



Altonaer  
Kinderkrankenhaus

Ein Unternehmen des UKE

# Skeletal dysplasias – Hamburg experience



R. Stücker



Department of Pediatric Orthopaedics

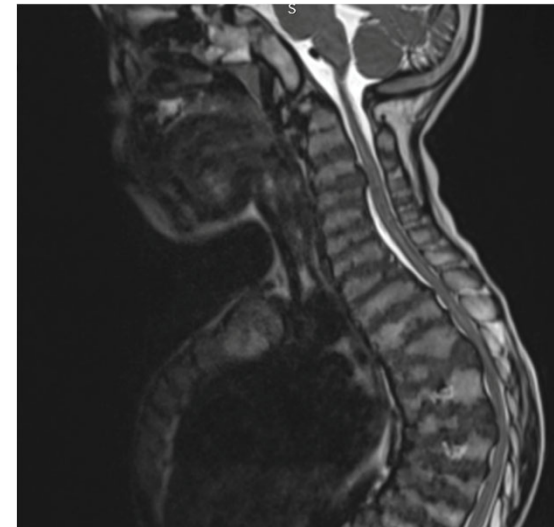
Pediatric Spine Center

The Childrens Hospital Hamburg Altona  
University Clinic Hamburg

- Travel expenses and speaker fees
  - Nuvasive
  - Depuy / Synthes

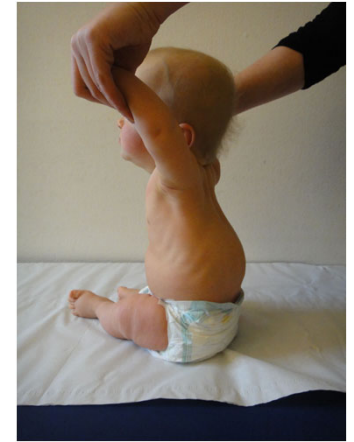
- Heterogenous group of about 400 diseases affecting development of cartilage and bone formation
- Incidence: 2-8 / 10.000
- Restrictive lung disease not uncommon
- Often accompanied by spinal involvement
  - Instability, spinal stenosis, deformities in coronal and sagittal plane
- Other problems of musculoskeletal system
  - Small stature, deformities of extremities

- Common spinal problems
  - Cervical spine instability and stenosis
  - Restrictive lung disease
    - Maintaining thoracic growth important
  - Spinal stenosis
  - Development of deformities (kyphosis, scoliosis)





- Early deformity correction and fusion
  - Achondroplasia
- Experience with growth preservation
  - VEPTR, GR
- The problems associated with kyphosis

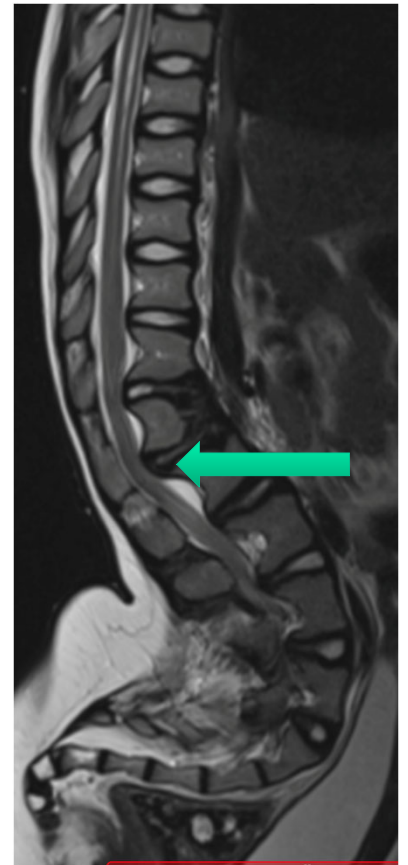
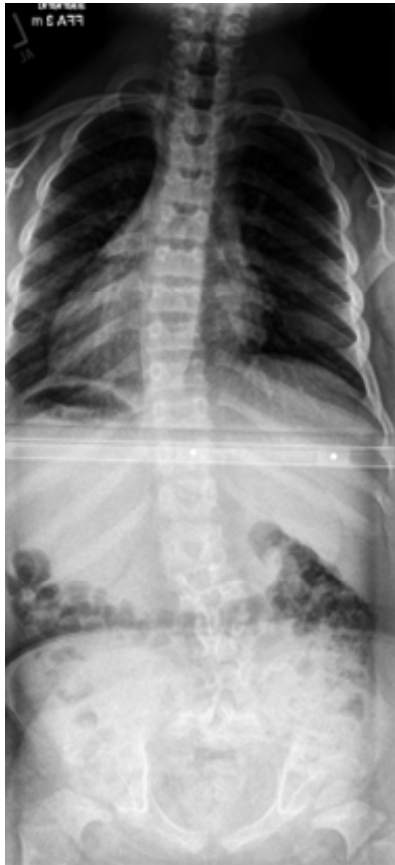




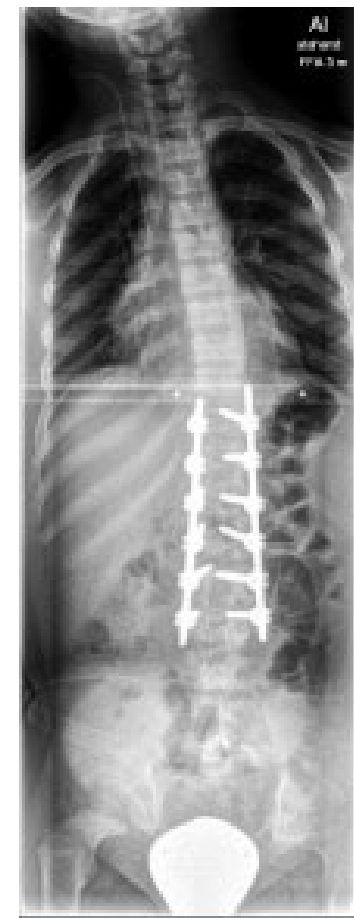
Altonaer  
Kinderkrankenhaus

Ein Unternehmen des UKE

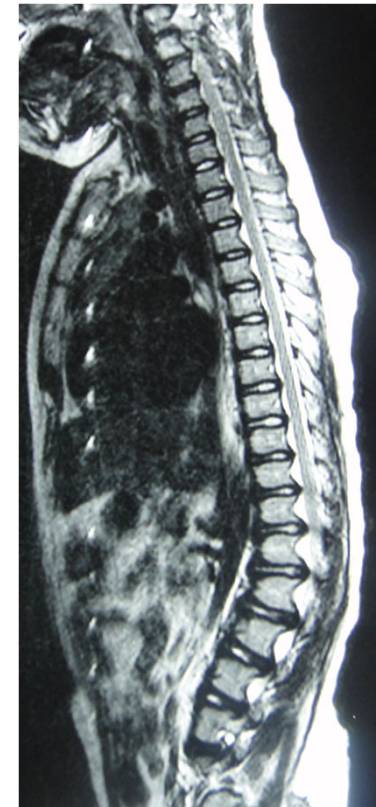
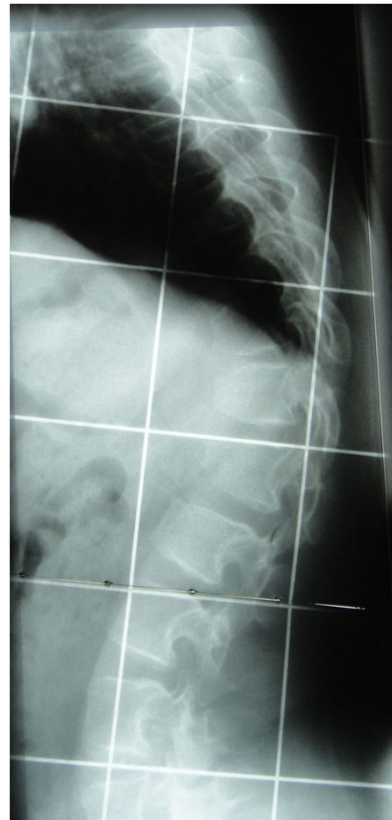
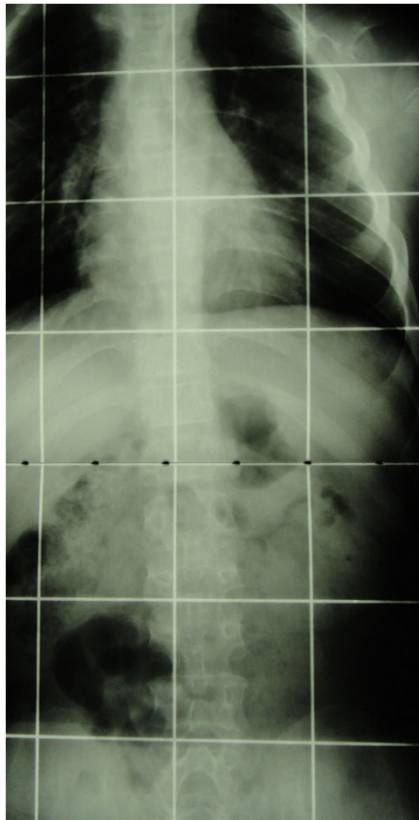
# 6 year old male patient with ACH, progressive kyphosis and claudication



- Had anterior release at 3 levels followed by posterior decompression and instrumentation



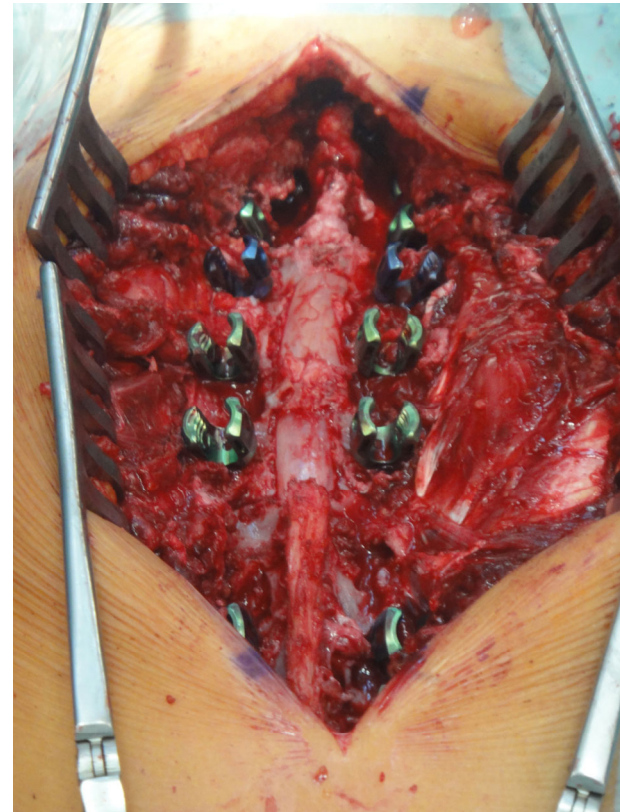
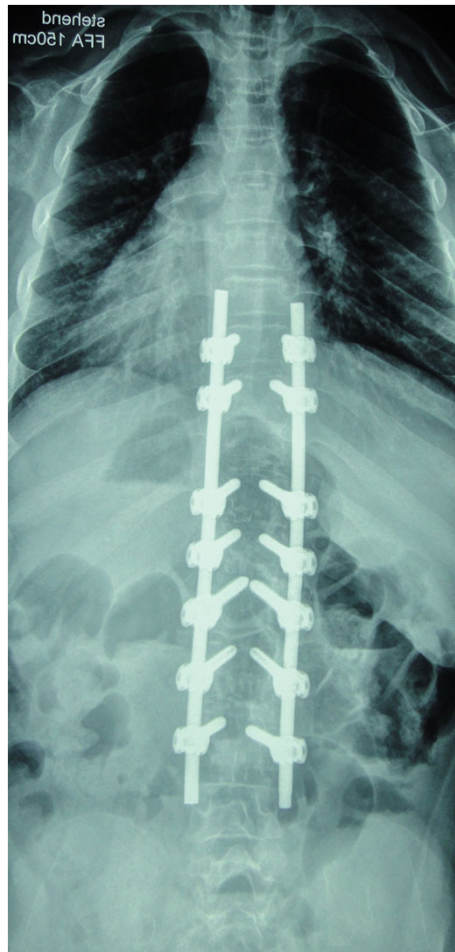
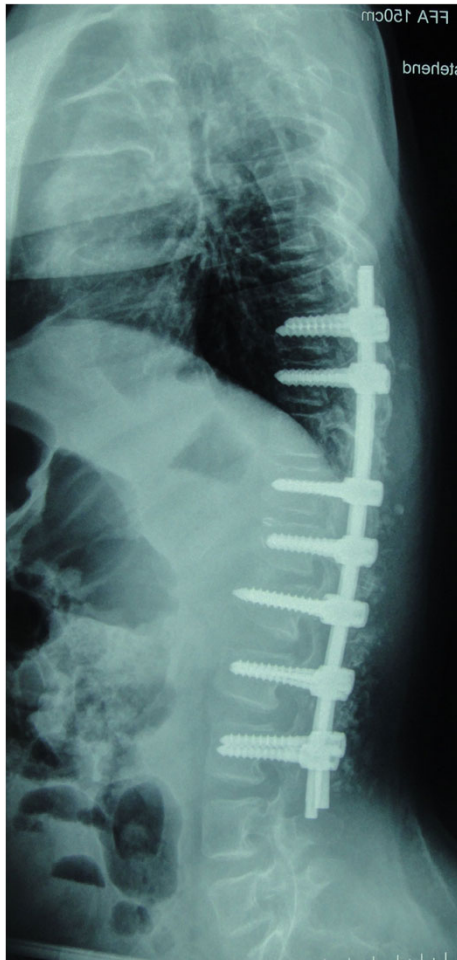
## 8 year old boy with Achondroplasia and symptoms of spinal canal stenosis



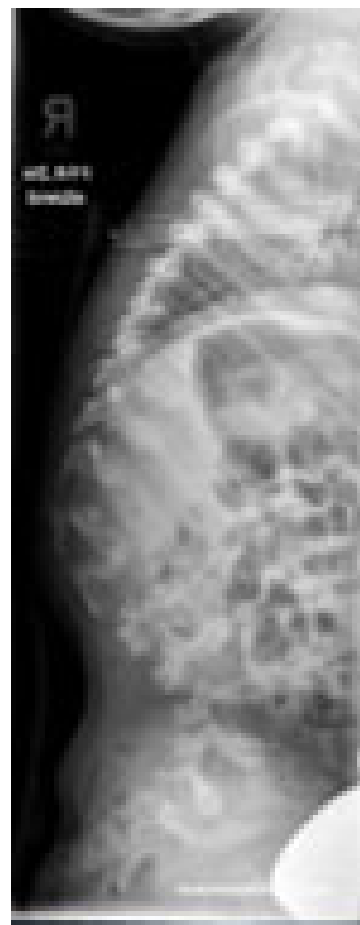
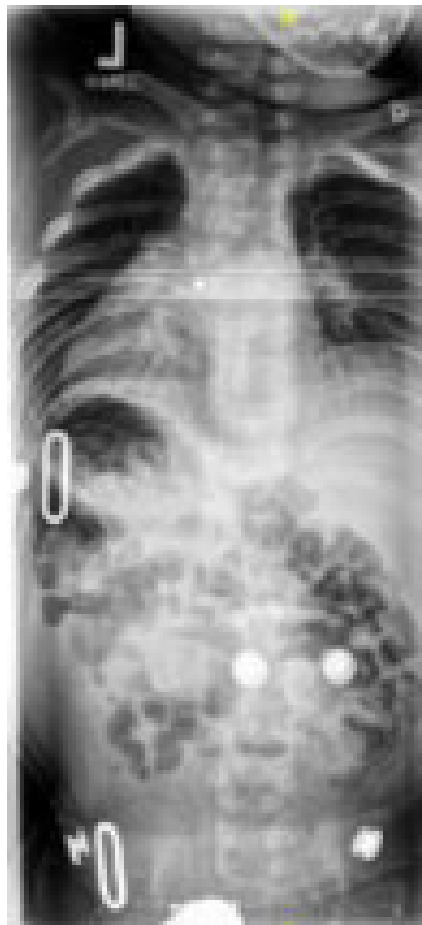
2 reasons for anterior surgery: remove bulging disks, facilitate fusion



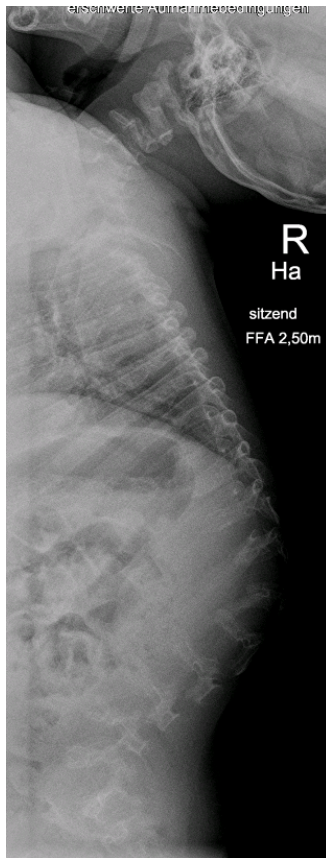
Approach: decompression by removal of 3-4 apical discs, followed by posterior decompression and fusion



3+4 years old boy with undefined skeletal dysplasia and incomplete paralysis of lower extremities

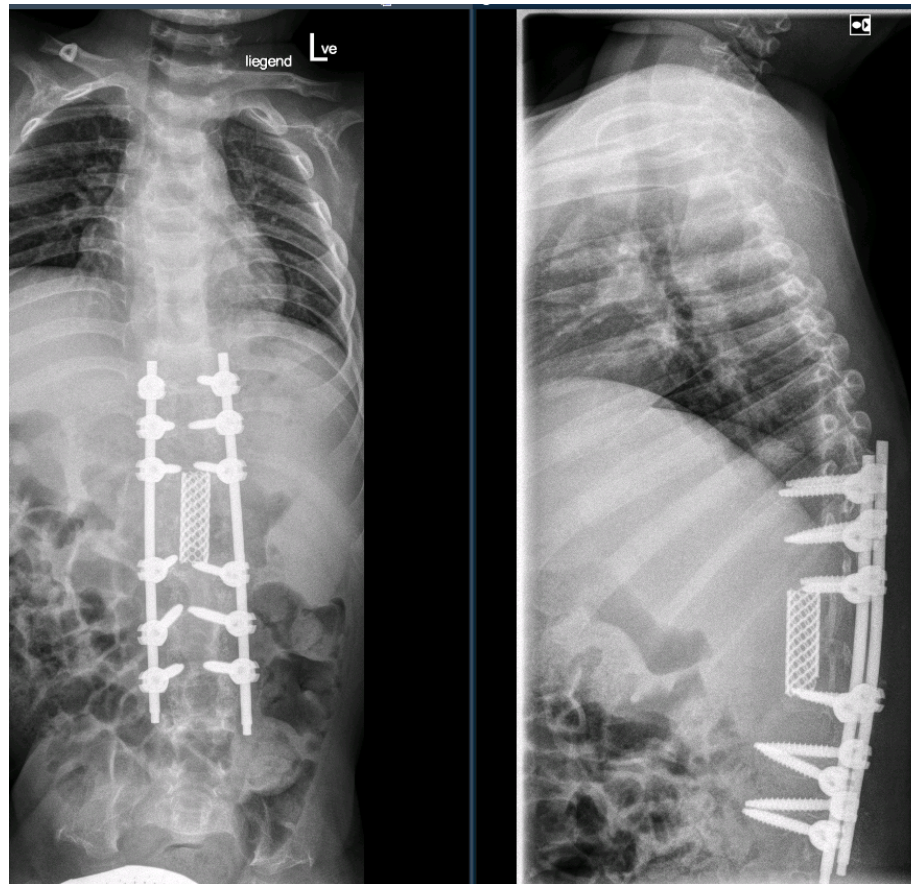


## Deterioration of paraplegia after posterior decompression incl. laminectomies





## Treatment by combined anterior and posterior approach T10-L3 with minor neurological improvement



- Progressive kyphosis
- Failed bracing
- Claudication
- In ACH anterior multilevel decompression recommended
- Development of thoracic lordosis

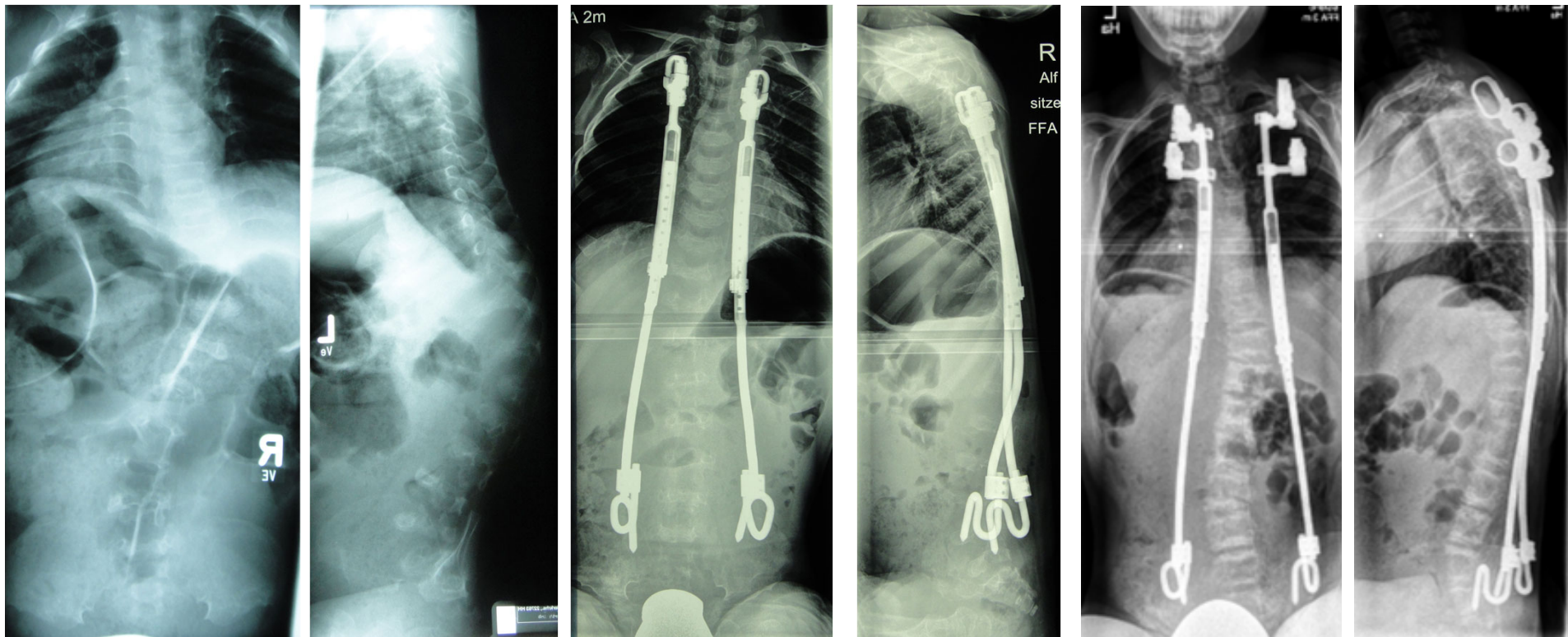
- Progressive scoliosis
- Thoracic kyphosis at young age
- Pulmonary issues



Altonaer  
Kinderkrankenhaus

Ein Unternehmen des UKE

5 year old male patient with spondylo-  
metaphyseal dysplasia, mental retardation,  
not ambulant

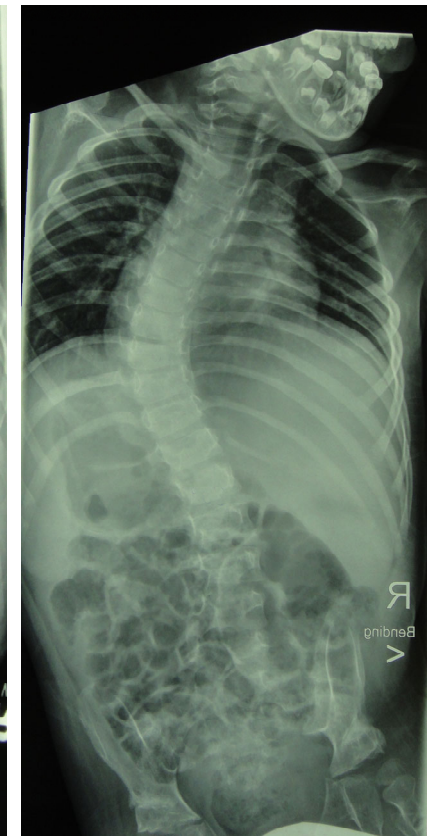
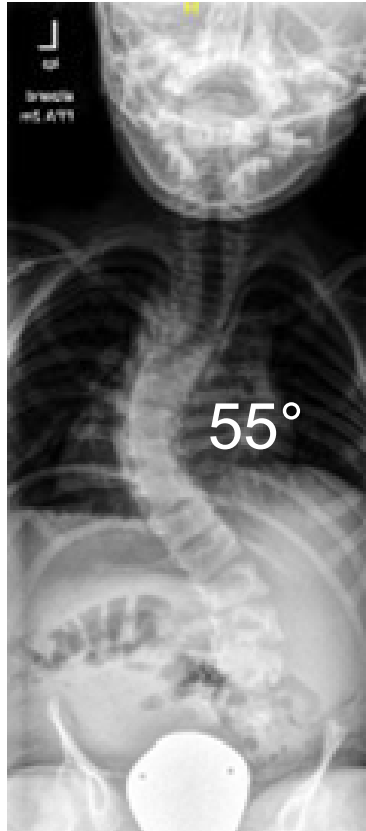


postop

6 years f/u

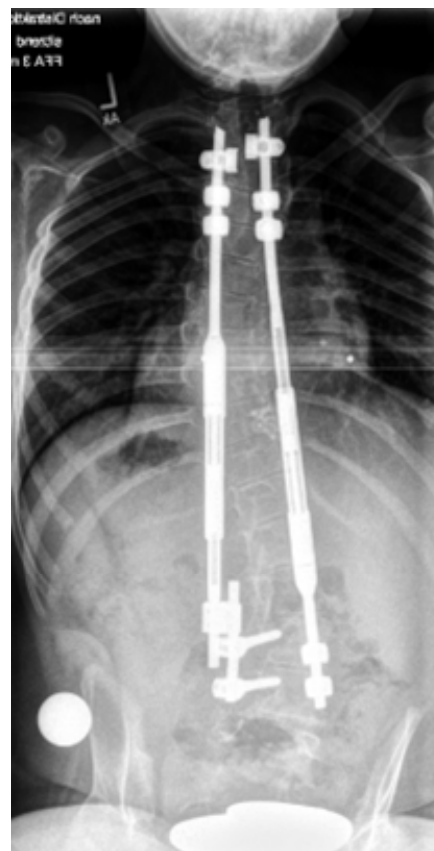


# 5+7 year old girl with spondyloepi-metaphyseal dysplasia and progressive scoliosis



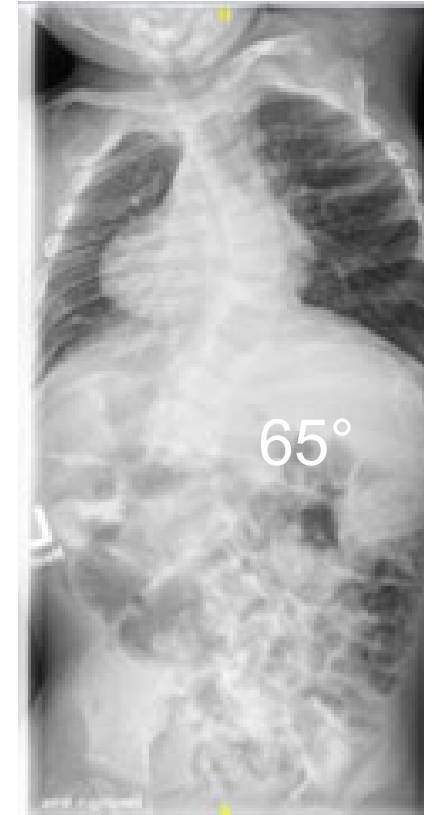
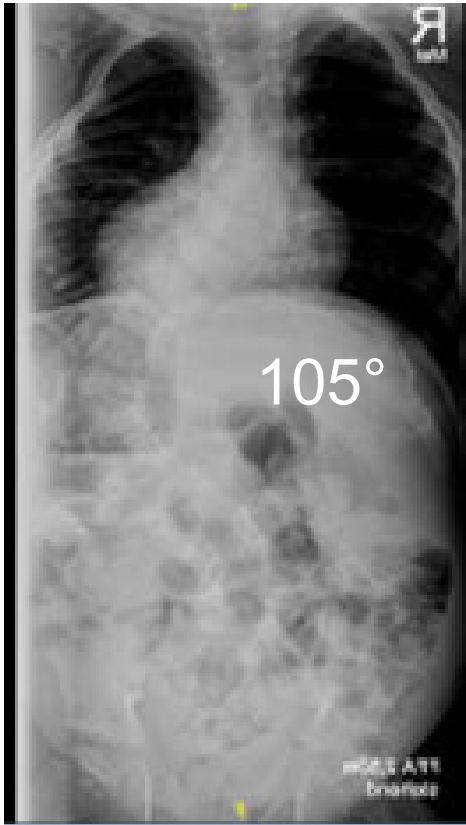
Bending





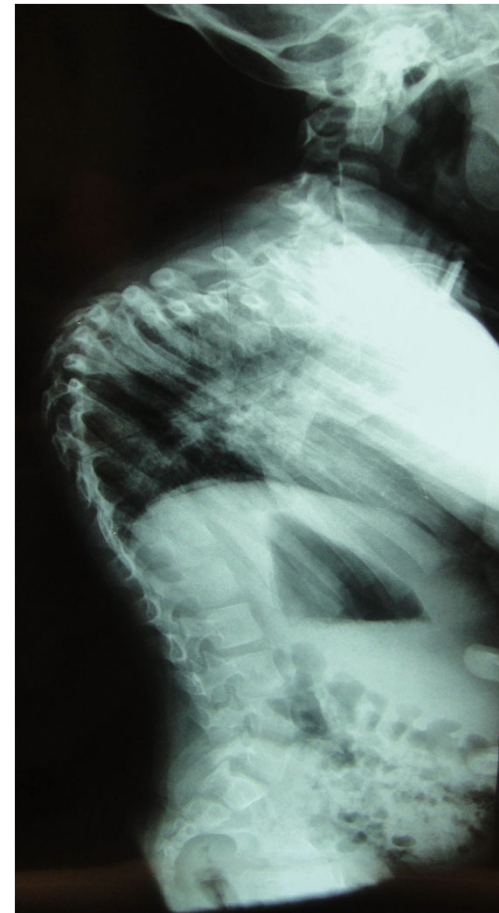
4 years f/u



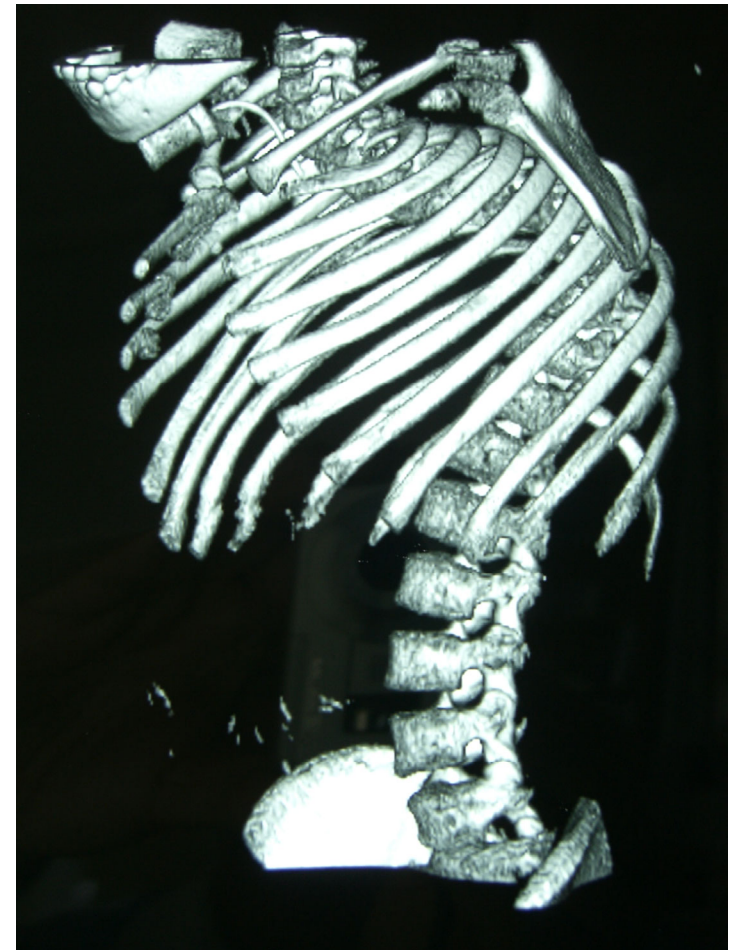
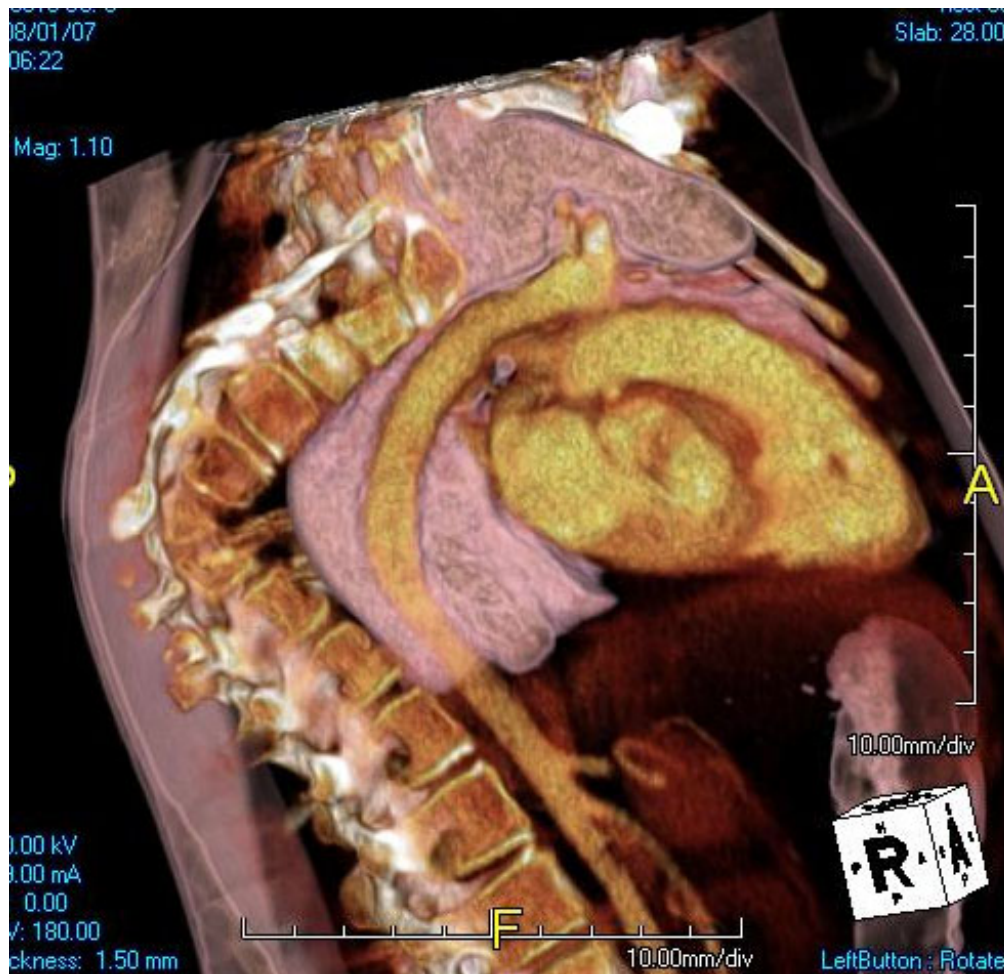




4 year old girl with campomelic dysplasia, 115° kyphosis.  
Treatment with initial halo traction, then anterior VEPTR for 6 years



**Severe vertebral dysplasia with  
segments of failure of formation,  
severe chest deformity**

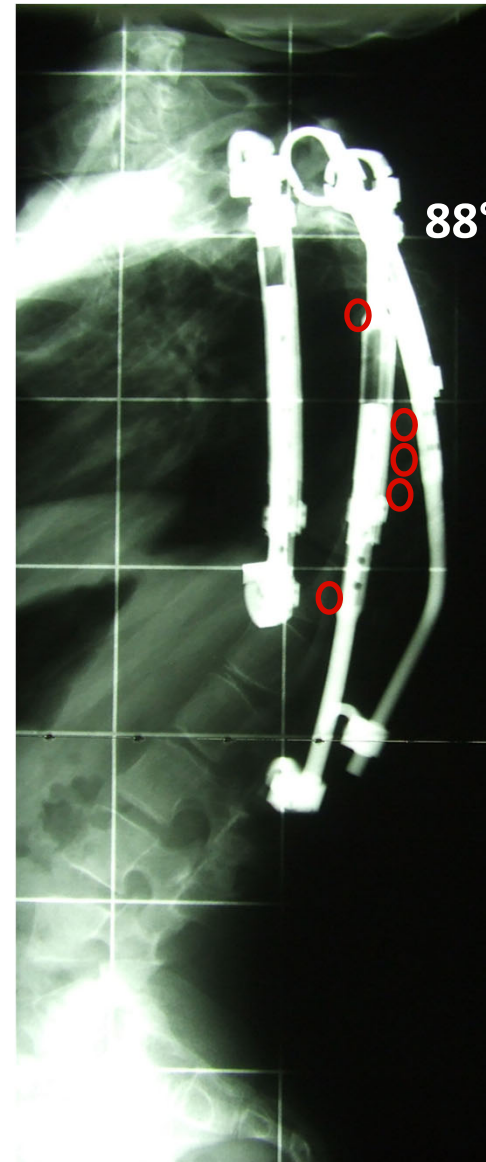
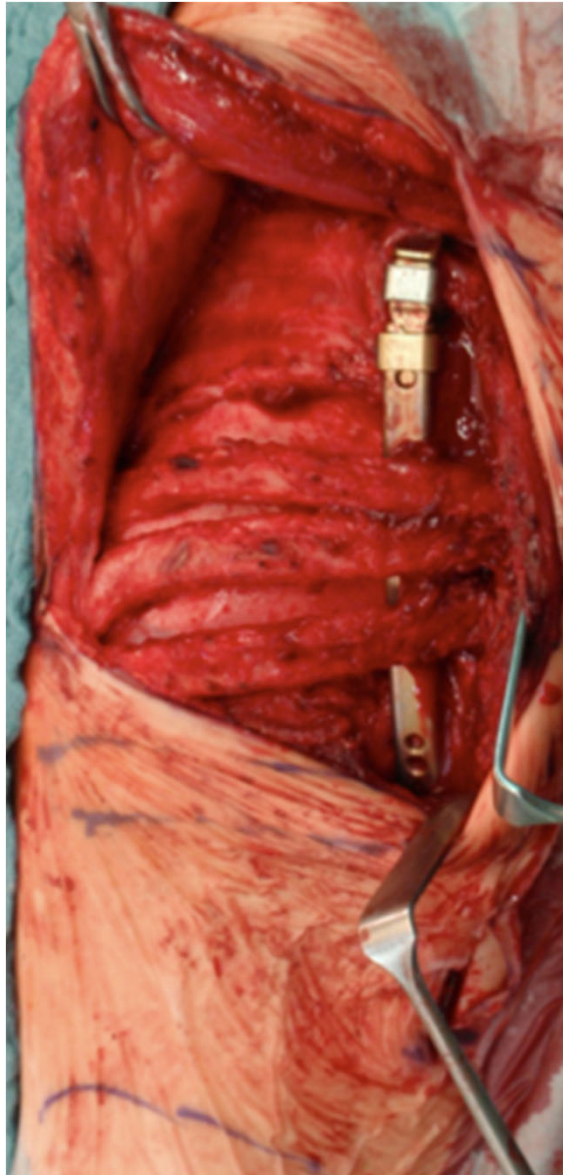






Altonaer  
Kinderkrankenhaus

Ein Unternehmen des UKE

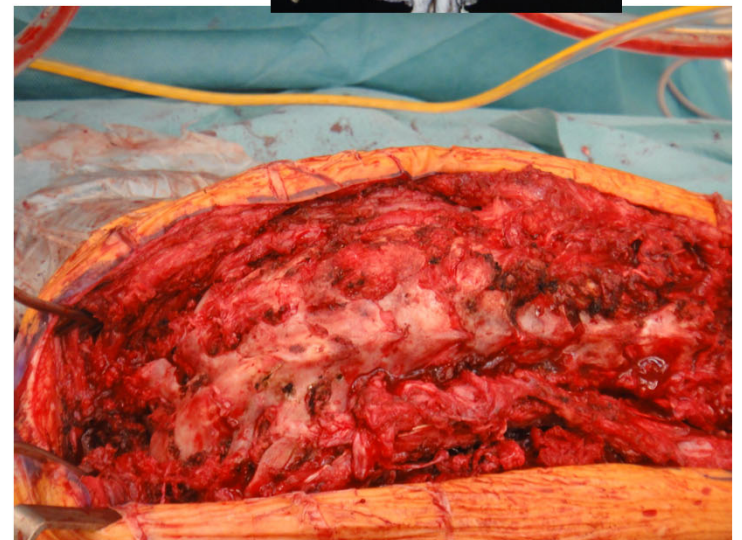
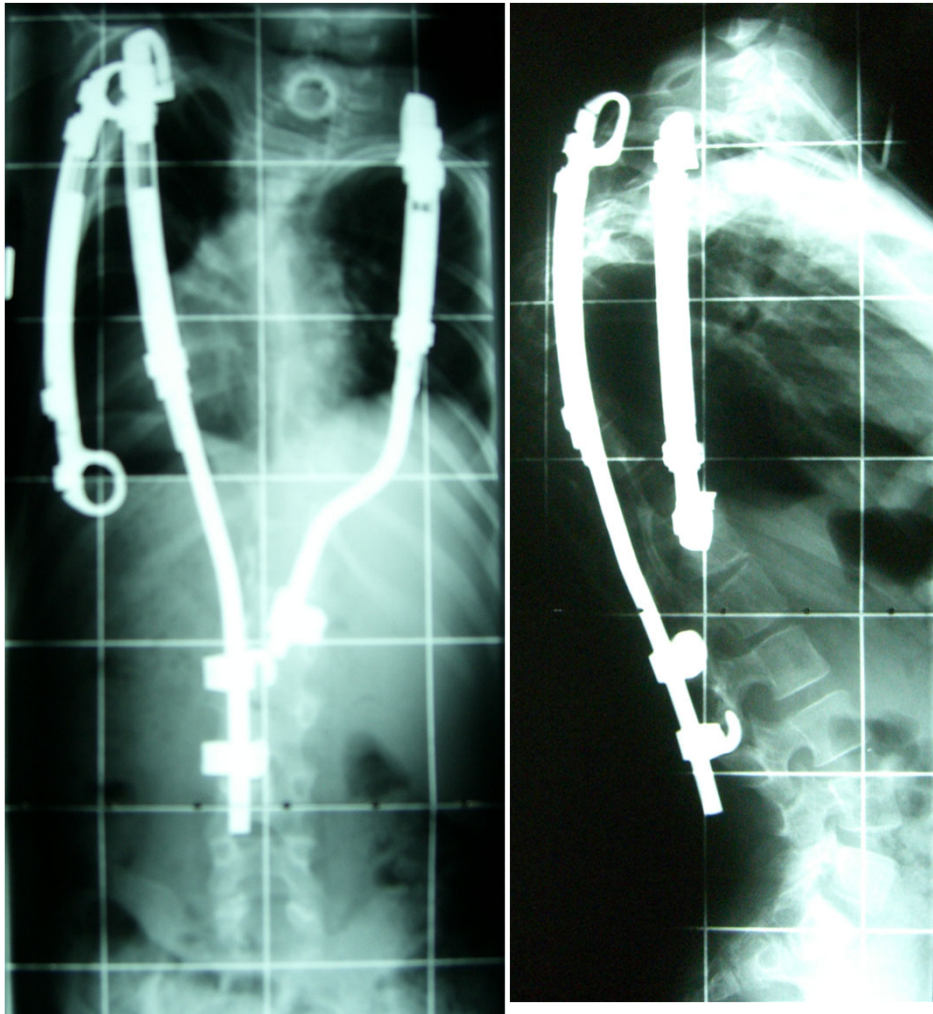




Altonaer  
Kinderkrankenhaus

Ein Unternehmen des UKE

The situation after 6,5 years  
had 3 revisions



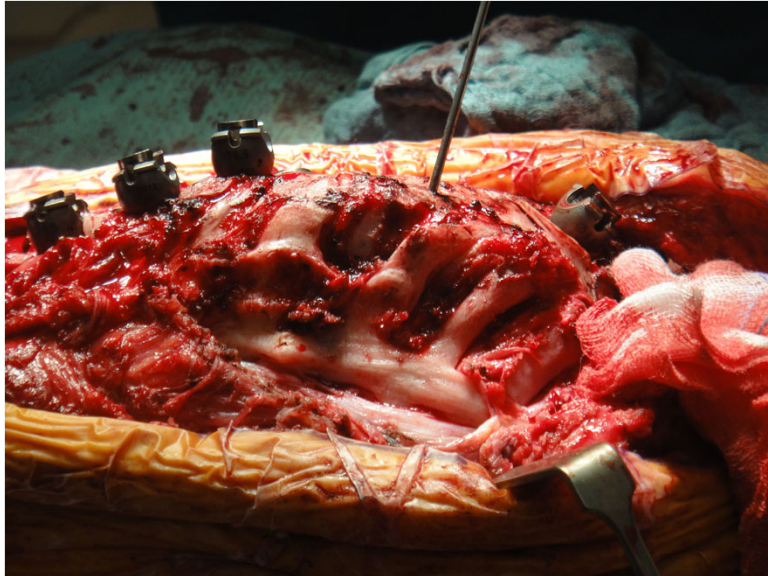




Altonaer  
Kinderkrankenhaus

Ein Unternehmen des UKE

## Final correction with 11 years



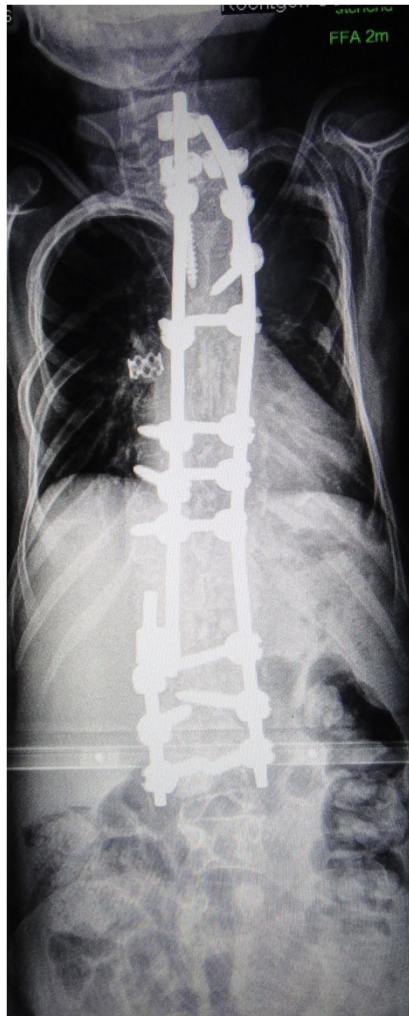




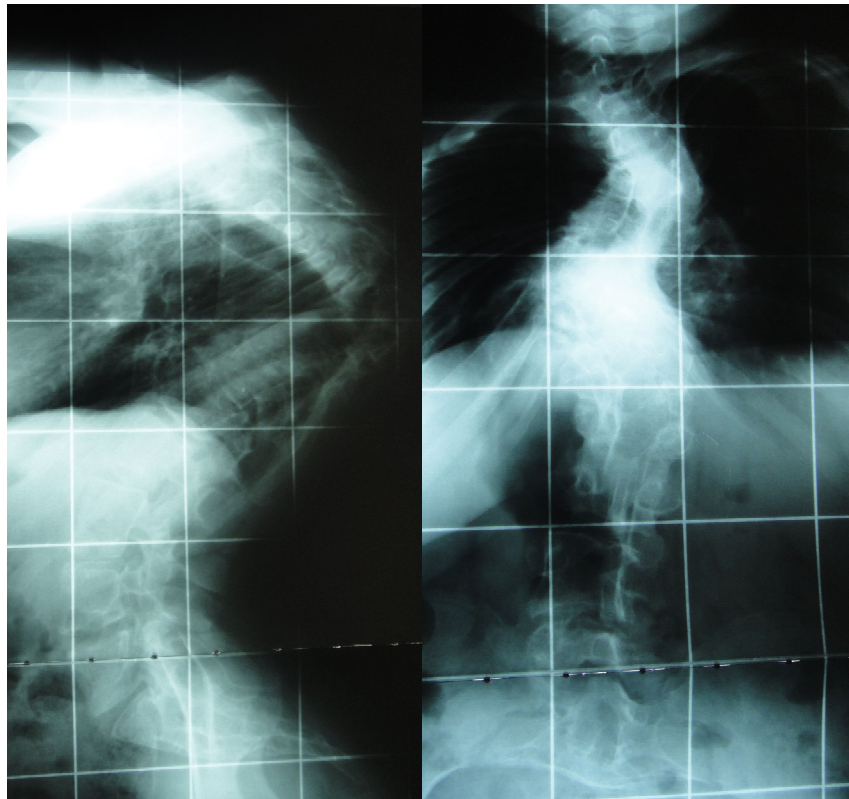
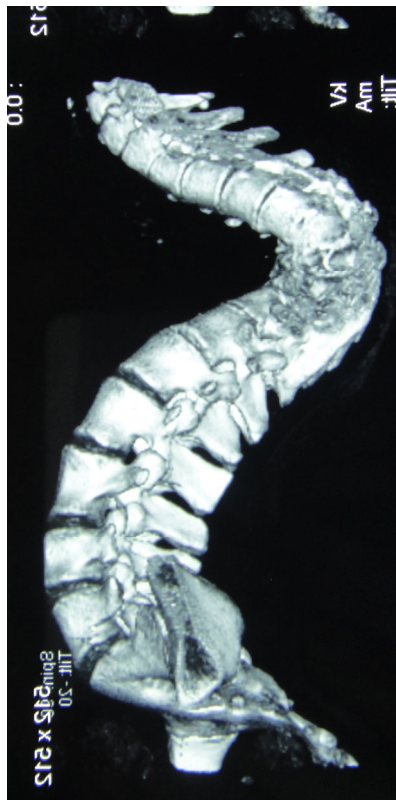
Altonaer  
Kinderkrankenhaus

Ein Unternehmen des UKE

## Final correction by VCR + multiple osteotomies



# 6 year old patient with Conradi Hunermann Syndrome Had apical posterior spine fusion to halt progression - failed



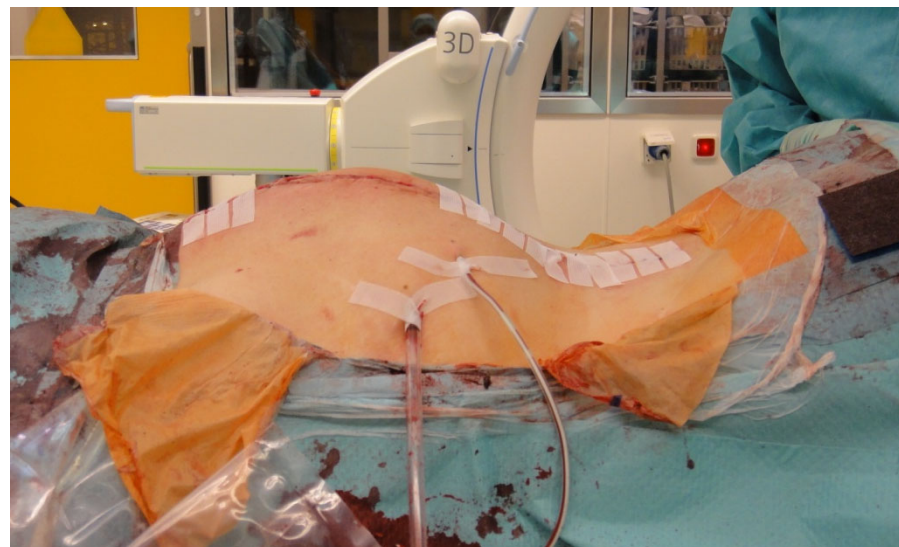
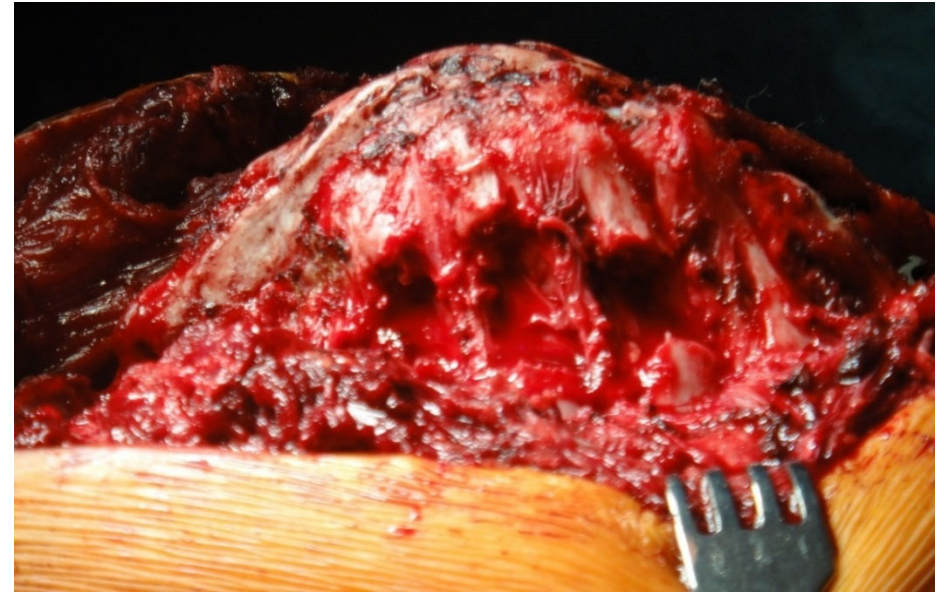




Altonaer  
Kinderkrankenhaus

Ein Unternehmen des UKE

## Correction by VCR and multiple osteotomies

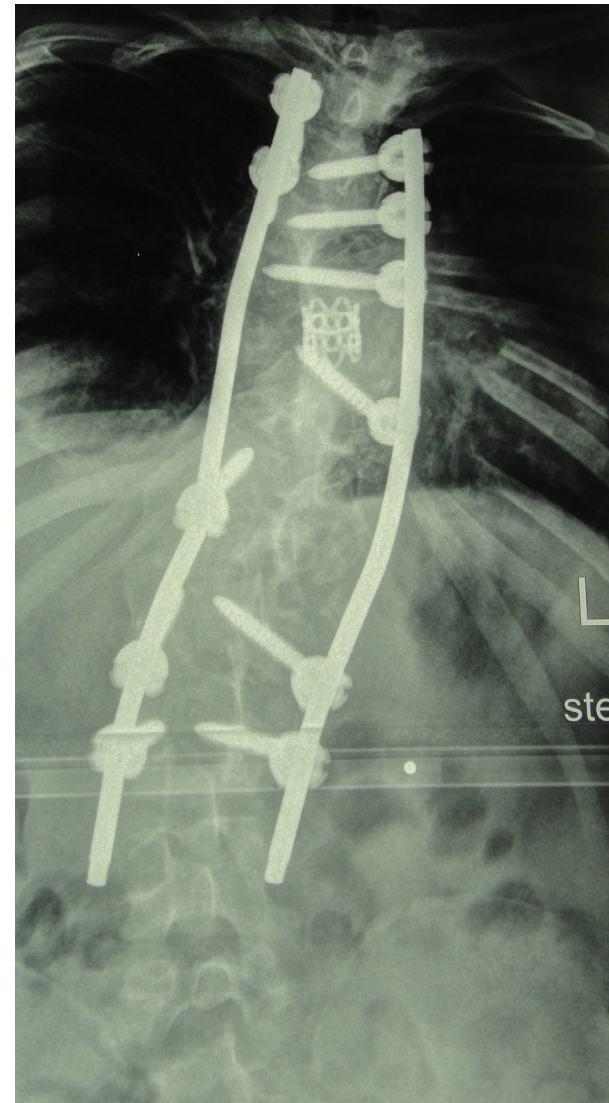
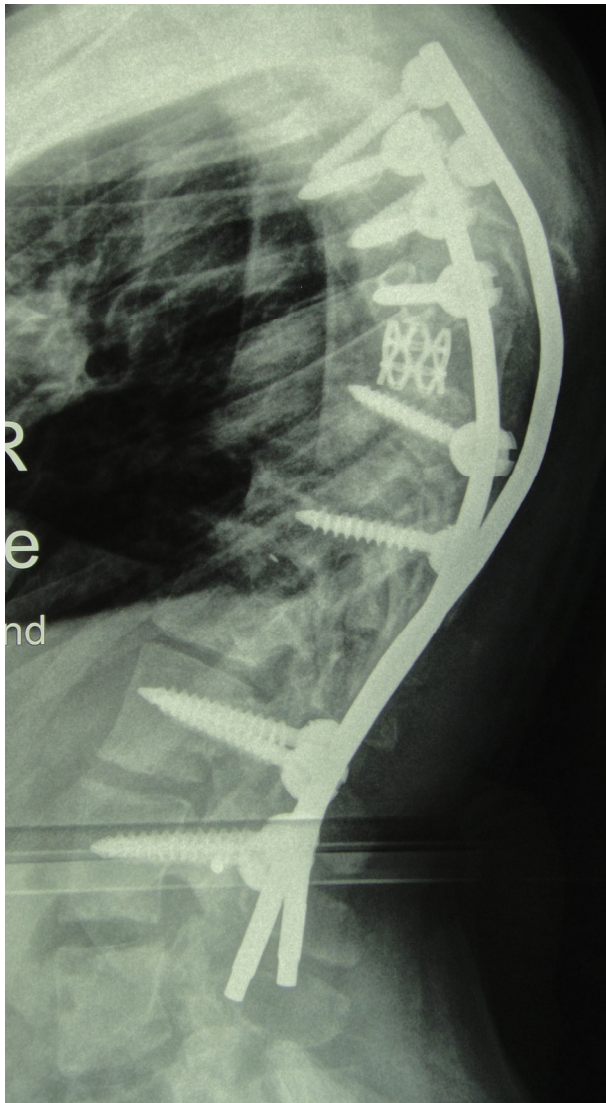




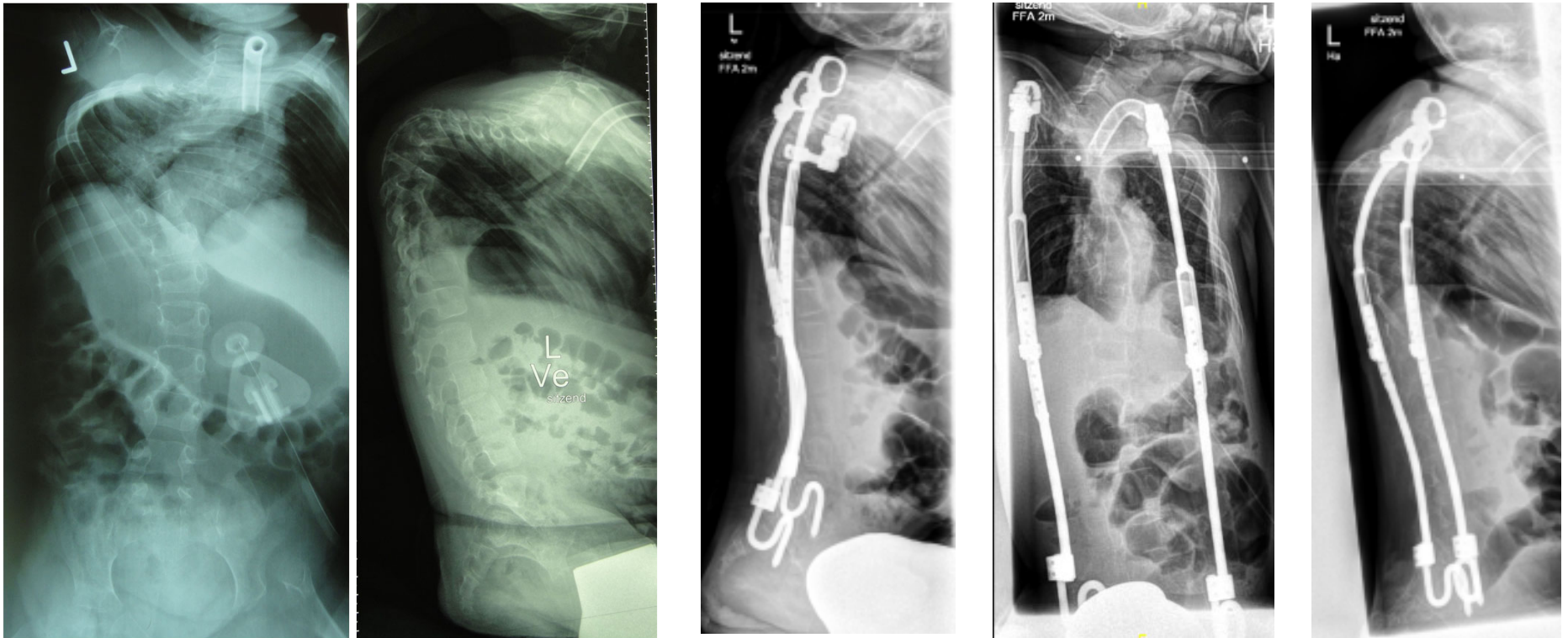
Altonaer  
Kinderkrankenhaus

Ein Unternehmen des UKE

Was corrected to 40°







6 year follow up after Halo traction, VEPTR  
instrumentation, posterior apical fusion, revision due to  
infection and new VEPTR implantation  
Kyphosis was well controlled

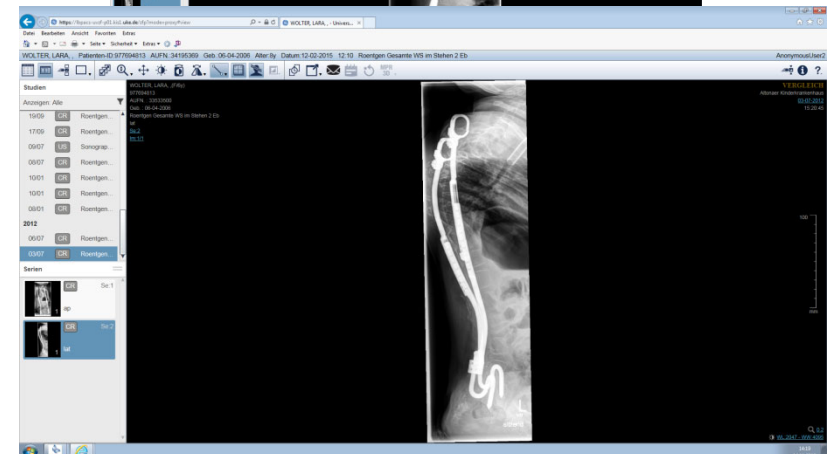
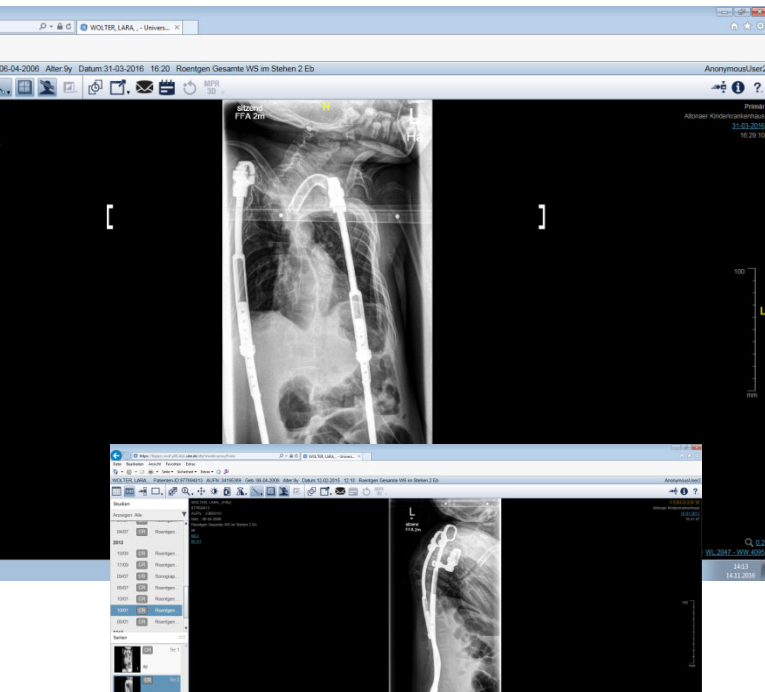
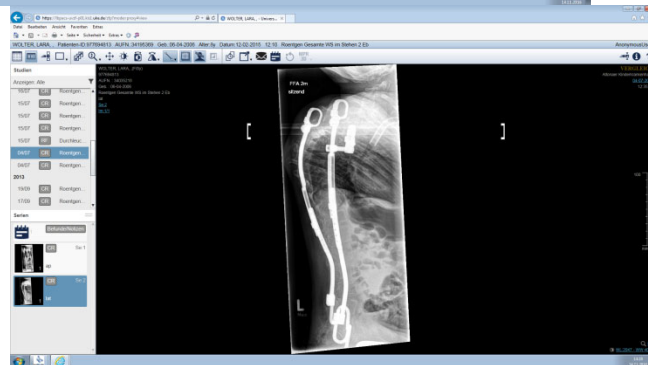
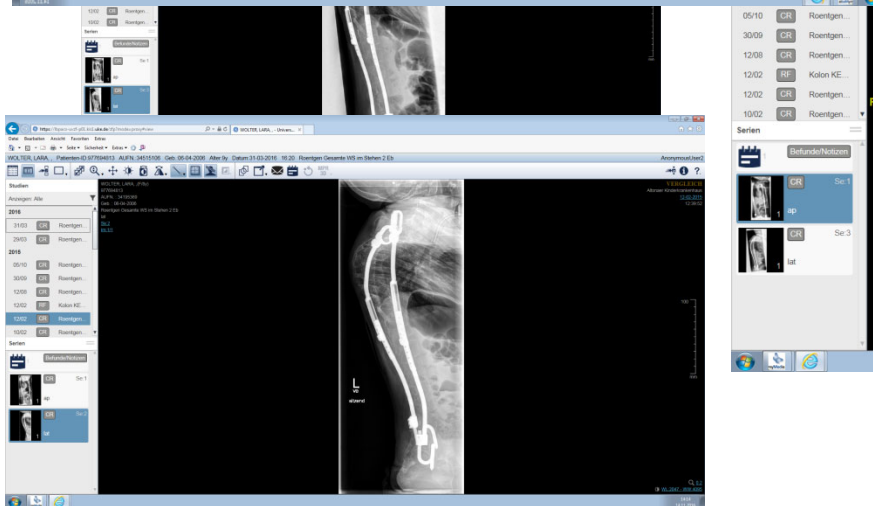
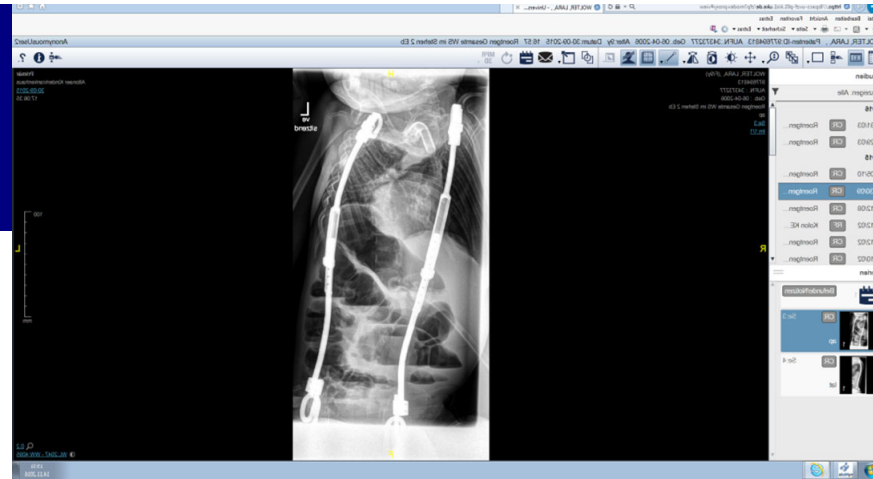
- Thoracolumbar kyphosis best treated by short fusion +/- decompression
  - Anterior decompression for some cases (ACH)
- Early onset scoliosis (EOS) can be treated by VEPTR or GR
- Kyphosis still a major challenge
  - Best treatment has still to be determined
  - Currently we perform VEPTR + posterior apical fusion with promising results

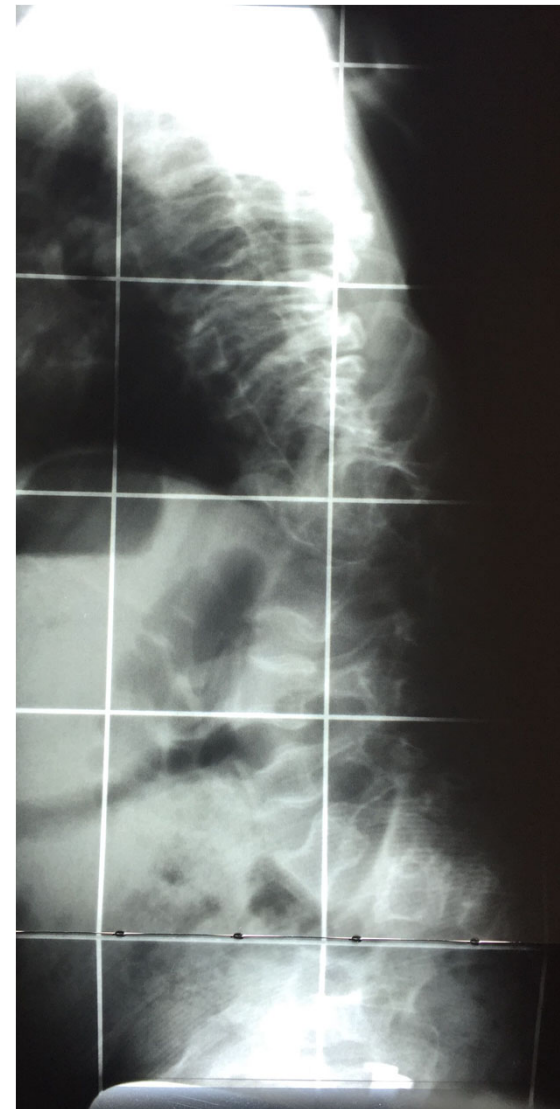
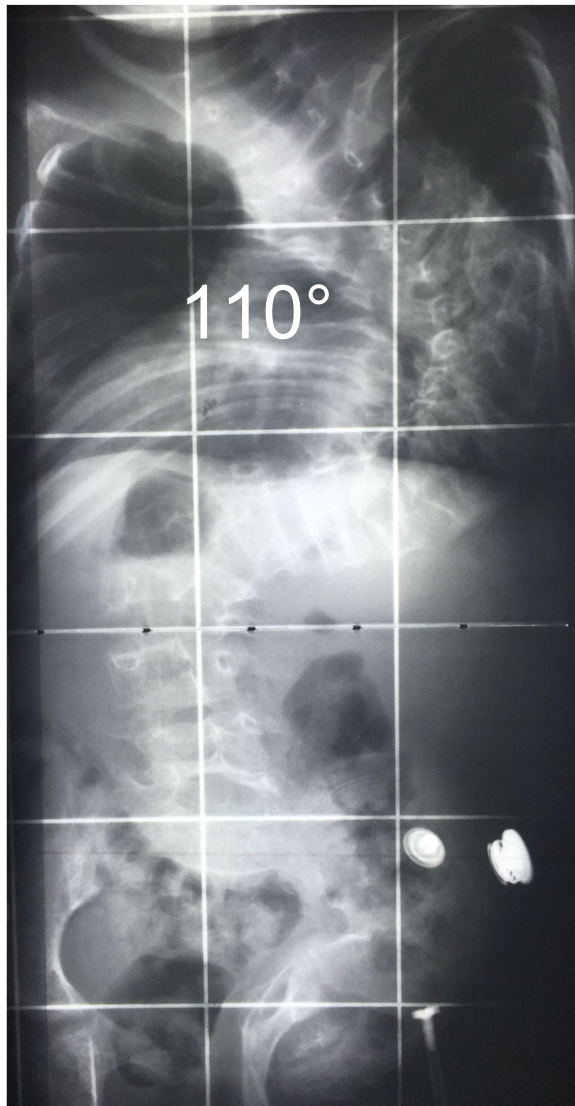


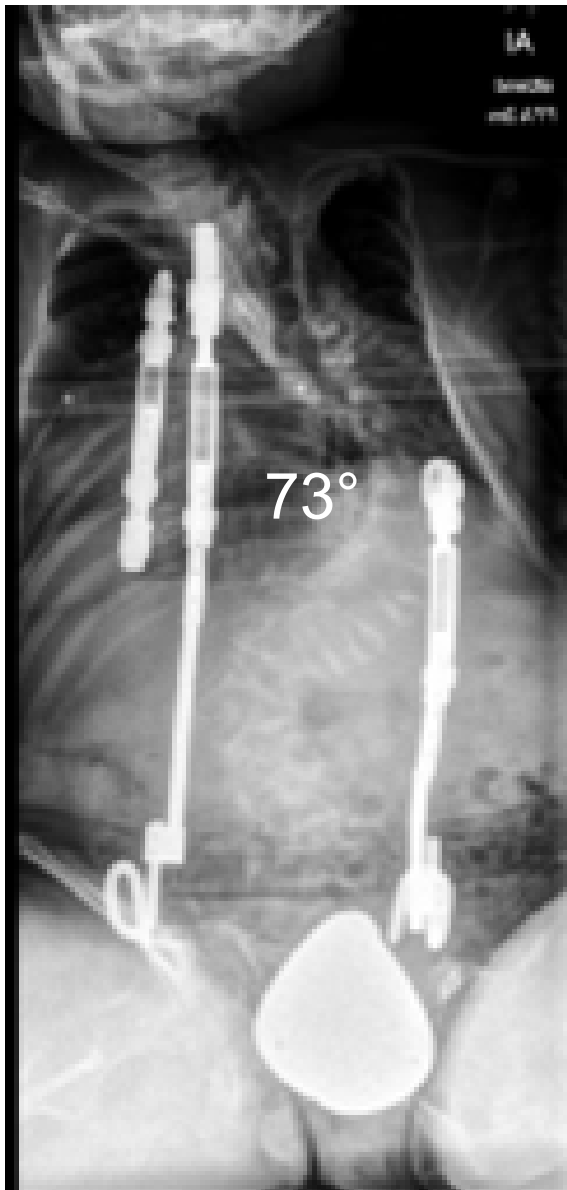




# Wolter, Lar

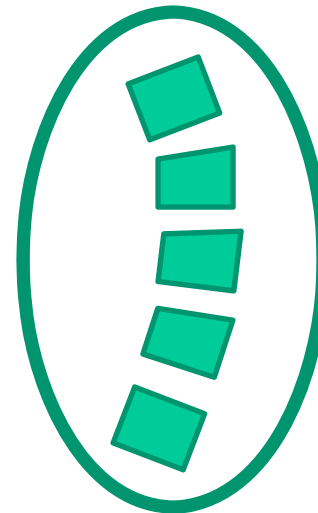






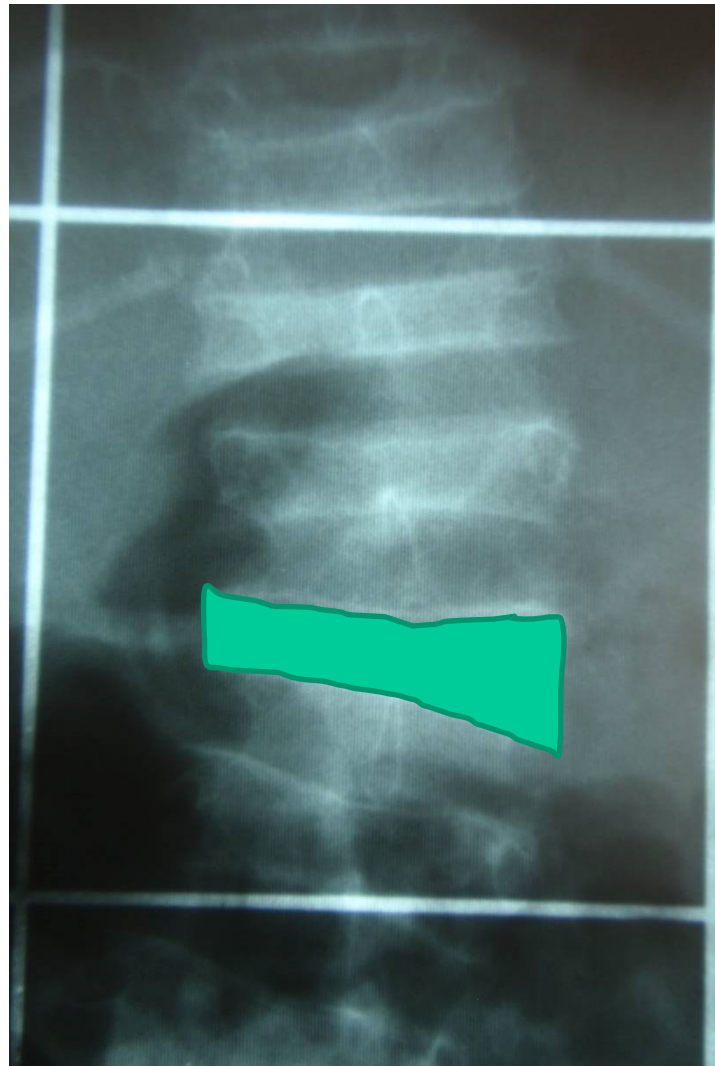
# Scoliosis and Osteogenesis Imperfecta

- Incidence: 20-80%
- Incidence increases with severity of disease
- Risk factors: ligamentous laxity and bone mass loss



An important reason for development of scoliosis  
is vertebral deformity







Altonaer  
Kinderkrankenhaus

Ein Unternehmen des UKE

Growth hormone treatment not  
effective for

- Metaphyseal chondrodysplasia



Altonaer  
Kinderkrankenhaus

Ein Unternehmen des UKE

Until recently it was unclear whether  
bisphosphonates are able to decrease  
the incidence of scoliosis

- 157 of 315 Patienten had scoliosis
- Type III: 68% prevalence und progression rate of  $6^{\circ}$  / year
  - If bisphosphonates-treatment begins before age 6, prevalence decreases to  $3,8^{\circ}$ /year
- Type IV: 54% prevalence, progression rate  $4^{\circ}$  / year
- Type 1: 39 % prevalence, progression rate  $1^{\circ}$  /year



- Prevalence of scoliosis
  - Type 3: 89%
  - Type 4: 61%
  - Type 1: 36%
- Bisphosphonates decrease progression rate in type 3 only
- No difference in progression rate in type 1 and 4
- The prevalence of scoliosis at maturity was not influenced by the bisphosphonate treatment history in any OI type.

- Non fusion techniques
  - New methods
  - For younger children
- Fusion techniques
  - Established methods
  - For children  $> 11$  years

- Curves usually stiff
  - Little correction on side bending
- Look at spondylolysis and spondylolisthesis
- Avoid large diameter screws (pedicle fractures)
- Consider sublaminar bands at apex of deformity
- Watch for thoracic deformities making approach to spine impossible

## Spondylolysis and Spondylolisthesis in O.I.

- Spondylolysis:
  - 8,2 % at 7,5 years
- Spondylolisthesis:
  - 10,9 % at 6,5 years



Prevalence of spondylolysis and  
spondylolisthesis about 20%





Altonaer  
Kinderkrankenhaus

Ein Unternehmen des UKE



# Chest wall deformities in O.I

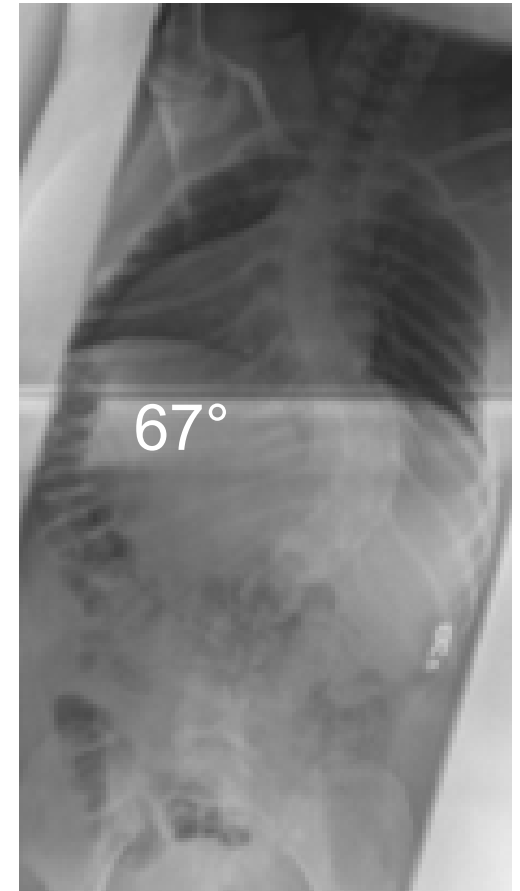
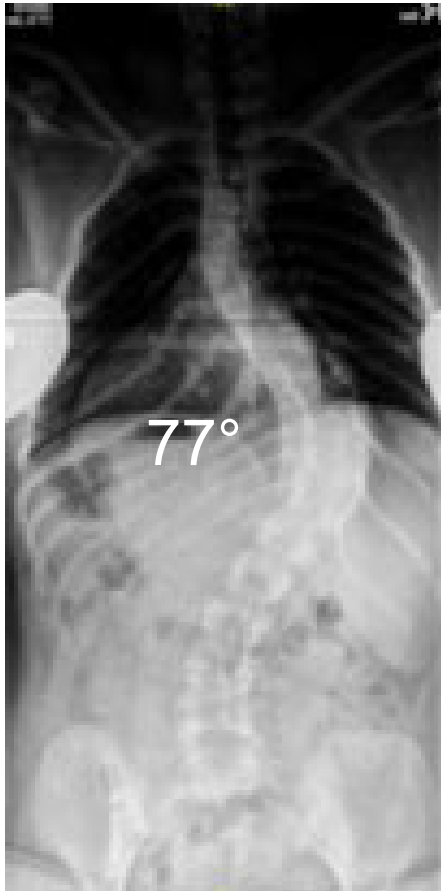


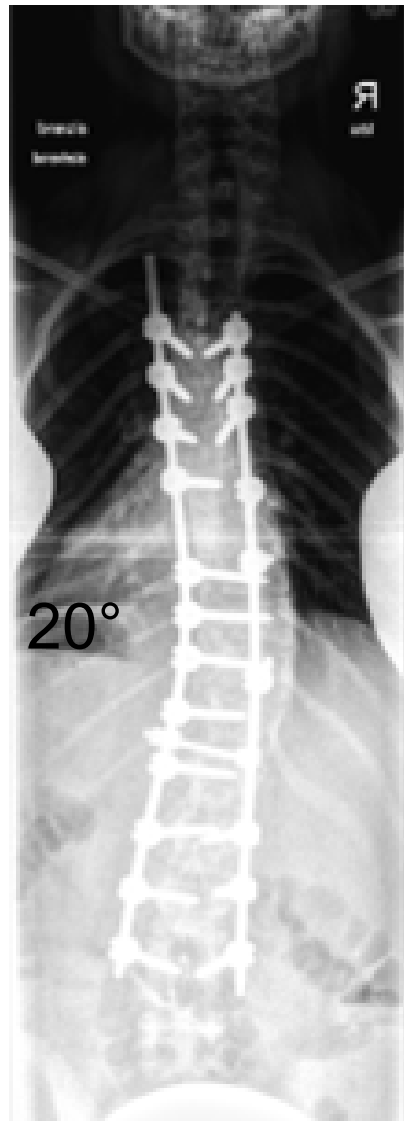


Altonaer  
Kinderkrankenhaus

Ein Unternehmen des UKE

# 17 year old girl with O.I., type 1



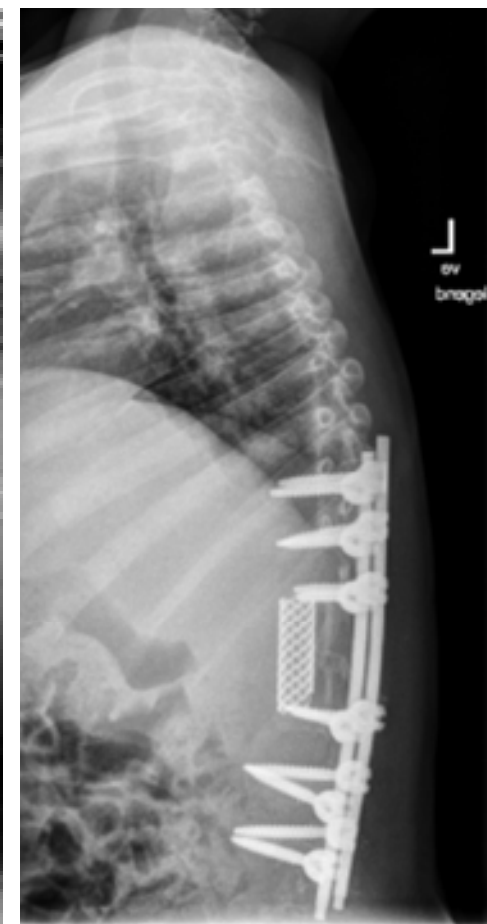
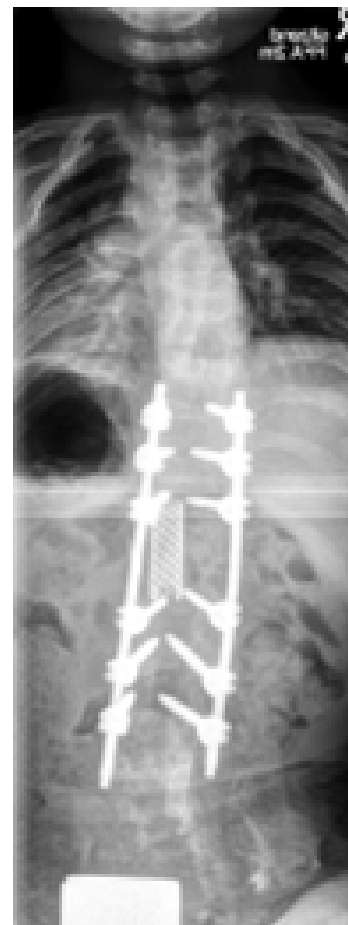
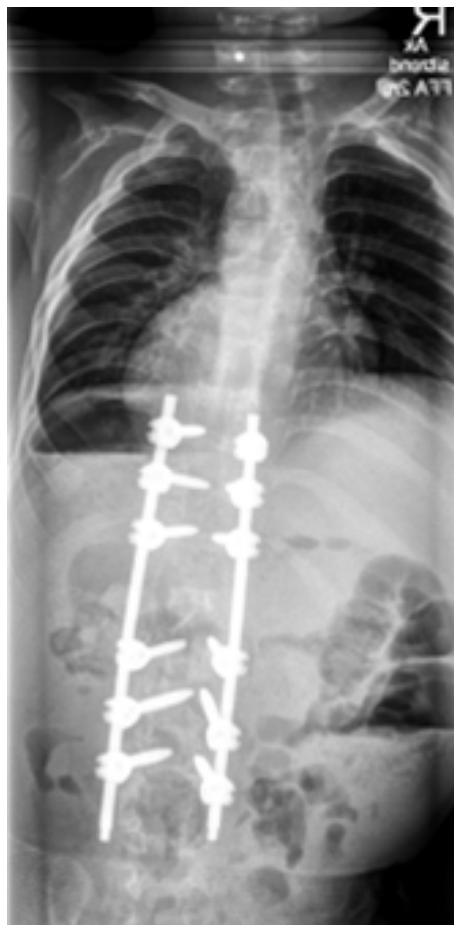






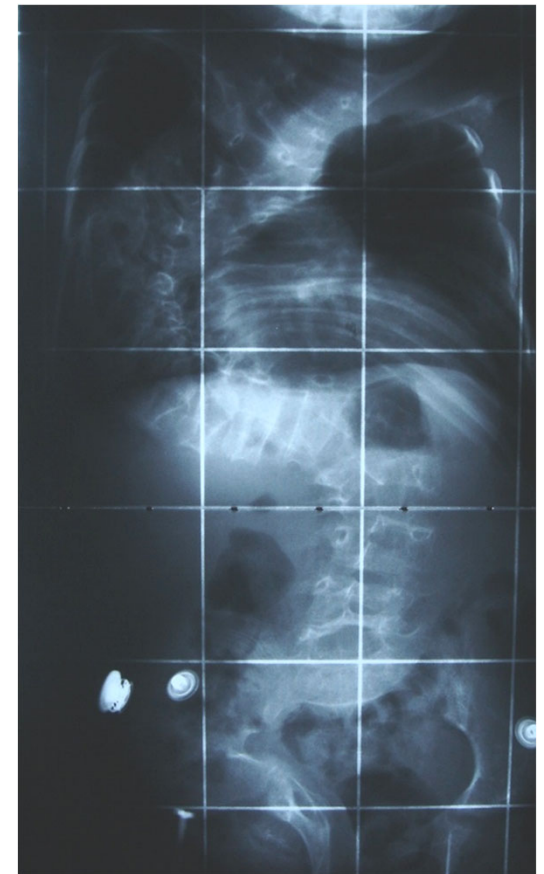
Altonaer  
Kinderkrankenhaus

Ein Unternehmen des UKE



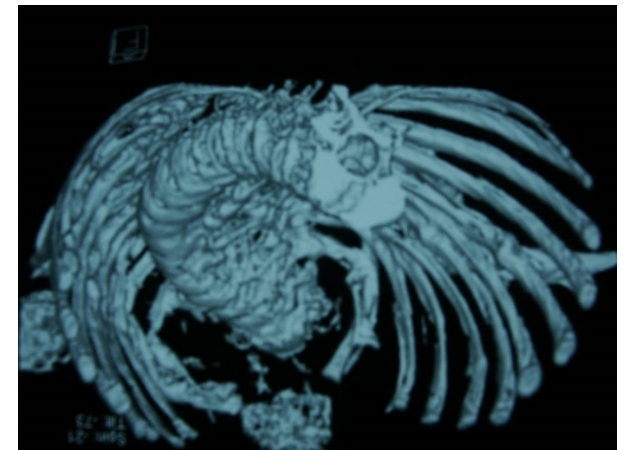
- Pulmonary failure leading cause of death in adults
- Vital capacity  $< 50\%$  if thoracic curve  $> 60^\circ$
- Early development of motor milestones inversely related to severity of scoliosis

- Deformities usually progressive
- Accompanied by chest wall deformities
  - Reduction of chest height and width
- May lead to thoracic insufficiency syndrome (TIS)
- But develop barrel chest deformity as a rescue strategy
- Often painful
- Further loss of mobility and loss of bone mass





- Curves usually rigid
  - Especially thoracic region with rigid rib cage
- Pedicles often brittle
- Often severe rotation
- Often severe kyphosis
- Possible correction of curve: 30-50%



- Physical therapy
  - No evidence of effectiveness
  - Respiratory training and exercises recommended
- Brace
  - Only for few patients with type 1
  - May lead to chest wall and further rib deformities
  - May lead to impaired breathing function



# Treatment of scoliosis in patients with O.I.

Yong-Hing, MacEwen, 1983

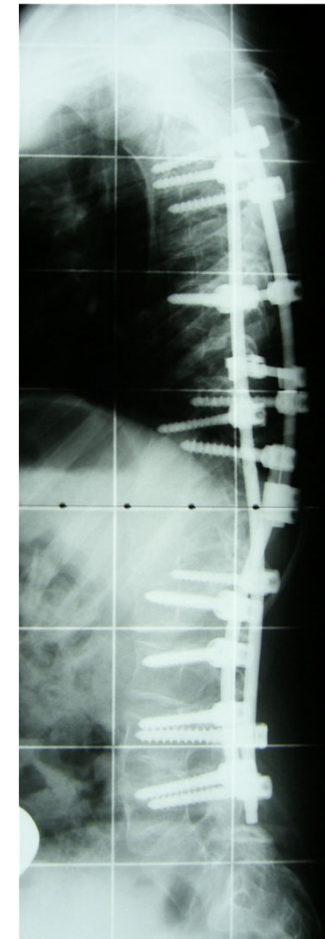
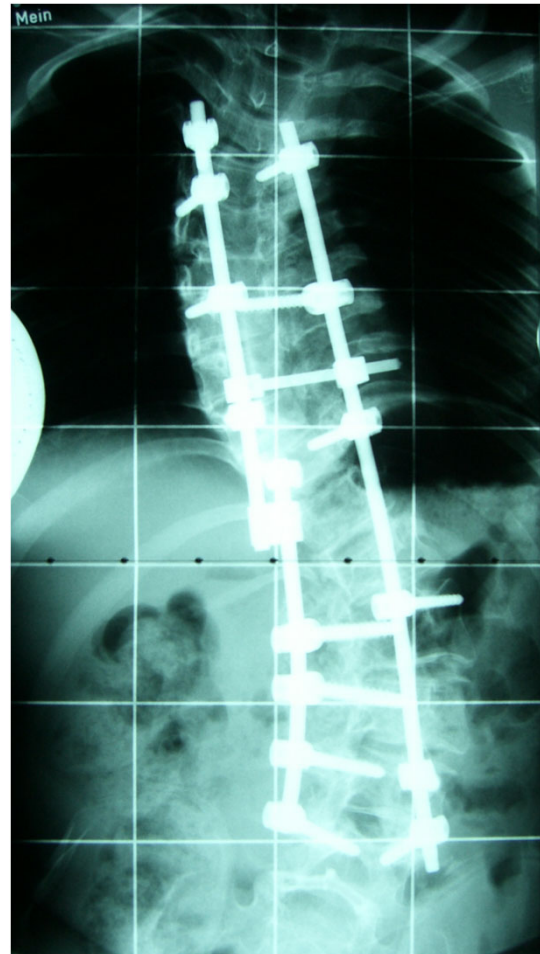
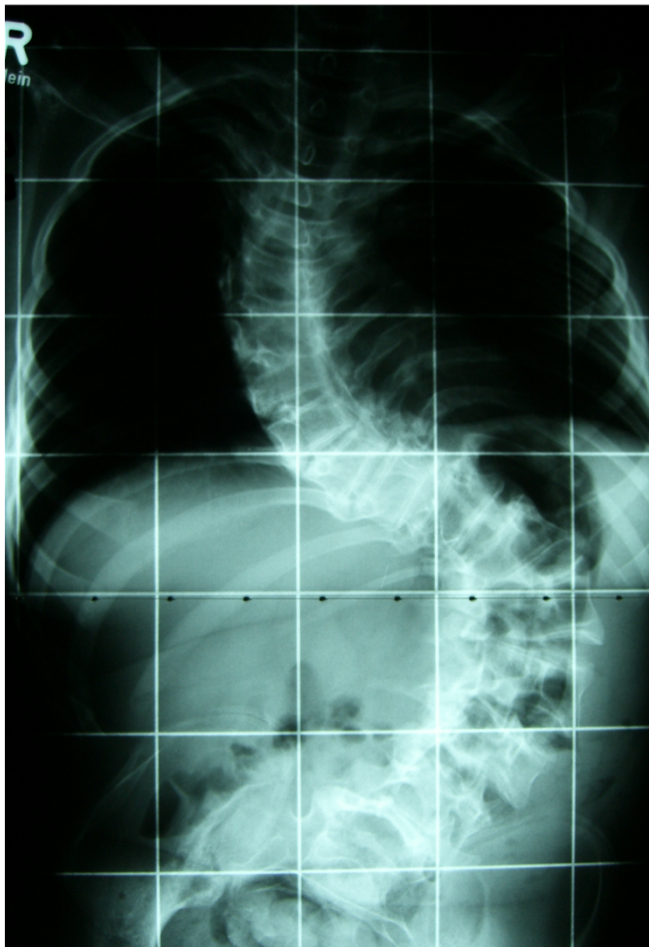
- 121 patients with O.I.
- Braces were not able to prevent progression
- Recommendation: all deformities  $> 50^\circ$  should be fused



Altonaer  
Kinderkrankenhaus

Ein Unternehmen des UKE

# 16 year old girl with O.I. type 1





- Few studies available
- It is not generally effective for treatment of short stature in skeletal dysplasias
- Patients with achondroplasia have normal GH production
- Some studies show increased growth in achondroplasia and hypochondroplasia with improved body proportions
  - The higher the dose and the younger the patient at onset of treatment, the better the achieved growth.



Altonaer  
Kinderkrankenhaus

Ein Unternehmen des UKE





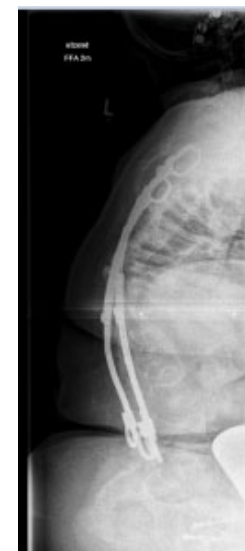
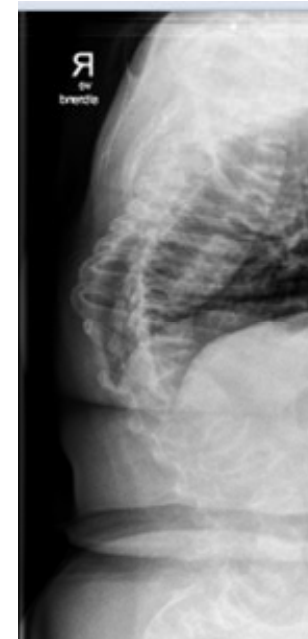
Note: correction of apical deformity by sublaminar bands. No possibility for implants at convexity



Altonaer  
Kinderkrankenhaus

Ein Unternehmen des UKE

# 7 year old boy with O.I. type 3 and progressive deformity





- VEPTR: n=4
  - 2 went to final fusion
  - 2 still expanding (every 9 months)
- Spine fusion: n=17
- 8 type 1
- 7 type 3 and 4



Altonaer  
Kinderkrankenhaus

Ein Unternehmen des UKE

# Kayi, Pinar 13 J. spond- metaph-Dys

