Early Onset Spinal Deformity: Growing Rods or VEPTR – How to decide?

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Disclosures:

• Conflicts of Interest:
  - Helped design VEPTR II
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  - Consultant:
    - Synthes spine
    - Medtronics spine
• Off-Label devices discussed:
  - All pedicle screws in children and growing rods (and staples and tethers) are off-label
  - VEPTR is FDA approved for Thoracic Insufficiency syndrome in growing patients. HDE approval is required

Growing Rods or VEPTR:

• Are they really very different?
  - Both distraction based:
    • Skaggs classification useful:
      • Rib-based distraction
      • Spine-based distraction
      • Combinations
    - Both have same major disadvantage:
      • Repetitive surgical lengthening required
      • Distraction-bases Rx not very good for kyphosis

Growing Rods or VEPTR

• Same treatment goals in early onset deformity:
  - At maturity try to achieve:
    • Maximum
      • Spine length, flexibility
      • Thoracic function (volume, movement)
      • Lung growth
    • Minimum
      • Surgery
      • Complications
      • Hospitalizations, disability

Growing Rods or VEPTR

• The difficult decision is when to intervene surgically?
  - Use evolution of chest deformity as a guide to timing of first surgery?
    - Rationale:
      • Surgical intervention can usually correct/control worsened spine deformity.
      • Surgical intervention less effective for established chest deformity
    - The dilemma:
      • Don’t wait to intervene – the chest deformity may be too severe to reverse
      • Don’t intervene too early – may get spontaneous fusion beneath growing rods after 7 years causing early termination of growth-friendly treatment.

Things we think we know: (maybe)

• Our EOS operations don’t correct severe or established 3-D chest deformity
  - Goal is therefore not to let severe thoracic deformity occur or progress
  - When is this? How much is too much?
  - Thorax shape more relevant than the Cobb angle
GR or VEPTR for Early onset deformity

- **Factors to consider**, ways to decide
  - **Etiology** of deformity
    - *Spine or chest* dominant or primary
  - Associated:
    - Bone quality
    - Kyphosis
    - Rigidity of deformity
    - Soft tissue coverage
    - Complications of treatment

Growing Rods or VEPTR

- **Etiology of deformity**
  - Is the *spine the primary deforming force*?
  - Is the *chest wall the primary deforming force*?
  - Are the *chest and spine* both etiologic factors?

Early Onset Deformity. – etiology as a factor:

- **Chest wall is primary problem**
  - Massive rib fusions
  - Other purely thoracogenic scoliosis
    - Chest wall tumors
    - Esophageal atresia,
    - Multiple thoracotomies
    - Some congenital diaphragmatic hernias
  - **Expansion thoracostomy, VEPTR best choice**
  - ‘Spine-only’ treatment will be defeated by chest wall tether
- Thoracogenic scoliosis may need skin expanders, staged procedures, flaps,

GR or VEPTR - VEPTR and **expansion thoracostomy** for rib fusions and congenital scoliosis

- **GR or VEPTR?**
  - Thoracogenic Scoliosis after chest wall tumor

Spine and Chest are both etiologies

- Spine and chest wall *both etiologic factors*
  - Rib fusions with congenital scoliosis
    - **Expansion thoracostomy with VEPTR** or expansion thoracostomy with GR best choices
  - **Established, severe chest wall deformity plus severe spine deformity**
    - Severe windswept thorax
      - GR approaches primary spine problem
      - Is VEPTR any better?
GR or VEPTR Spine and chest are both etiologies

- 2 y.o with progressive curve, increasing nighttime O2 requirement
- Multiple unilateral rib fusions
- Vertebral bars
- Note worsening ‘normal’ hemithorax

GR or VEPTR Spine and chest are both etiologies

- Early intervention for the sake of chest shape
  - Three thoracostomies
  - VEPTR
  - ‘normal’ side of chest improving slowly?

Age 11, PFT’s 45%

- Partial control of spine deformity, chest deformity

GR or VEPTR Spine and chest are both etiologies

Example: progression after in situ fusion

- Fusion at Age 6 months
- Curve Progression at Age 16 months

GR or VEPTR Spine and chest are both etiologies

Age 2 – s/p in situ fusion age 6 mos

- Curve Progression at Age 2 years

GR or VEPTR Spine and chest are both etiologies

- Two rib osteotomies
- One rib-to-rib device
- One rib-to-spine device

Curve Progression at Age 16 months

- VEPTR, expansion thoracostomy, prior fusion

- Age 2 years
Spine growth – 4 yrs after first VEPTR

- Continued growth - 6 lengthenings
- One device exchanged for growth

Age 13 – two years post menarchal - 11 years after first VEPTR

- PFT’s 55%
- Devices left in place.
- Thorax far from normal
- No fusion needed?

GR or VEPTR – Chest and Spine both a problem

- 3yo with progressive thoracic deformity
  - Congenital rib fusions
  - Multiple vertebral anomalies.
- VEPTR and expansion thoracostomies age 3

GR or VEPTR – Chest and Spine both a problem

- Multiple lengthenings
- Exchange x 2
- Age 14, mature
- Unhappy with waist asymmetry
- Active as cheerleader
- VEPTR removal, osteotomies, final fusion

GR or VEPTR – Chest and Spine both a problem

- Final spine fusion easier with VEPTR?
- Rib fusions, scarring expected
- Spontaneous spine fusions:
  - Below rod at lower end
  - In normally segmented part of curve
- Far from easy after VEPTR

GR or VEPTR – Spine is the problem

- Infantile idiopathic scoliosis (normal segmentation)
  - Moderately severe deformity
    - (Spine still worse than chest deformity)
  - Dual Growing rods
  - Rationale for choice:
    - Chest wall, although deformed, is mobile.
    - Expansion thoracostomy, VEPTR, may stiffen chest wall?
Infantile idiopathic scoliosis, moderately severe rotational chest deformity – Age 5

Infantile Idiopathic Scoliosis, dual growing rods – earlier would have been preferable? – Age 5

Age 12, after 7 years of lengthening, stopped by spontaneous fusion. Osteotomies, definitive fusion

GR or VEPTR? Infantile idiopathic Age 3, age 4 in brace, age 5 after casting

GR or VEPTR? More casting, bracing – Age 7, Age 9

GR or VEPTR – IIScasted until age 9
Preop Windswept chest. Note severe thoracic lordosis
GR or VEPTR? - Infantile idiopathic, Age 9

Windswept Chest

IIS - VEPTR first stage

VEPTR fixation lost. GR inserted. Spine under control, chest irrevocably altered.

After 3 years lengthening, Age 12. FEV1 65% Chest still windswept

GR or VEPTR? - Infantile idiopathic, Age 9

Growing Rods or VEPTR
Neuromuscular deformity – VEPTR or GR?

- High-tone (‘CP’)
  - Neither very good. Rarely indicated in ‘CP’ high tone
  - Personal opinion – more trouble with rib drifting in ‘CP’

Growing Rods or VEPTR
Neuromuscular deformity – VEPTR or GR?

- Low-tone SMA, arthrogryposis, myopathy
  - VEPTR directly treats the ‘parasol’ deformity
  - Multiple fixation points – tandem connectors
  - Xrays better with VEPTR, but are the patients any better?
  - Minimal chest wall movement – breathing is all diaphragmatic, hence GR may be as good as VEPTR!

GR or VEPTR?
5 yo with SMA trach but not vent dependent
GR or VEPTR?
SMA ages 5, 10, 12

GR or VEPTR – usefulness of tandem VEPTR II anchors to horizontalize parasol deformity?

GR or VEPTR - *Myelodysplasia* with collapsing kyphosis or scoliosis

- Age 5, thoracic level.
- Worsening deformity, impending skin breakdown

GR or VEPTR - *Myelodysplasia* with collapsing kyphosis or scoliosis

- VEPTR II rib to pelvis.
  - Better able to maintain distraction, deformity control without involving dysplastic lower spine
  - Control of spine without instrumentation of spine
  - Tandem rib connectors useful.
    - Distribution of force
    - Resistant to pull out

GR or VEPTR in Infection

- Infection – VEPTR or GR?
  - Either is a good rescue for infection in the other!
  - Soft tissues a critical component

- Implant removal/retention with deep infection?
  - Depends upon
    - Extent
    - Duration after index procedure
    - Skin, soft tissues

GR or VEPTR in Infection
Fetal alcohol syndrome Complex congenital vertebral anatomy - age 6
GR or VEPTR in Infection
Fetal alcohol syndrome – long segment of spine

GR or VEPTR in Infection
Fetal alcohol syndrome

GR or VEPTR? – Upper thoracic kyphosis

- Kyphosis (upper thoracic) problematic for both growing rods and VEPTR
  - Multiple factors:
    • Weak paraspinals
    • Junctional stresses above stiff segment
    • GR - Disruption of posterior elements.
    • VEPTR – lack of direct sagittal spine control
    • Both GR and VEPTR – distraction-based

GR or VEPTR? – collapsing deformity, arthrogryposis age 4. Coronal deformity controlled

GR or VEPTR? arthrogryposis – age 4 to 8 – initial control then progressive PJK

VEPTR in arthrogryposis – poor control proximal kyphosis.

- Cervico thoracic junction collapsed further into kyphosis, rotating around VEPTR attachments
- Distraction based systems poor for upper thoracic kyphosis
- GR better than VEPTR?
  - Can extend more proximally with GR
GR or VEPTR for upper thoracic kyphosis? 7 yo with familial dysautonomia

- GR or VEPTR for upper thoracic kyphosis
  - 7 yo with familial dysautonomia age 7, 8, 16

GR or VEPTR for upper thoracic kyphosis
- neither is perfect
- GR has some advantages

- Strategies:
  - Leave some kyphosis
  - Pre-op halo gravity traction may facilitate device insertion by diminishing kyphosis
    - Tendency to recur
  - Growing rods can extend more cephalad than VEPTR – ? Past the kyphosis?

- GR preferable:
  - More cephalad extent possible
  - More contouring options
  - Direct control of spine

GR or VEPTR – Bone dysplasias

- Bone dysplasias
  - Spine or ribs better bone for anchor points?
    - Small soft vertebra? – VEPTR may be preferable
  - Spinal stenosis or hypoplastic pedicles? Prior laminectomy? VEPTR may have advantage
  - Beware waiting too long to establish control over curves in spinal stenosis – neurologic risk with progression, correction.

Spondyloepiphyseal dysplasia

- Age 14 mos.
  - C1-C2 fusion
  - Growing rod for scoliosis, kyphosis
- Age 30 months – proximal disengagement
- Age 10 – paraplegia following hip osteotomies, epidural post-op. Slow resolution

SED
Pre-traction

Halo gravity for 7 weeks

Post VEPTR

- Kyphosis well controlled
- One rib sleeve bent to accommodate kyphosis

Early Onset Deformity. – etiology as a factor:

- Bone dysplasias:
  - If upper thoracic kyphosis, GR has an advantage over VEPTR
  - GR can be extended as far cephalad as needed

Campomelic Dysplasia

- Vent dependent
- Rapid progression after 6 months
- Age 18 months – minimal deformity

Age 30 months – severe kyphosis
Age 30 months – severe kyphosis

- Age 30 months – severe kyphosis – stiff
- GR or VEPTR?

- GR enables more cephalad purchase

Age 34 months – growing rods, extending to C7 with sublaminar cable at C7

Age 7 symptomatic spondylolisthesis required extension to pelvis

GR or VEPTR – Osteopenia in Early Onset Deformity

- Osteopenia, poor bone quality
  - VEPTR may have an advantage?
  - ‘more ‘give’ in ribs than spine?
  - Experience with osteogenesis imperfecta?
  - GR allows staged anchor placement

VEPTR or GR? Osteopenia and kyphosis

- 5 yo with recurrent TEF
- Failed VEPTR with severe osteopenia
- Progressive scoliosis
- Rigid upper thoracic kyphosis

VEPTR or GR? Osteopenia and kyphosis:
Insert anchors, 2mos, apply H-G tx, 1 1/2 mos
VEPTR or GR? Osteopenia and kyphosis Age 6, 8

- Worsening deformity
- Recurrent breakdown when attempting to sit or with a brace
- Severe osteopenia, recurrent fractures
- Increasing respiratory distress (secondary TIS)

4 y.o. with Ehlers Danlos variant

GR or VEPTR - complications

- GR – growth stopping complication
  - Lengthening may not be possible indefinitely
- Skaggs and GSSG data:

GR or VEPTR - complications

- Recurrent rib fusion after thoracostomy

GR or VEPTR - complications

- Inadvertent fusion of scapula to ribs:
  - 2 patients with solid fusion of scapula to ribs
  - Numerous with limited scapulothoracic function

GR or VEPTR - Final fusion easier?

- GR and VEPTR:
  - Spontaneous stabilization may have occurred by end of growth
  - No fusion may be needed if:
    - Modest deformity
    - Implants not troublesome
    - Likely underlying fusion
- Conversion to final instrumented fusion?
  - GR challenging conversion
  - Scar, spontaneous fusions, distorted anatomy
  - VEPTR easier conversion?
    - Spine “untouched”?
      - Spine spontaneously fused, esp lower spine anchor area
      - Rib fusions beneath device
# VEPTR or Growing Rods?

How to choose between GR and VEPTR:

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<tr>
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<th>Growing Rods</th>
<th>VEPTR</th>
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<tbody>
<tr>
<td>Spine growth preservation</td>
<td>+</td>
<td>+</td>
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<tr>
<td>Chest deformity correction</td>
<td>+/-</td>
<td>++</td>
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<tr>
<td>Ease of use, familiarity</td>
<td>±</td>
<td>±</td>
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<tr>
<td>Multiple ops</td>
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<tr>
<td>Final fusion needed</td>
<td>+/-</td>
<td>+/-</td>
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<tr>
<td>Complication which limits distraction</td>
<td>Fusion underneath rod</td>
<td>Chest wall stiffness, rib re-fusions</td>
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<tr>
<td>Solution to complication?</td>
<td>Early fusion</td>
<td>Repeat thoracostomy</td>
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<td>Common device problems, failures</td>
<td>Rods break</td>
<td>Rib attachments drift</td>
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<tr>
<td>Upper thoracic kyphosis</td>
<td>Better</td>
<td>Poor</td>
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<tr>
<td>Osteopenia</td>
<td>Poor</td>
<td>Poor</td>
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## Strong indications for VEPTR:

- Primary Chest wall problem
  - Massive rib fusions
  - Thoracogenic scoliosis
- Failed Growing rods
  - Infected spine anchors
- Poor spine anchors
  - Bone dysplasia with spinal stenosis
  - Spina bifida

## Strong indications for growing rods

- Primary spine deformity with lesser, flexible chest deformity
- Normally segmented, unscarred chest wall
- High thoracic kyphosis
  - (GR/local fusion can extend into the cervical spine if needed)