

THE BANDWAGON I JUMPED ON AND THEN OFF...

No financial disclosures



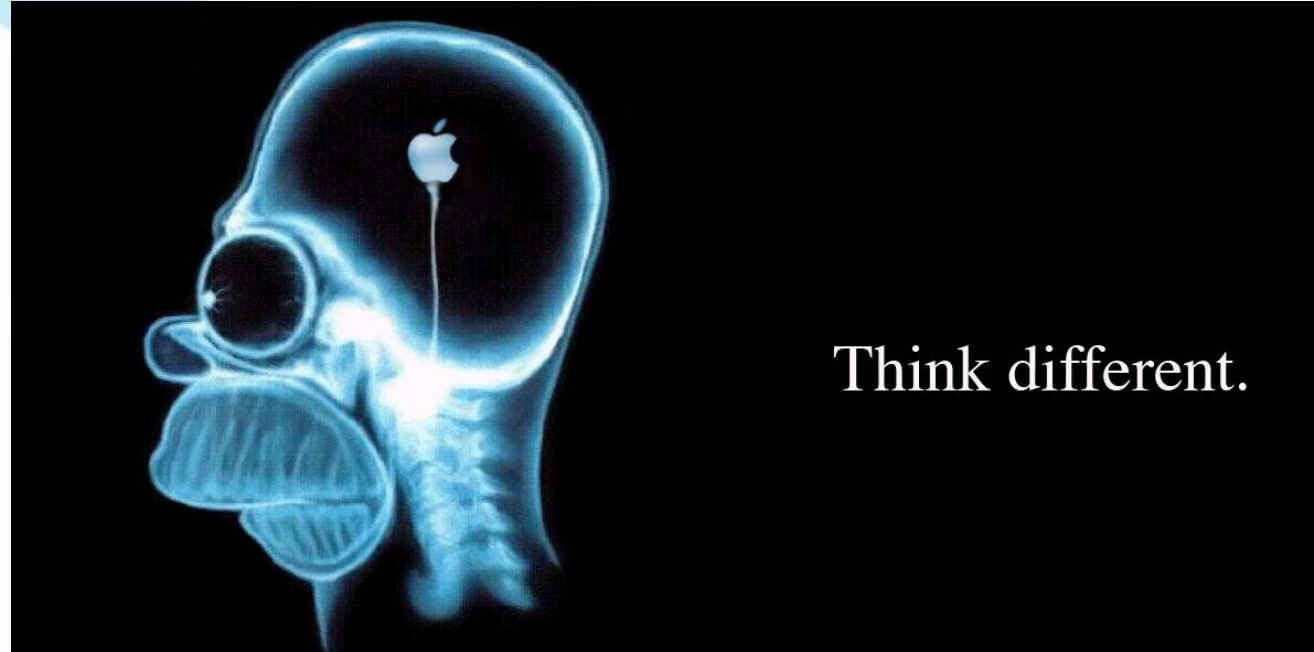
If the woman is always right,
and a man is always wrong.
Then, if a man tells a woman
that she's right.



Is the man right
or wrong?



**THE BANDWAGON
I JUMPED ON
AND THEN OFF...**



**EARLY CORRECTION
OF KYPHOSIS IN MPS I**



EARLY CORRECTION OF KYPHOSIS IN MPS I

The
expert in
anything
was
once a
beginner.

Join the Fun @ www.alldayjokes.com

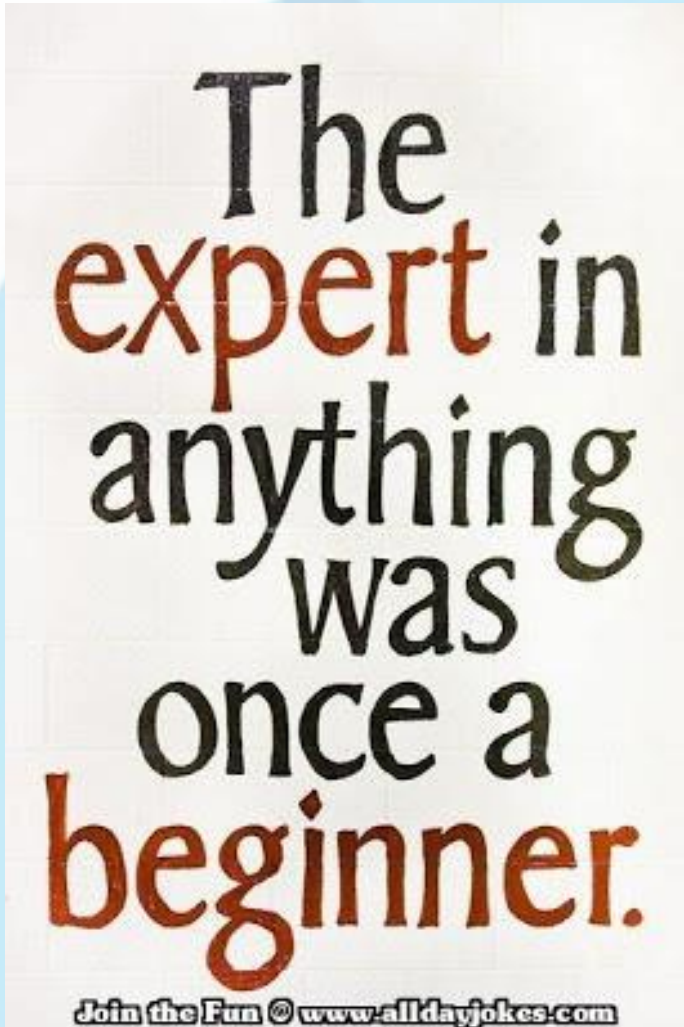
WITHOUT GETTING NERVOUS, HOW
MANY LEGS DID YOU HAVE WHEN
YOU WENT INTO THE OR?

TWO

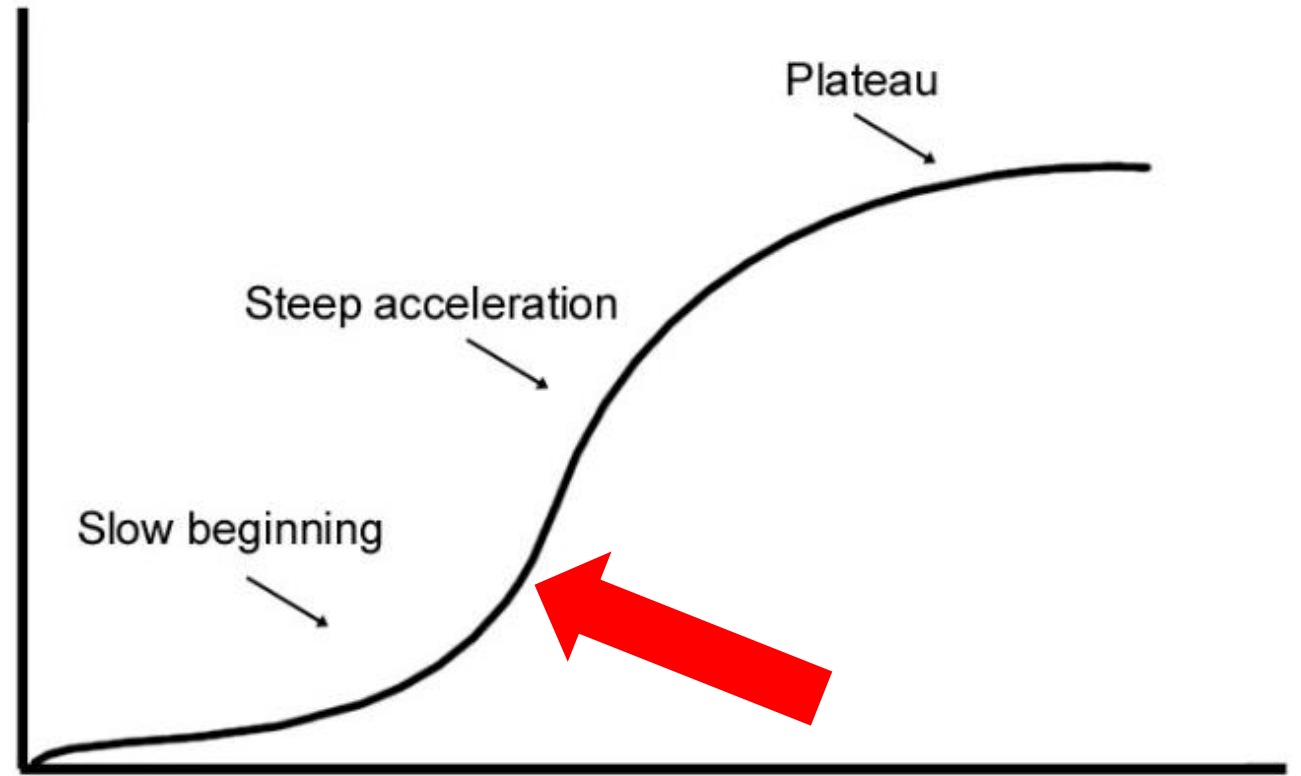
NO WAY



EARLY CORRECTION OF KYPHOSIS IN MPS I



Performance measure



Number of trials or attempts at learning



**“I LIKE TECHNICALLY
CHALLENGING SURGERIES”**



 OPEN ACCESS

466

Acta Orthopaedica 2017; 88 (5): 466–471

Risk preferences and attitudes to surgery in decision making A survey of Swedish orthopedic surgeons

Andreas MEUNIER ¹, Kinga POSADZY ², Gustav TINGHÖG ^{2,3}, and Per ASPENBERG ¹

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EARLY CORRECTION OF KYPHOSIS IN MPS I

**“I LIKE TECHNICALLY
CHALLENGING SURGERIES”**

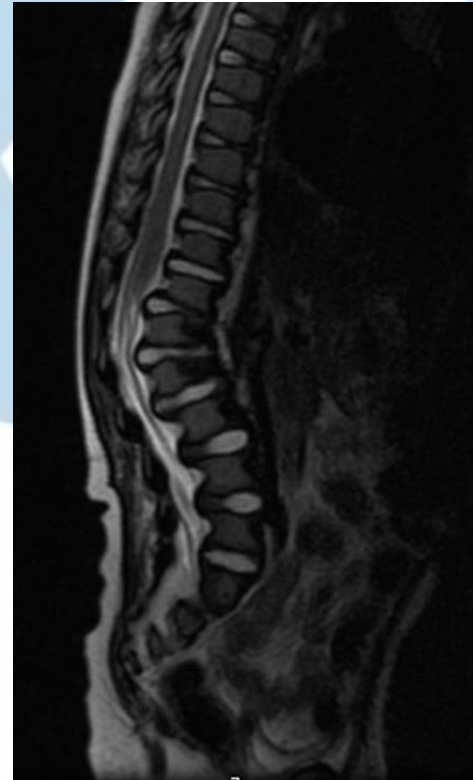


Hazardous Attitudes

MACHO



EARLY CORRECTION OF KYPHOSIS IN MPS I



EARLY CORRECTION OF KYPHOSIS IN MPS I

✓ **BRACING SEEMS NON EFFECTIVE**



EARLY CORRECTION OF KYPHOSIS IN MPS I

- ✓ **BRACING SEEMS NON EFFECTIVE**
- ✓ **PROGRESSIVE $> 50^\circ$ → SURGERY**



EARLY CORRECTION OF KYPHOSIS IN MPS I

- ✓ **BRACING SEEMS NON EFFECTIVE**
- ✓ **PROGRESSIVE $> 50^\circ$ → SURGERY**
- ✓ **MYELOPATHY → SURGERY**



EARLY CORRECTION OF KYPHOSIS IN MPS I

- ✓ **BRACING SEEMS NON EFFECTIVE**
- ✓ **PROGRESSIVE $> 50^\circ$ → SURGERY**
- ✓ **MYELOPATHY → SURGERY**
- ✓ **↑ RISK ANESTHESIA RELATED COMPLICATIONS**



EARLY CORRECTION OF KYPHOSIS IN MPS I

- ✓ **BRACING SEEMS NON EFFECTIVE**
- ✓ **PROGRESSIVE $> \angle 50^\circ$? \rightarrow SURGERY**
- ✓ **MYELOPATHY \rightarrow SURGERY**
- ✓ **\uparrow RISK ANESTHESIA RELATED COMPLICATIONS**
- ✓ **\angle ANTERIOR ? + POSTERIOR FUSION**



EARLY CORRECTION OF KYPHOSIS IN MPS I

- ✓ **BRACING SEEMS NON EFFECTIVE**
- ✓ **PROGRESSIVE $> \angle 50^\circ$ → SURGERY**
- ✓ **MYELOPATHY → SURGERY**
- ✓ **↑ RISK ANESTHESIA RELATED COMPLICATIONS**
- ✓ **\angle ANTERIOR? + POSTERIOR FUSION**
- ✓ **3 MONTHS PO BRACING**

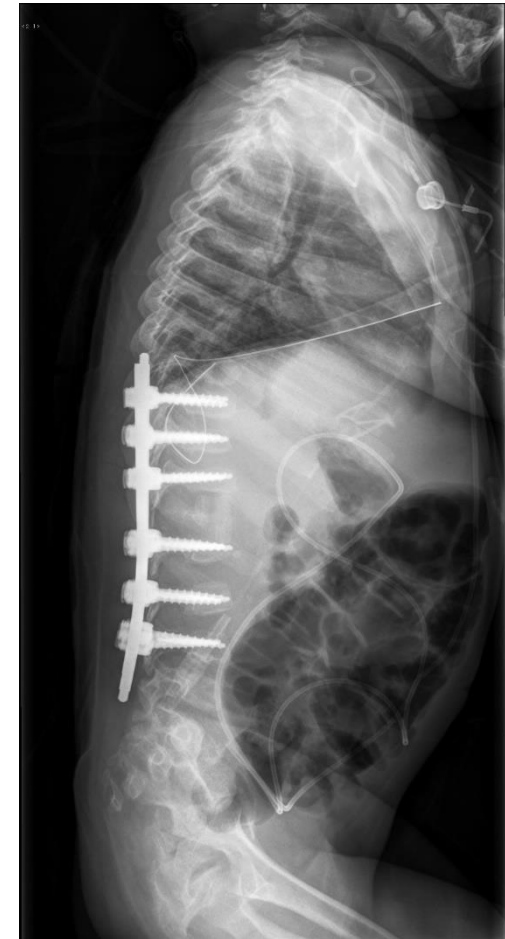
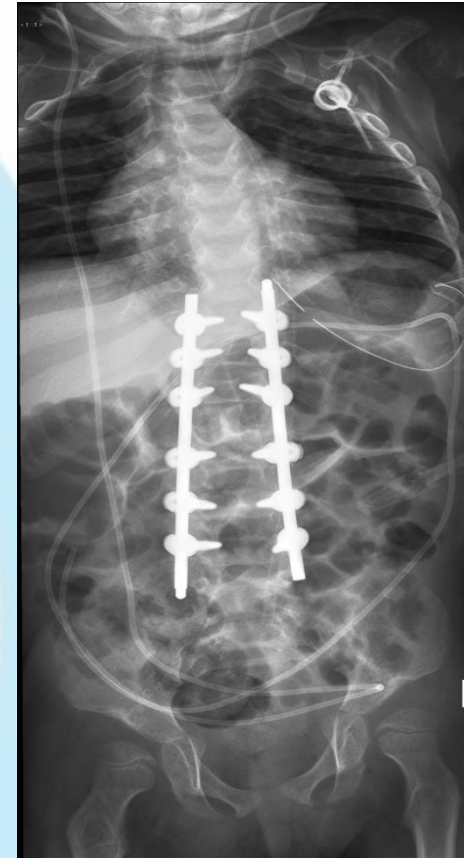
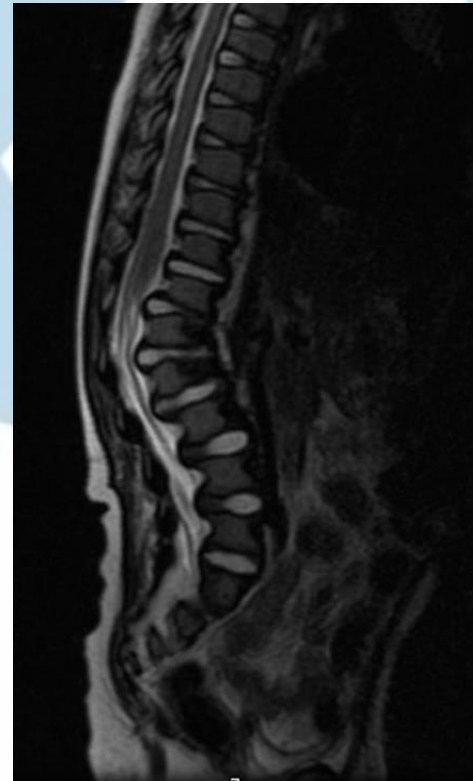


EARLY CORRECTION OF KYPHOSIS IN MPS I

- ✓ **BRACING SEEMS NON EFFECTIVE**
- ✓ **PROGRESSIVE $> \angle 50^\circ$ → SURGERY**
- ✓ **MYELOPATHY → SURGERY**
- ✓ **↑ RISK ANESTHESIA RELATED COMPLICATIONS**
- ✓ **\angle ANTERIOR? + POSTERIOR FUSION**
- ✓ **3 MONTHS PO BRACING**
- ✓ **OSTEOPENIC, SMALL & DYSPLASTIC BONE → RISK OF FAILURE**



EARLY CORRECTION OF KYPHOSIS IN MPS I



EARLY CORRECTION OF KYPHOSIS IN MPS I

**CLARA
3+5**

**MPS I, ERT, WALKS
THORACOLUMBAR GIBBUS
NO SIGNS OR SYMPTOMS
OF MYELOPATHY
RX: L1-L3 55° KYPHOSIS**



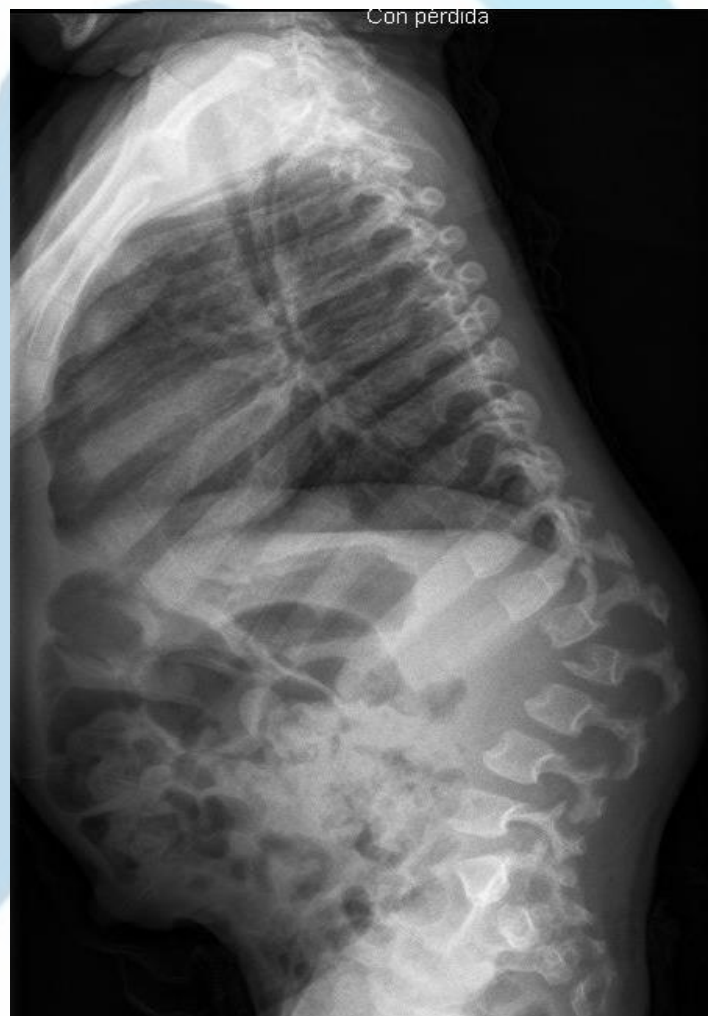
EARLY CORRECTION OF KYPHOSIS IN MPS I



CLARA

3+5

**MPS I, ERT, WALKS
THORACOLUMBAR GIBBUS
NO SIGNS OR SYMPTOMS
OF MYELOPATHY
RX: L1-L3 55° KYPHOSIS**



5+10

STILL WALKING

↑↑ GIBBUS

**NO SIGNS OR SYMPTOMS
OF MYELOPATHY
RX: L1-L3 70° KYPHOSIS**

EARLY CORRECTION OF KYPHOSIS IN MPS I



CLARA

3+5

**MPS I, ERT, WALKS
THORACOLUMBAR GIBBUS
NO SIGNS OR SYMPTOMS
OF MYELOPATHY
RX: L1-L3 55° KYPHOSIS**

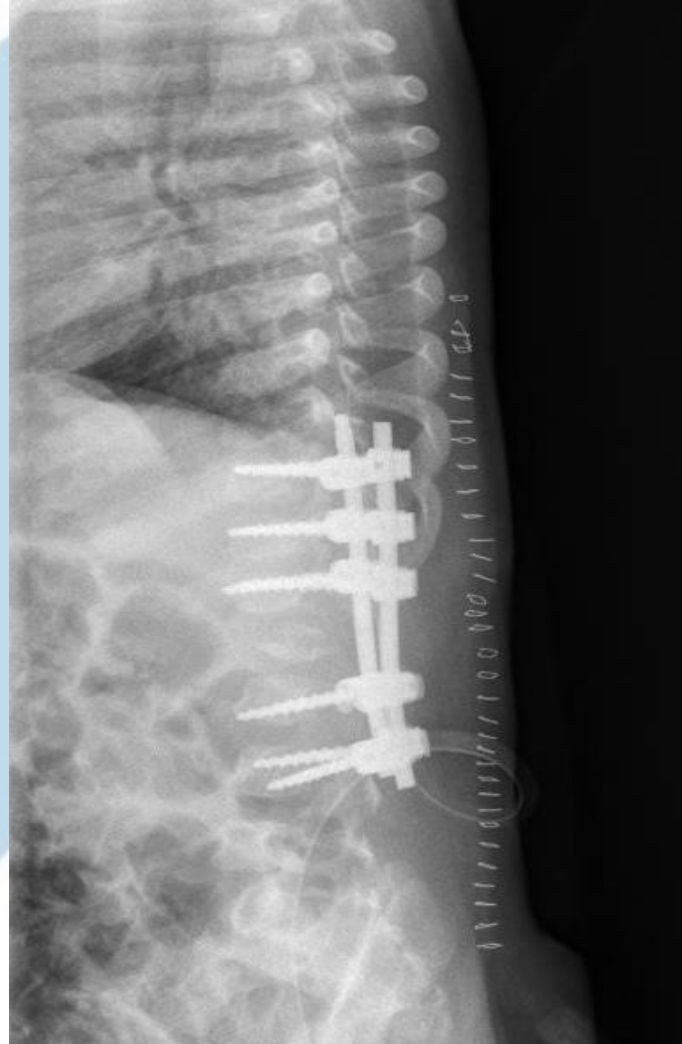
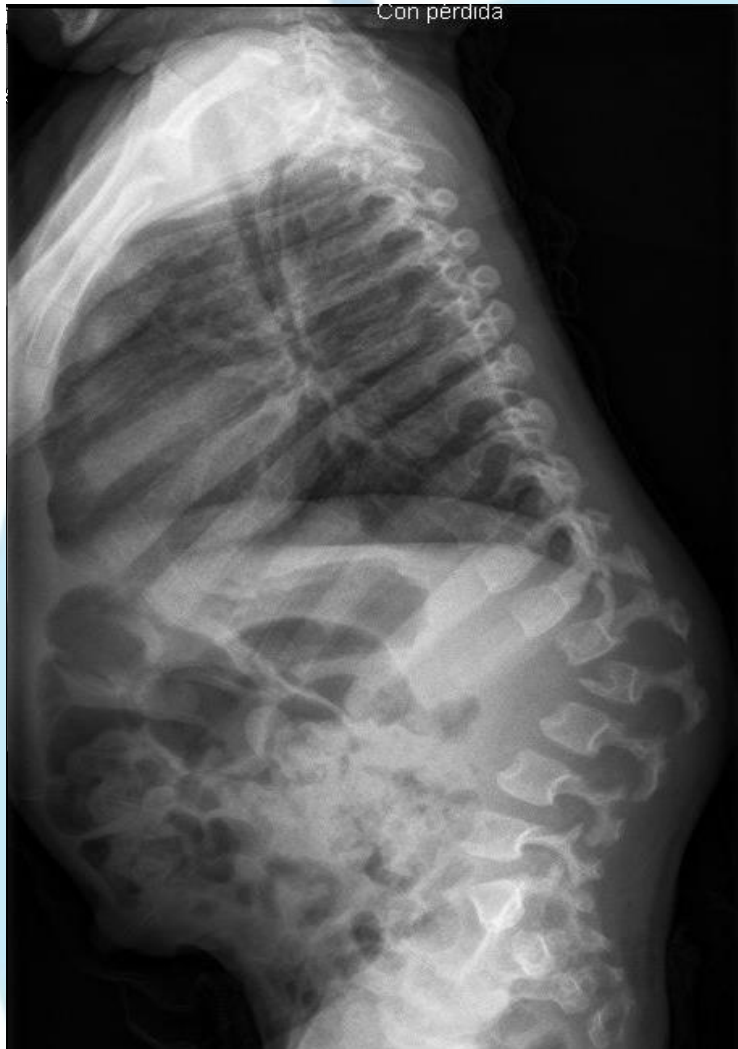
5+10

**STILL WALKING
↑↑ GIBBUS
NO SIGNS OR SYMPTOMS
OF MYELOPATHY
RX: L1-L3 70° KYPHOSIS**

6+0

**PSF T11-L4+L2 PSO
IOM OK**

EARLY CORRECTION OF KYPHOSIS IN MPS I



**CLARA
3+5**

**MPS I, ERT, WALKS
THORACOLUMBAR GIBBUS
NO SIGNS OR SYMPTOMS
OF MYELOPATHY
RX: L1-L3 55° KYPHOSIS**

5+10

STILL WALKING

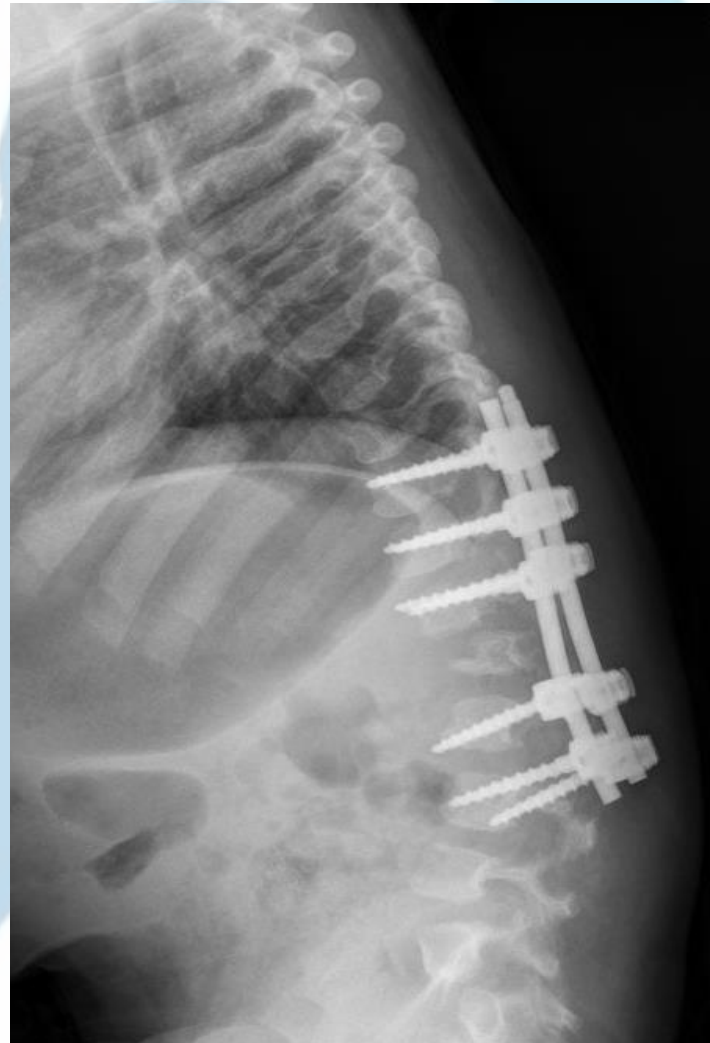
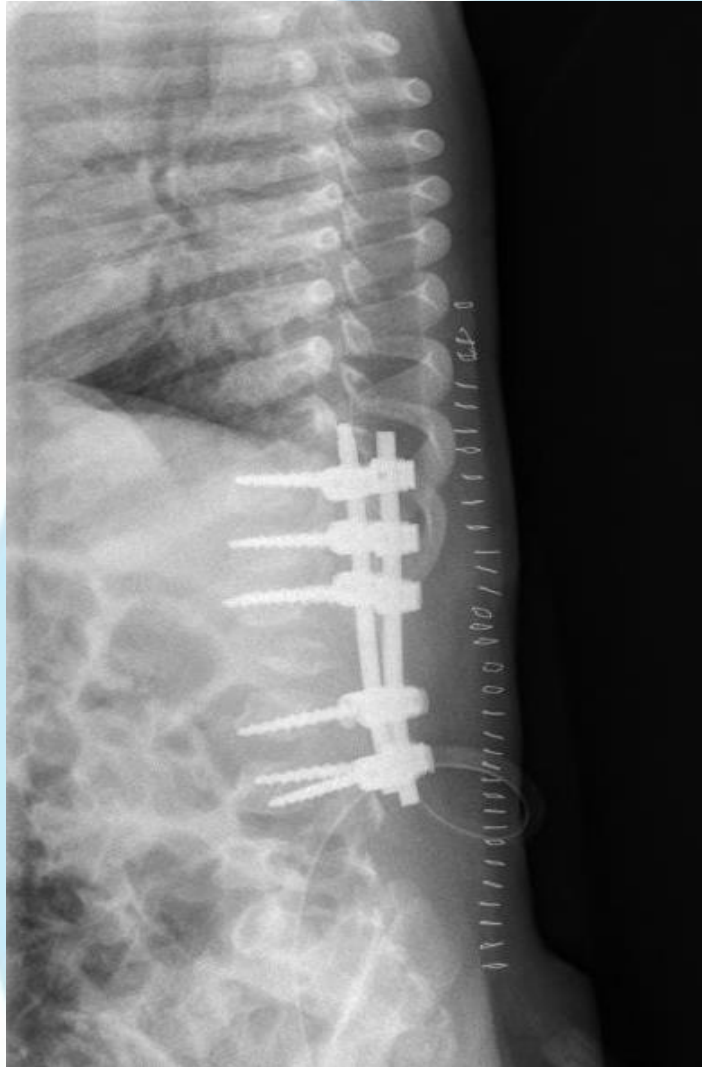
↑↑ GIBBUS

**NO SIGNS OR SYMPTOMS
OF MYELOPATHY
RX: L1-L3 70° KYPHOSIS**

6+0

**PSF T11-L4+L2 PSO
IOM OK**

EARLY CORRECTION OF KYPHOSIS IN MPS I

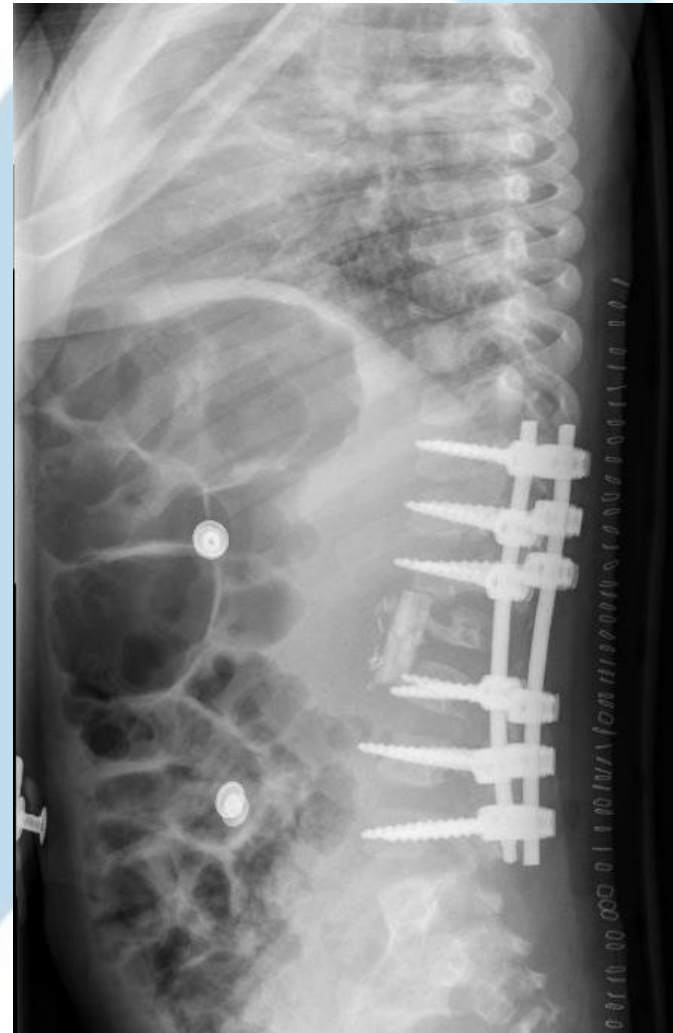
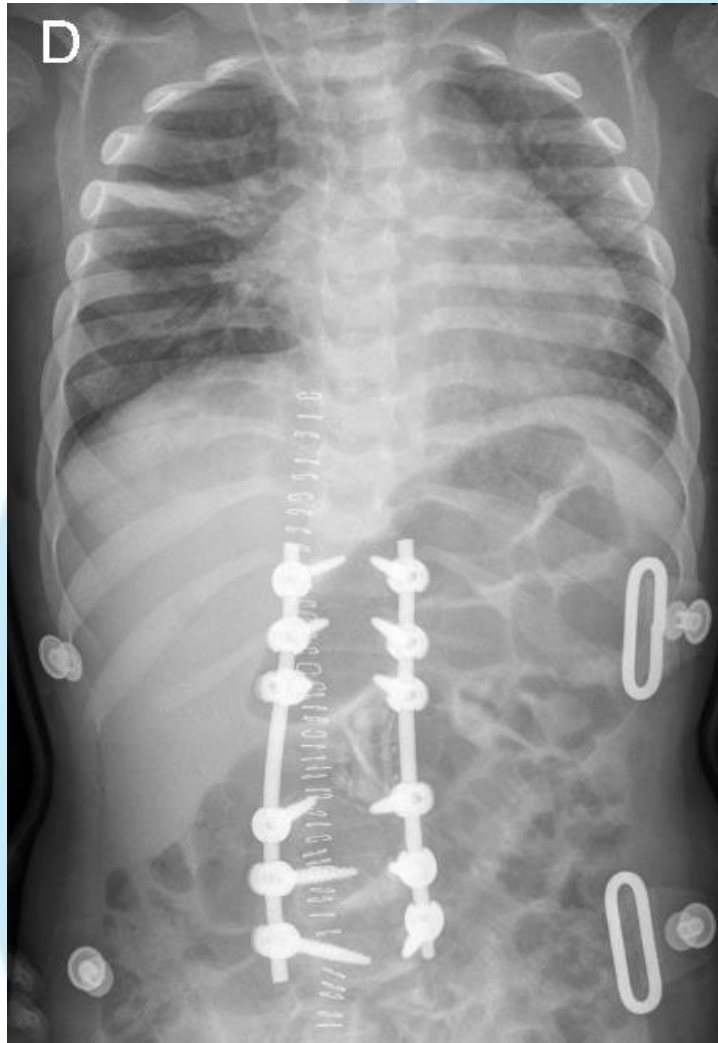


CLARA

6+3

GIBBUS "IS BACK"
NO SIGNS OR SYMPTOMS
OF MYELOPATHY
RX: L4 DISTAL PULL OUT

EARLY CORRECTION OF KYPHOSIS IN MPS I



CLARA

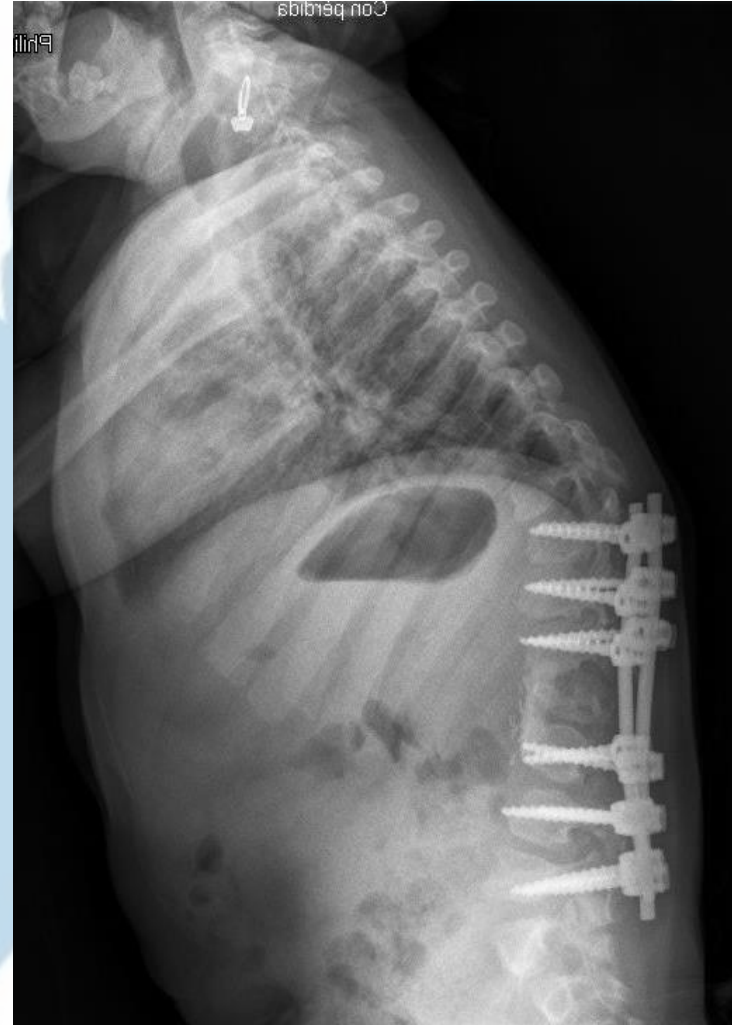
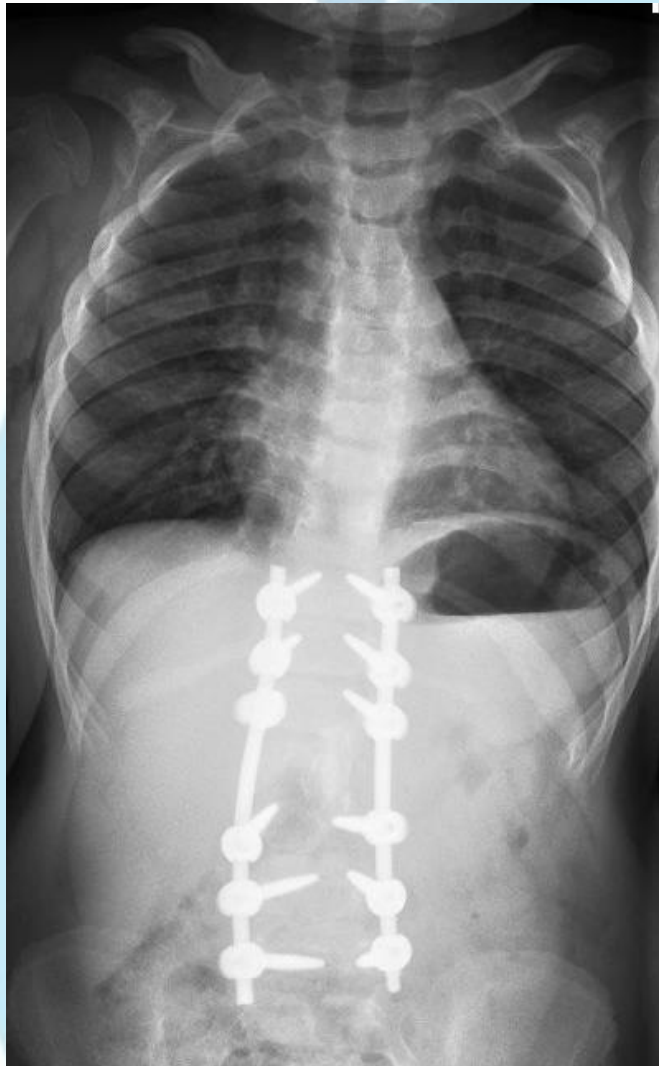
6+3

GIBBUS "IS BACK"
NO SIGNS OR SYMPTOMS
OF MYELOPATHY
RX: L4 DISTAL PULL OUT

6+4

ASF L1-L3+PSF T11-L5
IOM OK

EARLY CORRECTION OF KYPHOSIS IN MPS I



CLARA

6+3

**GIBBUS “IS BACK”
NO SIGNS OR SYMPTOMS
OF MYELOPATHY
RX: L4 DISTAL PULL OUT**

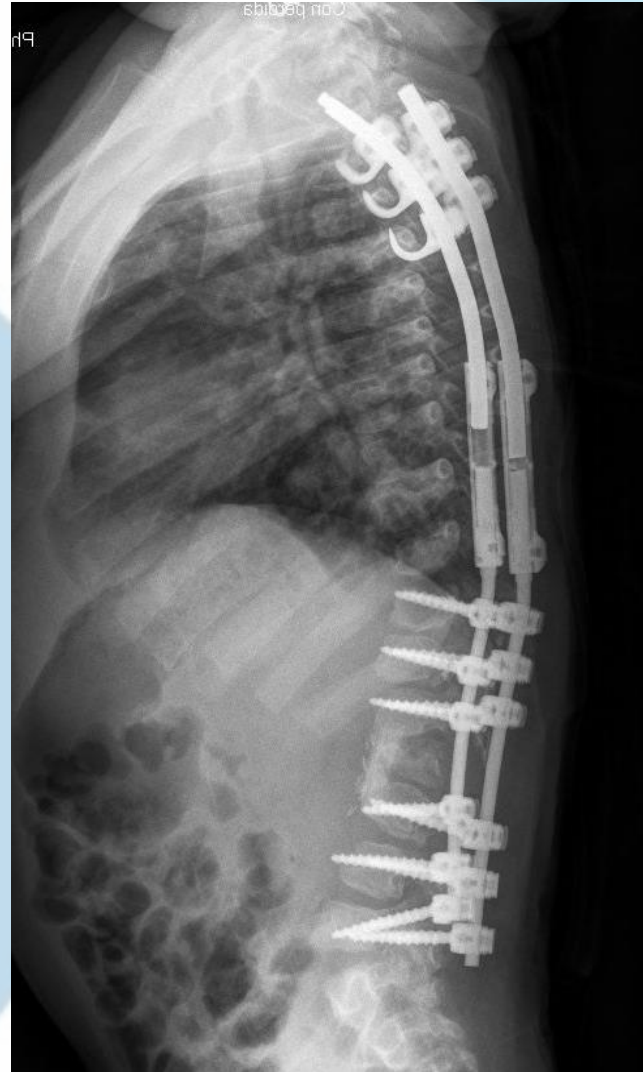
6+4

**ASF L1-L3+PSF T11-L5
IOM OK**

7+3

**STARTING TO LEAN
FORWARD
NO SIGNS OR SYMPTOMS
OF MYELOPATHY
RX: PJK**

EARLY CORRECTION OF KYPHOSIS IN MPS I



CLARA

6+3

**GIBBUS “IS BACK”
NO SIGNS OR SYMPTOMS
OF MYELOPATHY
RX: L4 DISTAL PULL OUT**

6+4

**ASF L1-L3+PSF T11-L5
IOM OK**

7+3

**STARTING TO LEAN
FORWARD
NO SIGNS OR SYMPTOMS
OF MYELOPATHY
RX: PJK**

7+6

**EXTENDED GROWING RODS
T2/3/4-L5
IOM OK**

EARLY CORRECTION OF KYPHOSIS IN MPS I

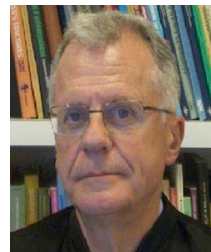
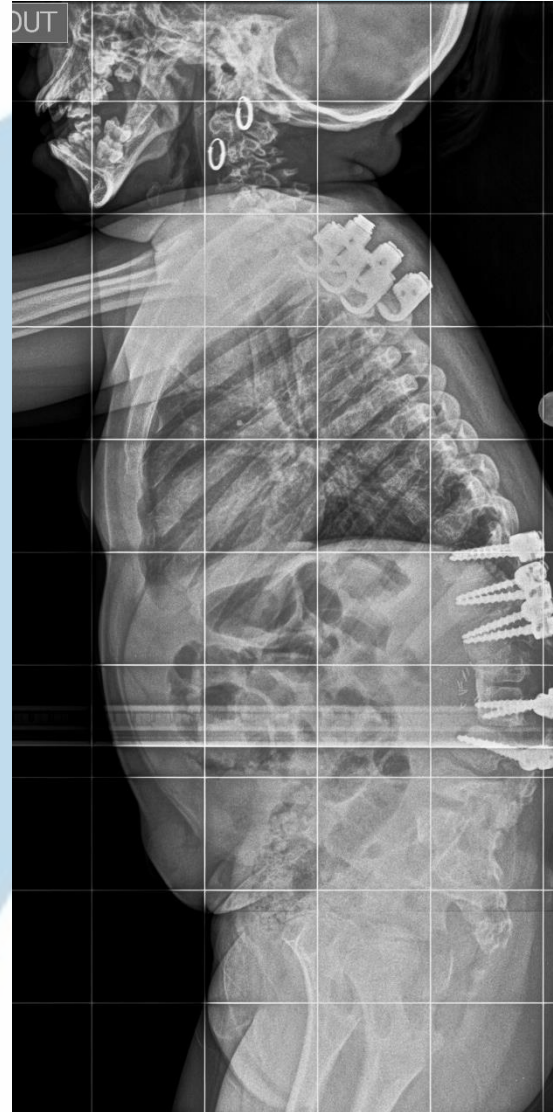
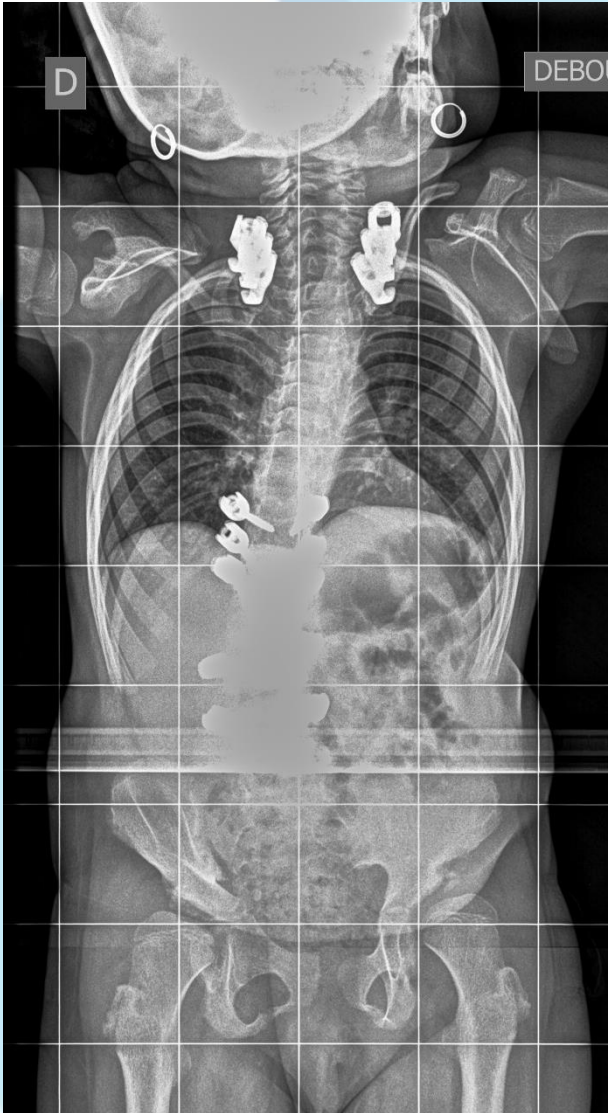
CLARA
PATIENT “FLEES” TO FRANCE



EARLY CORRECTION OF KYPHOSIS IN MPS I

CLARA
PATIENT “FLEES” TO FRANCE

9+3
RODS + L5 SCREWS REMOVAL

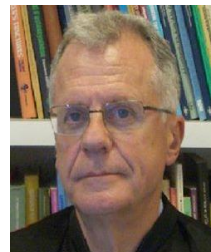
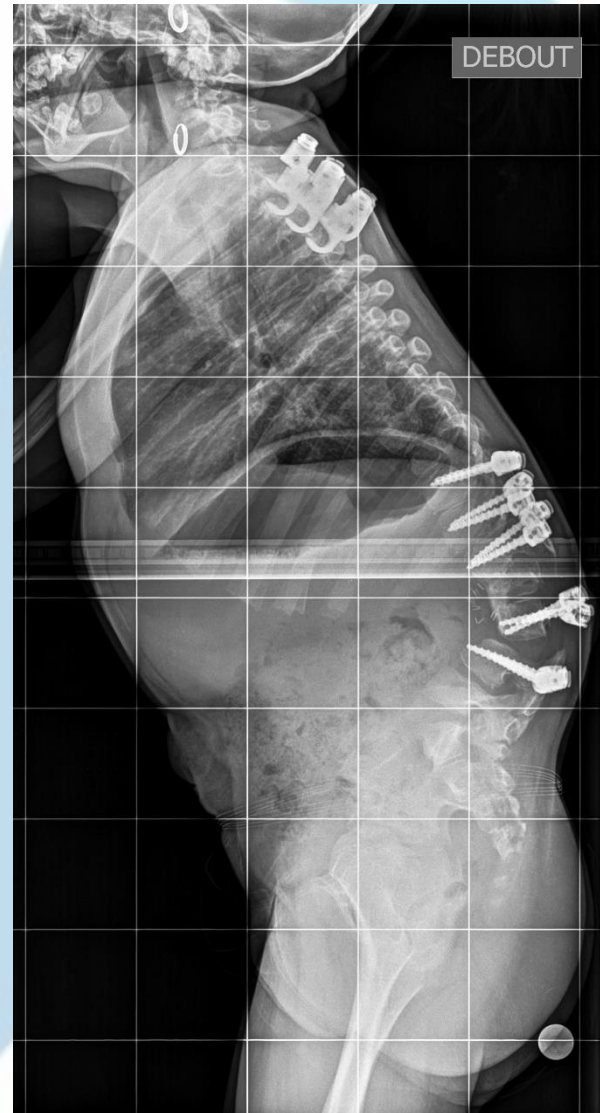
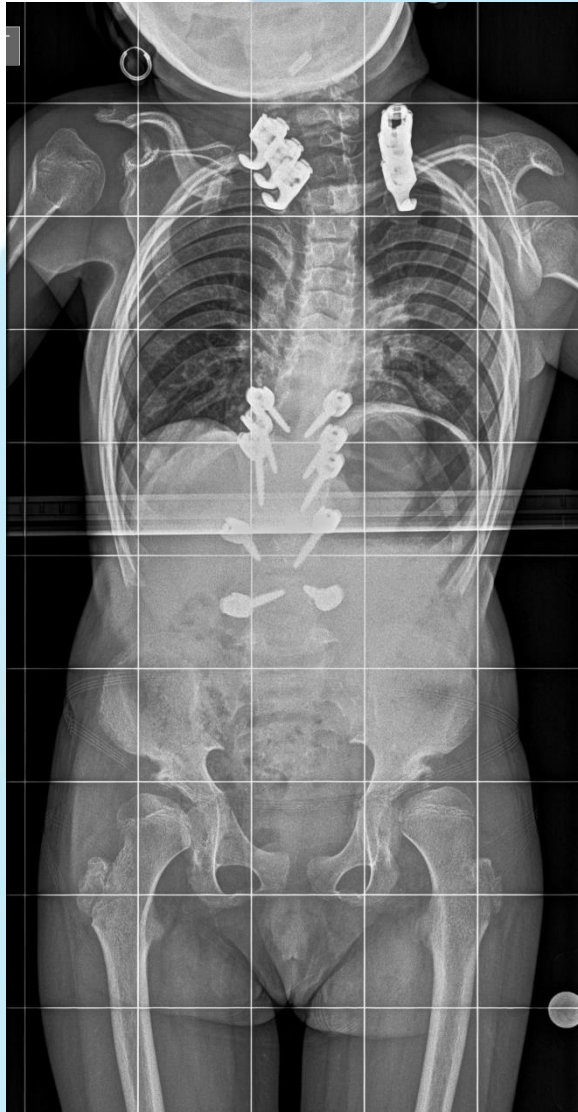


EARLY CORRECTION OF KYPHOSIS IN MPS I

CLARA
PATIENT “FLEES” TO FRANCE

9+3
RODS + L5 SCREWS REMOVAL

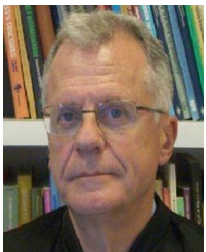
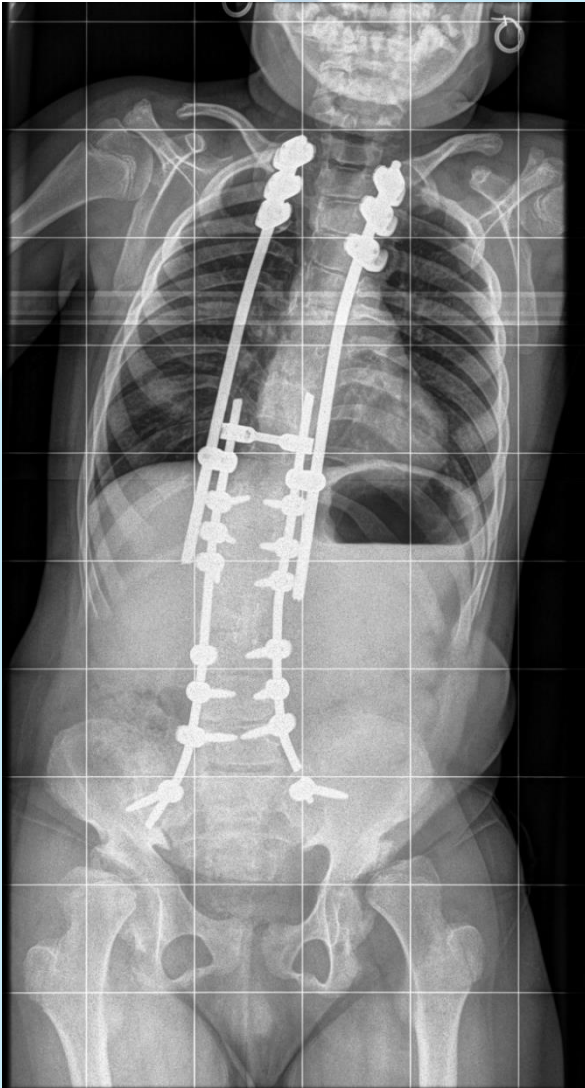
10+0
L3-L4 ↑ KYPHOSIS



EARLY CORRECTION OF KYPHOSIS IN MPS I

CLARA
PATIENT “FLEES” TO FRANCE

9+3
RODS + L5 SCREWS REMOVAL



EARLY CORRECTION OF KYPHOSIS IN MPS I



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PATIENT “FLEES” TO FRANCE

9+3
ROD+ L5 SCREWS REMOVAL

10+0
L3-L4 ↑ KYPHOSIS

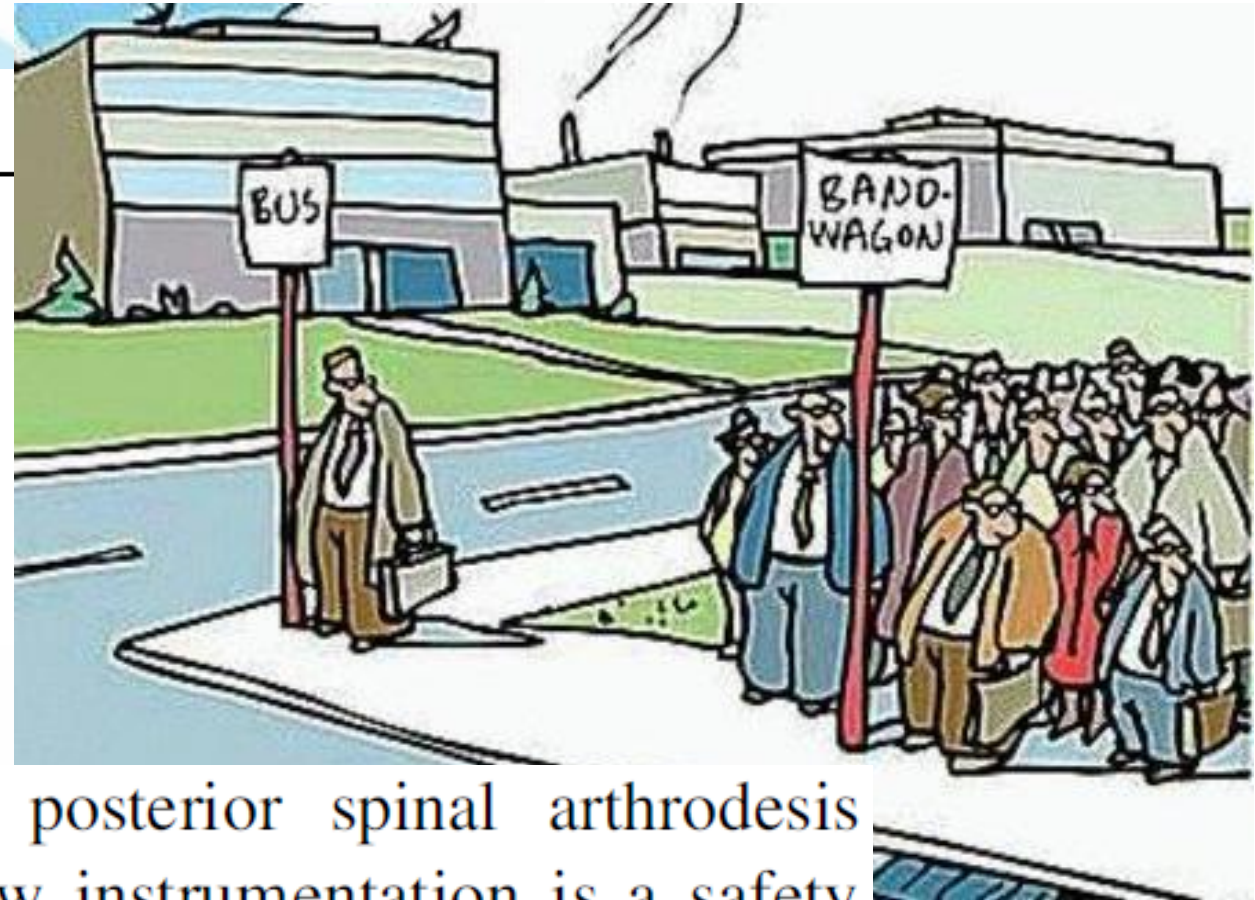
12+1
LAST FOLLOW UP

REFLECTION WITH
PROF. BOLLINI



Combined spinal arthrodesis with instrumentation for the management of progressive thoracolumbar kyphosis in children with mucopolysaccharidosis

Enrique Garrido · Félix Tomé-Bermejo ·
Christopher I. Adams



Conclusions Anterior and posterior spinal arthrodesis with segmental pedicle screw instrumentation is a safety and reliable technique for the treatment of severe thoracolumbar kyphosis in children with MPS. This technique achieves excellent correction of the deformity with adequate decompression of the spinal canal. The fusion is



Surgical Management of Progressive Thoracolumbar Kyphosis in Mucopolysaccharidosis: Is a Posterior-only Approach Safe and Effective?

Senol Bekmez, MD,* Halil G. Demirkiran, MD,† Ozgur Dede, MD,‡ Vusal Ismayilov, MD,§
and Muharrem Yazici, MD†



Conclusions: Posterior-only corrective techniques and instrumented fusion with pedicle screws for progressive thoracolumbar junctional kyphosis in MPS patients are safe and effective methods. Its results are comparable to those achieved with conventional circumferential fusion. However, patients should be monitored closely for adjacent-segment problems.

ORIGINAL ARTICLE

Surgical management of thoracolumbar kyphosis in mucopolysaccharidosis type 1 in a reference center

Kariman Abelin Genevois • Christophe Garin •
Federico Solla • Nathalie Guffon • Rémi Kohler



Conclusion Surgery is necessary when kyphosis is progressive despite orthopedic management, aggravating the multifactorial trunk imbalance. Regarding our experience, circumferential arthrodesis should be recommended to achieve stable correction. Surgical management requires a multidis-



Spine challenges in mucopolysaccharidosis

Marco Crostelli¹ • Osvaldo Mazza¹ • Massimo Mariani¹ • Dario Mascello¹ • Carlo Iorio¹



Conclusions Bracing is a viable treatment strategy in thoracic lumbar kyphosis and can obtain good clinical results at medium terms follow-up even if kyphosis deformity remains in radiographs. Surgical treatment is effective in severe evolving cases both at cervical and thoracic lumbar level, main difficulties arose from unavailability of dedicated instrumentation in very young patient, as even smallest devices available are often too big.



Spine

LITERATURE REVIEW

Surgical Management of Thoracolumbar Kyphosis in Patients With Mucopolysaccharidosis

A Systematic Review

Nicole Williams, BMed, FRACS,* Peter J. Cundy, MBBS, FRACS,* and Deborah M. Eastwod, MB, ChB, FRCS†

➤ Key Points

- ❑ Systemic therapies in mucopolysaccharidosis (MPS) have permitted more patients to be considered surgical candidates.
- ❑ Surgery for thoracolumbar kyphosis is most commonly performed in MPS I, but preoperative neurological compromise in this MPS type was not reported.
- ❑ There is insufficient evidence to recommend thoracolumbar spine surgery in MPS patients to improve function and/or quality of life.
- ❑ MPS patients are at risk of peri-operative thoracic spinal cord ischaemia and paraplegia when undergoing both spinal and lower limb orthopaedic surgery.
- ❑ Caution must be exercised when extrapolating surgical indications from the non-MPS population.



SLOW DOWN
CALM DOWN
DON'T WORRY
DON'T HURRY
TRUST THE PROCESS

Spine

LITERATURE REVIEW

Surgical Management of Thoracolumbar Kyphosis in Patients With Mucopolysaccharidosis

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EARLY CORRECTION OF KYPHOSIS IN MPS I

Procrastination:



Just Do It...Later

50

70

Age?

MYELOPATHY!!!!



THANK YOU

IT'S OKAY IF YOU
DISAGREE WITH ME

I CAN'T FORCE YOU TO BE RIGHT

