EOS: The Biggest Challenge in Spine !

…and finding solutions

Paul D Sponseller MD
Johns Hopkins Medical Institutions
Baltimore MD

Disclosures

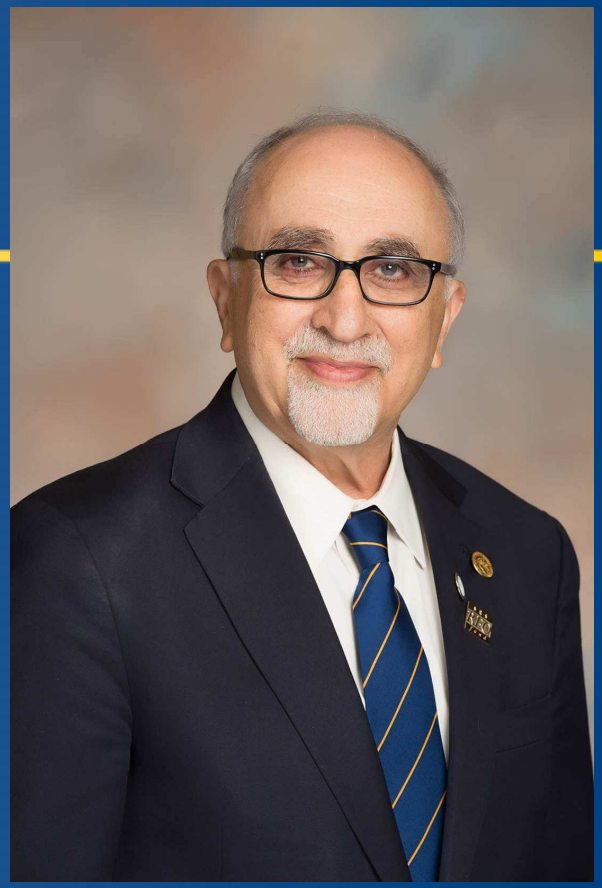
- JBJS
  - Pediatrics Editor
- Pediatric Spine Foundation-BOD
- Thieme
- DePuy Spine
- Globus
- OrthoPediatrics

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**

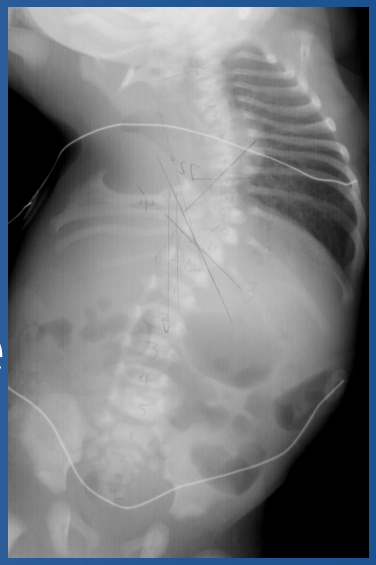
EOS = Not a license for early surgery

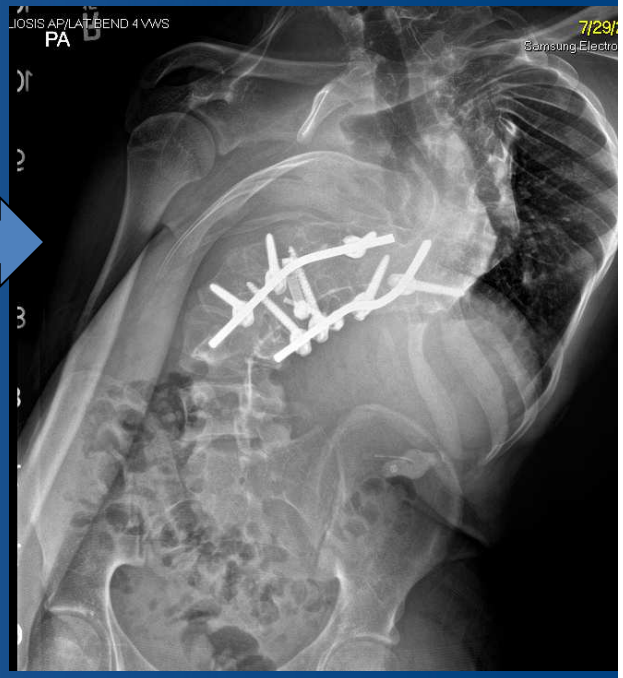
Outline

- 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

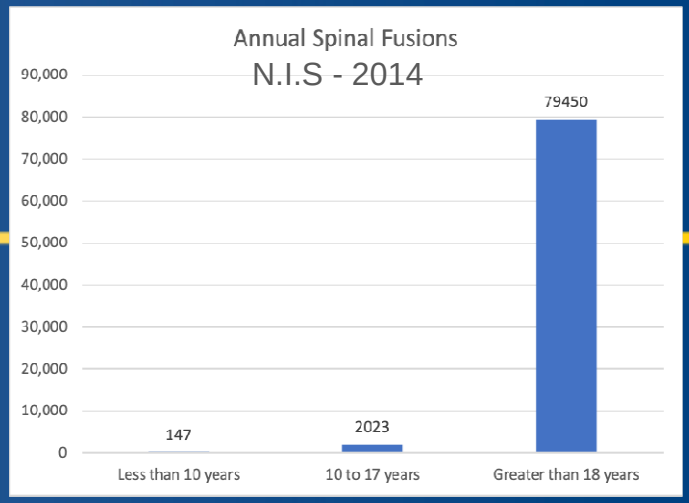
1. Why EOS is the Biggest Challenge
EOS vs Adult

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





EOS vs Adult Spine

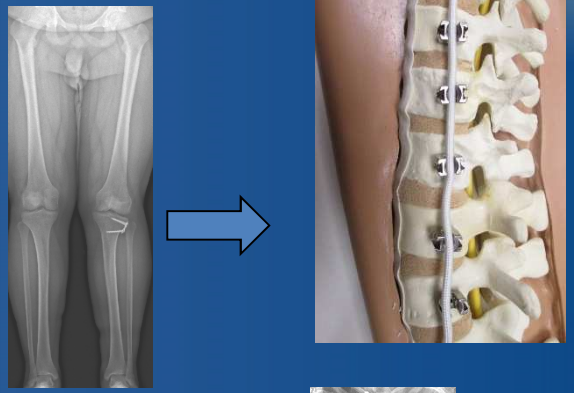


- Smaller market
  - Less funding, interest
- Higher risk
- Different physiology
- 70+ years' Life demands
- Prognostic & proactive vs reactive
  - Need certainty when advising risky procedure
  - "She's doing fine now"

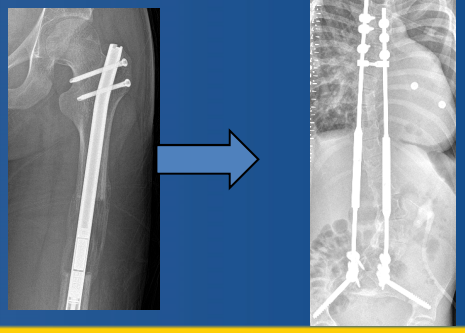
Pediatrics is our “roots”…

- Bracing, Ponseti vs Mehta casting

- Growth guidance

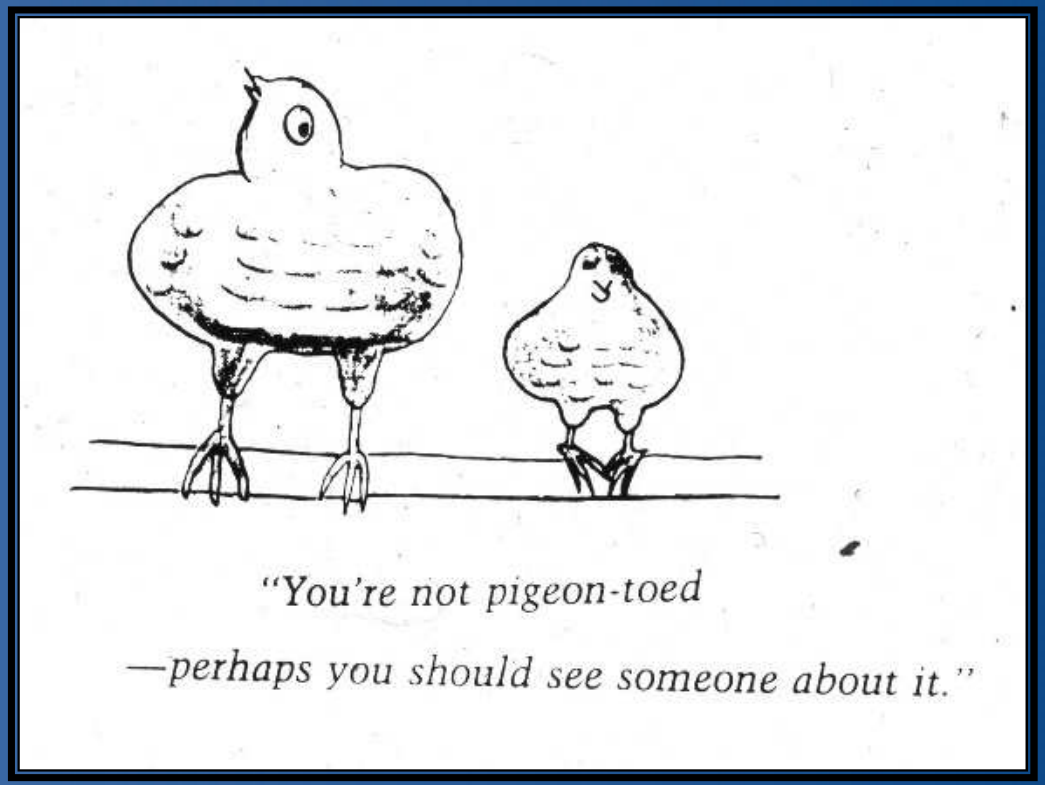


- Lengthen & Stabilize



Pediatrics is our “Roots”…

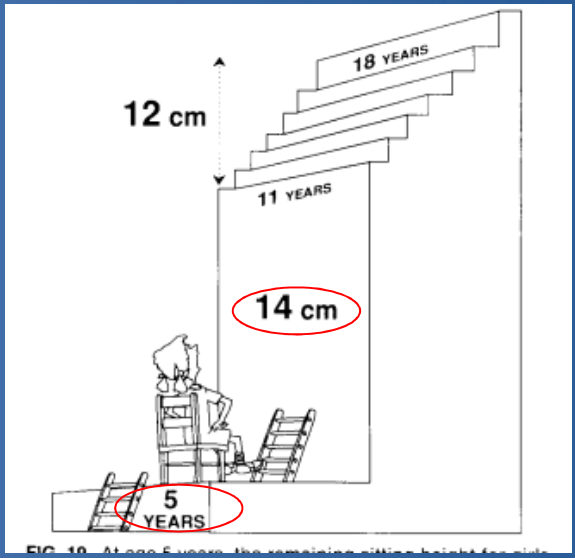
- Function versus appearance
- Respect for self-image as part of health



Mercer Rang MD

Definition of Early Onset Scoliosis

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



ELSEVIER
Spine Deformity
Volume 3, Issue 2, March 2015, Pages 105-106

Letter to the Editor

Early Onset Scoliosis - Time for Consensus

Ron El-Hawary MD, MSc, FRCS(C), Behrooz A. Akbarnia MD

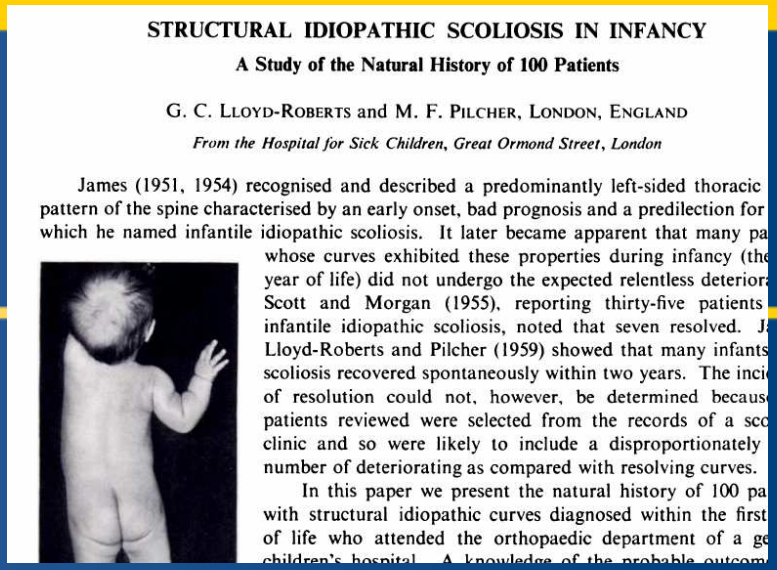
ELSEVIER
Spine Deformity
Volume 3, Issue 2, March 2015, Page 107

Consensus Statement

Early Onset Scoliosis Consensus Statement, SRS Growing Spine Committee, 2015

David L. Skaggs MD, MMM, Tenner Guillaume MD, Ron El-Hawary MD, John Emans MD, Michael Mendelow M
John Smith MD, SRS Growing Spine Committee

Natural History



STRUCTURAL IDIOPATHIC SCOLIOSIS IN INFANCY

A Study of the Natural History of 100 Patients

G. C. LLOYD-ROBERTS and M. F. PILCHER, LONDON, ENGLAND

From the Hospital for Sick Children, Great Ormond Street, London

James (1951, 1954) recognised and described a predominantly left-sided thoracic pattern of the spine characterised by an early onset, bad prognosis and a predilection for which he named infantile idiopathic scoliosis. It later became apparent that many pa whose curves exhibited these properties during infancy (the year of life) did not undergo the expected relentless deterior Scott and Morgan (1955), reporting thirty-five patients infantile idiopathic scoliosis, noted that seven resolved. J Lloyd-Roberts and Pilcher (1959) showed that many infants scoliosis recovered spontaneously within two years. The inci of resolution could not, however, be determined becaus patients reviewed were selected from the records of a sc clinic and so were likely to include a disproportionately number of deteriorating as compared with resolving curves.

In this paper we present the natural history of 100 pa with structural idiopathic curves diagnosed within the first of life who attended the orthopaedic department of a g children's hospital. A knowledge of the probable outcom

- GC Lloyd-Roberts
- 100 babies at Great Ormond St
- 92 resolved
  - Not always bad
- Molding of head, ribs and pelvis

SPINE Volume 28, Number 20, pp 2397–2406

Respiratory Function and Cosmesis at Maturity in Infantile-onset Scoliosis

C. J. Goldberg, M.D.,* I. Gillic,† O. Connaughton,† DP Moore, F.R.C.S.I., M.Ch.Orth.,‡

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Natural History of IIS (C. Goldberg)

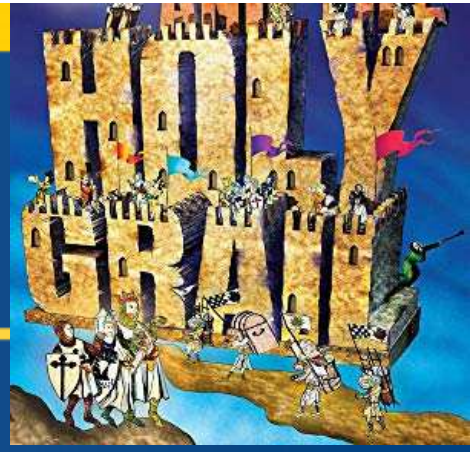
"Those managed by Risser jacket and brace until >10y before surgery have fair …respiratory function and cosmesis

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 ".

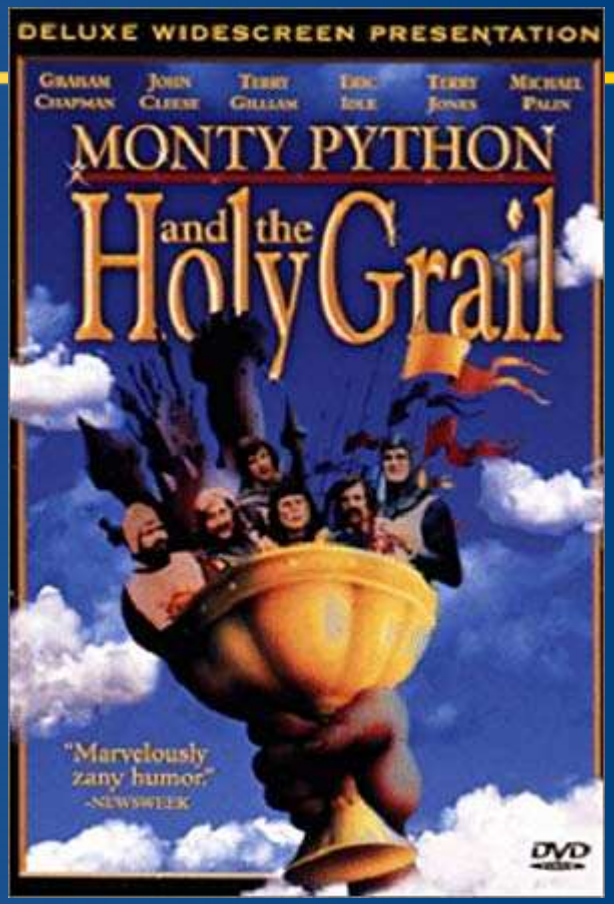
“Holy Grails” of EOS



- *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

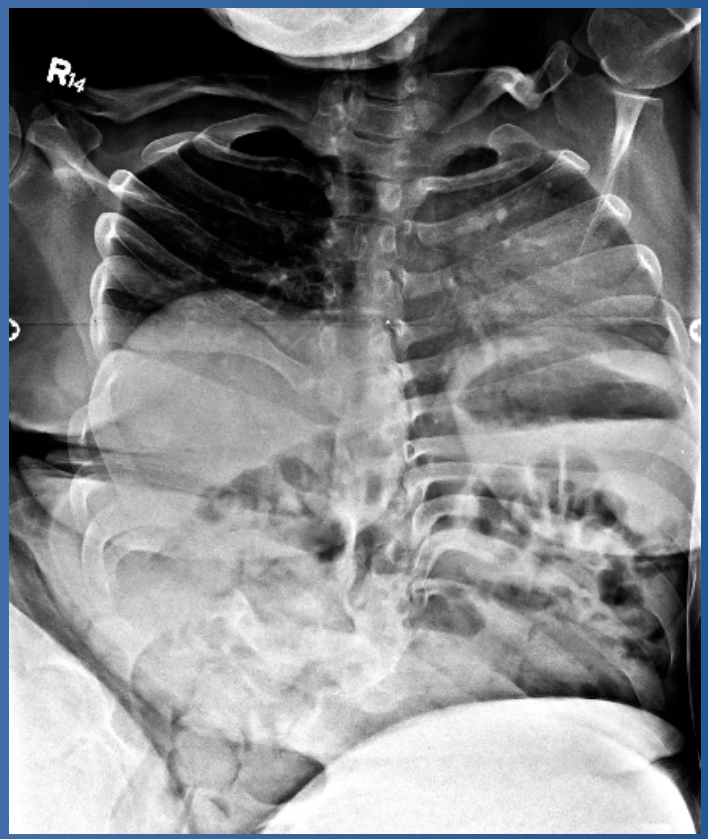


2. Ideas which Fell Short
Old Maxim

- *“ A short straight spine is better than a short crooked spine”*
  - R. Winter 1986

But far from ideal!

- Sometimes achieve neither!

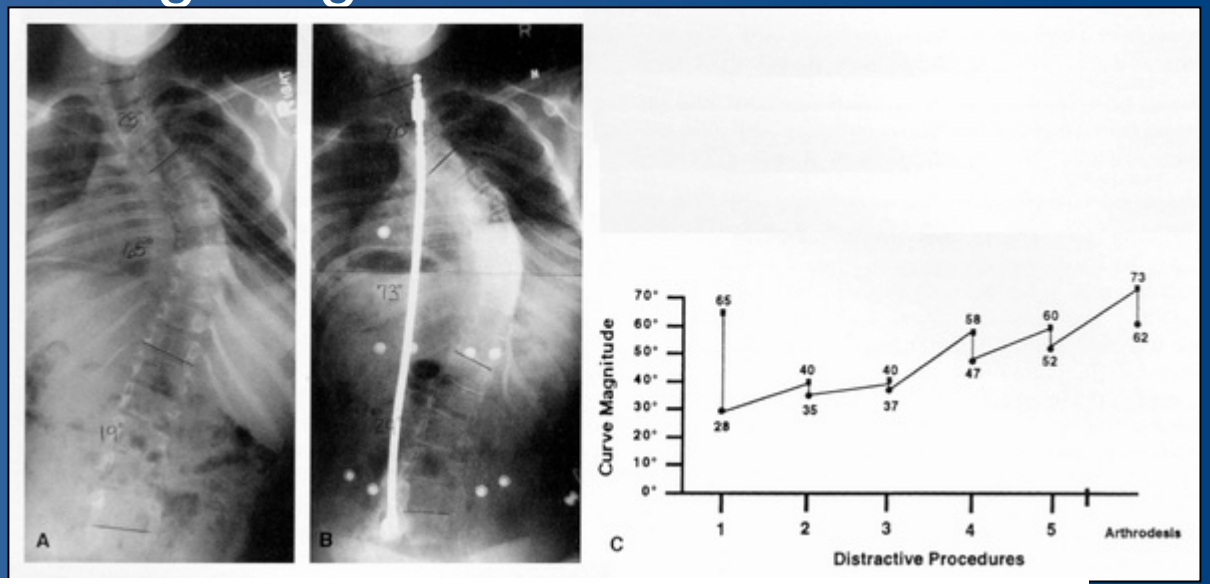


2. Ideas which fell short

- 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



Spinal Instrumentation Without Fusion for Progressive Scoliosis in Young Children

Klemme, William R. MC, USA; Denis, Francis M.D.*; Winter, Robert B. M.D.*; Lonstein, John W. M.D.*; Koop, Steven E. M.D.+

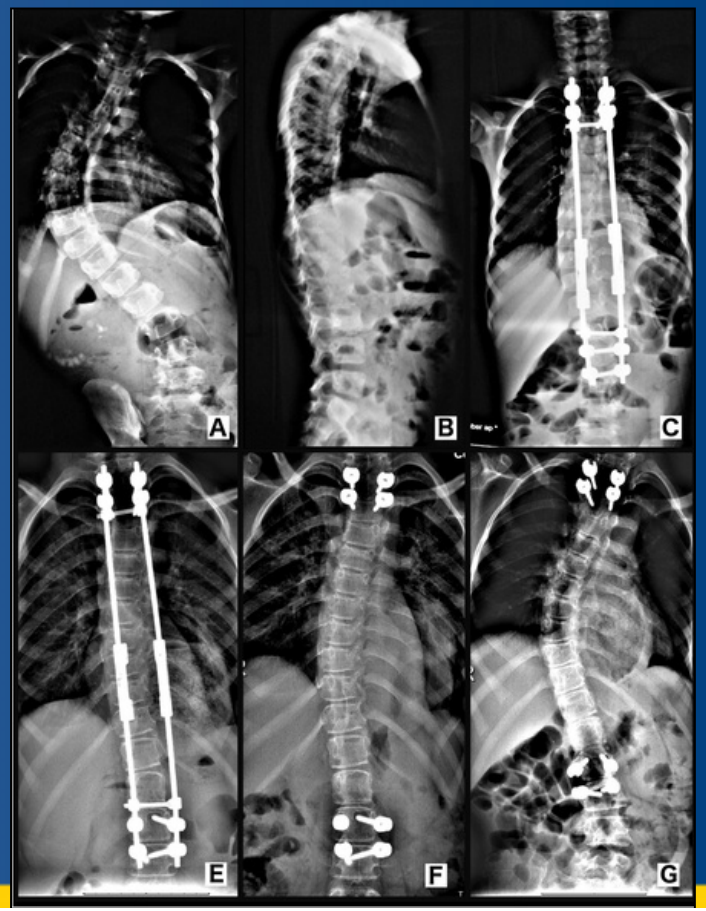
2. Ideas Which Fell Short

- Temporary Instrumentation

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

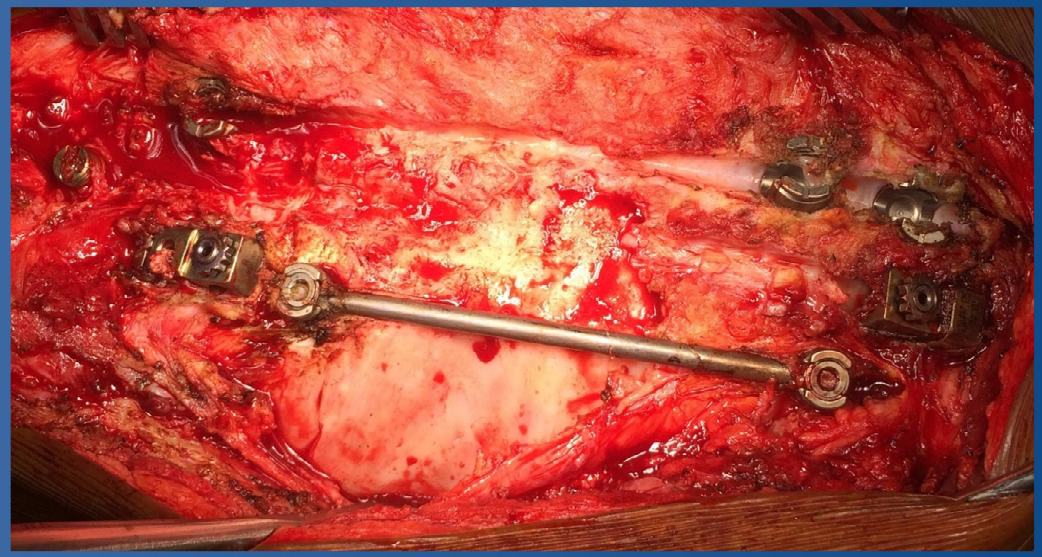
Ismail Aykut Kocyigit, MD, Z. Deniz Olgun, MD, H. Gokhan Demirkiran, MD, Mehmet Ayvaz, MD, and Muharrem Yazici, MD

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

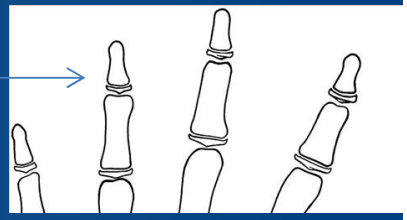
- Concepts:
  - Spinal Growth
  - T.I.S.
  - Diminishing Returns
  - C-EOS
- Techniques:
  - Mehta Casting
  - Growing implants
  - Growth guidance (Trolley, Shilla, tether)

3. Game-Changing Concepts:
Assessment of Growth & Maturity

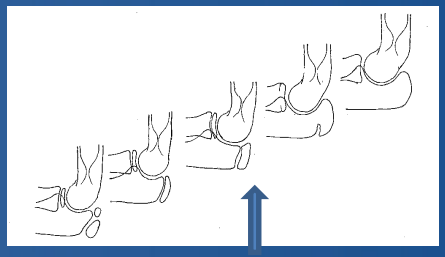
- Growth occurs in two peaks: 0-2 and PHV
- Significance of peak height velocity (PHV)
- What are markers of PHV?

Estimate of Peak Growth Velocity

- Premenarche
  - ≤11 years of age
- Risser 0



- **<u>Better:</u>**
- Open triradiate cartilages
- Sanders DMS stage 3 (capped not fused)
- Sauvegrain –rectangular olecranon
- Blood test?



Game-Changing Concept:
Thoracic Insufficiency Syndrome (TIS)

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The Inability of the Thorax to Support

Normal Respiration

or

Lung Growth

Campbell, Smith, et al.
J Bone Joint Surg, Mar , 2003
J Bone Joint Surg, Aug, 2004

Thoracic Insufficiency Syndrome (TIS)

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- 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"

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Spine stiffens with time

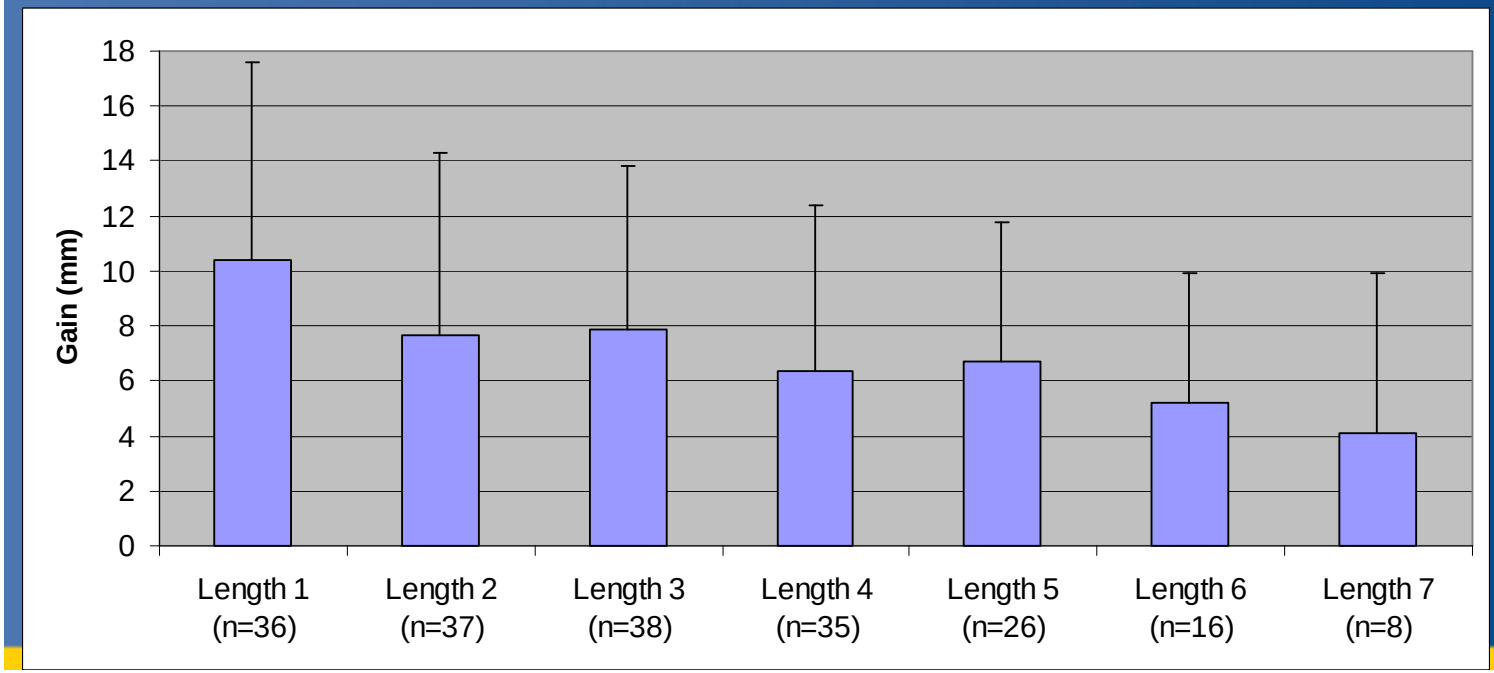
Rigid Immobilization?

Surgical exposure?

Surgical stretch?

Implication; surgery to be used sparingly & timely

Sankar and Skaggs



hp1

Concept Game Changer: C-EOS

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

Etiology	Cobb Angle (Major Curve)	Maximum Total Kyphosis	Progression Modifier (optional)
Congenital/Structural	1: <20º	(-) <20º	P0: <10º/yr
Neuromuscular	2: 21-50º	N: 21-50º	P1:10-20º/ yr
Syndromic	3: 51-90º	(+): >50º	P2: >20º/yr
Idiopathic	4: >90º		

hp1 hyp2102, 8/30/2012

Treatment Game-Changer
Mehta Casting

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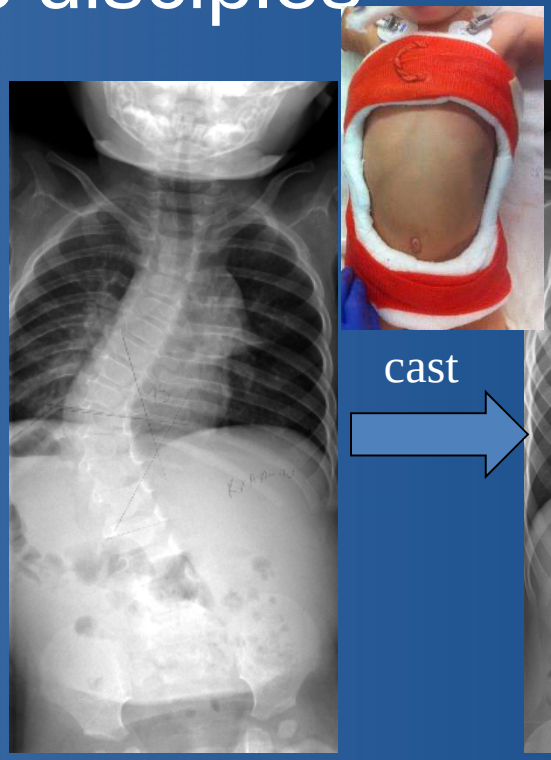
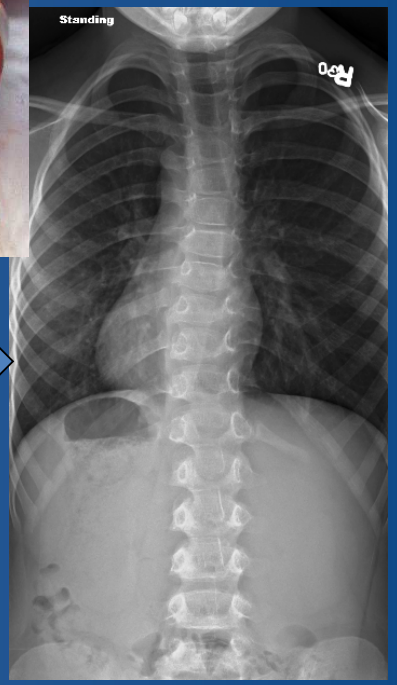
- Like Ponseti, it took a while for us to catch on
- Enthusiastic disciples
  - Sanders
  - D'Astous



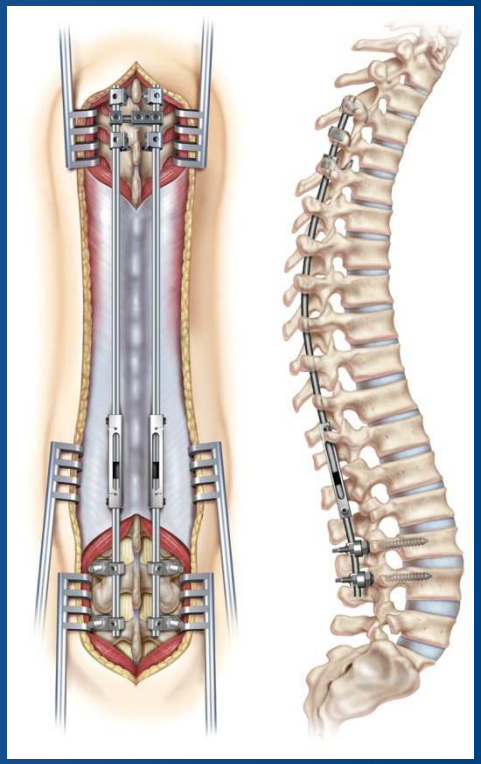
cast



Straight at 6 yrs old

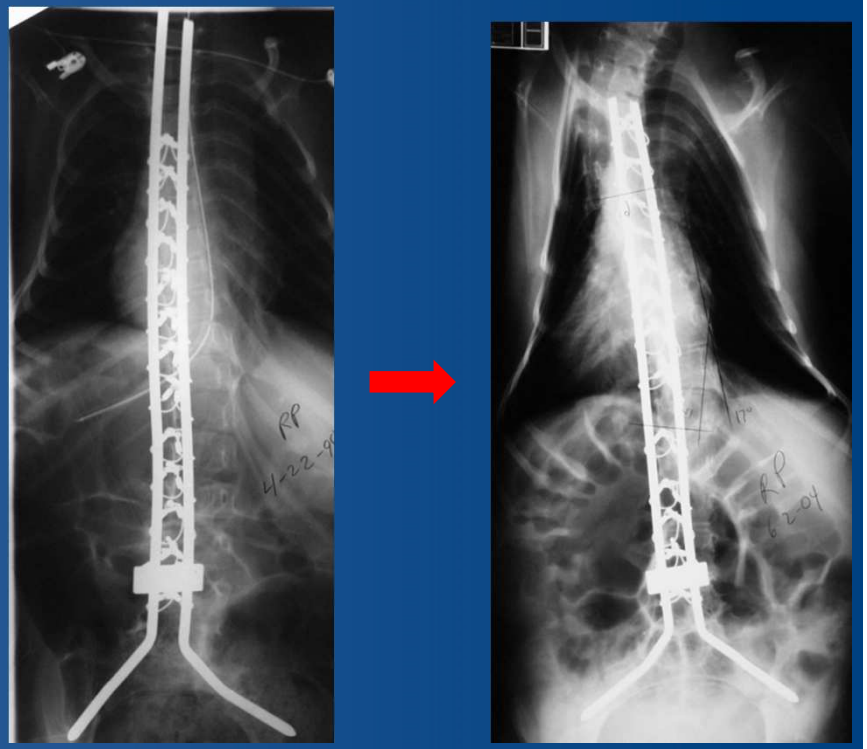
Treatment Game Changers: Dual (Growing) Rods

- 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

Dedicated meeting: ICEOS

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4. The Role of SRS and Societies in Advancing EOS care

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- SRS Mission: *To Foster the Optimal Care of <u>All</u> 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

POSNA/SRS PEDIATRIC DEVICE TASK FORCE

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:

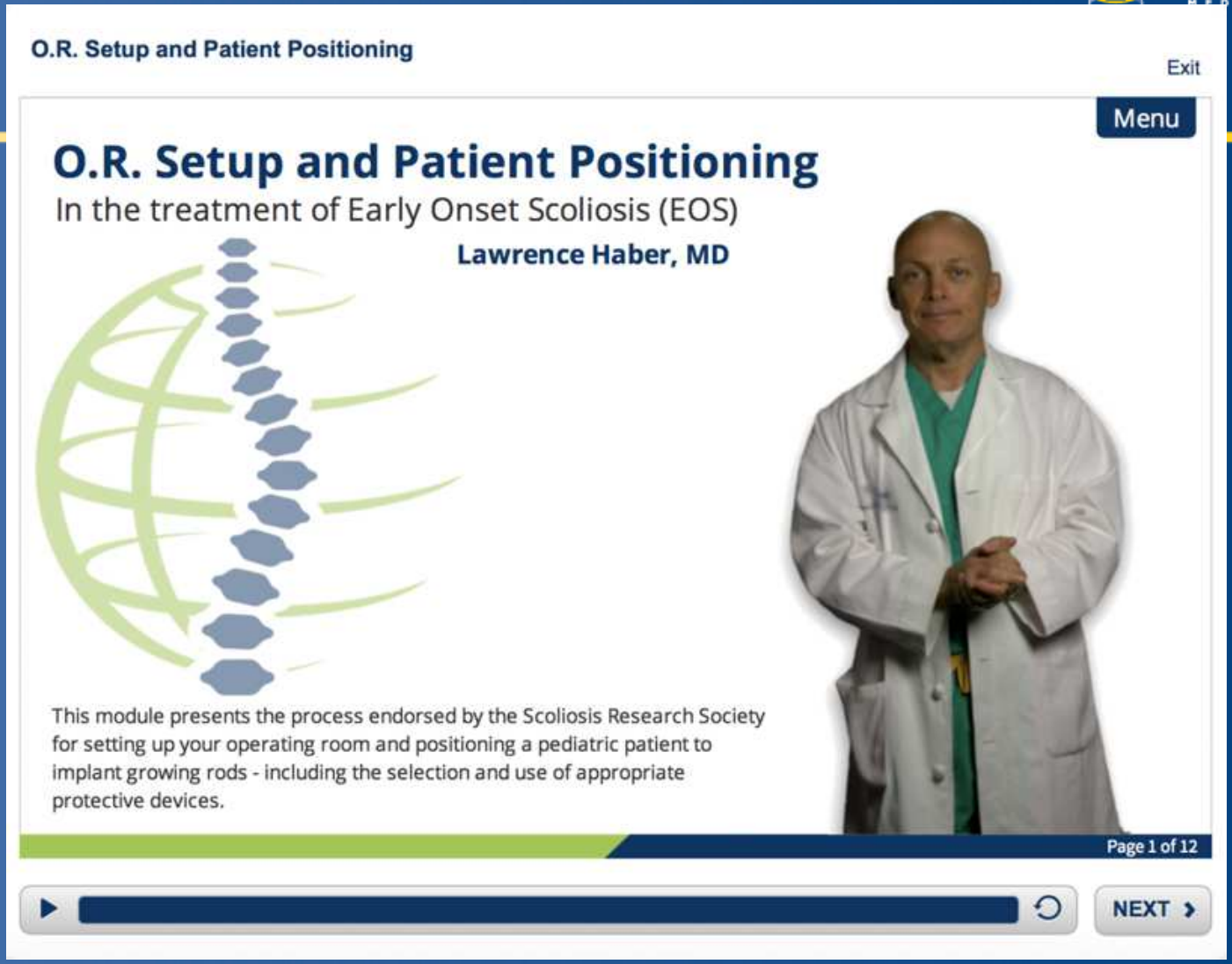
- OREF/SRS/POSNA solicit investigator-initiated research proposals focused on the development of new research knowledge that will change treatment for **growing spine conditions** within the next five (5) years. This funding opportunity should be uniquely important to early onset/growing spine research 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

SRS –EOS Learning module

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

Muharrem Yazici MD
Chairperson EOS Editorial Team

  - Muharrem Yazici, Laurel Blakemore, Larry Haber
  - HVR
  - GR
  - Mehta Cast


O.R. Setup and Patient Positioning
Exit
Menu
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.
Page 1 of 12
NEXT

Start

5. Relationship with the Family

Mercer Rang, MD

EOS Parent Decision Tool & Ed.

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Treatment of Early Onset Scoliosis (EOS): What Families Need to Know

Table of Contents

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?

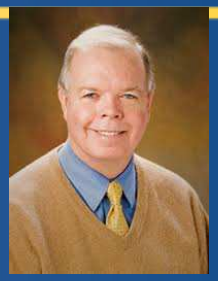
There are many causes for EOS. They are divided into four categories. Treatment choices differ depending on which category your child's scoliosis is in.

- EOS parent videos
  - PSSG website
  - SRS/POSNA

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

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Discipline

- Anesthesiology

What we can teach

- Antibiotics
- Blood management
  - Smaller = riskier
- Cord perfusion
- Relevant neurology
- Working with IONM

Improve Education Outside Spine World

Discipline	What we can teach
• ID	• Organisms by Diagnosis • Early vs late infection mgmt. • Is it osteo? • We can usually save the implants!

Improve Education Outside Spine World

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Discipline	What we can teach
• Pulmonary	• Axial growth principles • 25º vs 95º curve • Lordosis vs kyphosis • Large airway blockage • Spinal intrusion • Sternal deformities

Improve Education Outside Spine World

Discipline	What we can teach
• GI / nutrition	• Spine/ GI physiology • Lumbar lordosis – Spine-based obstruction • Improving nutrition – Pre-op – Post-op

Improve Education Outside Spine World

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Discipline

- IONM, Neurology

What we can teach

- Different monitoring for different deformity
- Deformity/risk relationships
- Working together

Understand Science of Disordered Axial Physiology

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- Airway
- Viscera
- Ribs/sternum

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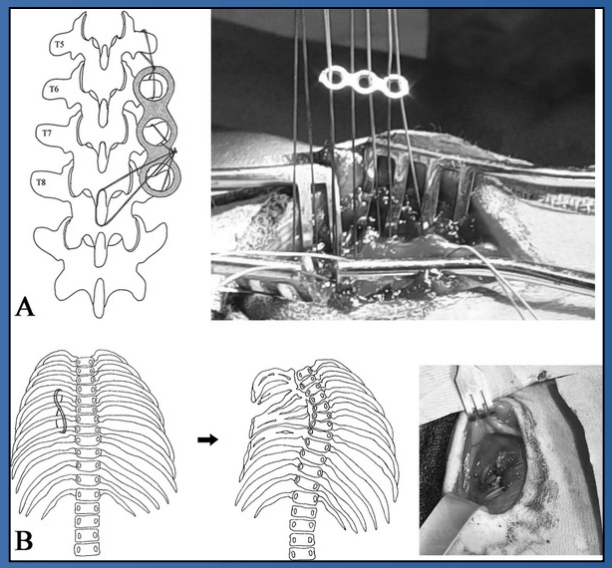
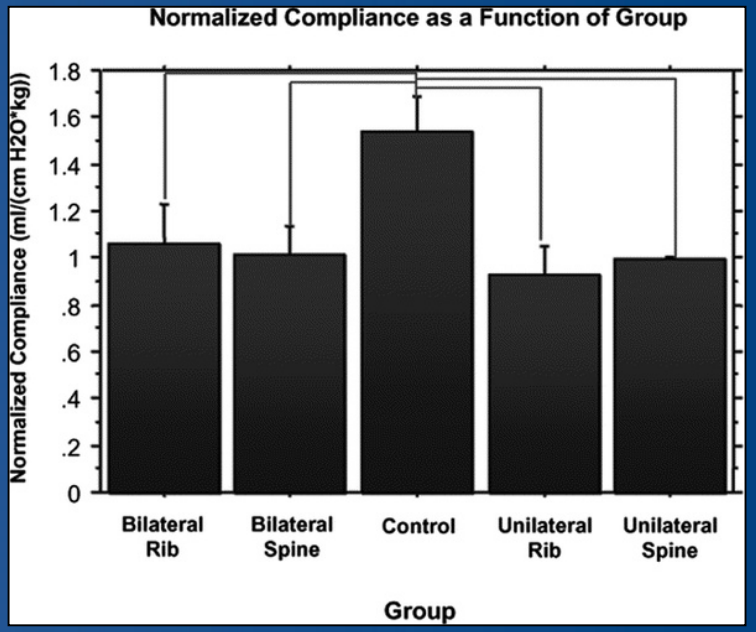
The Reciprocal Relationship Between Thoracic and Spinal Deformity and Its Effect on Pulmonary Function in a Rabbit Model

A Pilot Study

Hemal P. Mehta, MS,*† Brian D. Snyder, MD, PhD,*‡ Natasha N. Callender, BS,†
Carissa L. Bellardine, MS,† and Andrew C. Jackson, PhD†



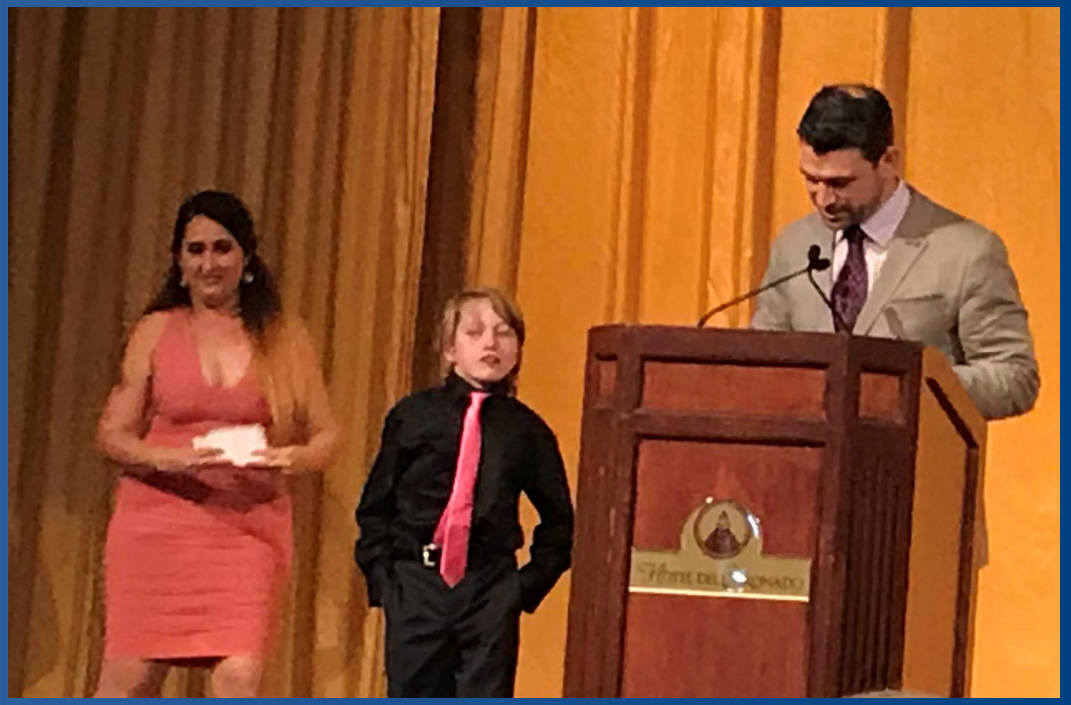


Translational Therapies

- 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

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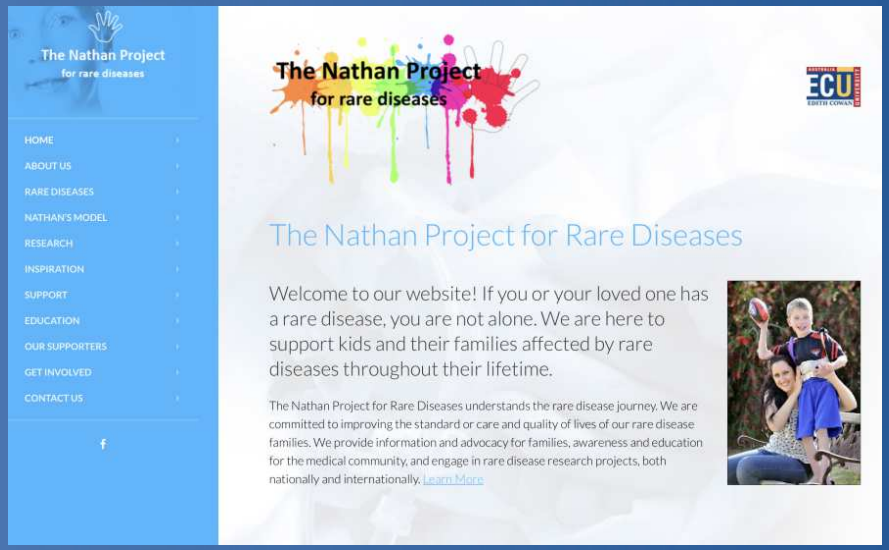
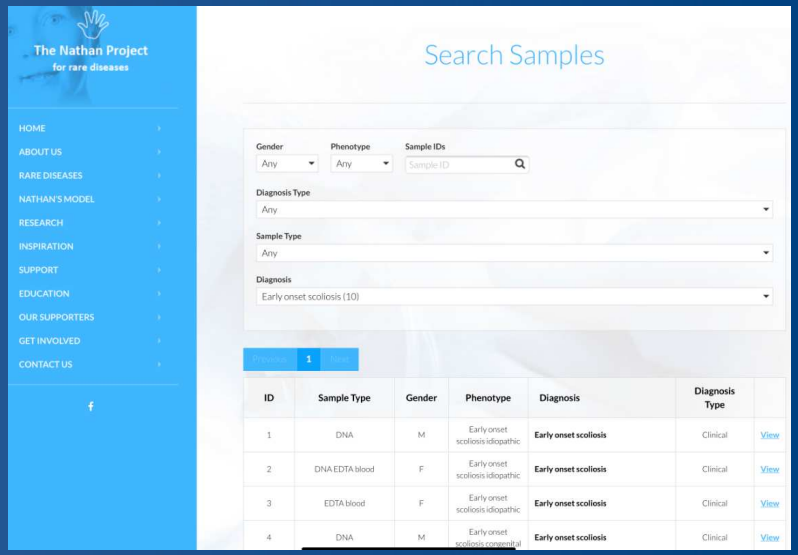
Aims

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

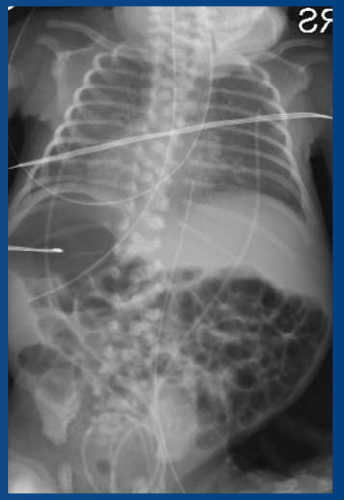
- Accelerate research advancements for EOS
- Translate discoveries into diagnostic tools and therapies





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?

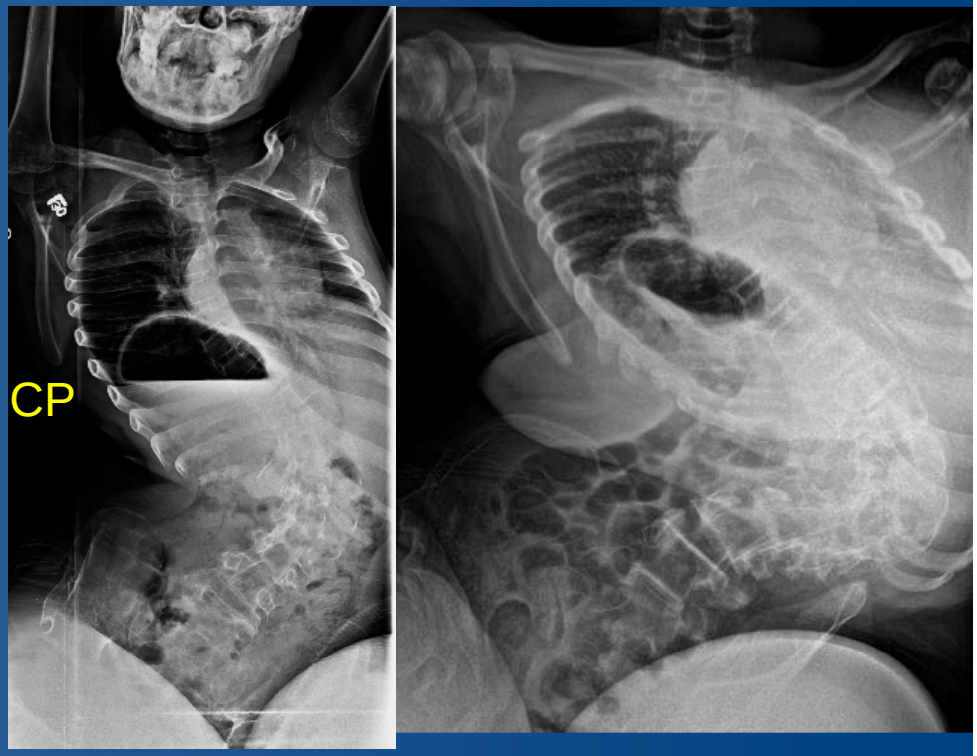


2013 2019

Better Understanding of Natural History

– Which neuromuscular curves benefit from PSF

Same Current CP
CHILD score



2013 2019

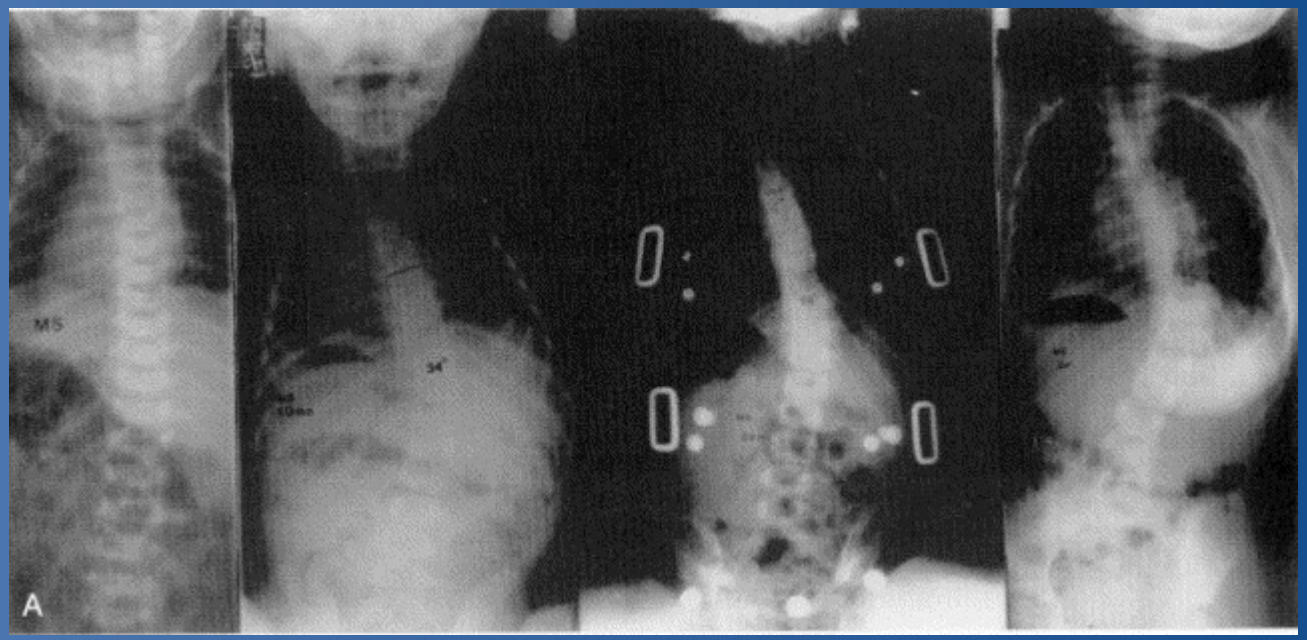
Summary

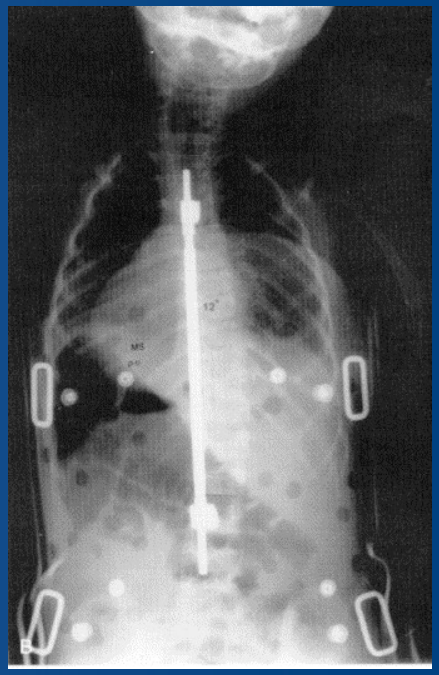
- Many factors have gotten us here
- Group wisdom & Societal resources will continue to guide progress
- To solve the biggest problem in Spine!

Thank You & Happy Thanksgiving!

2. Ideas which fell short

- Temporary Single rods





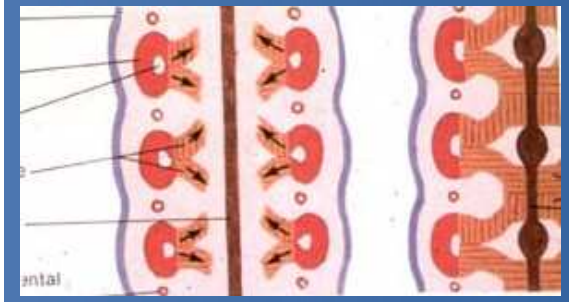
The Early Onset Scoliosis Biobank

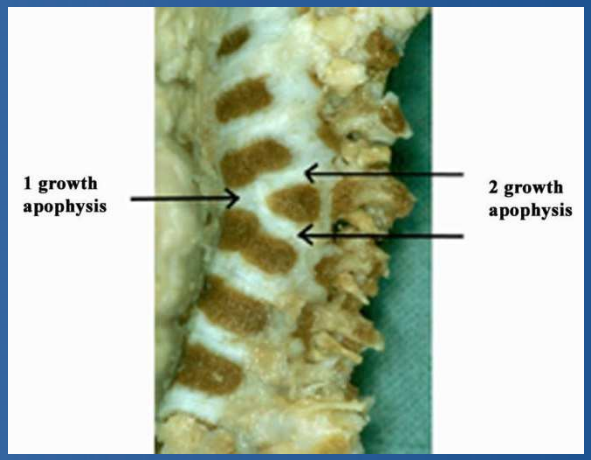
Spine Deformity

- 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





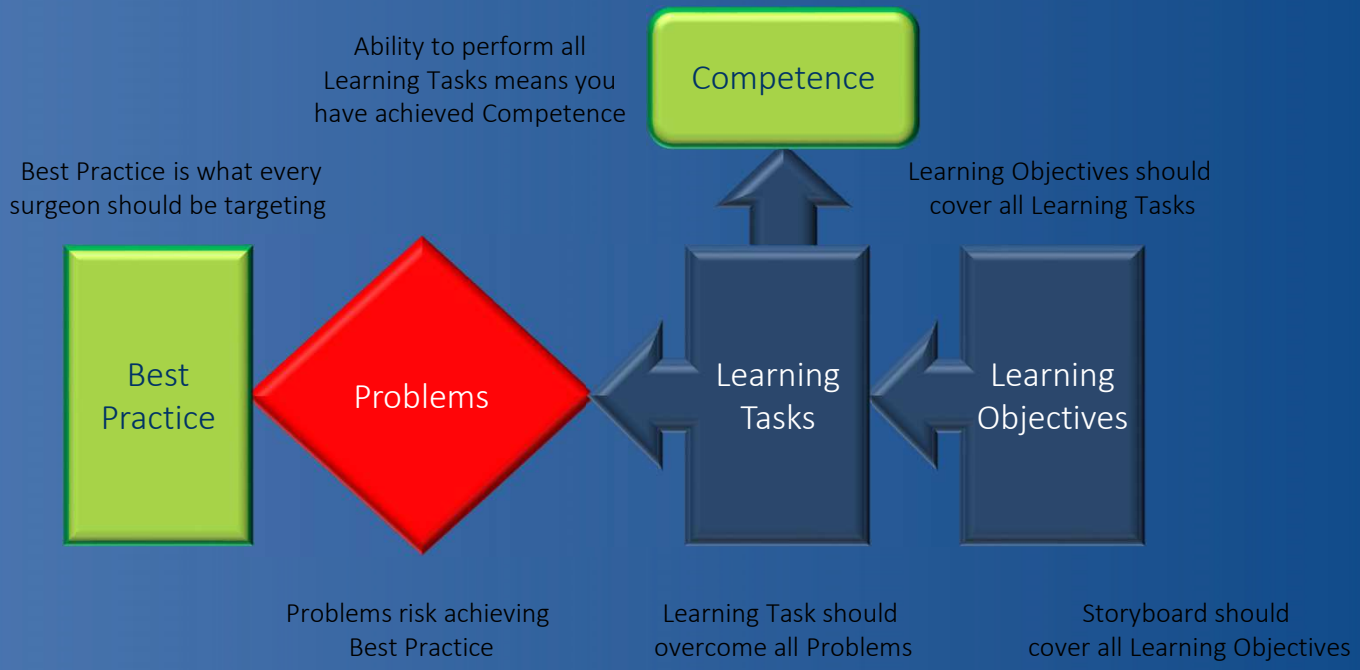
Previous Treatments

- Bracing
- Moe Rods
- Luque trolley

Infantile - Rx

- Mehta Cast, then brace for progressive type < 4 y.o.

Steps taken to reach Storyboarding…



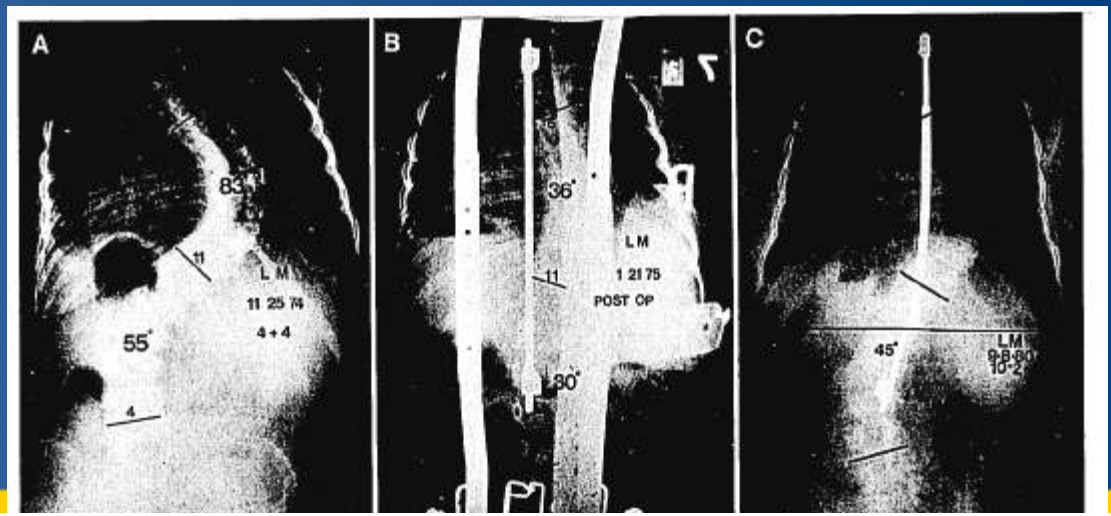
Ga

- 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 < 10º 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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Respiratory Function and Cosmesis at Maturity in Infantile-onset Scoliosis

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F. E. Dowling, F.R.C.S.I., B.Sc.‡

JOHNS HOPKINS MEDICINE

(C Goldberg)

- 23 pts- 6

g Volumes (N = 21), Surface Topography (N = 23), Latest Angle (N = 23), and Age at Surgery (N = 17)

	Age	FVC	FEV_1	VA	T_1S_1O	POTSI	SHS	AS1
Age:								
R	1.000	.653**	.653**	.670**	−.361	−.463	−.618**	−.796**
P		0.004	0.004	0.006	0.154	0.061	0.008	<0.001
N	17	17	17	15	17	17	17	17
FVC:								
r	.653**	1.000	.976**	.955**	−.627**	−.767**	−.696*	−.779*
P	0.004		<0.001	<0.001	0.001	<0.001	<0.001	<0.001
N	17	23	23	21	23	23	23	23
FEV_1:								
r	.653**	.976**	1.000	.948**	−.605**	−.749**	−.724**	−.756**
P	0.004	<0.001		<0.001	0.002	<0.001	<0.001	<0.001
N	17	23	23	21	23	23	23	23
VA:								
r	.670**	.955**	.948**	1.000	−.645**	−.730**	−.749**	−.721**
P	0.006	<0.001	<0.001		0.002	<0.001	<0.001	<0.001
N	15	21	21	21	21	21	21	21
T_1S_1O								
r	−.361	−.627**	−.605**	.645**	1.000	.705**	.263	.318
P	0.154	0.001	0.002	0.002		<0.001	0.226	0.140
N	17	23	23	21	23	23	23	23
POTSI								
r	−.463	−.767**	−.749**	−.730**	.705**	1.000	.583**	.684**
P	0.061	<0.001	<0.001	<0.001	<0.001		0.004	<0.001
N	17	23	23	21	23	23	23	23
SHS:								
r	−.618**	−.696**	−.724**	−.749**	.263	.583**	1.000	.826**
P	0.008	<0.001	<0.001	<0.001	.226	0.004		<0.001
N	17	23	23	21	23	23	23	23
AS1:								
r	−.796**	−.779**	−.756**	−.746**	.318	.684**	.826**	1.000
P	<0.001	<0.001	<0.001	<0.001	.140	<0.001	<0.001	
N	17	23	23	21	23	23	23	22
Cobb:								
r	−.574*	−.784**	−.746**	−.713**	.358	.578**	.644**	.896**
P	0.016	<0.001	<0.001	<0.001	.093	0.004	0.001	<0.001
N	17	23	23	21	23	23	23	23

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

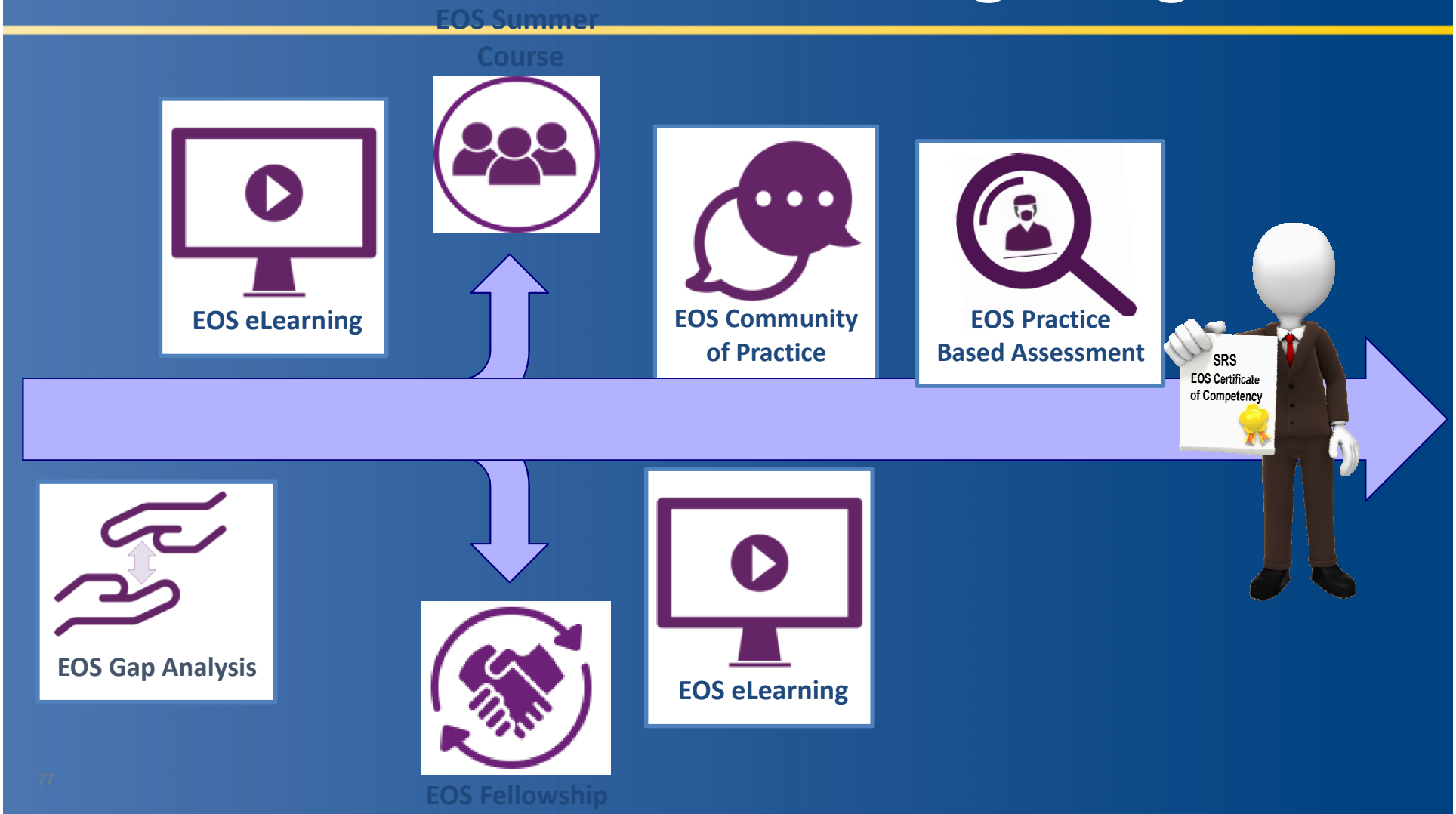
Outline

- 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; whe
  - 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

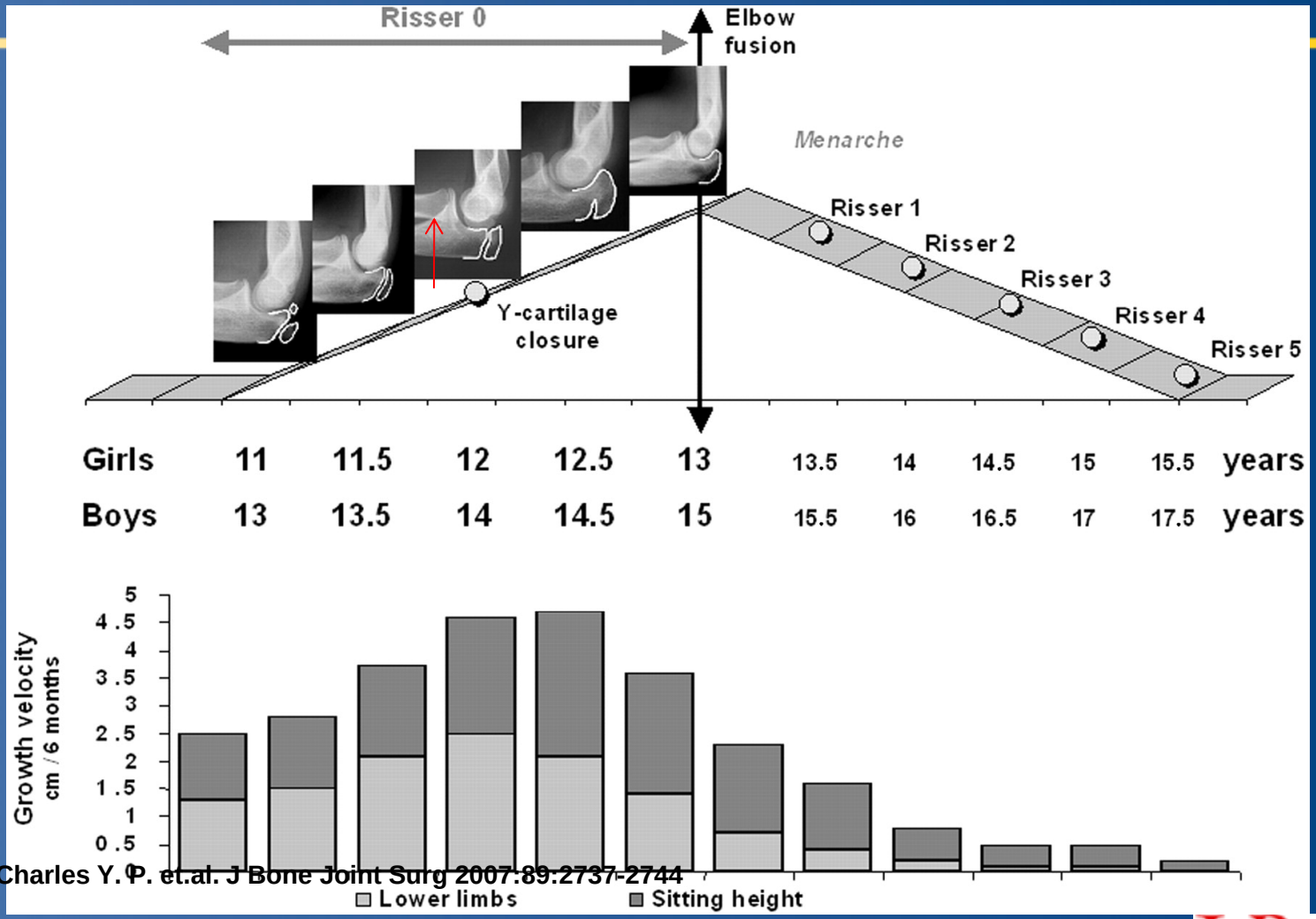
SRS EOS Blended Learning Program



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



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