EOS: The Biggest Challenge in Spine!

...and finding solutions

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Johns Hopkins Medical Institutions
Baltimore MD
Disclosures

- JBJS
  - Pediatrics Editor
- Pediatric Spine Foundation-BOD
- Thieme
- DePuy Spine
- Globus
- OrthoPediatrics
Kudos to...

• Program Committee

• Local Host
Behrooz A. Akbarnia

- Driving force for the field
- Founder of this meeting
- Icon of a leader
  - professionally
  - personally
Decisions, Decisions…Choose Wisely

Peter O. Newton MD
Rady Children’s Hospital & University of California, San Diego

- Often no perfect solution
- Buy time, even if it’s hard
- Choose wisely with an eye on the final solution

EOS = Not a license for early surgery
Outline

• 1. Why EOS is the biggest challenge!
• 2. Ideas which didn’t work well
• 3. Game-Changers
• 4. Role of SRS & Societies in helping
• 5. Relationship with patient/family
• 6. Ideas for the Future
1. Why EOS is the Biggest Challenge

EOS vs Adult

- Challenges of Spine Formation
- Central organizing structure against space and gravity
  - Congenital
  - Neuromuscular
  - Connective tissue
EOS vs Adult Spine

- Smaller market
  - Less funding, interest
- Higher risk
- Different physiology
- 70+ years’ Life demands
- Prognostic & proactive vs reactive
  - Need certainty when advising risky procedure
  - “She’s doing fine now”
Pediatrics is our “roots”…

• Bracing, Ponseti vs Mehta casting

• Growth guidance

• Lengthen & Stabilize
Pediatrics is our “Roots”...

- Function versus appearance
- Respect for self-image as part of health

“You’re not pigeon-toed
—perhaps you should see someone about it.”

Mercer Rang MD
Definition of Early Onset Scoliosis

- <3yrs- James
- <5yrs- Ponseti, Dickson
- <10yrs- Akbarnia, GSSG, CSSG
Natural History

- GC Lloyd-Roberts
- 100 babies at Great Ormond St
- 92 resolved
  - Not always bad
- Molding of head, ribs and pelvis
“Those managed by Risser jacket and brace until >10y before surgery have fair … respiratory function and cosmesis

Those who underwent spinal fusion before age 10 years all had major recurrence of the deformity and moderate-to-severe respiratory compromise.

Early surgery, even with anterior growth arrest, did not prevent deformity or arrest the decrease in pulmonary function

Theories of best treatment should be reevaluated “.
“Holy Grails” of EOS

- *Growing* while correcting the deformed spine
- Optimizing flexibility
- Knowing when to start and stop
- Minimizing Surgeries
- Preventing Iatrogenic Deformities
How do we get there?

- Individual insights
  - Campbell, McCarthy
- Study Group Efforts
  - PSSG
- Society involvement
  - SRS & others
2. Ideas which Fell Short
Old Maxim

- “A short straight spine is better than a short crooked spine”
  – R. Winter 1986
But far from ideal!

- Sometimes achieve neither!
2. Ideas which fell short

- Early PSF in situ
- “Thick Fusion Mass” often led to
  - Lordosis
  - Crankshaft
  - Shortening
2. Ideas which fell short

- Moe = Temporary Single H-rods
  - Poor anchorage
  - No means of storing length
2. Ideas Which Fell Short

- Temporary Instrumentation

Graduation Protocol After Growing-Rod Treatment: Removal of Implants without New Instrumentation Is Not a Realistic Approach

Ismail Aykut Kocygir, MD, Z. Deniz Olgun, MD, H. Gokhan Demirkiran, MD, Mehmet Ayvaz, MD, and Muharrem Yazici, MD

Investigation performed at the Department of Orthopaedics and Traumatology, Hacettepe University Hospitals, Ankara, Turkey
2. Ideas which fell short

- Rib based distraction for Early-Onset Idiopathic Scoliosis
2. Ideas which fell short

• Anterior fusion and posterior Growing Rods
  – Deformity continues
3. Game-Changers

- **Concepts:**
  - Spinal Growth
  - T.I.S.
  - Diminishing Returns
  - C-EOS

- **Techniques:**
  - Mehta Casting
  - Growing implants
  - Growth guidance (Trolley, Shilla, tether)
3. Game-Changing Concepts: Assessment of Growth & Maturity

- Growth occurs in two peaks: 0-2 and PHV
- Significance of peak height velocity (PHV)
- What are markers of PHV?
Estimate of Peak Growth Velocity

- Premenarche
  - $\leq 11$ years of age
- Risser 0

**Better:**
- Open triradiate cartilages
- Sanders DMS stage 3 (capped not fused)
- Sauvegrain – rectangular olecranon
- Blood test?
Game-Changing Concept: Thoracic Insufficiency Syndrome (TIS)

The Inability of the Thorax to Support Normal Respiration or Lung Growth

Thoracic Insufficiency Syndrome (TIS)

- Occurs with congenital, infantile or neuromuscular scoliosis
- Congenital anomalies of the ribs
- Acquired chest wall deformities
- Some skeletal dysplasias
- Need to incorporate dynamics
Game Changing Concept:
“Law of Diminishing Returns”

Spine stiffens with time
Rigid Immobilization?
Surgical exposure?
Surgical stretch?
Implication; surgery to be used sparingly & timely

Sankar and Skaggs
**Concept Game Changer: C-EOS**

- Etiologies are most studied
- Implication: use this to determine strategy

<table>
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<th>Etiology</th>
<th>Cobb Angle (Major Curve)</th>
<th>Maximum Total Kyphosis</th>
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Treatment Game-Changer
Mehta Casting

• Like Ponseti, it took a while for us to catch on
• Enthusiastic disciples
  – Sanders
  – D’Astous

Straight at 6 yrs old
Treatment Game Changers: Dual (Growing) Rods

- Improved mechanics
- Stable proximal & distal “foundations”
  - Hooks or screws cranially
  - Screws distally
- Dual Rods
- Lengthen every 6-12 months
  - “drive the spine”
- ?Final fusion at maturity?
3. Treatment Game Changers - Luque Trolley

- Great idea in concept
- Unpredictable in idiopathic EOS
- Best results in NM
- Modifications:
  - Ouellet
  - Miladi
Treatment Game-changer: Shilla

- “Self-guided” spinal growth
- Fuse/control apex
- Rods grow off screws
- 5 year follow up
  - Half as many procedures
    - less length gain
4. The Role of Organizations

- GSSG
- CSSG
- PSSG
- Supported by members + Industry
4. The Role of SRS and Societies in Advancing EOS care

- SRS Mission: To Foster the Optimal Care of All patients with Spine Deformities
SRS

• Relevant Efforts/Committees for EOS:
  – Growing Spine- define datasets
  – Education- EOS content for the masses
  – Health Policy- liaison with AAP
  – Pediatric Device TF
  – Patient Education
  – Non-operative Management- Evidence on brace/cast
  – Research Grants $; evaluate & elevate the science
  – QSVI- Make surgery safer- checklists & processes
Imagine a World Where...

- Pedicle screws not approved for kids
- No growth connectors
- Sliding screws not approved
- Need to go to Europe or Asia for MCGR
Devices Approved for Pediatric Applications During Tenure of Task Force

- Reclassification of pedicle screws
- Reclassification of cervical screws
- Reclassification of Non-fusion, “growth friendly” posterior-based spinal systems
  - Growth Connectors
  - MCGR
  - Sliding screws
  - VBT
SRS / OREF / POSNA Funding

- Announced 11/18
  - John Callaghan
  - Industry

OREF/SRS/POSNA Transform Practice – Spinal Growth Grant

Funding:
$240,000 grant ($80,000-$120,000 per year)

Grant Description / Eligibility:
- OREF/SRS/POSNA solicit investigator-initiated research proposals focused on the development of new research knowledge that will change treatment for growing spine conditions within the next five (5) years. This funding opportunity should be uniquely important to early onset/growing spine research and not readily fundable through another mechanism (NIH, PCORI, AHRQ).
- Areas of research focus may include: translational or clinical research to preferably include at least pilot human studies. Research should answer important translational or clinical questions to address unmet patient needs. Specific topics may include normative spinal growth, guided growth conditions, and other areas as determined by the OREF grant committee.
SRS – EOS Learning module

• Chosen as our first module in Learning Management System
  – $150K investment
  – High production value

– Muharrem Yazici, Laurel Blakemore, Larry Haber
– HVR
– GR
– Mehta Cast
O.R. Setup and Patient Positioning

In the treatment of Early Onset Scoliosis (EOS)

Lawrence Haber, MD

This module presents the process endorsed by the Scoliosis Research Society for setting up your operating room and positioning a pediatric patient to implant growing rods - including the selection and use of appropriate protective devices.
5. Relationship with the Family

Mercer Rang, MD
Treatment of Early Onset Scoliosis (EOS): What Families Need to Know

Table of Contents

Section I. Facts to Know
   A. What is early onset scoliosis (EOS)?
   B. Causes
      1. Congenital
      2. Neuromuscular
      3. Syndromic
      4. Idiopathic
   C. Why is EOS a problem for my child?

Section II. Treatment Options
   A. Background
   B. Information about treatment options
      1. Observation
      2. Bracing
      3. Casting
      4. Surgery
         a. distraction-based procedures
         b. growth guidance systems
         c. growth modulation procedures
         d. spinal fusions
   C. Summary of treatment indications

Section III. Benefits and Risks
   A. Summary of treatment benefits
   B. Summary of treatment risks and complications

Section IV. Risks of General Anesthesia in Children

Section V. Getting Ready To Choose
   A. Things to consider when choosing
   B. Feeling ready to Choose

Section VI. Glossary

Section VII. References

Choosing a treatment option for your child can be hard to do. This booklet gives you information and answers to questions you may have about this decision. You and the doctor will talk about how to make a good decision based on what is best for your child and you.

Section I: Key Facts to Know

In this section you will learn about what early onset scoliosis (EOS) is, what problems it causes, and what it may mean for both you and your child if it gets worse.

A. What is EOS?
   EOS is an abnormal curve of the spine in children younger than 10 years of age.

B. What caused my child to have EOS?
   There are many causes for EOS. They are divided into four categories. Treatment choices differ depending on which category your child’s scoliosis is in.
6. Ideas for the Future

- Improved education outside Spine world
- Understand science of disordered physiology
- Genetics of EOS
- More understanding of natural history
- Translational science
How Do We Make Progress?

- Unusual Vision
- Charges from mentors
- Inspiration from peers at meetings
- Trial and error
- Fortuitous
Increased Funding for Research

- NIH
- CIHR / IRSC
- Health/Quality Research Agencies

- We should advocate for it
  - Serve on study sections
- Patients should demand it
- We should elevate the science
Improve Education Outside Spine
World

- Anesthesiology
- ID
- Pulmonary
- GI, nutrition
- IONM
Improve Education Outside Spine World

**Discipline**
- Anesthesiology

**What we can teach**
- Antibiotics
- Blood management
  - Smaller = riskier
- Cord perfusion
- Relevant neurology
- Working with IONM
## Improve Education Outside Spine World

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<td>• Is it osteo?</td>
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<td>• We can usually save the implants!</td>
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### Improve Education Outside Spine World

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## Improve Education Outside Spine World

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Understand Science of Disordered Axial Physiology

- Airway
- Viscera
- Ribs/sternum

The Reciprocal Relationship Between Thoracic and Spinal Deformity and Its Effect on Pulmonary Function in a Rabbit Model

A Pilot Study

Homel P., Melto, MS.* Brian D. Snyder, MD, PhD.* Natasha N. Callander, BS.* Carissa L. Bellardine, MS, T. and Andrew C. Jackson, PhD.*
Translational Therapies

- Losartan for Marfan: helps the aorta; not the spine
- BMT & ERT for MPS: Helps the CNS; not the spine
- Nusinersen for SMA: Helps strength, not the spine
- Vasoritide for Achondroplasia?
- Need to start somewhere
Genetics of EOS

- Monique Garcia
- OS Biobank
The EOS Biobank

Aims
1. Collect blood samples and fibroblasts from EOS patients
2. Link samples to matched clinical data from the PSSG
3. To establish a multi-centre, prospective, longitudinal study for EOS

Expected Outcomes
- Accelerate research advancements for EOS
- Translate discoveries into diagnostic tools and therapies
Better Understanding of Natural History

– Which EOS-I will worsen
  • Moving beyond RVAD

– Which congenital curves will worsen
  • And worsen importantly?
  • Can we tell from advanced imaging in infancy?
Better Understanding of Natural History

– Which neuromuscular curves benefit from PSF

Same Current CP CHILD score

2013

2019
Summary

• Many factors have gotten us here
• Group wisdom & Societal resources will continue to guide progress
• To solve the biggest problem in Spine!
Thank You & Happy Thanksgiving!
2. Ideas which fell short

- Temporary Single rods
The Early Onset Scoliosis Biobank
Spine Deformity

- Difficult, risky to control
- Magnified 10X if occurs early!
Deformity
- Definition

- Deformity due to primary vertebral malformation
- Rule out infantile idiopathic scoliosis and neurogenic scoliosis
Previous Treatments

- Bracing
- Moe Rods
- Luque trolley
Infantile - Rx

- Mehta Cast, then brace for progressive type < 4 y.o.
Steps taken to reach Storyboarding...

- Best Practice
- Problems
- Learning Tasks
- Learning Objectives

Ability to perform all Learning Tasks means you have achieved Competence.

Learning Objectives should cover all Learning Tasks.

Learning Task should overcome all Problems.

Problems risk achieving Best Practice.

Best Practice is what every surgeon should be targeting.

Storyboard should cover all Learning Objectives.

Ability to perform all Learning Tasks means you have achieved Competence.

Learning Objectives should cover all Learning Tasks.

Learning Task should overcome all Problems.

Problems risk achieving Best Practice.

Best Practice is what every surgeon should be targeting.
• Grow the spine for health
  – Min Mehta
  – John Moe
  – Behrooz Akbarnia
  – George Thompson
Treatment Game Changers
Dual Growing Rod

- Subcu rod
  - endfusion (Moe 1984)
  - Distract if < 10° degrees correction
  - 3.8 cm mean
  - Added MMA and end fusion
Physical Examination

- **HT:** 79.2 cm  
- **WT:** 10.2 kg  
- **BMI:** N/A  
- **General:** Healthy, happy appearing toddler with obvious spine deformity.  
- **Spine:** Significant kyphosis and scoliosis. Kyphosis is major part of deformity. Midline incision and resection of right chest wall. Stretching prone recovers most of scoliosis and some of the kyphosis. Collapsed position when sitting.  
- **Neurologic Exam:** Lower extremities appear normal, neurologic appears normal.
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POSNA/SRS Pediatric Device Task Force Committee

- Brian D Snyder, MD, PhD. Co-Chair
- Michael Vitale, MD Co-Chair
- Behrooz A Akbarnia, MD
- B Stephens Richards, III, MD
- Richard E McCarthy, MD
- Robert M Campbell, Jr, MD
- David W Polly, Jr, MD, SRS Representative
- Laurel C Blakemore, MD
- Paul D Sponseller, MD
- Nigel J Price, MD
- William Hennrikus, MD, AAP Representative
- John (Jack) Flynn, MD
- Peter O Newton, MD
Outline

• Why EOS is the biggest challenge!
• Ideas which didn’t work well
  – Temporary instrumentation
  – Early PSF
  – Apical fusion (Roaf)

• Game-Changers
  – Crankshaft phenomenon / ASF
  – Diminishing Returns
  – Growth guidance (Trolley, Shilla, tether)
  – T.I.S.
  – Mehta casting
  – C-EOS

• Role of SRS & Societies
  – Help bring resources (research grant, priority setting, PDTF)
  – EOS LMS content
  – Put in context
  – Global
  – Learn from adult ideas
  – Role in teaching other specialists
  – Advocate for appropriate healthcare funding for our patients

• The larger relationship with the patient (and the disease)
  – Patient organizations

• Ideas for the Future
  – Refine ability to predict (when resolving, when congen scoli will worsen; when height is adequate; when CP is too severe)
  – Role of spine in health
  – Rib and sternum
  – Maximizing mobility
  – NIH funding
  – Improved growth guidance
  – Avoid stiffening of spine and rib cage
Charges

• Evaluate current state of medical device development for pediatric use to treat musculoskeletal diseases
• Identify stages of device development for clinical use in children and relevant regulatory pathways
• Describe specific barriers and propose solutions to facilitate development and approval for pediatric devices
Versus

- Challenges of wear, trauma and degeneration (gravity and homeostasis)
  - Variations in “normal” bone health
  - Variations in disc aging and composition
  - React to existing concerns
Fig. 1 Simplified skeletal age assessment with the olecranon method during the accelerating pubertal growth phase of peak height velocity and Risser grade 0 from the ages of eleven to thirteen years in girls and from thirteen to fifteen years in boys, with a decelerating growth phase after elbow fusion.