congenital spinal deformities

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Congenital scoliosis

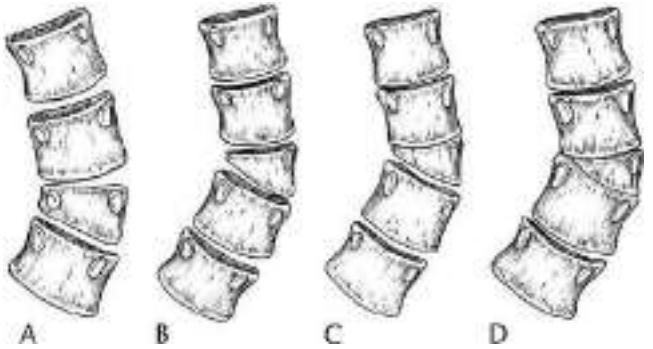
- Curvature of the spine caused by congenital anomalies of the vertebrae and supporting structures (ribs)

- Congenital scoliosis is a lateral deformity of the spine with a disturbance of **the sagittal profile** caused by malformations of vertebra and ribs.

Congenital scoliosis MacEwen classification

- Failure of formation
- partial wedge vertebra
- complete hemivertebra

- Failure of segmentation
- unilateral unsegmented bar
- bilateral block vertebra



Schematic representation of formation failures. A, Wedge vertebra. B, Fully segmented hemivertebra. C, Partially segmented hemivertebra. D, Unsegmented hemivertebra.

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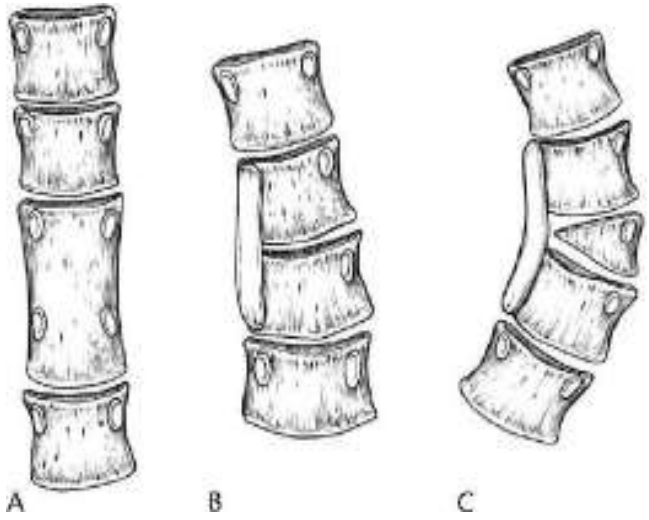
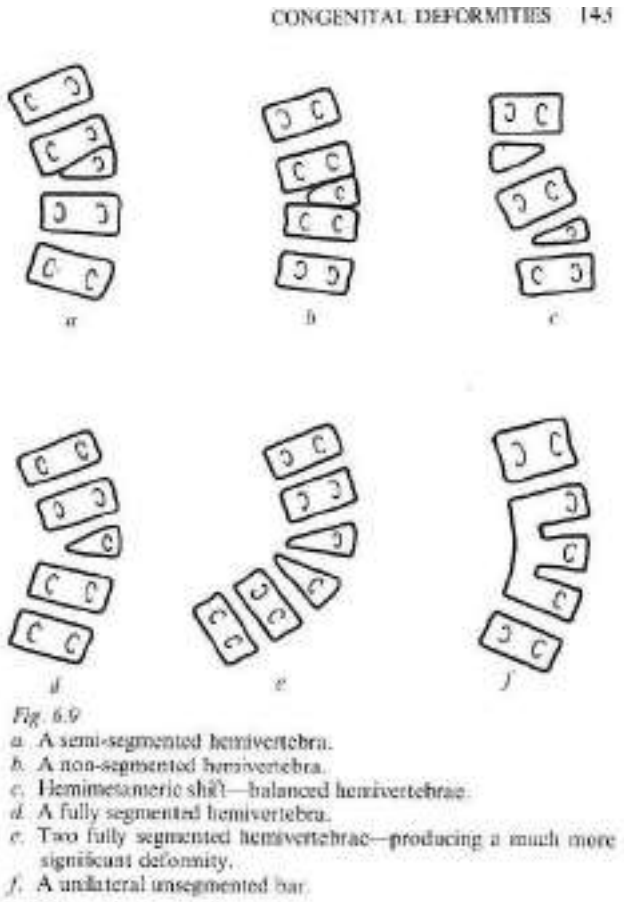


Failure of formation hemivertebra

- Lack of cartilaginous precursor due to notochord problem
- Lack of vascularization
- Reduction or absence of blood supply during surgery



Congenital scoliosis

- Neural element anomalies
- Diastematomyelia
- Spinal dysraphism

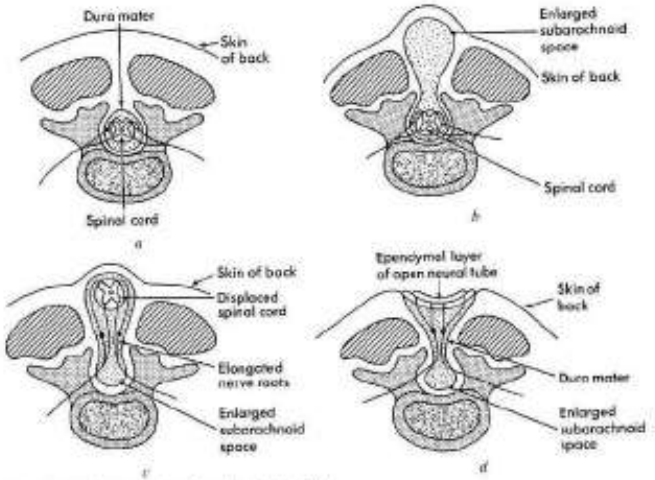
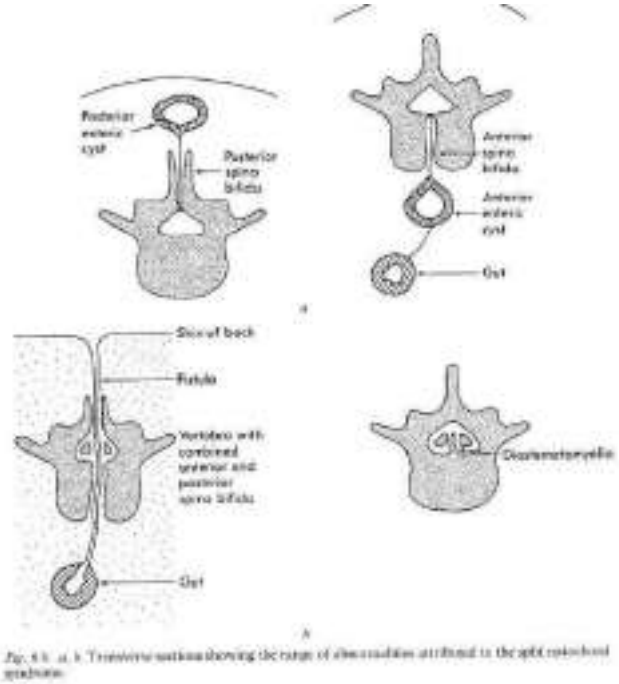


Fig. 6.7. The four categories of severity of spina bifida.
a. Spina bifida occulta.
b. Meningocele.
c. Myelomeningocele.
d. Myelocele.



Congenital scoliosis associated anomalies

- Genitourinary anomalies
- *Undifferenciated block of mesenchyme that differentiate medially into vertebra and ventrolaterally into mesonephros*
- Congenital heart disease
- Spinal dysraphism

Congenital scoliosis associated anomalies ribs fused

Congenital scoliosis associated anomalies
ribs fused
oesophagus stenosis

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Congenital scoliosis associated anomalies multiple vertebra , ribs and hydrocephalus

Congenital scoliosis associated anomalies Fragouli ma

Congenital scoliosis multiple hemivertebra butterfly

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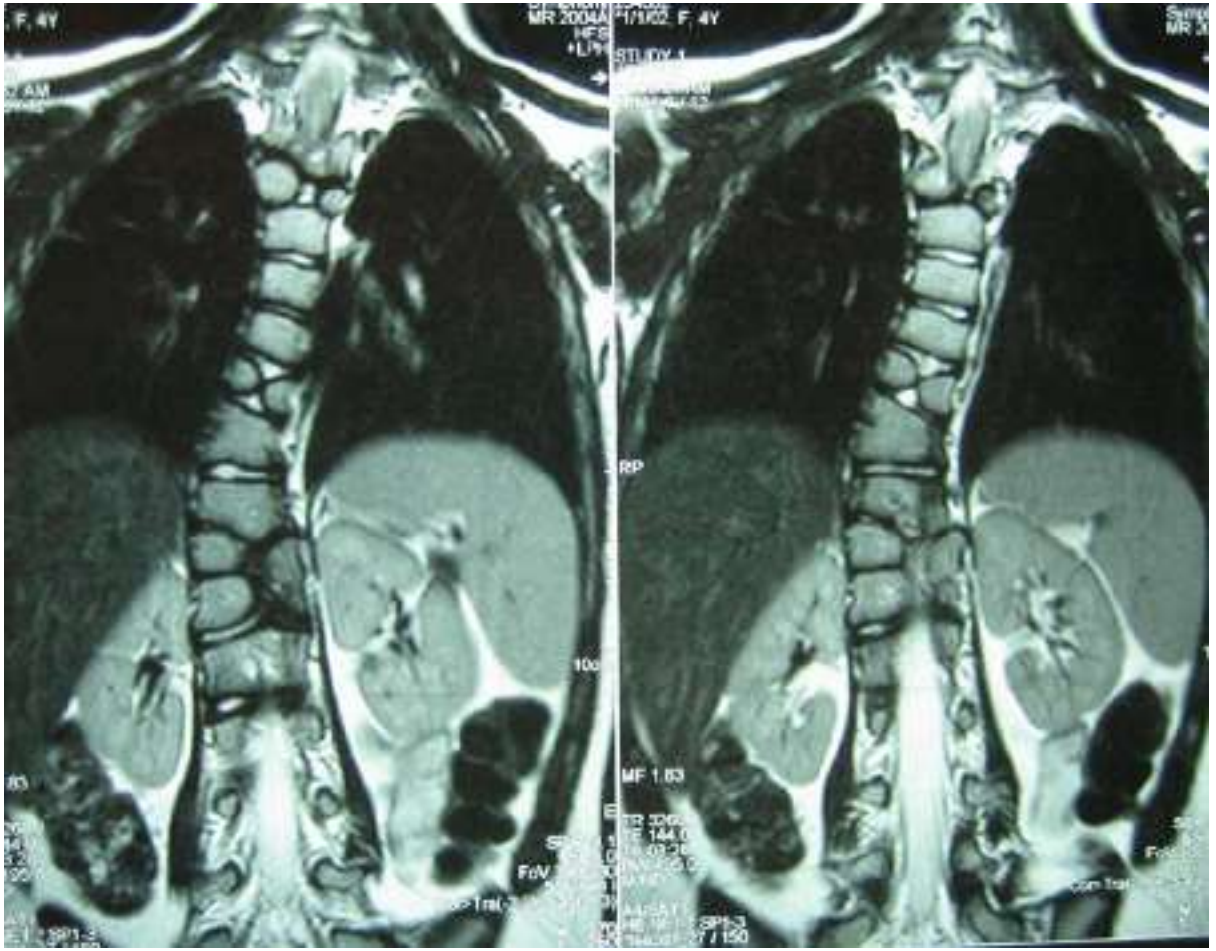
Congenital scoliosis multiple hemivertebra butterfly



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Congenital scoliosis pre natal diagnosis

Congenital scoliosis Spinal dysraphism

- Tethered spinal cord
- Fibrous dural band
- Intradural lipoma

- Hair patches, hemangiomas in the middle line in the back
- Foot deformities cavus feet, club feet, vertical talus

Congenital scoliosis clinical picture and xray

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Feet deformities and spine disorders

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Congenital scoliosis sacral dysplasia Vartso

The split notochord syndrome

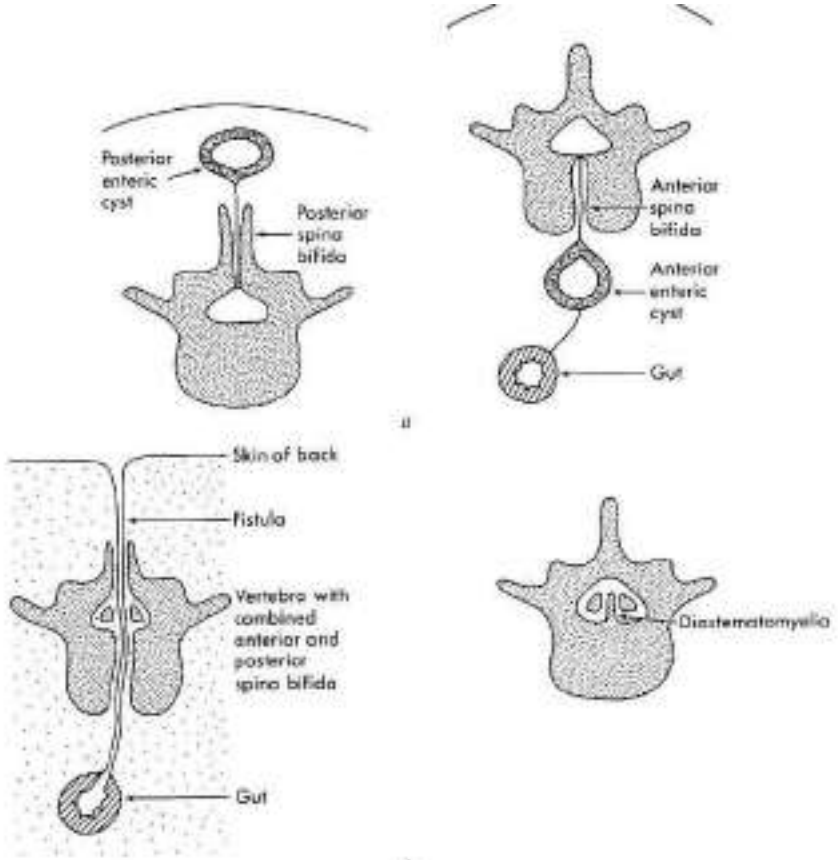
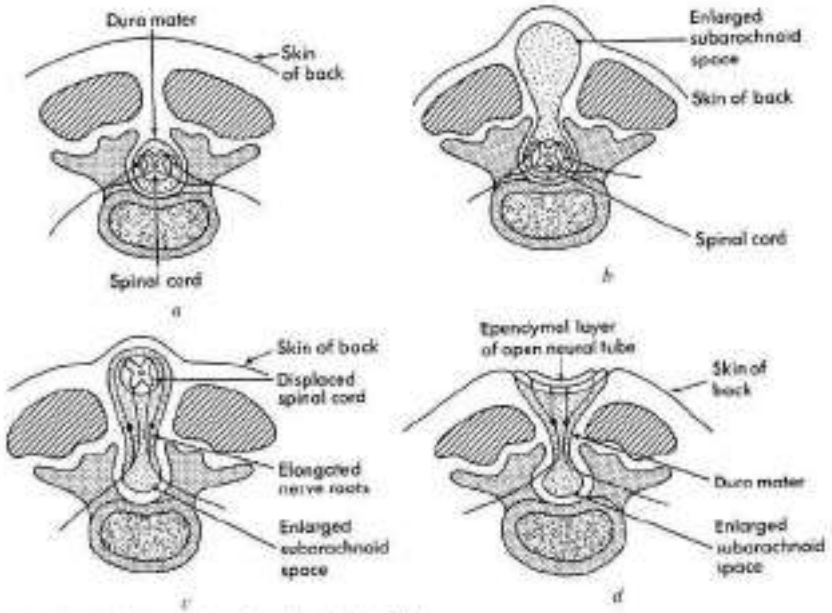


Fig. 6.5 a, b. Transverse sections showing the range of abnormalities attributed to the split notochord syndrome.

SPINA BIFIDA



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Congenital scoliosis radiological documentation

- Serial radiology every 6 months
- Documentation of progression BOTH on AP and Lateral x rays

- Scoliosis may be stable for years BUT progress rapidly during growth spurt

Congenital scoliosis curve progression

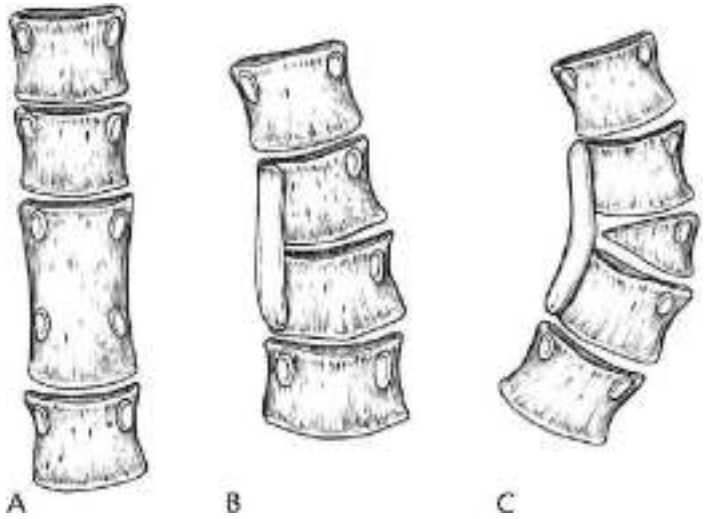
- 25% have stable curves with no progression
- 75 % have progression and 50% of them REQUIRE FUSION

Congenital scoliosis curve progression

- UNILATERAL unsegmented bar have the WORST prognosis
- The curvature is VERY STIFF that resists in correction, so it requires IMMEDIATE fusion, before any malicious progression

Congenital scoliosis curve progression

- UNILATERAL unsegmented bar have the WORST prognosis
- It is even more aggessive with contralateral hemivertebra



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hemivertebrae

- Single
- Multiple
- Balanced
- Unbalanced

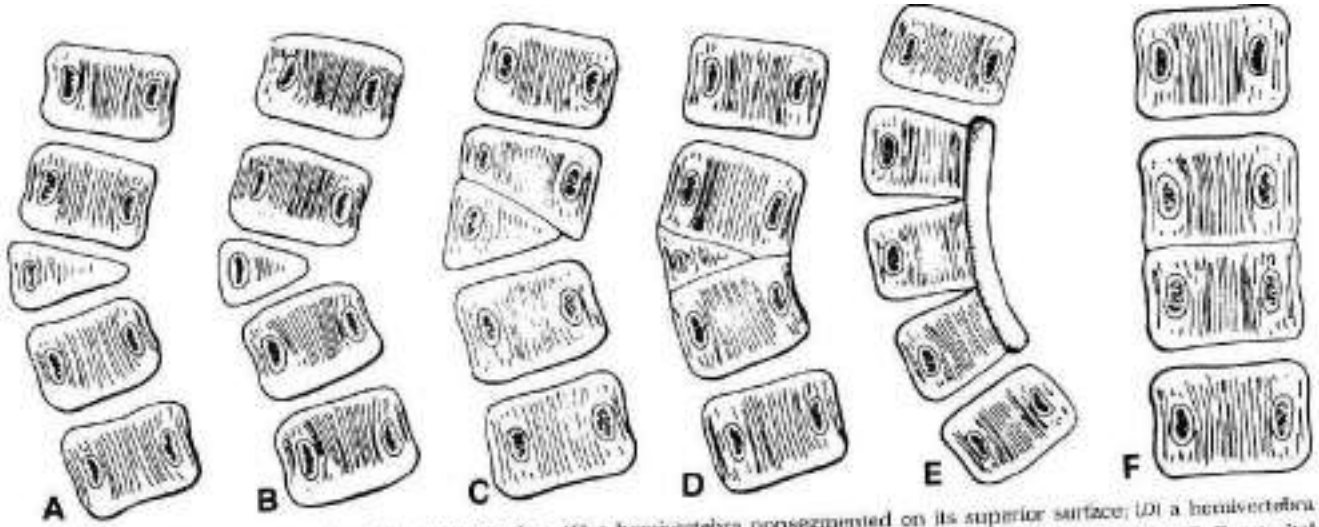

FIG. 18–34. (A) A wedge vertebra; (B) a hemivertebra; (C) a hemivertebra nonsegmented on its superior surface; (D) a hemivertebra nonsegmented on both sides; (E) a unilateral unsegmented bar; and (F) a nonsegmental vertebra (bloc vertebrae). (Winter R. Congenital deformities of the spine. New York: Thieme-Stratton, 1983:12–17)

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pine Diploma)

hemivertebrae

- Contralateral hemivertebrae separated with several segments may produce double curve, that require fusion for both
- Contralateral hemivertebrae that are close, are usually well balanced

single hemivertebra

- Difficult to predict the spinal curvature progression
- Lumbar sacral level may produce decompensation

- Incarcerated non- incarcerated

single hemivertebra

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- Lumbar sacral level may produce decompensation
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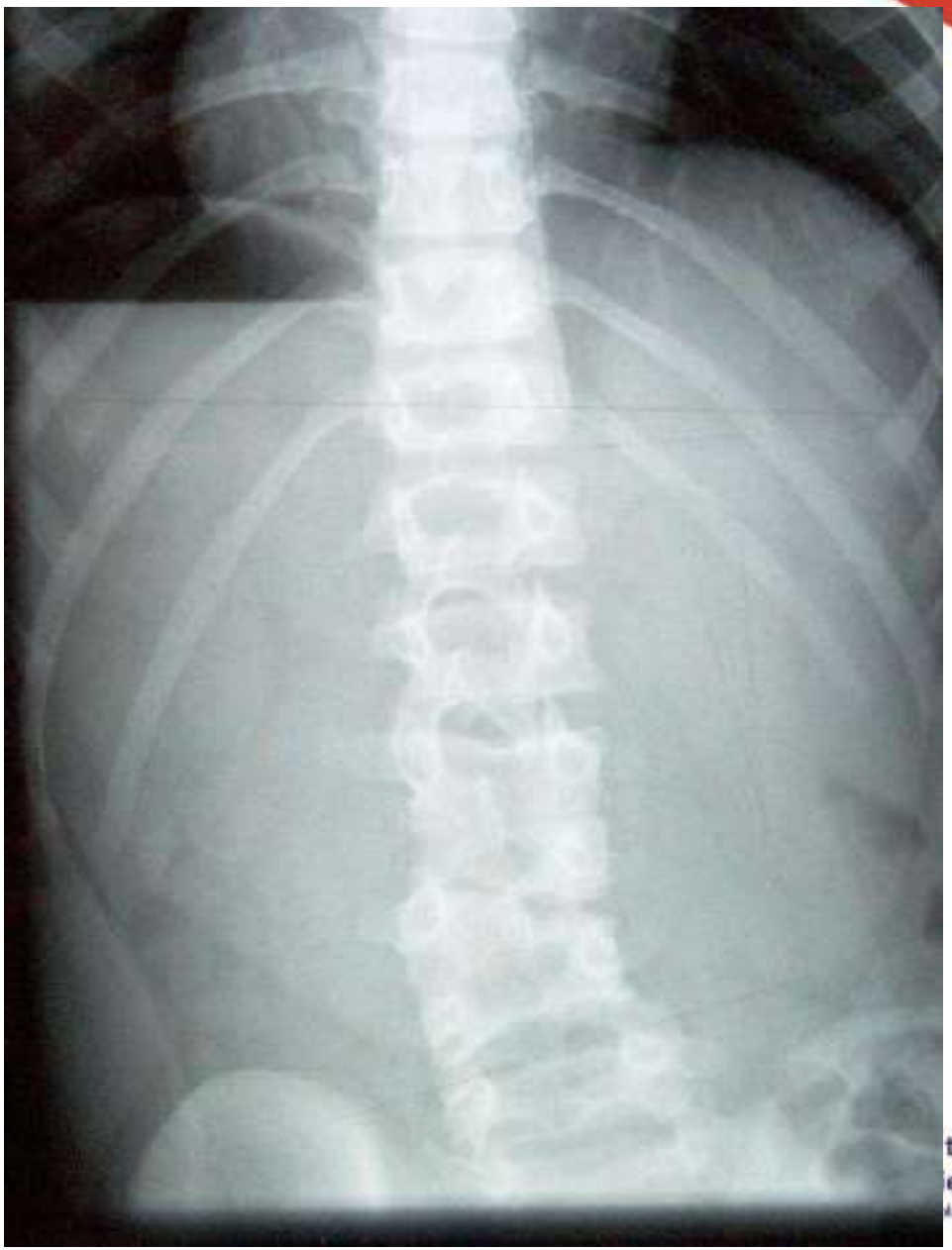

Orthotic treatment

- Effective in certain FEW indications
- Flexible curve below a proximal congenital anomaly
- Compensated curve for the stiff congenital curve
- Prevent progression OF BALANCE shift

- Useless to treat RIGID curves with an orthosis

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Thoracic hemivertebra tsak maria

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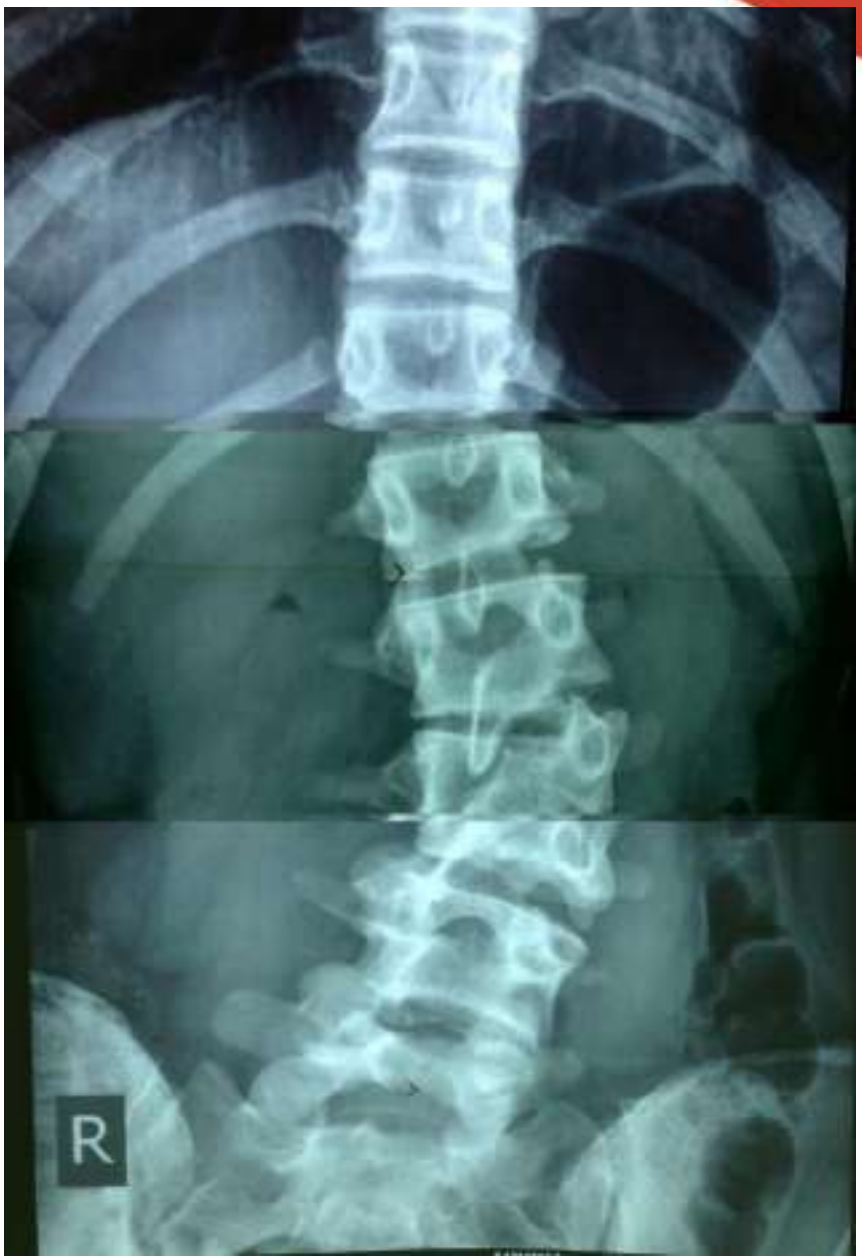
lumbar hemivertebra kont



lumbar hemivertebra
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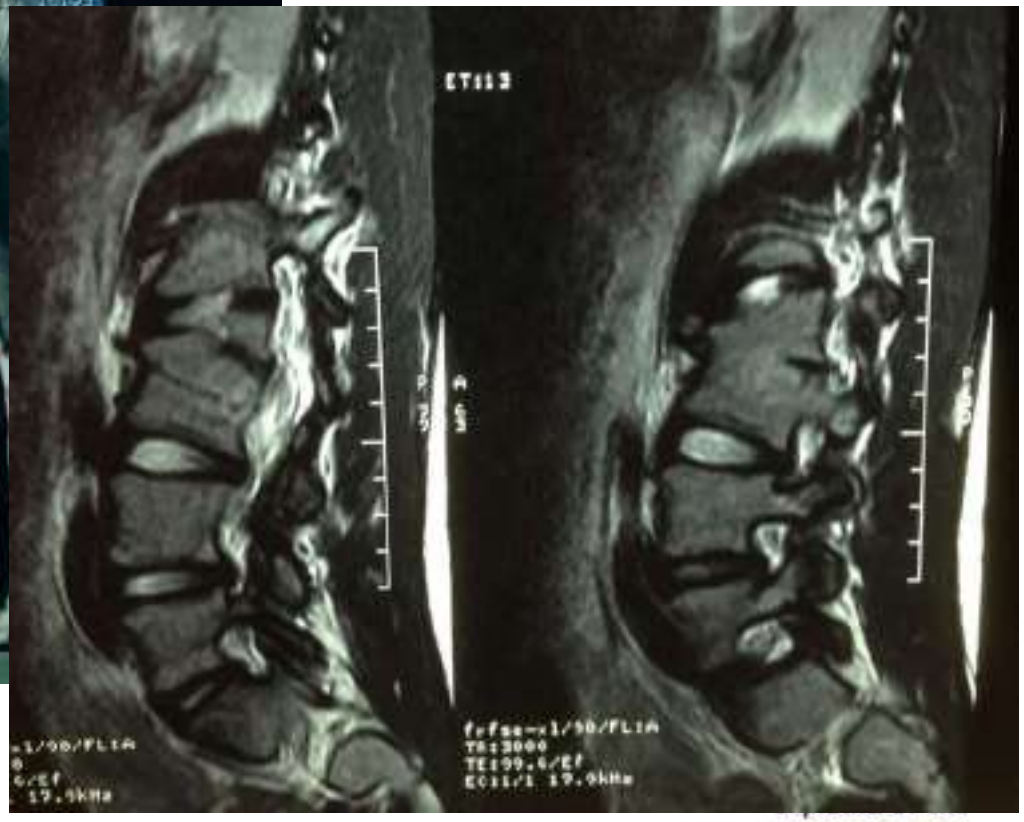

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lumbar hemivertebra tsantz

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3 d reconstruction

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lumbar
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lumbar hemivertebra
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multiple hemivertebra in a distance malliar

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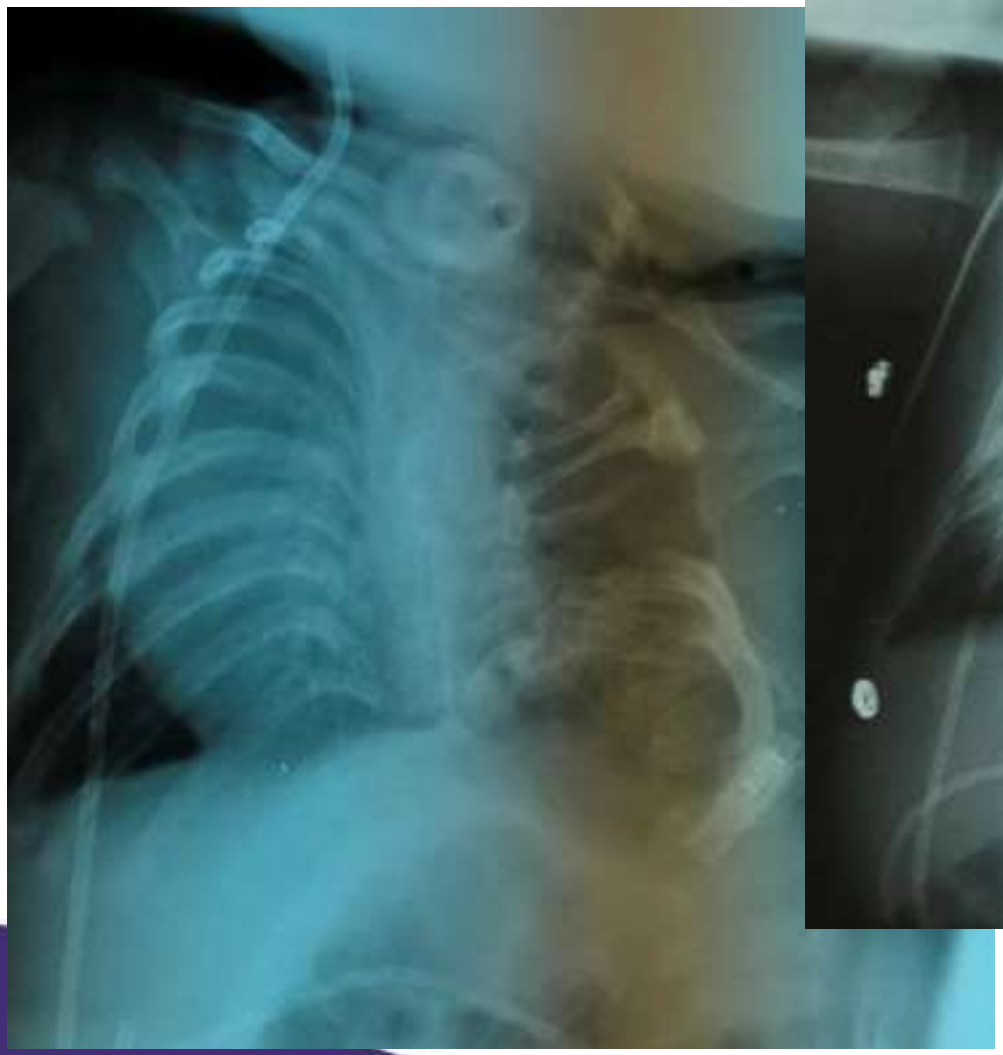
Spine dislocation



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congenital scoliosis and syringomyelia

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SURGICAL TREATMENT

- SURGERY IS IMPERATIVE for severe and progressive congenital scoliosis

- Fusion or hemivertebra excision

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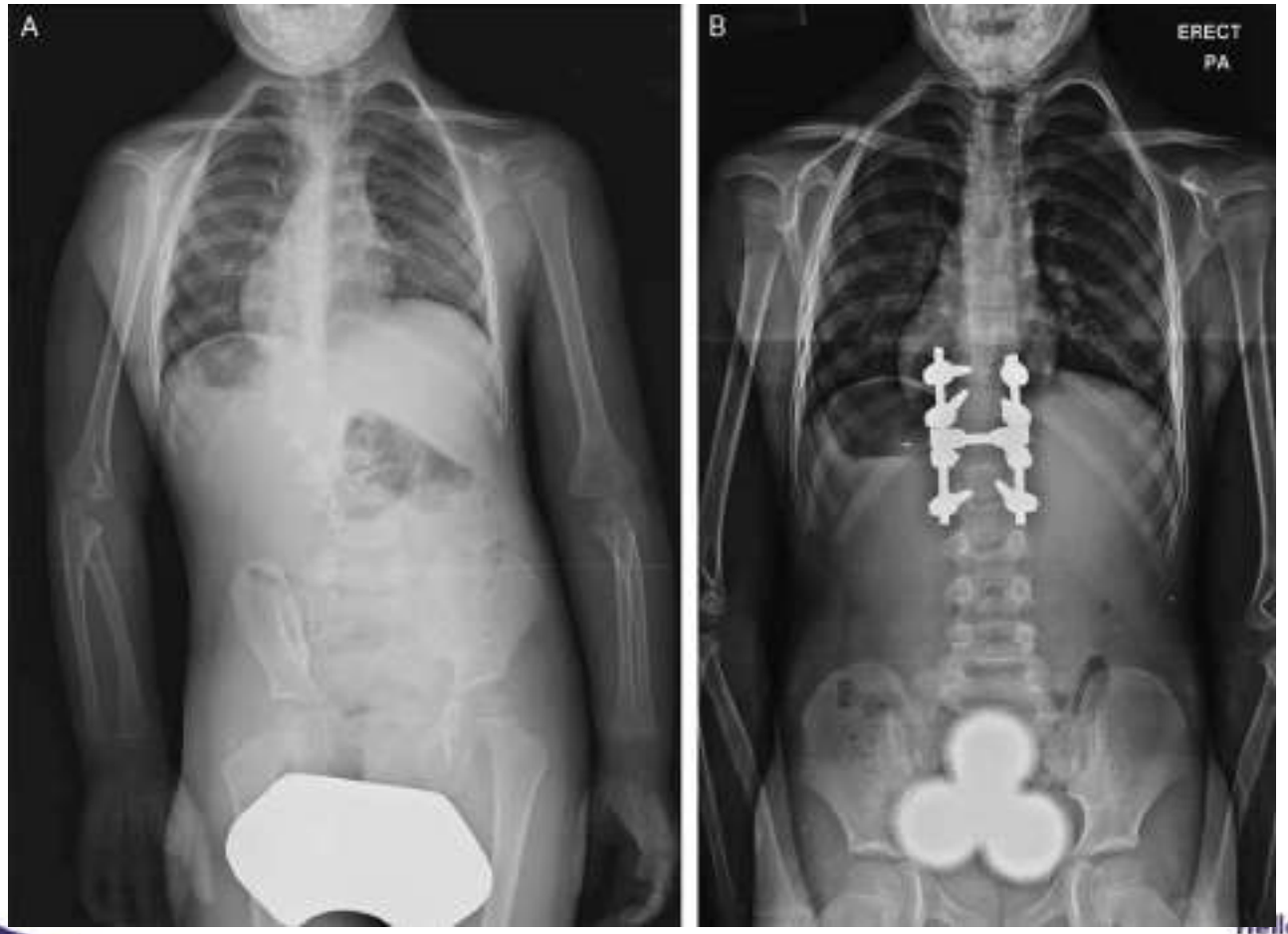
SURGICAL TREATMENT

- Early surgical treatment is required when a curve progress rapidly, even in an attempt to use an orthotic

- Anterior growth plate
- Hemivertebra excision is difficult and is dangerous to cord

What's New in Congenital Scoliosis?

Joshua M. Pahys, and James T. Guille,
J Pediatr Orthop, '' 2016



What's New in Congenital Scoliosis?
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- Growth friendly surgery with either spine-based or rib-based anchors has demonstrated good results with curve correction while maintaining spinal growth.
- Rib based anchors are typically more commonly indicated in the setting of chest wall abnormalities and/or when spinal anatomy precludes placement of spinal instrumentation.
- Recently, magnetically controlled growing rods have shown promising results in several studies that include a small subset of congenital scoliosis cases.

What's New in Congenital Scoliosis?

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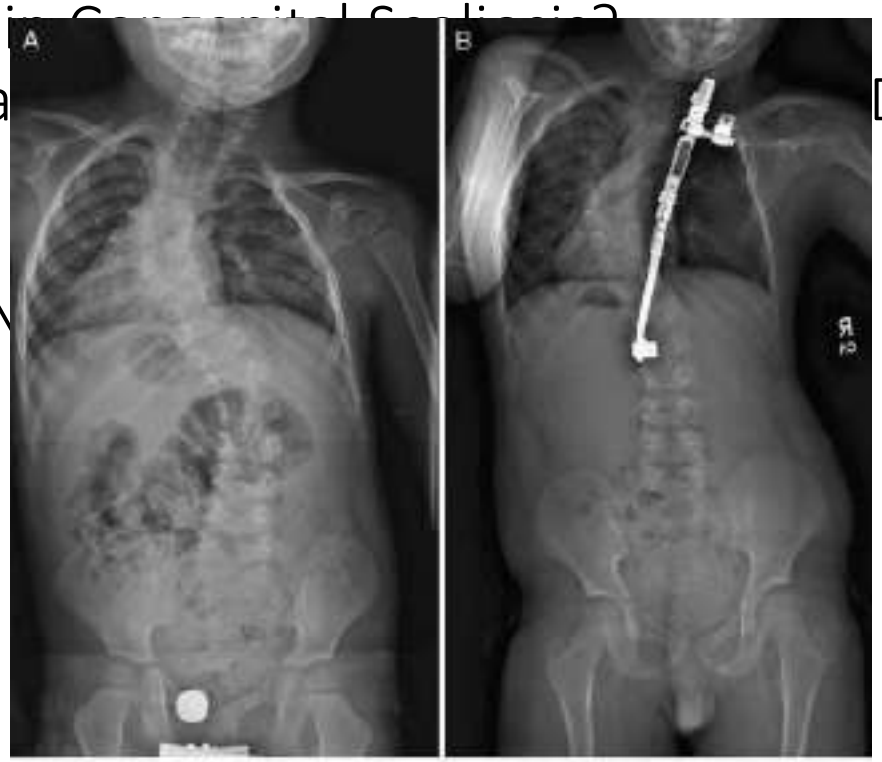

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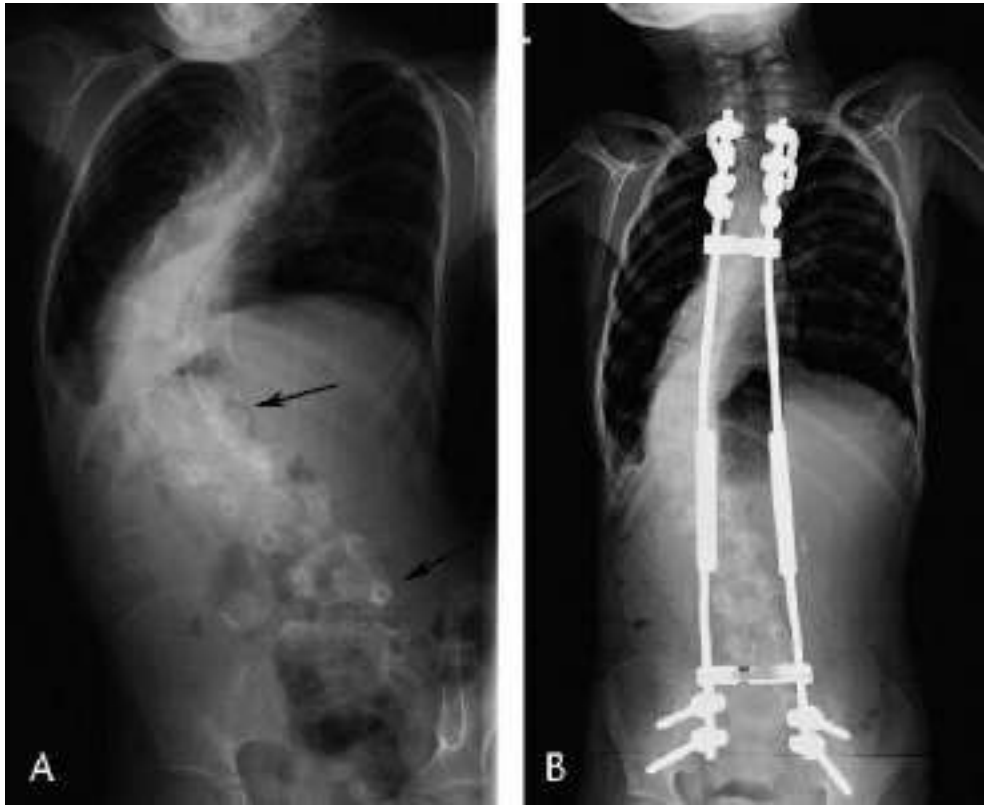
Congenital Scoliosis
A Review and Update

Daniel Hedequist, MD and John Emans, MD



Congenital Scoliosis
A Review and Update
Daniel Hedequist, MD and John Emans, MD

- Vertebral anomalies causing congenital scoliosis are classified on the basis of failures of formation, segmentation, or both. The natural history depends on the type of anomaly and the location of anomaly. Patient evaluation focuses on the history and physical examination, followed by appropriate imaging modalities. **The hallmark of surgical treatment is early intervention before the development of large curvatures**. The surgical treatment of a congenital deformity mandates the use of neurological monitoring to minimize the risk of perioperative neurological deficit. Modern surgical techniques have evolved to include the routine use of spinal instrumentation. Patients with associated chest wall deformities or large compensatory curves may be candidates for vertical expansion prosthetic titanium rib placement or growing rods insertion to maximize growth

Curr Pediatr Rev. 2016;12(1):43-7.

Congenital Scoliosis (Mini-review).

Weiss HR[1], Moramarco M.

- Congenital scoliosis is a lateral deformity of the spine with a disturbance of the sagittal profile caused by malformations of vertebra and ribs. Typically, early surgical intervention is the suggested treatment (before three-years-old) for young patients with congenital scoliosis.
- While a previous study was conducted in 2011 to investigate long-term studies supporting the necessity for this recommendation **and no evidence was found**, this current review, is an updated search for evidence published from 2011 through March 2015. This **also failed to find any prospective or randomized controlled studies** to support the hypothesis that spinal fusion surgery in patients with congenital scoliosis should be considered as evidence-based treatment.

Spine Deform. 2018.

Long-term Outcome of Early Fusions for Congenital Scoliosis.

Lonstein JE[1]

- The average scoliosis at presentation was 52°, 56° at surgery and 47° at follow-up. At follow-up, the average FVC was 53% of predicted, with an average PaO_2 of 86 and $PaCO_2$ of 44 mm Hg. The mean T1-T12 length was 20 cm.

- At a long-term average follow-up of 37 years, the patients had a low FVC of 53%, with one on permanent oxygen and 5 with dyspnea. The patients were functioning well at follow-up, but it is unknown what their function would be without surgery or with longer follow-up.

Klippel Feil syndrome

- Congenital failure of segmentation affecting the cervical spine and cervicothoracic junction (Klippel and Feil 1912)

- Short neck, low hairline, limited cervical spine motion

- Sprengel deformity

Klippel Feil syndrome

- Local scoliosis
- Scoliosis below the congenital anomalies

Klippel Feil syndrome

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Klippel Feil syndrome

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  Congenital Scoliosis (Mini-review).
  Weiss HR[1], Moramarco M.
- Congenital scoliosis is a lateral deformity of the spine with a disturbance of the sagittal profile caused by malformations of vertebra and ribs.
- An updated search for evidence published from 2011 through March 2015, **failed to find** any prospective or randomized controlled studies to support the hypothesis that spinal fusion surgery in patients with congenital scoliosis **should be considered as evidence-based treatment.** Contradictory results exist on the safety of hemivertebra resection and segmental fusion using pedicle screw fixation.
- When using the VEPTR (vertical expandable prosthetic titanium rib) device, **studies show a high rate of complications** exist. It is difficult to predict the final outcome for patients with congenital scoliosis.
- *However, it is possible that many patients with congenital scoliosis may be able to avoid spinal surgery with the application of advanced bracing technology. Therefore, it is only prudent to advocate for conservative management first before spinal surgery is considered.*