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Chapter 46.3.6 Paediatric and adolescent spinal deformity

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Part of "Chapter 46.3 - Paediatric orthopaedics"

Introduction

Spinal deformity can develop during embryogenesis (congenital scoliosis, kyphosis, myelodysplasia) or during growth in association with disordered development, neuromuscular disorders, or bony dysplasia. Table 1 lists the more notable causes of spinal deformity. Many spinal deformities worsen with growth, but some also spontaneously improve or stabilize. Although often asymptomatic during growth, spinal deformities may have profound implications for adulthood. Progressive spinal deformity may lead to severe cosmetic distortion, pain, difficulty with seating balance, respiratory failure from restrictive lung disease, and in some congenital deformities, spinal cord impingement. Understanding the natural history of untreated spinal

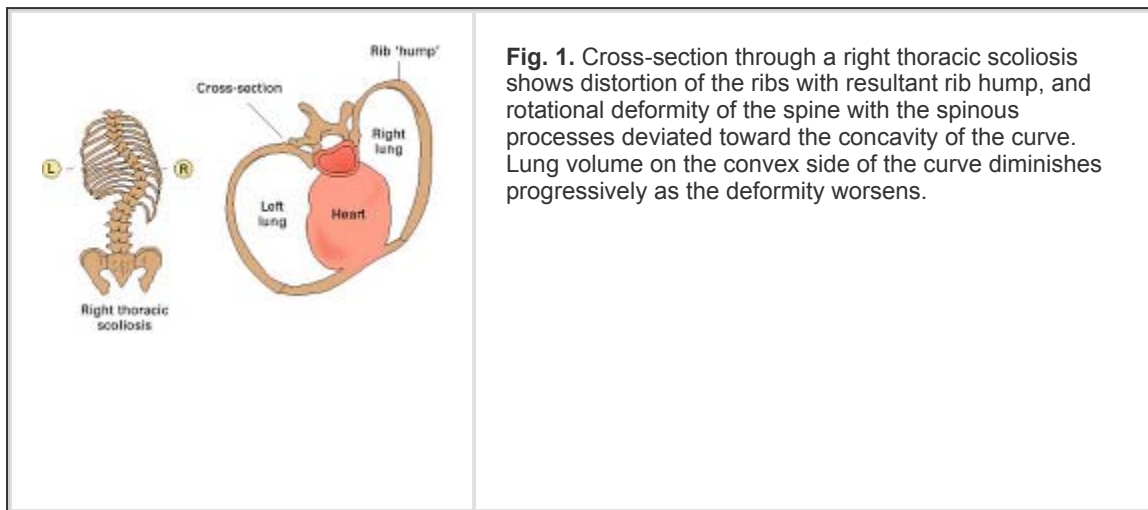
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deformity is crucial to choosing appropriate treatment, as deformities of differing etiologies and differing severity vary widely in their behavior during growth and adulthood as well as their response to treatment.

<p>Congenital</p> <p>Idiopathic or developmental</p> <p>Neuromuscular</p> <p>Bone dysplasia</p> <p>Associated with:</p> <p>Secondary to:</p>	<p>Scoliosis Kyphosis Klippel-Feil deformity Sacral agenesis Segmental spinal dysgenesis</p> <p>Mitochondrial scoliosis Scheuerman's kyphosis Spina bifida Spina bifida</p> <p>Cerebral palsy Myelodysplasia Muscular dystrophy Spinal muscular atrophy Friedreich's ataxia Charcot-Marie-Tooth hereditary sensorimotor neuropathy</p> <p>Spina bifida Dysostosis Osteogenesis imperfecta</p> <p>Neurofibromatosis</p> <p>Laminectomy Thoracotomy Burs</p>	<p>Table 1 Partial list of childhood and adolescent spinal deformities</p>
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Spinal deformities are complex three-dimensional disorders (Fig. 1) with deformity in the coronal (scoliosis) and sagittal (kyphosis, lordosis) planes as well as axial rotational and sometimes translational deformity. Multidimensional classifications and measurement systems exist, but in clinical practice deformities are referenced in the coronal and sagittal planes and simple estimates of rotational deformity made. Scoliosis refers to curvature with

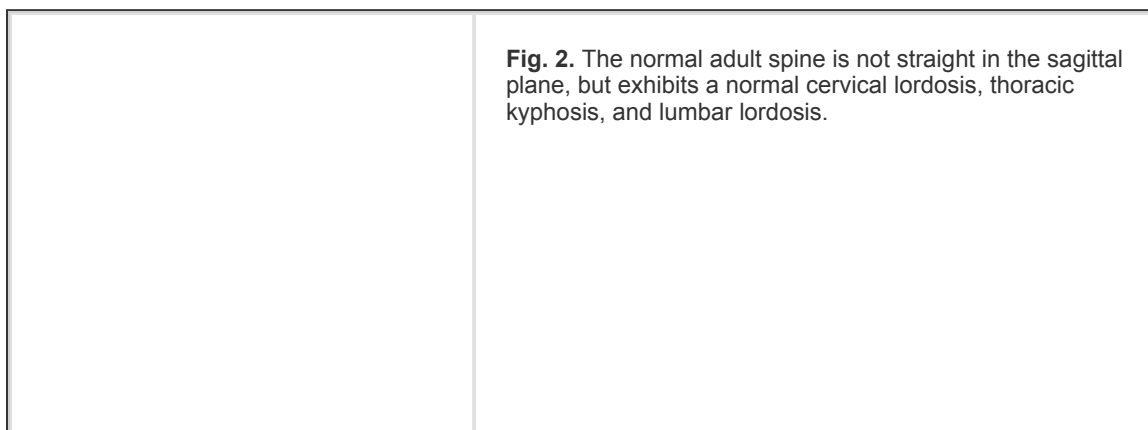
the apex laterally, kyphosis posteriorly, and lordosis anteriorly. For a given curve the etiology, direction, and apical vertebra define the curve. A typical idiopathic scoliosis might be described as 'right thoracic idiopathic lordoscoliosis with an apex at T5'.



Normal spinal growth

Formation of the spinal column is an early and fundamental embryologic event. Somite and vertebral development are intimately related, hence visceral or neural malformations are commonly associated with congenital vertebral anomalies. Most vertebrae form from several discrete ossification centers. The cartilaginous neurocentral synchondroses joining early vertebral body ossification centers with those of the lamina are visible in infant spine radiographs and may be confusing. By the age of 5 years neurocentral SYNchondrosis closure is completed and the diameter of the spinal canal has reached nearly adult dimensions.

The normal adult spine is not perfectly straight, but exhibits a normal cervical lordosis, thoracic kyphosis, and lumbar lordosis (Fig. 2). The infant spine is a long, gradual, C-shaped kyphosis. Normal adult cervical lordosis, thoracic kyphosis, and lumbar lordosis develop in sequence as the infant progressively develops head control, sitting, and standing. Thoracic kyphosis increases through childhood to reach a normal adult range of 20 to 50°. Mild scoliosis of less than 10° in the adult is common and considered normal.





Assessing spinal deformity

Physical findings

Severe spinal deformity with distortion of the chest and waist is immediately apparent to the observer. Subtle signs of spinal deformity include asymmetry in neck or waist contour, shoulder height, scapula position and prominence, and apparent leg length difference (Fig. 3). Single curves are more visible, as are curves with large rotational deformities and rib 'humps' (Fig. 4). Imbalance may occur and refers to a shift of the head or trunk laterally as compared with the pelvis.

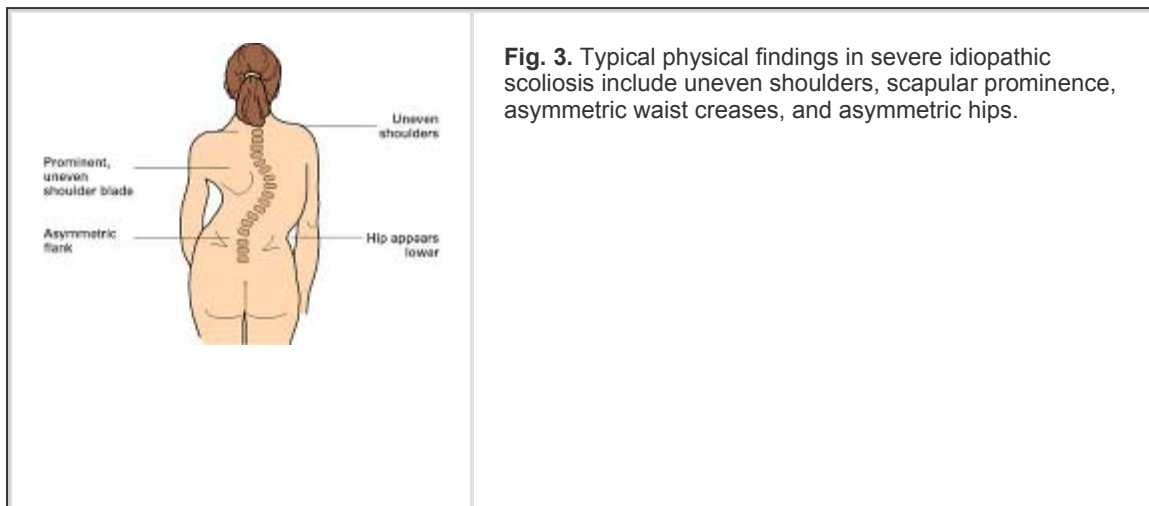


Fig. 3. Typical physical findings in severe idiopathic scoliosis include uneven shoulders, scapular prominence, asymmetric waist creases, and asymmetric hips.

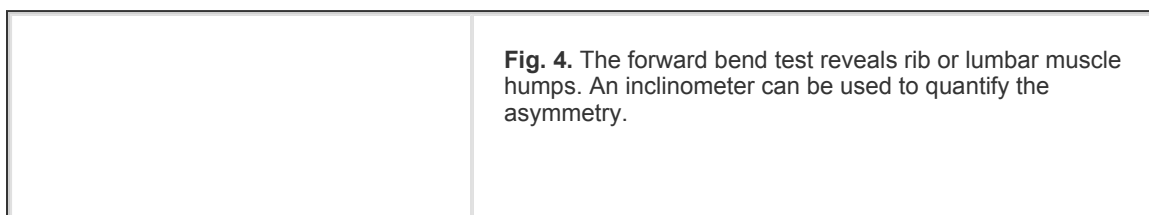


Fig. 4. The forward bend test reveals rib or lumbar muscle humps. An inclinometer can be used to quantify the asymmetry.



Physical examination for spinal deformity includes an assessment of asymmetry and a forward bend test in which the subject bends forward, with hands together, allowing the examiner to see along the back, searching for rib or lumbar humps to measure with an inclinometer (Fig. 4). The magnitude of the rotational asymmetry or hump usually reflects the severity of the curvature, but severe curves may have minimal rotational deformities and vice versa. Deviations from the normal cervical lordosis, thoracic kyphosis, and lumbar lordosis are noted. Leg length is assessed. A screening neurologic examination of reflexes and strength is completed. Signs of root tension such as pain on neck flexion or straight leg raising, tight hamstrings, foot deformity, atrophy of one leg, or associated hairy patches or dimples over the spine may be signs of an underlying spinal cord abnormality. Hip contractures may accompany long-standing severe scoliosis, particularly neuromuscular curves. Flexibility of a scoliosis is assessed by sideways bending, kyphosis and lordosis by flexion and extension.

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Radiographic interpretation

Radiographs are an essential part of spinal deformity evaluation. Most simple idiopathic deformities need only standing posteroanterior views, but severe, non-idiopathic, or painful scoliosis and deviations in the sagittal plane (kyphosis, lordosis) also require lateral views. Preoperative planning may dictate bending views. CT scan with three-dimensional reformatting better defines complex deformities. MRI is useful for assessment of neuroanatomy or disc disease. Posteroanterior are generally preferred to anteroposterior views as the absorbed dose of radiation in growing breast and thyroid tissue is diminished with the former exposure. Full-length films, high-speed films, and special grids enhance visualization of the vertebrae.

Curve severity is measured using the Cobb angle in both the coronal and sagittal plane. A perfectly straight spine has a Cobb angle of 0° . The Cobb angle refers to the angle formed between the top of the uppermost tilted vertebra in a curve and the bottom of the lowermost tilted vertebra. Intra- and interobserver error with Cobb angle measurement can be large: 7 and 13° in idiopathic and congenital scoliosis, respectively.

Prediction of progression in scoliosis depends in part on the magnitude of growth remaining. The 'Risser sign' refers to the extent of ossification of the iliac crest apophysis and is used as a guide to growth remaining when treating idiopathic scoliosis. A hand and wrist radiograph may be obtained for assessment of bone age.

Principles of non-operative treatment

Non-surgical treatment of spinal deformity during growth may have different goals, methods, and efficacy depending on the etiology, severity, or location of the deformity. Active exercises are used in postural disorders and as an adjunct to scoliosis or kyphosis bracing. Exercises may be directed at the spine itself or at extremity contractures such as hip flexion deformities or hamstring contractures. Exercises may consist of stretching or strengthening or postural awareness.

Spine braces or external orthoses such as the Milwaukee or Boston brace are used in progressive idiopathic scoliosis to halt worsening (progression) of the deformity. Except in milder kyphotic deformities and some idiopathic scoliosis it is less common to achieve substantial long-term correction of deformities through orthoses alone. Semi-rigid or total-contact external orthoses are used in neuromuscular scoliosis such as cerebral palsy to facilitate seating and function. Rigid lumbosacral orthoses are used in spondylolysis to relieve pain or encourage healing of a pars interarticularis stress fracture. Orthoses act on the spine via pressure applied to the skin, soft tissues, ribs, and abdomen and the corrective force transmitted to the spine is limited by the tolerance of these tissues. In spinal deformities where patient sensation or neuromuscular control is limited, external orthoses may be poorly tolerated.

Principles of operative treatment

Indications

Pain, continued worsening (progression), neurologic compromise, predicted pulmonary compromise, or severe cosmetic deformity may all be indications for operative treatment of a spinal deformity. Operative treatment may be performed for present problems in the growing spine or in anticipation of problems with the spinal deformity as an adult.

Spinal fusion

Spinal fusions are commonly performed. By achieving a solid fusion or arthrodesis the deformed section of spine is permanently stabilized and will not continue to deform with growth or aging. Choosing the extent of the fusion is critical. The length of spine fused is kept to a minimum to preserve as much normal spinal motion as possible, yet the fusion must include enough of the deformed spine to preclude future worsening of the deformity. The extent of fusion depends upon etiology, curve type, curve severity, and correction gained with instrumentation. Fusion is achieved by exposing and denuding (decorticating) the surfaces of adjacent vertebrae and applying bone graft. Instrumentation to stabilize and correct the deformity is often applied at the same time.

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Fusions can be performed posteriorly, anteriorly, or both. Posterior fusions include exposure of the entire lamina and transverse processes, while anterior fusions involve complete intervertebral disc and endplate excision to expose the adjacent raw cancellous surfaces of vertebral bodies. The choice of posterior, anterior, or combined anterior and posterior fusion is determined by curve severity, growth remaining, and the likelihood of

fusion. Anterior fusion in addition to posterior fusion may be mandated by a great deal of remaining growth or by severe deformities where excision of discs and associated soft tissue structures such as the costovertebral joints is required to enhance correction of the spine at the time of instrumentation. Absence of posterior bony elements associated with myelodysplasia or after laminectomy mandates anterior fusion and often anterior instrumentation. If the underlying condition makes it less likely that fusion will be achieved (conditions such as neurofibromatosis, myelomeningocele, or post-radiation deformity), then combined anterior and posterior exposure increases the surface area of the fusion mass and likelihood of fusion. The quality of the underlying bone, the closeness and surface area of adjacent vertebrae, rigidity of fixation, and source of bone graft all contribute to the likelihood of fusion. Autogenous iliac crest cancellous or corticocancellous strips are the most osteogenic bone graft available, but freeze-dried allograft bone is utilized when iliac autogenous bone is not practical. Recombinant bone morphogenic protein and other osteo-inductive substances hold promise for the future as bone graft substitutes.

Correction, immobilization, and instrumentation of spinal deformity

Spinal fusion can be performed without deformity correction (*in situ* fusion) but usually correction and immobilization are desired. Pre-or postoperative correction by casting or traction has some special applications, but generally instrumentation is used at the time of spinal fusion for both correction and immobilization. Harrington rod instrumentation, which initiated the modern era of spinal instrumentation, has been supplanted by 'segmental' instrumentation and multihook contourable rods. The Harrington rod had two points of fixation, at either end of the spinal curve, while segmental instrumentation has multiple points of fixation either through multiple wires, hooks, or screws and often involves more than one rod. Segmental instrumentation produces greater correction and more rigid immobilization and usually eliminates the need for a postoperative brace or cast. Multiple hook types and wires allow fixation posteriorly to the lamina or pedicle or vertebral body via the pedicle, while screws permit fixation to the anterior vertebral body when used with anterior instrumentation or via the pedicle to the vertebral body when used posteriorly.

Sublaminar wires passed at every level are frequently used in paralytic conditions with soft bone. In most paralytic conditions when fusion to the sacrum is desired the instrumentation is placed in the ilium, rather than sacrum. A variety of fixation techniques to the ilium such as screws or rods placed between the inner and outer tables of the ilium ('Galveston' fixation) allow indirect fixation of the sacrum.

The Harrington rod or other non-segmental instrumentation drew the deformed spine toward a straight rod. Modern segmental fixation is contourable and a combination of distraction, compression, and translatory forces along with rod contouring make it possible to achieve more normal thoracic kyphosis and lumbar lordosis.

Instrumentation without fusion is occasionally performed in the very young. By periodically lengthening the instrumentation, correction of the deformity can be maintained while allowing for some spine growth and awaiting definitive fusion at a later age.

Thoracoplasty with resection of the protuberant posterior ribs is performed for cosmetic improvement of severe deformities. By resecting a section or the proximal end of the rib and allowing the thoracic cage to shift to a more neutral position, an improved cosmetic appearance is achieved.

Spinal release, spinal fusion, and anterior instrumentation can be accomplished via video-assisted thoracoscopic surgery, diminishing the morbidity normally associated with thoracotomy.

Complications of spinal fusion

Spinal fusion eliminates motion and growth over the fused section of the spine. If a long fusion is performed in a very young child the loss of spinal height may be profound. However in those conditions needing early spinal fusion such as severe congenital scoliosis or kyphosis, loss of spine height from fusion is minimal compared with worsening of the deformity through growth. In most circumstances fusing the spine in a shorter, straighter position is preferable to allowing the evolution of a severe, uncorrectable deformity. An approximation of the growth remaining in a normal thoracic or lumbar spine is given by the formula 0.05 cm.(thoracic) or 0.07 cm. (lumbar)/segment per year of growth remaining. Fusing 8 thoracic vertebrae in a 10-year-old girl might result in $0.05 \text{ cm} \times 8 \text{ segments} \times 6 \text{ years} = 2.4 \text{ cm}$ lost, less than one might suspect. Inhibition of growth in diameter of the spinal canal is rarely a problem as the neurocentral synchondroses have finished growth early in childhood.

Fusion concentrates stress over the remaining mobile segments of spine. If the thoracic and most of the lumbar spine are fused leaving few motion segments in the lower lumbar spine, there can be degenerative changes, stress fractures, or spondylolysis below the fusion. Thus unwarranted extent of the fusion is avoided.

If a posterior fusion is performed in a young child with substantial growth remaining, persisting growth in the anterior vertebral bodies may cause the anterior spinal column to lengthen. With the length of the posterior column fixed and tethered by the fusion, the fusion mass may bend into a progressive lordosis or increasing scoliosis and rotation, a phenomenon referred to as the 'crankshaft phenomenon'. To avoid this side-effect of persisting anterior spinal growth, both anterior and posterior fusions are performed when a great deal of growth remains.

Pseudoarthroses or failures to fuse are a common complication of spinal fusion. Pseudoarthroses occur more commonly in metabolically unfavorable situations, when bone surfaces are inadequate, and when immobilization is incomplete. Repair of the pseudoarthrosis with better bone graft and better fixation often results in a fusion.

In any spinal procedure, a neurologic injury may occur. The origins of neurologic injury associated with spinal instrumentation and fusion are varied. Direct intrusion into the spinal canal by instrumentation or severe translation of one vertebra relative to another can result in compromise of the canal. A change in position of an abnormal or scarred spinal cord such as in myelodysplasia or tethered spinal cord may be associated with neurologic loss. Elongation of the spinal canal may not be tolerated and traction on the

vessels supplying the spinal cord or ligation of the segmental vessels associated with anterior exposure may account for some cases of neurologic loss. Neurologic loss can be immediate or delayed, reversible or irreversible, complete or incomplete. Typical precautions taken at the time of spinal instrumentation and fusion include neurophysiologic monitoring of somatosensory evoked potentials or in some cases motor evoked potentials. In co-operative patients the so-called Stagnara 'wake up' test allows assessment of function under anesthesia, after instrumentation but before the procedure is completed. Postoperative neurologic loss is rare in idiopathic spinal deformities and common in some congenital deformities and bone dysplasias with spinal stenosis.

Idiopathic spinal deformity

Idiopathic scoliosis

Idiopathic scoliosis is the most commonly encountered spinal deformity requiring treatment.

Infantile and juvenile idiopathic scoliosis

Infantile idiopathic scoliosis, occurring under the age of 3 years, is rare and may spontaneously resolve. Juvenile idiopathic scoliosis, occurring in the age range 3 to 10 years, may be associated with an underlying neuropathic abnormality, particularly Chiari I malformation and associated syringomyelia. Juvenile idiopathic scoliosis severe enough to treat warrants a screening MRI of the intraspinal contents to screen for these abnormalities. Juvenile idiopathic scoliosis may respond well to bracing or may relentlessly progress to a more severe deformity needing fusion.

Adolescent idiopathic scoliosis

Etiology, incidence, and natural history

Although 'idiopathic' in origin there appear to be multiple possible etiologies for adolescent idiopathic scoliosis. The genetics of adolescent idiopathic scoliosis are poorly defined but presumed to be variable in inheritance and expression. A neuropathic etiology is suggested by studies showing differences in vibratory sense, postural equilibrium, and brain stem asymmetries. Adolescent idiopathic scoliosis typically worsens during the rapid early pubertal growth spurt and disordered growth has been cited as an etiology. Most thoracic idiopathic scoliosis is associated with thoracic hypokyphosis or lordosis. The persistence of hypokyphosis late in childhood, particularly in girls, may be associated with the development of progressive adolescent idiopathic scoliosis. An underlying connective tissue or collagen disorder has been suggested by studies demonstrating platelet dysfunction in adolescent scoliosis. It seems unlikely that any one etiology will prevail as the sole cause of 'idiopathic' scoliosis, rather adolescent idiopathic scoliosis with its typical curve patterns and worsening during the early pubertal growth phase may be a common expression of several subtle disorders.

The incidence of adolescent idiopathic scoliosis depends upon definition. Many normal individuals have a lateral curvature of 10° or less and a high percentage of school children screened for spinal asymmetries may demonstrate a mild scoliosis. Approximately 1 in 500

to 1 in 2000 adolescents may need brace or surgical treatment for idiopathic scoliosis. Adolescent idiopathic scoliosis is more common in girls, appears as a smaller curve in late childhood, and generally worsens with growth. Progression (worsening) of the curvature parallels growth rate but is variable in the individual. Milder curves under 20° often progress no further and even curves as large as 30° with substantial growth remaining may spontaneously stabilize. Most curves over 30° continue to progress with growth. Small curves at maturity usually remain stable through adulthood, while larger curves in excess of approximately 50° frequently continue to worsen slowly during adulthood. Adolescent idiopathic scoliosis is rarely SYmptomatic during adolescence but large progressive curves during adulthood can cause disfigurement and persistent pain, not easily treated by surgical or non-surgical means.

Observation and treatment

The screening, observation, and treatment of adolescent idiopathic scoliosis are directed toward preventing the adult sequelae of idiopathic scoliosis. School screening of 5th to 9th grade children is suggested to search for potentially progressive scoliosis in a phase when curves are still treatable by non-operative means. School screening has become controversial, has been abandoned in some countries, and has been the subject of a United States Public Health Service task force report on preventative services. Unnecessary referral of small curves for radiography can be reduced by use of the 'inclinometer'. Inclinometer readings of less than 5° angle of trunk rotation on the forward bend test are usually less than 20° Cobb angle of curvature on radiography and do not need radiographic evaluation. If physical findings suggest non-idiopathic scoliosis or there is associated back pain, even smaller curves should be examined radiographically. The natural history of adolescent idiopathic scoliosis during growth is unpredictable in the individual with smaller curves. Follow-up observation of curves in growing children depends upon the magnitude of the curve and growth remaining. Frequent radiographic follow-up is appropriate for larger curves with growth remaining.

Based on natural history studies, brace treatment of idiopathic scoliosis during growth is indicated for juvenile curves of more than 20° , for progressive adolescent curves of more than 25° , and for curves between 30 and 45° with growth remaining. Spinal bracing is of limited effectiveness in curves of greater than 45° , curves high in the thoracic spine, or those associated with hypokyphosis.

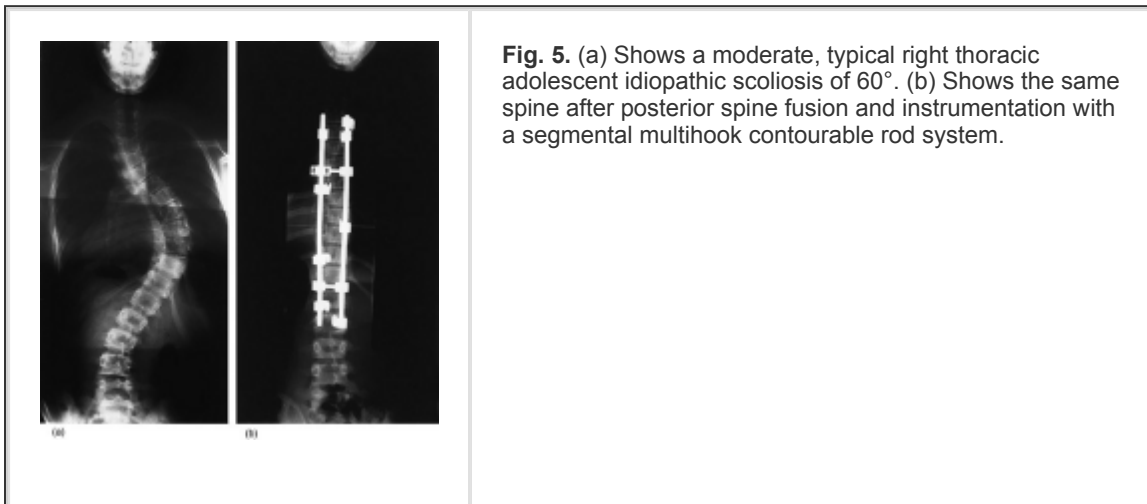
The goal of non-operative treatment of adolescent idiopathic scoliosis is cessation of curve progression or improvement such that a fusion is not required. A wide variety of braces are available with different designs and varied degrees of documentation. The Milwaukee brace, which includes a metal superstructure, has the longest track record, followed by the Boston brace. Nighttime only 'unbending' braces such as the Charleston brace have gained popularity. A recent meta-analysis suggests that the effectiveness of brace treatment of adolescent idiopathic scoliosis is dose related and that part-time braces may not be as effective as full-time braces. A typical 'full-time' brace program involves wearing a brace for 20 out of 24 h with time out of the brace allowed for activities such as sport, bathing, and exercises. Bracing is continued until skeletal maturity. While wearing the brace there is substantial curve reduction and around 50 per cent reduction of a typical scoliosis curve

should be sought in the initial brace application. In-brace curve correction is generally lost over time. After discontinuing the brace at maturity the average result of bracing is cessation of

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curve progression, not correction. About 30 per cent of patients experience substantial lasting correction, about 50 per cent are the same as at the initiation of bracing, and fully 10 to 20 per cent continue to experience curve progression in spite of bracing and may require spinal fusion. Non-compliance with requested brace wear is a major problem. The efficacy of bracing has been questioned. A prospective study undertaken by the Scoliosis Research Society demonstrates a large, statistically significant difference between brace treatment and observation alone. Electrical stimulation for scoliosis was shown to be statistically ineffective.

Operative treatment for adolescent idiopathic scoliosis is indicated for curves predicted to be troublesome as an adult or severe curves in the growing child uncontrolled by bracing. A curve magnitude of more than 50° is often cited as an indication for fusion, but this indication is balanced with curve appearance, progression after maturity, or pain as additional indications for fusion. Posterior fusion with multihook contourable rod instrumentation is standard (Fig. 5(a and b)), but anterior instrumentation and fusion may be preferable for single thoracolumbar or lumbar curves. Usually two-thirds correction of the curve and 50 per cent correction of the associated rib hump or asymmetry can be achieved. Typical risks include a 0.02 rate of pseudoarthrosis and 0.001 risk of paralysis. Autologous predonation of blood and intraoperative spinal cord monitoring are usually employed. The extent of fusion varies according to severity, type, and location of deformity.



Idiopathic kyphosis (Scheurmann's kyphosis)

Kyphosis is normal in the thoracic spine, with 20 to 45 or even 60° cited as normal thoracic kyphosis. Normal kyphosis is evenly distributed in the thoracic spine and centered in the mid-thoracic spine. An apex of kyphosis in the low thoracic spine or at the thoracolumbar junction or a magnitude of kyphosis in excess of 45 to 60° is considered abnormal.

Idiopathic adolescent thoracic hyperkyphosis (Scheurmann's kyphosis) occurs more in

males and shows typical vertebral endplate irregularity and wedging. Scheuermann's kyphosis is less flexible than normal thoracic kyphosis. On forward bending an accentuated hump is noted and on extension of the thoracic spine the kyphosis does not reverse as it would in a normal adolescent. The etiology of Scheuermann's kyphosis is unclear but is typically associated with tight hamstrings and may be associated with back pain.

The natural history of untreated Scheuermann's kyphosis in adulthood is not well documented. Generally, abnormal kyphoses at the thoracolumbar junction are felt to be associated with progressive deformity and pain during adulthood. Evenly distributed mid-thoracic hyperkyphosis may not be a major source of pain during adulthood but may be associated with pain in the lumbosacral area or cervical spine because of compensatory cervical or lumbar hyperlordosis. The incidence of L5 spondylolysis is higher in individuals with Scheuermann's thoracic hyperkyphosis than the general population.

Non-operative treatment for mild Scheuermann's kyphosis includes exercises with variable success. Braces, particularly Milwaukee braces, can be successful for moderate kyphosis between 50 and 70°, but the long-term effectiveness of kyphosis braces has been questioned.

Operative intervention for Scheuermann's kyphosis is generally anterior and posterior fusion although posterior fusion alone may suffice. The indications for prophylactic fusion are unclear but include severe pain, an unacceptable cosmetic deformity, or a severe deformity at the thoracolumbar junction or upper lumbar spine.

Congenital spinal deformity

Congenital spinal deformities originate from an early embryologic malformation and can be subdivided into failures of segmentation, failures of formation, or combinations. Other musculoskeletal, visceral, and neurologic anomalies frequently accompany vertebral anomalies. Patients with congenital spine deformities should have a screening renal ultrasound to rule out an asymptomatic renal collecting anomaly with obstruction. An MRI or other neural imaging study is needed prior to operation on congenital kyphosis or scoliosis or if there are signs of spinal cord tethering or neurologic abnormality. Intraspinal anomalies such as low lying conus, fatty filum terminale, tethered spinal cord, diastematomyelia, diplomyelia, syringomyelia, dermoid, and lipoma are not rare. VACTERL (Vertebral, Anal, Cardiac, Tracheo-Esophageal, Renal, Limb) association includes congenital vertebral anomalies. Sprengel's deformity with a high-riding scapula is commonly accompanied by upper thoracic or cervicothoracic vertebral anomalies.

Congenital scoliosis and kyphosis

Congenital scoliosis and kyphosis can be classified as complete or incomplete failures of segmentation or formation and many combinations thereof. The natural history of congenital scoliosis is variable. If growth potential is unbalanced with more growth potential

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on one side of the spine than the other, then the curve will progress with growth. If a tethering structure such as a rib fusion or congenital bony intervertebral bar is present,

then progression may be rapid. Balanced growth potentials such as found in block vertebrae, balanced hemivertebrae, or fully incarcerated, non-segmented hemivertebrae may show no worsening with growth. Risk of progression is highest during the rapid growth periods of infancy and the first 2 years and again during the preadolescent growth spurt.

Progressive congenital scoliosis should be fused, as brace treatment is generally ineffective. For simple congenital scoliosis, *in situ* fusion without instrumentation is appropriate. For advanced deformity, hemivertebra excision or osteotomy of existing bony bars or areas of congenital fusion coupled with instrumented correction may be more appropriate. Hemivertebra excision is particularly effective at the lumbosacral junction or thoracolumbar junction in young children. Any correction of congenital spinal deformity carries an elevated risk of neurologic injury. Early *in situ* fusion of any progressive curve is generally the preferred treatment.

Congenital kyphosis nearly always worsens with growth. Type I congenital kyphosis with failure of formation may be associated with the early onset of paraplegia when progressive. Type II congenital kyphosis associated with failure of formation is less commonly associated with paraplegia but also worsens with growth. Congenital kyphosis less than 40° in children less than 5 years of age can generally be treated with a low-risk posterior *in situ* fusion and cast immobilization. Continued anterior spine growth after posterior fusion may provide further correction. More severe deformities or those with spinal canal compromise need anterior decompression, fusion, and posterior fusion and instrumentation. Surgery for congenital kyphotic deformities entails a high risk of neurologic injury. Traction for congenital kyphotic deformities is contraindicated, as longitudinal traction draws the already compromised spinal cord more tightly across the apex of the kyphosis. As for congenital scoliosis there is no effective non-operative treatment for congenital kyphosis and early *in situ* fusion is preferable where possible.

Other congenital spine deformities

Klippel–Feil deformity consists of multiple congenital fusions of the cervical and upper thoracic vertebrae. Typically there is an associated short neck and commonly associated Sprengel's deformity of the scapula and webbing of the neck. Because of the restricted cervical motion the remaining mobile cervical segments are subjected to extra stress and may become unstable. Protection from violent activities is often necessary to avoid neurologic injury in children and adults with Klippel–Feil deformity. Follow-up for the development of potentially dangerous cervical instability should continue through adulthood.

Partial or complete sacral or lumbosacral agenesis consists of a complete absence of the bony elements of the sacrum and lumbar vertebrae. Typically the pelvis is diminutive and as a result diverting colostomy or urinary drainage may be necessary. Lower extremity sensation is often intact but motor level paralysis reflects the level of absent vertebrae. Other congenital spinal anomalies include segmental spinal dysgenesis and Jarcho–Levin SYndrome with multiple vertebral anomalies and abnormal ribs. Many other congenital spinal abnormalities may occur singly or in connection with other anomalies

Neuromuscular spinal deformity

Spinal deformity commonly develops in association with neuromuscular conditions occurring during growth. Virtually every neuromuscular condition can be associated with spinal deformity. Abnormal muscle tone, strength, or balance combined with growth can produce profound deformities of the vertebral column. In idiopathic and congenital spinal deformity, treatment is generally directed toward preventing the long-term, adult consequences of deformity. Treatment of neuromuscular deformity emphasizes functional results. Bracing may be directed toward functional gain rather than cessation of curve progression. Spinal fusion will prevent further deformity but also may enhance use of the upper extremities or simplify seating management.

Cerebral palsy

Spinal deformity is common in patients with severe cerebral palsy who are non-ambulatory, but uncommon in the milder, ambulatory patient. Spinal deformity may profoundly affect the non-ambulatory patient, complicating seating and positioning, causing pain or skin break down, or in its most severe forms causing respiratory distress. There is a common and direct relationship between hip deformity and spinal deformity. Asymmetric hip contractures contribute to development of the spinal deformity and vice versa. Commonly a 'windswept' deformity of the hips with unilateral or bilateral subluxation and an oblique pelvis is associated with the scoliosis. Non-operative treatment of milder deformities consists of positioning and stretching. In moderate deformities an external orthosis, such as a semirigid orthosis, will facilitate seating and function but is probably ineffective in controlling progressive curvature. The natural history of spinal deformity in cerebral palsy is variable. Skeletal maturity is often achieved later and curve progression may continue relentlessly through adulthood. When deformity becomes severe enough to be functionally troublesome in spite of bracing, spinal fusion is indicated. Anterior and posterior fusion with posterior segmental sublaminar wire fixation and dual rods (Fig. 6(a and b)), such as a Luque'-Galveston instrumentation, will withstand the abnormal muscle tone and permit early postoperative mobilization. Spinal fusion in cerebral palsy requires close attention to nutrition, as complication rates are directly correlated with preoperative nutritional status. Anterior and posterior surgery can generally be done on the same day, diminishing time in hospital and improving nutritional status.

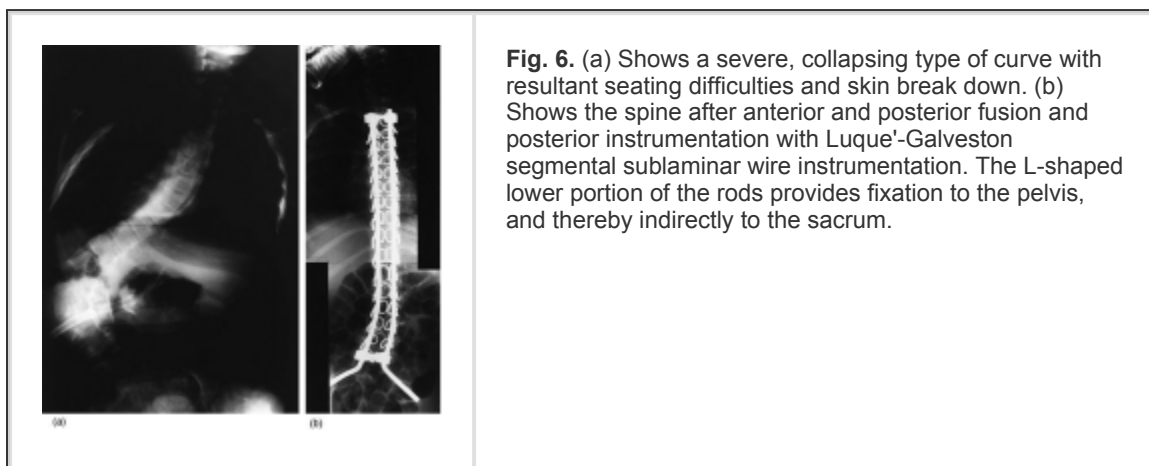


Fig. 6. (a) Shows a severe, collapsing type of curve with resultant seating difficulties and skin break down. (b) Shows the spine after anterior and posterior fusion and posterior instrumentation with Luque'-Galveston segmental sublaminar wire instrumentation. The L-shaped lower portion of the rods provides fixation to the pelvis, and thereby indirectly to the sacrum.

Myelodysplasia

Spinal deformity associated with myelodysplasia and other neural tube defects presents a wide spectrum of deformity. Some patients with asymmetric levels of innervation, a tethered spinal cord, or asymmetric hip contractures develop a long, collapsing deformity while others with associated congenital vertebral anomalies may develop a rigid, localized deformity. Non-surgical treatment of myelodysplasia-related spinal deformity is directed toward facilitating function. Adaptive seating, wheelchair modifications, external spinal orthoses, and exercises are designed to improve independence, mobility, and upper extremity function. When deformity becomes large and functionally troublesome, fusion is usually indicated. Because the skin overlying most myelodysplasia spines is abnormal,

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preoperative tissue expanders or planning for local skin flaps is required. Because the accompanying neural elements are abnormal, simultaneous or concomitant spinal cord detethering may be required and because the posterior bony elements of the spine are usually deficient, combined anterior and posterior fusion is usually indicated. Instrumentation is variable depending on what bony elements are present. The functional effect of spinal fusion can be profound, helping the patient who regains the use of an upper extremity previously needed to maintain seating support, or adversely affecting ambulatory patients who lose trunk motion needed for ambulation. Risks of spinal fusion in myelodysplasia include a high incidence of infection, pseudarthrosis, and neurologic injury.

Muscular dystrophy

Nearly all patients with Duchenne's muscular dystrophy experience troubling spinal deformity during their progressive decline in function. As the patient with Duchenne's dystrophy ages and loses ambulatory ability, a spinal deformity usually appears and slowly progresses. Spinal deformity has been particularly troublesome for keeping patients seated comfortably during the non-ambulatory, progressively dependent phase of Duchenne's muscular dystrophy. Attempts to deal with the deformity through adaptive seating or bracing have been less successful than early spine fusion and instrumentation. By the time the spinal deformity is troublesome, respiratory and cardiac function have usually declined too much to tolerate the spinal fusion operation. Current practice in most clinics for Duchenne's muscular dystrophy is to offer spinal fusion at the time the patient either first ceases to walk or when pulmonary function declines to 50 per cent of predicted.

Spondylolysis and spondylolisthesis

Spondylolysis refers to a disruption in the continuity of the vertebral arch at the pars interarticularis (the portion of the vertebral arch joining the superior and inferior articular processes). Spondylolisthesis refers to the forward displacement of one vertebra upon another. Both occur in the lower lumbar spine with the greatest prevalence at L5 and both are developmental in origin.

Spondylolysis

Spondylolysis of L5 (or much less commonly L4) occurs in up to 5 per cent of the normal adult population, and is more common among relatives, certain ethnic groups, and in association with spina bifida occulta of L5 or S1. Spondylolysis is not present at birth, but first develops in the school age child. It is presumed that spondylolysis occurs by a mechanism of pars interarticularis stress, stress fracture, and finally complete fracture with spondylolysis. Fortunately most spondylolysis in adults is asymptomatic, the incidence of low back pain associated with spondylolysis not being different than the general population. Spondylolysis is a common source of pain in the older child and adolescent. Typical physical findings include lumbosacral pain with or without radiation to the buttocks, worse with sitting, back bending, and athletic activities. The incidence of spondylolysis seems higher in athletes involved in activities involving spinal hyperextension such as gymnastics. Plain radiographs may be negative, but spondylolysis may be seen on oblique radiographs and can be definitively diagnosed with a CT scan. If the spondylolysis is still in the 'stress reaction' or stress fracture stages, before established lysis

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has occurred or if the lysis has occurred recently, bone scan will reveal increased uptake at the pars interarticularis.

Treatment for symptomatic spondylolysis or stress fracture of the pars interarticularis in the adolescent includes activity modification and temporary brace application. Many stress fractures with impending spondylolysis can be healed in this manner. Spondylolysis in the child and young adolescent may evolve into spondylolisthesis and should be followed periodically with repeat lateral radiographs to rule out progressive slippage. Surgery is rarely indicated in the child or adolescent with spondylolysis, as most can be rendered asymptomatic or minimally symptomatic with non-operative treatment. If required, surgery consists of an *in situ* fusion of L5 to S1.

Spondylolisthesis

Spondylolisthesis is less common than spondylolysis, but also is not present at birth and gradually develops during childhood and adolescence. Spondylolisthesis is classified according to etiology (see adult spinal deformity chapter), but in the child and adolescent two types are encountered. The 'isthmic' type involves elongation or disruption of the pars interarticularis allowing the L5 vertebral body to displace progressively forward while the posterior elements of L5 remain in place. The 'dysplastic' type involves an insufficiency of the posterior elements and facets between L5 and the sacrum, allowing the L5 vertebral body and its posterior elements to displace forward on the sacrum. Radiographic measurements include slip angle (a measure of kyphosis at the lumbosacral junction) and forward displacement of L5 forward on S1 (grade I to V, or 0 to 100 per cent). Physical findings are highly variable. In mild slips there may be no positive physical findings. More severe symptomatic slips are associated with a palpable step-off between the spinous processes of L4 and L5, a shortened waist, visible lumbosacral kyphosis with flattened buttocks, severely tight hamstrings, and a 'jump' posture consisting of hip and knee flexion. Symptoms include tight hamstrings and a variable degree of back and leg pain. The natural history of spondylolisthesis in the child and adolescent is variable. Progression of the

slippage may be relentless during childhood and adolescence or may stabilize. Treatment for milder degrees of slip (less than 50 per cent displacement) is observation and non-operative treatment including bracing if SYmptomatic. Brace treatment is not felt to prevent further displacement. Treatment of more severe degrees of slippage depends upon growth remaining and symptoms. Asymptomatic severe slips in adults may be observed. Progressive slips in excess of 50 per cent and particularly those that are symptomatic in the adolescent are treated with L4 to the sacrum fusion with or without reduction, nerve root decompression, or instrumentation.

Further reading

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