



Altonaer
Kinderkrankenhaus

Ein Unternehmen des UKE

Skeletal dysplasias – Hamburg experience



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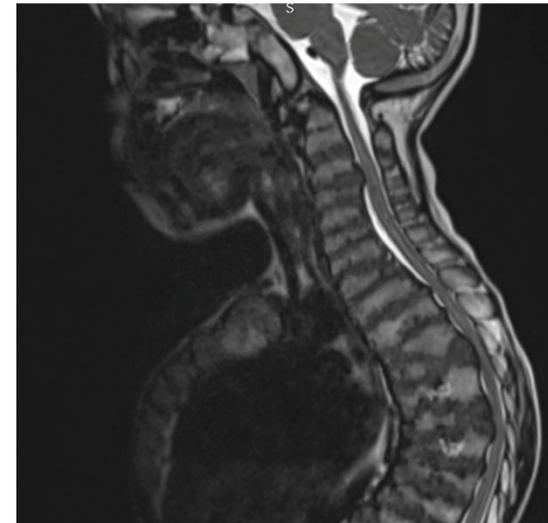
Pediatric Spine Center

The Childrens Hospital Hamburg Altona
University Clinic Hamburg

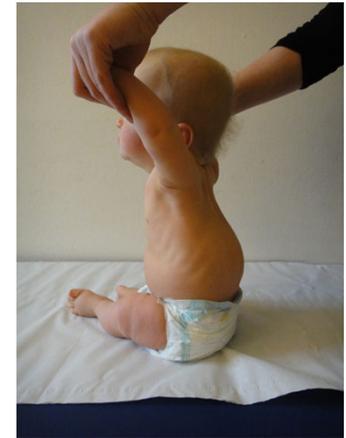
- Travel expenses and speaker fees
 - Nuvasive
 - Depuy / Synthes

- 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

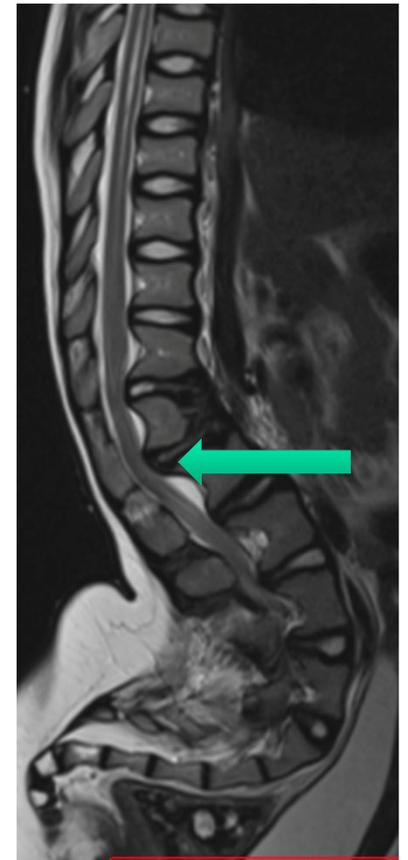
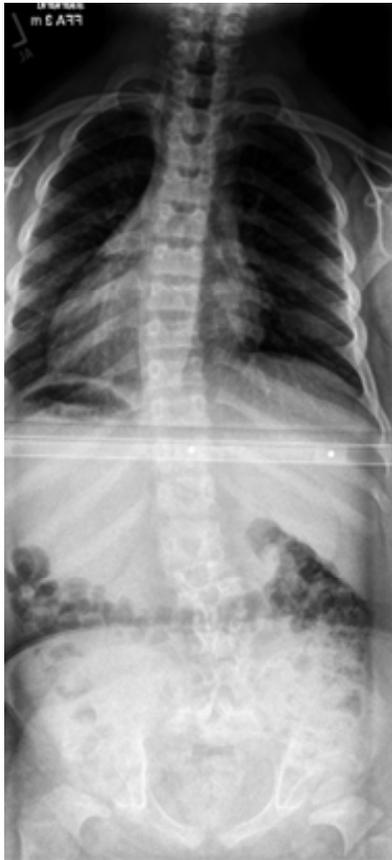




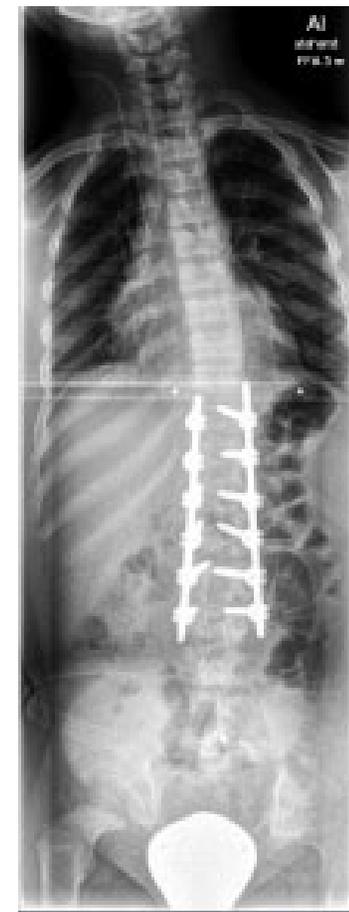
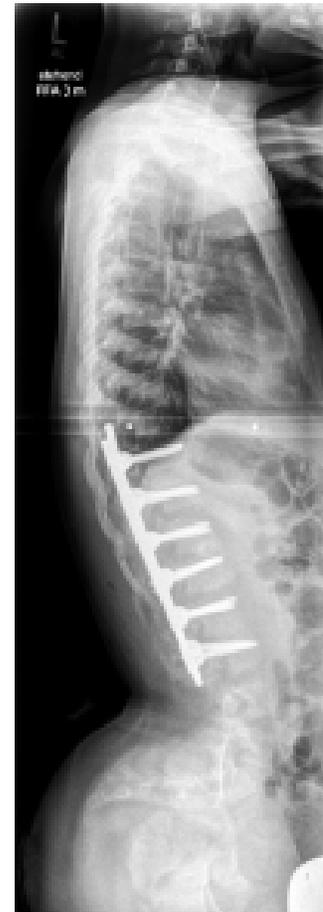
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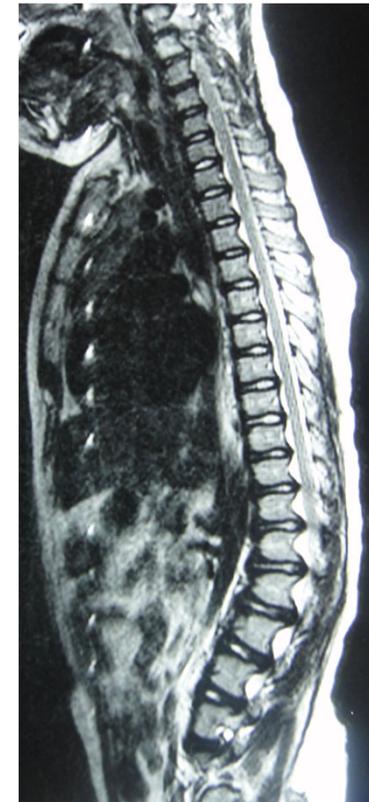
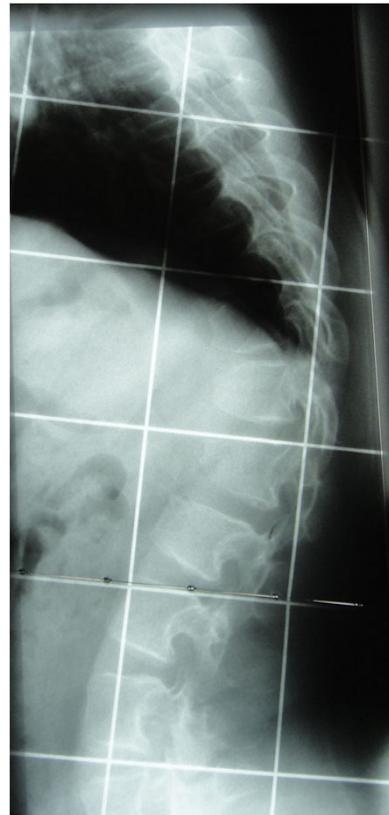
6 year old male patient with ACH, progressive kyphosis and claudication



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

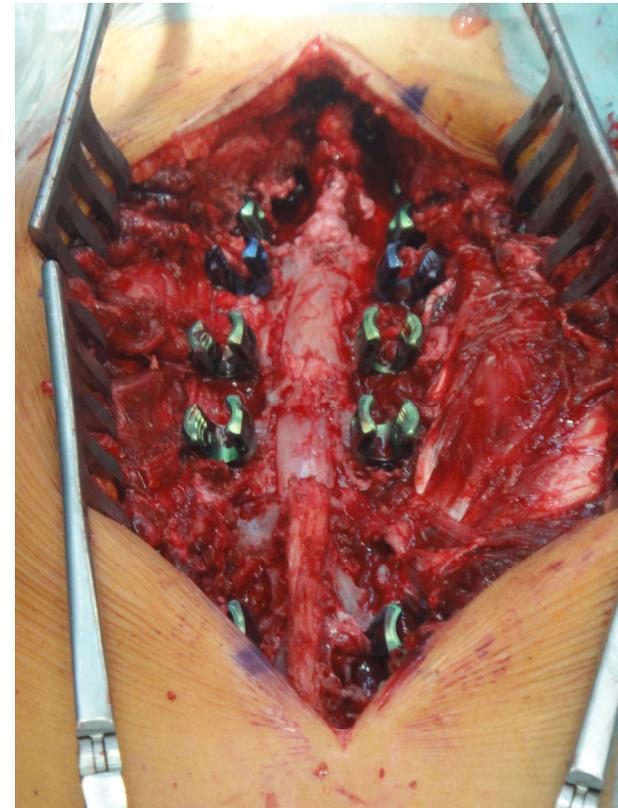
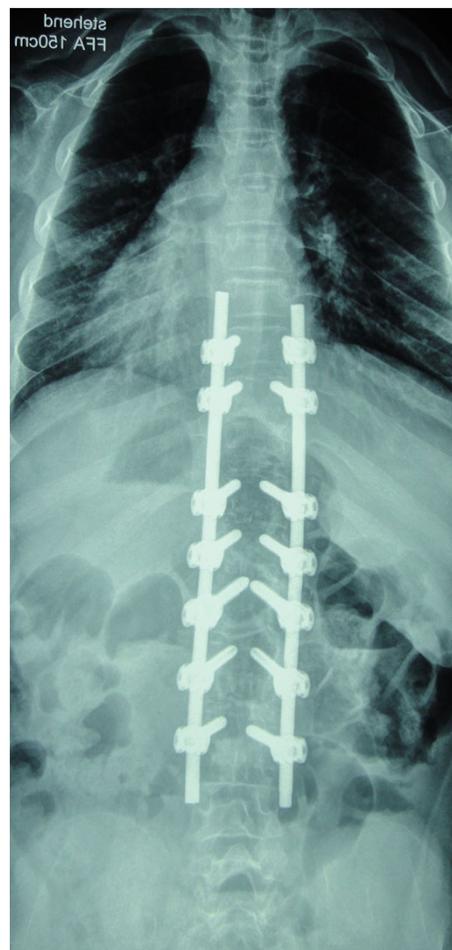
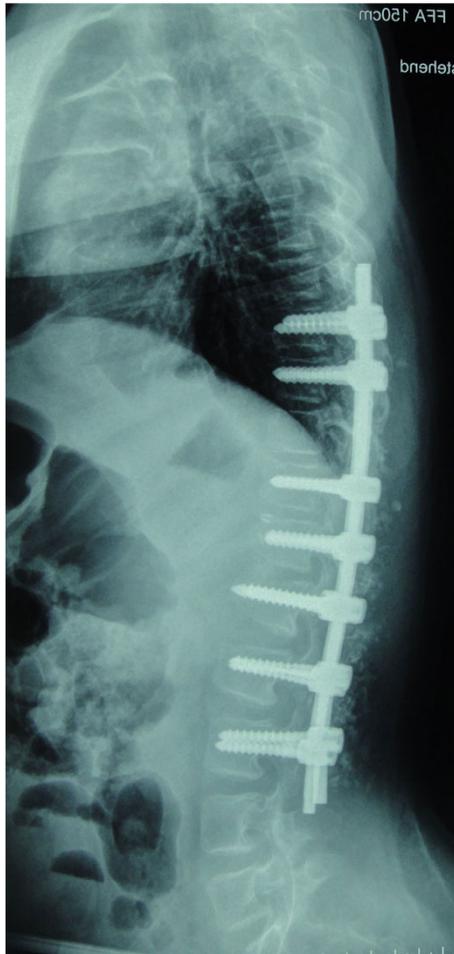


8 year old boy with Achondroplasia and symptoms of spinal canal stenosis

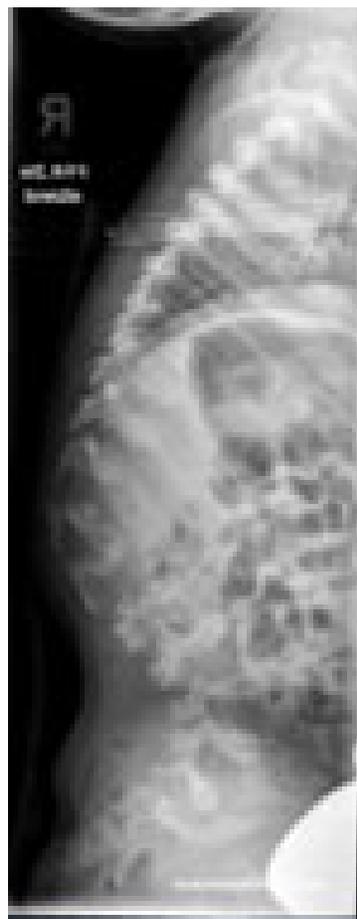


2 reasons for anterior surgery: remove bulging disks, facilitate fusion

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



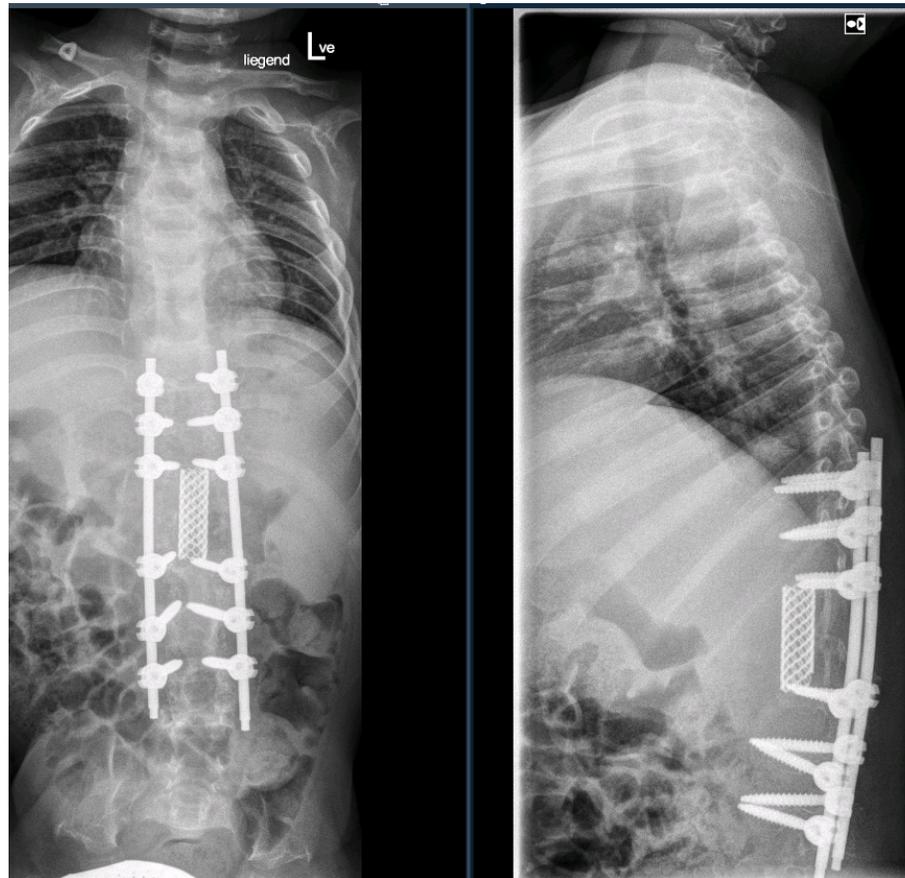
3+4 years old boy with undefined skeletal dysplasia and incomplete paralysis of lower extremities



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



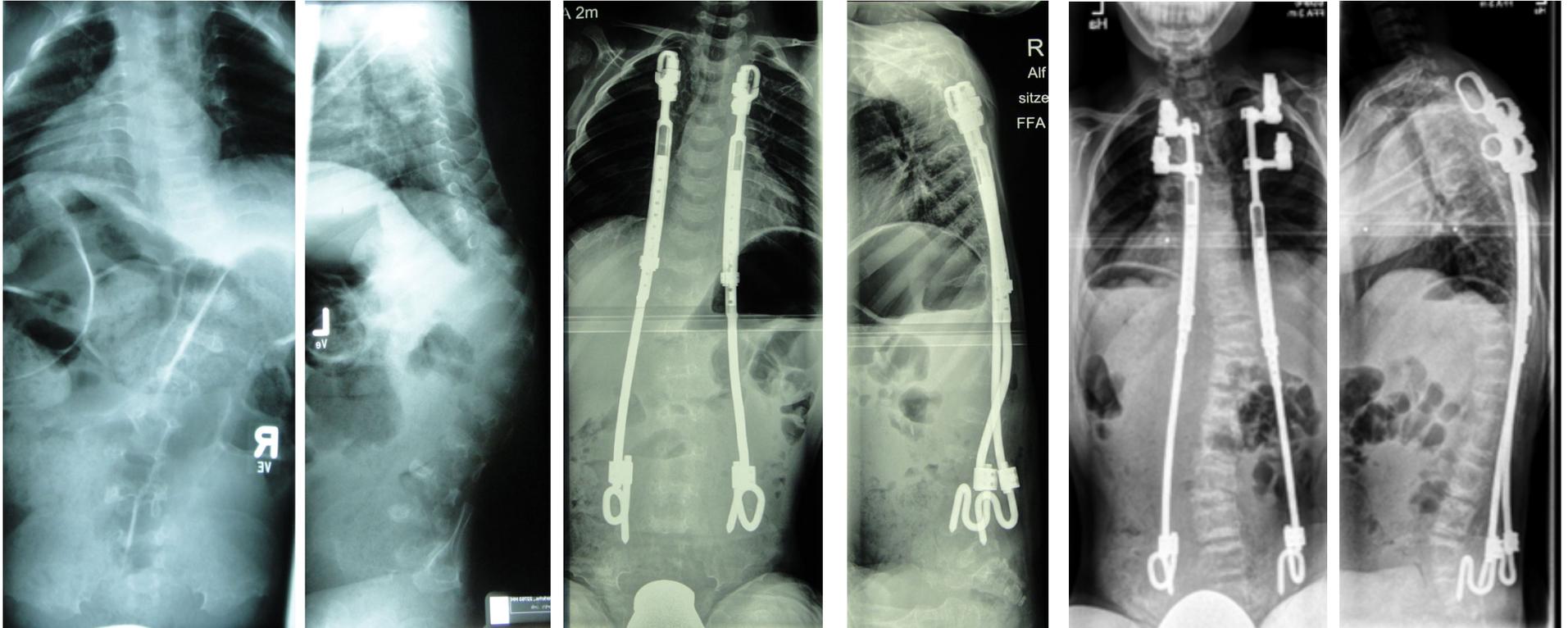
- Progressive scoliosis
- Thoracic kyphosis at young age
- Pulmonary issues



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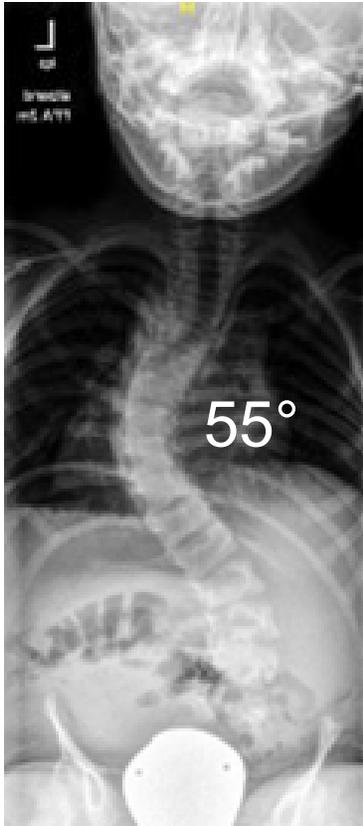
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5 year old male patient with spondylo-
metaphyseal dysplasia, mental retardation,
not ambulant



postop

6 years f/u



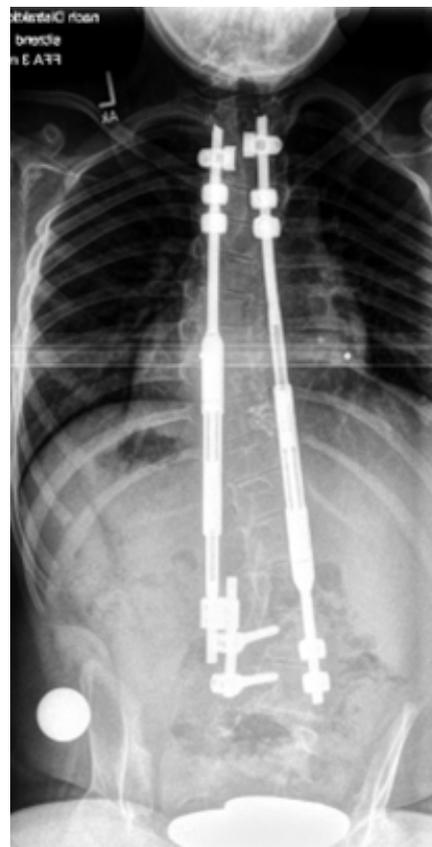
Bending



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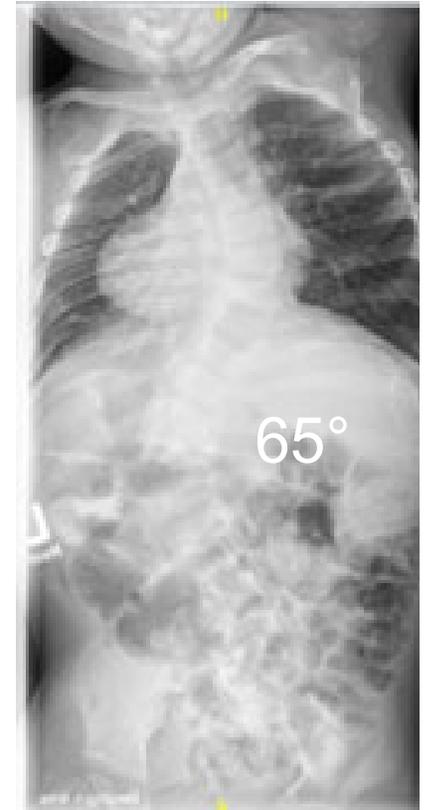
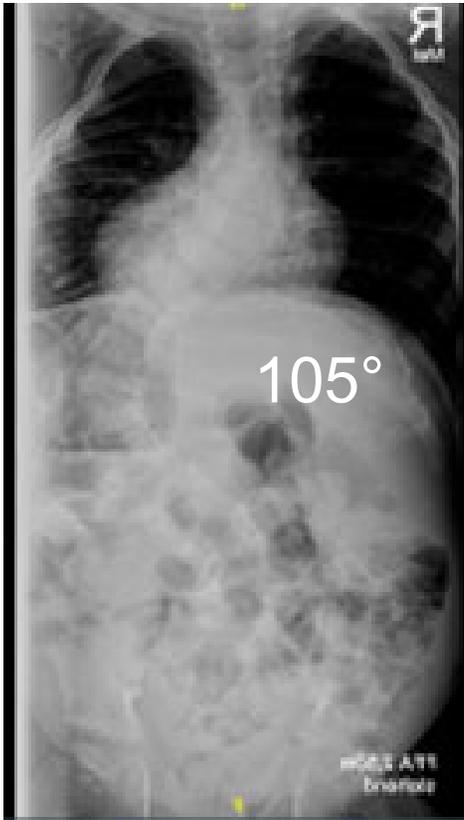
4 years f/u

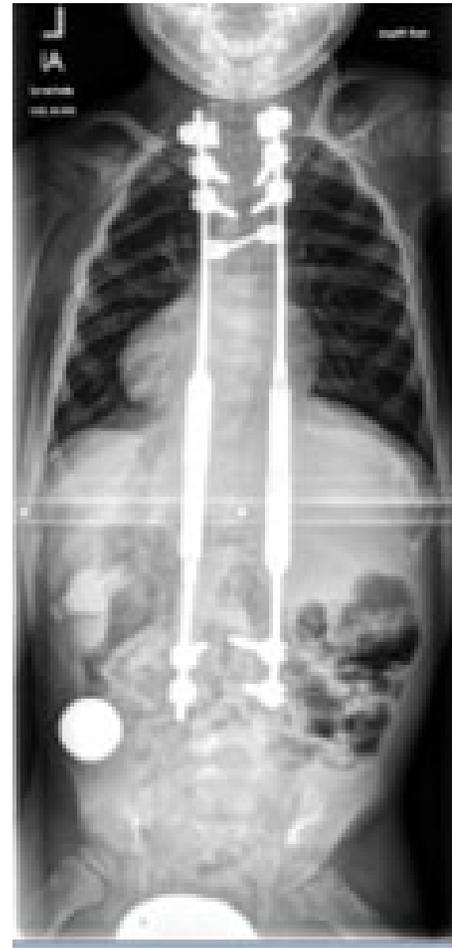
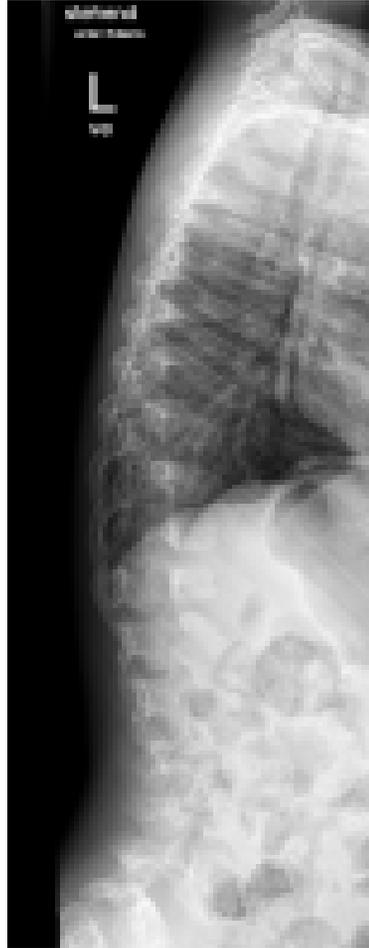


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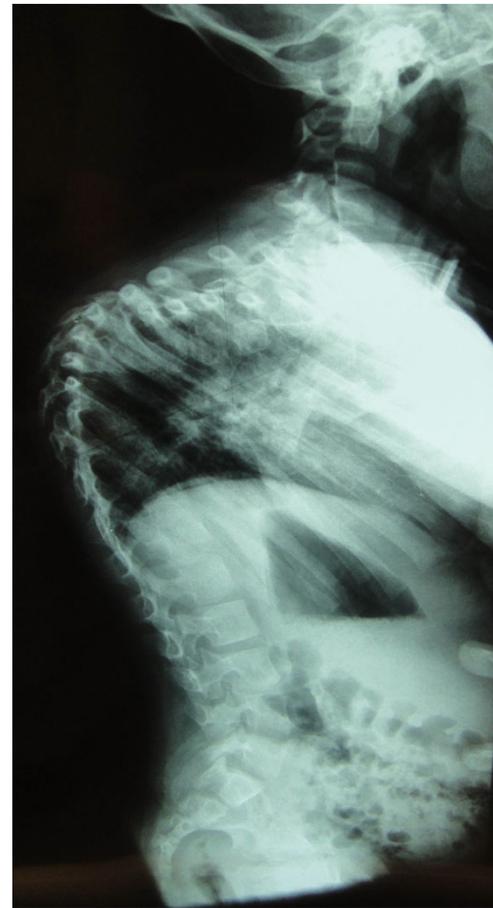
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7 year old patient with SED





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

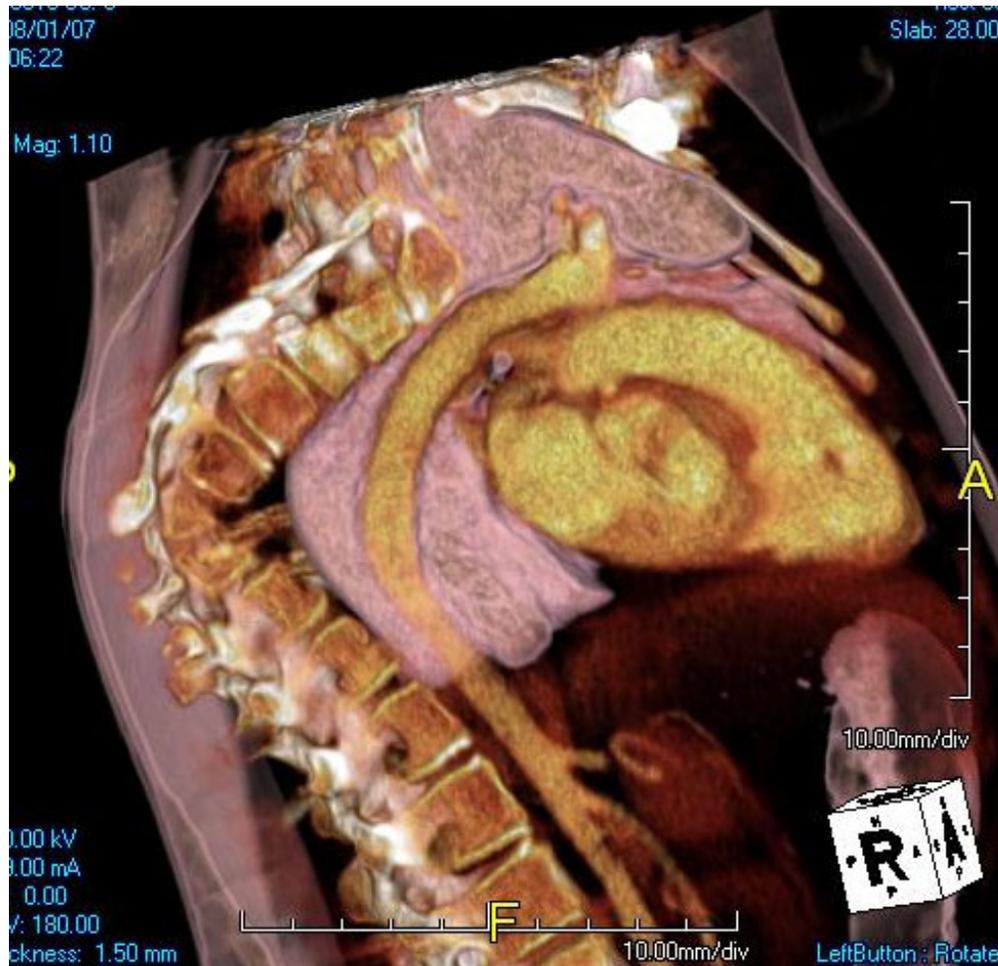




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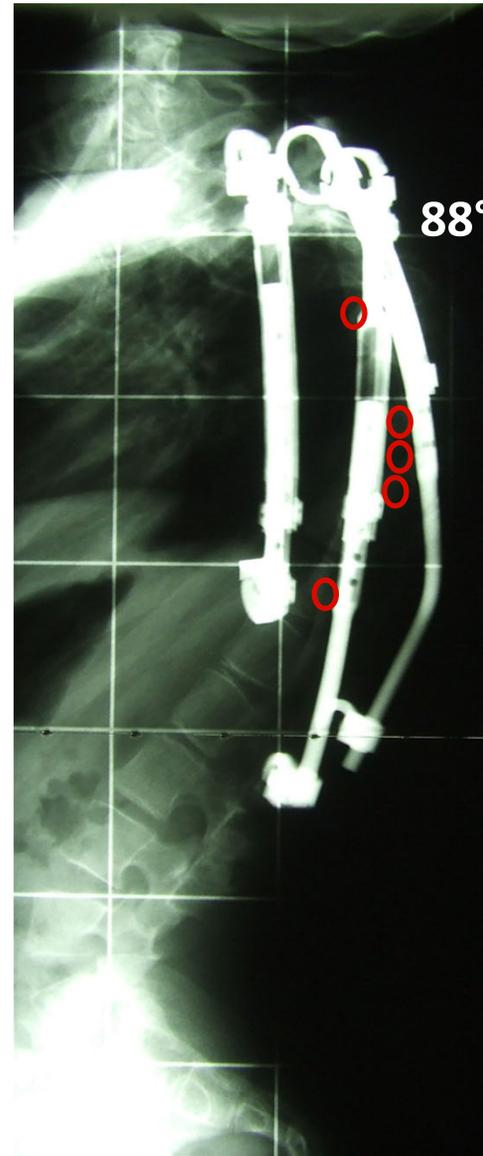
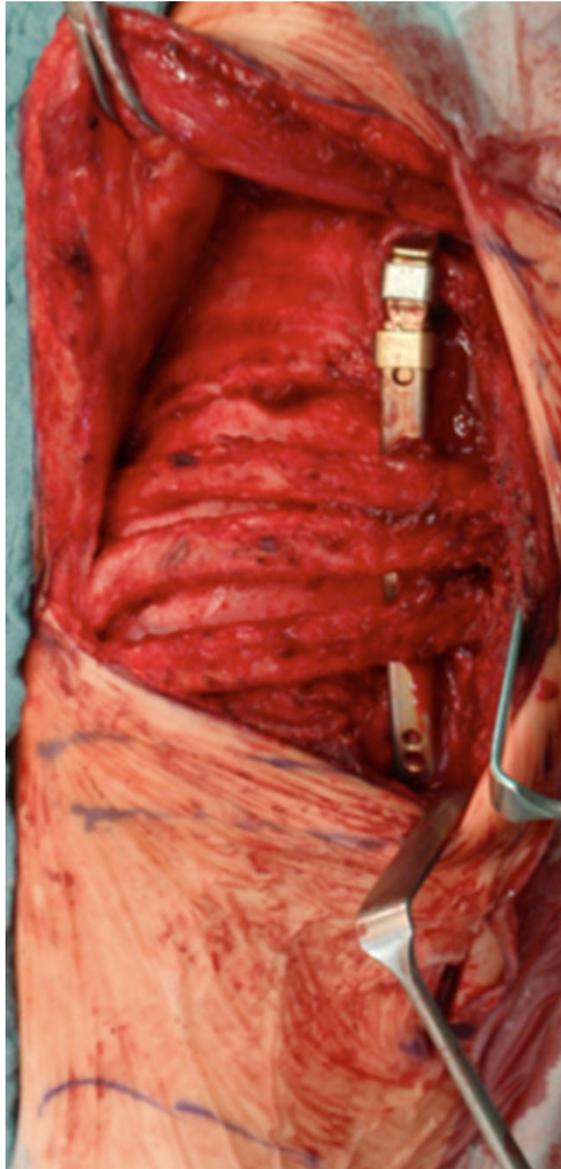
Severe vertebral dysplasia with segments of failure of formation, severe chest deformity





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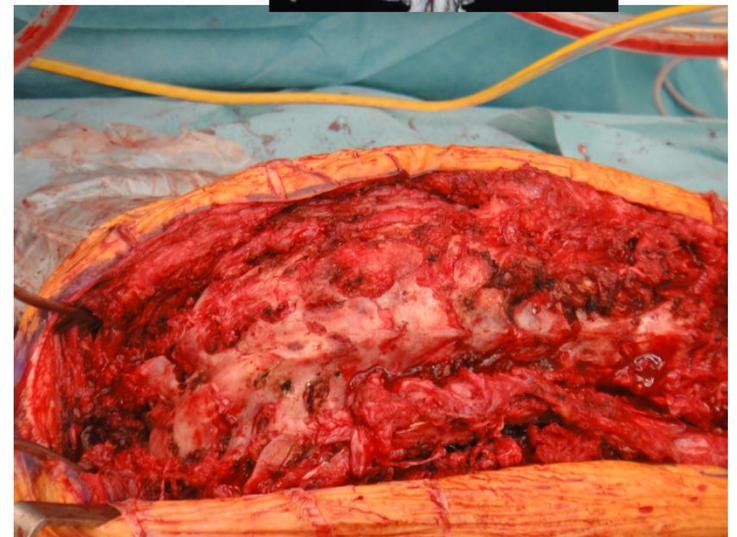
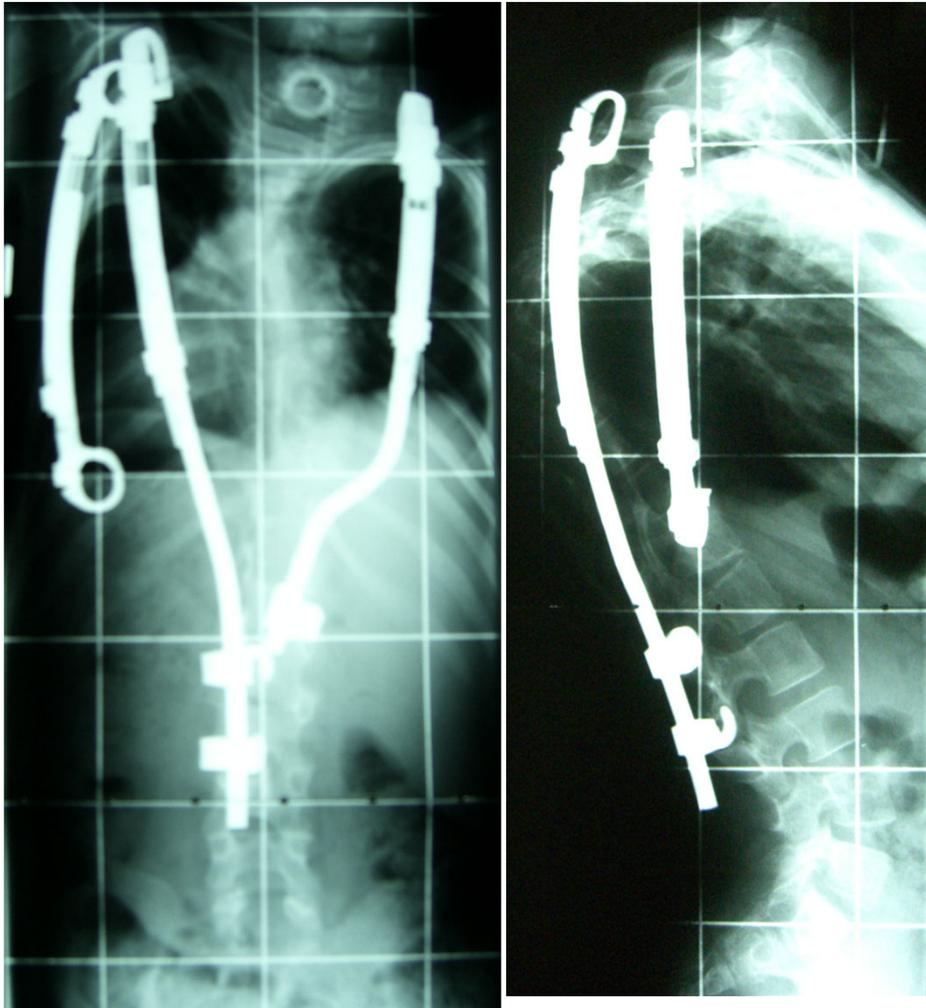




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The situation after 6,5 years
had 3 revisions

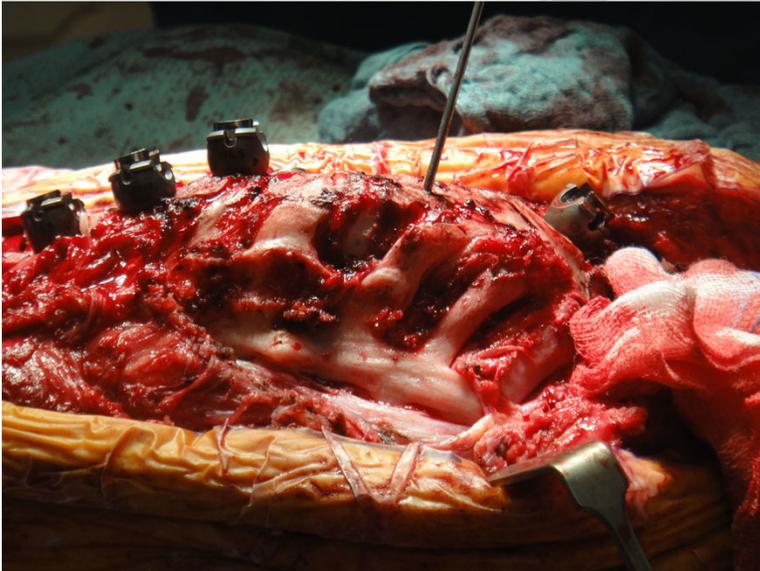




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Final correction with 11 years

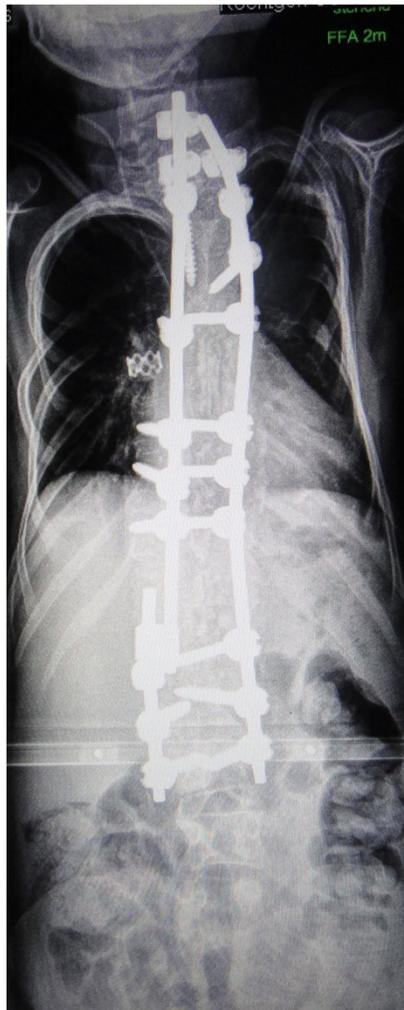




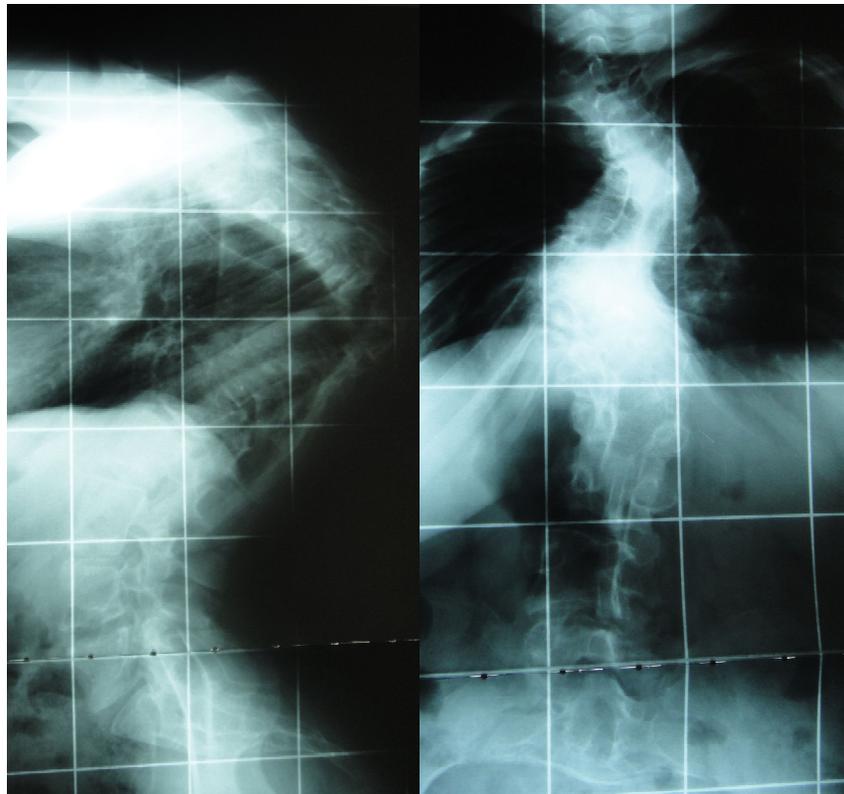
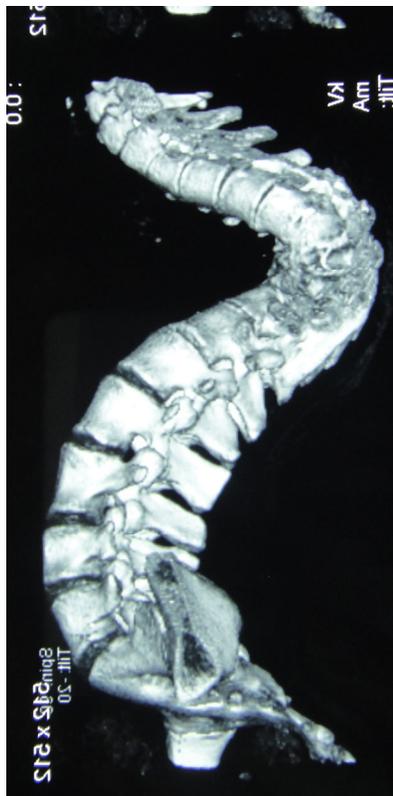
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Final correction by VCR + multiple osteotomies



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

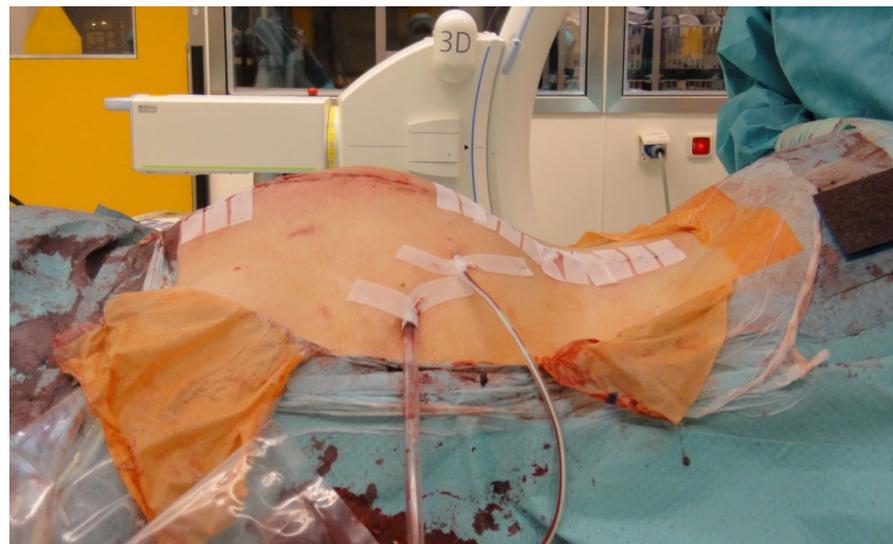




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Correction by VCR and multiple osteotomies

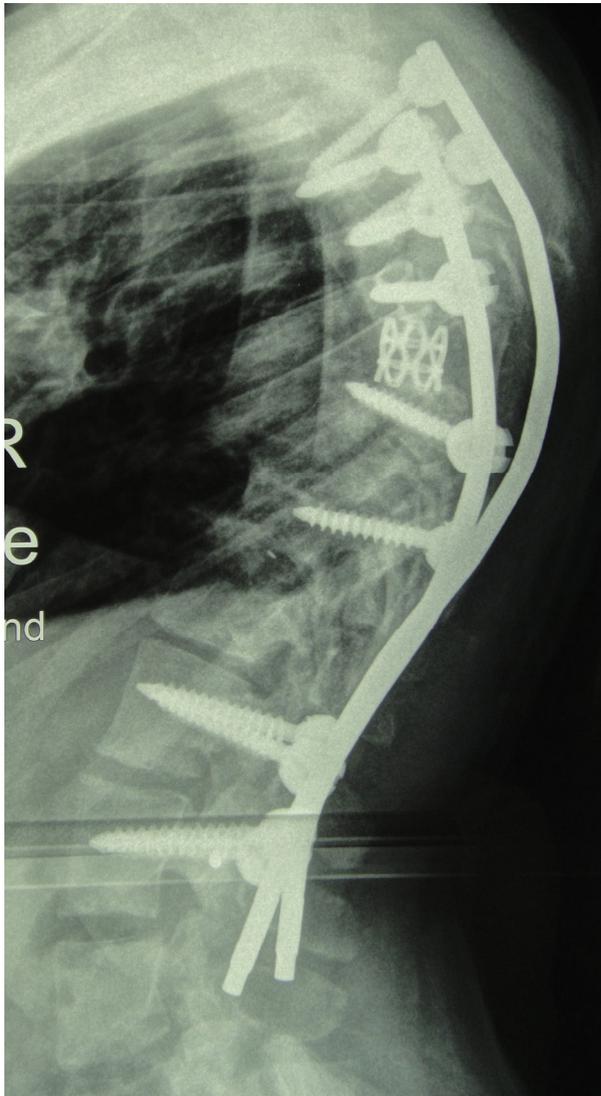


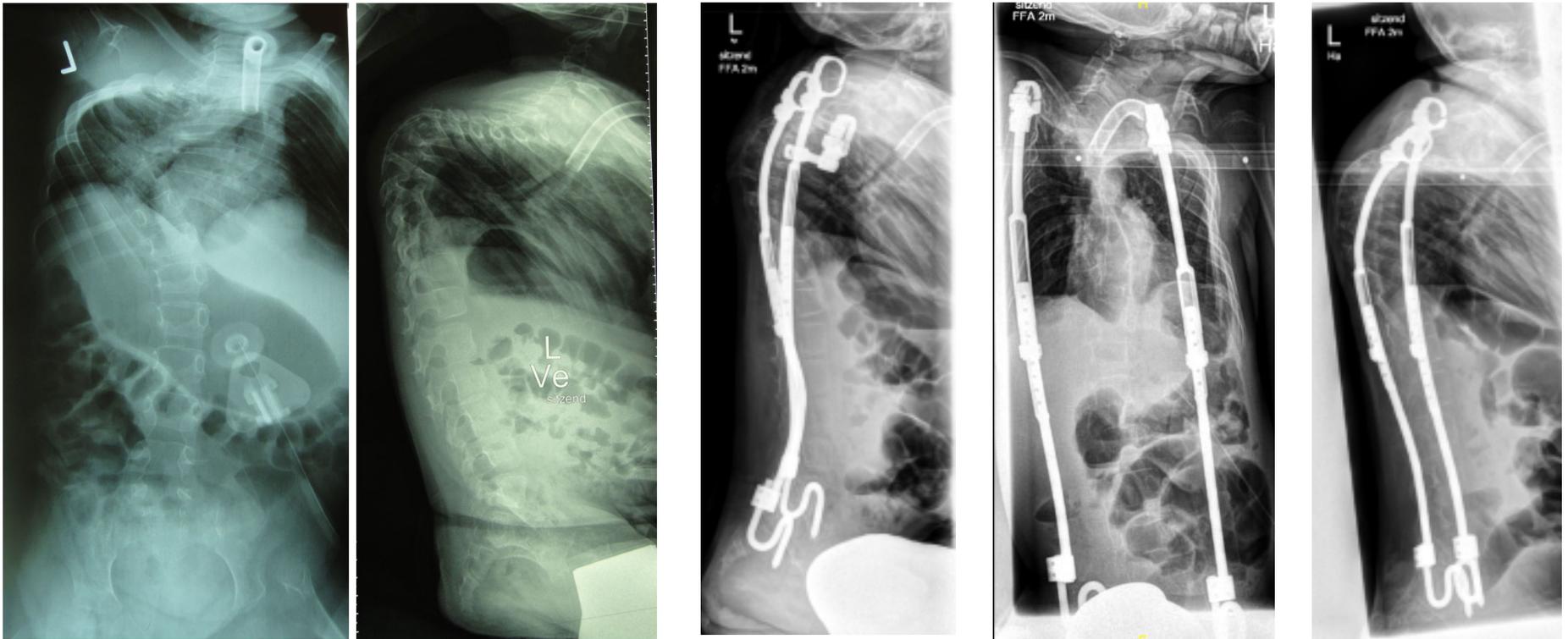


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Was corrected to 40°



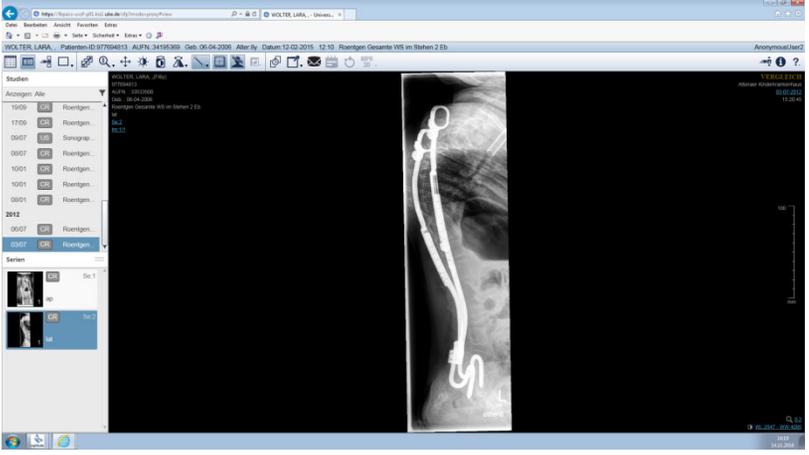
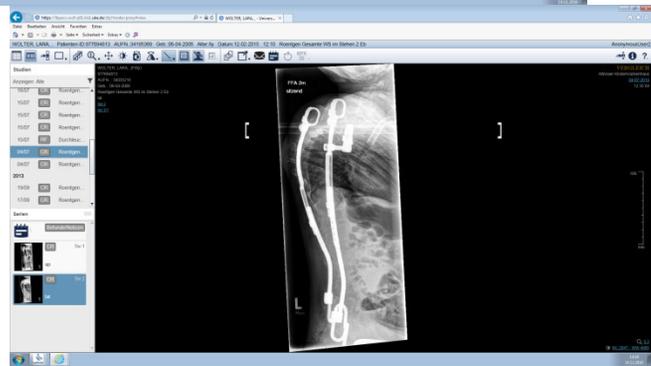
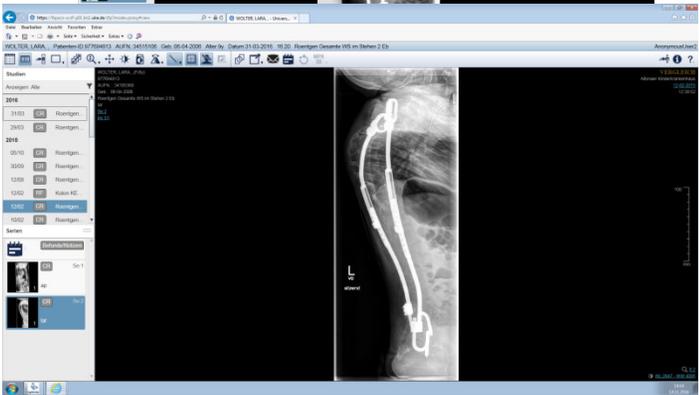
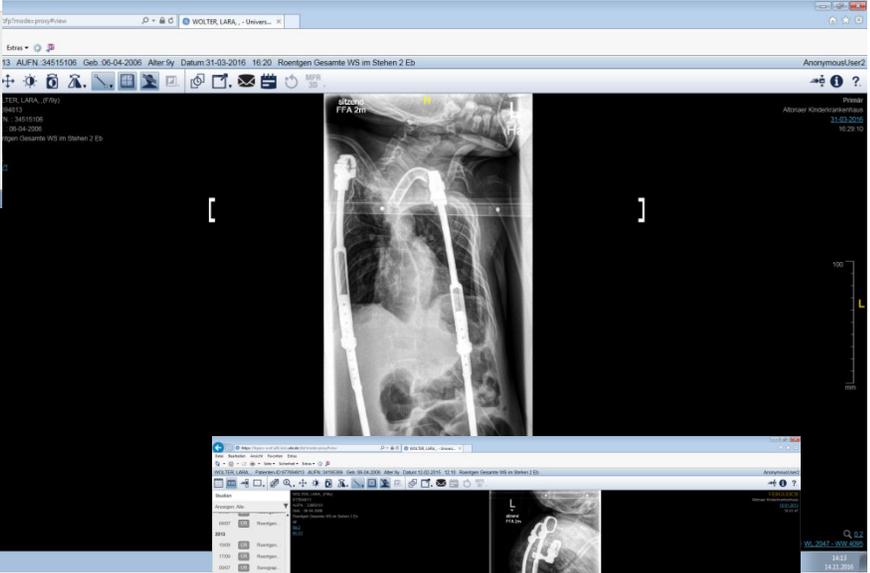


6 year follow up after Halo traction, VEPTR
instrumentation, posterior apical fusion, revision due to
infection and new VEPTR implantation
Kyphosis was well controlled

- 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

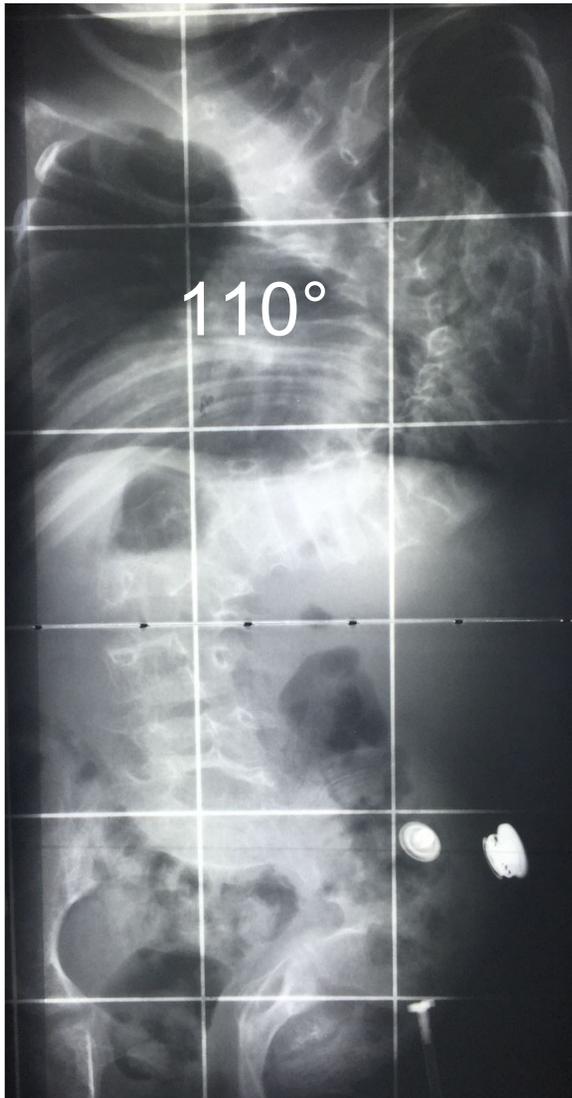


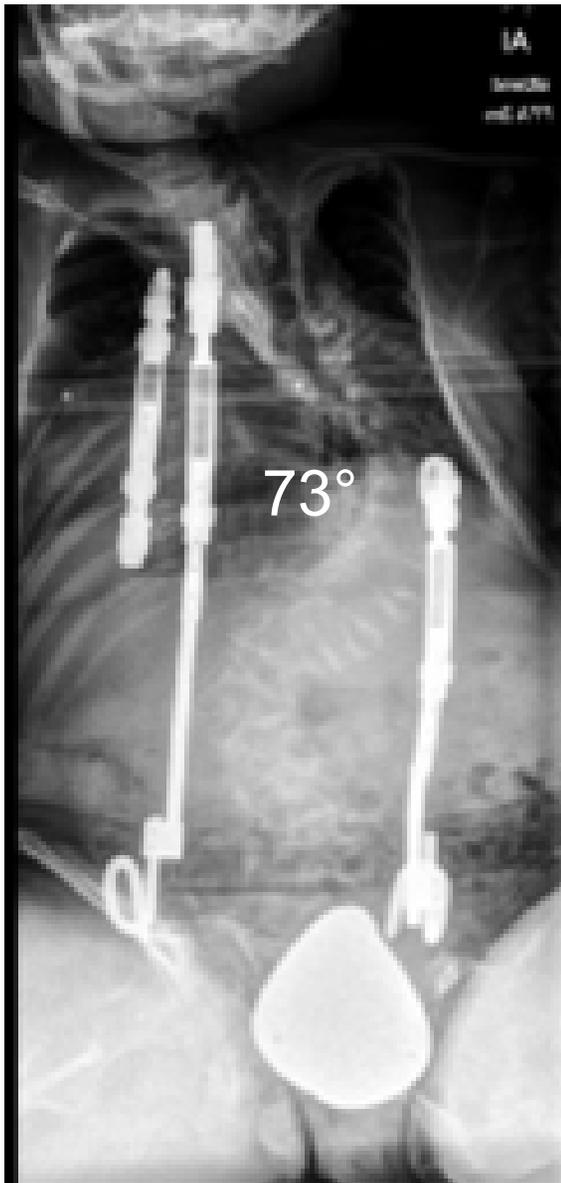


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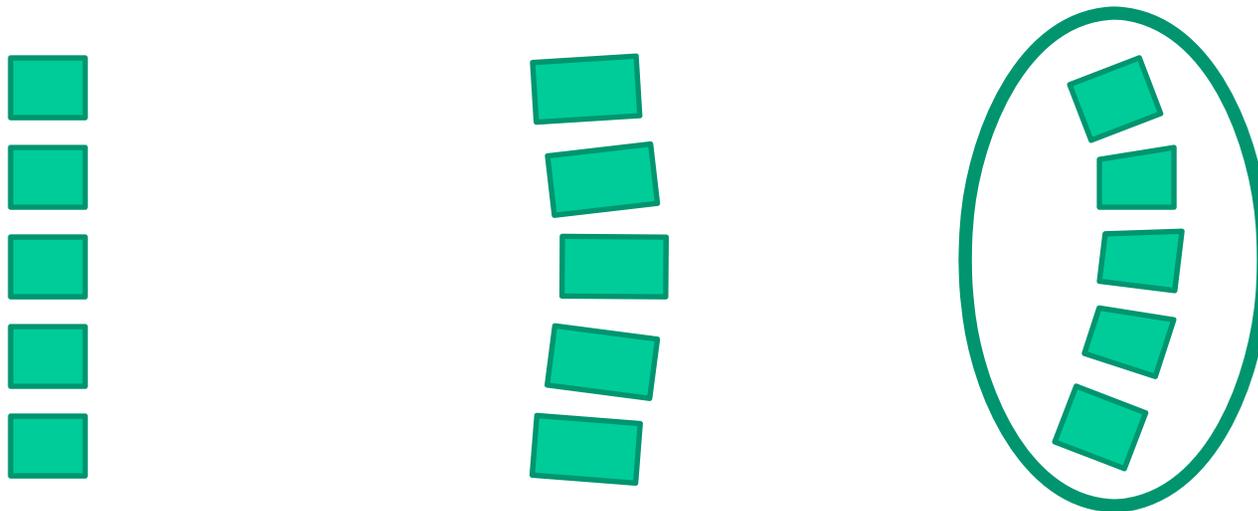
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S.E., O.I. type 4, 8 years

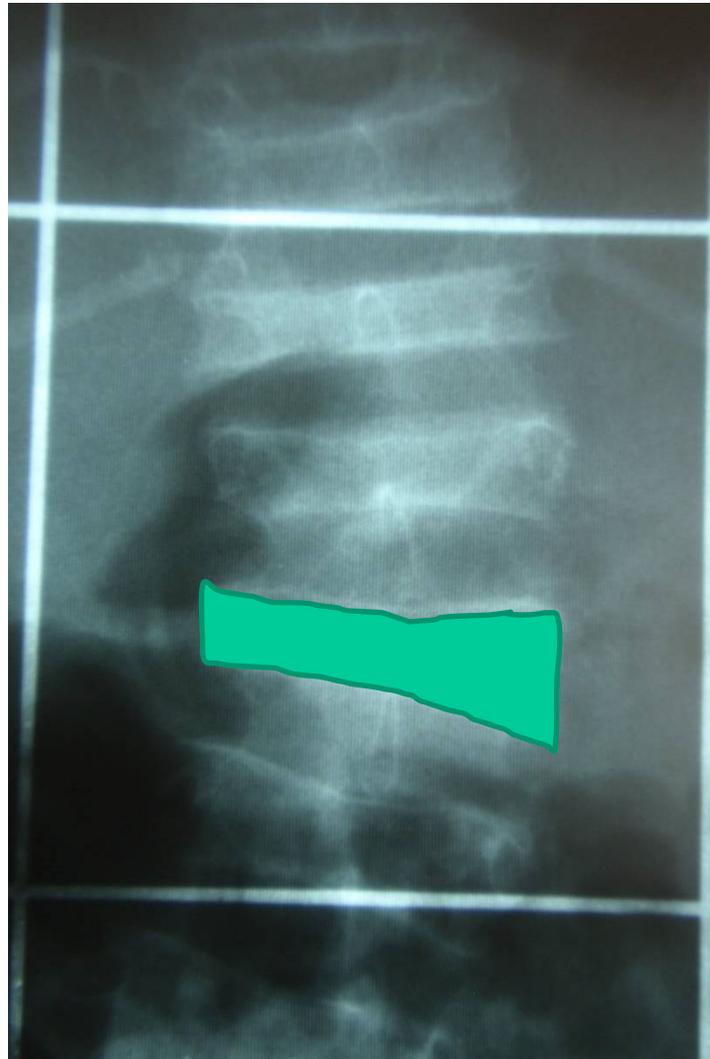




- Incidence: 20-80%
- Incidence increases with severity of disease
- Risk factors: ligamentous laxity and bone mass loss



An important reason for development of scoliosis
is vertebral deformity





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Growth hormone treatment not
effective for

- Metaphyseal chondrodysplasia



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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° / year
 - If bisphosphonates-treatment begins before age 6, prevalence decreases to $3,8^{\circ}$ / year
- Type IV: 54% prevalence, progression rate 4° / year
- Type 1: 39 % prevalence, progression rate 1° / 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:
 - 8,2 % at 7,5 years
- Spondylolisthesis:
 - 10,9 % at 6,5 years

Prevalence of spondylolysis and
spondylolisthesis about 20%





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Chest wall deformities in O.I

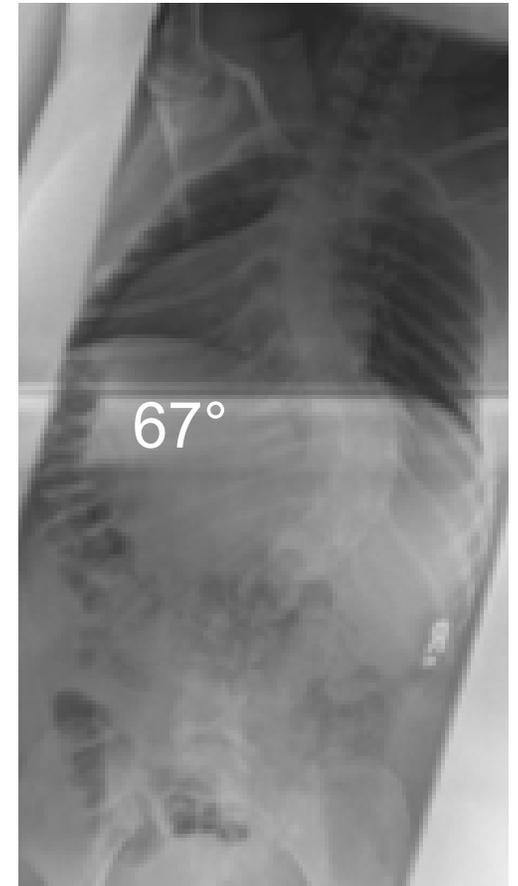
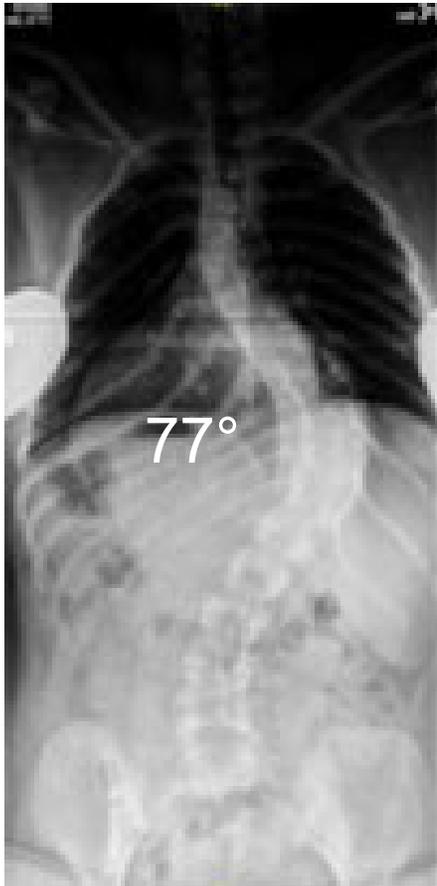


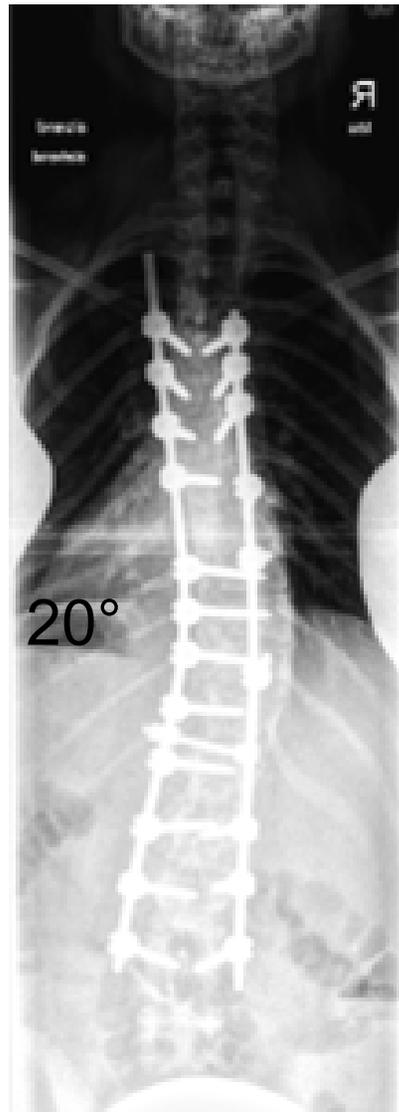


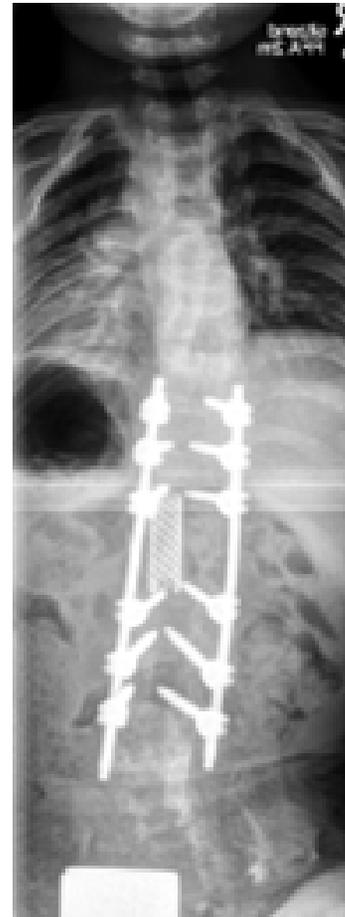
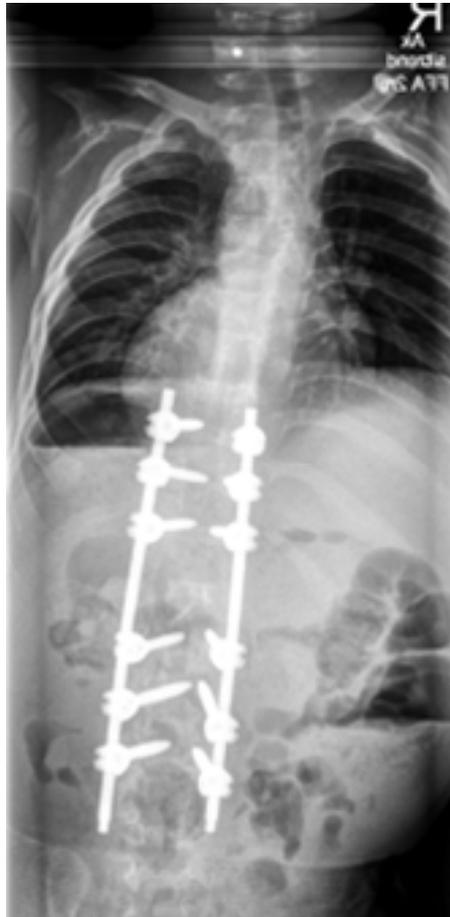
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17 year old girl with O.I., type 1



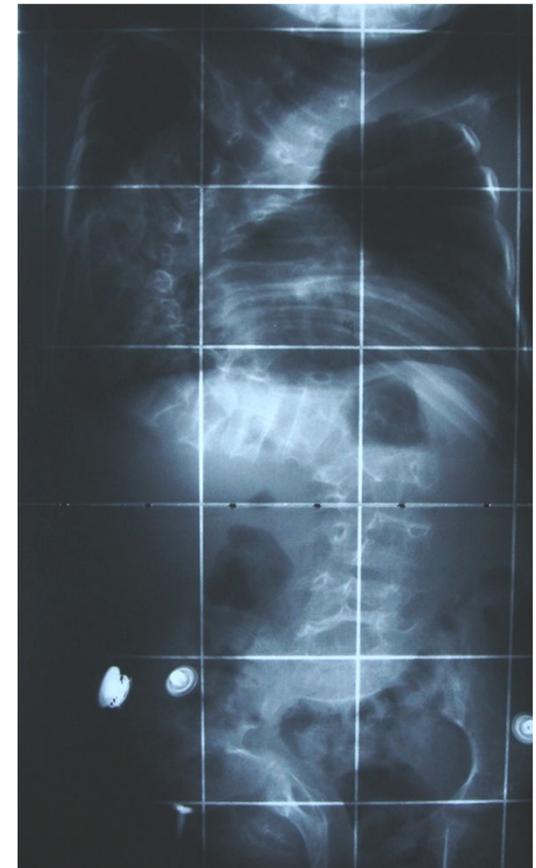






- Pulmonary failure leading cause of death in adults
- Vital capacity $< 50\%$ if thoracic curve $> 60^\circ$
- Early development of motor milestones inversely related to severity of scoliosis

- 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

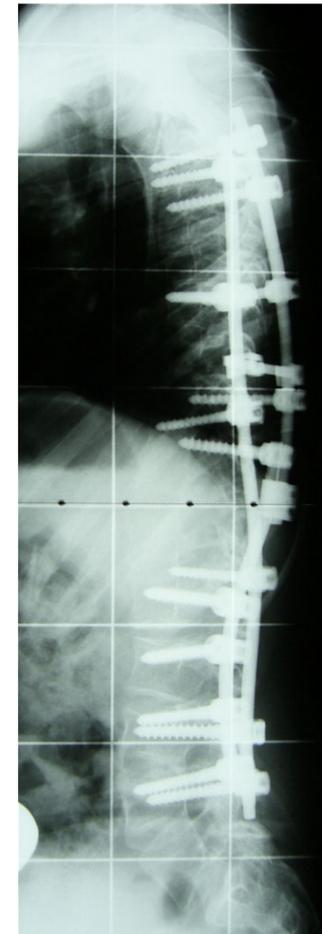
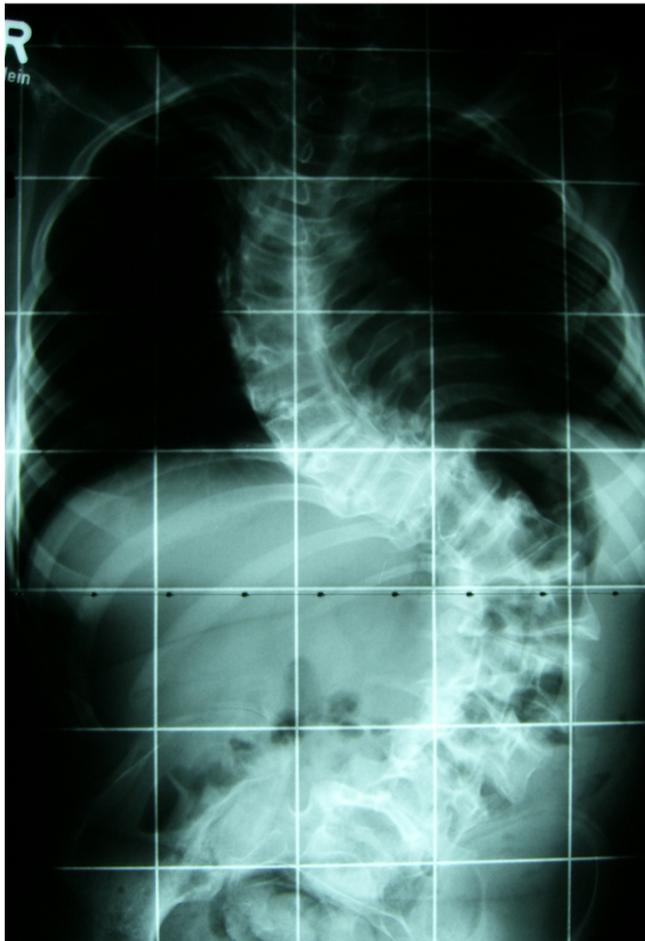
- 121 patients with O.I.
- Braces were not able to prevent progression
- Recommendation: all deformities $> 50^\circ$ should be fused



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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.



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



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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



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Kayi, Pinar 13 J. spond- metaph-Dys

