Idiopathic Scoliosis Update

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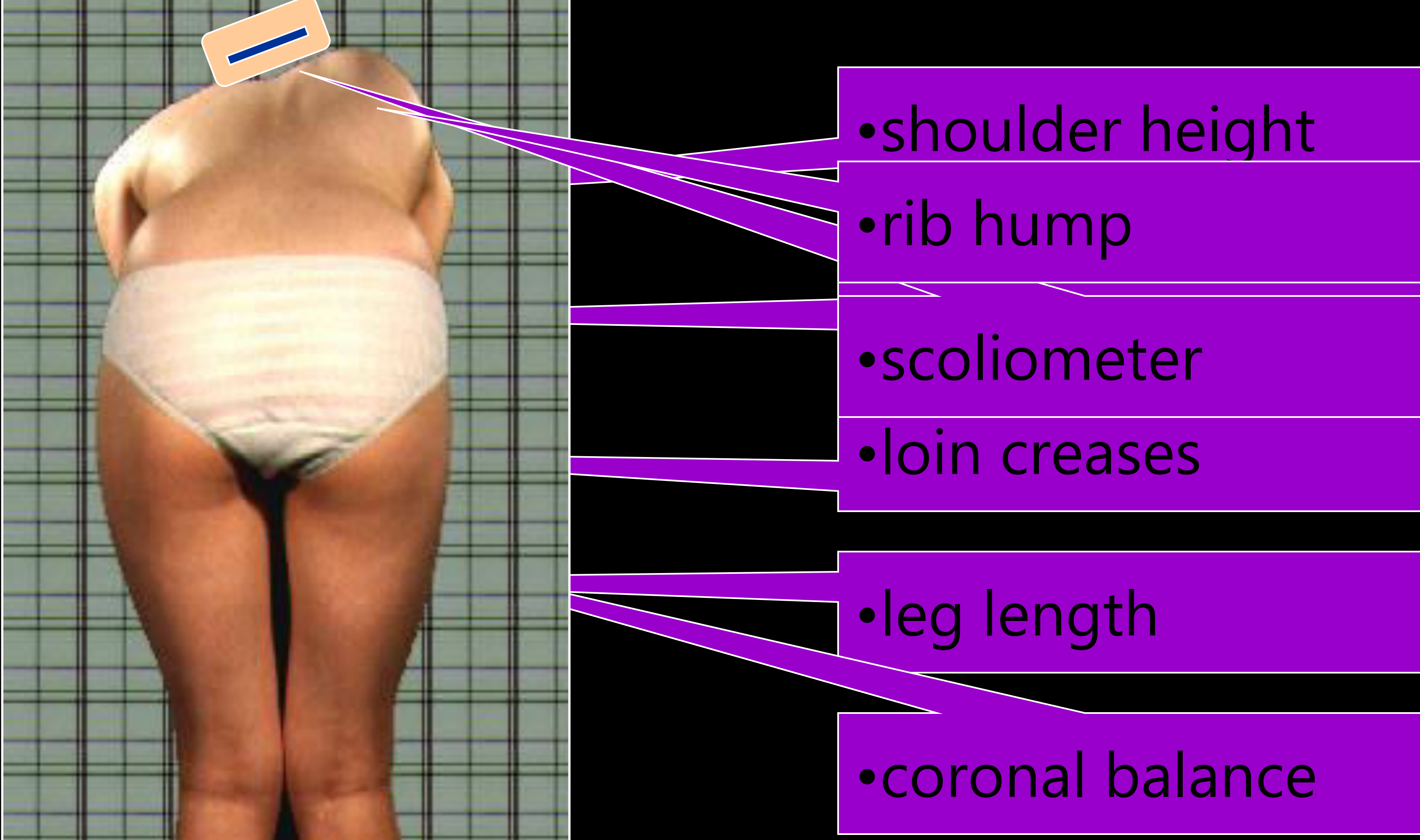
Introduction

- What is Scoliosis
- What are the causes of scoliosis
- When do we treat scoliosis

Scoliosis – coronal plane deformity

Structural vs non structural

Lateral curvature of the spine exceeding 10 deg.

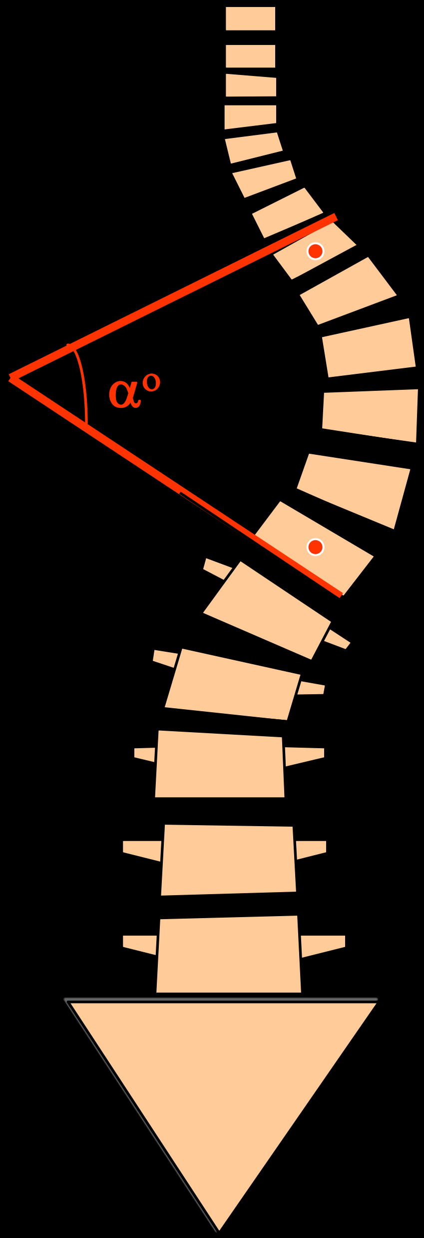


Clinical Features

Radiological features

Lateral curvature

Vertebral rotation



- identify curve(s)

- choose end vertebrae
  - most tilted from horizontal or last to converge
  - upper end plate upper

- measure angle between them
  - lines along endplates
  - superior endplate of cranial vertebra
  - inferior endplate of caudal vertebra

INCIDENCE

overall 25 per 1000
10 º-19º 23 per 1000
20 º-29º 5 per 1000
30 º-39º 2 per 1000
40 º + 1 per 1000

females > males
10 º-19º F:M 2:1
20 º-29º F:M 5:1
30 º-39º F:M 10:1
40 º + F:M 20:1

Effects of scoliosis

- Deformity
- Back Pain
- Cardiopulmonary function
- Loss of seating balance

MRI

- Neurology
- Excessive kyphosis
- Early onset
- Rapid progression
- Associated syndromes
- Left thoracic/ thoraco-lumbar curves

Scoliosis Aetiology

- Idiopathic
- Neuromuscular
- Congenital
- Syndromic

Idiopathic Scoliosis

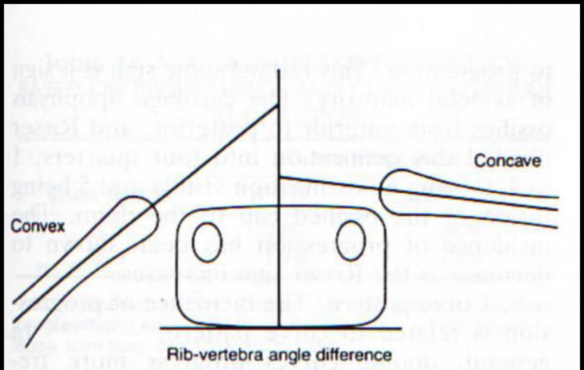
- Early onset
- Late onset

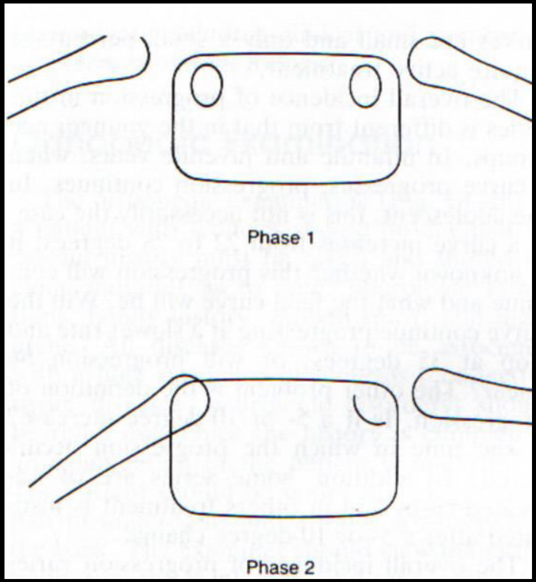
- Infantile
- Juvenile
- Adolescent

Early onset idiopathic scoliosis

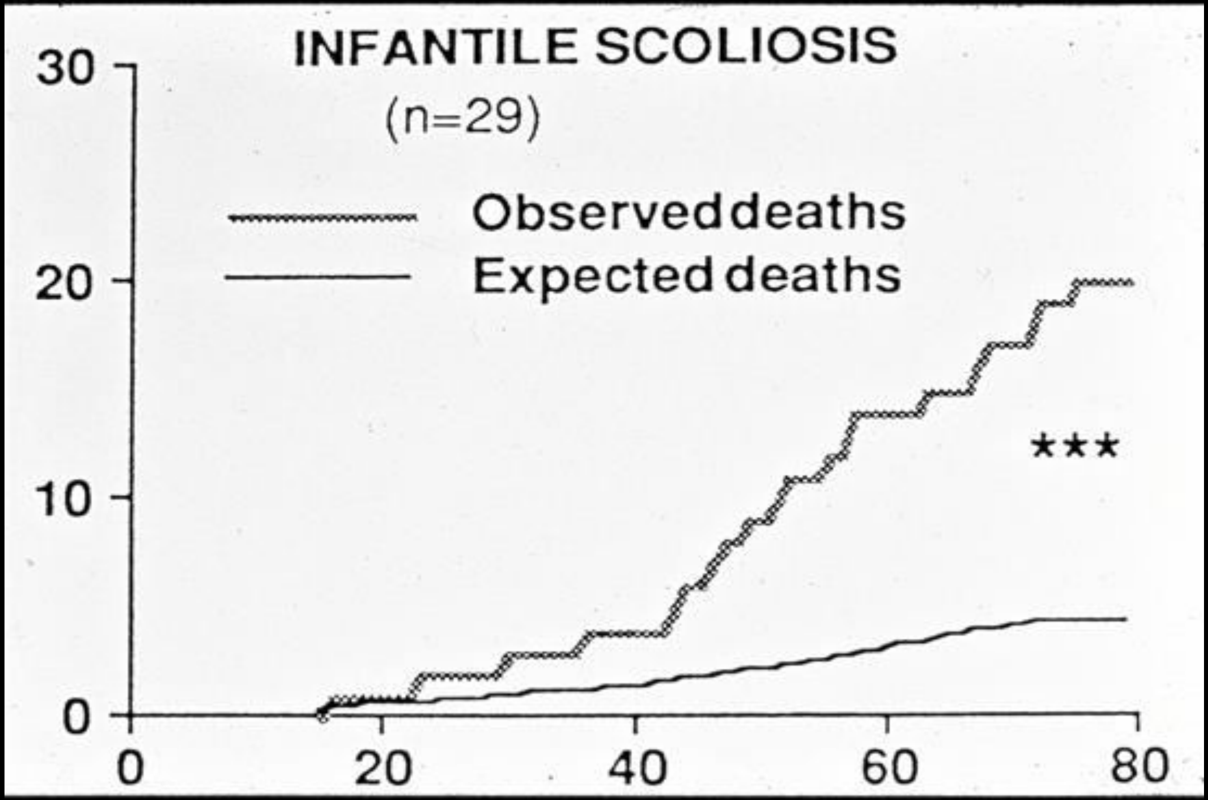
Infantile Idiopathic

- Different to adolescent type
- 60% boys, 75% left thoracic curves
- Girls with right curves worst outlook
- Only 10% progressive



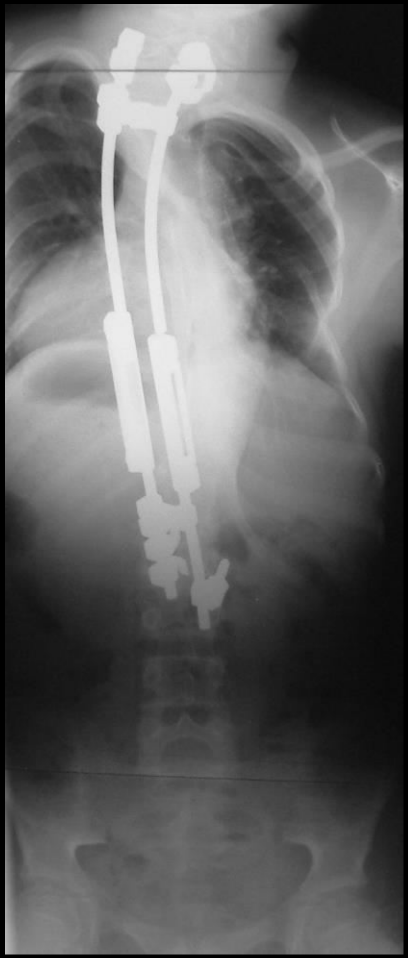


Early onset Scoliosis



EO scoliosis management

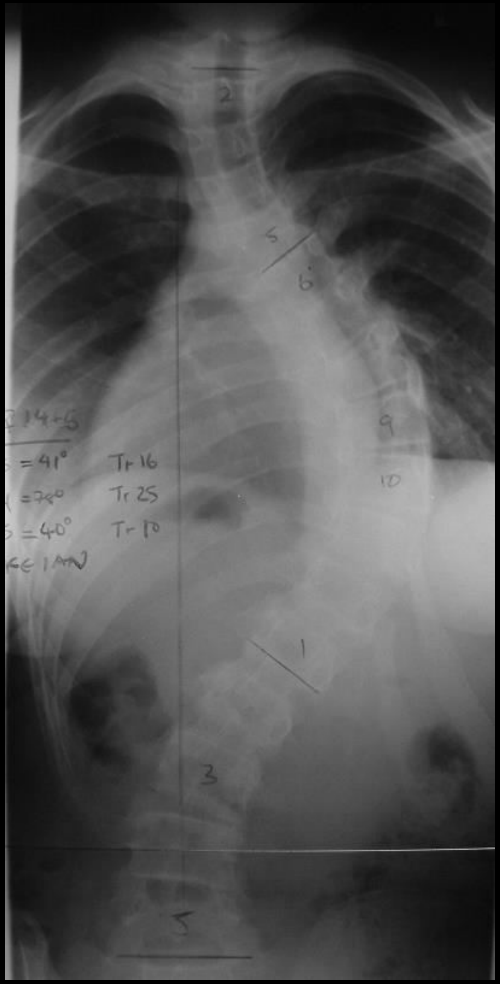
- Serial plasters
- Bracing
- Growing rods



Juvenile Idiopathic

- Girls with right thoracic curves predominate
- 70% need some form of treatment
- 50% are braced successfully
- Delay to surgery to allow growth allowable to 60 deg curve

Late Onset Idiopathic Scoliosis



Mainly girls

Probably genetic

Often painless

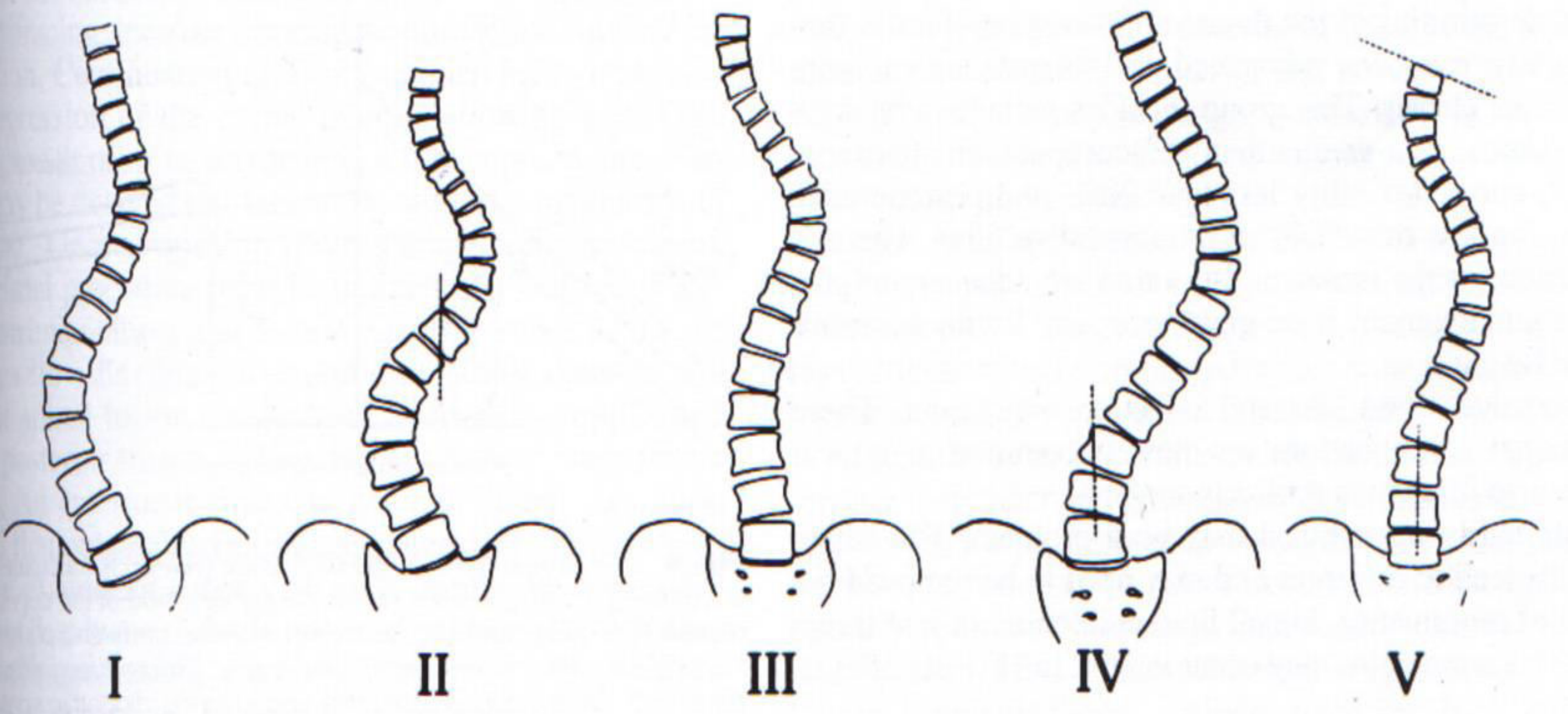
Curve types

Thoracic scoliosis

Lumbar scoliosis

Adolescent idiopathic

King-Moe Classification



Curve Type

Type	Proximal Thoracic	Main Thoracic	Thoracolumbar / Lumbar	Curve Type
1	Non-Structural	Structural (Major*)	Non-Structural	Main Thoracic (MT)
2	Structural	Structural (Major*)	Non-Structural	Double Thoracic (DT)
3	Non-Structural	Structural (Major*)	Structural	Double Major (DM)
4	Structural	Structural (Major*)	Structural	Triple Major (TM)
5	Non-Structural	Non-Structural	Structural (Major*)	Thoracolumbar / Lumbar (TL/L)
6	Non-Structural	Structural	Structural (Major*)	Thoracolumbar / Lumbar - Main Thoracic (TL/L - MT)

*Major = Largest Cobb Measurement, always structural
Minor = all other curves with structural criteria applied

STRUCTURAL CRITERIA
(Minor Curves)

Proximal Thoracic: - Side Bending Cobb ≥ 25°
- T2 - T5 Kyphosis ≥ +20°

Main Thoracic: - Side Bending Cobb ≥ 25°
- T10 - L2 Kyphosis ≥ +20°

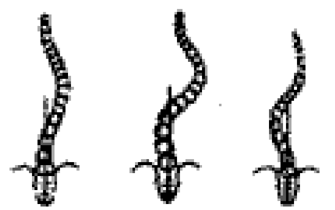
Thoracolumbar / Lumbar: - Side Bending Cobb ≥ 25°
- T10 - L2 Kyphosis ≥ +20°

LOCATION OF APEX
(SRS definition)

CURVE	APEX
THORACIC	T2 - T11-12 DISC
THORACOLUMBAR	T12 - L1
LUMBAR	L1-2 DISC - L4

Modifiers

Lumbar Spine Modifier	CSVL to Lumbar Apex
A	CSVL Between Pedicles
B	CSVL Touches Apical Body(ies)
C	CSVL Completely Medial



A B C

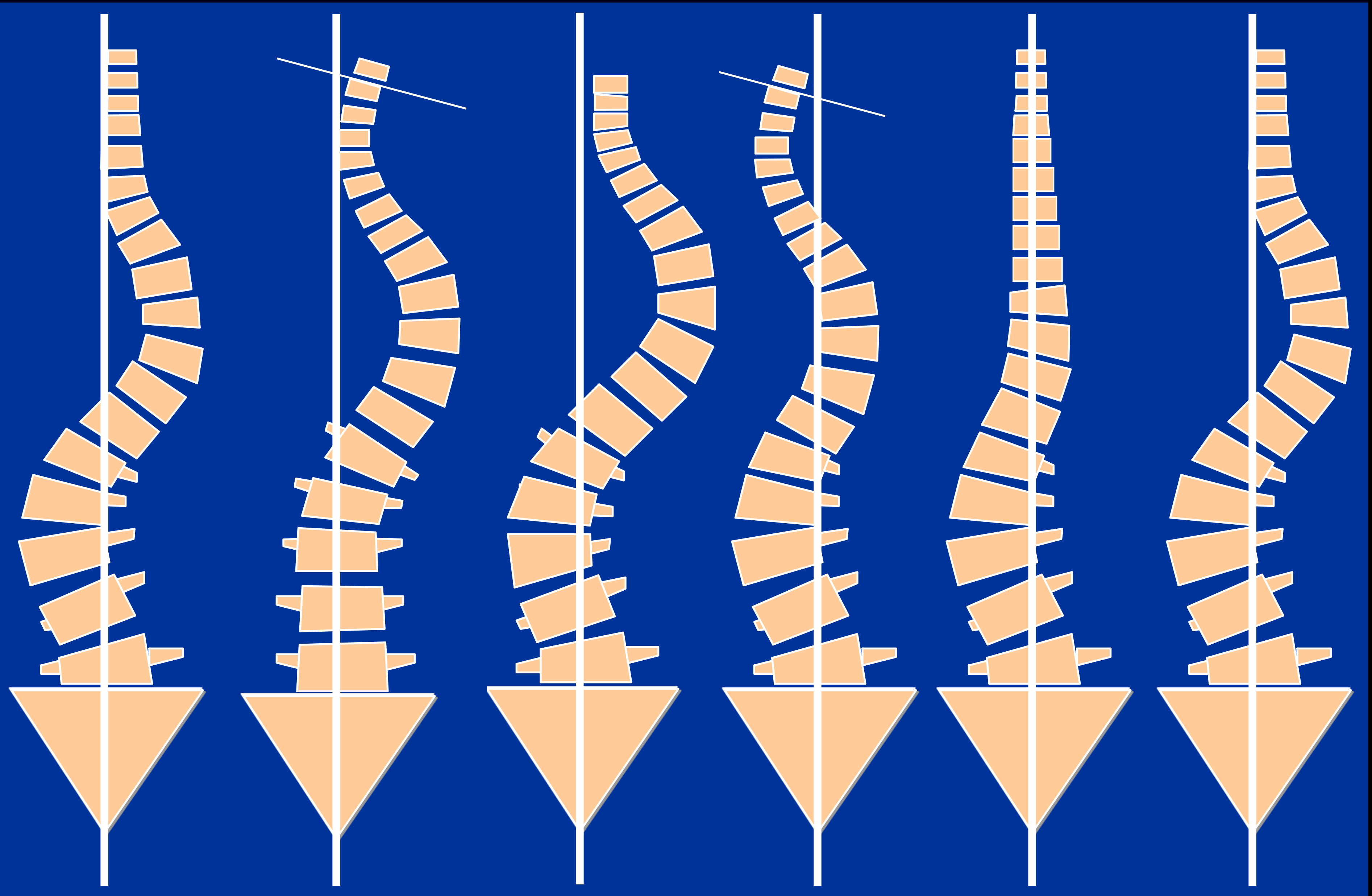
Thoracic Sagittal Profile T5 - T12	
– (Hypo)	< 10°
N (Normal)	10° - 40°
+ (Hyper)	> 40°

Curve Type (1-6) + Lumbar Spine Modifier (A, B, or C) + Thoracic Sagittal Modifier (-, N, or +)
Classification (e.g. 1B+):__________

Fig. 2 Synopsis of all necessary criteria for curve classification. SRS = Scoliosis Research Society, and CSVL = center sacral vertical line.

J Bone Joint Surg [Am] 2001; 83-A; 1169-81

SRS - LENKE



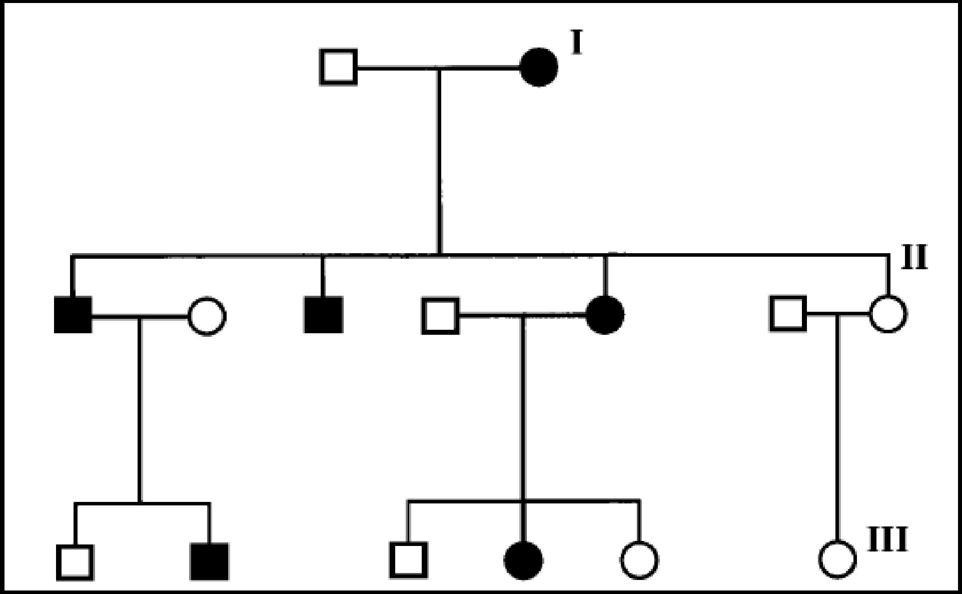
Genetics update

2 types of genetic diseases

- Single gene (Mendelian):
  - Rare diseases…….runs in families
  - GENETIC + environmental
- Complex trait / diseases:
  - Common
  - Familial inheritance not obvious
  - Polygenic
  - GENETIC + ENVIRONMENTAL

Genetic inheritance patterns

- Dominant-, multi-gene
  Wynne-Davies JBJS B 1968
- Dominant-, x-linked (paucity of male to male transmission)
  Cowell Clin Orthop 1972
- Multi-factorial: ↓ frequency 1st (11.1%) ---- 3rd degree (1.4%)



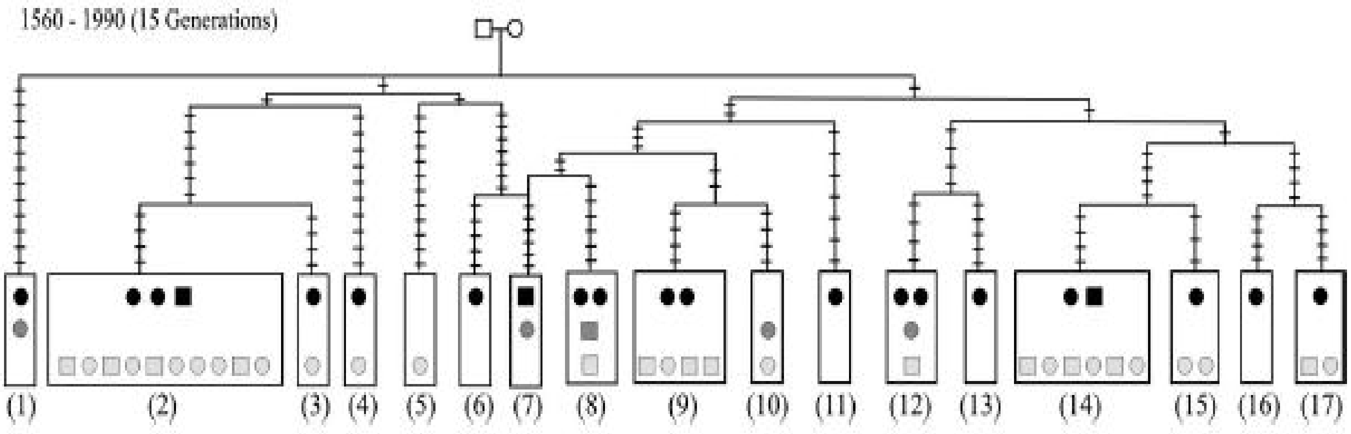
Family history

- Faber (1935): 600 AIS
  - 14% parents; 7% siblings
- Wynne-Davies (1968): 180 AIS
  - 25% in family members
- Riseborough, Wynne-Davies (1973): 207 AIS
  - 1st deg 2nd deg 3rd deg
  - Brother 7%; Sister 42%

Founder effect:

- 145 AIS probands
- Family history & Genealogy records
- 97% connectedness (major scoliosis gene)
- 70% connected with families in England
  (Essex, 1520 AD; Kent 1560 AD)

Figure 1. Pedigree of 17 scoliosis families connected to one founder in Kent, England circa 1560. The GenDB database uses unique identification numbers to identify relationships to other participating scoliosis families.



Twin studies

- 37 monozygotic 73% concordance
- 31 dizygotic 36% concordance
- Curve patterns in monozygotic r = 0.399
- Lack of 100% concordance:
  - Older studies (miss-diagnosis)
  - Differences in intra-uterine environment
  - Uneven clevage
  - Differences in external environment
  - Mosaic genotype (not completely identical)

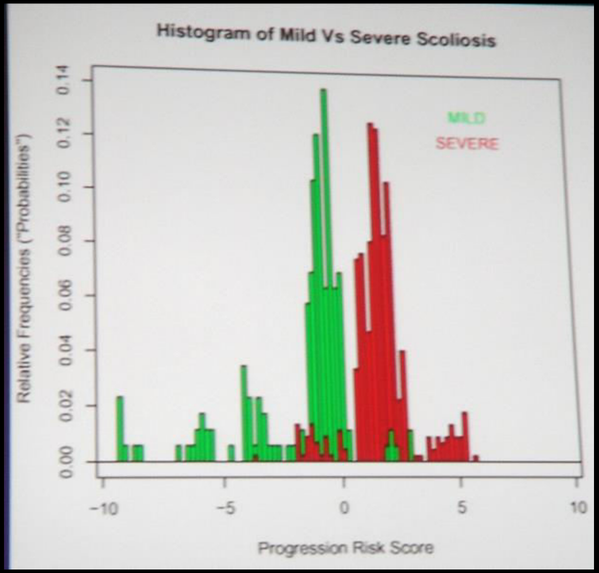
12 DNA markers to assess progression risk

Braun et al SRS 2007

- 118 AIS + 125 controls (Utah)
  675 AIS (US) .....454 severe!

- Whole genome scan (blood and saliva)
  Affymetric 100 K genechip

- 12 markers:
  - Sensitivity
  - Specificity
  - Odds ratio



Progression risk score: $p < 2.2 \times 10^{-16}$

Minneapolis school study: Lonstein JBJS 1982

1.5 million children screened

↓

3.4% (51,000) referred for X ray evaluation

↓ ↓

1.2% (18,000) diagnosed (> 10°)

2.2% (33,000) < 10°

↓ ↓

0.1% (1,500) Braced

0.01% (150) Fused

Minneapolis school study: Lonstein JBJS 1982

1.5 million children screened

Genetic testing ⇩

1.2%

~~3.4% (51,000)~~ referred for X ray evaluation

⇩

~~1.2% (18,000) diagnosed (> 10°)~~

⇩

0.11% referred for treatment

Prognostic factors

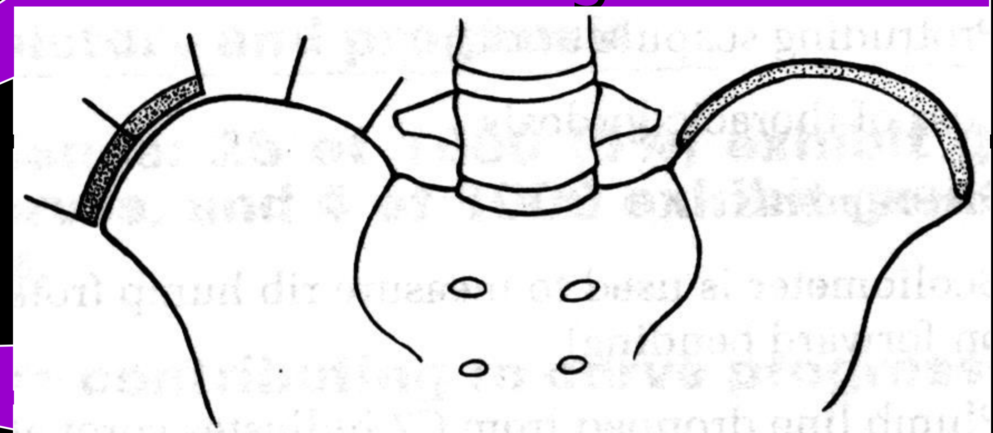
growth potential
- age
- menarche
- Risser sign

sex

curve severity

curve pattern

- risk of progression >10°
  - three times greater



to

progression factor	risk
1	20%
1.5	50%
2	80%

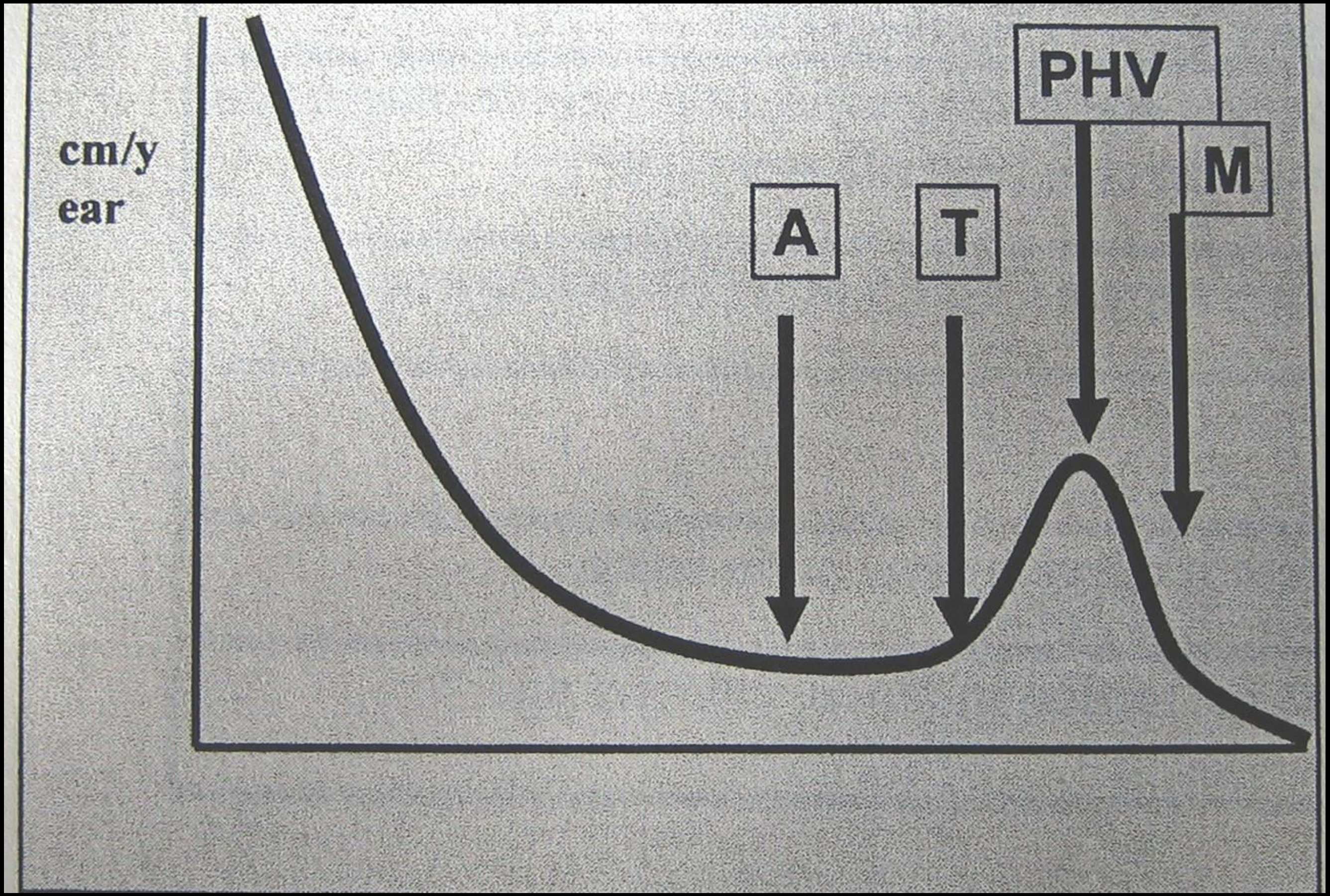
- thoracic curves more likely to progress than lumbar curves

Maturity assessment update

Peak height growth velocity (PHV)

- Infants rapid
- 7yrs - 5cms/yr
- Puberty – 8.3cms/yr Growth spurt spans 2 yrs, 1 yr before and after the peak with some continued growth 2-3 yrs after the peak

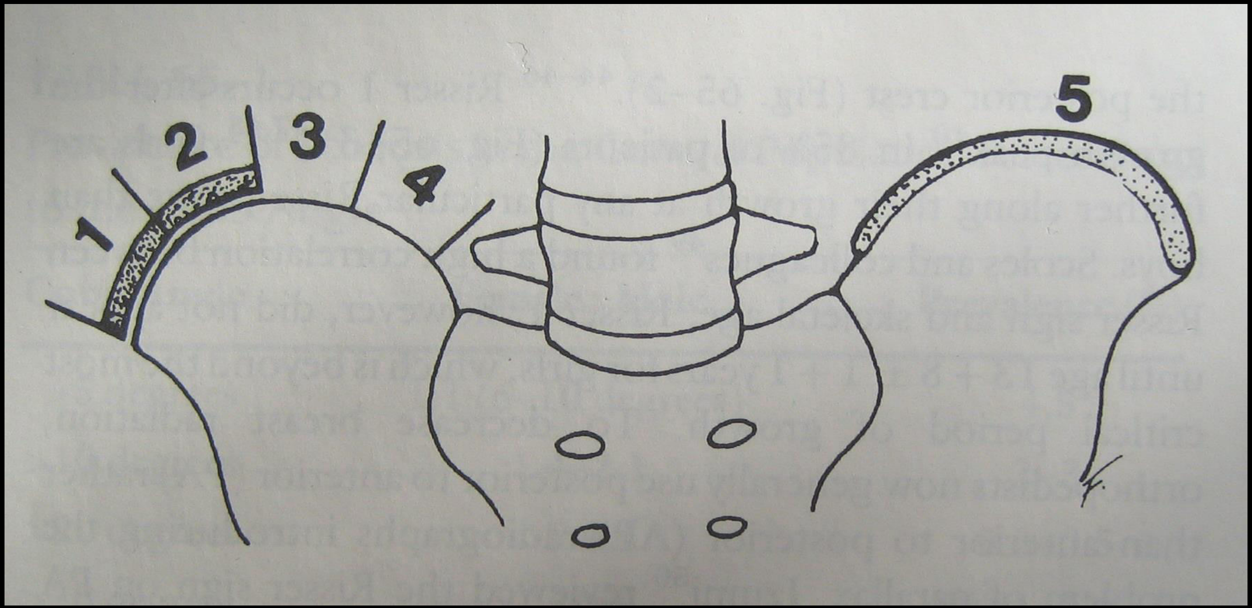
PHV vs age

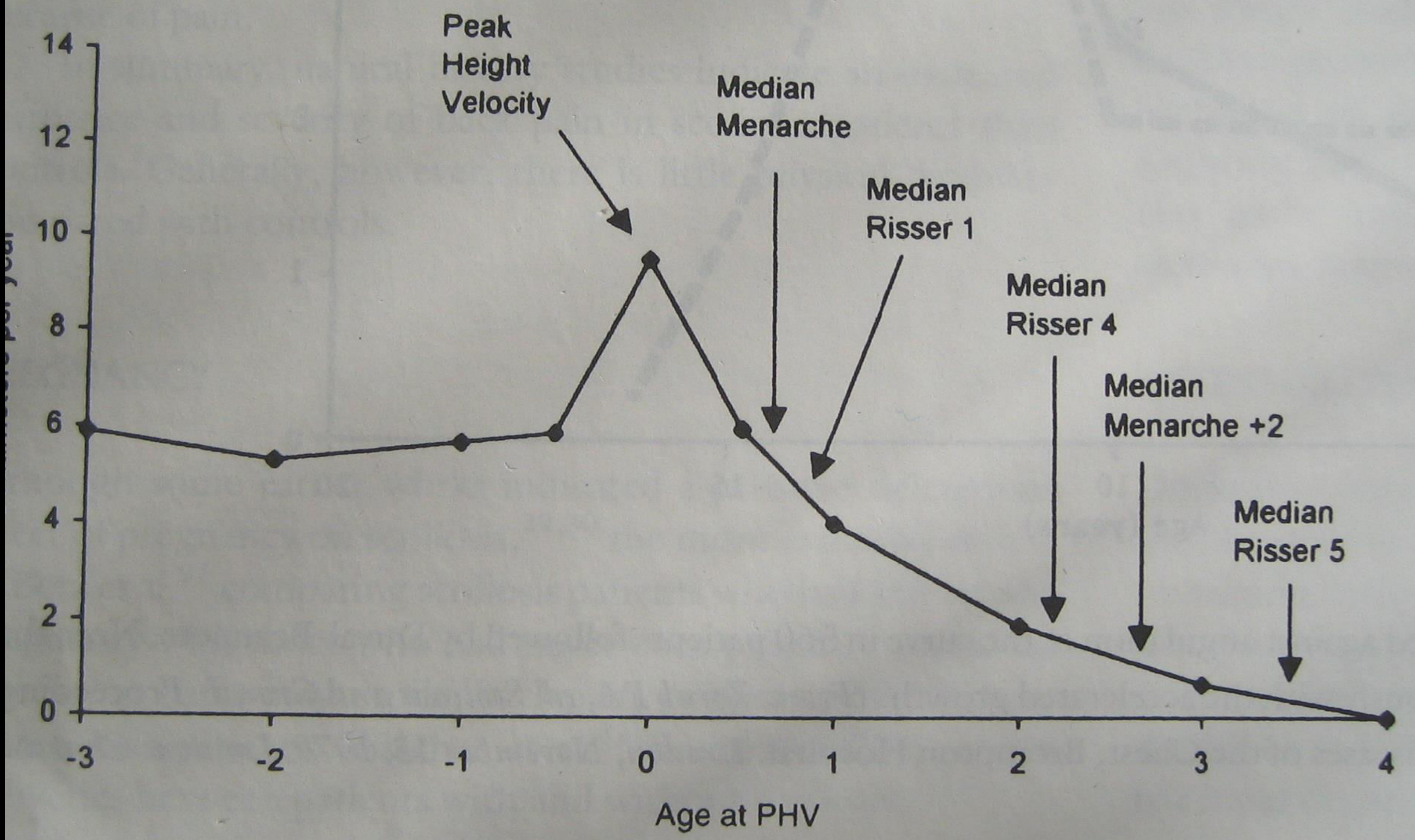


PHV and curve progression

- Girls: <30 deg at PHV – 4% surgery
  >30 deg at PHV – 83% surgery
- Boys: <30 deg at PHV – 14% surgery
  >30 deg at PHV – 100% surgery

Risser sign





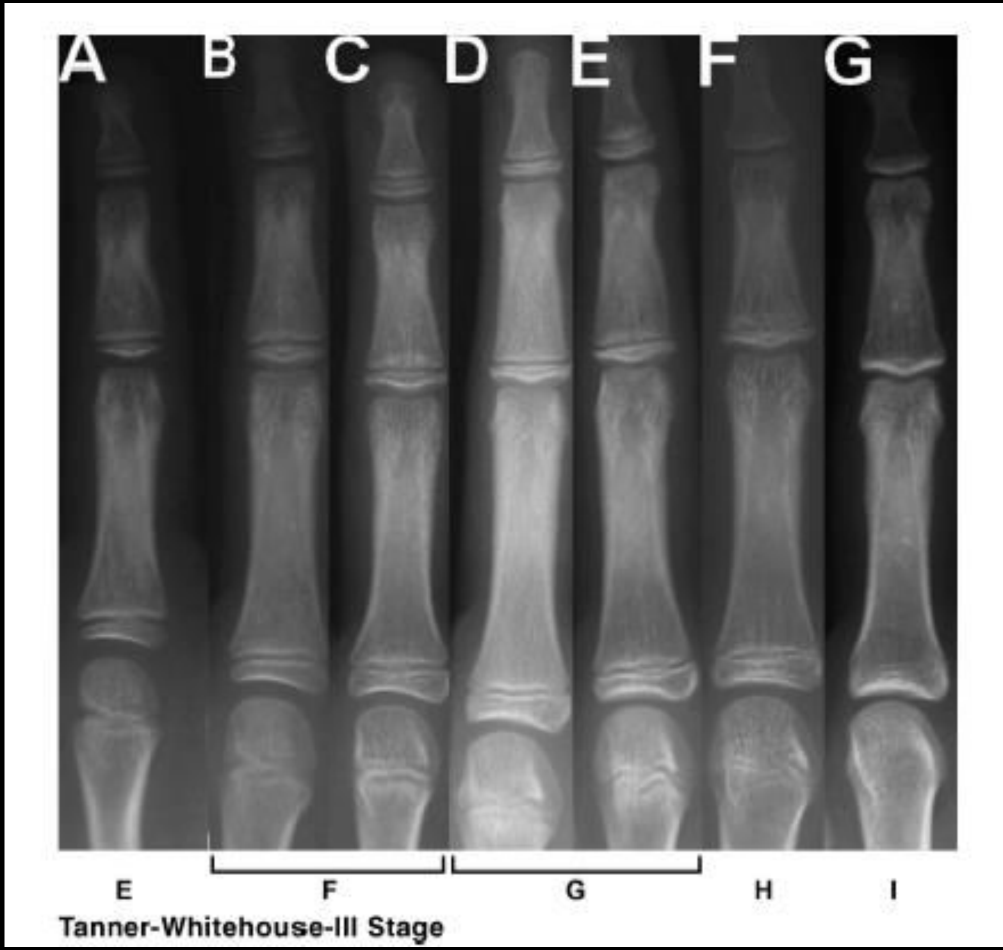
Peak Height Velocity
Median Menarche
Median Risser 1
Median Risser 4
Median Menarche +2
Median Risser 5
14
12
10
8
6
4
2
0
-3
-2
-1
0
1
2
3
4
Age at PHV

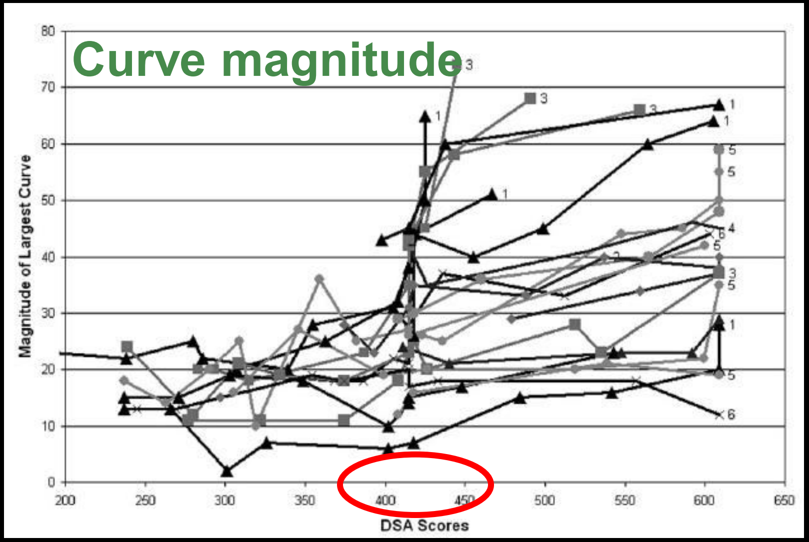
Maturity indicators

Correlation with Curve Acceleration Phase	Pearson R Value	P Value
Tanner-Whitehouse-III RUS Score	0.93	<0.001
Greulich and Pyle bone age	0.90	<0.001
Chronological age	0.89	<0.001
Timing relative to peak height velocity	0.88	<0.001
Tanner staging	0.82	<0.001
Triradiate cartilage stage (Oxford)	0.78	<0.001
Tanner-Whitehouse-III CARP score	0.77	<0.001
IGF-1 levels	0.75	<0.001
Modified Oxford score	0.75	<0.001
Risser stage	0.60	<0.001
Femoral head stage (Oxford)	0.55	<0.001
Ischial stage (Oxford)	0.54	<0.001
Greater trochanter stage (Oxford)	0.39	<0.001
IGFBP-3 level	0.35	<0.001
DHEA-S level	0.28	0.001
Osteocalcin level	0.23	0.0063
Estradiol level	0.13	0.1452
Bone-specific alkaline phosphatase level	−0.10	0.2281

Sanders et al JBJS Am 2007

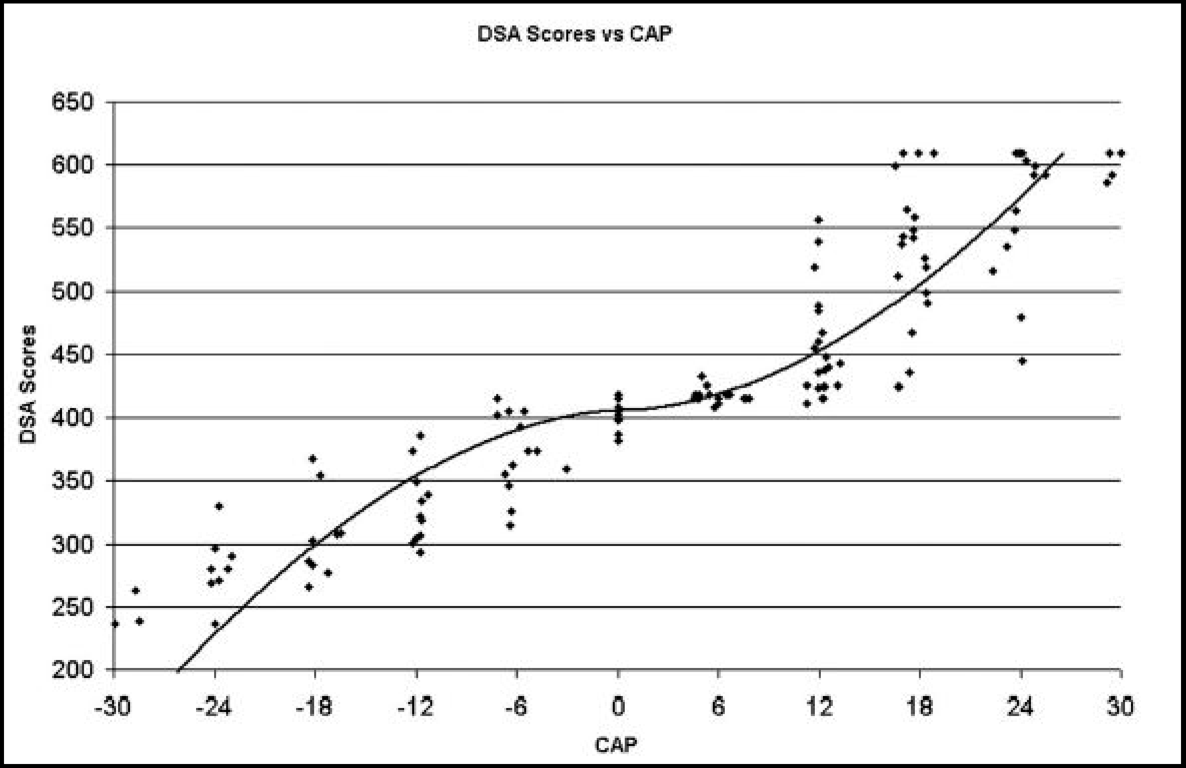
Digital skeletal age





Sanders et al JBJS Am 2007

Digital skeletal age



The equation that was generated for the relationship between the curve acceleration phase (CAP) and the digital skeletal age (DSA) was

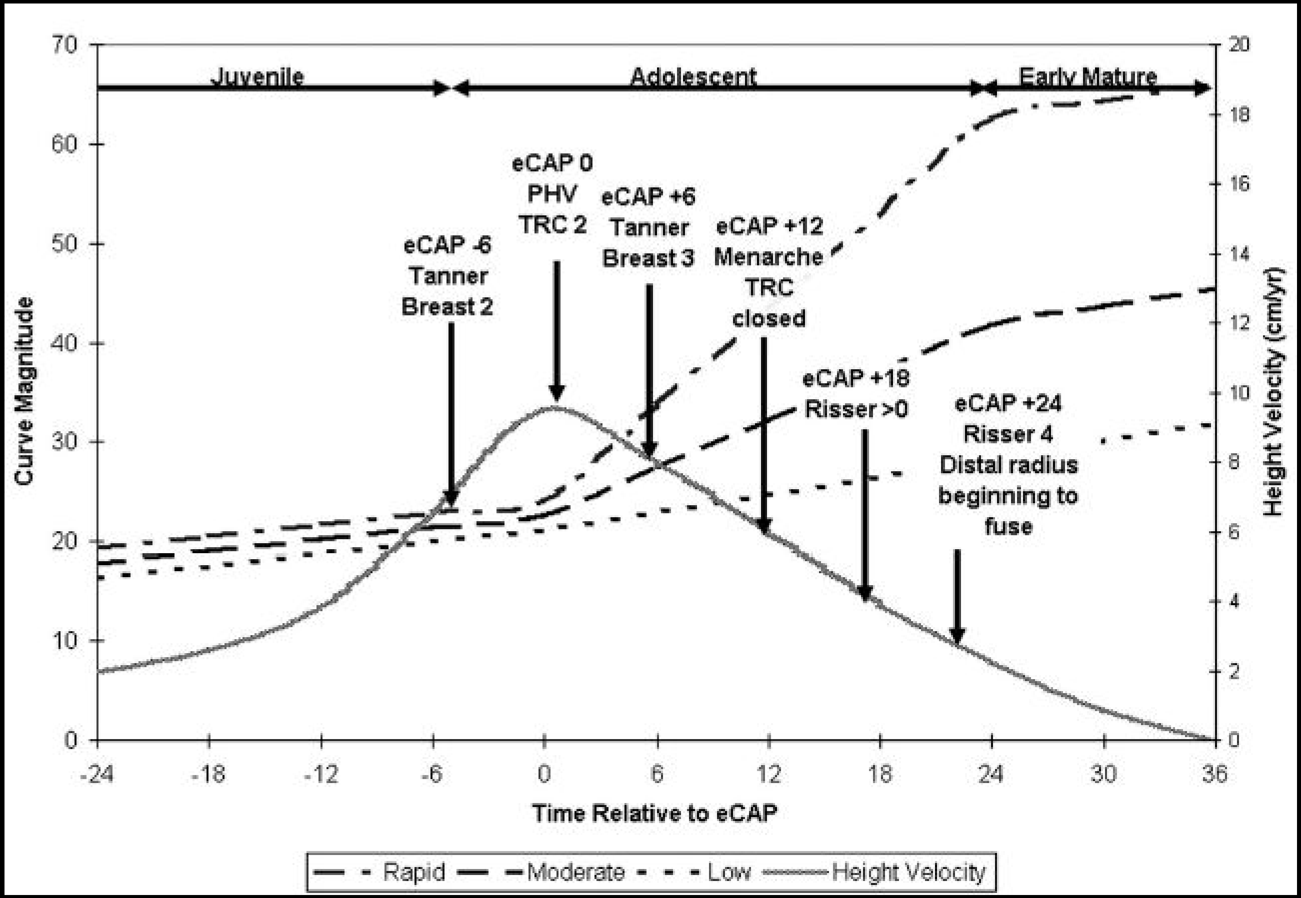
$$CAP = a + b\left(\frac{DSA - c}{d}\right)\left(\frac{|DSA - c|}{d}\right)^{-e}$$

where a = 0.3479, b = 24.91, c = 406.04, d = 185.3, e = 0.4473 and $r^2 = 0.90$ (r = 0.95).

r = 0.93

Sanders et al JBJS Am 2007

Curve progression and maturity markers



Sanders et al JBJS Am 2007

Management

- 0-20 degrees observation
- 20-40 degrees bracing
- 40+ degrees surgery

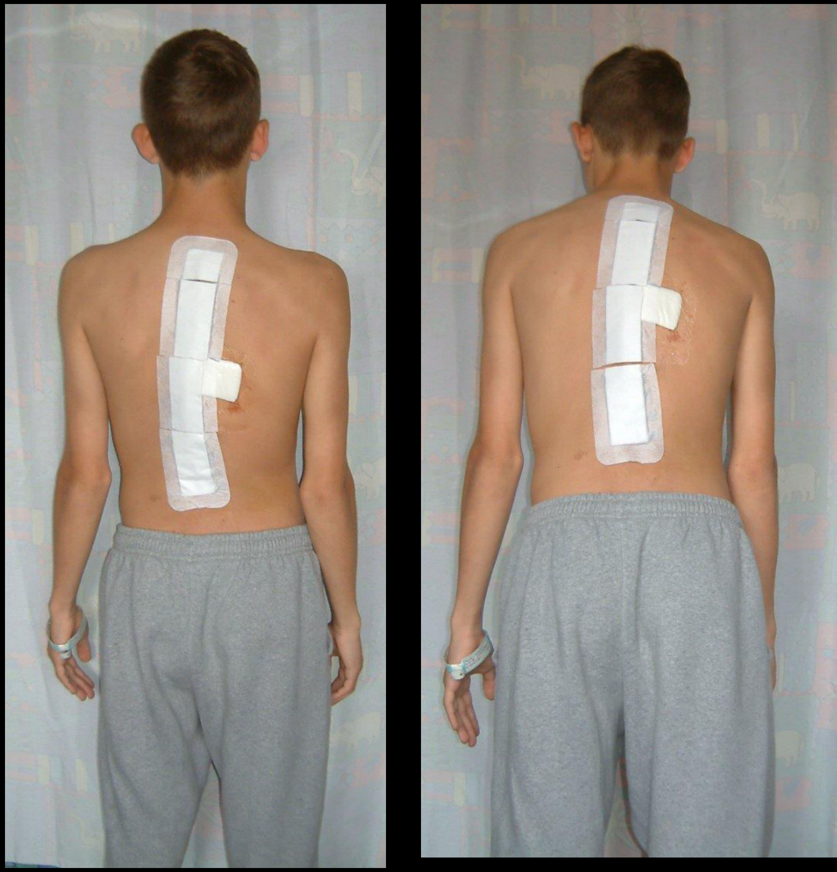
Surgical correction

Thoracic scoliosis
Post instrumantation

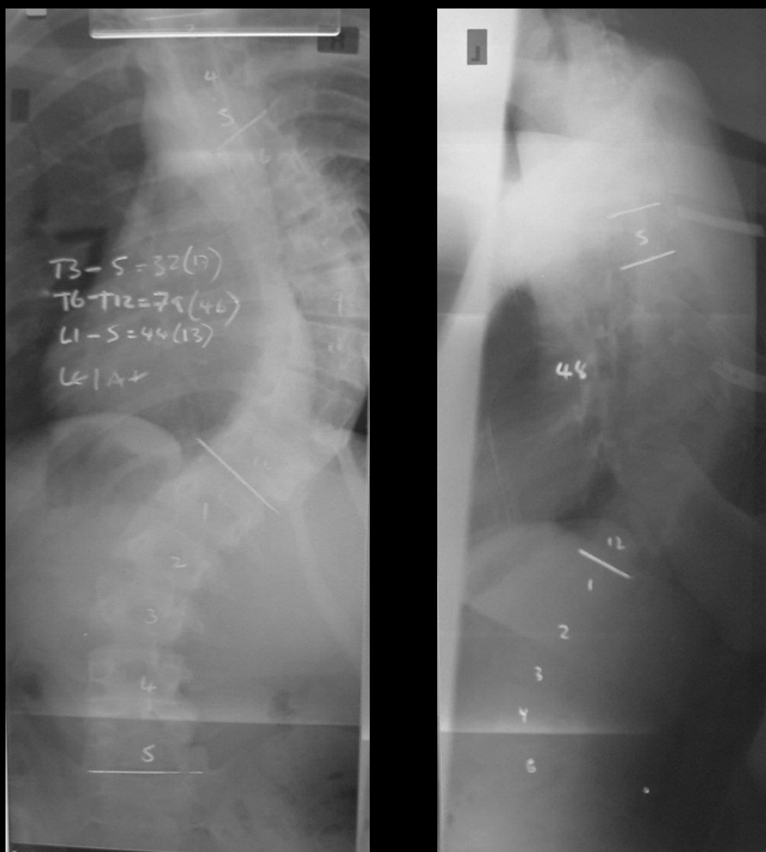
Lumbar scoliosis
Antr instrumention

Pre op

Post op

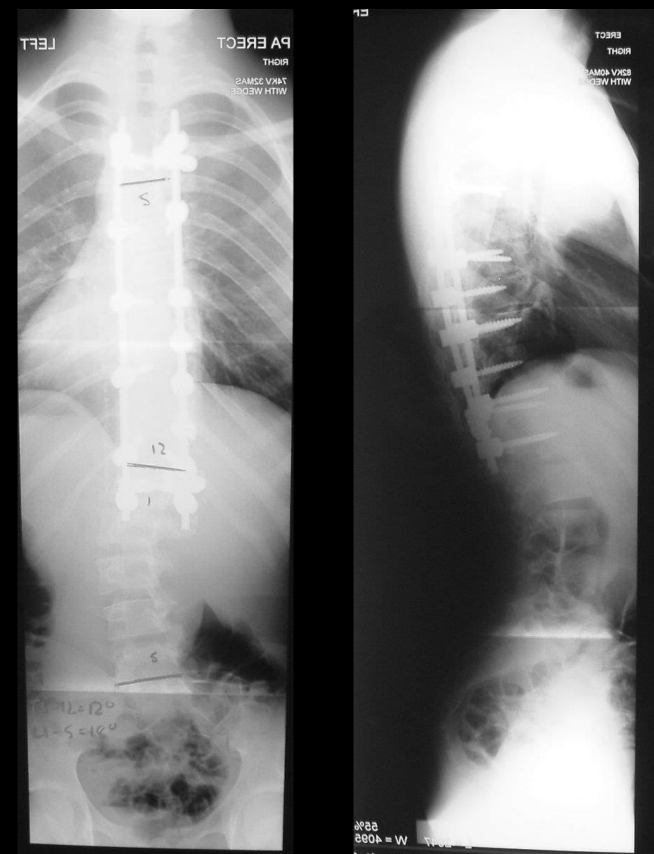


Pre op



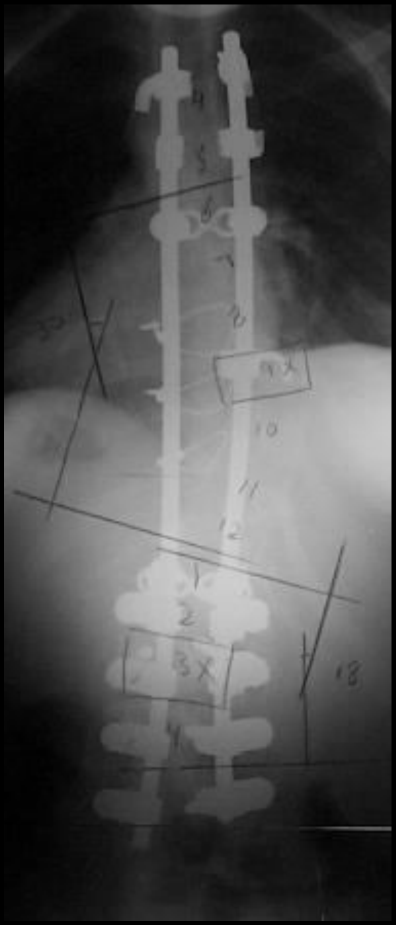
T5-12 79, L1-5 46

Post op

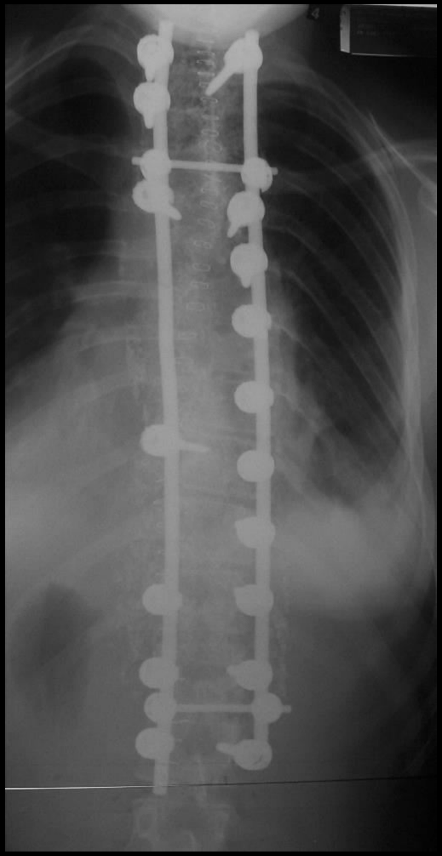


T5-12 12, L1-5 18

Posterior instrumentation

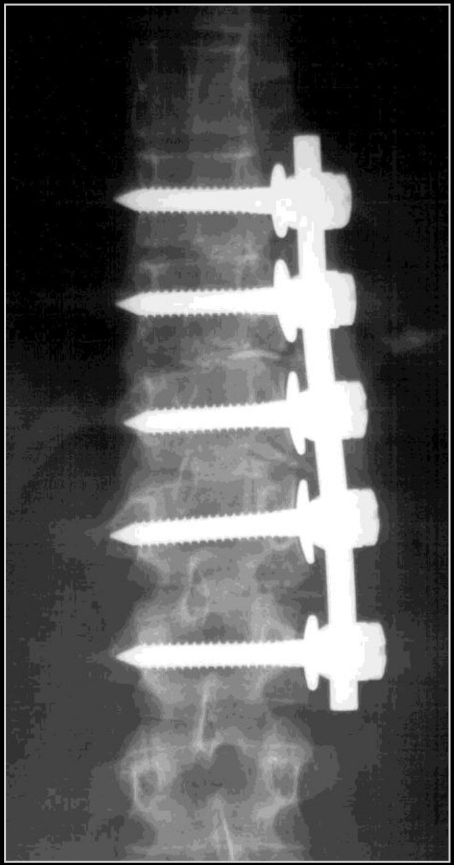


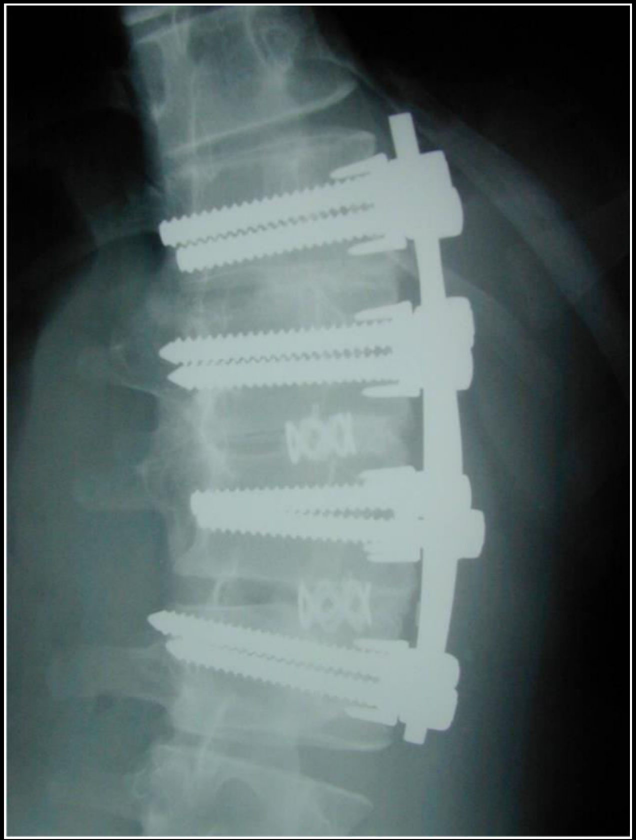
Hybrid construct



Screw only construct

Anterior instrumentation

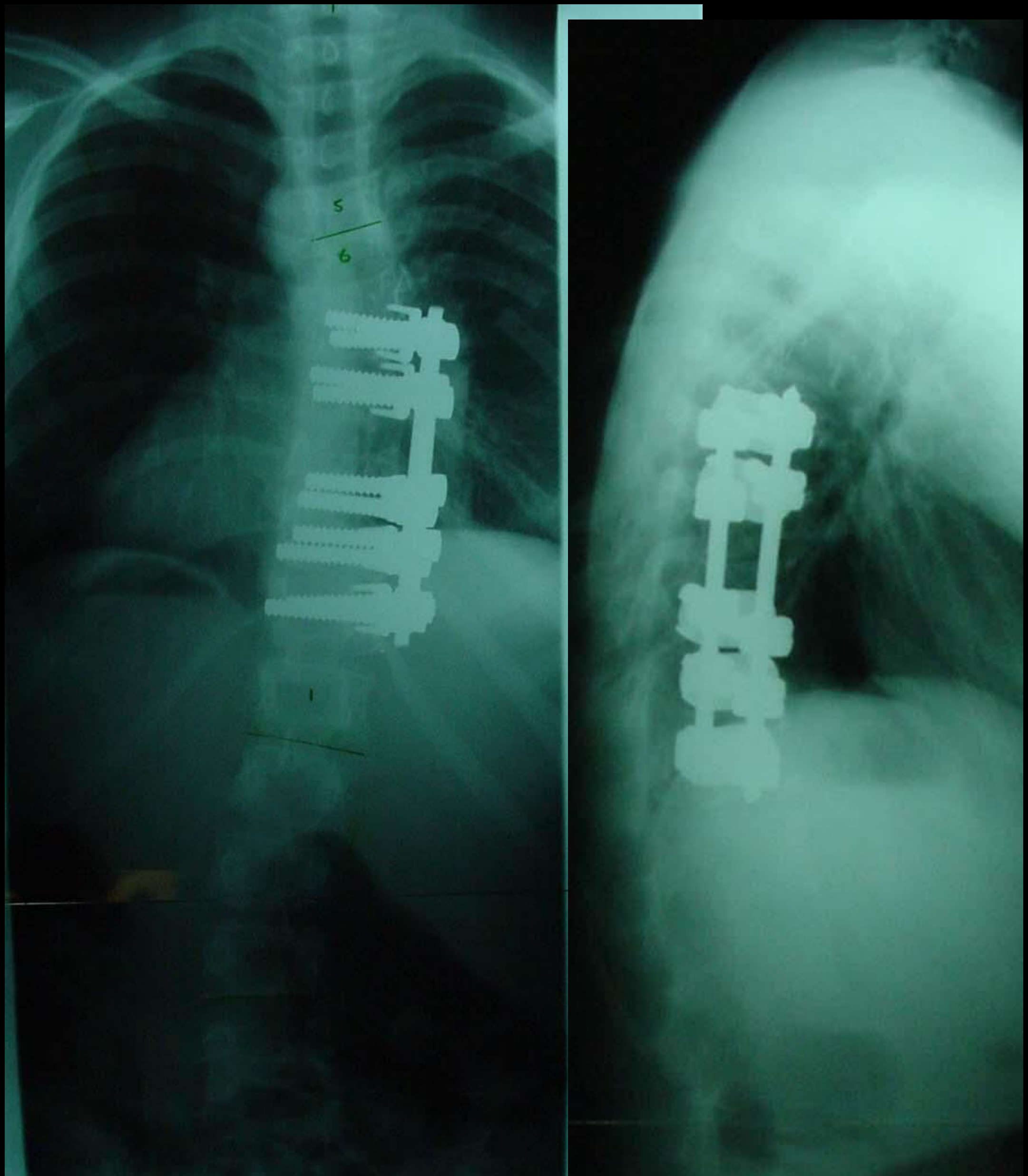

Single rod


Dual rod

SURGERY

Anterior surgery

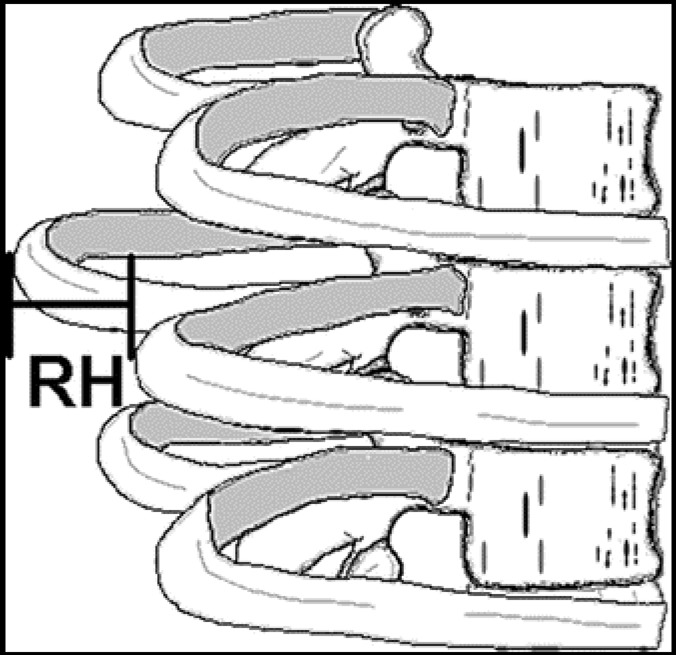
KASS



Radiographic outcomes of ASF versus PSF with thoracic pedicle screws for treatment of Lenke Type 1 AIS curves

Potter et al Spine (2005) 30:1859-66

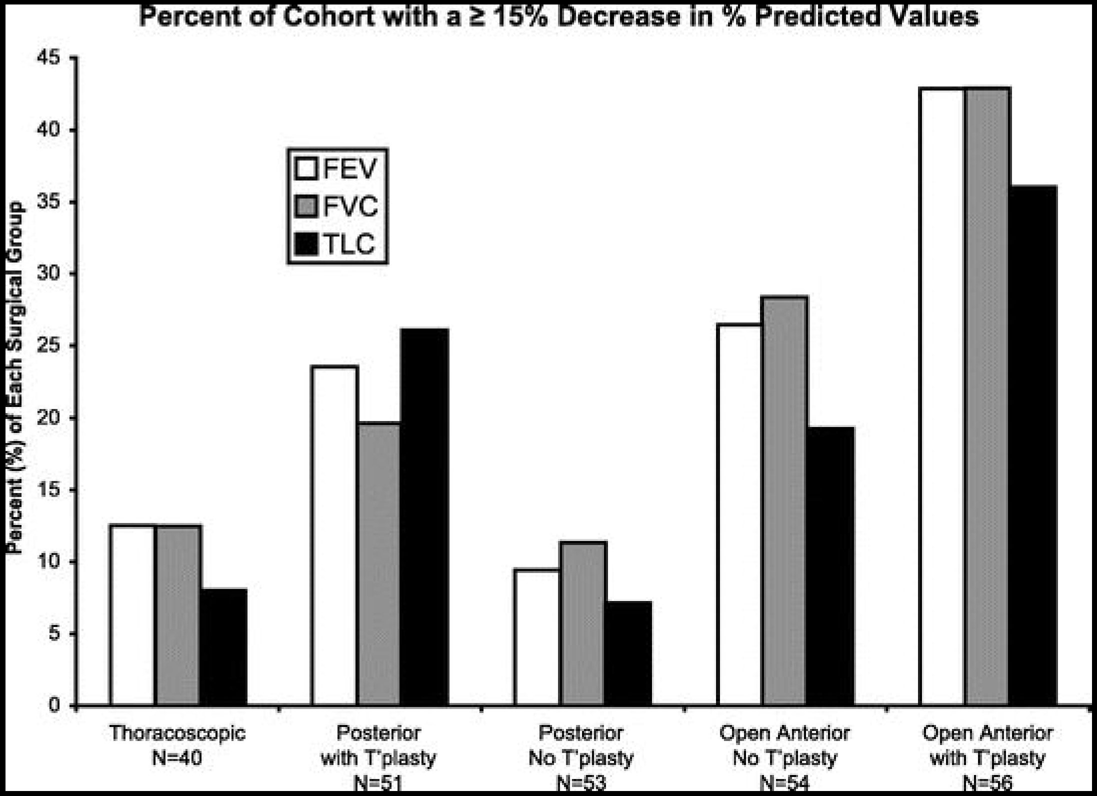
- Retrospective 40pts
- 20 each Gp
- PSF group better RH and XR correction
- 1 extra level fused with PSF gp



Predictors of change in Postoperative Pulmonary Function in AIS

Newton et al Spine (2007) 32:1875-1882

- Prospective multicentre study 254 pts



A pedicle screw construct gives enhanced posterior correction of AIS when compared with other constructs

Myth or Reality

Vora et al Spine (2007) 32:1869-74

- Retrospective tricentre cohort 72 pts
- 3 Gps- Gp 1 hook/wire/hook, Gp2 hook/wire/screw, Gp3 all screw
- Preoperative curve flexibility predicts correction rather than construct type