THE BANDWAGON
I JUMPED ON
AND THEN OFF...

No financial disclosures
THE BANDWAGON
I JUMPED ON
AND THEN OFF...

EARLY CORRECTION
OF KYPHOSIS IN MPS I
Without getting nervous, how many legs did you have when you went into the OR?

Two

No way
The expert in anything was once a beginner.
Risk preferences and attitudes to surgery in decision making
A survey of Swedish orthopedic surgeons

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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
BRACING SEEMS NON EFFECTIVE
EARLY CORRECTION OF KYPHOSIS IN MPS I

✓ BRACING SEEMS NON EFFECTIVE
✓ PROGRESSIVE >\(50^\circ\)? → SURGERY
✓ BRACING SEEMS NON EFFECTIVE
✓ PROGRESSIVE >50°? → SURGERY
✓ MYELOPATHY → SURGERY
BRACING SEEMS NON EFFECTIVE
PROGRESSIVE >50°? → SURGERY
MYELOPATHY → SURGERY
↑ RISK ANESTHESIA RELATED COMPLICATIONS
✓ BRACING SEEMS NON EFFECTIVE
✓ PROGRESSIVE >50°? → SURGERY
✓ MYELOPATHY → SURGERY
✓ ↑ RISK ANESTHESIA RELATED COMPLICATIONS
✓ ¿ANTERIOR?+POSTERIOR FUSION

EARLY CORRECTION OF KYPHOSIS IN MPS I
BRACING SEEMS NON EFFECTIVE
PROGRESSIVE >¿50°? → SURGERY
MYELOPATHY → SURGERY
↑ RISK ANESTHESIA RELATED COMPLICATIONS
¿ANTERIOR?+POSTERIOR FUSION
3 MONTHS PO BRACING
EARLY CORRECTION OF KYPHOSIS IN MPS I

- Bracing seems non effective
- Progressive >50°? → Surgery
- Myelopathy → Surgery
- ↑ Risk anesthesia related complications
- ¿Anterior? + Posterior fusion
- 3 months po bracing
- Osteopenic, small & dysplastic bone → Risk of failure
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

LAST FOLLOW UP
EARLY CORRECTION OF KYPHOSIS IN MPS I

CLARA

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
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.
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 multidisciplinarily-based approach and commitment by the patient and family.
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.
Surgical Management of Thoracolumbar Kyphosis in Patients With Mucopolysaccharidosis

A Systematic Review

Nicole Williams, BMed, FRACS,* Peter J. Cundy, MBBS, FRACS,* and Deborah M. Eastwood, MB, ChB, FRCS†
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!!!!
EARLY CORRECTION OF KYPHOSIS IN MPS I

THANK YOU

IT'S OKAY IF YOU DISAGREE WITH ME
I CAN'T FORCE YOU TO BE RIGHT