

Congenital deformity of the spine-classification, diagnosis and therapy

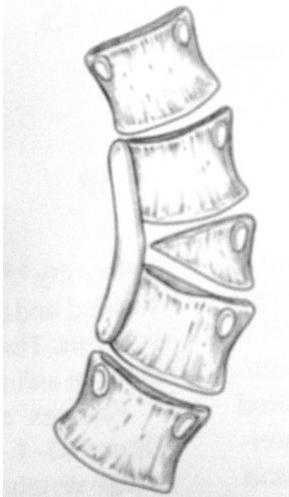
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Classification of congenital scoliosis

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

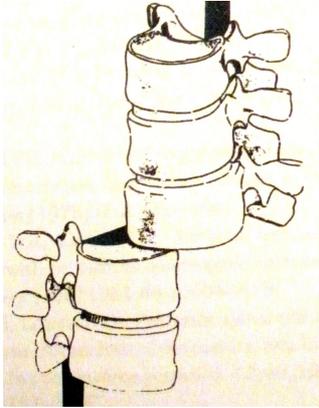
hemivertebra thoracolumbar 2-3,5°/year
2 hemivertebrae 5°
unilateral unsegmented bar 6-9°
unilateral unsegmented bar with contralateral hemivertebra >10°
most severe progression in thoracolumbar region!
most severe progression up to 5th year and in adolescence/puberty

Classification of congenital kyphosis (Mc Master)

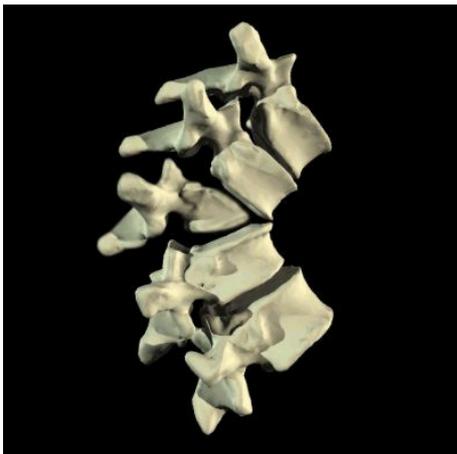
failure of formation
 posterolateral quadrant hemivertebra
 butterfly vertebra
 posterior hemivertebra
 wedge vertebra
failure of segmentation
 anterior unsegmented bar
 block vertebra
mixed anomalies
unclassifiable anomalies



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



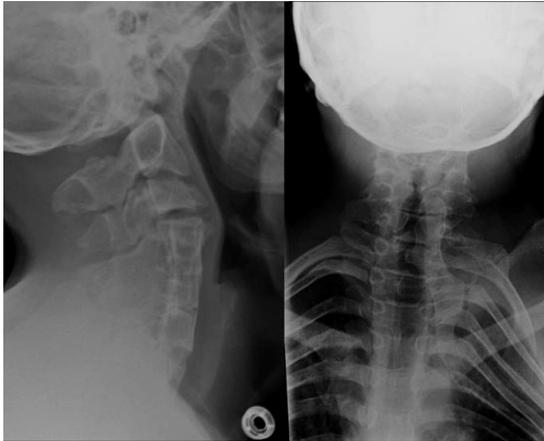
Picture: posterior half vertebra

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

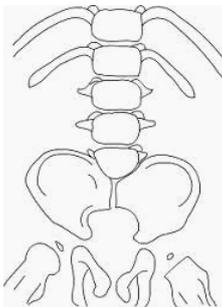
lumbosacral abnormalities (partial, total, asymmetric, symmetric)
sacral obliquity
sacral dysplasias up to sacral agenesis
sacral dysraphism (MMC)

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

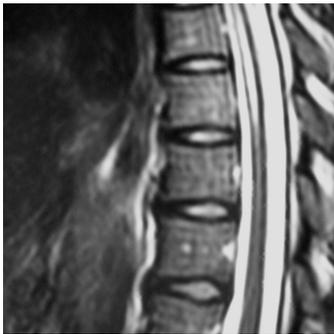
mostly in combination with congenital scoliosis, rarely with congenital kyphosis
70% had thoracic or thoracolumbar concomitant congenital scoliosis

mostly on concave side of unilateral failure of segmentation

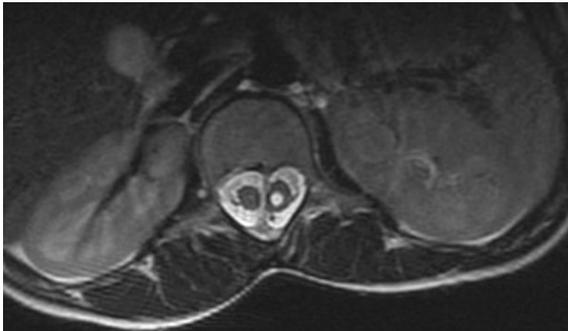
40% with Sprengel deformity
prognosis without difference to scoliosis without rib fusions

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, diplomyelia-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, hypospadias...
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

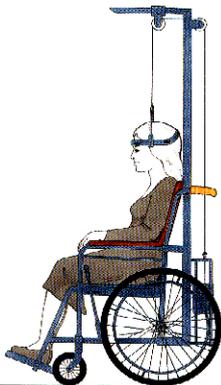
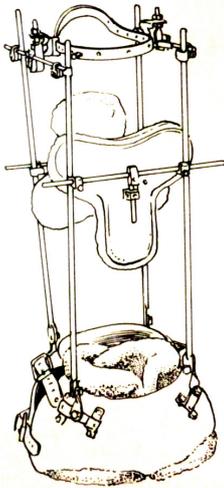
pyramid signs - Babinsky, abdominal reflex

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!

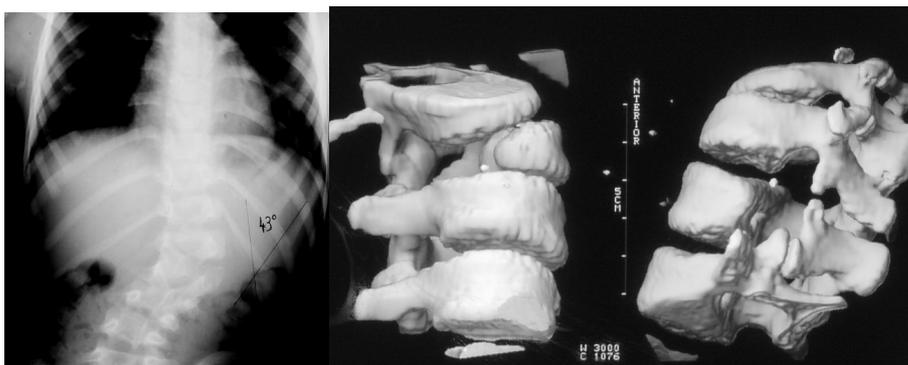
Convexsided hemiepiphysodesis

- 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

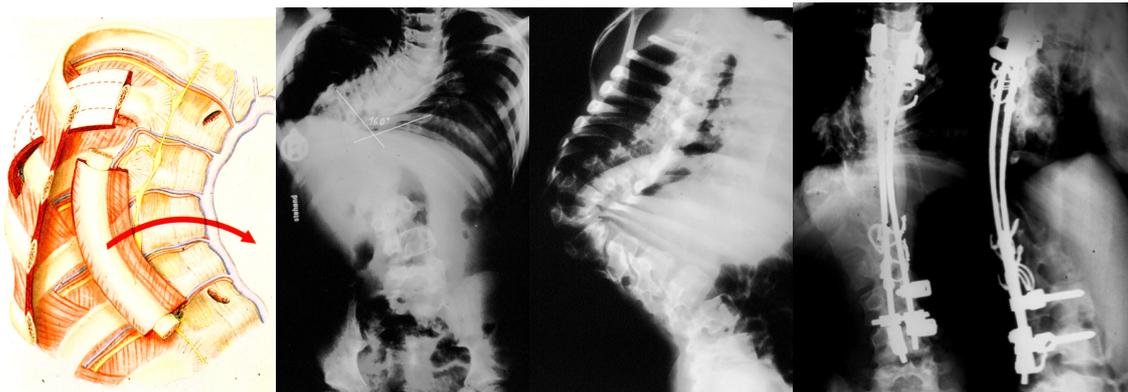
in combination with osteotomies-eventually with intermittent Halo traction (cave-no traction in rigid apex of kyphosis!)

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

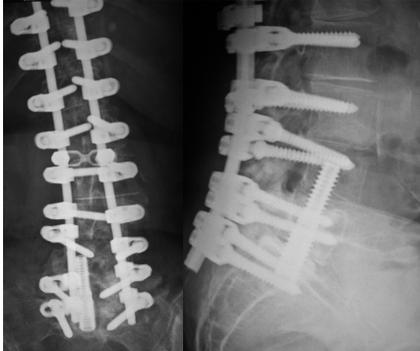
thoracotomy, thoracophrenolumbotomy Hodgson, double-thoracotomy Bauer, extreme lateral approach

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