

congenital spinal deformities

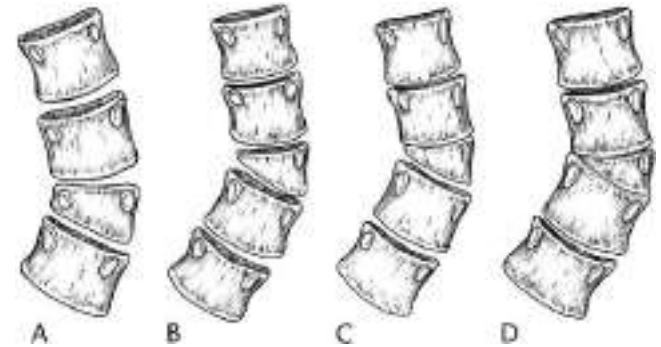
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 - Pediatric Orthopaedic Surgeon
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 - Interbalkan Medical Center

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.

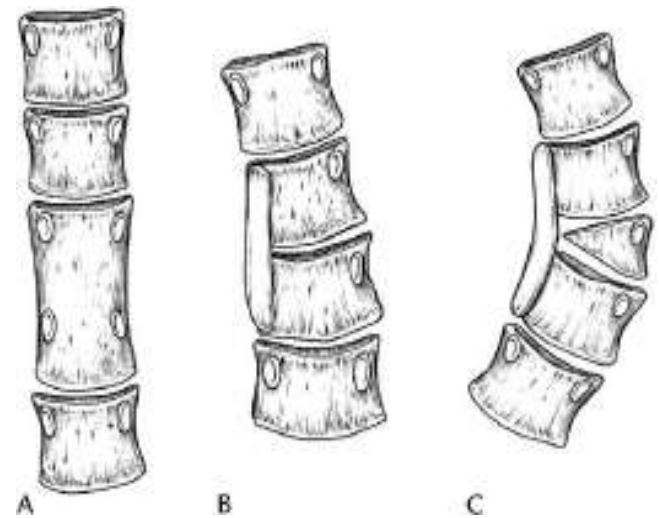
B, Fully segmented hemivertebra.

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 DEFORMITIES 145

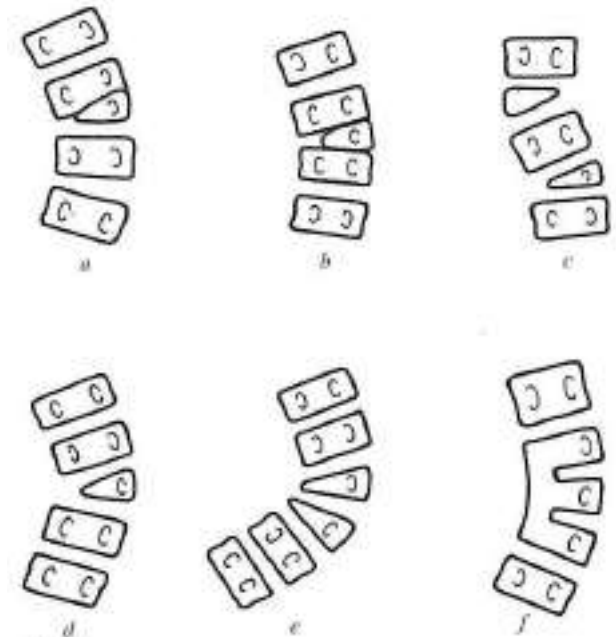


Fig. 6.9

- A semi-segmented hemivertebra.
- A non-segmented hemivertebra.
- Hemimetameric shift—balanced hemivertebrae.
- A fully segmented hemivertebra.
- Two fully segmented hemivertebrae—producing a much more significant deformity.
- A unilateral unsegmented bar.

Congenital scoliosis

- Neural element anomalies
- Diastematomyelia
- Spinal dysraphism

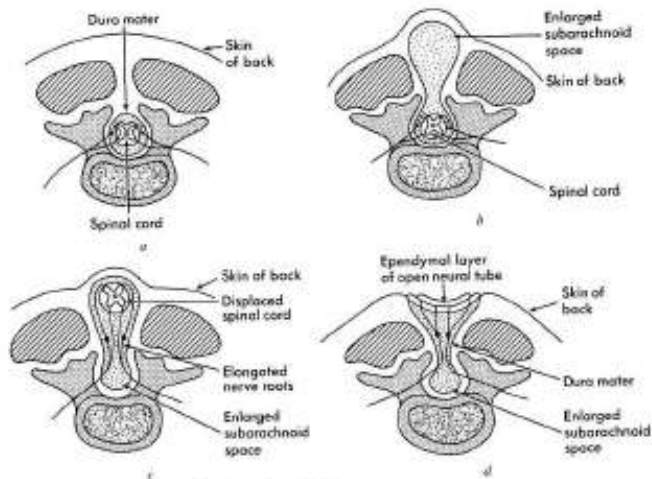


Fig. 6.7. The four categories of severity of spina bifida.
a. Spina bifida occulta.
b. Meningocele.
c. Myelomeningocele.
d. Myelocele.

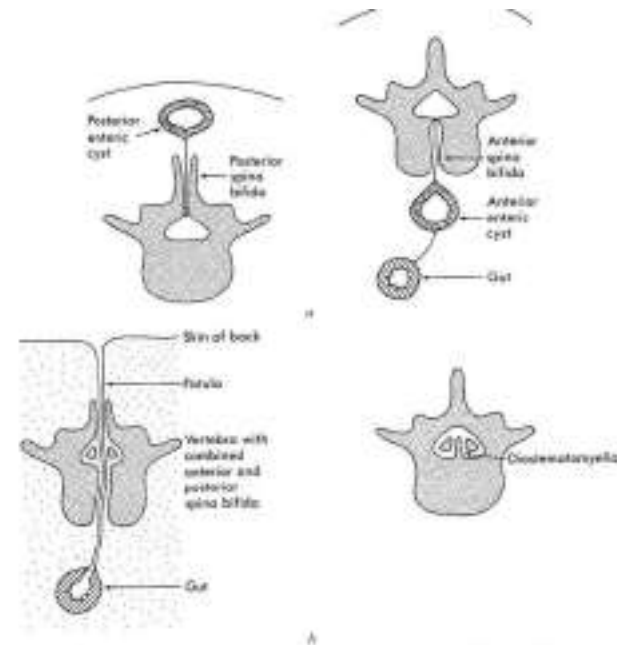


Fig. 6A. A. Transverse sections showing the range of abnormalities attached to the split vertebral syndrome.

Congenital scoliosis associated anomalies

- Genitourinary anomalies
- *Undifferentiated 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



Congenital scoliosis associated anomalies multiple vertebra , ribs and hydrocephalus



Congenital scoliosis associated anomalies

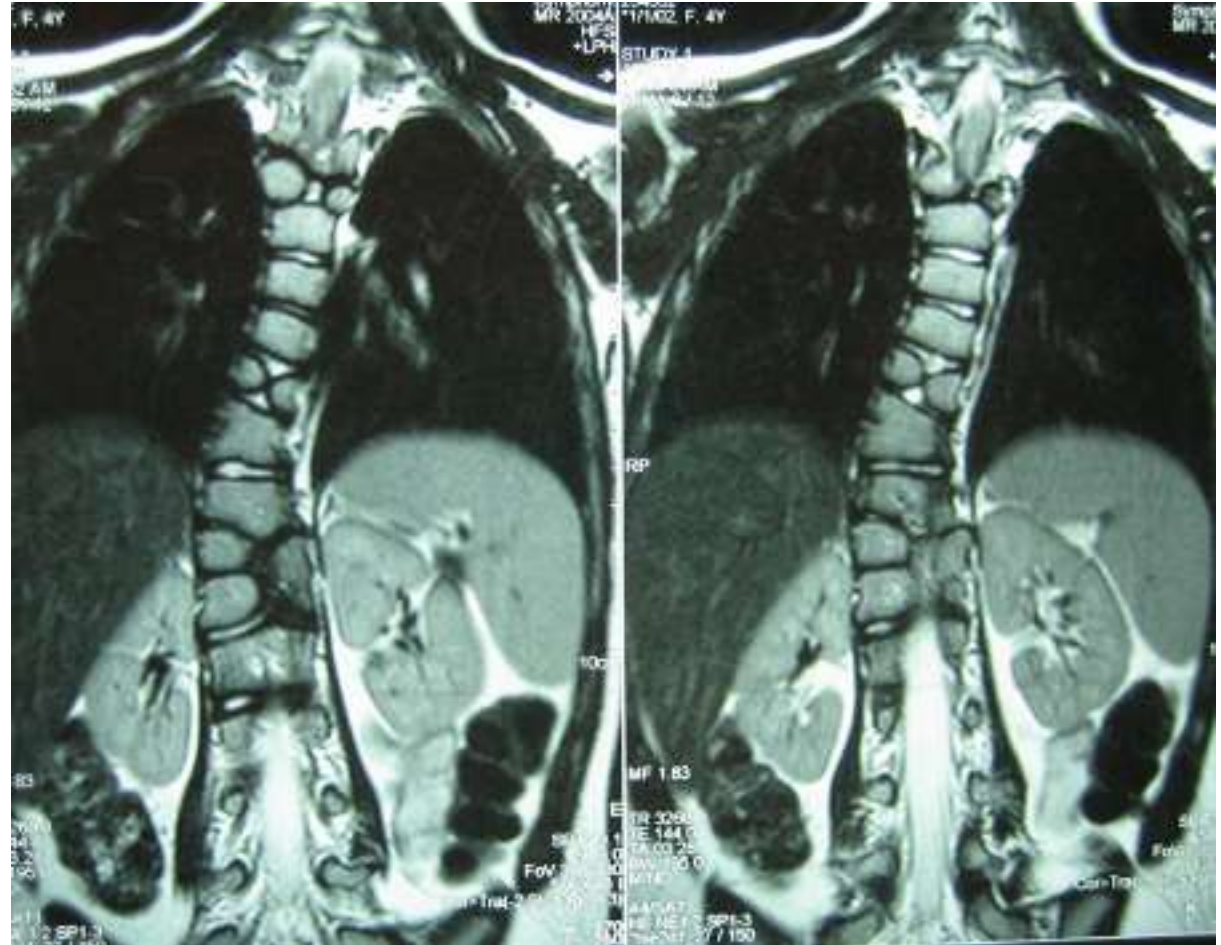
Frangouli ma



Congenital scoliosis multiple hemivertebra butterfly



Congenital scoliosis multiple hemivertebra butterfly



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



Feet deformities and spine disorders



Congenital scoliosis sacral dysplasia Vartso



SPINA BIFIDA

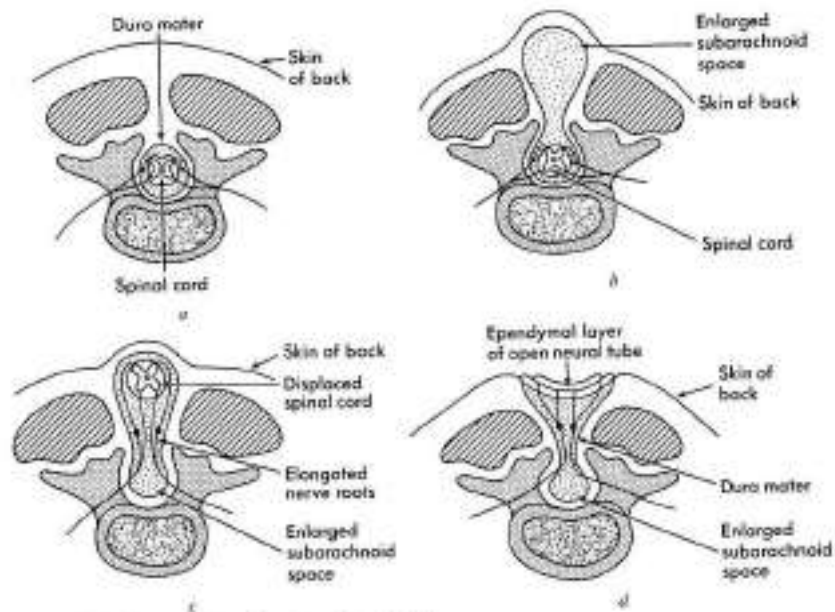


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Congenital scoliosis radiological documentation

- 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

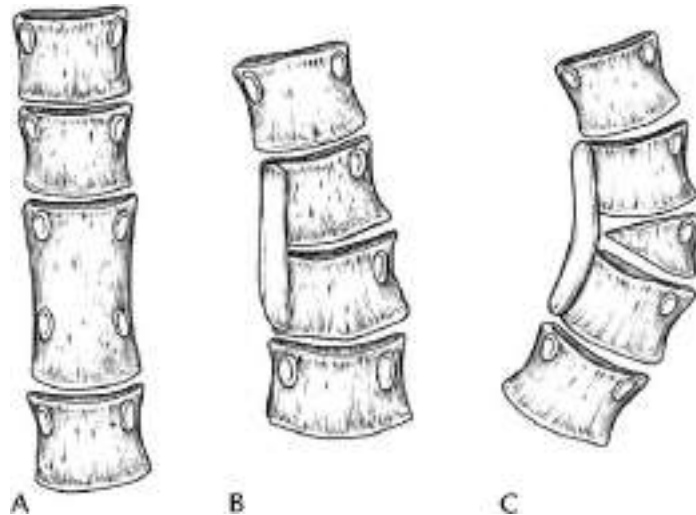
- 25% have stable curves with no progression
- 75 % have progression and 50% of them **REQUIRE FUSION**

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 aggressive with contralateral hemivertebra



hemivertebrae

- Single
- Multiple
- Balanced
- Unbalanced

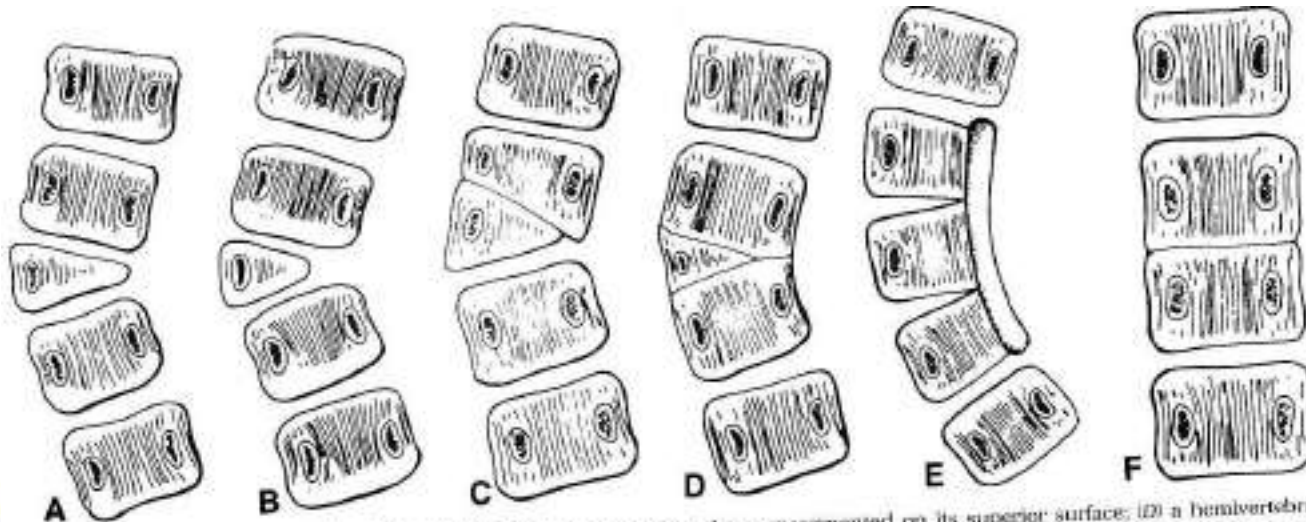


FIG. 18-34. (A) A wedge vertebra; (B) a hemivertebra; (C) a hemivertebra nonsegmented on its superior surface; (D) a hemivertebra nonsegmented on both sides; (E) a unilateral unsegmented bar; and (F) a nonsegmental vertebra (bloc vertebra). (Winter B. Congenital deformities of the spine. New York: Thieme-Stratton,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

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

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



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

Thoracic hemivertebra tsak maria



lumbar hemivertebra kont



lumbar hemivertebra tsantz



3 d reconstruction



lumbar man



lumbar hemivertebra maragou olga



multiple hemivertebra in a distance maliar



Spine dislocation



Del eleni congenital scoliosis and syringomyelia



Del eleni congenital scoliosis and syringomyelia



Congenital kyphosis mil



Congenital kyphosis mil



Congenital kyphosis milonas



SURGICAL TREATMENT

- SURGERY IS IMPERATIVE for severe and progressive congenital scoliosis
 - Fusion or hemivertebra excision

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.



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Congenital
Scoliosis?
Joshua M.
Pahys,
MD* and
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Pediatr
Orthop

Volume
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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

- 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

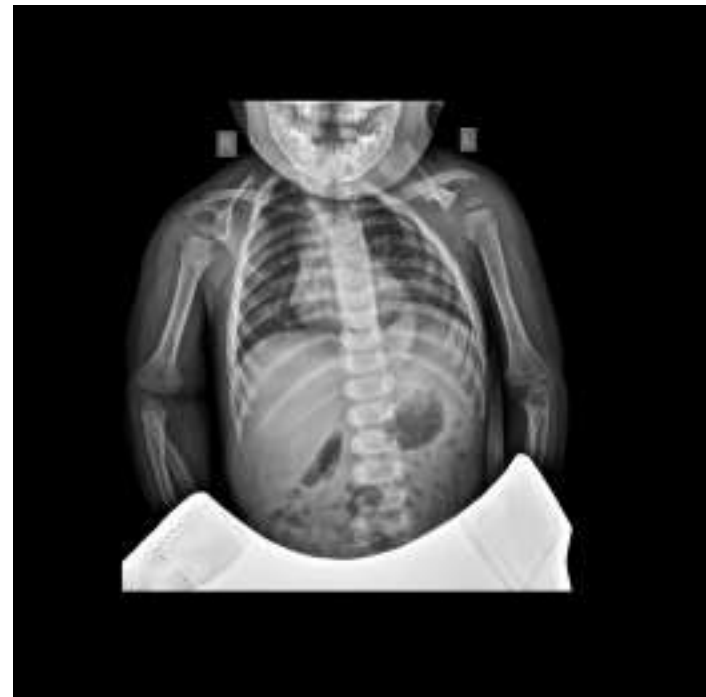
- The average scoliosis at presentation was 52°, 56° at surgery and 47° at follow-up. At follow-up, the average FVC was 53% of predicted, with an average PaO₂ of 86 and PaCO₂ of 44 mm Hg. The mean T1-T12 length was 20 cm.
- At a long-term average follow-up of 37 years, the patients had a low FVC of 53%, 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



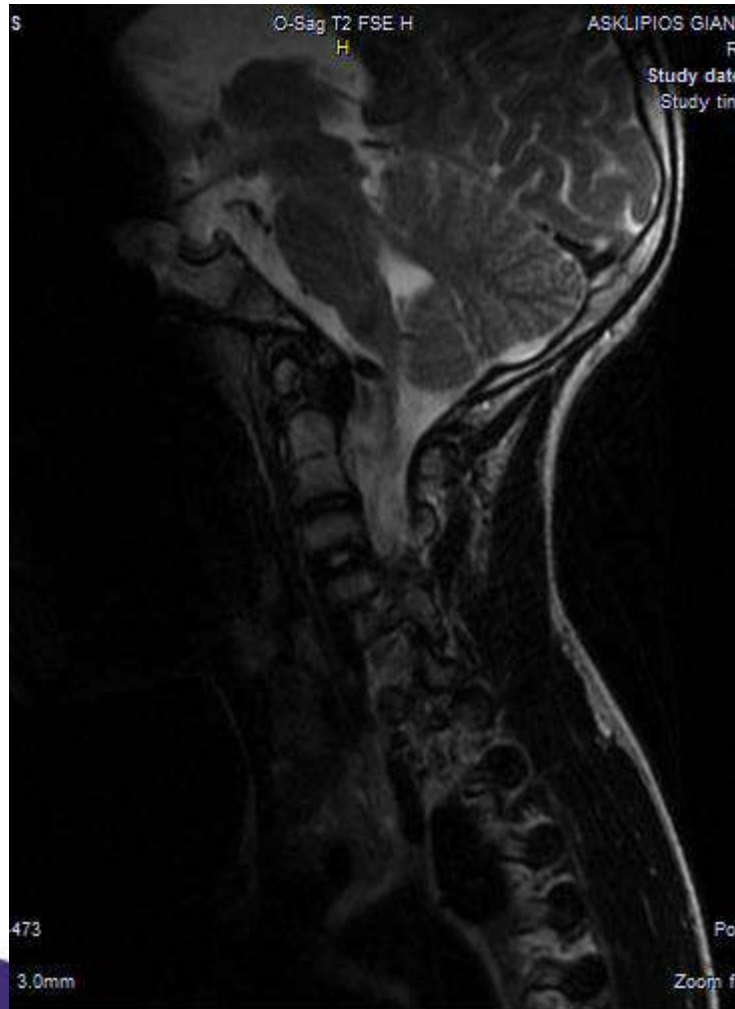
Klippel Feil syndrome

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Klippel Feil syndrome

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Klippel Feil syndrome

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Klippel Feil syndrome

- kais



- [Curr Pediatr Rev. 2016;12\(1\):43-7.](#)
Congenital Scoliosis (Mini-review).
[Weiss HR¹](#), [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.***

- J Pediatr Orthop. 2018 Mar;38(3).
What's New in Congenital Scoliosis?

[Joshua M Pahys¹](#), [James T Guille](#)

Bracing for congenital scoliosis is rarely indicated, while recent reports have demonstrated the **utility of serial derotational casting** for longer curves with multiple anomalous vertebrae as an effective "**time buying strategy**" to delay the need for surgery.

Earlier hemivertebra excision and short-segment posterior spinal fusion have been advocated to prevent future curve progression of the deformity and/or the development of large compensatory curves. It has been shown in recent long-term follow-up studies that growth rates of the vertebral body and spinal canal are not as dramatically affected by pedicle screw instrumentation at a young age as once thought.

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.

- Childs Nerv Syst 2018 Nov;34(11):2155-2171).
A comprehensive review of the diagnosis and management of congenital scoliosis

Mackel CE, Jada A, Samdani AF, Stephen JH, Bennett JT, Baaj AA, Hwang SW.

Congenital scoliosis represents 10% of pediatric spine deformity and is a developmental error in segmentation, formation, or a combination of both leading to curvature of the spine. **Treatment options are complicated by balancing growth potential with curve severity.** Often associated abnormalities of cardiac, genitourinary, or intraspinal systems are concurrent and should be evaluated as part of the diagnostic work-up. Management balances the risk of progression, growth potential, lung development/function, and associated risks. **Surgical treatment options involve growth-permitting systems or fusions.**

Congenital scoliosis is a complex spinal problem associated with many other anomalous findings. **Treatment options are diverse but enable optimization of management and care of these children**