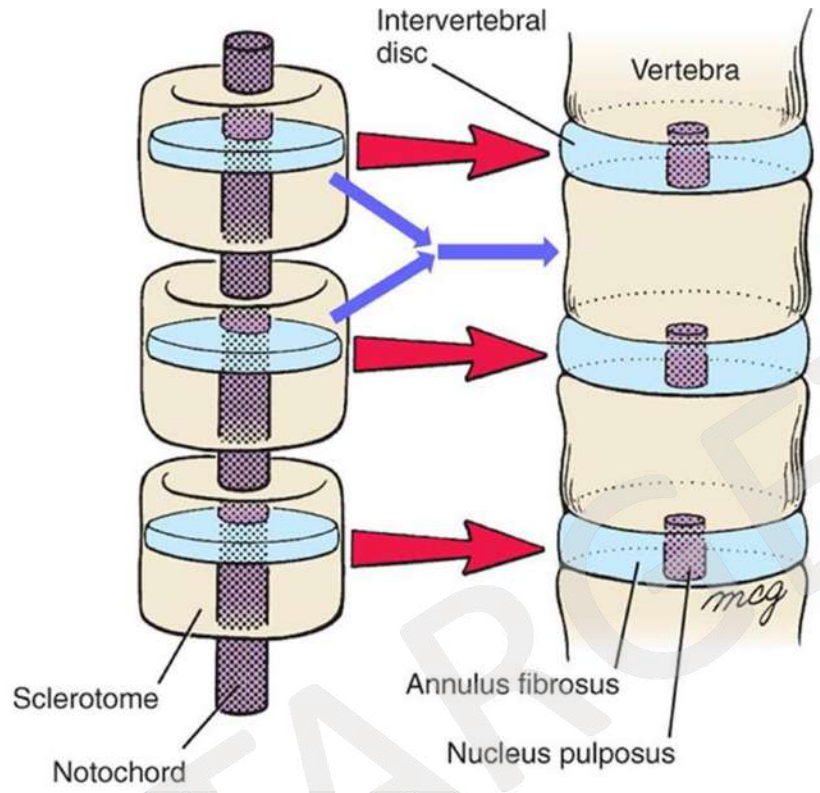
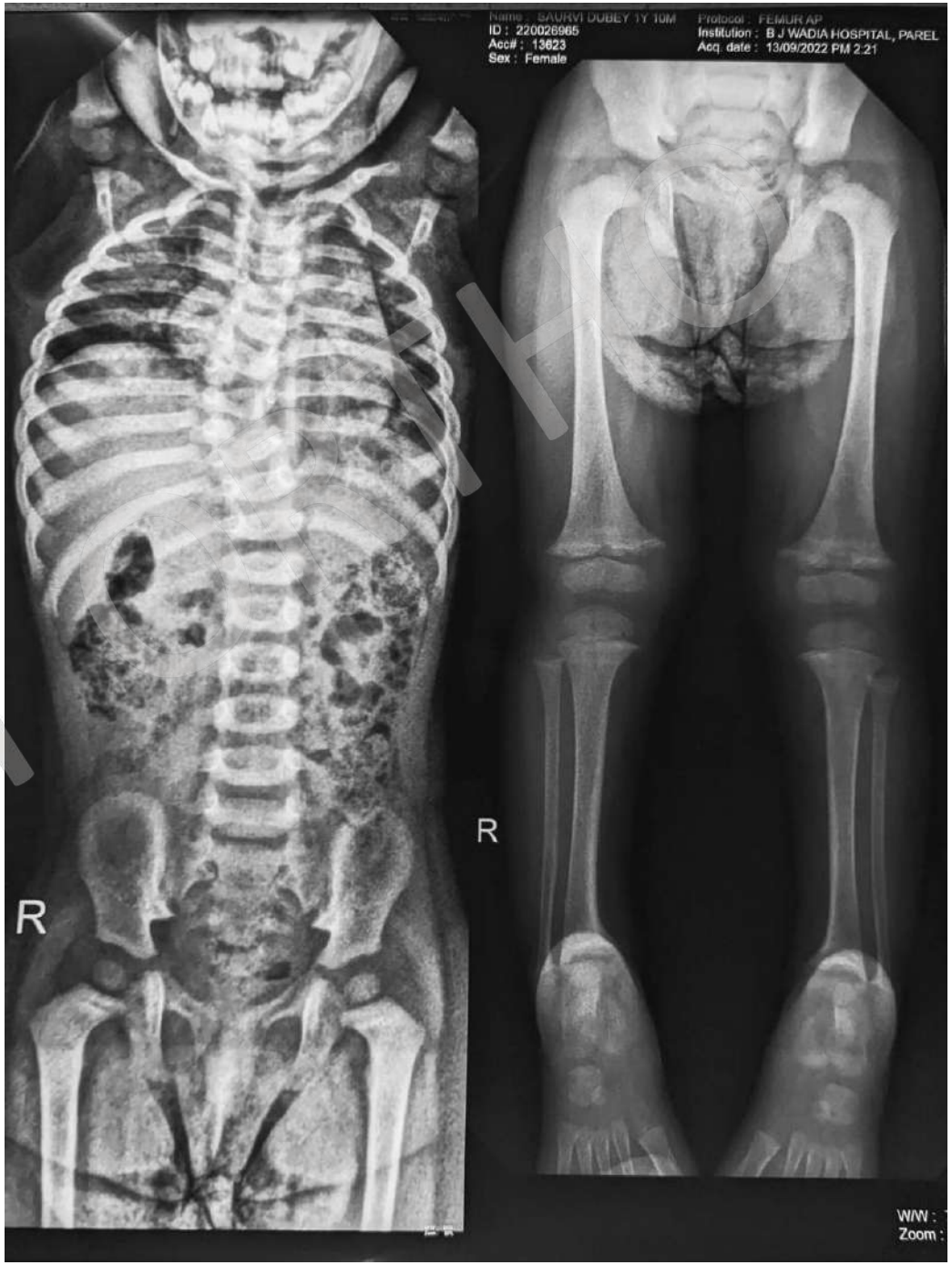


Congenital and Early onset scoliosis

Shalin Shah

Fellow in Pediatric Orthopaedics

M.S. Ortho, DNB Ortho



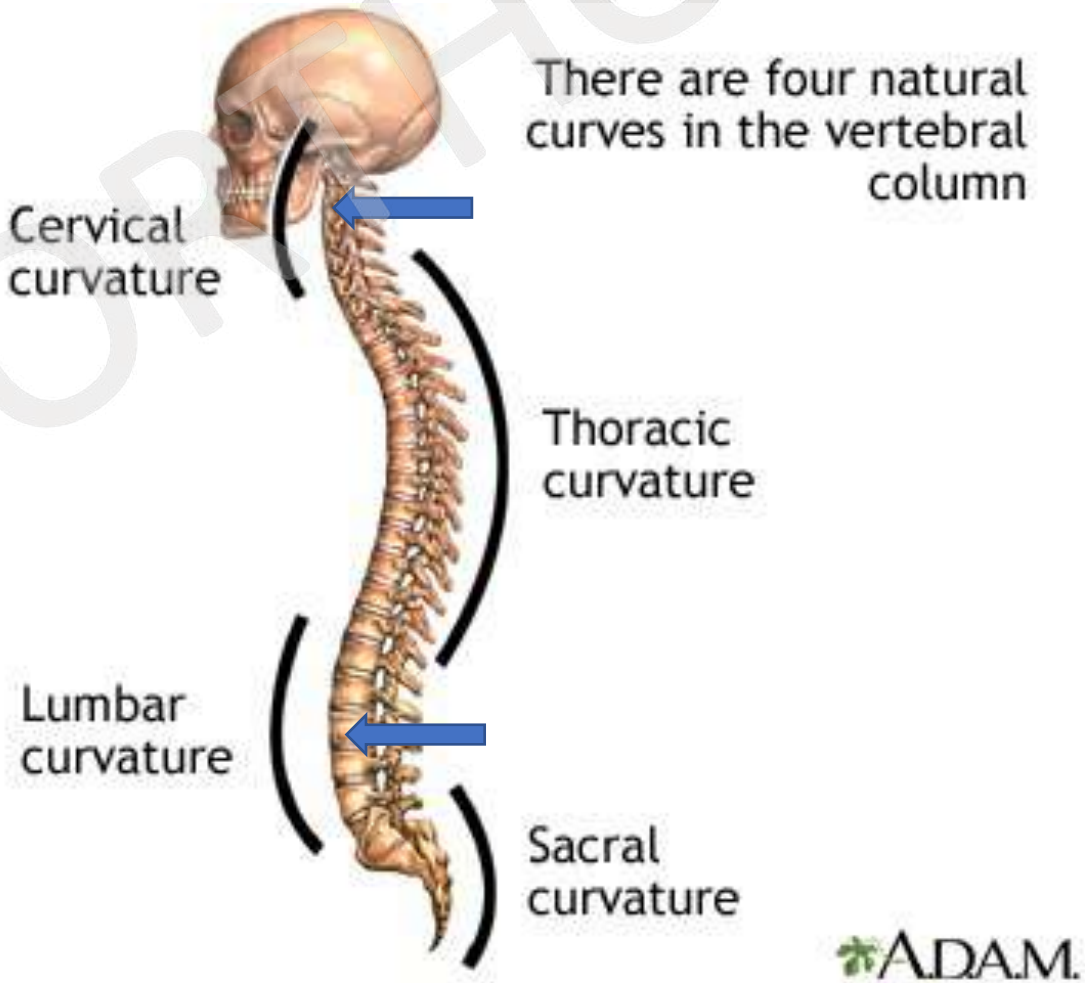
Orthopedics-

Making the child straight

Opportunities in EOS

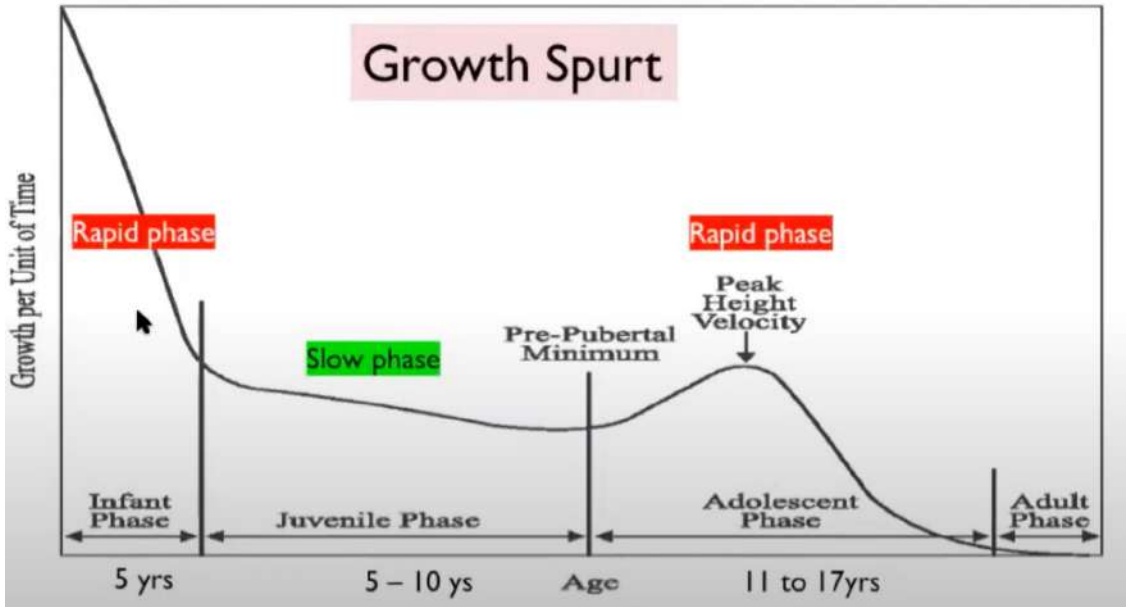
Normal Spine Curvatures

- Cervical lordosis develops with head holding
- Lumbar lordosis develops with sitting and walking



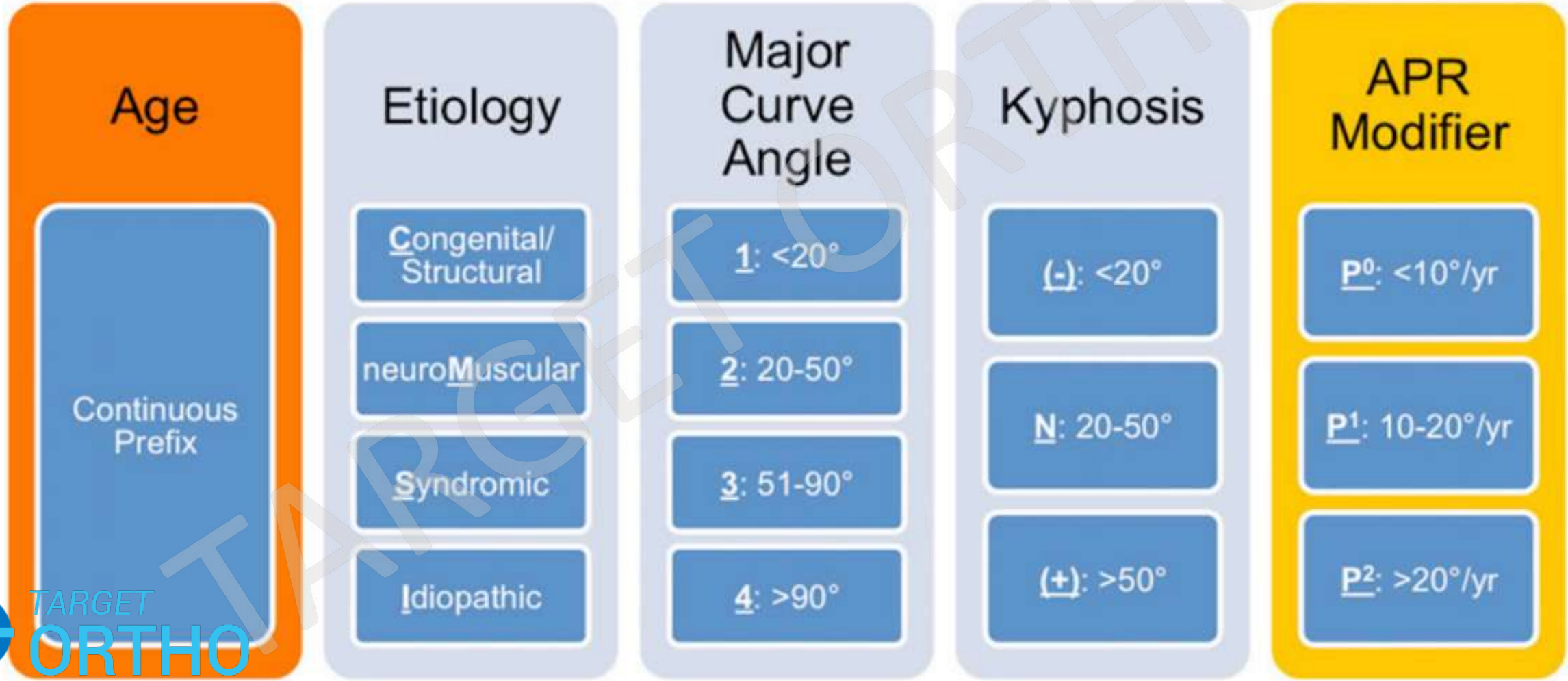
History of the terminology

- Ponseti & Friedman : EOS = <10 Years
- James: <3 Year **Infantile**; 4-9 Years **Juvenile**;
  >10 Year **Adolescent**
- Dickson **Early onset** <5 yrs, **Late onset** >5 years
- Growing Spine Study Group (GSSG) and the Children Spine Study Group (CSSG) : Any deformity before 10 years



GSSG/ CSSG-

APR= Annular Progression Rate



Etiology includes :

- Infantile Idiopathic Scoliosis
- Congenital Scoliosis.
- Syndromic/ Neuromuscular Idiopathic Scoliosis

Most commonly presents in children ages 3 years or less.

Due to the failure of normal vertebral development during 4th to 6th week of gestation

Associated NM/ Systemic disorder

Difference from AIS

	EOS	AIS
Onset of deformity	<10 Yrs	>10 Yrs
Etiology	Varies: Cong, Syn, NM, Id	Id
Spinal Height	<50% developed	>50% Developed
Height of Patient	<50%	>80%
Thoracic Volume	Less	Fully developed
Lung Development	<80%	>80%
Respiratory capacity	Less	Normal
Mortality	High	Low
Curve magnitude at maturity	High	Low
Surgical difficulty, Complication	High	Less

Pathophysiology

- **Most of the curves during first year of life,**
- **Infants have a natural tendency to turn toward the right side,**
- **Plasticity of the infant's axial skeleton, this can lead to :**

Plagiocephaly,
Bat ear on the right side,
& curvature of the spine toward the left side

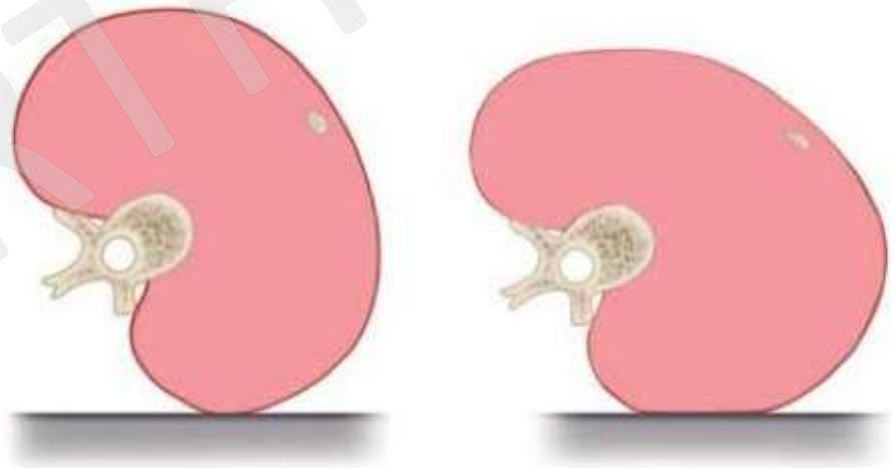


FIGURE 44-1 Diagram illustrates postural molding of thorax when infant is laid supine and partly turned toward the side.

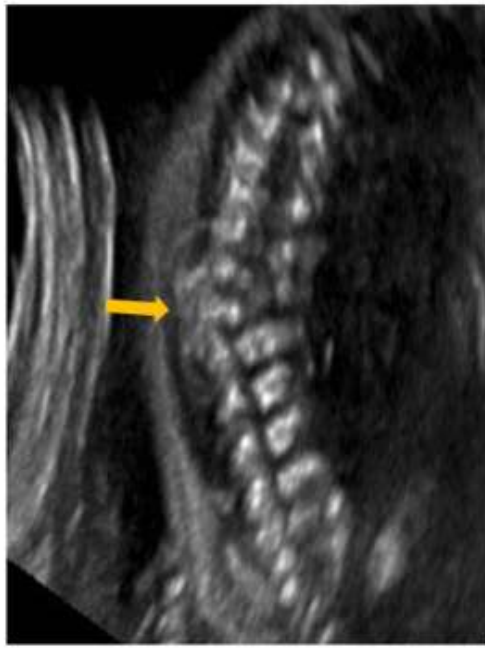
Etiology

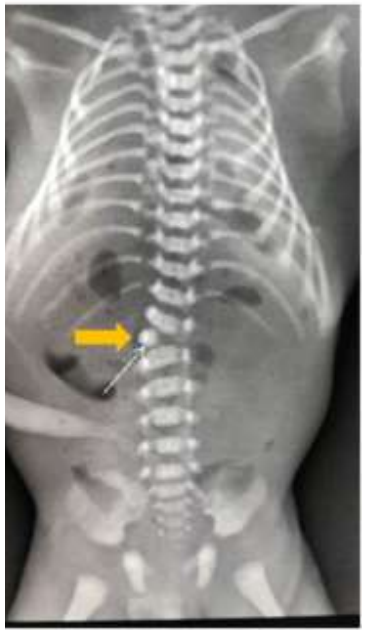
1. Mechanical factors during intrauterine life are responsible for the higher incidence
2. A second hypothesis suggests multifactorial causes, : genetic factors that are either facilitated or inhibited by external factors
3. Other associations include older mothers from poorer families, breech presentation (CO Exposure), and premature and male low-birth-weight babies.

History and Physical examination

History

- Antenatal Scan
- Exposure to teratogens/ Carbon monoxide
- DM/ Hyperthermia/ Anticonvulsant
- Postural
- Repeated infection

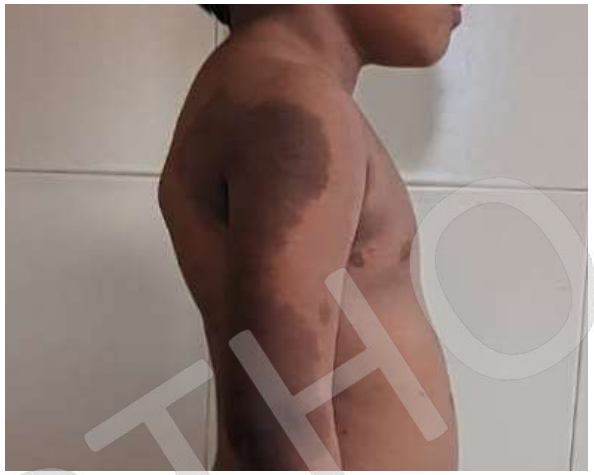


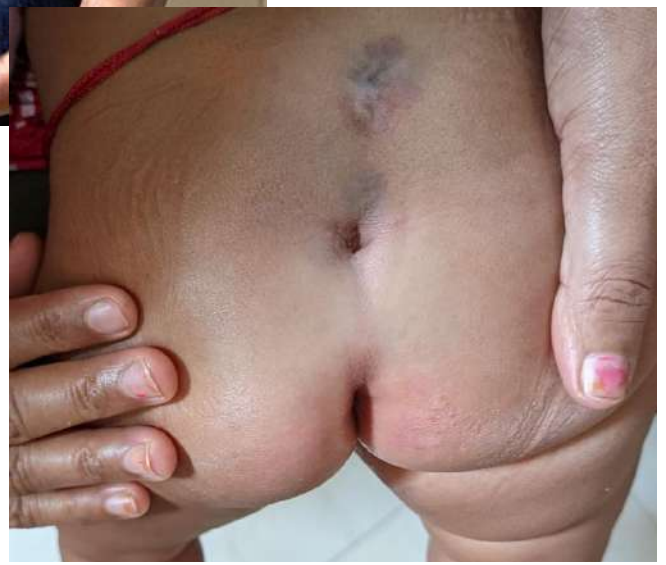


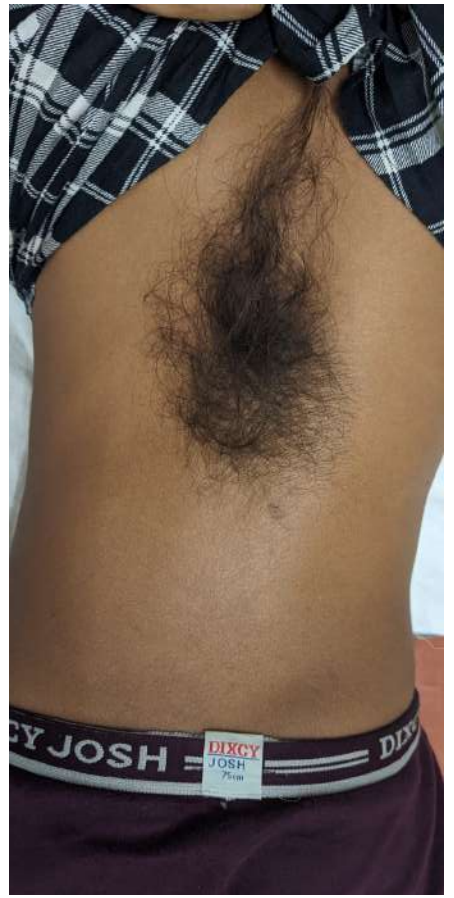
Examination

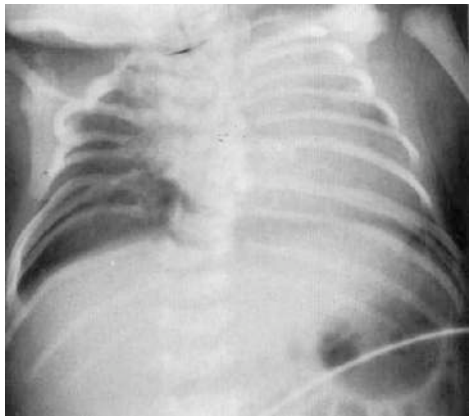
- Head to toe exam
- Neurologic examination
- Neurocutaneous markers
- Chest wall examination

- Scoliosis special tests
- Assesment of Renal/ Cardiac system





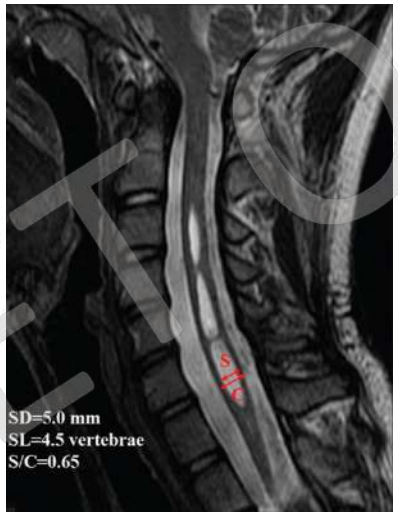


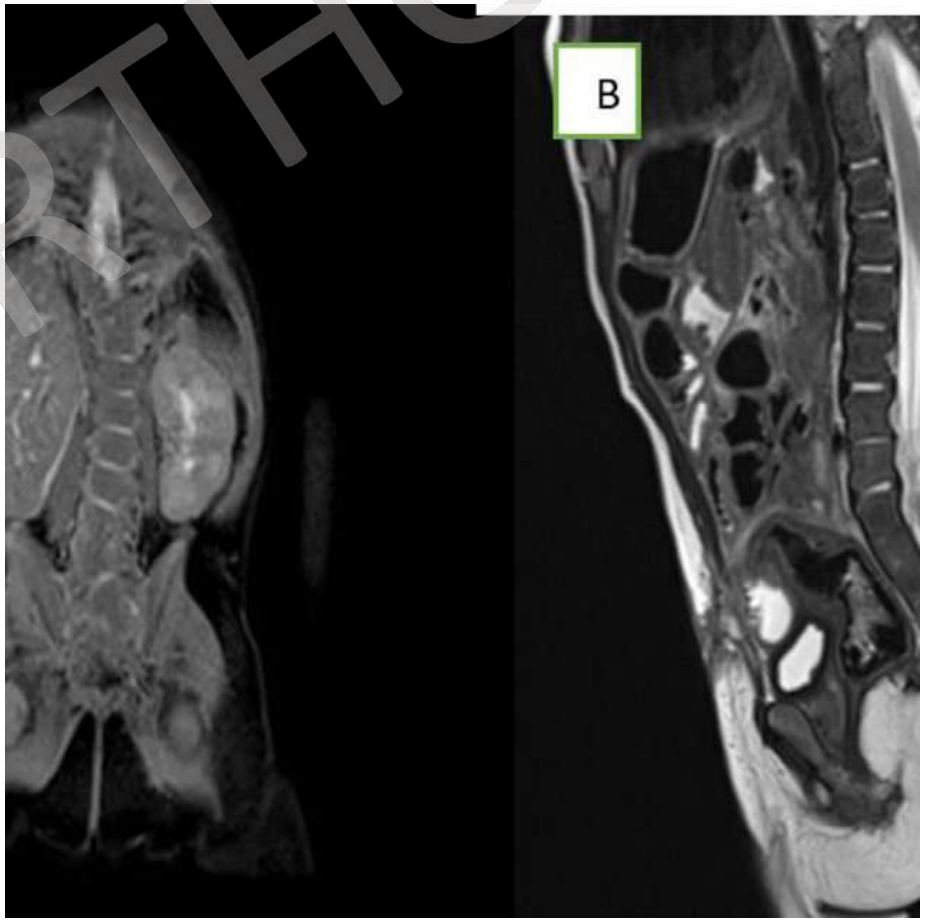


Associated anomalies

- Present in upto 60% patients

- Renal 20% (USG +/- Doppler)
- Cardiac 25% (2D Echo)
- Neurologic 10-35% (MRI)





Thorax assesment

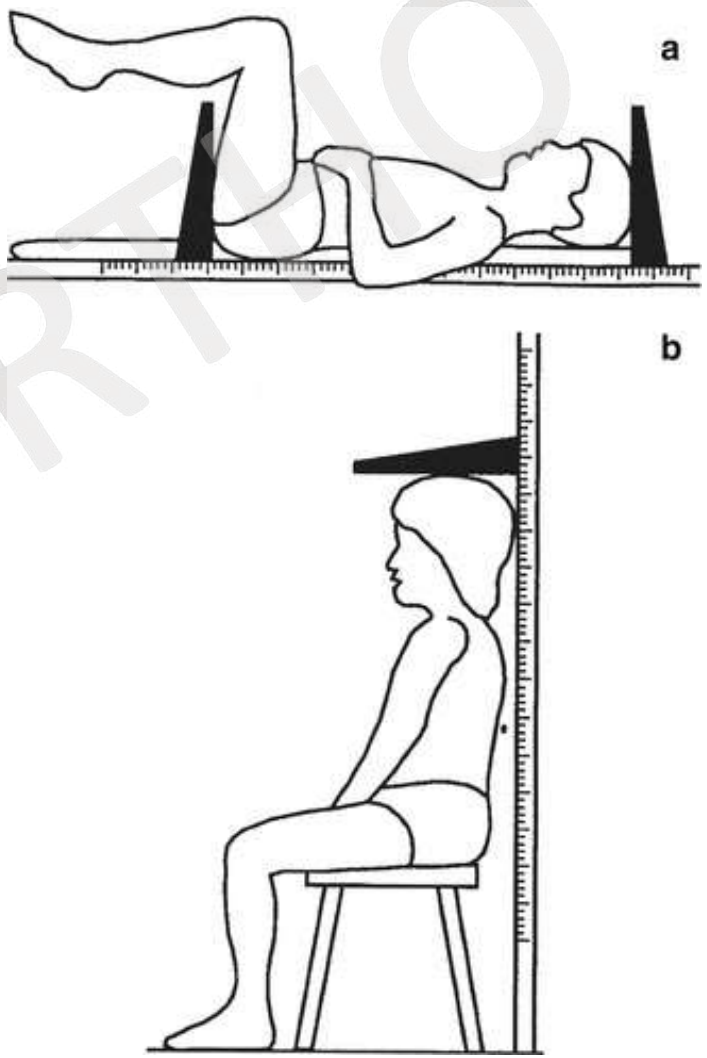
- Sitting Height
- Thorax abnormalities
- TIS

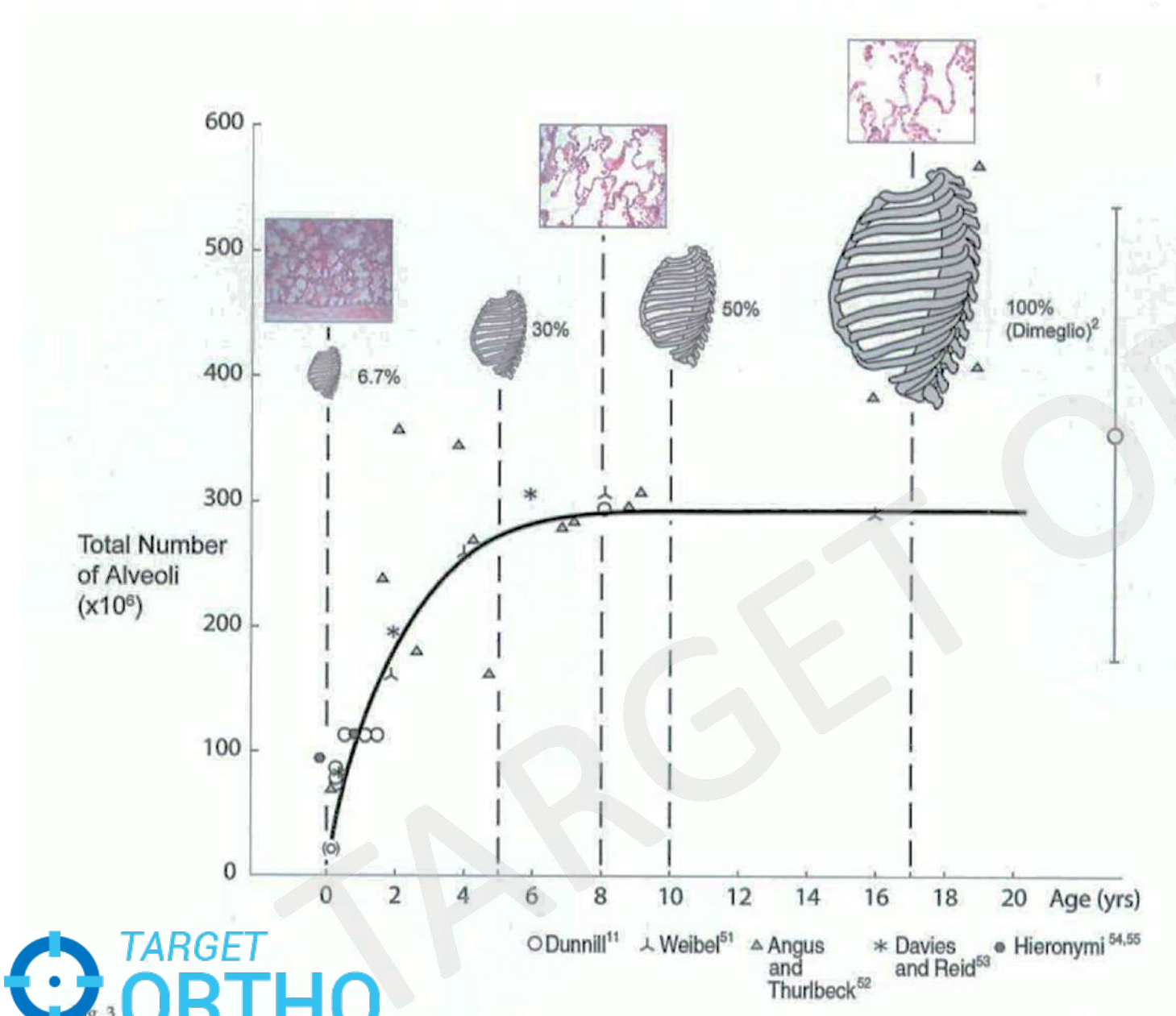
Aim in infantile scoliosis:

Wt >40 Kg,

Vital Capacity > 50%,

Length 22cm thoracic spine,
T1-S1 30 cm



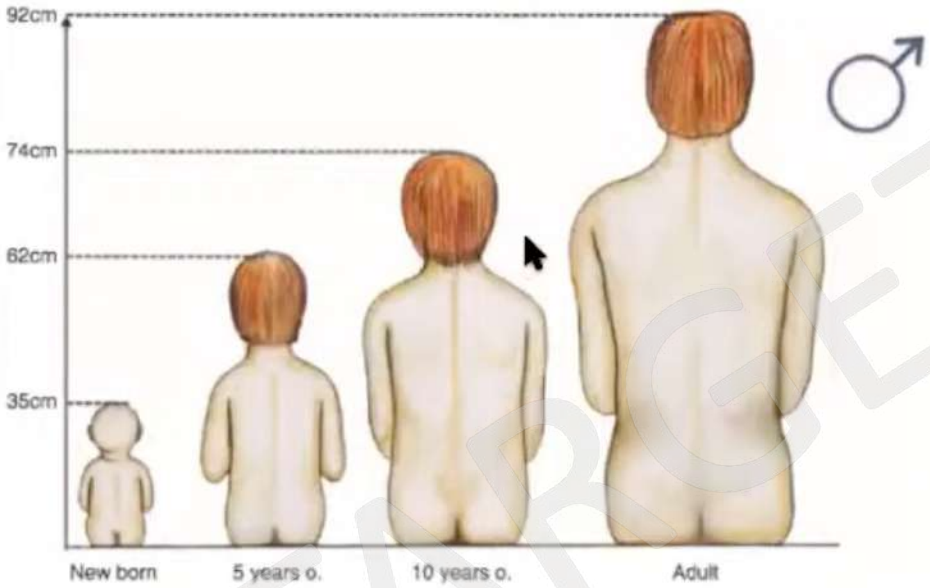


Vital capacity 50% at 10 years

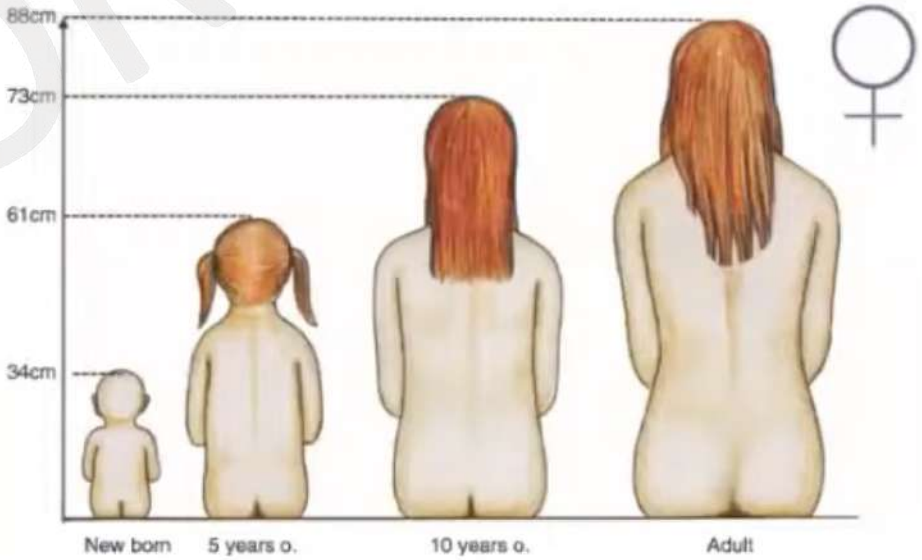
Sitting height

T1-S1 Length
1st 5 years: 10 cm (2cm/yr)
5-10 years: 5 cms (1cm/ yr)
Adolescence: 10 cm (2 cm/yr)

Thoracic spine height
At birth 11 cm
At 5 years 18 cm
At 10 years 22 cm



Average Sitting Height in Boys



Average Sitting Height in Girls

Scoliosis at early age

↓

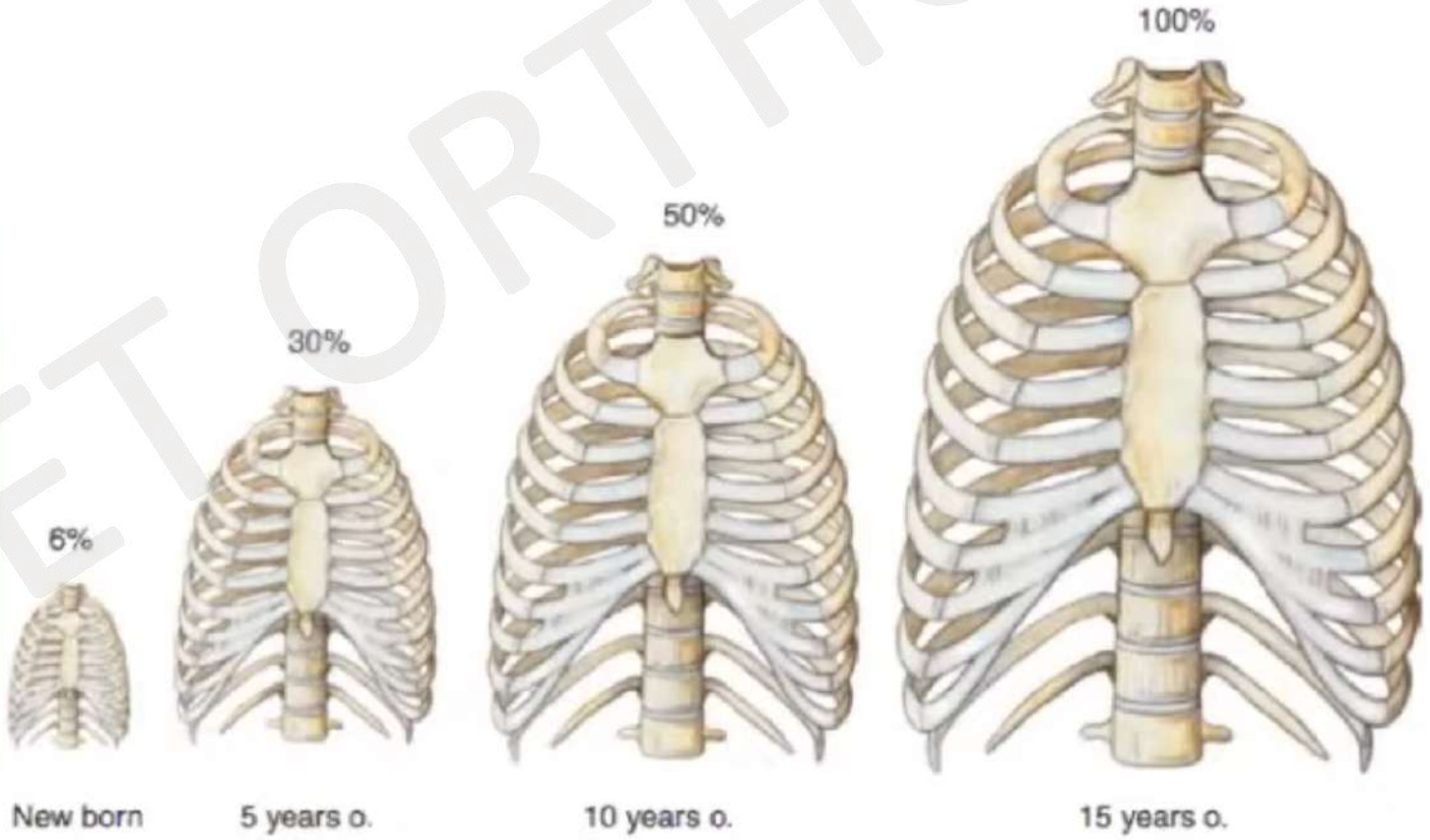
Thoracic development affected

↓

Lung development and volume affected

↓

Restrictive lung disease



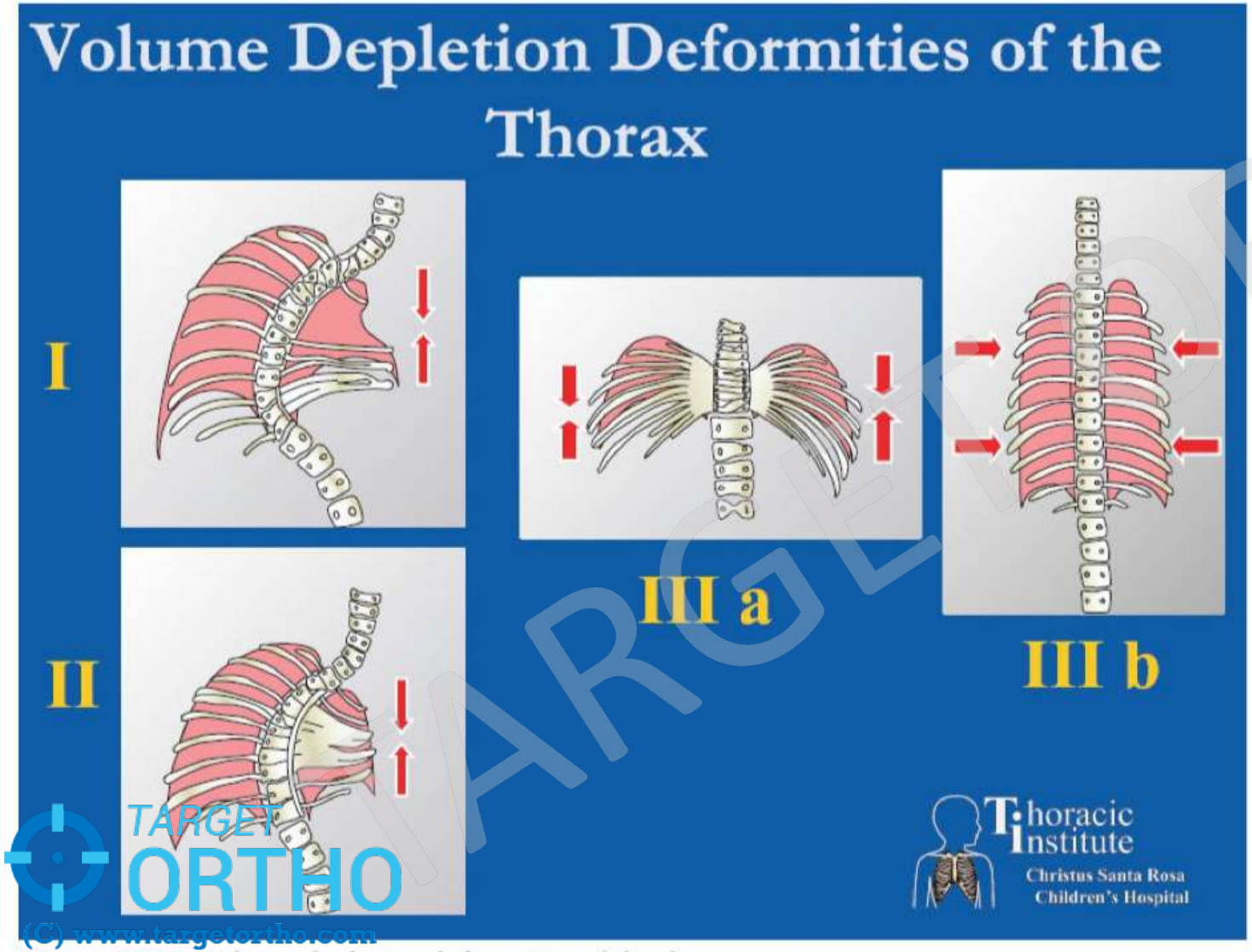
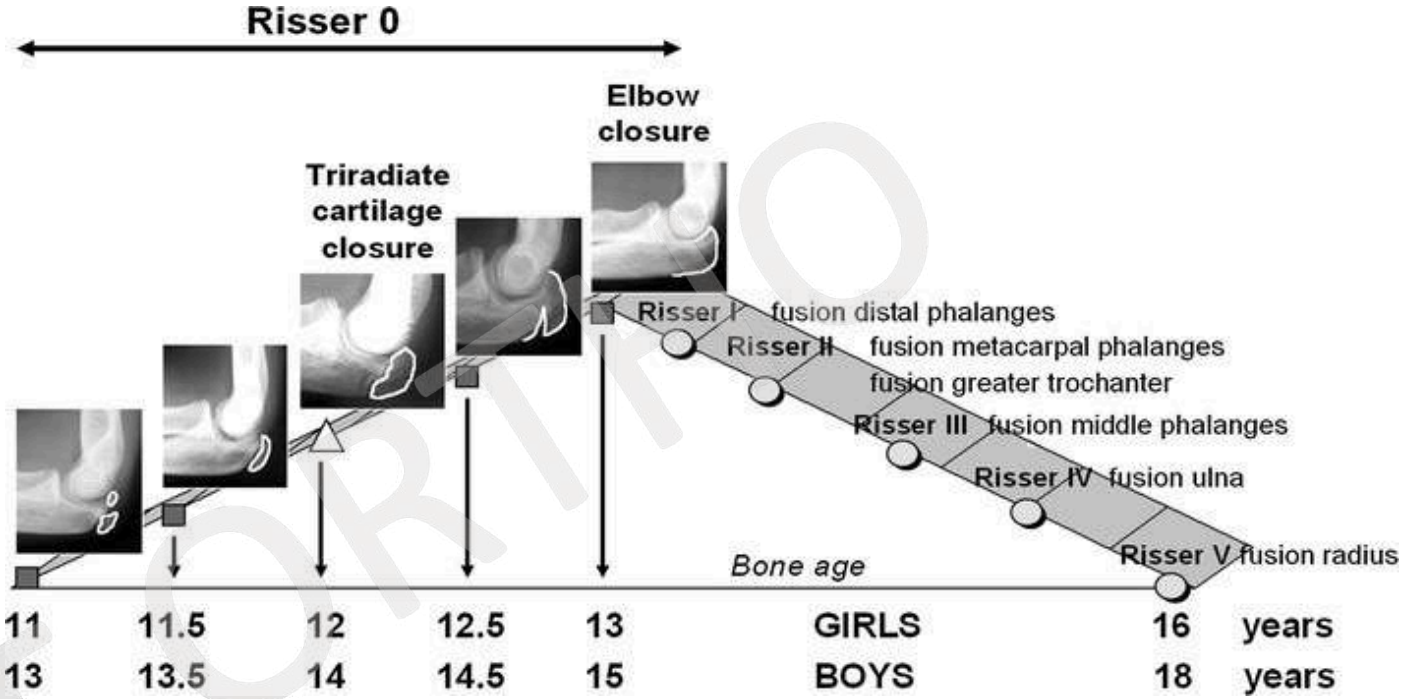
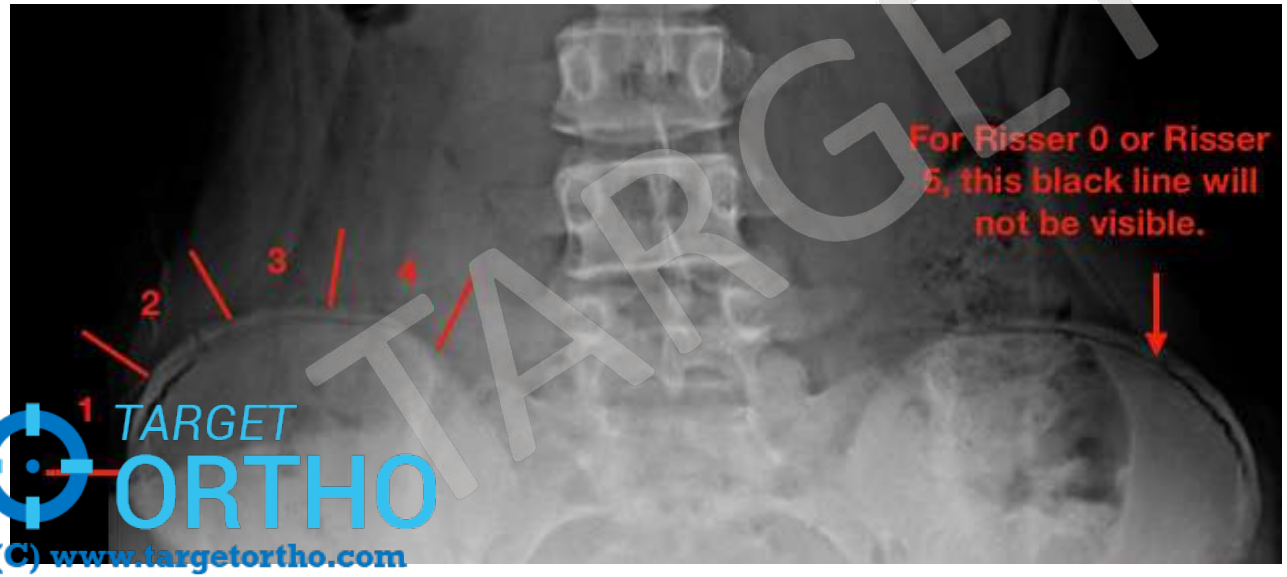


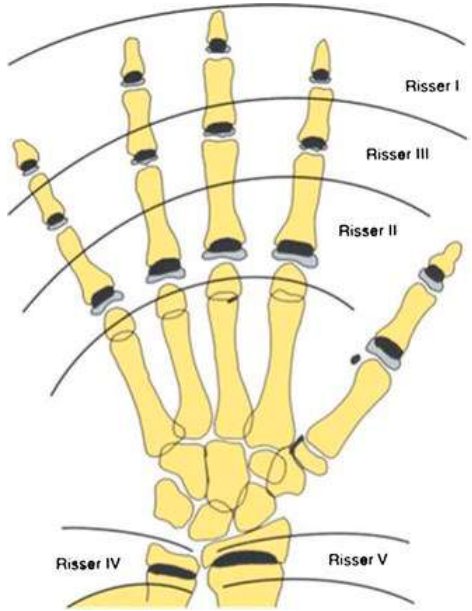
TABLE 109.1 Volume Depletion Deformities (VDDs) of the Thorax[3]

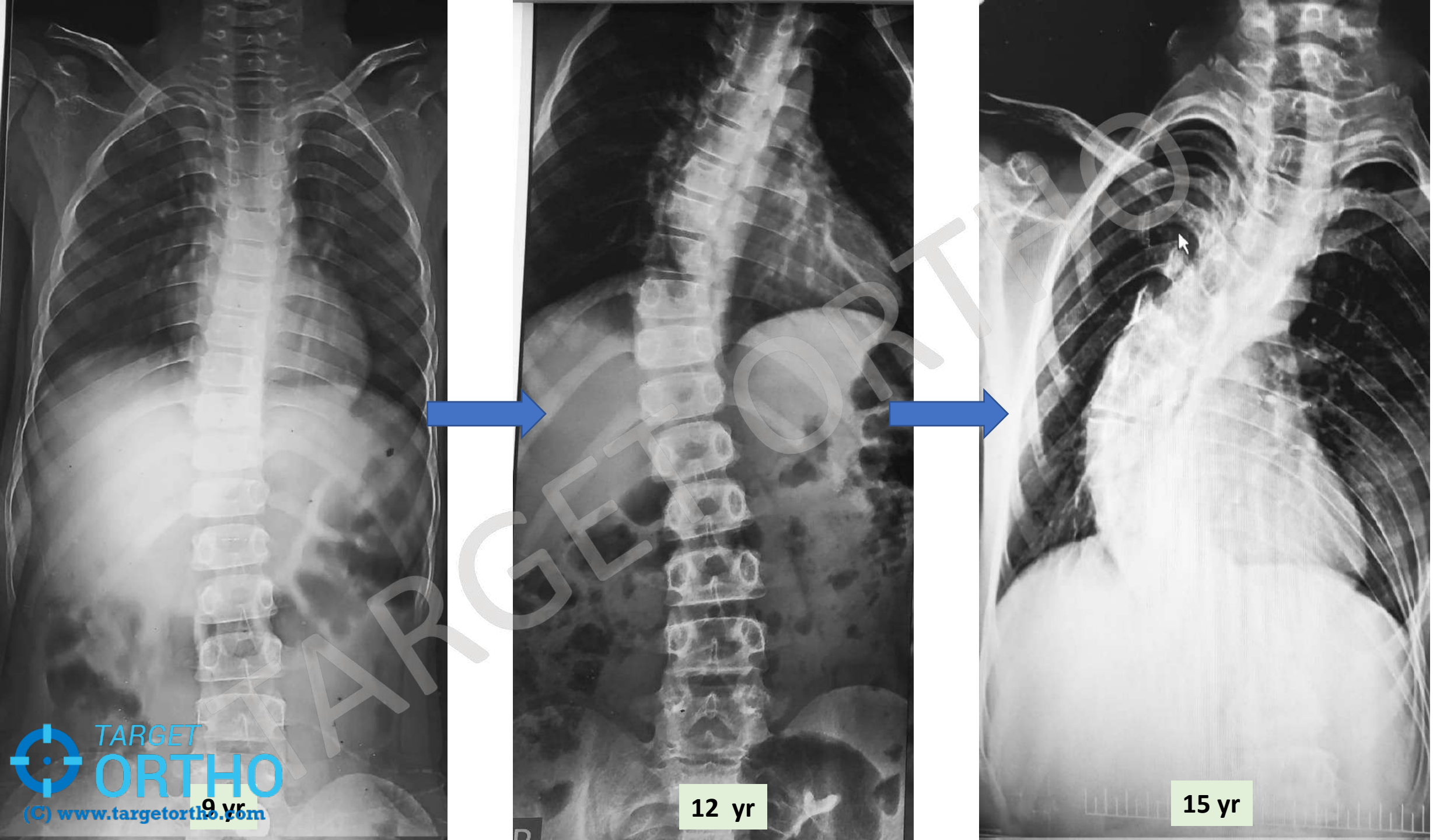
Unilateral thoracic volume depletion deformity	
Type I	**Absent ribs and scoliosis**
Type II	**Fused ribs and scoliosis**
Global thoracic volume depletion deformity	
Type IIIa	**Jarcho–Levin syndrome**
Type IIIb	**Jeune syndrome, early-onset scoliosis**

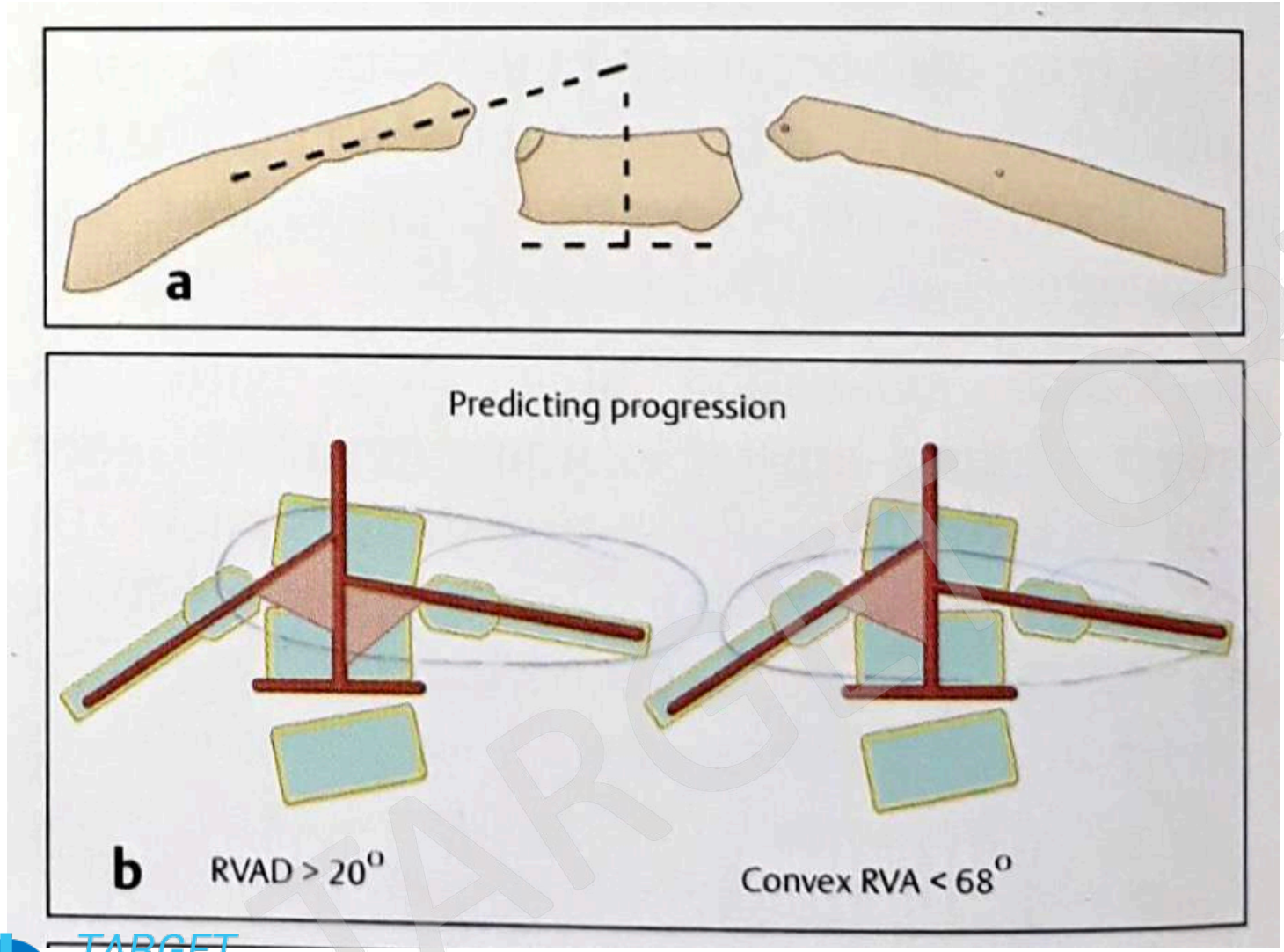
Radiological assessment of remaining growth





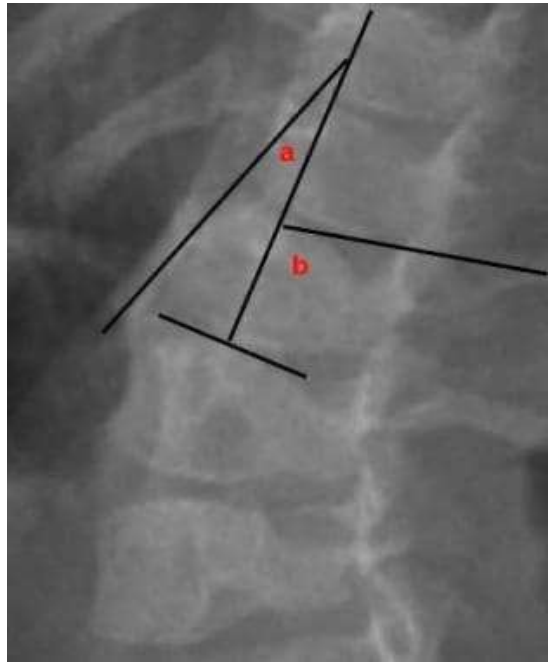






Rib Vertebral angle difference (RVAD)

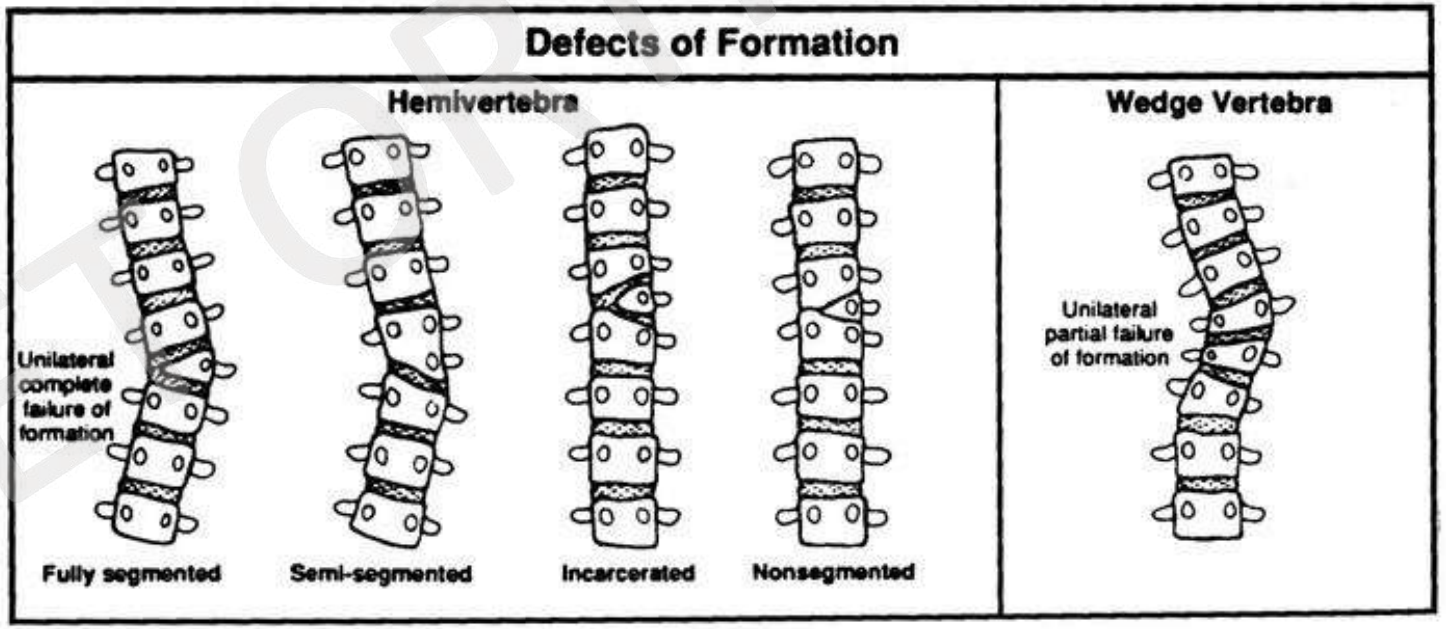
Mehta's RVAD >20* is an indication for interventions as these curves are likely to progress



Congenital: Formation/ Segmentation defects

1. Wedge
2. Hemi

- Segmented: Disc above, Below
- Semi Segmented: Fused above/Below
- Incarcerated: Pedicle lines with above and below
- Non-Segmented: Fused above and below

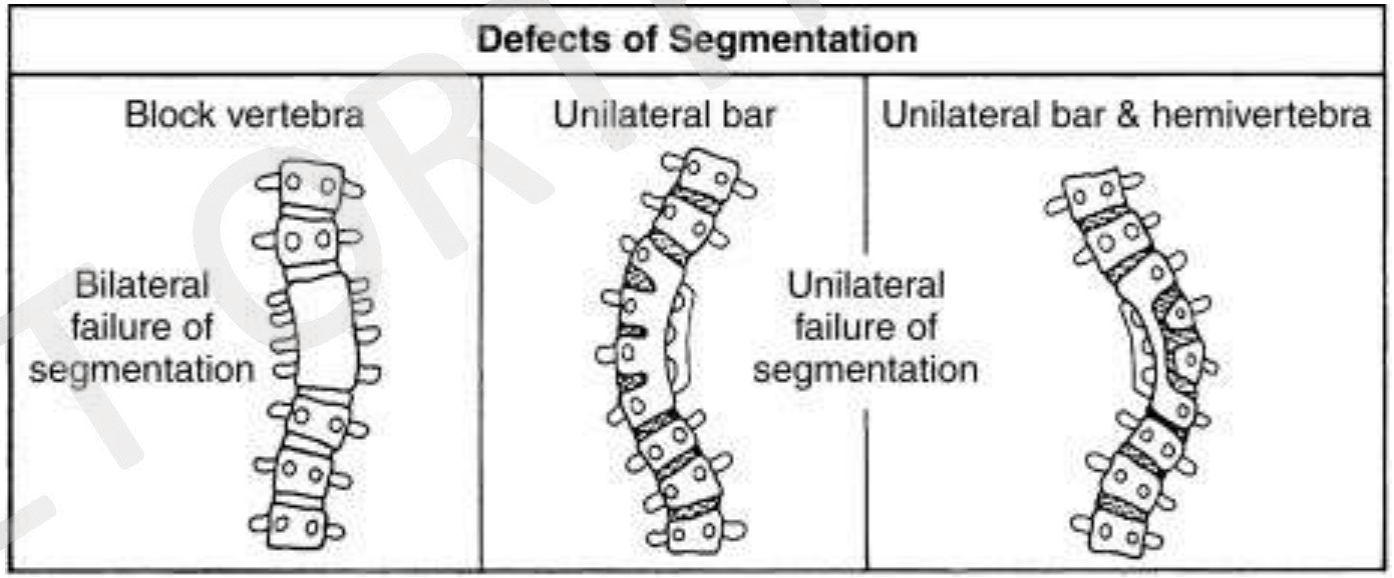


Open disc= Growth Potential ++

Congenital: Formation/ Segmentation defects

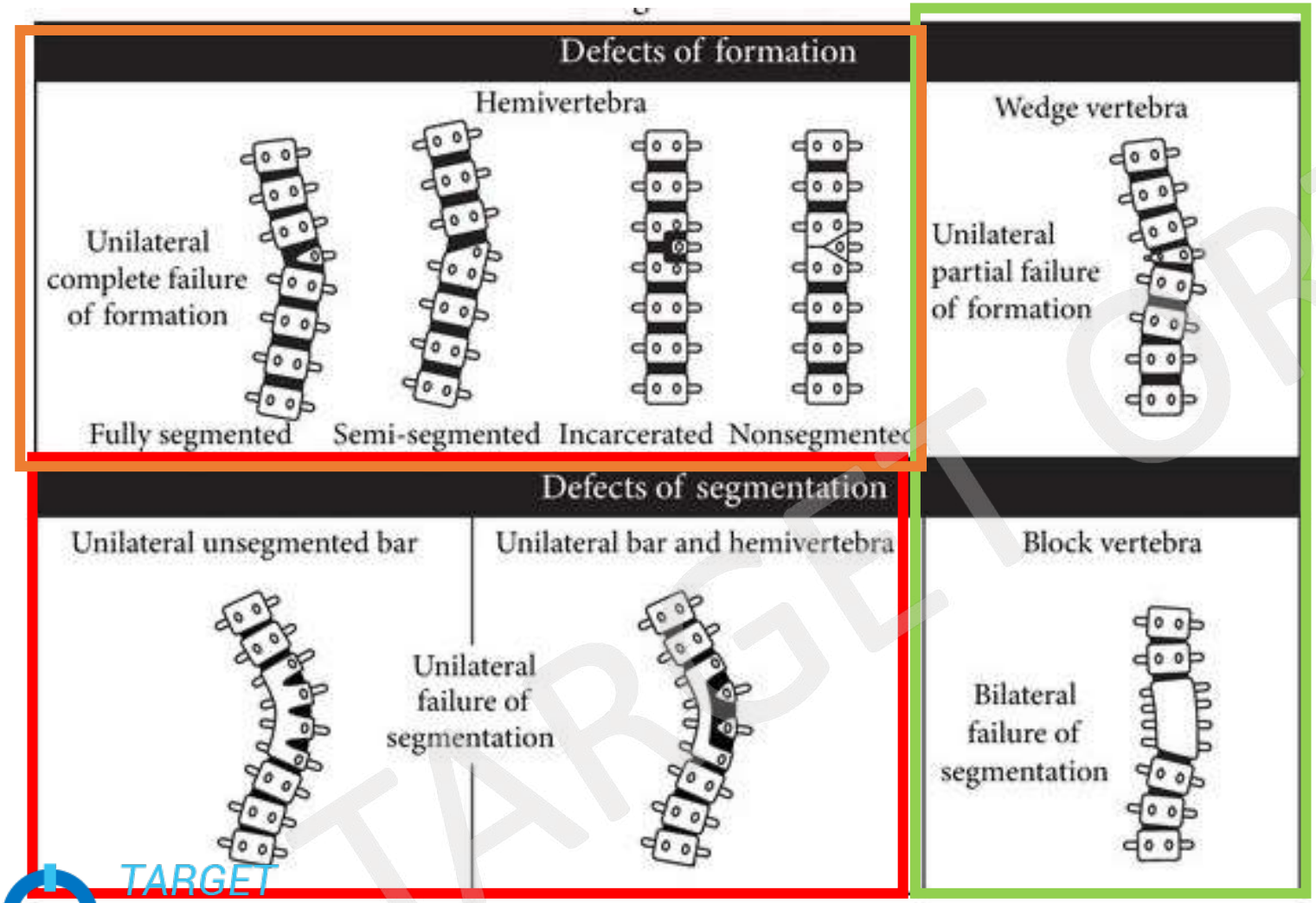
1. Block
2. Bar

- With/ Without hemivertebra



Hemi vertebra = Growth Potential ++

Types of Congenital Scoliosis



Unilateral unsegmented bar with contralateral hemivertebra (5 to10 degrees/year) >

Unilateral unsegmented bar >

Fully segmented hemivertebra >

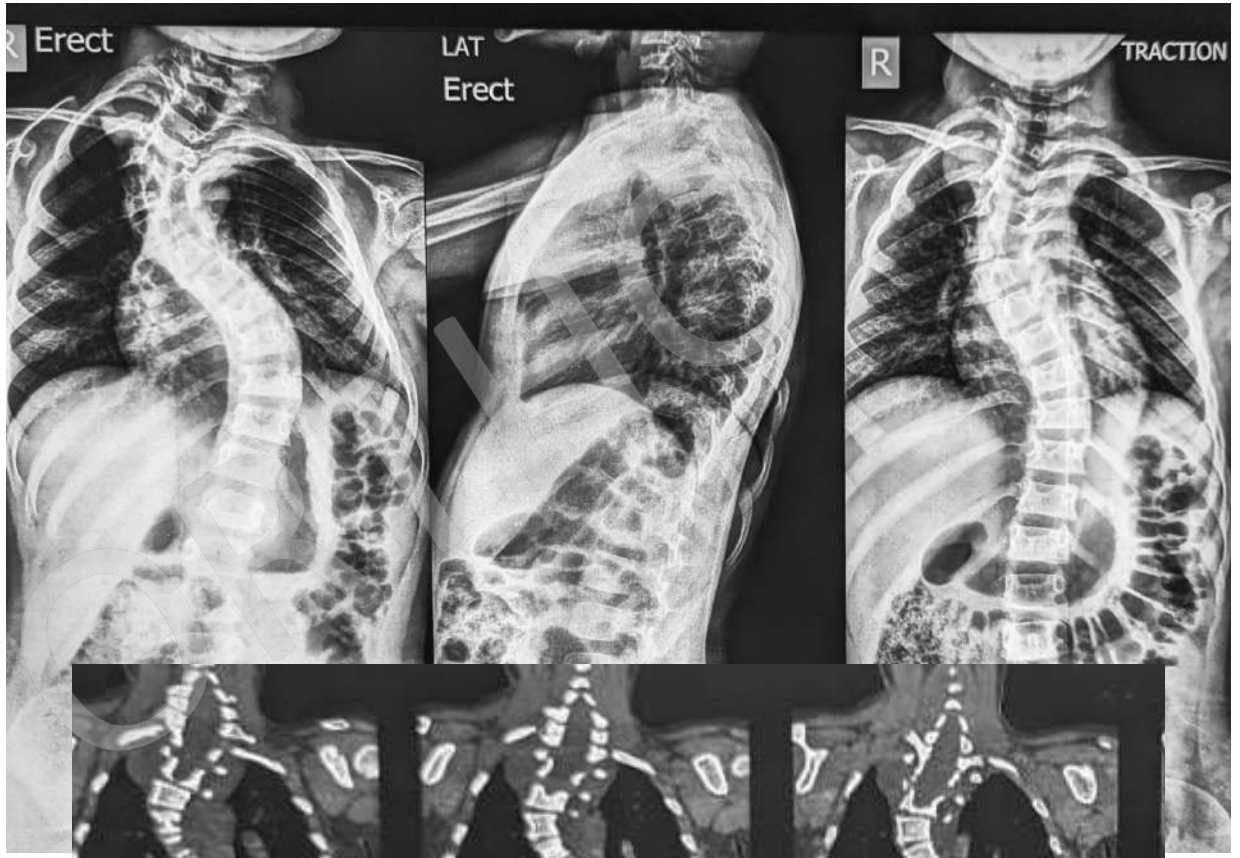
Unincarcerated hemivertebra >

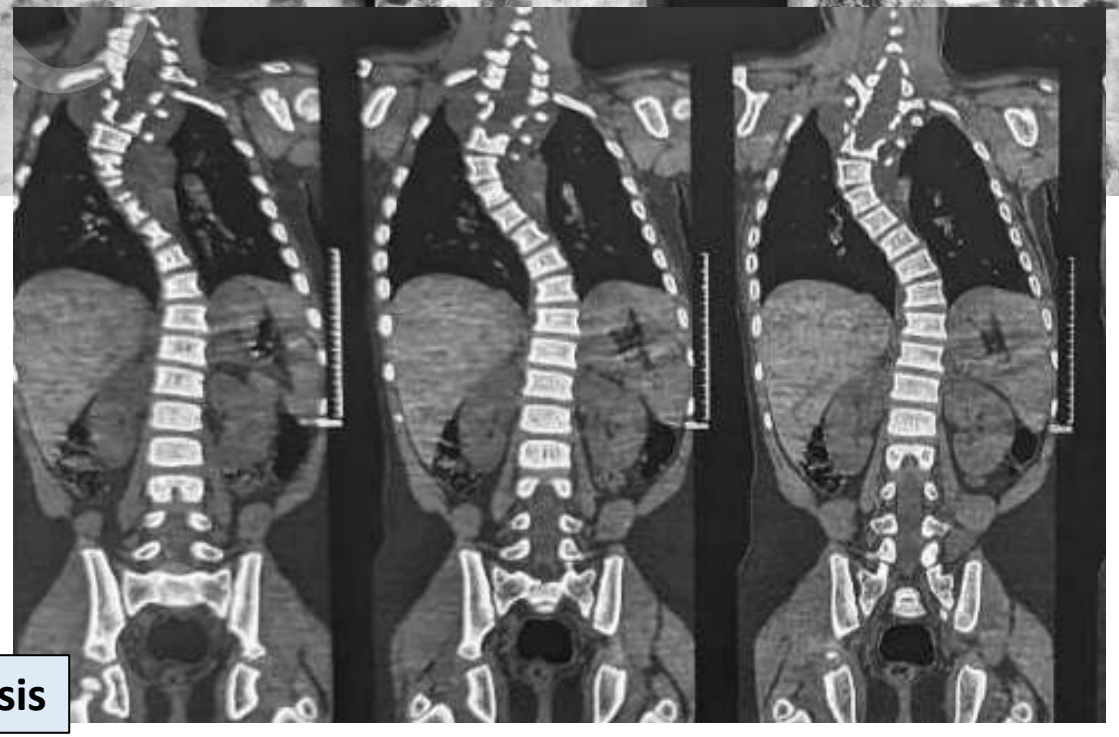
Incarcerated hemivertebra >

Unsegmented hemivertebra >

Block vertebrae (<2 degrees/year)

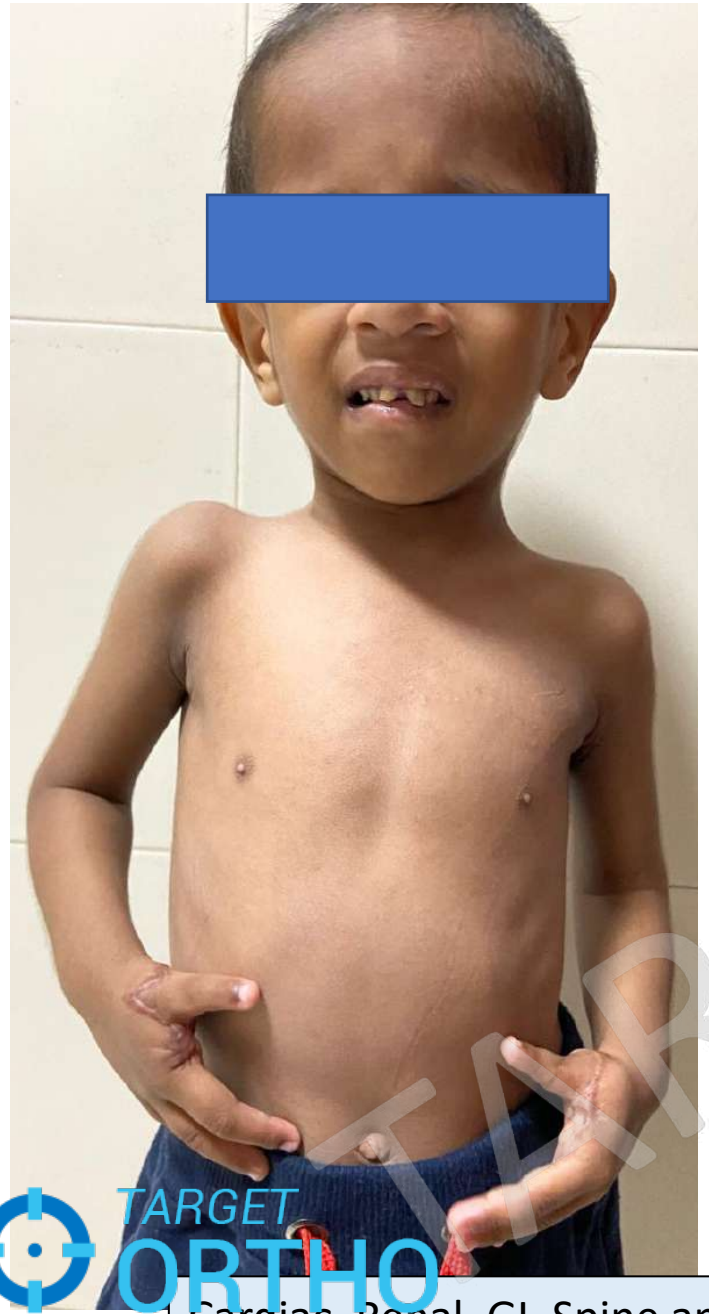
Presence of fused ribs increases risk of progression

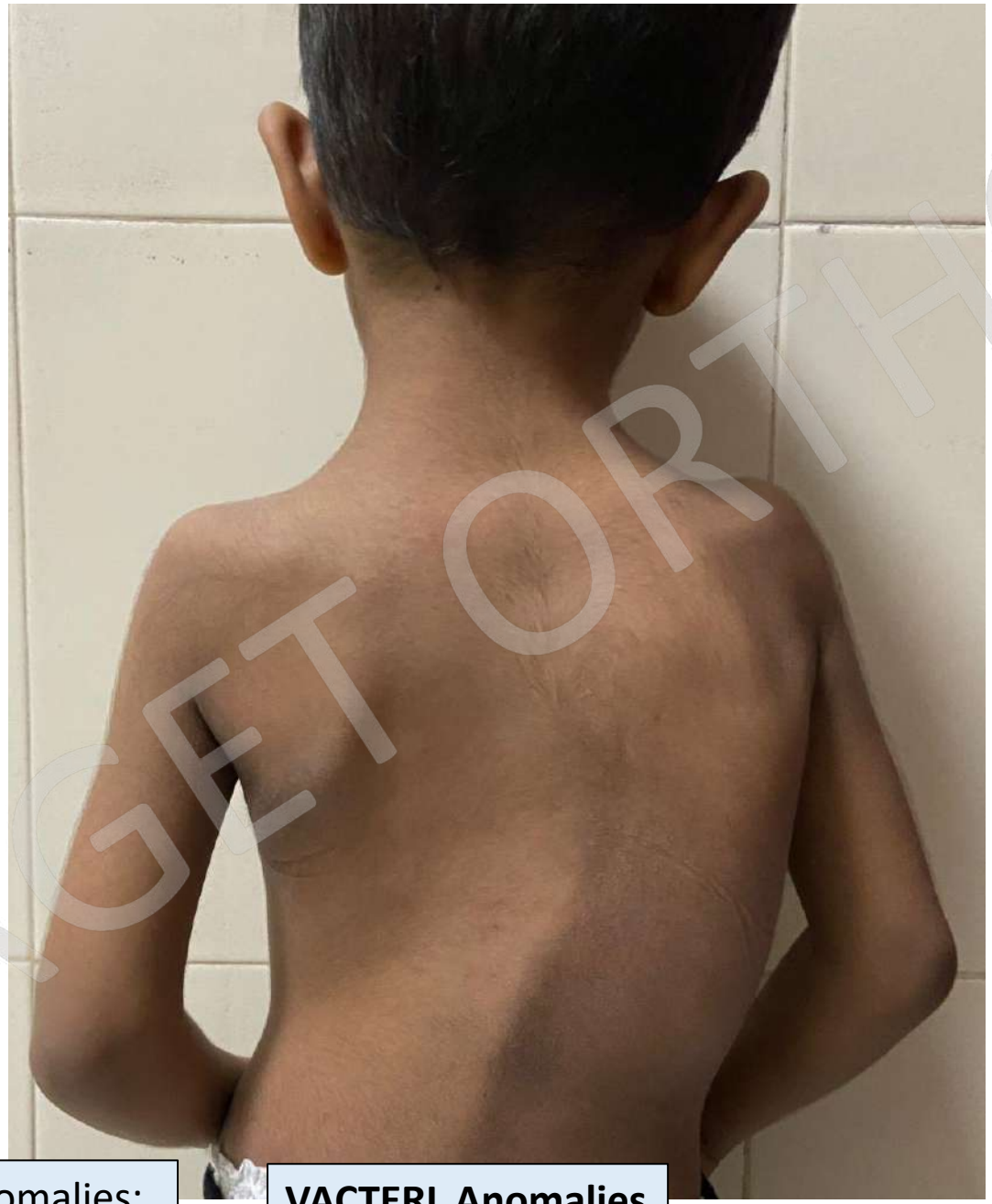


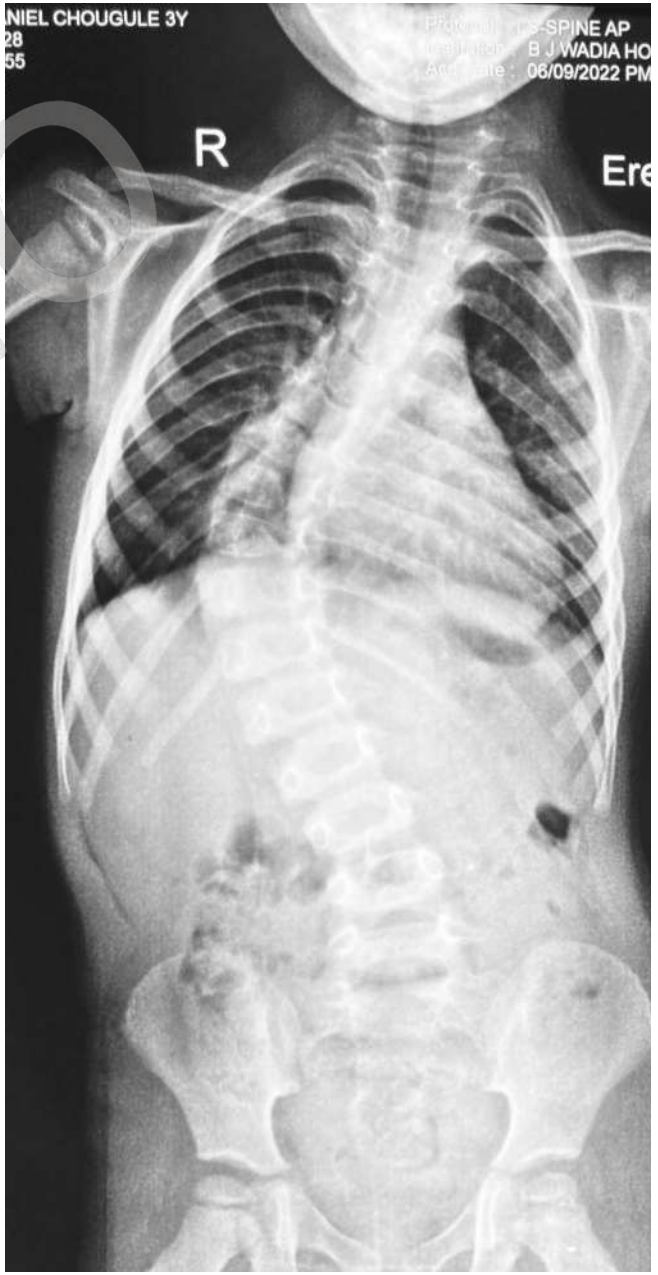


Café au lait spots, Neurofibromas :

NF with scoliosis

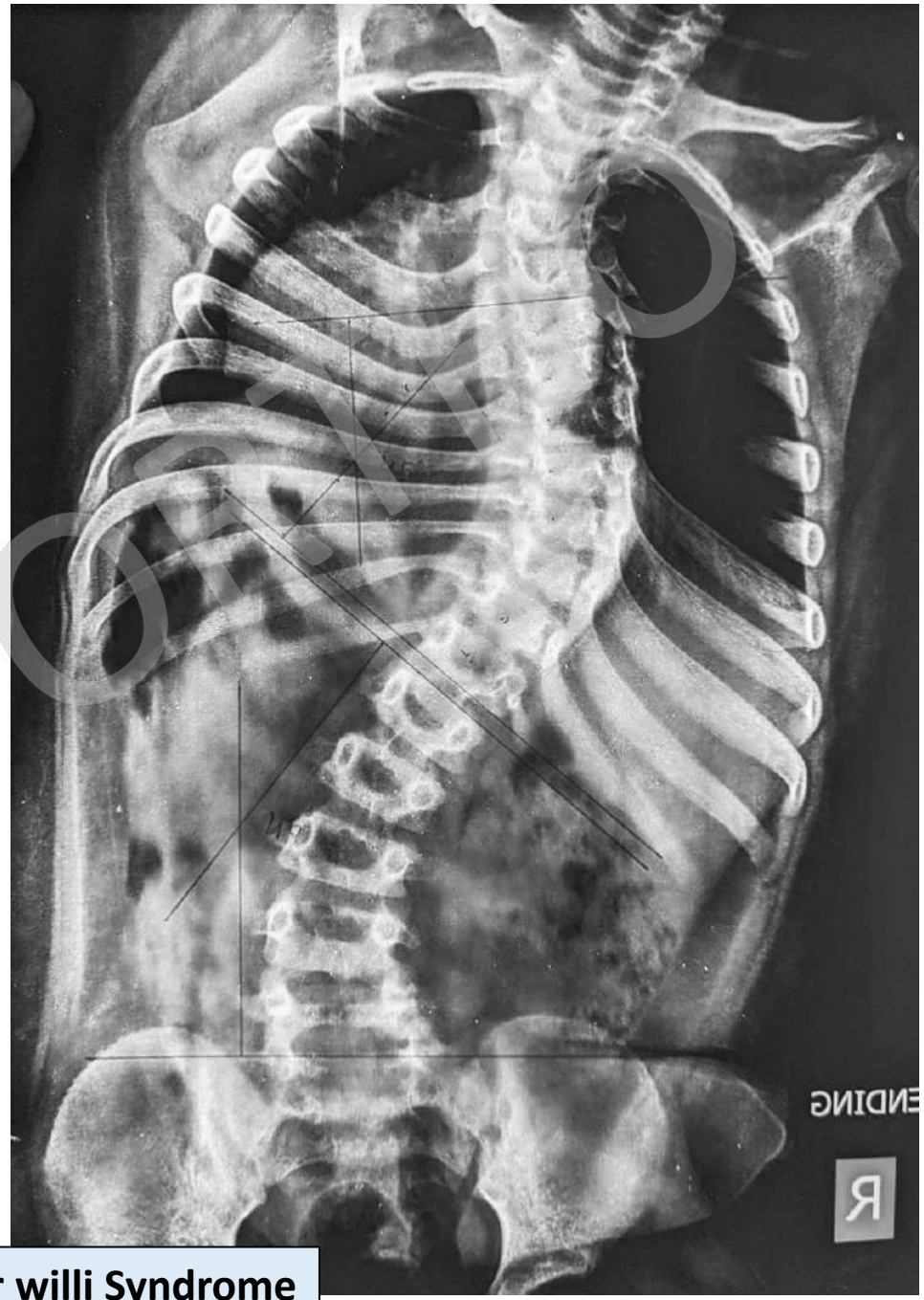






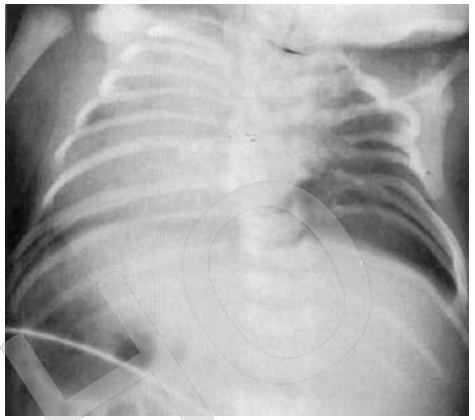
Cardiac, Renal, GI, Spine anomalies:

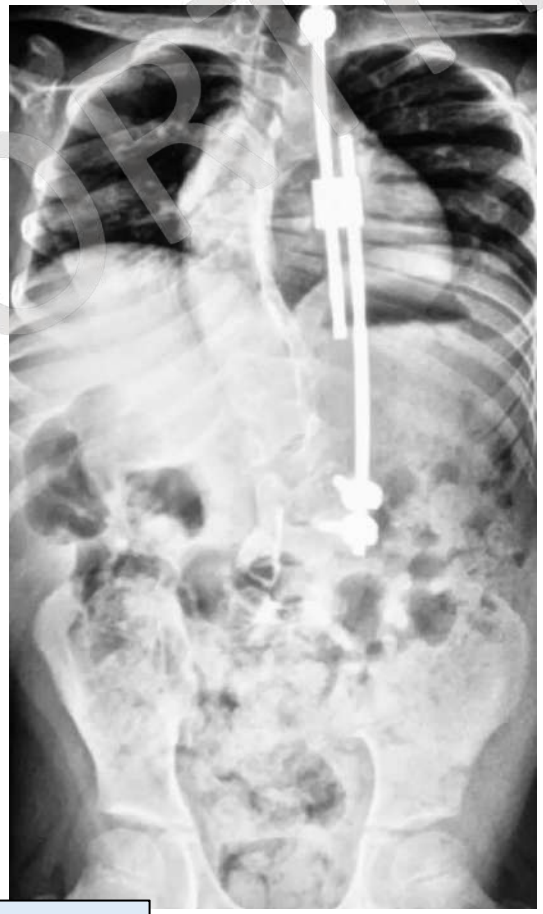
VACTERL Anomalies

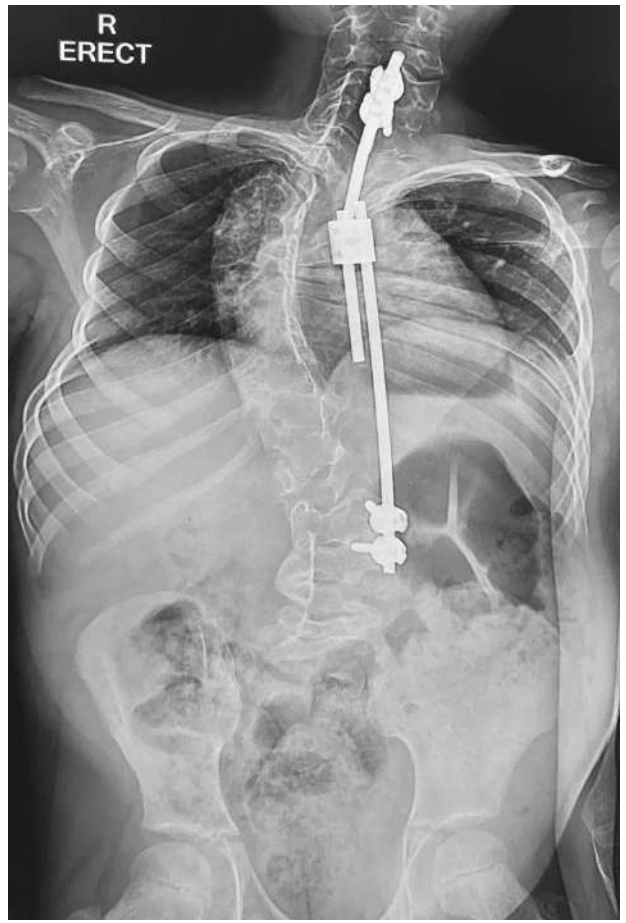


Obesity, Polyphagia, DM, Delayed puberty

Prader willi Syndrome



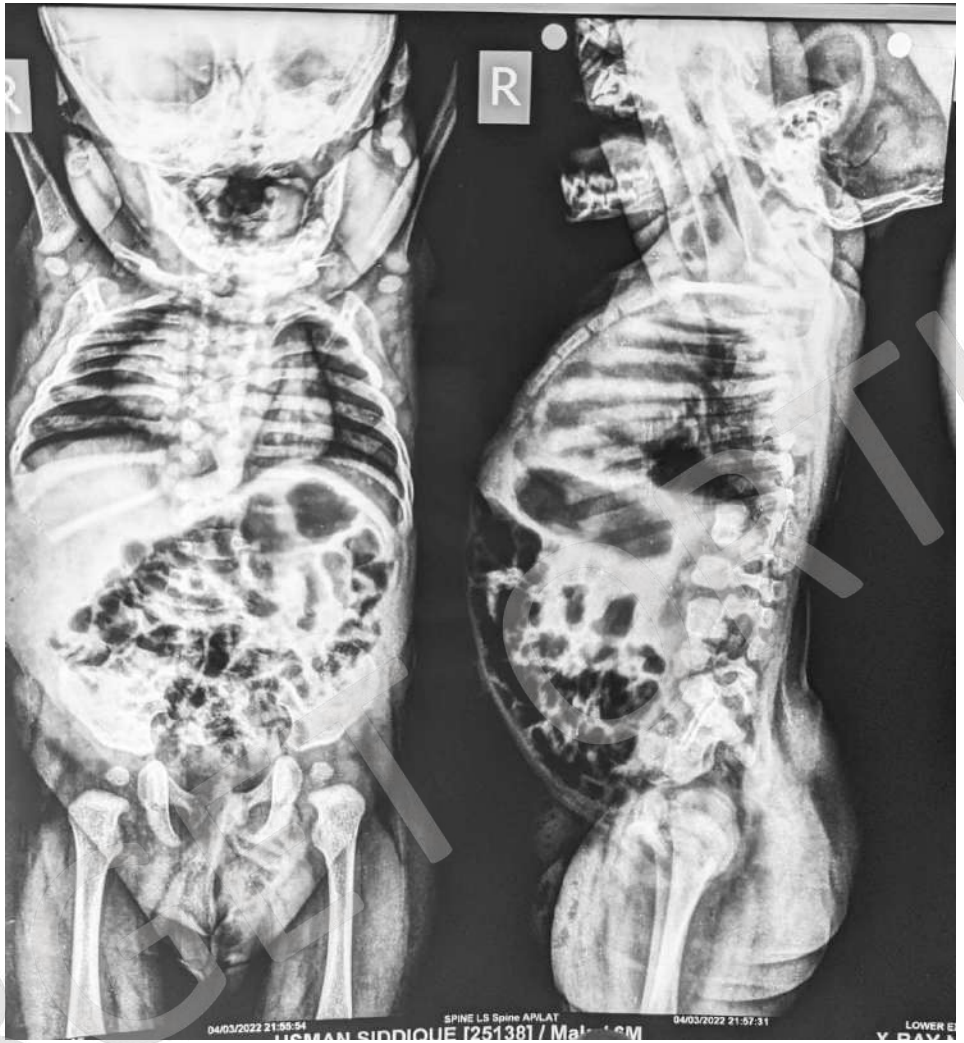




Short neck, short trunk and a constricted thorax
Multiple Rib and vertebral anomalies
Multiple childhood respiratory infection

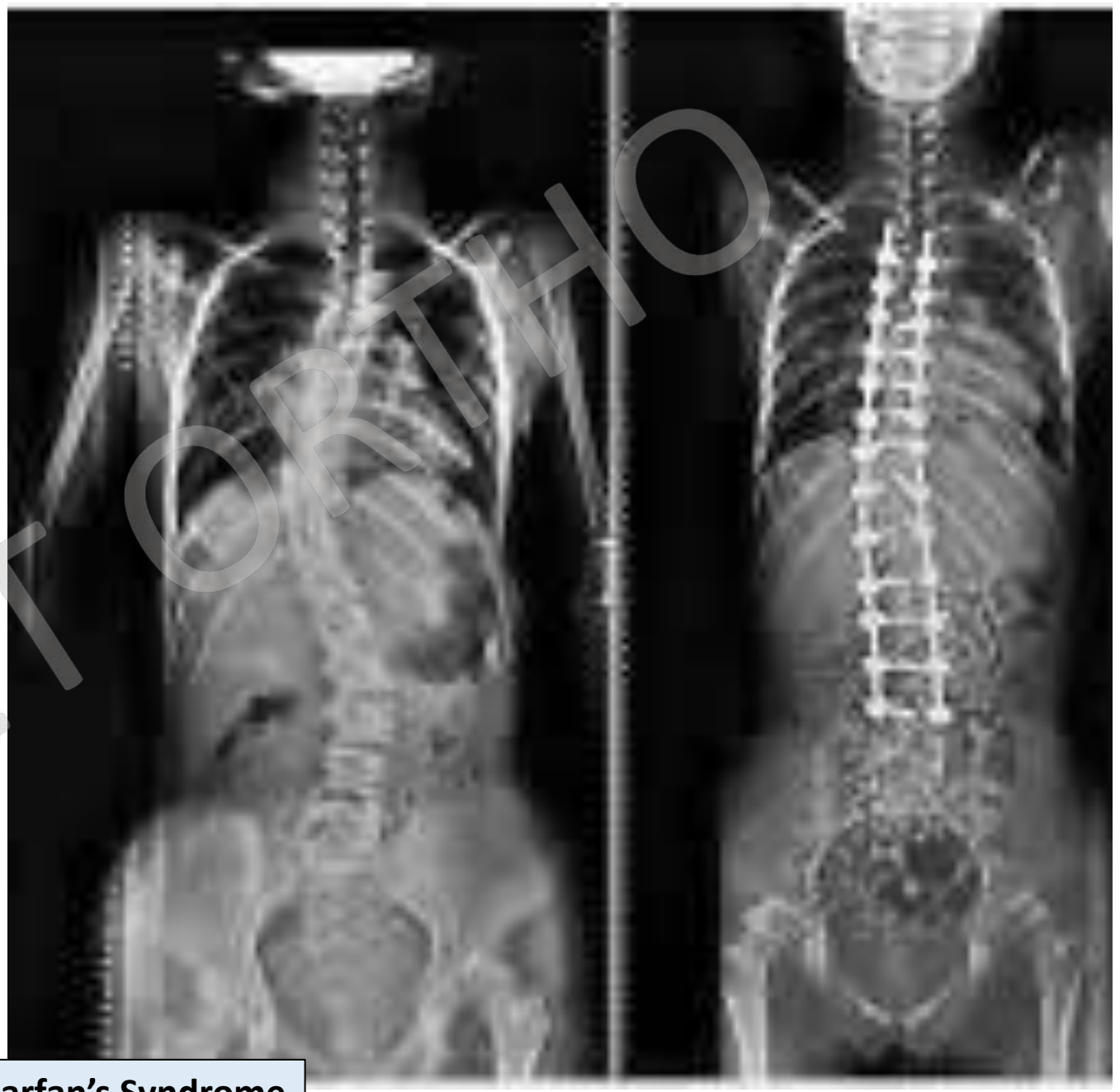
Spinocostal Dysplasia

Short neck, short trunk and a constricted thorax
Multiple Rib and vertebral anomalies
Multiple childhood respiratory infection

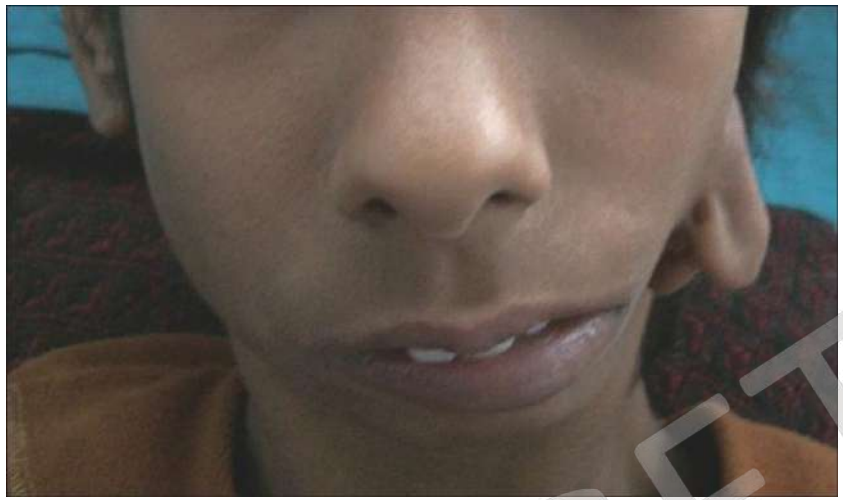


Jarcho Levine Syndrome

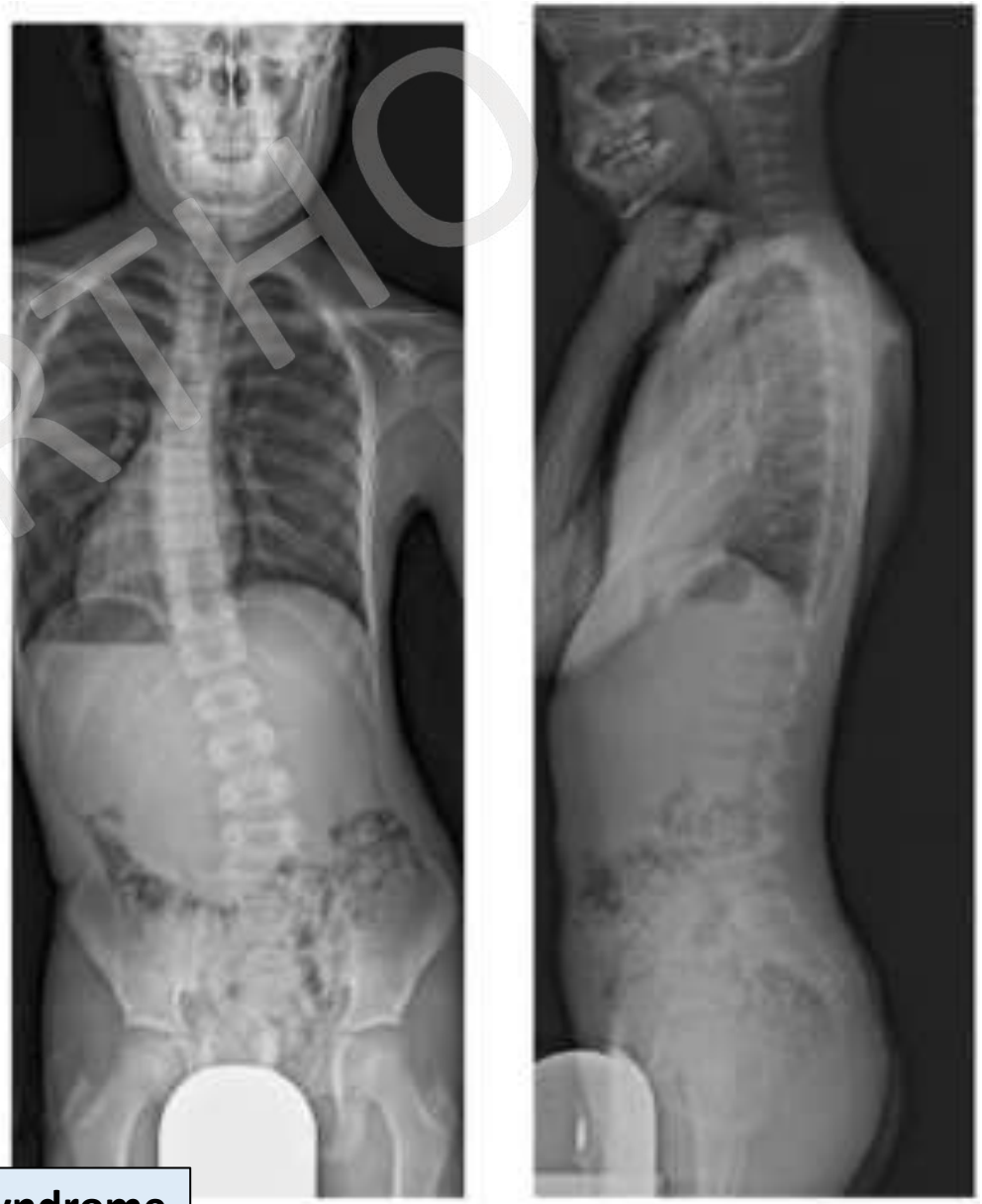
Hyperlaxity,
Increased arm span,
Caardiac defects



Marfan's Syndrome



Abnormal development of the:
eye, ear and spine.
Also known as
"oculo-auriculo-vertebral spectrum"
- No scoliosis in Treacher collins



Goldenhaar Syndrome

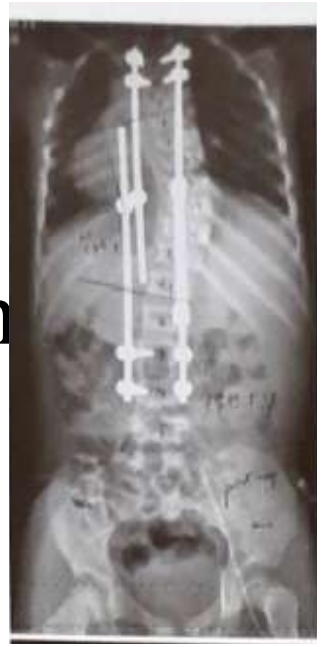
1. Hairy patches - Spina bifida or diastematomyelia
2. Subcutaneous tumors - Neurofibromatosis
3. Pigmented lesions - Neurofibromatosis
4. Excessive flexibility - Ehlers–Danlos or Marfan syndrome
5. Joint contractures- Cerebral palsy or arthrogryposis
6. Craniofacial abnormalities- Syndromic scoliosis
7. Weakness of extremities- Neuromuscular scoliosis.

To Conclude

- Interrelated growth of spine and thorax
- Associated Comorbidities

COMPLEX Management

atric scoliosis: Managen



Dr Hriday Acharya

Consultant Spine Surgeon

MS Ortho(Gold medalist)

Fellowship in spine surgery

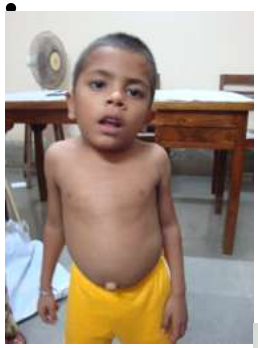
ASSI – Mumbai

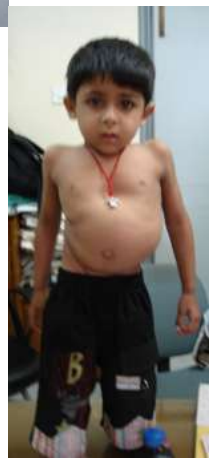
Pediatric spine fellowship – B J Wadia

Goals of Management

- Minimize spinal deformity over the life of the patient
- Maximize thoracic volume and function over the life of the patient
- Minimize the extent of any final spinal fusion
- Maximize motion of the chest and spine
- Minimize complications, procedures, hospitalizations, and burden for the family
- Consider overall development of the child

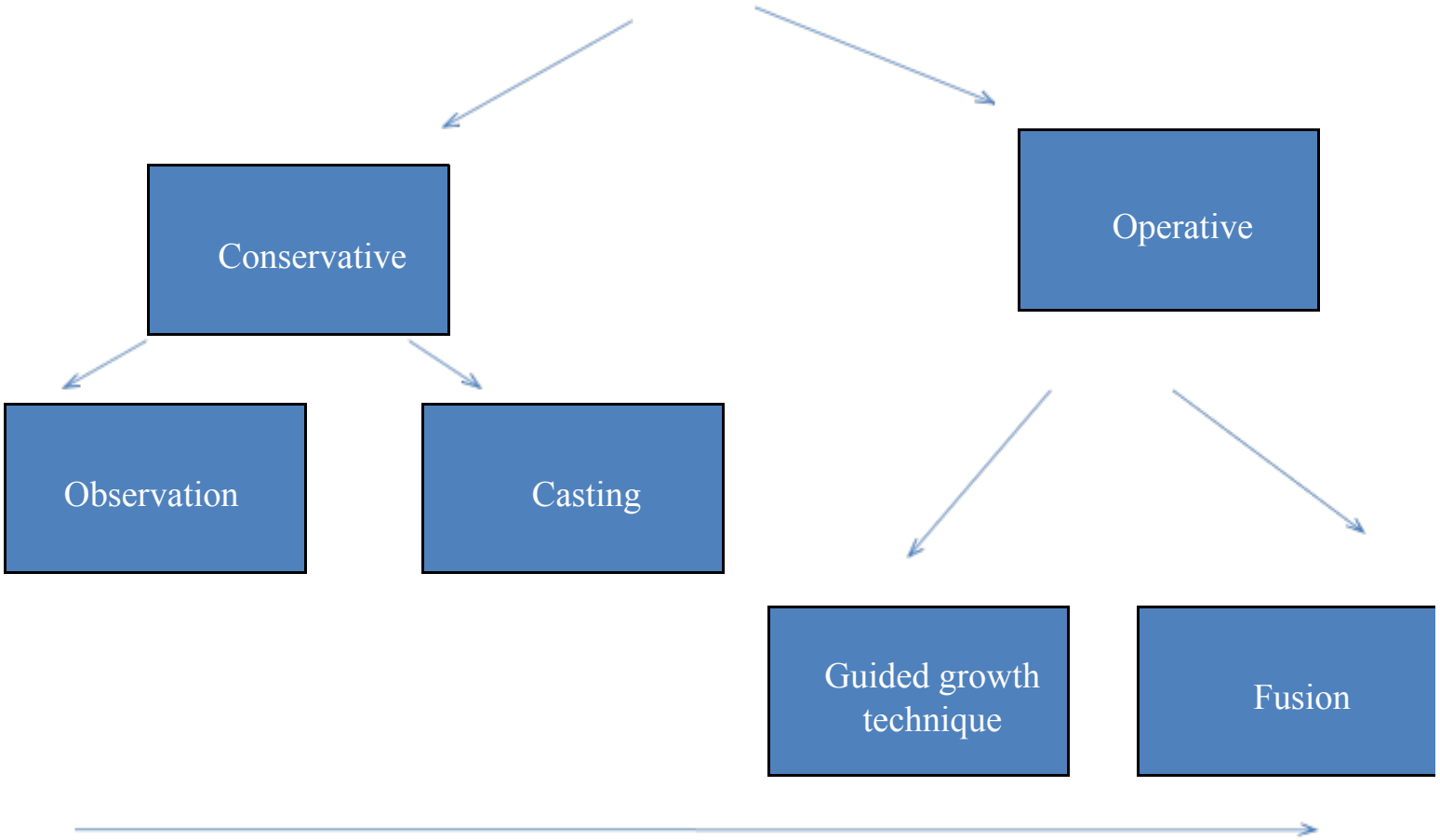
How do I decide?



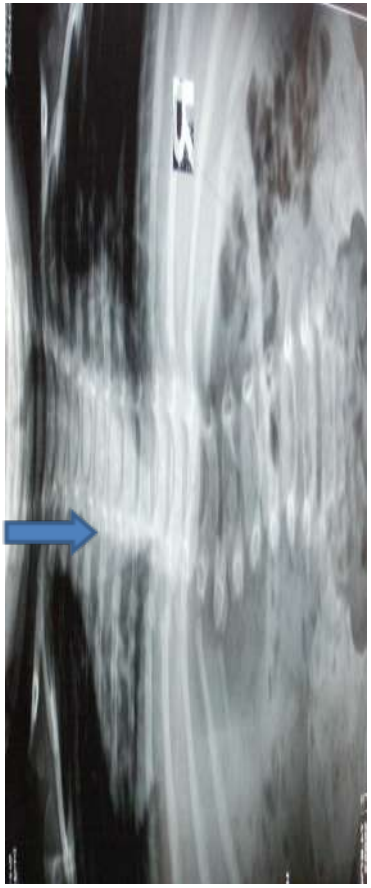


- Know the natural history
- Overall health / prognosis
- Size / stature of the child
- Walker ?
- Associated overt anomalies / syndromes
- MRI
- USG KUB, 2D Echo
- Document progress

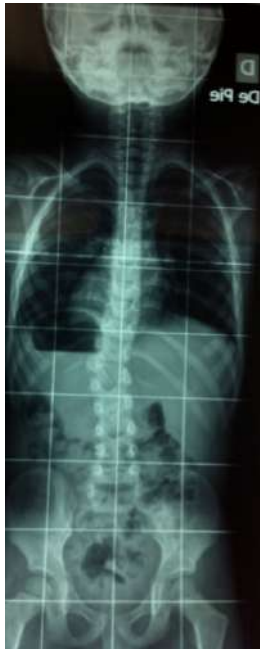
Management



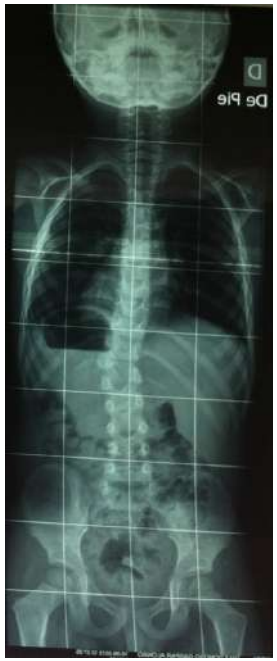
DIAGNOSIS?



2010



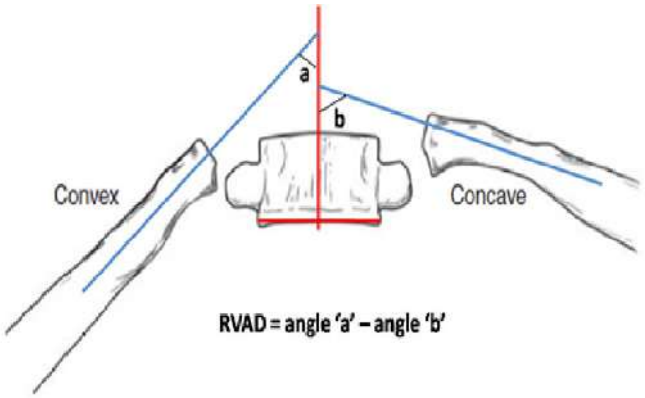
After 2 years



After 3 years

Observation

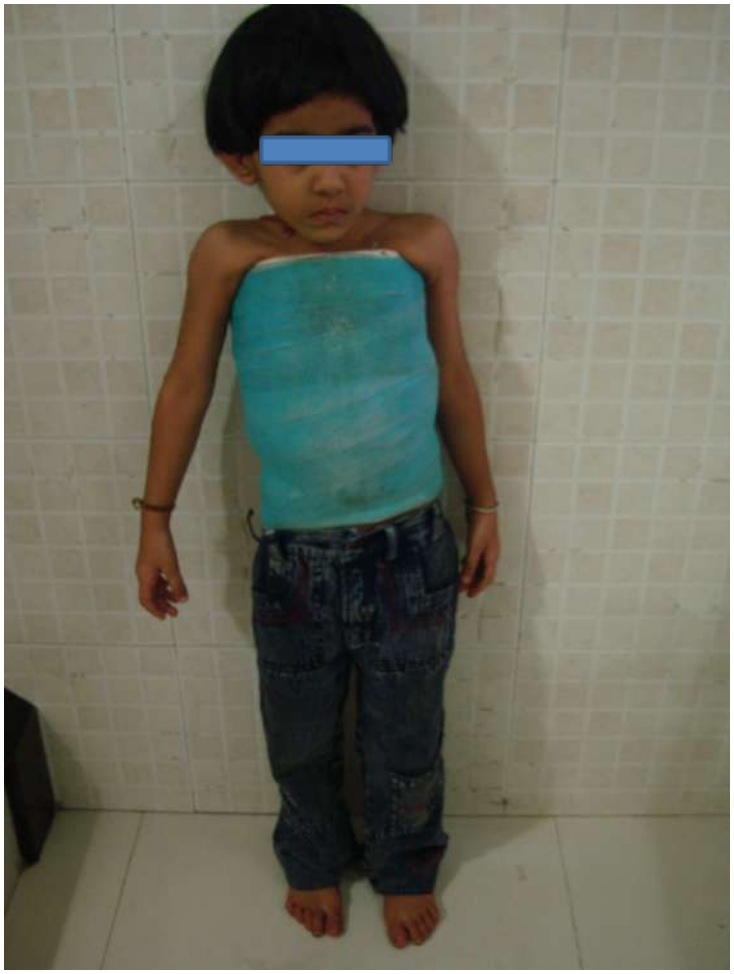
- Cobbs < 25
- RVAD < 20



- Serial radiographs : 6 monthly
- Progression < 5
- Cont to observe

If not go to the next step : Casting

Casting



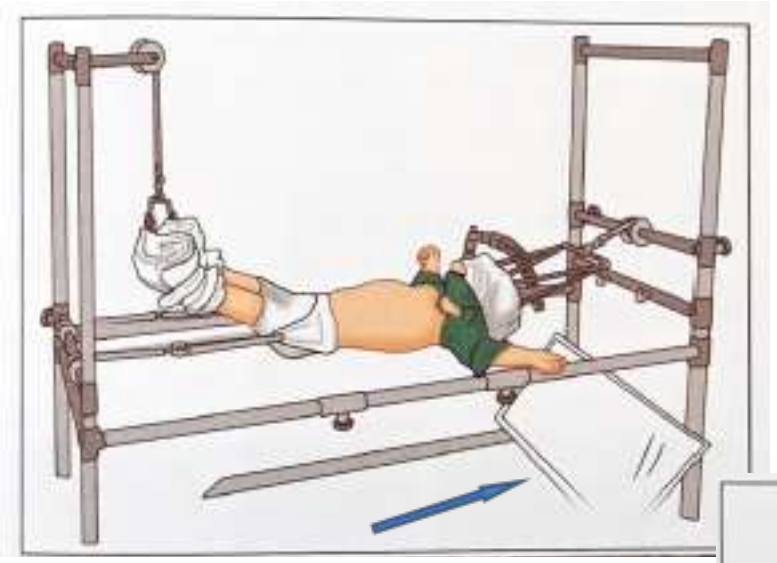
Casting

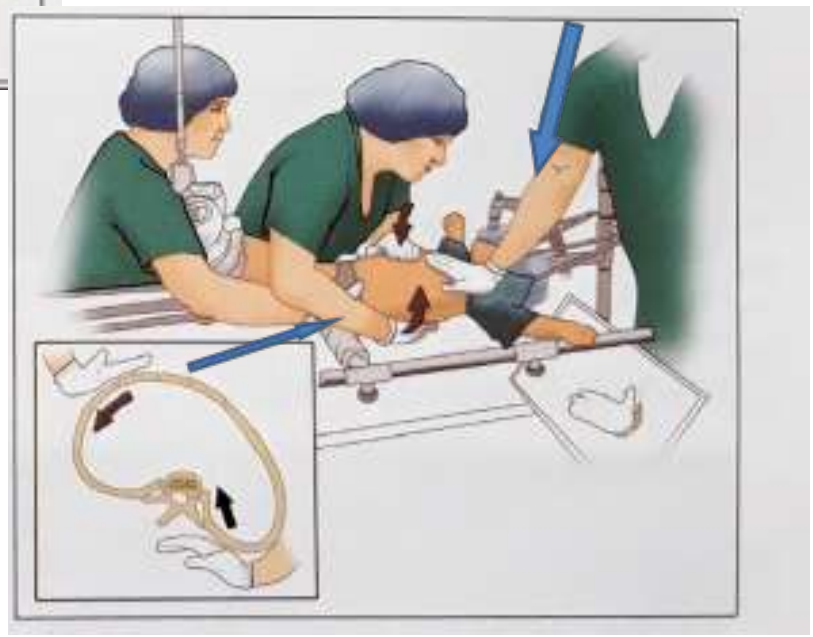
- Cobbs > 35
- RAVD >20

- Can start as early as 1 years (when the skin can tolerate cast)
- Done under GA (better muscle relaxation, better tolerated by patients)

Casting

- Risser developed first cast but no roational correction
- Cotrel and Morel – EDF (elongation, derotation & flexion cast)
- Mehta casting done by Dr Min Mehta popularized the EDF technique
- Limited for apex above T9





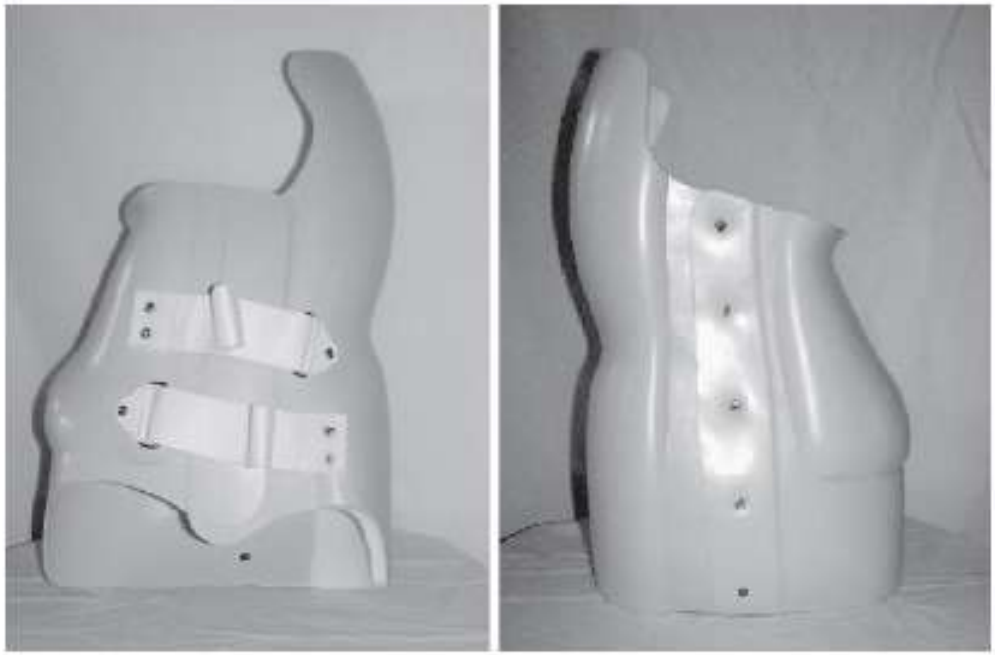
Bracing

- Done as a weaning method
- Shift from cast to brace with 1-2 hr of brace free period
- Later shift to night bracing (Providence Nocturnal Scoliosis system)

Boston Brace :
TLSO
Worn full day

Fig. 18.25 Milwaukee brace.

Milwaukee Brace :
From neck to pelvis
Best for curves above T8

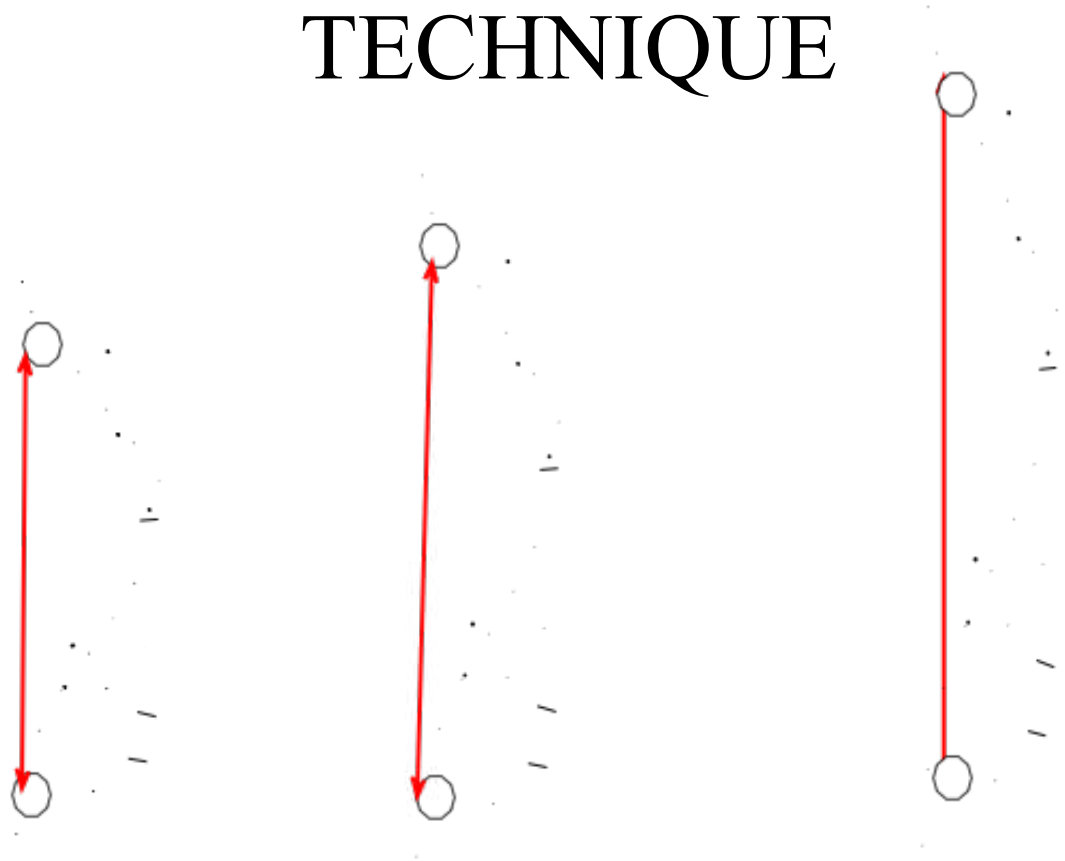


Charlston Brace :
Designed for night wear as a
weaning off brace

Growth Guided procedure

- Distration based: Growth rods /VEPTERS / MAGEC rods
- Compression based : VBT / VBS
- Growth guided technique – Shilla technique

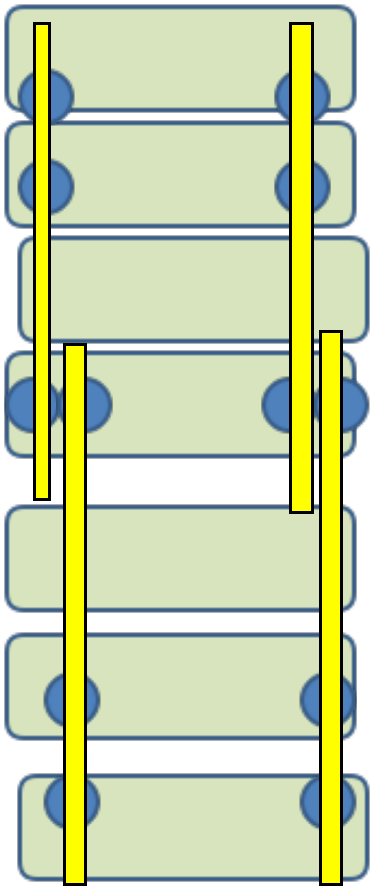
DISTRACTION BASED TECHNIQUE



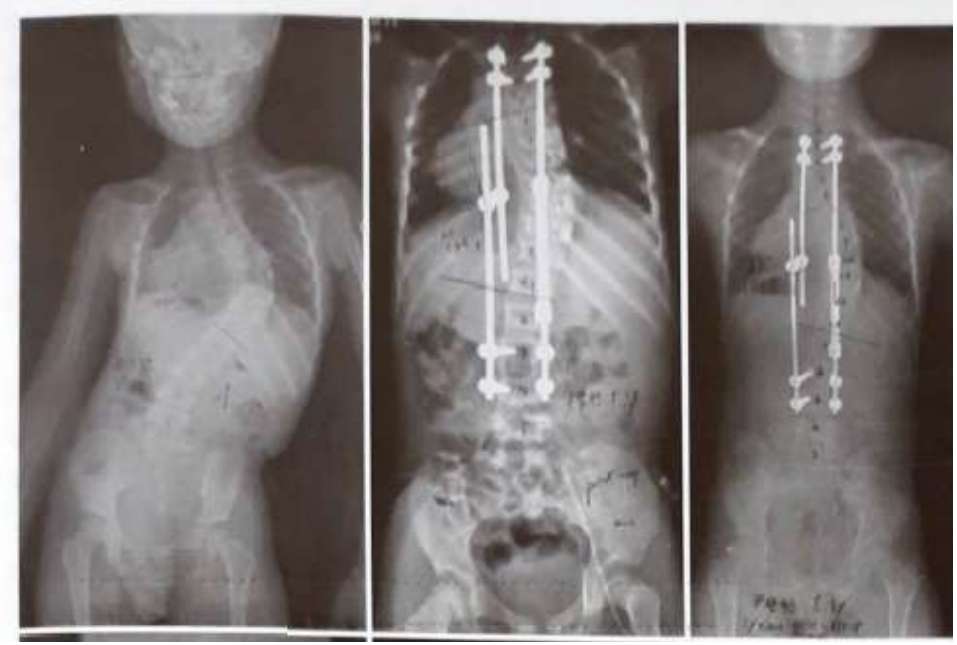
Convetional Growth Rods.

- Curve >60 degree
- Age less than 10 years
- Flexible curve
- All etiology
- Screw and rod contruct
- Better hold then hooks
- Can be from pedicle to vertebra,

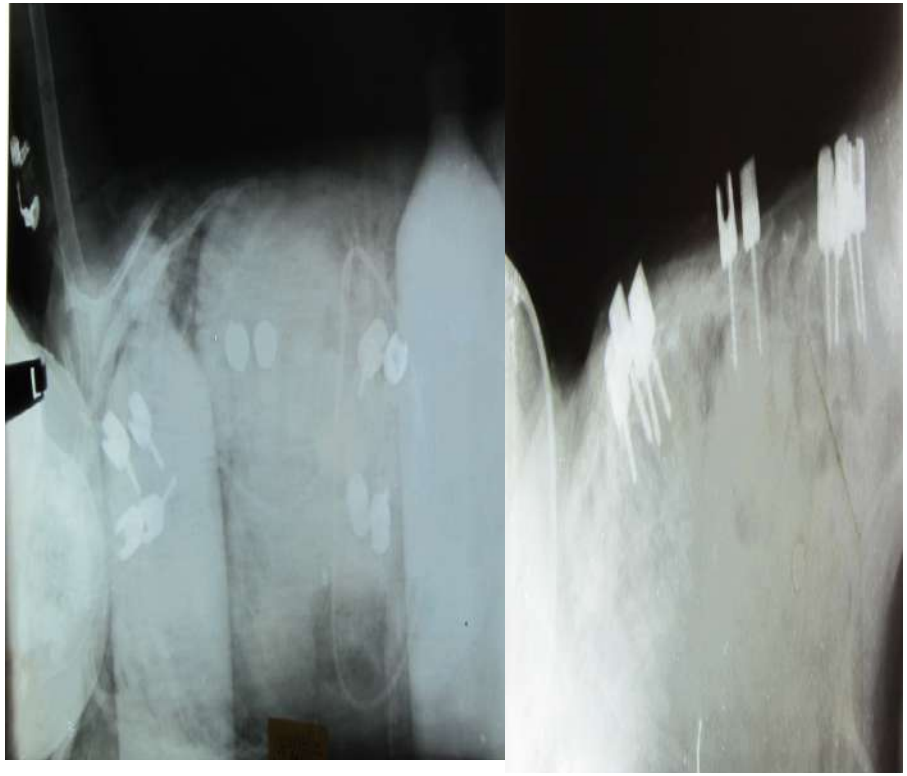
Or rib or pelvos



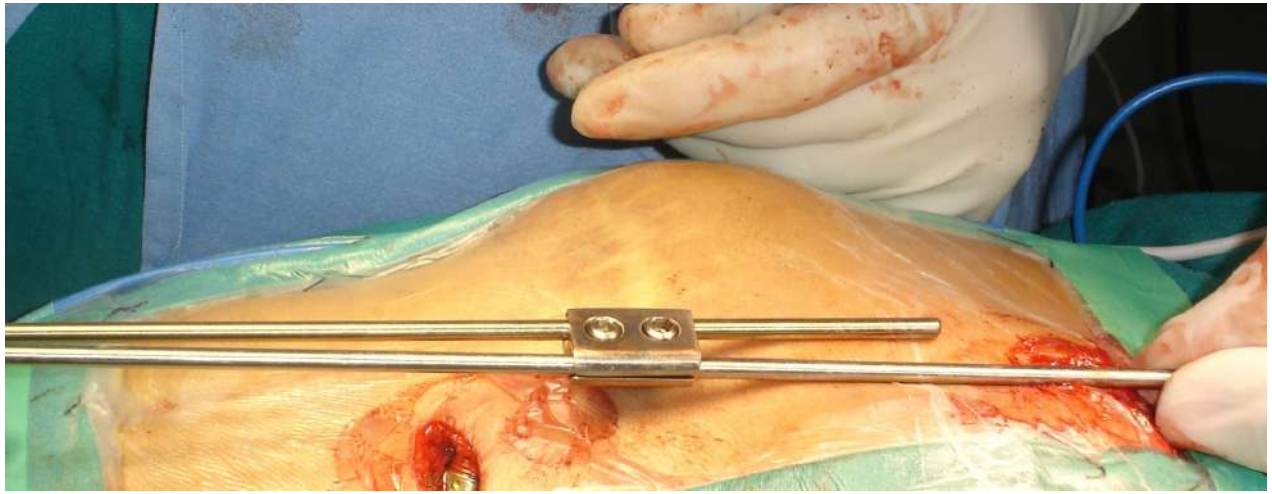
Convetional Growth Rods.

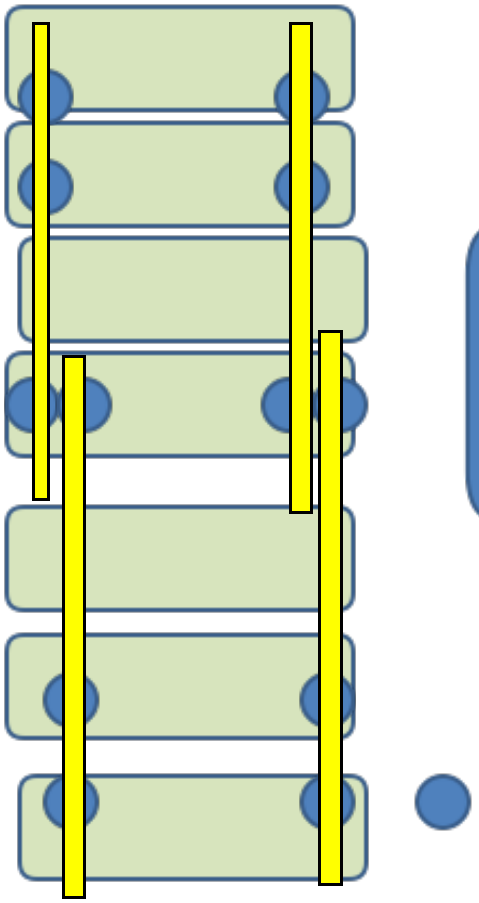


3.5mm poly axial screws, 4mm rods



‘domino connector’





Go in and distract at the apex

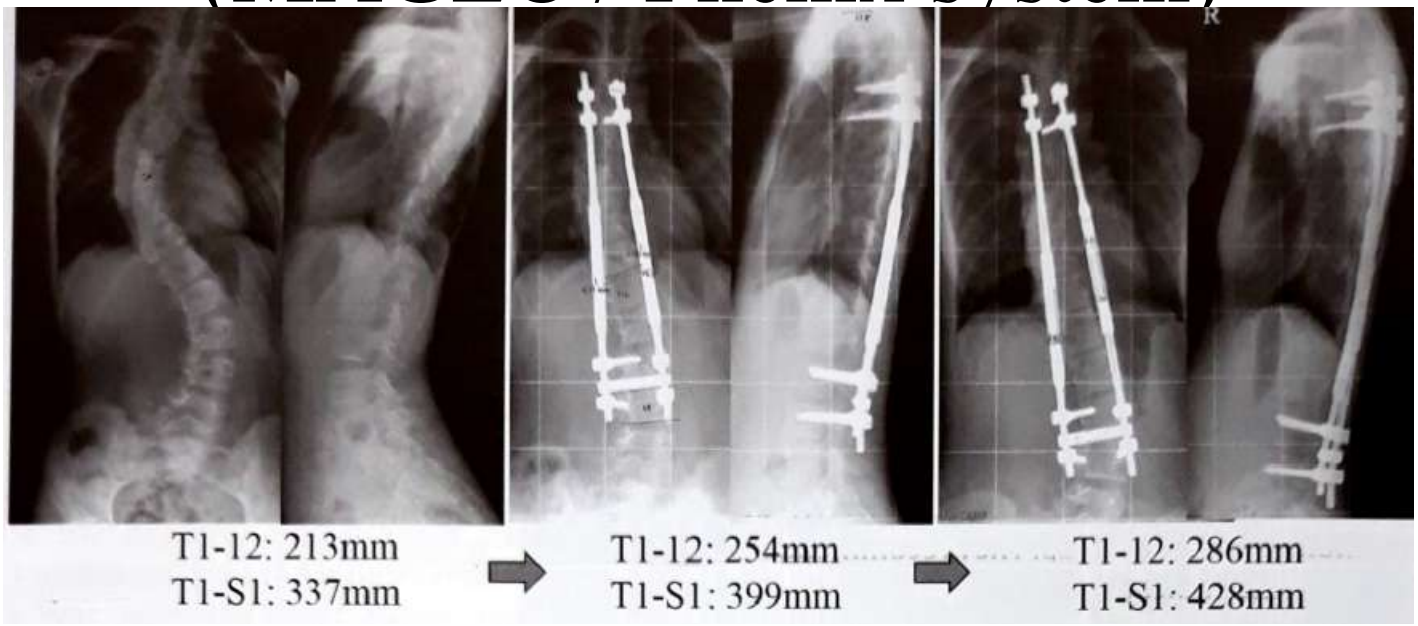
- Dual rods better than single rod
- Better purchase
- Better correction
- Less failure

- Every 6 monthly distraction
- **Law of diminishing returns**

Disadvantage:

- Each time patient need to undego anesthesia
- Implant failure
- Not a good idea in patients with kyphosis (can be a contraindication of GR)
- Insufficient chest wall expansion

MCGR – Magnetic Controlled GY (MAGEC / Phenix system)



- Usg guided distraction
- Use in dillema due to metallosis, implant failure

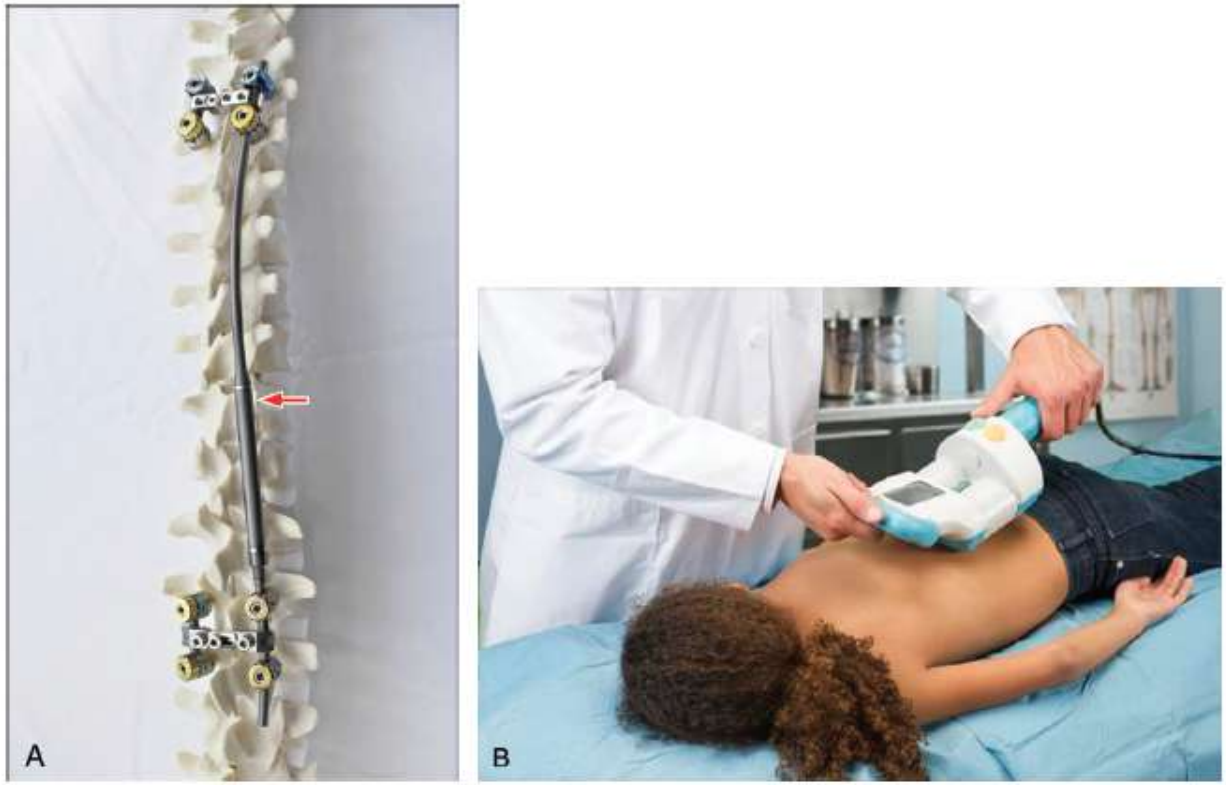


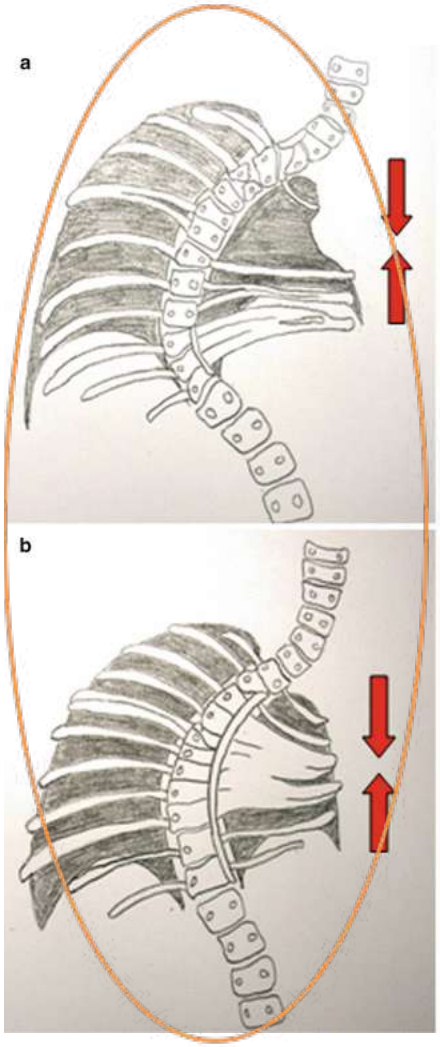
Fig. 37.6 (A) A single magnetically controlled growing rod depicted on a spine model. The enlarged midportion of the rod contains the magnetically controlled distraction mechanism *(red arrow)*. (B) Lengthening of the magnetically controlled growing rod in the office. (A: From Cheung KM, Cheung KP, Samartzis D, et al. Magnetically controlled growing rods for severe spinal curvature in young children: A prospective case series. Lancet 2012;379:1967–1974, Fig. 1, p. 1968. B: Courtesy Behrooz A. Akbarnia, MD, San Diego, CA.)

A major miss ?

One of the assicated anomaly with EOS is abnormal development of lung function.

And in all these technique there is no solution if the lung anatomy is abnormal

THORACIC INSUFFIENCY SYNDROME - A CHEST WALL ABNORMALITY WHICH INTERFERES WITH RESPIRATORY FUNCTION OF THE CHILD.

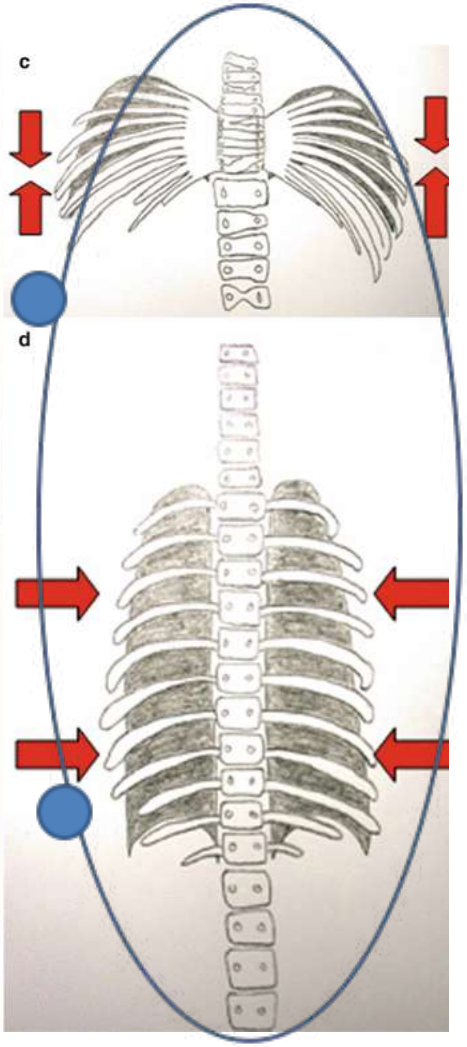


Unilateral

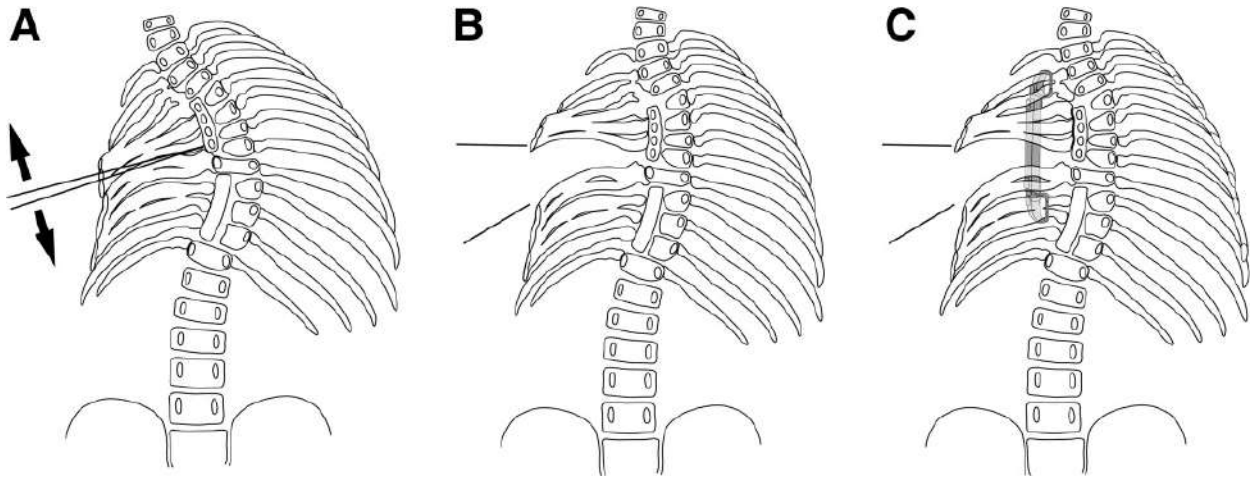
Type 1 – Absent rib and scoliosis
Type 2 – Fused ribs and scoliosis

Bilateral

Type 3a – Foreshortened thorax (Jarcho Levin Syndrome)
Type 3b – Transervese shortened thorax (Jeune syndrome)



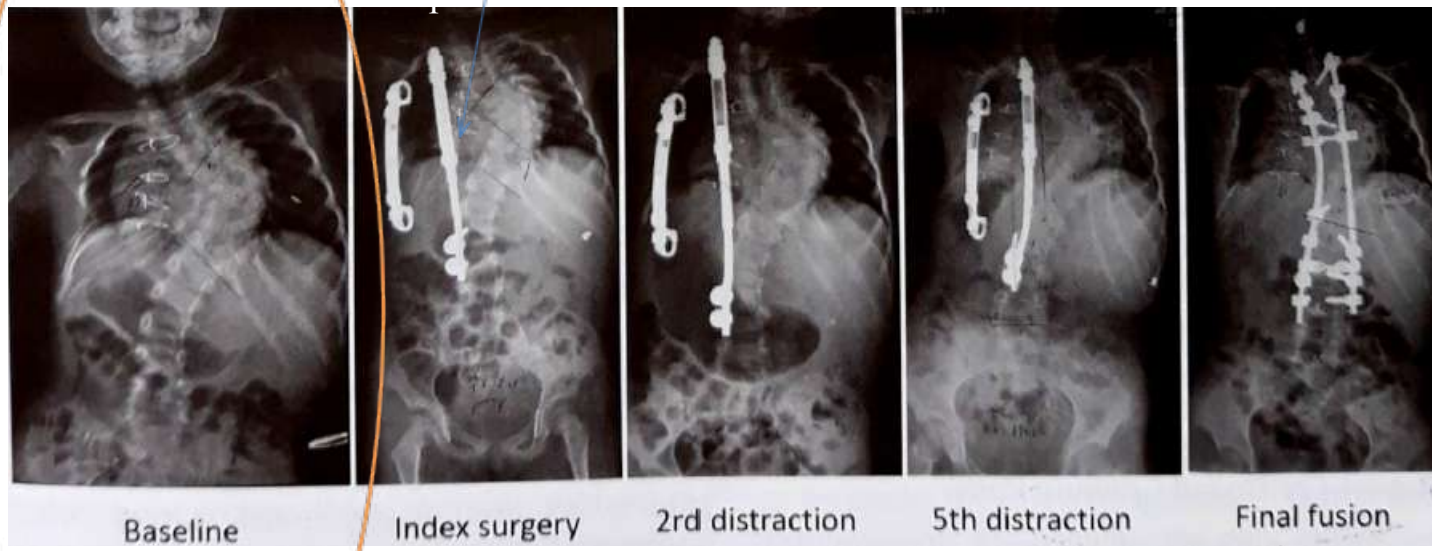
Vertically Expandible Prosthetic titanium Rib



One segment from rib-rib

Other segment from rib – spine or rib

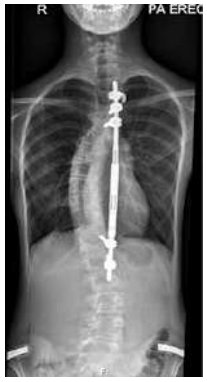
VEPTR

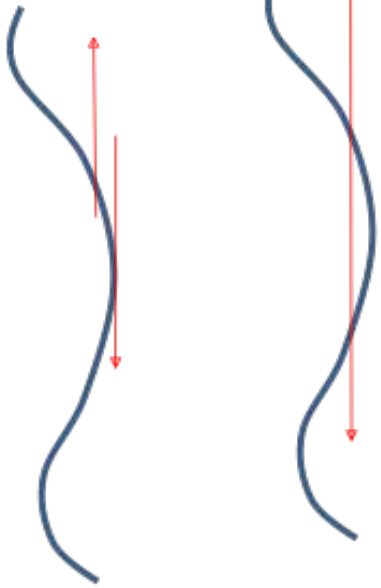


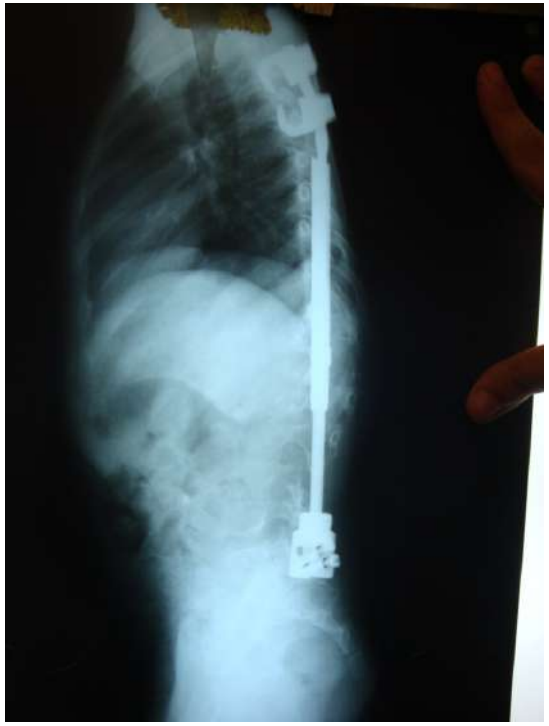
- Higher complication rates
- Implant failure
- Complications like lung injury

Not an implant of choice of routine use

Flat back syndrome



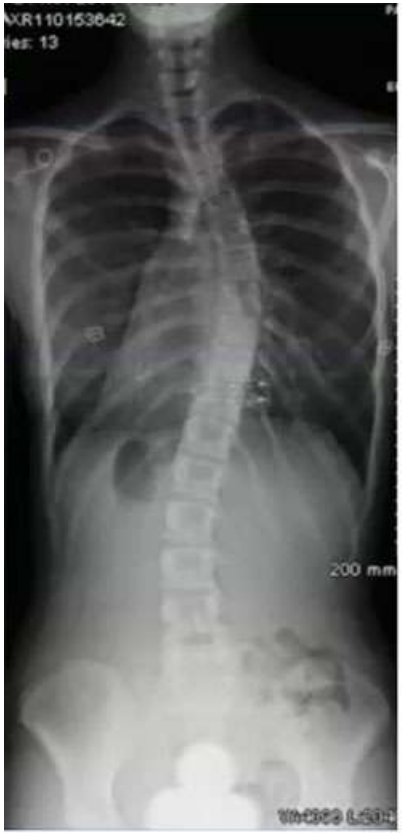


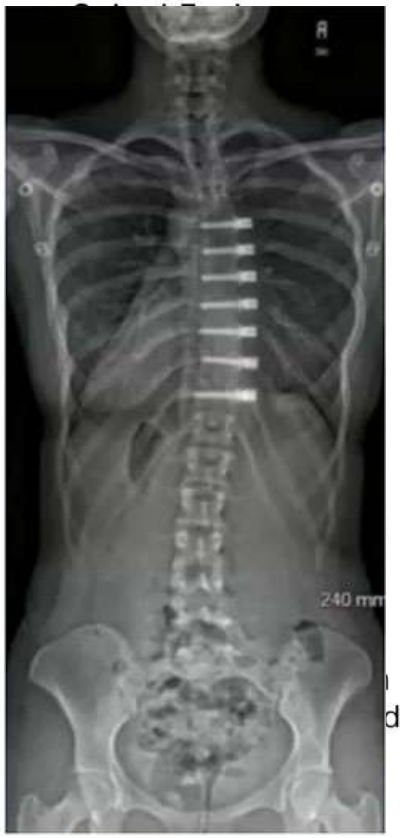


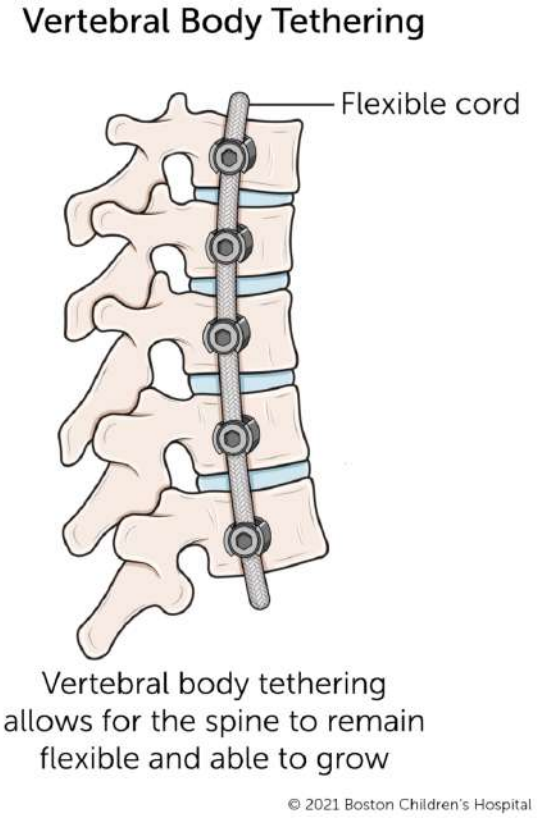
Compression Based system

- VBS / VBT
- Anterior procedures
- <13 girls/ <15 boys
- Risser 0 or 1 or 1 year of growth remaining
- Thoracic curve <35 and lumbar curve <45
- Saggital thoracic curve <40
- Skeletally immature patients

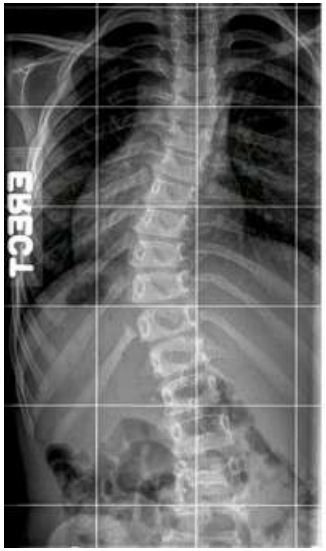
VBT

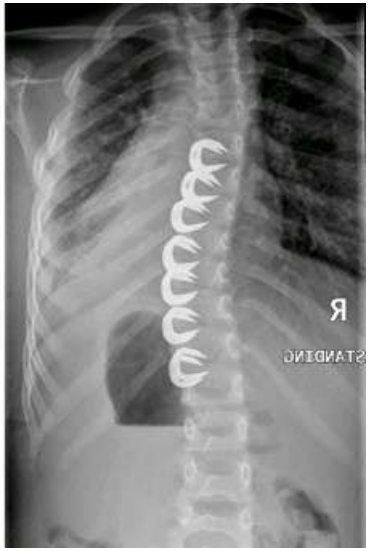






VBS





Growth Guided Technique

- Luque Trolley – rods and sublaminar wires

(fusion, implant failure, curve progression)

- Shella Technique(McCarthy)

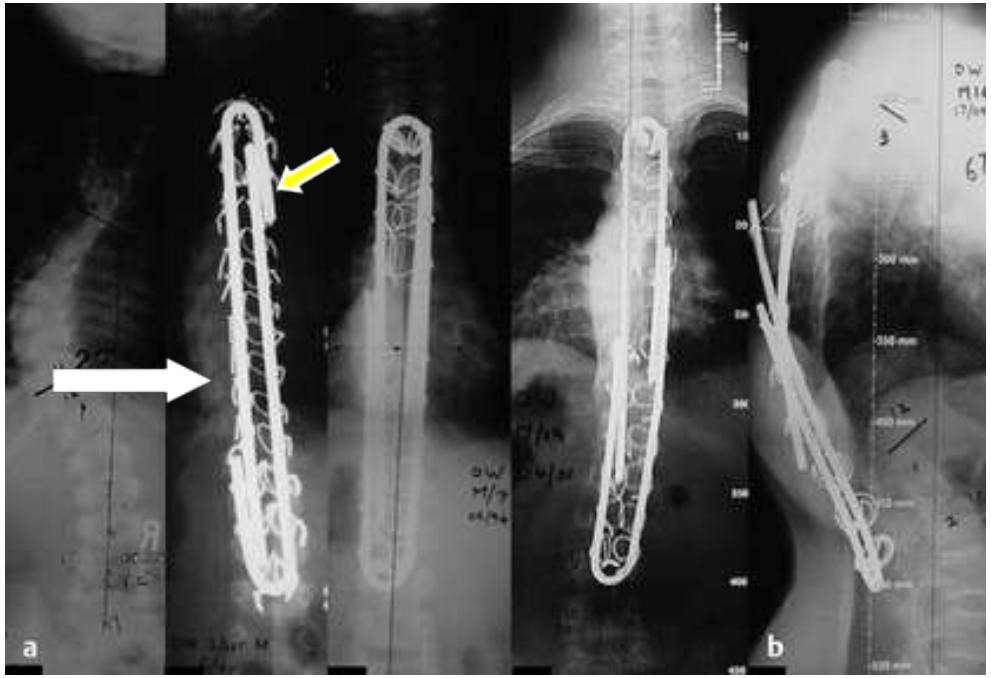
Dual growth rods

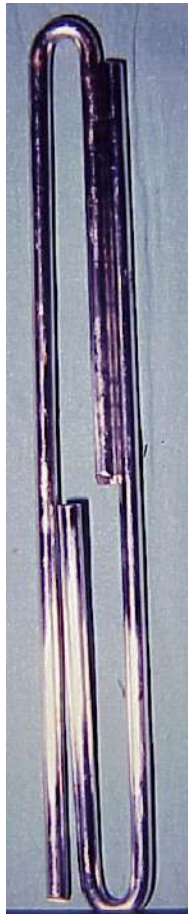
Apical fusion

And ends kept loose for rods to grow.

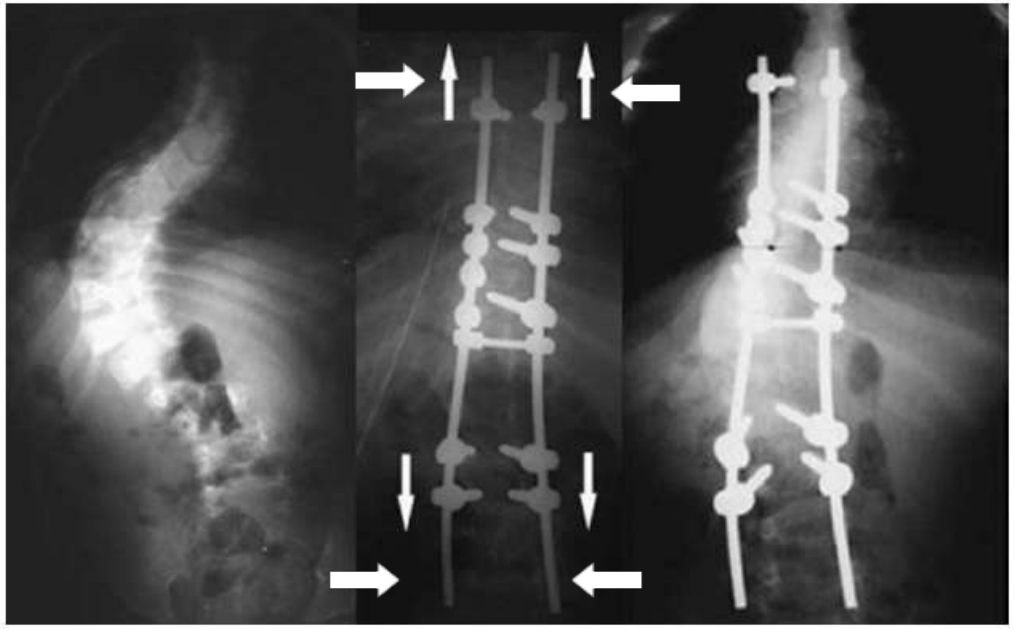
- Convex growth rod arrest

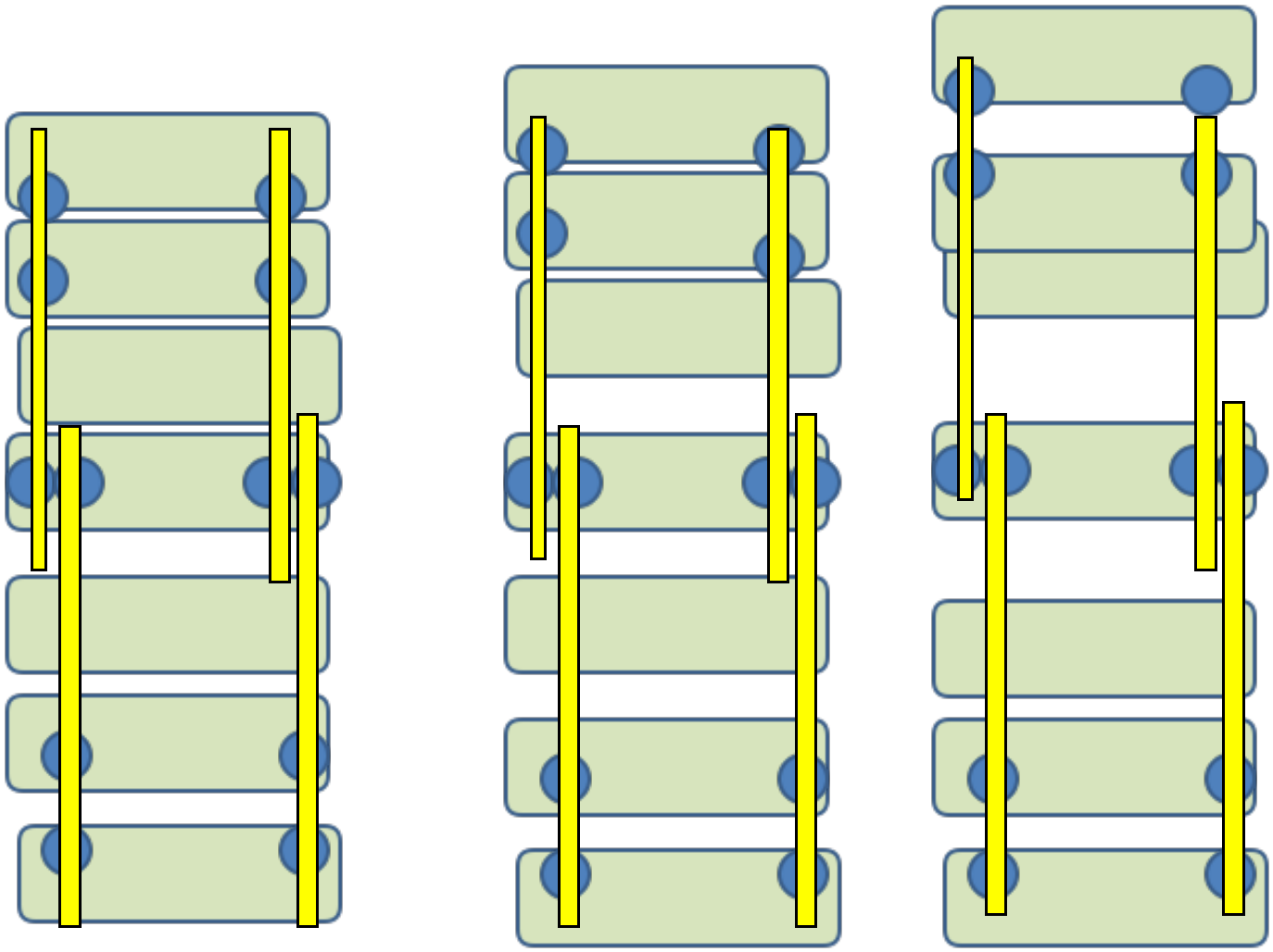
Luque trolley





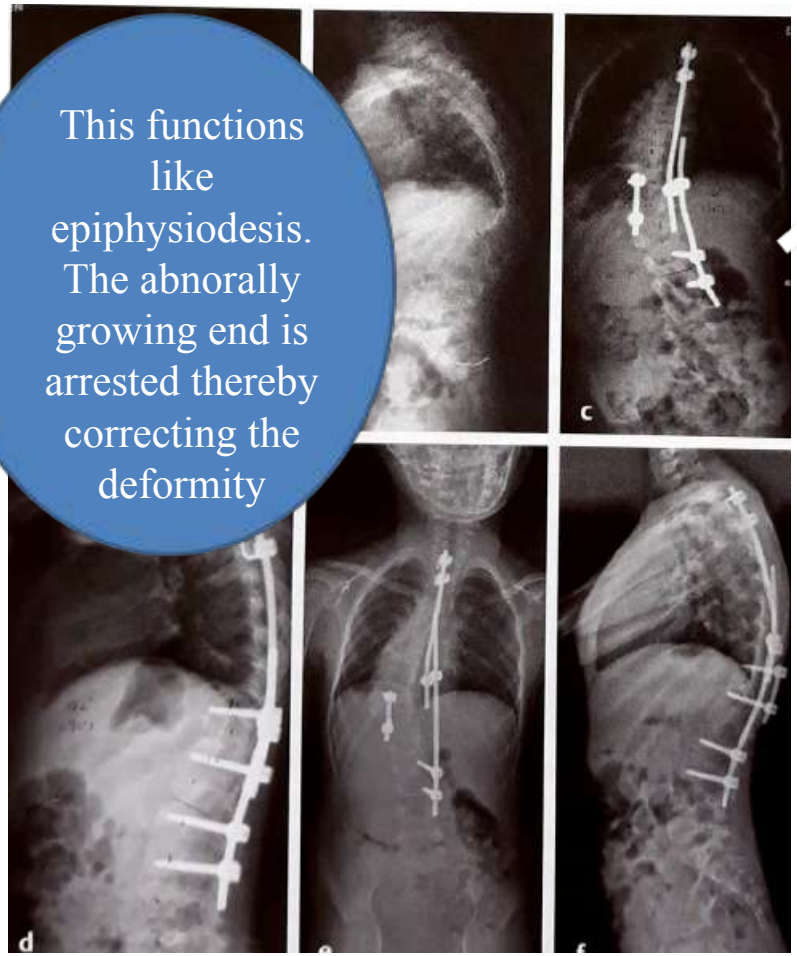
Shella Technique

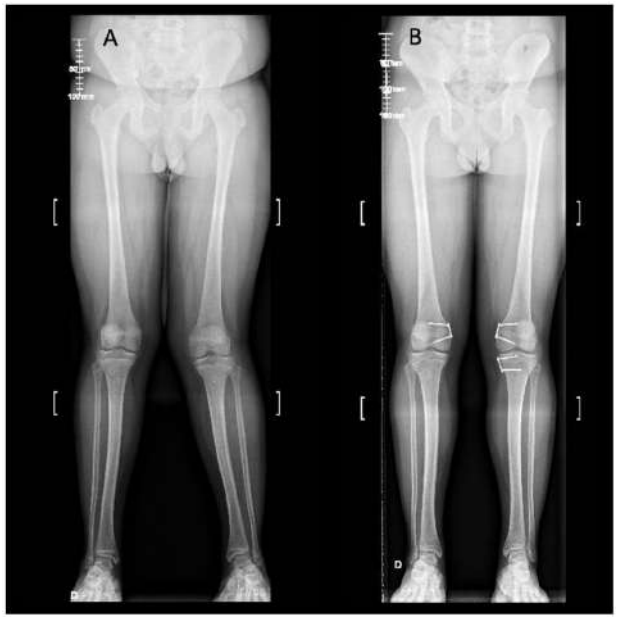




Convex Growth rod arrest

This functions like epiphysiodesis. The abnorally growing end is arrested thereby correcting the deformity

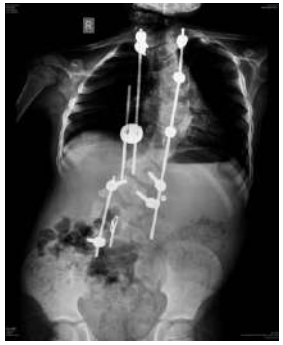




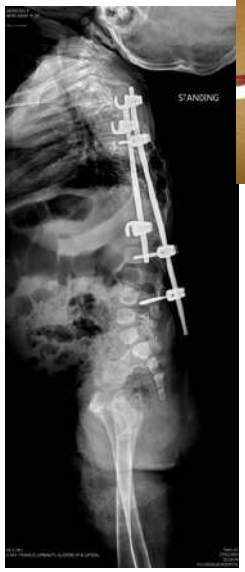
Final Fusion

- If deformity remains during the treatment of child undergoing treatment
- Preferably after 10 yrs
- After adequate pulmonary function has been achieved
- If too frequent complications with growth guided system
- Patient not fit for repeated anesthesia

Always be careful



- Almost always associated with complications
- Law of diminishing returns
- Mc is skin related complication
- Implant failures, backout
- Curve progression – careful in syndromic cuves – behave differently – Marfan, EDS, NF



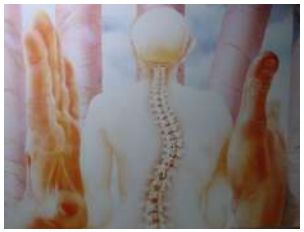
EOS

No surgery

Observe / brace

Surgery

Apical pathology



No apical pathology

Courtesy : Dr Abhay Nene

No global deformity

Global deformity

'small' child

'big' child / rigid deformity

Apical fusion

Apical fusion + Growth rod

Growth rod +/- Rib

Primary fusion