PLEASE CLICK ON THE FOLLOWING LINK TO WATCH THE LECTURE ONLINE:-

- https://www.youtube.com/watch?v=XjGQsIj-VX0&list=PLuBRb5B7fa_embZp8jWG_hG8_o1JXLEeo&index=10

Early Onset Scoliosis

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Orthopedic Surgery Specialist

Spine Orthopedic Fellow

Early Onset Scoliosis

- Definition
- Types
- Thoracic Insufficiency Syndrome
- Initial Evaluation-Diagnosis
- Prognosis
- Natural history of untreated EOS
- Discussion each type alone
- Outcome and Mortality
- Classification
- Goals of Management
- Nonsurgical management
- Surgical management

Definition

- lateral (side-to-side) curve of the spine
- Cobb angle more than 10 degree
- three dimension deformity
- presenting in children younger than 10 years of age.
- Severe complex deformity of the spine and thorax
- 10% of all pediatric scoliosis cases.
- Cognitive, functional, and medical involvement within this population varies from normal to severely impaired.

Types

- Early Onset Scoliosis
  1. Congenital Scoliosis
  2. Neuromuscular Scoliosis
  3. Syndromatic Scoliosis
  4. Idiopathic Scoliosis: cause is unknown, child is otherwise healthy, (including “infantile” (0-3 years old) or “juvenile” (4-10 years old) scoliosis.
- Late Onset Scoliosis

Types

Review > World J Orthop. 2013 Oct 18;4(4):167-74. doi: 10.5312/wjo.v4.i4.167.

Normal and abnormal spine and thoracic cage development

Federico Canavese [1], Alain Dimeglio

- Why early and late onset ?
  - Spinal growth
    - Pathological alteration of Spine due to flexible spine which make it sensitive to deformity during growth ... affect thorax
    - Spine is mostly cartilaginous before age of 5, Birth 30% ossification and 5 years ossification 65%
    - Spine growth is not linear: 2cm / year from 1-5 year, 1.2 cm / year from 5 to 10 year and 1.8 cm / year from 10 to 15
    - Deformity progression: during these two period 5 and 10 years of age
  - Thoracic cage
    - 6.7% of the adult at birth
    - 30 % at 5 years
    - 50 % at 10 years
    - 100 % skeletal maturity

Types

Thorax (1962), 17, 329.

POSTNATAL GROWTH OF THE LUNG

BY

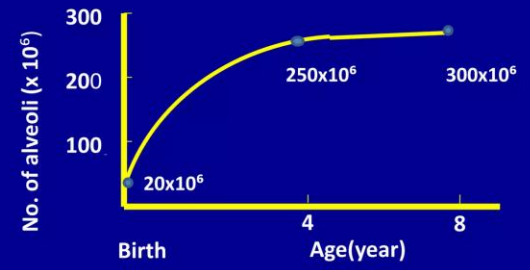
M. S. DUNNILL*

From the Department of Medicine, Columbia University, and the Cardiopulmonary Laboratory, Bellevue Hospital, New York

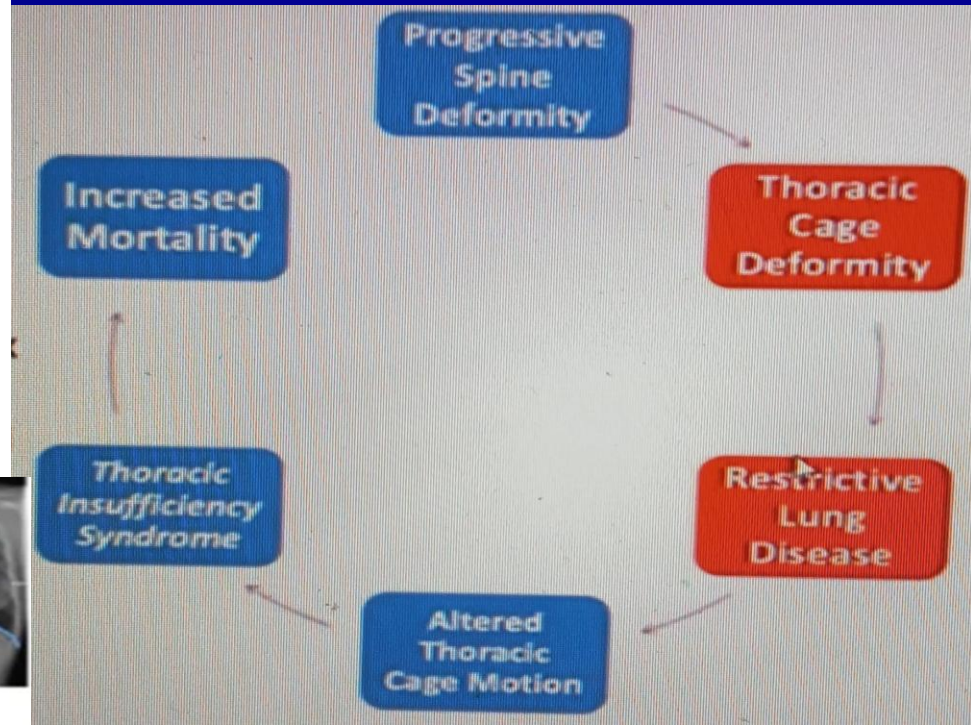
- Why early and late onset ?
  - Lung maturation
    - **First five years is crucial for lung maturation as the lung is growing dramatically**
  - Alveolization
    - **Alveolus multiples by the age of 8 years**
    - **Lung maturation continues**
    - **At birth 20 million alveoli**
    - **At 5 years 300 million alveoli**
  - T1-T12 thoracic height (target >18cm.. 80% of expected thoracic height HrQOL improves)
    - **12 cm at birth**
    - **18 cm at 5 years**
    - **27 cm adult**

Early Onset Scoliosis

Lung development



Early growth disturbance compromises thoracic volume

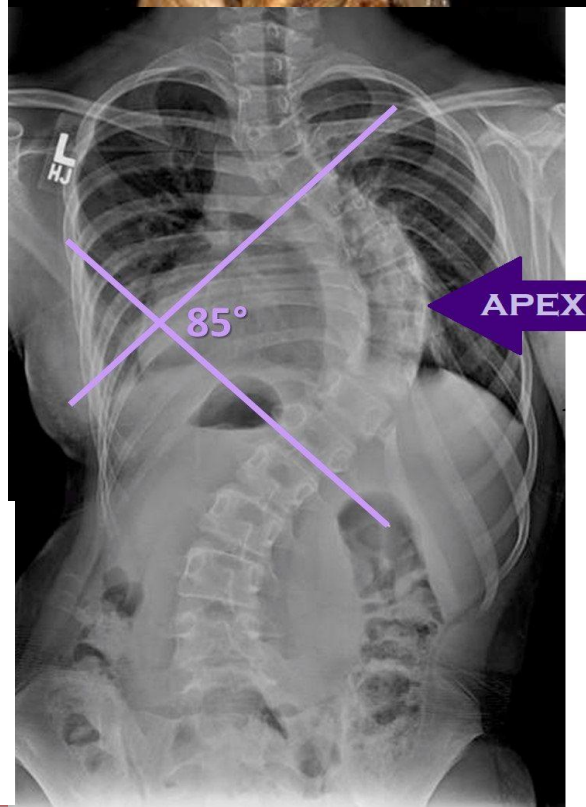


Types

- Why early and late onset ?
  - Early onset scoliosis
    - **Rare and severe**
    - **Male 2:1**
    - **Left sided**
    - **Systemic anomalies**
    - **Mostly syndromatic**
  - Late onset scoliosis
    - **Adolescent idiopathic**
    - **Female 6:1**
    - **Right thoracic**
    - **No other anomalies**
    - **Fhx common**

Thoracic insufficiency Syndrome

- Inability of the thorax to support normal lung growth and breathing
- Characterized by decreased thoracic growth and lung volume
- Deficiencies in chest volume, symmetry and lung function
- It's a 3D thoracic deformity (Respiratory Engine: Sternum, Ribs, Spine)
  - Kyphosis and / or scoliosis
  - Ribs anomalies
  - Lung disorders



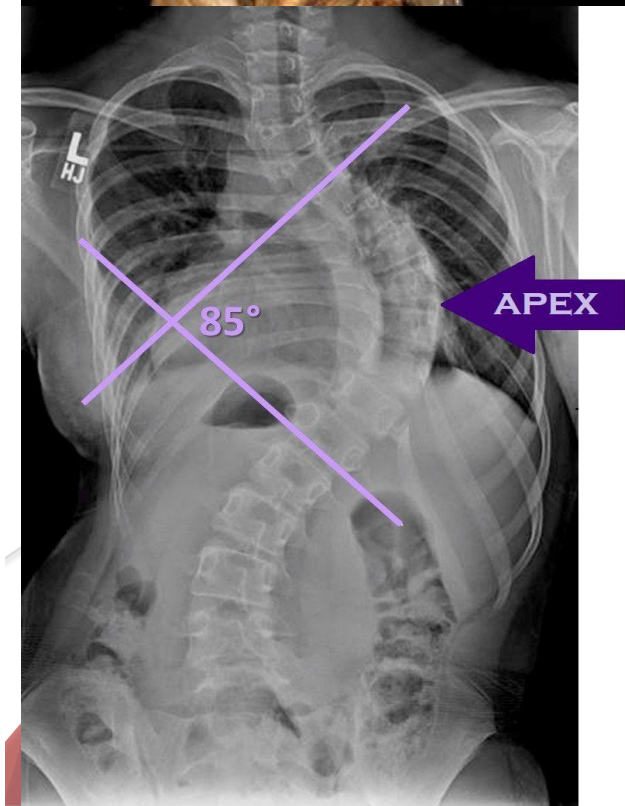
SCIENTIFIC ARTICLE

The Characteristics of Thoracic Insufficiency Syndrome Associated with Fused Ribs and Congenital Scoliosis

Campbell, Robert M. Jr. MD; Smith, Melvin D. MD; Mayes, Thomas C. MD; Mangos, John A. MD; Willey-Courand, Donna B. MD; Kose, Nusret MD; Pinero, Ricardo F. MD; Alder, Marden E. DDS; Duong, Hoa L. MD; Surber, Jennifer L. BS

Thoracic insufficiency Syndrome

- Leads to pulmonary hypertension and Cor pulmonale
- Pulmonary function impairment associated with curve >60 degree
- Cardiopulmonary issues associatedwith curve >90 degree
- O2 dependant, NIPPV,ventilator and reduced life span
- Poor quality of life compare to child with epilepsy, heart disease and cancer (vitale J,2008)



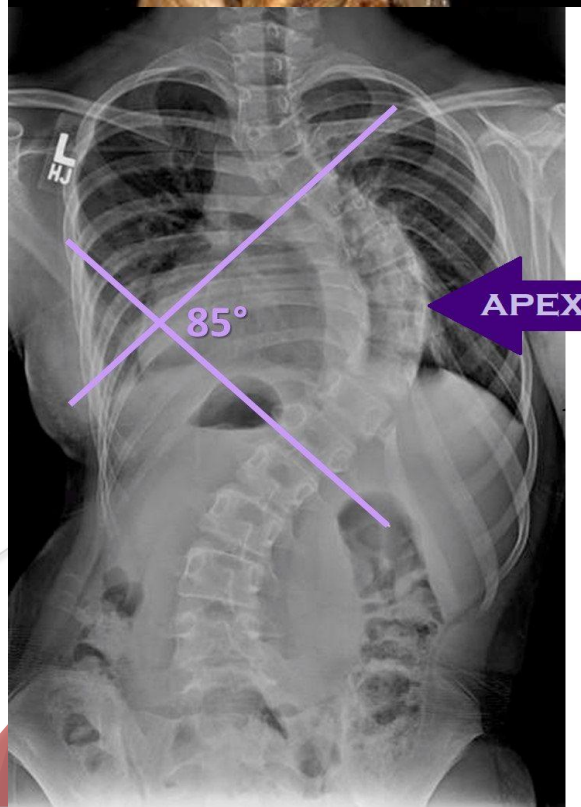
Multicenter Study > J Pediatr Orthop. 2008 Mar;28(2):239-43.
doi: 10.1097/BPO.0b013e31816521bb.

Health-related quality of life in children with thoracic insufficiency syndrome

Michael G Vitale [1], Hiroko Matsumoto, David P Roye Jr, Jaime A Gomez, Randal R Betz, John B Emans, David L Skaggs, John T Smith, Kit M Song, Robert M Campbell Jr

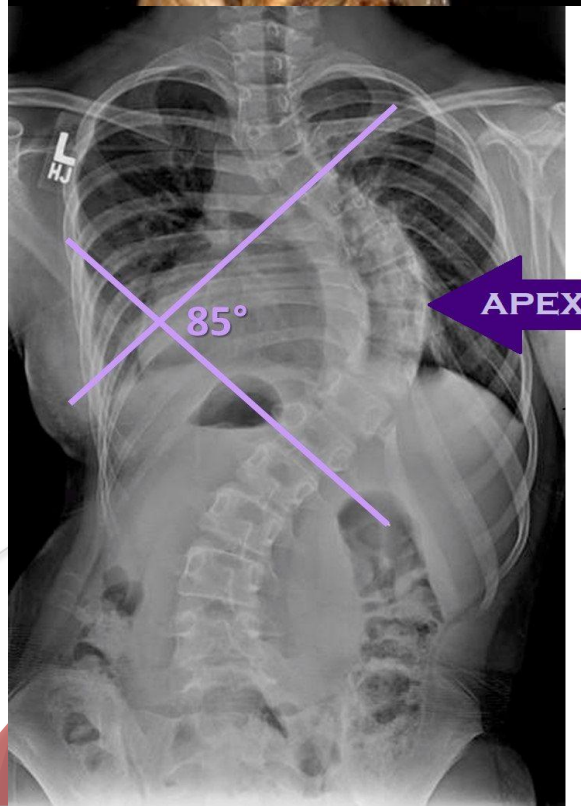
Thoracic insufficiency Syndrome

- For normal respiration
  - Diaphragm contract …air entered into the lung
  - Diaphragm relax …air exit from the lung
  - Ribs muscle
  - Symmetrical ribs
  - Ideal chest volume
- In congenital scoliosis…..missing (Flial chest) or fused ribs.
  - Side of fused or missing ribs …Lung collapses and constricted …in the concave side of curve
  - Other side continue of growth of convex curve of scoliosis…spine protrude into the chest at the convex side of curve what is called (sway back) which cause lung constriction at the other side of fused ribs.



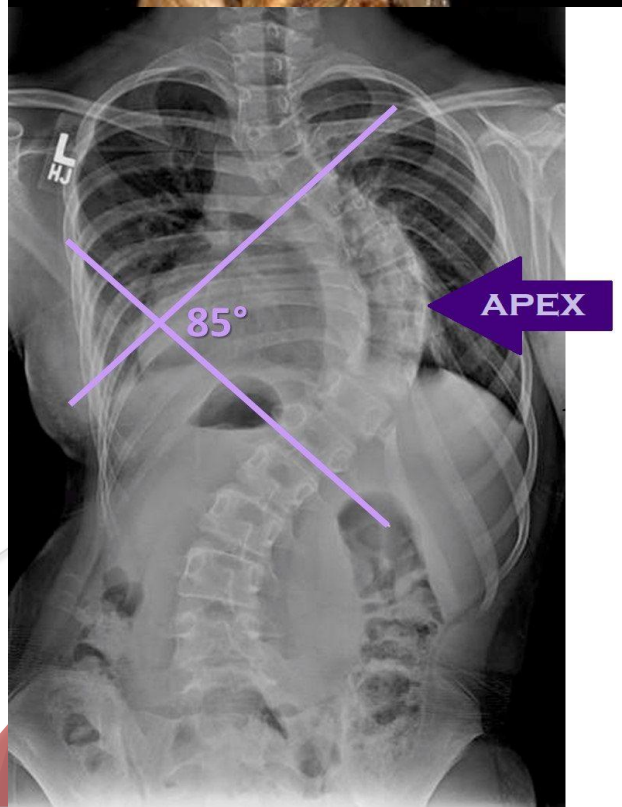
Thoracic insufficiency Syndrome

- Intrinsic alveolar hypoplasia
  - That is why the 5 first years of life have been considered ‘The Golden Period’
  - In conjunction with direct spinal elongation, the thoracic circumference doubles in magnitude after age 10 years.
- Extrinsic disturbance of chest wall function
  - In NMS , muscular problems exacerbates chest wall dysfunction
  - In non congenital deformity , scoliosis play a role in lung constriction at the concave side



Thoracic insufficiency Syndrome

- Diagnosis is complex
  - Detailed history
  - Physical examination
  - X-rays, CT scans
  - Pulmonary function studies, if obtainable
- EFFECT ON RESPIRATORY SYSTEM IS CORRELATED WITH AGE OF ONSET, SEVERITY OF THE DEFORMITY, ASSOCIATED RIB ANOMALIES, PRESENCE OF MUSCLE WEAKNESS



Initial Evaluation-Diagnosis

- History
  - Age deformity first noticed
  - Any progression
  - Perinatal history
  - Developmental milestones
  - Systemic illness
  - Family history

Initial Evaluation-Diagnosis

- **Physical Examination**
- **Height, Weight, Arm Span, Sitting Height**
  - **General appearance**
  - **Syndromic features**
  - Curve pattern
  - ➤ Right or left
  - Shoulder asymmetry
  - Truncal shift
  - Waist asymmetry
  - Costo pelvic impingement
  - Rib hump

- Spinal dysraphism
- Café-au-lait spots
- LLD, Feet deformity
- Ligamentous laxity
- **flexibility**
- **Neurological examination**
- **Abdominal reflex**

➢ Absent SAR

Initial Evaluation-Diagnosis

- Spine Xray (PA, Lateral and dynamic)
  - The details of vertebral abnormality are best seen on films obtained before the development of significant deformity often during infancy.
  - The initial radiograph must serve as the baseline study against which further curve progression is measured.
- CT with 3D is helpful for visualizing of vertebral anomalies and for preoperative planning
- MRI
  - As a rule of thumb, about 20% of children who are younger than 10 and who have a curve greater than 20º will have an underlying spinal condition.
  - MRI IS INDICATED during the initial evaluation and for all patients who are undergoing surgical intervention because up to 37% of patients have an intraspinal abnormality.

Initial Evaluation-Diagnosis

- ECHO
- PFT
  - FVC, FEV1
- Nutritional status
- MULTI DISCIPLINARY TEAM (MDT) APPROACH

Prognosis of EOS

- Severity-Early Pulmonary Function Compromises and increasing adulthood problem
- Neglected/untreated- Increased risk of early death
- Thoracic Insufficiency Syndrome (TIS)
- Depends on aetiology
  - ✓ Idiopathic/syndromic/underlying disorders or co-morbidities

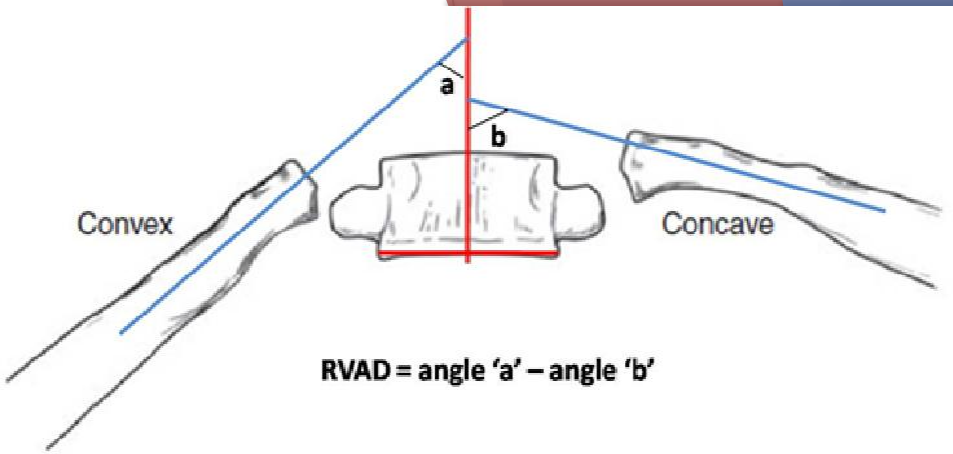
Prognosis of Idiopathies EOS

- Most children <2 years with infantile idiopathic curve <35 degree resolve spontaneously
- If progressive by age 5, 50% will have curve >70 degree
- **Mehta Predictor of progression**
- Cobb angle >20 degree
- RVAD >20 degree
- Phase 2 Rib-Vertebral relationship

Rib Vertebral Angle Difference (RVAD, Mehta angle)

- Measure angle between the endplate and rib (line between midpoint of rib head and neck)
- **RVAD = difference of 2 rib- vertebral angles**
- RVAD= Angle at concave- angle at convex area
- Apical vertebra
- > 20° is linked to high rate of progression
- <20° is associated with spontaneous recovery

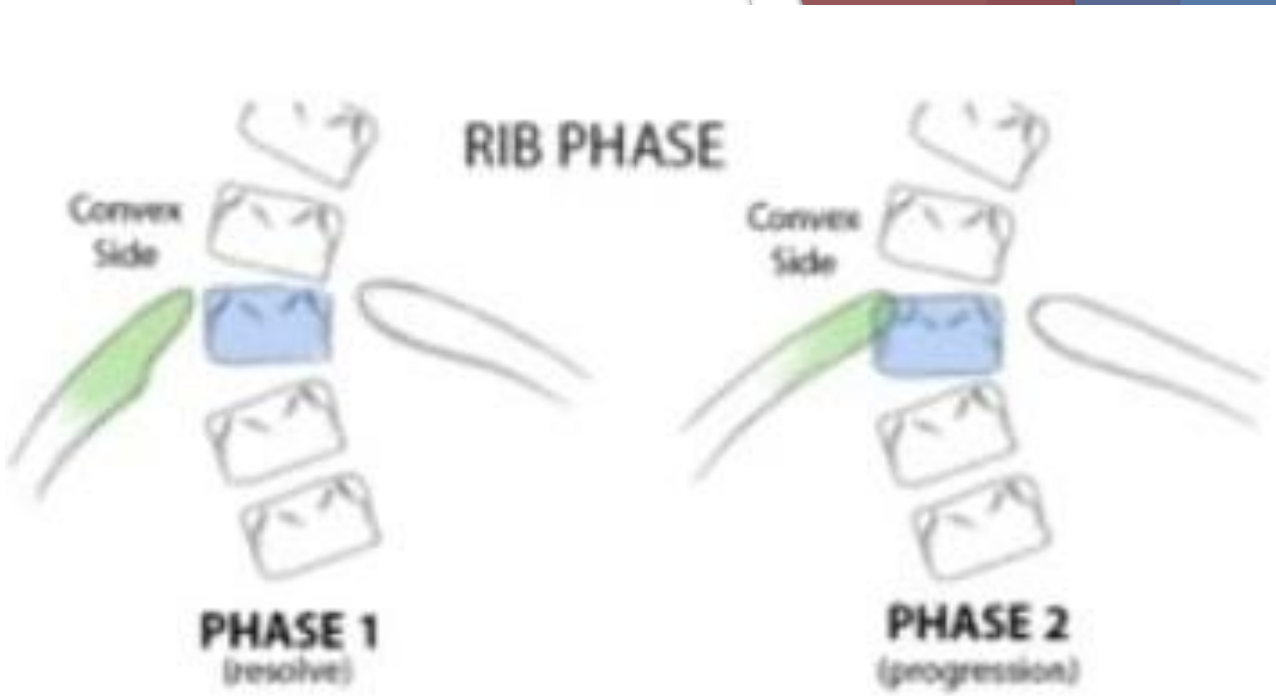
Mehta vs Kristmundsdot



- Mehta MH. The rib-vertebra angle in the early diagnosis between resolving and progressive infantile scoliosis.J Bone joint Surg Br 1972;54:230-243 PubMed
  - In a retrospective review of 138 patients with idiopathic EOS diagnosed before age 2, 80% of patients with progressive curves had an RVAD of> 20 degrees at initial presentation.
  - In contrast, 80% of patients with resolving curves had an initial RVAD of< 20 degrees.
  - Min meta brings RVAD in 1972
  - 80% predicting progression of infantile EOS
  - RVAD important implication in infantile EOS as it differentiate between progressive and resolving types of scoliosis during diagnosis
  - RVAD of 20 degrees and more is considered progressive
- Kristmundsdottir
  - use Convex RVA in 1980
  - 85% predicting progression of infantile EOS
  - Convex RVA of 68 degree and more is considered progressive.

Rib Phase

- Convex rib head position with respect to the apical vertebrae
- **Phase 1 - no rib overlap**
  - ✓ resolve
- **Phase 2 - rib overlap with the apical vertebrae**
- high risk for curve progression



Predicting progression of EOS

- Age
- Curve pattern (primary thoracic curve),
- Cobb angle (Infantile curves that reach 30 degree tend to continue to worsen without treatment)
- curve progression (6 to 7 degrees increase considered progressive curve)
- Cause (In curves associated with tethered spinal cord, early detethering has been suggested to reduce the scoliosis progression rate)

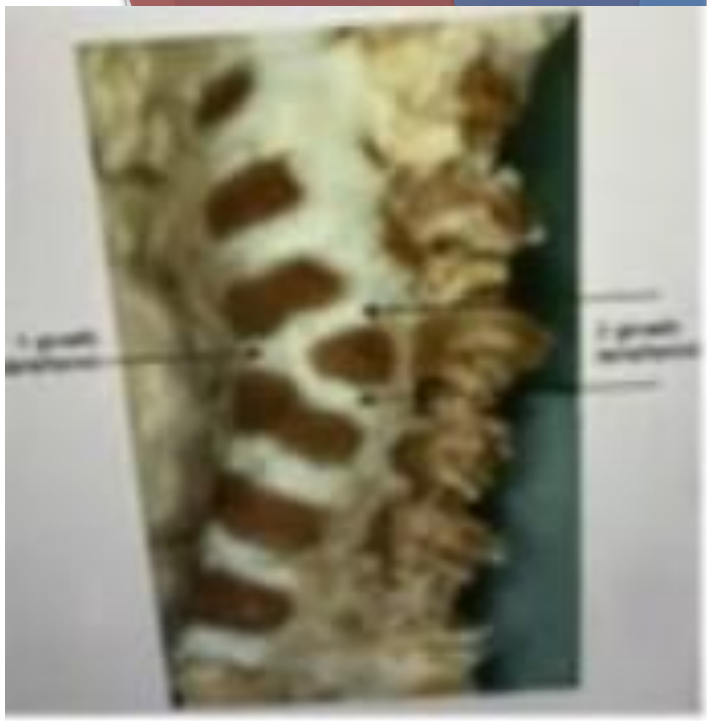
DOI: 10.1016/j.spinee.2022.03.016 • Corpus ID: 247944150

The Incidence and Prevalence of Early-Onset Scoliosis: A Regional Multicenter Epidemiological Study.

Mason AlNouri, Kanichiro Wada, +6 authors Y. Ishibashi • Published in The spine journal 1 April 2022 • Medicine

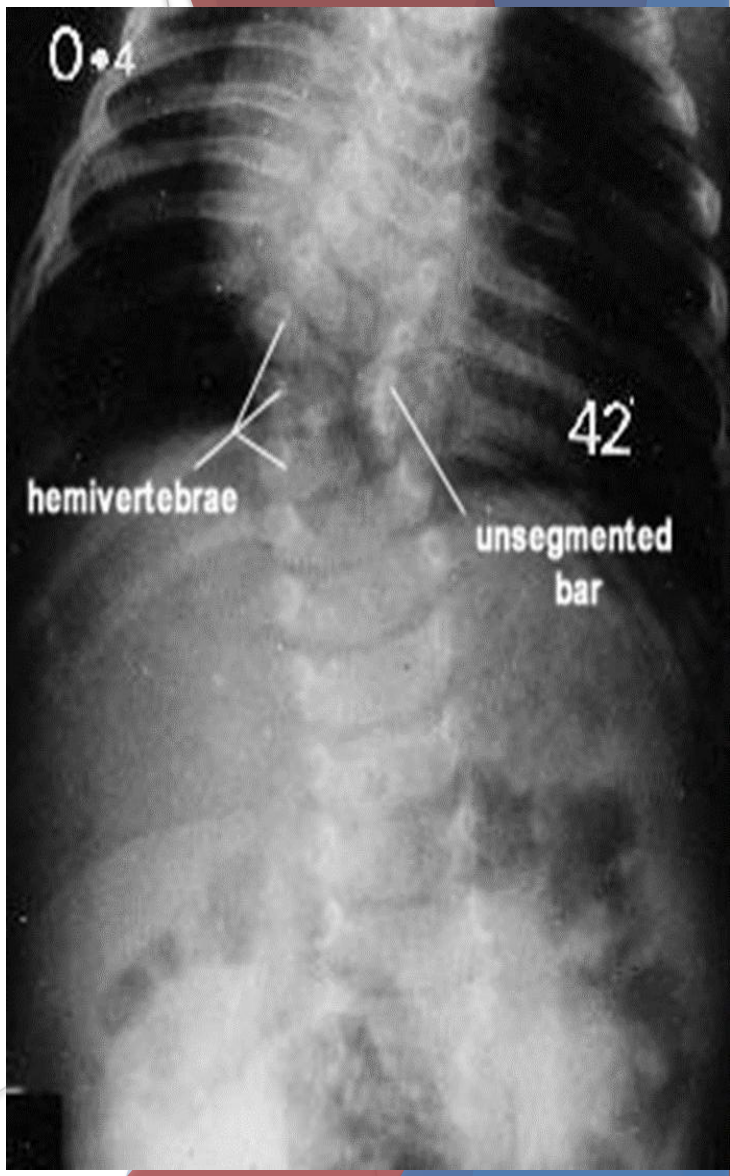
Prognosis of Congenital EOS

- **Rapid in the first 3 years of life**
- 70% progress during growth
  - Type of anomaly
  - Location
  - Number of anomalies
  - Initial severity of curve .. unilateral unsegmented bar with contralateral hemivertebra >>>>block vertebra
  - Global growth potential balance between each side of spine…Patients remaining growth at the time of diagnosis (two period of accelerated growth)
- Presence of fused ribs increases risk of progression



Rate of Progression Congenital EOS

- **Unilateral unsegmented bar with contralateral hemivertebra >**
  - greatest potential for rapid progression (5 to10 degrees/year)
- **Unilateral unsegmented bar**
- **Fully segmented hemivertebra**
- **Unincarcerated hemivertebra**
- **Incarcerated hemivertebra**
- **Unsegmented hemivertebra Block vertebrae**
  - little chance for progression (<2 degrees/year)



Prognosis of Neuromuscular EOS

- More rapidly progressive
- Progress even after skeletal maturity
- Ambulant Vs Non ambulator
- Long C curve in non ambulatory
- Pelvic obliquity affect sitting (esp non ambulator)
- Higher rate of pulmonary complication
- poor balance and poor coordination of their trunk, neck, and head.
- high frequency of concurrent kyphosis

Natural History of Untreated EOS

- Significant morbidity and often profound cardiopulmonary compromise
    - **Respiratory failure and Cor Pulmonale.**
- TIS poor quality of life compared to childhood epilepsy, heart disease and cancer **(Vitale J Paedtr Orthop 2008)**
- Swedish study reported twice the mortality rate by age 40 in patients with EOS compared with that of the general population (**Pehrrson et al Spine 1992**)

Idiopathic EOS

- Approximately 80% of patients with Scoliosis have Idiopathic Scoliosis
- Idiopathic scoliosis occurs in otherwise healthy individuals without a discernible cause
- Multifactorial : familial, molecular, biomechanical and neurogenic
- 75% of the curves resolves spontaneously
- 25% of the curves progress rapidly (malignant curve)
- Identification of malignant curves prevent respiratory compromise

Infantile Idiopathic Scoliosis

- Less than 1% of idiopathic scoliosis
- left-sided curves in the thoracic (chest) area.
- It is more common in boys
- Infants are healthy and normal, and simply have a small curvature of the spine.
- 90% OF CURVE SHOW SPONTANEOUS RESOLUTION
- RVAD and PHASE OF RIB HEAD can predict at risk of progression curve with angles difference more than 20 or phase 2 rib are very likely to progress

Juvenile Idiopathic Scoliosis

- 20% of idiopathic scoliosis
- More common in female
- Convex right thoracic curve is the commonest pattern
- More likely to progress, less likely to respond to brace treatment and more likely to require surgical treatment
- NEURAL. axis abnormalities is 18% to 26%, most of them are asymptomatic and have no signs of the underlying neural abnormality
- MRI IS RECOMMEDED during the initial evaluation…high incidence of ArnoldChiari malformation and syringomyelia.
- RVAD dose not predict curve progression but progressive curves have a steady increase in RVAD,WHERAS resolving curves have a steady decrease in RVAD.

Neuromuscular Scoliosis

Diagnosis	Incidence of Scoliosis
Cerebral palsy (2-limbs involved)	25%
Myelodysplasia (lower lumbar)	60%
Spinal muscle atrophy	67%
Friedreich ataxia	80%
Cerebral palsy (4-limbs involved)	80%
Duchenne muscular dystrophy	90%
Myelodysplasia (thoracic level)	100%
Traumatic paralysis (<10 years)	100%

- Involves neurologic or muscular deficiencies that produce progressive multiplanar spine deformities.
- These patients have an impaired ability to control the muscle that support the spine.
- They require a multidisciplinary team of doctors and health professionals working together to provide optimal care for them.
  - Pulmonary dysfunction
  - Cardiomyopathy
  - Urinary tract disease
  - Pressure sore
  - Hip dislocation

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Traumatic paralysis (<10 years)	100%

- Result from
  - neuropathic (nervous system related) disorders like Cerebral Palsy (CP), Spinal Muscle Atrophy (SMA), or trauma to the spinal cord
  - myopathic (muscular system-related) disorders like Muscular Dystrophy, Poliomyelitis, or Arthrogryposis.
- Children with spina bifida can also develop varying forms of spinal deformity depending on the level of involvement.

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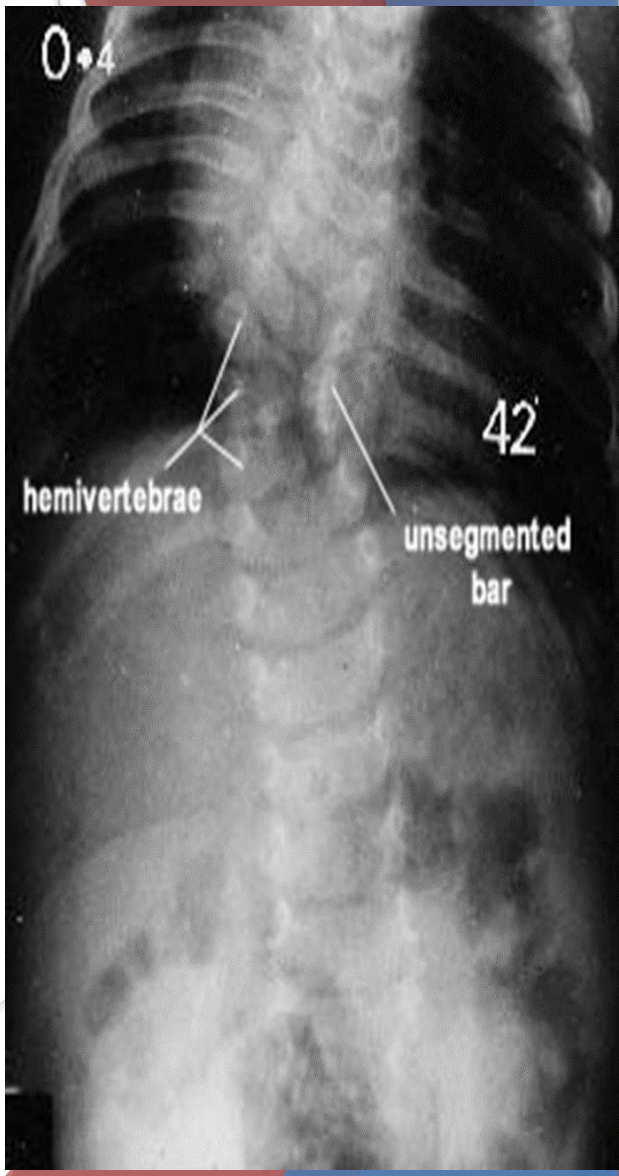
- Characteristic
  - Large curve early in life
  - Stiff curves: because of the early onset of neuromuscular deficiency resulting in limited mobility and secondary contracture.
  - Progressive curves which is greatest during rapid growth and loss of ambulation.
  - Long curves
  - Pelvic obliquity: resulted from lower extremity contracture and imbalanced spinal deformity which may impair comfortable sitting for these patients.
  - Sagittal balance deformity including thoracic and lumber hyperkyphosis or lumber hyperlordosis.

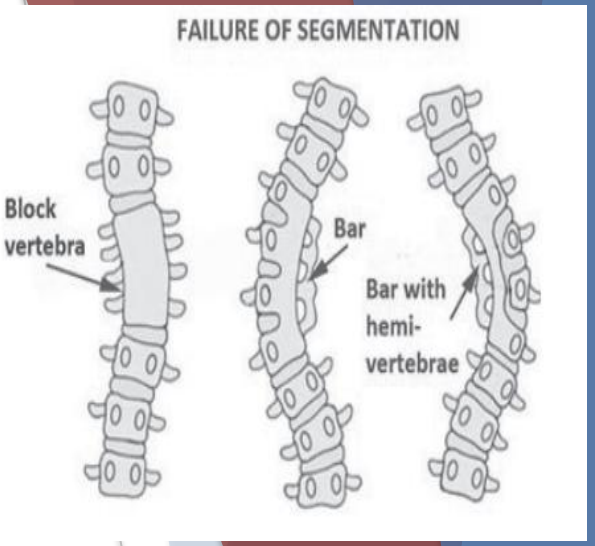
Congenital Socliosis

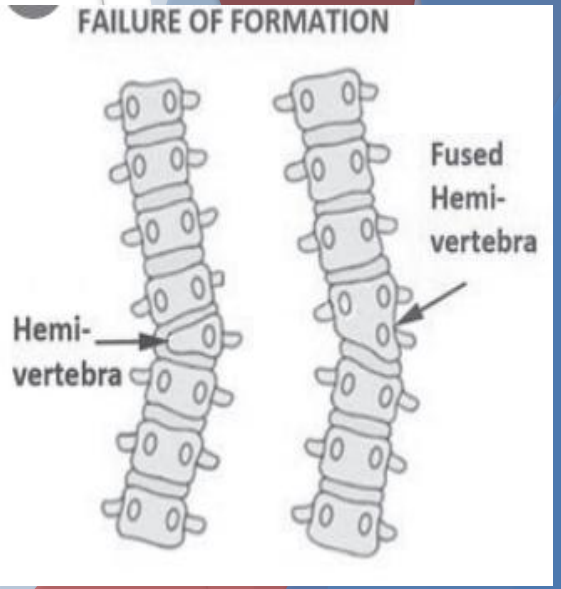
- A spinal deformity caused by vertebrae that are not properly formed
- Occurs very early in development between the 4th and 6th weeks of embryonic formation and often before the mother knows she is pregnant, and the cause is not known.
- Congenital scoliosis does not seem to run in families.
- Genetic studies to date have not yielded much evidence that this condition can be inherited.
- Although congenital scoliosis is often discovered during the infant or toddler period, in some children it does not appear until their adolescent years.
- The incidence of congenital scoliosis is 1 in 1 ,000 live births.
- Associated with systemic anomalies (61%): heart (10%), Genitourinary (25%) and spinal cord malformation (20-40%).



Congenital Socliosis

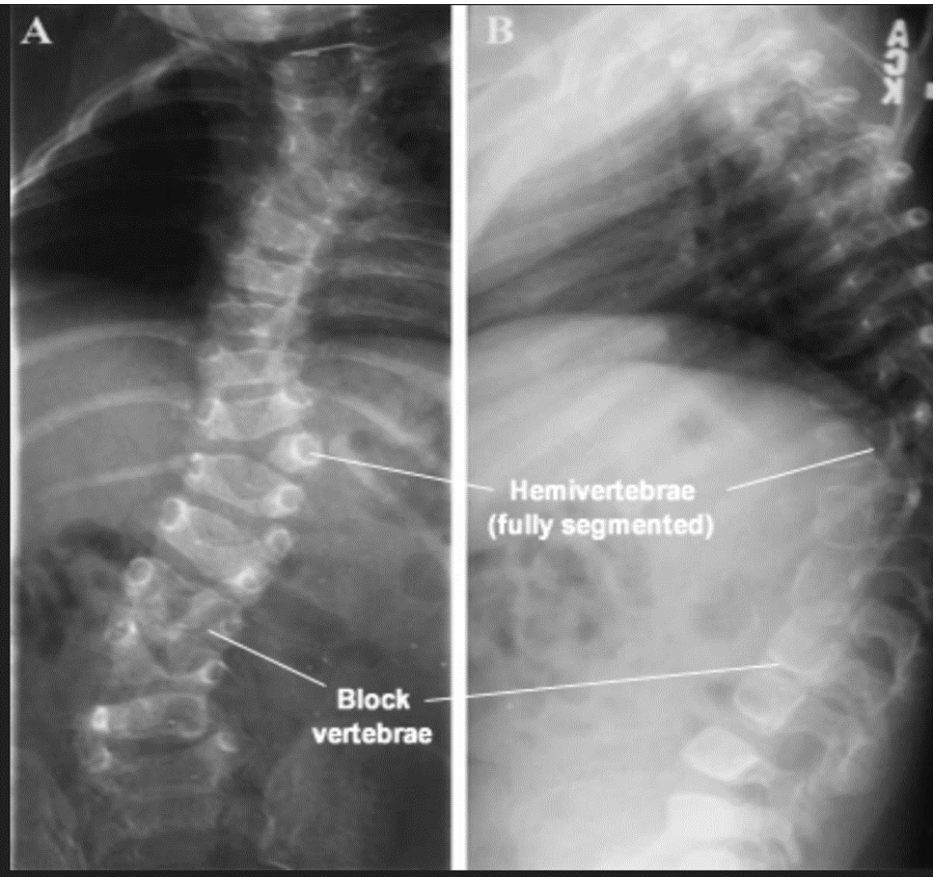
- Depending on the structure of the anomaly, the child may exhibit scoliosis (a curve to the right or left), kyphosis (rounded back), or lordosis (sway back) or combined as kyphoscoliosis.
- Anomalies are classified on the basis of failures of segmentation, formation, or both (mixed pattern).
  - “failure of segmentation.” This means that one or more vertebrae are abnormally connected together on one side. This connection will slow growth on that side of the spine
  - “Failure of formation,” the most common type of congenital problem, means that the normal shape of the vertebra is disrupted, these vertebra will look like triangles instead of rectangles. The abnormally shaped vertebra may cause a wedge in the front, back, either side, or a combination, tilting the spine at that level.

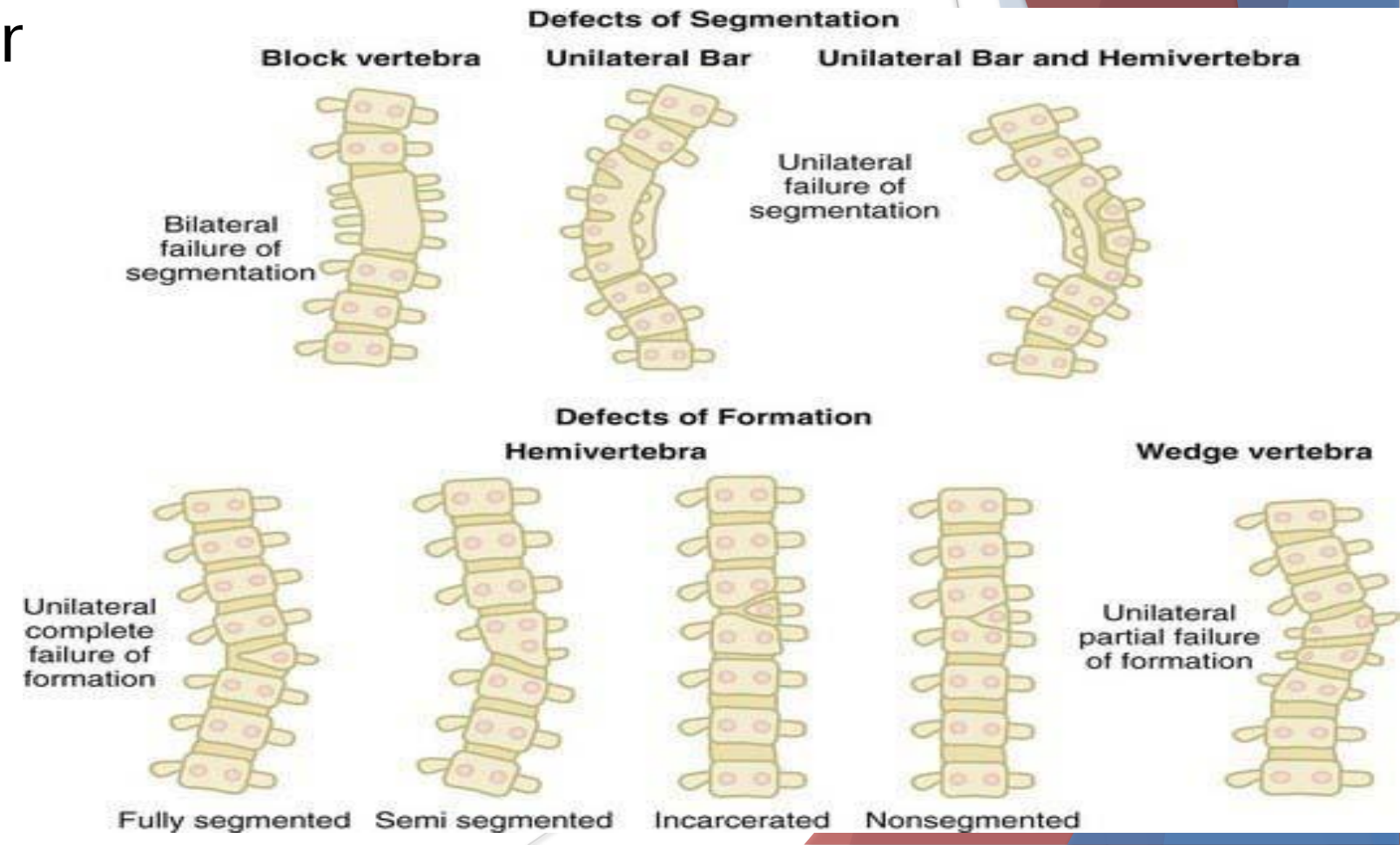




Congenital Socliosis

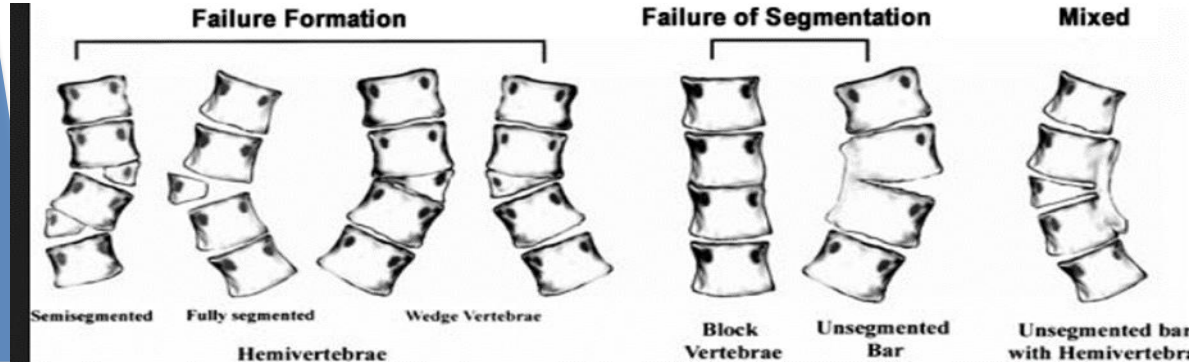
- Winters Classificatior

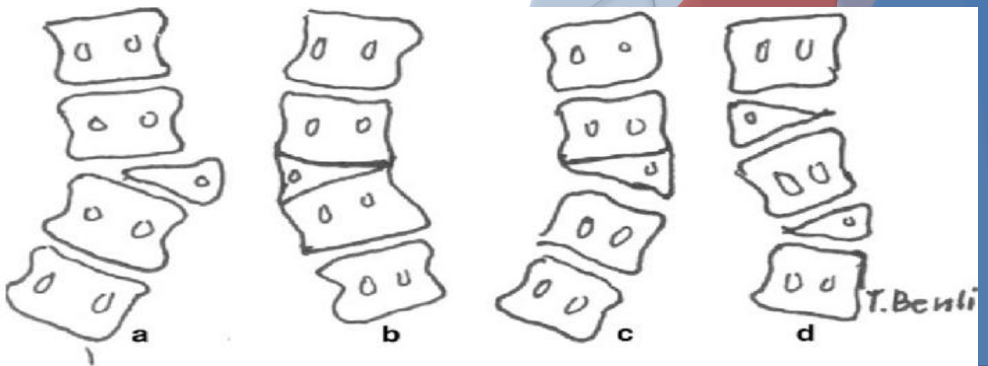




Congenital Socliosis

- Anomalies are classified on the basis of failures of segmentation, formation, or both (mixed pattern).
  - “Hemivertebra” vertebra had one pedicle, the contralateral pedicle is hypoplastic.
  - “fully segmented hemivertebra” means that there is a growth plate and a normal disk on both the top and bottom of the abnormal vertebra.
  - “Semi-segmented hemivertebra” have a normal disk and growth plate either above or below. Hemivertebra fused to adjacent vertebra on one side with disk on the other
  - “Non-segmented hemivertebra” hemivertebra are fused to the vertebra above and below.
  - “block vertebra” means that there is a missing disk space. Bilateral bony bars
  - “Bar body / unilateral unsegmented bar” is common and likely to progress
  - “Mixed / unsegmented bar with hemivertebra”
  - “Incarcerated hemivertebra” found within lateral margins of the vertebra above and below
  - “Unincarcerated hemivertebra” laterally positioned
  - “hemimethameric segmental shift” two or more hemivertebra exists on both left and right sides of the spine, where the hemivertebras are separated by at least 1 normal vertebra





Syndromatic Scoliosis

- Neurofibromatosis
- VACTERL syndrome
- VATER Syndrome
- Marfan Syndrome
- Ehlers Danlos syndrome
- Osteochondrodystrophy (dwarfism)
- Noonan Syndrome
- Angelman Syndrome
- Rett Syndrome
- bone dysplasia
- prader willi syndrome
- Klippel feil syndrome

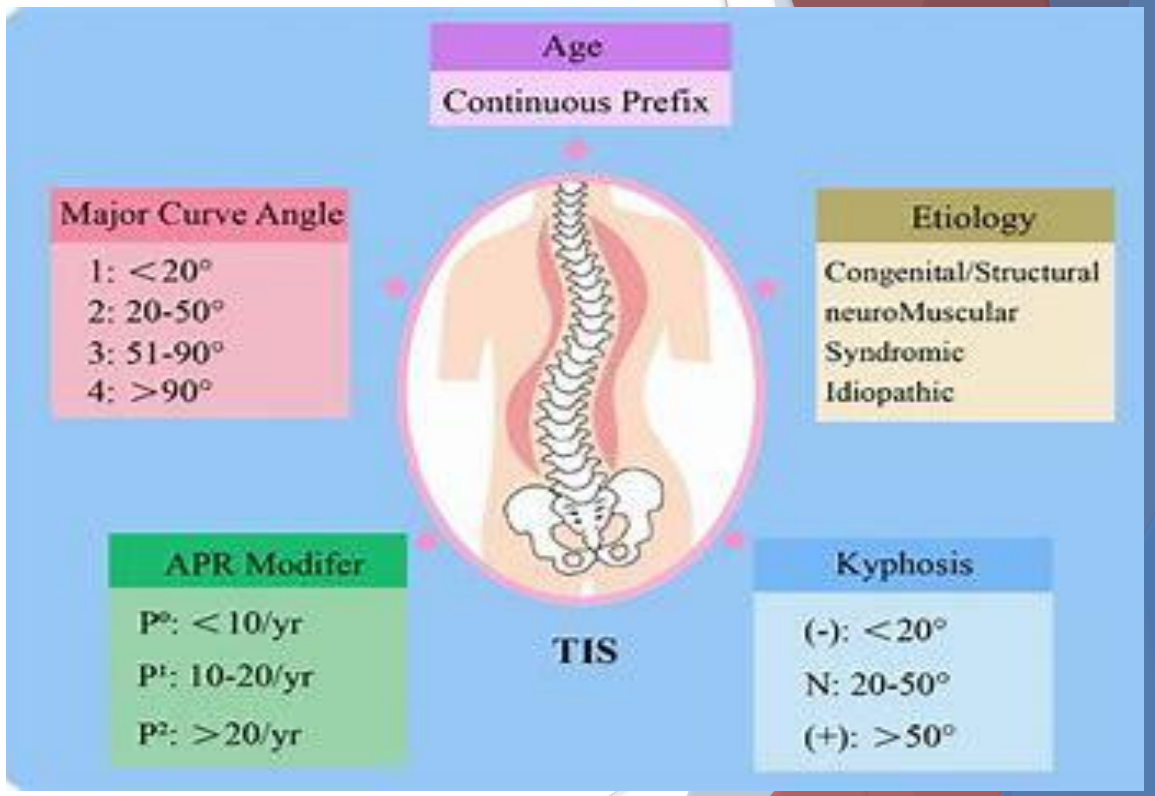
Outcomes and Mortality

- Pehrsson K. Larsson S, Oden A, Nachemson A. Longterm follow-up of patients with untreated scoliosis. A study of mortality, causes of death, and symptoms. Spine 1992;17:1091-1096 PubMed
  - Our best understanding of mortality in EOS comes from a long-term and much cited retrospective study by Pehrsson et al of 115 women in Sweden.
  - The authors report a 50% increase in mortality for patients with EOS from age 4 onward.
  - Mortality was most significantly increased for the subset of patients with a diagnosis of scoliosis before the age of 3 and was not increased in patients with an onset of scoliosis after age 10.
  - Patients with severe scoliosis (defined as curve > 70 degrees) had significantly increased mortality, whereas this was not true for patients with mild or moderate curves.

Classification

- Members of the Growing Spine Study Group (GSSG) and the Children's Spine Study Group (CSSG) collaborated to create the Classification of Early-Onset Scoliosis (C-EOS)

- APR =[(Major curve magnitude at time point 2)-(Major curve magnitude at time point 1) / (Time between tl and t2 in years)

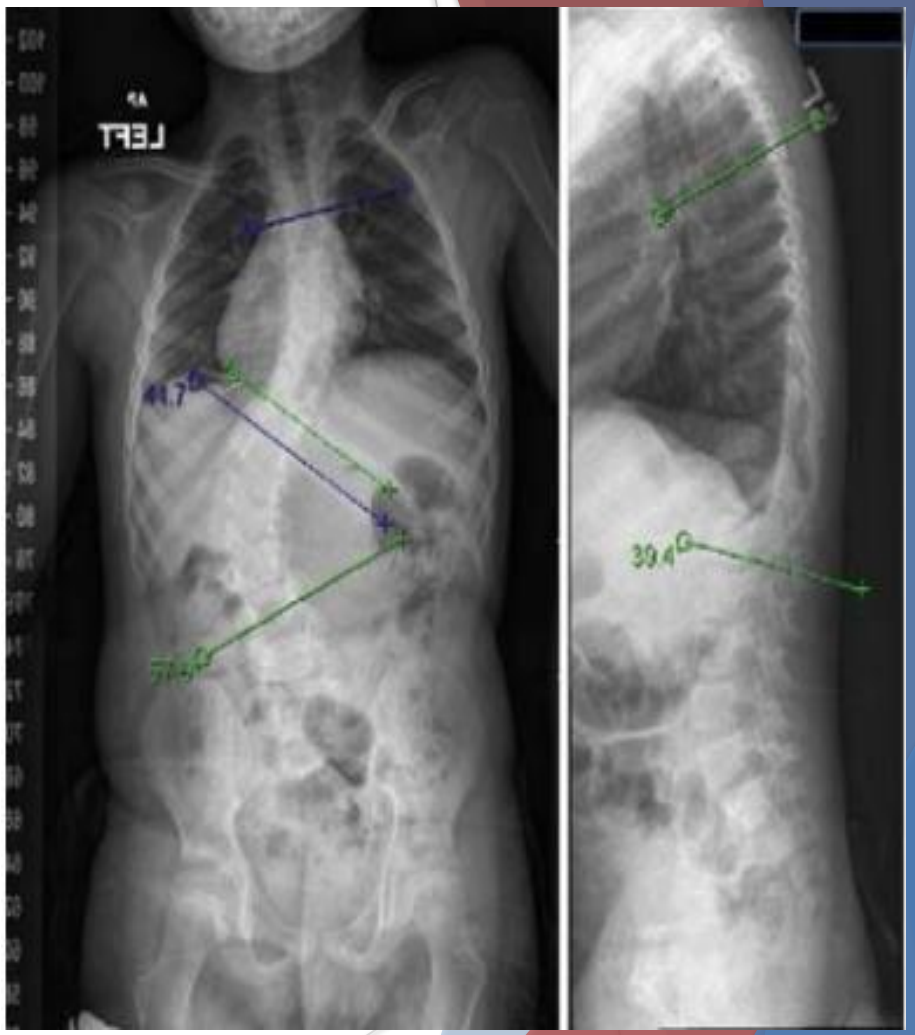


Classification

- Case example 1
- Posteroanterior (PA) and lateral radiographs of a 4-year-old patient with spinal muscular atrophy (SMA).
- The magnitude of the major curve in the coronal plane is 60 degrees.
- The maximum regional kyphosis is 35 degrees.
- The major coronal curve was 35 degrees 6 months ago.
- The annual progression ratio = (60 degrees-35 degrees/6 months)= (25 degrees/6 months)= (50 degrees/year).
- The final C-EOS classification Is 4M3NP2.

Classification

- Case example 2.
- PA and lateral radiographs of a 5-year-old patient with Angelman syndrome.
- The magnitude of the major curve in the coronal plane is 57 degrees.
- The maximum regional kyphosis is 39 degrees.
- The major coronal curve was 50 degrees 6 months ago.
- The annual progression ratio = (57 degrees - 50 degrees/6 months)= (7 degrees/ 6 months) = (14 degrees/year).
- The final C-EOS classffication Is 5S3NP 1.



Goals of Management

- Regulating progression of spinal deformity
- Maximizing thoracic volume
- Optimizing function and health-related quality of life
- Minimizing complications and negative effects of treatment

Non Surgical Management

- Observation:
  - Monitoring behaviour of the curve
  - Serial screening
    - clinical and radiological
    - 4-6 months interval
  - Whether static/resolving/progressing
  - Intervene if documented progression

Observation of EOS

4yrs Dec 2013

6yrs Apr 2015

10 yrs Apr 2019

Favourable Natural Course of Idiopathic EOS

Non Surgical Management

- Bracing/Casting
  - To allow spinal growth
  - Minimize curve progression
  - Buy time/ delay surgery
  - Avoid surgery in idiopathic infantile curves
  - Rarely effective in congenital scoliosis
  - Can avoid secondary curve development

Non Surgical Management

- Bracing

S Afr J Physiother. 2021; 77(2): 1573.

Published online 2021 Oct 26. doi: 10.4102/sajp.v77i2.1573

Brace treatment for patients with scoliosis: State of the art

Hans-Rudolf Weiss,[1] Tuğba Kuru Çolak,[2] Manuel Lay,[3] and Maksym Borysov[4]

Figure 5 – *The Wilmington brace is a custom-molded thora-columbosacralorthosis that has molds to push and correct the curve.*

Figure 4 – *The Kalabas brace has several straps that are applied over the shoulder and bend the child in the opposite direction of the curve*

Figure 6 – *Boston brace or thoracolumbar sacral orthosis (TLSO)*

Figure 7 – *The Milwaukee brace, with an extension to the chin, is able to control curves in the top part of the spine.*

Non Surgical Management

- Casting
  - Effective in small kids(uncooperative child)
  - Threshold to initiate cast varies with institute
  - >25degree/documented progression of 10 degree
  - Under GA…>flexibility curve
  - Parents education of cast care(Bathing)
  - Frequent changing Q2-4M, minimum 5 casts

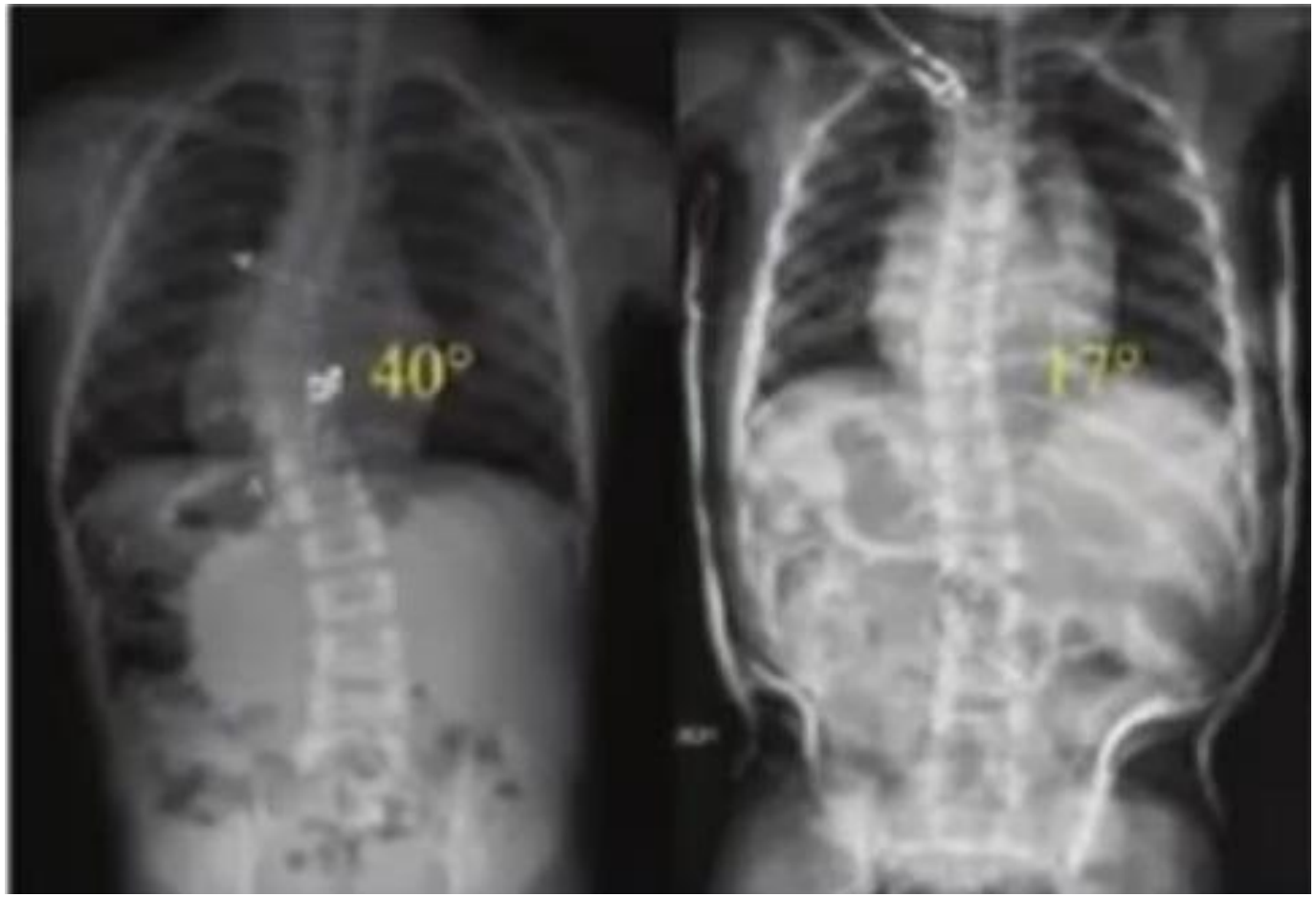
J Clin Orthop Trauma. 2020 Sep-Oct; 11(5): 810–815. PMCID: PMC74522
Published online 2020 Jul 8. doi: 10.1016/j.jcot.2020.06.034 PMID: 32879

Evolution of casting techniques in early-onset and congenital scoliosis

Rajat Mahajan,[a,*] Shyam Kishan,[b] Abhinandan Reddy Mallepally,[a] Cody Shafer,[b] Nandan Marathe,[a] and Harvinder Singh Chhabra[a]

Casting EOS

Casting/Brace EOS



Non Surgical Management

- Traction
- Preoperative halo-pelvic traction achieved significant improvements in spinal deformity and pulmonary functions, with minor and curable complications. Thus, it is an effective and safe solution before surgery and may be the optimal choice for severe scoliosis

Review > Eur Spine J. 2023 Mar;32(3):874-882. doi: 10.1007/s00586-023-07525-7. Epub 2023 Jan 9.

Halo-pelvic traction in the treatment of severe scoliosis: a meta-analysis

Yan Sun [1], Yong Zhang [1], Haoning Ma [2], Mingsheng Tan [3], Zhihai Zhang [4]

Affiliations + expand

PMID: 36622456 DOI: 10.1007/s00586-023-07525-7

Non Surgical Management

- Exercise Treatment
  - There are the following Physiotherapeutic scoliosis-specific exercise (PSSE) physiotherapy schools in Europe: Schroth, SEAS, BSPTS, FED, FITS, Lyon, Side Shift, and DoboMed
  - The methodologies of these schools are similar, in that they focus on applying corrective exercises in three planes, developing stability and balance, breathing exercises, and posture awareness
  - Schroth is the most widely studied and has been proven to be effective.

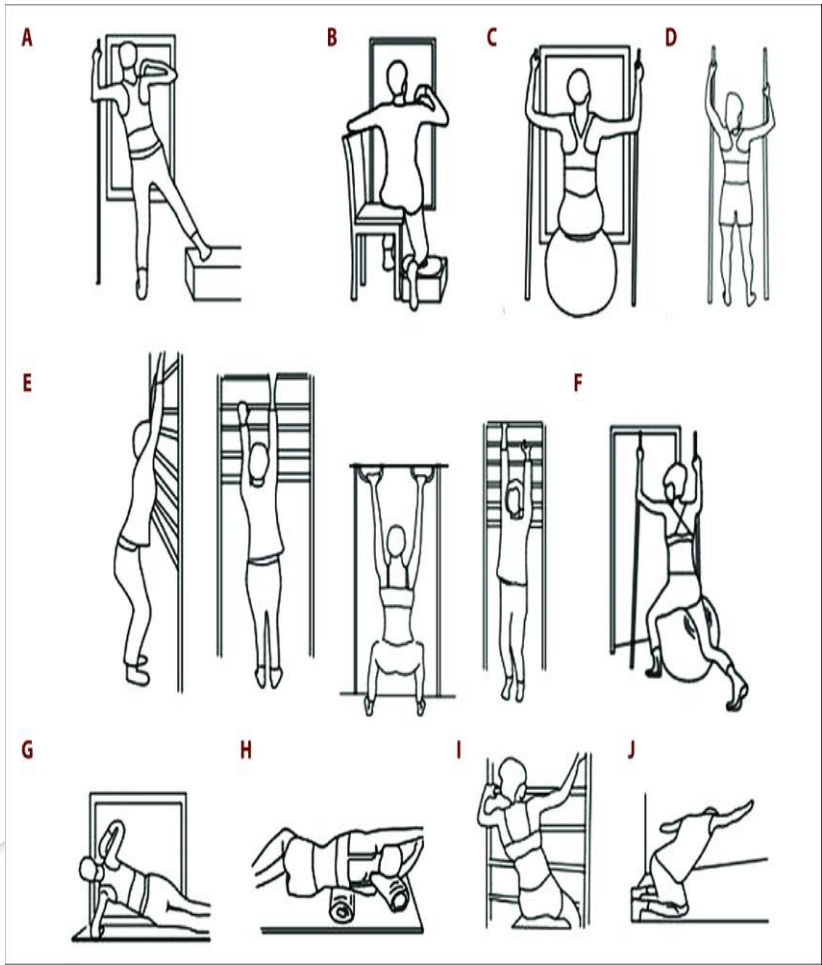
Review > Int J Environ Res Public Health. 2022 Jul 28;19(15):9240. doi: 10.3390/ijerph19159240.

Physiotherapeutic Scoliosis-Specific Exercise Methodologies Used for Conservative Treatment of Adolescent Idiopathic Scoliosis, and Their Effectiveness: An Extended Literature Review of Current Research and Practice

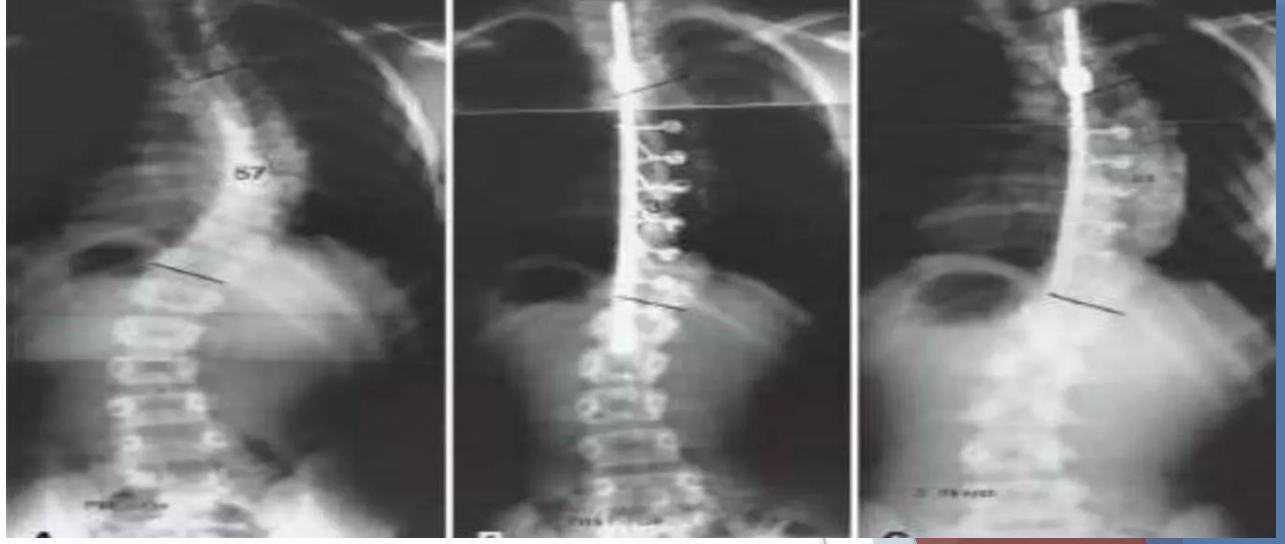
Vaiva Seleviciene [1 2], Aiste Cesnaviciute [3], Birute Strukcinskiene [4], Ludmiła Marcinowicz [5], Neringa Strazdiene [6], Agnieszka Genowska [7]

Affiliations + expand

PMID: 35954620 PMCID: PMC9368145 DOI: 10.3390/ijerph19159240



Historical Surgeries



- **<u>Early Spinal Fusion</u>**
  - **Crank Shaft Phenomenon**
  - **Disaster with Restrictive Lung Disease (Goldberg et al)**
    - Fusion at a mean age of 4.1 years, drop of pulmonary function to below 41%FVC.

Exception for Early Spinal Fusion

Hemivertebra

(unpredictable,prog,rot,tis,crank)

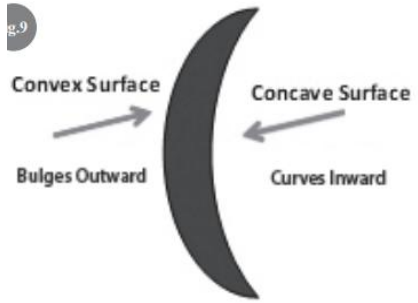
Smaller curve (40)less number of vertebra

early fusion (+/- hemivertebra excision)

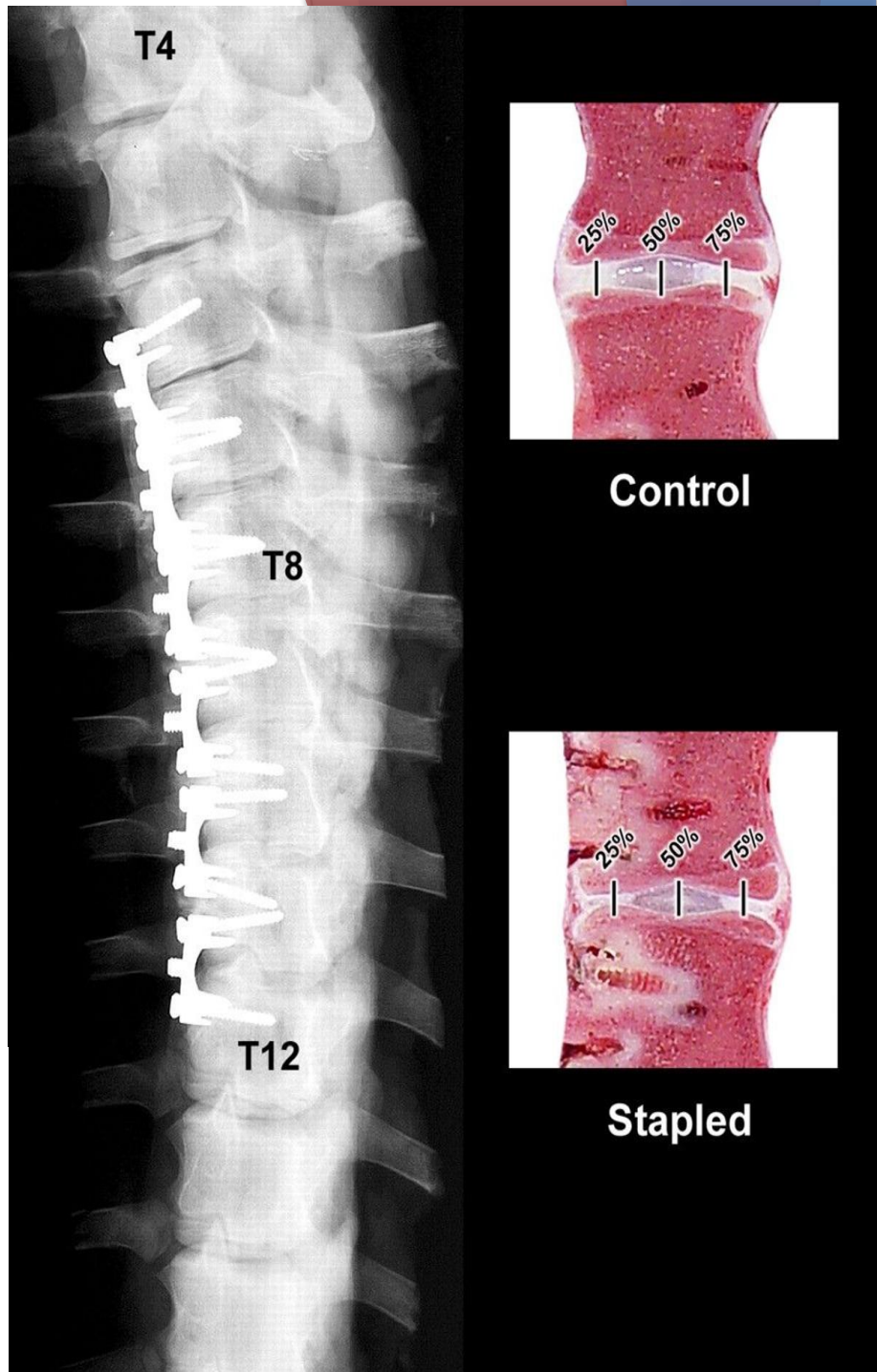
Surgical Managment

- Failure of cast/brace to control curve
- Curve not amenable for cast/bracing
  - Malignant curve
- The dilemma faced by the surgeon is how to stop the progression of a curve without adversely affecting future growth.
- Harrington first introduced instrumentation without fusion in 27 post-polio and idiopathic patients (rod era 1960s)

Surgical Managment



- Hemi-epiphysiodesis
  - If the growth centers are removed and spinal fusion is performed on the convex side, the concave side might continue to grow, possibly improving the curve.
  - As noted, these procedures can be unpredictable in young children with abnormal vertebrae in their back….hemivertebra
- good curve correction while maintaining the growth of thorax



SPINE

Convex Instrumented Hemiepiphysiodesis With Concave Distraction

A Treatment Option for Long Sweeping Congenital Curves

Demirkiran, Gokhan MD; Dede, Ozgur MD; Ayvaz, Mehmet MD; Bas, Can E. MD; Alanay, Ahmet MD; Yazici, Muharrem MD

Author Information

Journal of Pediatric Orthopaedics 36(3):p 226–231, April/May 2016. | *DOI:* 10.1097/BPO.0000000000000441

Surgical Managment

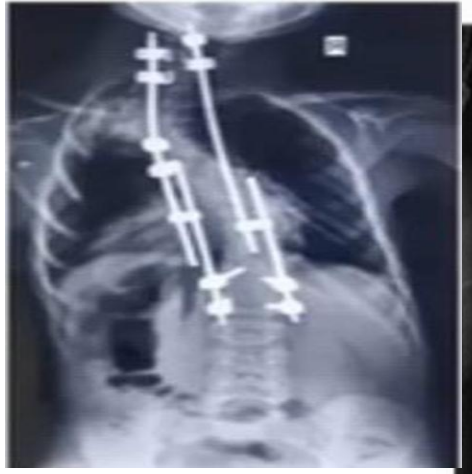
Multicenter Study > Spine Deform. 2021 May;9(3):851-858. doi: 10.1007/s43390-020-00270-7. Epub 2021 Feb 8.

Growth-friendly surgery results in more growth but a higher complication rate and unplanned returns to the operating room compared to single fusion in neuromuscular early-onset scoliosis: a multicenter retrospective cohort study

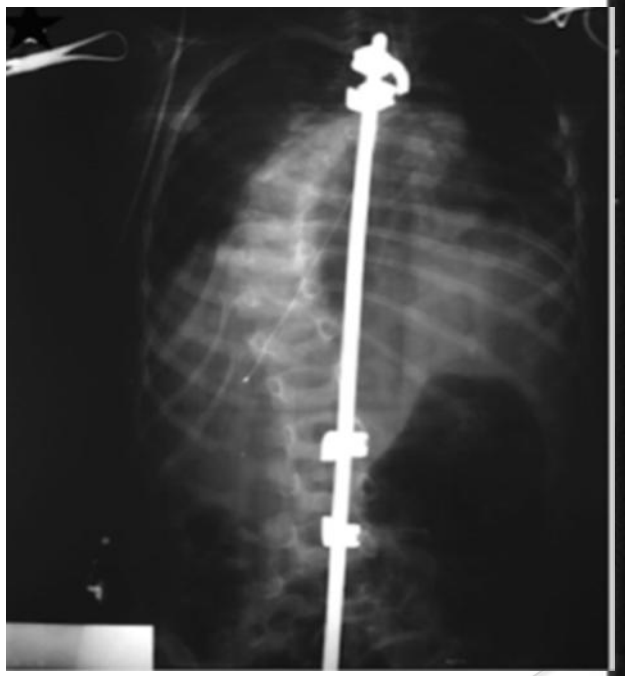
Ying Li [1], Jennylee Swallow [2], Joel Gagnier [3], Patrick J Cahill [4], Paul D Sponseller [5], Sumeet Garg [6], George H Thompson [7], Brandon A Ramo [8]; Pediatric Spine Study Group

- Growth Friendly Surgery
  - avoid severe respiratory problems, allow spinal growth
- Growth friendly surgery patients achieved more spinal growth but eight times more complications and nine times more return to operation room
  - Distraction Based
  - Guided Growth
  - Compression-Based

Surgical Managment



- Growth Friendly Surgery-Distraction Based
  - Traditional Growing rods (tGR)
    - Proximal and distal solid foundation
    - Growth of segment between anchors
    - Serial distraction
    - Dual rod (more stable)
    - Repeated surgeries
    - Psychological stress
    - Not for congenital scoliosis
    - Not for complete growth patient
    - not for chest wall abnormalities or thoracic insufficiency syndrome

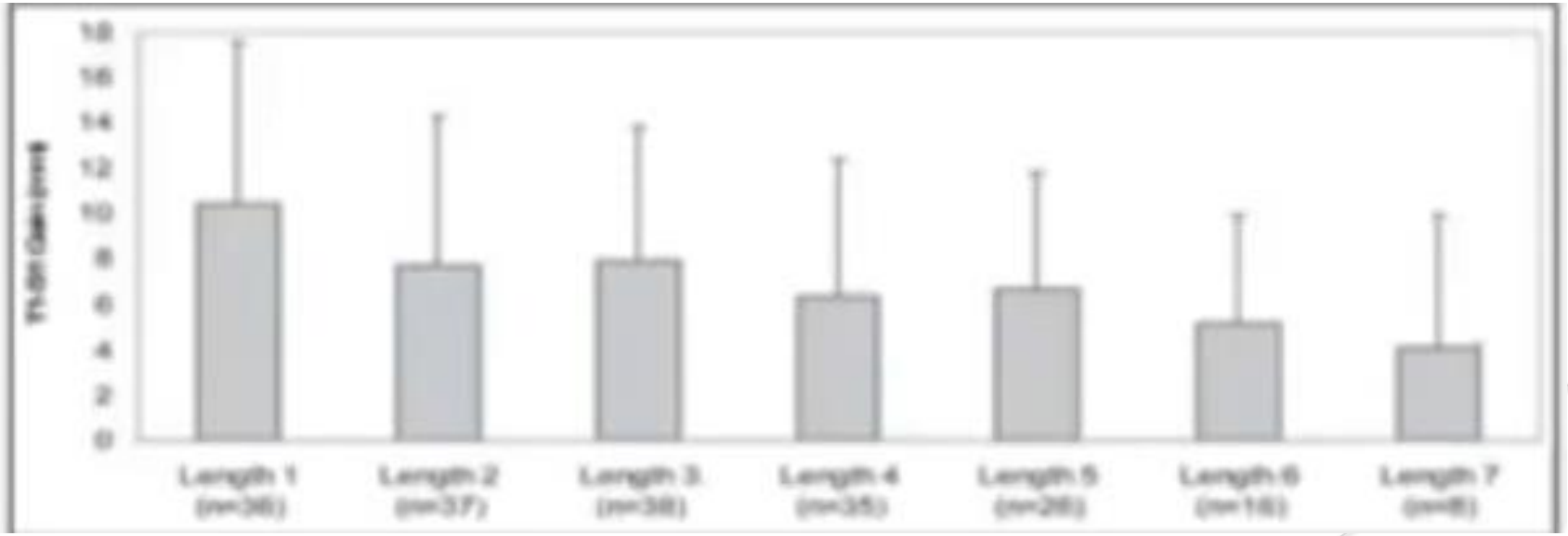


Surgical Managment

- Growth Friendly Surgery-Distraction Based
  - The curve can usually be corrected by fifty percent at the time of the first operation.
  - After the rods are implanted, patients are prescribed a special brace to wear for several months.
  - The child then returns every six months to have the rods “lengthened” by minor procedure(OP through small incision) until the spine is closer to maturity.
  - Most commonly, complications are related to spontaneous fusion, surgical site problems, or mechanical failure (proximal junctional kyphosis, implant loosening/pullout, metal debris, hook dislodgement, rod breakage)

Law of Diminishing Returns (Sankar et al)

- The number of lengthening episodes that a traditional growth rod (TGR) can undergo before stiffness across the construct prevents further increases in overall length
- Progressively less increase of T1-S1 length over time



Surgical Managment

- Growth Friendly Surgery-Distraction Based
  - Vertical Expandable Prosthetic Titanium Rib (VEPTR),
  - Rib based
  - Treatment of TIS.
  - Rib/pelvis +/- spine anchors
  - Thoracic expansion/growth
  - Serial distractions
  - Surgical morbidity
  - Spinal growth with serial expansions (mean 71 mm over 4 lengthenings)
  - A recent meta-analysis has shown that VEPTR was inferior in comparison to other correction techniques in terms of Cobb angle andT1-S1 height rates at all time points
  - VEPTR is associated with higher complication rate compared to TGR including infection, implant failure, and pneumothorax

Ann Transl Med. 2020 Jan; 8(2): 25. PMCID: PMC6995913
doi: 10.21037/atm.2019.09.158 PMID: 32055616

Long term outcome of vertical expandable prosthetic titanium rib treatment in children with early onset scoliosis

Daniel Studer and Carol-Claudius Hasler

Surgical Managment

- Growth Friendly Surgery-Distracti Based
  - Magnetically Controlled Growing Rods (MCGR)
    - Initial implantation under GA
    - • Serial non-invasive distractions
    - • Telescoping rod/ magnet/External remote device
    - • Dual rod > single rod
    - • Expensive

Early Experience of MCGR ORIGINAL ARTICLE

Eur Spine J
DOI 10.1007/s00586-015-3163-0

ORIGINAL ARTICLE

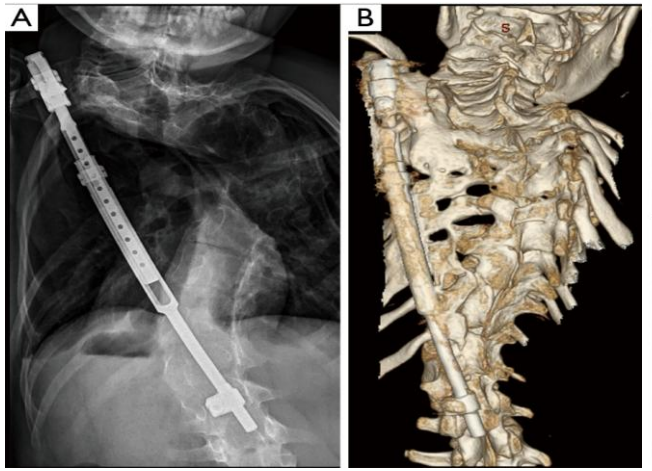
Early experience of MAGEC magnetic growing rods in the treatment of early onset scoliosis

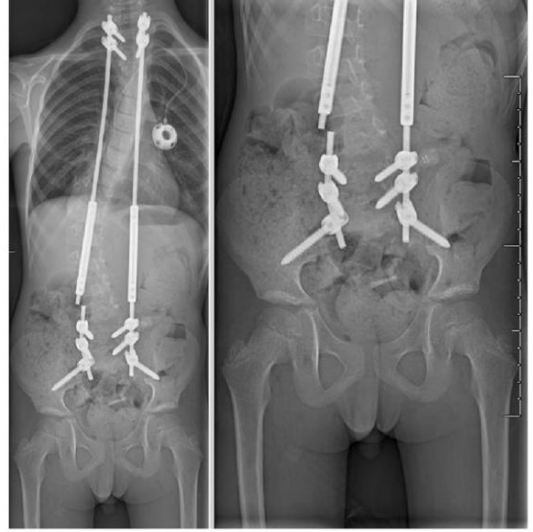
B. A. Hickey · C. Towriss · G. Baxter · S. Yarsu · S. James · A. Jones · J. Howes · P. Davies · S. Ahuja

AT 2 YEARS Appears Promising/Attractive alternative to CONVENTIONAL GROWTH ROD

Conclusion MAGEC growing rod system effectively controls early onset scoliosis when used as either a primary or revision procedure. Although implant-related complications are not uncommon, the avoidance of multiple surgeries following implantation is beneficial compared with traditional growing rod systems.

Surgical Managment





- Growth Friendly Surgery
  - The most frequent complications were categorized as implant (54%), general (17%), wound (15%) and alignment (12%)
  - About 54% of patients required unplanned surgeries due to complications, which comprised 15% of all surgeries.

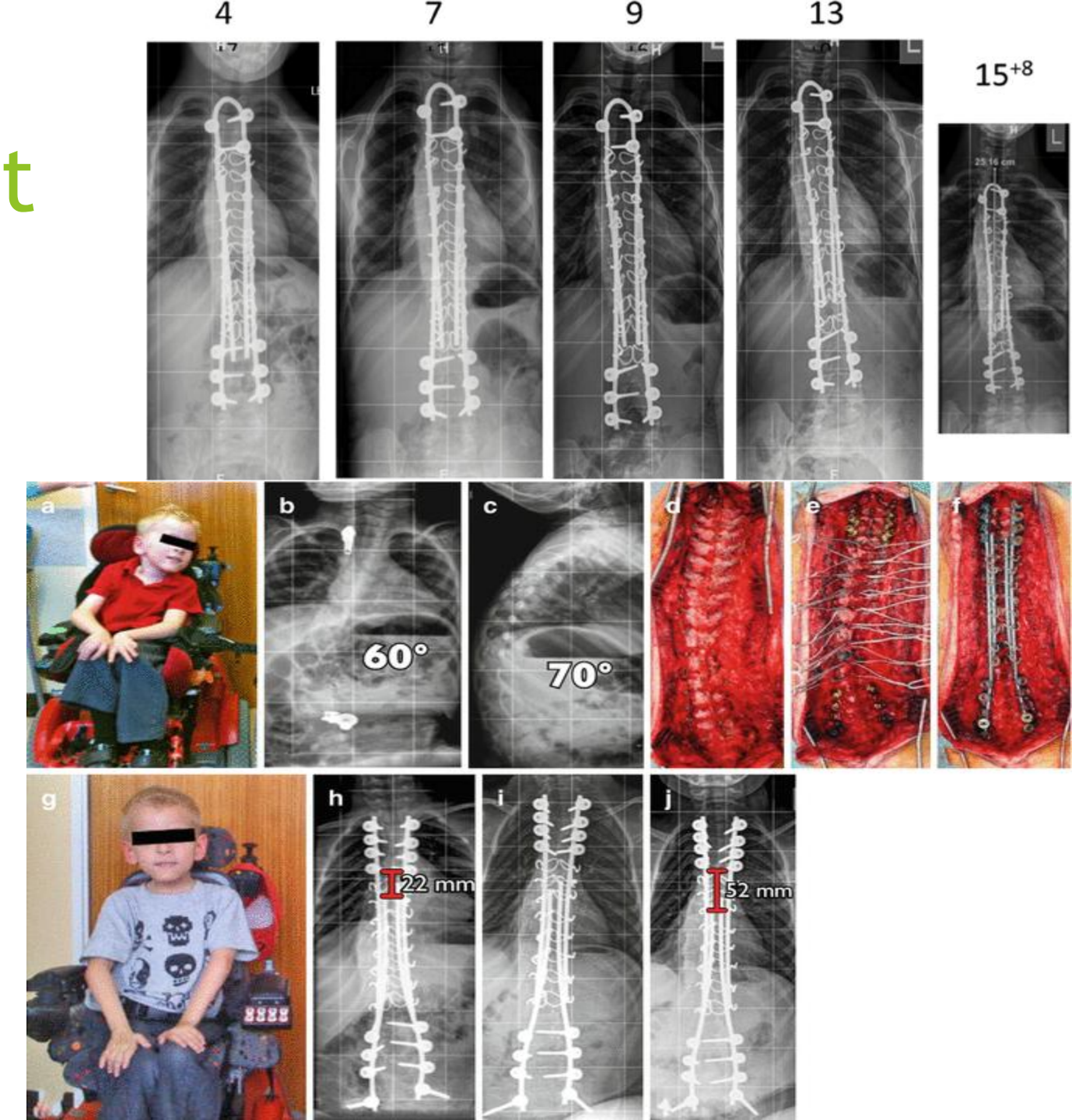
World J Orthop. 2021 Aug 18; 12(8): 584–603.
PMCID: PMC8384615
Published online 2021 Aug 18. doi: 10.5312/wjo.v12.i8.584
PMID: 34485105

Complications in growth-friendly spinal surgeries for early-onset scoliosis: Literature review

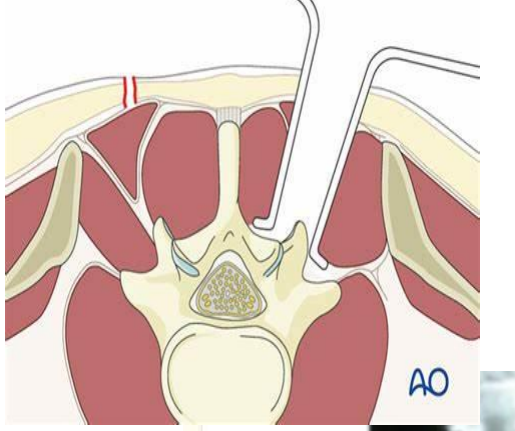
Michał Latalski, Marek Fatyga, Ireneusz Sowa, Magdalena Wojciak, Grzegorz Starobrat, and Anna Danielewicz

Surgical Managment

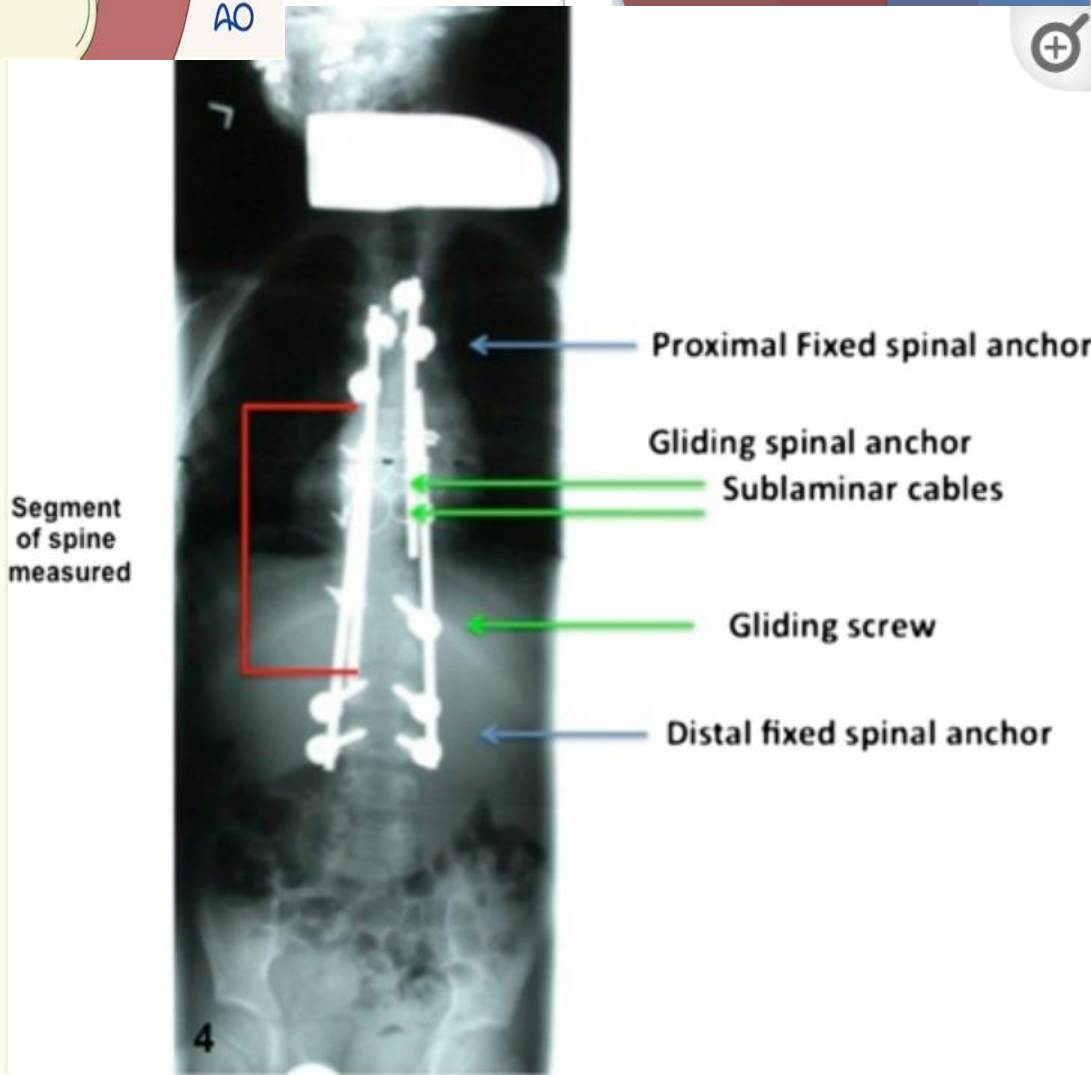
- Growth Friendly Surgery- Guided Growth
  - L-shaped rod
  - Sublaminar wires
  - Segmental fixation
  - Wires slid along rods
  - Spontaneous fusion
  - Limited spinal growth



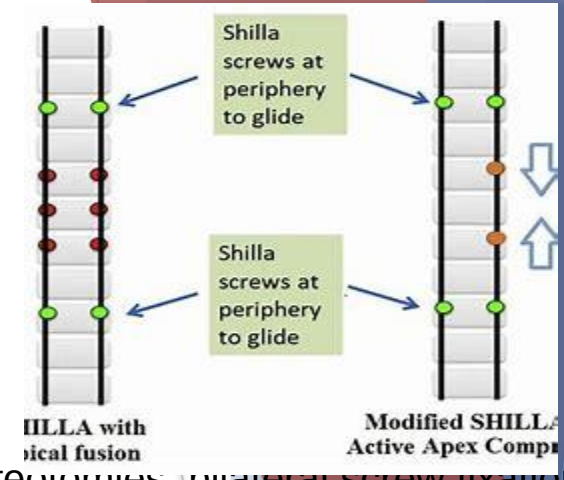
Surgical Managment



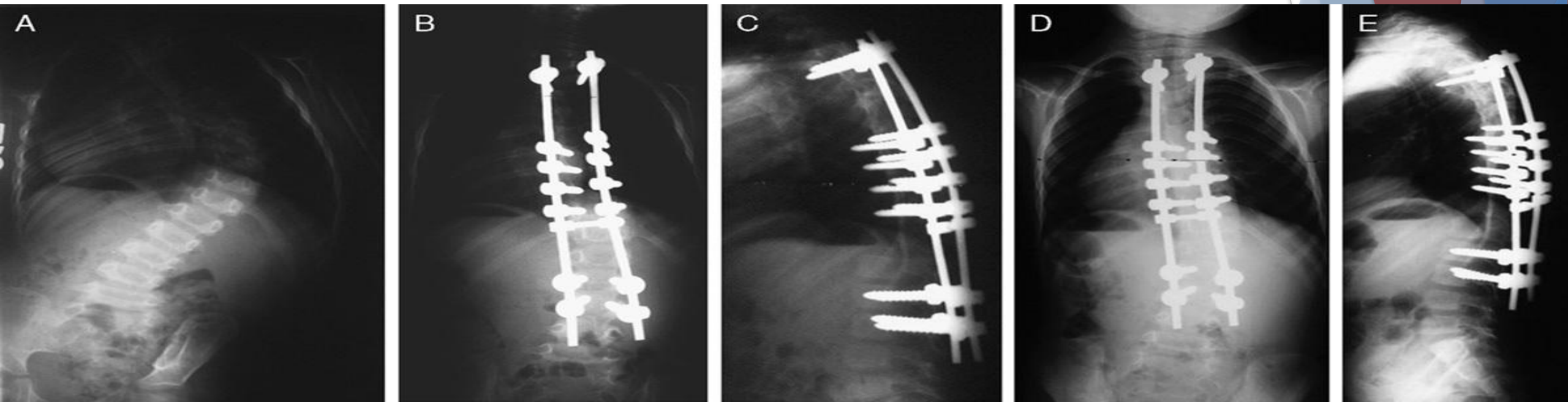
- Growth Friendly Surgery-Guided Growth
  - Modern Luque Trolley technique



Surgical Managment

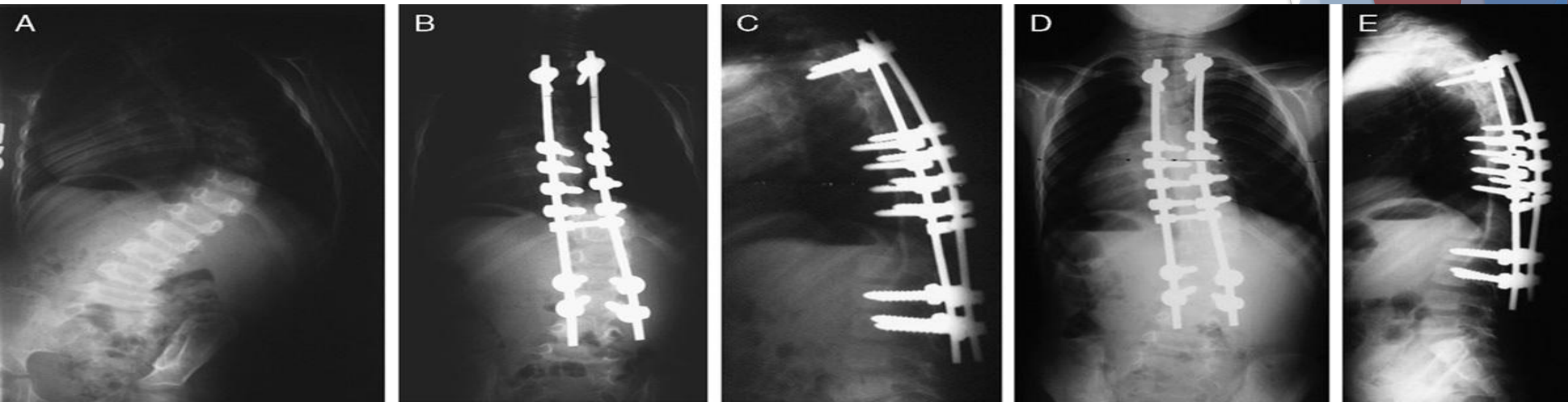


- Growth Friendly Surgery-Guided Growth
  - The center of the deformity (apex) is corrected in all planes (coronal, sagittal, and axial) with osteotomies, bilateral screw fixation, and corrective maneuvers.
  - Dual rods are fixed to the corrected apex with fusion of the apical segments.
  - Transmuscular sliding pedicle screws are placed above and below the apex without disrupting the periosteum;
  - The Shilla screw cap (which tightens to the screw and not to the rod) then allows vertebral growth in a cephalad and caudad direction.



Surgical Managment

- Growth Friendly Surgery-Guided Growth
  - Kim et al. found that Shilla obtained better initial coronal correction with similar correction rates than TGR and MCGR at final follow up. The overall rate of infection was lower for Shilla than VEPTR and TGR.
  - T1-S1 length gain was lower than TGR. Interestingly,
  - Baliog˘lu et al. evaluated the effect of complications on spinal growth and deformity correction. They found that the lordosis angle and T1–S1 length were significantly lower in the early and final postoperative controls of those who developed complications compared to those who did not



Surgical Managment

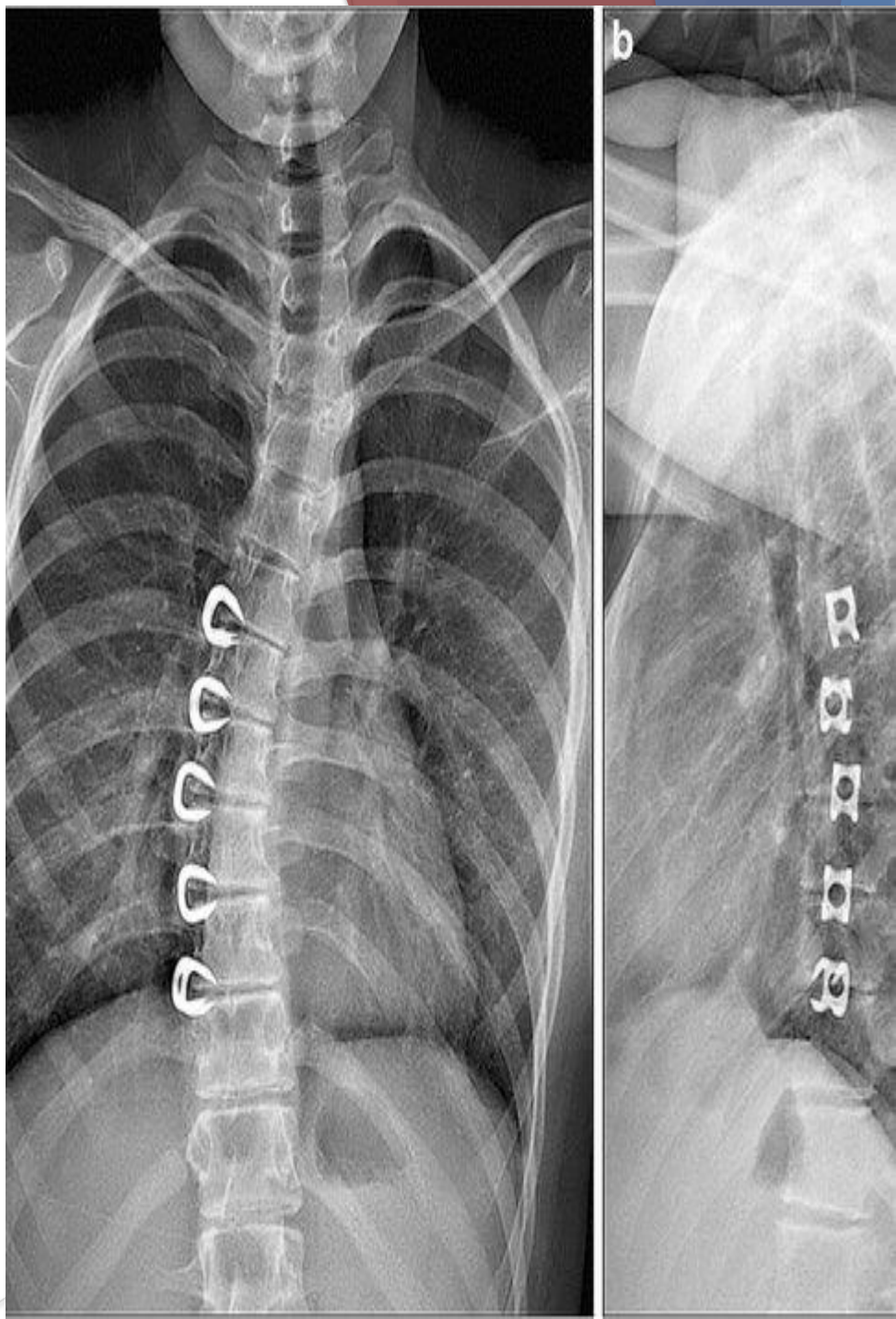
- Growth Friendly Surgery-Guided Growth
  - sliding H bar construct

Compression Based Techniques

- Stop growth on convex side
- Allow growth on concave side
- Hemiepiphysiodesis
- No Fusion

Surgical Management

- Growth Friendly Surgery-Compression-Based
  - Growth inhibition on the convex side of the curve.
  - less than 35 degrees, and who are able to tolerate open or endoscopic exposure of the spine.
  - Overcorrection Ideal for children with limited growth remaining



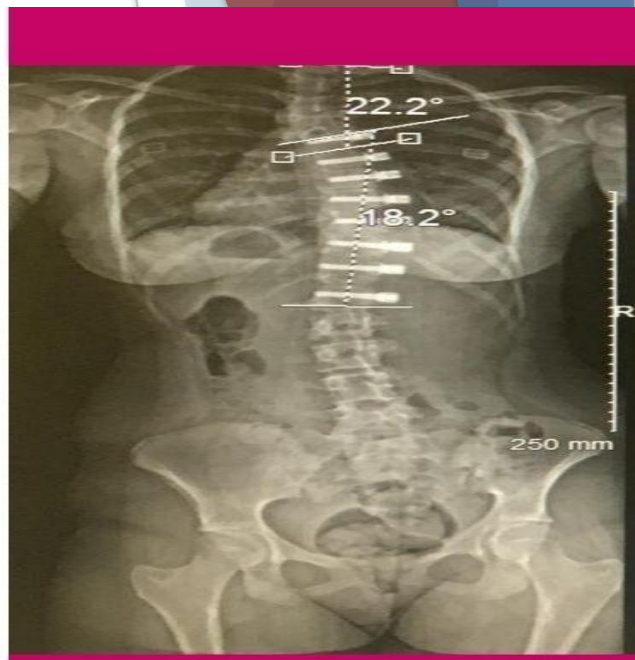
Surgical Management

- Growth Friendly Surgery- Compression-Based

Before VBT, Nov. 2016

1st X-Ray After VBT, May 2017

2nd X-Ray After VBT, Dec. 2017



3rd X-Ray After VBT, June 2018

Thank You