ARTHROGYROSIS AND VEPTR
Surgery on the cutting EDGE
CLINICAL CASE

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Introduction

**ARTHROGRYPOSIS**

- Rare syndrome
- Kyphoscoliosis is the most common deformity
- Very early onset, rapidly evolving and become very stiff from early age.
  - Incidence: from 30% to 67%.

- Multidisciplinary approach (paediatric orthopaedic surgeon, anaesthesiologist, pulmonologist).
- Few literature arthrogryposis and EOS surgically treated.

**AIM of the study**

- We reviewed our children affected by EOS in arthrogryposis and surgically treated to describe the efficacy of rib-based distraction systems.
- We use Vertical Expandable Prosthetic Titanium Rib (VEPTR1 and 2) device. (DePuy J&J)
Material and Methods

• 4 cases (1 male, 3 females; mean age at surgery 5.75 years) affected by arthrogryposis multiplex congenita were reviewed
• From 2011 to 2014.

Each patient was studied from the genetic point of view, pft, Cardio-US and abdominal US, neuropsychiatric evaluation.

brain-spinal MRI: NO Malformation

Surgery was performed using VEPTR system in all cases, spinal-rib construct only, 1 case with a double construct, 1 case underwent at final fusion with posterior instrumented arthrodesis.
1st Case

2 yy and 10 mm. Body growth at the age of 2: less than 3rd centile.
Respiratory deficit with frequent infections of the upper respiratory pathways.
Full time brace treatment since the age of 1 year

W = 6 Kg
H = 72 cm
Post-OP x-Rays II

6 yy  After 3 lengthening

Good sagittal correction
2nd Case

Severe Early onset scoliosis in Arthrogryposis
Frequent Pneumoniae
7 yy
Pre-OP x-Rays I

Post-OP x-Rays I

VEPTR after 2 lengthening

Complication: pneumonia
3rd Case

Scoliosis in arthrogryposis

VEPTR
After 1 lengthening
Good sagittal correction
Less respiratory disease
FU x-Rays I and Clinical

Complication: PJK

Revision + Lengthening
4th Case: Final Arthrodesis

Early onset scoliosis in Arthrogryposis. Cardiopathy, renal malformation

9 yy

VEPTR during the growth

13 yy

T2-L3 arthrodesis

Hands surgery

After 2 lengthening
Results

First surgery:
• Scoliosis correction: 24.7%, from 113.2° to 85° (mean value)
• Kyphosis correction: 24.9% from 87.2° to 65.5°

After 6 lengthening procedures, 1.5 for each patient:
• gradual improvement of scoliosis correction of 6.1%
• loss of correction of kyphosis of 24%.

Complications: a case of pneumonia and 1 asymptomatic PJK
• Mean follow-up was 24 months (12-36).

• Children are grown on average of 2.5 cm, each follow up year.
• One patient underwent to final fusion at the age of 13 years old with a pedicle screw and hooks instrumentation from T2 to L3.
Conclusion

• Arthrogryposis is one of the most severe causes of stiff and rapidly evolved kyphoscoliosis.

• Prompt action should be taken, even with early surgery to limit the progression of scoliosis and pulmonary failure.

• In arthrogryposis brace treatment is mostly ineffective

• In very young children rib-based distraction (VEPTR) seems to be an effective treatment method to limit the curve progression and maintaining thoracic growth considering the particular stiffness of this kind of deformities;

• On the other hand we can confirm that at follow up there is a little loss of correction, mostly in the sagittal plane correction.