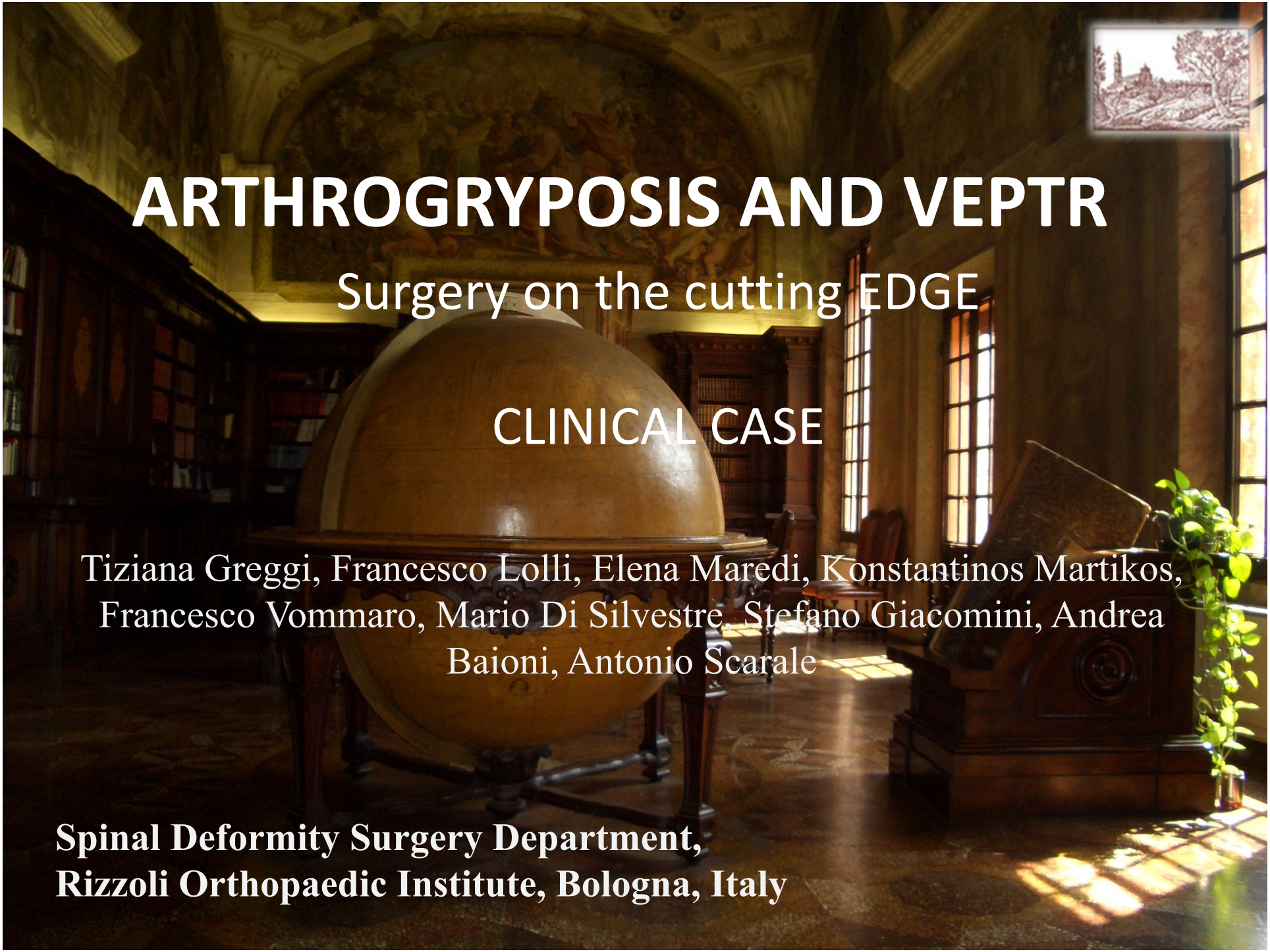

# ARTHROGRYPOSIS AND VEPTR

## 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. T

Incidence: from 30% to 67%.

- multidisciplinary approach (paediatric orthopaedic surgeon, anestesiologist, pulmonologist).
- Few letterature arthrogryposis and EOS surgically treated.

## AIM of the study

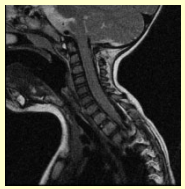
- We reviewed our children affected by EOS in arthrogryposis and surgically treated to describe the efficacy or rib-based distraction systems.
- We use Vertical Expandable Prosthetic Titanium Rib (VEPTR1 and 2) device. (DePuy J&J)

# 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.
<u>brain-spinal MRI: NO Malformation</u>





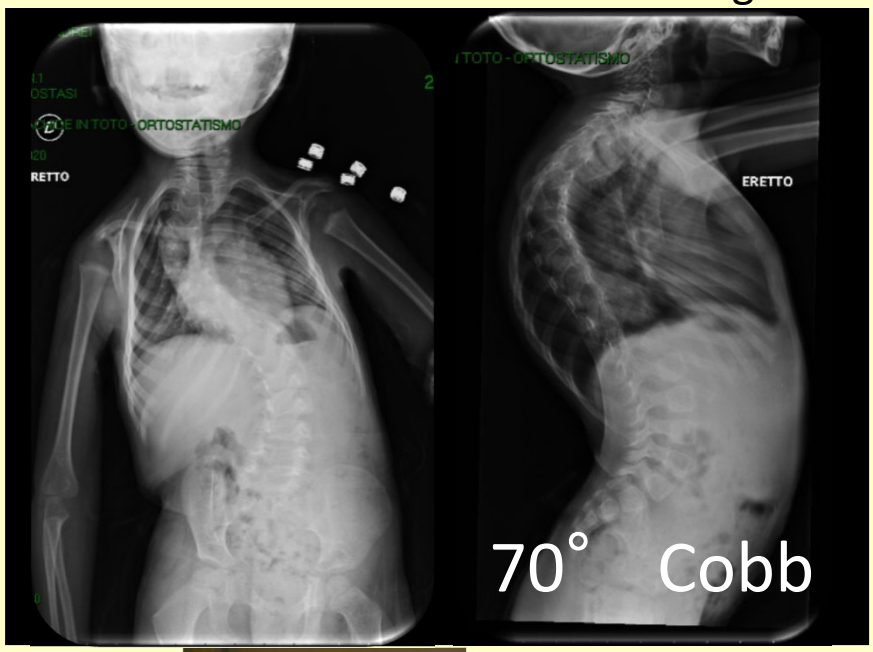
Surgery was performed using VEPTR system in all cases, spinal-rib construct only, 1 case with a double construct, 1 case underwent at final fusion with posterior instrumented arthrodesis.

# ☑ 1st Case

2 yy and 10 mm.  Body grouth at the age of 2: less than 3rd centile.

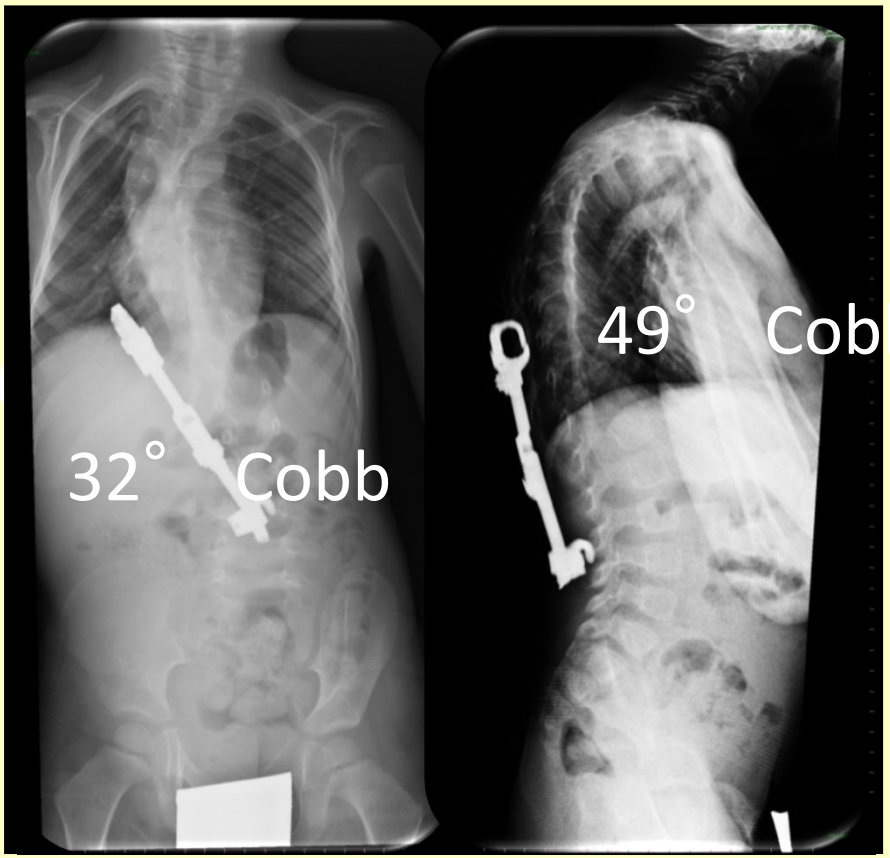
Respiratory deficit with frequent infections of the upper respiratory pathways.

Full time brace treatment since the age of 1 year




**W = 6 Kg**
**H = 72 cm**

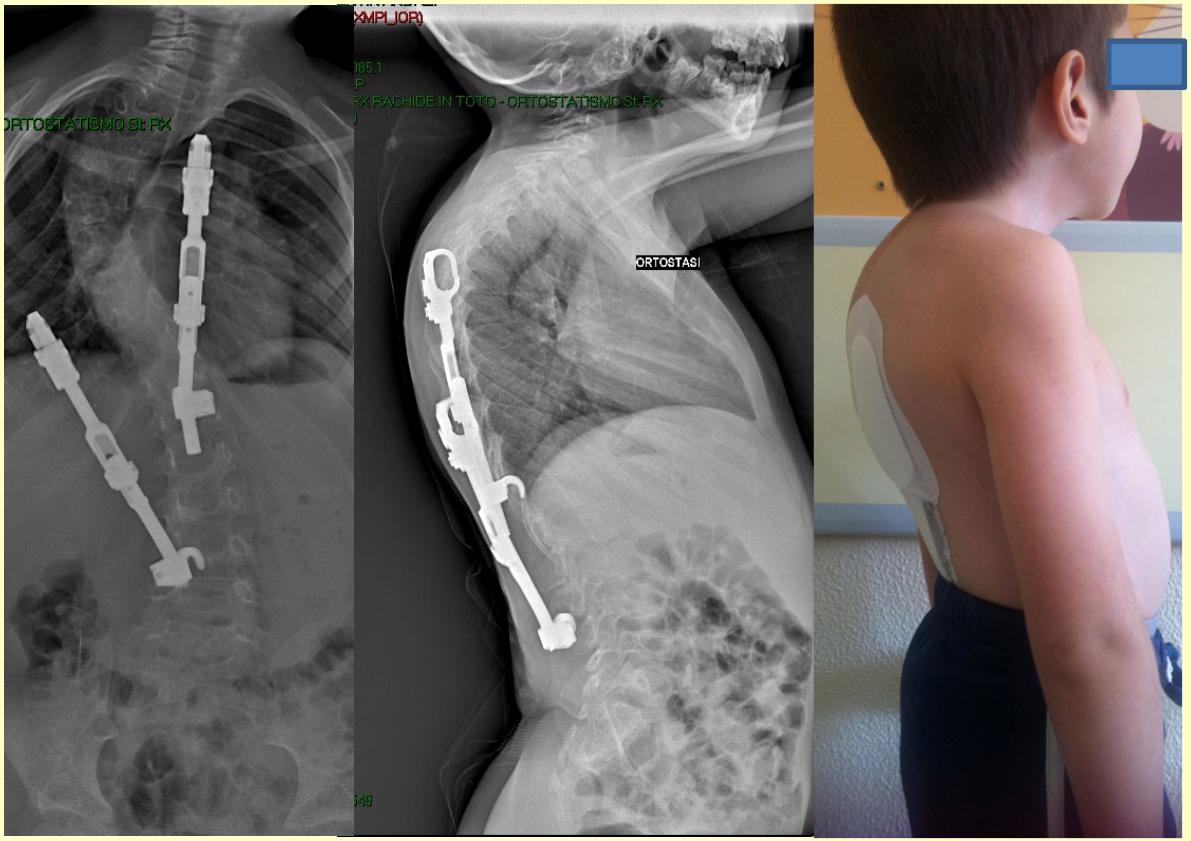
1 st VEPTR



# Post-OP x-Rays II

6 yy

After 3 lengthening



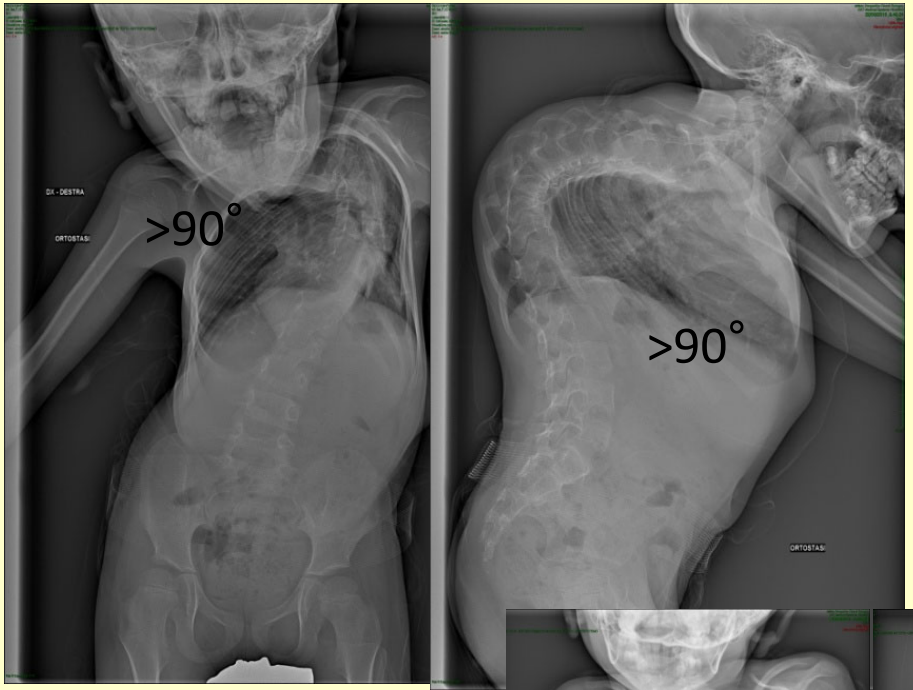

Good sagittal correction

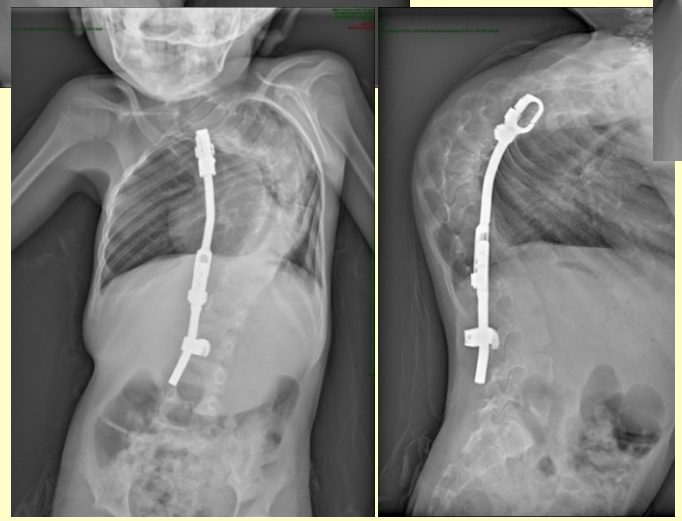
# ☑ 2nd Case

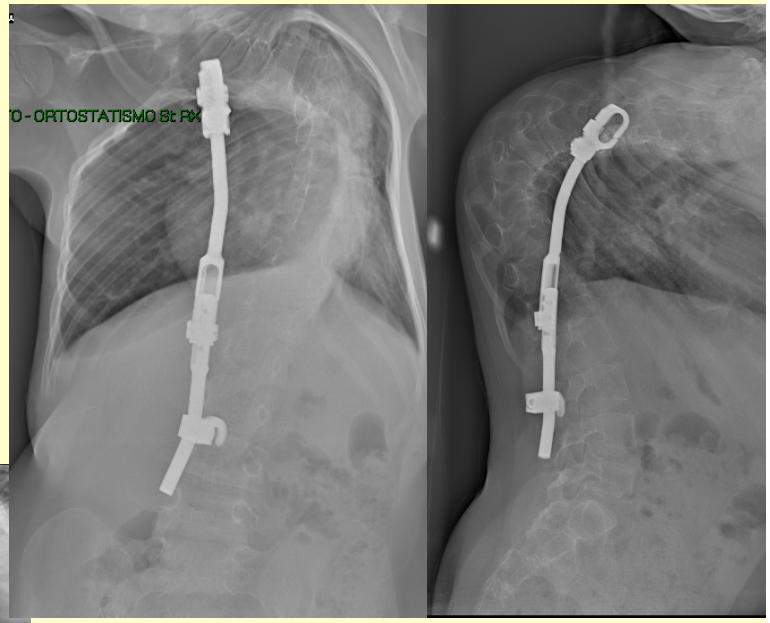
Severe Early onset scoliosis in Arthrogryposis
Frequent Pneumoniae
7 yy

# Pre-OP x-Rays I

# Post-OP x-Rays I



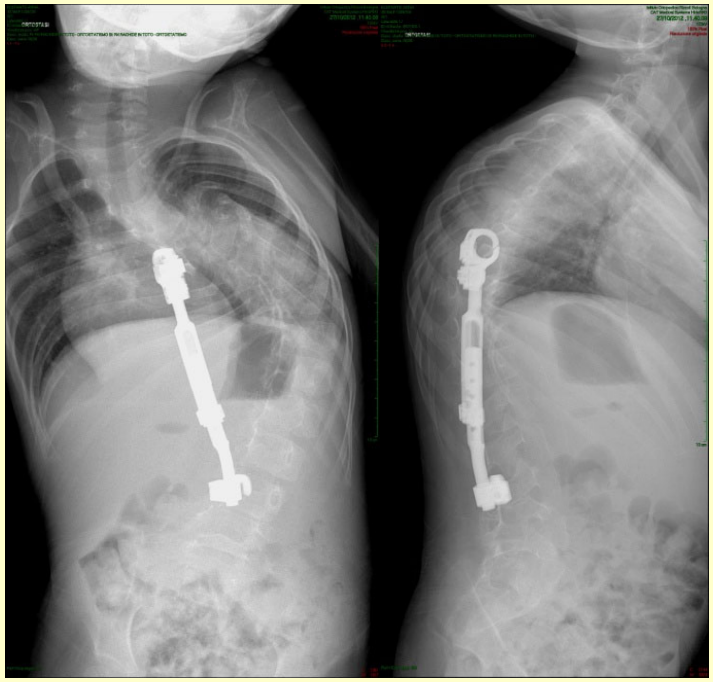


VEPTR after 2 lenghthening

Complication: pneumonia

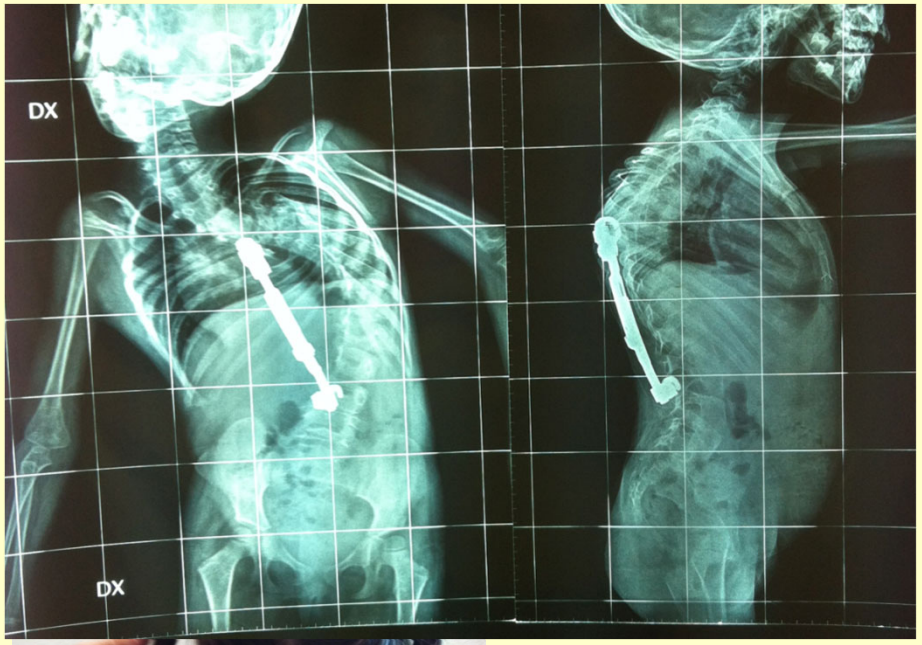
# ☑ 3rd Case

4 yy
Scoliosi in arthrogryposis



VEPTR
After 1 lengthening
Good sagittal correction
Less respiratory disease

# FU x-Rays I and Clinical



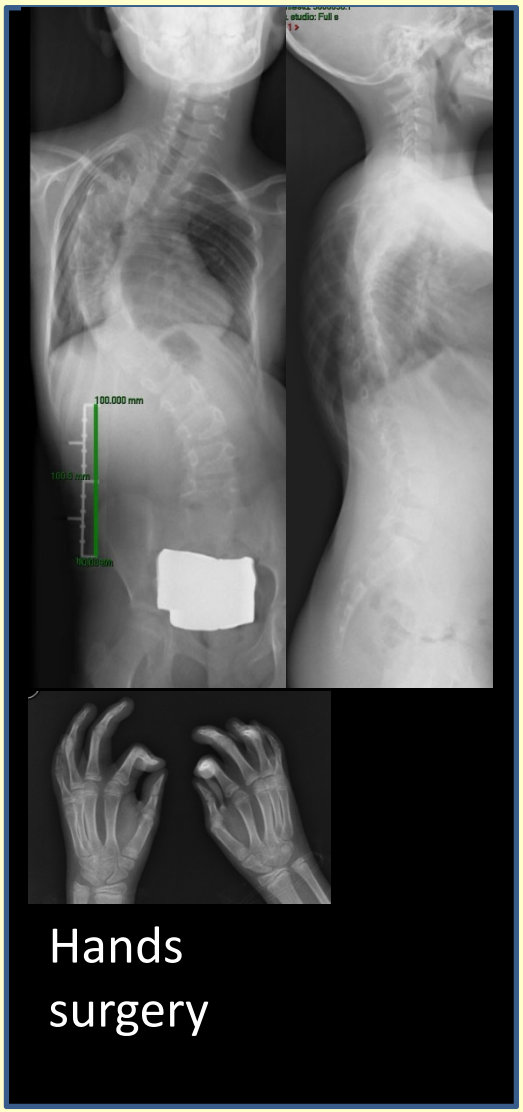

Complication:
PJK

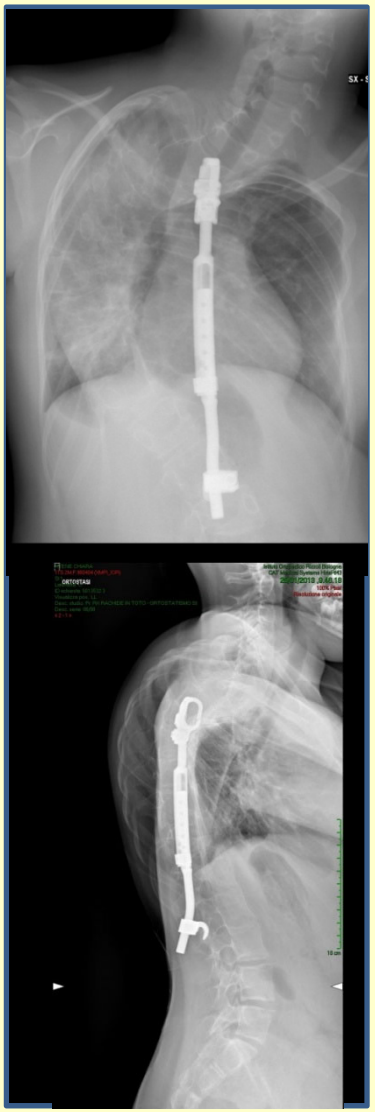
Revision +
Lenghthening

# ☑ 4st Case: Final Arthrodesis

Early onset scoliosis in Arthrogryposis. Cardiopathy, renal malformation

9 yy



VEPTR during the growth

13 yy

T2-L3 arthrodesis

# Results

First surgery:

•Scoliosis correction: 24.7%, from 113.2° to 85° (mean value)

•Kyphosis correction: 24.9% from 87.2 to 65.5°

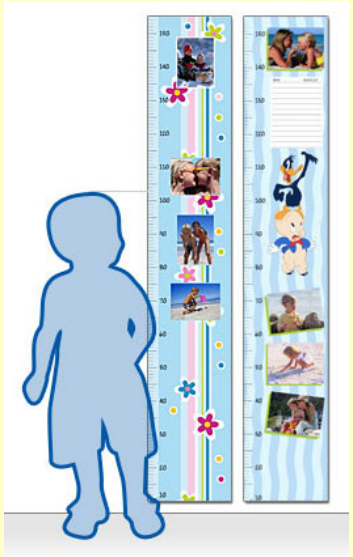
After 6 lenghthening procedures, 1.5 for each patient:

•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 arthogryposis 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.