

## Congenital deformity of the spine-classification, diagnosis and therapy

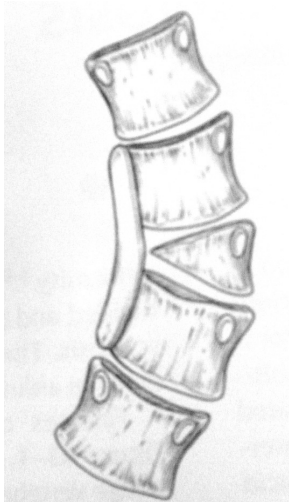
Author: Univ. Doz. Dr. Werner Lack

### Classification of congenital scoliosis

failure of formation-wedge vertebrae (2 pedicles), half vertebrae...  
     unsegmented half vertebra: fused to the vertebral body above and below  
     partially segmented half vertebra  
     fully segmented half vertebra-separated above and below by disc space  
 failure of segmentation-block vertebrae, unilateral bar...  
 combination

### Special combinations

hemimetameric shift-hemivertebra counterbalanced by another one on the contralateral side  
     in the same region  
 unilateral bar and contralateral hemivertebra-"worst case scenario", progression per year up  
     to 10° Cobb!



Picture: worst case scenario: unilateral bar and contralateral half-vertebra

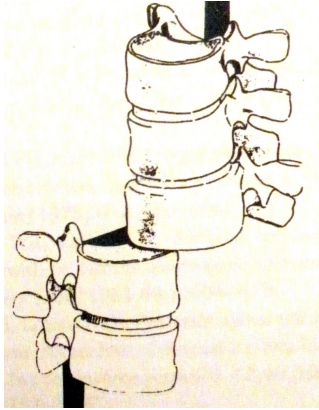
### Severe progression

hemivertebra thoracolumbar 2-3,5°/year  
 2 hemivertebrae 5°  
 unilateral unsegmented bar 6-9°  
 unilateral unsegmented bar with contralateral hemivertebra >10°  
 most severe progression in thoracolumbar region!  
 most severe progression up to 5th year and in adolescence/puberty

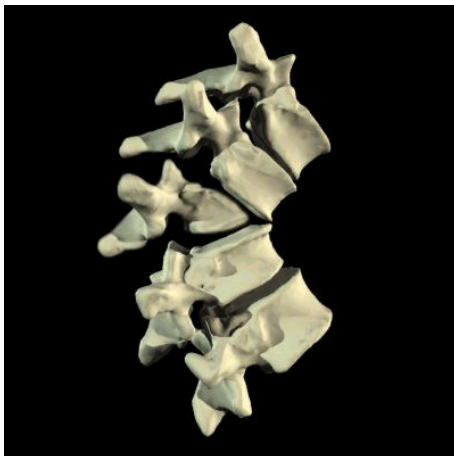
### Classification of congenital kyphosis (Mc Master)

failure of formation  
     posterolateral quadrant hemivertebra  
     butterfly vertebra  
     posterior hemivertebra  
     wedge vertebra  
 failure of segmentation  
     anterior unsegmented bar  
     block vertebra  
 mixed anomalies  
 unclassifiable anomalies

### **Congenital vertebral displacement-sudden sagittal kyphotic displacement, most severe of congenital kyphosis**



Picture: there exist no known genetic abnormalities associated with the development of congenital kyphosis



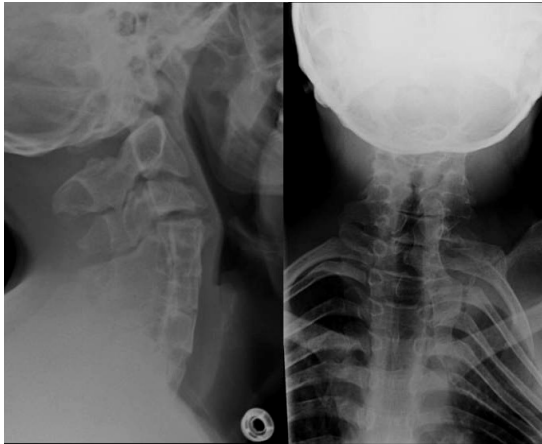
Picture: posterior half vertebra

### **Klippel-Feil-syndrome**

- segmentation defects of the cervical spine
- plagiocephaly
- reduced cervical spine mobility
- anomalies of urinary system in one third
- cong. Heart anomalies in 15%
- Sprengel-deformity in 20-30%
- problems in hearing in one third

### **Sprengel deformity**

- congenital failure of descendance of one or both shoulders; mostly left shoulder elevated
- „omovertebral bone“ (connection scapula-C5 or C6)
- often Klippel-Feil, rib anomalies...
- 75% girls
- scapula small and rotated



Picture: Klippel-Feil with Sprengel deformity

### **Congenital sacral problems**

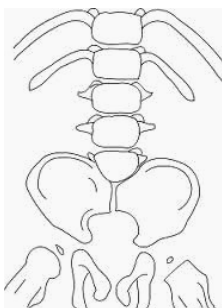
lumbosacral abnormalities (partial, total, asymmetric, symmetric)  
 sacral obliquity  
 sacral dysplasias up to sacral agenesis  
 sacral dysraphism (MMC)

### **Congenital sacral obliquity**

angular deformation of the sacrum from a horizontal line drawn parallel from a line across the femoral heads from congenital origin!  
 it is not a pelvic obliquity!  
 mostly sacral endplate elevated at the right side  
 max. 20°, but can cause lumbar scoliosis up to 50°-effect like a lumbar hemivertebra

### **Congenital sacral agenesis (Renshaw 1978)**

- 1) unilateral agenesis, partial or total
- 2) symmetrical partial with stable articulation iliac bone-S1
- 3) variable lumbar and total sacral agenesis with articulation iliac bone-lowest lumbar vertebra
- 4) variable lumbar and total sacral agenesis with either fused iliac bones or amphiarthrosis



### **Symptoms of severe forms**

often maternal diabetes  
 in severe cases bowel and bladder dysfunction  
 deformities of hips (flexion, abduction, outward rotation), knees (flexion contracture), feet, atrophy of the legs, motoric palsy („Buddha-like-position“)

### **Congenital rib deformities**

mostly in combination with congenital scoliosis, rarely with congenital kyphosis  
 70% had thoracic or thoracolumbar concomitant congenital scoliosis

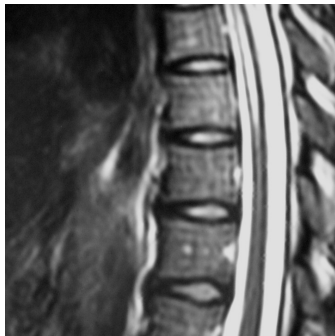
mostly on concave side of unilateral failure of segmentation

40% with Sprengel deformity

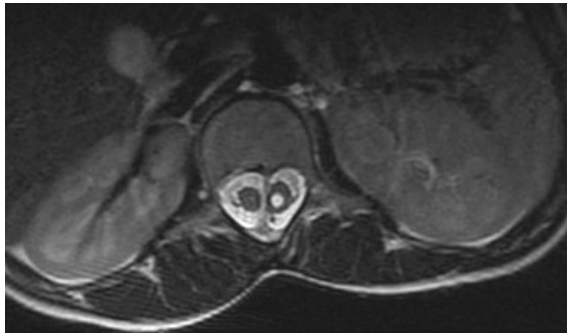
prognosis without difference to scoliosis without rib fusions

### Associated anomalies

syringomyelia (abnormal fluid collection in the medullary canal of the spine, caused by Arnold-Chiari-syndrome, basilar invagination, cord compression, trauma, arachnoiditis, can cause scoliosis, kyphosis, motor weakness, dyscoordination, neuropathic arthropathy, pain)



Picture: Chiari malformation (caudal dislocation of cerebellar tonsils below the foramen magnum), diastematomyelia, diploemyelia-fibrous ligament or osseous bar, mostly in lumbar spine



Picture: diastematomyelia

tethered cord (filum terminale pulls cord down to L4)...MRI brain stem and complete **spine**; release always before spine surgery, better not at the same operation; seldom spontaneous release of symptoms in scoliosis >40°

congenital heart disease (25%)-septum defects, hypoplastic left heart, transposition...  
**echocardiogram**

genitourinary anomalies (20%)-renal aplasia, duplicate ureters, hypospadia...**renal ultrasound**

### anamnesis for neurologic infantile symptoms

- is the child toilet trained ?
- bed wetting problem ?
- bowel or bladder „accidents“ ?
- limping ?
- shoe size difference ?

### clinical evaluation

- spinal dysraphism ?
- asymmetric calves, cavus feet, clubfeet, vertical tali ?
- truncal or pelvic imbalance ?

spinal balance frontal and sagittal

rib cage deformity ?  
in-and expiration capacity  
neurologic deficit ?  
standing and sitting size  
rib hump  
asymmetry of lumbar height  
height of shoulders  
height of pelvic rim  
vertical centre line

#### **radiologic parameters ap**

Cobb - angle (main-curve, secondary - curve)  
rotation (Nash and Moe 0 - 4)  
deviation of perpendicular line  
pelvic inclination  
Risser-sign (0-5)

#### **radiologic parameters sagittal**

evaluation of perpendicular line S1  
evaluation of L4- (should be horizontal)  
measurements

Cobb-angle thoracic kyphosis  
Cobb-angle lumbar lordosis  
Cobb-angle Th11-L1 (should be 0°)

#### **CT in congenital deformities**

bony details  
good details of apical hemivertebra (for hemivertebra excision)  
myelo-CT for complex dysraphic problems  
CT-models for complex deformities

#### **MRI in congenital deformities**

for detection of neural axis abnormalities  
necessary MRI from brain stem to sacrum  
evaluation of disks and growth potential  
evaluation of kidneys and lungs...

#### **Lung function**

„of all spinal deformity patients having early death due to cor pulmonale , congenital scoliosis patients probably have the highest frequency“ (Winter 1983)  
lung function analysis (total capacity, vital capacity, Tiffeneau-test...)  
blood gas analysis

#### **Conservative therapy**

in clear cases of progression (2 hemivertebrae, unilateral bar etc no waiting !  
watching and control  
brace therapy only in cases of compensatory curves, very infrequent indicated!

#### **Operative therapy-general rules**

better a short and straight than a long and curved spine  
at optimal time in most cases small operations are sufficient; in late cases frequently long operations with high complication risk are necessary  
**waiting up to the end of growth in progressive congenital deformities is a severe mistake!**

#### **Preoperative therapy**

in case of severe deformity, especially with severely reduced lung function Halo-extension (Halo-gravity, Halo-wheelchair, Halo-pelvic)

exact neurologic examination during Halo-extension:

**eye muscles** – looking at moving finger

**n. facialis** – closing eyes, showing the teeth

**n. accessorius** – lifting of shoulders

**n. hypoglossus** – showing the tongue, speech!

motor and sensoric testing of upper and lower extremities

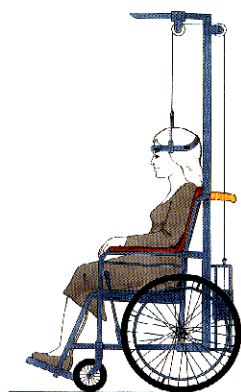
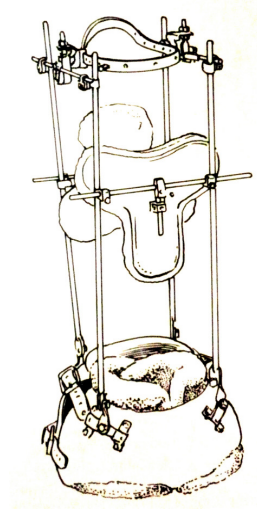
**pyramid signs** - Babinsky, abdominal reflex

### Severe complications in Halo-Pelvic-extension

peritonitis by perforation of iliac screws

cervikal problems >50%, degenerationen of cervikal spine, avascular necrosis of dens,

spontaneous fusion (Dove et al. 1980)



Picture: [Halo-wheelchair-extension](#)

### Complications of Halo-ring

pin-infection – change of screw position, oral antibiotics

pin-loosening

nerve irritation (n.supraorbitalis)

intracerebral pin penetration-bleeding, pneumocephalus, brain abscess..

Halo-extension is contraindicated in rigid kyphosis apex-by extension of the proximal and distal spine in a rigid apex the myelon can be bent over the apex with following paraplegia!

### Operation techniques for congenital deformity

- posterior fusion in situ
- convex hemiepiphysodesis
- resection of half vertebra (anterior and posterior, from posterior alone)
- instrumented correction and fusion
- concavesided osteotomy and distraction (D.Jesensky)
- growing rods (single rod concavesided, Luque Trolley...)
- VEPTR (vertical expandible prosthetic vertebra rib)
- combined operations

### Fusion in situ

- in segmentation defects (unilateral bar)
- in fully segmented hemivertebrae (as soon as diagnosis is clear!)
- use of implants is recommended already in toddlers
- posterior fusion alone mainly in kyphotic deformities
- additional anterior fusion depends on the quality of disks-cave Crankshaft-phenomenon, mainly in lordotic deformities!

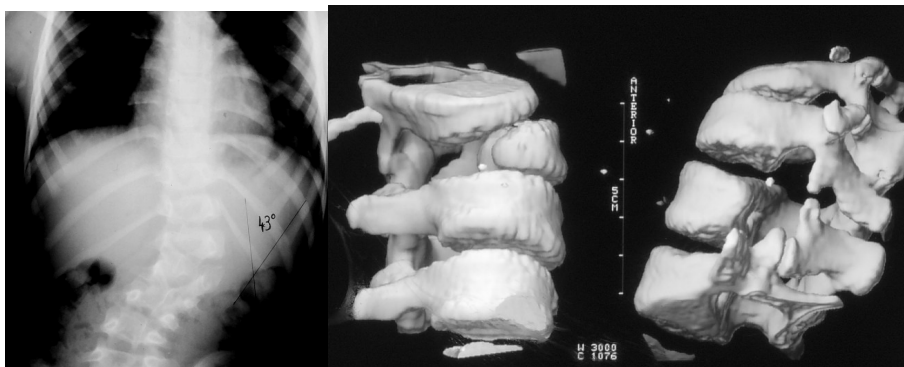
### Convexsided hemiepiphysodesis

- in cases of expected growth at the concave side (hemivertebra)
- optimal in children <5 years
- correction av. <15°
- anterior and posterior approach
- anterior: disc resection cranial and caudal of hemivertebra only to midline with bone grafting
- posterior only approach with use of pedicle screws to obtain anterior growth arrest by transpedicular convexsided disc resection and bone transplantation

### Resection of half-vertebra

- anterior-posterior**-in lateral decubitus position, fixation by hooks or pedicle screws
- posterior approach alone-blood loss and complication risk higher!

**posterior** : in cases with good flexibility and relatively normal segmentation







Picture: half vertebra resection

### Correction and instrumented fusion

**anterior-posterior:** 1) in cases of less mobility in bending films  
2) at risk of Crankshaft

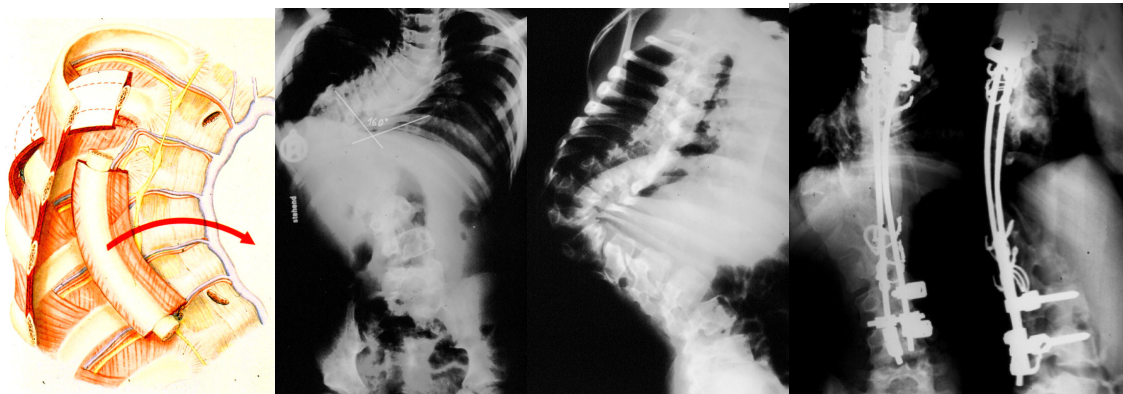
**in combination with osteotomies**-eventually with intermittent Halo traction (cave-no traction in rigid apex of kyphosis!)

**as posterior correction alone by pedicle subtraction osteotomy**

### Anterior support in remaining kyphosis

#### Bradford-technique of vascularized rib graft

preparation of elected rib under remaining intercostal muscles cranial and caudal;  
anterior ligation of intercostal vessels, posterior cautious deperiostation and cut of rib  
under care of vessels; preparation of intercostal artery and vein to the foramen, then  
creation of holes into the end-vertebral bodies of kyphosis and implantation of  
vascularized rib; osseous integration within of 2 months!



Pictures: nonon-fusion-techniques  
growing rods-subcutaneous or submuscular rods





Picture: [Luque-Trolley](#)

Picture: [VEPTR-vertical expandable prosthetic vertebral rib](#)



#### **anterior approaches in congenital deformity**

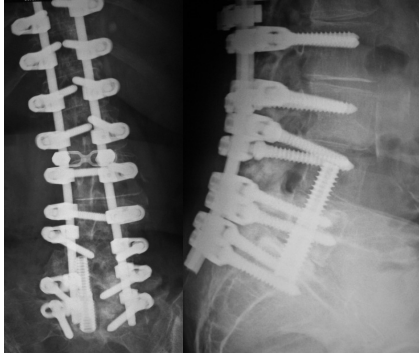
thoracotomy, thoracophrenolumbotomy Hodgson, double-thoracotomy Bauer, extreme lateral approach

#### **iliosacral fixation**

**Galveston technique (rods 6-8cm into the iliac bone)**



[Iliac screws](#)  
[S1+S2-screws](#)  
[AxiaLIF L5/S1](#)



Picture: AxiaLIF als anterior lumbosacral support in long-distance fusion