CHAPTER 3

SPINE

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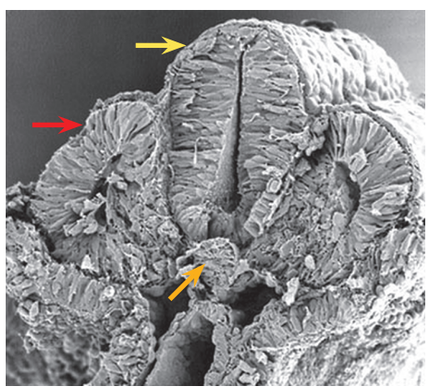
Converse to adults, deformity commands the largest proportion of spine care in children. Back pain, while not infrequent, in most children will have no identifiable cause.

EMBRYOLOGY AND DEVELOPMENT

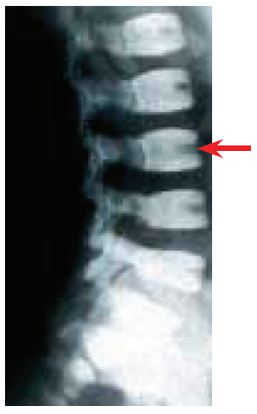
At the end of the 2nd week, the embryo is trilaminar, composed of endoderm (which gives rise to the gut) adjacent to the yolk sac, ectoderm adjacent to the amnion, and mesoderm in between.

Ectoderm gives rise to the nervous system. The notochord appears during the 3rd week, at the beginning of *gastrulation*. The notochord (of which the vestige is the nucleus pulposus) induces the formation of the neural plate. The neural plate begins to fold into the neural tube in the 4th week from its center rostrad and caudad, a process known as *neurulation*. During neurulation, there is *disjunction* of the superficial ectoderm, which goes on to form the skin.

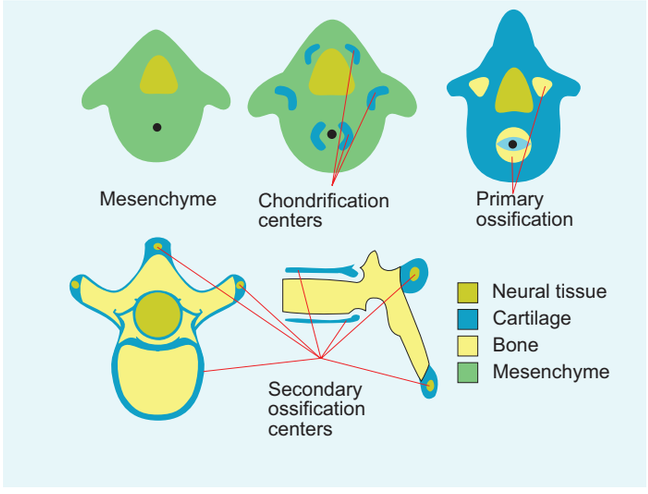
Paired blocks of mesoderm, known as somites, give rise to the musculoskeletal system [A]. The part of a somite that forms bone is known



A Scanning electron micrograph of an embryo Somite, *red*. Neural tube, *yellow*. Notochord, *orange*. [http://php.med.unsw.edu.au]



B Vertebral intersegmental development The vertebral bodies form as intersegmental structures. As blood vessels grow between somites, their final position is midvertebral. The site of blood vessel entry and somite fusion may appear radiographically as an anterior notch in the vertebral body of the child (*red*).



C Vertebral development Chondrification leads to ossification, which is complete by maturity. From Moore KL. *The Developing Human*. 4th ed. Philadelphia, PA: WB Saunders; 1988.

as a sclerotome and that which forms muscle is a myotome. During the 4th week, the sclerotome grows around the notochord to become the vertebral body and anulus fibrosus and around the neural tube to form the vertebral arches.

The rostral half of one sclerotome fuses with the caudal half of the adjacent one to form each vertebra. This intersegmentation means that original vascular ingrowth between somites ends in midvertebral segmental blood vessels. Anterior notching of vertebrae in the immature spine marks the site of somite fusion [B].

During the 7th to 8th weeks, paired *chondrification* centers appear around the notochord, which will fuse during the process of *ossification* to form the centrum of the vertebral body; around the neural tube, where they fuse to form the neural arch; and at each transverse and spinous processes, where their rôle is apophysial [C]. Fusion of the neurocentral junction occurs by 5 years of age, at which time the final capacity of the vertebral canal is determined.

Differential growth of the spinal cord and vertebral column leads to *ascension* of the conus medullaris to its adult level at L1 by the first few months of age. This is accompanied by formation of the cauda equina, as lumbosacral nerves travel to reach their respective intervertebral foramina. The spinal cord is anchored to the coccyx by the filum terminale.

In the coronal plane, the spine is straight throughout growth. In the sagittal plane, the spine evolves from a single curve at birth to a triple curve.

A knowledge of embryology and its nomenclature aids an understanding of disease.

- Dysrrhaphism refers to a "bad" "groove," a general term for midline, that is, spine anomaly
- Schisis refers to "splitting, cleavage, or *schism*" of the spine. Two types are distinguished, myeloschisis: "splitting of the medulla, or spinal cord," and rhachischisis: "splitting of the osseous spine."
- Spina bifida is the general term for "splitting of the spine." This may be aperta: "open," in which nerve has escaped the confines of bone, or occulta: "closed," characterized by retention of nerve within an open osseous canal (a benign, often radiographic finding most frequently located at the lumbosacral junction that affects up to ¼ of the normal population).
- Spina bifida aperta includes myelomeningocœle, which results from incomplete neurulation.
- Premature disjunction leads to migration of mesoderm into the neural tube, where the mesodermal cells are induced to form fat and ultimately a lipomyelomeningocœle.
- Failure of disjunction accounts for cutaneous signs of dysrrhaphism, such as hairy patch or sinus.
- Failure of ascension may result in tethered cord.
- Adhesion between endoderm and ectoderm results in split cord malformation, such as diastematomyelia or neurenteric cyst.
- Hyperkyphosis may be observed for the first 2 years for the normal development of sagittal contour during growth.
- Fusion of the spine before 5 years of age, that is, before ossification of the neurocentral junction, risks iatrogenic stenosis of the vertebral canal.
- Development of the vertebrae is associated most closely with that of the neuraxis, genitourinary system, and heart. As a result, an insult to one system may insult them all, and evaluation of one should include evaluation of the others.

EVALUATION

History and Physical Examination

Sit down, and take some time to gain the confidence of the child (and family): the spine is a site of awkwardness and vulnerability. Examine the whole patient. Is this an isolated problem of the spine, or part of a generalized disorder? Look at other systems, such as the skin, where *café au lait* spots suggest neurofibromatosis or a midline stigma points to dysrrhaphism.

History Directly and independently include the child, who may express a different complaint from parent or referring physician. For example, the child may be more concerned about the appearance of spine deformity than the pædiatrician, who may be focused on long-term consequences. Ask the family, which may reveal a genetic disorder. Because of the gravity of diagnosis, many other disciplines have become involved in spine care; be considerate as you navigate this world with patient and family.

Physical examination Develop a standard approach, including the following:

- Age. This is the most important patient characteristic in any disease.
- Other measure(s) of maturity [A].

Progression of spine deformity is proportional to growth.

- Gender. Certain conditions are more frequent or have a different natural history according to gender.
- Gait. This may be influenced by pain or neural function.
- Pain. This traditionally has been underestimated in children and in certain conditions (e.g., scoliosis).
- Palpation. In particular tenderness, and step-off (e.g., spondylolisthesis).
- Lower limb length discrepancy. This may produce an apparent or secondary spine deformity.
- Plane of deformity. Evaluate spine in three dimensions: coronal, transverse, and sagittal.
- Trunk asymmetry. Include the shoulder, scapula, ribs, breast, flank, iliac crest.
- Spine balance. Plumb line dropped from inion to natal cleft [B].
- Spine motion, including Adams forward bend test.
- Neural examination. Spine deformity may be primary or secondary to a neural lesion.
- Skin. Look for cutaneous signs of decompensation in the setting of deformity [B] and for a midline sign of dysrrhaphism.

Imaging

Röntgenogramme is the fundamental screening modality for spine deformity and for back pain. The latter also may be screened with scintigramme, which may detect a radiographically occult lesion [C].

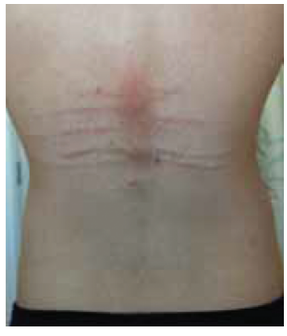
Scintigramme allows a dynamic or physiologic assessment of bone: the phosphorus attached to technetium (^{99m}Tc) medronic acid is taken up by bone undergoing turnover, as in an active spondylolysis that has potential to heal or in inflammation induced by infection.

Single photon emission computed tomography (SPECT) allows localization of scintigraphic uptake in a vertebra, for example, the pars interarticularis in spondylolysis.

Computed tomography (CT) gives the best detail of vertebral architecture [D]. It confirms spondylolysis. In neurofibromatosis, it reveals osseous erosion. In tumors, it defines margins. In complex deformity, it allows understanding of the three-dimensional shape of the spine and aids planning of operative treatment.

Magnetic resonance imaging (MRI) shows the soft tissue, in particular neural, intervertebral disc, muscle, ligament. Indications for MRI include abnormality on physical examination, such as weakness or upper motor neuron sign, as well as symptoms or signs of atypia such as thoracic hyperkyphosis, early age of presentation.

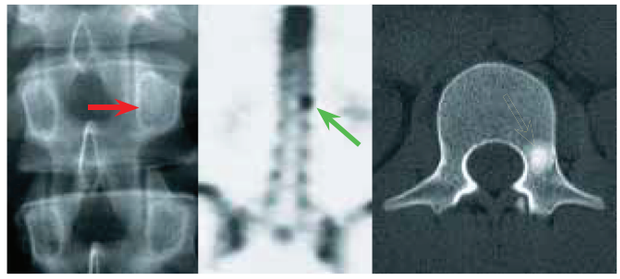
A Maturity Maturation is variable. Girls mature earlier and more predictably than boys. The ethnic spectrum is broad. These two boys are 16 years old.



B Skin decompensation The skin shows stretch marks and callus over an inflexible kyphosis. This is an objective sign that the deformity is significantly impacting (and will impact) the patient's function, such as sitting against the back of a chair.

Imaging method	Indication
Röntgenogramme	Osseous deformity
Oblique röntgenogramme	Lumbosacral spondylolysis
Ferguson view röntgenogramme	Spondylolysis Lumbosacral fusion
Scintigramme	Back pain—initial screening Osseous activity—spondylolysis, infection, tumor
SPECT (single photon emission CT)	Spondylolysis
CT (computed tomography)	Osseous deformity—operative planning Osseous architecture—fracture, spondylolysis, tumor
MRI (magnetic resonance imaging)	Soft tissue detail—neural lesion, disc, tumor, infection Osseous activity—spondylolysis, infection, tumor

C Imaging methods and their indications These are not a substitute for history and physical examination. Obtain judiciously: incidental findings may lead to unnecessary intervention.



D Osteoid osteoma of the spine Distortion of pedicle on röntgenogramme (*red*) was associated with focal increased uptake on scintigramme (*green*). CT scan showed a geographic, sclerotic lesion in the pedicle (*yellow*), guiding excision.

BACK PAIN

Prevalence

Drawing upon engineering and aviation, in medicine, back pain resembles a "black box." Up to 1/3 of children complain of back pain; the incidence increases in adults to >2/3 in adults.

Causes

In more than three-quarters of these, no cause can be identified; such children are said to have "overuse" or "growing" or idiopathic pain, in the same way they may complain of other sites in the skeleton [A]. The most common identifiable cause is spondylolysis, followed by deformity such as hyperlordosis. Less than 10% of children will present with a significant arthropathy, infection, neural lesion, or tumor, which should assure the physician to take a measured approach to evaluation and management.

CANDIF

One CANDIF (ferentiate) "good," or benign or idiopathic, from "bad" back pain, which should raise concern for an identifiable cause.

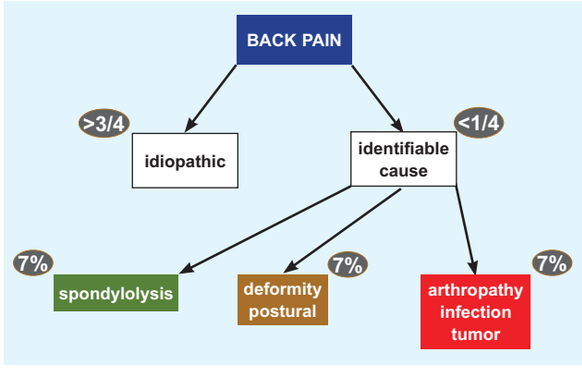
- Constant. Come and go pain typically is mechanical, from overuse.
- Associated symptoms or signs. This includes abnormality detected on physical examination, such as neural deficit or deformity.
- Nocturnal. Pain that arouses a child from sleep is more concerning than pain that interferes with falling asleep, which is typical of overuse pain.
- Duration. Follow-up a patient with pain <3 months. The physician may be at odds with the patient, who may regard duration >3 months to be evidence that something is amiss, since the pain has not resolved spontaneously. Educate the patient that a significant morbid process accounting for pain would manifest in other ways after several months or years.
- Intensity. Take note of pain that is worsening inexorably, as opposed to pain that may wax and wane within a stable range.
- Focal. Focal pain is more likely to have a structural cause. Pain that is diffuse, or that radiates in a nonanatomic distribution, is more typical of overuse.

Evaluation

History and physical examination are the foundation. Image judiciously: results may be misleading, they may raise anxiety unnecessarily, they may lead to unhelpful "treatment", and they add cost.

- Röntgenogramme. This is the first-line modality, particularly useful for spondylolysis and deformity. It also shows loss of height in discitis.
- Scintigraphy (including SPECT). For spondylolysis and occult process. This may be a screening tool for "serious" disease.
- MRI. For tumor, infection, or neural lesion (in particular in association with radiculopathy).
- CT. In the evaluation of back pain, this often serves a confirmatory rôle, such as to define spondylolysis or to characterize extent of osseous involvement by tumor.

Laboratory analysis is useful for infection, inflammatory arthritis, and blood tumors. C-reactive protein is the best measure of acute infection. Inflammatory arthritis may affect other joints (in particular knee and hand) and shows elevation of erythrocyte sedimentation rate. Complete blood count with differential screen for leukæmia.



A Back pain A simplified ætiologic algorithm.

Management

Idiopathic back pain is best managed according to general principles.

- Establish the diagnosis by exclusion.
- Educate and reassure the patient and family. Education may include a physiotherapist on modalities for back health.
- Activity modification. Avoid what makes it worse, such as a back pack that is >15% body weight.
- Light aerobic exercise, including participating in P.E.
- Weight reduction and control.
- Stretching. This may begin under the guidance of a physiotherapist but should continue as part of a more "holistic" approach, such as yoga.

Encourage "lifestyle" techniques that may be adopted by a child into adulthood, rather than medical interventions with which the child may not be compliant or which will be regarded as temporary. This is of particular importance because the majority of adolescents who complain of back pain and have a family history of such will experience back pain as adults.

Juvenile Idiopathic Arthritis

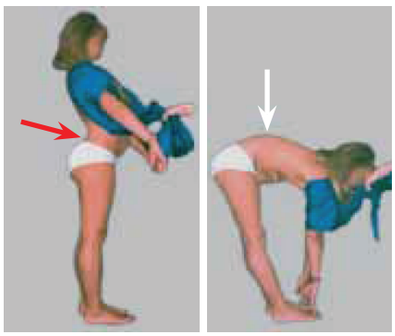
The majority of patients will be HLA-B27 positive (ANA and RF negative). Up to a third of patients with seronegative spondyloarthropathy will have a family history. Other characteristic signs include the following:

- Schober test for spine stiffness. Forward bend is associated with <4 cm of lumbar spine excursion due to syndesmophytes.
- Enthesitis, for example, at tendo-Achillis
- Cutaneous stigmata, for example, psoriasis
- Visceral involvement, for example, uveitis, enteritis

Management of back pain in this setting is secondary to management of the primary disease, by a rheumatologist.

Developmental Lordosis

Strain on facet joints in the hyperlordotic lumbar spine is a cause of back pain around the turn of the decade [B]. It is flexible, transient, and resolves spontaneously. Treatment consists of education and assurance, with postural training by antilordotic exercises.



B Physiologic lordosis of puberty This results from hyperflexibility of the lumbar spine as the trunk settles upon the pelvis.

DISCITIS

Discitis represents infection of the metaphysial region of a vertebra. By contrast with tumor, to which the intervertebral disc may be impervious, infection will traverse and erode the disc. The term places emphasis on the loss of disc height on röntgenogrammes [A].

Clinical Features

Presentation may be obscure and result in delayed diagnosis. It is one of the causes for refusal to walk in an infant. The preadolescent may complain primarily of abdominal pain; only in the second decade does the complaint reliably localize to the spine. Constitutional symptoms and signs, stiffness of the spine, and elevated inflammatory markers suggest the diagnosis. Paraspinous muscle splinting may be assessed in the prone position with the hips extended and the knees flexed: moving the pelvis from side to side causes a synchronous movement of the lumbar spine (Goldthwaite sign). Severe or untreated disease may be complicated by extension to paravertebral soft tissues, such as psoa abscess.

Imaging Röntgenogrammes take more than a week to show reduced disc height [A]. Scintigramme shows increased uptake of adjacent vertebrae at onset of disease [A] and will hasten diagnosis. MRI is indicated only if uncertainty persists, as it may introduce confusion with more morbid process such as tumor and lead to unnecessary intervention such as aspiration or biopsy.

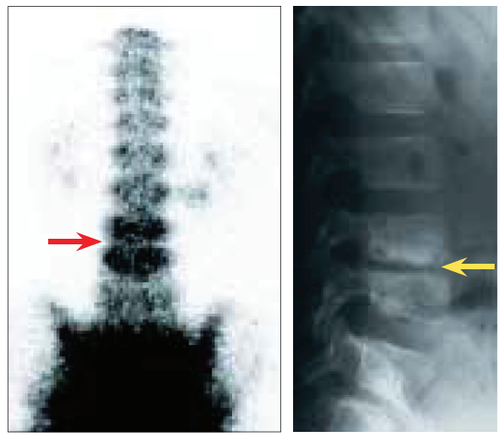
Management

If the child does not present with systemic illness, immobilization alone in brace or pantaloon cast may be sufficient [B], while the spine, well vascularized, in a healthy child recovers spontaneously. Antibiotics may be added to shorten disease duration and avoid complications; they are indicated unequivocally in systemic illness. Regimen follows general principles for musculoskeletal infection.

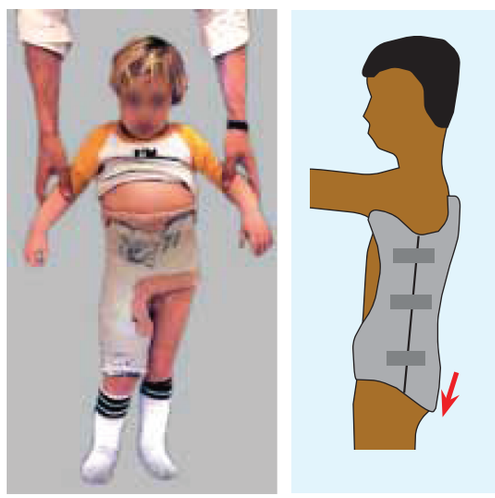
- Initially target gram-positive organisms.
- Venous antibiotics for systemic illness and until the disease is controlled.
- Conversion to oral antibiotics after clinical and laboratory response.
- Continuation of treatment until normalization of ESR.
- Change of antibiotics, further imaging, aspiration, or biopsy for atypical course or lack of response.

Prognosis

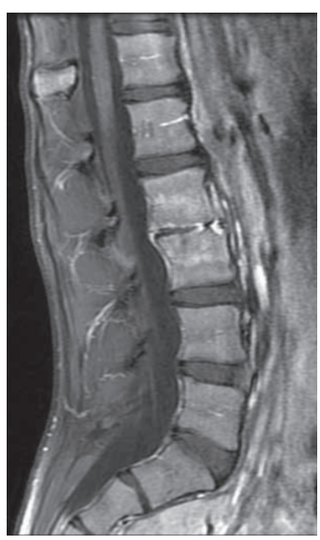
Disc recovery is inversely proportional to age at presentation [C]. Despite outcomes that include residual disc narrowing, sagittal plane deformity, and spontaneous fusion, function is not significantly impacted during childhood.



A L3-L4 discitis Scintigramme at presentation shows increased uptake (*red*). After 2 weeks, lateral röntgenogrammes shows narrowing of the disc (*yellow*).



B Immobilization reduces discomfort Most complete immobilization includes the back and one limb to immobilize the lumbosacral spine (**left**). A "long" (*red*) TLSO is better tolerated.



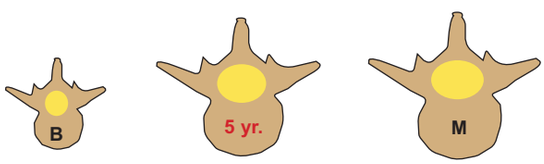
C Outcome of discitis Persistent loss of disc height and mild focal kyphosis 2 years after successful treatment.

Category	Comment
Idiopathic	Cause unknown Most common type Subdivided by age: infantile, juvenile, adolescent
Congenital	Vertebral anomaly
Neuromuscular	Upper neural lesions—cerebral palsy Lower neural lesions—spina bifida
	Myopathies—muscular dystrophy
Syndromic	Skeletal dysplasias Marfan syndrome Neurofibromatosis (of von Recklinghausen)
Secondary	Pain Lower limb length discrepancy

A Classification of scoliosis.

Abnormality	Comment
Genetic	Up to 75% concordance in monozygotic twins 10% in first–degree relatives Chromosome mapping to multiple loci
Hormonal	Growth leads to progression ↓ Melatonin ↑ Calmodulin
Neural	Dorsal column: abnormal proprioception Oculovestibular system: abnormal nystagmus to caloric testing cerebral cortex: abnormal evoked potentials ↑ Chiari malformations
	Myopathies—muscular dystrophy
Structural	Growth disturbance of anterior column leads to loss of sagittal contour until buckling lateralward and rotation
Serologic	Abnormality of platelets

B Causes of idiopathic scoliosis Scoliosis is multifactorial, and most cases are new spontaneous.



C Growth of the vertebral canal Volume doubles from birth (0) to 5 years of age, at which point volume is 95% of maturity (M).

Characteristic	Comment
Specific and sensitive	Nonspecific: normal variation in population Too sensitive: referral of untreated curves
Applied to at risk population	Applied to all children
Cost:benefit ratio	~33% increased cost of management ~0.2% indication for bracing
Early intervention alters natural history	Treatment effect of bracing difficult to quantify Manipulative and alternative modalities debatable

D Screening of scoliosis Essential characteristics of an effective screening test. Screening for scoliosis is recommended twice for girls and once for boys in the first half of the second decade.

SCOLIOSIS

Scoliosis is ancient. It is named and discussed extensively in the Hippocratic Corpus, including the scamnum for correction. It permeates Western literature and art: Shakespeare recounts how the Duke of Gloucester lamented his back, "Where sits deformity to mock my body." While it is defined as angulation in the coronal plane >10 degrees, scoliosis is a three-dimensional deformity including rotation in the transverse plane and alteration of sagittal alignment. It may be divided into causative categories [A].

IDIOPATHIC SCOLIOSIS

Idiopathic refers to "disease" (Greek παθος) that arises on its "own" (Greek ιδιος), that is, cause unknown. Despite the name, several causes have been speculated [B]. Idiopathic scoliosis is the most common deformity of the back. It affects approximately 1% of children, approximately 90% of whom never require active treatment. Progression is proportional to growth, necessitating follow-up through maturity. It is defined as ≥5 degrees, and averages 1 degree *per mensem* during growth and 1 degree *per annum* after maturity for curves >50 degrees.

Evaluation

Essential to evaluation is exclusion of other cause, determination of growth potential, characterization of curve (including magnitude and flexibility), estimating risk of progression, and by extension likelihood of treatment.

History

- Age. This is one measure of growth, which correlates with progression of deformity. Children in the first decade have a higher incidence of neuraxis lesion. Fusion arrests growth. Significant growth of the spinal canal concludes by 5 years of age [C], before which focal operation for vertebral anomaly should be postponed if possible in order to avoid stenosis. Clinically significant growth of the thoracic spine concludes as the child transitions into the second decade, by which point the 50% threshold of thoracic volume has been traversed and before which fusion will have a deleterious effect on pulmonary function.
- Gender. Scoliosis affects both genders equally, but progression in girls is approximately five times than in boys.
- Other measures of growth. Growth amplifies deformity. Girls grow fastest in the 7 months before menarche and stop significant growth 2 years postmenarche. Menarche is variable, including according to activity and ethnicity. It occurs after peak growth velocity, that is, after the curve acceleration phase, the time of greatest risk of progression. It applies to only half of children. Boys grow later and less predictably, in particular a greater variability in cessation of growth.
- Complaints. Pain traditionally has been underestimated in children, and in scoliosis in particular, where it affects 2/3 (twice the background rate). This may be related to fatigue of muscles attempting to rectify the spine. On the other hand, abnormal pain or other complaint (e.g., neural change) may be a sign that the scoliosis is not idiopathic.
- Family. Such a history may suggest greater risk of progression. A susceptibility to develop scoliosis has been linked to the gene GPR126 on chromosome 6, which is highly expressed in cartilage and is associated with height and trunk growth.
- Ethnicity. Caucasians are most affected.
- Screening [D]. This developed from the observation that the thoracic spine and its deformity may be seen on röntgenogramme of the chest obtained during tuberculosis screening. It no longer is practised in the United Kingdom or Canada, and in only half the United States. Greater than 99% of referrals consist of normal variants, scoliosis <10 degrees ("schooliosis") or <25 degrees (no treatment).

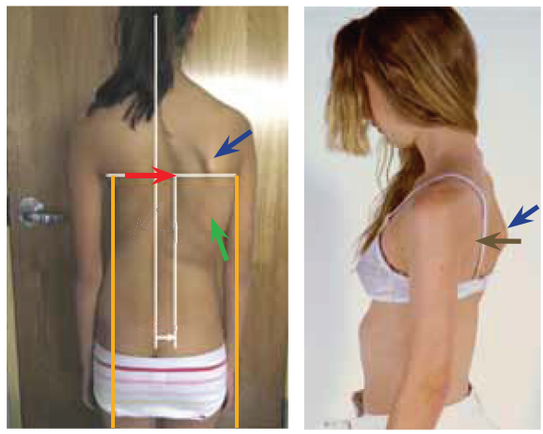
Physical examination Develop a standard approach, including the following:

- Gait. This may indicate pain (antalgic), a generalized disease or associated findings (e.g., ataxia).
- Maturity. Tanner scale of secondary sex characteristics has not been bettered by any other system [A]. It is the simplest, fastest, and cheapest method. It may be performed in office (not off-site) and is least morbid (e.g., no radiation from röntgenogrammes). Progression is greatest in the first two stages. Stage 3 coincides with menarche. Stage 4 marks end of significant spine growth. Plane of deformity. Examine coronal (from back and front), transverse (forward bend), and sagittal (from side) planes.
- Palpation. For tenderness, step-off, crepitus, and including the paraspinous soft tissues.
- Trunk asymmetry, including the shoulder, scapula, rib, breast, flank, iliac crest. Asymmetry may be a secondary sign of spine deformity, or it may be a normal variant (5 to 15 mm). Correlate trunk asymmetry with other signs, including direct palpation of the spinous processes. Coronal curvature in the chest elevates the shoulder and below the chest will indent the flank [B]. Rotation will push the scapula, breast, and ribs out, which may be accentuated by sagittal plane deformity such as loss of thoracic kyphosis. Rotation is proportional to risk of deformity progression. Angle of trunk rotation may be measured on forward bending with a scoliometer [C].
- Trunk shift [B]. Project the surface of thorax vertically relative to the iliac crests. This carries an increased risk of deformity progression.
- Spine balance [B]. Even in the setting of trunk shift, a child with idiopathic scoliosis will bring the head back toward midline to balance it on the pelvis. Imbalance may be a subtle indicator of neural abnormality.
- Spine motion, including Adams forward bend test. This is performed ideally from the front (not the back) so that the entire coronal contour may be evaluated by varying the degree of bend without changing the examiner's position, as well as from the side. Asymmetry or limitation may be a sign of pain, neural lesion, or associated abnormality such as hamstring tightness.
- Neural examination, including central signs such as altered gait, lower limb hyperreflexia, clonus, and abnormal abdominal reflex [D]. The latter is a superficial reflex for the thoracic spinal cord. Presence or absence is normal; asymmetry suggests a spinal cord lesion. It is explained evolutionarily as a mechanism to thicken the abdominal wall to protect subjacent viscera.
- Other deformity. For example, in cavus, the arch may point to the primary problem (proximalward to the spinal cord).
- Skin. Note the apothegm that "the skin tells the story." Look for cutaneous signs of decompensation in the setting of deformity, such as stretch marks (rapidity of change in back shape) and callus (in response to unrelieved pressure due to lack of flexibility). Look for a midline stigma of underlying neural lesion, such as hairy patch or sinus. The former may be regarded as an early functional outcome sign, which may influence intervention. The latter is an indication for magnetic resonance imaging.
- Laxity. Signs of hyperlaxity may be present, including elbow extension <0 degrees, placement of the thumb on volar forearm, extension of metacarpophalangeal joints >90 degrees, extension of the knee <0 degrees, flexion of the ankle >30 degrees, and the ability to place palms flat on floor during forward bending with knees straight. As an example, an increased incidence of private variants in fibrillin-1 gene, causing nonsynonymous amino acid substitutions, has been found in a subset (4%) of children with adolescent idiopathic scoliosis who present with more severe and more progressive curves, as well as hyperlaxity without other sign of Marfan syndrome (*cf.* Syndromes chapter).

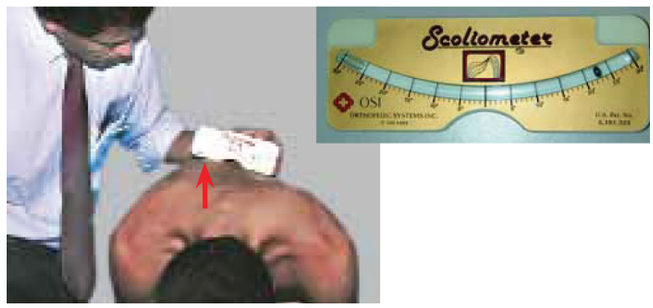
Stage	Girl	Boy
1	Prepubertal	
2	Breast bud	Sparse + downy hair scrotal skin texture + pigment
3	Breast elevation subjacent contour	Dense + coarse hair
4	Areolar mound	Triangulation of hair
5	Adult	Hair extends to thighs

P

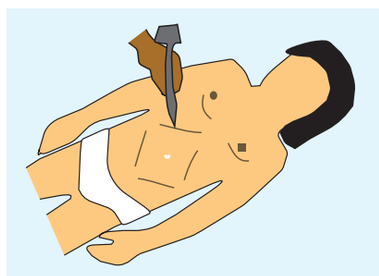
A Tanner scale consists of five stages Distinctive and practical features. P: progression of spine deformity.



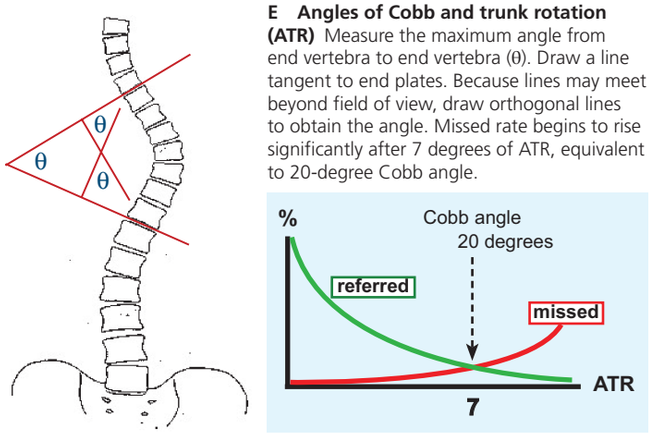
B Trunk asymmetry Apex dextrad curvature (*red*) produces a right posterior thoracic prominence (*green*) and scapular protraction (*blue*). The trunk shifts dextrad (*orange*) relative to the pelvis. Despite trunk shift, balance is maintained, as evidenced by a plumb line (*white*) dropped from inion to natal cleft. Sagittal alignment is changed as the thoracic spine rotates out of kyphosis (*brown*).



C Trunk inclination The scoliometer is grooved in its center to accommodate spinous processes as the examiner runs it continually along the entire spine. Observe inclination of a silver ball along a curvilinear slot marked in degrees.



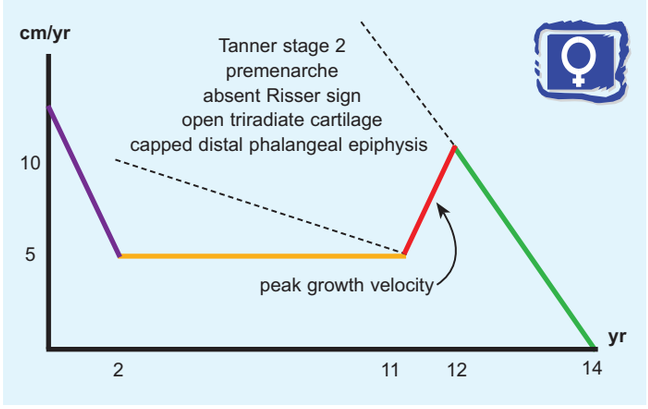
D Abdominal reflex Stroke each quadrant of the relaxed abdomen. Reflex contraction of abdominal muscles draws umbilicus toward the stimulation.



E Angles of Cobb and trunk rotation (ATR) Measure the maximum angle from end vertebra to end vertebra (θ). Draw a line tangent to end plates. Because lines may meet beyond field of view, draw orthogonal lines to obtain the angle. Missed rate begins to rise significantly after 7 degrees of ATR, equivalent to 20-degree Cobb angle.

Sign	Comment
Risser	Divide iliac apophysis into quarters 1–4; fusion of apophysis is 5. Most progression occurs before appearance of iliac apophysis.
Triradiate cartilage	Closed marks cessation of spine growth. Open allows no differentiation of maturation stage.
Distal phalangeal epiphysis	Capping of distal phalanges by epiphysis marks beginning of curve acceleration phase. Fusion marks end of significant progression. Fusion coincides with menarche, triradiate closure, absent Risser.
Tanner-Whitehouse RUS	Complex method based upon multiple sites in radius, ulna, and small bones of the hand. Selection of distal phalangeal epiphysis is a simplification.

F Radiographic signs of maturity This is in constant flux. Risser noted that the iliac apophysis could be seen serendipitously at the bottom of spine röntgenogrammes. Evaluation of the distal phalangeal epiphysis is the current favorite method.



G Growth curve for a girl Note the features associated with peak growth velocity, during which the spine goes through the curve acceleration phase.

H Vertebral rotation This may be graded according to location of spinous process and of pedicles as viewed in coronal plane. Beyond d, or grade ++++, spinous process projects beyond vertebral body. In grade +++ and ++++, concave pedicle has disappeared.

Imaging Röntgenogrammes are the foundation. Measure curve magnitude in degrees after the method of Cobb [E]. Note that measurement error is approximately 5 degrees, which has not been improved by digital imaging. In order to reduce referrals for which orthopædic management is not indicated, a minimum of 7 degrees on scoliometer has been identified to correspond with a Cobb angle of 20 degrees.

Obtain full-length röntgenogrammes to determine spine balance. Sagittal balance is represented by a vertical line between midpoint of C7 and superoposterior corner of S1. In the coronal plane, this vertical line should pass through S1 spinous process. Imbalance is measured in centimeters: ≤2 cm is within normal range.

Posteroanterior projection reduces breast exposure to radiation, which, after reflection of its majority off the skin, declines with travel through tissue, reaching the breasts last. Shield the breasts when it does not impact analysis of the deformity. Include the clavicles to assess shoulder height, which influences operative technique and outcomes. Include the iliac apophysis and triradiate cartilages to aid determination of maturity [F]. Add röntgenogramme of the hand for evaluation of the distal phalangeal epiphysis. Note that calculation of maturity is nuanced and multifactorial [G]. The iliac crests also allow assessment of limb length discrepancy.

Röntgenogrammes identify stable and neutral vertebrae. The former is bisected by the central sacral line. The latter show no rotation, defined as symmetric pedicles and midline spinous process [H]. Rotation is a risk factor for progression and must be considered in addition to curve magnitude. The apical vertebra is least stable and most rotated. Limit fusion to vertebrae that at least are crossed by the central sacral line. Pedicle screws have promoted fusion before reaching the neutral vertebra, because of direct vertebral rotation.

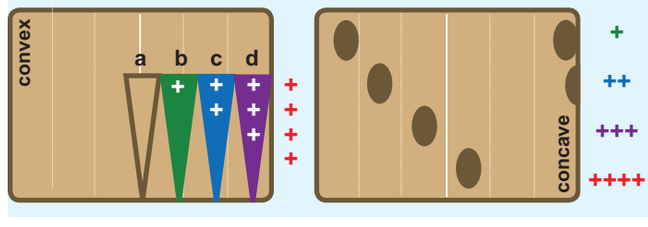
Determine flexibility on röntgenogrammes. Prone posteroanterior view simulates the operating room. Bending views will show opposite inclination at flexible discs, which may be spared fusion. Bending views also show whether the planned lowest instrumented vertebra can become <15 degrees oblique (greater associated with progression) and enters the stable zone.

Computed tomography is useful for complex osseous architecture. In idiopathic scoliosis, this applies to severe deformity rather than vertebral anomaly, which is not a feature. CT provides detail in the postoperative spine, for example, for pseudarthrosis. CT with myelogramme evaluates the instrumented spine for vertebral canal encroachment, for example, by implants in the setting of neural compromise. For other neurologic evaluation, myelography has been replaced by magnetic resonance imaging. Pedicle dimensions may be measured on CT to aid operative planning.

Magnetic resonance imaging is indicated when an abnormality of the neuraxis is suspected in the so-called "idiopathic" setting. Principal risk factors are as follows:

- abnormal physical examination
- age <10 years
- kyphosis >40 degrees for thoracic curves

There is debate about association between neural lesion and other signs of atypia, such as apex left thoracic curve, rapid progression (moderate estimate >3 degrees *per mensem*), male gender, dystrophic curve (short and sharp, Harrington factor >5 degrees/segment), or otherwise unusual appearance curve (e.g., long and sweeping akin to a neuromuscular presentation).



Classification

Morphologic Curves are divided into six types [I]. Main refers to a structural thoracic curve in the setting of an upper structural thoracic curve. Major refers to a structural curve when thoracic and lumbar regions are affected independently, and the largest of two or three structural curves. Nonstructural refers to a compensatory curve that may be spared fusion.

- Main thoracic. Structural thoracic curve.
- Double thoracic. Two structural thoracic curves.
- Double major. Structural thoracic + lumbar curves.
- Triple major. Two thoracic + thoracolumbar/lumbar structural curves.
- Thoracolumbar or lumbar curve.
- Thoracolumbar or lumbar + thoracic structural curves.

Thoracic curves are modified based upon kyphosis:

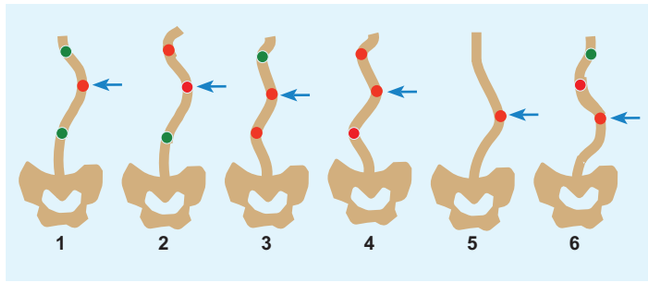
- − hypo-, defined as <10 degrees
- N, normal
- + hyper-, defined as >40 degrees

Lumbar curves are modified based upon whether central sacral line:

- A bisects the apical pedicles.
- B touches apical vertebral pedicles.
- C is medial to the apical vertebra.

Classification applies order based upon pattern recognition, but variability argues against dogma. An alternative is the descriptive approach.

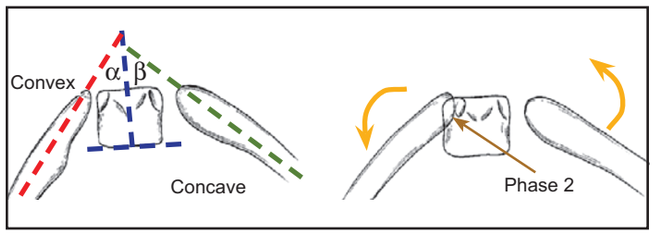
- Curve magnitude. This is defined in the coronal plane. Sagittal plane is most predictive of functional outcome. Appearance is most influenced by rotation in the transverse plane.
- Type of curve. This is a geographic assessment based upon location of apical vertebra: thoracic, which may be single or double (termed main and upper); lumbar; combined thoracic and lumbar; and thoracolumbar (T12-L1 apex).
- Flexibility of curve. This is based upon intervertebral disc inclination. Discs that are inflexible are part of a structural curve. Flexible discs are part of what may be a compensatory curve that rectifies or does not progress upon treatment of the structural curve and as such may be spared direct treatment. Curves flexible to <25 degrees also may be excluded.
- Relation to midline. A curve that crosses the midline is structural. A curve that is returning to the midline from a structural curve, for example, the lumbar spine returning to the pelvis, represents compensation by the flexible spine.
- Sagittal contour. Thoracic curves may be associated with hypokyphosis, lordosis, or hyperkyphosis. This is more difficult to correct than coronal curvature. Increased power of instrumentation exposes anterior overgrowth and loss of thoracic kyphosis with increasing coronal correction. Sagittal contour is essential to determination of end instrumented vertebrae. Sagittal balance correlates more directly with functional outcome than coronal balance. Hyperkyphosis of the thoracic spine may be the most sensitive predictor of associated neural lesion.
- Coronal and sagittal balance. Imbalance is associated with poor functional outcome, most in the sagittal plane.
- Rotation in the transverse plane. This is predictive of progression, in addition to curve magnitude. It may serve as an alert that a flexible curve may not be compensatory.
- Atypia. This may be an indication for MRI of the neuraxis.



I Classification of scoliosis—morphologic Fusion is indicated for structural curve, represented by *red*. *Green* signifies a compensatory curve that may be spared. Main and major curves are structural and largest (*blue*).

Type	Comment
Infantile <3 years	• <5% • Boys 2 times girls • Most thoracic curves apex left • Extrinsic cause suggested by associated "packaging" signs including torticollis, plagiocephaly, hip dysplasia • Resolving or progressive—calculate Rib–Vertebral Angle Difference and Phase
Juvenile 3–10 years	• High rate of associated neural lesion ≤1/3 • High rate of progression ≤2/3
Adolescent >10 years	• >80% of all cases

J Classification of scoliosis—clinical Three types are distinguished according to age.



K Rib–vertebral angle difference (RVAD) and phase Determine angle subtended by the axis of the rib (*red, green*) and vertical axis of the body of the apical vertebra (*blue*). RVAD = α – β. The difference results from hinging of the ribs as the spine curves away from the convex set toward the convex set. Greater than 80% of curves with RVAD <20 degrees will not progress. RVAD >20 degrees is associated with >80% progression.

On the right, the curve is in phase 2, which is defined as overlap of the convex rib head with apical vertebra (*brown*) and in which progression is certain. Such a curve is termed "progressive" type. Phase 2 is a manifestation of rotation displacing the apical vertebra anterior to the apical rib so far that the overlap becomes radiographically visible even in the incompletely ossified skeleton. Phase 1, exemplified by the figure on the left, is defined as no overlap: such curves do not progress and are known as "resolving."

Clinical Idiopathic scoliosis may be classified according to age [J].

Infantile This is rare and distinct from juvenile, which may be considered a graver form of adolescent. It may be divided into two types: resolving and progressive. The former is unique in potential for spontaneous correction, which along with associated packaging signs suggest a unique cause, at least for this subgroup. Progression may be predicted by the rib–vertebra angle difference and phase [K].

Juvenile This is distinguished by a high rate of associated neural lesion (up to 1/3) and the highest rate of progression (up to 2/3). The former is an indication for MRI of the neuraxis. The latter adds urgency: more patients will require treatment, earlier and more aggressively.

Natural History

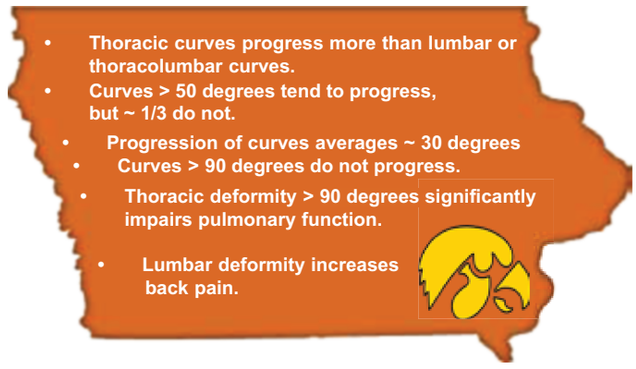
Understanding of natural history is founded in the classic studies from the University of Iowa [A]. While they are revered due to their 50-year follow-up, which is unrivaled in orthopædic surgery, there is not absolute clarity.

Progression The principal determinants of progression are growth and curve magnitude. Risk of progression to surgery is what patients and parents most want to know. Progression has been the most consistent outcome measure in the literature of idiopathic scoliosis. Deformity has the potential to progress during growth. All juvenile idiopathic scoliosis >20 degrees progresses [B]. If the threshold of 30 degrees is crossed in the first decade, scoliosis will progress beyond 50 degrees (to fusion) despite nonoperative treatment (bracing).

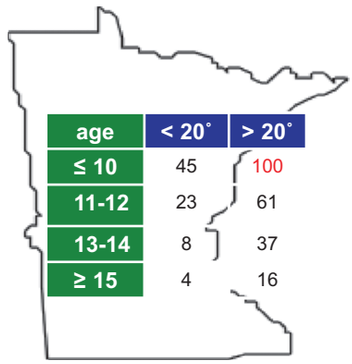
Pulmonary function This is the principal concern of thoracic scoliosis. Curve magnitude, location, and rotation conspire to produce pulmonary disease, of which essential features are spinal encroachment upon thoracic viscera, reduced thoracic dimensions, and disruption of mechanics of ribs and respiratory muscles (intercostal and diaphragm). Alveolar and pulmonary vascular hyperplasia cedes to hypertrophy toward the end of the first decade, as growth moves from increasing numbers to increasing size of preexisting structure.

Diminution in pulmonary function testing, in particular total lung capacity and forced vital capacity as measures of restrictive lung disease, may be observed at 60 degrees of scoliosis. Clinically apparent pulmonary dysfunction, such as dyspnœa on exertion and sleep hypopnœa or apnœa, occurs at 90 degrees. Spine fusion with instrumented correction of deformity in idiopathic scoliosis may arrest pulmonary decline or even improve function by up to 10%. By contrast, costal osteotomy or thoracotomy, for example, for anterior approach, may reduce pulmonary function by a similar amount, although the effect may be temporary.

Back pain This is the principal concern of lumbar scoliosis. Back pain affects 2/3 of children with scoliosis. It is related to fatigue of muscles resisting progressive deformity and imbalance. Spine fusion reduces pain but not for every patient and not always completely. Long-term pain is related to degenerative changes at facet joints and intervertebral discs that are malaligned (abnormal force vector) or spared fusion (force concentration). Untreated severe scoliosis is associated with increased pain quantitatively; for example, in the Iowa experience, it occurred "frequently or daily" 50% more than controls. There is a qualitative increase in back pain as well, such as less physically demanding employment and more frequent disability claims. Controversy and lack of further granularity remain, in large part due to the difficulty of measuring an effect when back pain is so prevalent in the general population.



A Natural history of scoliosis The Iowa experience.



B Progression is influenced by growth and curve magnitude The Minnesota experience drew attention to the at-risk group of juvenile scoliosis with significant curvature. This has been refined to progression in <10 years of age >30 degrees to severe deformity and fusion.

Appearance The Iowa studies noted that untreated patients with scoliosis were more self-conscious of appearance. For surgically treated patients, the Spinal Appearance Questionnaire and the Scoliosis Research Society Appearance domain show large effect size, demonstrating that they are sensitive enough to measure treatment effects. By contrast, Scoliosis Research Society Activity, Pain and Mental domains show small effect sizes and standardized response means, demonstrating an immeasurable effect on these functional outcomes by operation. Appearance matters, both to patients and because it is the factor most effectively treated.

Management

The pillars of management are education, observation, bracing, and surgery. Education is all encompassing. For example, educate parents that "doing nothing" by observation is a legitimate form of treatment, as most curves do not progress significantly and as such do not cause long-term disability. Patient characteristics that drive management are curve magnitude and age.

Observation 10 to 30 degrees Obtain röntgenogrammes every 6 months through maturity.

Bracing 30 to 50 degrees Institute at 25 degrees for the high-risk child, in particular in the first decade. This is the standard of care for the growing skeleton. The principle that underpins bracing is application of contralateral forces through the ribs or flank against the limbs of the spine on either side of the apex. The multiplicity of braces and regimens exemplifies the axiom that permeates surgery: "Where there is variability there is uncertainty." The Boston brace as well as full-time wear (as opposed to night time) have the broadest experience [A]. While a curve may be corrected in a brace, success of bracing is defined as arrest of progression. Factors associated with brace failure include thoracic lordosis, high thoracic apex, obesity and poor compliance, as well as curve >30 degrees in the first decade.

- Custom brace fabrication to the patient and röntgenogramme.
- Wean to full-time wear, defined as >20 hours *per diem*.
- Doff brace day before serial follow-up röntgenogrammes.
- Don brace until maturity.

Such treatment at such a critical developmental period carries a psychosocial cost that, while difficult to measure (unlike, e.g., angle of scoliosis), is real and may endure after cessation of active care.

Casting 30 to 50 degrees This is indicated for infantile idiopathic scoliosis. Serial casts are applied every 2 to 4 months from 2 to 4 years of age under general anæsthesia. Reduction of spine deformity is based upon derotation as opposed to direct force. Distinctive features of the cast include minimal padding, stable purchase on the pelvis as the foundation, and windows to allow for dynamic correction. Success is defined as continual curve correction to resolution (cure), arrest of progression that is maintained by casting followed by bracing, or delay of operation. Assessment of efficacy is impacted by limited experience given rarity of condition, uncertainty about natural history, as well as significant intra- and interobserver error in RVAD and phase determination.

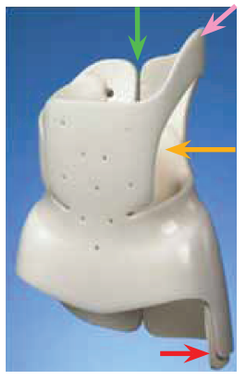
Operation 30 to 50 degrees fusionless Patients in the first decade with a curve >30 degrees, and premenarcheal girls with curves >40 degrees, have a high likelihood of progression to fusion despite bracing. In the former group, fusion in the first decade limits growth of the spine and thereby reduces thoracic volume enough to negatively impact clinical pulmonary function. The latter group is approaching the magnitude for fusion with significant growth remaining, including the curve acceleration phase before menarche. For these groups, operation is an alternative. The goal of fusionless surgery is to tether the convexity of a curve, which is growing more rapidly. This may arrest progression (as an internal brace) or allow for correction with concave growth, at which point the tether may removed. The latter distinguishes this modality from bracing.

Growth modulation by physial tethering is well established, in particular at the distal femur and proximal tibia for *genu valgum et varum*. Application of the technique in the spine differs in that implants are placed between bones across a motion segment, which theoretically increases the risk of failure. There are two types of fusionless operations, performed by telescope or open on the anterior spine, which may restore kyphosis in the thoracic region but has the potential to reduce lordosis in the lumbar region. As with *genu valgum et varum*, implant removal is indicated if there is anatomic correction.

VERTEBRAL BODY STAPLING [B] This is indicated for juvenile idiopathic scoliosis 30 to 39 degrees. Vertebral end plates and intervertebral discs are spanned by a two- or four-pronged NiTiNOL staple, which is inserted ice-cold and with straight tines that return to the original curved shape as the memory metal warms to body temperature. The innovation of curved NiTiNOL has eliminated the complication of staple back out. The procedure has the advantage of sparing the intercostal vessels, which may be mobilized away during insertion of, and rest between, staples.

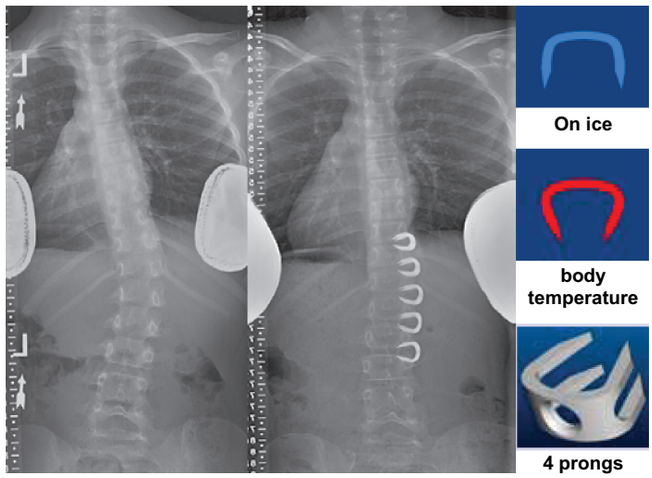
SCREW AND CORD [C] The ideal candidate is a child with a curve 40 to 60 degrees and having more than 2 years of growth remaining. The latter may be estimated as premenarche in a girl and no facial hair in a boy, or according to the arithmetic method as under 12 years for a girl and under 14 years for a boy. Other measures of significant growth potential include open triradiate cartilage and no Risser sign.

Hydroxyapatite coating of vertebral body screws is designed to reduce pullout. Screw insertion requires intercostal vessels ligation. Washers support screw fixation. A flexible polyester cord fixed to the screws is tensioned to effect a partial reduction and resist convexity growth.

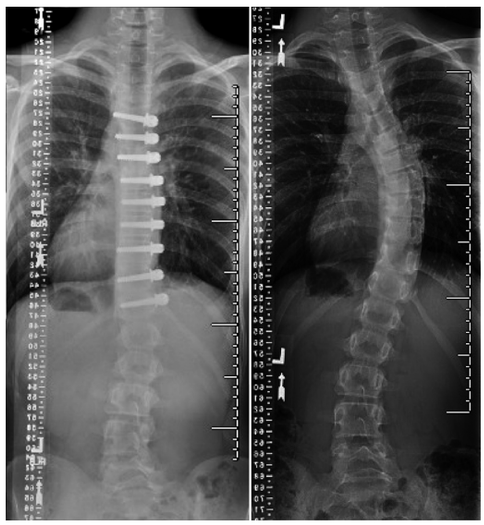


A Boston brace Thoracolumbosacral orthotic (TLSO) is custom fabricated based upon patient shape and röntgenogramme from hard plastic lined by dense foam with pads placed according to morphology of curve. It opens in the back (*green*), is indented to fit the waist above the iliac crests, and includes opening (*orange*) and greater trochanteric extension (*red*) to allow three-point bend. Note that the highest extension is axillary (*pink*) reducing effectiveness for curves with apex above T6.

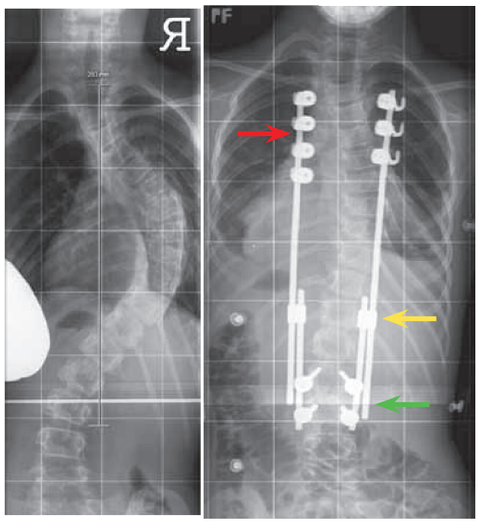
Wean to full-time wear, defined as >20 hours per diem. Doff brace day before serial follow-up röntgenogrammes. Don brace until maturity.



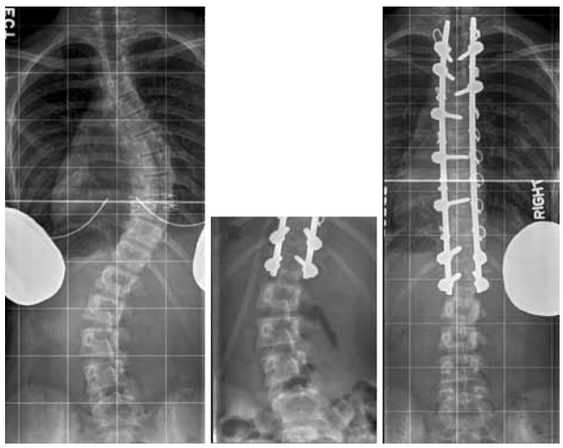
B Vertebral body stapling Correction in 7-year-old girl with 32-degree thoracolumbar curve. NiTiNOL (NIckel–TItanium Naval Ordnance Laboratory), a memory metal, has eliminated implant back out.



C Screw and cord Correction by tethering the convex side of the spine, which is growing more rapidly. While overall correction is complete, segmental correction varies because segmental growth – normal and asymmetric – is variable.



D Growing rods A 7-year-old girl with 75-degree thoracic curve. Two rods and pedicle screws at a lower end fusion (*green*) improve stability. Modification of upper anchors to the ribs (*red*) increases number of anchors at flexible sites and reduces spine dissection, potentially reducing implant failure and fibrosis. Use of standard implants lengthened at side–side connectors (*yellow*) simplifies index procedure and procedure at maturity.



E Selective fusion The lumbar curve was spared because it was flexible <25 degrees. Early residual curvature spontaneously corrected by 1 year. Transverse process wires at the top instrumented vertebra avoid interference with superjacent facet joints and midline dissection. Screws at the ends reduce construct pullout. Apical screws aid reduction of deformity. There are two anchors at every level, including transverse process wires that are safe and cheap.

Operation >50-degree fusionless Indication for fusionless operation is juvenile scoliosis having curve magnitude too great for vertebral body stapling or screw and cable.

Growing rods [D] This is the original fusionless operation. The goal is deformity correction and its maintenance while allowing growth of the spine. It is mechanically disadvantaged: end fixation with intervening motion fails approximately 15% of the time. Advances to enhance stability include the following:

- Two contoured rods with or without cross-links, compared with one straight rod
- End fusion of two vertebrae, compared with anchoring to a single vertebra at each end
- Pedicle screws, compared with laminar hooks

Unlike vertebral body stapling or screw and cable, which rely on spontaneous growth of the concave spine, growing rods lengthen the spine by active distraction. It is biologically disadvantaged: after implantation and initial distraction (which achieves the most elongation), the rods are distracted every 4 months through the same incision. Repeated opening of a scarred wound results in an infection rate up to 10%. Multiple procedures, prolonged instrumentation, and infection lead to fibrosis, possible heterotopic ossification, and stiffening of the spine. At maturity, or as close as is reasonable based upon patient tolerance of multiple procedures and complications, definitive fusion is performed.

Operation >50-degree fusion Indication for fusion is risk of progression regardless of growth, as the spine decompensates into advancing deformity. The goals of operation are safety, fusion, and correction.

- Safety. "To do no harm" is the primary ethical imperative. Pertinent factors are discussed under *Complications*.
- Fusion. This is augmented by osseous graft and decortication.
  — Autogenous graft from iliac crest adds time and pain (although the latter may not be clinically significant compared with overall pain of operation) and may add a separate incision. Autogenous graft also may be obtained by harvest of spinous processes, and from ribs, as part of thoracoplasty. Allogenous graft is as effective as autogenous, saves time but adds cost (although the latter may not be significant relative to overall cost of operation, including implants) and does not require second incision. Bone graft substitutes and adjuvants are not indicated in primary operation.
  — Decortication simulates fracture to stimulate adjacent vertebrae to heal to one another. This may be performed with burr or gouge. The latter lifts strips of bone from transverse processes and laminæ that may bridge vertebrae without the heat of a motorized tool.
  — Facet joint excision aids fusion and correction.
  — Selective fusion maximizes remaining motion of the spine and reduces risk. As in the assessment of maturity, selection of fusion levels cannot rely on any single factor nor can it be formulaic; rather, it is a synthesis of several concepts [E, F].

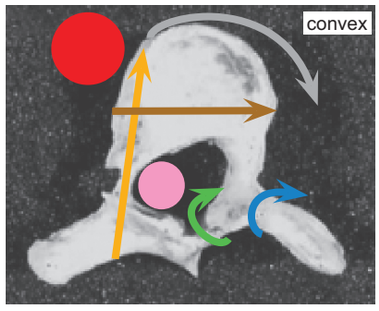
Principle	Comment
Structural curve	***Fuse.***
Compensatory curve	Bending röntgenogrammes show opposite bending of intervertebral discs and curve <25 degrees. ***Spare.***
Stable vertebra	Each end vertebra must be crossed by center sacral line.
Neutral vertebra	Pedicle screw instrumentation may shift this by vertebral derotation to limit fusion.
Sagittal contour	End vertebrae must be out of kyphosis.
Rotation	May represent a structural component independent of coronal magnitude or flexibility
Shoulder height	Extend fusion proximalward in order to pull down a shoulder elevated opposite apex of thoracic curve.
Thoracolumbar junction	T12 lowest instrumented vertebra associated with increased risk of junctional kyphosis–stop at L1
Obliquity	Obliquity of lowest instrumented vertebra >15 degrees may be associated with progression and poor long term outcome.

F Selection of fusion levels

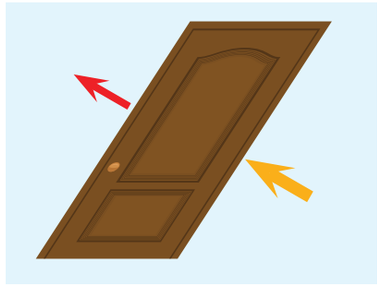
- Correction. This is achieved by instrumentation, osteotomy, and vertebral resection.
  — Akin to cement technique in arthroplasty, posterior instrumentation has developed in generations [G]. Harrington introduced a rod with lamina hooks at end ratchets for reduction of scoliosis by distraction. Luque used lamina wires for segmental fixation and translation. Wires also may be applied to spinous and transverse processes. Cotrel and Dubousset reduced the spine by rod rotation applied through lamina and pedicle hooks and added cross-links between two rods to enhance construct stability. Pedicle screws are most stable, which reduces anchor failure at ends of fusion to; in addition, they enable correction of the third dimension of deformity by vertebral derotation, both at the apex to improve appearance and at the ends to shift the neutral vertebra in order to minimize fusion and maximize preservation of motion.
  — While pedicle screws are the most stable anchor, correction is more related to ante-operative flexibility and number of anchors. Optimal correction for ante-operative flexibility is ≥ 1.5 anchors/level.
  — In scoliosis correction, Ponte modification of Smith-Petersen osteotomy enhances restoration of lumbar lordosis. Pedicle subtraction osteotomy includes the pedicle as it extends across the three columns of the spine to correct focal kyphosis in revision operation. Vertebral resection is reserved for severe and stiff deformity and for congenital disease.
  — It remains debated whether rib osteotomy, for example, thoracoplasty to reduce posterior prominence and improve appearance, and rib resection, for example, in concavity to aid curve correction, improve functional outcome, particularly in light of potential deleterious impact on pulmonary function.

Posterior approach to the spine is the oldest, most familiar and most versatile. Posterior osteotomies have equalized levels of fusion with anterior approach. Posterior osteotomies have reduced the need for anterior operation to loosen a stiff curve. Posterior circumferential dissection makes vertebral resection possible without, and even easier than, adding an anterior approach. Anterior approach has a rôle.

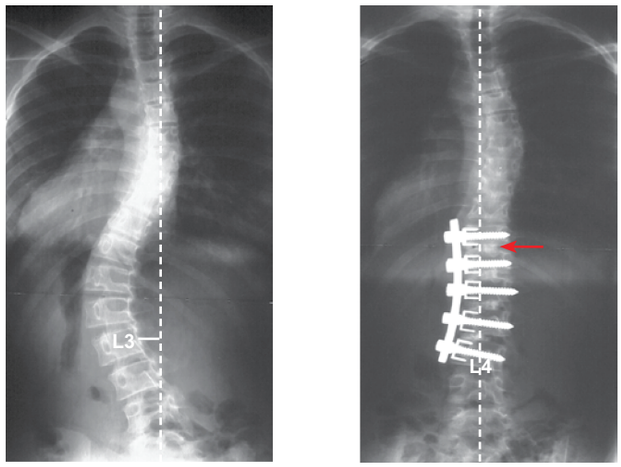
- It may enable better correction and even overcorrection to limit fusion levels, as it allows pushing against the deformity in contrast with pulling from behind [H]. Pushing more readily translates a lowest instrumented vertebra that is not crossed by the center sacral line and reduces obliquity of the subjacent spared vertebra. The isolated major thoracolumbar or lumbar curve is ideal for anterior operation [I].
- Absence of posterior elements for sufficient surface area for fusion, or previous posterior pseudarthrosis.
- In the immature spine, continued anterior growth about a posterior fusion axis may produce the "crankshaft phenomenon." Pedicle screws, which lock the anterior column to posterior instrumentation, have made this obsolete. In addition, fusionless modalities have reduced indications for fusion in the first decade.
- The increased primary morbidity of the anterior approach must be balanced by its significantly lower rate of infection compared with the posterior approach.



G Vertebral anchors Vertebral body is rotated (*gray*) toward convexity. Concave pedicle is narrower and shorter. Pedicle screw (*orange*) navigates between spinal cord (*pink*) and great vessel (*red*). Laminar hook or wire (*green*) enters canal. Transverse process hook or wire (*blue*) avoids canal. Vertebral body screw (*brown*) serves as an anterior anchor.



H Spine as door Leaning door is more easily rectified by pushing (*orange*), akin to anterior correction of the spine, than pulling (*red*), as is the mechanism in posterior correction.



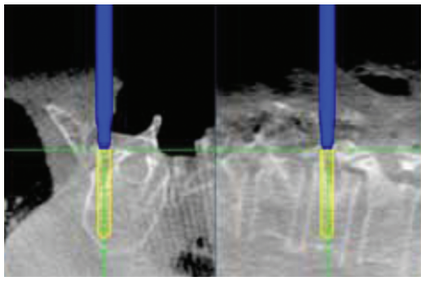
I Anterior operation Overcorrection spares L4 and brings it horizontal (*red*). One or two rods may be used, with vertebral body screws and staples. Intervertebral cages, in this case allogenous fibula (*green*), support sagittal contour to maintain lordosis.

Complication	Rate
Hæmorrhage requiring transfusion	<10%
Infection	1%
Neural injury	<1%
Superior mesenteric artery syndrome	<<1%
Pancreatitis	<10%
Pseudarthrosis	2%
Reoperation	10%

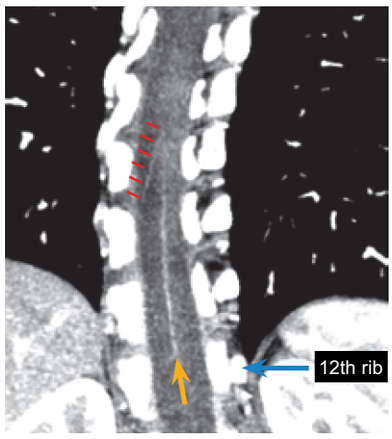
A Complication rate Primary spine fusion with instrumentation for adolescent idiopathic scoliosis is safe.

Modality	Comment
Somatosensory-evoked potential	Spinal cord—dorsal column and medial lemniscus pathway. Continuous. **Alarm**: ↓ 50% amplitude, ↑ 10% latency
Motor-evoked potential	Spinal cord—corticospinal tract. Triggered. Most sensitive to injury. Most sensitive to blood pressure, inhalation anaesthetic, body temperature, younger age. **Alarm**: ↓ 80% from 1 muscle site
Electromyography	Nerve root. • Spontaneous. **Alarm**: high frequency high amplitude trains • Triggered by direct implant stimulation. **Alarm** : ≤6 mA suggests osseous breach
Wake up test	Non-specific assessment of motor function. *Post facto.* Requires patient comprehension and risks excessive movement.

B Neural monitoring during spine surgery There are four modalities.



C Operative CT Navigation aids instrumentation. Drill sleeve is *blue*, length is measured by *green* ruler, and location of selected width pedicle screw is projected as *yellow*.



D CT angiogramme Segmental medullary artery of Adamkiewicz (*red*) arises from the 10th intercostal segmental vessels and travels proximalward to the spinal artery (*orange*). This patient underwent anterior convex instrumentation that skipped the 10th vertebra.

Complications

Complications are rare but potentially spectacular [A].

Hæmorrhage This may be limited by:

- Positioning with the abdomen free to reduce external pressure on the vertebral venous plexuses (e.g., Relton-Hall frame).
- Hypotensive anæsthesia. This must be balanced with normal blood pressure or hypertension to protect spinal cord perfusion, in particular during correction and fluid shifts over time.
- Surgical technique, including subperiosteal dissection, diathermy, wound packing.
- Antithrombolytic agents *per venam*, as well as gelatin, thrombin, and other agents *in situ*.
- Autogenous blood recovery system.

Infection The skin is the single greatest barrier to infection, and spinal deformity wounds are large. Infection rate is approximately 1%, compared with up to 15% for neuromuscular scoliosis. Infection may be limited by:

- Sterile technique, including Chlorhexidine and bleach for skin.
- Antibiotics. Administer within 30 minutes of incision and every 4 hours thereafter, to account for hæmorrhage. Postoperative administration may be single dose and should not exceed 24 hours. Mixture of antibiotics in allogenous osseous graft or lavage may be effective.
- Efficacy of other practices, such as limiting personnel, use of a sterile adhesive drape, changing of clothing and instruments, occluding suction when not in use, is difficult to demonstrate.
- Treatment includes incision and drainage, irrigation, and débridement of wound and spine. Remove loose osseous graft. Replace loose anchors. Insert drains, which are removed when the wound is sealed. Retain implants until robust fusion, at which point implant removal allows cure.

Neural injury This is the gravest concern for the patient and family.

- Minor injury includes positional neurapraxia. Most frequent is compression of the lateral femoral cutaneous nerve at the anterior iliac crest, but motor neurapraxias may occur, such as femoral nerve compression at the brim of the pelvis.
- Major injury may be central, at the spinal cord level, or peripheral, at the nerve root level. Neural monitoring includes somatosensory and motor-evoked potentials and electromyography [B]. Wake-up test may supplement these modalities. Keep implants out of the canal. Operative imaging, with image intensifier, röntgenogrammes, or CT, aids instrumentation [C]. Consider CT angiography in preparation for anterior instrumentation in order to look for dominant perfusion of the spinal cord from the side of operation [D]. Temporarily and atraumatically clamp intercostal vessels before ligation to ensure no change in neural signals.
- The risk of major neural injury is under 1%. Most children will demonstrate at least partial recovery. Ischæmic injury, such as after intercostal vessel ligation, shows least recovery.
- Treatment is complex and multifactorial. Observe positional neurapraxias, which recover spontaneously. In the event of neural signal changes, consider reversal of operative steps, for example, replace implant or relieve correction. Raise blood pressure, blood count, and body temperature. Modify anæsthetic agents. Administration of steroids *per venam* is controversial. CT myelogramme will outline vertebral canal in case of encroachment. Lumbar catheter will decompress vertebral canal in the event of spinal cord swelling.

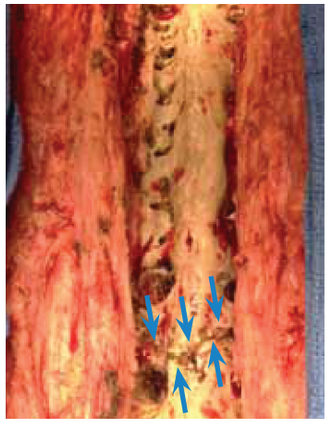
Superior mesenteric artery syndrome This also is known as "cast syndrome," from its occurrence after the application of extension casts for spinal fracture. This is caused by compression of the third part of the duodenum between superior mesenteric artery and aorta (angle <25 degrees) when the trunk is stretched after the spine is corrected. It typically occurs in tall and thin girls (less mesenteric fat), who present with nausea and vomiting, epigastric pain, and abdominal distension. Evaluation includes a dynamic contrast radiography. Treatment consists of left decubitus or prone positioning, intravenous fluids, abdominal rest and decompression, and in persistent cases total parenteral nutrition. Extremely rarely, division of the ligament of Treitz to release the duodenum, or duodenojejunostomy to bypass the point of obstruction, may be necessary.

Pancreatitis This is underdiagnosed after spinal fusion, because pain, nausea, loss of appetite are nonspecific. Theories include pancreatic hypoperfusion, altered anatomy, or complement activation, for example, by an autogenous blood recovery system. The presentation may resemble that of superior mesenteric artery syndrome, from which it is distinguished by elevated serum lipase (more sensitive and specific) and amylase. Treatment includes abdominal rest, intravenous fluids and, if persistent, hyperalimentation.

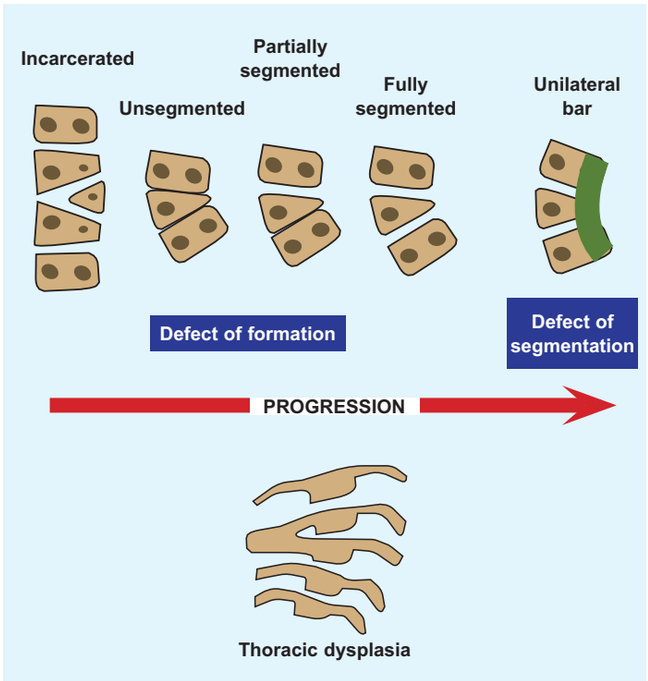
Progression of deformity In the instrumented curve, this may be due to pseudarthrosis or the crankshaft phenomenon. In the latter, posterior fusion acts like an axis about which the spine may turn with continued anterior growth to produce progressive deformity. The phenomenon does not occur after peak growth velocity and curve acceleration phase. Operations on children before these time points should include anterior fusion to avoid crankshaft. In the spared spine, curve progression may be seen in younger patients in whom fusion was performed in the early stages of development of structural curvature. Alternatively, this may occur after zealous correction that exceeds the degree to which the spared spine can compensate. This prioritizes spine balance over curve correction.

Pseudarthrosis The rate of pseudarthrosis after spinal deformity surgery is up to 2% in children, compared with up to 10 times in adults [E]. Principal causes are surgical technique and infection. Most present within the first 3 years, including persistent pain, progression of deformity, and implant failure. CT gives detail of pseudarthrosis and implant failure. Management consists of exploration of the entire spine, as multiple pseudarthroses may be present, possible opposite approach for a fresh bed for grafting, and revision instrumentation as necessary.

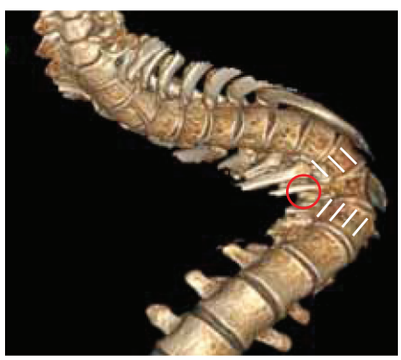
Venous thromboembolism, a serious complication of adult spine surgery, occurs so rarely in children that it does not warrant prophylaxis. Overall rate of secondary operation during childhood is approximately 10%.



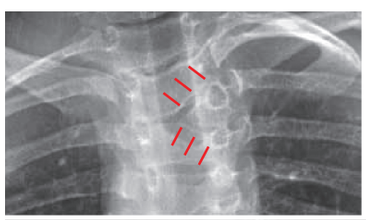
E Pseudarthrosis This was discovered at implant removal for chronic infection.

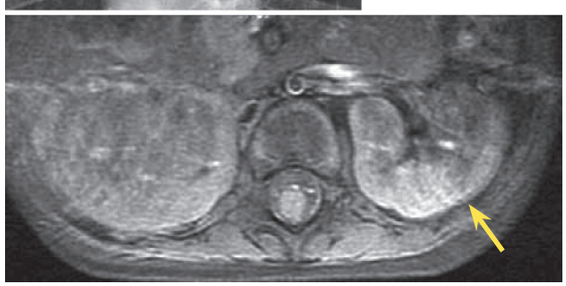


A Simplified classification of congenital scoliosis Heterogeneity and multiplicity undermine utility, leading to continual efforts to subclassify.



B CT of congenital scoliosis Unilateral bar (*red*) opposite hemivertebra (*white*) is associated with greatest progression.





C Incarcerated hemivertebra Outlined by red. No progression over 3 years. MRI spine also showed solitary kidney (*yellow*).

CONGENITAL SCOLIOSIS

This is defined as scoliosis produced by vertebral anomaly. It is classified based upon presence of tether around which growth may accelerate deformity and growth potential at vertebral physis [A].

Evaluation

Because development of the vertebrae is associated most closely with that of the neuraxis, genitourinary system, and heart, evaluation must encompass all these systems, including appropriate subspecialist referral. Vertebral anomaly may be seen in association with other anomalies, such as exemplified by VACTERL (Vertebra, Anal atresia, Cardiac anomaly, Tracheo-Esophageal fistula, Renal anomaly, Limb anomaly). In addition, congenital scoliosis may represent one part of a greater regional disorder that results in thoracic dysplasia, including chest wall and viscera.

Imaging Röntgenogrammes provide the initial assessment of type and magnitude and subsequent assessments of progression. CT defines osseous anatomy and is essential during operative preparation [B]. MRI evaluates the neuraxis for associated lesion, which is seen in approximately 1/3 of patients [C]. Abdominal ultrasonogramme screens the renal system. Echocardiogramme screens the heart.

Management

Observation Progression depends upon growth, in particular during infancy and puberty, and type of anomaly. Follow progression with serial röntgenogrammes. Three-fourths of curves will progress significantly, and up to 1/2 will require operative treatment. Postpone operative treatment, if necessary, until after 5 years of age, if possible: consider observation of the supine rather than the upright spine, following flexibility and ensuring that compensatory curves have not become stiff. Bracing is ineffective.

Operation After 5 years of age, balance the goal of further longitudinal growth with focal operation on an isolated lesion before secondary structural deformation of the spine occurs. The former approach will consist of a limited fusion while the latter may require extension over a broad segment of the spine.

Posterior fusion This is safest. The ideal indication is a second decade child and a curve that is sufficiently flexible and mild–moderate that a fusion will not be too long to balance the spine.

Anterior and posterior fusion This is indicated for a child in the first decade. It may be performed to obtain greater correction and thereby limit levels of fusion, and in order to prevent crankshaft, although the latter is less of a concern in congenital scoliosis due to circumferential growth disturbance compared with idiopathic scoliosis.

Vertebral resection This may be performed by simultaneous anterior and posterior approach performed with the young patient in the decubitus position when the anomaly is discrete or staged anterior followed by posterior in the older patient with severe deformity. Alternatively, this may be performed by a posterior approach [D].

- Insert anchors before resection.
- Identify and excise anomalous posterior elements.
- Disarticulate and excise corresponding rib heads.
- Excise corresponding transverse processes.
- Ligate thoracic nerve roots as necessary.
- Develop interval between spine and pleura or peritoneum, placing circumferential retractors.
- Identify cranial and caudal intervertebral discs. Discs serve as limits of resection, which begins by discectomy.
- Insert temporary rod to stabilize spine.
- Resect bone. This may be *in toto* in a young patient with a discrete anomaly, or sequentially starting with pedicles and removing posterior cortex of vertebral body last to protect spinal cord.
- Complete instrumentation, including anterior support as indicated.
- Reduction must be controlled and must not lengthen the spine.

THORACIC INSUFFICIENCY SYNDROME

The chest is deformed *in toto*: it can support neither pulmonary function nor pulmonary growth. Thoracic dysplasia with pulmonary insufficiency drive management in the first decade of life, which is aimed at restoring and maintaining thoracic volume in order to support lung development and function. Congenital scoliosis (incorporated in the term "spondylothoracic dysplasia") may be one component and is treated secondarily.

Evaluation

Röntgenogrammes measure thoracic dimensions as well as spine curvature. CT allows calculation of chest and lung volume. Spirometry may be limited by patient participation given age and disease burden.

Management

Thoraco(s)tomy and expansion The thorax is opened according to location of deformity, including costal and spine anomalies [A]. Cranial hook and cap are attached to the ribs, circumferential and forgiving to tolerate motion during respiration. Caudal implants may be attached to the ribs, lumbar spine by means of lamina hooks, or pelvis by looping over and into iliac crest.

While the procedure has the potential to rescue affected children from pulmonary collapse, it carries the significant costs of multiple procedures, implant failure, infection, and fibrosis of the chest wall with time.

Secondary Scoliosis

Scoliosis represents the effect on the spine of a primary problem [A] and as such is not structural. This type is distinguished by:

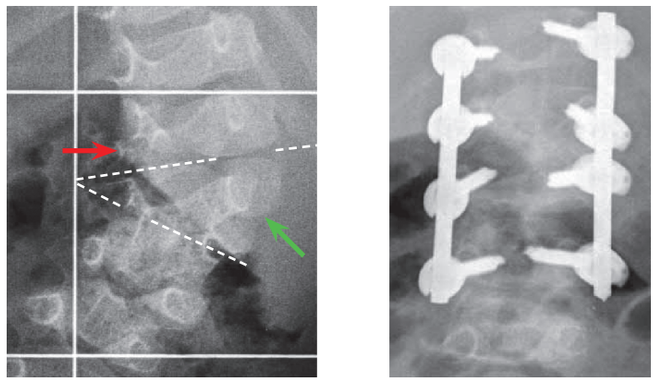
- Complete resolution ("cure") by complete treatment of the underlying disorder
- Absence of vertebral rotation

Lower limb length discrepancy Tilting of the pelvis results in obliquity of takeoff of the lumbar spine, which curves back toward the midline in order to balance the head on the pelvis. Curvature of the spine will not become structural because not enough time in a day is spent erect with both feet on the ground to tilt the pelvis.

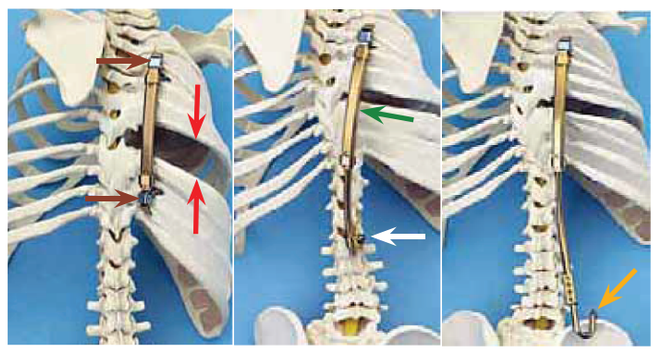
EVALUATION Place a block under the respective foot to bring the pelvis horizontal. Alternatively, examine the spine sitting or in the prone position. The deformity will thereby be eliminated. Side bending shows that the spine can be curved opposite to the presenting deformity. Röntgenogrammes with a block under the foot equal to the discrepancy will show a straight spine.

MANAGEMENT This follows general principles for lower limb length discrepancy. Do not let concern for scoliosis lead to unnecessarily treatment, for example, shoe lift.

Pain Scoliosis may be the presenting sign for several inflammatory disorders. Curvature of the spine splints against pain. In contrast to scoliosis secondary to lower limb length discrepancy, the deformity may not be influenced by position (if pain is not) and may not be flexible (opposite bending may exacerbate pain).

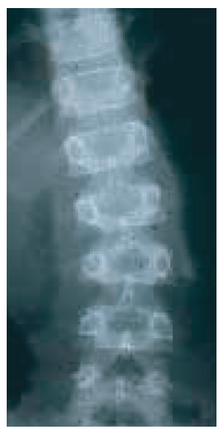


D Posterior vertebral resection for congenital scoliosis MRI confirmed incompletely ossified unilateral bar (*red*) and hemivertebra (*green*). After removal of posterior elements, anterior hemivertebra was resected *in toto* by following planes through intervertebral discs (*white*).

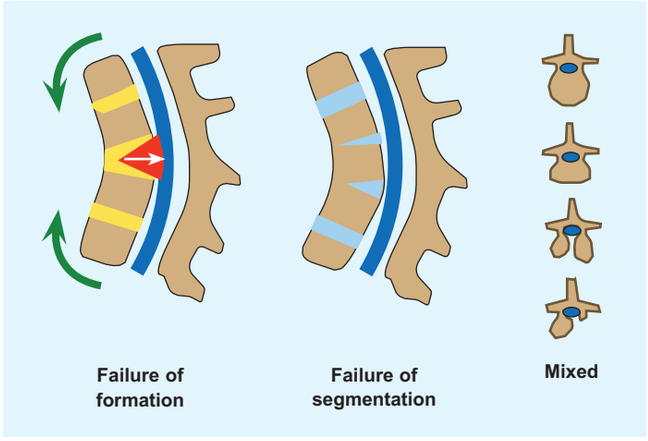


A Vertical Expandable Prosthetic Titanium Rib (VEPTR) Thoracotomy permits expansion of the chest by a growing implant (*green*) that anchors to the ribs (*brown*), lumbar lamina (*white*), or iliac crest (*orange*). [www.pmda.go.jp].

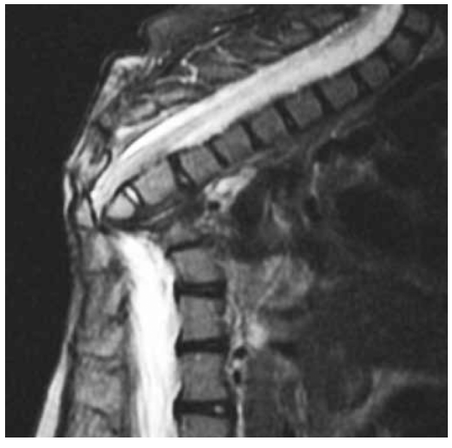
Primary cause		Comment
Lower limb length discrepancy		Compensatory to lumbosacral obliquity produced by pelvic tilt
Pain	Infection	Discitis
Pain	Deformity	Spondylolisthesis
Pain	Tumor	Osseous, e.g., osteoid osteoma Neural



A Secondary scoliosis The deformity is not structural and resolves with resolution of the primary disorder. It is distinguished by limitation to a coronal plane deformity with absence of transverse plane deformity. Imaging shows no vertebral rotation.



A Classification of congenital kyphosis Increasing flexion of the spine can propel a hemivertebra into the vertebral canal, where the spinal cord is draped over and restricted by the gibbus. Variability of anomalies is captured under the mixed type.



B Congenital kyphosis This presents the highest neural risk of spine deformity in children.

CONGENITAL KYPHOSIS

Like congenital scoliosis, it is defined as kyphosis produced by vertebral anomaly. It is classified as failure of formation, failure of segmentation, and mixed, including rotatory type [A]. It may be distinguished from congenital scoliosis in being rarer and graver, because the plane of deformity deflects the vertebral column directly toward the spinal cord while simultaneously restricting its motion.

Evaluation

Like congenital scoliosis, evaluation must encompass neuraxis, genitourinary system, and heart. Failure of formation and mixed types are more progressive, and present a greater neural risk, than failure of segmentation. While the tether in failure of segmentation creates deformity, the lack of tether in failure of formation allows sagittal displacement of the hemivertebra toward the neuraxis. Mixed lesions may include rotatory subluxation or dislocation, which also compromises the vertebral canal. Most patients with congenital kyphosis are asymptomatic in the first decade. Adolescents may complain of back pain due to subjacent lumbar hyperlordosis, which is compensatory to maintain sagittal balance. Progressive deformity, or trauma in the setting of severe deformity, may present with neural deficit. Neurologic examination is essential. Evaluate flexibility by draping the small child over the knee or by asking the older child to push up with the hands in the prone position. Associated scoliosis tends to be mild and is treated secondarily.

Imaging Röntgenogrammes provide the initial assessment of type and magnitude, and subsequent assessments of progression. Hyperextension röntgenogrammes evaluate flexibility. CT defines osseous anatomy and is essential during operative preparation. MRI evaluates the vertebral canal for encroachment and neuraxis for injury [B]. Urodynamic testing is helpful in the setting of neural changes.

Management

Observation Progression depends upon growth and type of anomaly. Follow progression with serial röntgenogrammes. Bracing is ineffective.

Operation The young deformed spine is technically challenging to instrument. Incomplete ossification makes excision difficult: gristle is tougher than bone.

Posterior fusion This is safest. It is indicated in the child <5 years of age with curve <50 degrees. The tether produced by fusion may lead to spontaneous improvement of deformity with anterior growth, although this is unpredictable as it is inherently abnormal. A hooked construct does not lock the anterior column, thereby not interfering with its potential for growth. Hooks by definition are placed in the vertebral canal, where there is less space available for cord than in scoliosis.

Vertebral resection This is indicated for severe deformity and in the event of neural deficit, in which decompression is necessary. Posterior approach with circumferential dissection is more direct than the addition of an anterior approach, which is farthest from the site of deformity, and safer, as the great vessels and viscera fall forward away from the deformity.

KYPHOSIS OF SCHEUERMANN

Normal thoracic kyphosis is 20 to 50 degrees. Increasing lumbar lordosis compensates for thoracic hyperkyphosis, as in Scheuermann ("Shoyer-man") disease [A]. Hip flexion contracture will drive the spine into lordosis. Sagittal balance is more directly related to functional outcome than coronal balance.

Clinical Features

Pathogenesis This is an osteochondrosis, a growth disturbance producing vertebral wedging, shortening of the anterior column, and kyphosis. Because this is an undergrowth phenomenon, the spine does not buckle to produce concomitant rotation. Scheuermann implicated manual labor in pathogenesis ("apprentice kyphosis"), although the rôle of abnormal load is unclear. There is an undefined heritable component.

Evaluation Onset is in the second decade. Boys are affected more. The disease occasionally occurs in the lumbar spine. Pain is characteristic: this occurs at the apex of deformity and/or at an associated lumbar spondylolysis. Patients are significantly disturbed by the appearance. Scheuermann kyphosis "peaks" with forward bending [B], in contrast with postural round back, which is less severe, more flexible, less acute, and not painful. The skin at the apex of deformity may be roughened due to prominence and lack of flexibility of the spine: this is a sign of decompensation and serves as a functional extraskeletal indication for treatment. There may be associated scoliosis: this tends to be small, not significantly progressive, and as such does not drive management. Compensatory hyperlordosis of the lumbar spine to maintain sagittal balance increases the risk of spondylolysis.

Imaging Röntgenogrammes:

- Allow measurement of kyphosis, after the method of Cobb. This includes lateral fulcrum hyperextension to assess flexibility.
- Show end plate irregularity associated with anterior wedging of vertebrae. The end stage is Schmorl nodes [C]: these represent vertical herniation of intervertebral disc through end plate into vertebral body, where bone may be necrotic.
- Show associated spondylolysis and scoliosis.
- Must be full length standing to enable assessment of spine balance, which is of particular importance in operative cases.

MRI provides detail but may not influence management.

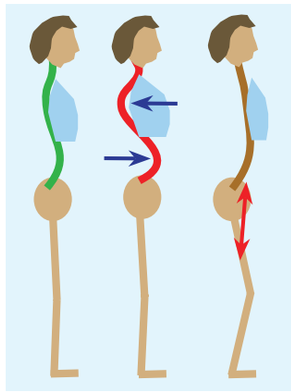
Management

Natural history is not well understood, making treatment recommendations variable. Nonoperative treatment includes the following:

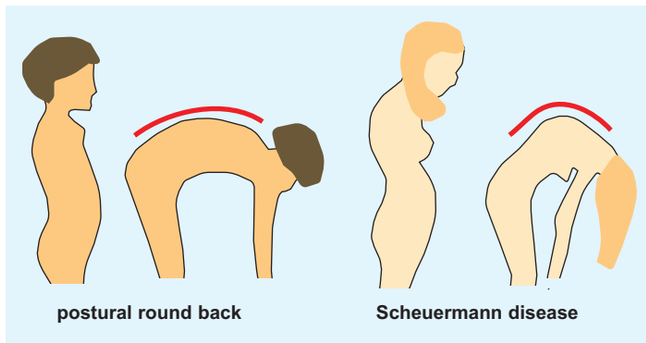
- Symptom control, including activity modification and nonnarcotic analgesics.
- Stretching exercises, focused on the spine (hyperextension), proximal muscles, and posture ("tuck" chin "don't poke" nose).
- Thoracolumbosacral orthotic (TLSO), although evidence for effectiveness, and compliance, are poor.

Operative treatment is indicated for progressive and severe deformity. There is no consensus, influences including pain, patient's perception of appearance, and degree (>75 degrees is a moderate guideline). Issues of relevance to spine fusion with instrumentation include the following:

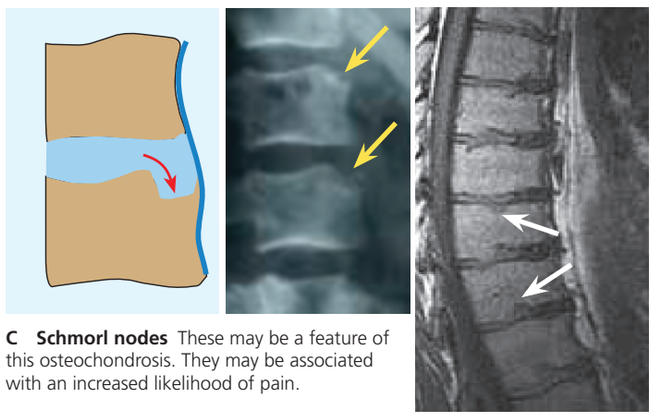
- Anterior approach for stiff curves. The rate has been reduced by greater understanding and acceptance of posterior osteotomies, in particular Ponte modification of Smith-Petersen [D].
- Multiple (this is diffuse deformity) posterior osteotomies to allow shortening of posterior column by compression instrumentation. This overcomes severity and stiffness of curves, reduces risk of lengthening of the anterior column and associated neural risk, and decreases cantilever force thereby reducing implant failure (which is more frequent than in scoliosis operation).
- Fusion from lordosis to lordosis and stable to stable vertebra. Stopping fusion at a kyphotic level, or posterior to sagittal balance line, risks implant failure and adjacent segment kyphosis.



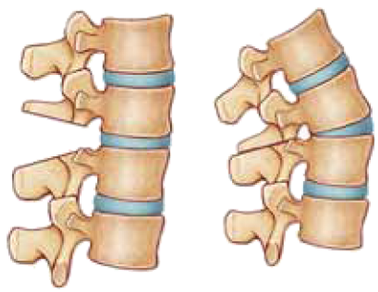
A Spine alignment Spine and pelvis alignment is interconnected. The spine compensates to maintain balance. Note chest dimensions: Scheuermann kyphosis increase thoracic volume unless severe, by contrast with lordosing conditions of the thoracic spine, which reduce pulmonary space.



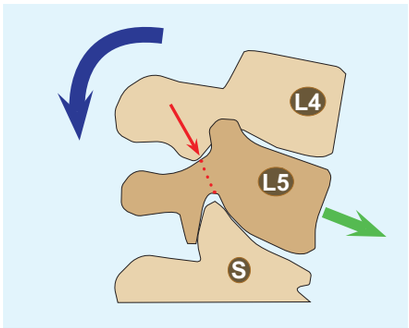
B Postural round back from Scheuermann kyphosis Scheuermann kyphosis is exacerbated to a focal angulation with spine flexion.



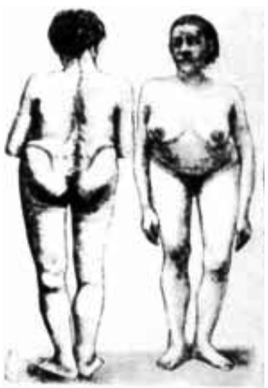
C Schmorl nodes These may be a feature of this osteochondrosis. They may be associated with an increased likelihood of pain.



D Posterior osteotomy for Scheuermann kyphosis Resection of adjacent articular processes, ligamentum subflavum with or without spinous processes allows segmental correction (1 degree/mm bone up to 10 degrees/level) of nonfocal kyphosis by shortening posterior column without lengthening anterior column.
[www.srs.org]

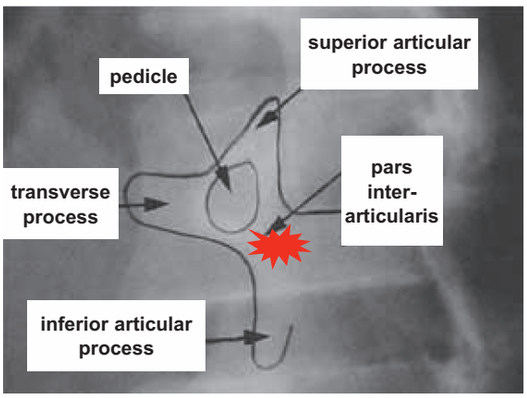


A Spondylolysis With hyperextension of the spine, inferior articular process of superjacent vertebra strikes and (eventually) fractures pars interarticularis of affected vertebra. If both parts are fractured, the affected vertebra can slip forward on subjacent spine, taking with it the entire trunk.

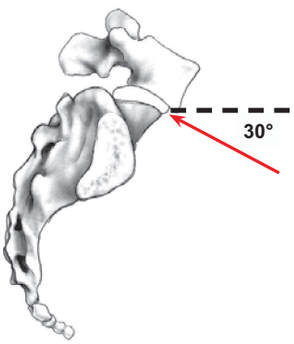


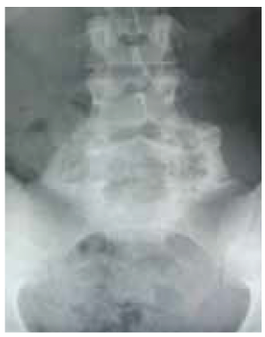
B Spondylolisthesis Neugebauer's original sketch (1895):

- lumbar hyperlordosis
- vertical sacrum
- lumbosacral kyphosis
- lumbosacral step off
- cordiform buttocks
- hip flexion
- knee flexion
- secondary painful scoliosis



C Scotty dog Oblique view röntgenogramme exposes pars interarticularis.





D Ferguson view X-ray beam (*red*) is directed tangent to L5-S1 intervertebral disc for a true orthogonal view of the lumbosacral junction, thereby highlighting fusion mass on röntgenogrammes.

SPONDYLOLYSIS AND SPONDYLOLISTHESIS

Greek σπονδυλη: "*spondyle,*" is Latin *vertebra*. Greek λυσις: "*lysis*," is equivalent to Latin *fractura*. Greek ολισθεσις, anglicized as "*olisthy*," means "slipping." Greek πτωσις: "*ptosis,"* means "falling down," used of both the eyelid and a vertebra.

Pathogenesis

Spondylolysis refers to a fracture of the pars interarticularis caused by the inferior articular process of the superjacent vertebra during hyperextension of the lumbar spine [A]. In 2/3 of patients, this represents a stress or fatigue type of fracture; however, up to 1/3 of athletes recall a sudden event. Unilateral spondylolysis is stable and more likely to heal with nonoperative treatment. Bilateral spondylolysis disengages the neural arch from the body, which may allow the body with superjacent vertebral column to "slip" relative to the ipsilevel neural arch and remaining subjacent vertebral column. Hyperlordosis and horizontalization of sacrum increase traction and likelihood of olisthy. The traumatic mechanism is reinforced by the fact that spondylolysis occurs with increased frequency in children who participate in certain sports that involve hyperextension with axial loading of the lumbar spine, such as gymnastics, and the fact that it has not been described in the nonambulatory. A genetic predisposition is suggested by familial and racial features (e.g., Eskimos an order of magnitude more than Whites) and by increased incidence in certain diseases (e.g., Marfan syndrome).

Evaluation

History and physical examination Spondylolysis occurs in approximately 5% of the population. Boys are two to three times more affected than do girls. In >80%, L5 is affected. The majority are asymptomatic. However, it is the most common identifiable cause of back pain, which results from micromotion and the inflammatory response of attempted healing. Pain is exacerbated by hyperextension of the lumbar spine, which when performed in single limb stance helps localize a unilateral lysis as the child cannot accommodate for pain by shifting away. Causes of pain in spondylolisthesis include tension on the broken bone, neural compression (foraminal or central), and shear on the intervertebral disc. Spondylolisthesis is two to three times more frequent in girls, which is consistent with the principle that natural history is worse in the gender in which a disease is rarer. Its *forme pleine* is well illustrated in the original sketch of Neugebauer [B]. Phalen and Dickson (1961) described hamstring tightness limiting swing phase to produce a short-stride, shuffling, toe–toe, crouched gait. Children are at highest risk of olisthy during growth acceleration of puberty, although clinically significant progression is <10%.

Imaging Oblique röntgenogrammes bring the pars interarticularis orthogonal to the x-ray beam, thereby exposing spondylolysis. In the Scotty dog representation [C], the transverse process is the nose, the ear is the superior articular process, the eye is the pedicle, the forelimb is the inferior articular process, and the neck is the pars interarticularis. Spondylolysis is said to appear as a collar or a broken neck. Bilateral spondylolysis may be seen on lateral projection röntgenogramme.

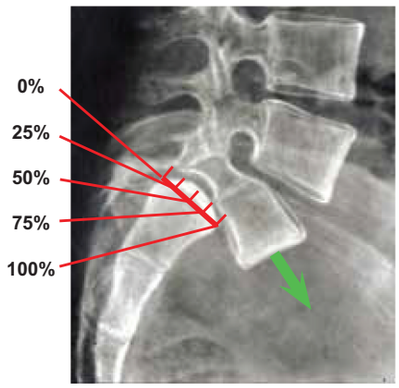
Ferguson view, taken with beam directed tangent to L5-S1 intervertebral disc, is a true orthogonal view of the lumbosacral junction and is essential to evaluation of L5-S1 fusion [D]. Standard anteroposterior projection may produce an upside down "Napoleon hat" superimposition of L5 as it has slipped in front of S1.

For spondylolisthesis, several measurements aid assessment [E–G].

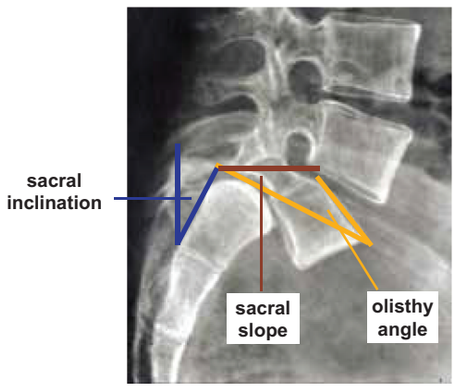
- Grade (severity). Myerding divided the top of S1 into quarters. Taillard distinguished olisthy <50% as stable from olisthy >50%, which is unstable. Such reductiveness is functional and akin to, for example, dividing physial fractures after Salter and Harris into without (I + II) and with (III + IV) articular extension. When the affected vertebra slips off the subjacent one (>100%), this is known as spondyloptosis.

- Sacral inclination. Angle subtended by a tangent to posterior S1 relative to the vertical. The greater the sacral inclination, the more vertical is the axis of body weight, which increases shear on L5 pars interarticularis and L5-S1 intervertebral disc, thereby increasing risk of olisthy.
- Sacral slope. Angle subtended by the horizontal and a line tangent to the top of S1. Increasing sacral slope is associated with increasing shear force. As L5 disengages from S1—in the extreme in spondyloptosis—sacral slope and inclination reduce as the pelvis retroverts.
- Olisthy angle. Angle subtended by a tangent to L5 and a perpendicular to the posterior S1 tangent. This is a measure of lumbosacral lordosis (positive value) or kyphosis (abnormal, stated as a negative value). The latter is an indication of instability, as L5 has begun its fall off S1, and is associated with >50% olisthy.
- Pelvic incidence [G]. Angle subtended by a perpendicular to the top of S1 and a line drawn from the center of S1 to the center of rotation of the hips. If femoral heads are not superimposed, take the midpoint between their centers. Assuming pelvis rotation centered at the hips, pelvic incidence is constant and as such is a patient characteristic. The normal range is 45 to 60 degrees. Increasing pelvic incidence means increasing lordosis and increasing shear force leading to tensile failure. Decreasing pelvic incidence means decreasing lordosis and compressive failure ("nut-cracker" mechanism). Ideal balance is achieved when pelvic incidence equals lumbar lordosis, akin to a thoracic inlet angle equal to cervical lordosis at the upper end of the spine.
- Pelvic tilt [G]. Angle subtended by the vertical and a line drawn from the hip center(s) to the center of the top of S1. By contrast with pelvic incidence, reduction in pelvic tilt brings the lumbosacral junction more vertical, thereby increasing shear force on pars interarticularis and intervertebral disc. Unlike pelvic incidence, pelvic tilt can be altered, for example, with physiotherapy targeted at retroversion of the pelvis to reduce lumbar lordosis to relieve back pain.
- Flexion and extension lateral views aid in determination of flexibility of the olisthy.
- Full length lateral röntgenogramme assesses spine sagittal balance. This is essential to operative planning. For a balanced spine, spondylolisthesis needs no reduction. By contrast, pull L5 back and restore lumbosacral lordosis if that is what is necessary to correct sagittal imbalance. If sagittal balance is not restored, a flexible pædiatric spine may compensate by increasing lumbar lordosis; however, a degenerating, stiffening lumbar spine as the child transitions to adult will expose sagittal imbalance, which now is locked by a fusion at the lumbosacral junction.

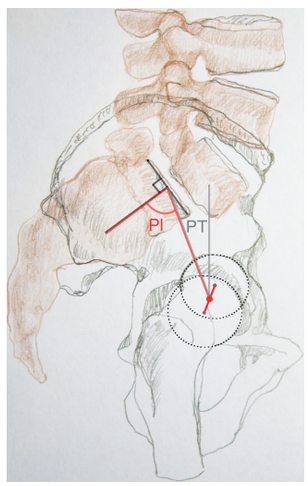
Scintigraphy provides a measure of osseous metabolism; as such, it may detect an active spondylolysis that has potential to heal, which may influence decision to brace [H]. Single photon emission computed tomography (SPECT) allows localization of scintigraphic uptake in a vertebra, that is, to the pars interarticularis [I]. Positive scintigramme defines an "acute" spondylolysis.



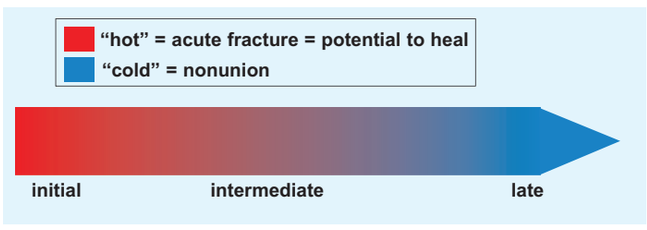
E Spondylolisthesis—grading after Myerding Types I-IV according to % olisthy.



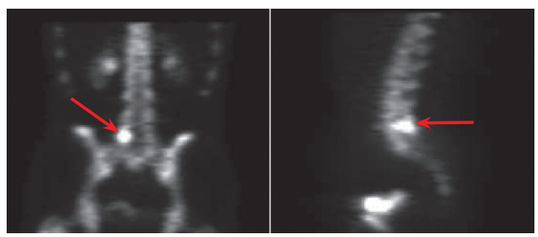
F Spondylolisthesis—angular assessment Note that secondary osseous deformity in chronic conditions introduces imprecision to measurements based upon tangents.



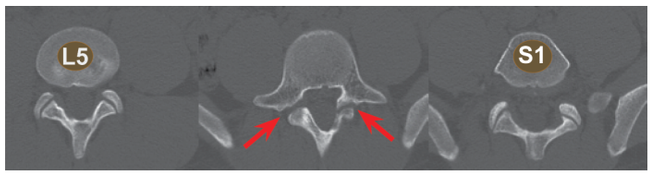
G Pelvic incidence (PI) and pelvic tilt (PT) The assessment of spondylolisthesis has expanded to include the hips. [https://neurosurgerycns.wordpress.com]



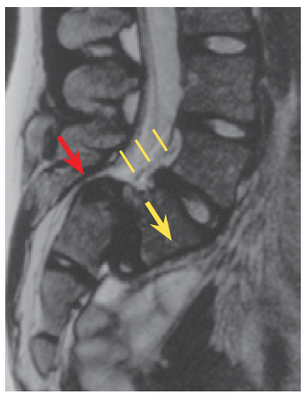
H Scintigraphic activity of spondylolysis Increased uptake (*red*) and/or early detection is an indication for bracing to allow healing of lesion without operative intervention.



I Single photon emission computed tomography SPECT shows that increased uptake occurs focally in the posterior elements on the right side of L5, at the pars interarticularis.



J CT of Spondylolysis Bilateral atrophic L5 spondylolysis, which appears as an "extra facet joint sign" between L5-L4 and L5-S1 facet joints.



K MRI of spondylolisthesis Note course of L5 nerve root (*yellow*), visible in its descent as part of the cauda equina and in cross section resembling a "target" as it exits the neural foramen. Note severe central canal stenosis (*red*), which influences decompression in risk of neural or dural injury and in indication for sacral ostectomy.

Factor	Comment
L5 lesion	Increased lordosis leads to increased shear force at fracture
Bilateral lesion	Increased motion and instability at fracture
Chronic	Fracture nonunion

L Factors associated with nonhealing of spondylolysis

Factor	Comment
Olisthy > 50%	Unstable
Lumbosacral kyphosis	Unstable
Growth acceleration of puberty	Growth amplifies deformity Follow patients through maturity
Gender	Female
Increased sacral inclination Increased sacral slope Decreased pelvic tilt	Radiographic assessment essential

M Factors associated with progression of spondylolisthesis

N Brace for spondylolysis This differs from a deformity brace in being antilordotic, fabricated to 15 degrees, and simple, having no pads or openings.

Computed axial tomography provides the best osseous detail in spondylolysis [J]. It has a rôle when the clinical presentation is consistent with spondylolysis but scintigramme, as the initial screening test, is negative. It also can definitively follow healing of spondylolysis.

Because of availability, broad scope, and lack of radiation, MRI often is used as a screening test for back pain in children. Its greatest utility lies in the discovery of a "prelysis" state, as evidenced by increased T2 signal in the pars interarticularis without fracture. Such patients are braced to heal the lesion as prophylaxis against spondylolysis. Like positive scintigramme, high signal change defines an acute spondylolysis. MRI is most utilized in operative preparation [K].

Management

General principles

- Successful treatment is defined as resolution of symptoms, even in the setting of persistent spondylolysis or pseudarthrosis after spondylolisthesis fusion.
- Healing of spondylolysis is least likely for L5, bilateral, chronic spondylolysis [L].
- Nonoperative management suffices for the majority (>80%) of patients with stable lesions.
- Several factors are associated with olisthy and its progression [M].
- Decompression is indicated for neural symptoms and signs. These may be central, such as bladder dysfunction assessed by cystometrogramme, or foraminal, such as radiculopathy. They tend to be features of unstable olisthesis.
- Instrumentation aids fusion and enables controlled reduction. It is essential for unstable lesions, and after decompression, which is destabilizing.
- Circumferential fusion with instrumentation of unstable olisthesis reduces pseudarthrosis and saves a level.
- Reduction includes both translation and angulation. The former is more likely to result in stretch neuropathy. Goals of reduction are restoration of sagittal balance and moving from instability to stability. Complete reduction is not necessary, and increases neural risk. Reduction also reduces pseudarthrosis by restoring stability as well as providing compressive force and greater surface area for osseous graft. Operative casting and traction for reduction may be regarded as historical, although proponents of the latter point to reduced neural risk due to its gradual nature. Reduction must be preceded by decompression to reduce neural risk.
- Sagittal balance is the most sensitive measure of quality of life. Röntgenogrammes must include full length standing views. Operation must restore global alignment in addition to addressing focal oilsthesis.

Spondylolysis

Management depends upon two features: activity of lesion and symptoms.

- Symptomatic care.

This is in dictated for:

— Chronic lesion
— Acceptable symptoms and signs

This consists of:

— Activity modification, for example, stop offending sport
— Exercises, including pelvic tilt and others antilordotic to the lumbar spine, hamstring, and iliopsoa stretching
— Nonsteroidal anti-inflammatory agents

Brace [N].

This is indicated for:

— Hyperintensity on scintigramme or MRI, suggesting potential to heal. This represents immobilization of an acute fracture.
— Unacceptable pain.

The protocol consists of two consecutive phases:

— Full time for 3 months with no sports
— Full time for 3 months with sports

- Operative treatment.

This is indicated for unacceptable pain despite symptomatic care or bracing. It consists of the following:

— usion *in situ* [N]. This is advocated for L5 lesion. Motion at L5-S1 is least in the lumbar spine. L5-S1 is most affected by degenerative disc disease. Repair of L5 is technically difficult because transverse processes are small (if used as site for cranial anchor) and because this is the most lordotic part of the lumbar spine, making access difficult. A midline posterior approach or a paramedian approach may be utilized [O]. The latter consists of bilateral incisions in deep fascia followed by splitting of erector spinæ for a direct approach to partes interarticulares, facet joints, and transverse processes. It is said to be easier than retraction of the robust column of muscle at the lumbosacral junction against the iliac crests. In addition, disruption of midline soft tissues may be destabilizing. Excise L5-S1 facet joint. Place osseous graft obtained from ilium through same cutaneous incision from sacral alæ to L5 transverse processes with or without interlaminar grafting. Instrumentation may aid fusion and substitutes for postoperative immobilization. These must be balanced against increased risk, time, and cost. Absent instrumentation, supplement with postoperative brace, including the hip to immobilize the lumbosacral junction.

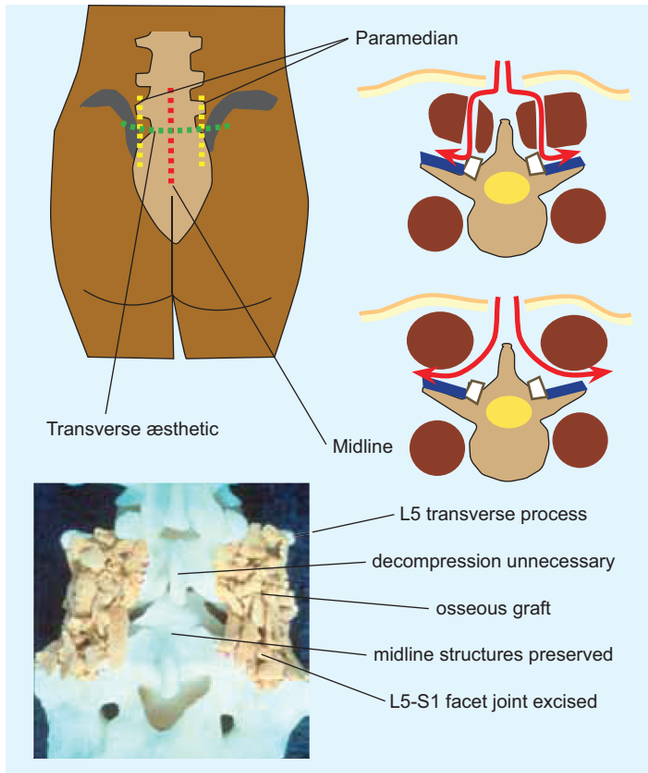
— Repair. No motion is sacrificed. Midline posterior approach, for any level. Anchor cranial (pedicle or transverse process) and caudal (lamina or spinous process) to the spondylolysis, or place a screw through the spondylolysis [P]. Débride spondylolysis. Lay graft obtained from the ilium through same cutaneous incision across spondylolysis. A wire may be looped around transverse process as a cranial anchor; this has evolved to a screw in the pedicle, which preserves the transverse process for complete decortication and fusion. The screw may be connected with a rod to a laminar hook as a caudal anchor or to a spinous process wire or cable as the caudal anchor. The latter caudal anchor achieves compression across the spondylolysis while trading rigidity for lower profile to permit a wider surface area for fusion.

Spondylolisthesis <50% This is stable. As such, treatment follows spondylolysis.

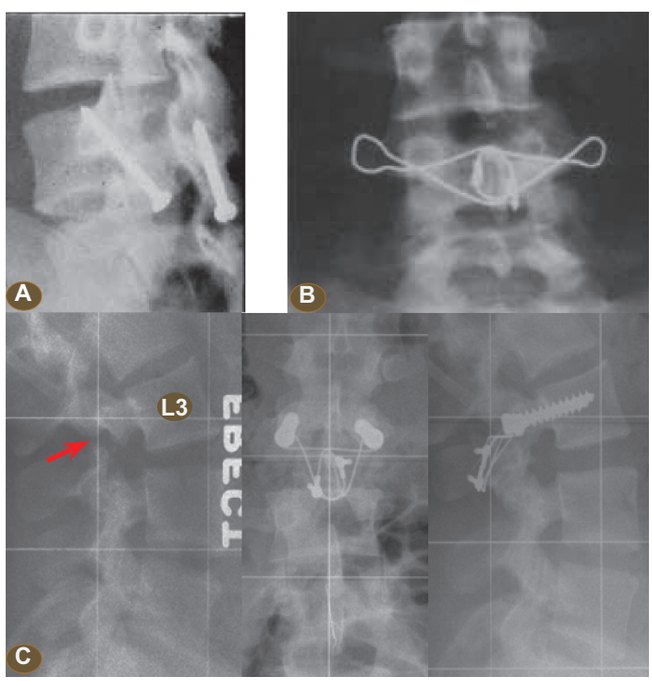
- An exception is bracing based upon osseous activity, because there is no expectation of healing.
- Repair is controversial, with the balance of operative treatment weighted toward fusion.
- Instrumentation is controversial. Stability argues against. Stability means lordosis is preserved, obviating the need for reduction. Instrumentation may aid fusion across a wide gap under tension.

Spondylolisthesis >50% The following characteristics dictate management.

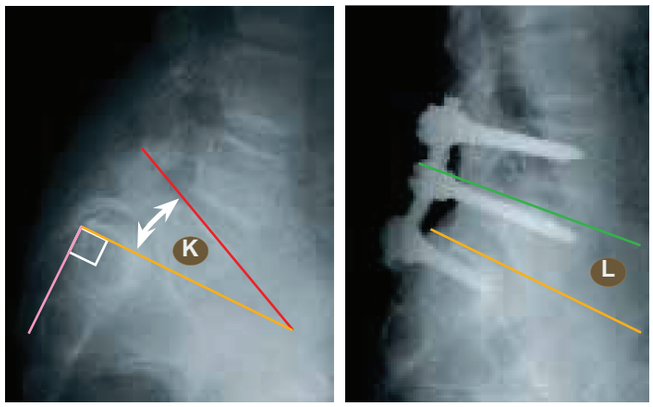
- Instability. Operative treatment is indicated primarily.
- Instability. Instrumentation and reduction are stabilizing.
- Severity of olisthy leads to kyphosis and increases likelihood of sagittal imbalance. Instrumentation with reduction correct angular deformity and restore balance.
- Severity of olisthy is associated with significant central and foraminal neural compression, which requires decompression and in turn instrumentation. Excision of the posterior superior corner of S1 allows for adequate decompression without the need for excessive reduction.



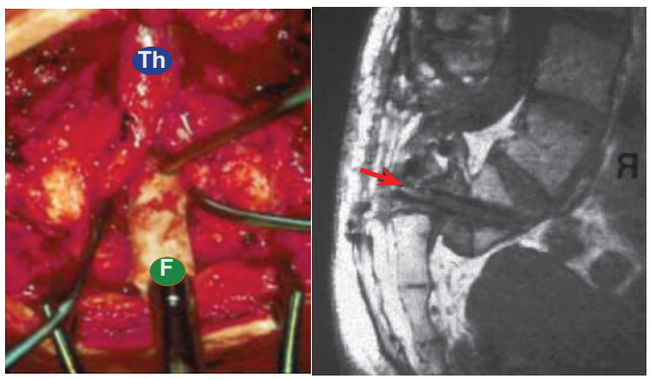
O Surgical approach for spondylolysis fusion Midline is the standard. It is extensile and provides full exposure for fusion for decompression and for instrumentation. Paramedian give direct access for fusion *in situ* but is not extensile and does not allow decompression.



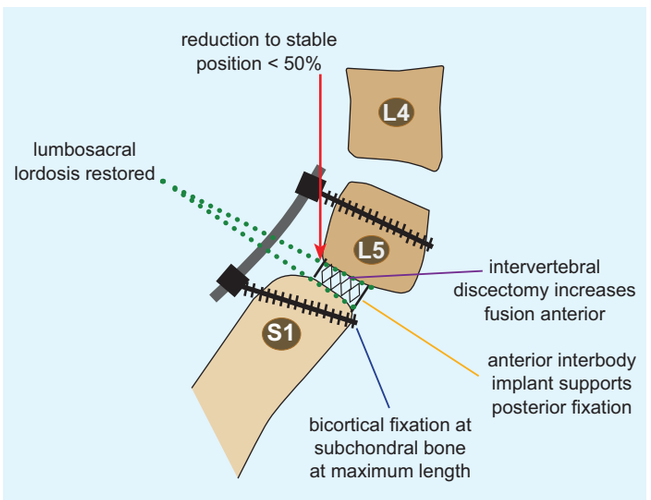
P Repair of spondylolysis.
A—Buck placed a screw through and across the pars interarticularis.
B—Scott compressed the fracture by a wire passed around transverse and spinous processes.
C—Débridement, compression, and grafting of L3 spondylolysis (*red*) resulted in successful repair (*orange*).



Q L4-S1 posterior fusion with instrumentation L4 provides fixation stability to aid reduction and increased surface area to aid fusion in compression. Lumbosacral kyphosis (k) has been reduced to lordosis (L).



R L4-S1 posterior fusion with instrumentation Fibula allograft (F) as anterior instrumentation via posterior approach to enhance construct stability. After decompression, cannulated drill prepares central channel into which fibula is tamped between neural elements (Th).

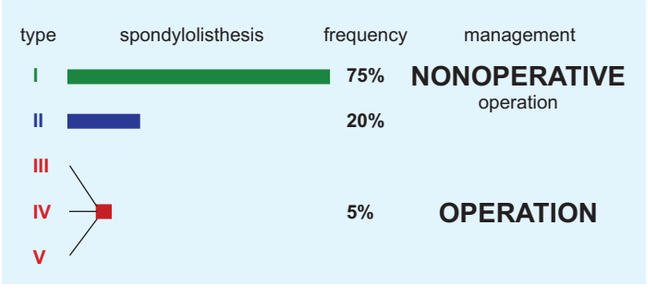


S L5-S1 circumferential fusion with instrumentation This allows saving of a motion segment.

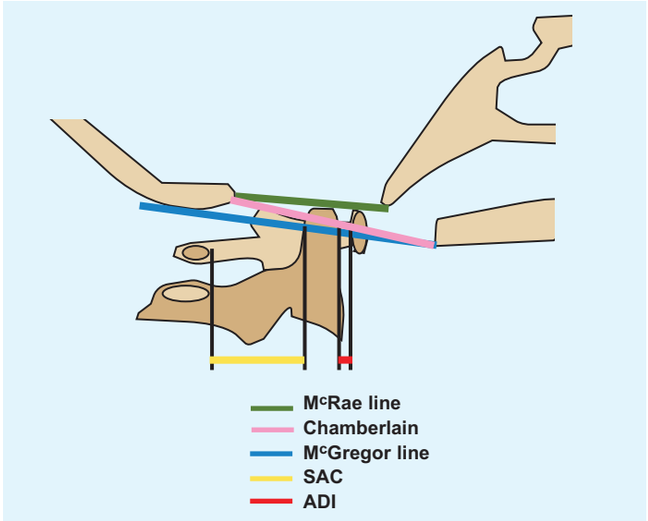
For L5-S1 spondylolisthesis >50%, there are two principal approaches.

- Posterior fusion with instrumentation. This extends from L4 to S1 [Q]. L4 is indicated for construct stability, which aids fusion and reduction, as well as reducing implant failure. Distal fixation may be enhanced by anchoring into the ilium and by anchoring through S1 into the body of L5 [R]. Extension to L4 increases surface area for fusion, in particular after decompression removes posterior elements of L5, and allows compression upon graft rather than tension to L5.
- Circumferential fusion with instrumentation. This saves a motion segment by limiting to L5-S1. Stability is enhanced by circumferential instrumentation, including anterior interbody support [S]. Fusion is enhanced by providing interbody surface area under anterior compression.

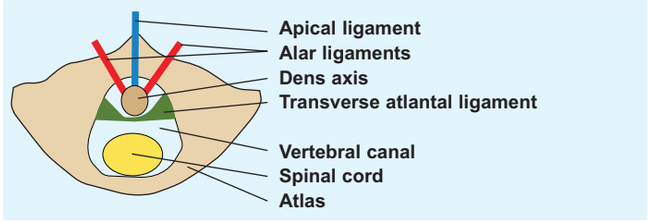
Summary of management [T]



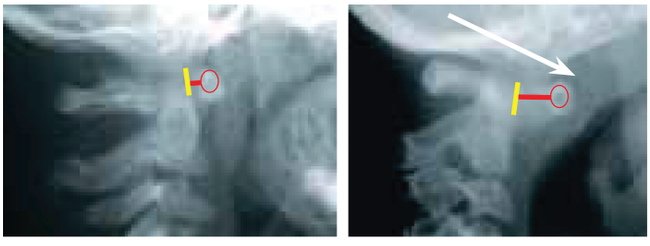
T Spondylolisthesis Most cases are stable, and nonoperative management is effective in most cases.



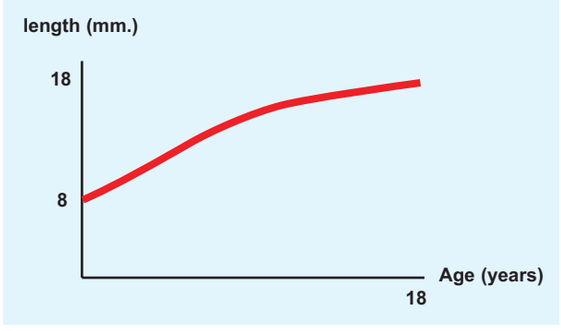
A Relational lines of the upper cervical spine They describe relationship of the spine to the skull and the spine to the spinal cord.



B Ligaments of the dens They allow the atlas to rotate about the dens for neck motion but restrict dens translation to protect the spinal cord.



C Atlantoaxial instability Neutral and flexion lateral views of the cervical spine show an increase of the interval between posterior surface of the anterior arch of atlas (*red ring*) and the anterior dens axis (*yellow line*) due to rupture or laxity of the transverse atlantal ligament.

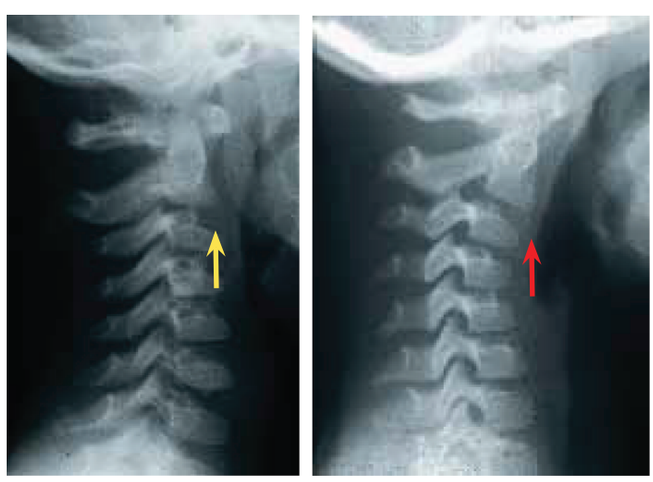


D Growth of dens axis The dens axis ossifies mostly over the first decade but continues over the second decade.

CERVICAL SPINE

Essential to understanding the cervical spine are anatomic and radiographic relationships [A].

- McRae, Chamberlain, and McGregor lines describe the entrance to the skull. Chamberlain and McGregor lines relate to the hard palate, which may be difficult to see. In addition, they allow for the dens to be above in the normal state (≤1.5 mm and ≤4.5 mm, respectively). McRae line is most reliable: it is drawn from opisthion to basion and defines the foramen magnum, above which the dens never reaches in the normal state. Basilar impression is defined as tip of dens above McRae line. Chiari malformation is defined as >5 mm downward displacement of cerebellar tonsils; ≤5 mm is considered benign tonsillar ectopia.
- SAC. Space available for cord. Posterior dens to anterior surface of posterior arch. Normal is ≥13 mm.
- ADI. Atlantodental interval, measured from posterior surface of anterior arch of atlas to anterior dens. The dens is waisted where it is retained by the transverse atlantal ligament against the anterior arch of atlas to within 5 mm. [B]. Flexion is a stress view [C]. Atlantoaxial instability is defined as ADI >5 mm. This may be due to anomaly of the dens axis (*q.v.*) or to ligamentous laxity, as typified by Down syndrome (*q.v.*).
- The alar ligaments connect dens to occipital condyles and check side–side movement of the skull when it turns. The apical ligament connects the tip of dens with the anterior margin of foramen magnum; it is a vestigial intervertebral structure containing elements of notochord.
- Dens axis development. The tip of dens may not reach the arch of atlas until the end of the first decade and continues to grow through the second decade. Early imaging may mistake normal development for hypoplasia [D].
- Pseudosubluxation. Ligamentous laxity allows <5 mm of anterior translation of C2-C3 and less frequently C3-C4 in the first decade [E].



E Pseudosubluxation Anterior translation of C2 on C3, and less frequently C3 on C4, <5 mm is normal in a child, due to ligamentous laxity.

Anomaly of the Dens Axis

Aplasia is rare [A].

Hypoplasia is associated with skeletal dysplasia, in particular the type II collagenopathies and the mucopolysaccharidoses [B].

Os odontoideum refers to separation of a well-developed rounded bone from the body of axis [B]. It represents a nonunion of fracture of the dens. The mechanism is hypothesized to include disruption of a tenuous blood supply to the base of the dens together with traction applied to the fragment by the alar ligaments. Indications for treatment include pain, evidence of instability, and neural change. Direct osteosynthesis with a dens screw if the fragment is sufficiently large stabilizes the fracture; posterior atlantoaxial fusion sacrifices motion but is technically easier.

Klippel-Feil Syndrome

This is a generalized disorder defined by Klippel and Feil (1912) based upon anomaly of the cervical spine.

Type	Features
I	Autosomal dominant mutation in growth differentiation factor 6 gene on chromosome 8q22.1. GDF6 is a member of the transforming growth factor β superfamily, regulating neural induction and patterning of ectoderm by interaction with bone morphogenetic proteins.
2	Autosomal recessive mutation in the mesenchyme homeobox 1 gene on chromosome 17q21. MEOX1 plays a rôle in mesoderm induction and regional specification, including somitogenesis as well as myogenic and sclerotomal differentiation.
3	Autosomal dominant mutation in GDF3 gene on 12p13.1. Distinguished by skeletal and ocular manifestations, including microphthalmia, iritis, and retinal coloboma.

Pathogenesis Klippel-Feil, Sprengel anomaly and Chiari malformation have been linked as defects in postotic neural crest cells, which give rise to the osseous and muscular connexions between head and shoulder girdle.

Evaluation

- Failure of segmentation of the cervical spine is the essential feature, which results in
- Short webbed neck with low hairline
- Reduced cervical motion, with or without torticollis

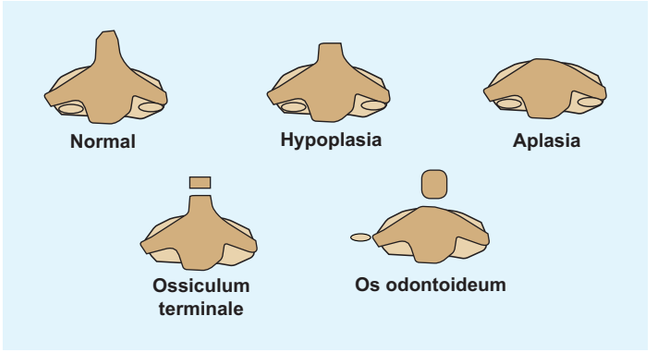
While the triad focuses on the neck [C], associated findings in up to 1/3 of patients include the following:

- Genitourinary anomalies. Screen with renal ultrasonogramme.
- Cardiac anomalies. Refer to cardiologist.
- Sensorineural and conductive deafness. Audiology testing.
- Synkinesia (involuntary or "mirror" movement associated with voluntary movement of a remote part of the body).
- Foramen magnum dysplasia associated to Chiari malformation.
- Other skeletal abnormalities, including extracervical spine deformity, facial asymmetry, Sprengel anomaly, carpal and tarsal coalition.

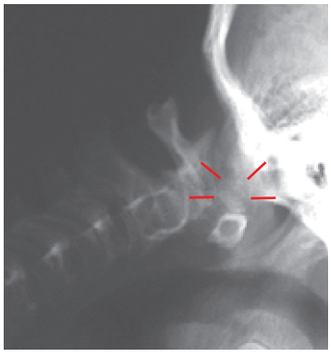
Röntgenogrammes establish failure of segmentation [D]. CT defines anomalous architecture. MRI characterizes the occipitocervical junction and associated neural lesions.

Management Educate the family to modify activity, that is, avoid contact sports. Surgical treatment is targeted at:

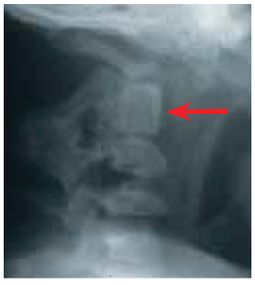
- Deformity. Due to location, rigidity, and potential for adjacent decompensation, intervention is indicated for any progression.
- Brainstem compression, as evidenced by neural dysfunction, in particular Chiari malformation and basilar impression.
- Painful adjacent segment hypermobility/instability. This rarely occurs during childhood.



A Odontoid dysplasia



B Os odontoideum Dens separate (*red*) from the body of axis retained against anterior arch of atlas. Discontinuity allows migration of atlas anteriorward.



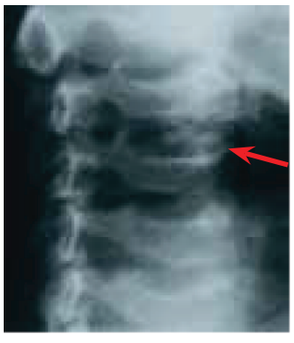
D Radiographic features of Klippel-Feil syndrome Failure of segmentation of upper cervical spine (*red*).

Cervical Intervertebral Disc Calcification

Cervical disc calcification is a rare, idiopathic condition characterized by neck pain and stiffness, with or without torticollis and fever. Röntgenogrammes show calcification. Pain resolves spontaneously by 1 month on average but may persist for several months. Calcification resolves over several years with no significant sequelæ.

Evaluation Röntgenogrammes are characteristic [A].

Management This is symptomatic, including rest, collar, and nonsteroidal anti-inflammatory agents. Recognize the benign course and resist aggressive evaluation or intervention.



A Cervical disc calcification Although cause is unknown, the disorder has a more benign clinical course than imaging might suggest.

TORTICOLLIS

Torticollis describes a "twisted neck." It has multiple lay names—*wryneck, cock-robin deformity*—because it long has been recognized, including as a symbol of beauty in Western art after the bust of Alexander the Great. It may be divided into several types [A].

Type	Comment		
Muscular torticollis	contracture of SCM		neck stiffness
Osseous	normal SCM		neck stiffness
Traumatic	pain	new onset	neck stiffness
Inflammatory	pain	new onset	neck stiffness
Tumor	pain	new onset	neck stiffness
Gastrointestinal	pain	systemic signs	normal motion
Ocular	normal SCM		normal motion
Neural	normal SCM		normal motion
Other	"idiopathic"		

A Types of torticollis SCM: sternocleidomastoid muscle.

Osseous

Pathogenesis Vertebral anomaly mechanically tilts the head.

Evaluation Röntgenogrammes of the spine early in life are difficult to interpret and will not provide sufficient detail to characterize an osseous anomaly, such as hemivertebra. CT or MRI is necessary, which will require anæsthesia. As a result, this diagnosis should be one of exclusion that is pursued if deformity is severe or progressive, if it is otherwise complicated, for example, in the setting of a syndrome, or if it will affect management.

Management This follows principles for congenital deformity correction.

Ocular

Pathogenesis This is produced by paralysis of extraocular muscle, most often obliquus oculi superior innervated (trochlear nerve), of which the primary action is intorsion (or internal rotation) to maintain horizontal vision during looking up and down. The child turns the head to keep the field of vision horizontal in compensation for the palsy.

Evaluation Diagnosis typically is made after the first year of age, after sufficient head and postural control have become established. The neck is normal, including normal motion and sternocleidomastoid muscles. On physical examination, covering the uninvolved eye will rectify a hypertropia in the affected eye; by contrast, covering the affected eye will correct a hypotropia in the normal eye.

Management Refer to an ophthalmologist.

Neural

Pathogenesis Abnormal sensory input, in particular visual in a posterior fossa lesion, or asymmetric neural signals to cervical paraspinous muscles.

Evaluation Physical examination reveals associated primary neural or secondary other system abnormalities. The difficulty of performing an accurate neurologic examination in an infant delays diagnosis. Neck motion and sternocleidomastoid muscles are normal. MRI of the head and cervical spine is the imaging modality of choice.

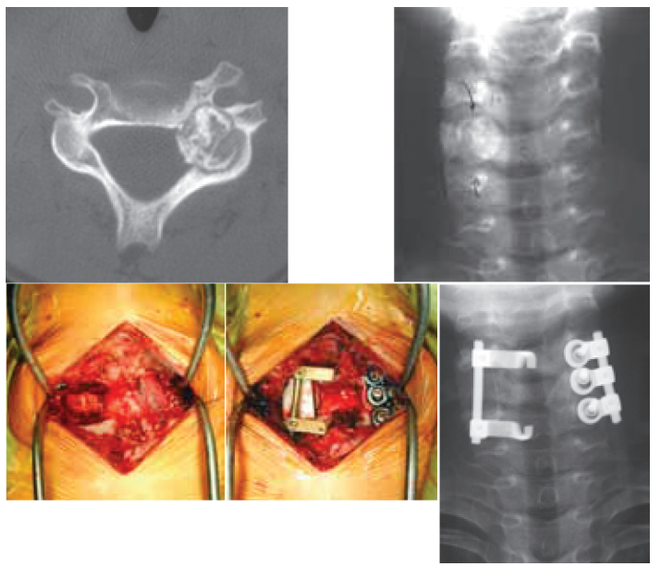
Management Refer to a neurologist.

Tumor

Pathogenesis Vertebral deformation mechanically, or inflammatory pain secondarily, tilts the head.

Evaluation Eosinophilic granuloma collapses bone into vertebra plana. This may be asymmetric, producing torticollis. Osteoid osteoma or osteoblastoma may cause a child to splint because of pain. As with osseous anomaly, röntgenogrammes of the spine may not provide sufficient detail to characterize the morbid process: CT and MRI are necessary.

Management This follows principles for benign tumors [B].



B Tumor causing torticollis Cervical osteoblastoma. Preoperative imaging shows destruction of the lateral mass, abutting the canal for the vertebral artery. This was excised *en bloc*. Spine was fused and instrumented for stability.

Gastrointestinal

This is known as Sandifer syndrome, a gastrointestinal disorder with neurologic features of which torticollis is one.

Pathogenesis Abnormal posturing occurs with gastroœsophageal reflux (e.g., hiatal hernia), against which a child splints into torticollis.

Evaluation The condition must be considered in the setting of:

- No sternocleidomastoid contracture
- Normal cervical motion
- Extracervical dystonia
- Irritability
- Other signs of gastrointestinal dysfunction such as failure to thrive

Diagnosis may be established by pH study for œsophagitis, motility study for reflux, and endoscopy with biopsy as necessary.

Management Refer to a gastroenterologist.

Other

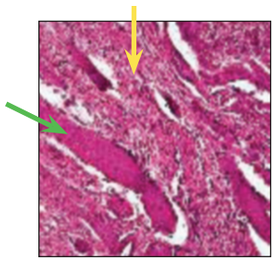
This category includes paroxysmal type.

Pathogenesis Vestibular system dysfunction has been implicated but not proven.

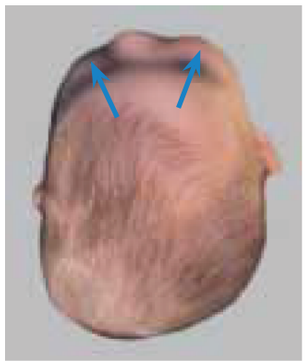
Evaluation The characteristic profile is as follows:

- Head and neck examination is normal.
- Girls are more often affected.
- It occurs in the first 3 years of life.
- The course is episodic.
- The torticollis may be bilateral.

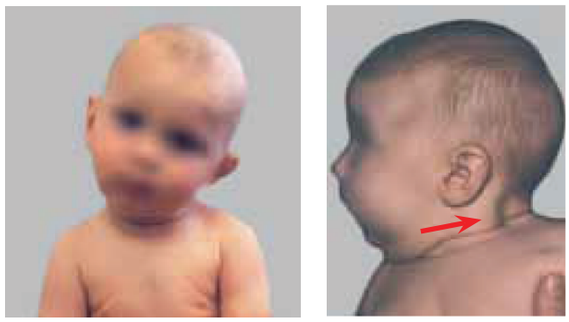
Management Educate parents and assure them that this is a benign process that resolves spontaneously without sequelæ.



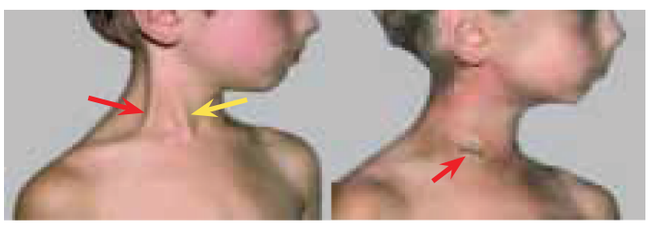
A Pathogenesis Fibrosis (*yellow*) of muscle (*green*) has led to the concept of *in utero* compartment syndrome.



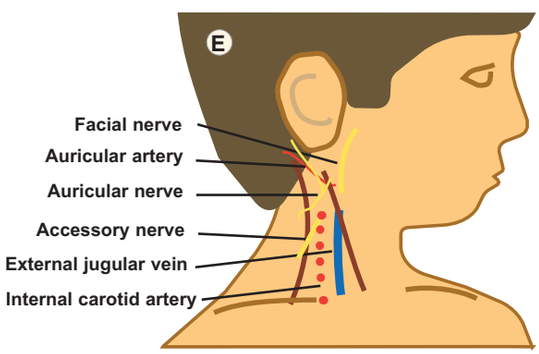
B Plagiocephaly Plagiocephaly and other "packaging" problems are associated with muscular torticollis. Arrows show malar blunting, also seen on CT performed to evaluate for osseous cause.



C Muscular torticollis The head tilts toward and rotates away from the contracted sternocleidomastoid muscle (*arrow*)



D Surgical approach Bipolar release through discreet incisions.



E Surgical anatomy This includes major and minor blood vessels, as well as motor and sensory nerves.

Muscular Torticollis

This is distinguished as

- Congenital
- Painless
- Associated with other deformities of the head, in particular plagiocephaly

It is the most common form seen by a surgeon.

Pathogenesis Compartment syndrome has been implicated, due to swelling and fibrosis of the sternocleidomastoid [A]. Evidence of a problem of "packaging" includes high prevalence in breech babies of primiparous mothers and associated signs of intrauterine crowding such as plagiocephaly [B], hip dysplasia, and metatarsus adductus.

Evaluation Contracture of sternocleidomastoid tilts the head toward and rotates the head away, in the process limiting neck motion [C]. The muscle is prominent and firm. Screen hips with ultrasonogramme or röntgenogrammes after 3 months of age. Packaging problems resolve spontaneously, as does muscular torticollis in >90% in the first year of life.

Management The benignity of natural history should temper treatment. Physiotherapy and other manipulative techniques (e.g., place cot against the wall to force the child to rotate against contracted sternocleidomastoid to look into room) in the first year are unproven. Surgical release should be delayed to allow natural history to play out. Operation consists of sternocleidomastoid section, which may be percutaneous or open, located at either or both poles, and involving excision of a small segment or Z-lengthening.

Surgical considerations There is no consensus, and approach is influenced by the following:

- Recurrence. Some believe that this may be reduced by delaying intervention. Others regard delay to be associated with poor outcome due to secondary contracture and deformity that may become permanent and that are not addressed by operation on the sternocleidomastoid, such as facial asymmetry from chronic traction. Recurrence is the reason given for bipolar (sternal, clavicular, as well as mastoid ends) section. Others counter by exsection of 1 cm of the distal poles, and include all surrounding fascia and other adhesions, which also conspire to contract the neck. Removing the entire muscle is excessive.
- Cosmetic concerns. "Percutaneous," endoscopic or open approaches are equivalent, as incisions are small and may be made transverse to the muscle along von Lager lines [D]. Concern that distal polar section or exsection may leave an ugly divot in the neck has led to Z-lengthening of the sternal head of the muscle; however, this technique adds complexity and may increase the risk of recurrence.

Anatomic considerations

- Auricular artery and nerve are at risk with release of the proximal pole [E].
- Facial nerve limits the anterior extent of a proximal incision
- The spinal accessory nerve pierces the muscle in its middle, away from any incision.
- At the heads of the sternocleidomastoid, the external jugular courses in the superficial interval. The deep interval lodges the internal carotid artery: complete release of the distal poles should leave only pulsating fat in the surgical bed, demonstrating release of surrounding fascia and adhesions, which are thought to account for recurrence after, and therefore dissatisfaction with, this unipolar approach. This may be unfamiliar territory, intimidating the orthopædic surgeon from dissecting deep.
- Raise flaps in this region of mobile skin and release all regional adhesions that may not be part of the muscle *per se* but have developed secondarily.
- Do not repair the deep fascia: augment closure of only superficial fascia and skin with glue.

Controversy continues in aftercare. Place the patient in a soft collar. Allow the wound 1 week to heal, then mobilize the neck under supervision of a physiotherapist until the patient has comfortable and full motion.

Atlantoaxial Rotatory Displacement

This is distinguished as:

- Acquired. It may follow inflammation of soft tissue of the neck, or trauma.
- Painful.
- Involvement of the contralateral sternocleidomastoid muscle, which contracts to rectify the neck and is elongated.

This is typed [A]:

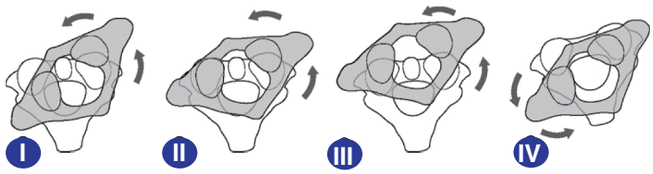
- I. Rotation without displacement.
- II. Rotation with ≤5-mm displacement.
- III. Rotation with >5-mm displacement.
- IV. Rotation with posterior displacement. This may include fracture of dens axis.

Pathogenesis Venous drainage of the pharynx toward the periodontoidal vertebral plexus, where there are no lymph nodes to divert flow, leads to inflammatory hyperæmia that produces ligamentous laxity and spasm of paraspinous muscles. Nontraumatic torticollis first was described by Bell (1830); it is named after Grisel (1930), who implicated venous drainage and described enucleation of the atlas to correct the deformity. Traumatic extreme motion of the neck may shift and squeeze meniscus-like synovial membrane between atlas and axis, resulting in pain inhibition and a mechanical block to reduction.

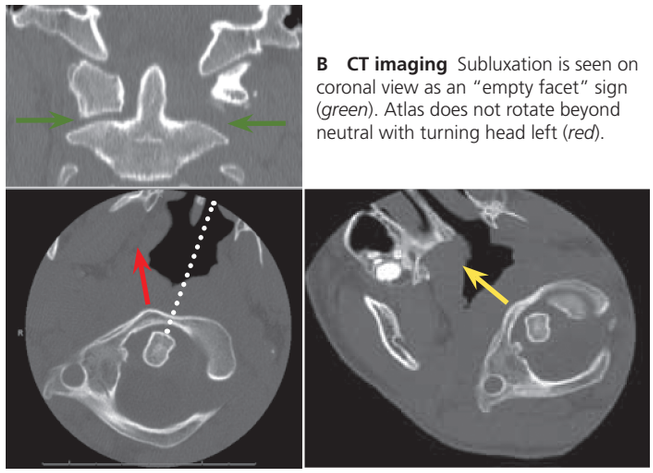
Evaluation There is a history of infection or trauma; the latter may be activity related (e.g., wrestling) or may occur after head and neck surgery. The contralateral sternocleidomastoid muscle is contracted, painful, and elongated. Neck twisting may distort röntgenogrammes: the most useful view is a lateral of the skull (with which atlas is reduced), which may show increased atlantoaxial interval. CT will show displacement, and dynamic CT will show fixed or incomplete rotation of atlas relative to axis [B].

Management Infectious AARD resolves with treatment of the infection. Support the neck in a collar. This may be the most common form of torticollis, as it often is managed by the pædiatrician without involvement of the orthopædic surgeon. Traumatic AARD may persist. Correction or stabilization of deformity is indicated to eliminate risk of neural injury [C].

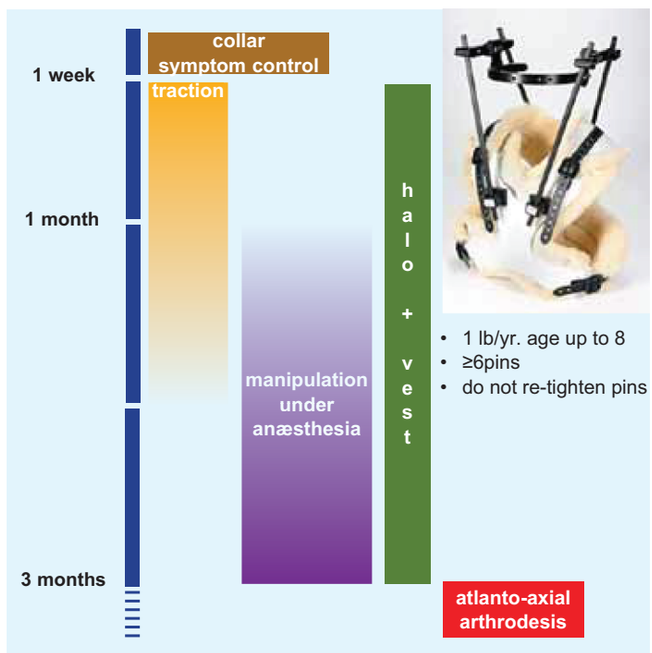
- 0 to 1 week. Soft collar and symptom control, such as rest and anti-inflammatory medication. The majority resolves clinically; imaging is not necessary given the benign and typical course.
- 1 to 4 weeks. Reduction. Start with traction in hospital, aided by relaxants. While halo is more invasive, it allows rotational as well as longitudinal force, and halter risks chin ulcer. Confirm reduction with CT. After reduction, halo–vest is worn for 3 months to reduce resubluxation.
- >1 month. Chronic AARD is unlikely to resolve. There is a gray zone of time during which some advocate manipulative reduction under anæsthesia. The head is held by halo and rotated gently, slowly, and fully with neural monitoring.
- >3 months. Persistent AARD despite closed methods is treated by atlantoaxial fusion. Sublaminar wiring of an H-graft at atlas with axis spinous process wiring (Gallie) or atlantoaxial sublaminar wiring (Brooks) introduces implants into a canal of which the volume may be reduced by the subluxation. Transarticular screws from axis into atlas (Magerl) avoid this, are more stable, and may be reductive [D].



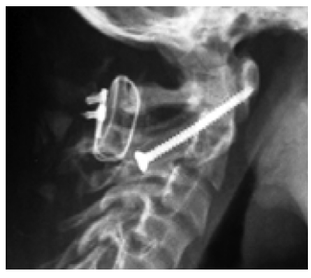
A Types of atlantoaxial rotatory displacement Scintigramme at presentation shows increased uptake (*red*). After 2 weeks, lateral röntgenogrammes shows narrowing of the disc (*yellow*).



B CT imaging Subluxation is seen on coronal view as an "empty facet" sign (*green*). Atlas does not rotate beyond neutral with turning head left (*red*).



C Treatment protocol for atlantoaxial rotatory displacement



D Atlantoaxial arthrodesis Brooks fusion combined with Magerl transarticular screws.

ASSOCIATED NEURAL DISORDERS

Neural disorders may be associated with spine and other musculoskeletal deformity, such as cavus or unilateral calf asymmetry, vertebral anomaly, pain, and mixed upper and lower neuropathy.

Evaluation After physical examination, MRI is the imaging modality of choice. CT (with myelography) characterizes osseous architecture, including vertebral canal and for severe deformity, in preparation for operation.

Management Neurosurgical. Occipitocervical decompression of Chiari malformation. Section of thickened and contracted filum terminale to release tethered cord. Fenestration and drainage of syrinx. Resection of septum and release of adhesions for diastematomyelia.

Chiari Malformation

Brainstem compression associated with hindbrain and other neural anomalies [A]. Type I is defined as displacement of cerebellar tonsils >3 mm below foramen magnum. Type II is associated with myelomeningocœle.

Syrinx

This is defined as a morbid cavity in the spinal cord distinct from dilatation of the central canal, which is termed hydromyelia. They may occur in conjunction with Chiari malformation and will resolve with foramen magnum decompression. Treatment depends upon symptoms and size. The asymptomatic patient with a small syrinx is followed with serial MRI. Large syringes may require fenestration and subarachnoid, peritoneal, or pleural shunting [B].

Tethered Cord

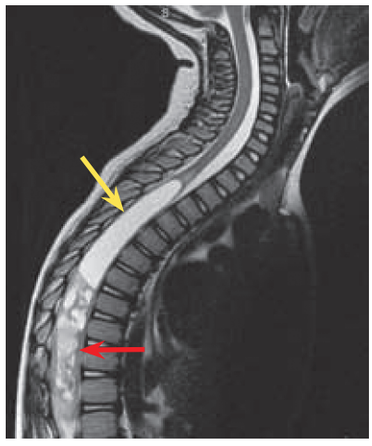
The conus medullaris is retained below L2 by a contracted filum terminale [C]. The filum terminale is thickened such that it can indent the theca, and fatty, including formation of a fibrolipoma that appears as a mass on MRI.

Diastematomyelia

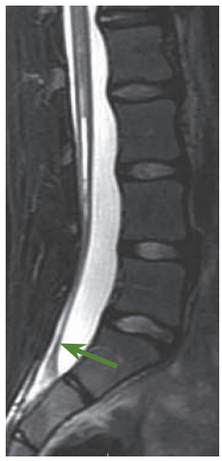
The term refers to a "through" "split" of the "spinal cord." There are two hemicords, in contrast with diplomyelia, in which the cord is duplicated. Girls are affected thrice as often. Most lesions occur in the thoracic cord and are connected with a stigma of dysrrhaphism such as hairy patch [D].

Chiari type	Features
I	Cerebellar tonsils below foramen magnum
II	Multiple hindbrain anomalies
III	Occipitocervical encephalocœle
IV	Absence of cerebellum

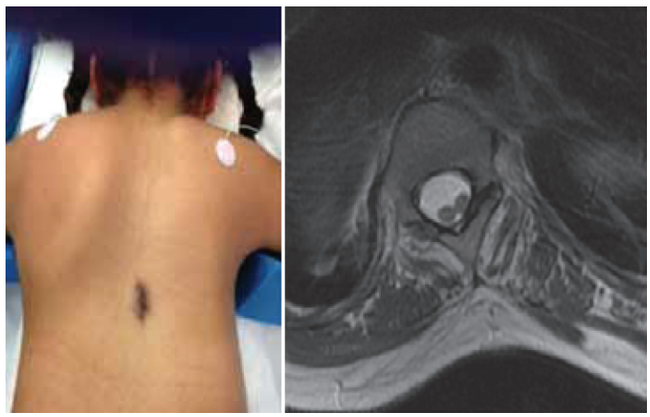
A Chiari malformation Types and features.



B Syrinx and tumor Six-year-old boy presented with scoliosis associated with hyperkyphosis. MRI shows tumor (*red*) and syrinx (*yellow*).



C Tethered cord Conus medullaris, with syrinx, is tethered to the level of L4 by a thickened filum terminale (*green*).



D Diastematomyelia Hairy patch associated with thoracic lesion.

TUMOR

Primary tumors may be divided into benign or malignant and may affect the hard tissue—bone—or soft tissue—neural elements [A]. Less than 10% of primary bone tumors occur in the spine, of which half occur in childhood. Vertebral tumors may affect the body or the neural arch [B]. The latter may produce radiculopathy, thereby mimicking disc herniation. Tumors of the spine may produce deformity, in particular scoliosis, which may be distinguished by:

- Focal pain
- Marked reduction in motion
- Rapid progression
- Absence of rotation

Cervical lesions may present with torticollis (*q.v.*).

Surgical treatment may be:

- Intralesional excision, such as curettage and bone grafting. While appropriate for benign tumors, this may be complicated by recurrence in aggressive lesions such as osteoblastoma.
- *En bloc* resection. This reduces recurrence but may risk surrounding neurovascular structures (e.g., vertebral artery in the cervical spine, the dura mater requiring patch grafting) and may produce instability (in particular kyphosis) necessitating fusion.

Consider contemporaneous spine fusion with instrumentation for *en bloc* resection, complete laminectomy, facetectomy. Follow partial excision for postsurgical deformity, in particular kyphosis in the thoracic spine.

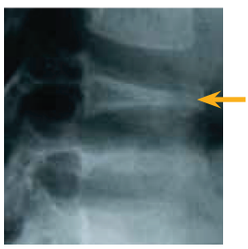
Benign Osseous Tumors

Osteoid osteoma and osteoblastoma Osteoid osteoma is defined as <2 cm and is less aggressive than osteoblastoma, which most often is located in the spine. Pain is worst at night. The tumor releases prostaglandins, which explains its inflammatory nature and pain; prostaglandin inhibitors (e.g., ibuprofen) are both diagnostic and therapeutic, although not in all children. Delay in diagnosis can be longer than a year, in part because more than half are invisible on röntgenogrammes. SPECT shows intense uptake. CT will show the pathognomonic target lesion, consisting of a sclerotic margin surrounding a separate sclerotic nidus (hence the old term "sclerosing nonsuppurative osteomyelitis"). This modality localizes the lesion for radioablation or laser coagulation [C] and defines architecture for surgical resection.

Aneurysmal bone cyst Röntgenogrammes show an expansile heterogeneous geographic lesion [D]. Preoperative embolization reduces operative blood loss.

Eosinophilic granuloma Ten percent of lesions occur in the spine. Replacement of vertebral body by tumor results in collapse into vertebrae plana [E]; recognition of this deformity will aid in differentiation from more aggressive tumors, such as Ewing sarcoma. Indication for surgical intervention is neural deficit resulting from large or multiple lesions. The natural history of the majority of lesions, which are solitary and associated with normal neural function, is spontaneous regression with reconstitution of vertebral height. Management is supportive, including bracing. Residual osseous deformity usually is acceptable.

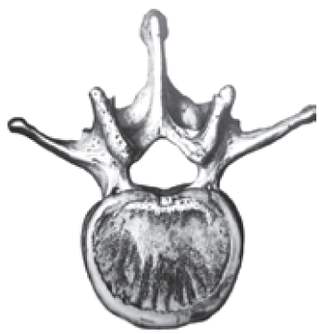
Osteochondroma This has a predilection for the cervical spine. Ten percent of patients with hereditary multiple exostosis have spine involvement. Symptoms are due to mass effect, including pain and neural deficit, which may be successfully treated with resection.



E Eosinophilic granuloma Vertebra plana, with preservation of the intervertebral disc.

Osseous Tumors	Neural Tumors
Benign	**Benign**
Eosinophilic granuloma	Neurofibroma
Osteoid osteoma	Lipoma
Aneurysmal bone cyst	Spinal cysts
Osteoblastoma	**Malignant**
Neurofibromatosis	Astrocytoma
Osteochondroma	Ependymoma
Malignant	Mixed glioma
Ewing sarcoma	Ganglioglioma
Lymphoma of bone	
Leukæmia	

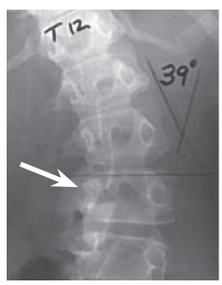
A Vertebral (bone) and spinal cord tumors

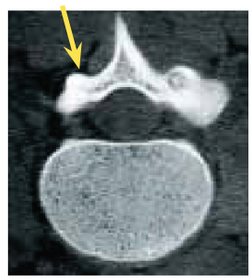


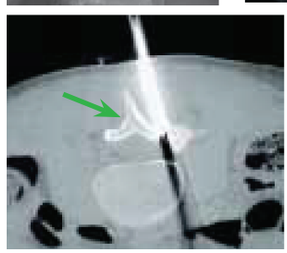
Posterior elements
Osteoid osteoma
Osteoblastoma
Aneurysmal bone cysts
Osteochondroma

Anterior elements
Eosinophilic granuloma
Giant cell tumor
Hæmagioma

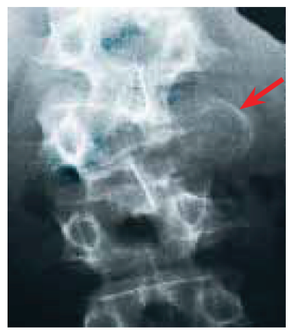
B Geographic distribution of benign osseous tumors Tumors may affect the thoracolumbar spine (*green*) or the cervical spine (beige). Other tumors that affect the spine do not occur in children (*gray*).



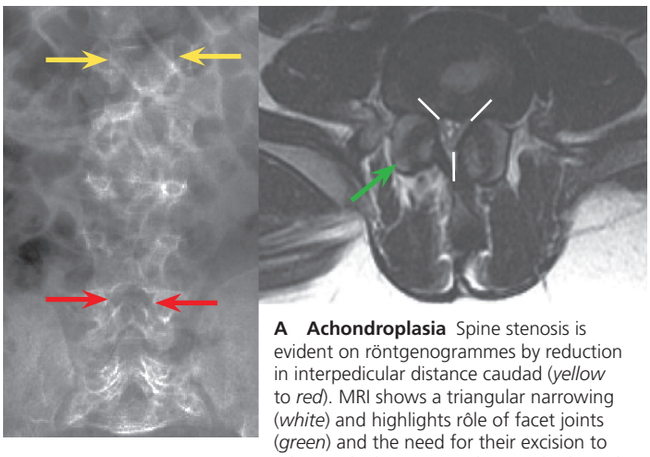




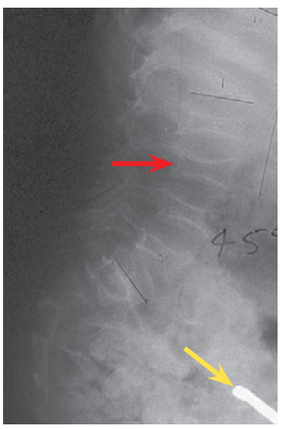
C Osteoid osteoma The lesion produces nonspecific sclerosis of the pedicle on röntgenogramme (*white*) but is well defined on CT (*yellow*) and is treated with radioablation (*green*).



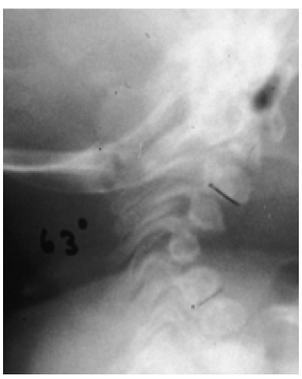
D Aneurysmal bone cyst Expansile lesion with sclerotic margin located at the concave apex of a painful secondary scoliosis.



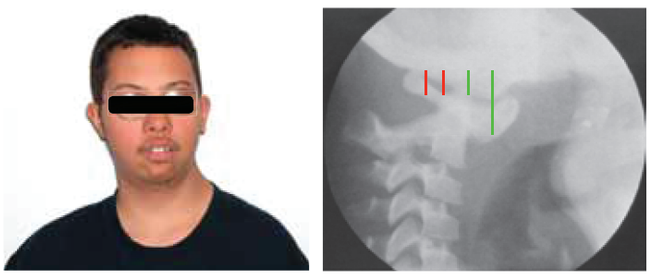
A Achondroplasia Spine stenosis is evident on röntgenogrammes by reduction in interpedicular distance caudad (*yellow* to *red*). MRI shows a triangular narrowing (*white*) and highlights rôle of facet joints (*green*) and the need for their excision to adequately decompress the vertebral canal.



B Osteogenesis imperfecta Codfish vertebrae (*red arrow*) are characteristic. Note the femoral Rush rod (*yellow*).



C Diastrophic dysplasia Cervical kyphosis is characteristic and grave. Neural risk is an indication for early surgical intervention.



D Down syndrome Torticollis is a sign of atlantoaxial instability, as patient splints to stabilize cervical spine. Operative image shows space available for cord is reduced (*red*) as atlantodental interval increases (*green*).

Spine in Other Disease

The spine is affected characteristically in the skeletal dysplasias and in several syndromes. General principles of spine deformity include the following:

- Increased incidence
- Increased magnitude
- Increased progression, earlier onset, more and more rapid
- Increased neural risk, from disease and operative treatment
- Increased technical demand, including more frequent anterior fusion in addition to posterior fusion and more complications.

Achondroplasia

Lumbar spine Stenosis is the most common spine problem in achondroplasia. It results from hypoplasia of the neural arch and may be seen on röntgenogrammes as a reduction in interpedicular distance caudad in contrast with normal increase [A]. Reduction in vertebral canal volume and intervertebral foramen diameter is exacerbated by lumbar hyperlordosis, which in turn is worsened by thoracolumbar kyphosis or hip flexion contracture. Children typically present in the second decade: there is a spectrum of pain, claudication, radiculopathy, and neural deficit, including abnormal urodynamics. Flexion of the lumbar spine, such as interrupting play to squat, provides relief by increasing space available for nerves. Treatment consists of decompression and fusion with instrumentation to the pelvis. There is not enough space for laminectomy without disruption of facet joints to suffice.

Foramen magnum By contrast with the flat bones of the skull, which form by intramembranous ossification, this part of the skull forms by endochondral ossification, which is abnormal in achondroplasia. Foramen magnum stenosis may be treated based upon absolute diameter compared with published normative CT data for achondroplasia. Functional treatment is based upon demonstration of an abnormal sleep apnea study. Foramen magnum decompression with ventricular drainage improves sleep study metrics.

Kyphosis This most commonly affects the thoracolumbar junction. The natural history includes spontaneous improvement during infancy for flexible deformity. Kyphosis (no consensus on magnitude, certainly >50 degrees) persistent beyond 5 years of age is treated with single-stage circumferential fusion performed *via* a posterior approach and including circumferential instrumentation.

Osteogenesis Imperfecta

Severity and incidence of spine deformity is proportional to severity of disease. Biconcave vertebral bodies are characteristic and known as "codfish vertebra" [B].

Basilar impression This is a feature of type III. It is defined as tip of dens above McRae line on lateral röntgenogramme. The base of the skull settles from the level of the atlanto-occipital joints to the level of atlantoaxial joints on open mouth view. Because this occurs gradually, most patients are asymptomatic. Symptoms and signs relate to brainstem and upper cervical cord compression. Fundamental to treatment is surgical decompression.

Scoliosis Cause relates to loss of extracellular matrix integrity, including osseous fragility and ligamentous laxity. There is a relative lack of rotation and associated hyperkyphosis [B]. Early operative intervention for progressive curves <50 degrees is motivated by poor purchase of anchors and primary distortion of spine architecture.

Fragilitas ossium In its most subtle form, osteogenesis imperfecta may be silent during childhood and manifest as premature osteoporosis in the adult, including vertebral compression fracture.

Diastrophic Dysplasia

The name was proposed to highlight "twisting" of feet and spine, including scoliosis, lumbar hyperlordosis, and kyphosis. Risk of neural injury from cervical kyphosis necessitates early surgical intervention [C]. Anterior fusion with strut grafts addresses kyphotic tension on posterior fusion and frequent deficiency of posterior elements.

Atlantoaxial Instability

Altlantoaxial instability (AAI) is a feature of Down syndrome, several skeletal dysplasias (pseudoachondroplasia, type II collagenopathies), and the mucopolysaccharidoses. Pathogenesis includes connective tissue laxity and odontoid dysplasia.

Down syndrome In 1983, Special Olympics recommended radiographic screening before participation. This position was endorsed in 1984 by the American Academy of Pediatrics. Because patients with Down syndrome can move spontaneously between radiographic stability and instability without clinical manifestation and because all reports of neural injury from AAI demonstrated at least several weeks of prodrome, in 1995, the recommendation for screening was retired. In addition, surgical treatment has a high complication rate, making clear indications essential. Educate caregivers about symptoms and signs of AAI, including deformity (in particular torticollis), stiffness, myelopathy [D]. Follow patients clinically. Absent clinical manifestation, screening is not recommended lest it yield a radiographic false positive. The moderate position consists of clinical observation for ADI 5 to 10 mm and atlantoaxial posterior spine fusion with instrumentation for ADI >10 mm.

Type II collagenopathies and mucopolysaccharidoses While these two groups contain distinct diseases, which may include other spine deformity such as scoliosis and kyphosis [E], odontoid dysplasia resulting in AAI is the most significant spine abnormality, both in its high incidence (up to 1/2 of patients) and treacherous consequences. Abnormality of type II collagen produces spondyloepiphysial dysplasia and Kniest syndrome; lysosomal enzyme deficiency forms the basis of Morquio and Hurler syndromes. Upper cervical cord compression may be mistaken for typical delay in motor milestones resulting from generalized skeletal dysplasia, including lower limb deformity. Abnormal ossification makes röntgenogrammes difficult to interpret. Dynamic MRI shows the dens and vertebral canal, including constriction and any signal change in the spinal cord. Because the dens is dysplastic, follow reduction of atlas on axis during tightening of implants with image intensification to guard against posterior displacement of atlas.

Marfan Syndrome

Scoliosis This is a major diagnostic criterion. Approximately 1/2 of patients will develop scoliosis >30 degrees. Bracing is ineffective. Treat according to general surgical principles.

Dural ectasia This is one of the major diagnostic criteria. It is significant clinically in genesis of back pain. Its surgical significance rests in the fact that it results in osseous erosion, which undermines anchors and may account for high historic rates of pseudarthrosis due to instability of constructs. MRI is essential to evaluate this [F].

Spondylolisthesis This is part of the differential diagnosis of back pain. Olisthy is attributed to connective tissue laxity. Treat according to general principles.

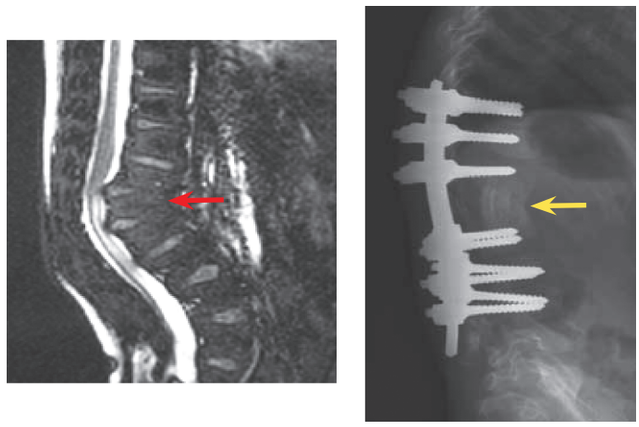
Neurofibromatosis

The spine is affected in type 1 (of von Recklinghausen).

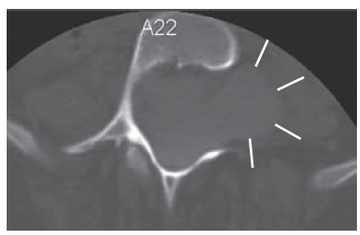
Scoliosis Scoliosis affects approximately 1/3 of patients. It may resemble idiopathic scoliosis, or it may be dystrophic [G], of which characteristics include the following:

- Short sharp curvature.
- Severe rotation.
- Osseous erosion by neurofibromata, which distorts anatomy, is destabilizing to the extreme of dislocation and impairs fixation.
- Rapid progression, which dictates early surgical intervention.

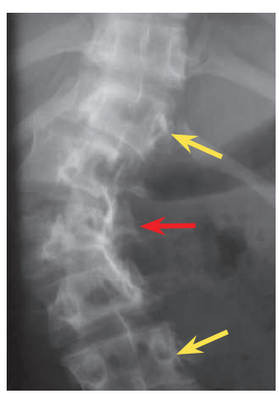
Kyphosis This is associated with dystrophic scoliosis. It also may occur in the cervical spine, where potential for severe angulation and instability dictates aggressive surgical management.



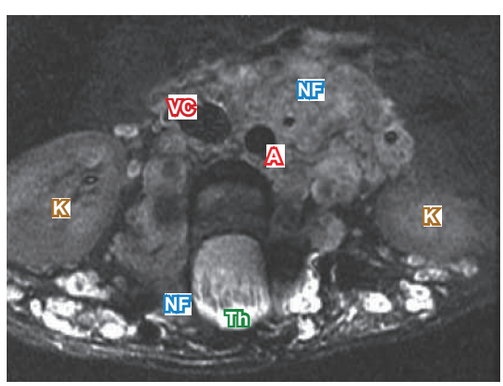
E Hurler syndrome Six-year-old girl with progressive thoracolumbar kyphosis (*red*) was treated by posterior approach vertebral column resection, anterior strut grafting (*yellow*), and posterior instrumentation.



F Marfan syndrome Dural ectasia eroding pedicles, thereby compromising fixation.



G Neurofibromatosis Note the severe rotation (*red*) two segments away from relatively neutral vertebrae (*yellow*) in this dystrophic scoliosis.



H Neurofibromatosis Paraspinous neurofibroma (NF) has eroded posterior elements such that they cannot accept implants for fixation, and allowing theca (Th) to balloon. K: kidney, A: aorta, VC: vena cava.

Pseudarthrosis, a fundamental feature of neurofibromatosis, necessitates circumferential fusion for spine deformity, in particular the dystrophic type. CT defines osseous architecture, while MRI defines neurofibromata and neural architecture [H]. The latter is essential to reducing risk during spine manipulation from traction upon intraspinal lesions.

Caudal Dysgenesis

This also is known as caudal regression syndrome and sacral agenesis. It represents a heterogeneous constellation of anomalies affecting the caudal spine and spinal cord, pelvic and abdominal viscera, and the lower limbs. It may be divided into without or with abnormal sacrum, the latter into without or with instability [I].

Evaluation One-fifth of mothers of affected children have insulin-dependent diabetes mellitus. Presentation includes multiple contractures of the lower limbs, dislocation of the hips, clubfoot, scoliosis, and kyphosis. Musculoskeletal deformities vary in severity with level of agenesis and resulting neural loss. Motor loss is disproportionate to sensory loss, which can remain protective despite significant severity. Cognition is spared. Appearance of severe contractures of the lower limbs has been likened to Buddha posture. There are associated anomalies of the hindgut and the urogenital system. Currarino syndrome represents an autosomal dominant form of the disease, caused by a mutation in the HLXB9 homeobox gene on chromosome 7q36.

Management Neural status guides management. While the presentation can be dramatic, the most prudent approach is restraint. Painless instability is better than painful stiffness. Deformities recur despite operative correction because contractures are tenacious. Aggressive surgical treatment is indicated for the ambulatory. For the others, limited operative procedures to aid posture and care, combined with orthotic support and mobility aids are tailored to the child.

Sacral Dimple

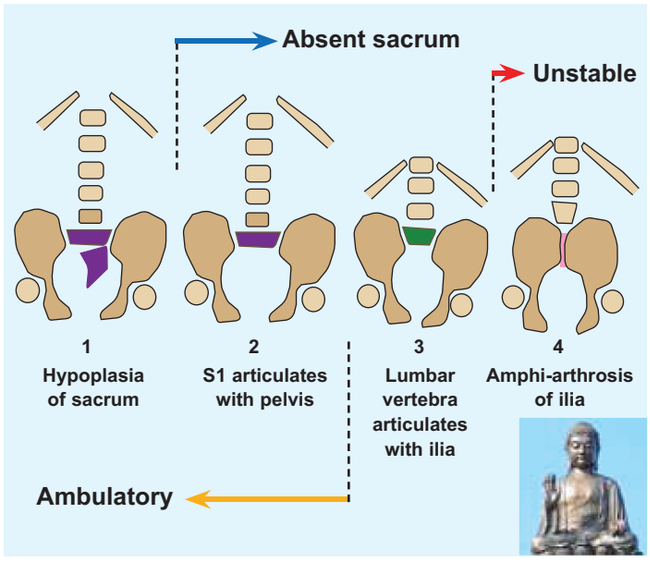
This represents an indentation, or "pit," that may be observed during the neonatal physical examination at or within 1 inch of the natal cleft, which may be distorted. It affects 3% of children.

Evaluation The benign anomaly must be differentiated from a midline sign of dysrrhaphism, such as hair, sinus, cutaneous tag, altered pigmentation, drainage.

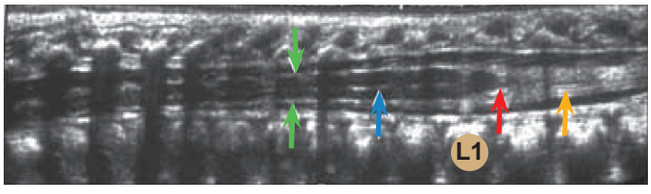
Management Diagnostic dilemma may be resolved in the neonate by ultrasonogramme to evaluate the subjacent neural elements [J], in particular level of the conus medullaris and thickening of filum terminale tethering the spinal cord. After 4 months of age, ossification of the posterior elements of the spine obscures ultrasonogramme, necessitating MRI. Because the latter requires general anæsthesia in this age, be prudent in obtaining this.

Spinal Cord Injury

Spine deformity is the rule if spinal cord injury occurs before puberty. All children develop scoliosis, while 2/3 will develop kyphosis and 1/5 will develop lumbar hyperlordosis. The spine deformity tends to be progressive and most will require spine fusion with instrumentation including the pelvis. Less than 1/5 of children who sustain spinal cord injury after puberty will develop a paralytic deformity.



I Sacral agenesis The sacrum may be hypoplastic or absent (*blue*). The spine–pelvis relationship may be stable or unstable (*red*).



J Ultrasonogramme of neonatal spine for sacral dimple Spinal cord (*green*) and central canal (*blue*) are normal, as is level of conus medullaris (*red*) between L1 and L2. Cauda equina (*orange*) shows no thickened filum terminale.

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