SURGICAL TREATMENT FOR EARLY ONSET SCOLIOSIS ASSOCIATED WITH RARE DISEASE

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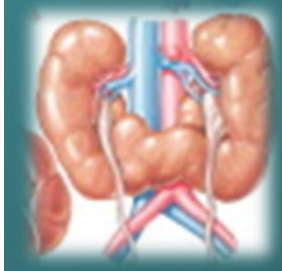
BACKGROUND

Kyphoscoliosis is the most common spinal deformity in rare syndromes.



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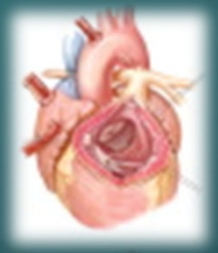
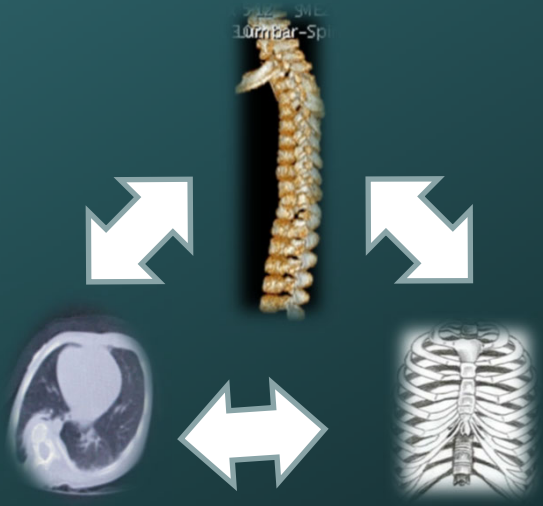


- Rapidly evolving
- Early Onset or Congenital
- Difficult to manage
- Numerous malformations
- Interdisciplinary approach



Spine Deformities in Rare Congenital Syndromes Clinical Issues. SPINE Volume 34, 2009. Robert M. Campbell, Jr, MD

Profiling Scoliosis in Rett Syndrome. Pediatr Res. 2010 April. Alan K. Percy, Hye-Seung Lee, et al.

Treatment of scoliosis in intermediate spinal muscular atrophy (SMA type II) in childhood. Orthop Traumatol Rehabil, 2005. Fujak A, Ingenhorts A et al

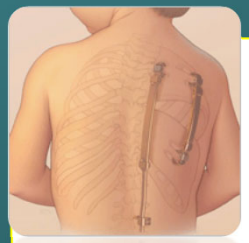
MATERIAL AND METHODS

From 2006 to 2011
retrospective study
11 paediatric patients

♂ 7 ♀ 4

INCLUSION CRITERIA

1) EOS or congenital scoliosis in rare syndrome
2) Growth sparing systems
3) Follow-up 24 months (12-36)



VEPTR
4 patients

Mean age at 1st visit : **3 yrs** (1-7)
Mean age at surgery: **5 yrs** (3-9)

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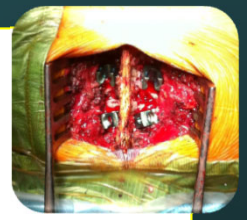
GROWING ROD
7 patients

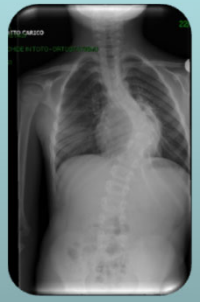


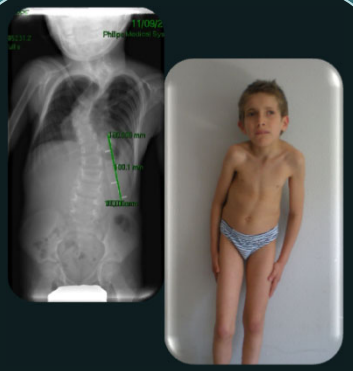
Mean age at 1st visit : **6 yrs** (3-10)
Mean age at surgery: **9 yrs** (5-12)

AIM OF THE STUDY: to describe an approach to spinal deformities in rare syndromes and to assess the effectiveness of growth sparing systems

OUR SYNDROMS

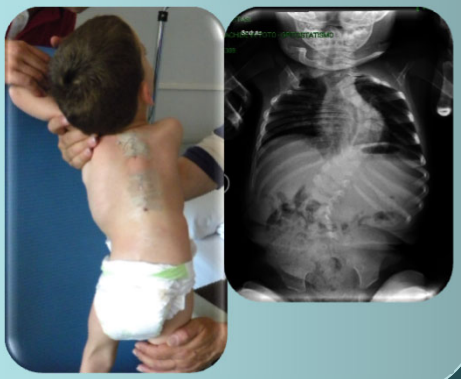
type 1 neurofibromatosis (2)

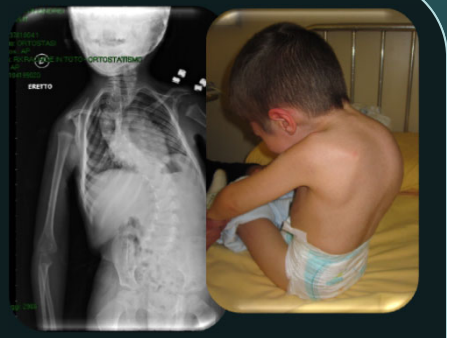




trisomy 8 (1)

Spondylo-rib dysplasia (1)

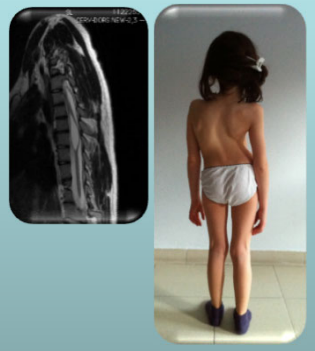




arthrogryposis (2)

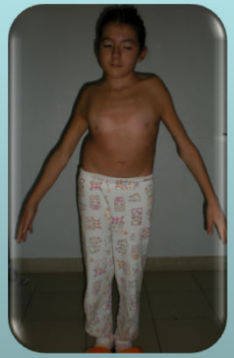
Prader Willi (1)

syringomyelia (1)

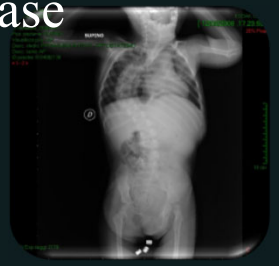


Arnold Chiari type I (1)

Escobar (1)

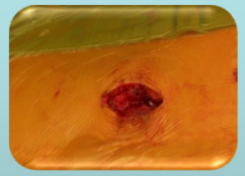


congenital heart disease (1)

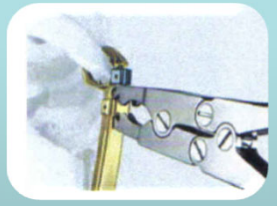


RESULTS

Preoperative treatment with Bivalve or Milwaukee brace



11 lengthening procedures



average of 11 months after

Mean thoracic curve correction: 50%

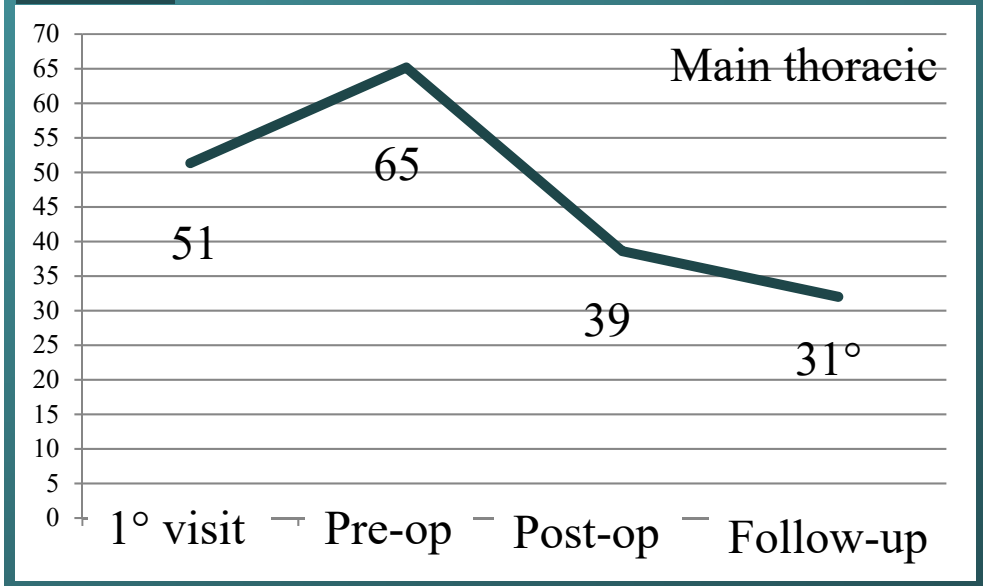
61° to 31°

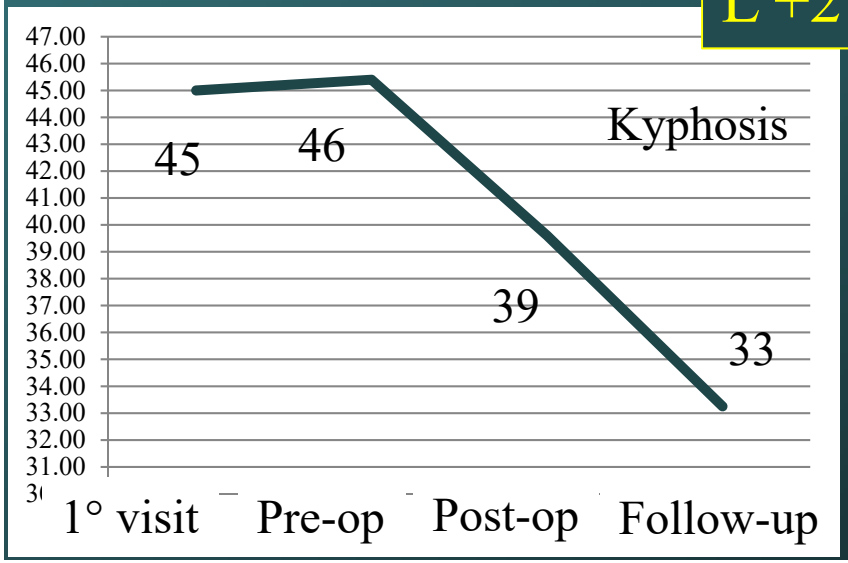
Lumbar curve: loss of correction of 50% at follow up.

8 postoperative complications (mechanicals in all cases)
2 revision surgeries (not originally planned)

RESULTS

GR





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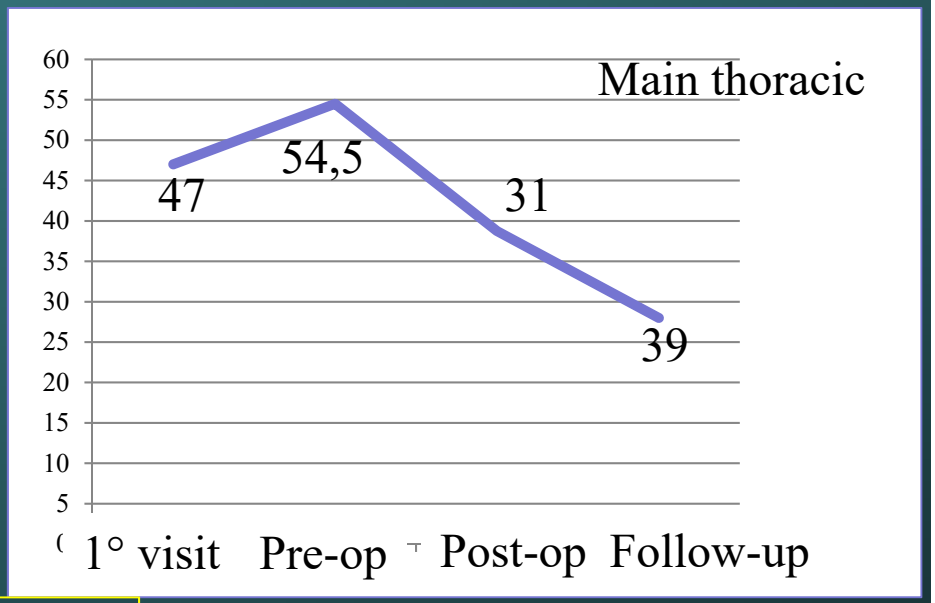
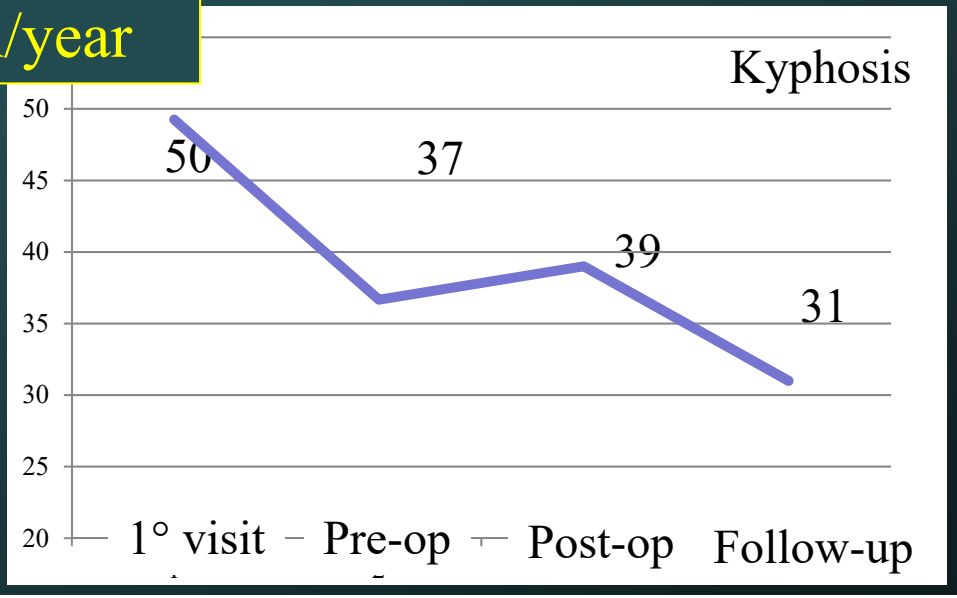


L +2 cm/year



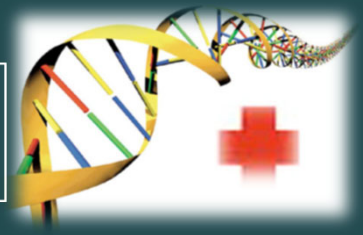
CONCLUSIONS

TEAM

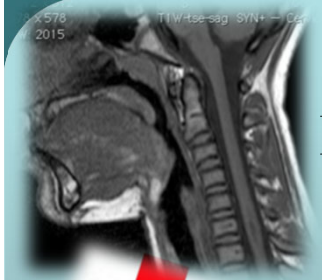
OUR STRATEGY

Ambulatory for rare diseases

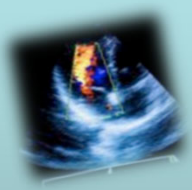
Genetic diagnosis



Pediatric Orthopaedic Ambulatory



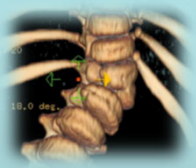
MRI

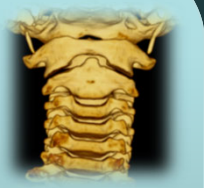


Echocardiogram

Abdominal ultrasound

CT



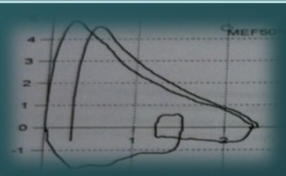


C0-C2 CT scan

Neuro Surgeon

Anesthesiologist evaluation

Neuropsychiatrist



PFT

Lung CT scan

Pneumologist

Quality of life

SPINE Volume 34, Number 17, pp 1815–1827 ©2009 Spine Deformities in Rare Congenital Syndromes Clinical Issues _ Robert M. Campbell, Jr, MD

CONCLUSIONS

- Very aggressive deformities
- Hard to manage it conservatively

→ **Growth sparing systems**

TEAM

Clinical and instrumental evaluation **specific to each patient**

Without a confirmed genetic diagnosis of a suspected syndrome is impossible to plan preoperative strategy

Robert M. Campbell, Jr, MD

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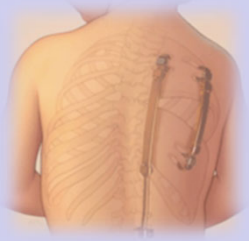


< 5 years

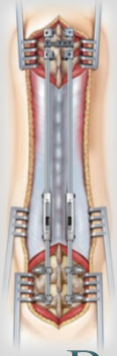
Every diagnosis

Correction of chest cage

Little thoracic stiffness

Muharrem, Emans MD, Spine 34

GR



> 5 years

Only for spinal deformities

Cervical extension

Better control of upper thoracic kyphosis