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

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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

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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 $\rightarrow$ enlarge thorax **early** ($<$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
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** - early 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
- **Defirmity** - 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 cm³ (200-1200)
- FVC abs vol: 1100 cm³ (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)
Pre rx: 9 cm

Last f/u: 9 cm
Summary: Just Keeping Up….

• Th spine length gain (mean 9 cm) parallels normal growth pattern – initial <5\textsuperscript{th} \%\textsubscript{ile}, end up w/ same \%\textsubscript{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

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- “in situ fusion” for deformity is obsolete

FVC = 2.63 l. 56% pred
T1-12 20cm
Case in point - potential TIS?

- Cxray 18 mo male otherwise healthy
- Austin patient
T1-T7 congenital w/ rib fusions
Types of Volume Depletion
Deformities of the Thorax

→ Veptr Bait
Clinical - what do we treat?

Age 7
Subjective Pulmonary

PFT's

- FVC %prd
- FEV1 %prd

<table>
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<tr>
<th>Date</th>
<th>Age</th>
<th>FVC</th>
<th>FEV1</th>
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<tbody>
<tr>
<td>4/14</td>
<td>6yr</td>
<td>1.06</td>
<td>121</td>
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<tr>
<td>5/16</td>
<td>8yr</td>
<td>1.38</td>
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</table>

Tf - tech fail

19/12 625 cc
Rx: observation..... in spite of Canavesese/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

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

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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 have 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 (pointillé) 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
Postop Correction / 2D
3 D Visual Correction

Preop
Cvx/Cav = 46.5/144 = 0.32
AVR = 43°
A/P cav = 5  A/P cvx = 3.2

Postop
Cvx/Cav = 63/100 = 0.63
AVR = 25°
A/P cav = 3.1  A/P cvx = 2.6
Early rx must correct or prevent progressive spinal deformity producing windswept thorax.
Age 4 s/p
de-tether
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° (12.9, 82° 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

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<th>Pre-implant</th>
<th>1st Expansion</th>
<th>Last FU</th>
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<tr>
<td>Cobb (degrees)</td>
<td>80</td>
<td>68</td>
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<td>Maximum thoracic kyphosis</td>
<td>57</td>
<td>50</td>
<td>66</td>
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<td>kyphosis (degrees)</td>
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<td>T1-T12 height (mm)</td>
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<td>131</td>
<td>149</td>
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T1-12=14.9 cm ......NOT NEARLY ENOUGH  (Karol et al JBJS ’08)
EOS & Pulmonary Function - Summary

- Lengthen spine (serial procedures) -> objective
  \[ \uparrow \text{FVC or FEV}_{1} \text{ pred. not seen} \]
- Lengthen/expand chest wall -> definitely less compliance, less length, \[ \downarrow \text{‘d FVC/FEV}_{1} \text{ pred.} \]
- 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