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Hope lives here.

Center for Thoracic Insufficiency Syndrome

Novel Approaches for Assessing Diaphragmatic Contribution to Thoracic Insufficiency Syndrome: SMA as an Illustration

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For
RM Campbell, MD
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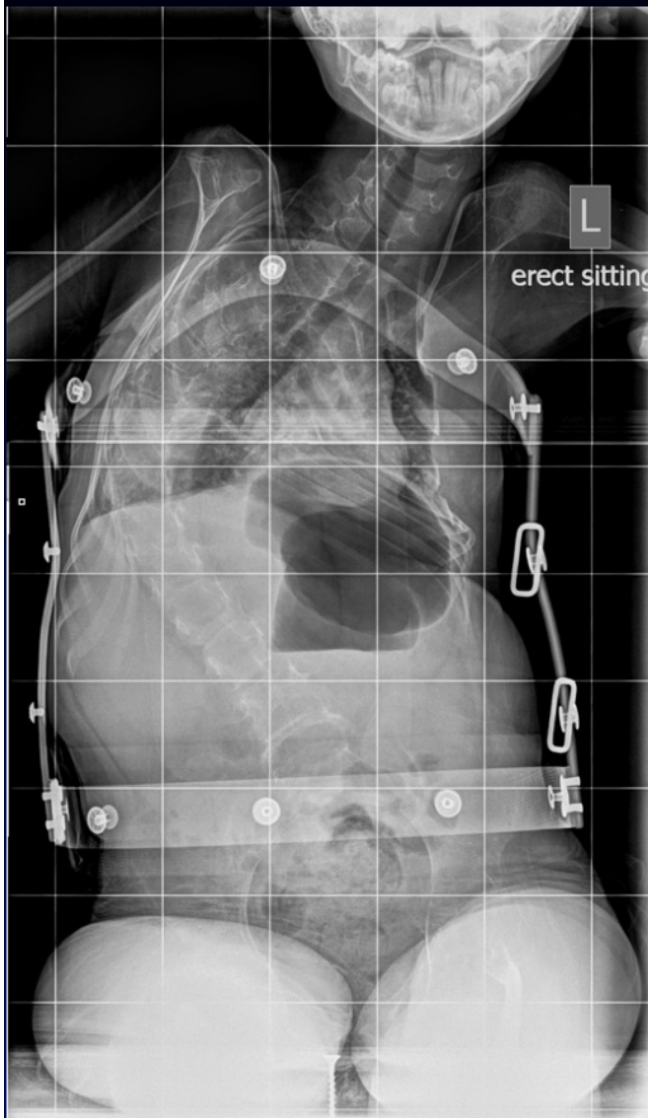
Division of Orthopaedics, CHOP
Professor of Orthopaedic Surgery
University of Pennsylvania Perelman School of Medicine



Key Clinical Question:

How do chest wall, **muscle dysfunction**, and spine deformity affect respiration in SMA?

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Goals

- Contribution of chest wall, diaphragm, and spine deformity in SMA to development of respiratory failure
  - Static /dynamic factors
- The effect of a novel Prosthetic Rib technique
- Novel Approaches for Assessing the Chest Wall and Spine in SMA

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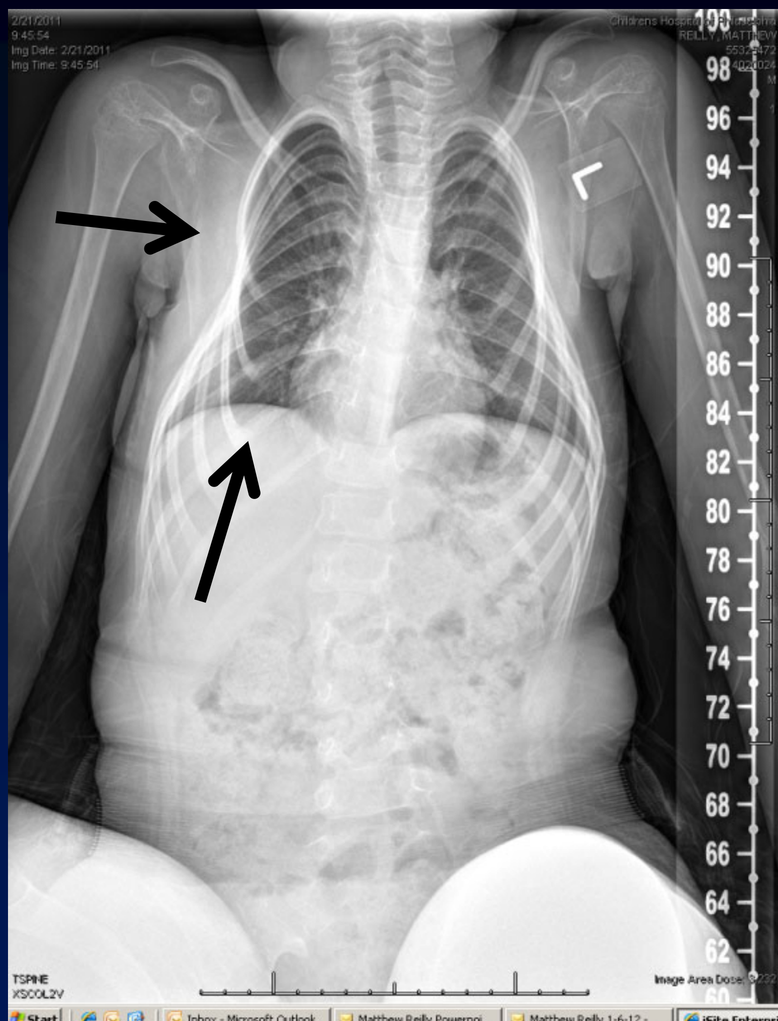
SMA Chest Wall Deformity



- Bell shaped thorax
  - Perez, et al. CHEST, 1996
- Collapsing Parasol Deformity of Dimeglio
  - Campbell and Smith, JBJS 2007
- Parasol rib deformity
  - Livingston, Snyder, et al, SPINE 2015

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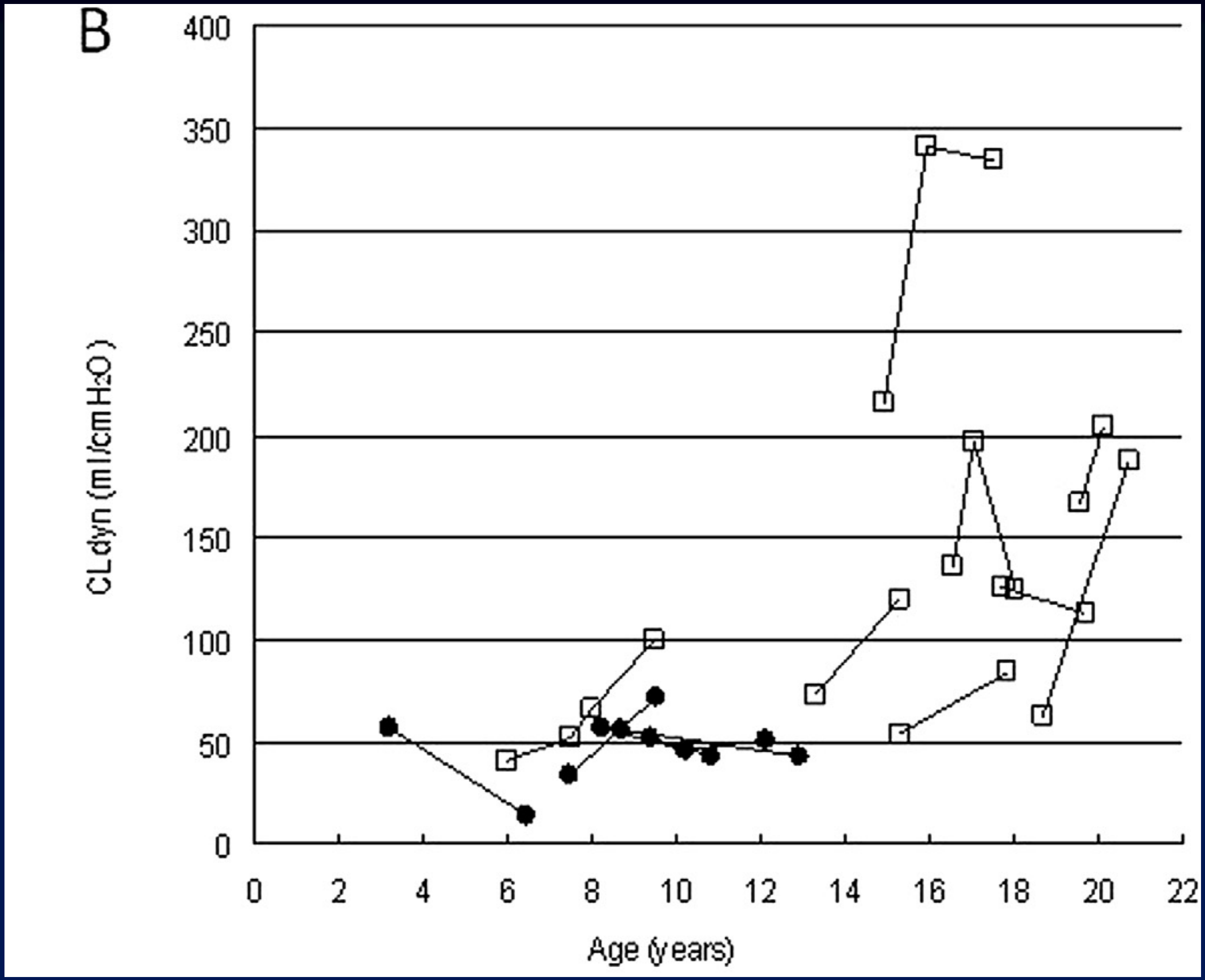
Chest Wall :SMA II, III



-Kihrani, et al, Europ J of Paed Neurology 2013

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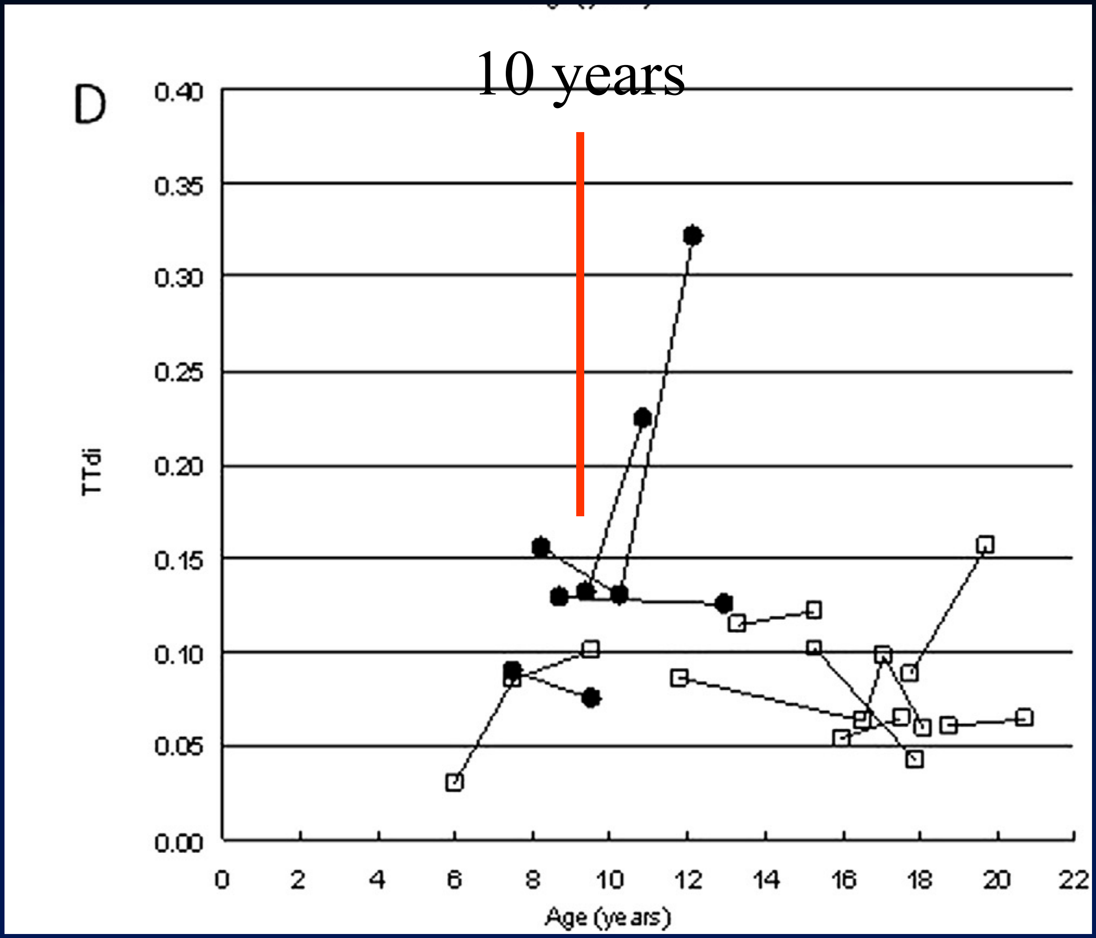
Diaphragm Fatigue: SMA II, III



-Kihrani, et al, Europ J of Paed Neurology 2013

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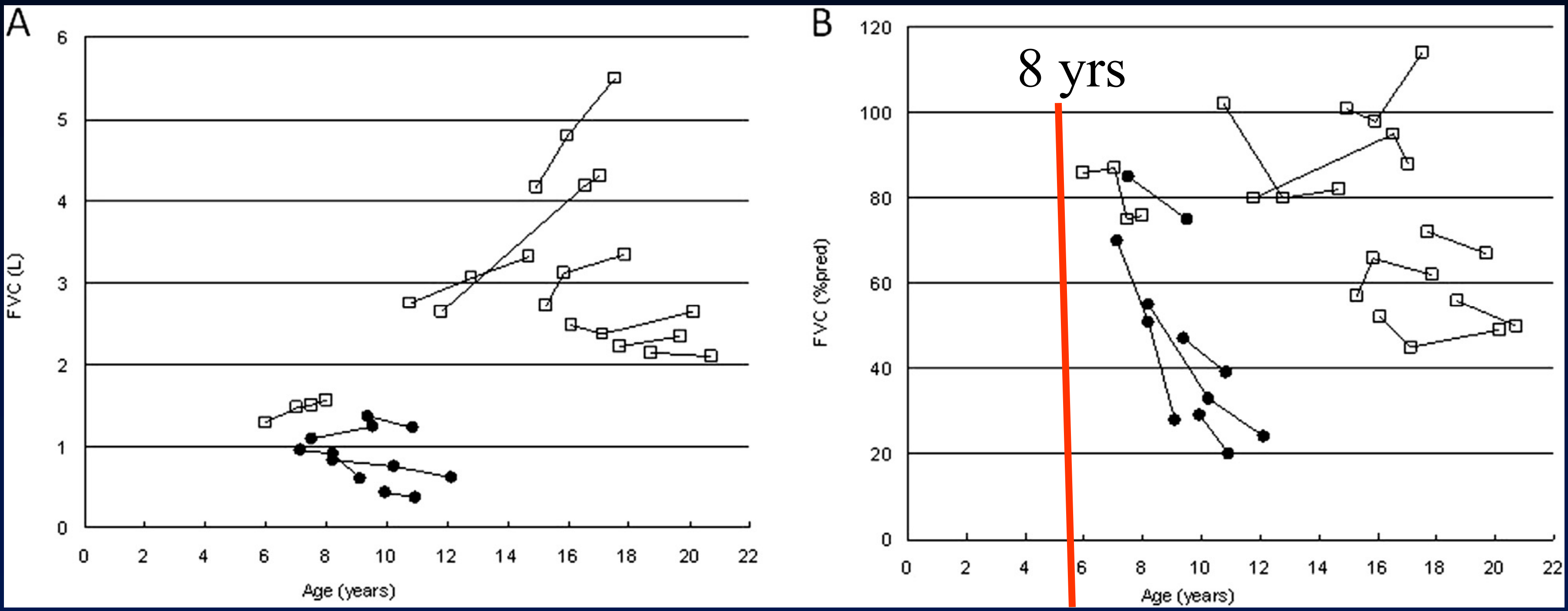
SMA II and III

FVC % pred FVC



-Kihrani, et al, Europ J of Paed Neurology 2013

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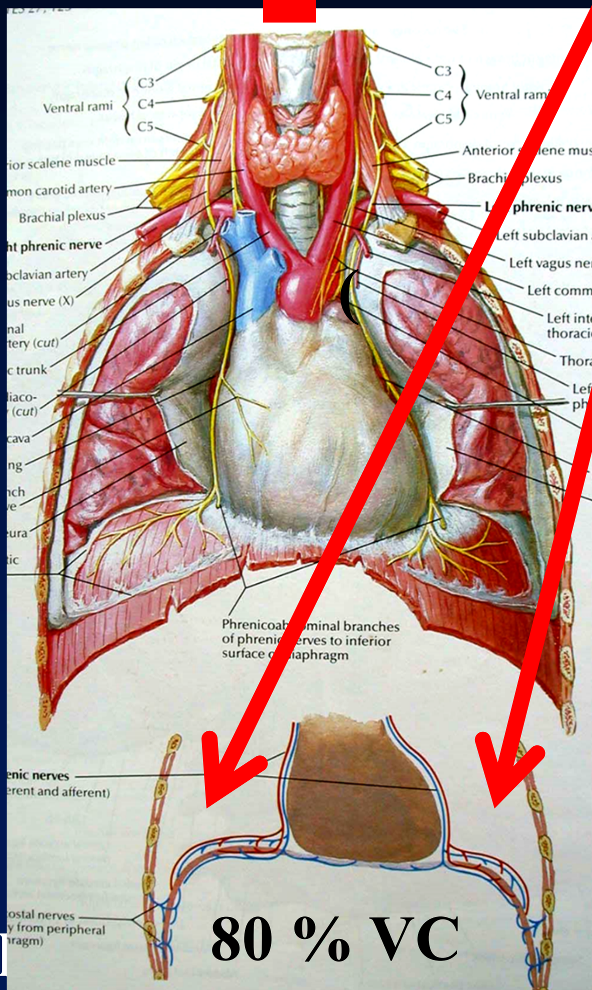
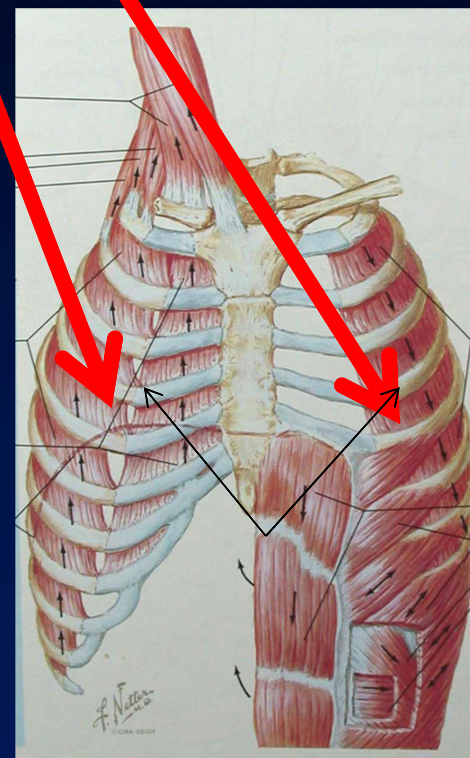
The 4 determinants of the thoracic respiratory pump

VC



80 % VC



20 % VC

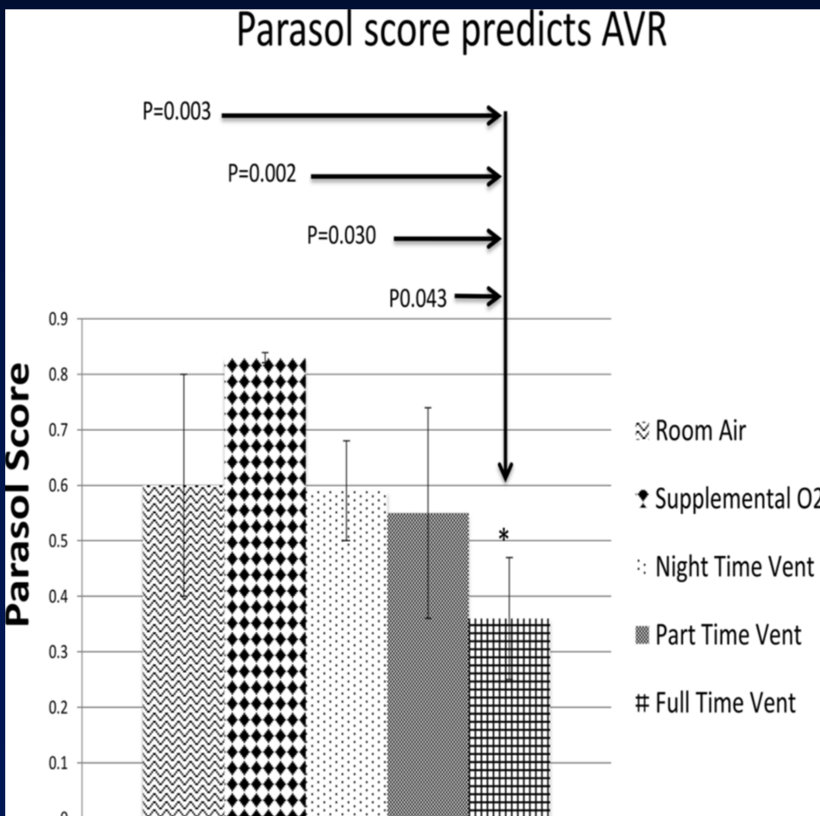
CT


2/21/2011
9:45:54
Img Date: 2/21/2011
Img Time: 9:45:54
RELLY, MATTHEW
TSPINE
XSCOL2V
Start
Inbox - Microsoft Outlook
Matthew Reilly Powerpoi...
Matthew Reilly 1-6-12 - ...
iSite Enterpri


es lung view
e #1/30
DOB 8/22/2004; Age 0

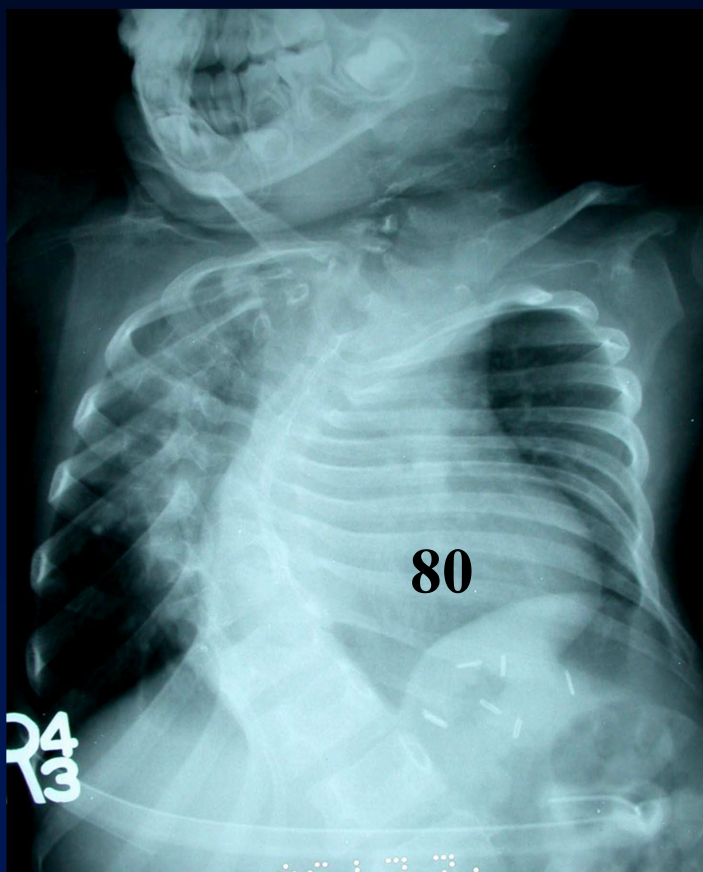
Parasol Score

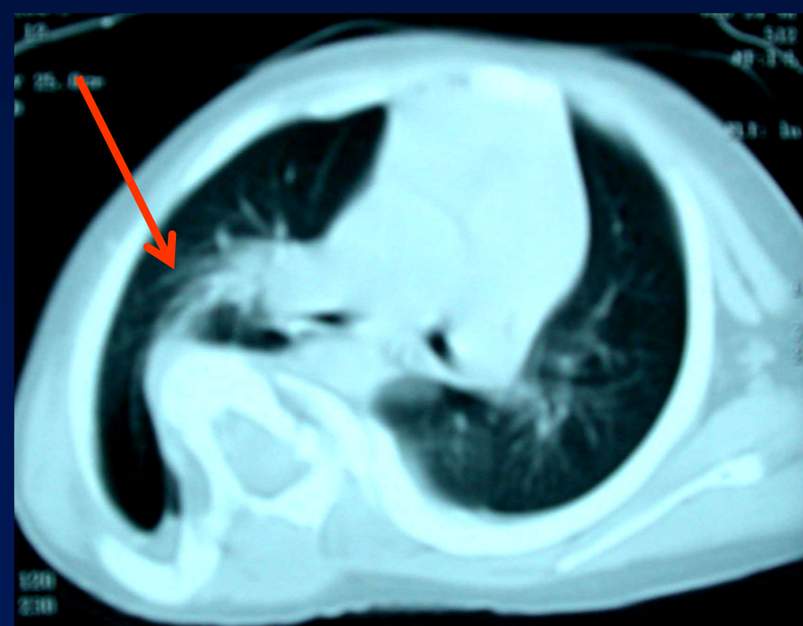
$$\frac{\text{T6 convex hemithoracic width}}{\text{T6 concave hemithoracic width}} \times \frac{\text{T6 thoracic width}}{\text{T12 thoracic width}}$$



-Livingston, Snyder, et al. Spine, 2015

The Hidden Volume Problem





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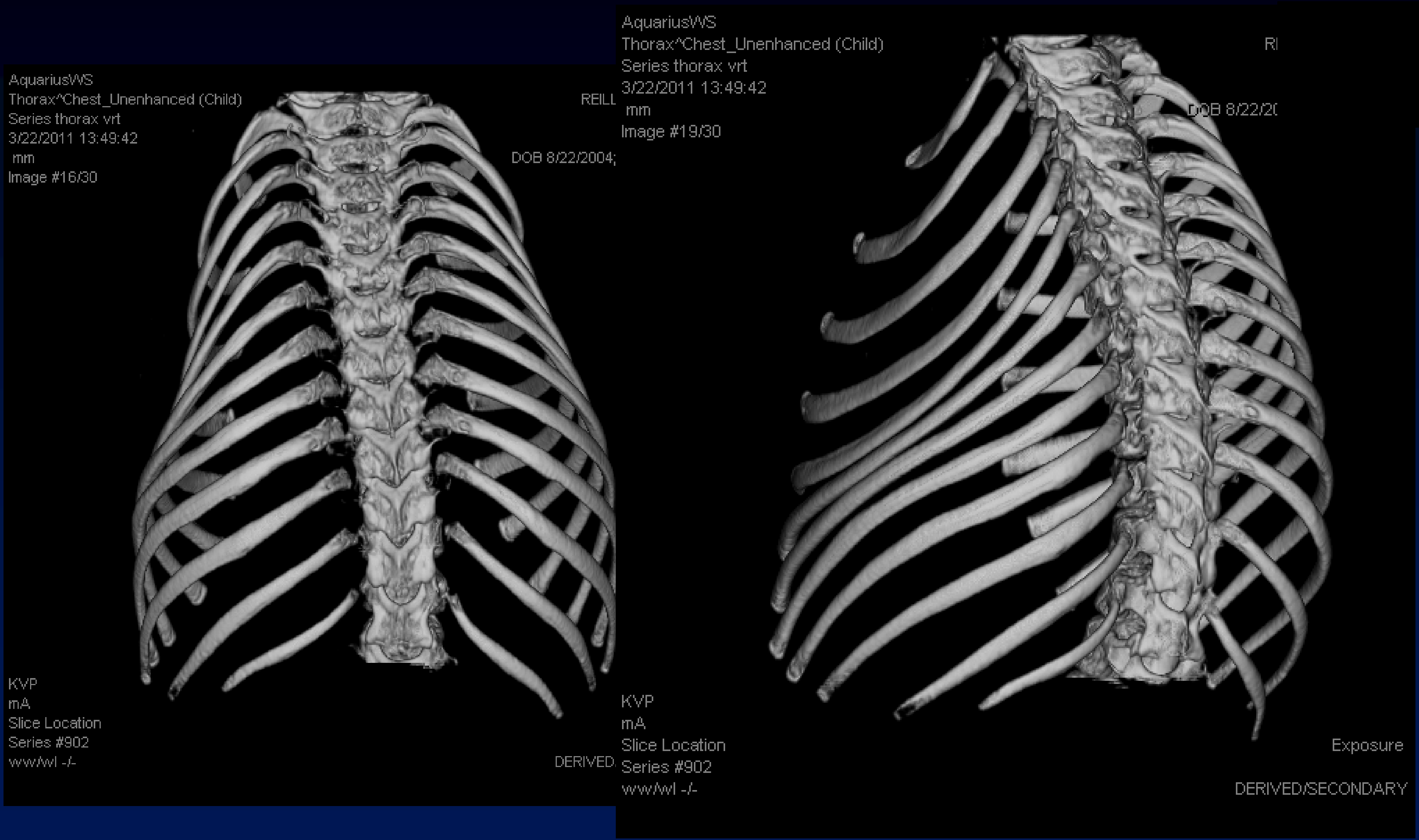
AquariusWS
Thorax^Chest_Unenhanced (Child)
Series thorax vrt
3/22/2011 13:49:42
mm
Image #16/30
REILL
DOB 8/22/2004;
KVP
mA
Slice Location
Series #902
ww/wl -/-
DERIVED
AquariusWS
Thorax^Chest_Unenhanced (Child)
Series thorax vrt
3/22/2011 13:49:42
mm
Image #19/30
RI
DOB 8/22/20
KVP
mA
Slice Location
Series #902
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Exposure
DERIVED/SECONDARY

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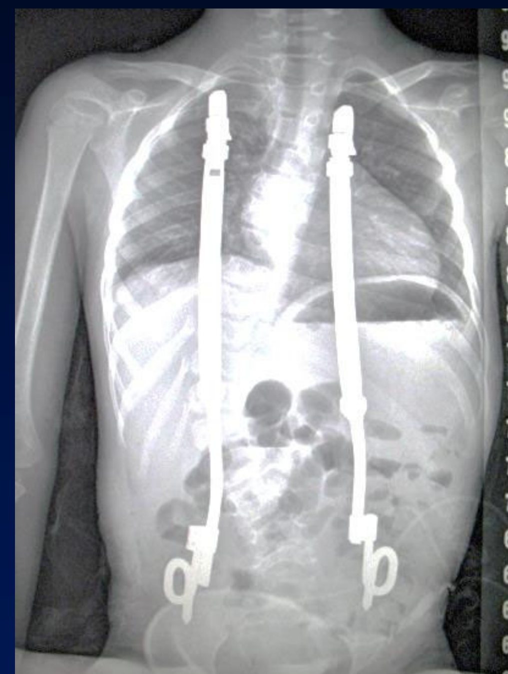
SMA spine treatment



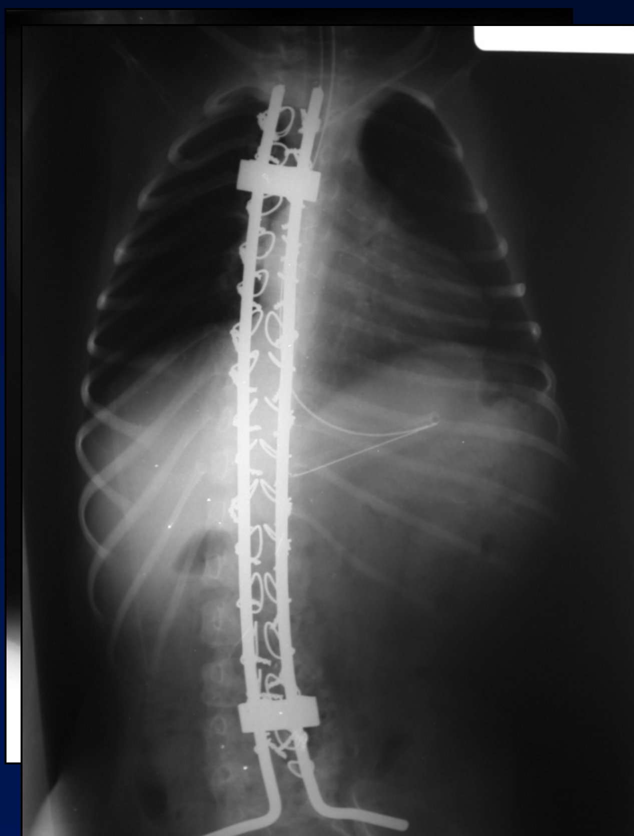
Prosthetic Rib

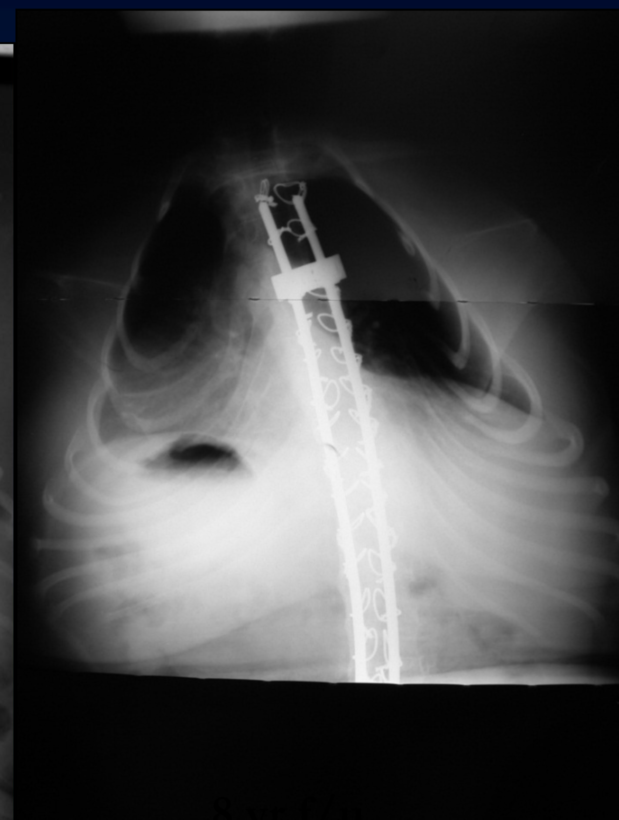
C

-Mesfin, Sponseller, Leet JAAOS 2012

Spine stabilization does not treat the Collapsing Parasol Deformity of SMA

- Campbell/Smith JBJS, 2007





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VC 40 % nl

SMA II/III: natural history and fusion FVC

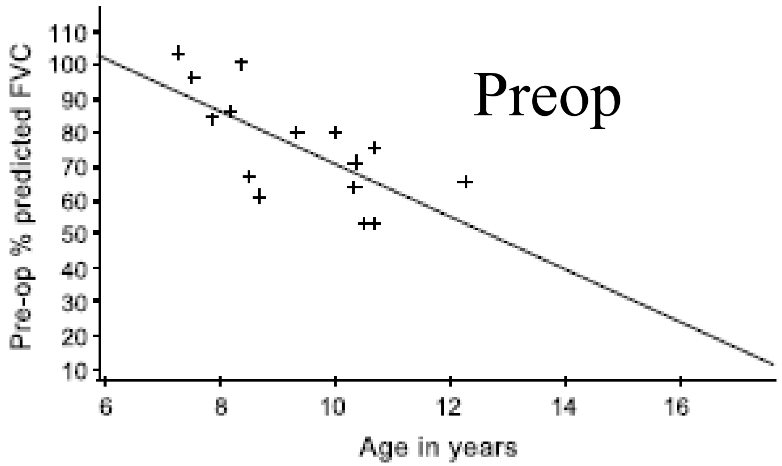

Fig. 1 Pre-operative percentage of predicted forced vital capacity (FVC) versus age in years.

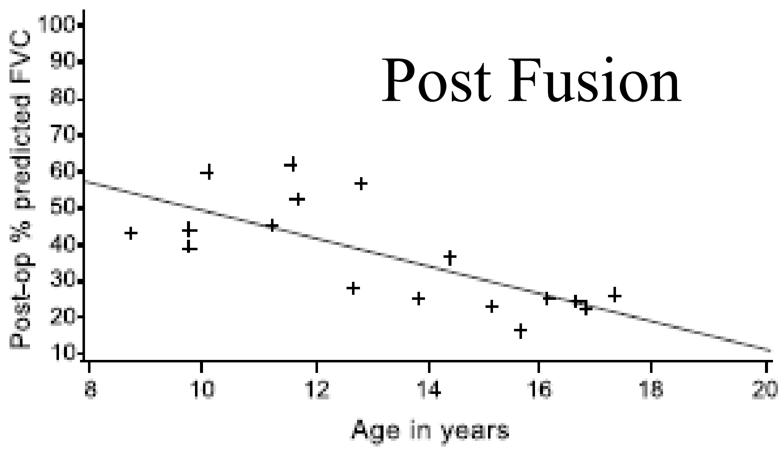

Fig. 2 Post-operative percentage of predicted forced vital capacity (FVC) versus age in years.

-Chng, et al, J Ped Child Health, 2003

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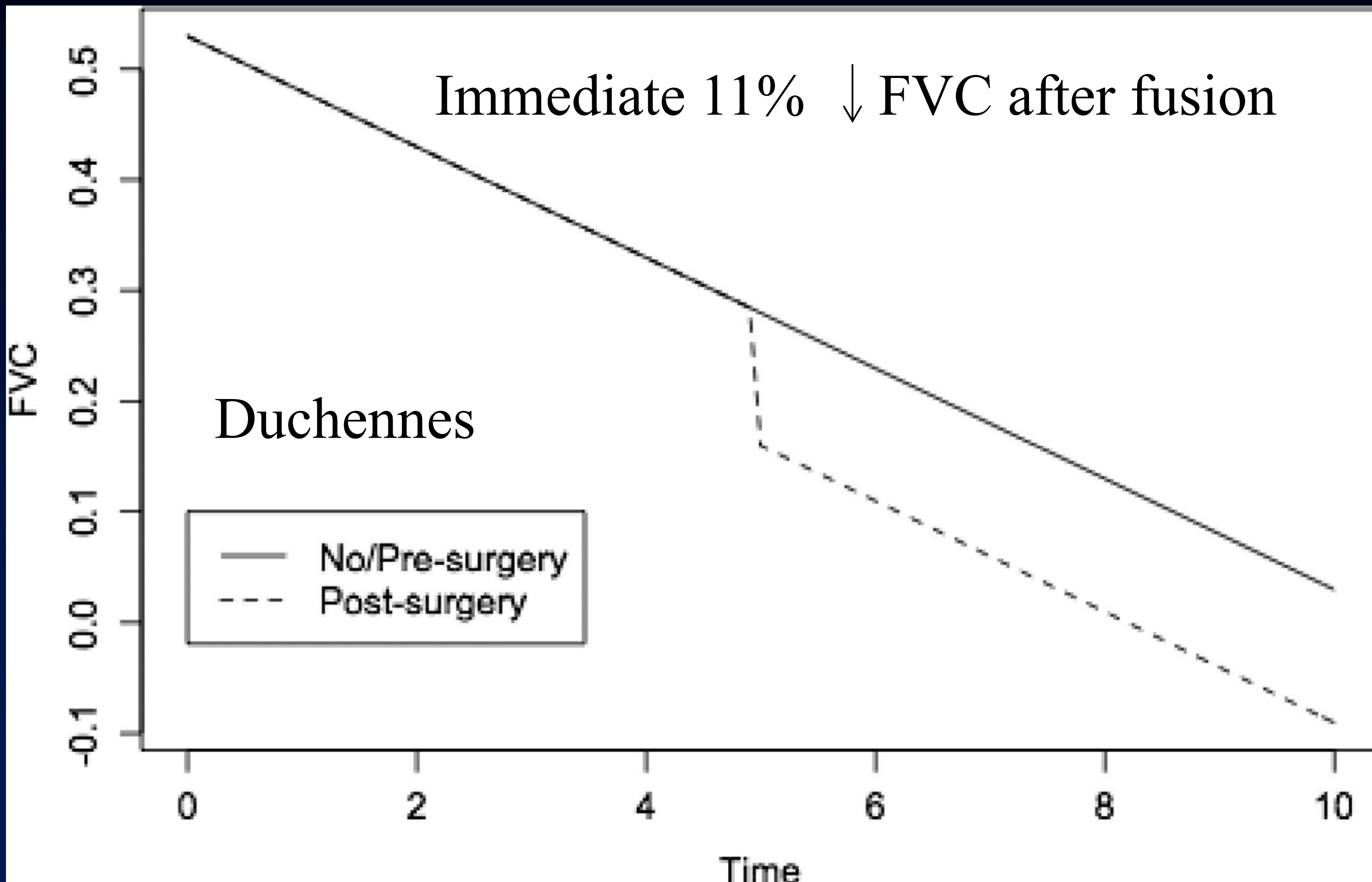
Effect of posterior spine fusion



-Roberto, et, SPINE, 2011

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Pulmonary Function Testing

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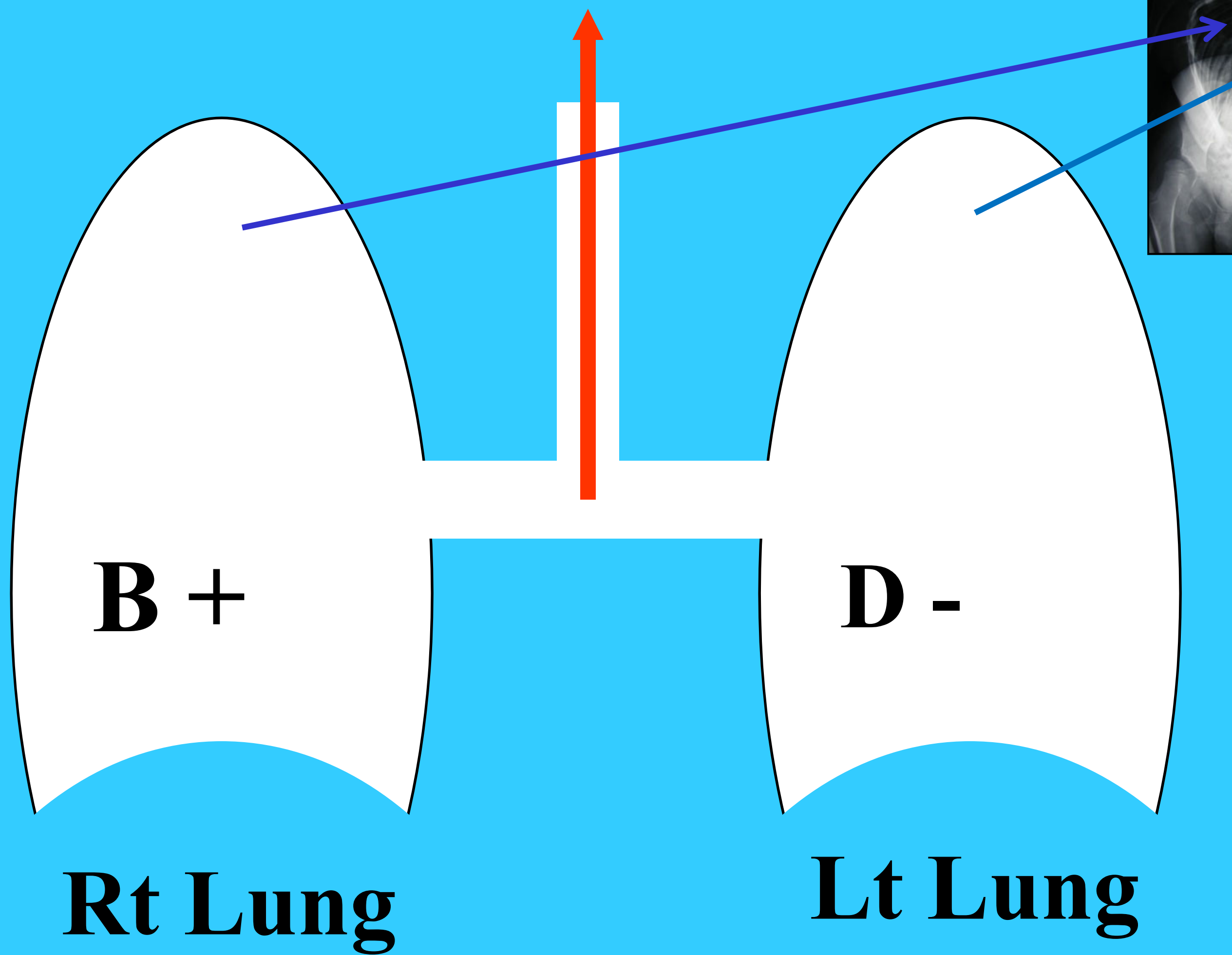
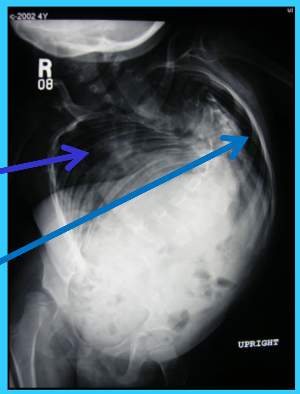
PFT <u>averages</u> these : FVC, TV, Etc .. = C + ?

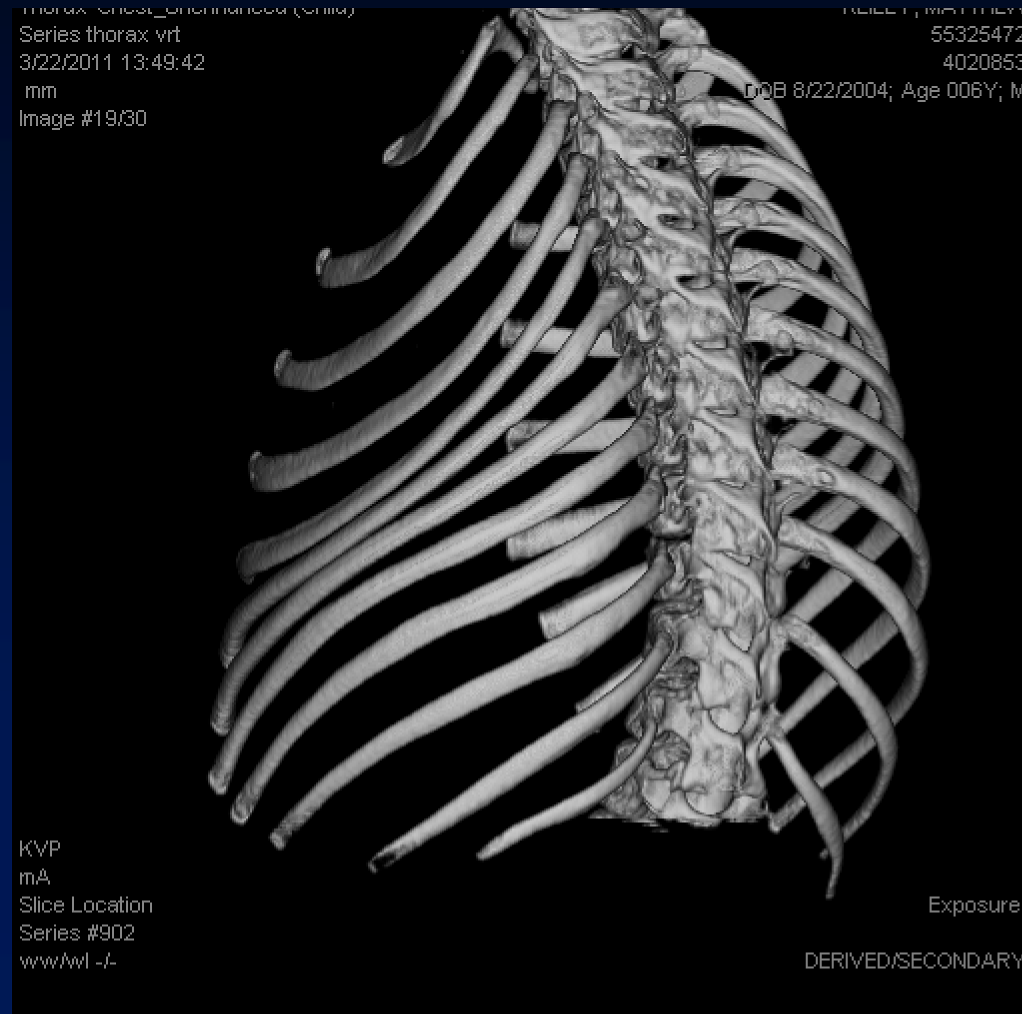




PFT’s are unfortunately, summative,
And hard to do under age 5 years

What’s the problem?

- Intercostal muscle weakness only?
- Collapsing parasol deformity contributes?
- How to better understand this?



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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

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Dynamic Lung MRI

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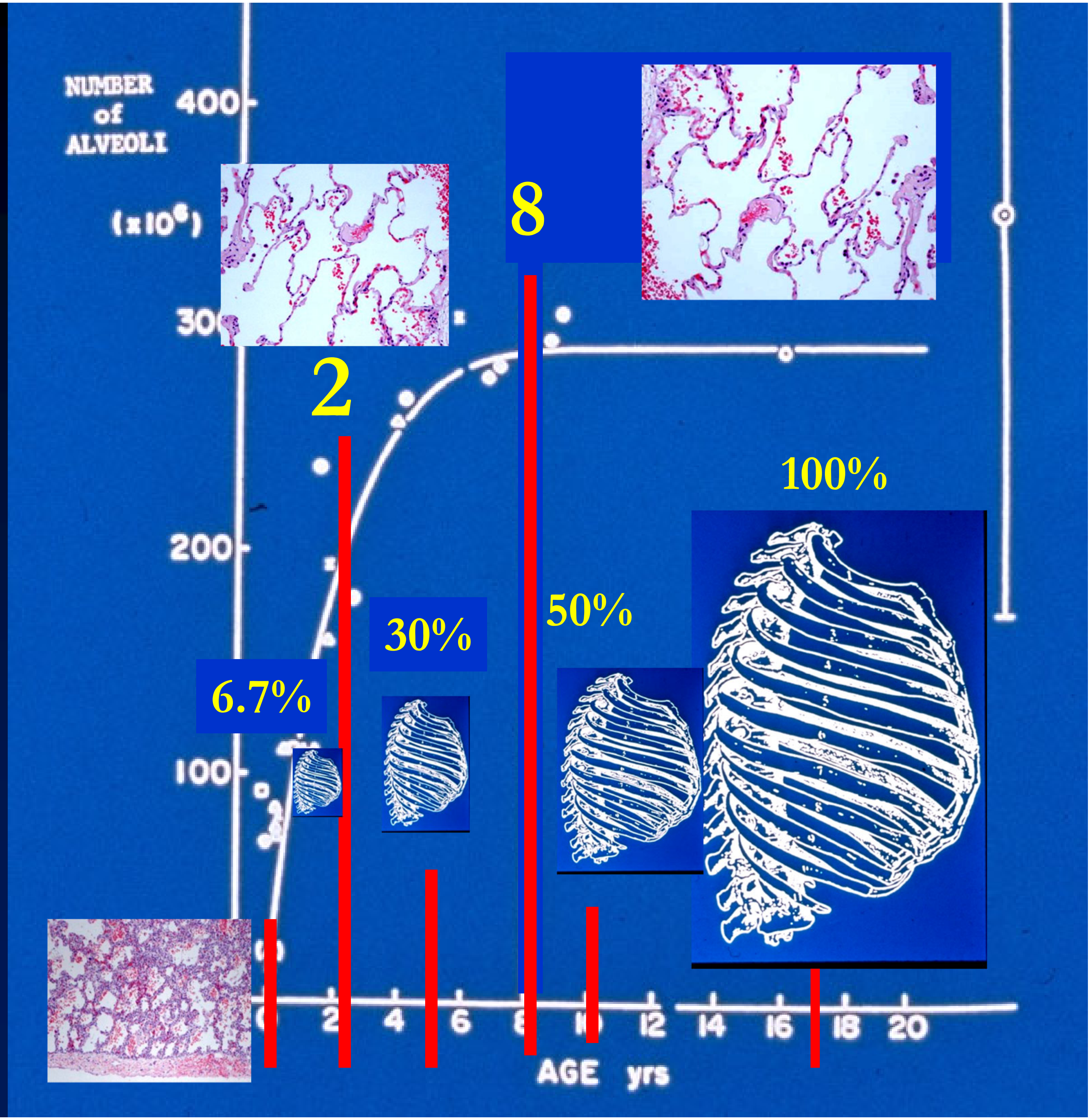
Lung and Thoracic Growth are Inter-Dependent



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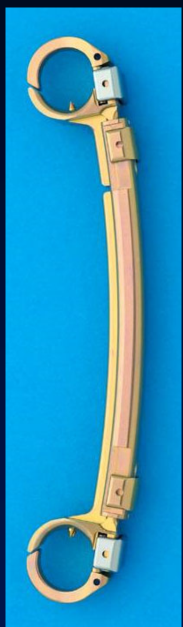
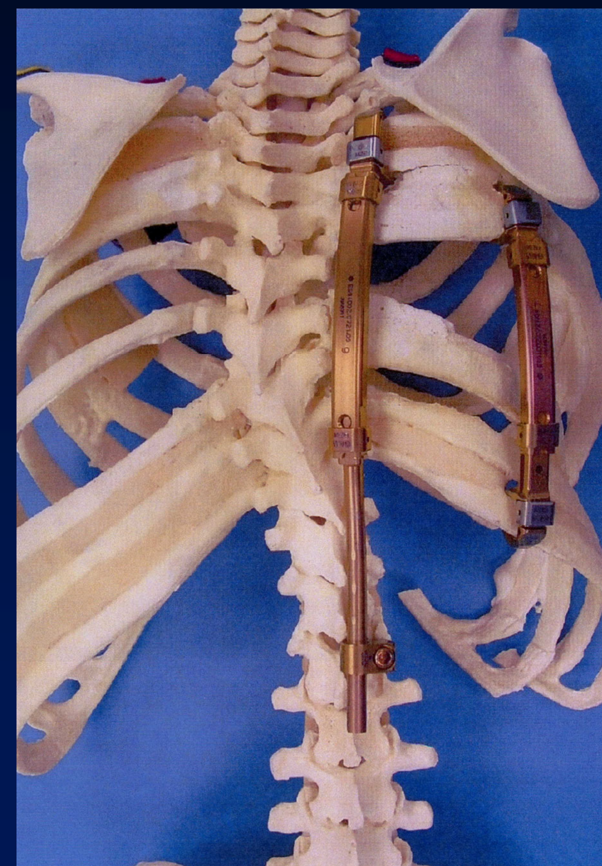
Treatment of TIS:
(Prosthetic Rib) FDA approved 2004 for HDE





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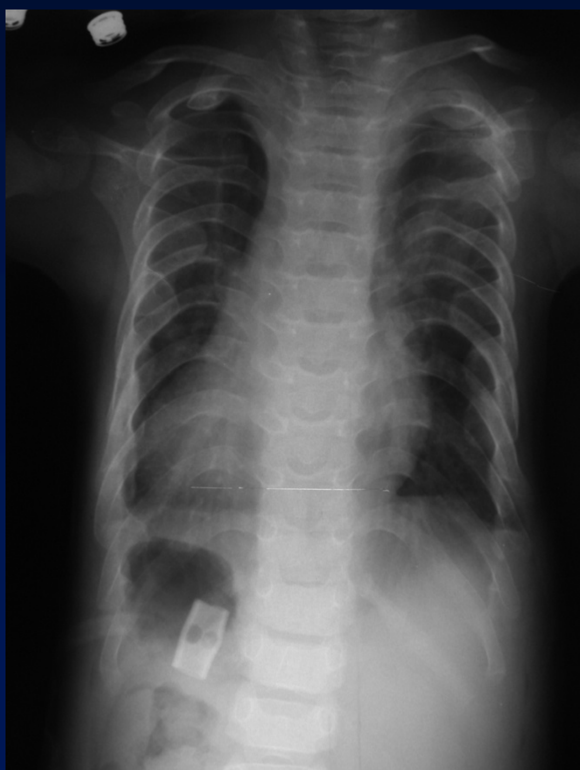
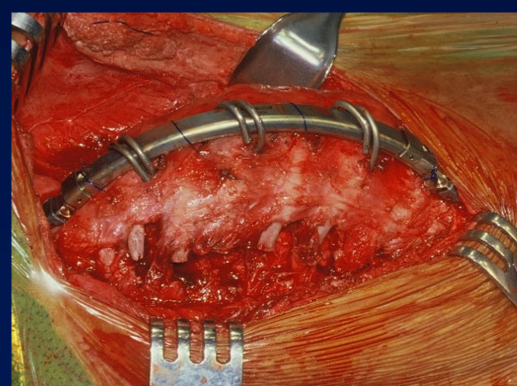
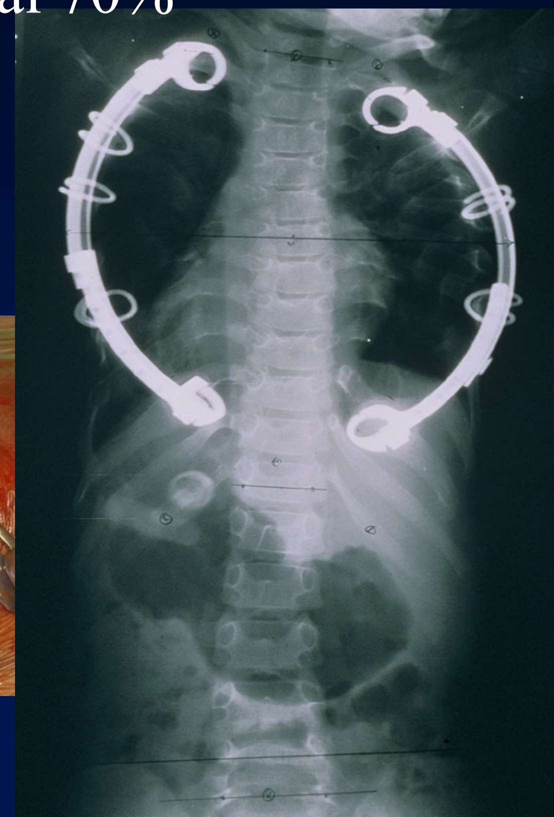
Prosthetic Rib Postero-Lateral Dynamic Expansion Thoracoplasty for Jeune's Syndrome

Natural History Mortality 80%
Prosthetic Rib Treatment Survival 70%







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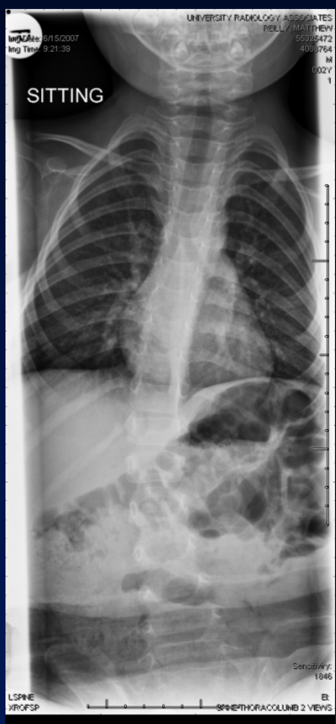
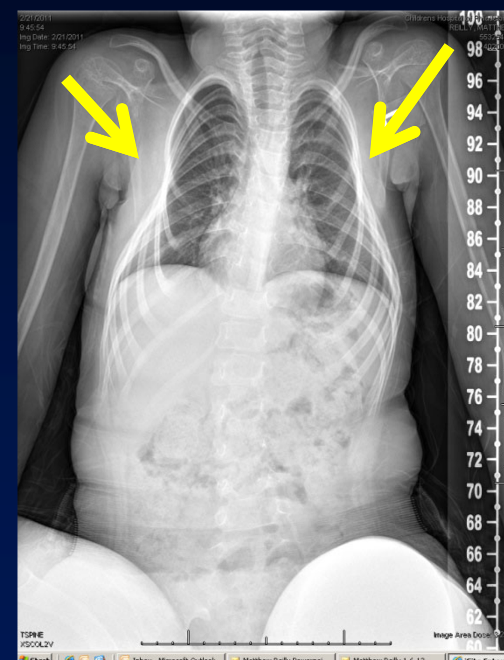
Spinal Muscular Atrophy

2007 2009 2010 2011





7 y/o

3 y/o

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Respiratory Rate 40/min, Vital Capacity dropping


AquariusWS
Thorax^Chest_Unenhanced (Child)
REILL
Series thorax vrt
3/22/2011 13:49:42
mm
DOB 8/22/2004;
Image #16/30
KVP
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Slice Location
Series #902
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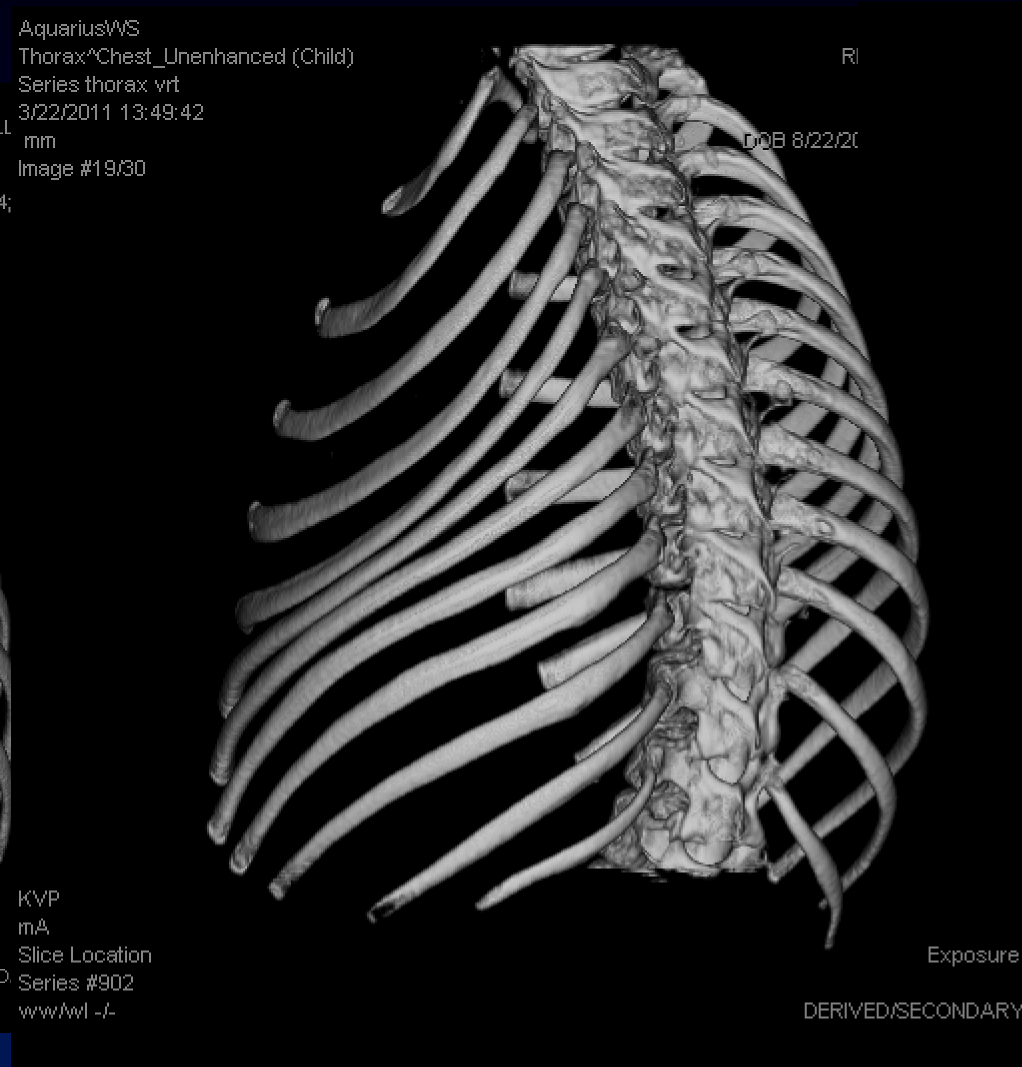
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Image #19/30
KVP
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Slice Location
Series #902
ww/wl -/-
Exposure
DERIVED/SECONDARY

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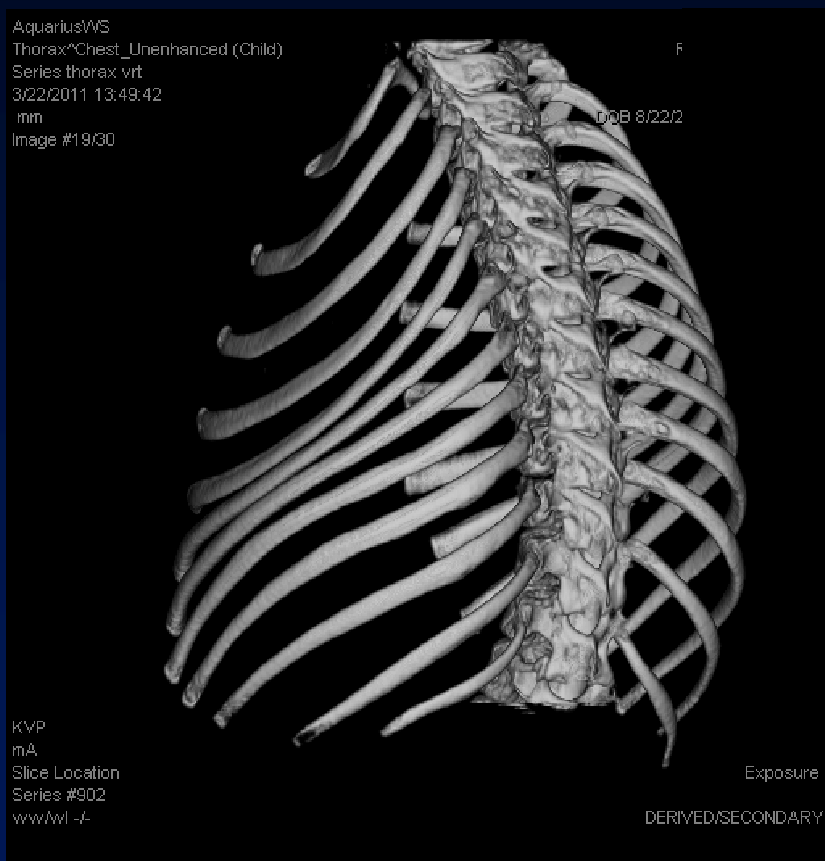
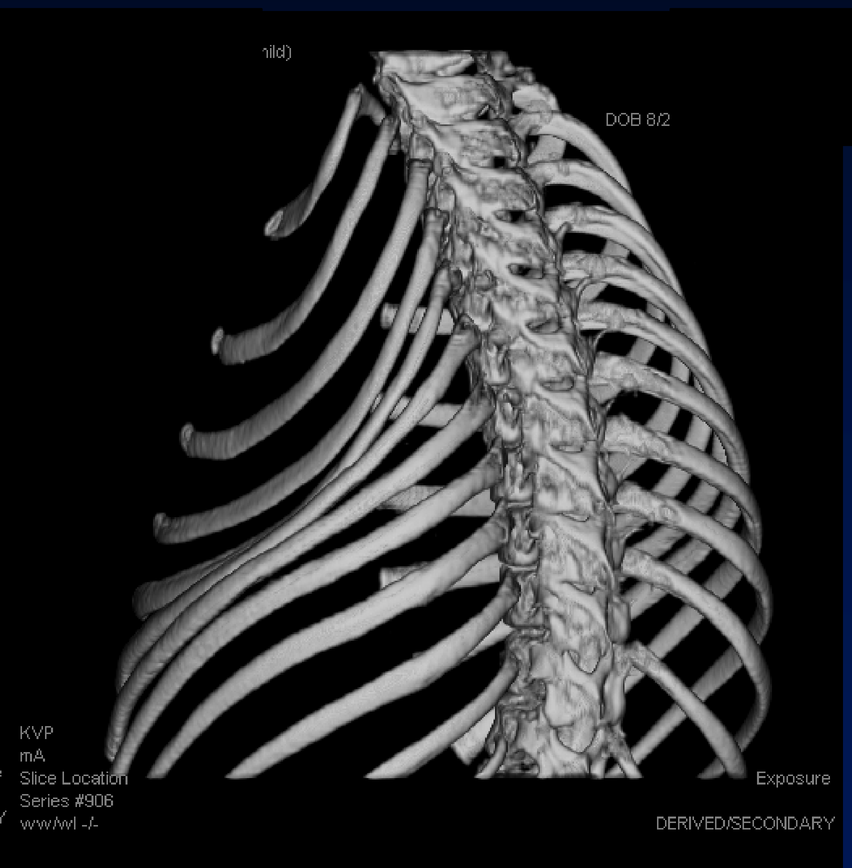
Increased Thoracic Deformity

Presentation

9 month F/U





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Natural History

Age 7 : Presentation

9 months later

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Dynamic Lung MRI

Presentation age 7

9 month later

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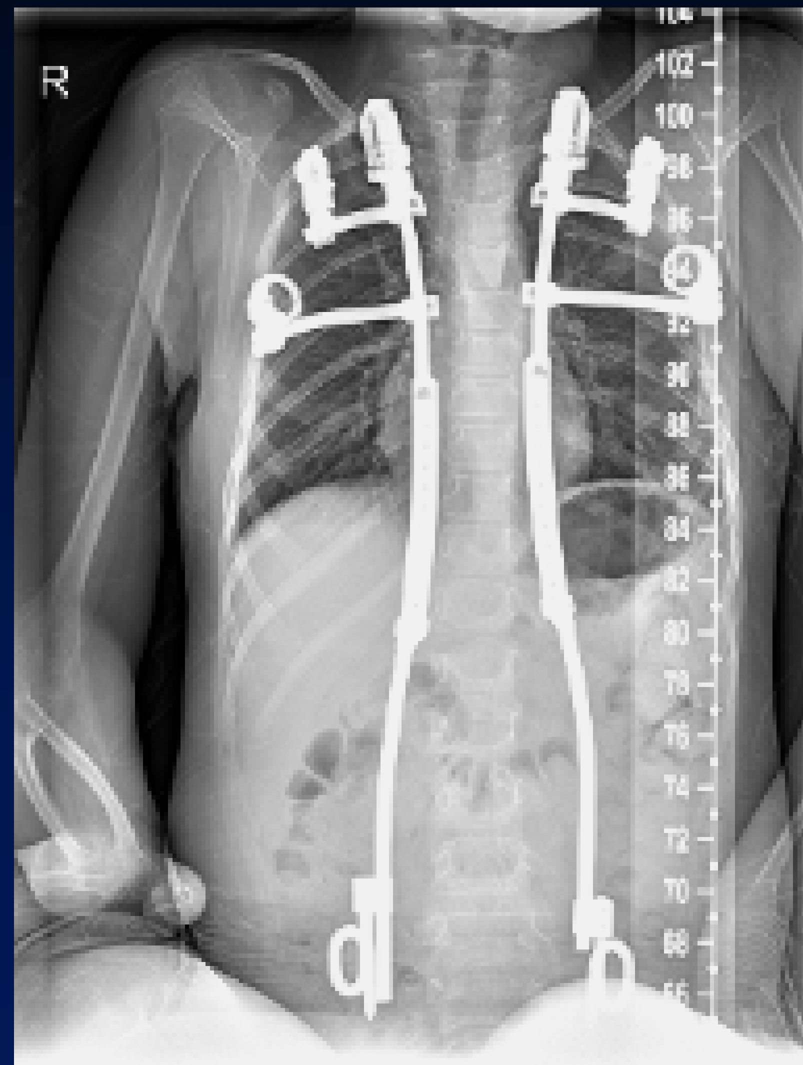
Expanded Thoracic Volume



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Subjective Increase in Thoracic Function

Pre op

6 months Post op

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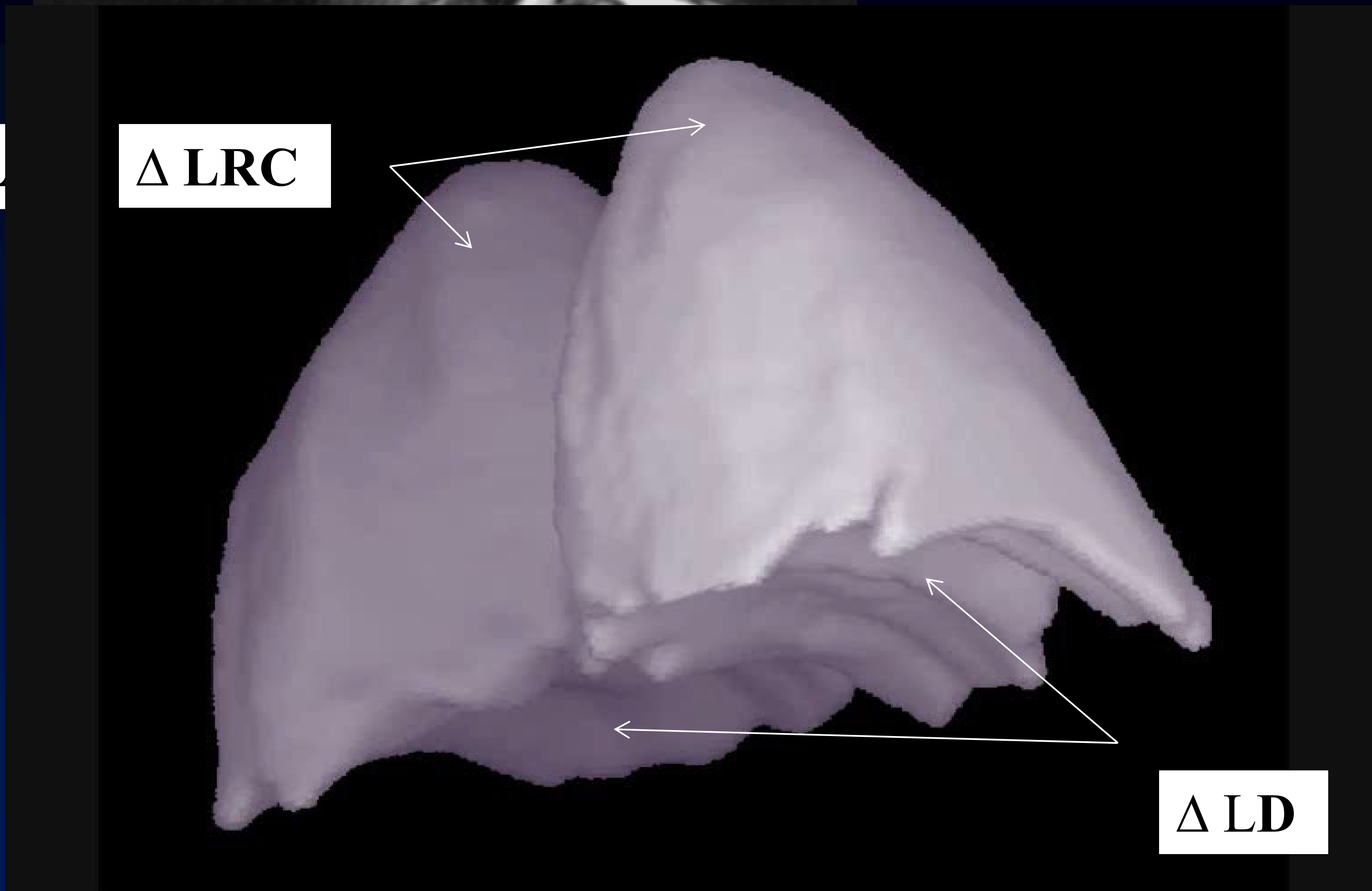
How can we quantify this so we get some numbers?

Dynamic Lung MRI (dMRI)

Δ L



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Medical Image Analysis 35 (2017) 345–359

Contents lists available at ScienceDirect

Medical Image Analysis

journal homepage: www.elsevier.com/locate/media

ELSEVIER

Retrospective 4D MR image construction from free-breathing slice Acquisitions: A novel graph-based approach

CrossMark

Yubing Tong[a], Jayaram K. Udupa[a,*], Krzysztof C. Ciesielski[a,b], Caiyun Wu[a], Joseph M. McDonough[c], David A. Mong[c], Robert M. Campbell Jr.[c]

[a] Medical Image Processing Group, Department of Radiology, University of Pennsylvania, Philadelphia, PA, 19104 United States
[b] Department of Mathematics, West Virginia University, Morgantown, WV, 26505 United States
[c] Center for Thoracic Insufficiency Syndrome, Children's Hospital of Philadelphia, Philadelphia, PA, 19104 United States

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4D image construction approach*

- A purely image-based, retrospective 4D Image construction from “free-breathing” MRI slice acquisitions

- Expected features
  - no need for external tidal volume measurement
  - no requirement on breath holding
  - no need to estimate respiratory signal and sort images with respiratory signal (no registration or curve fitting operation)

*Tong, Udupa, et al.. MeDIA, 35 (2017) 345–359.

Acquiring 2D slices

Gather all slices with anatomic spatial and dynamic information about the free-breathing thorax

Constructing optimal 3D scenes

Determine breathing phases & label reference nodes

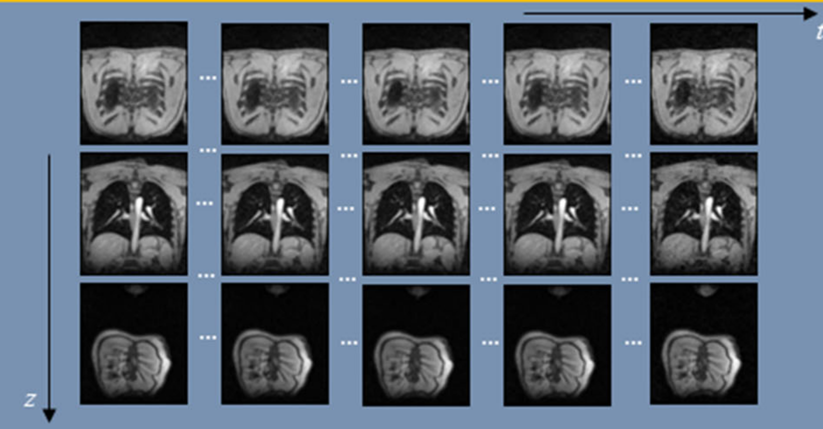
Build a weighted graph, $G = (A, P, w)$

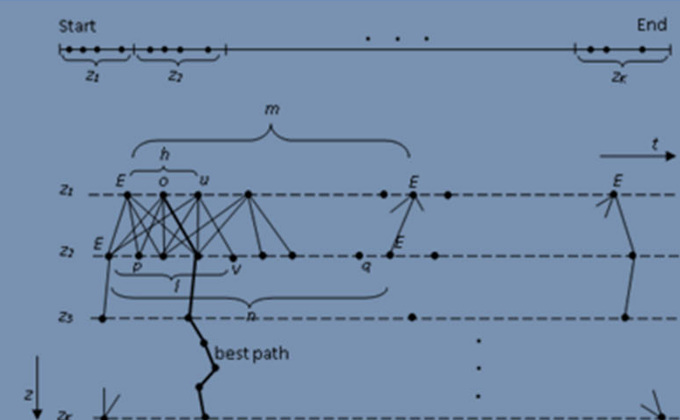
Search optimal paths on graph & each best path for one best 3D scene

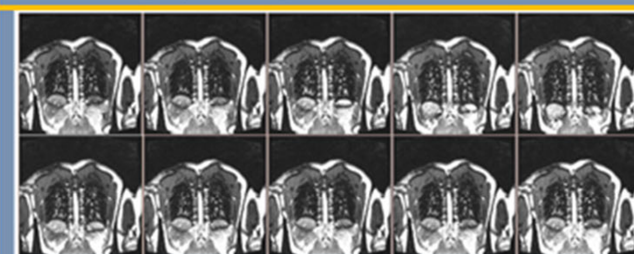
Constructing an optimal 4D image

Select one subset of 3D scenes with smallest cost to construct an optimal 4D image over one respiratory period

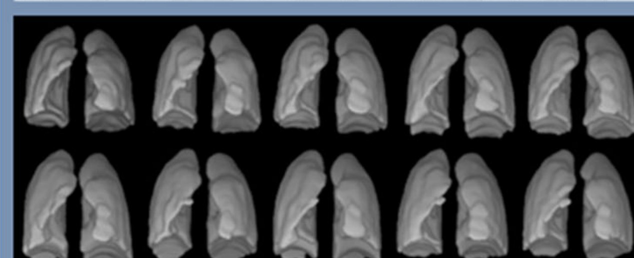
Graph $G = (A, P, w)$
A: set of all 2D images with each image as a node in G
P: set of arcs
w: weight between two nodes at neighbor locations





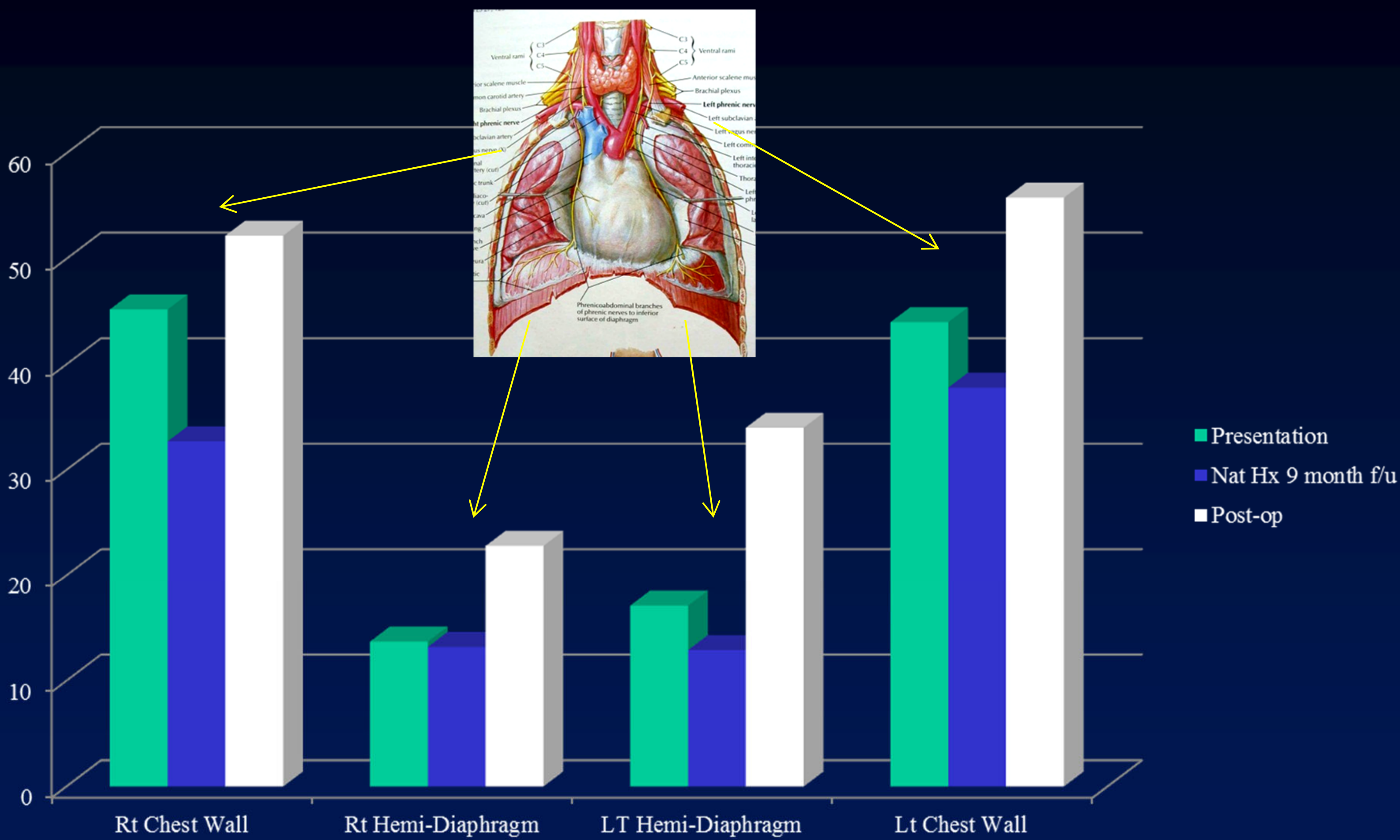


Constructed 4D Image over one period (10 time points).



Surface renditions of 10 volumes.

Thoracic Performance Index



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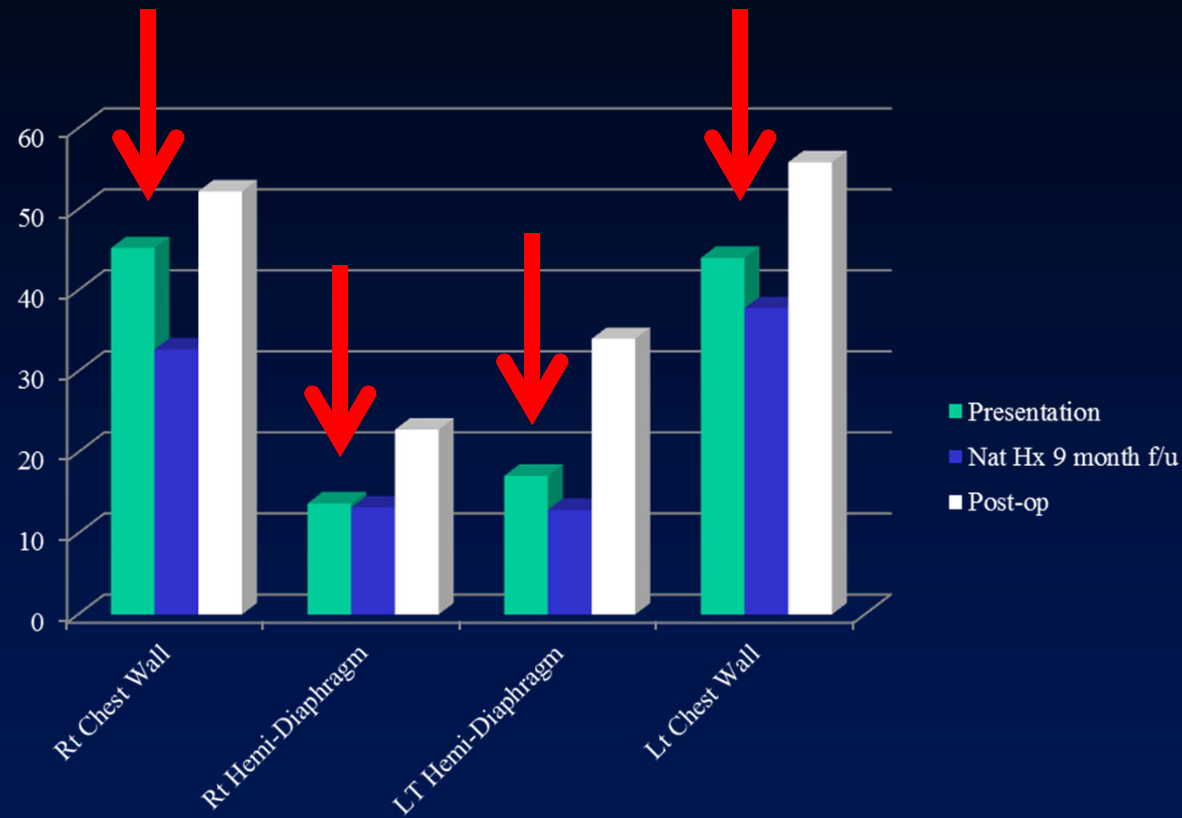
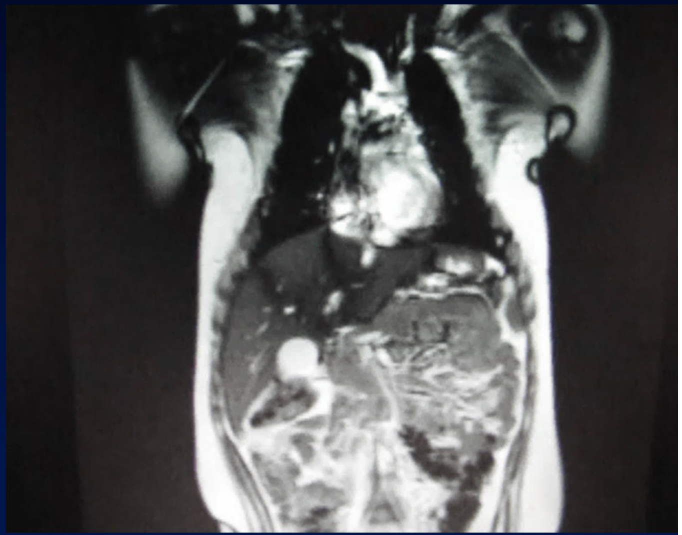

60
50
40
30
20
10
0
Rt Chest Wall
Rt Hemi-Diaphragm
LT Hemi-Diaphragm
Lt Chest Wall
Presentation
Nat Hx 9 month f/u
Post-op



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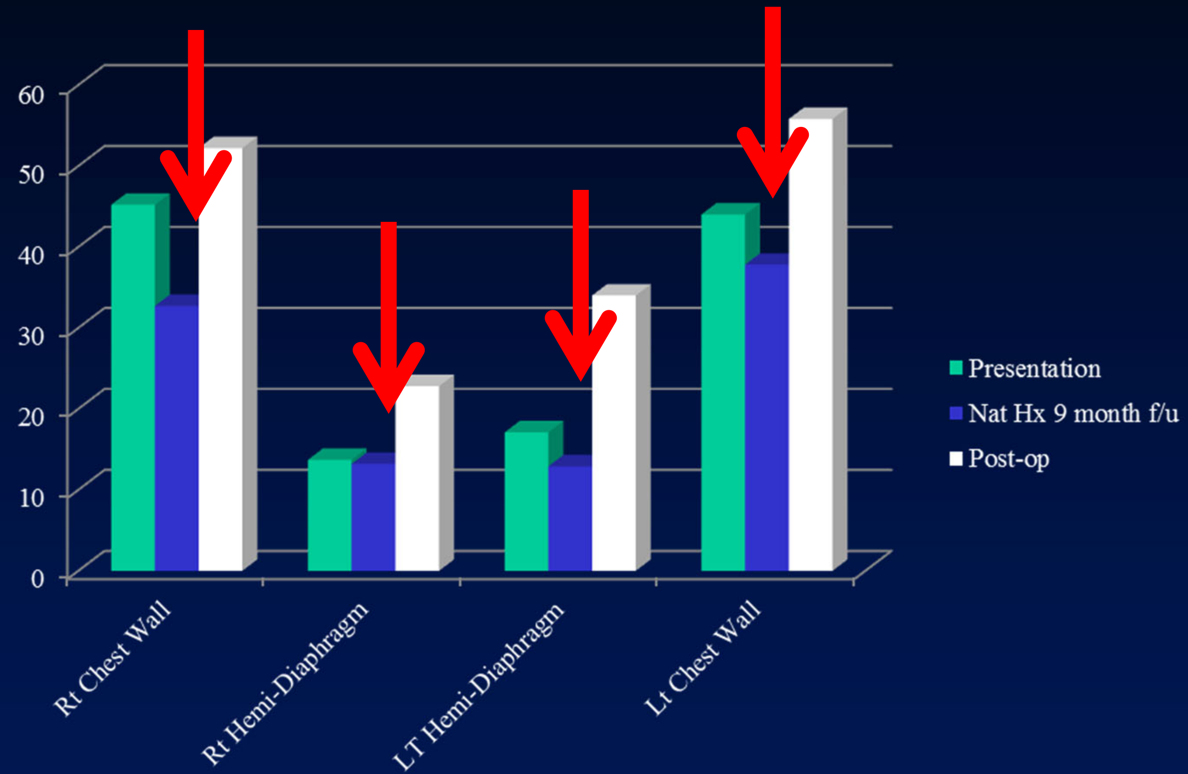
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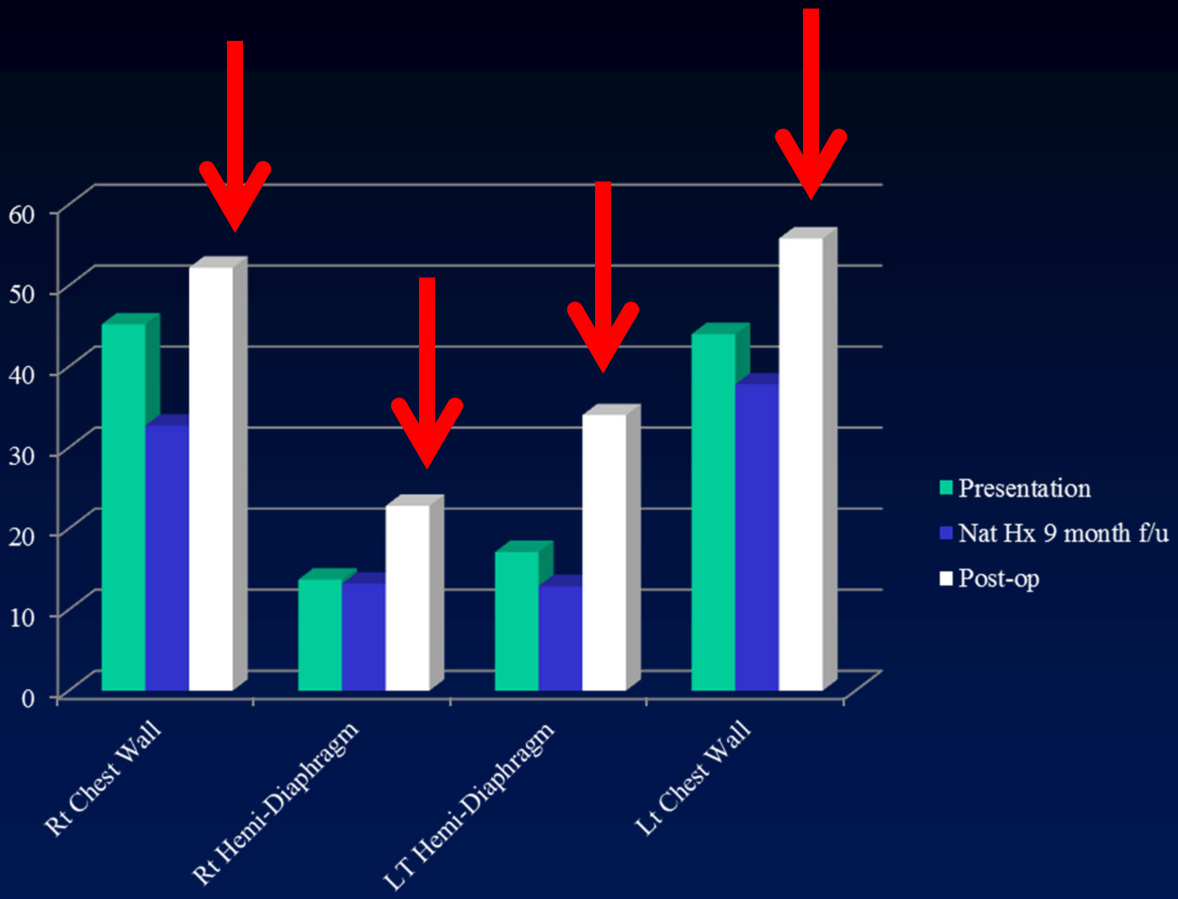
Rt Hemi-Diaphragm
LT Hemi-Diaphragm
Lt Chest Wall
Presentation
Nat Hx 9 month f/u
Post-op

CTIS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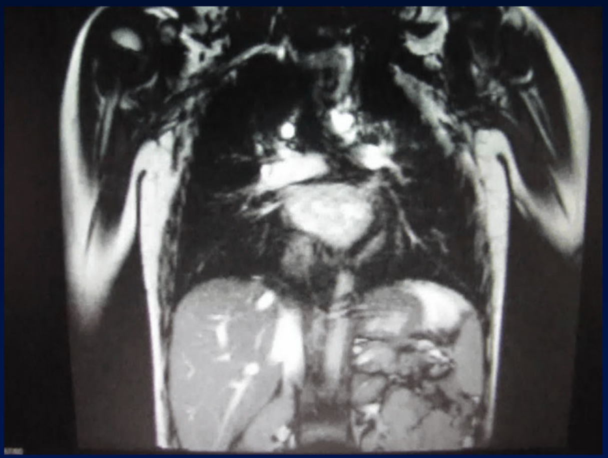
LT Hemi-Diaphragm
Lt Chest Wall
Presentation
Nat Hx 9 month f/u
Post-op



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Thoracic Performance Index

	Chest Wall Expansion	**Diaphragm Excursion**
Presentation	89.2 cc (74.3%) Total 120cc	30.8cc (25.7%)
9 month f/u pre-op	70.5cc (73%) Total 96.6cc	26.1cc (27%)
	↓ 21%	↓ 15.3%
6 months post-op	108cc (65.5%) 164.8cc	56.6cc (34%)
	↑ 53.2%	↑117.6%

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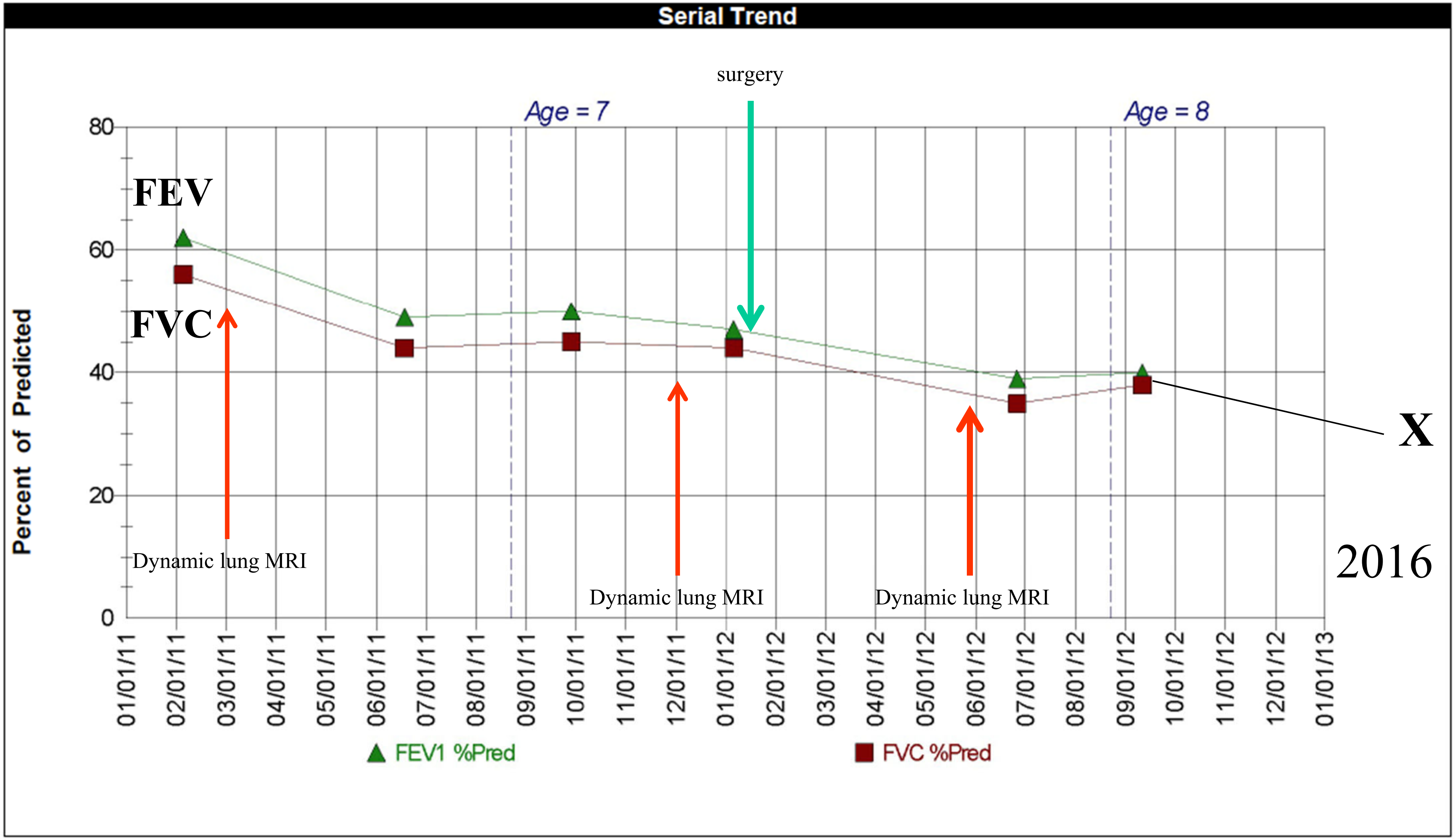
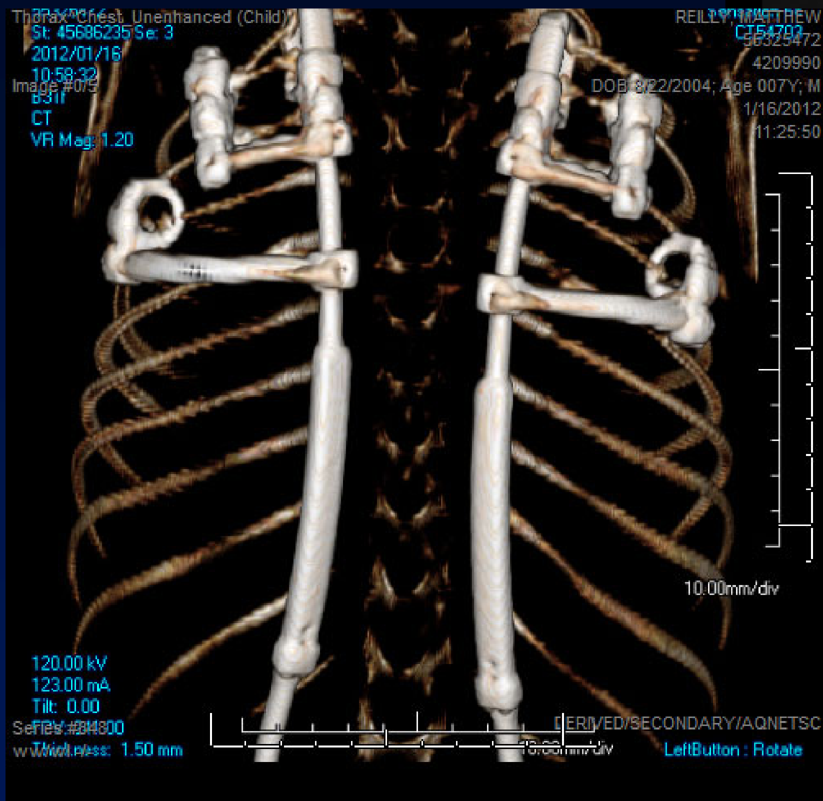
Pulmonary Function tests: FVC, FEV



5 yr f/u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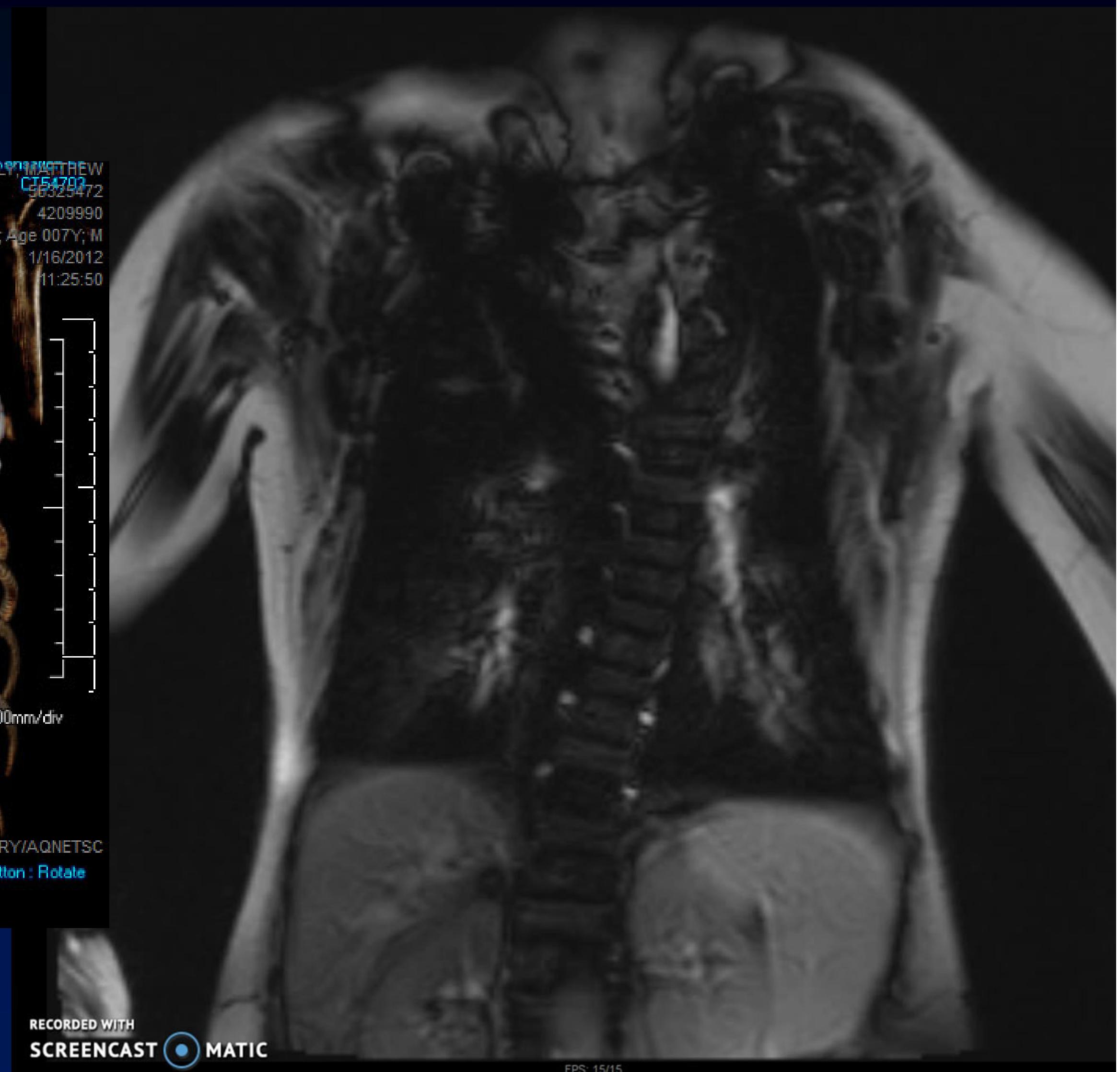
10:58:32
CT
VR Mag: 1.20
REILLY, MATTHEW
4209990
DOB: 8/22/2004; Age 007Y; M
1/16/2012
11:25:50
10.00mm/div
120.00 kV
123.00 mA
Tilt: 0.00
Thickness: 1.50 mm
DERIVED/SECONDARY/AQNETSC
LeftButton : Rotate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SCREENCAST MATIC
FPS: 15/15

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Final Thoughts

- A dynamic thoracic model of chest/spine abnormalities is needed
- Dynamic Lung MRI assessment is useful for monitoring static/dynamic components of these complex contributions to thoracic insufficiency

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Center
for
Thoracic Insufficiency
Syndrome

Thank You!