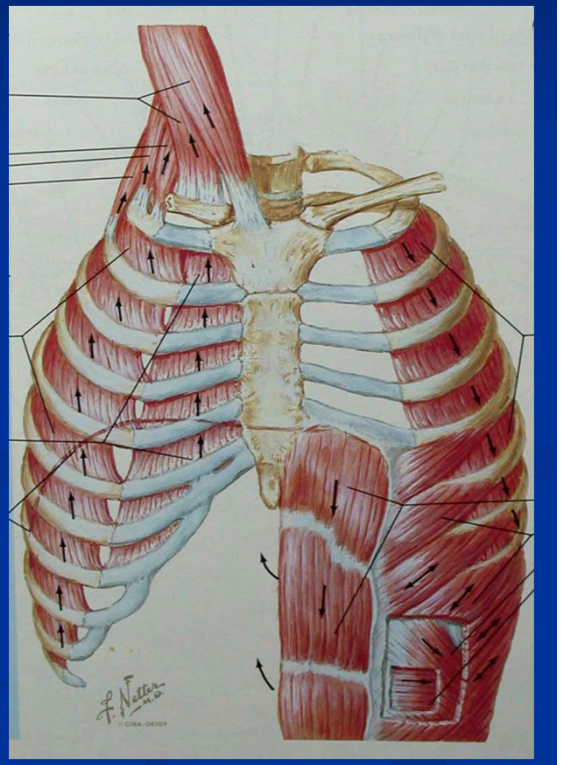
-Campbell, Smith, et al.
J Bone Joint Surg, Mar , 2003
J Bone Joint Surg, Aug, 2004

Thoracic Institute
Christus Santa Rosa Children's Hospital

The Normal Thorax: The Engine of Respiration

- **Volume**
- **Ability to change Volume**
  - diaphragm
  - chest wall expansion





Problems:

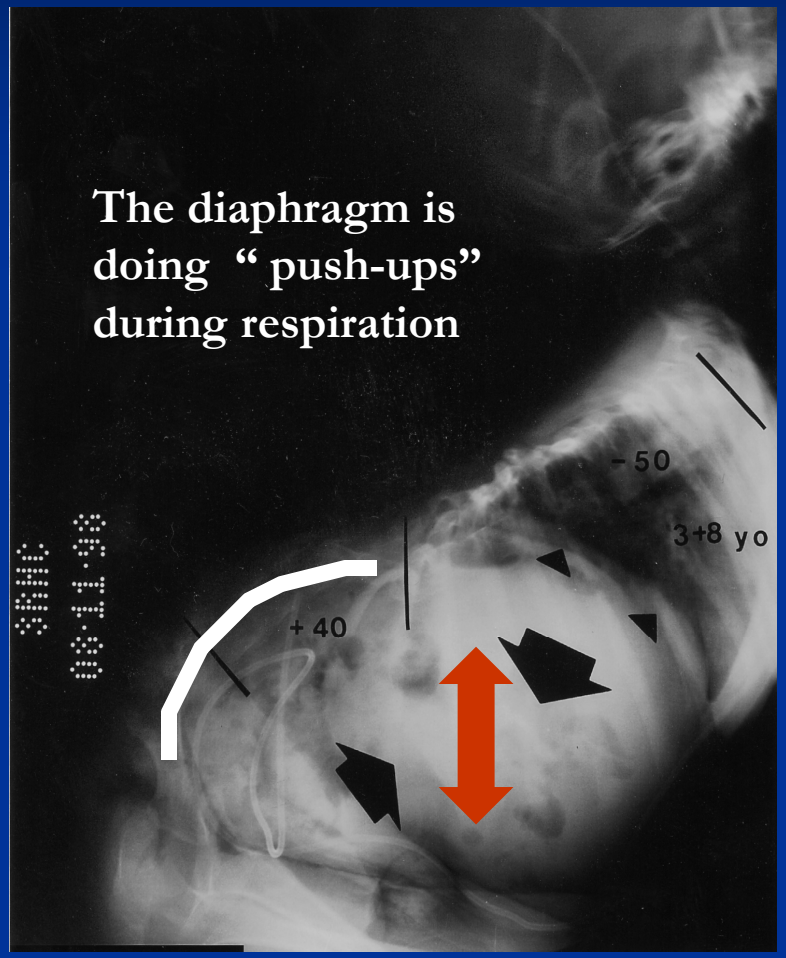
Primary Thoracic Insufficiency Syndrome

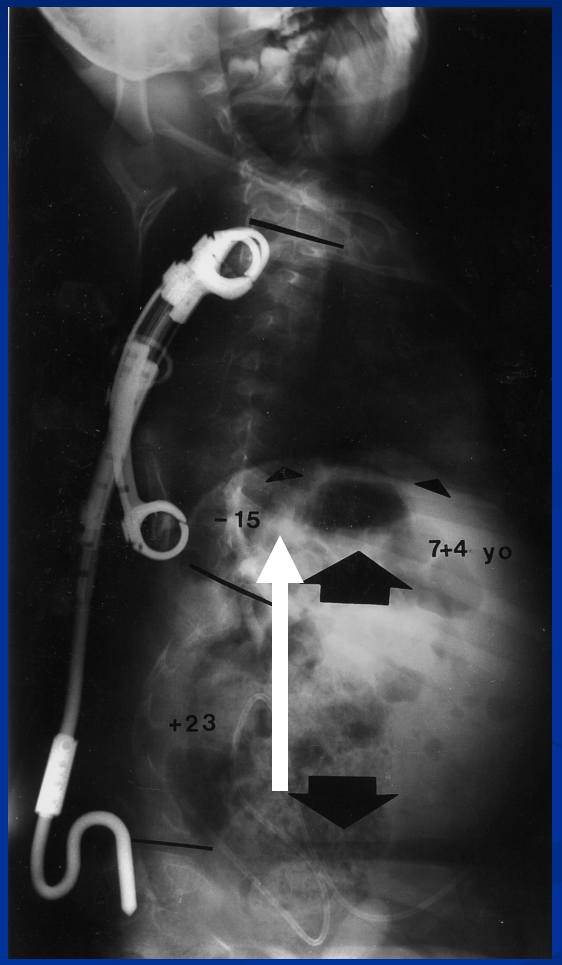
Campbell, Smith, et al. JBJS 2003, 2004

Thoracic Institute
Christus Santa Rosa Children's Hospital

Secondary Thoracic Insufficiency Syndrome

-Campbell, Smith, et al
JBJS, 2004



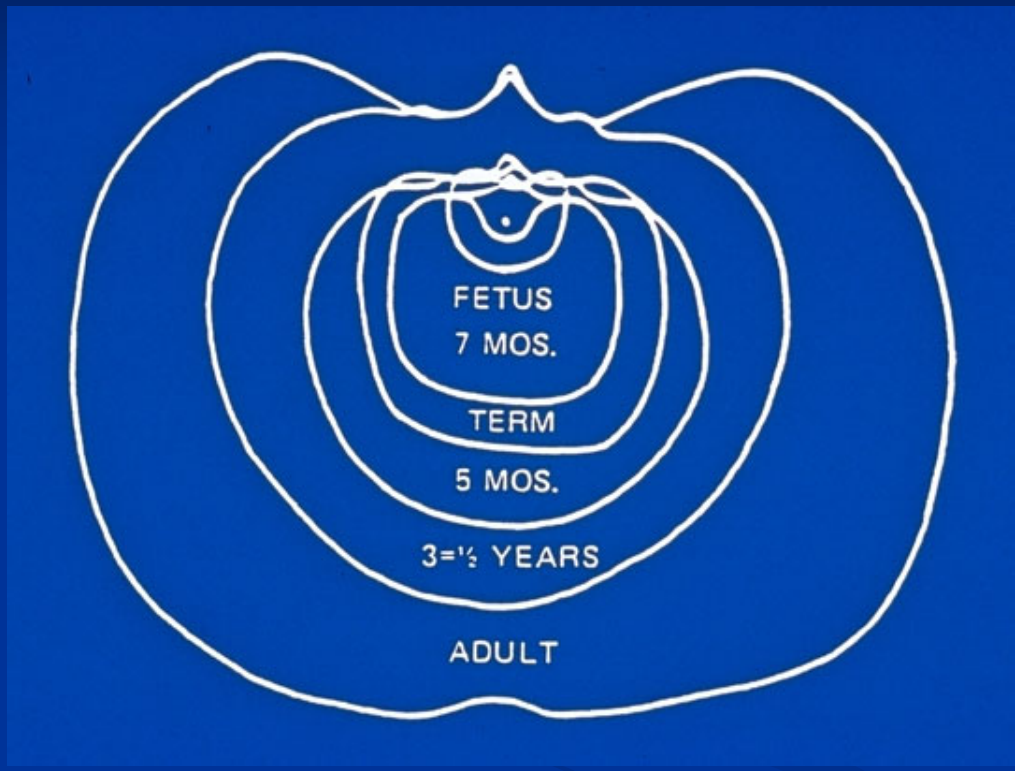


Positive " Marionette " Sign

Thoracic Institute
Christus Santa Rosa Children's Hospital

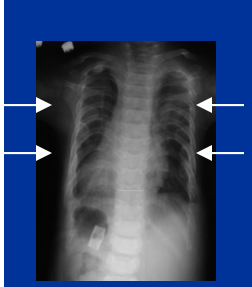
Lung Growth

Growth of the Thorax must occur during the *Golden Phase* of Lung Growth

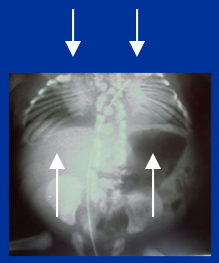


Thoracic Institute
Christus Santa Rosa Children's Hospital

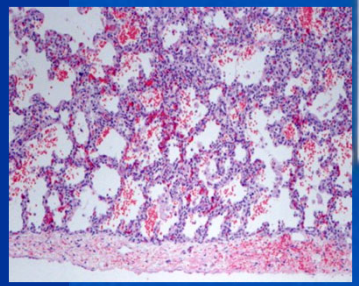
Lung and Thoracic Growth are Inter-Dependant

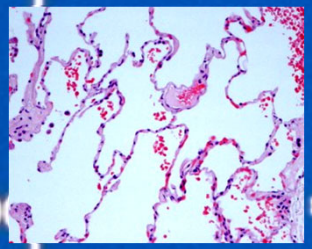


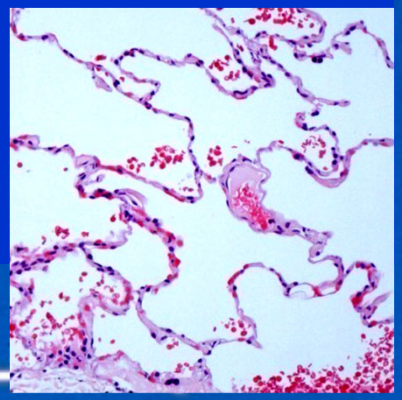
Jeunes



Jarcho-Levin

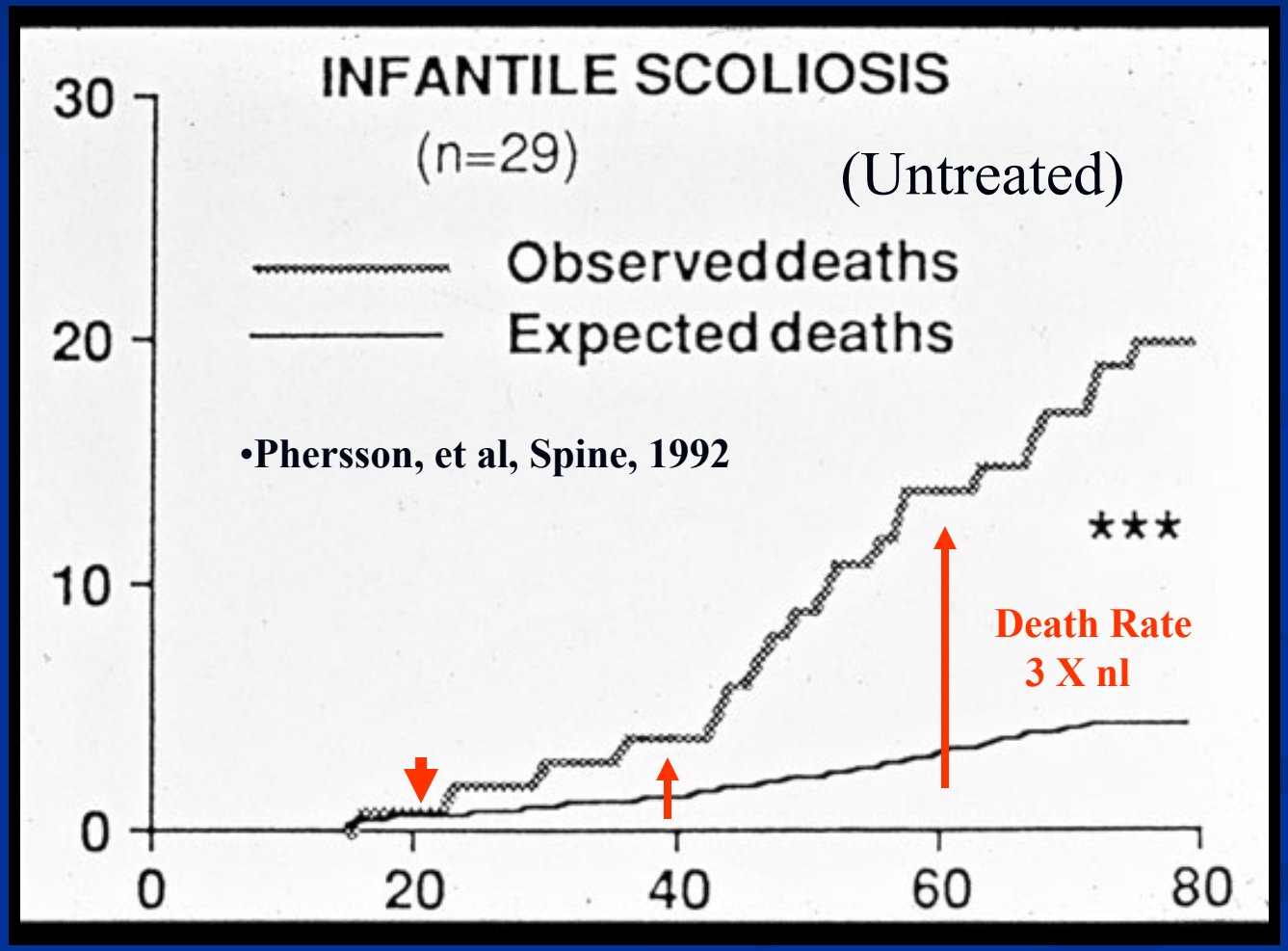






Long term Natural History of Untreated Scoliosis:

Probable Untreated Thoracic Insufficiency Syndrome



Thoracic Institute
Christus Santa Rosa
Children's Hospital

Why did these children die?

TIS

Absent Ribs/Scoliosis.
Died age 14 months

Jeune's Syndrome
Died age 18 months

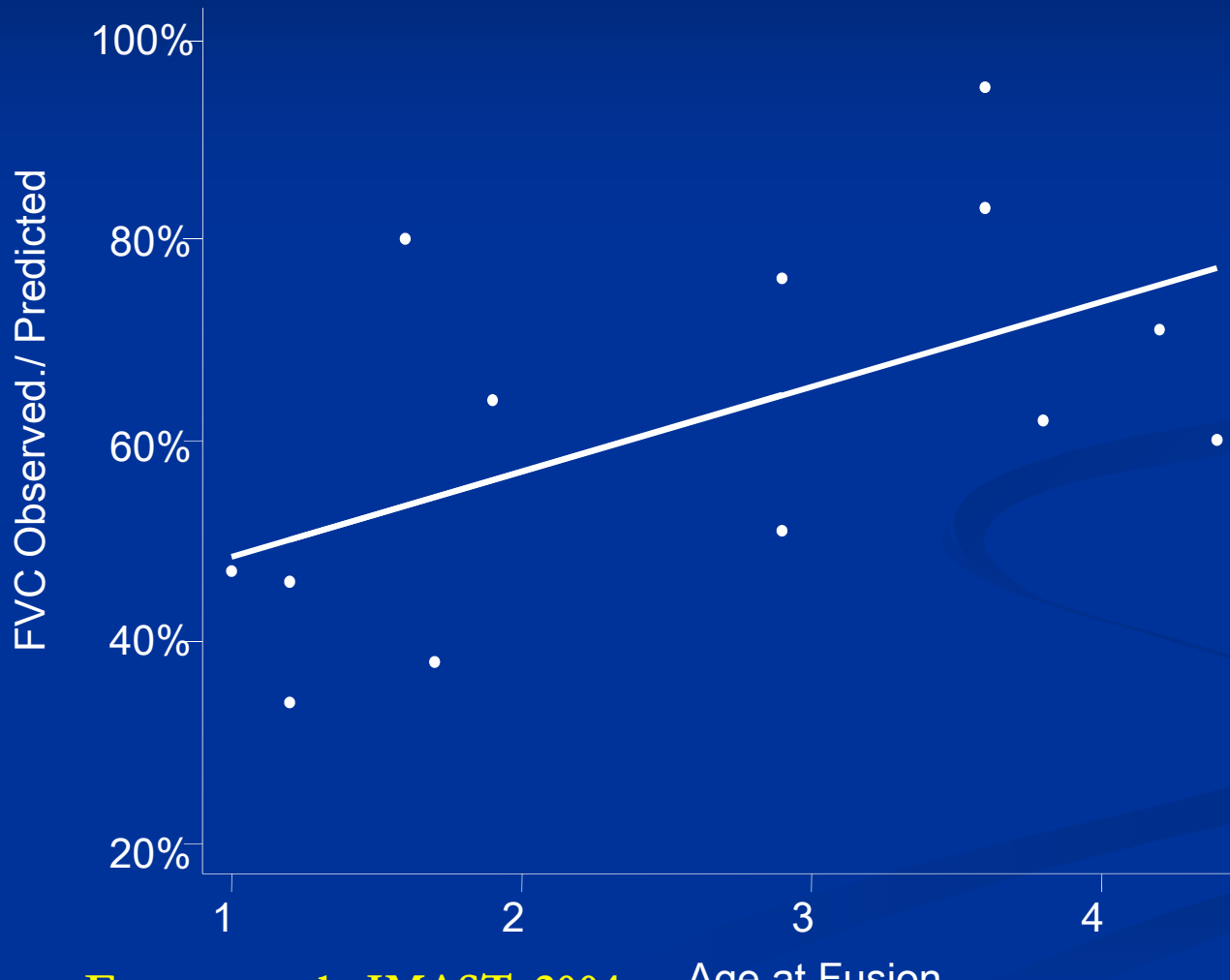
The pleural of anecdote is not data.

- FDA Chief of Devices comment to Dr. Campbell , 2002

Thoracic Institute

Christus Santa Rosa Children's Hospital

FVC Observed/Predicted is *more severely decreased* when *fusion is performed at younger age* (P=0.046)



-Emans, et al., IMAST, 2004

Thoracic Institute
Christus Santa Rosa Children's Hospital

Early Fused patients can do well

Cases Reports -Winter and Lonstein

(1999) Post fusion cong scoliosis age 3 yrs, agenesis lt lung (VC 70% nl at 41 yr f/u)

(2004) Post fusion cong scoliosis, C7-T8

(VC 70% nl age 18 yrs)

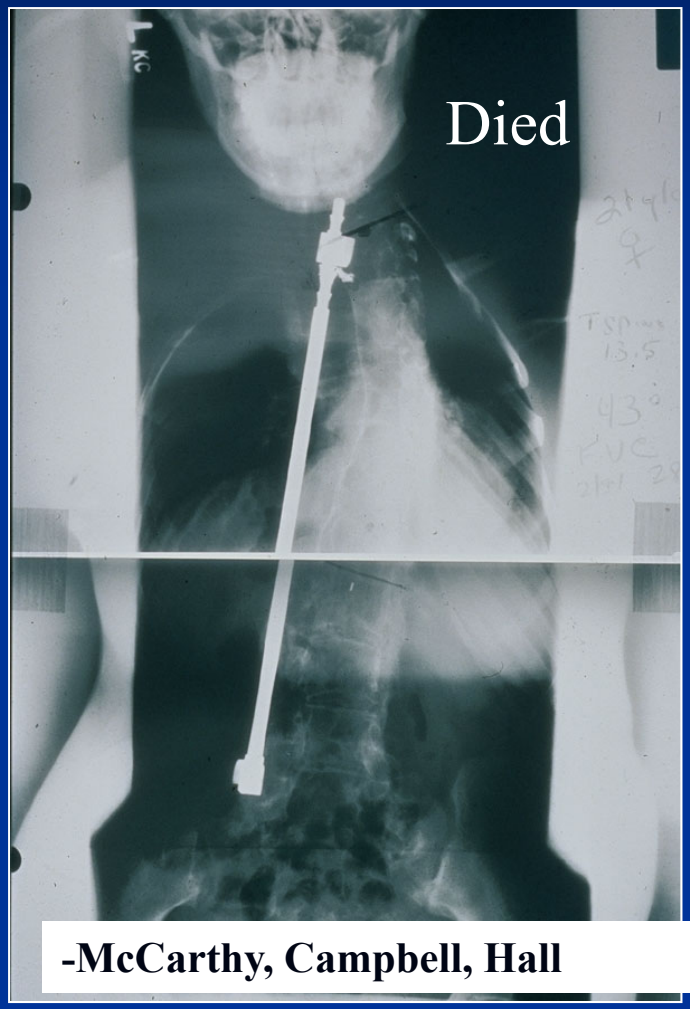
(2007) Post fusion/rib osteotomy age 1 yr, A/P fusion age 5 yrs

(VC 42% nl at 36 yr f/u)

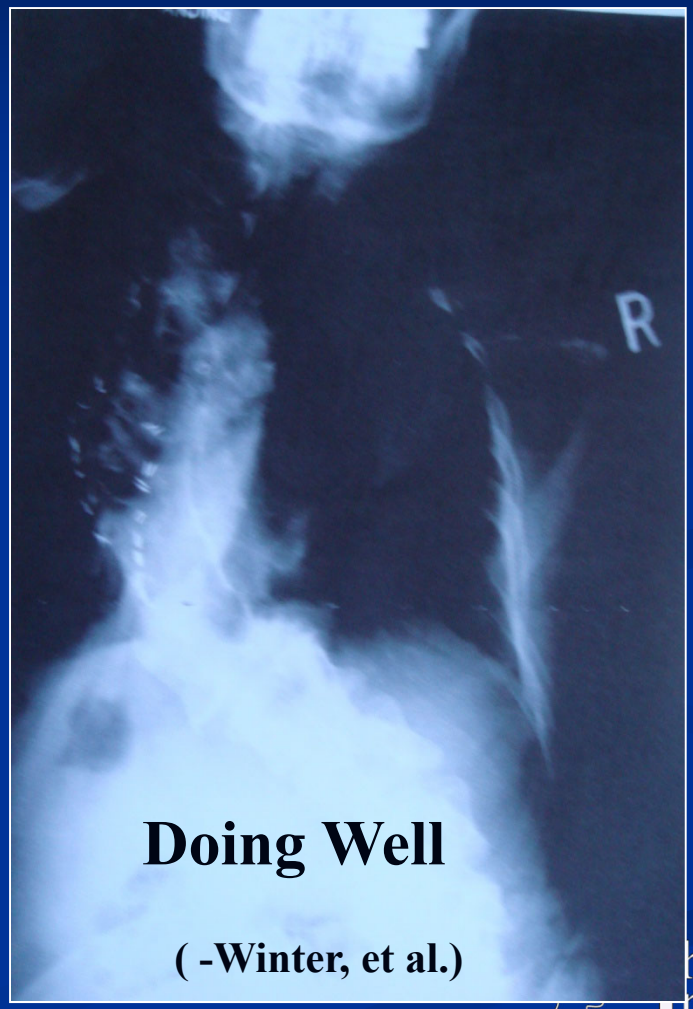
Thoracic Institute

Christus Santa Rosa Children's Hospital

What's the difference?



-McCarthy, Campbell, Hall



Doing Well

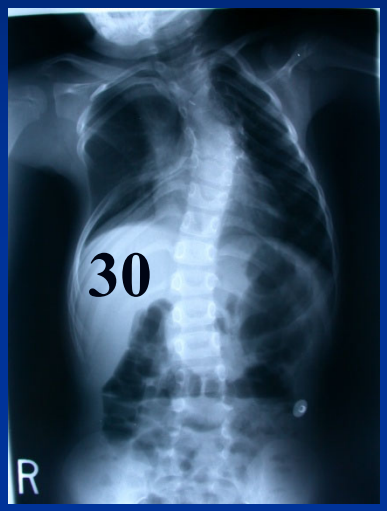
(-Winter, et al.)

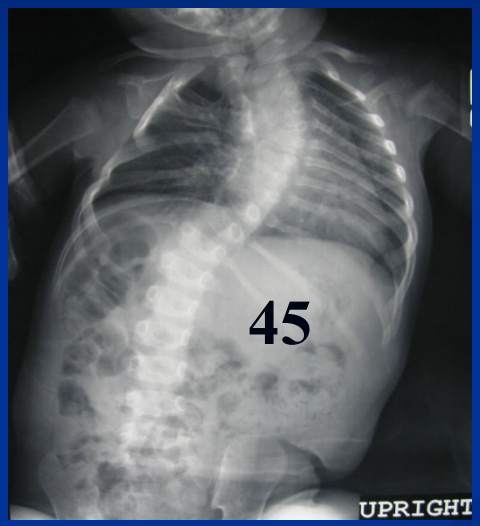
horacic
nstitute
Christus Santa Rosa
Children's Hospital

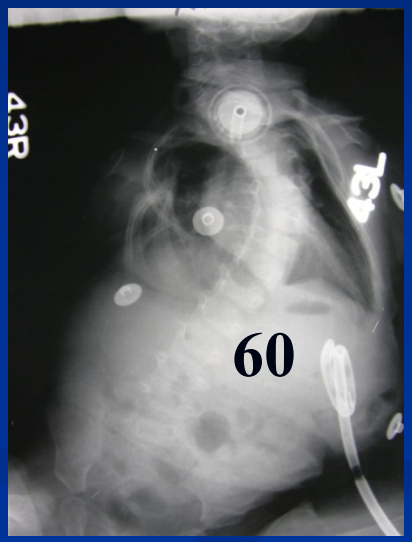
What's the Denominator ?

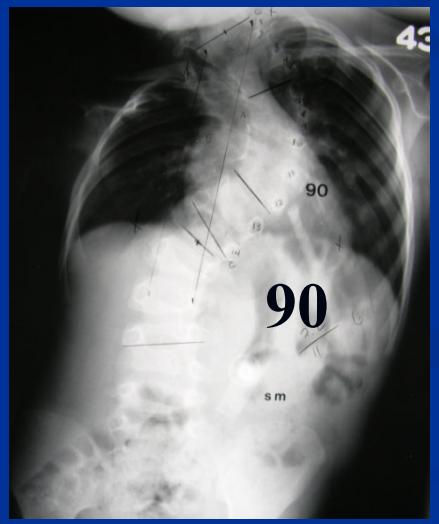
Thoracic Institute
Christus Santa Rosa
Children's Hospital

Beware of studying differences in small, heterogeneous patient populations









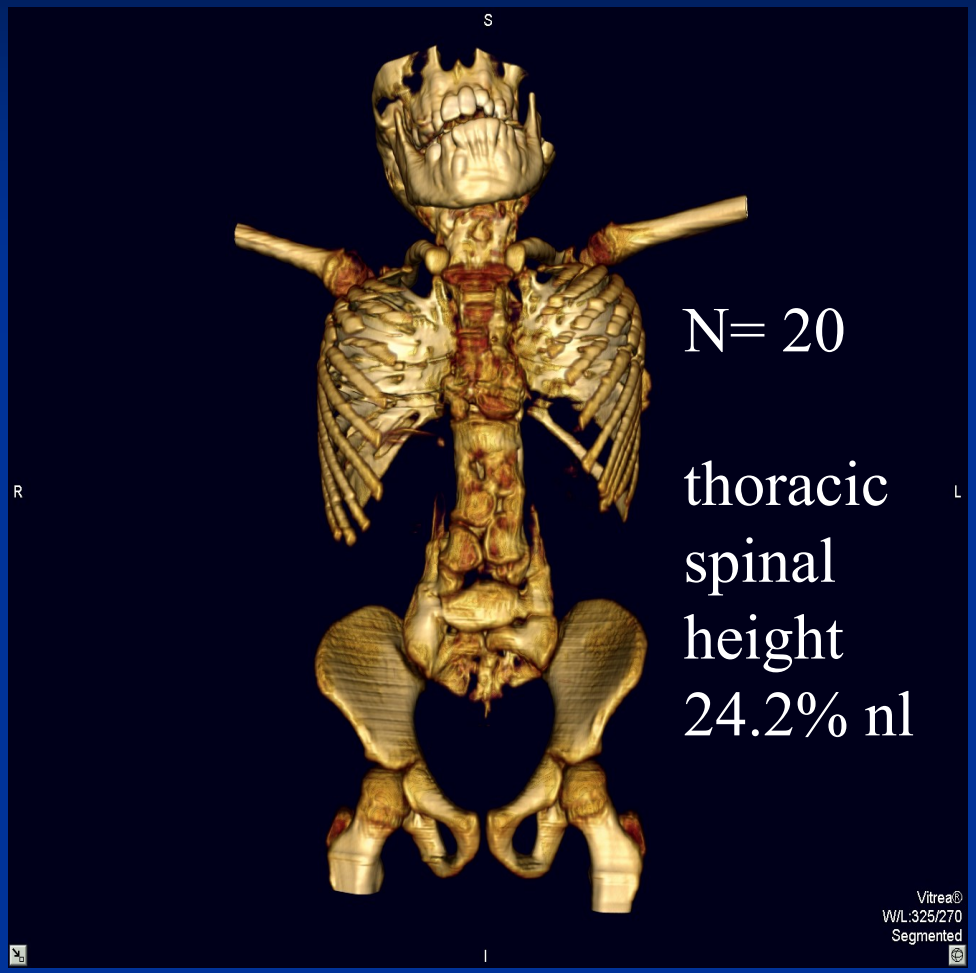
cic
ute
s Santa Rosa
en's Hospital

Time Honored Scoliosis Treatment

- The surgical treatment outcome is better than natural history **?**
- The gold standard for scoliosis treatment is spine fusion
  - "A short, straight spine is better than a crooked one" **?**
- Is growth inhibition of the thorax due to spine fusion a good trade-off ?

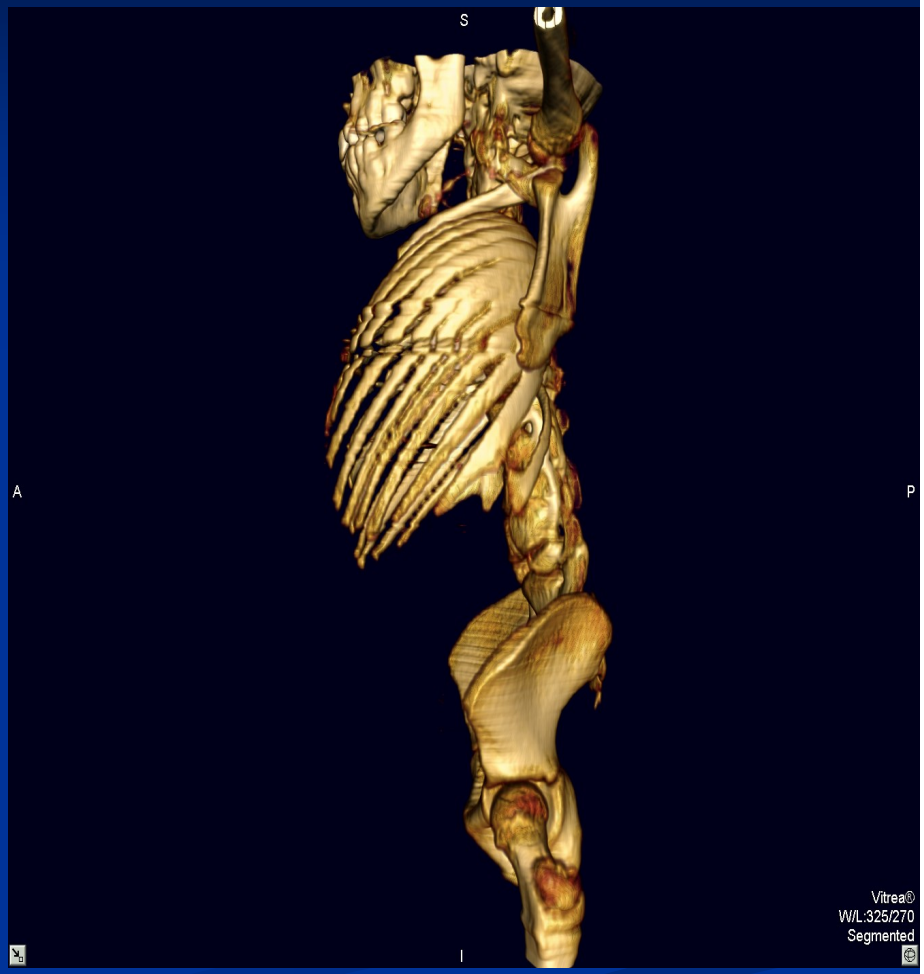
Thoracic Institute
Christus Santa Rosa Children's Hospital

Natural history model of early spine fusion: Jarcho-Levin Syndrome



N= 20

thoracic spinal height 24.2% nl

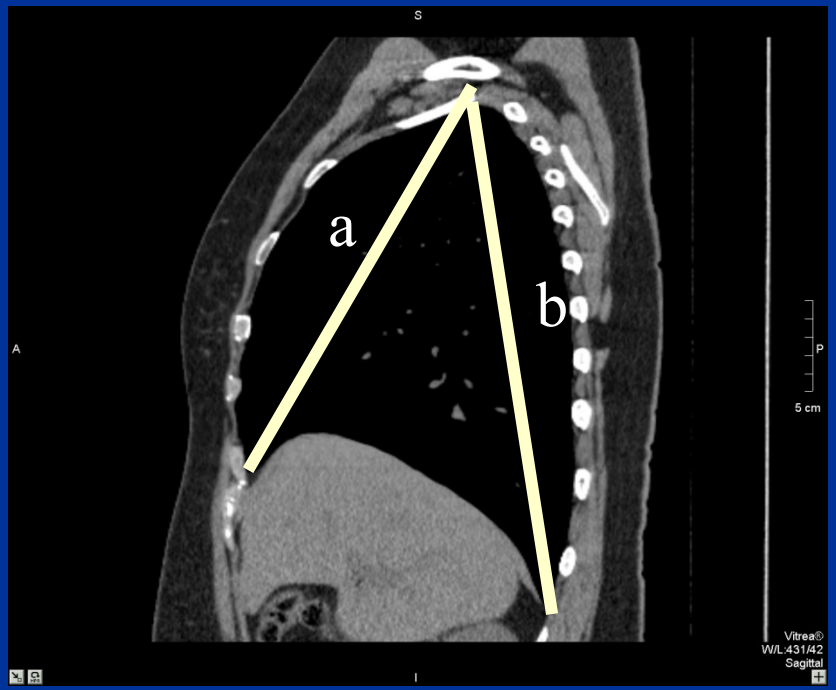


Ramirez, et al. JBJS, 2007

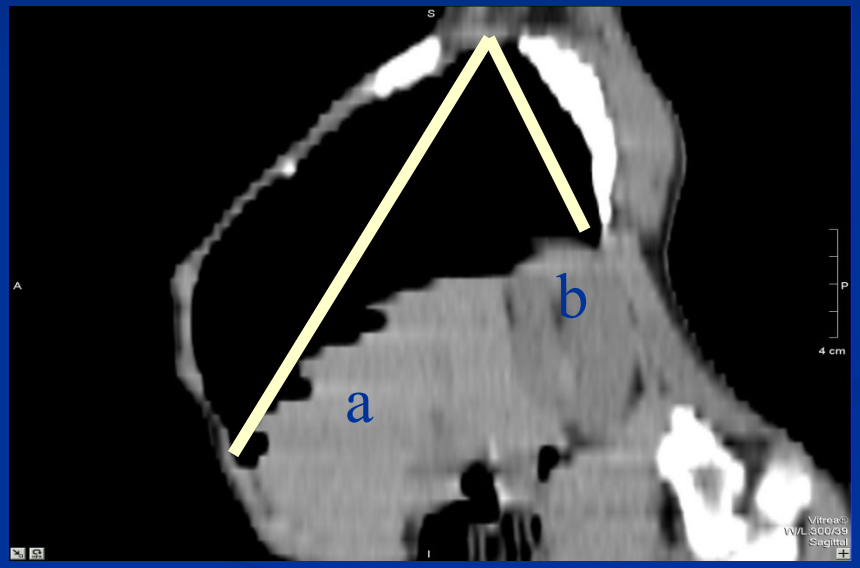
Thoracic Institute
Christus Santa Rosa Children's Hospital

Sagittal Patho-Anatomy

Ramirez, et al. JBJS, 2007



Normal sagital costophrenic depth ratio.



Abnormal sagital costophrenic depth ratio in Spondylothoracic dysplasia.

Thoracic Institute
Christus Santa Rosa Children's Hospital

Survivors of Jarcho-Levin Syndrome

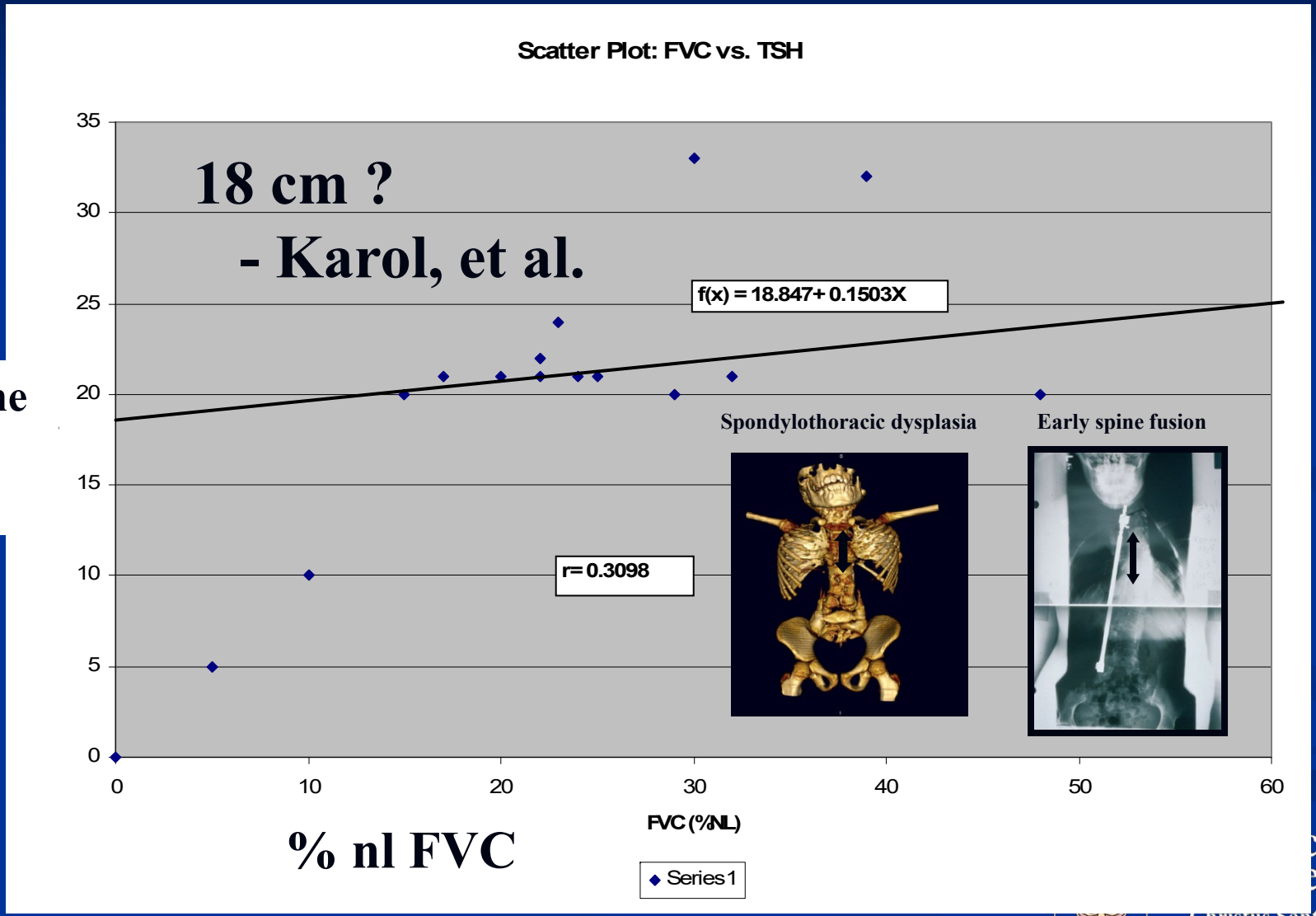
- Thoracic Spinal Height 24.2% nl

- CT lung scan volume 23.4 % nl
- FVC 27.9 % nl
- FEV1 29.5 % nl
- FEV1/FVC 0.92

- Is their a direct relationship between per cent normal thoracic spinal height and per cent normal vital capacity?

Thoracic Institute
Christus Santa Rosa Children's Hospital

% nl thoracic spinal ht vrs VC % nl



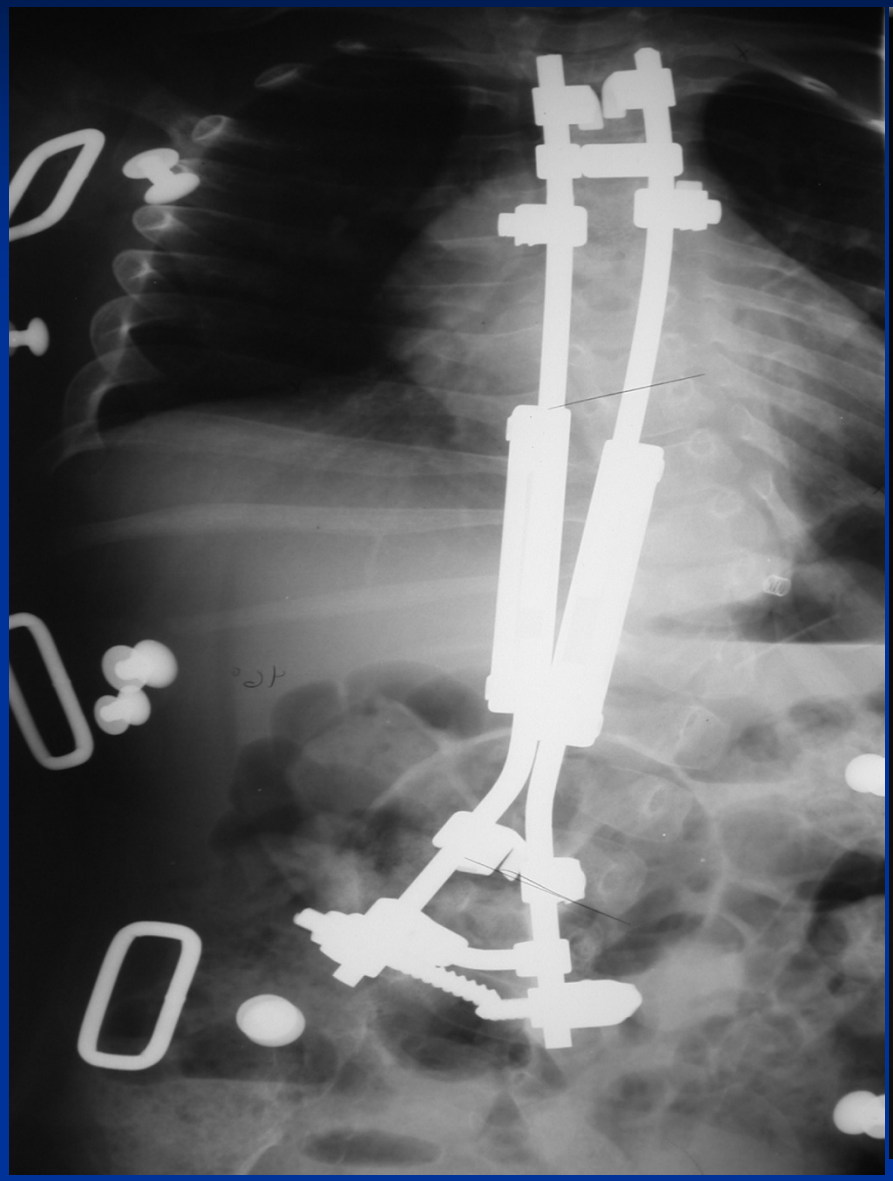
The Thorax: Surgical Treatment Viewpoint and TIS

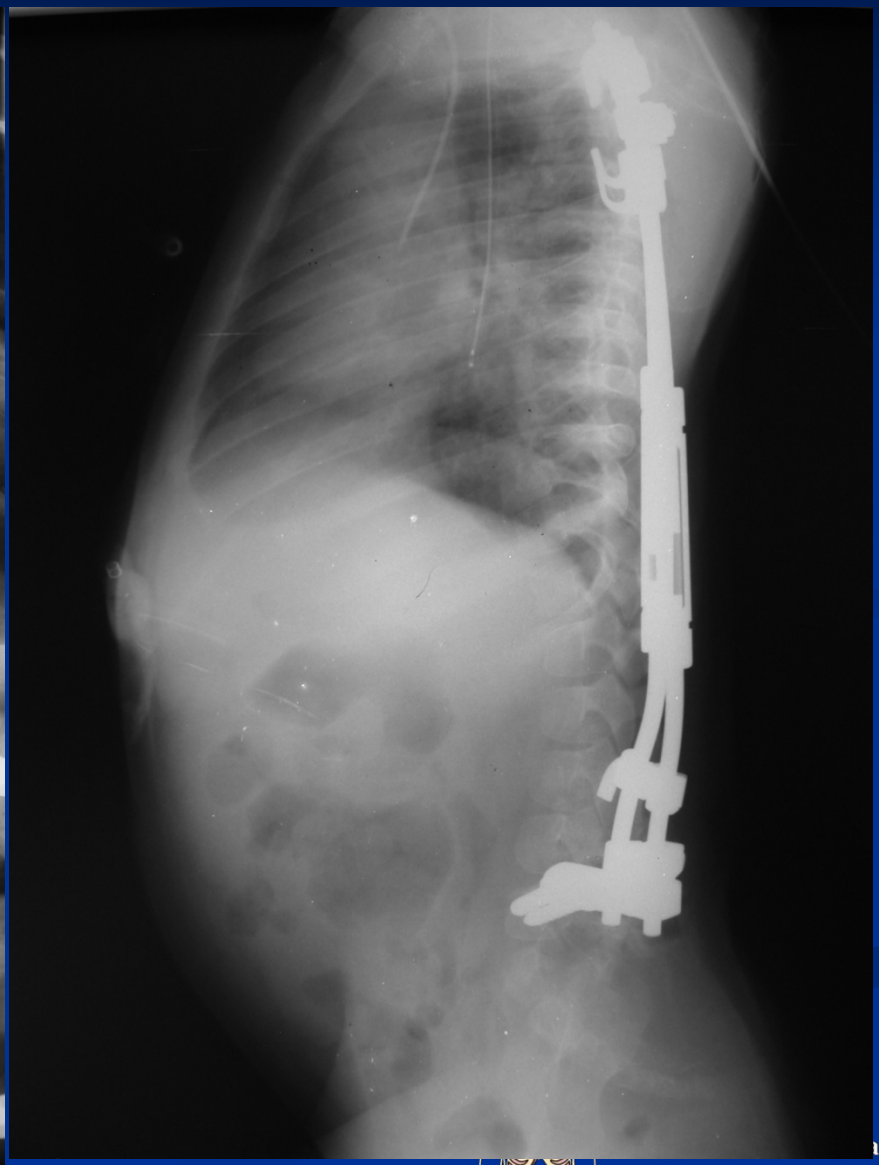
- The thorax is a *dynamic* chamber of respiration, composed of the spine , the ribs , and the sternum
- The spine is the *posterior pillar* of the thorax
- Treatment Restores the Characteristics of the Thorax:
  - *It must have normal , stable, symmetrical volume*
  - *It must be able to change this volume*

-Campbell, Smith, et al
J Bone Joint Surg, 2003

Thoracic Institute
Christus Santa Rosa Children's Hospital

Growing Rods

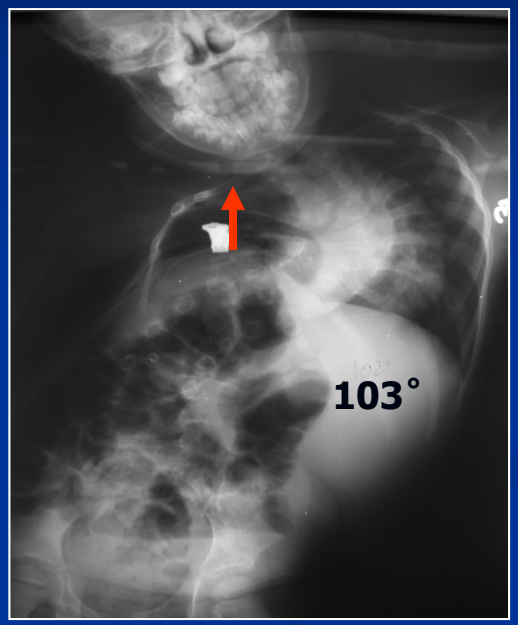




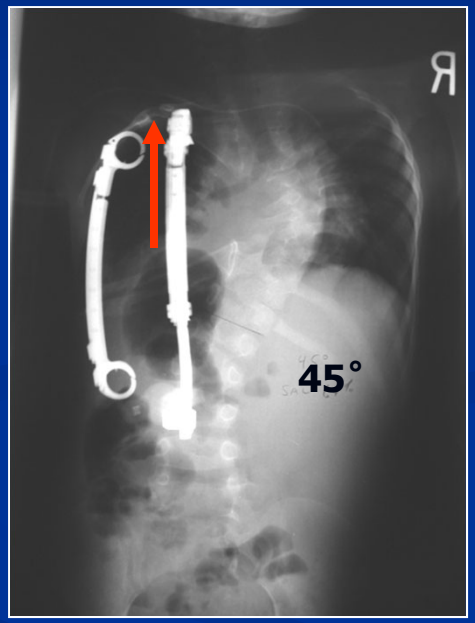
Children's Hospital

What does VEPTR treat?

- Thoracic Insufficiency syndrome
  - Primary
  - Secondary
- Volume depletion deformities of the thorax
- Indirectly, spine deformity
  - Cobb angle Correction without growth inhibition



SAL 21%

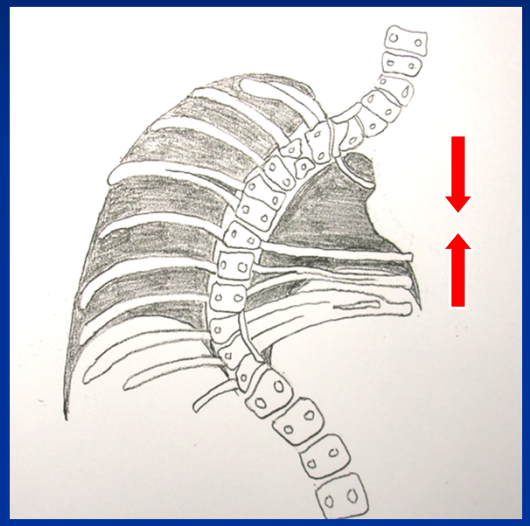


SAL 64%

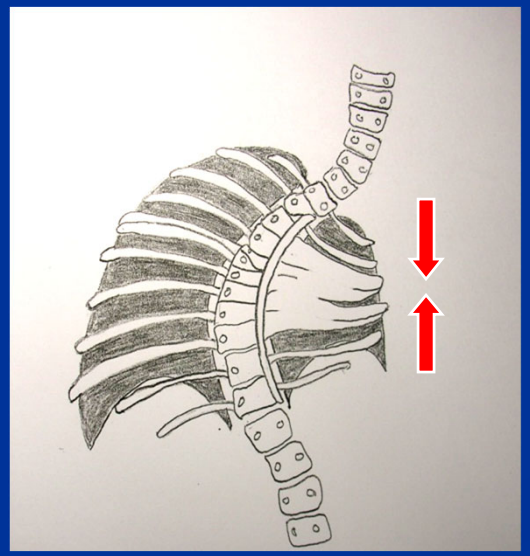
Thoracic Institute
Christus Santa Rosa Children's Hospital

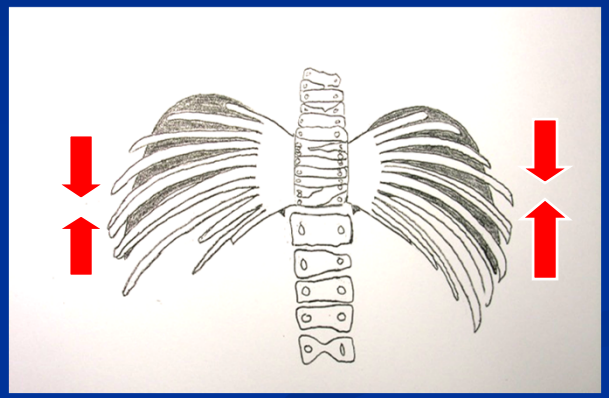
Volume Depletion Deformities of the Thorax - Campbell Smith, JBJS, 2007

I

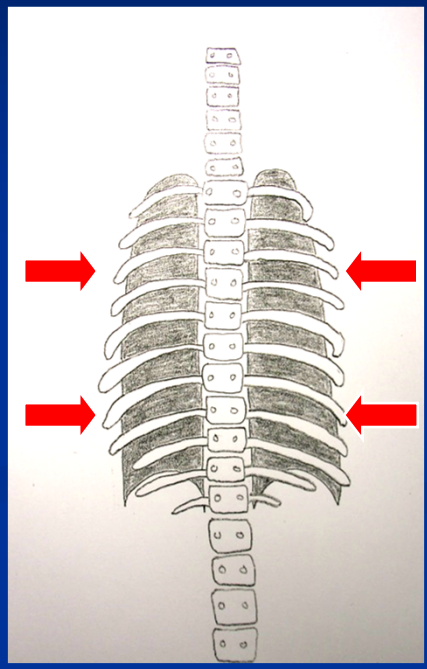


II



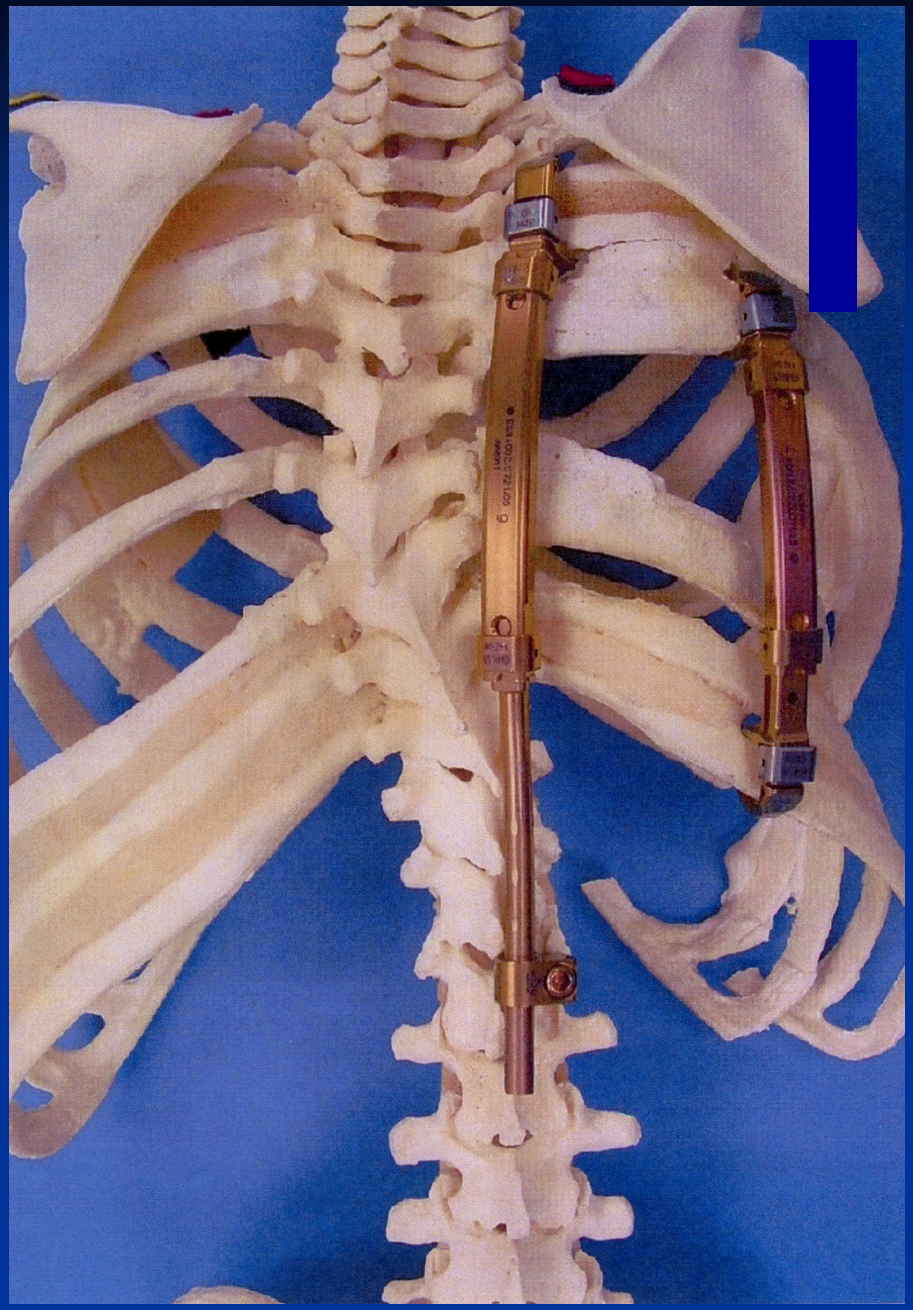


III a



III b

Thoracic Institute
Christus Santa Rosa Children's Hospital



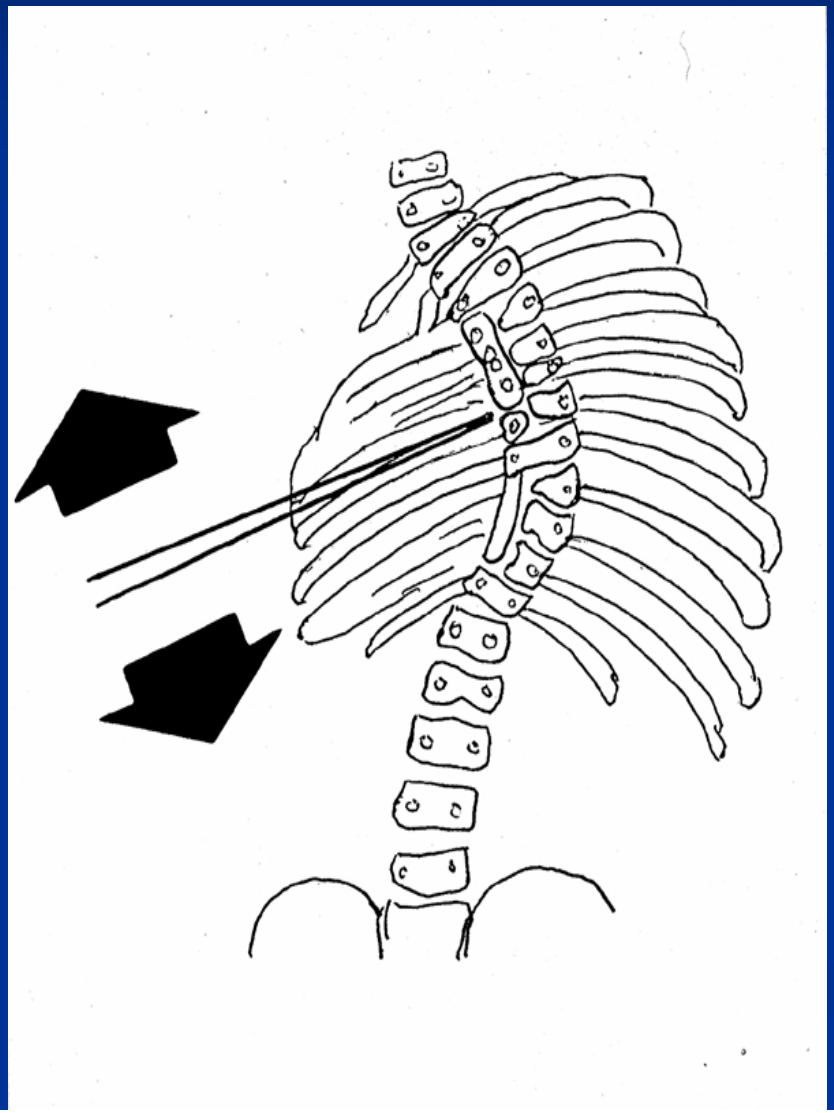
VEPTR Treatment

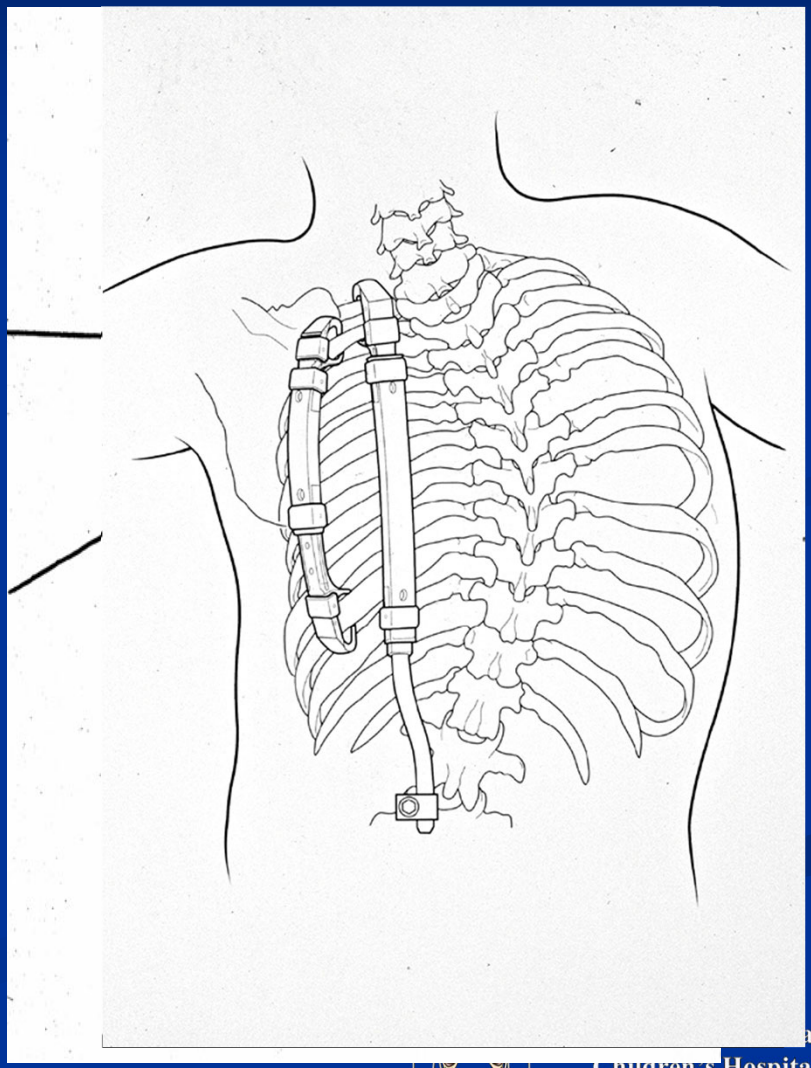
for Type II VDD

Thoracic Institute

Christus Santa Rosa
Children's Hospital

VEPTR Opening Wedge Thoracostomy





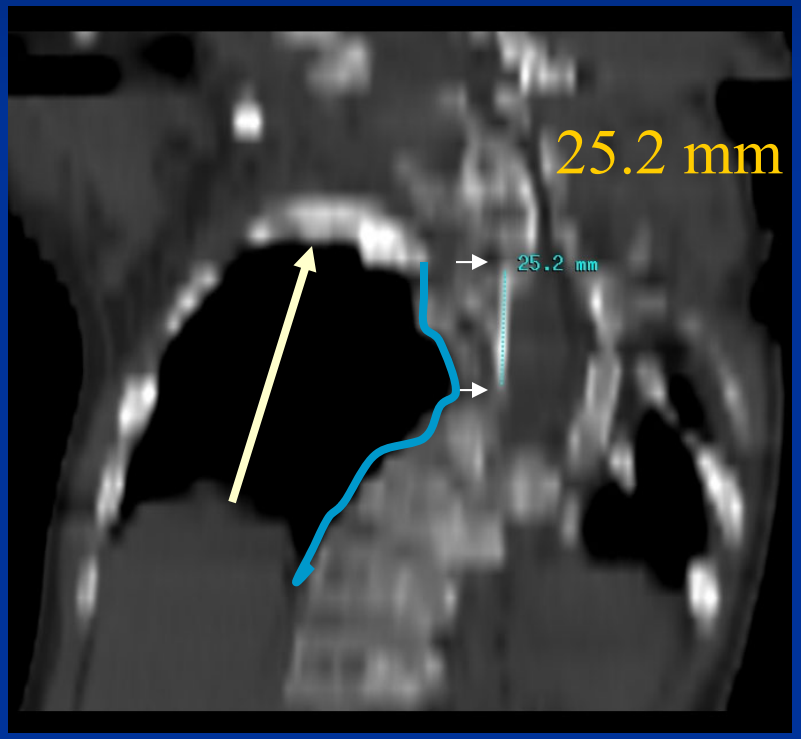
Results

- Avg. preop curve 74 º (35 - 140º)
  (avg. progression 15º / yr)
- Avg. post op curve 49º (4 - 84º)
  Avg. 25º correction
- Avg. thor. spine height growth 0.71 cm/yr
- Avg. increase thor. width 0.51 cm/yr

-Campbell, Smith, JBJS, 2004

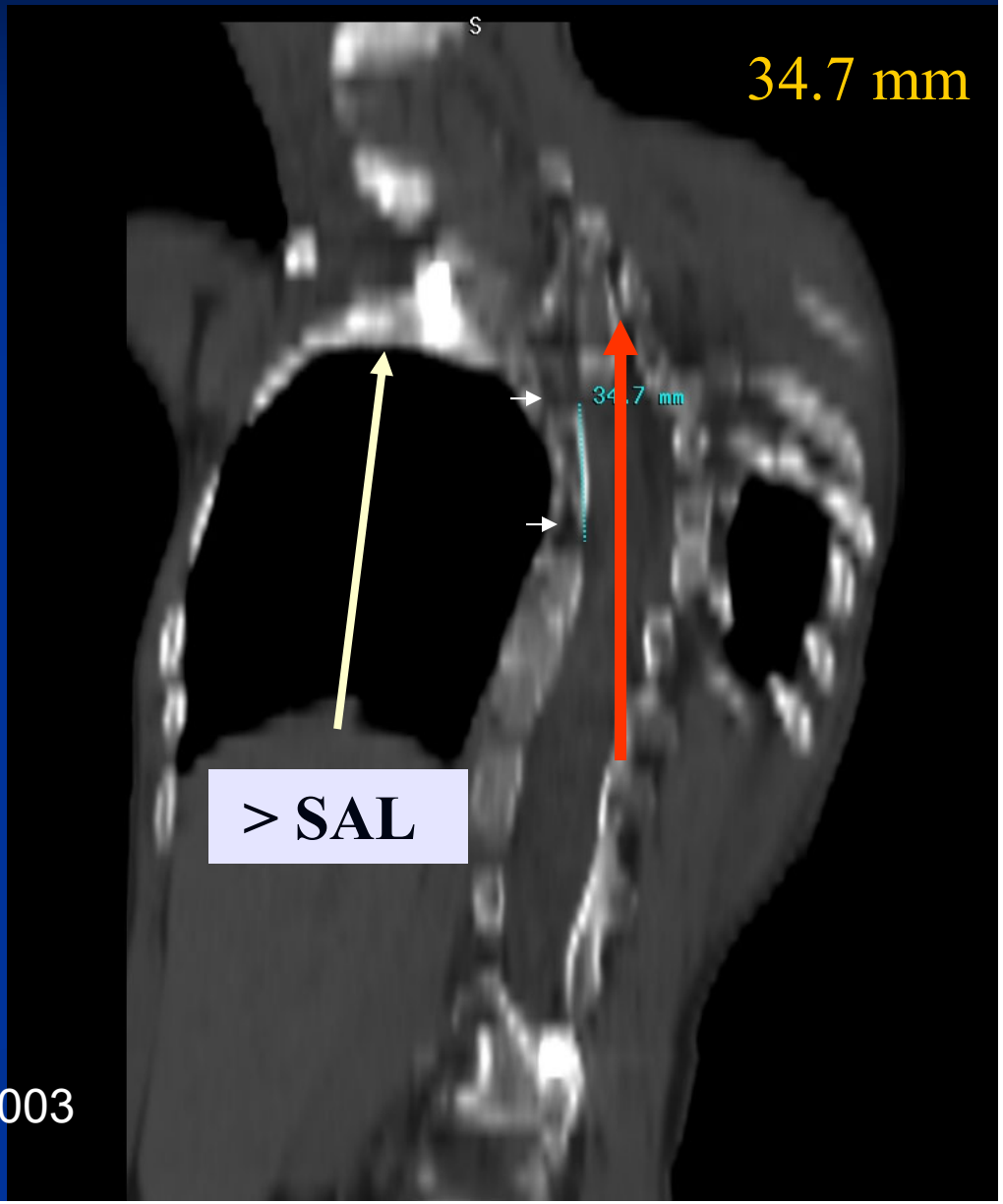
Thoracic Institute
Christus Santa Rosa Children's Hospital

Growth of Bars / Concave side of Spine

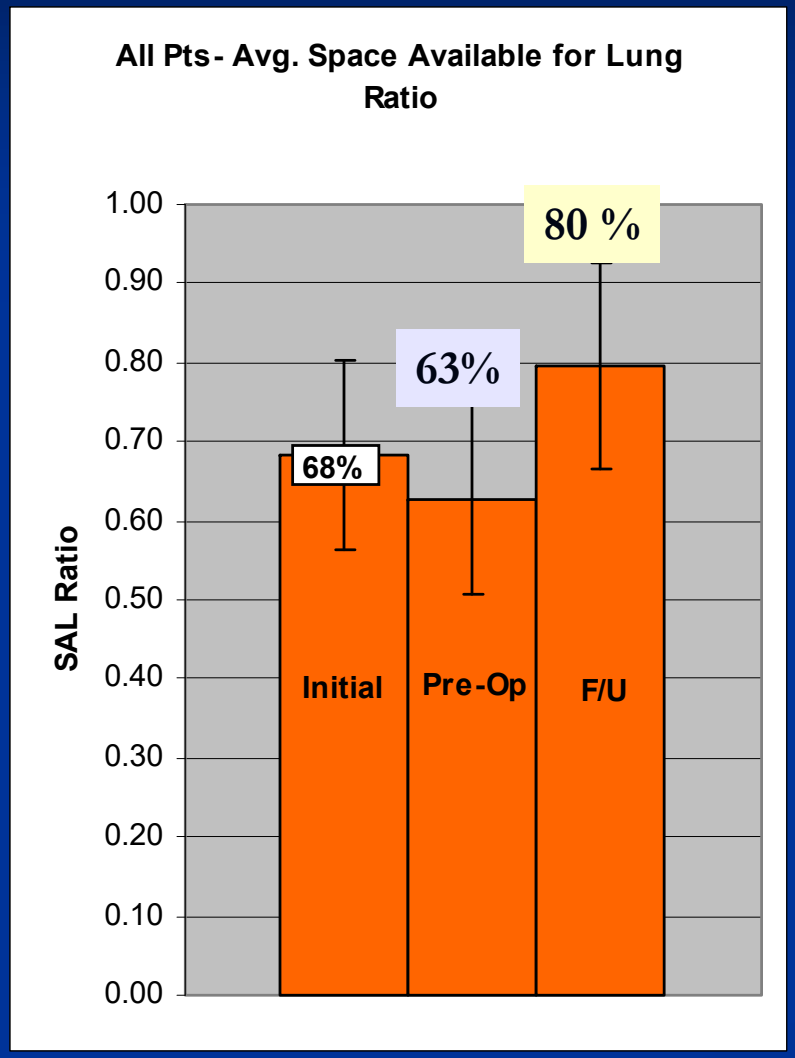


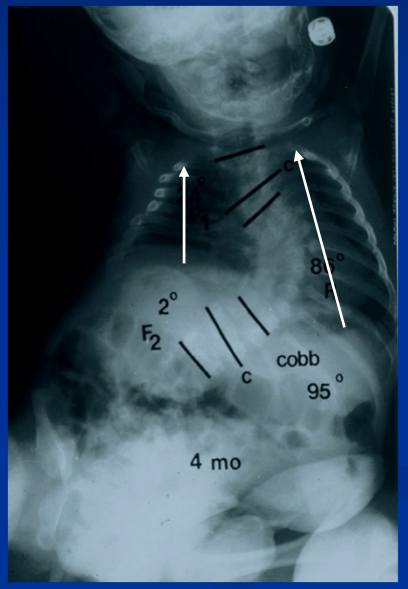
-Avg 7% increase length concave side and bars

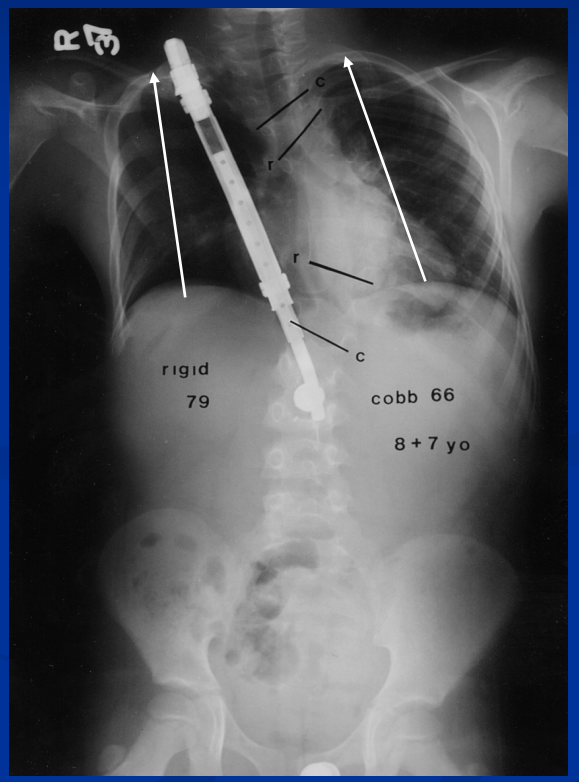
P < 0.0001 -JBJS, 2003



Space Available for Concave Lung







Campbell , Smith et al., JBJS, 2004

Thoracic Institute
Christus Santa Rosa Children's Hospital

Emans, et al. Spine, 2005

Lung volume by CT:

Pre-op:	369 ± 279 cm3
First Post-op	394 ± 289 cm3
Last Follow-up:	736 ± 462 cm3

Lung on side of VEPTR increased:

219% ± 306% (range, 13%–1,160%)

Lung not on side of the VEPTR increased:

147% ± 176% (range, 24%–731%)

The ratio of right to left lung volume compared with a normal value of 0.85 improved by 13%

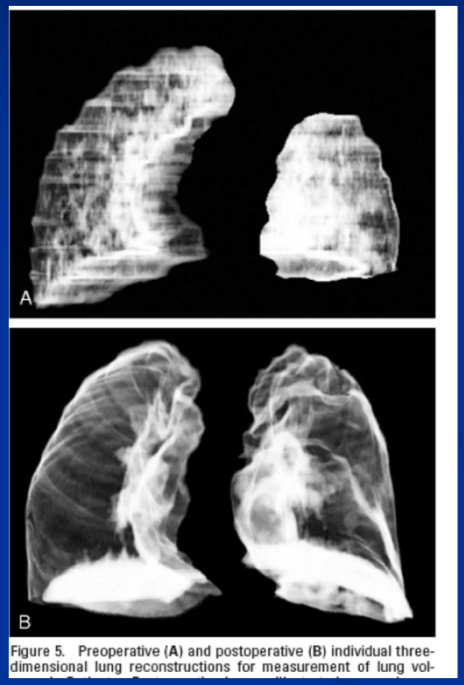
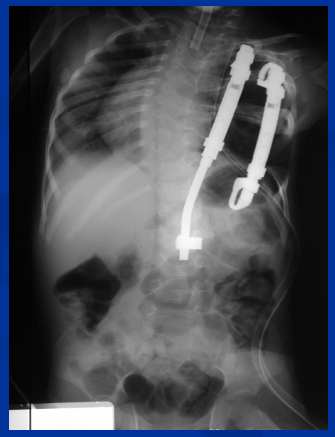


Figure 5. Preoperative (A) and postoperative (B) individual three-dimensional lung reconstructions for measurement of lung vol-



Thoracic Institute
Christus Santa Rosa Children's Hospital

How does VEPTR affect pulmonary function?: An *in vivo* assessment using the rabbit scoliosis model

H Mehta MS[1,2], B Snyder MD/PhD[1,3], A Jackson PhD[2],
S Baldassarri[4], M Hayward MD[5], M Giuffrida MD[5], J Wilson MD[5]

[1] Orthopedic Biomechanics Laboratory, Beth Israel Deaconess Medical Center, Boston, MA

[2] Department of Biomedical Engineering, Boston University, Boston, MA

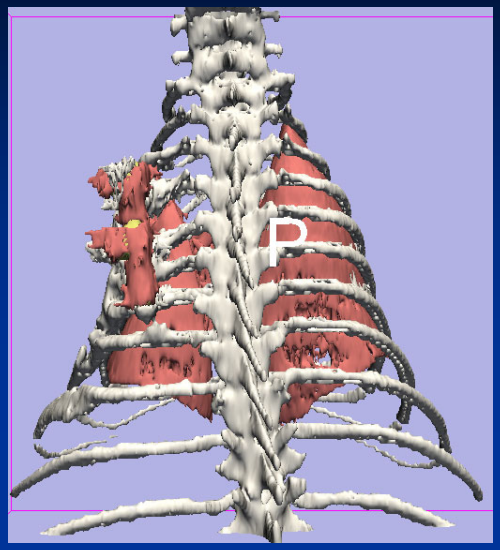
[3] Department of Orthopaedic Surgery, The Children's Hospital, Boston, MA

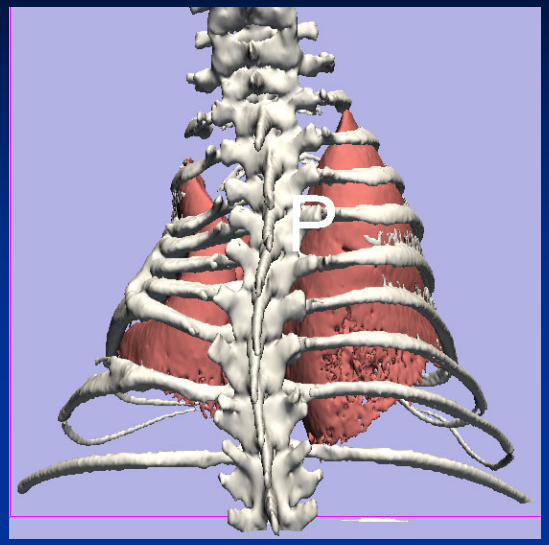
[4] School of Medicine, Boston University, Boston, MA

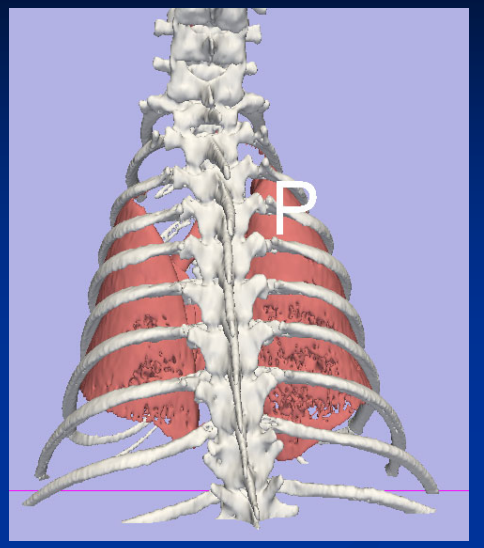
[5] Department of General Surgery, The Children's Hospital, Harvard Medical School, Boston, MA

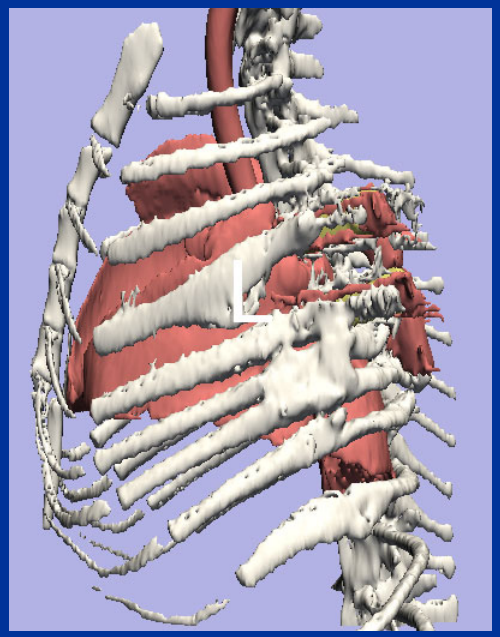
Thoracic Institute
Christus Santa Rosa Children's Hospital

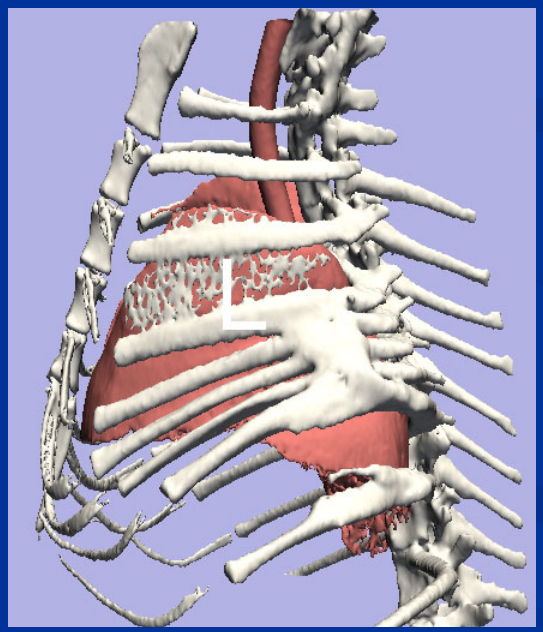
Comparison of Thoracic Cage at 18 wks

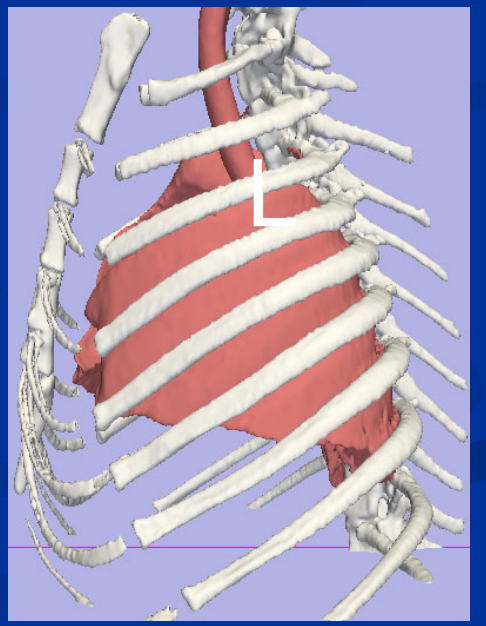






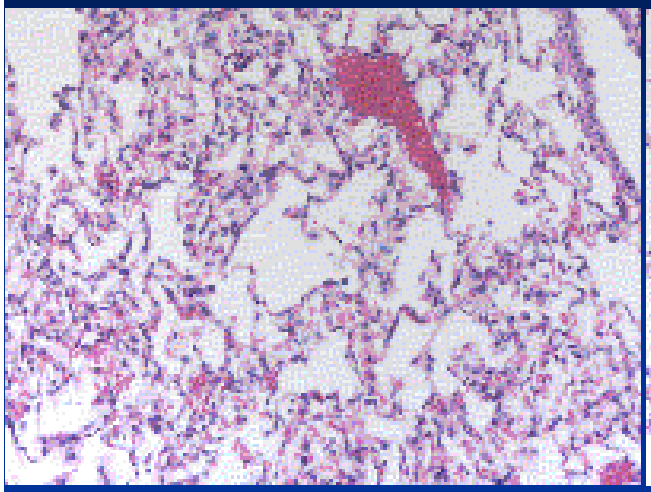
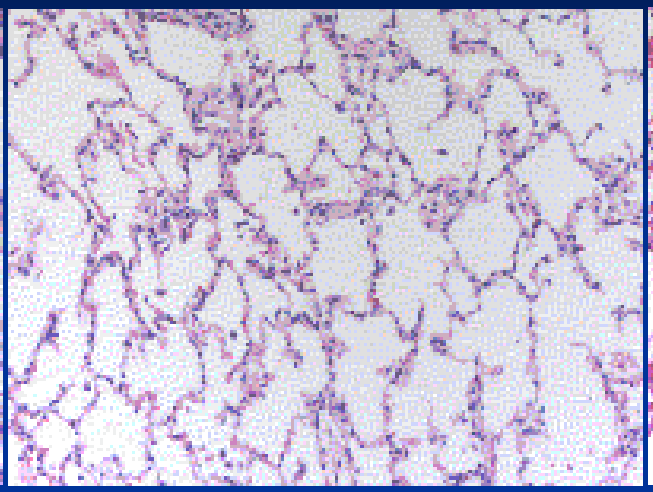
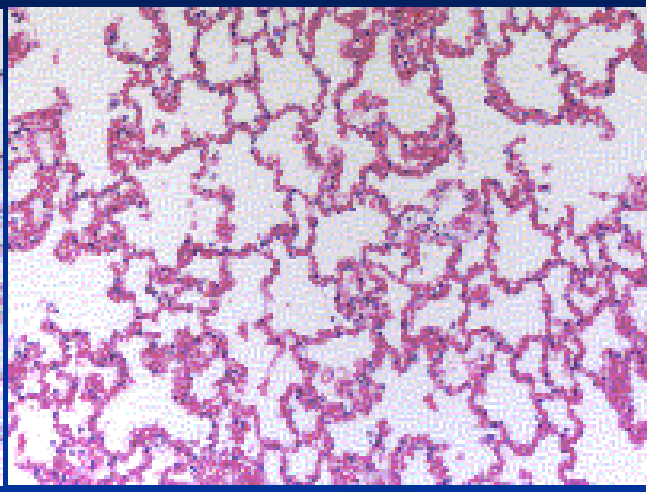
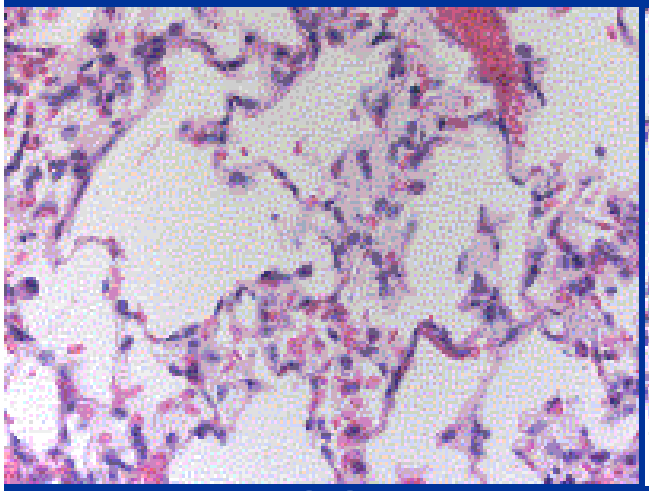
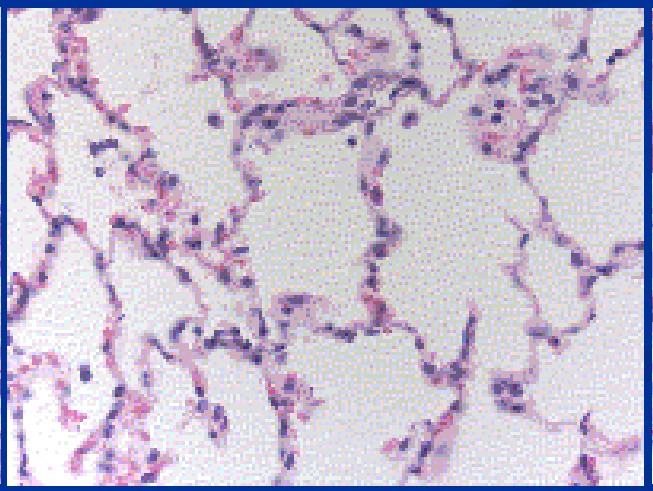
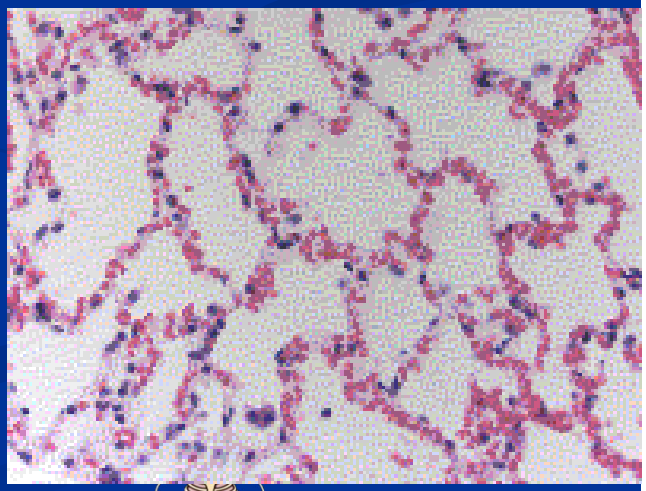






VEPTR Treated **+ Disease Control** **Normal Control**

Histology

+ Disease Control	Normal Control	VEPTR Treated
		
200x	200x	200x
		
400x	400x	400x

Courtesy Santa Rosa Children's Hospital

VEPTR Tx
Fused Ribs/Scoliosis

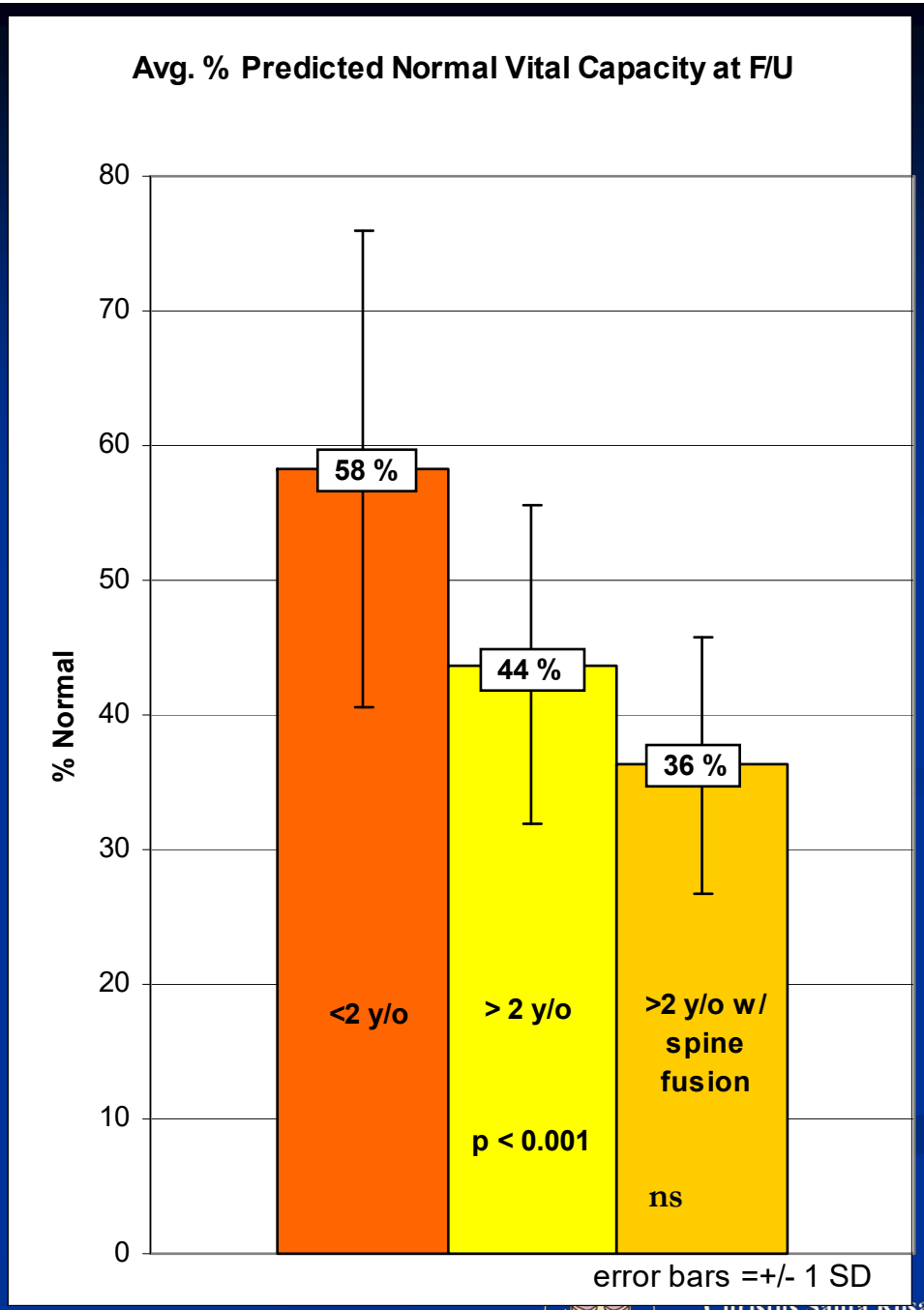
Avg. Predicted Normal Vital Capacity at follow-up:

< 2 yr

>2yr

>2 yr w fusion

- Campbell, Smith JBJS 2004



Christus Santa Rosa Children's Hospital

San Antonio VEPTR device complications
All patients (n=220) , 1989-2004

	San Antonio	Klemme et al.	Tello
f/u	6 yrs	3.1 yrs	4.75 yrs
Migration index	0.09 mig/yr	0.1 mig/yr.	0.029 mig/yr
Percentage pts	27%	31%	14%
Time	3.2 yrs	?	?
Infect rate/proc	3.3%	1.5%	5.3%
Skin slough	8.5%	4.5%	13.6%

- JBJS, 2007

Thoracic Institute
Christus Santa Rosa Children's Hospital

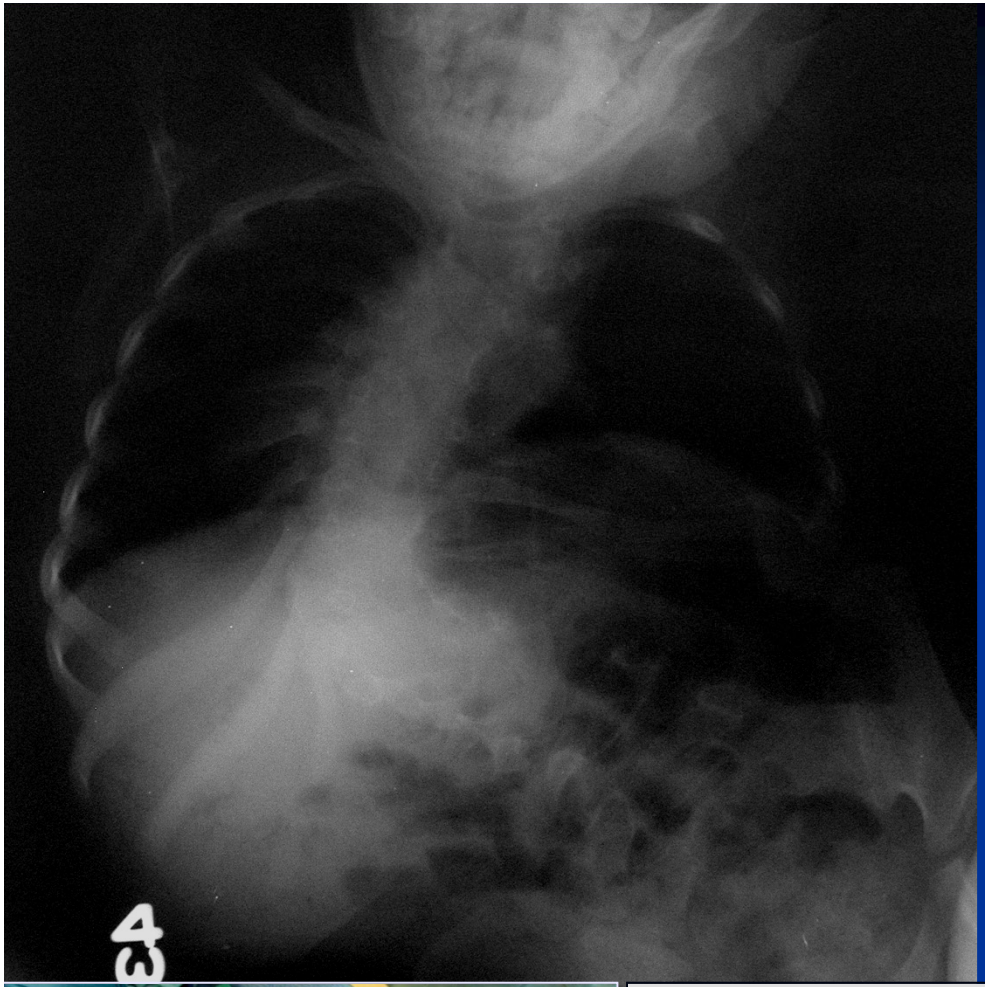
SA VEPTR Neurological Problems 1987-2007

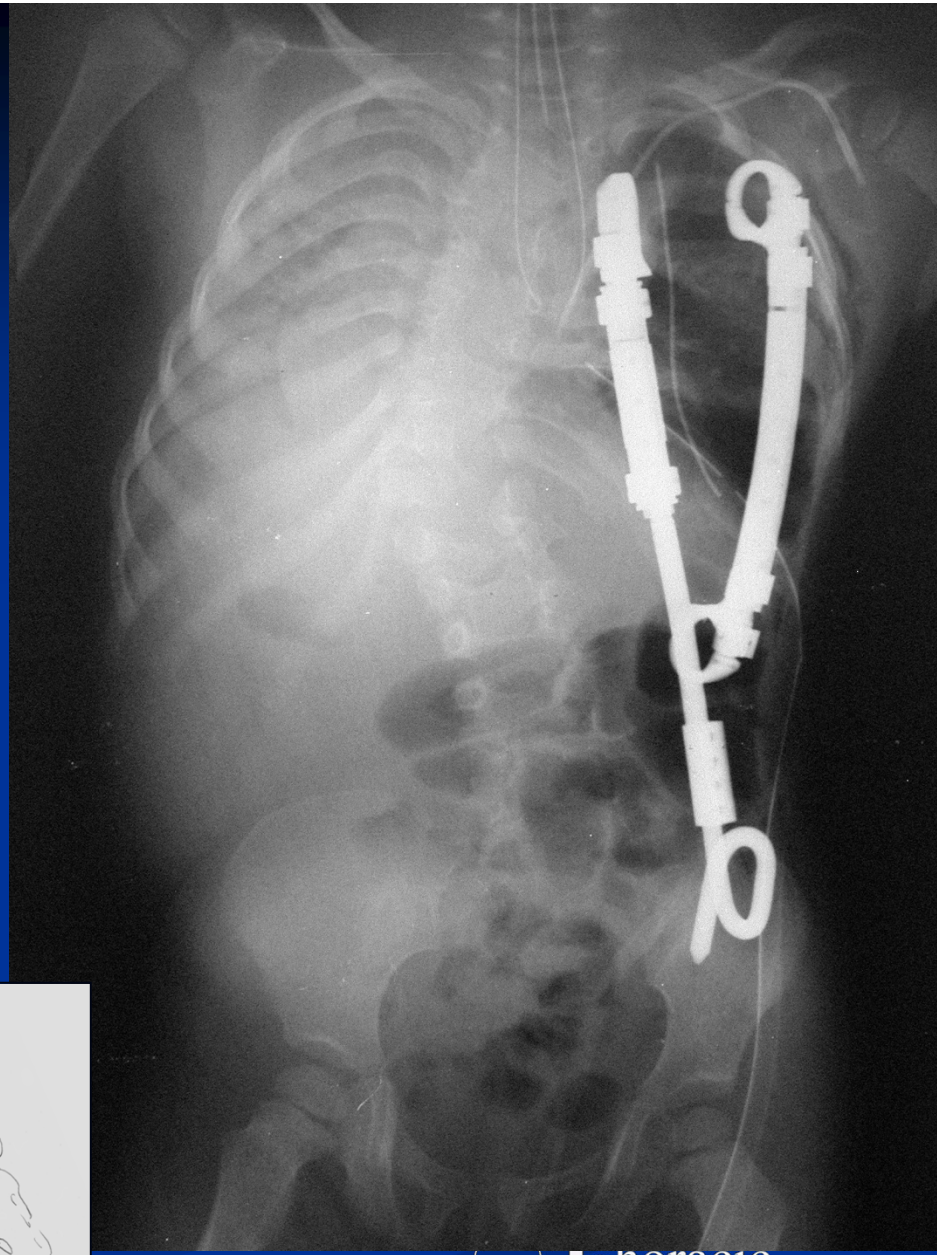
n > 270

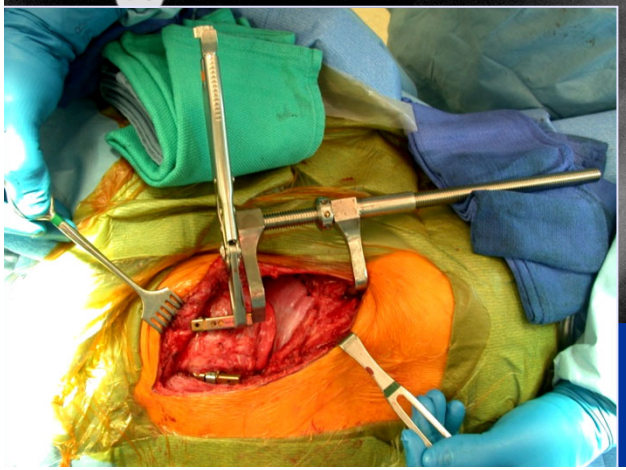
- UE's
  - Handful of transient brachial plexopathies
- LE's
  - 1 pt: Monoplegia, transient, canal violation
  - 1 pt: Severe Congenital kyphosis, paraplegia, traction injury
  - 1 Pt: Transient "paraplegia", VP shunt problem ?

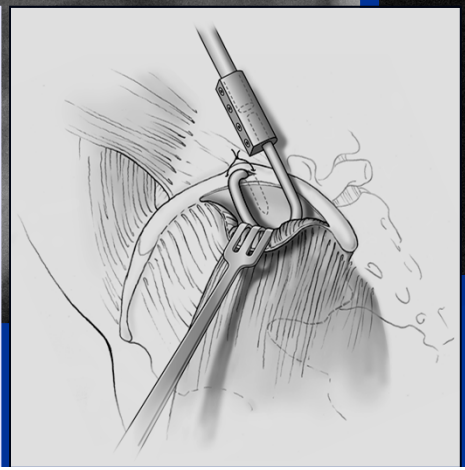
(predicted neurologic injury in this population - 10%
-personal communication, Dean MacEwen)

Thoracic Institute
Christus Santa Rosa Children's Hospital

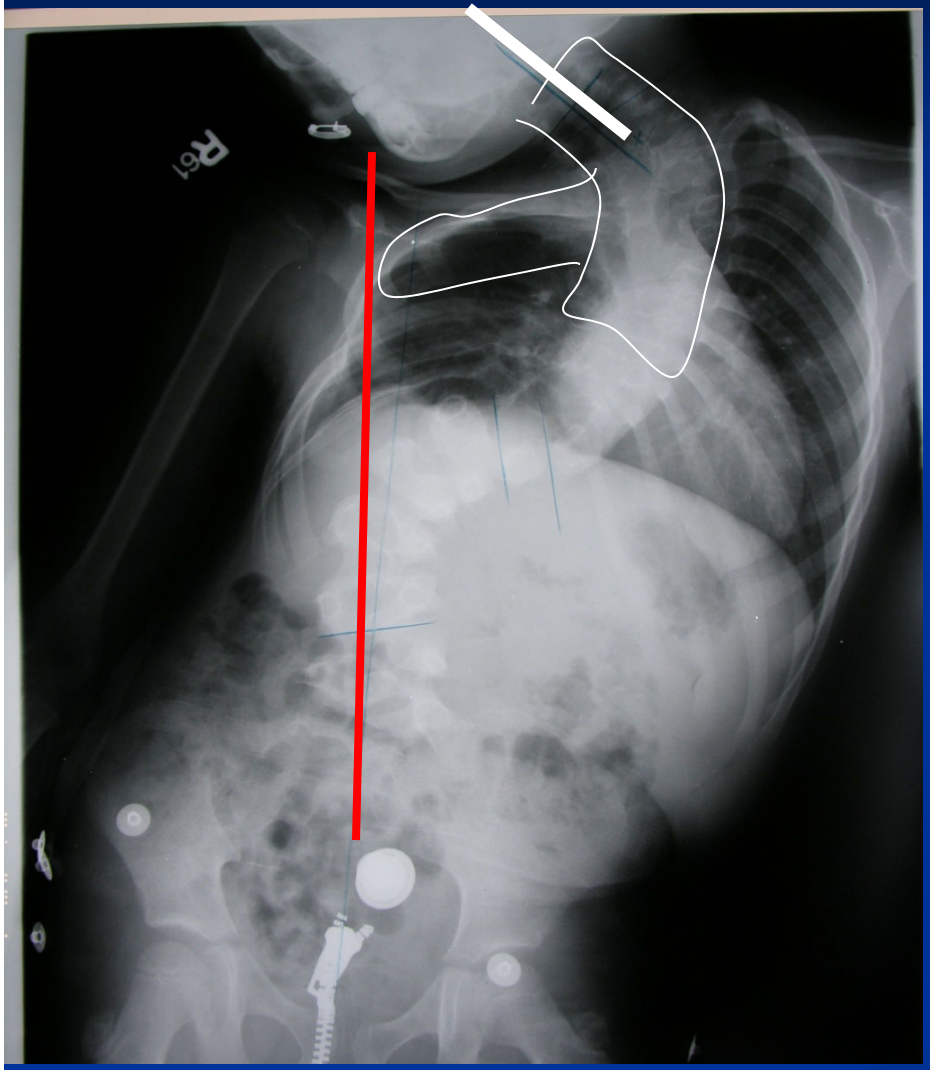


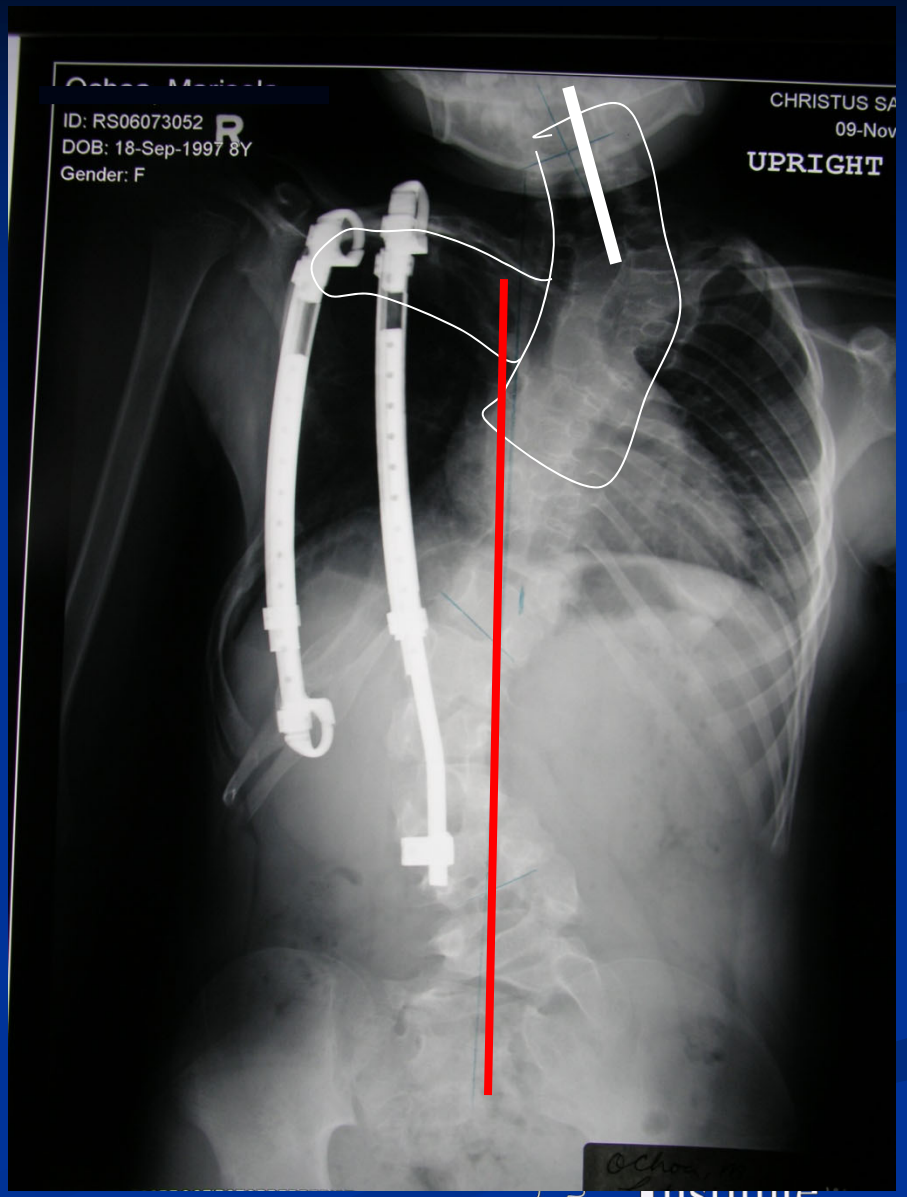






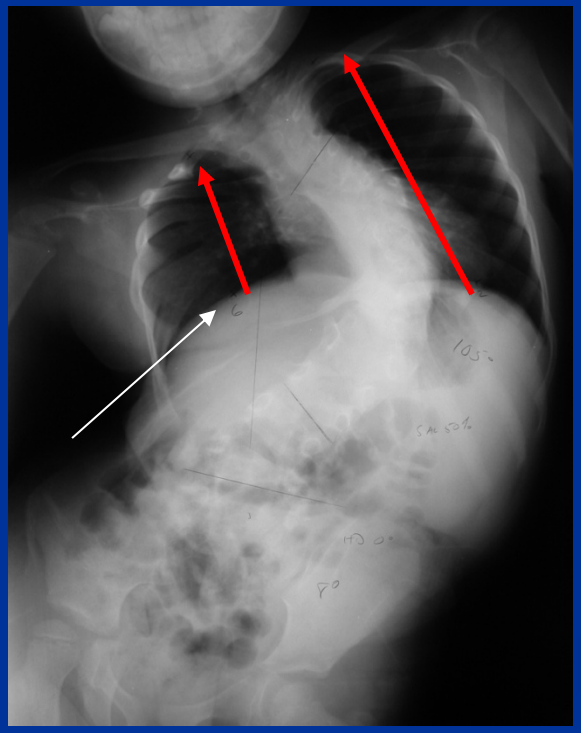
Thoracic Institute
Christus Santa Rosa
Children's Hospital

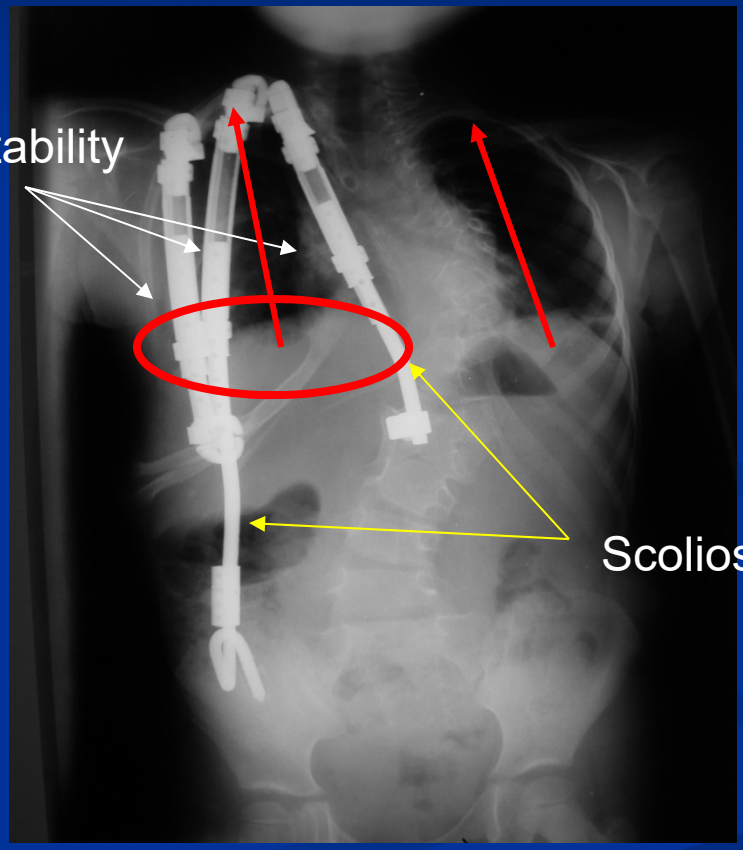




Institute
Christus Santa Rosa
Children's Hospital

VEPTR Procedure for Type I VDD

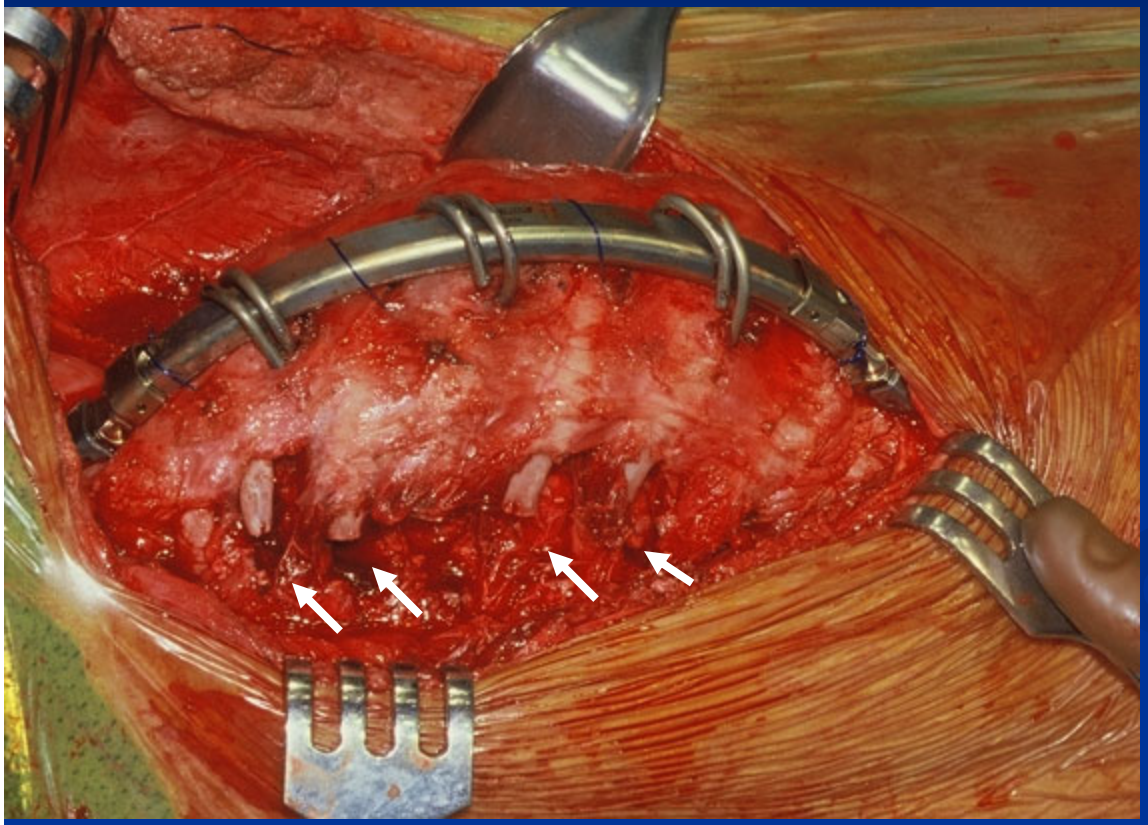




Thoracic Institute
Christus Santa Rosa Children's Hospital

VEPTR for Type III a VDD: Jarcho-Levin Syndrome

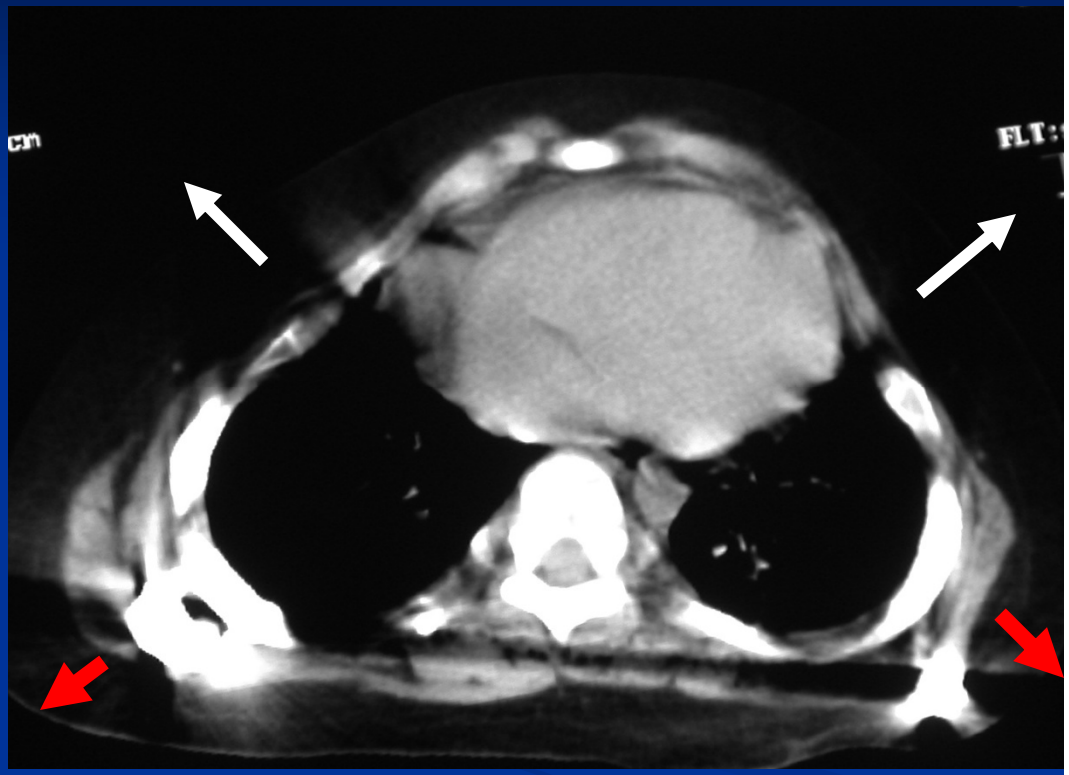
Jeune's Syndrome



Staged Procedures

Christus Santa Rosa
Children's Hospital

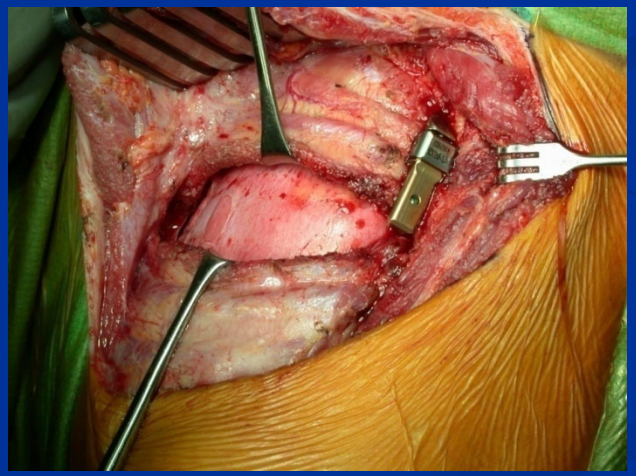
Increased Thoracic Volume and Improved Thoracic Symmetry

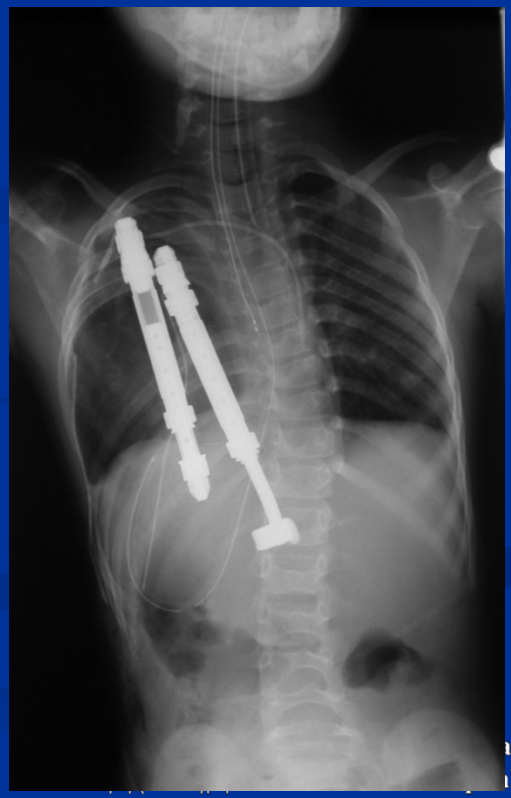


4 Year f/u

Thoracic Institute
Christus Santa Rosa Children's Hospital

Type IIIb VDD:
Early Onset scoliosis with chest wall constriction syndrome



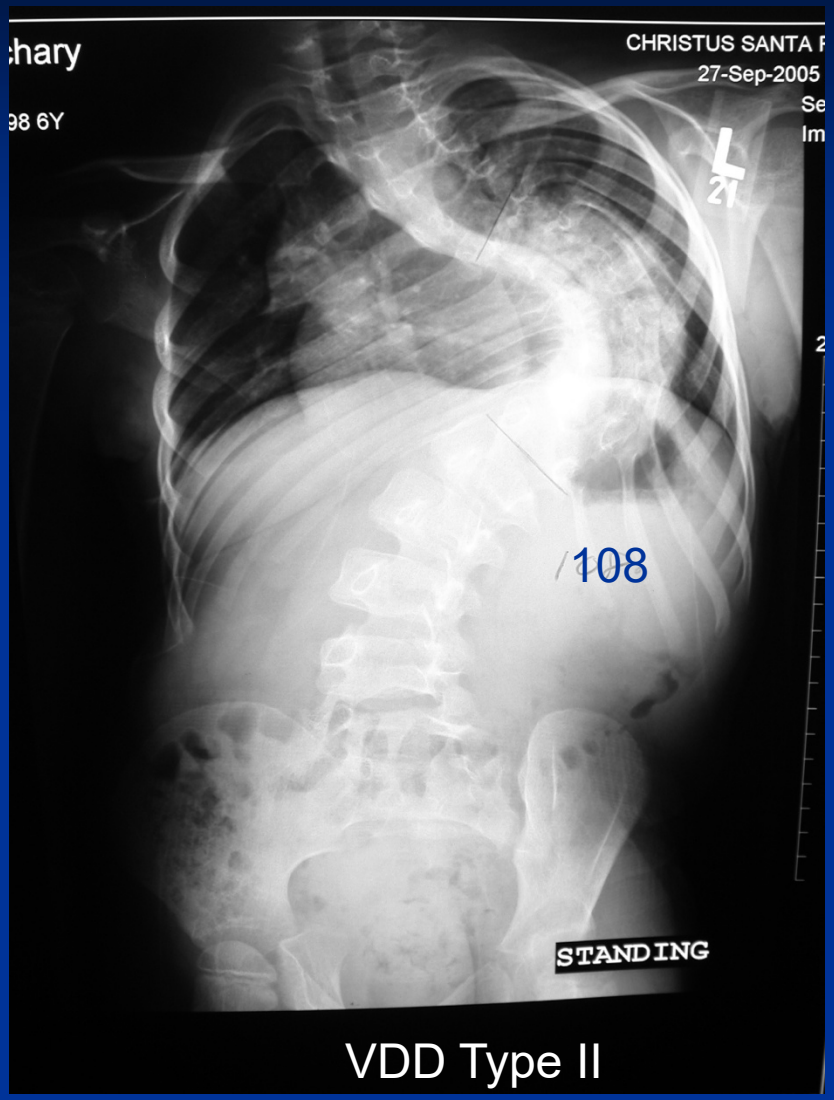


Percutaneous Bilateral Rib to Pelvis VEPTR without thoracostomy

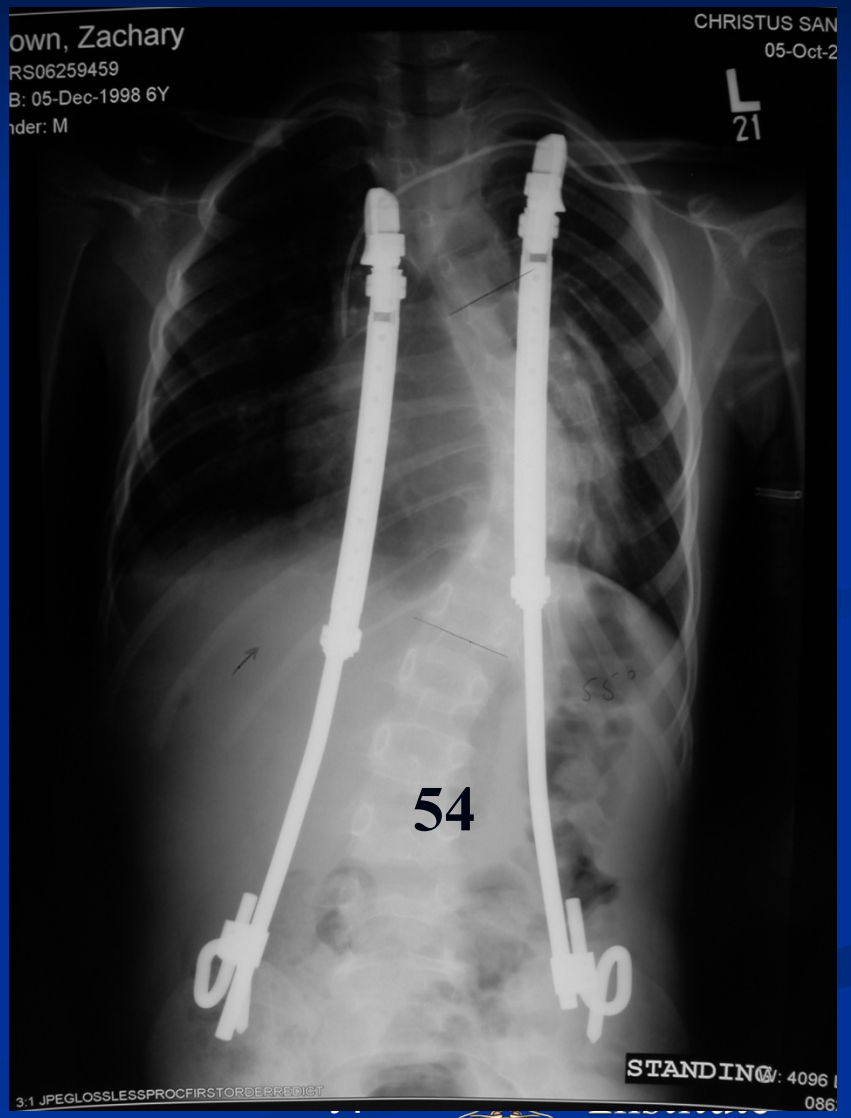
-John Smith, MD , Utah

Thoracic Institute
Christus Santa Rosa Children's Hospital

Hybrid Approach

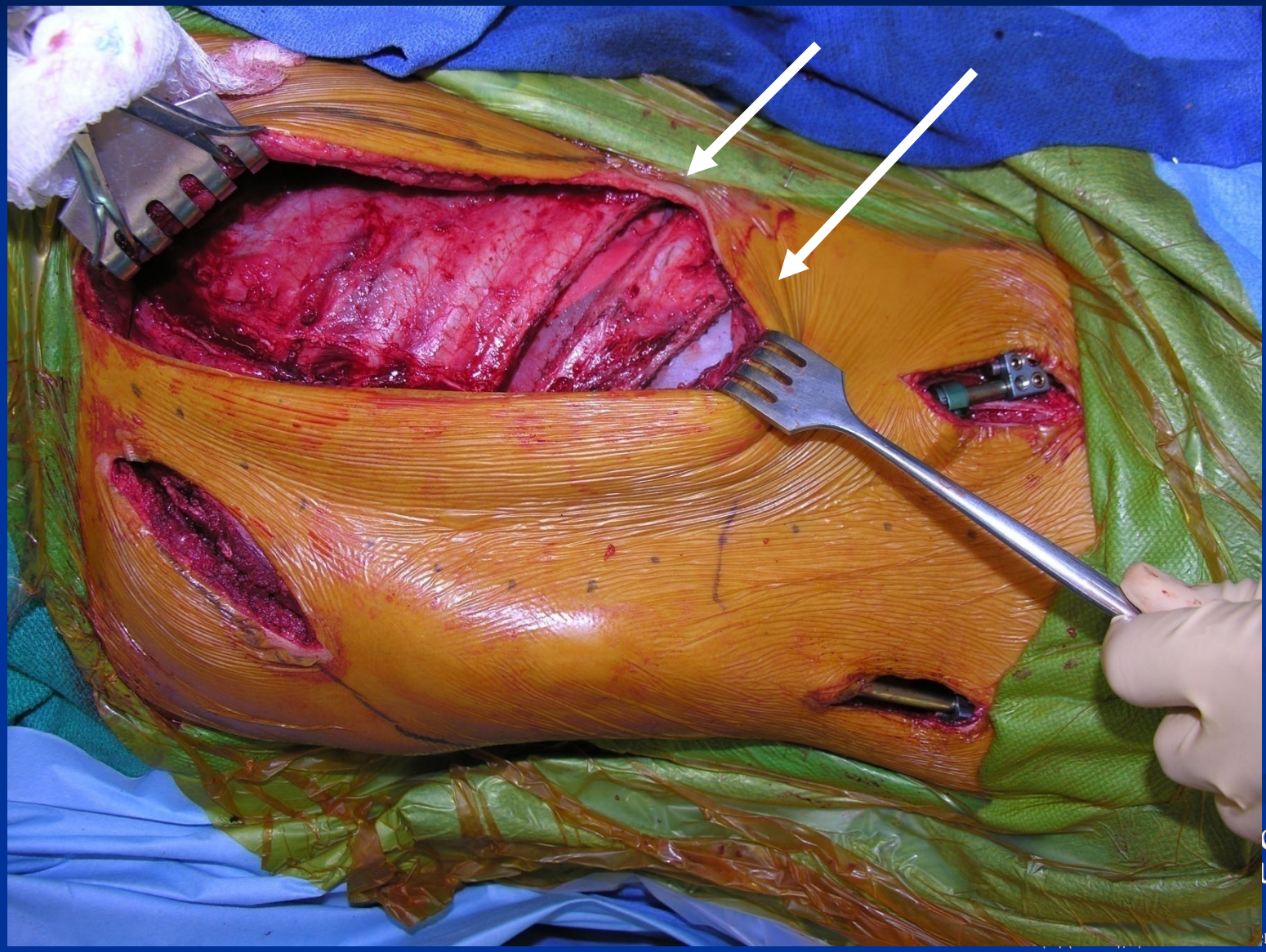


VDD Type II



Christus Santa Rosa
Children's Hospital

Hybrid EOS Approach

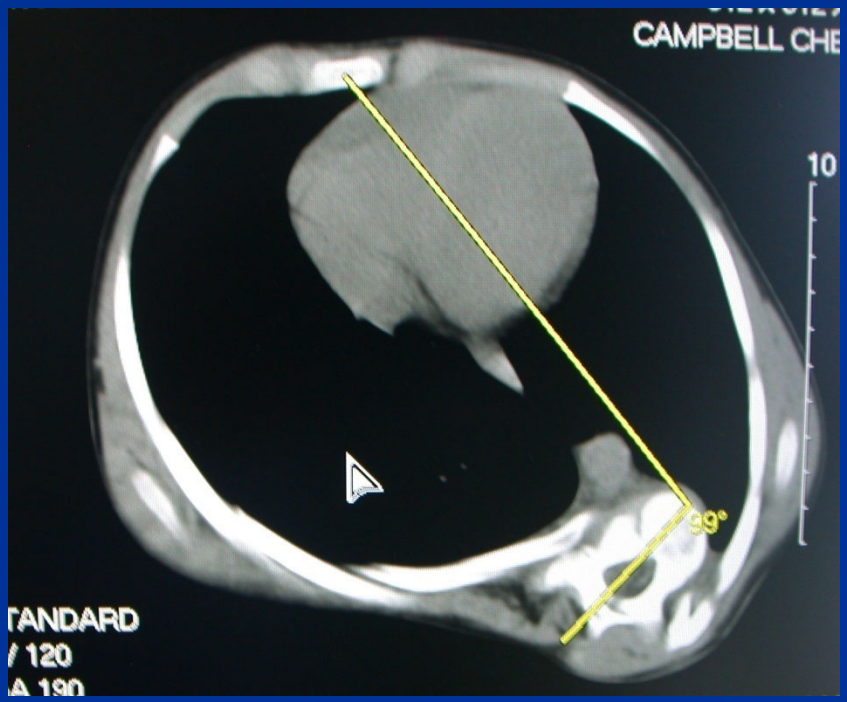


cic
ute
Santa Rosa
n's Hospital

Thoracic Rotation

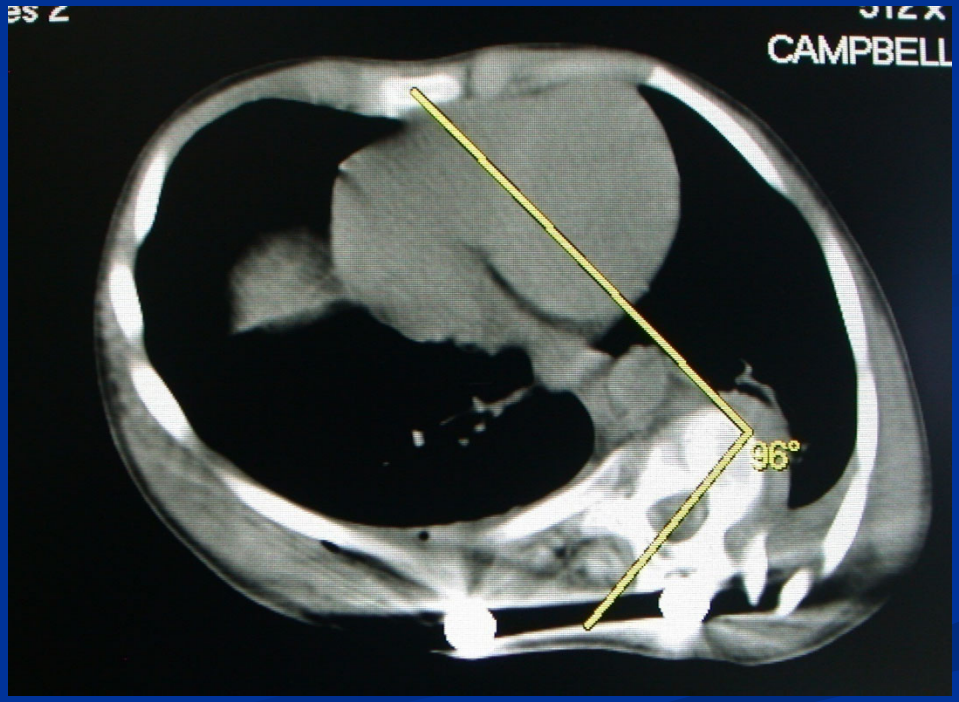
Pre op

99

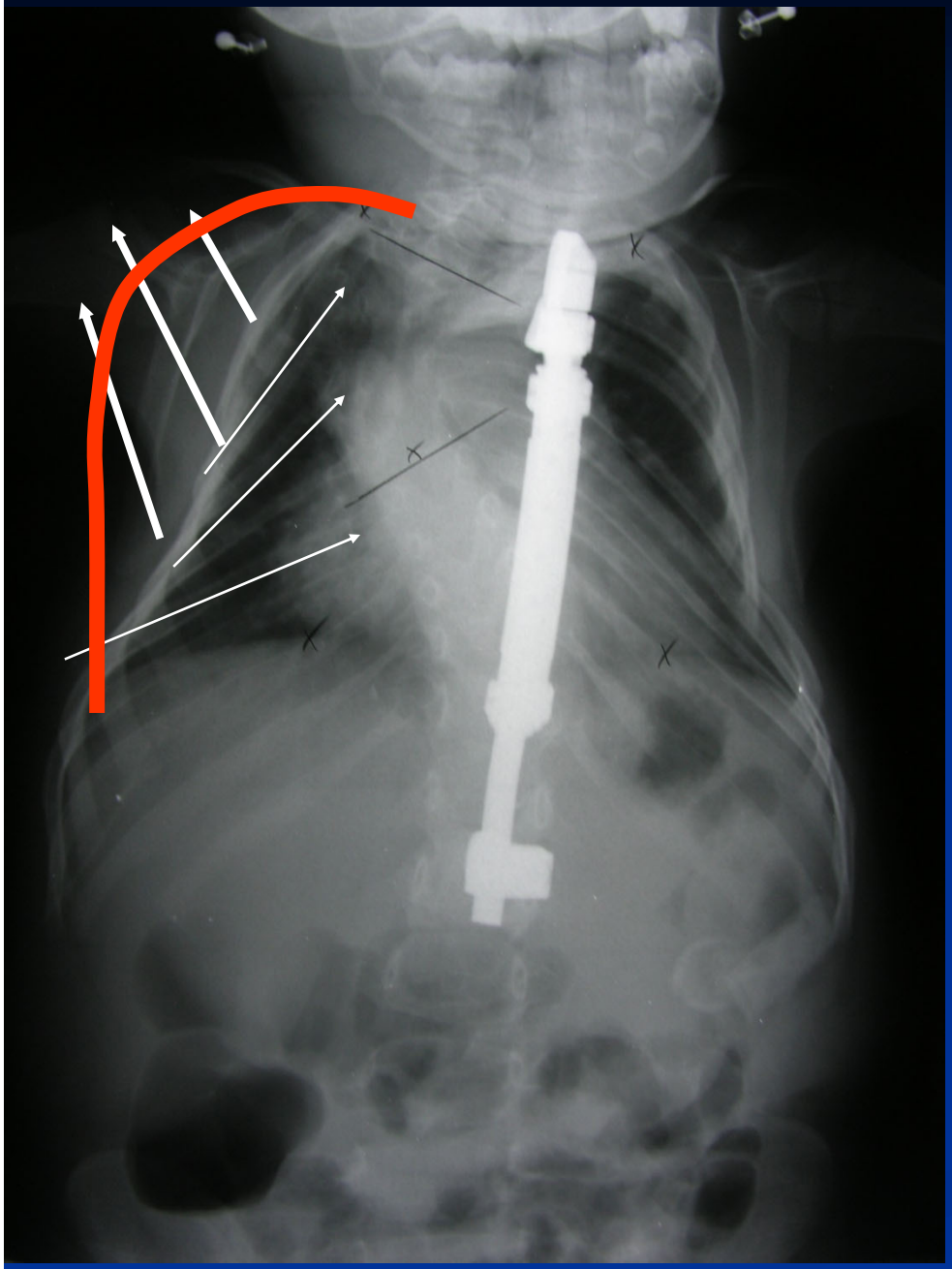


Post op

96



Thoracic Institute
Christus Santa Rosa
Children's Hospital

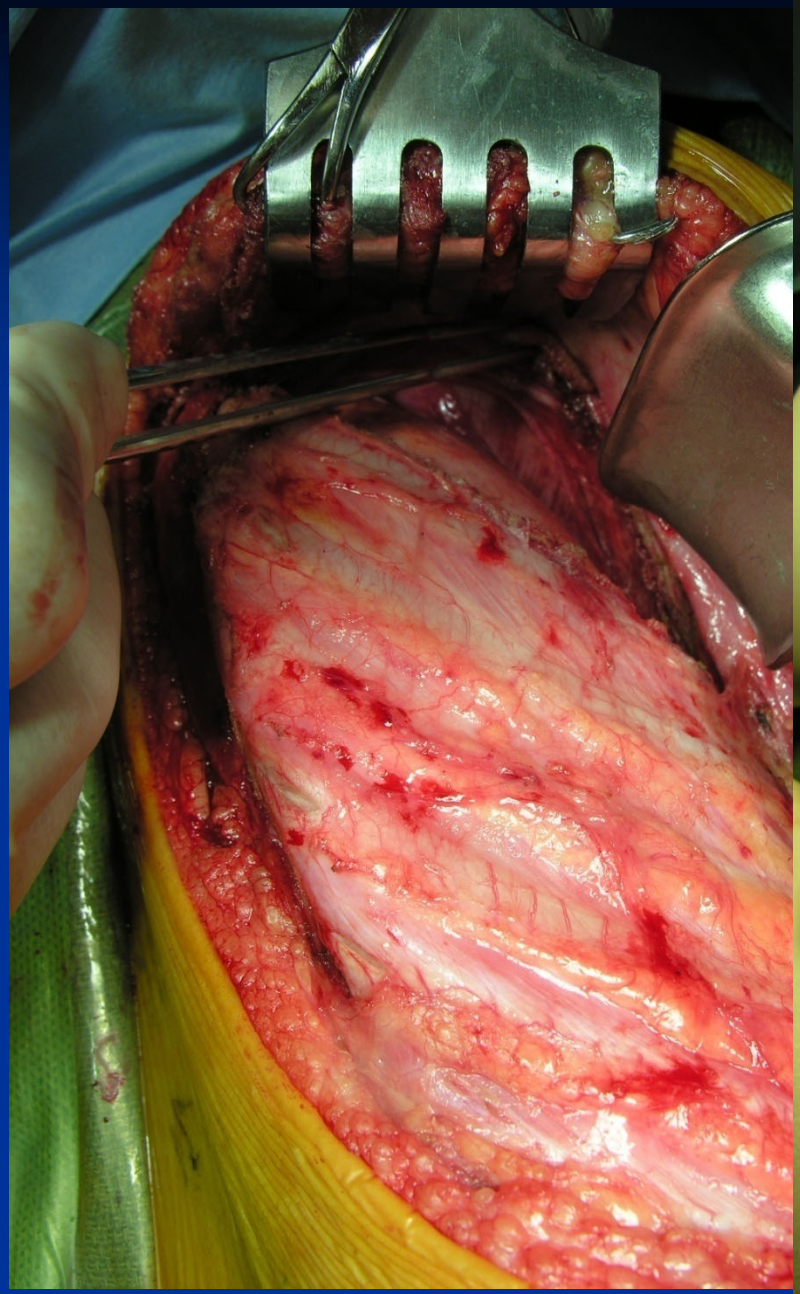


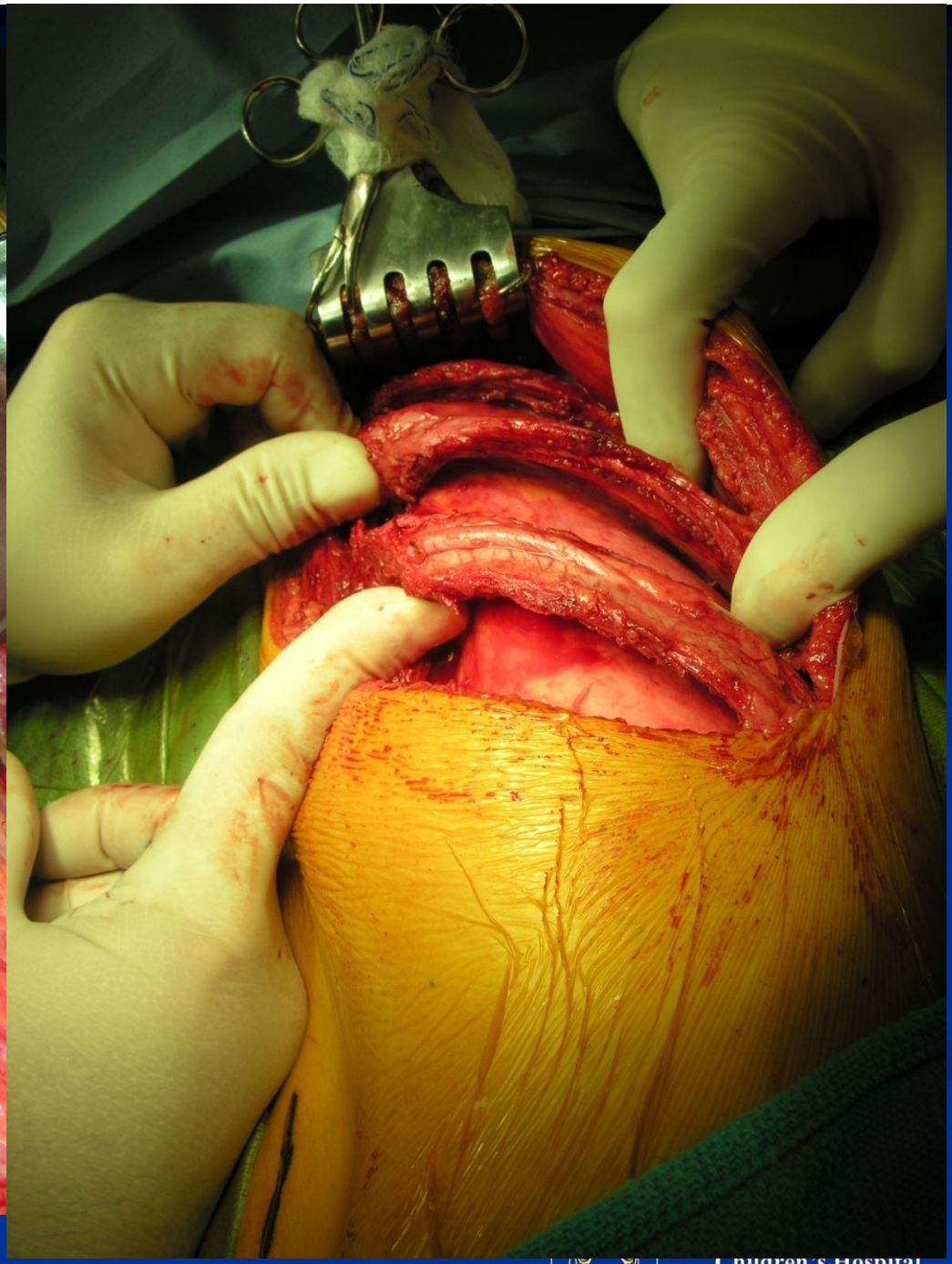
The VEPTR parasol procedure

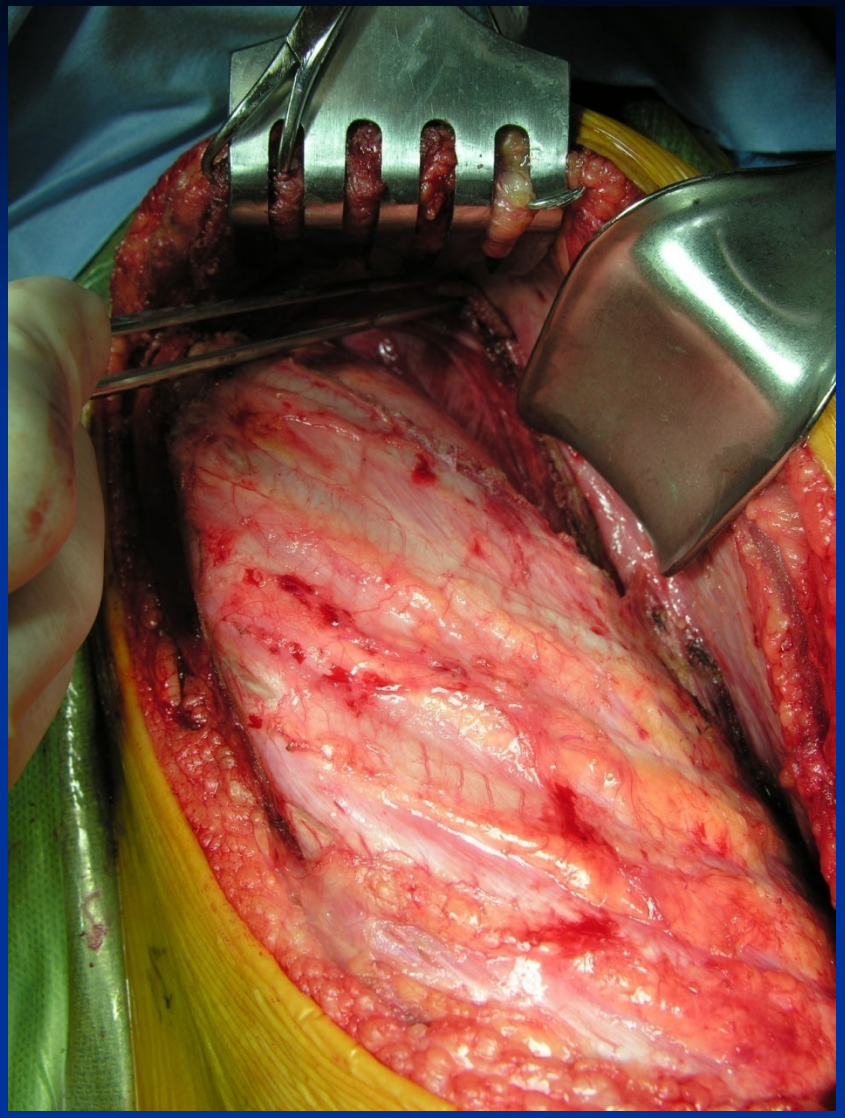
Thoracic Institute
Christus Santa Rosa Children's Hospital

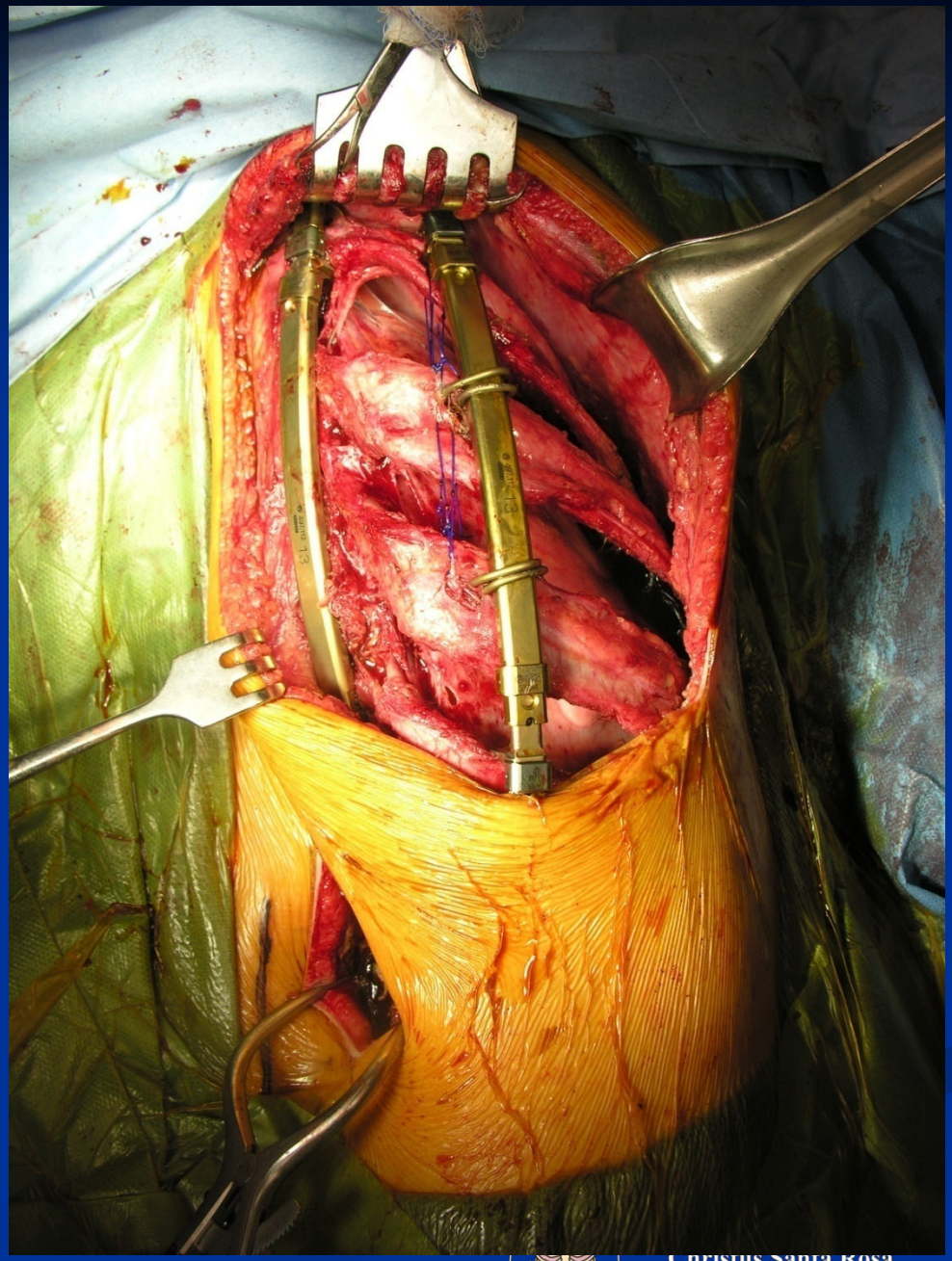
The Dimeglio Parasol Analogy

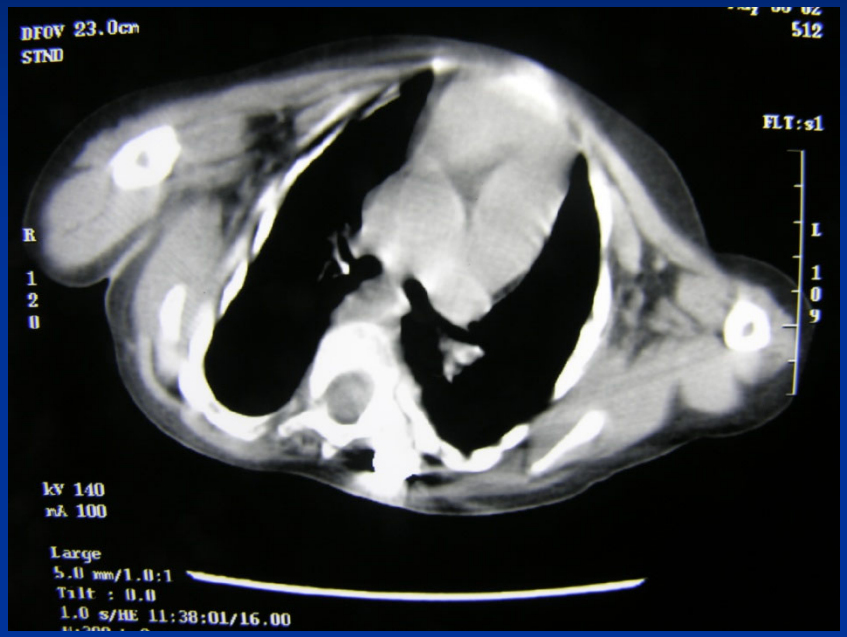
Institute
Christus Santa Rosa
Children's Hospital

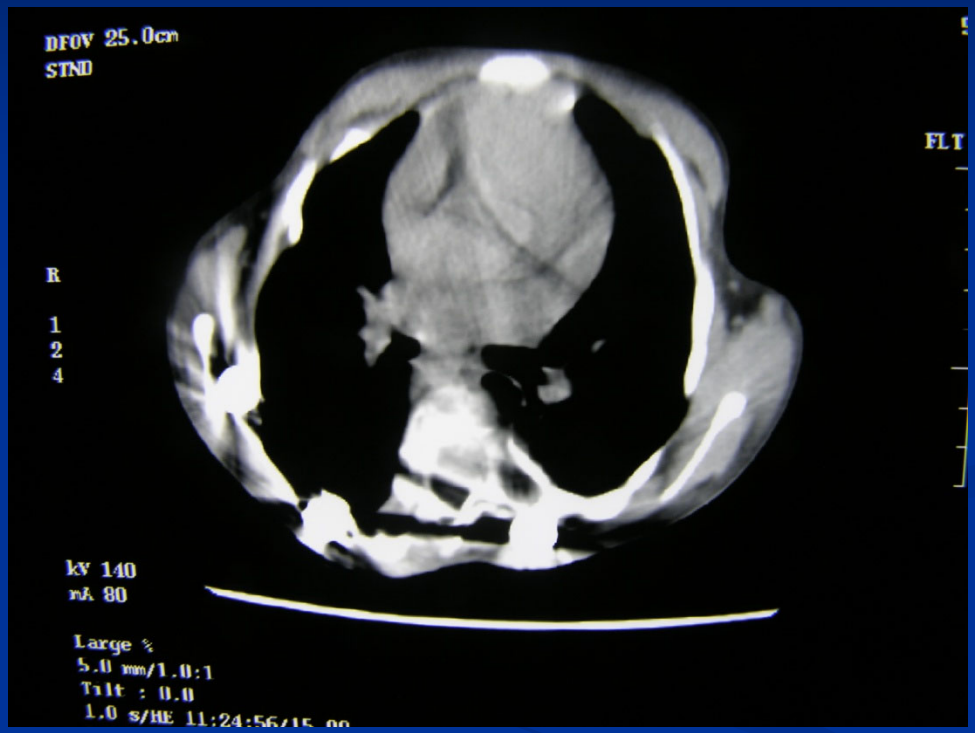












Thoracic Institute
Christus Santa Rosa
Children's Hospital

VEPTR and Myelomeningocele Flexible Lumbar Kyphosis : Five pts

"Eiffel Tower" Construct

-Secondary thoracic insufficiency

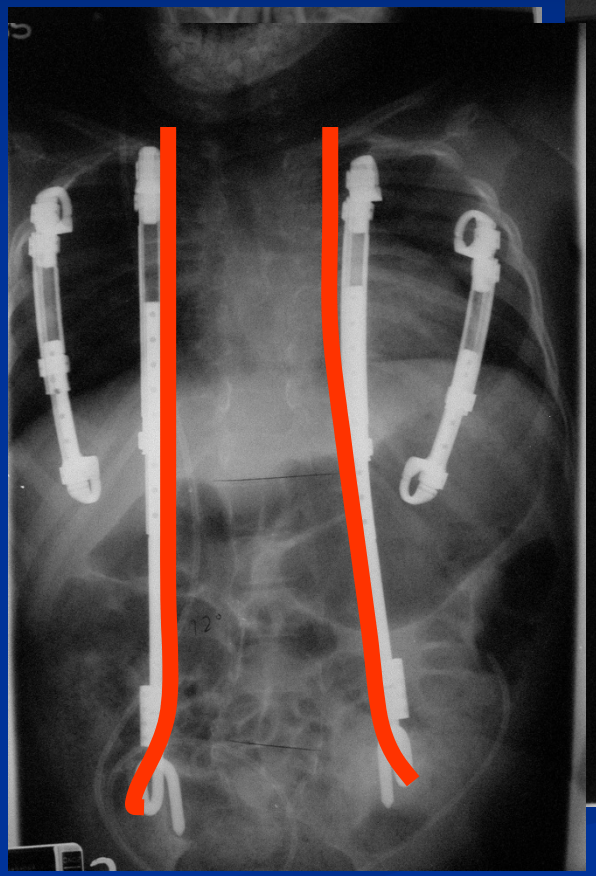
-lumbar kyphosis of 65°

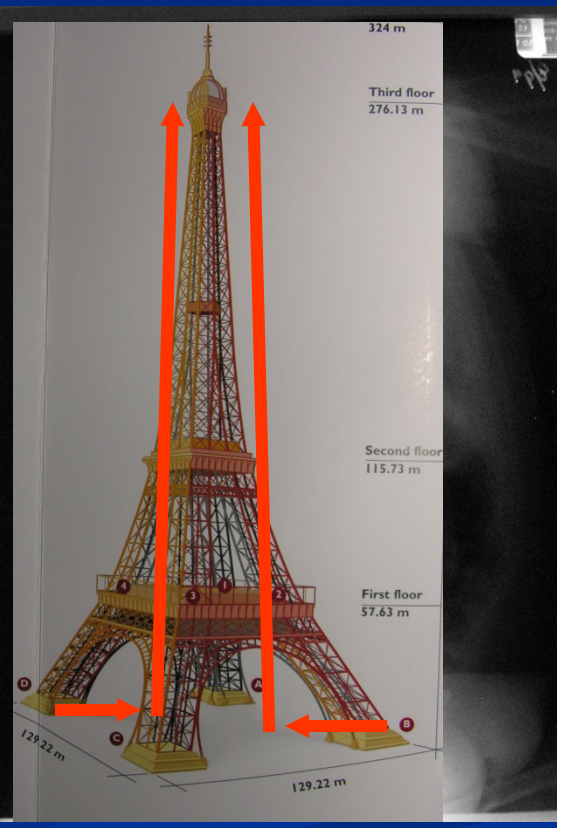
- All resolved their marionette signs with hybrid treatment

-decrease of the kyphosis to 42°.

-1 pt stable lumbar kyphosis of 24°.

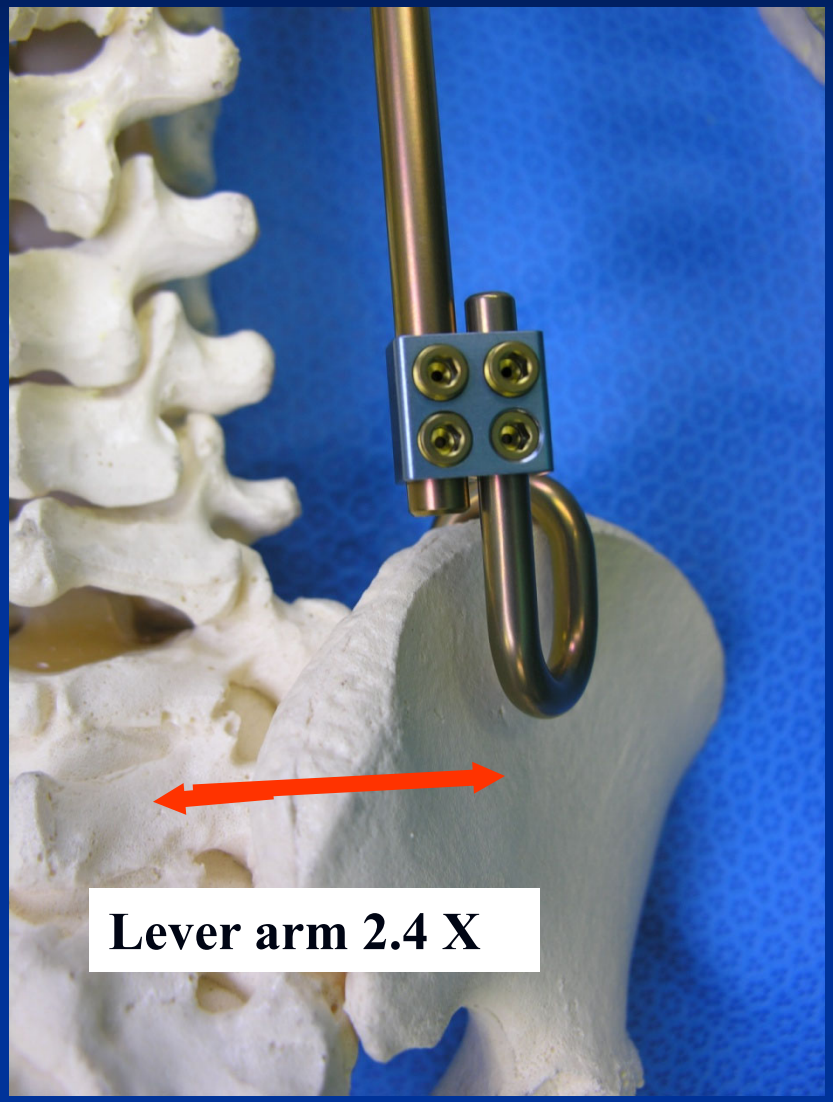
-1 pt , preop 20° , 32° f/u





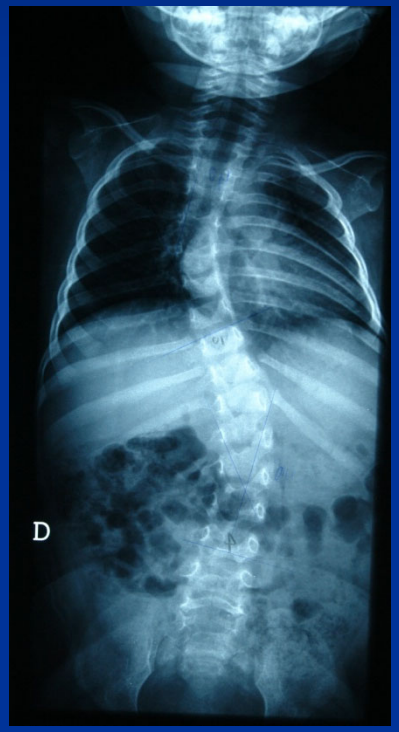
Thoracic Institute
Christus Santa Rosa Children's Hospital

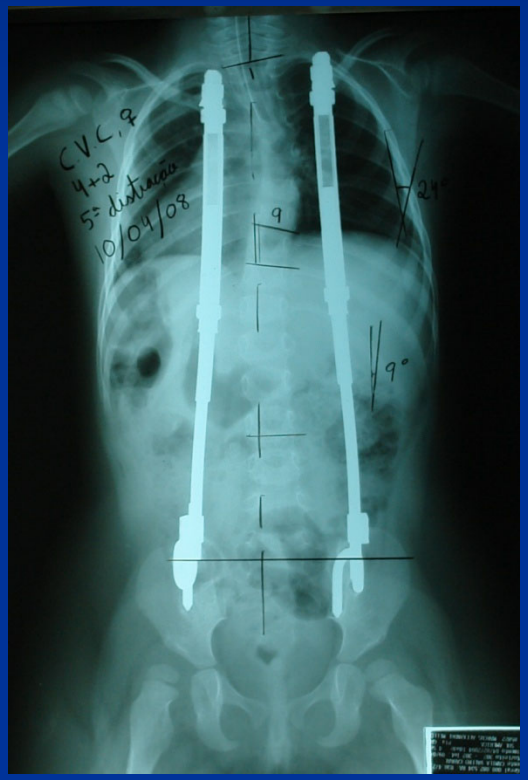
3 cm safe zone
Unilateral S hook : 7.9 mm/yr
Bilateral S hook : 8.4 mm/yr

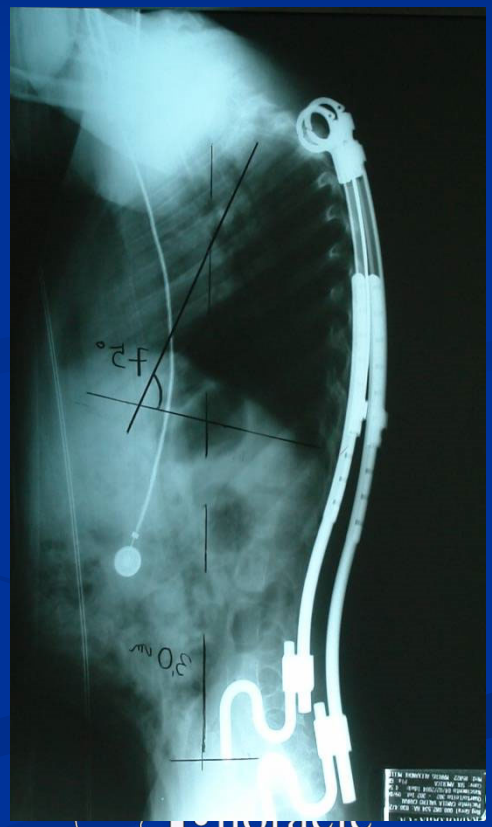


PRADER WILLI
(Paul Cavali)

2 yr f/u





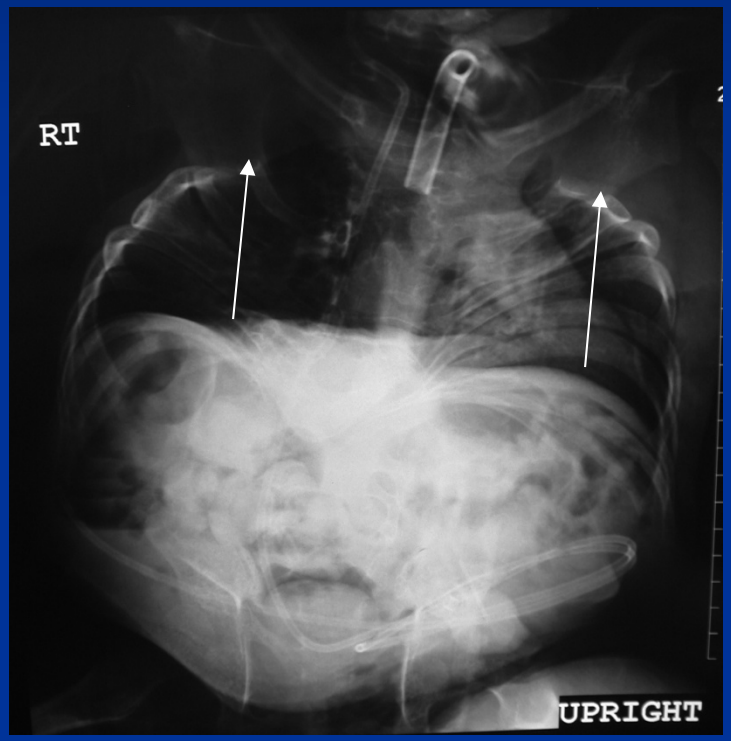


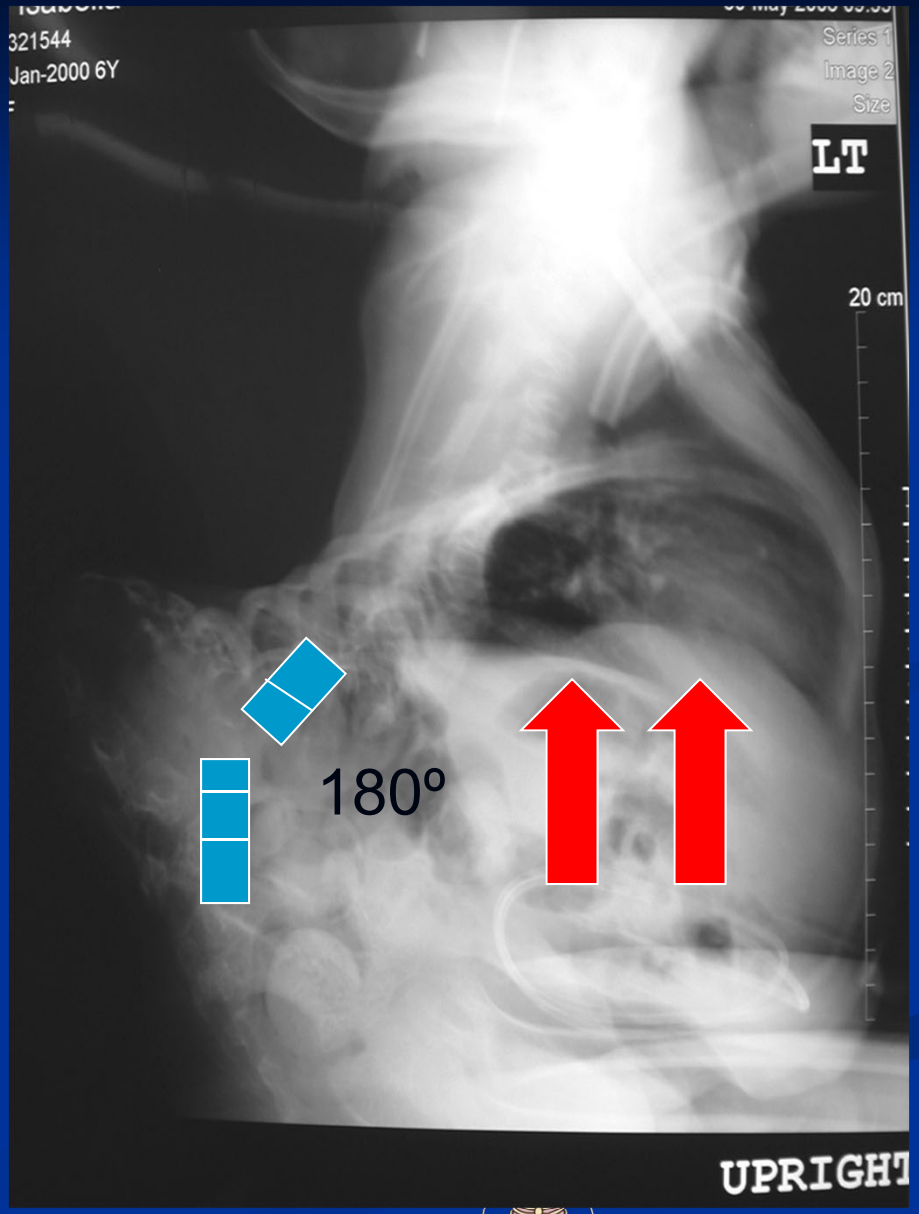
19 m /o

Institute
Christus Santa Rosa
Children's Hospital

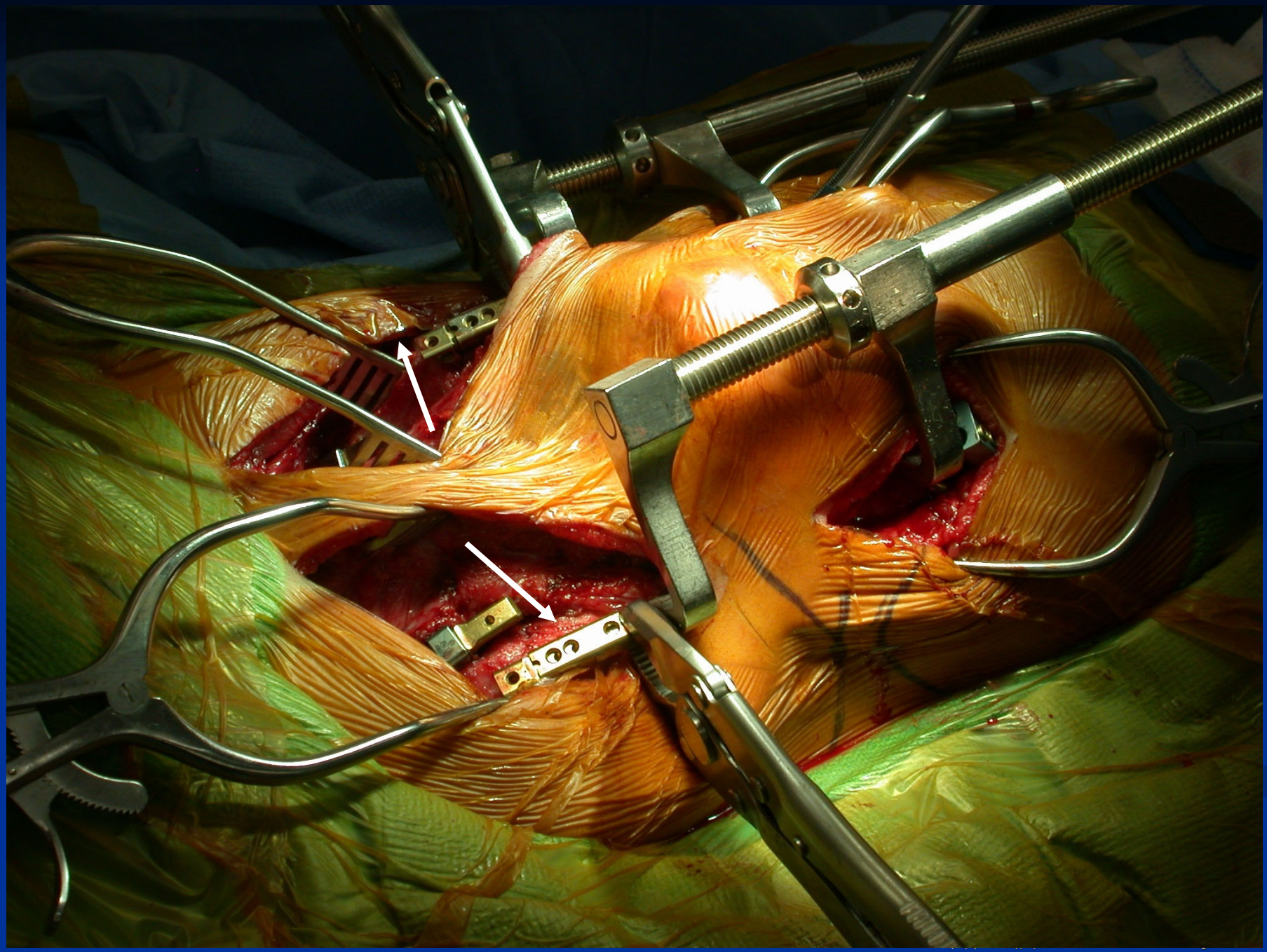
Myelominigocele Gibbus Treatment

6 y/o myelominingocele ventilator dependant

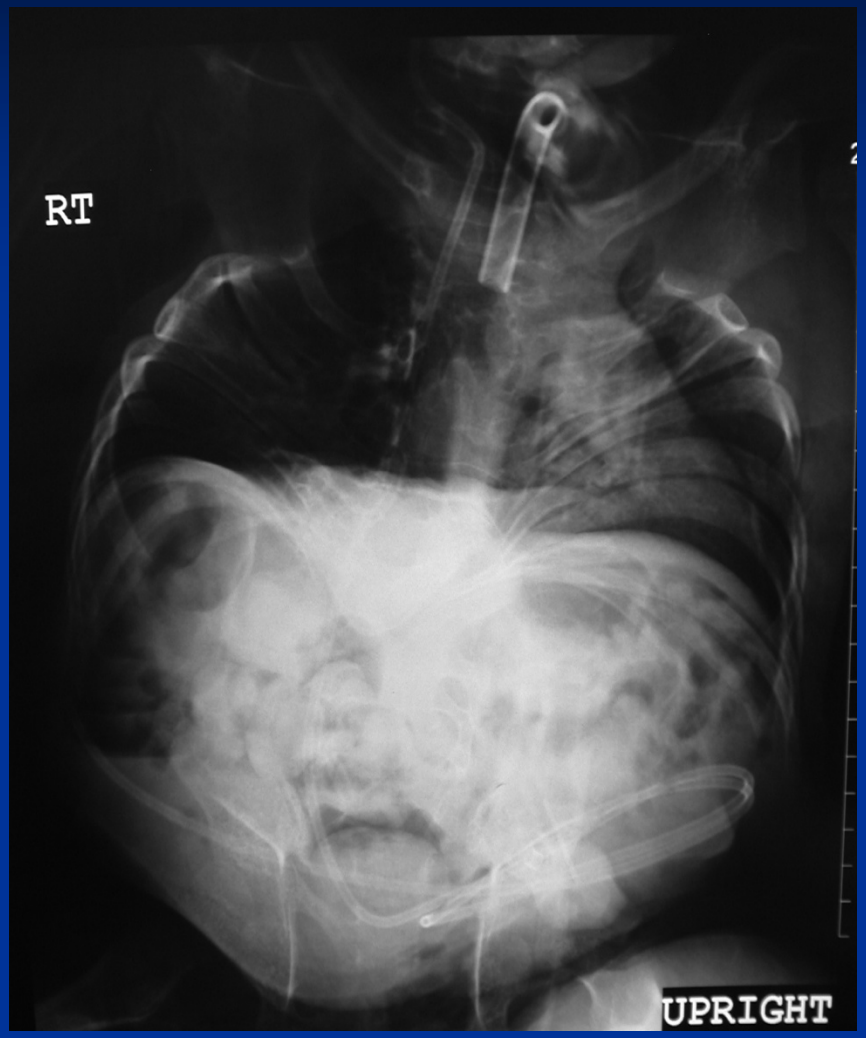


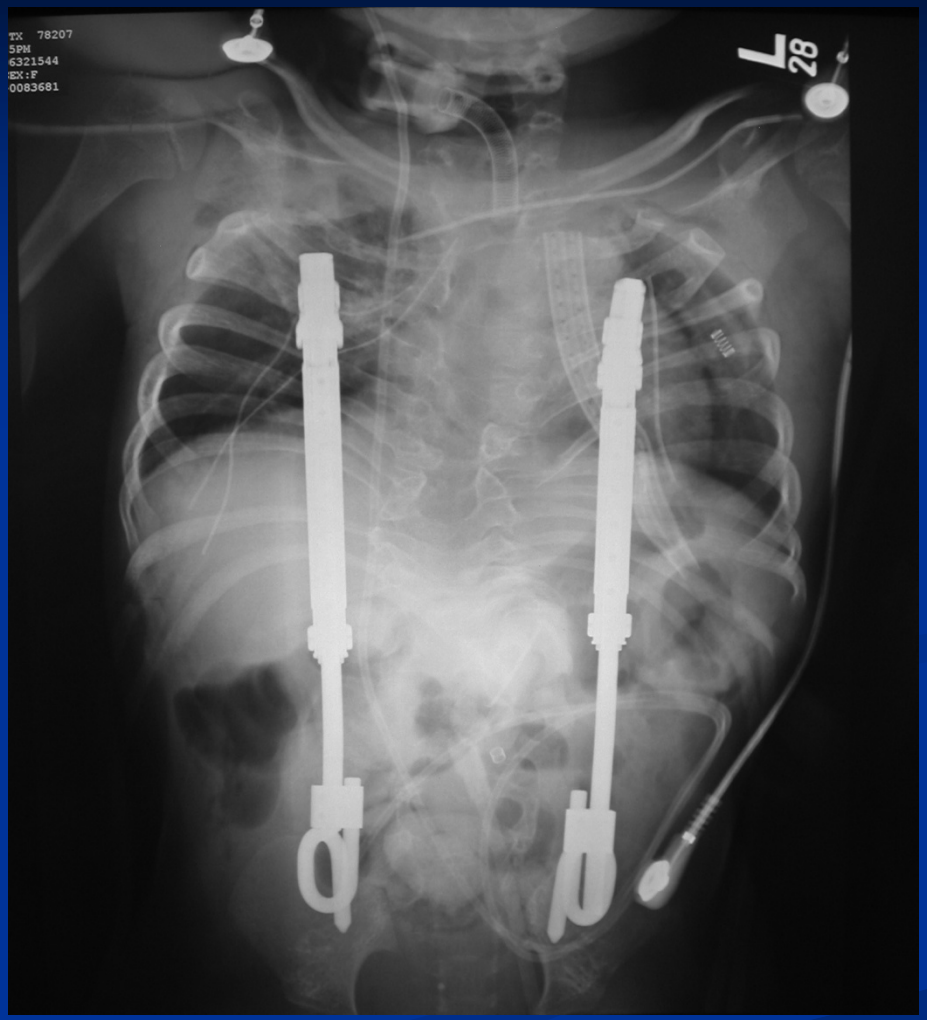


Christus Santa Rosa
Children's Hospital



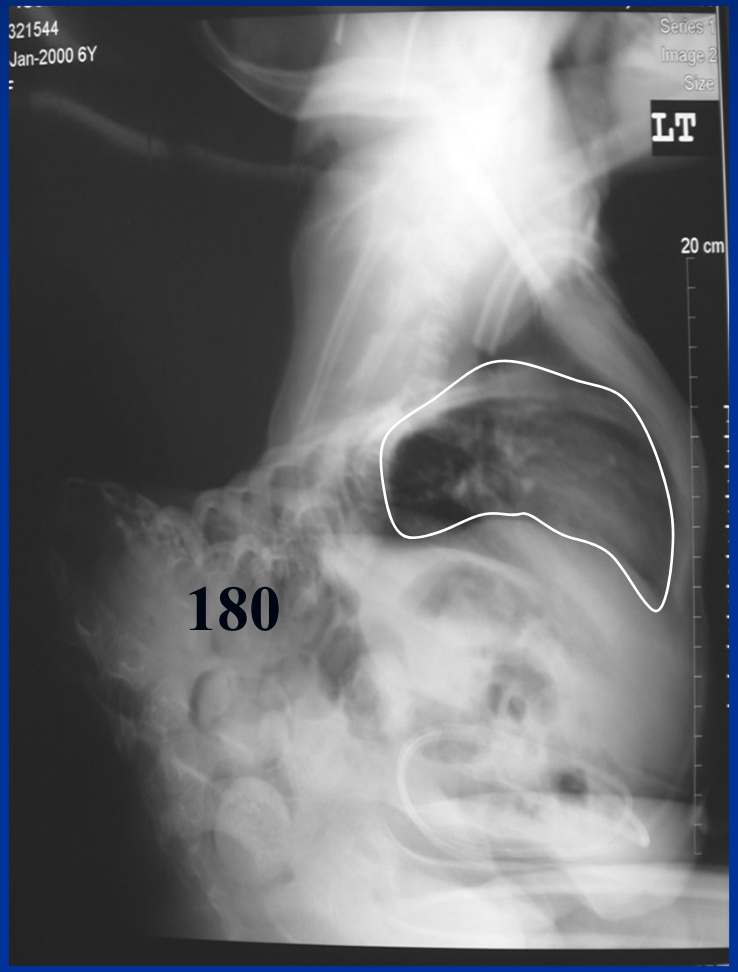
Thoracic Institute
Christus Santa Rosa
Children's Hospital

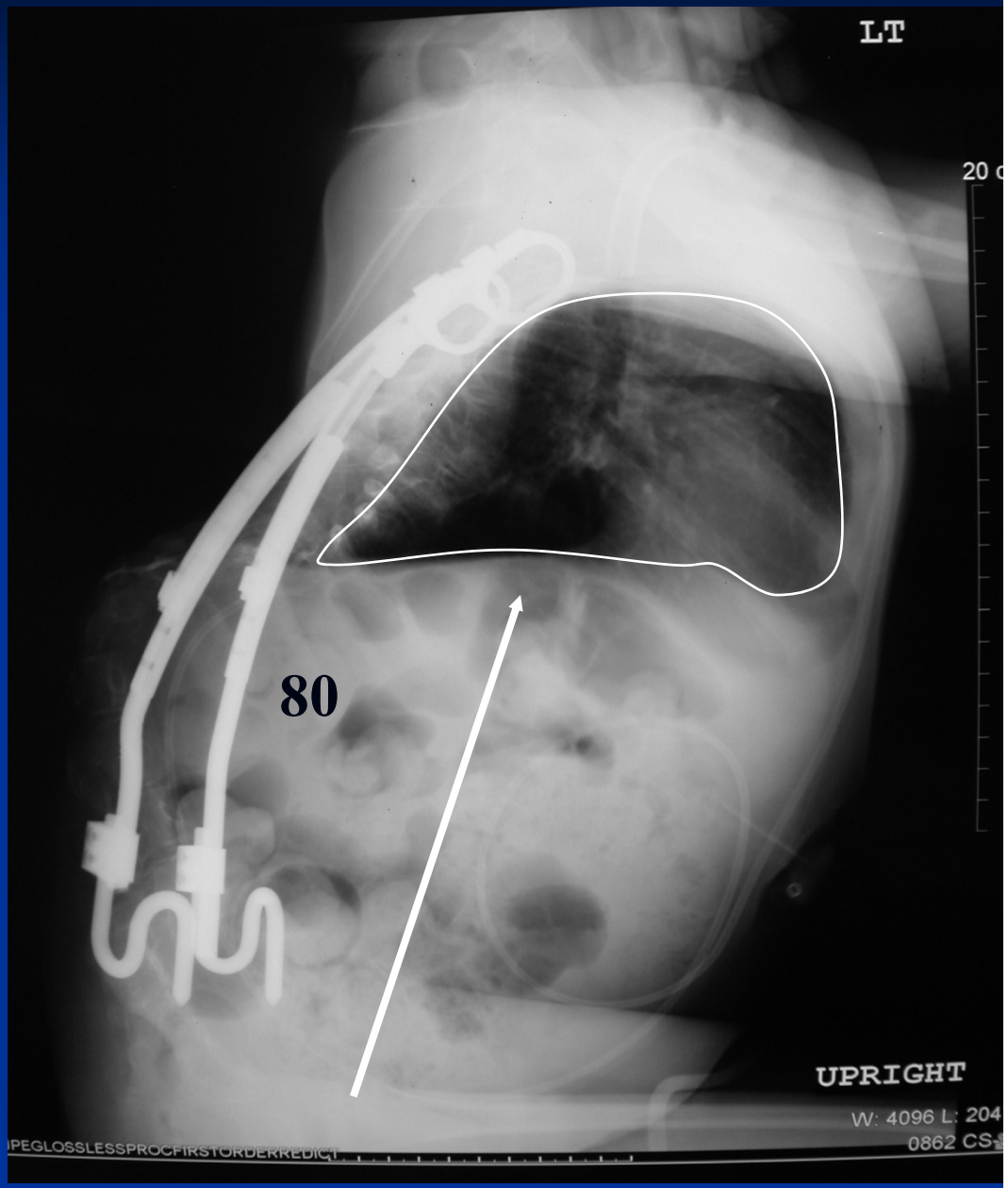




Thoracic Institute
Christus Santa Rosa
Children's Hospital

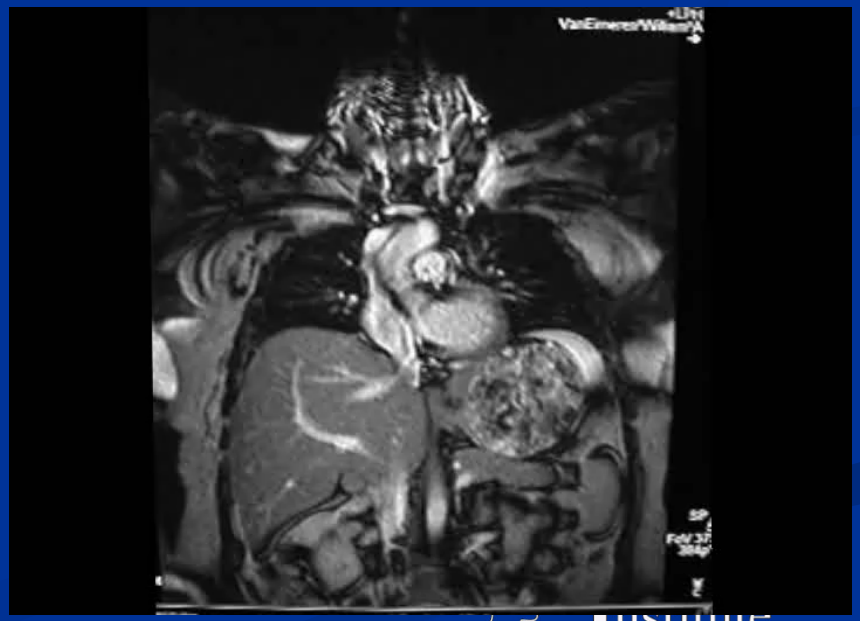
2 yr follow-up





The Future

- We need to define the natural history of TIS
- We need new ways to assess the results of all treatments
  - The Cobb angle will become obsolete at some point
  - Dynamic MRI of the thorax has the potential to characterize the anatomic basis of thoracic insufficiency syndrome



- VEPTR is a good first step

-Self Expansion VEPTRs

-New devices to treat TIS

Institute
Christus Santa Rosa
Children's Hospital

Thank You!

Thoracic Institute
Christus Santa Rosa
Children's Hospital