Have We Improved Pulmonary Function? Outcomes to Date, Future Directions

Charles E Johnston MD

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TIS in Texas - after Bob Left

Melvin Smith, MD

1941-2008

The Characteristics of 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

The inability of the thorax to support normal respiration and lung growth

What does this really mean ?

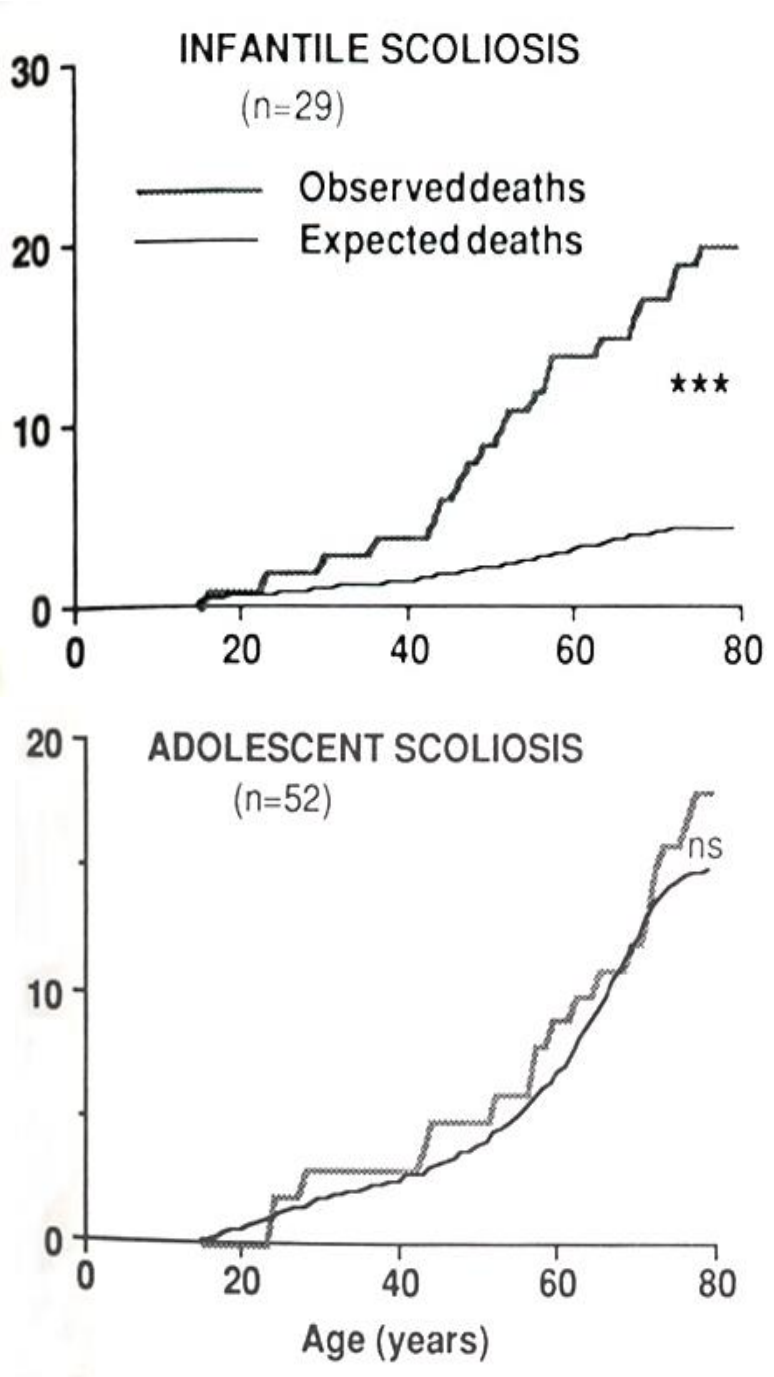
What we know ….

- Natural Hx untreated EOS not good

↓

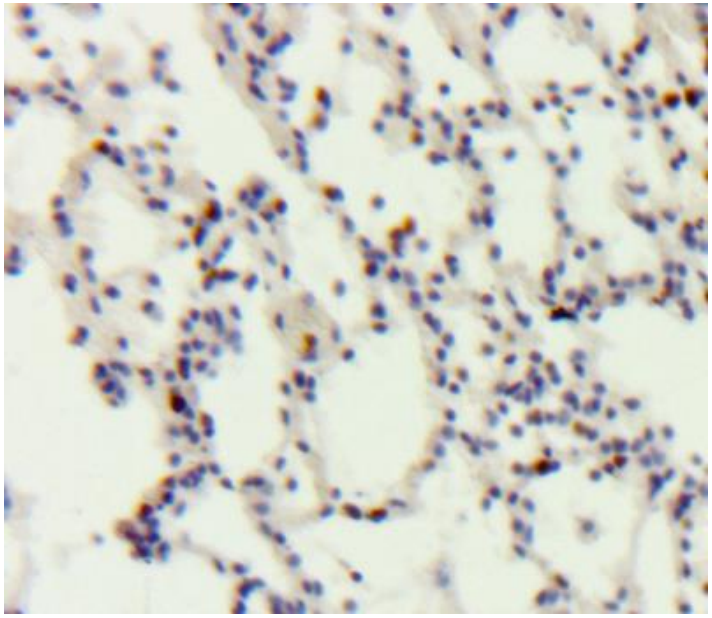
increased mortality
(Pehrsson, Branthwaite)

Remember –>
it's not good data

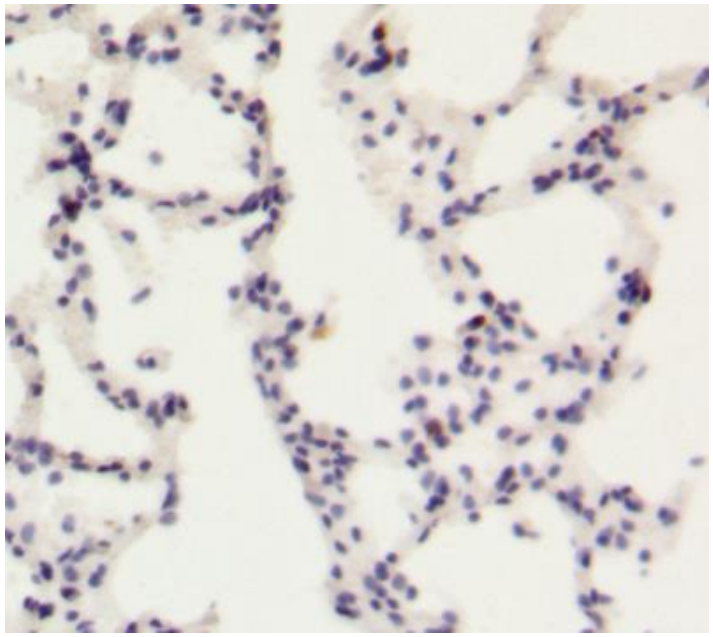


Not Enough Alveoli

- Post-mortem studies
- **Intrinsic** problem of EOS
- Apparent RX –> enlarge thorax <u>early</u> (<age 2?)



Normal



rabbit TIS Olsen et al

Reality ?

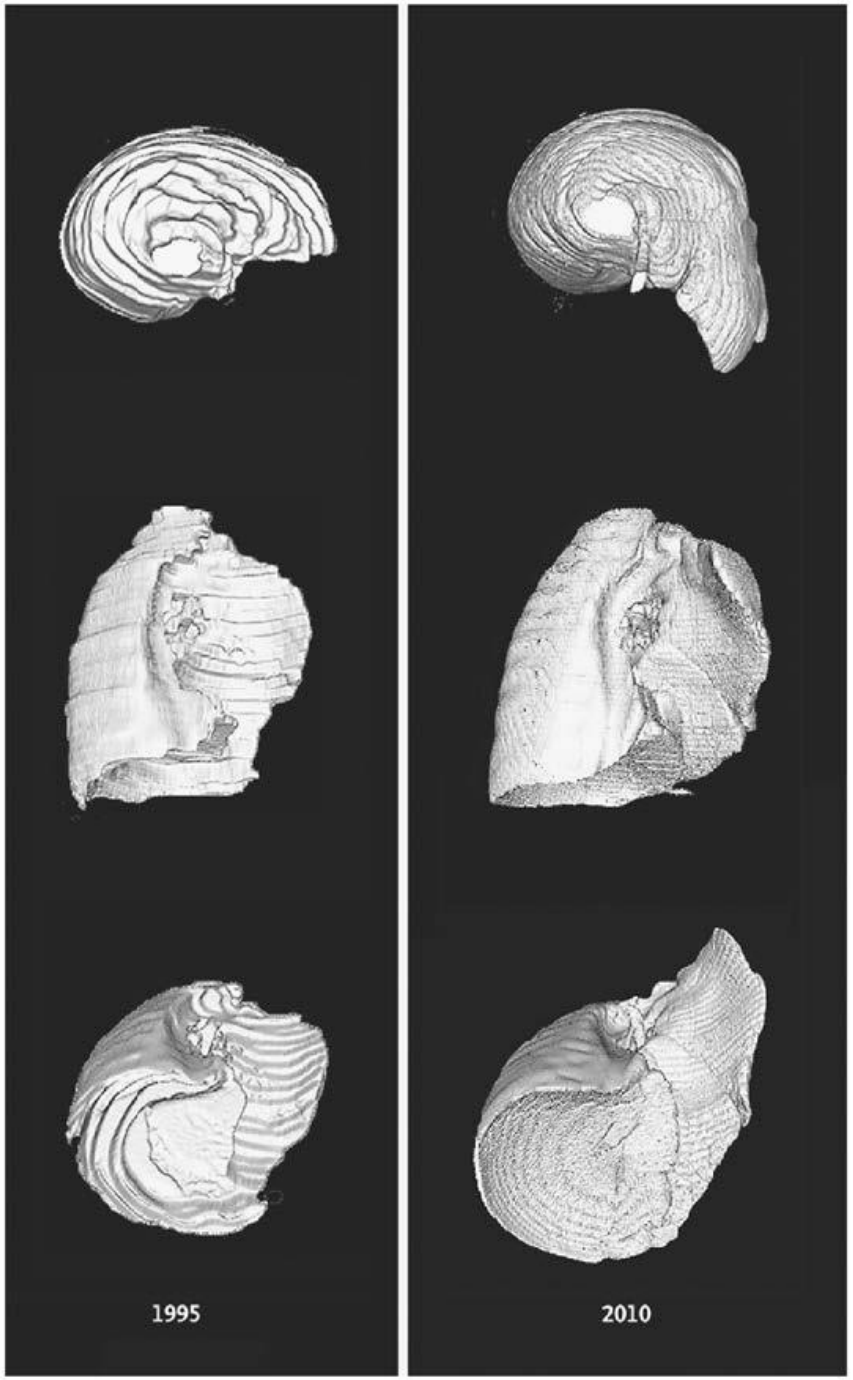
- No convincing histologic or clinical evidence that alveolar hyperplasia actually effected/enhanced by repeated expansions or lengthenings (Snyder models)
- Recent literature suggesting alveolar hyperplasia may occur in adolescence (Brown, Butler, Narayanan) - not necessarily terminated at age 8
- Doubling of thoracic volume > age 10 -> perfect time to exploit normal growth with expansion/lengthening techniques

Newer Data (Nopp et al 1997)

Parameters	0–1 months	1–3.5 months	4–12 months	13–24 months	25–38 months	Adult
Extracapillary vessels: radii, μm	30	38	42	46	50	70
Extracapillary blood volume, ml	17	40	60	80	118	300
Number of alveoli	50 M	60 M	70 M	80 M	90 M	300 M
Size of alveoli, μm	124	126	135	145	155	180
Lung filling factor	1.35	1.55	1.65	1.8	1.9	2.3
Tortuosity	1.5	1.8	1.8	1.8	1.85	1.9
Proportions of blood, membranes and intercellular fluid in alveolar walls	67/24/9	80/16/4	81/15/4	82/14/4	83/14/3	85/12/3

Brown et al (Am J Resp Crit Care 2012) : # alveoli increases from 90 million (age 2-3) to 300 million (adult)

"Alveolar dimensions determined by ^{3}HeMR ….is best explained by postulating that lung grows largely by neoalveolarization through childhood and adolescence. This contradicts the prevailing hypothesis that alveolarization is restricted to fetal life and early childhood"



Butler et al, *NEJM 2012* "Evidence for Adult Lung Growth in Humans"
15 year f/u pneumonectomy in 33 yo F

3D CT (shown)
Helium-3 MRI

"We hypothesize that, reminiscent of the role of stretch in lung development, **cyclic stretch** as such may be an important trigger for new lung growth."
[cyclic stretch = cycling, yoga]

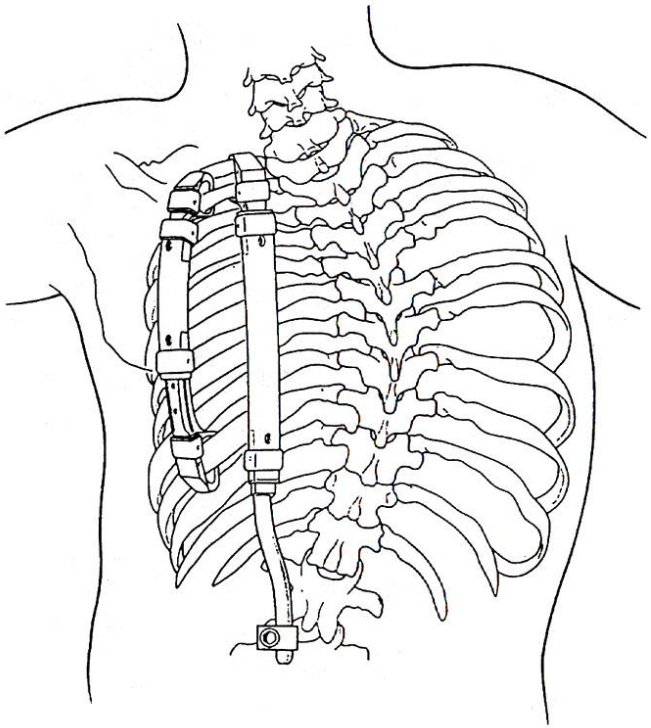
? Implication for chest walls that are frozen

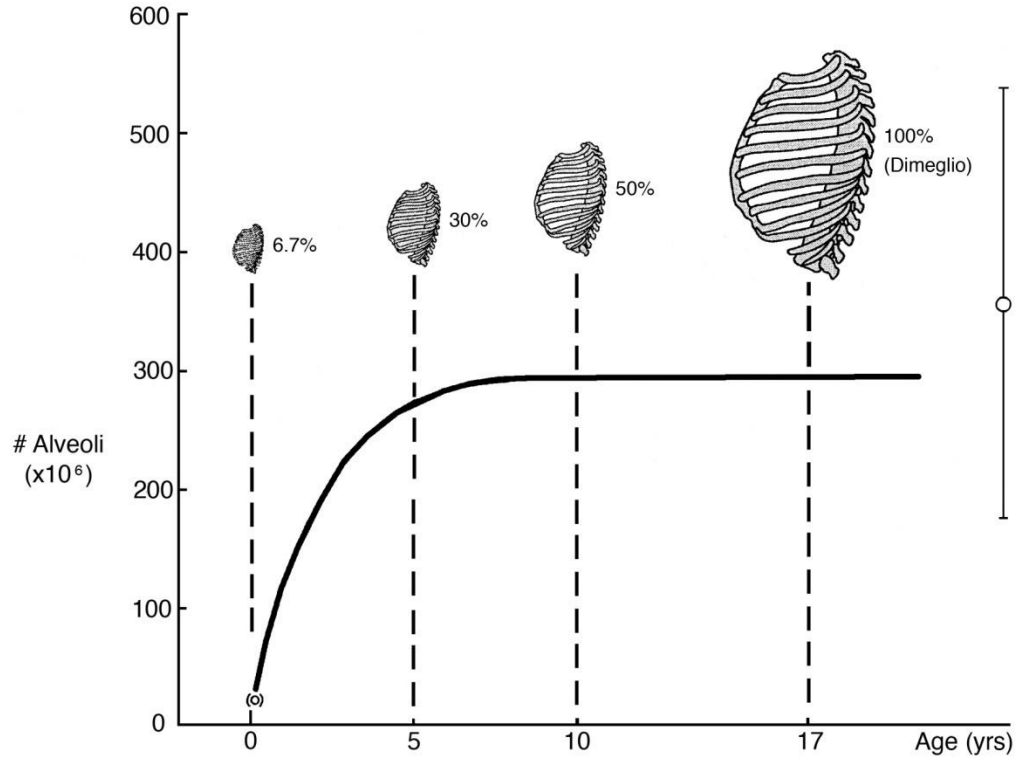
MRI with He-3 gas showed overall acinar-airway dimensions consistent with an **increase in alveolar number** rather than the enlargement of existing alveoli

? Implication if expansion could be proven to increase # alveoli

Growth of Thorax > age 10
Can chest wall implant impair normal circumferential growth?

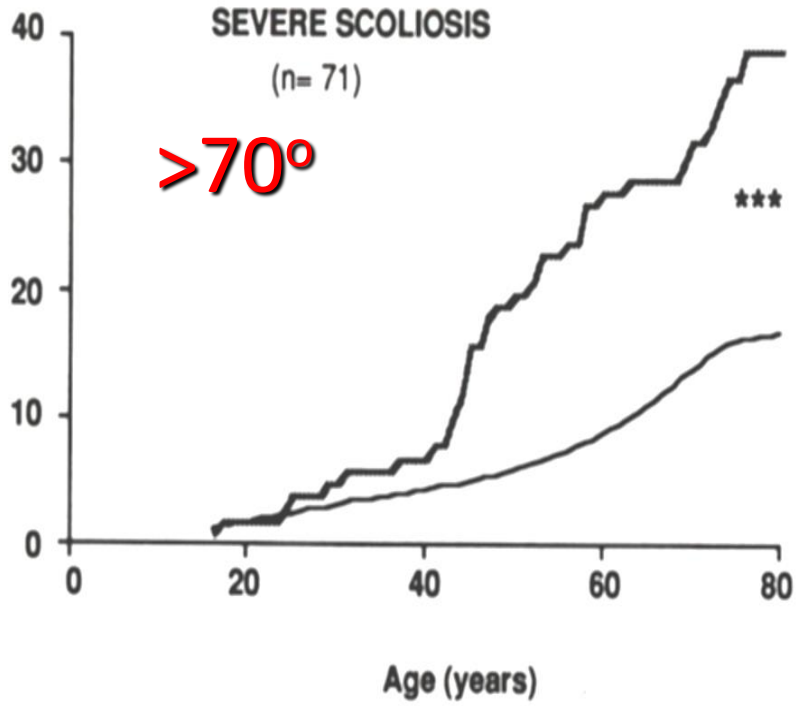
Charles, Dimeglio Spine '08; Dede et al JBJS '14





EOS RX – Prevention of T.I.S.

- Intrinsic – <u>early</u> thoracic enlargement
- **Extrinsic – control/correct deformity w/o growth inhibition**

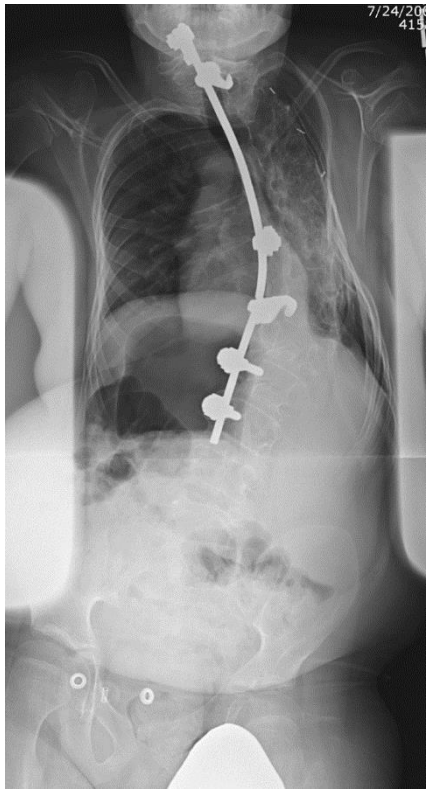


- Pehrsson
- Branthwaite
- Bergofsky

Nat'l Hx Ominous for PFT's <45% pred

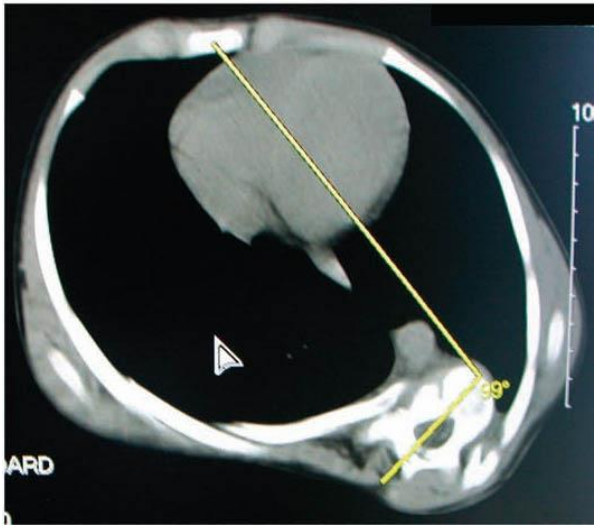
What Causes T.I.S. and its Respiratory Morbidity ?

- Early onset – intrinsic lack of alveoli
- Deformity – extrinsic chest wall dysfunction – attention to apex



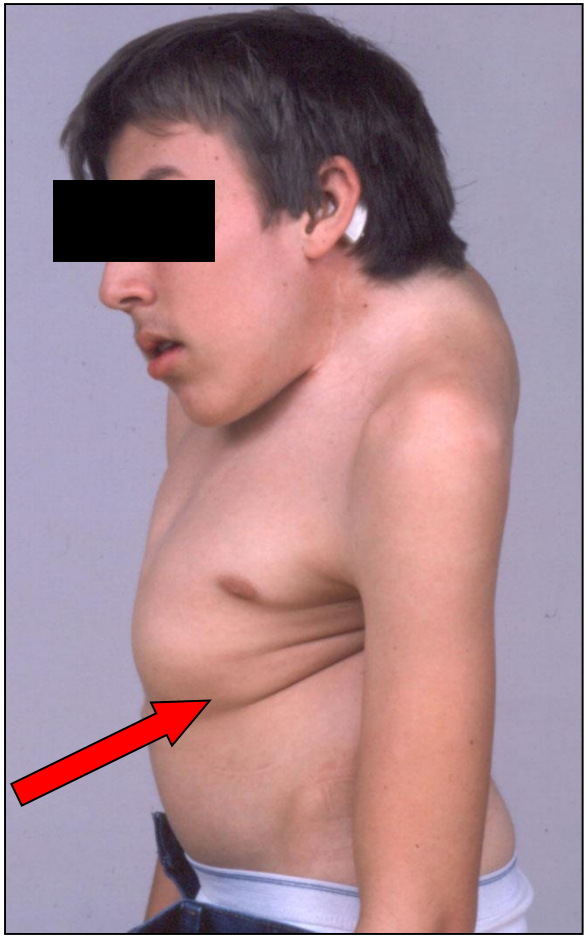
Extrinsic deformity of EOS impairs normal respiration

- Narrowing/stiffening of convex chest wall as rib hump increases – loss of compliance (=inability to change volume)



Spinal penetration

Windswept thorax



Concave intercostals non functional –volume not expandable

Caution – simply lengthening thorax / improve Cobb doesn't necessarily improve FVC (Mayer JPO '09)

PFT Summary – GR "graduates"

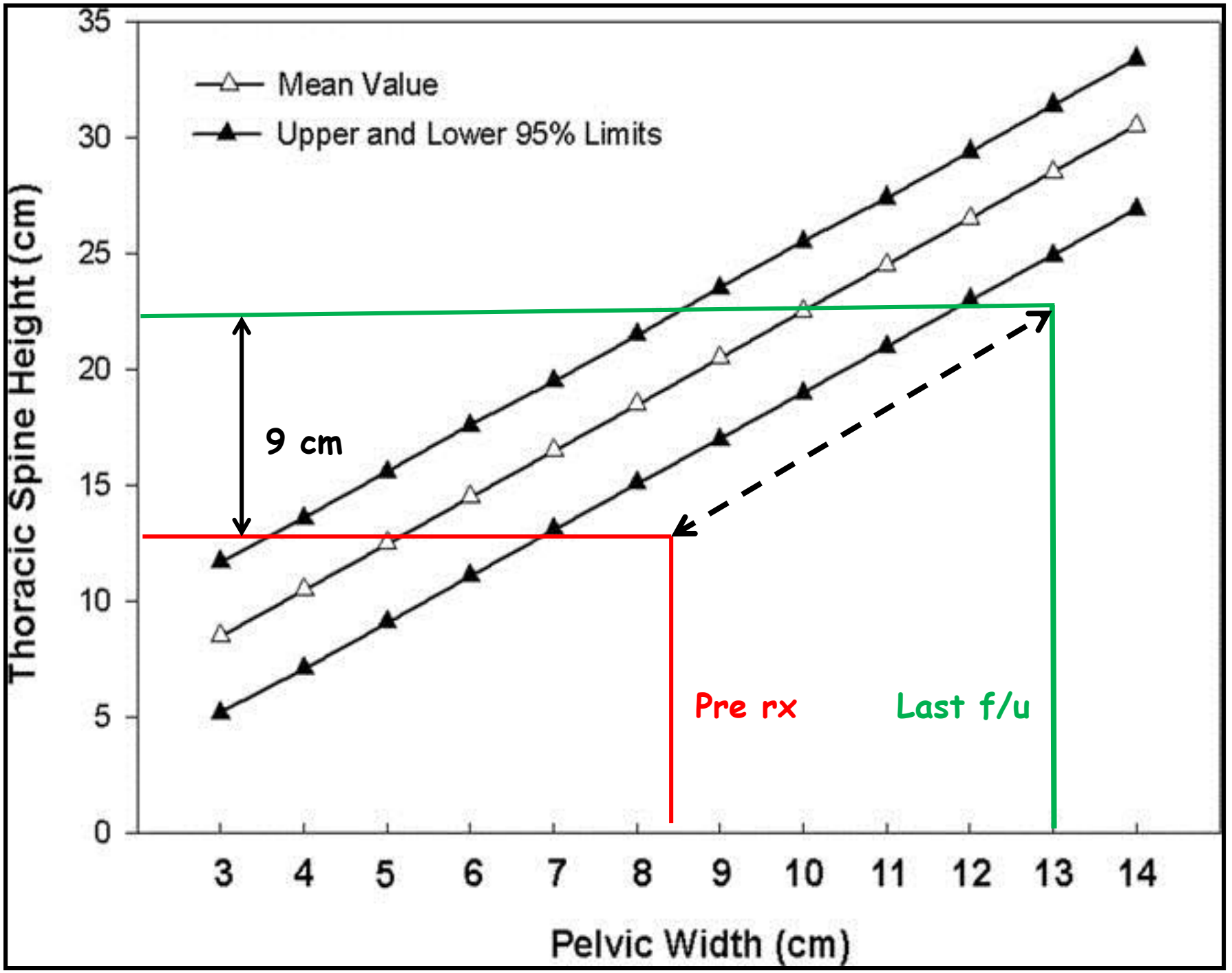
Johnston, JBJS 99-A:1036,2017

- FEV1 abs vol ↑ 900 cm^3 (200-1200)
- FVC abs vol ↑ 1100 cm^3 (100-1800)

- FEV1 %pred ↓ 1.7 % (52.1%)
- FVC %pred ↑ 1.8% (55.3%)

= no change

- over 6.7 yr f/u (5-11 yr)


Mean Value
Upper and Lower 95% Limits
Thoracic Spine Height (cm)
Pelvic Width (cm)
9 cm
Pre rx
Last f/u
0
5
10
15
20
25
30
35
3
4
5
6
7
8
9
10
11
12
13
14

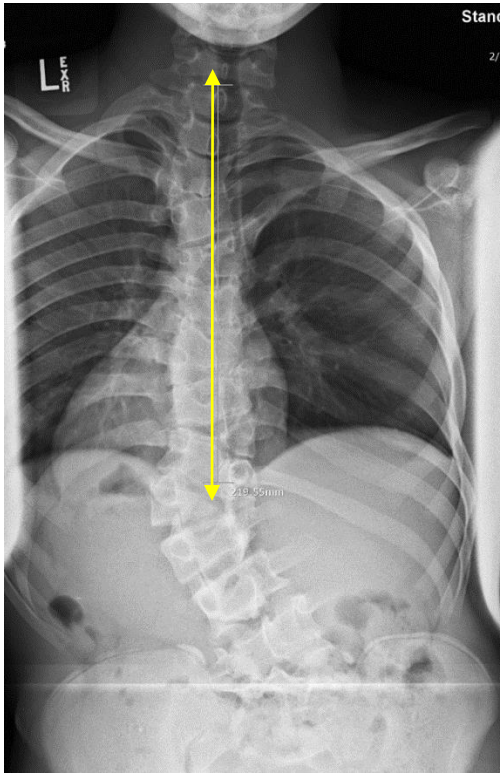
Summary : Just Keeping Up….

- Th spine length gain (mean 9 cm) <u>parallels</u> normal growth pattern –
  initial <5th %ile, end up w/ same %ile
- PFT's at f/u same % pred values as earliest measurement in spite of 1 L increase in absolute volume
- **Stretching length-wise isn't enough**
- **Circumference needed**

"Short & straight is better than long and crooked"

Short & crooked -> thoracic insufficiency

Just short/not crooked…….may be OK



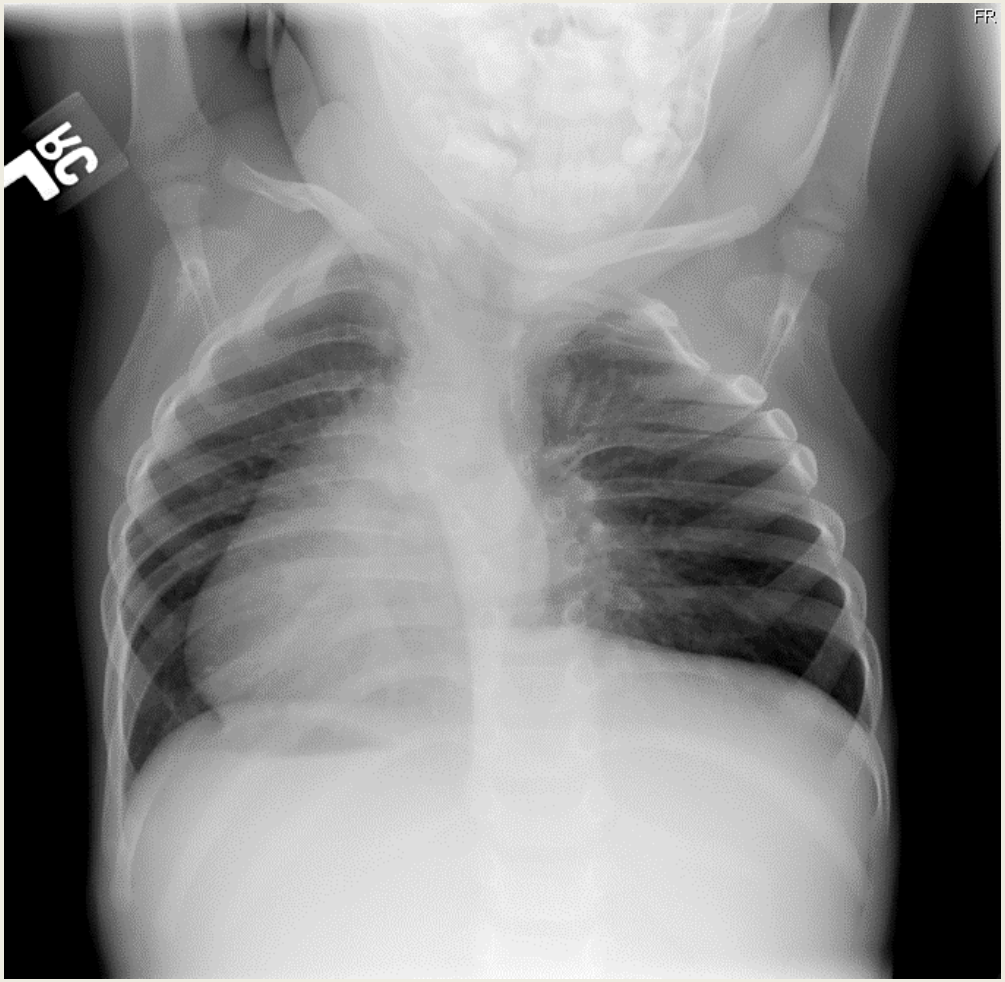
FVC = 2.63 l. 56% pred

T1-12 20cm

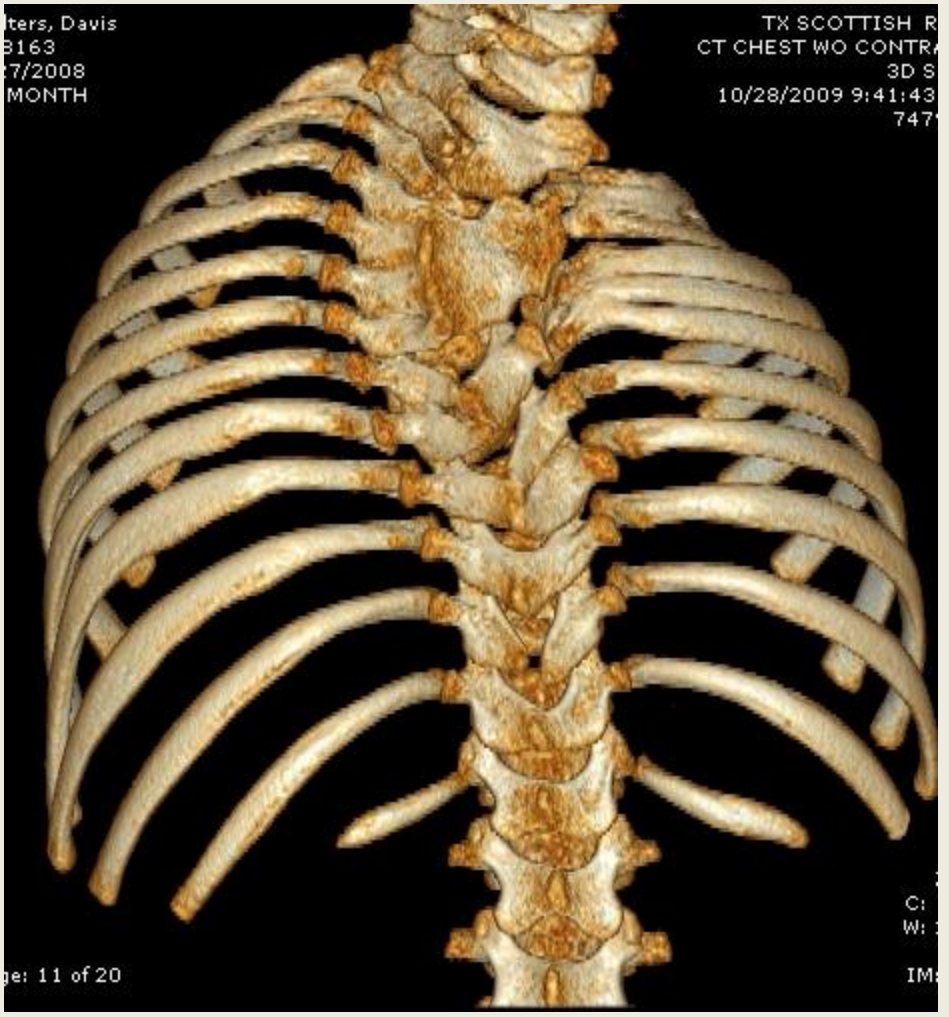
- Absence of rib/chest wall dysfunction (circumferential growth)
- Ineffective early deformity surgery = culprit (? early in general)
- "in situ fusion" for deformity is obsolete

Case in point – potential TIS ?

- Cxray 18 mo male otherwise healthy
- Austin patient

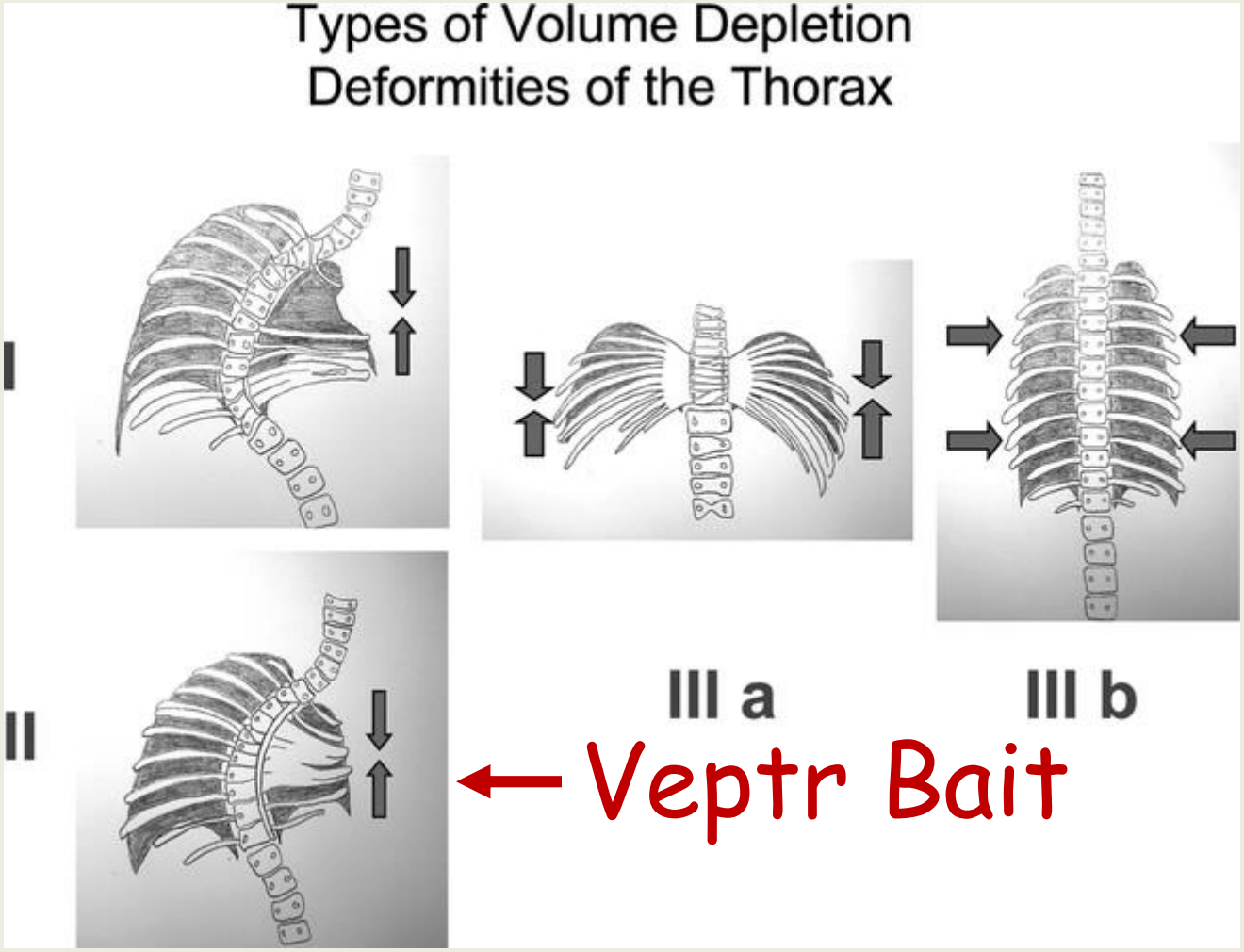


CT



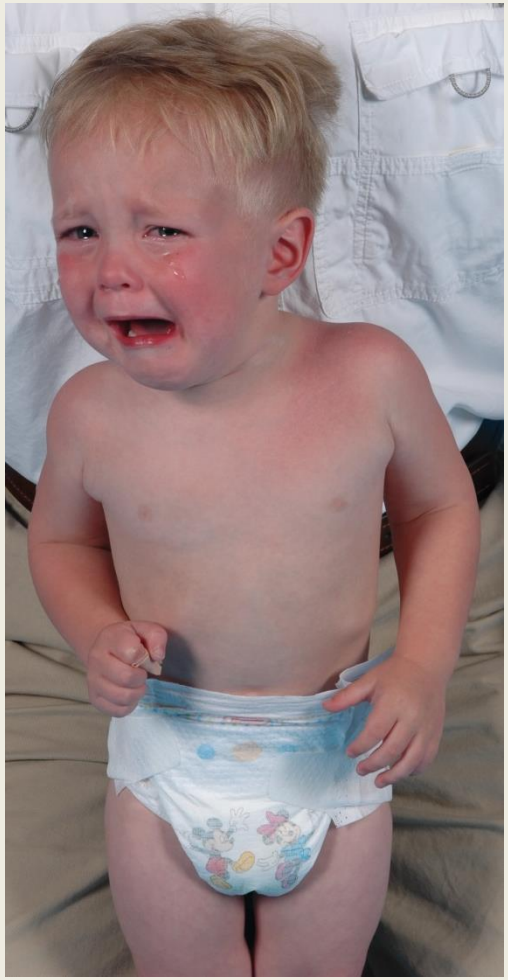
T1-T7 congenital w/ rib fusions

Campbell

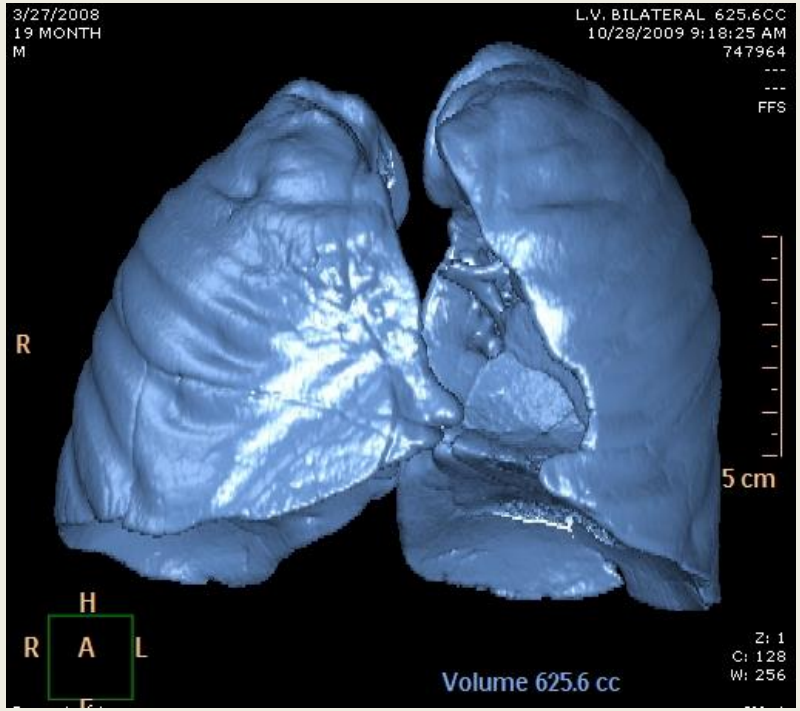


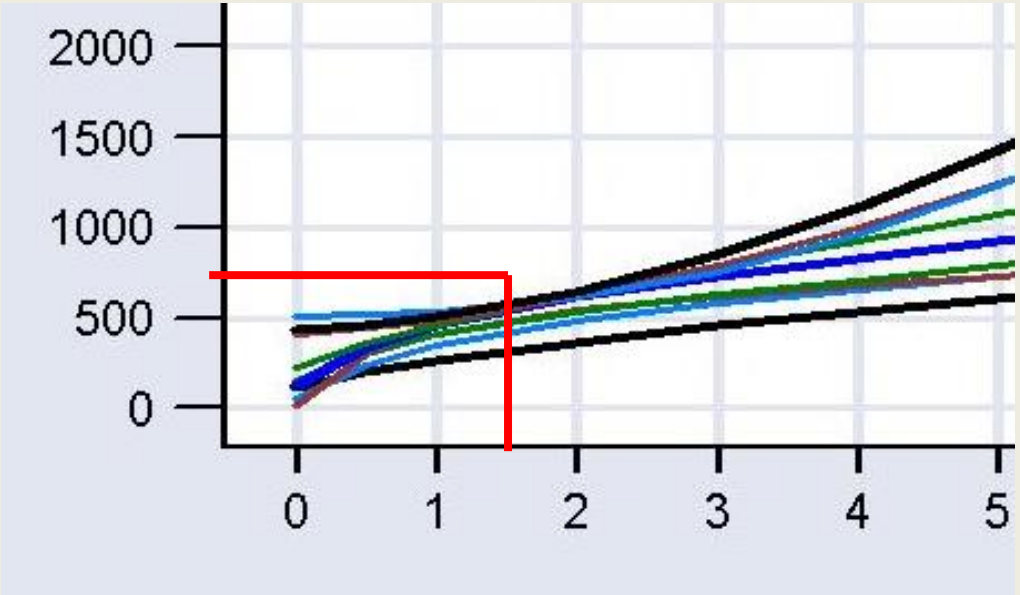
← Veptr Bait

Clinical – what do we treat?



~~Subjective~~ Pulmonary





19/12 625 cc

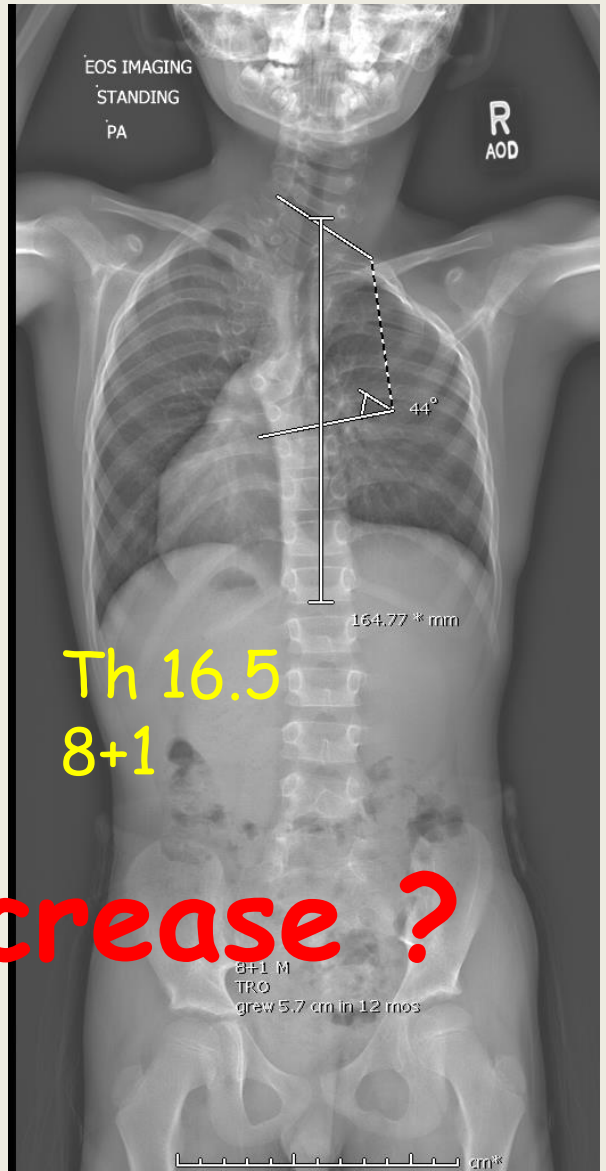
		PFT's FVC	%prd	FEV1	%prd
4/14	6yr	1.06	121	0.98	123
5/16	8yr	1.38	106	tf	

Tf – tech fail

Rx: observation….. in spite of Canavese/Karol death trap

25 mo

Short…..
But it's not crooked



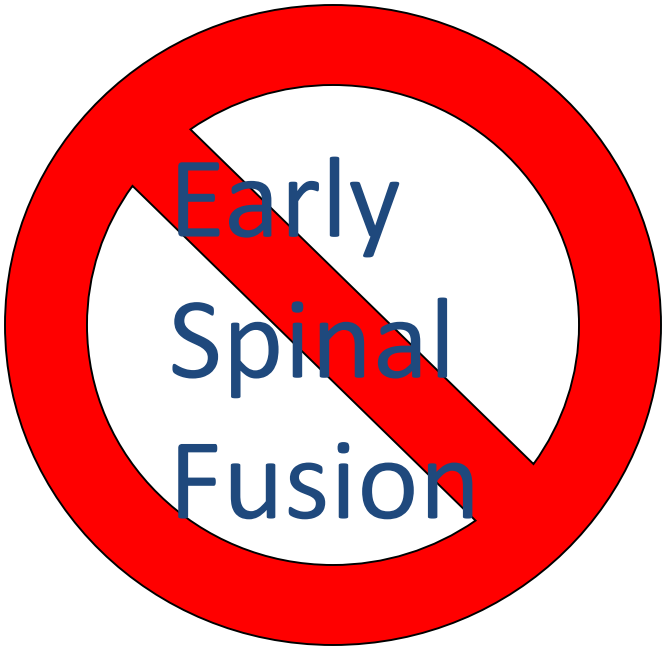
Circumference increase ?

The basics - Fusion prior age 4-5

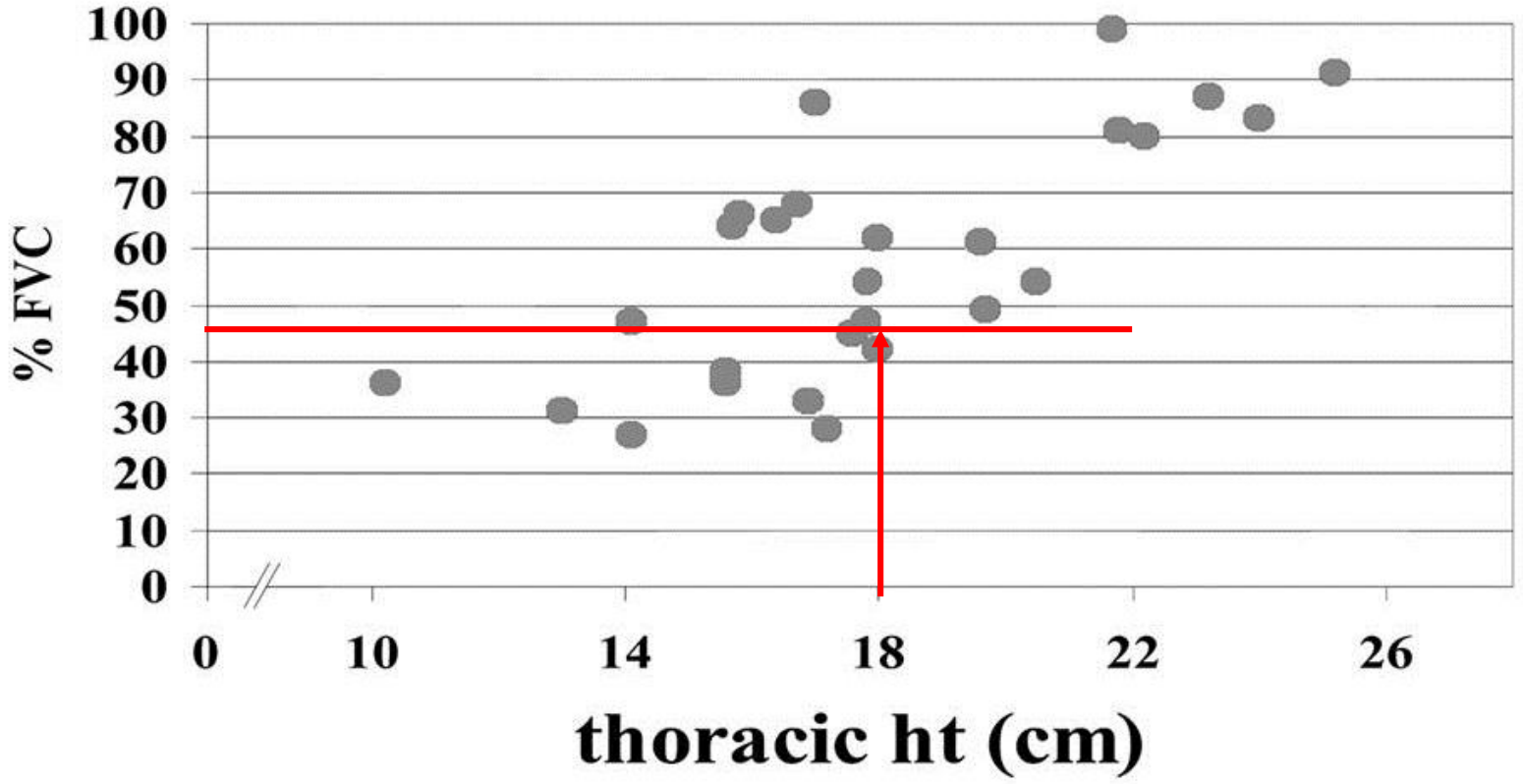
- Goldberg ('03) – "….early surgery, even with anterior growth arrest…did not halt the deformation of scoliosis and did not reliably preserve respiratory function in this group whose scoliosis presented before age 4."

- Emans ('04)
- Karol ('08)
- Vitale ('08)
- Typical PFT's 20-50% pred. when tested 10 yr later

Critical fact : little or no correction of the scoliosis… in situ fusion did not correct the spine and chest wall deformities



Goal of RX: T1-12 length > 18 cm
But deformity correction also critical



Karol L. A. et.al. J Bone Joint Surg 2008:90:1272-1281

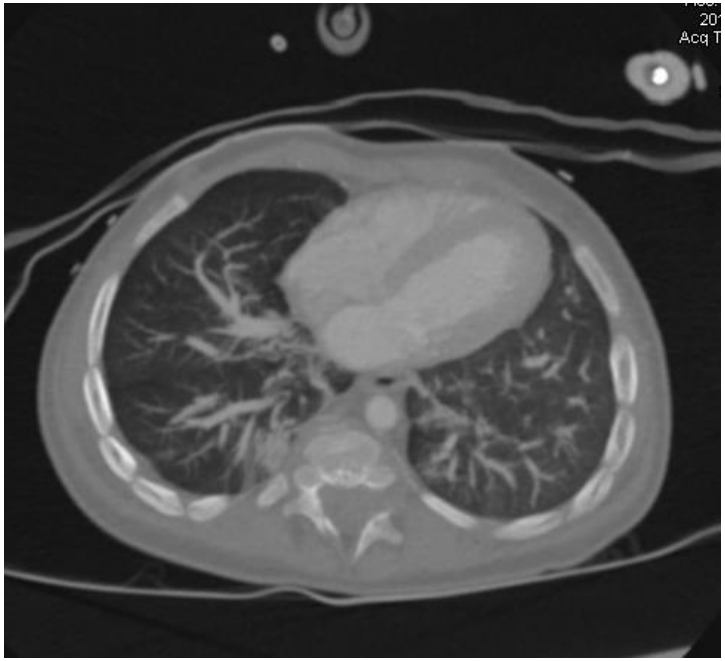
Apert case 10 mo

Trach'd as newborn for cranio-facial surgeries

T1-12=8.4 cm

Is this thoracic insufficiency syndrome ?

Normal chest wall / ribs

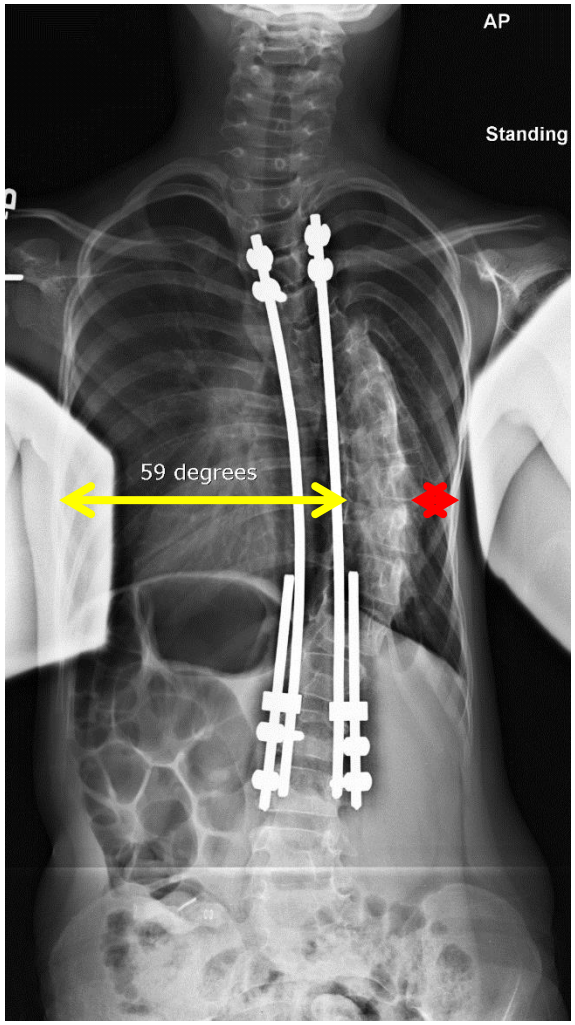


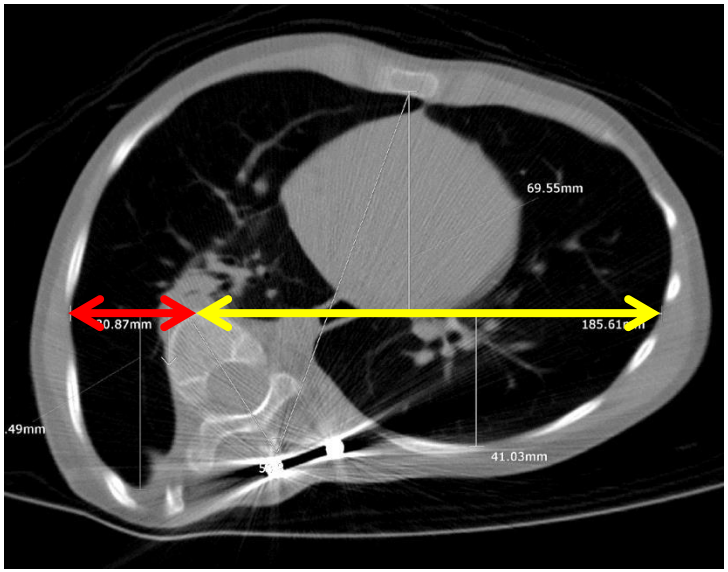
Spine lengthening -> minimal increase possible, patient will be short-stature

Leave chest wall alone, let circumferential growth occur

Jeune approach – expand thorax transversely?

Apical Windswept Deformity = TIS in Idiopathic Disguise

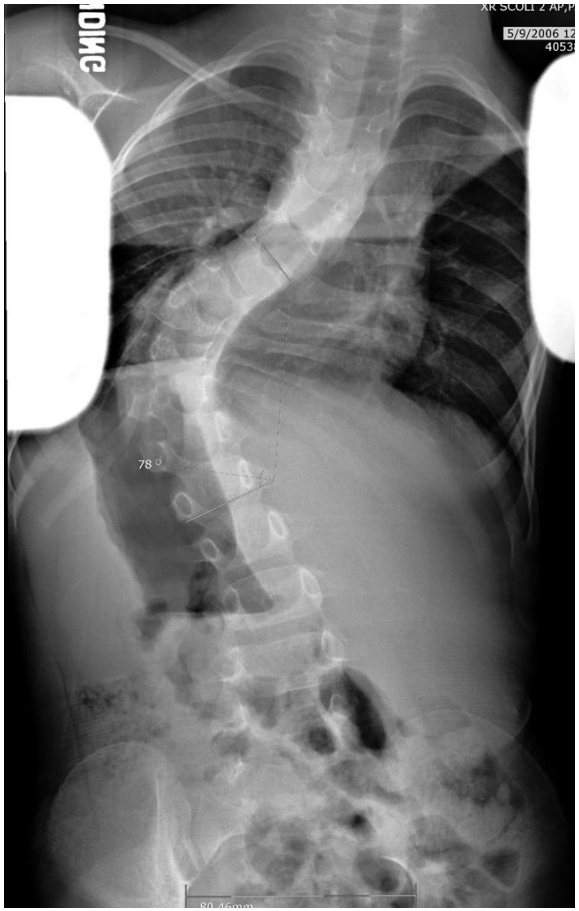


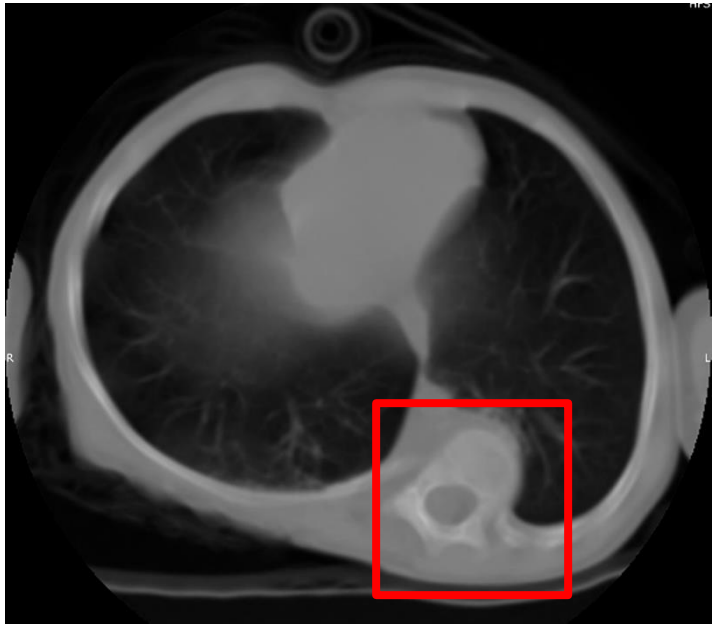


Apical penetration into convex hemithorax

Hemithoracic Ratio should be ~ 1

What is Chest Penetration ?
Attempt to understand Dubousset





Scolioses thoraciques : les gibbosités exo et endo thoraciques et l'index de pénétration rachidienne

Thoracic lordoscoliosis: exothoracic and endothoracic de and the spinal penetration index

J. Dubousset *, Ph. Wicart *, V. Pomero **, A. Barois ***, B. Estourn

* Hôpital Saint-Vincent-de-Paul, 82, avenue Denfert-Rochereau, 75014 Paris.
Université René Descartes, 12, rue de l'Ecole de médecine, 75006 Paris.
** Ecole Nationale des Arts et Métiers, 63, rue Olivier de Serres, 75015 Paris.
*** Hôpital Raymond Poincaré, 104, boulevard Raymond Poincaré, 92380 Garches.

Rev Chir Orthop 2002

ABSTRACT

Purpose of the study

We reviewed retrospectively our patients with thoracic lordoscoliosis and with airway compression and atelectasia due to anterior protrusion of pathological conditions involved and the management methods used. quantifying thoracic deformation. The individual cases discussed here hav analysis to date.

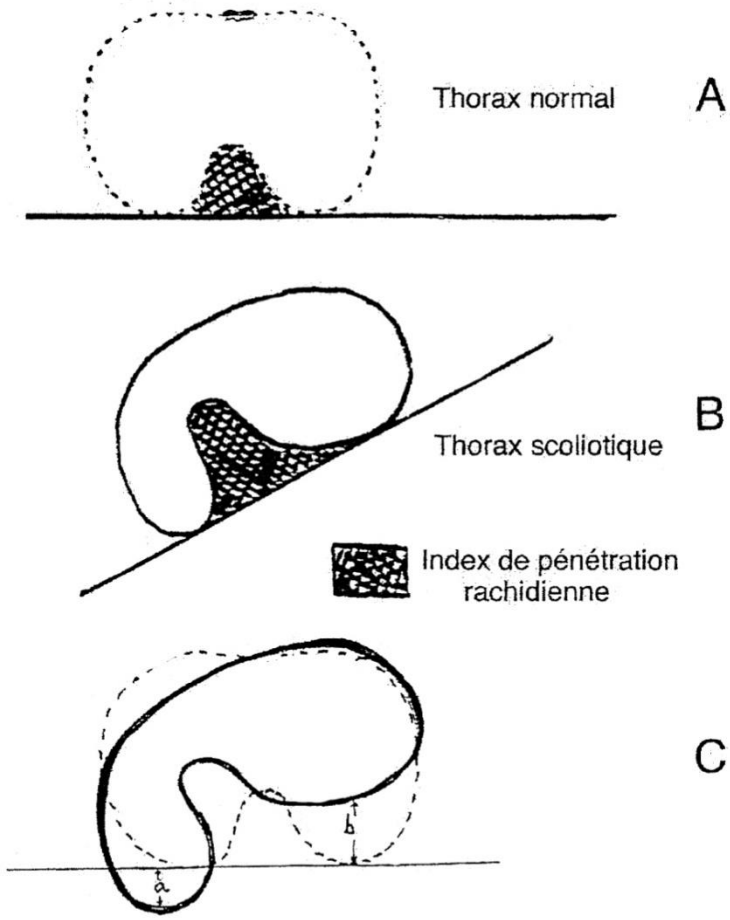
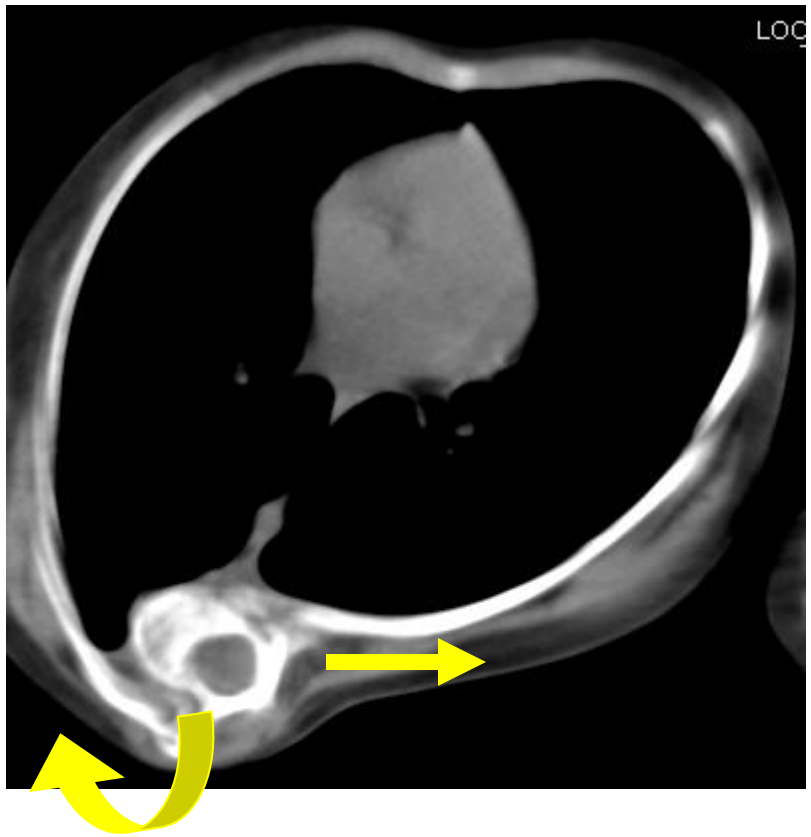


FIG. 1. – *A. Thorax normal en pointillé. B. Thorax scoliotique en trait plein. L'index de pénétration rachidienne = % de surface ou volume occupé par la pénétration des corps vertébraux et des structures attenantes rapporté à surface ou volume théorique calculé à partir d'une tangente au bord postérieur des côtes droites et gauches. C. Comparaison entre le contour thoracique normal (pointillés) et un contour thoracique scoliotique où l'on peut constater : a) La gibbosité exo thoracique convexe (en plus). b) Le manque thoracique concave (en moins).*

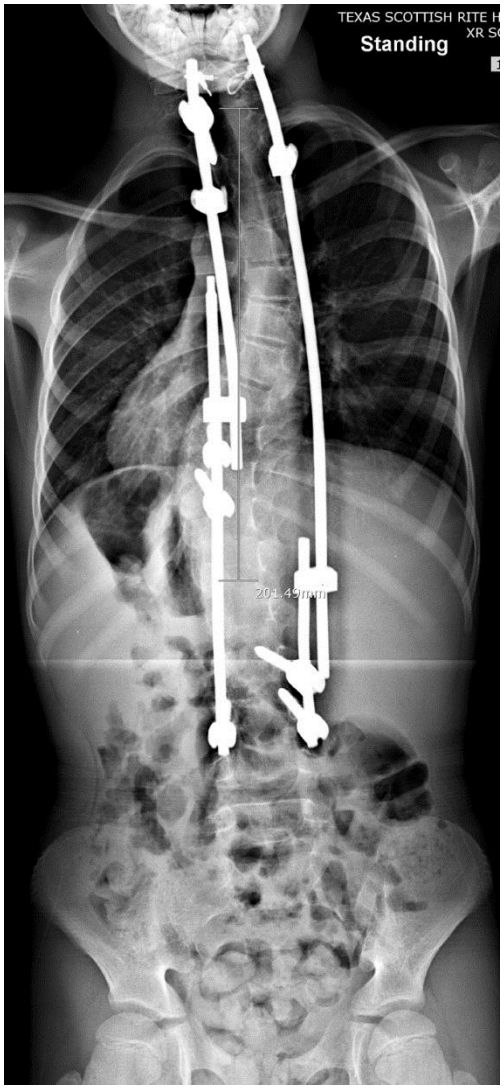
GR's + apical fusion -> poor outcomes (Thompson,Akbarnia)

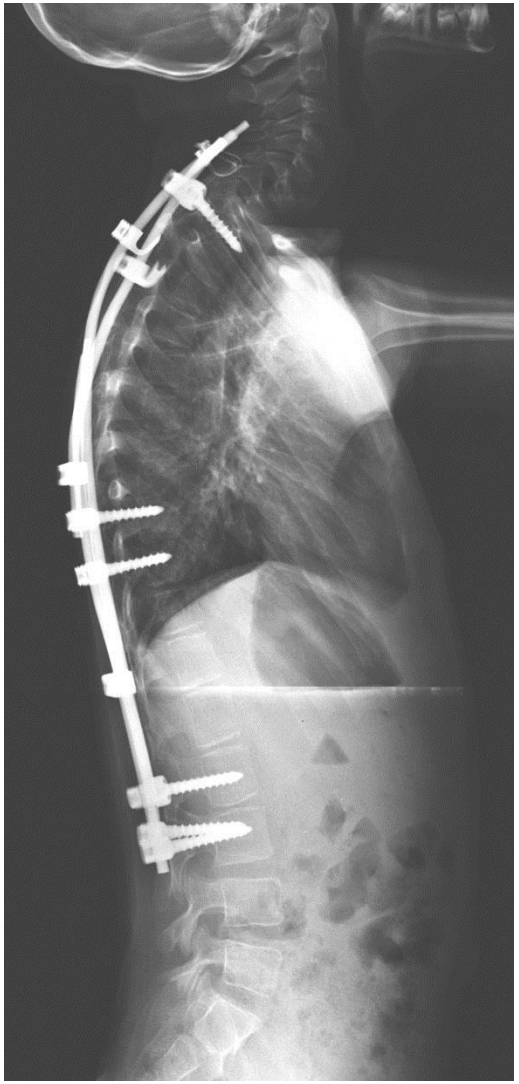
1. Lack of apical control by implants
2. "in situ" fusion of most deformed part -> ? ineffective to control deformity (= crankshaft)
3. Apical fusion is NOT apical control due to lack of correction



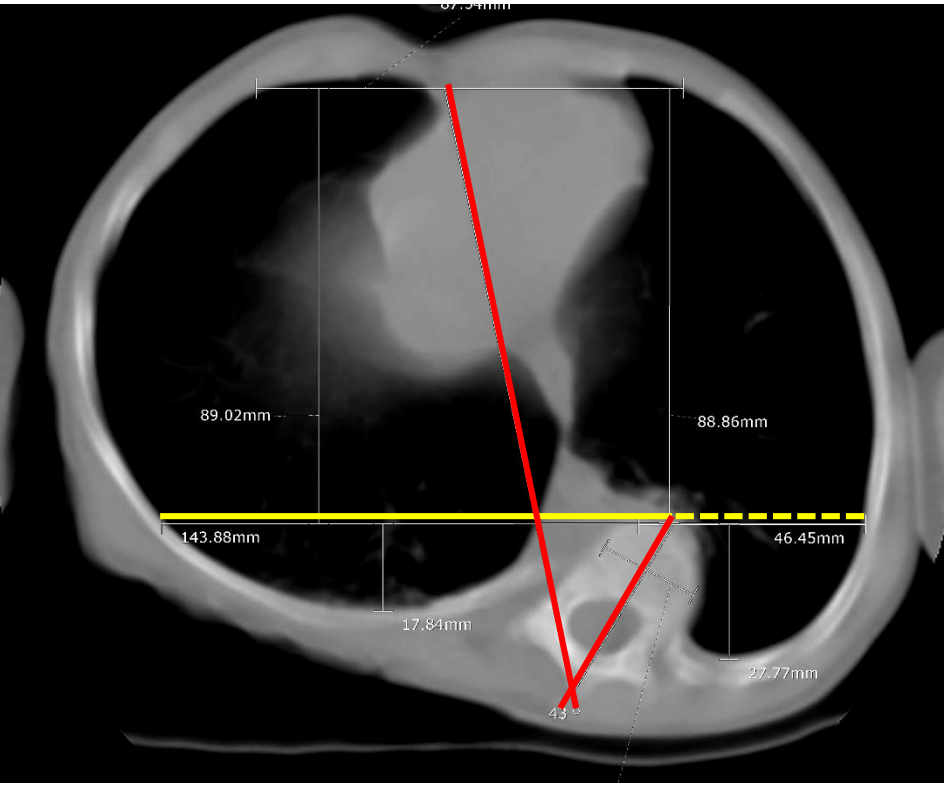
APICAL CONTROL

Postop Correction / 2 D





3 D Visual Correction

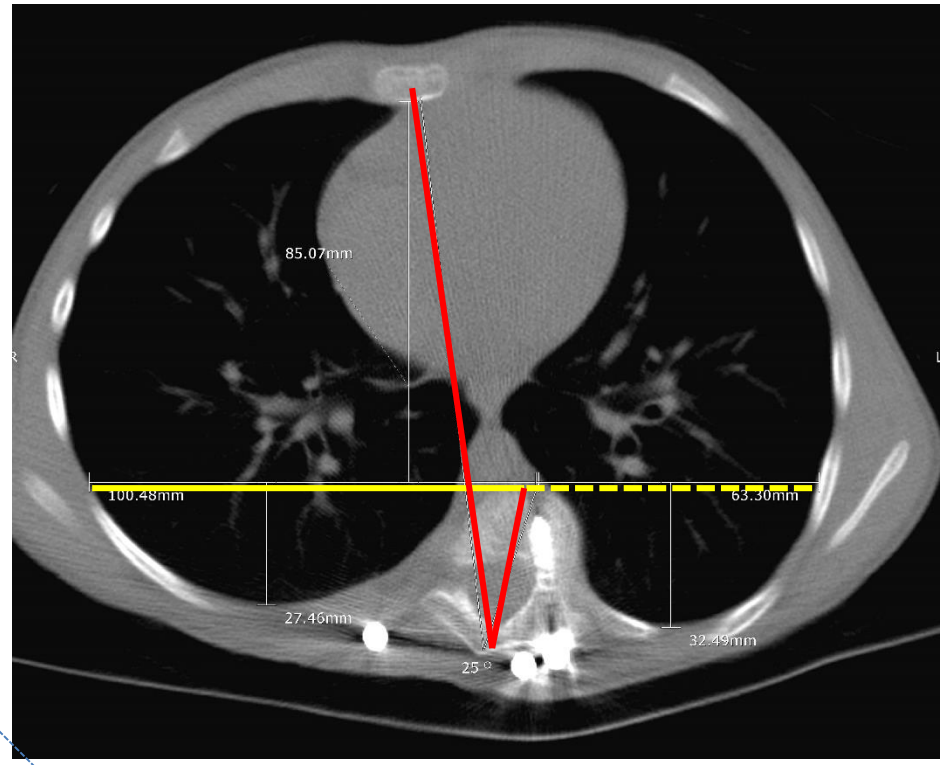


Postop

Cvx/Cav = 63/100 = .63

AVR = 25°

A/Pcav = 3.1 A/P cvx = 2.6



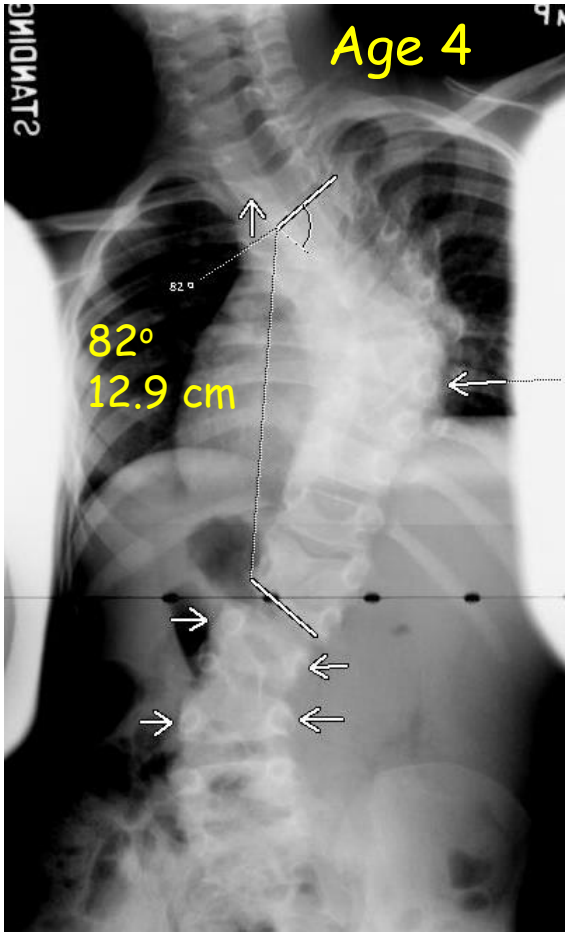
Preop

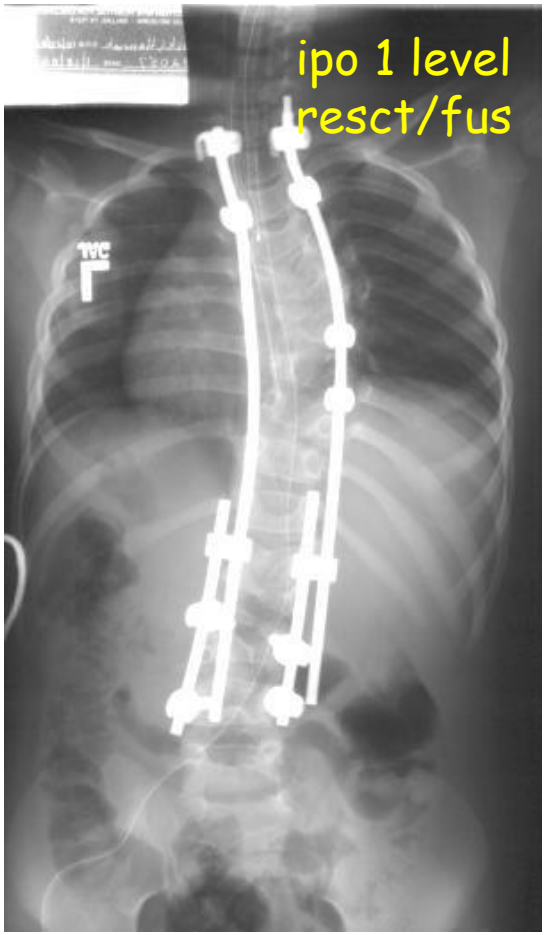
Cvx/Cav = 46.5/144= .32

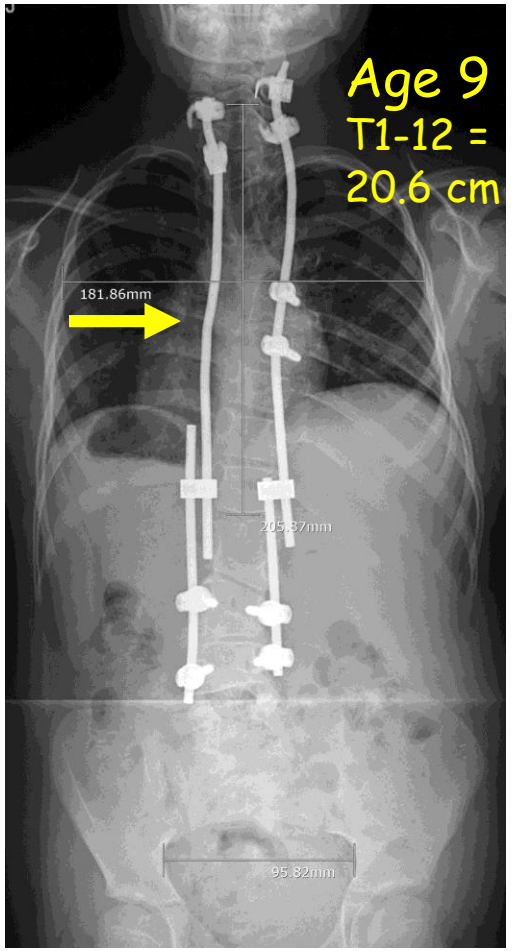
AVR = 43°

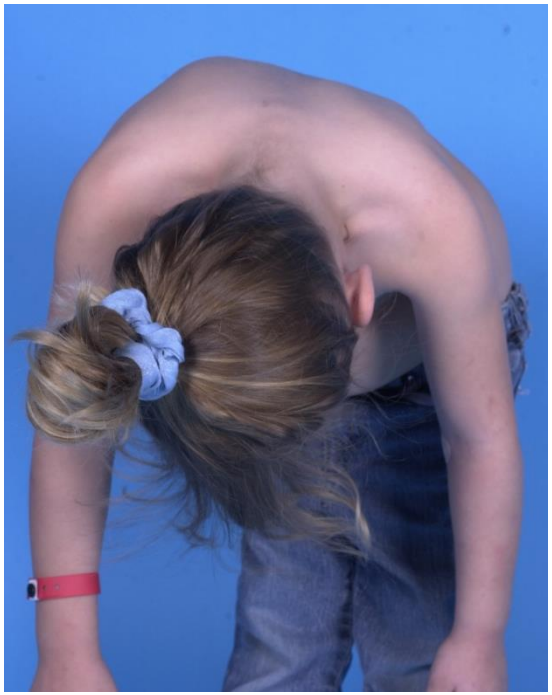
A/P cav = 5 A/P cvx = 3.2

Early rx must correct or prevent progressive spinal deformity producing windswept thorax







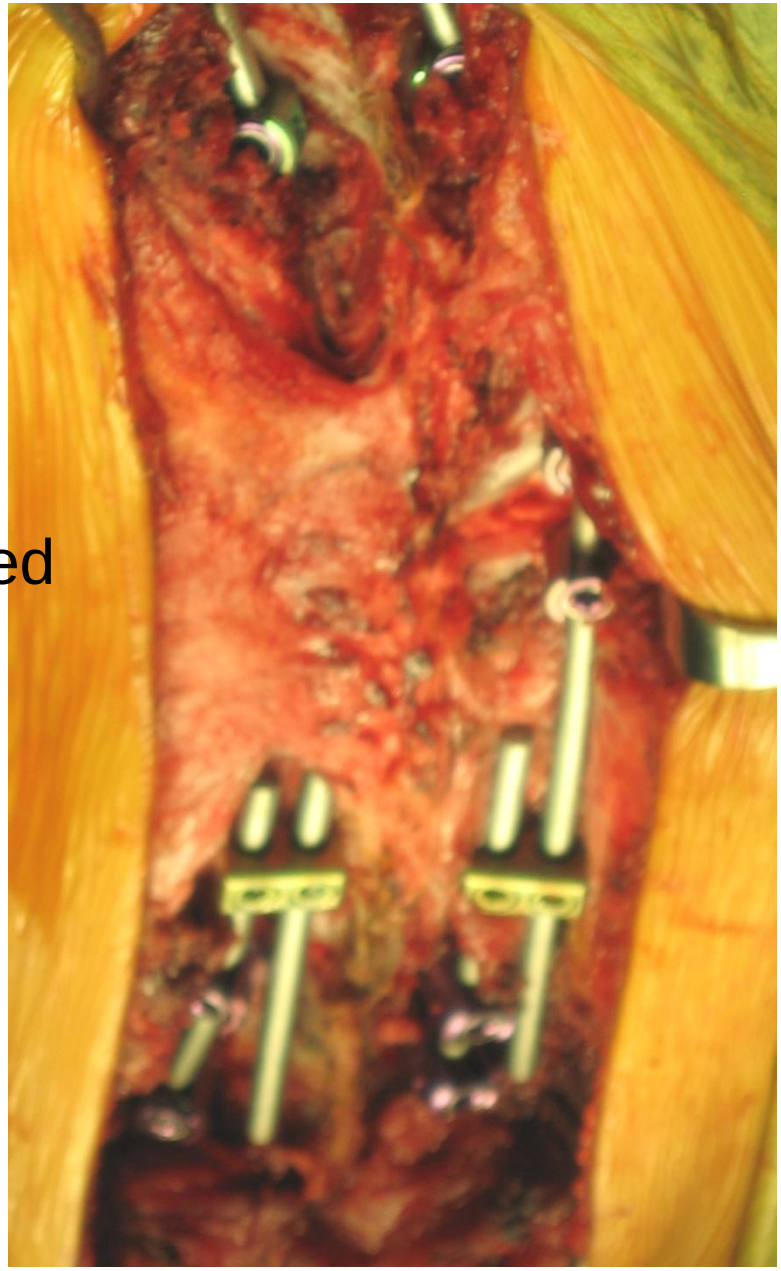


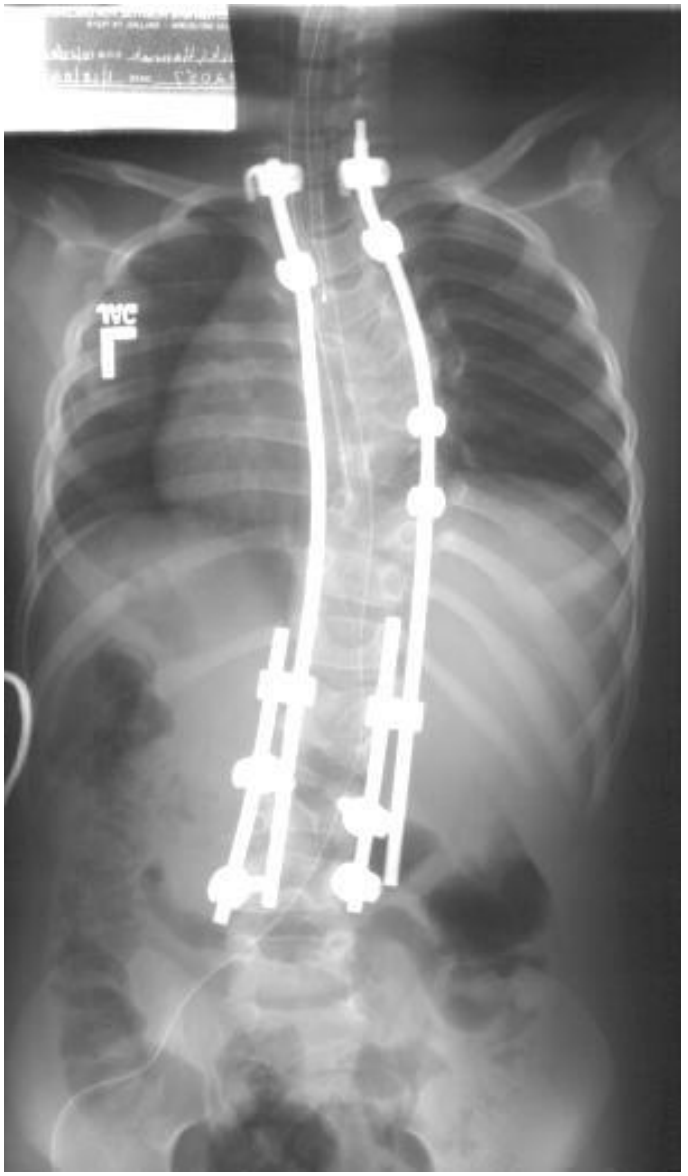
Age 4 s/p
de-tether

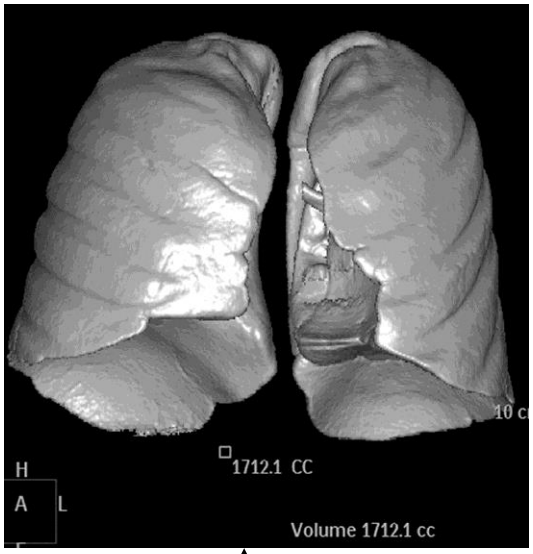
i.p.o.

Concavity not exposed

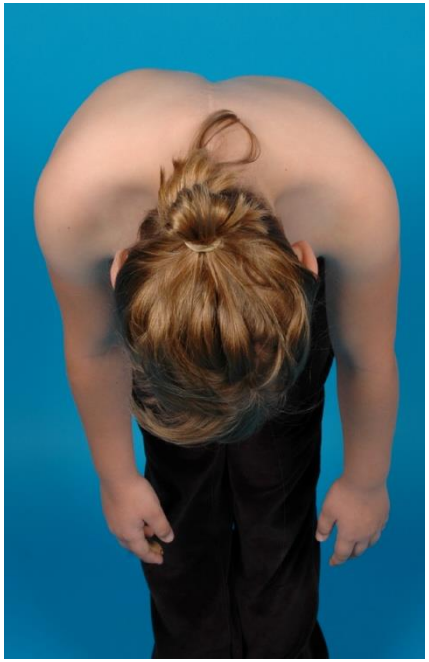
Minimal acute distraction (mep Δ)

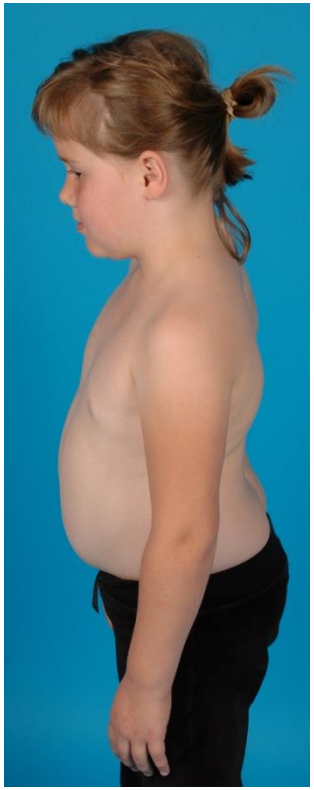


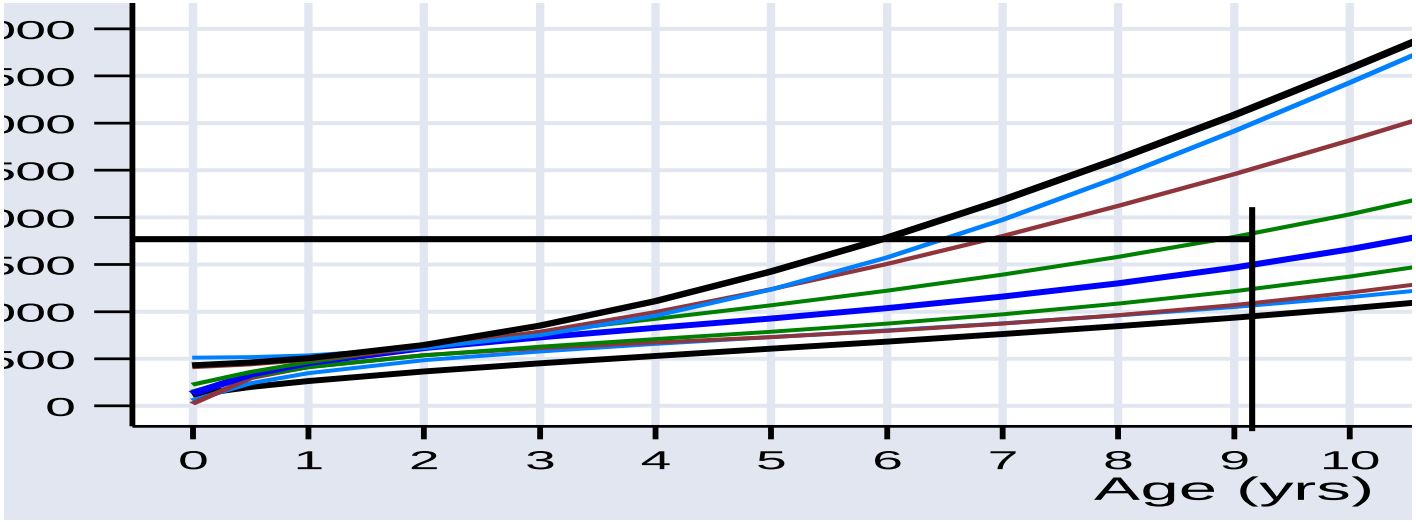




CT volume age 9+2





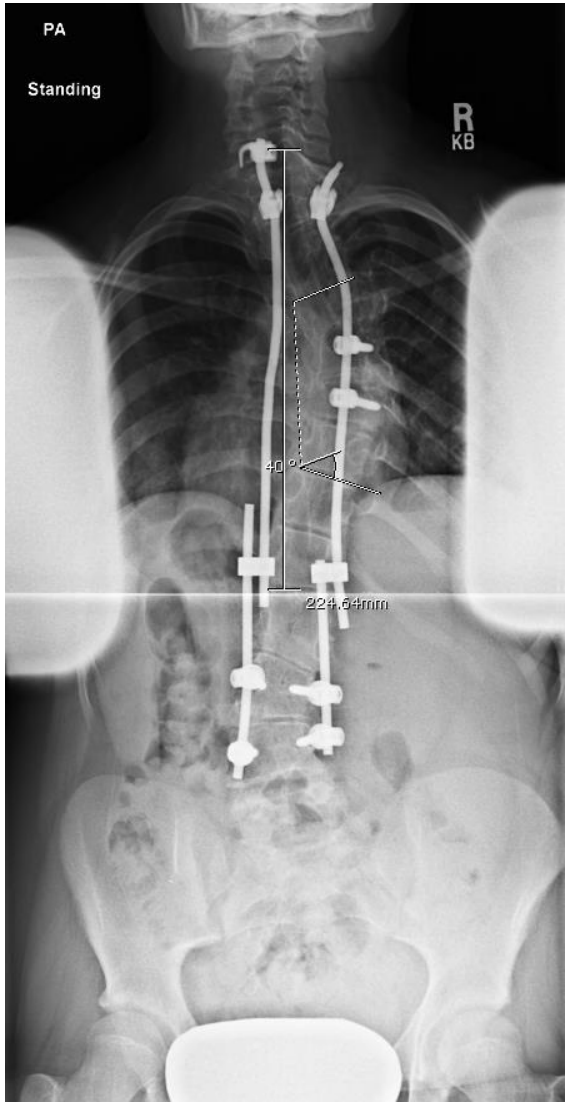


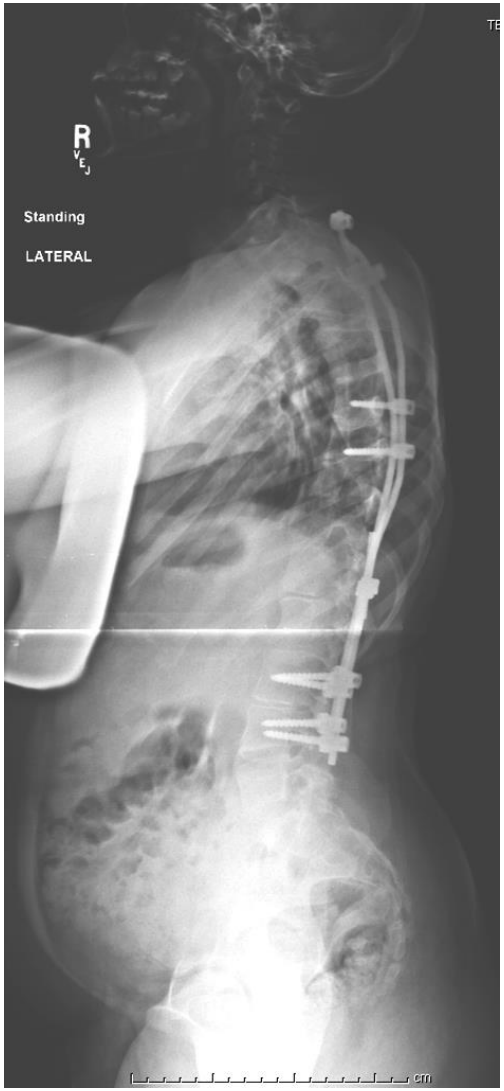
FVC = 1.05 L. (70% pred)
FEV1 = 1.05 L. (78%)
ETCO2 38 (nl.)

[age 12]

f/u 11/12, age 12+1

T1-12 = 22.4, 40^{o} (12.9, 82^{o} to start)





Last lengthening 4/27/10

No change in Cobb since ipo

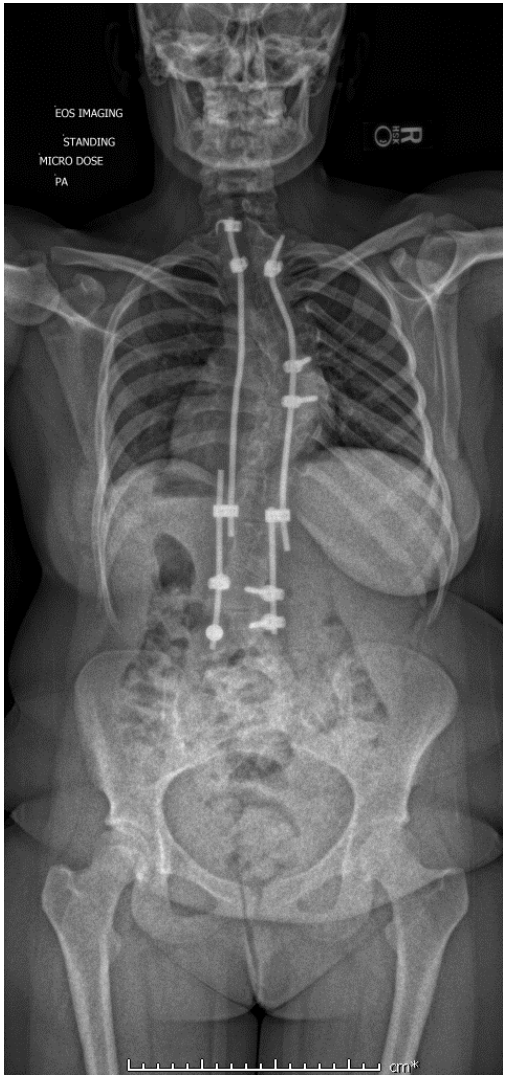
TRC now closed

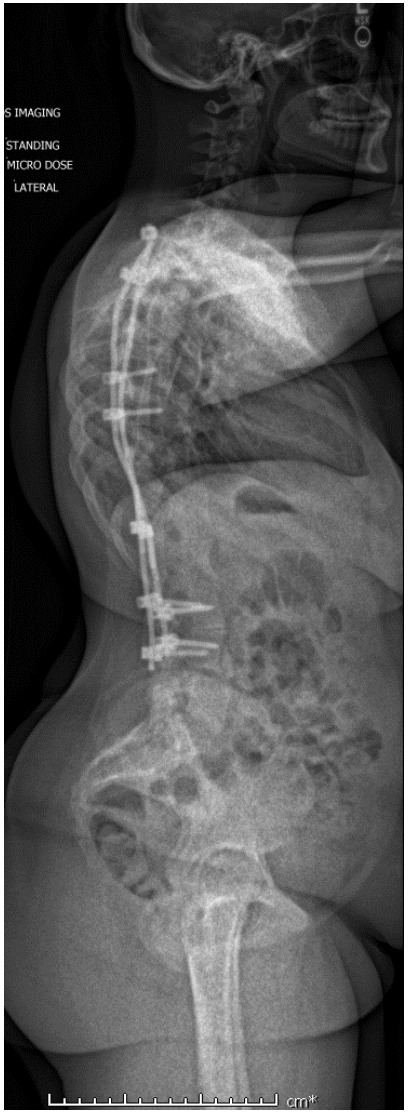
T1-12 gain > 9 cm

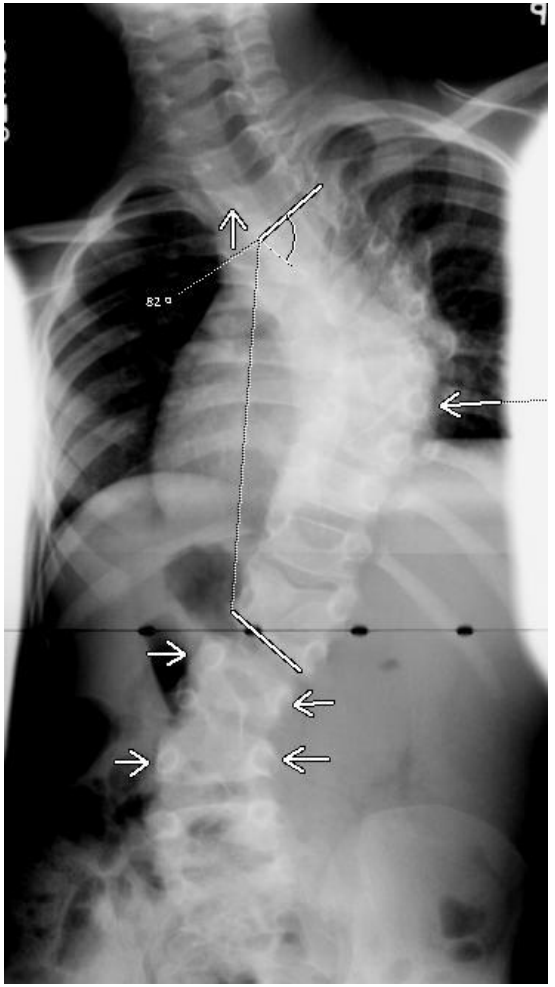
Never touched chest wall

No final fusion necessary to date

No further lengthening age 16



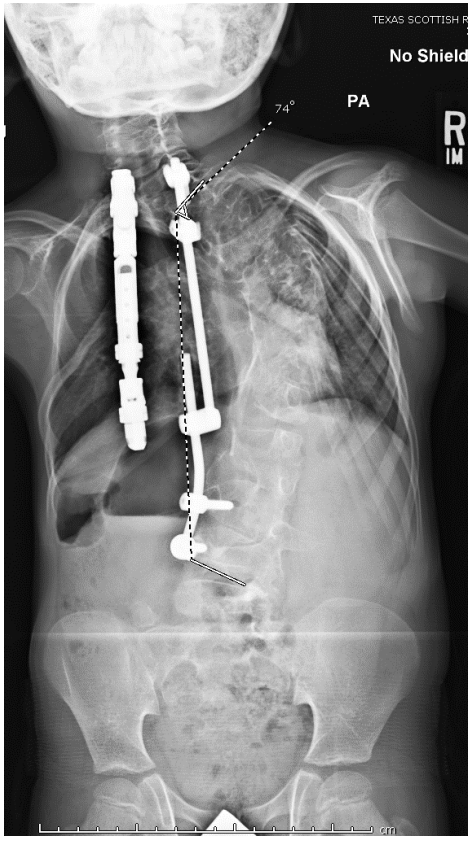




Age 4

Chest Wall Expansion (?) w/o Curve Correction/apical control -> No Improvement in PFT = Fusion in situ w/o correction

- If rx starts early and we're ineffective -> **inflict pain w/ no gain**



Age 7 – 10 surgeries later

Dede, Motoyama et al JBJS 2014
Pulmonary and radiographic outcomes of VEPTR
Age 4.8 yr /11 expansions/ 6 yr f/u

	Pre-implant	1st Expansion	Last FU	P
Cobb (degrees)	80	68	67	0.002
Maximum thoracic kyphosis (degrees)	57	50	66	0.08
T1-T12 height (mm)	123	131	149	0.054
Crs/kg	1.4	1.2	0.9	0.0006
FVC (L)	0.65	0.68	0.96	<0.0001
FVC% arm	77	77	58	0.0001
SAL	0.77	0.80	0.87	0.006

T1-12=14.9 cm ……NOT NEARLY ENOUGH (Karol et al *JBJS '08*)

EOS & Pulmonary Function - Summary

- Lengthen spine (serial procedures) -> objective ↑ FVC or FEV_1 pred. not seen
- Lengthen/expand chest wall -> definitely less compliance, less length, ↓ 'd FVC/FEV_1 pred.
- More attention to apical correction, circumference / diaphragm P.T. (cyclic motion)???
- MCGR be a game-changer re: satisfaction, emotion/mental health?
- McCarthy's solution

Greetings from Big D

TEXAS
SCOTTISH RITE HOSPITAL
FOR CHILDREN