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**NATIONAL CONVENTION**

**June 21 – 24, 2023 • Orlando, FL**

# ORTHOPAEDIC ISSUES

AND

# PWS

**Harold J. P. van Bosse, MD, FAAOS**

**Pediatric Orthopaedic Surgery**

**SSM Health Cardinal Glennon Children's Hospital**

**Saint Louis, Missouri**



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# Introduction

- **Most common genetic obesity syndrome**
- **1887 Langdon-Down**
  - ⊕ **Adolescent girl**
- **1956 Prader, Labhart and Willi**
  - ⊕ **Series of patients**
- **1981 Ledbetter, Riccardi and Airhart**
  - ⊕ **Microdeletions of chromosome 15**



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# Prader-Willi Syndrome

- **Genetic syndrome**

- **Chromosome 15**

- ✦ **Prader-Willi region**

- ✦ **Lacking from father's chromosome**

- ✦ **If lacking from mother's chromosome:**

**Angelman syndrome**

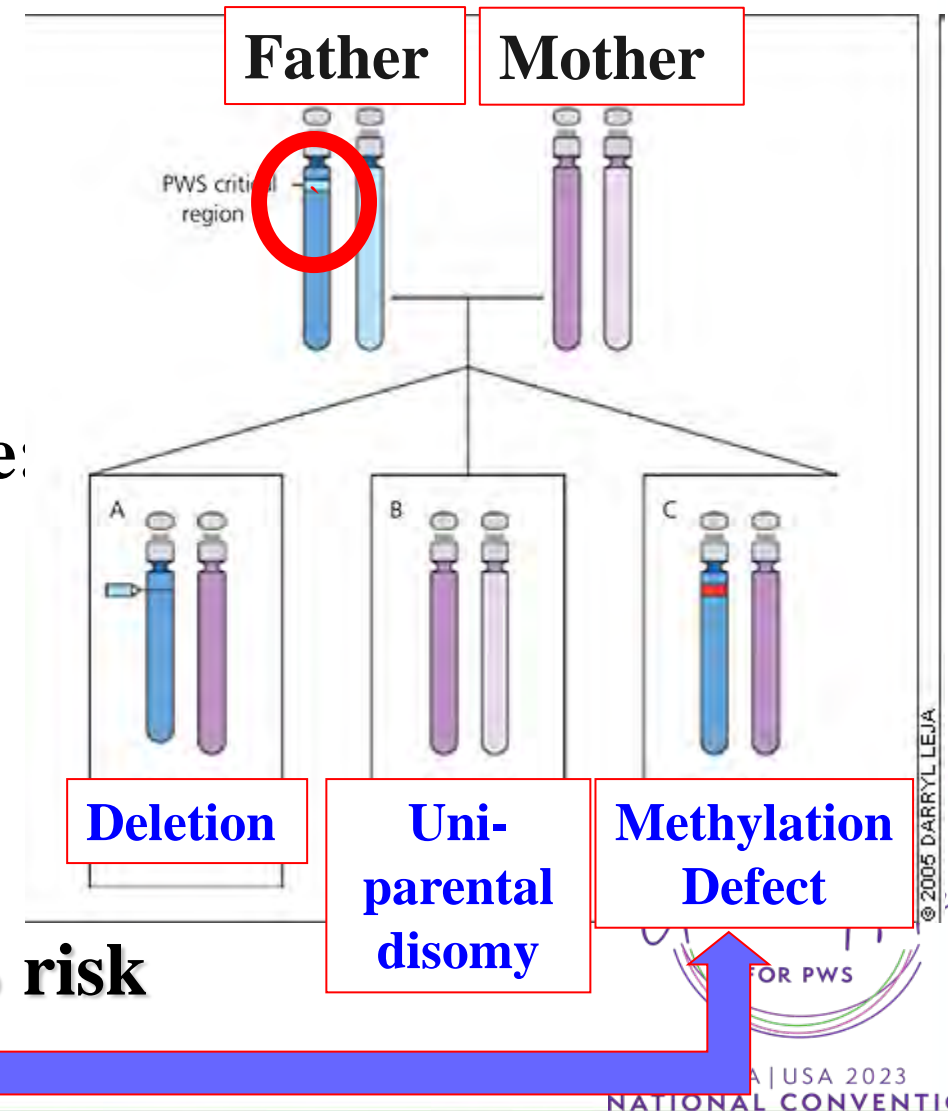
- **Genetic syndrome**

- ✦ **Unlikely to “run in the family”**

- ✦ **Most causes of PWS run <1% risk**

- ✦ **“Imprinting defect” runs about 50% risk**

- ✦ **Accounts for <5% of PWS cases**



# Musculo-skeletal Characteristics

## ● **Hypotonia**

- ⊕ **Especially in the infant**
- ⊕ **Developmental delay**
- ⊕ **Contributes**
  - ✦ **Scoliosis**
  - ✦ **Hip dysplasia**
  - ✦ **Flat-footedness (pes planus)**



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# “Why Doesn’t My Practitioner Know PWS?”

## ● **Rare: Occurs about 1:10,000 to 1:30,000 births**

Compare with more “common” orthopaedic conditions

⊕ Cerebral palsy 3:1000

⊕ Spina bifida 7:10,000

⊕ Hip dysplasia 1:1000

⊕ Clubfoot 1:2000

## ● **Generalized treatment thinking**

⊕ But children with PWS follow different rules

⊕ Need to treat as a “child with PWS who also has scoliosis, hip dysplasia, pes planus, etc”

⊕ Not “Child with scoliosis who also has this PWS thing”



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# Musculo-skeletal Issues in PWS

- **Developmental delay (milestones)**
- **Flatfoot deformity (pes planus)**
- **Osteopenia (low bone calcium)**
  - ⊕ **Frequent fractures**
- **Hip dysplasia**
- **Spine deformities**



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## ● Hypotonia early on

### ⊕ Pre-natal and delivery history

- ✦ Decreased fetal movement

- ✦ Open cesarean section for delivery

### ⊕ Newborn

- ✦ Poor sucking ability

- ✦ Weak cry



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# History

## ● **Infants and Toddlers**

- ⊕ **Poor head control**
- ⊕ **Poor postural control (sitting)**

## ● **Milestones can take twice as long**

- ⊕ **Sitting 12 months average (typical 6 months)**
- ⊕ **Walking 27 months average (typical 12 months)**



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# Treatment

## ● **Therapy, Therapy, Therapy**

- ⊕ **Balance and motor abilities**

## ● **Bracing**

- ⊕ **Ankle-knee control**

- ⊕ **Ankle-Foot Orthosis (AFO)**

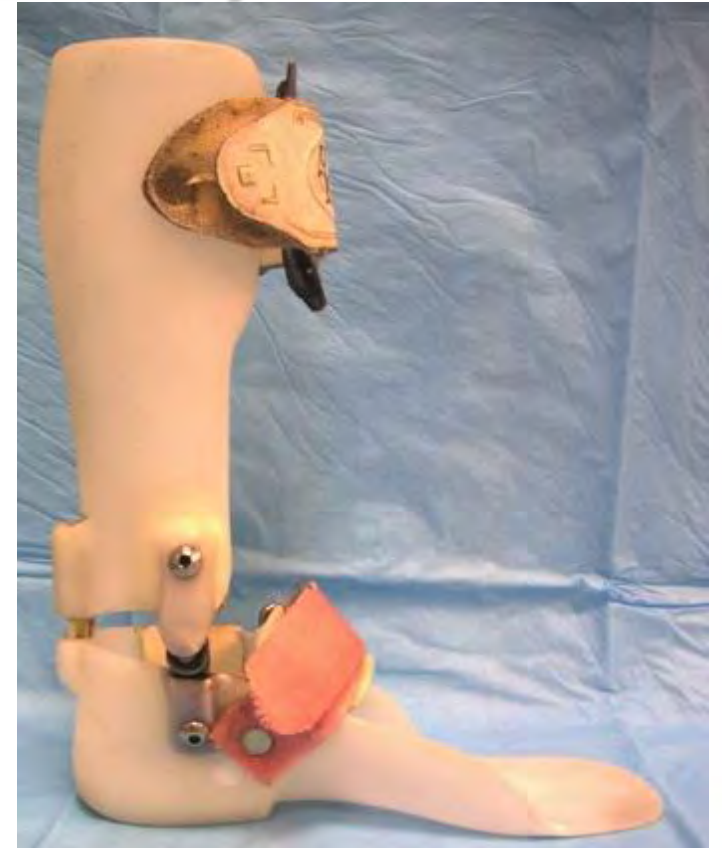
  - ✦ **Solid ankle braces – stable foundation**

- ⊕ **Purpose – “Get them up walking any way possible...  
then work on points for style”**



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# Ankle Foot Orthosis (AFO)



● **Solid versus hinged**

⊕ **More ankle stability with solid braces**

United in Hope  
FOR PWS

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# PES PLANUS (FLAT FOOT) DEFORMITIES



# PES PLANUS DEFORMITIES

## ● **Flat feet occur frequently in PWS ~41%**

- ⊕ **Laxity in ligaments and low-tone musculature**
- ⊕ **Children generally “fall” into pronation**
- ⊕ **Poor foot positioning for walking/running**

## ● **Mechanical Effects**

- ⊕ **Poor foot/ankle position, heel rolls to the outside**
- ⊕ **Hard to generate “push off” or get up on toes**
- ⊕ **Shortening of the Achilles tendon**
- ⊕ **Develop a crouched position, knees slightly bent**



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# Pes Planus Deformities

## ● Residual Effects

- ⊕ Poor balance, wide stance and walking base
- ⊕ Prolonged cruising, delayed reaching with hands and running
- ⊕ Leg fatigue
- ⊕ Forward leaning posture with high guard (hold hands up)

## ● Treatment

### ⊕ Bracing

✦ SMOs or UCBLs

✦ If using AFOs, transition once walking well



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# Braces



## ● **Supra-malleolar orthotics (SMO)**

- ✦ **Start just above ankle malleoli**
- ✦ **Usually not full foot length**
- ✦ **Soft malleable plastic**



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# My bias against SMOs



## ● Forefoot abduction

- ⊕ Lack of full lateral border
- ⊕ Front of foot still drifts out

## ● Ankle motion

- ⊕ Design inhibits full ankle motion

## ● Heel stability

- ⊕ Sub-optimal control of heel valgus



# aces



**Arch support  
won't work**



## ● Osteoporosis – weak bone

- ⊕ Low bone mineral density

## ● Different studies show different rates

- ⊕ 9% osteoporosis (PWSA-USA survey)

- ⊕ 29% history of fractures (British adult study)

  - ✦ Decreased pain sensitivity

- ⊕ 45% fractures (Philadelphia adult study)

- ⊕ No recent studies

  - ✦ Probably better results due to growth hormone



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# Osteoporosis and PWS

● **20% lower Bone Mass Density (BMD) vs. typical (2004)**

⊕ **Dual Energy X-Ray Absorptiometry (DEXA)**

● **Significantly lower BMD**

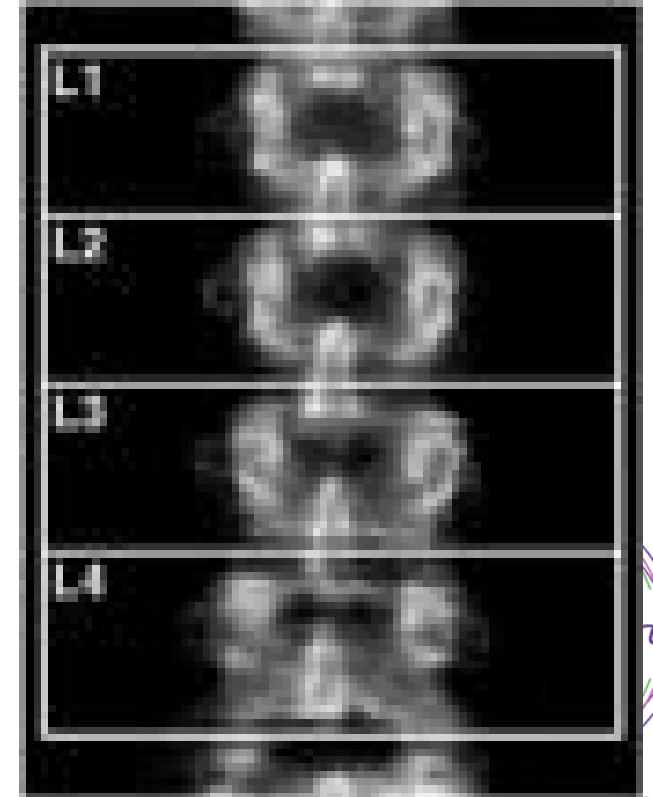
⊕ **Lumbar spine**

⊕ **Pelvis**

⊕ **Lower extremities**

● **Not significantly lower BMD**

⊕ **Head, upper extremities, thoracic spine**



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# Osteoporosis and PWS

## ● Treatment

- ⊕ Growth hormone

- ⊕ Vitamin D and calcium

- ⊕ Activity

- ⊕ Awareness

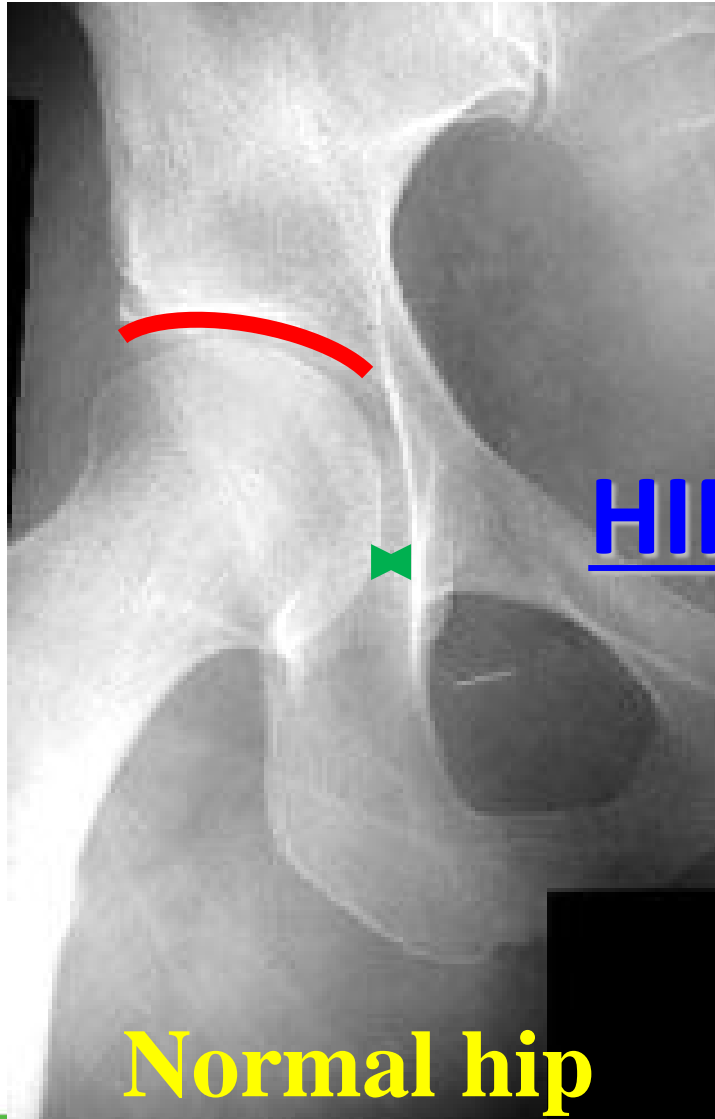
  - ✦ Injury appreciation

    - ⊕ Especially with characteristic PWS higher pain threshold

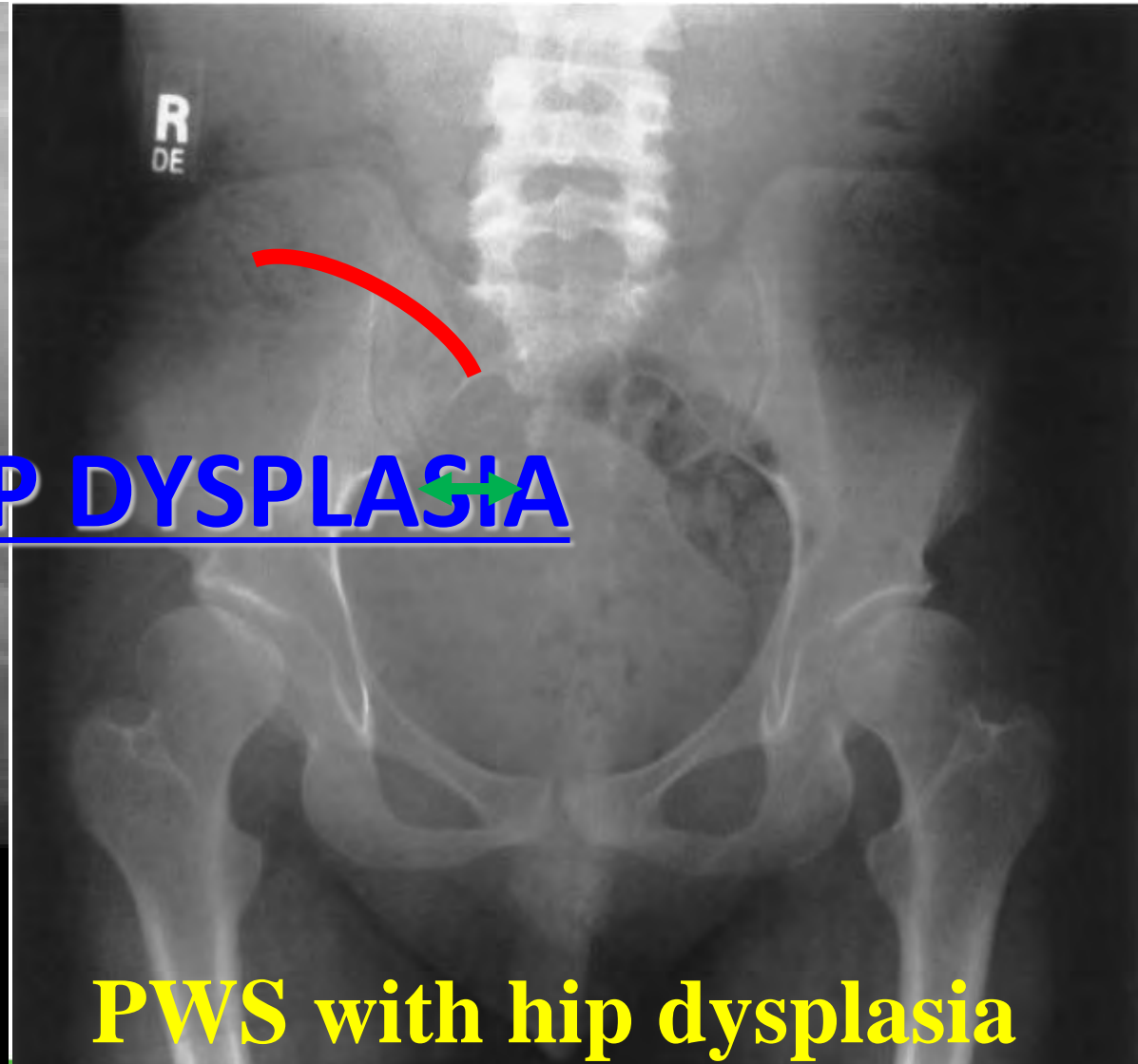
  - ✦ Surgical planning



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**Normal hip**



**PWS with hip dysplasia**

**HIP DYSPLASIA**



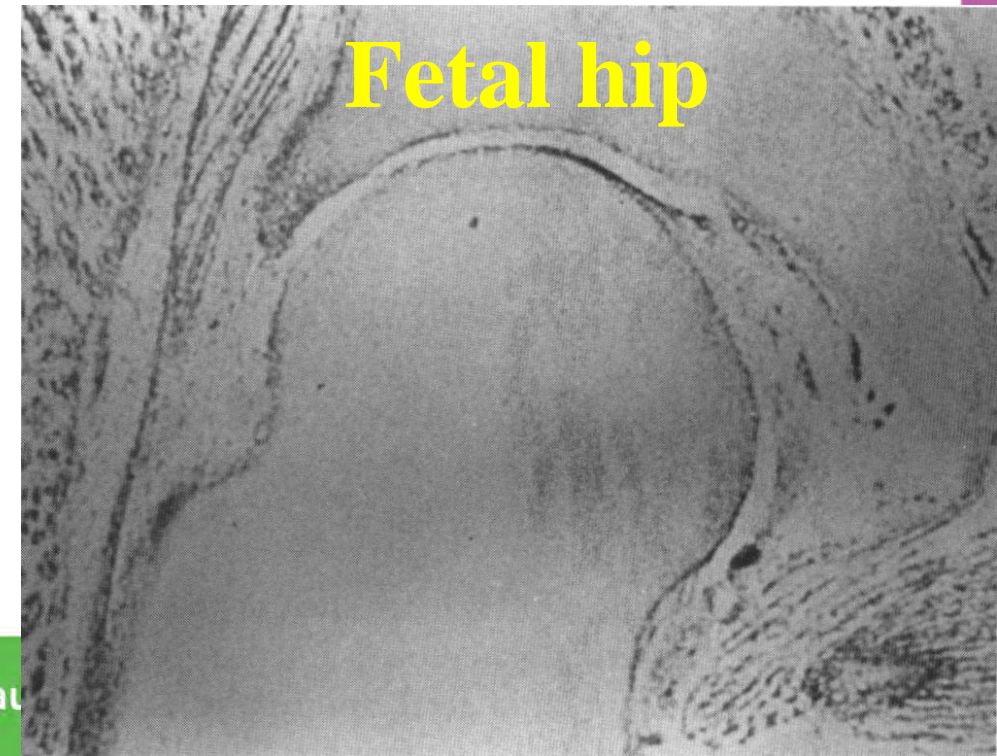
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# Hip Dysplasia

- **Dysplasia** - Abnormal development or growth
  - ⊕ Presumably normal during early development
  - ⊕ Deforms with growth in late pregnancy or after birth
    - ✦ Hip needs movement and muscle forces to develop correctly
  - ⊕ Hip dysplasia can lead to hip arthritis

- **Congenital Dislocation of Hip**

- ⊕ Hip born out of socket
- ⊕ Very rare in PWS (to me)
  - ✦ I only know of 3 cases
- ⊕ Other centers have reported higher incidence



# Hip Dysplasia

## ● Incidence 8 – 30%

- ⊕ ~1% for typically developing children

## ● Prevention

- ⊕ Early screening of hips (when start sitting independently)
- ⊕ Activity
- ⊕ Continue to screen every 1-2 years for at risk hips

## ● Hip arthritis

- ⊕ No reported cases
- ⊕ ?Too few cases per surgeon



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# National Hospital Discharge Database

- **2004-2014: 3.1 million (weighted) THAs**
  - **39 total with PWS**
  - **68% under 50 years old**
- **THA incidence rate in PWS was 1:80,000**
- **PWS incidence 1:30,000**
- **People with PWS ~2½ times less risk of hip replacements**
- **If dysplasia rate so high, why is arthritis rate so low?**

**REMODELING!**



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# 5.5 year old girl, PWS/UPD



**5.5 years old**



**7 years old**



**10 years old**

Prader-Willi Syndrome

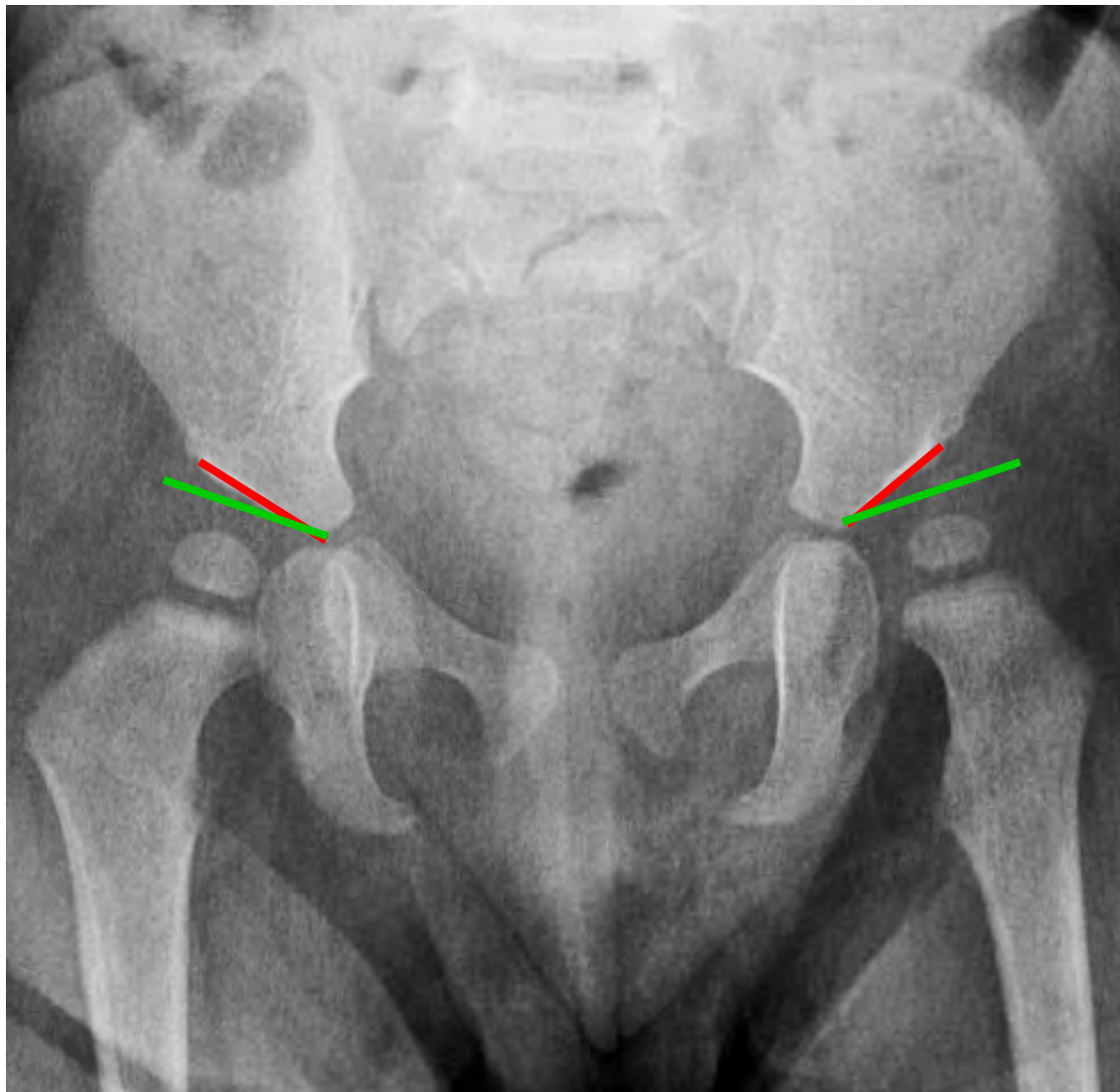


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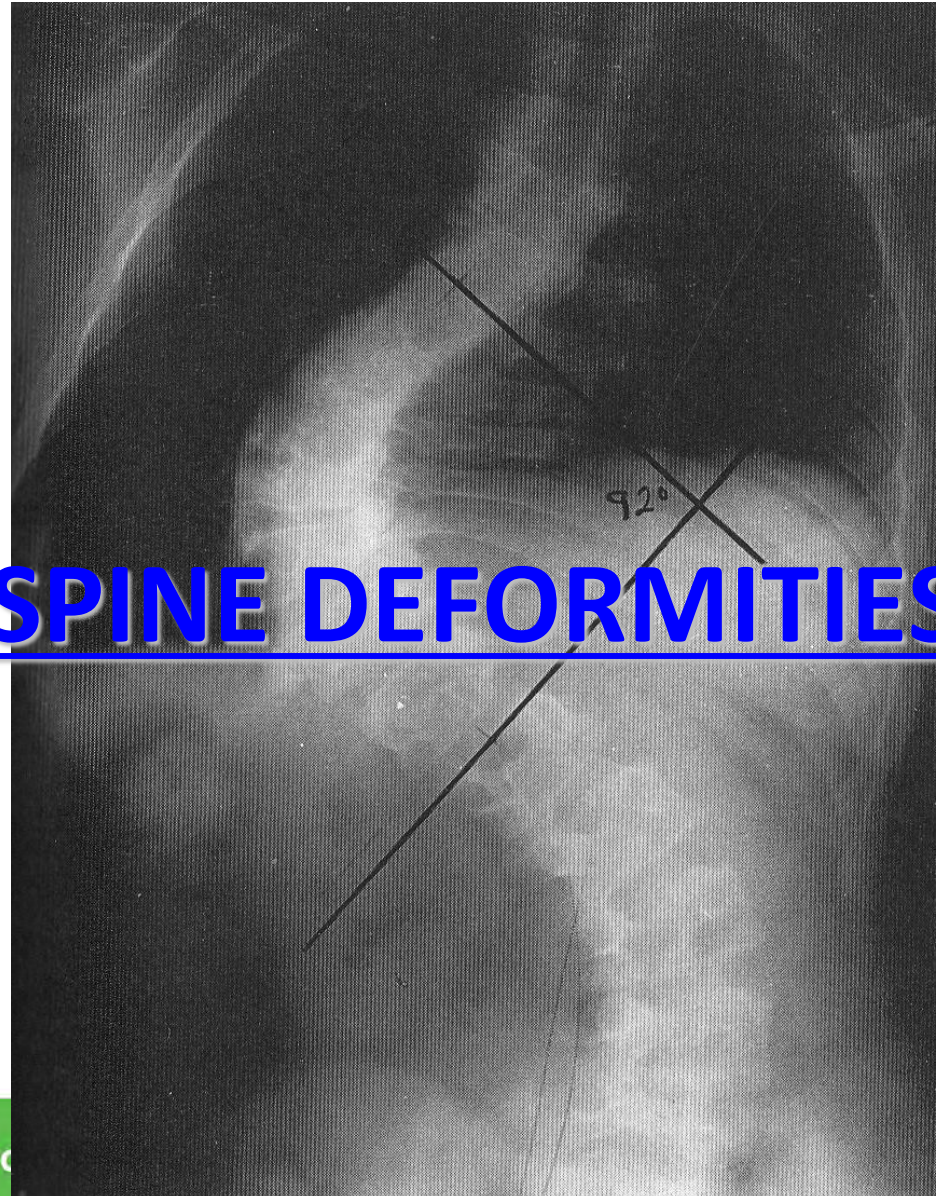
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# 13 month old girl



# 5 years old



# SPINE DEFORMITIES

Prader-Willi Syndrome

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# Spine Deformities

## ● **Scoliosis**

⊕ Any curve seen from behind

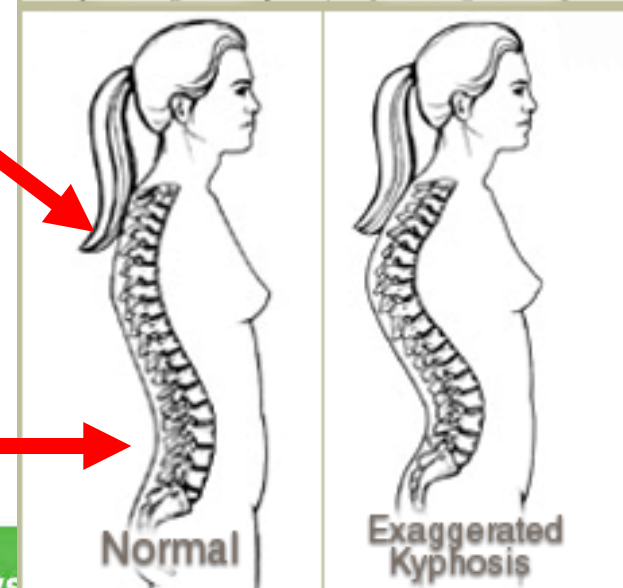
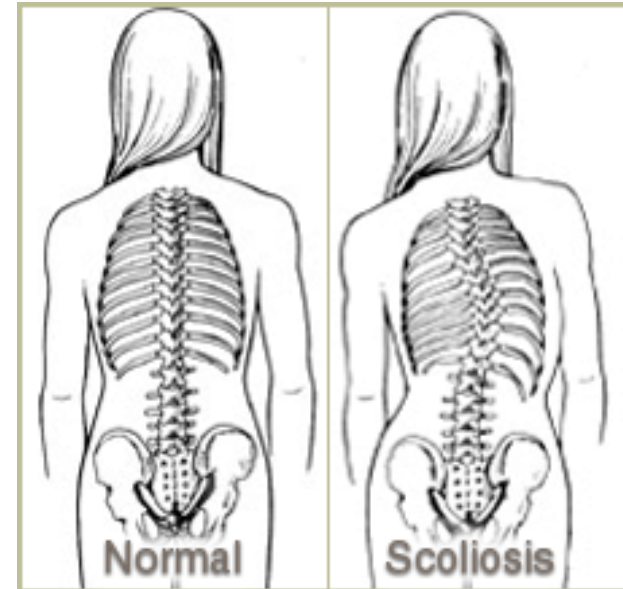
## ● **Kyphosis and Lordosis**

## ● **Hyperkyphosis**

⊕ Exaggeration of the normal postural upper back curve, seen from the side

## ● **Hyperlordosis**

⊕ Exaggeration of normal lower back curve (small of back)



**Kyphosis**

**Lordosis**

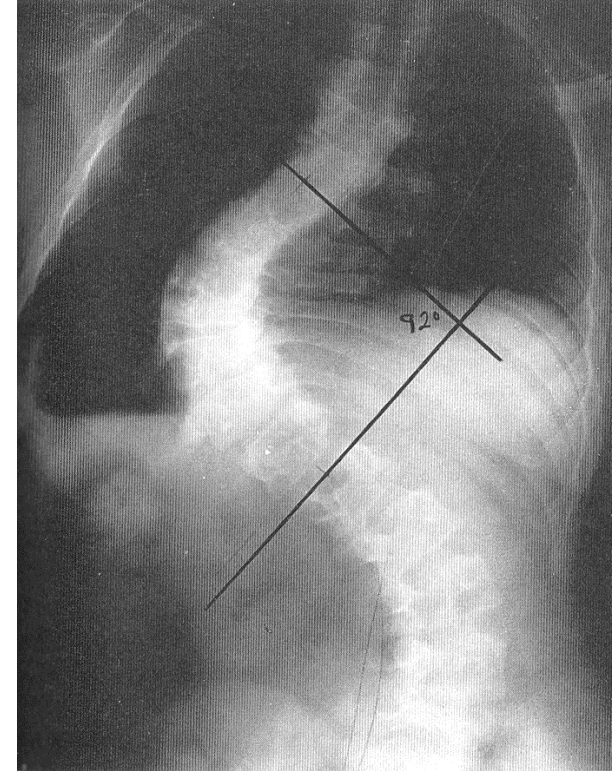


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# Let's do the numbers!

- **Spine deformity prevalence in PWS**
  - **Approximately 60%-70%**
- **Bimodal age distribution**
  - **23% of children before 4<sup>th</sup> birthday**
  - **Second (bigger) peak is in the adolescent period**
- **15% of PWS children will need spine surgery**
  - **Complication rates from surgery ~56%**



# More numbers

## Treatment rationale for scoliosis

### ● **At maturity**

- **Curves  $\leq 40^\circ$ : 95% will *not* progress in adulthood**
- **Curves  $\geq 50^\circ$ : 95% will progress**
- **Between  $40^\circ$  and  $50^\circ$  - a grey zone**

### ● **Curves under $25^\circ$ - observe**

### ● **Surgery is indicated for curves $> 40^\circ$ to $50^\circ$**



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# SCOLIOSIS

## Treatment Rationale

### ● **Cardiopulmonary Compromise**

#### ⊕ **Pulmonary insufficiency**

✦ **Lungs too squooshed to get enough oxygen into the bloodstream for the body**

#### ⊕ **Cor pulmonale**

✦ **Heart has to work too hard to push blood through the squooshed lungs: overwork**

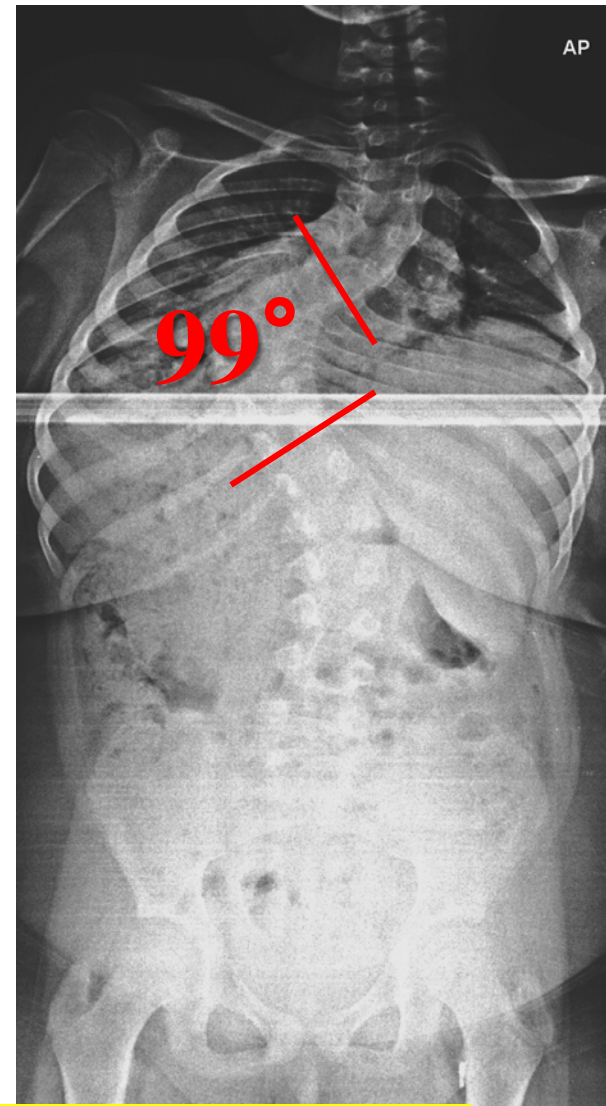
#### ⊕ **Curves over 80° to 90°**

#### ⊕ **Smaller curves can cause breathing problems (curves over 60°)**



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# Hidden Spine Deformities



8 1/2 year old boy



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# Hidden Spine Deformities

4 year old with  
30° curve



# Scoliosis

## ● Obesity

- ⊕ No differences in Body Mass Index

  - ✦ Between children with or without scoliosis

- ⊕ More than 50% of all curves start before obesity onset

  - ✦ 2/3 of all severe curves start before obesity

## ● More likely due to hypotonia

## ● Obesity control

- ✦ Detection

- ✦ Bracing



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# PWSA-USA 2007 Survey

## ● Gender

- ⊕ Females have ~10% higher chance of developing scoliosis
- ⊕ Curves behave the same (risk of progression) for both genders

## ● Genetics

- ⊕ UPD had a slightly higher risk of developing scoliosis
- ⊕ No PWS type has a higher risk of progression

## ● Looking forward to FPWR online registry results



GLOBAL  
PRADER-WILLI SYNDROME<sup>s</sup>  
REGISTRY



# Treatment

- **Difficult to draw conclusions**
- **Few reported operative cases in literature**
- **These are my opinions based on accumulated experience**

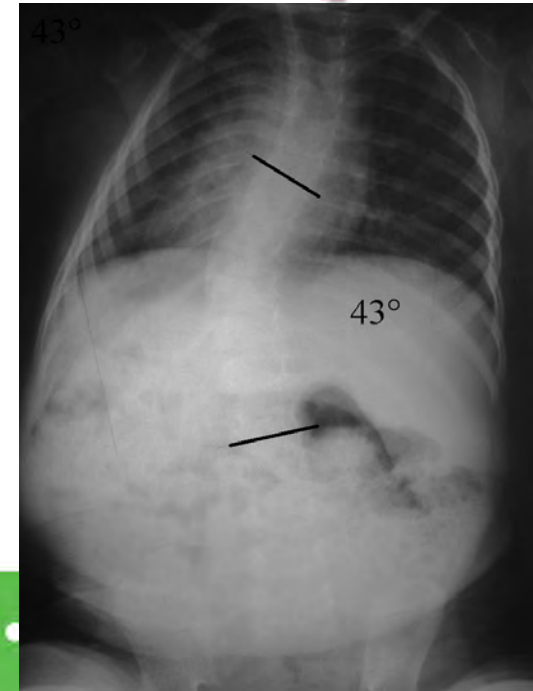


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# Prevention

- **Delay upright sitting until baby can pull to sitting position themselves**
  - ⊕ Prevents hypotonic slouch
  - ⊕ Seating devices tilted back about 30°
- **Emphasize tummy time activities as much as possible**

1 ½ year old girl



## ● Screening

# Treatment

- ⊕ Yearly screening/radiographs, once starts sitting

## ● Physical therapy

## ● Casting

- ⊕ Usually start before patients reach 3 years old

## ● Bracing

- ⊕ For curves larger than  $25^{\circ}$

- ⊕ Prevent curve progression when upright

## ● Surgery

- ⊕ For curves larger than  $45^{\circ}$



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# Reality Check

- **Important to note – some curves are destined to progress even with the best treatment**
- **Strategy then is to control the curve as long as possible**



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# Physical Therapy

## ● **Physical therapy**

- ✦ **Trunk strengthening**
- ✦ **Sensory integration**
- ✦ **Keep the young child down to develop normal gross motor skills**
- ✦ **Children with PWS develop their extremities before their trunk**



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# Janice Agarwal's Favorite Therapy



Prader-Willi Syn



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# Hippotherapy

- **Child who needs head, trunk, leg control**
- **Movement of the horse**
  - ⊕ **Encourages the child's body to exercise**
  - ⊕ **Correct alignment**
- **Therapist can provide**
  - ⊕ **Extra support**
  - ⊕ **Resistance**



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# Casting



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# Casting

## ● **Appropriate age has been increasing**

- ⊕ **Even effective in older children (up to even 7 years old)**
- ⊕ **Delay tactic before other treatments options: have problems**

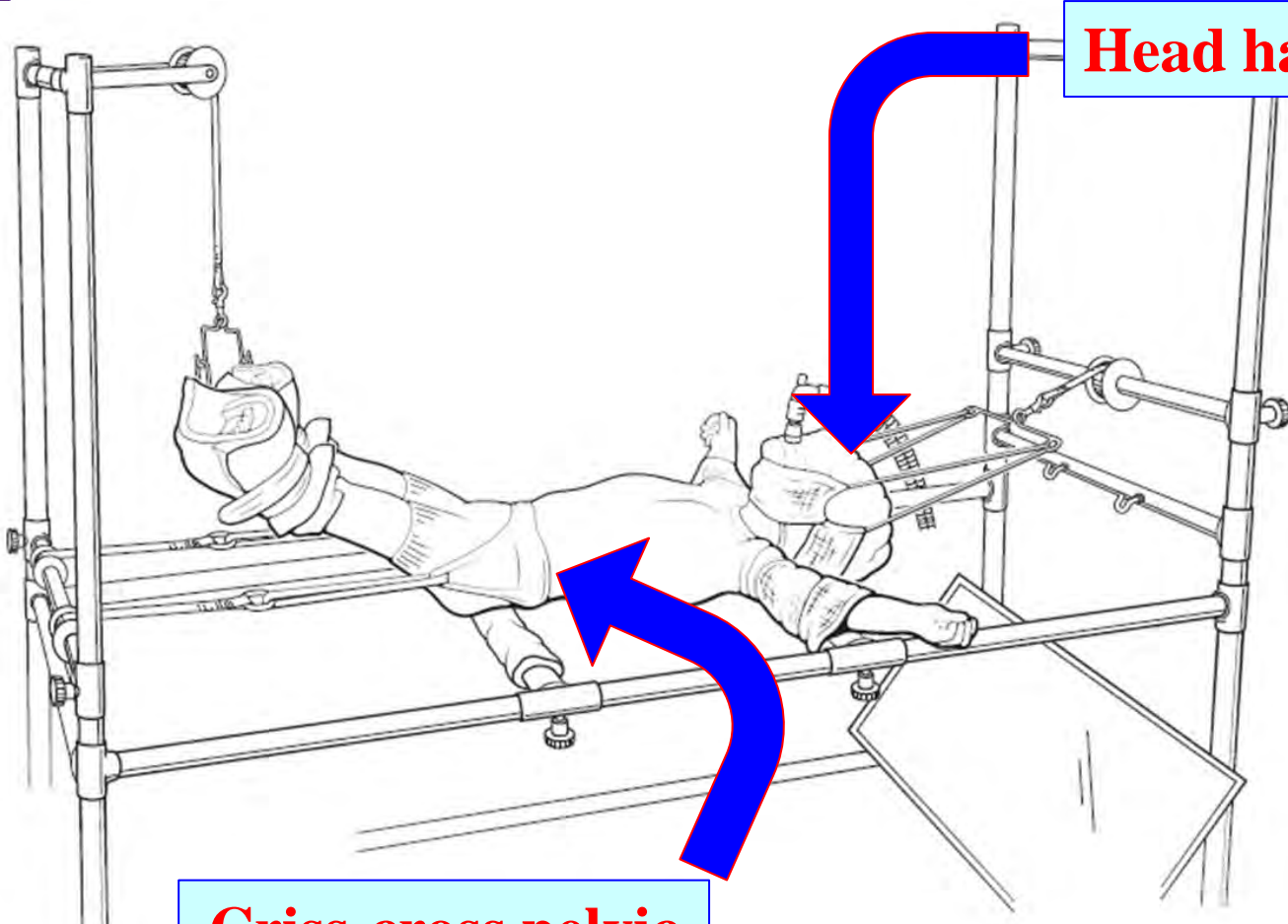
## ● **Cast under anaesthesia**

### ⊕ **Casting schedule**

- ✦ **Under 2 years, change every 2 months**
- ✦ **Over 2 years, change every 3 months**
- ✦ **Over 3 years, change every 4 months**

## ● **End casting when reach goal, or curve reaches a plateau**

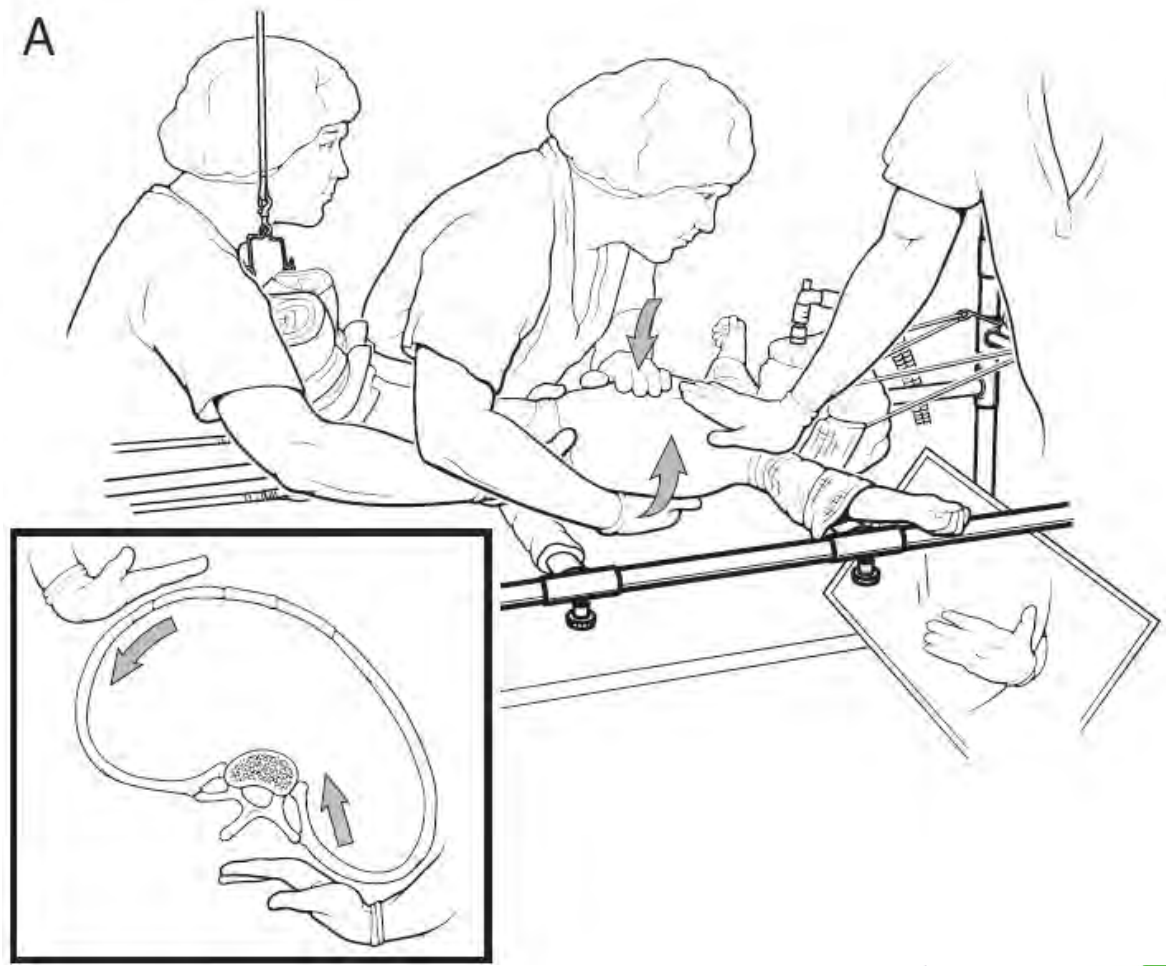
## ● **Post-treatment bracing**



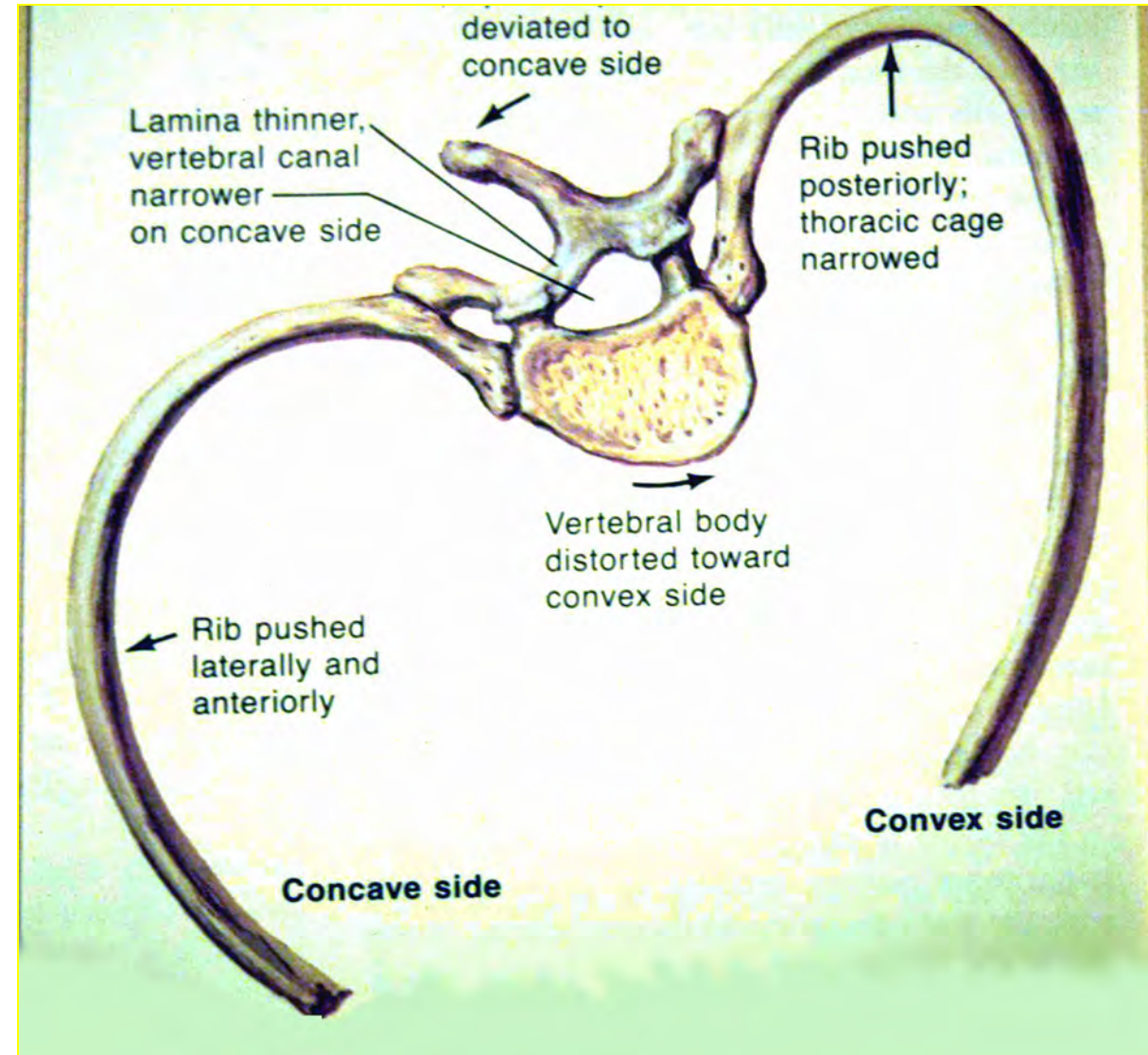
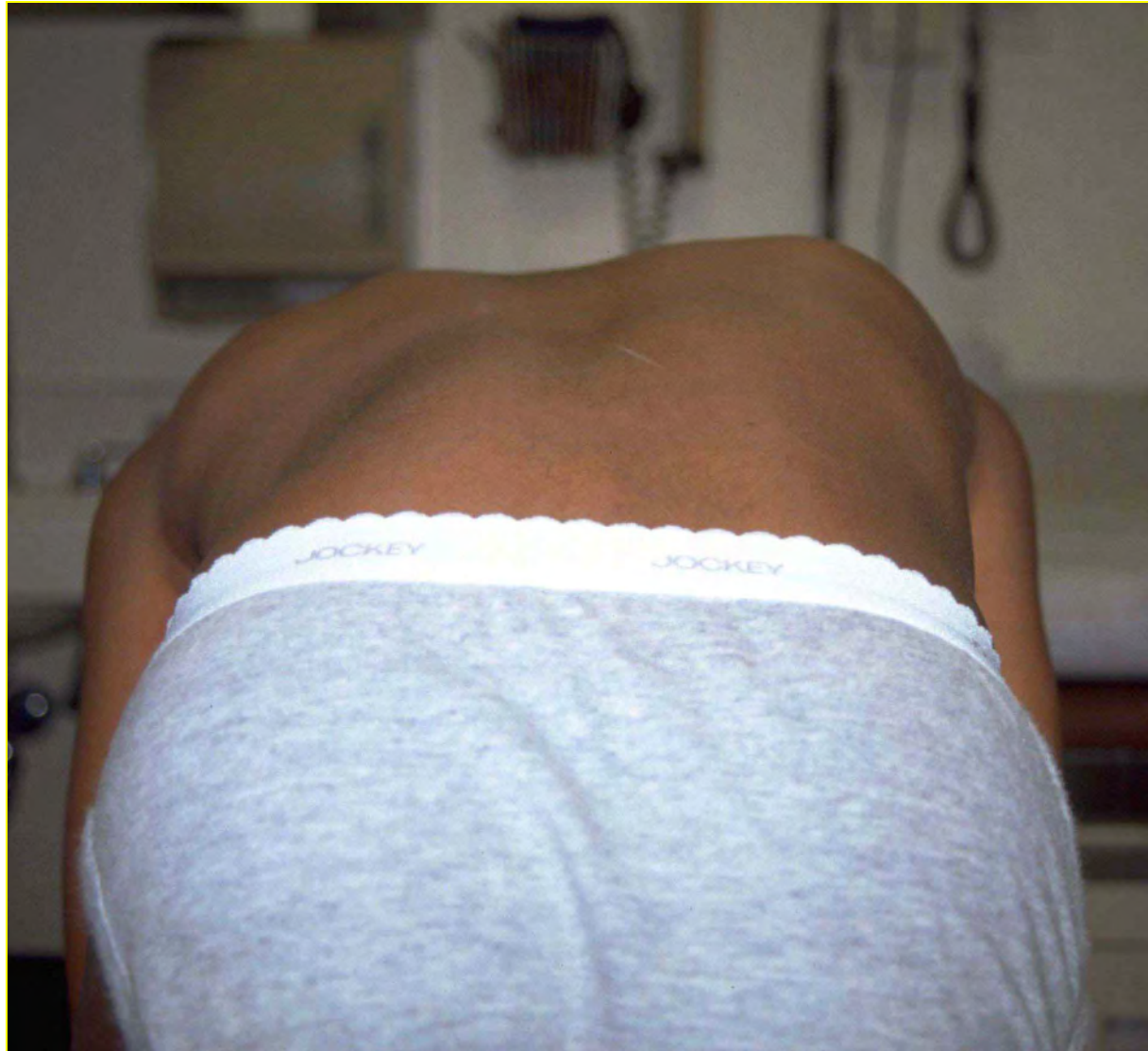
**Head halter traction**

**Criss-cross pelvic band traction**

*Sanders et al, JPO 2009*



# Derotate Chest to Correct Curve



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# PWS Spine Casting Study

## ● PWS spine casting

- ◆ 34 PWS spine casting patients

  - ◆ 2008-2018

- ◆ >24 months followup

## ● Criteria for starting casting

- ◆ Curve greater than 25°

- ◆ Age: Sitting age to 5 y.o.





# Endpoints

## ● “Cured”

- ⊕ Curve under  $15^\circ$  out of cast
- ⊕ Curve progression is likely halted
- ⊕ Transition to brace for 1 year, hopefully brace free afterwards

## ● Braced

- ⊕  $20^\circ$ - $50^\circ$  and over 5 years old – brace expectantly

## ● “Controlled”

- ⊕ Reached surgical criteria before started casting
- ⊕  $>50^\circ$  and over 5 years old – discuss expandable implants



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# Results



## ● Average age at first cast

✦ 32 months (range: 14-64 months)

## ● Average number of casts

✦ 8 casts (range: 3-18)

## ● Average followup

✦ 57 months (range: 25–103 months)



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# Results

## ● Cured ~1/3 of the group

- ⊕ PWS type: 7 UPD and 5 deletion
- ⊕ Curves went from 44° to 17° over 6 casts (17 months)

## ● Braced ~50% of the group

- ⊕ PWS type: 10 deletion, 7 UPD, 1 methylation defect
- ⊕ Initial curve 55° improved to 35° over 7 casts (27 months)

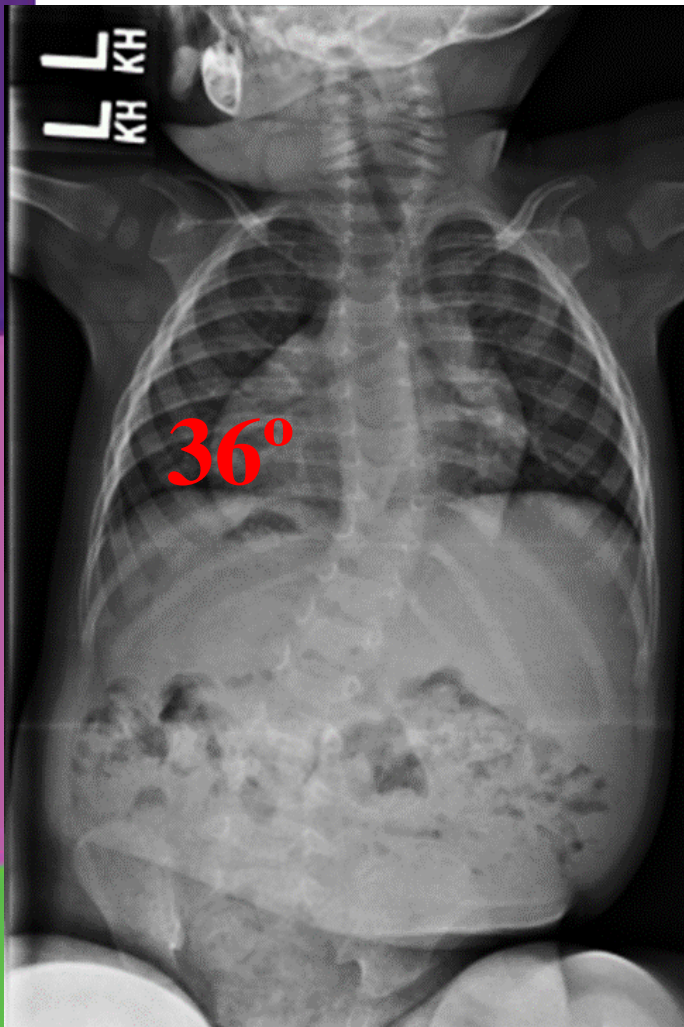
## ● Controlled 4 patients

- ⊕ PWS type: 3 deletion and 1 UPD
- ⊕ Pre-cast curve 85° improved to 54°
- ⊕ Surgery delayed ~ 4 years (from 22 months to 72 months)

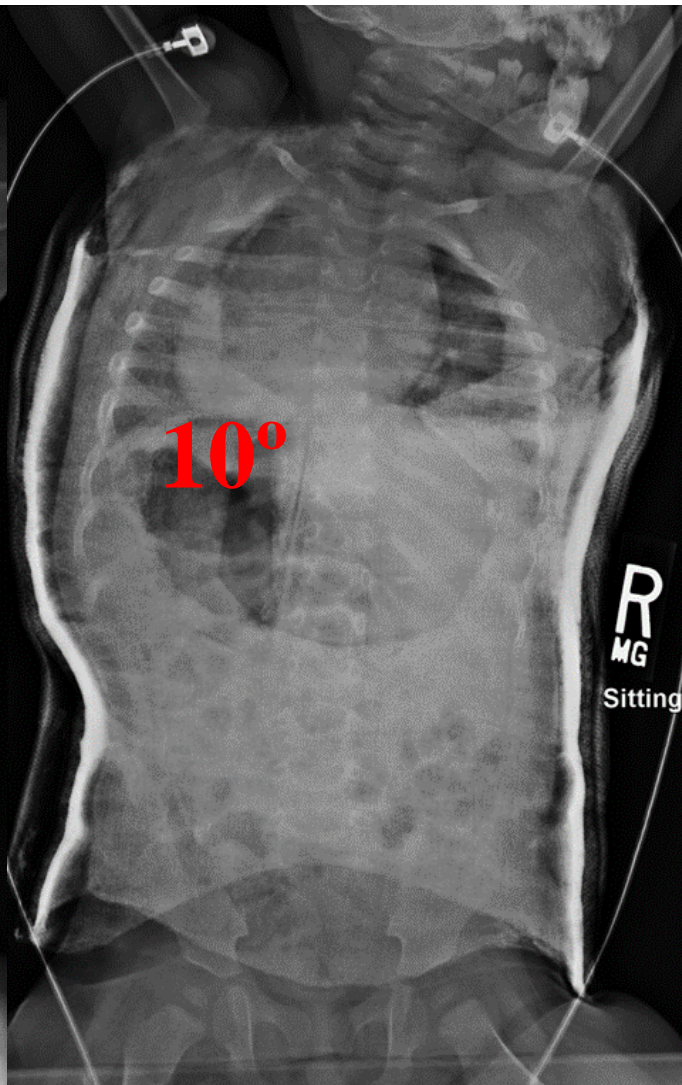


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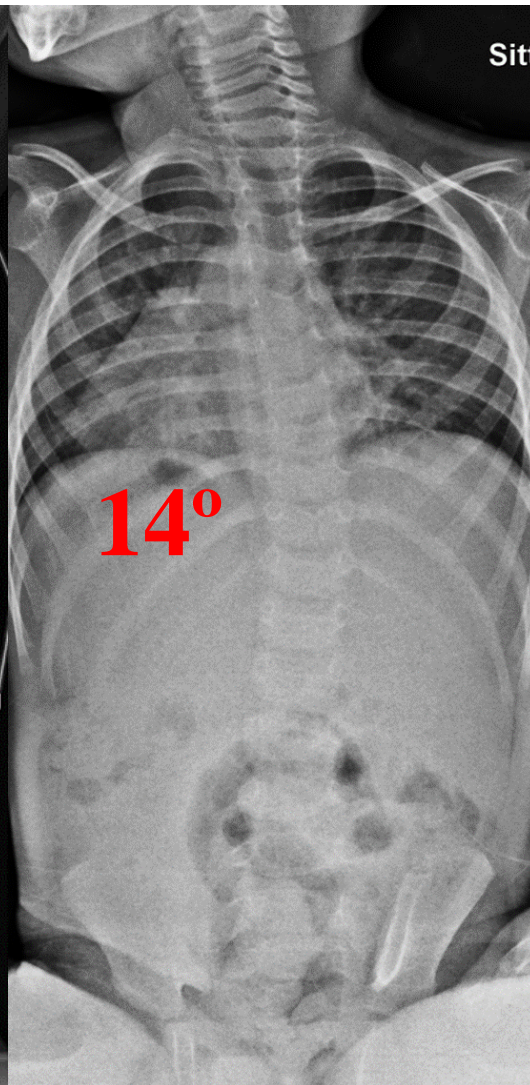
# Timeline of A Cured Curve



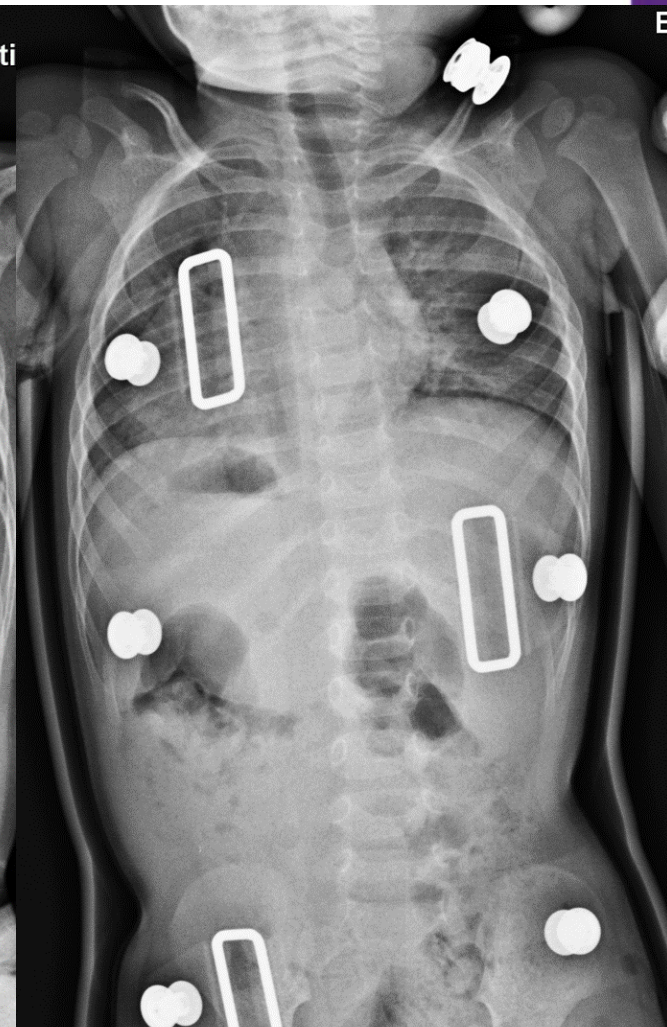
**17 mos old boy  
UPD**



**1<sup>st</sup> cast**



**After 4 casts**



**After 5 casts  
20° curve**

# Followup



4 years old  
2 1/2 years  
post-casting



8 years old  
6 years post-  
casting



9 years old  
25°, restart  
bracing



11 years old  
30° curve

● **New  
technique**  
● **Adolescence**

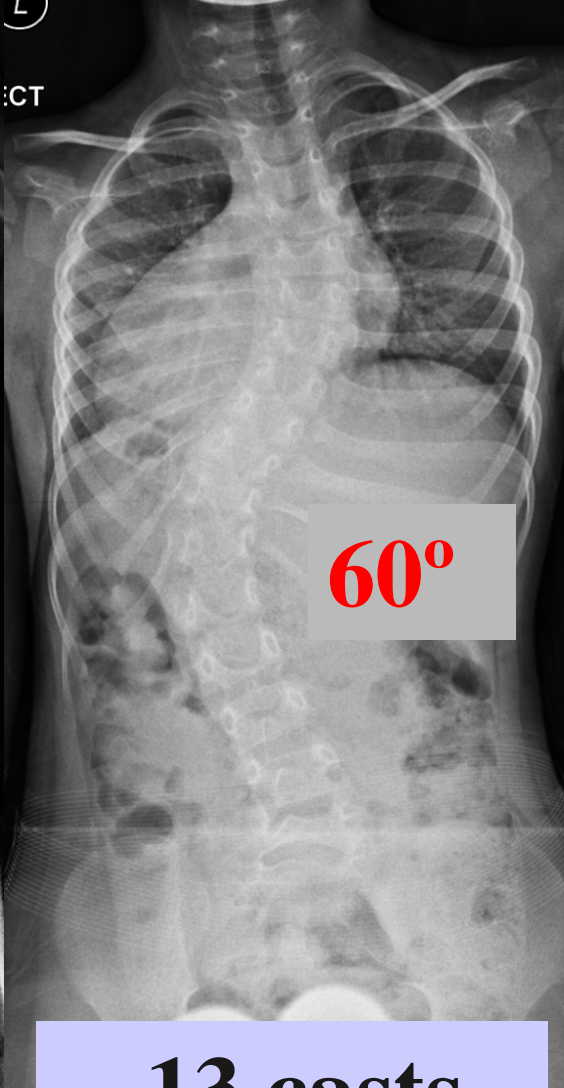
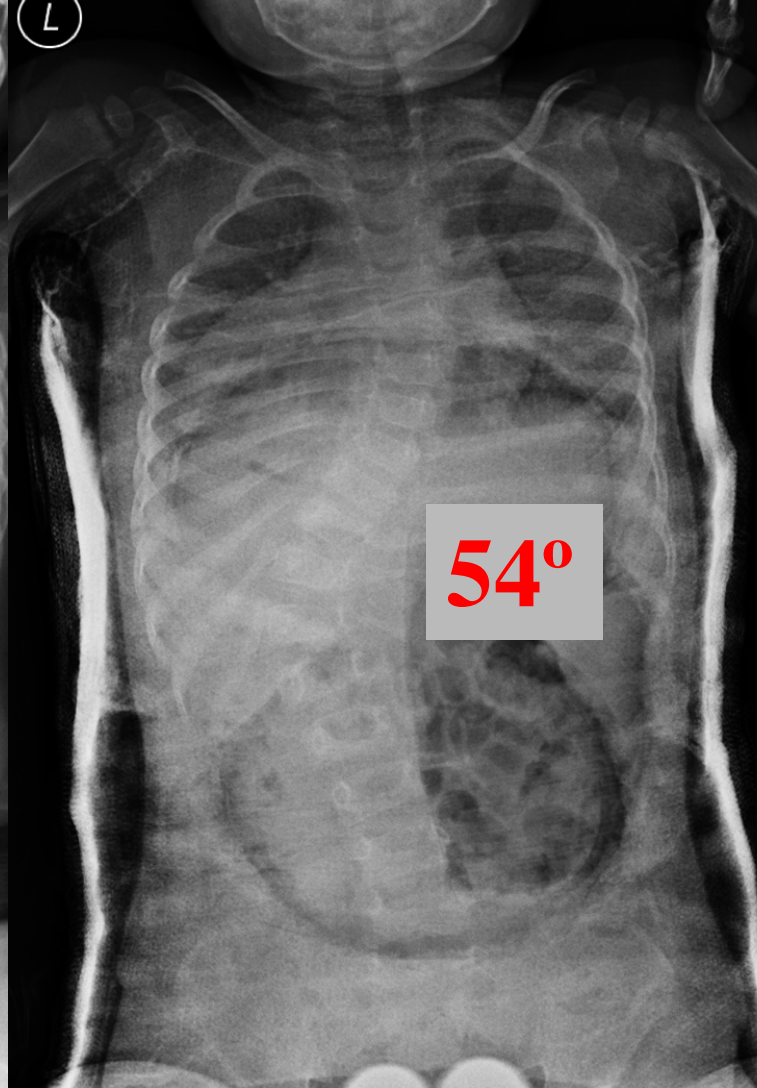
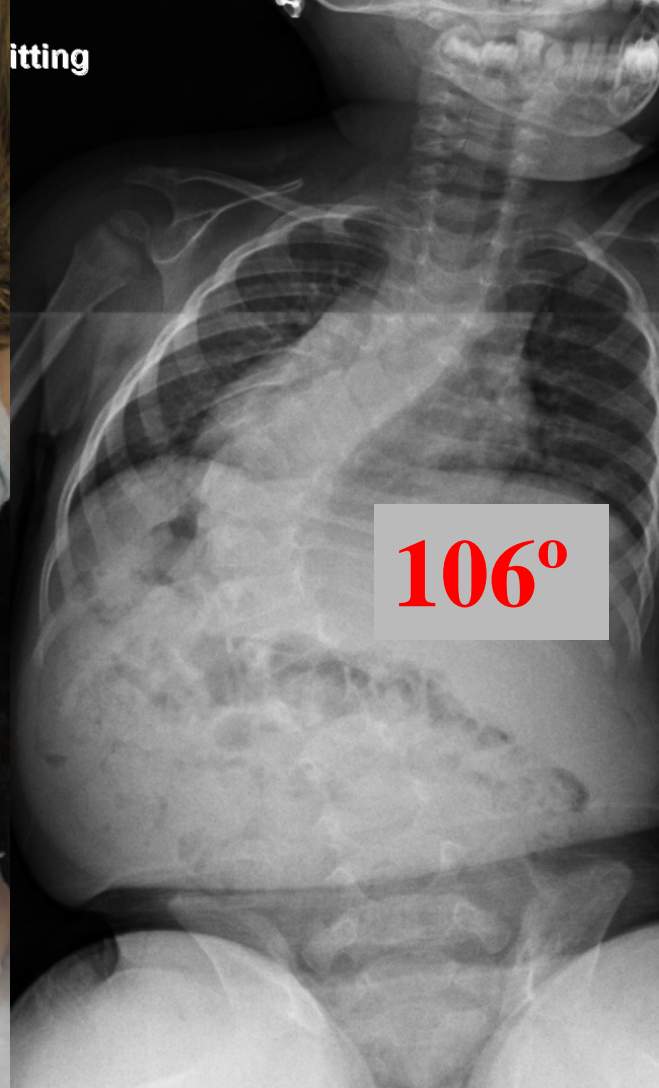


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# Timeline of a Controlled Curve



**18 month old with deletion**

**1<sup>st</sup> cast**

**13 casts  
4 years old**

# Followup



## ● Followup

- ⊕ 18 casts
- ⊕ 4 years later
- ⊕ Curves below 60° in cast
  - ✦ About 65° out of cast
- ⊕ At a good age for expandable implants

13 cast  
4 years old

18 cast  
6 years old



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# Predicting Factors

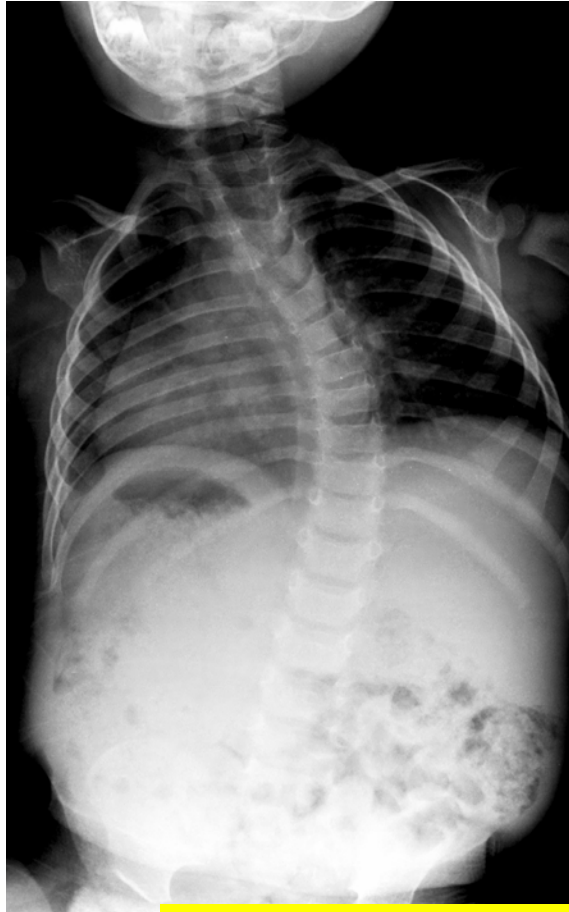
- **No differences seen between those with “Cured” curves and any of the other groups**
  - ⊕ **Gender**
  - ⊕ **Sidedness (right versus left)**
  - ⊕ **Curve region (thoracic versus lumbar)**
  - ⊕ **Age at cast initiation**
- **Initial curve  $<50^\circ$** 
  - ⊕ **Odds Ratio 9,  $p=0.008$**



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# Casting Is Survivable



Having scoliosis is tough...

...but I am tougher! 



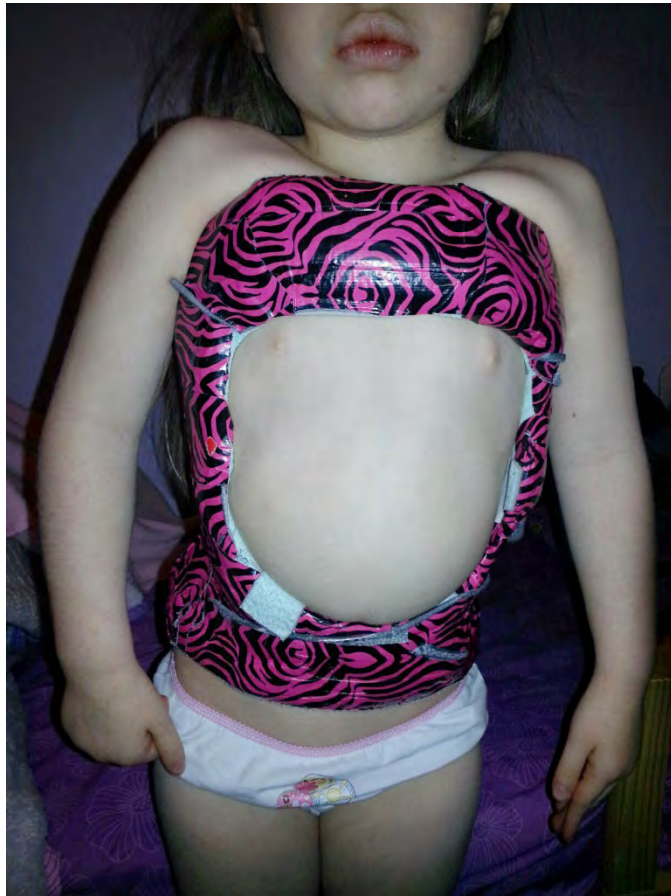
- 17 month old boy with  $55^\circ$  curve
- 5 casts over 15 months, braced for 12 months
- Now, 4 years old with  $13^\circ$  curve, no brace

Hope  
is

2023  
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 SLUCare  
Physician Group  
SAINT LOUIS UNIVERSITY

# Casting is survivable!



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# Bracing

- **For curves larger than 20° - 25°**
- **Prevent curve progression**
- **Cannot (usually) make a curve smaller**
- **Difficult to fit on obese children**



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# Bracing

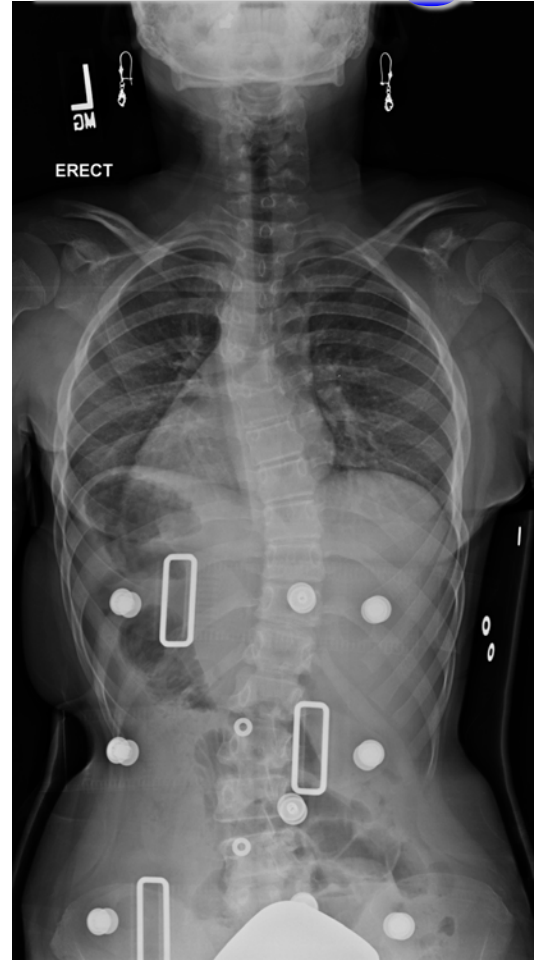


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# Bracing



**January 2009**  
**32°**



**January 2009**  
**First brace**



**March 2013**  
**44°**



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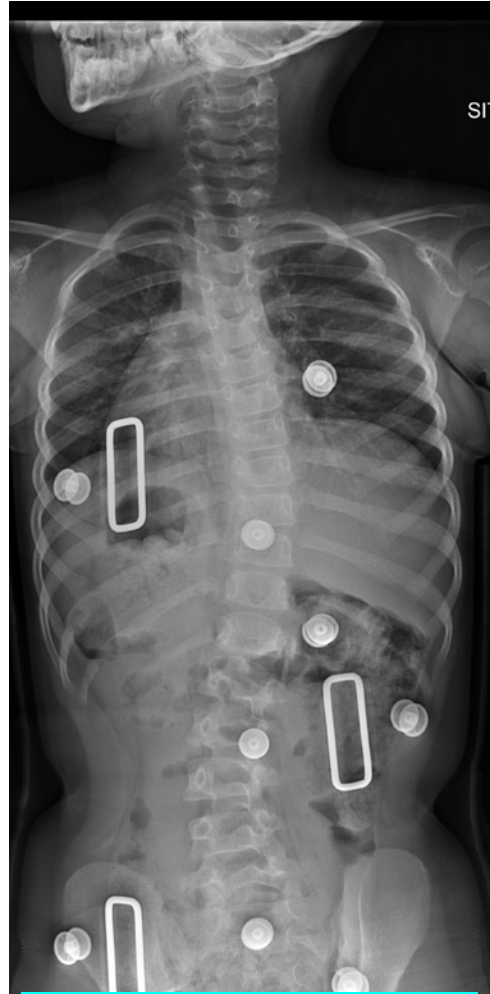


# Bracing

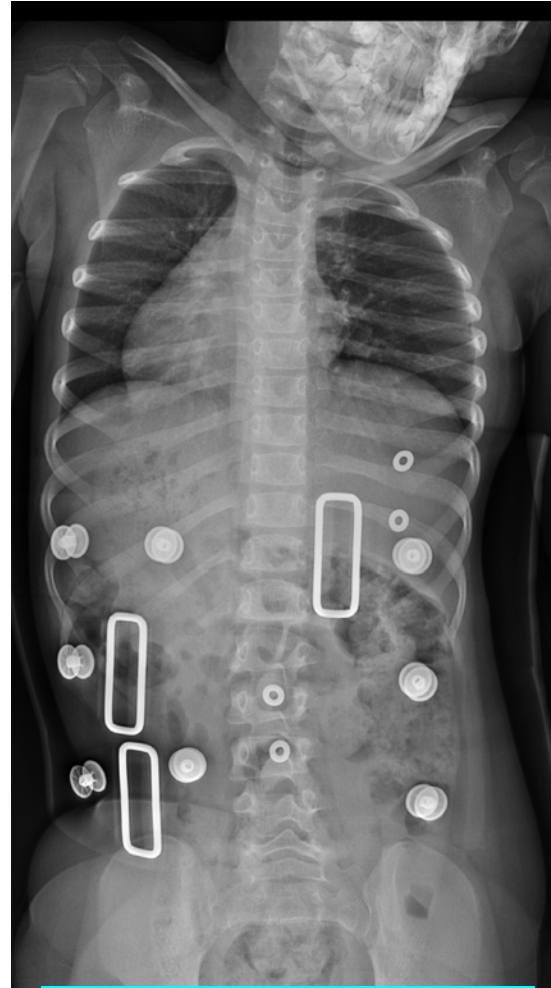
5 year old girl, PWS/imprinting defect



Dec 2014, 5 y.o. 46°



Jan 2015, 22°  
Daytime brace



Jan 2015, 2°  
Nighttime brace



Nov 2018, 9 y.o. 21°

Hope

2023  
CONVENTION

# Bracing

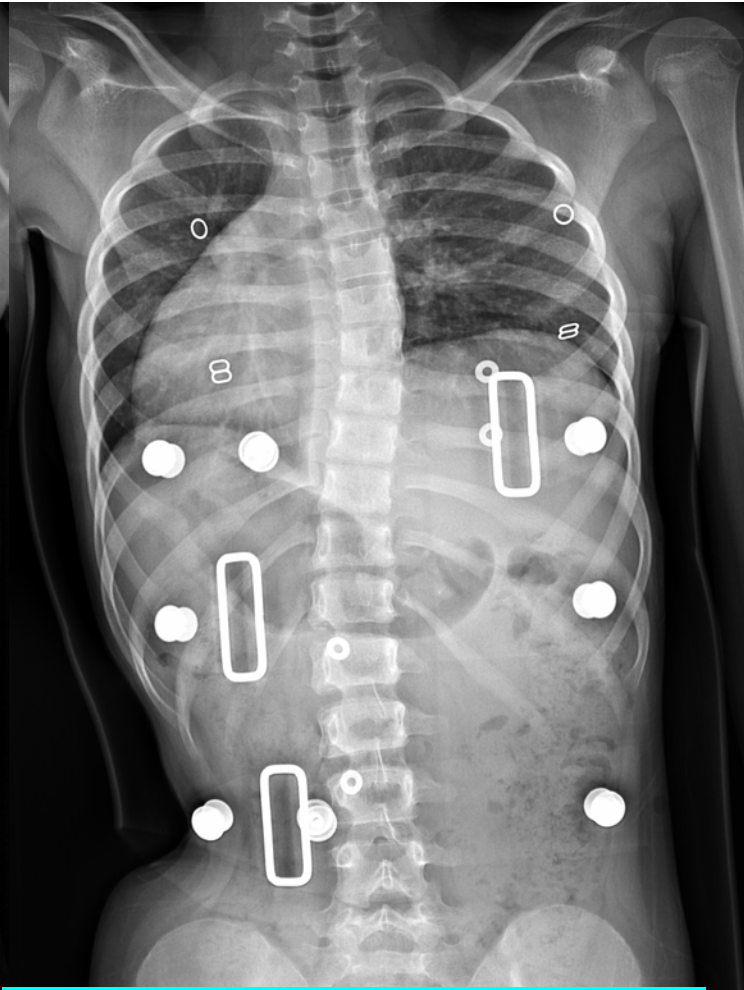
10 year old girl, PWS/del



10 years old  
37° and 41° curves



10 years old  
in TLSO



10 years old  
in Providence brace



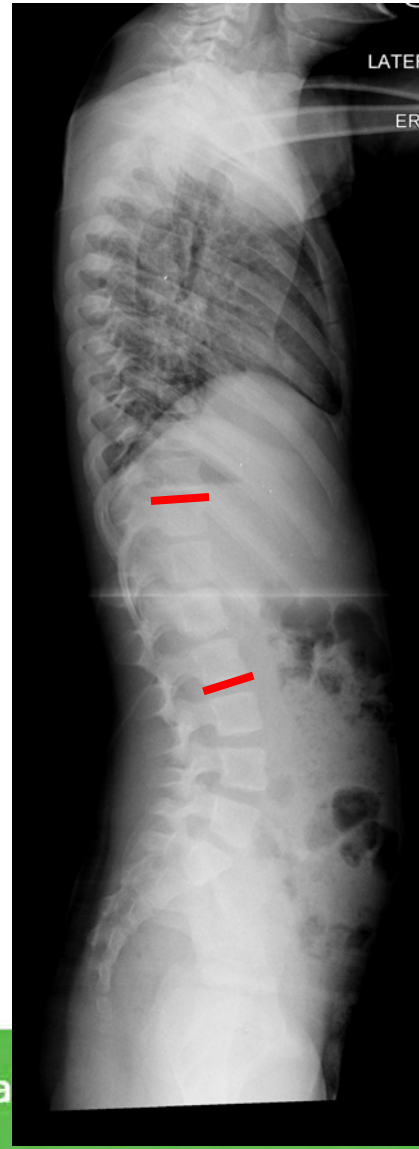
13 years old  
25° curve

-Willi S... on | USA

2-040

# Bracing

20 month old boy, PWS/del



- 20 months old  
51° lumbar kyphosis
- 2 1/2 years old, in brace
- 4 years old  
15° residual kyphosis



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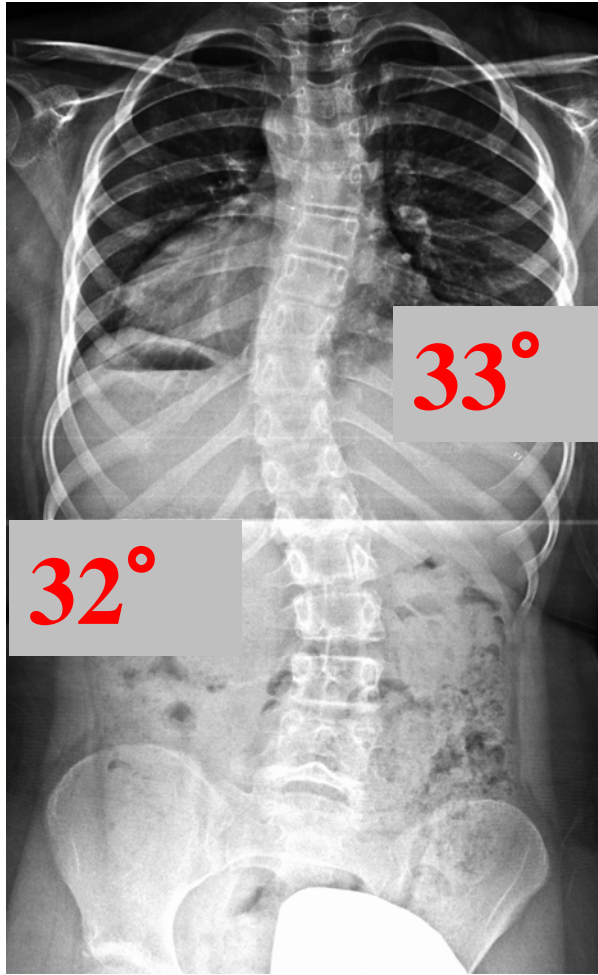
[pwsausa.org](http://pwsausa.org) • (941) 312-0400





# Bracing

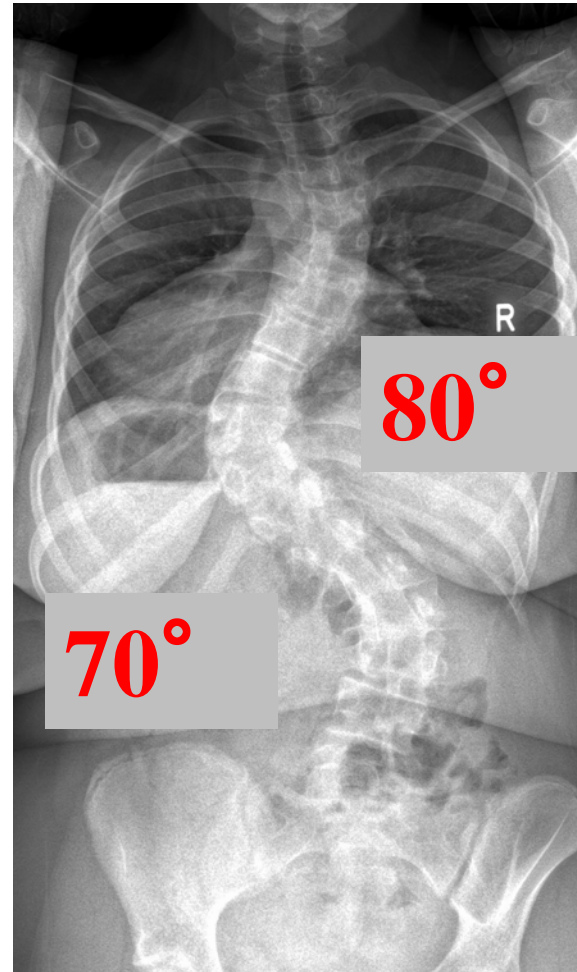
**10 year old girl, PWS/UPD**



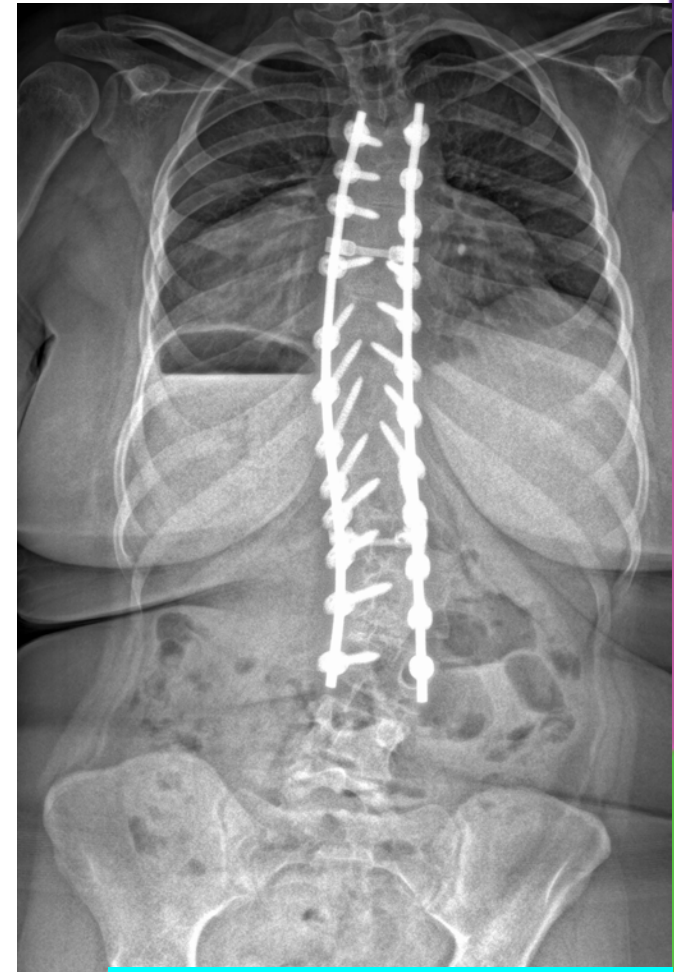
**10 years old  
33° and 32° curves**



**12 years old  
In brace**



**15 years old  
80° and 70° curves**



**19 years old  
4 years after fusion**

# Surgery

- **Surgery is indicated for curves between 40° and 50°**
  - ⊕ **Nearing maturity**
- **Align spine in best position**
  - ⊕ **Side to side curve (scoliosis)**
  - ⊕ **Front to back alignment (kyphosis/lordosis)**
- **Hold in position**
  - ⊕ **Rods**
  - ⊕ **Hooks, wires, and screws**



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# Surgery

## ● **High rate of complications in PWS**

### ⊕ **Infections**

✦ **Skin picking**

### ⊕ **Anaesthetic (intra or peri-operative)**

### ⊕ **Pulmonary/Respiratory**

✦ **Apnea**

### ⊕ **Hardware failure/pseudoarthrosis**

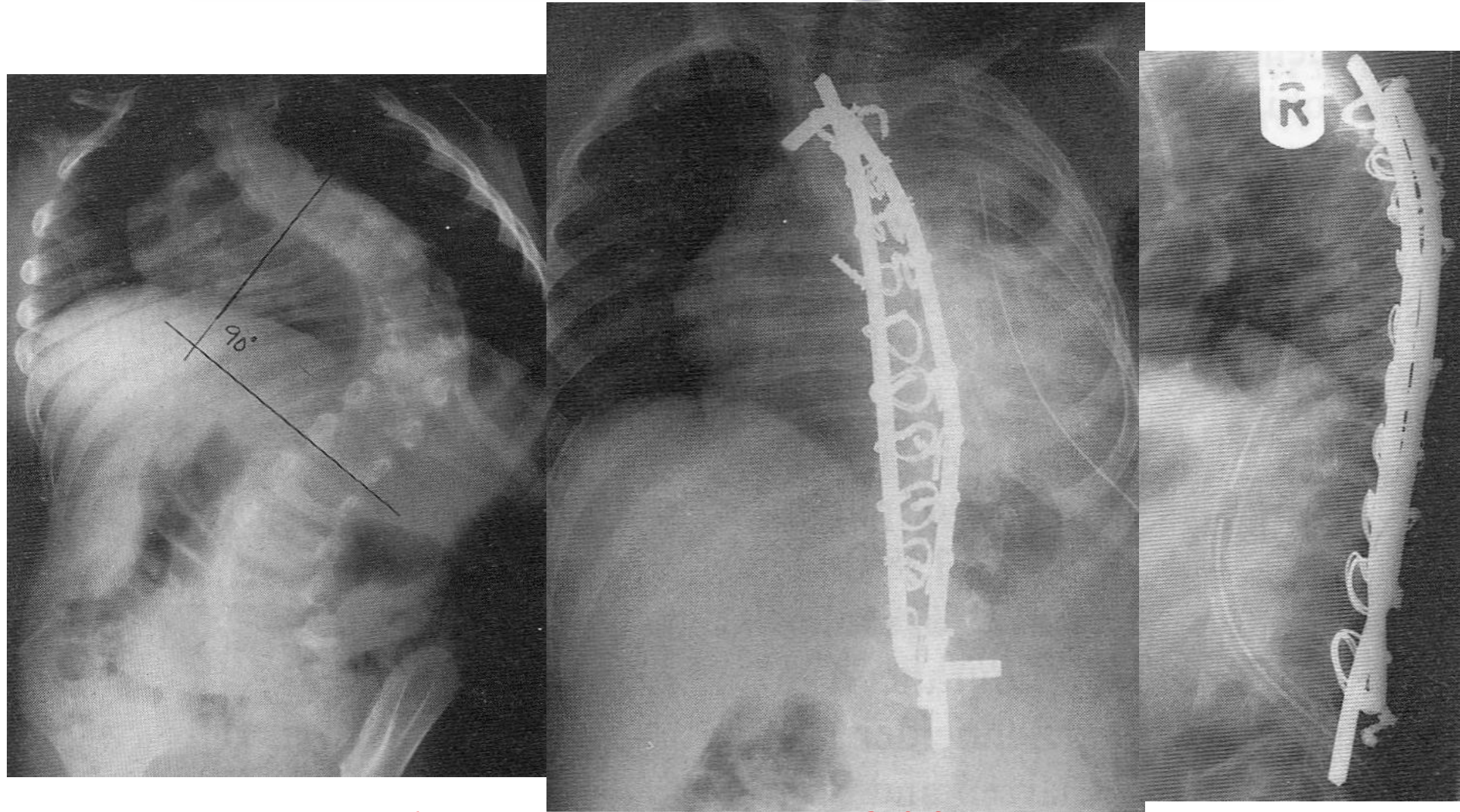
✦ **Osteoporosis**

### ⊕ **Need to continuously educate treating surgeons to these special risks**



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# Curves in Young Children

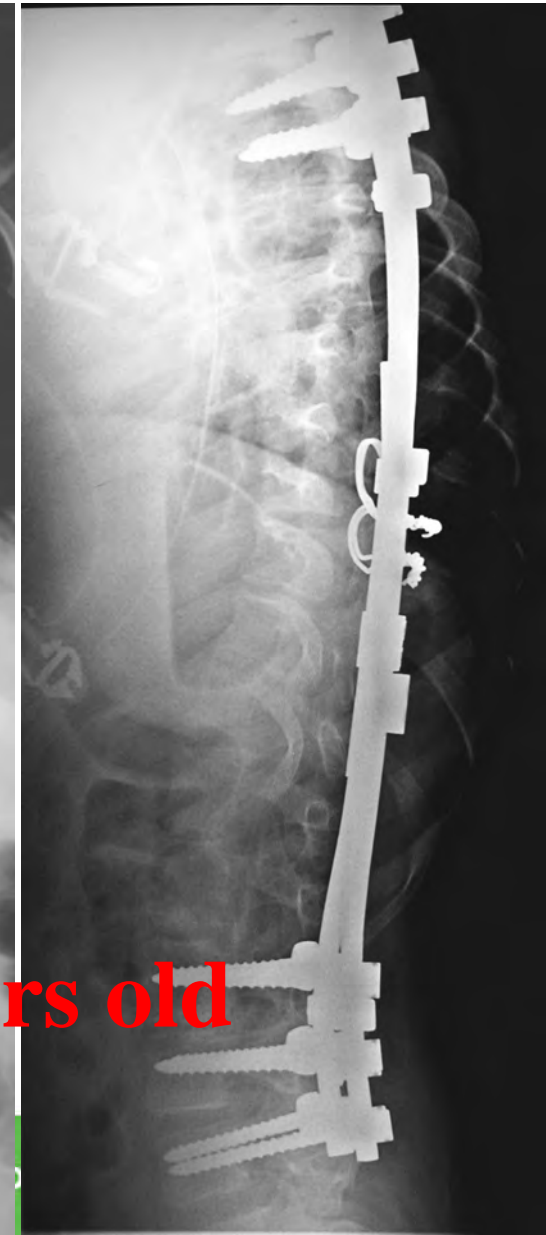
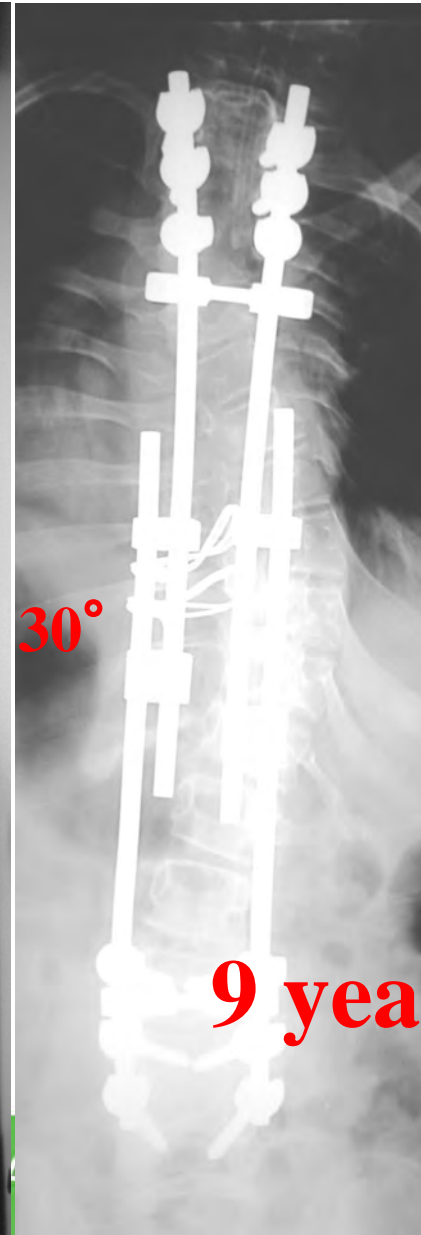
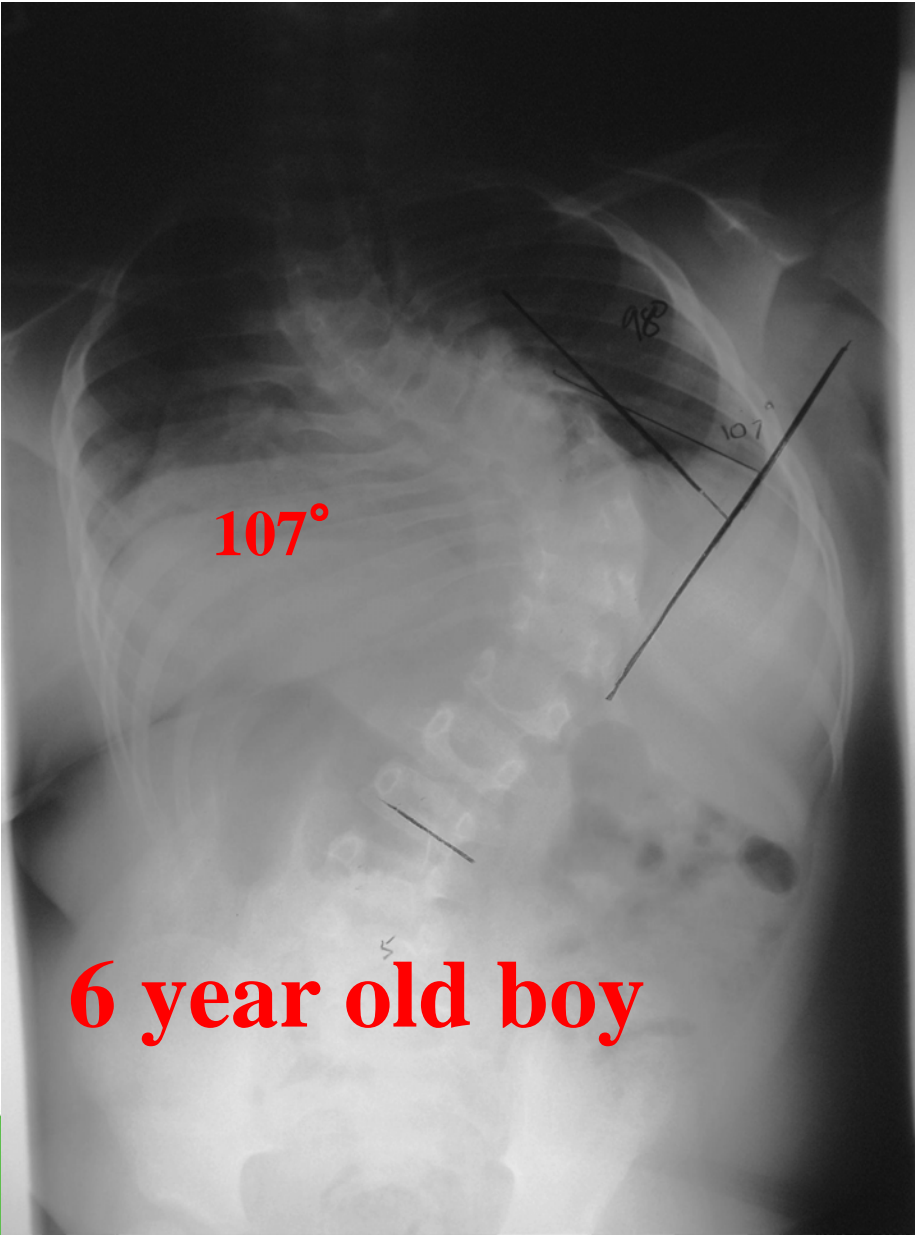


**6 year old boy, 90° curve**



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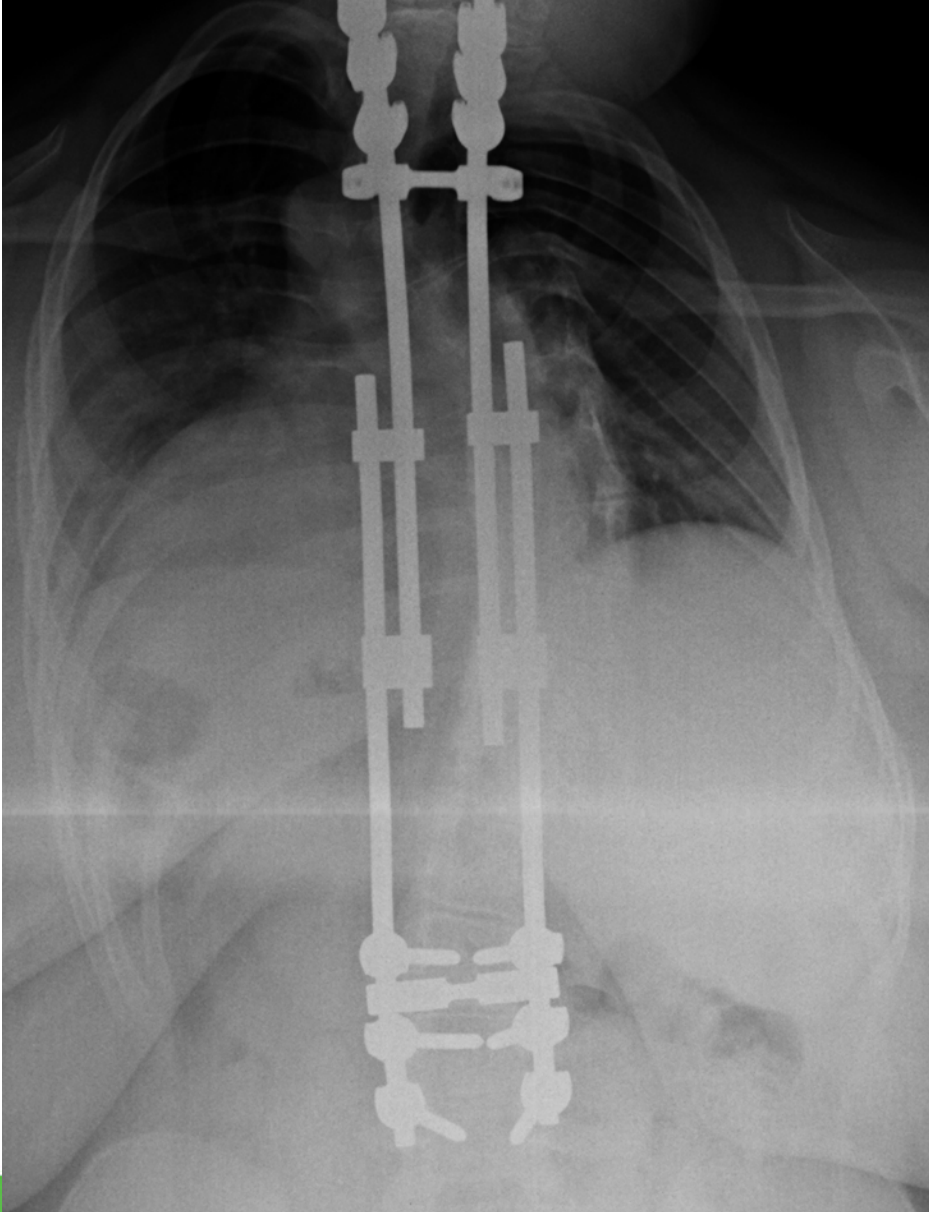
# Non Fusion Spinal Instrumentation



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# 6 year old with 107° curve



## ● Followup

⊕ 16 years old

⊕ ~50° curve

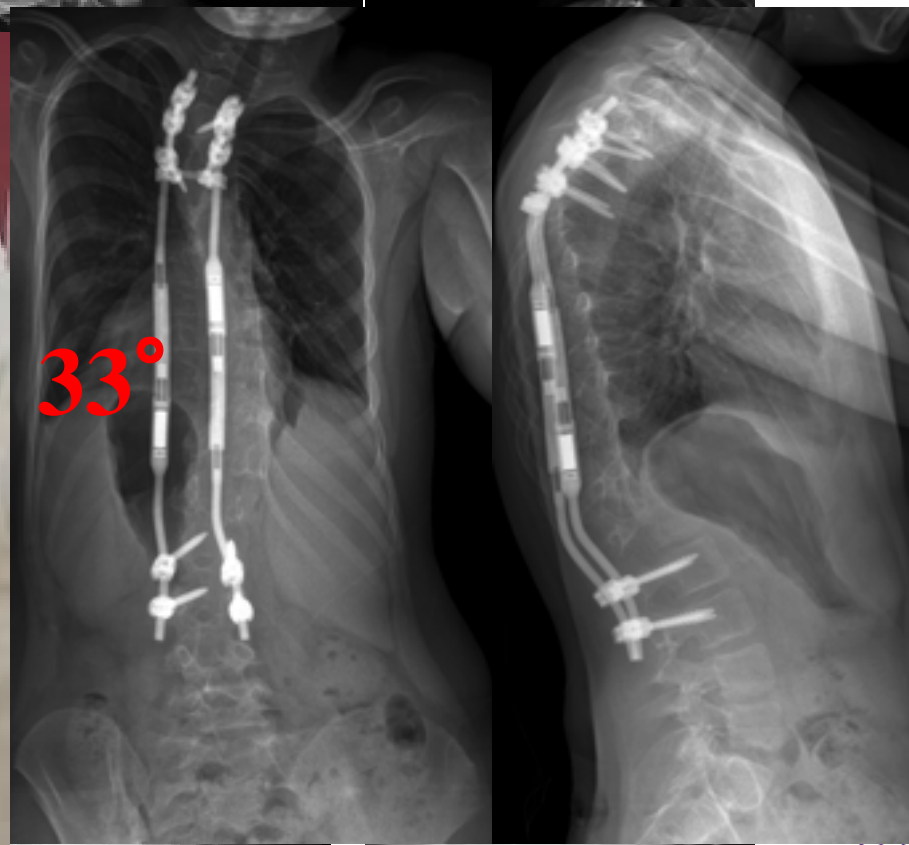
⊕ “Graduated”



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# MAGnetic Expansion Control MAGEC System

Electric  
Remote  
Control



**10 year old boy with PWS** After 2 lengthenings

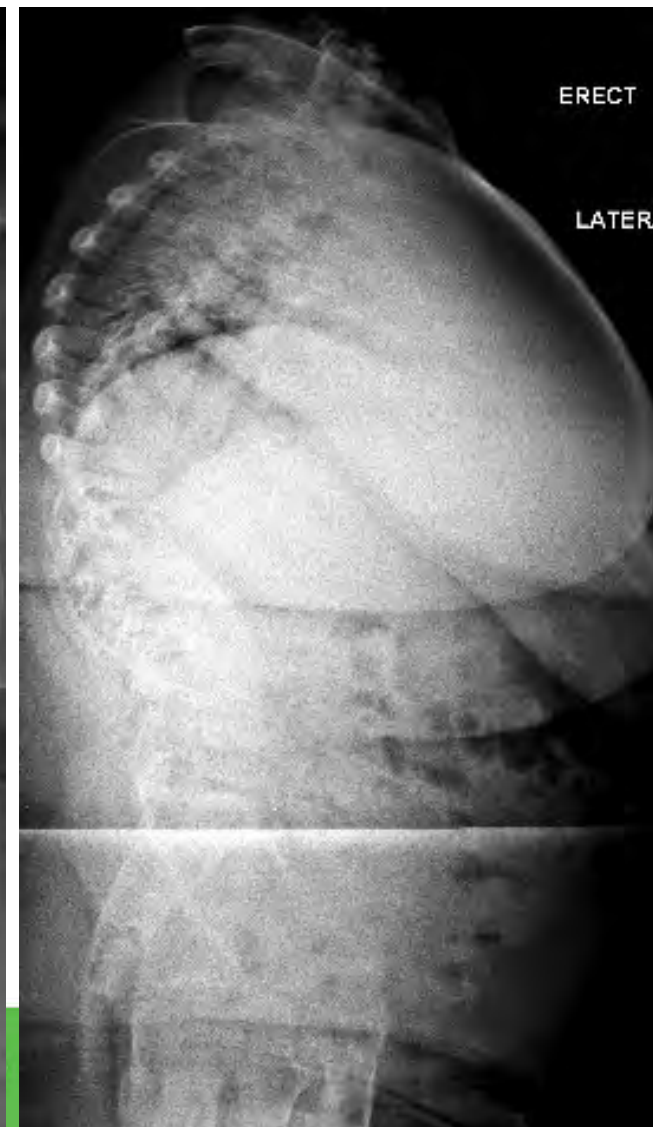
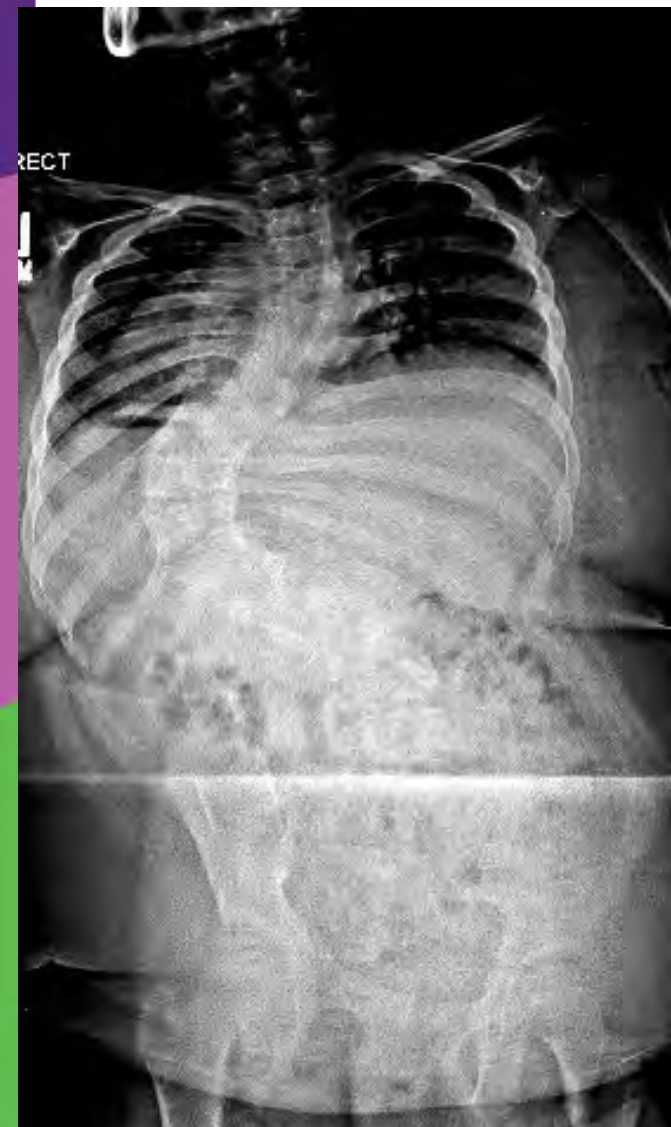


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10 y.o. girl, 103°

End-game  
Graduation

15 y.o.





# Spinal Fusion

- **For curves over 50° at maturity**
- **Timing of surgery**
  - ⊕ **Balance expected maturity with curve size**
  - ⊕ **My preference in younger patients:  
delay until the curve is over 50° in brace**
- **Avoid anterior approach**
- **Newer pedicle screw instrumentation**
  - ⊕ **Better in osteopenic bone**



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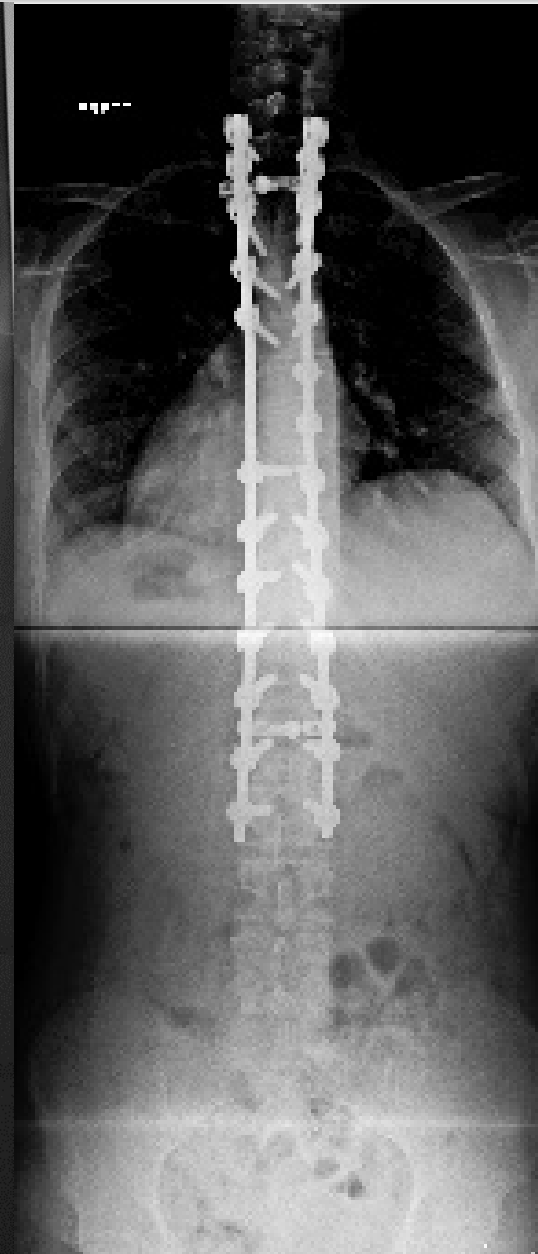
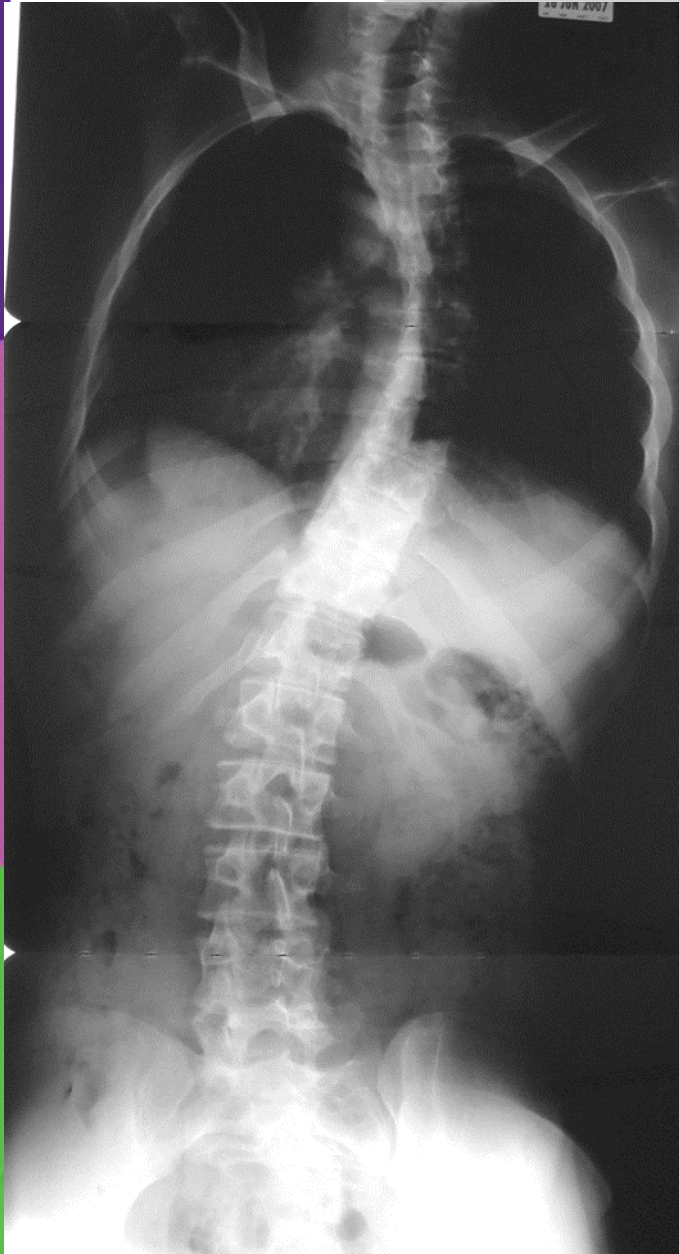
# 15 y.o. girl with 67° scoliosis



# 18 y.o. with kyphosis



# 25° scoliosis and 110° kyphosis



# Surgery and PWS

## Pre-operative

### ● **Pulmonology work-up**

- ⊕ **Sleep study for apnea (need post-op CPAP or BiPAP?)**
- ⊕ **Possible ENT evaluation**

### ● **Anaesthesia evaluation**

- ⊕ **Assess airway management**
  - ✦ **Ability to intubate**
  - ✦ **Thickened saliva**
  - ✦ **IV access – may need central line**



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# Surgery and PWS

## Other Factors

- **Be aware of food s(n)eeking behaviors**
- **High pain threshold**
  - ⊕ **May be difficult to awake**
  - ⊕ **Often more compliant with post-op therapy**
- **Airway management, apnea**
  - ⊕ **Consideration to delay extubation**
  - ⊕ **PICU for observation, CPAP or BiPAP**
  - ⊕ **Hypotonia – poor cough**



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# Surgery and PWS

## Other Factors

### ● **Skin picking**

- ⊕ **Infections**

### ● **GI complications**

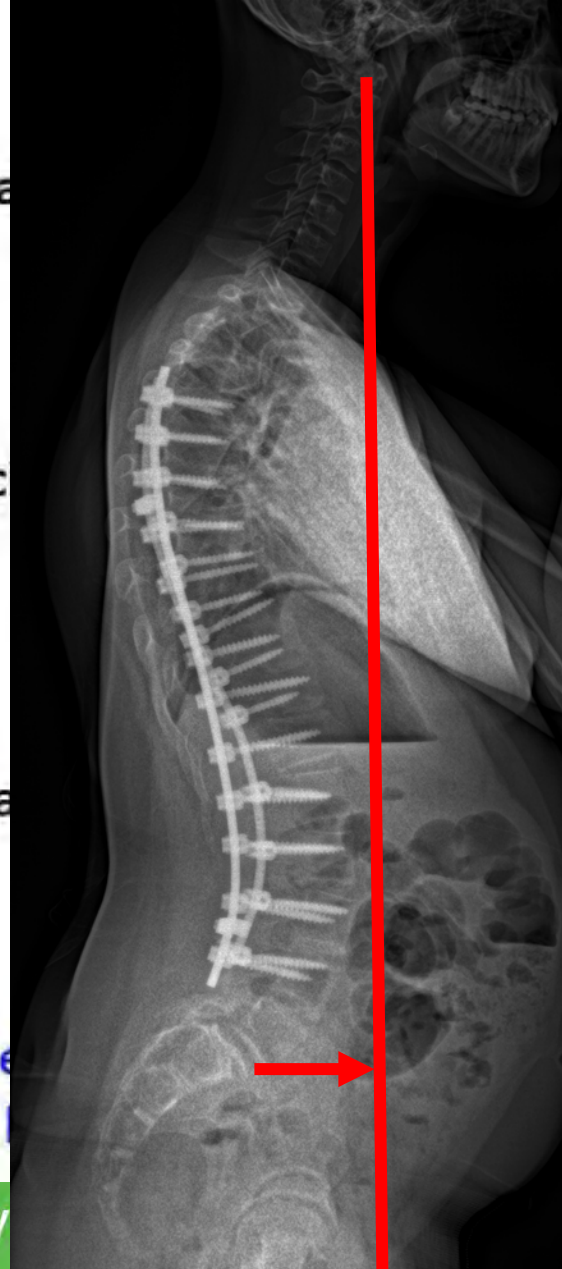
- ⊕ **Gastro-motility slows down**

- ⊕ **Very gradual increase in post-op diet**



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# Typical Sagittal Alignment



## ● Posture of the spine

- ⊕ A “plumb” line from the upper neck vertebra
- ⊕ Passes just front of sacrum
- ⊕ To the center of the hips

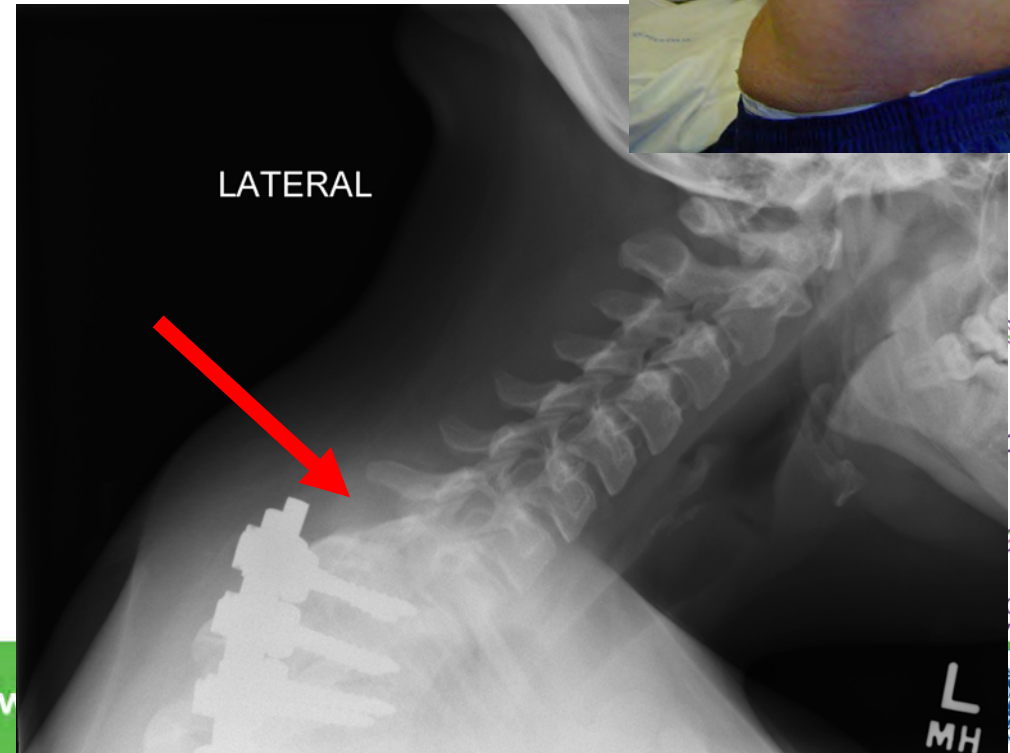
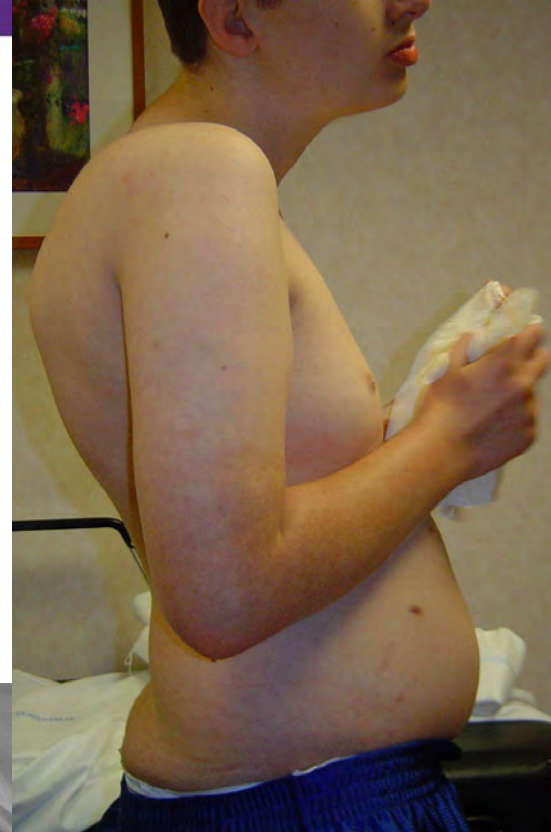
## ● PWS

- ⊕ More head forward posture
- ⊕ Plumb line more forwards
- ⊕ Need to preserve surgically



# Cervico-Thoracic Kyphosis

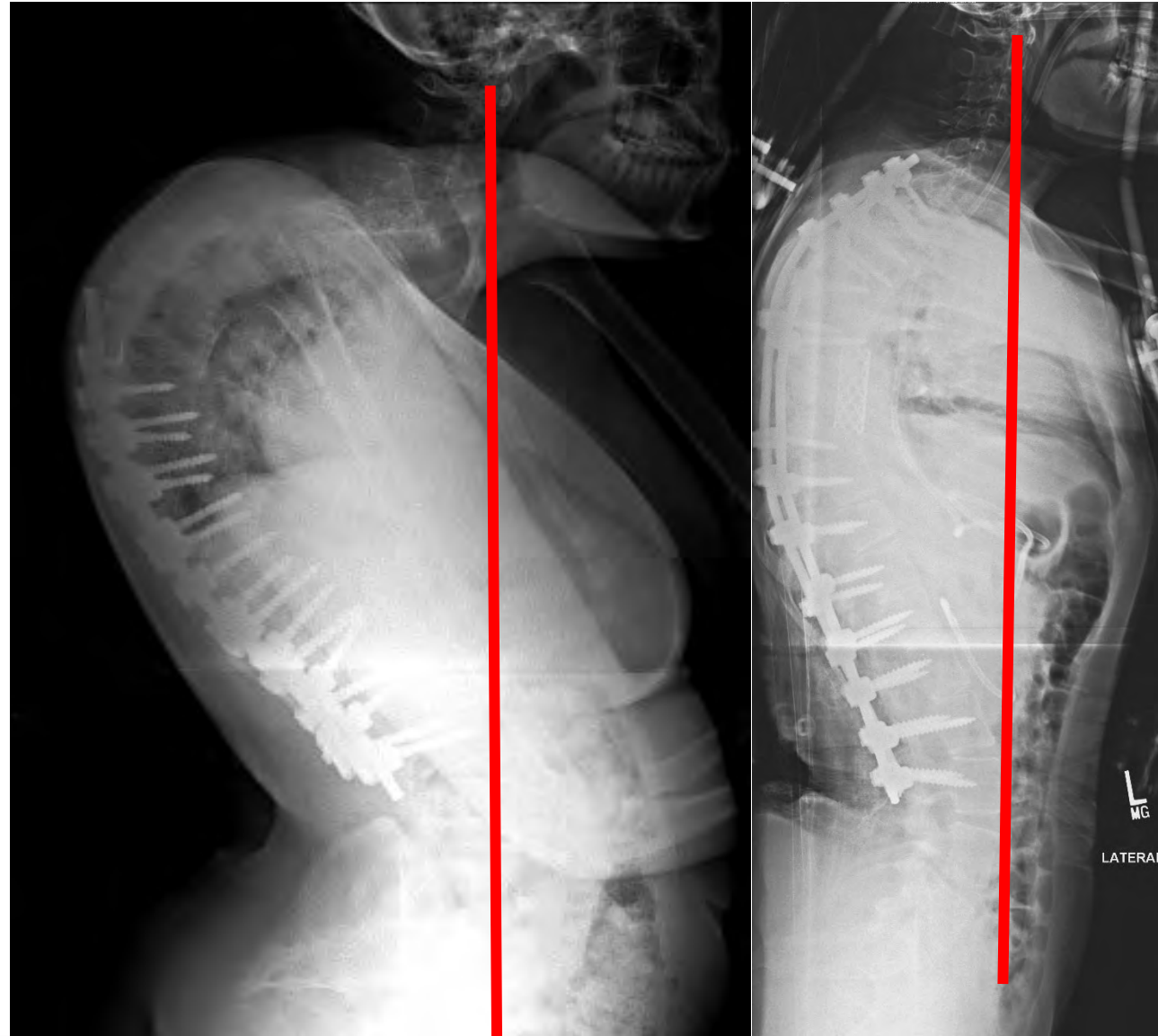
- **Exaggerated forward positioning of the head**
  - ◆ **Forward head thrust**
- **Characteristic of PWS posture**
- **Can be significant problem**
- **Can worsen after spine surgery**



# Sagittal Alignment



# Reconstructions



**17 year old girl**



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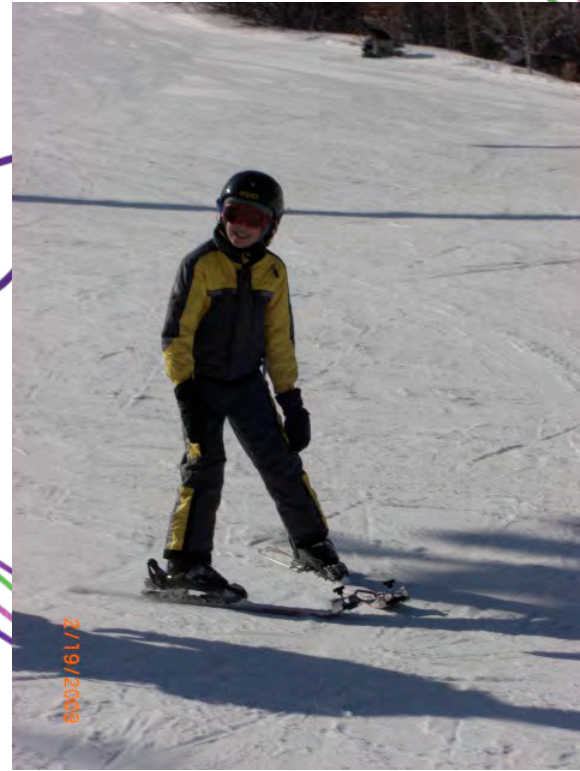
# Growth Hormone and Scoliosis

- **Concerns of worsening scoliosis with GH**
  - ⊕ **Turner syndrome**
    - ✦ **Scoliosis worsens with GH**
- **PWSA 2007 Survey**
  - ⊕ **For every month GH not started, risk of surgery increases 0.7%**
- **My “take” on it**
  - ⊕ **Scoliosis will progress comparative to growth**
  - ⊕ **GH has many important benefits**



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# People with PWS are living longer



USA | USA 2023



## We work together for a better quality of life

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