

AGAINST

Correction by traction followed by cast or brace in the treatment of

SYNDROMIC spine deformities such as.....

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MUCOPOLYSACCHARIDOSIS



2 mois



18 mois



5 ans



8 ans



13 ans

Early Kyphotic Deformity





Kyphosis: 50% of the patients

10 ans



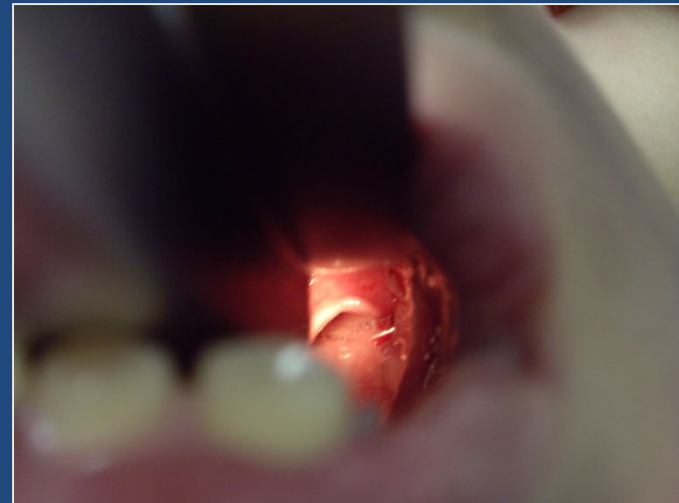
Scoliosis: 20% of the patients



For the anesthesiologists veinous access is difficult



Airway access is difficult



Intubation using fibroscope

Laryngeal mask

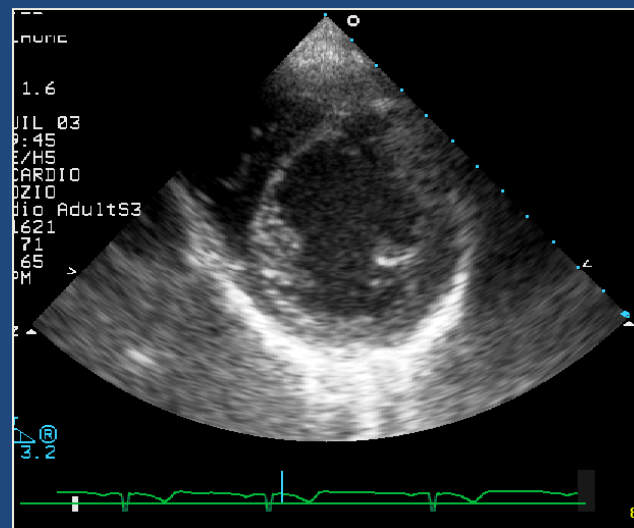
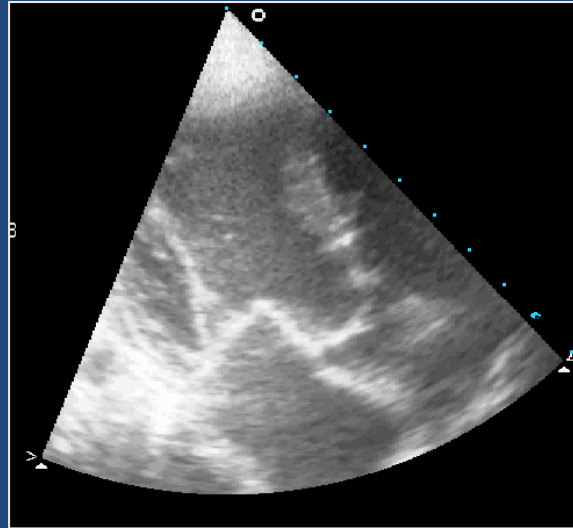




Ventilation difficulties

- Broncho-pulmonary obstruction
 - Asthma
-
- Broncho/laryngic spasma
 - High insufflation pressure
 - Lung oedema (Hypoxemia, obstruction)
 - Atelectasia

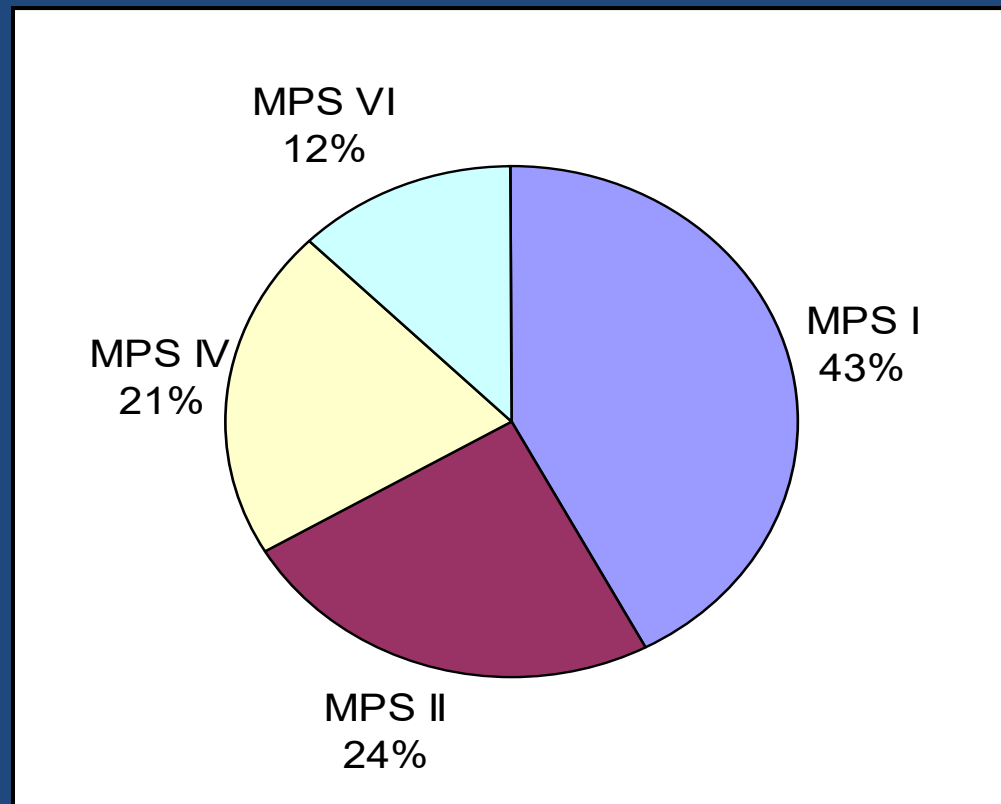
Cardiomyopathy



N= 175 patients

Maroteaux – Lamy disease

Morquio disease



*Hurler et Scheie
Disease*

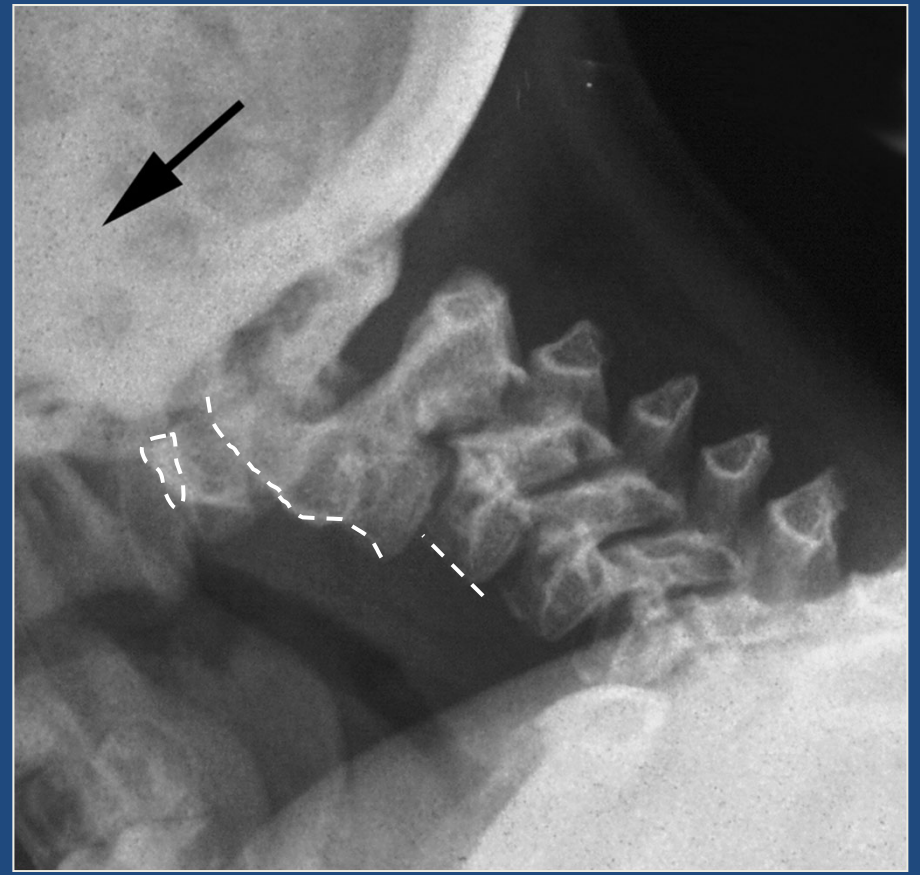
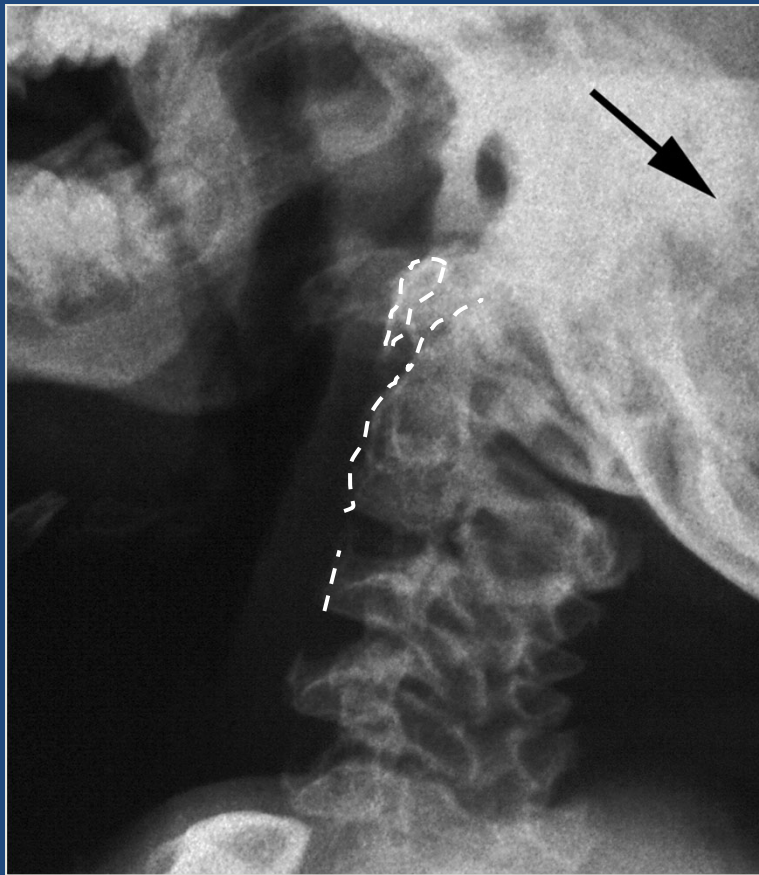
Hunter disease

Symposium of the French Society of Paediatric Orthopaedic 2006

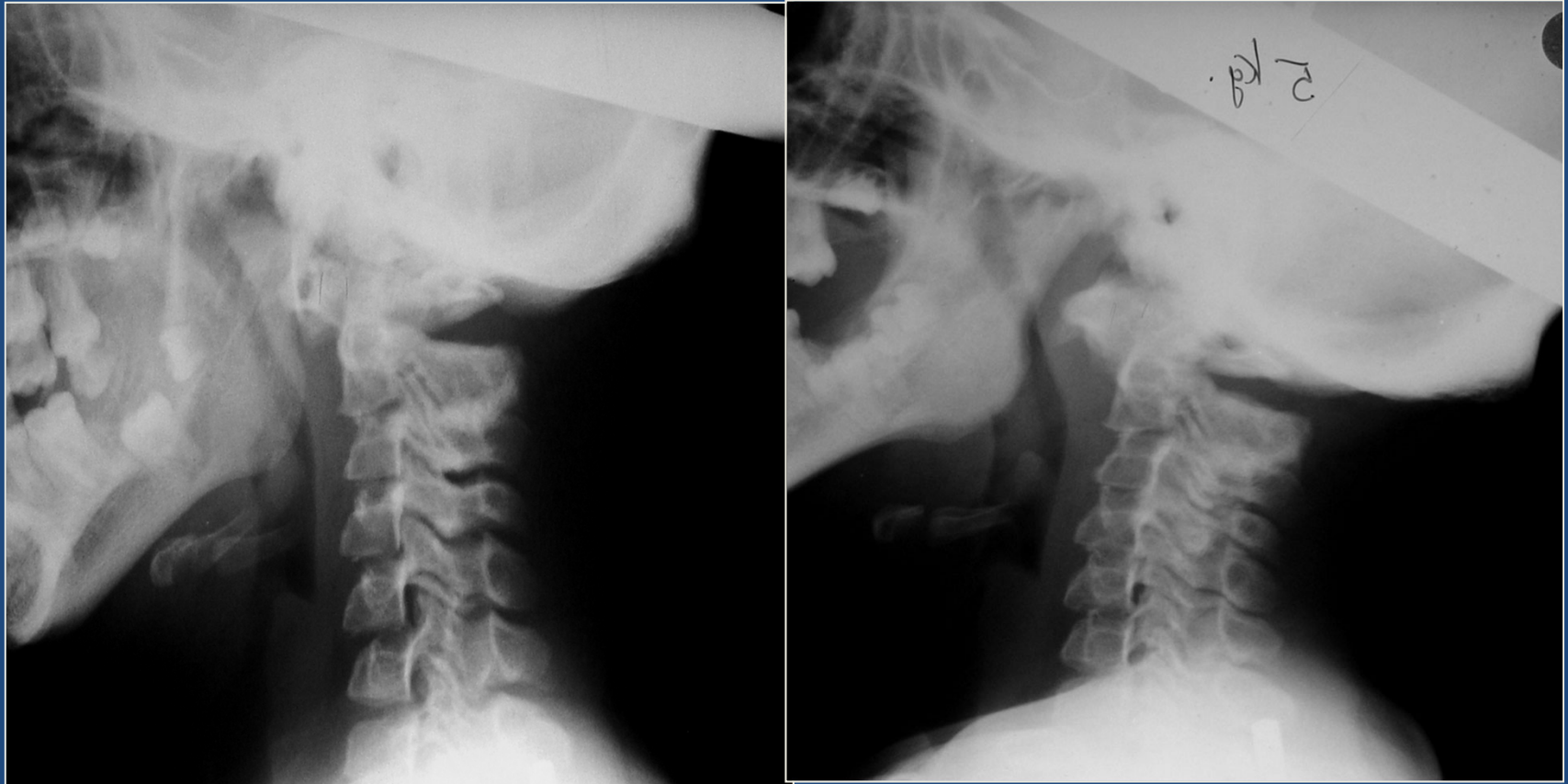
Instability

- 9 pts / 175 (5%)
 - 5 pts with medulla impingement and tetraparesia
 - 8 MPS IV (*Morquio*)
 - 1 MPS I (*Hurler*)
- Surgery for everyone
- 1 Death: respiratory failure
- 5 neurological improvement

Instability



Instability



Occipito-cervical stenosis

- 41 pts / 175 dossiers (23%)
 - 13 MPS I (*Hurler et Scheie*)
 - 1 MPS II (*Hunter*)
 - 19 MPS IV (*Morquio*)
 - 8 MPS VI (*Maroteaux – Lamy*)

Occipito-cervical stenosis



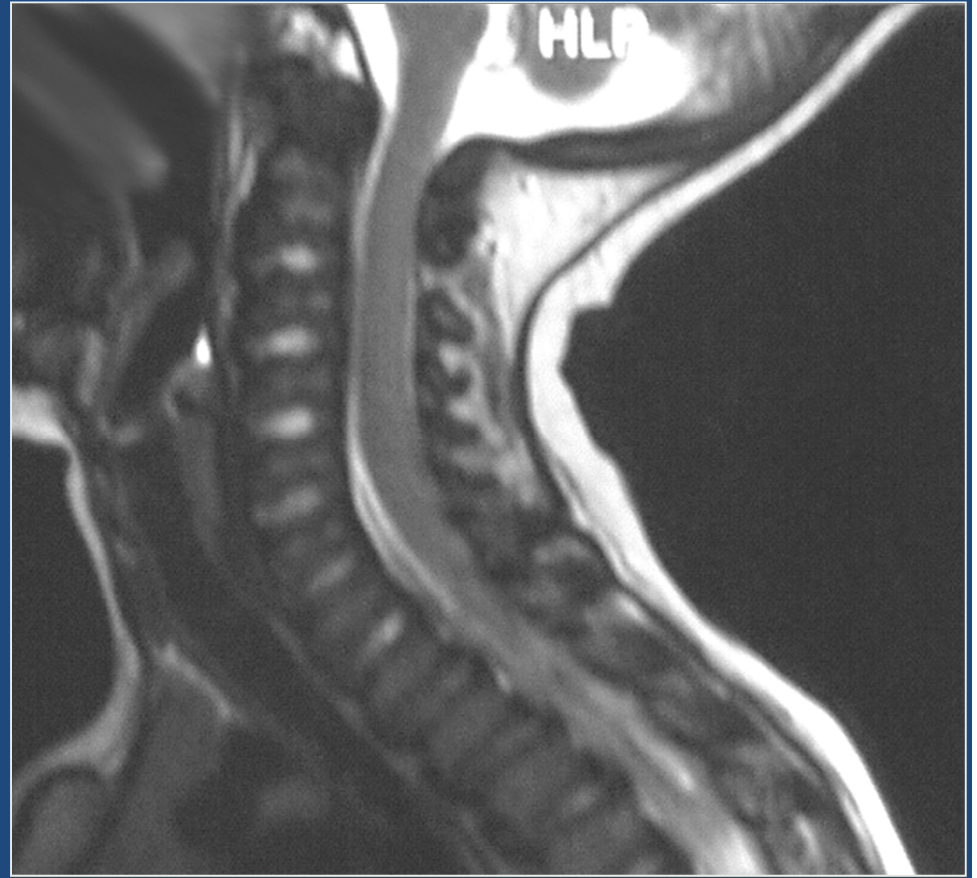
Distal cervical stenosis

- 5 pts / 175 (3%)
 - 3 MPS I (*Hurler et Scheie*)
 - 1 MPS IV (*Morquio*)
 - 1 MPS VI (*Maroteaux – Lamy*)

Distal cervical stenosis



Associated stenosis + instability



Strategy

- Sténosis = décompression
- Instability: Occipito-cervical fusion



Halo-Vest

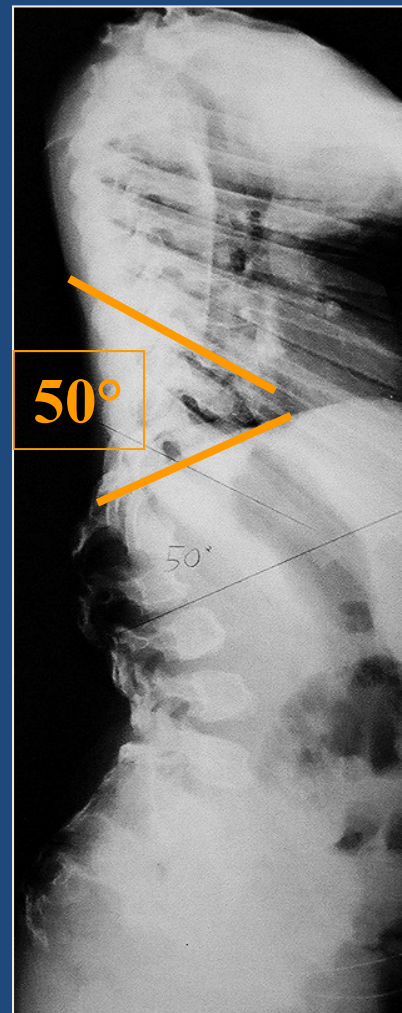
Fusion

Ant + Post Fusion

N= 18 pts

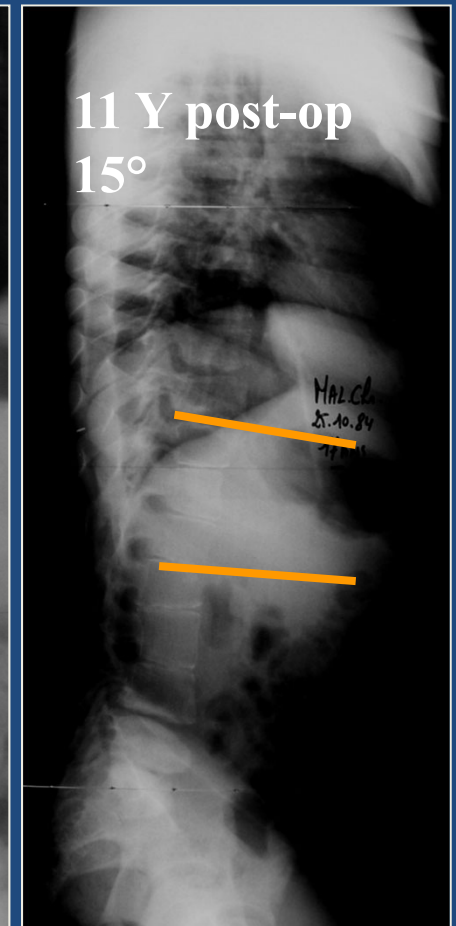
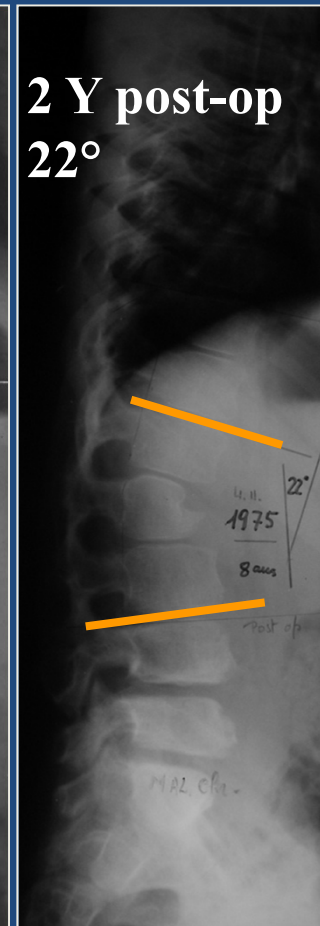
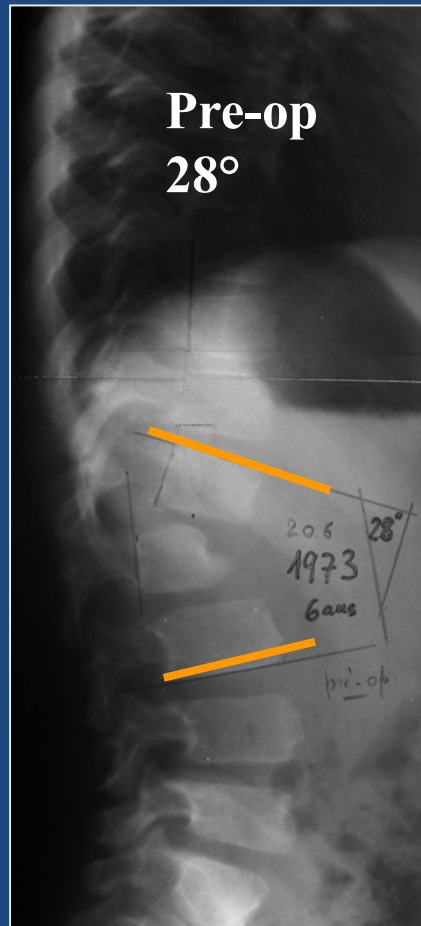
$50^\circ < \text{Cobb Angle} < 90^\circ$

Girl 12 Y
MPS I (*Hurler*)



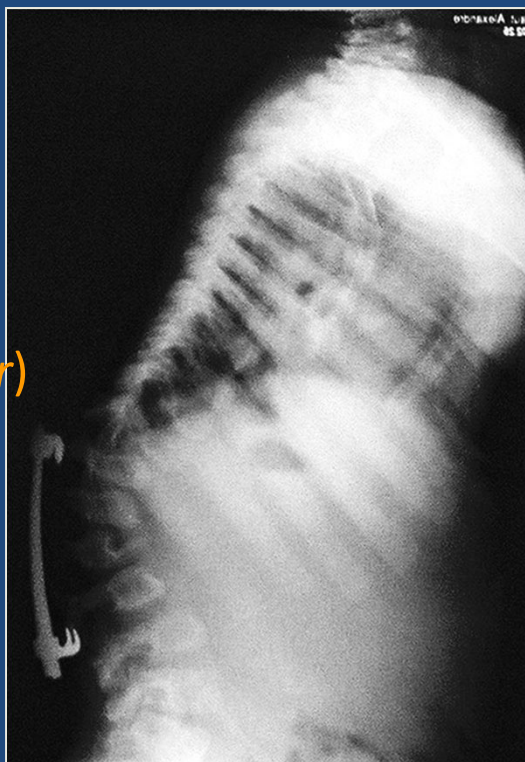
Post Epiphyseodesis

N= 2 pts

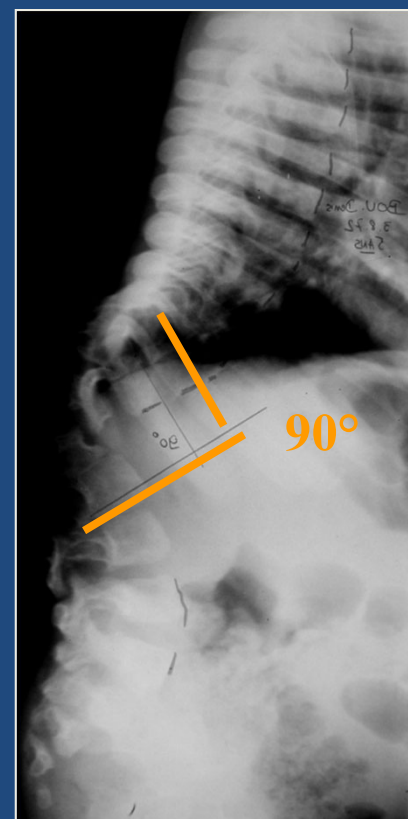


Boy 6 Y MPS II (Hunter)

9 Y
MPS I (*Hurler*)



14 Y
MPS I (*Hurler*)



Neurological Complications

Apex of the Kyphosis: 3 cas

Boy 14 Y, MPS IV (*Morquio*)

Spastic Tetraparesia

Ant medulla release + ant-post fusion

