Late [Surgical] Intervention for EOS
....but not too late

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ICEOS 2013 Rancho Bernardo
Disclosure : Medtronic, Elsevier
• Worthy opponent wants you to operate EARLY
• Problem….
• No objective evidence to start early - if it looks like a good idea, just do it

If it quacks......
Not Enough Alveoli

- Post-mortem studies
- Intrinsic problem of EOS
- Apparent RX $\rightarrow$ enlarge thorax early ($<\text{age 2?}$)
The inability of the thorax to support normal respiration & lung growth

Melvin Smith, MD
1941-2008
Rationale for Early Expansion

Reid (1971)

10 X increase in # alveoli up to age 5
Alveolar size increases once alveolar number has plateau'd
**PFT Results** *(Campbell, 8/04)*

*Age @ surgery 3.2 y.(6m.-12y.)  - f/u 5.7 y.*  

<table>
<thead>
<tr>
<th>Age @ surg.</th>
<th>Mean VC</th>
<th>% Pred VC</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 2 y.</td>
<td>0.83 L</td>
<td>58</td>
</tr>
<tr>
<td>&gt; 2 y.</td>
<td>0.88 L</td>
<td>44</td>
</tr>
<tr>
<td>&gt; 2 y. w/fusion</td>
<td>0.93 L</td>
<td>36</td>
</tr>
</tbody>
</table>

**Problems**

No preop values available

Improvement or deterioration?
### Motoyama et al 2013 (Paper #3)
Pulmonary and radiographic outcomes of VEPTR
Age 4.8 yr /11 expansions/ 6 yr f/u

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

T1-T12=14.9 cm ......NOT NEARLY ENOUGH (Karol et al JBJS '08)
Reality?

• No histological or clinical evidence that alveolar hyperplasia actually can be affected by surgery (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 fem

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]

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

Thoracic Volume

Birth  6.7% of final volume
age 5  30%
age 10 50%
Growth-friendly Surgery...not so friendly

• Many complications due to repeated surgeries
  - infection, rod fracture, anchor migration
• Spontaneous ankylosis (law of diminishing returns)
• Junctional kyphosis (esp. proximal)
• Poor axial plane correction (windswept deformity of chest)

• Complications ↑ in younger patients ➡ earlier start means earlier abandonment
Many reasons to delay surgery......

- Earlier start -> more lengthenings -> more complications (Bess)
- Law of diminishing returns (Sankar) applies quicker
- ↑'d risk premature finish 2o ankylosis or infection

On the other hand......

- Delay value dependent on tactic which is not detrimental to deformity control, spine + thoracic growth
Direct Comparison - Casts vs GRI 27 pairs
Johnston et al Spine Deform 2013

- Curve magnitude 67°
- GRI achieves better curve correction (33% vs 0)
- T1-S1 length no difference @ f/u
- 6 lengthenings vs 5 casts

- GRI treatment 2 yrs longer in duration
- Complication rate 44% vs 4% casts
- 15/27 casted patients → surgery after 1.7 yr (0-4.8) delay after last cast
Casting Beneficial

- Surgery successfully delayed even though surgical magnitude
- Spine length maintained (same as GRI), at least in short f/u (2 yr)
- No surgical complications to deal with -> no early termination of GRI
- Delaying tactic not detrimental to spine growth
SERIAL CASTING AS DELAYING TACTIC IN CONGENITAL SPINE DEFORMITY

DEMRKIRAN, YACIZI PAPER #14

16 pts, age 35 mos.
Casts x4
No change in scoliosis (58° -> 54°)
Kyphosis improved 85° -> 70°
4/16 progressed -> surgery
Surgery delayed 24 mos (12-41), remainder still under cast rx
JG : EO “Idiopathic”

1+6  70°

Early Surgery?
1+7
1\textsuperscript{st} cast

2+0
2\textsuperscript{nd} cast

2+7
1\textsuperscript{st} brace
Braced age 4+0
Most recent

8+2 T1-12 = 19 cm
• Age 15 mo
• MRI: Negative
• Weight: 5.2 kg (<5<sup>th</sup>%)
5 separate sessions of traction rx over 3 years, interval rx with brace
Growing rod insertion
2/06
(age 5)
status 9/08 - axial plane ?!

50° T1-12 = 15.2

6 planned lengthenings
2 unplanned broken rod revisions
Age 7+6
Jan '11 age 9+10  Broken rod #3
(+ 9 scheduled lengthenings)

Decision for final traction + fusion

Final improvement accepted
Open TRC
Rod removal, final HGT -> ASF/PSF

Best corrections still by HGT
ASF (vats)/PSF with extensive posterior facet ankylosis

T1-12 = 21.0 cm
T1-S1 = 32.3 cm
T4-L1 48°
4 3/4 yr delay
5 yr surgical rx
PFT's : FVC 46%
FEV1 50%

6 mo po
Axial plane = ▼ PFT ?
Early (<4 yr) Surgery - careful

- Law of diminishing returns (ankylosis)
- ↑'d rate complication as # procedures ↑
= the earlier you start, the earlier you will finish distraction-based methods

If you’re so good, why can’t you ever strike twice in the same place?”
How young can/should you start rx

Delay advantage

• Better anchors, esp spine
• Less # procedures over time -> less # complications
• Later occurrence of ankylosis (law of diminishing returns)

Delay disadvantage

• Miss window of best alveolar growth
  Not proven
• May require more complex procedure 2/2 more deformity
  Possible
Rib-based is not spine-based

• Less correction of scoliosis (non-congenital equivalent cases)
• More sagittal plane issues (esp PJK)
• More anchor migration
• Effect of ankylosed chest wall in patients with no rib abnormality to start

• Pick your window of active expansion/lengthening wisely
Growth of Thorax > age 10 - go for hyperplasia + hypertrophy

Brown 2012
Thanks for listening

“Today’s sermon will be followed immediately by a rebuttal from the opposition.”
Growth-friendly Surgery...not so friendly

• Many complications due to repeated surgeries
  - infection, rod fracture, anchor migration
• Spontaneous ankylosis (law of diminishing returns)
• Junctional kyphosis (esp. proximal)
• Poor axial plane correction (windswept deformity of chest)
The Irony of Lengthening

• The more we expand, the greater the lack of soft tissue coverage, prominence of implants, scarring from repeated use of same incisions
Dis-anchoring

Hooks

Cradles

S rod subsidence
Th pedicle screw dis-anchoring

2 yr later, uneventful lengthen + translate x3

Dos 9/06 - uneventful
Spastic paraparesis, urinary retention 5 mo after last uneventful lengthening

3/09 recovery after screw removal, subsequent re-implanted
Early Onset Spine Deformity:

What Do We Know?
How Are We Doing?

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Disclosures : Medtronic Sofamor Danek (research, consultant, royalties)
Mayer/Redding JPO '09

• Veptr in older (mean age 9) pts. increases residual volume with no change in FVC or FEV1 (actual %)
• RV increase in face of unchanged FVC or FEV1 suggests loss of lung compliance - additional volume functionally wasted?
• Caveats - short term, too late for effect in older pts?

Don’t delay too long
**EOS Dilemma - Early Surgery**

**PRO**
- Deformity control
- Permits ↑'d spine growth w/ earlier start
- Improved pulmonary function w/ earlier start *(VEPTRCampbell)*
- Nothing external

**CON**
- More surgeries q4-6 mo. → more complications (58%)
- Complications ↓ w/ delayed start *(Bess et al)*
- Law of diminishing return earlier *(Sankar)*
- ↑'d risk premature finish 2° ankylosis or infection
Halo-Gravity Traction

O. Boachie MD / SRS Ghana