CLINICALKEY

Scoliosis

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Key points

- Scoliosis is a three-dimensional spinal deformity affecting the axial, coronal, and sagittal planes
- Signs, symptoms, and physical examination findings are varied depending on the age group and medical history of the patient
- In idiopathic scoliosis, the period of greatest concern is the adolescent growth spurt, when scoliotic curves rapidly progress
- The appropriate diagnostic workup of some scoliotic patients involves multiple organ systems and varied imaging modalities
- Non-operative and operative interventions may be instituted based upon the degree of the spinal deformity, etiology of the scoliosis, the curve's rigidity, and the progression of the deformity
- The prognosis is variable, depending upon the patient's age and the etiology of the condition

Background

Description

- Scoliosis is a three-dimensional deformation of the spine that affects the axial, coronal, and sagittal planes
- There are multiple etiologies, including idiopathic, degenerative, and neuromuscular
- Disease presentation and progression differ in the adult and pediatric populations
- Other organ systems, especially pulmonary, may be affected
- A thorough evaluation of the patient by a multidisciplinary team is necessary in the decision-making process to correct the deformity

Normal anatomy

- Normal anatomy of the spine is defined by a coronal curvature of 0º to 10º, a thoracic kyphosis (T2 to T12) of approximately 20° to 40°, and a lumbar lordosis (T12 to S1) of approximately 30° to 60°

Definitions

- Idiopathic scoliosis (IS): spinal deformity without known etiology. May be present in the skeletally immature or mature spine

- Idiopathic scoliosis is a coronal plane spinal deformity with a Cobb angle of greater than 10°, with or without sagittal imbalance (kyphosis or hyperlordosis), in children up to 18 years of age at time of diagnosis. The Cobb angle is determined by drawing tangential lines through the superior endplate of the most rostrally involved vertebra and the inferior endplate of the most caudally involved vertebra. The angle of the intersection of perpendicular lines to the tangential lines mentioned above is the Cobb angle

- Idiopathic scoliosis is differentiated from other forms of childhood scoliosis based on etiology, with congenital scoliosis arising from vertebral abnormalities (*eg*,hemivertebra, malformed vertebra)

  - Infantile idiopathic scoliosis (IIS): children from birth to 3 years

    - Boys have IIS more frequently than girls

  - Juvenile idiopathic scoliosis (JIS): children aged 3 to 10 years

    - JIS is associated with a right thoracic curvature and occurs more frequently in girls

    - Progress before 10 years of age has been reported at 1° to 3° per year, with rapid increase to 4.5° to 11° per year after that age. The majority of patients will require surgical fusion despite early diagnosis and bracing

  - Adolescent idiopathic scoliosis (AIS): children and adolescents aged 10 to 18 years

    - Typically presents with a right-sided thoracic curve or a left-sided thoracolumbar curve

    - In smaller-curve AIS, there is an equal male-to-female distribution; however, larger curves are associated with a female predominance

    - Natural history studies have shown that the majority of curves diagnosed from ages 10 to 18 will not significantly progress with time, and more than 90% will completely stabilize by skeletal maturity

    - Overall progression rate has been quoted at 0.5° to 1.0° per year

    - Severe curves (greater than 110°) may be associated with cardiopulmonary abnormalities and remain difficult to treat secondary to long-lasting deformity with full potential of future skeletal growth remaining

  - AIS that progresses or is uncorrected results in adult idiopathic scoliosis

- Adult degenerative scoliosis (ADS), or de novo scoliosis: degenerative change occurring in adulthood without pre-existing deformity

- ADS pertains to a coronal plane spinal deformity in the skeletally mature, with a Cobb angle greater than 10° with or without sagittal imbalance and axial rotational deformity (a three-dimensional deformity)

- It is postulated that ADS begins with asymmetrical intervertebral disc degeneration that leads to degeneration and instability of the posterior elements, especially in the facet joints. This results in axial rotation, lateral listhesis, and ligamentous laxity. Osteoporosisand micro/macro vertebral compression fractures likely contribute to the degenerative process and formation of ADS

- Neuromuscular scoliosis: a coronal plane spinal deformity, with or without sagittal imbalance (kyphosis or hyperlordosis) in patients with defects in either the muscular or neuronal pathways of the body, such as cerebral palsy and muscular dystrophies

- Neuromuscular scoliosis is known for the early age of onset with rapid progression during skeletal growth, and continued progression after skeletal maturity. Curves are generally long and are associated with pelvic obliquity

- The earlier onset of scoliosis has been associated with more severe spinal curvatures. Additionally, with increasing age comes increased curve stiffness and rigidity, making it more difficult to achieve adequate surgical correction

Lenke classification system

- Multiple systems exist based upon patient age, scoliotic curve, spinal abnormalities, and overall spinal alignment

- The Lenke classification system is the most commonly used

- Takes into account the curvatures of the proximal thoracic spine, mid-thoracic spine, and the thoracolumbar junction. Modifiers are also used to describe the coronal imbalance of the lumbar spine and the degree of sagittal imbalance of the thoracic spine

- Curve type (1-6) + Lumbar coronal modifier (A, B, C) + Thoracic sagittal modifier (-, N, +) = Curve classification (*eg*, 1B+)

- The following information and tables were modified from Lenke LG, Betz RR, Harms J, et al. Adolescent idiopathic scoliosis: a new classification to determine extent of spinal arthrodesis. J Bone Joint Surg Am. 2001;83A:1169-81,Figure 41-30: Curve types and criteria for structural curves and location of apex

- Structural criteria (minor curves):

  - Proximal thoracic

    - Side bending Cobb greater than or equal to 25°

    - T2-T5 Kyphosis greater than or equal to+20°

  - Main thoracic

- Side bending Cobb greater than or equal to 25°
- T10-L2 Kyphosis greater than or equal to+20°

  - Thoracolumbar/lumbar
    - Side bending Cobb greater than or equal to 25°
    - T10-L2 Kyphosis greater than or equal to+20°
- Location of apex (SRS definition):

  - Thoracic curve: T2 to T11/12 disc
  - Thoracolumbar curve: T12/L1
  - Lumbar curve: L1/2 disc to L4

Table 1. Lenke classification system for AIS

Curve type	Proximal thoracic	Main thoracic	Thoracolumbar/lumbar	
1	Nonstructural	Structural[a]	Nonstructural	
2	Structural[b]	Structural[a]	Nonstructural	D
3	Nonstructural	Structural[a]	Structural[b]	
4	Structural[b]	Structural[c]	Structural[c]	
5	Nonstructural	Nonstructural	Structural[a]	Tl
6	Nonstructural	Structural[b]	Structural[a]	Th ma

[a]Major curve: largest Cobb measurement, always structural;[b]Minor curve: remaining structural curves; [c]Type 4: MT or TL/L can be the major curve

Table 2. Lumbar coronal modifier

Lumbar coronal modifier	Center sacral vertical line to lumbar apex
A	Between pedicles
B	Touches apical body(ies)
C	Completely medial

Table 3. Thoracic sagittal profile T5-T12

Modifier	Cobb angle
- (Hypo)	<10°
N (Normal)	10°-40°
+ (Hyper)	>40°

Epidemiology

- Approximately 2% to 3% of children under age 16 have a Cobb angle of more than 10°
- Only 0.3% to 0.5% have a Cobb angle of more than 20°
- With increasing age, the gender ratio in the adolescent population is 3.6:1.0 female-to-male. However, in adolescents with curves less than 10°, the female-to-male ratio is approximately 1:1
- Prevalence of ADS is between 1% and 10%
- ADS is found roughly equally among men and women and typically presents in the sixth and seventh decades, with a mean age of onset of 70.5 years. ADS is rarely found before the age of 40

Causes and risk factors

- No single etiology for the development of scoliosis has been identified. It is a multifactorial process. However, current research has identified associations that seem to be linked

- Osteopenia and osteoporosis , both in the adolescent and adult populations, have been associated with scoliosis. Lower bone mineral densities have been identified in adolescents with more severe curves
    - Adolescent IS may result from abnormally low bone mineralization during a time of rapid bone growth
- Platelet calmodulin levels have been shown to be lower in patients with progressively worsening curves when compared to those with stable curves. It has been hypothesized that because platelets and muscles share similar contractile proteins, an underlying myopathy may contribute to the development of IS
- Melatonin deficiency has been associated with the development of scoliosis in animal models, but this has not yet been demonstrated in nonhuman primates or humans
- *CHD7*was the first gene identified to be associated with IS. It codes for chromodomain helicase DNA binding protein 7, which is involved in chromatin organization

Associated disorders

- Chiari I malformation with or without syringomyelia
- Ehler-Danlos syndrome
- Charcot-Marie-Tooth disease
- Prader-Willi syndrome
- Cerebral palsy
- Spinal muscular atrophy
- Muscular dystrophy
- Familial dysautonomia
- Friedreich ataxia
- Proteus syndrome
- Marfan syndrome
- Neurofibromatosis
- Connective tissue disorders
- Spinal dysraphism
- Scheuermann disease
- Spina bifida -myelomeningocele

Screening

Summary approach

- There is currently some controversy over the issue of routine scoliosis screening of children in both schools and physicians' offices
- Previous studies have both supported and refuted the need for screening, and there have been no recent studies to definitively address this issue
- Routine screening of asymptomatic adolescents for AIS, while still performed annually by many school systems, is not recommended by the U.S. Preventive Services Task Force as of 2004
- The task force found no good evidence that routine screening of this age group detects scoliosis at an earlier stage than it would have been detected without screening
- In a 2008 position statement developed as an educational tool based on the opinion of the authors, the American Academy of Orthopaedic Surgeons, the Scoliosis Research Society (SRS), the Pediatric Orthopaedic Society of North America, and the American Academy of Pediatrics did not support any recommendation against scoliosis screening. The position statement was not based on a systematic review
- Physicians should understand scoliosis screening methods and be prepared to use them if:
    - There are suggestive findings on physical examination
    - It is requested by the patient or a pediatric patient's parent as part of the physical examination
    - There is a family history of scoliosis

Population at risk

- If routine screening is undertaken, it should include the pre-adolescent and adolescent age groups
- Female adolescence, with its associated growth spurt, occurs about two years earlier than in males, so screening should also be done at earlier ages
- The American Academy of Orthopaedic Surgeons, SRS, Pediatric Orthopaedic Society of North America, and American Academy of Pediatrics recommend that screening of girls be done at age 10 (grade 5) and repeated at age 12 (grade 7)
- The recommended ages for screening boys are age 13 (grade 8) and 14 (grade 9)

Screening modalities

Physical examination

Description:

- The overall relationship of the cranium to the pelvis should be noted
- View the patient laterally, observing curvature of the cervical, thoracic, and lumbar regions. The postures of the pelvis and knees should be noted also, as these contribute to maintenance of sagittal balance
- The Adams forward bend test is a simple maneuver that can help detect coronal imbalance
    - The patient is positioned upright with arms extended and palms together. Then he or she is asked to bend forward at the waist until the thoracic spine is parallel to the floor
    - The examiner then looks for asymmetry in the shoulders, scapulae, rib cage, thoracic spine, and thoracolumbar regions. The examiner should observe the patient anteriorly, posteriorly, and from both the left and right sides. This is best done with the examiner's eyes at the patient's spine level
- A scoliometer can also be used to measure the coronal deformity during the Adams forward bend test. This device is placed on the spinous process at the apex of the rib hump and produces a value called the angle of trunk rotation (ATR), measured in degrees. It is recommended that the examiner have the patient return to an upright posture, then repeat the forward bend, and measure a second time to ensure accuracy
    - Use of the scoliometer in the lower thoracic and lumbar spines is less accurate, as the costovertebral attachments in this region do not produce a rib hump
- Leg-length discrepancies may cause positive results on the Adams forward bend test and scoliometer measurements. However, clinical methods of leg-length measurement are highly imprecise and inaccurate when the criterion for leg-length inequality is as low as 5 mm

Results:

- A "rib hump" seen on the Adams forward bend test is often found with a scoliotic curve greater than 10°
- An ATR value greater than 5° on the scoliometer correlates with a deformity of at least 10°. The positive predictive value of this correlation is 28%
- Using an ATR of 6° to 10° as the cut-point for obtaining x-ray imaging results in higher positive predictive values
- An ATR of 7° is a commonly used cut-point for progressing to further work-up, but the ideal cut-point is not known

Evidence

Scoliosis screening programs have a low positive predictive value for identifying scoliotic curves:

- A meta-analysis of 36 retrospective cohort studies of scoliosis screening programs was done to evaluate the clinical effectiveness of school screening for adolescent

idiopathic scoliosis. The pooled referral rate (using forward bend testing, angle of trunk rotation, or topography studies) for radiography was 5%. The positive predictive values for predicting curves greater than 10°, greater than 20°, and receiving treatment were 28%, 5.6%, and 2.6%, respectively. [1] *Level of evidence: 2*

References

Primary prevention

Not applicable.

Diagnosis

Summary approach

- When physical examination suggests scoliosis, the diagnosis is confirmed by radiography
- Long cassette (36-inch) posteroanterior and lateral plain films shot at a distance of 72 inches of the standing patient with hips and knees fully extended are the standard postures for examining overall spinal alignment
- The Cobb technique can be used to measure the magnitude of the curve
- Computed tomography (CT) , magnetic resonance imaging (MRI) , and/or bone densitometry may be needed diagnostically in complicated cases, as well as for preoperative imaging, but are not routinely necessary
- Other modalities exist that do not depend upon ionizing radiation
  - Rasterstereography casts a square grid of light at a known angle and distance. Digital images are captured that recreate the topography of a patient's back
  - Using skin markers, video-based systems gather data on spinal curvature
- Assessment of skeletal maturity and of other organ involvement may require additional studies, including radiography of the hand and pelvis

Clinical presentation

Children and Adolescents

Symptoms:

- Many patients, especially adolescents, may be asymptomatic and are referred to a physician after routine school screening detects an abnormality
- Some may be concerned only with cosmesis and body image
- Rarely, dyspnea
- Rarely, chest pain

Other historical information:

- Prenatal history of mother
- Birth history of child
- Cognitive and motor milestones
- Medical history
- A full review of systems. Pulmonary, renal, and cardiac anomalies are not uncommon and require additional evaluation
- Effect disease has upon quality of life, such as an inability to perform activities of daily living or participate fully in school activities
- If pain is present, ascertain quality, location, and any changes in pain over time

Signs:

- Adams forward bending test will be visually abnormal
- Flexibility of spine, especially at curve, may be diminished
- There may be a leg-length differential (LLD) with scoliosis, but the etiological significance of this finding is uncertain

Other physical examination factors:

- Examine skin for signs of anomalies which may coexist with scoliotic deformity
  - Axillary freckles
  - Café au lait spots
  - Midline dimpling on back
  - Tufts of hair on back
- Palpation of entire spine to elicit tenderness
- Neurologic examination
  - Muscle tone
  - Motor strength
  - Sensation
  - Reflexes
  - Coordination
  - Gait

Adults

Symptoms:

- Pain
  - Back pain, typically over apex of curve
  - Duration of symptoms is important, as this may help distinguish between idiopathic and de novo scoliosis
  - Radicular pain
  - Presenting symptoms in ADS are related to spinal stenosis and include axial back pain, radiculopathy, and neurogenic claudication due in part to a combination of lateral listhesis and/or subluxation, sagittal imbalance, facet arthropathy, and disc degeneration. More than 90% of patients with ADS will present with pain
- Rarely, dyspnea

Other historical information:

- Medical history
- Prior diagnosis and documentation of scoliosis
- Prior treatments for symptoms related to spine, back, chest
- Surgical history
- Effect disease has upon quality of life, such as inability to perform activities of daily living or go to work
- Note if the patient has had a historical change in posture, particularly loss of height

Signs:

- Adams forward bending test will be visually abnormal
- Flexibility of spine, especially at curve, may be diminished

Other physical examination factors:

- Palpation of entire spine to elicit tenderness
- Neurologic examination
  - Muscle tone
  - Motor strength
  - Sensation

- Reflexes
- Coordination
- Gait

- Surveys can help determine which patients would benefit from treatments, both operative and non-operative
- Oswestry disability index: evaluates back pain and the degree of disability caused by it. Usually more applicable to adults with degenerative scoliosis
- SRS outcomes score (SRS-22): evaluates pain, functionality, and self-image. Better for younger patients with AIS
- Short form-36: useful for all patients. Measures overall wellness

Diagnostic testing

Radiography of the spine

Description

- Plain radiographic films taken from several angles

Normal results

- Cobb angle of less than 10°
- Normal mobility on bending views
- Coronal curvature of 0º to 10º. In the sagittal plane, a plumb line dropped from the center of the C7 vertebral body should fall and intersect the posterior aspect of the S1 superior endplate on standing 36-inch (scoliosis) standard plain films
- The majority of patients maintain this intersection point within 2 cm either anteriorly or posteriorly as measured on the plain films
- Positive sagittal balance: C7 plumb line falls greater than 2 cm anterior to the posterior aspect of the S1 superior endplate
- Negative sagittal balance: C7 plumb line falls greater than 2 cm posterior to the posterior aspect of the S1 superior endplate
- Thoracic kyphosis (T2 to T12) is typically 20° to 40°
- Lumbar lordosis (T12 to S1) is typically 30° to 60°
- Pelvic incidence, or the angle between a line drawn from the center of the femoral head to the midpoint of the sacral endplate and the line perpendicular to it, ranges between 45º and 55º

Comments

- The American College of Radiology practice guidelines indications for radiography of the spine for scoliosis include, but are not limited to, the following:
  - Alterations in normal spinal alignment on physical examination
  - Alterations in normal spinal alignment detected on other imaging studies
  - Evaluation of spinal curvature progression
  - Follow-up of treatment (orthotic or surgical)
  - Evaluation of individuals with a history of scoliosis in immediate family member
- If not already obtained, standing 36-inch posteroanterior and lateral views of the thoracic spine, lumbar spine, and pelvis, to measure the Cobb angle
- The Cobb angle is determined by drawing tangential lines through the superior endplate of the most rostrally involved vertebra and the inferior endplate of the most caudally involved vertebra. The angle of the intersection of perpendicular lines to the tangential lines mentioned above is the Cobb angle
- 36-inch anteroposterior views with the patient bending to left and right to evaluate the mobility of the deformity may be needed in some cases but are not required for the initial workup

Computed tomography of the spine

Description

- Computer-generated cross-sectional images of bone and soft tissue

Normal result

- No abnormal curvatures in coronal, axial, or sagittal planes

Comments

- Rib vertebral angle difference (RVAD) values greater than 20º are associated with curve progression
- Studies have shown 84% of curves progressing with a RVAD of greater than 20°, as opposed to 83% of curves regressing if RVAD is less than 20° in IS2
- Elucidates the bony architecture
- Evaluate for aplastic vertebrae, dysplastic vertebrae, or congenitally fused vertebrae
- RVAD is formed between the superior endplate of the vertebral body and the rib head
- Rarely done in AIS, due to risks from radiation exposure

Magnetic resonance imaging of the spine

Description

- Sectional images of high resolution generated without ionizing radiation

Normal results

- Normal paraspinal neurovascular structures

Comments

- MRI should be done when the patient has atypical physical examination findings, such as pain, neurologic deficit, or atypical curves
- Particularly valuable for evaluating the neural structures
- If there is a contraindication to MRI, CT myelography may be performed

Dual energy x-ray absorptiometry scan

Description

- Dual energy x-ray absorptiometry (DEXA) scan for the measurement of bone density

Normal results

- Results should be within normal range for age

Comments

- Approximately 9% of adult patients with osteoporosis can also have a scoliotic deformity
- Should osteopenia or osteoporosis be identified, medical therapies can be instituted

Radiography of the hand and pelvis

Description

- Plain films of the hand to assess for skeletal maturity
- Plain films of the pelvis to examine Risser stage of iliac epiphysis ossification and to examine the state of the triradiate cartilage

Normal results

- A single anteroposterior view of the left hand and wrist can be compared to standards in the Greulich and Pyle hand atlas to determine bone age Risser stages:
  - Grade 1 corresponds to 0% to 25% skeletal maturity
  - Grade 2: 25% to 50%
  - Grade 3: 50% to 75%

- Grade 4: 75% to 99%
- Grade 5 indicates complete skeletal maturity

Comments

- Assessment of skeletal maturity may be necessary in AIS. Immaturity is associated with greater likelihood of curve progression
- Comparison to prior imaging is useful to evaluate the stability of the deformity
- Younger pediatric patients may require additional imaging of other organ systems to rule out undiagnosed anomalies that may coexist with deformity

Differential diagnosis

Not applicable.

Consultation

Diagnosis of scoliosis can typically be made by a primary care physician, based on physical examination and x-ray findings.

Treatment

Summary approach

- Treatment strategy depends on the degree and nature of the deformity and the age of the patient
- Therapeutic interventions may include bracing and a variety of surgical techniques
- Physical and aqua therapy may aid patients in accommodating to uncorrectable curves
- Pain management is often a necessary component of care and may involve use of oral analgesics (*eg*, acetaminophen , nonsteroidal anti-inflammatory drugs [NSAIDs] , opioids) or injection of anesthetic or steroids at vertebral facets or epidural space
- Surgical techniques include posterior instrumented fusions, with or without osteotomies ; combination anterior/posterior approaches ; anterior release, with or without instrumentation ; and decompression alone

Infantile idiopathic scoliosis:

- Most curves will resolve without treatment

Juvenile idiopathic scoliosis:

- Curves that reach 30° before age 10 will likely worsen and need treatment
- Bracing is sometimes used; others may require surgical treatment

Adolescent idiopathic scoliosis:

- The most important question when dealing with AIS relates to the potential for future curve progression and the adverse effects that will result in adolescence and in adulthood if the curvature goes untreated
- Risk for curve progression is related to skeletal growth potential and curve factors, such as curve magnitude, curve pattern (double greater than single curves, thoracic and thoracolumbar curves greater than lumbar curves), and apical vertebral location
- Small curves less than 25° in AIS are typically observed with serial radiographs; most will not progress to the point of needing treatment
- In cases that require intervention, treatment modalities include bracing and surgery
- Alternative measures, including physical therapy and inpatient scoliosis rehabilitation, have been used mainly in Europe as an adjunct to observation and are not typically used in the U.S.

Adult degenerative scoliosis:

- Curves found incidentally without pain or neurologic deficit are observed with serial radiographs to assess for progression
- The initial treatment of ADS in the setting of pain without neurologic deficit and curve magnitude less than 30° is non-operative conservative management
- Bracing has no role in the treatment of ADS
- Surgical indications are based on presence of neurologic deficit, failed prior conservative therapy, curve magnitude (Cobb angle), and probability of progression

Neuromuscular scoliosis:

- The aims of treatment in neuromuscular scoliosis are halting curve progression and correction of the spinal deformity, thereby achieving a balanced spine in both the coronal and sagittal planes over a leveled pelvis
- By correcting the deformity, sitting balance is re-established and maintained. This maximizes pulmonary function, sitting ability, functional independence, and ease in handling and transfers
- Due to the heterogeneity of diseases, there are no strict guidelines for the treatment of neuromuscular scoliosis. The majority of experts believe that spinal instrumentation and fusion should be undertaken for curves that progress beyond 50° or those that lead to deterioration in functional sitting

Medications

Acetaminophen

Indication

- Acetaminophen is indicated for mild pain

Dose information

Adults and children >12 years:

- 325 to 1,000 mg orally every 4 to 6 hours, when required
- Maximum: 4 g/d

Pediatric patients aged 6 to 12 years:

- 75 mg/kg/d or 4 g/d orally, whichever is less

Pediatric patients aged 2 to 6 years:

- 75 mg/kg/d or 4 g/d, whichever is less, of a suitable children's formulation, orally or per rectum

Pediatric patients aged <2 years:

- 75 mg/kg/d or 4 g/d, whichever is less, of a suitable children's formulation, orally or per rectum. Safety and efficacy of the intravenous formulation has not been established

Major contraindications

- Acetaminophen hypersensitivity

Comments

- Some over-the-counter formulations have lower maximum doses. See individual products. Extended-release oral product should not be used for pediatric patients

Nonsteroidal anti-inflammatory drugs

Indication

- NSAIDs are indicated for mild to moderate pain

Dose information

Diclofenac :

- Adults: 50 mg orally three times daily, when required, or 75 mg orally twice daily, when required. Maximum: 150 mg/d
- Neonates, infants, children, and adolescents: safety and efficacy have not been established

Ibuprofen :

- Adults: 200 to 800 mg orally every 4 to 6 hours, when required. Maximum: 3,200 mg/d

- Pediatric patients aged 6 to 8 years or weighing 48 to 59 lbs: 2 tablets (200 mg) orally every 6 to 8 hours as needed
- Pediatric patients aged 9 to 10 years or weighing 60 to 71 lbs: 2.5 tablets (250 mg) orally every 6 to 8 hours as needed
- Pediatric patients aged 11 years or weighing 72 to 95 lbs: 3 tablets (300 mg) orally every 6 to 8 hours as needed

Naproxen :

- Adults: 500 mg orally initially; followed by 250 to 500 mg orally twice daily, when required. Maximum: 1.25 g/d
- Adolescents and children: 2.5 to 5 mg/kg orally every 8 to 12 hours has been used. Manufacturer states that adequate effectiveness or dose-response data are not available for pain conditions, but the experience in juvenile arthritis and other use experience have established that single doses of 2.5 to 5 mg/kg (as naproxen suspension), with total daily dose not exceeding 15 mg/kg/d, are well-tolerated in pediatric patients >2 years

Major contraindications

- Bovine protein hypersensitivity (diclofenac)
- Burns (diclofenac)
- Coronary artery bypass graft surgery (diclofenac, ibuprofen, naproxen)
- Eczema (diclofenac)
- Exfoliative dermatitis (diclofenac)
- NSAID hypersensitivity (diclofenac, ibuprofen, naproxen)
- Salicylate hypersensitivity (diclofenac, ibuprofen, naproxen)
- Skin abrasion (diclofenac)

Comments

- Treat with the lowest effective dose and shortest possible duration

Opioids

Indication

- Moderate to severe pain

Dose information

Morphine :

- Adults: 10 to 30 mg orally (immediate-release) every 3 to 4 hours, when required
- Pediatric patients: Individualize dosage and titrate as appropriate

Oxycodone :

- Adults: 5 to 30 mg orally (immediate-release) every 4 to 6 hours, when required
- Pediatric patients: Individualize dosage and titrate as appropriate

Tramadol :

- Adults: 50 to 100 mg orally every 4 to 6 hours, when required. Maximum: 400 mg/d
- Neonates, infants, children, and adolescents <17 years: safety and efficacy have not been established

Major contraindications

- Acute abdomen (morphine)
- Acute intoxication of CNS depressants (tramadol)
- Alcoholism (morphine)
- Asthma (tramadol)
- Brain tumor (morphine)
- Cardiac arrhythmias (morphine)
- CNS depression (morphine)
- Ethanol intoxication (tramadol)
- GI obstruction (oxycodone)
- Head trauma (morphine)
- Hypovolemia (morphine)
- Ileus (morphine, oxycodone)
- Increased intracranial pressure (morphine)
- MAOI therapy (morphine)
- Opiate agonist hypersensitivity (morphine, tramadol)
- Respiratory depression (morphine, oxycodone, tramadol)
- Shock (morphine)
- Status asthmaticus (morphine, oxycodone)

Non-drug treatments

Epidural steroid injections

Description

- Epidural injection with anesthetics and/or steroids under fluoroscopic x-ray or computed tomography guidance

Indication

- Temporary pain relief if more conservative methods have failed

Complications

- Pain
- Bleeding
- Infection
- Intravenous or intra-arterial injection
- Spinal fluid leak
- Steroid accumulation

Comments

- May provide temporary relief of targeted pain
- Follow-up is required to assess effect of injection

Facet injection

Description

- Fluoroscopically guided injection of the lumbar facet joint

Indication

- Relief of chronic low back pain believed to be of facet joint origin

Complications

- Bleeding

Comments

- Patients should discontinue NSAIDs and anticoagulants for at least 1 week before the procedure
- Patients should maintain a pain diary in order to facilitate evaluation of the efficacy of the injection

Evidence

In ADS, there is low-quality evidence supporting injection therapy for radicular pain:

- A meta-analysis evaluated the evidence for efficacy and effectiveness of conservative treatments in adult (degenerative) scoliosis. Small case series and individual case studies were included in evaluation of several of the treatment modalities; the number of studies included and total number of patients varied by modality and were not specified for all modalities. There was low-quality evidence from one study supporting injection therapy as conservative treatment for ADS (61 patients, retrospective study). There was no evidence higher than case series to support physical therapy and chiropractic care for ADS. [2] *Level of evidence: 3*

References

Bracing

Description

- A variety of brace types are available, including the following:
  - Thoracolumbar-sacral orthosis (TLSO): molded, can be worn under clothing
  - Nighttime-only braces: the Charleston bending brace and the Rosenthal brace
  - Full-torso brace: Milwaukee brace

Indication

- Bracing in JIS and AIS is recommended for skeletally immature children with curves between 25° and 45°, with the expectation that bracing will not correct the deformity, but rather will halt its progression
- In specific situations, such as the very skeletally immature, bracing is recommended for curves less than 25° if likelihood of progression is high
- Nighttime-only braces (the Charleston bending brace and the Rosenthal brace) may be suitable for small curves in neuromuscular scoliosis. Bracing does not halt curve progression; it has been used as a temporary device to delay spinal surgery until skeletal maturity or for sitting balance assistance
  - Braces are best for hypotonic, ambulatory cerebral palsy patients with short thoracolumbar curves less than 40°
  - Braces universally fail in patients with long thoracolumbar curves or Duchenne muscular dystrophy
- Bracing has no role in the treatment of ADS

Complications

- Braces, especially full-body types, are uncomfortable and may lead to difficulties with self-image and social problems with peers

- TLSO braces reduce lung capacity by about 20%
- TLSO braces may also cause temporary, mild renal impairment
- Braces may result in skin breakdown, leading to pressure sores and ulcers

Comments

- It is recommended that most braces be worn for greater than 20 hours per day; however, it is not clear if there is a direct relationship between time in brace and curve progression
- Full-torso brace must be worn for 23 hours daily. It is now used only for severe curves
- In ADS, bracing may temporarily relieve pain, but its benefits are outweighed by the consequences of promoting muscle atrophy and deconditioning, thereby worsening curve progression

Evidence

Bracing is more effective than observation for female patients with Cobb angle of 25° to 35°:

- A meta-analysis included 20 studies (randomized, controlled trials [RCT]; nonrandomized, clinical, controlled trials; and case-control studies) comparing the results of bracing with either observation, conservative treatment, or surgical treatment. Most included studies were of poor methodologic quality. Data suggested that bracing is more effective than observation for the prevention of scoliotic curve progression and may not contribute negatively to quality of life, especially for female patients with a Cobb angle of 25° to 30°. Comparison of bracing with surgery was difficult due to large differences in curve magnitude between the groups at the onset of studies. [3] *Level of evidence: 2*

There is very low quality evidence favoring use of braces in AIS:

- A Cochrane meta-analysis included two studies: a prospective cohort study with 286 girls (low-quality evidence that bracing had reduced curve progression at skeletal maturity) and an RCT of 43 girls (low-quality evidence for improved outcomes with a rigid brace*vs*an elastic one in terms of Cobb angle). There were no significant quality-of-life differences in the brace versus control groups in the meta-analysis. The authors concluded that there is very low-quality evidence in favor of using braces and that treatment decisions should be made by multidisciplinary consultation. [4] *Level of evidence: 2*

References

Physical and aqua therapy

Description

- An individualized program of core strengthening with aerobic and flexibility exercises to prevent muscle deconditioning and improve muscle fatigue; these are likely inciters of axial back pain in ADS

Indication

- Mild back pain in ADS

Complications

- Generally, none

Comments

- Used in conjunction with other therapies, especially analgesia, and postoperatively
- Patient education, participation, and adherence are the keys to success with this therapy

Evidence

In ADS, there is indeterminate evidence to support conservative therapies, including physical therapy, bracing, and chiropractic care:

- A meta-analysis evaluated the evidence for efficacy and effectiveness of conservative treatments in ADS. Small case series and individual case studies were included in evaluation of several of the treatment modalities; the number of studies included and total number of patients varied by modality and were not specified for all modalities. There was low-quality evidence from one study supporting injection therapy as conservative treatment for ADS (61 patients, retrospective study). There was no evidence higher than case series to support physical therapy and chiropractic care for ADS. [2]*Level of evidence: 3*

There is good quality evidence supporting bracing, limited evidence to support the use of physical therapy, and no evidence supporting surgery for AIS:

- A European systematic review of treatment modalities included physiotherapy, scoliosis inpatient rehabilitation (SIR), bracing, and surgery. There is limited evidence to support physiotherapy and SIR. There is a prospective multi-center controlled study as well as a meta-analysis supporting bracing. No controlled studies of any post-surgical follow-up period revealed any substantial evidence to support surgical treatment for AIS. [5] *Level of evidence: 2*

References

Surgery: posterior instrumented fusions, with or without osteotomies

Description

- Open, posterior-approach procedure to correct and stabilize the spine with metal rods accompanied by spinal arthrodesis (fusion)

Indications

- AIS:
  - Curves greater than 50° are candidates for spinal instrumentation and fusion
  - In addition, curves that progress to 45° to 50° despite brace treatment are considered failures and necessitate surgical intervention
  - Posterior spinal instrumentation and fusion remains the primary approach for surgical correction of AIS
  - Untreated AIS extending into adulthood typically requires anterior release with or without instrumentation in addition to posterior instrumentation and fusion, secondary to loss of curve flexibility with age
- ADS:
  - Decompression with posterior full curve fusion is indicated for back and deformity pain, curves greater than 45°, and greater than 2 mm of lateral listhesis, with good coronal and sagittal balance
- NMS:
  - Posterior pedicle screw constructs with or without corrective osteotomies usually provide immediate and long-term deformity correction without the need for anterior release surgeries
  - Pelvic fusion should be performed in any case of neuromuscular scoliosis with pelvic obliquity greater than 15°, and it should be considered in non-ambulatory patients with pelvic obliquity less than 15°

Complications

- Correction of significant thoracic kyphotic deformity, large Cobb angle curves, use of osteotomies, and pre-existing neurologic deficit have been predictors of postoperative neurologic complication
- AIS:
  - Rate of non-neurologic complications after spinal instrumentation and fusion has been historically quoted to range from 0% to 10%
- ADS:
  - Overall morbidity and mortality associated with surgical correction of ADS is significantly higher compared to childhood scoliosis surgery, based on age, deformity characteristics, and associated medical comorbidities
  - Neurologic complication rates for the surgical correction of ADS range from 1% to 5%. Risk factors for neurologic complications include significant kyphotic deformity and combined anterior-posterior surgical approaches

- Non-neurologic complications after spinal instrumentation and fusion for ADS range from 10% to 40%

- Medical complications include respiratory issues (*eg*, atelectasis, pneumonia, prolonged intubation), superficial and deep wound infections, deep venous thrombosis, thrombophlebitis, coagulopathy, electrolyte imbalances, urinary tract infections, prolonged ileus, superior mesenteric artery syndrome, pulmonary embolus, pneumothorax, and myocardial infarction

- Infection rate is 3% to 5% and is typically treated with operative irrigation with debridement and prolonged intravenous antibiotics, though hardware is typically not removed

- Predictors of perioperative complications and poor clinical outcome include tobacco use, history of asthma or chronic obstructive pulmonary disease, coronary or cerebrovascular disease, diabetes, nutritional deficits, osteoporosis, depression, and significant life stressor

- Complication rate is directly proportional to age

- NMS:

- Implant-related complications include pseudoarthrosis (failure of fusion), rod fracture, and/or screw pullout. Rates have decreased with the technological improvements in spinal fixation. Implant failure, such as rod breakage or screw loosening, may indicate an established or pending pseudoarthrosis, however, surgical revision is recommended only for pain or clinically significant loss of correction over time

- Posterior-only approach with osteotomies typically results in significantly increased blood loss and necessitates postoperative intensive-care stays

Comments

- Pedicle-screw-based systems have led to greater corrective forces, greater correction of deformity in 3 dimensions, lower rates of hardware failure or pseudoarthrosis, reduced levels fused, and obviation of the need for postoperative bracing

- In younger children, growing rods are used in posterior constructs, due to the need to maintain the continued growing potential of the spine. These children typically undergo several operative procedures in which the rods are incrementally lengthened to accommodate spinal growth

- In ADS, referrals to spinal surgeons are for treatment of pain, not typically the deformity itself

- Rigid curves typically require combined anterior and posterior procedures, which may be staged depending on the operative length of each part

- Controversy exists as to whether the caudal extent of fusion to should end at L5 or S1, or extend to the pelvis

- The use of bone morphogenic protein (BMP), though controversial and off-label except for anterior lumbar interbody procedures, has resulted in improved fusion rates compared to the standard use of autograft with or without allograft. In fact, studies have shown 96% anterior and 93% posterior fusion rates when BMP was used alone or in conjunction with autograft

Evidence

No controlled studies reveal substantial evidence to support surgery for AIS:

- A European systematic review (the number of studies and participants was not published) examined treatment modalities including physiotherapy, SIR, bracing, and surgery. There is limited evidence to support physiotherapy and SIR. There is a prospective multi-center controlled study as well as a meta-analysis supporting bracing. No controlled studies of any post-surgical follow-up period revealed any substantial evidence to support surgical treatment for AIS. [5] *Level of evidence: 2*

Risk of non-neurologic complications for AIS surgery related to blood loss, renal disease, operative time, and anesthesia:

- A prospective study of a multicenter cohort of 702 cases of surgical fusion for AIS from 2002 to 2004 was undertaken to determine prevalence of non-neurologic complications (15.4%) and to identify preoperative and operative factors that can increase this risk. Increased surgical blood loss, history of renal disease, operative time, and total anesthesia time were associated with a higher prevalence of complications. [6] *Level of evidence: 2*

Surgery for ADS provides functional improvement and deformity correction:

- A meta-analysis of 16 studies (553 patients) to evaluate the role of surgery for treating degenerative scoliosis (DS) in terms of improved function and correction of deformity found that surgery was an effective and reasonable treatment for ADS, providing significant functional improvement and deformity correction, but with high incidence of complications and re-operations. [7] *Level of evidence: 2*

Surgery for ADS benefits the elderly:

- A retrospective review of a prospective multicenter database stratified by age group was performed to assess if elderly patients (aged 65 to 85 years) had comparable outcomes to younger patients undergoing surgical correction of ADS. Quantitative measures included the Oswestry Disability Index (ODI), SF-36, SRS-22, and a numerical rating scale for back and leg pain (NRS). Follow-up rates were below 50% for all age groups. At 2-year follow up, there was statistically significant improvement in ODI, SRS-22, and NRS in all age groups, with some improvements greater among the elderly. At the 2-year follow up, the ODI was equal among all 3 age groups, as was the back and leg pain NRS. [8] *Level of Evidence: 2*

References

Surgery: combination anterior/posterior approaches

Description

- Anterior release of the disc space (open or endoscopic) followed by anterior fusion, in addition to posterior instrumentation and fusion

Indications

- Untreated AIS extending into adulthood typically requires anterior release with or without instrumentation in addition to posterior instrumentation and fusion, secondary to loss of curve flexibility with age
- For ADS, decompression with anterior and posterior fusion is indicated in cases of absent thoracic hyperkyphosis, severe stenosis with back and deformity pain, greater than 2 mm of lateral listhesis, and the need for correcting the loss of lumbar lordosis and mild sagittal imbalance

Complications

- In ADS, risk factors for neurologic complications include significant kyphotic deformity and combined anterior-posterior surgical approaches
- Overall morbidity and mortality associated with surgical correction of ADS is significantly higher compared to childhood scoliosis surgery based on age, deformity characteristics, and associated medical comorbidities
- Neurologic complication rates for the surgical correction of ADS range from 1% to 5%

Comments

- Controversy exists as to whether the caudal extent of fusion should end at L5 or S1, or extend to the pelvis
- The use of BMP, though controversial and off-label except for anterior lumbar interbody procedures, has resulted in improved fusion rates compared to the standard use of autograft with or without allograft. In fact, studies have shown 96% anterior and 93% posterior fusion rates when BMP was used alone or in conjunction with autograft

Surgery: anterior release, with or without instrumentation

Description

- Open surgical or endoscopic procedure to surgically remove intervertebral discs and cut restricting ligaments. A contoured rod may be placed to manipulate the spine into better alignment

Indications

- Anterior release with or without instrumentation is indicated for isolated thoracolumbar or lumbar curves or cases that require significant improvement in sagittal balance

- Anterior procedures provide a means of correction for lumbar hypolordosis by increasing disc height and, consequently, indirectly decompressing the neural foramina; they reduce pseudoarthrosis and instrument failure

- For ADS, anterior release with or without instrumentation (and with or without combined posterior decompression and fusion) may be indicated, depending on the severity and flexibility of the deformity

- For neuromuscular scoliosis, anterior release surgeries may be indicated in some cases, but a posterior-only approach using pedicle-screw-based constructs is more common

Complications

- Coe et al. reviewed the morbidity and mortality database of the Scoliosis Research Society and found 6,334 patients who underwent surgery for AIS. Overall non-neurologic complication rate was 5.2% in anterior-only surgical approach

Comments

- The role for anterior release surgeries with or without instrumentation has decreased with the advancement of more powerful posterior pedicle-screw constructs and improved osteotomy techniques

- Rigid curves typically require combined anterior and posterior procedures, which may be staged depending on the operative length of each part

Surgery: decompression alone

Description

- Surgical removal of structures impinging on spinal nerve roots or the spinal cord

Indications

- Decompression alone is recommended for neurogenic claudication with need of 1 to 2 level laminectomy with Cobb angle less than 30°, less than 2 mm of lateral listhesis, good coronal and sagittal balance, and minimal or no back pain or deformity symptoms

Complications

- When performed for stenosis associated with scoliosis, scoliotic deformity may progress with symptom worsening

Comments

For ADS:

- Decompression with limited fusion is recommended for neurogenic claudication with need for extensive laminectomy in the setting of Cobb angle less than 30°, greater than 2 mm of lateral listhesis, absent back pain or deformity symptoms, and good coronal and sagittal balance

- Decompression with posterior full curve fusion is indicated for back and deformity pain, curves greater than 45°, greater than 2 mm of lateral listhesis, with good coronal and sagittal balance
- Decompression with anterior and posterior fusion is indicated in cases of absent thoracic hyperkyphosis, severe stenosis with back and deformity pain, greater than 2 mm of lateral listhesis, and the need for correcting the loss of lumbar lordosis and mild sagittal imbalance

Special circumstances

Comorbidities

- Predictors of perioperative complications and poor clinical outcome include tobacco use, history of asthma or chronic obstructive pulmonary disease , coronary or cerebrovascular disease, diabetes, nutritional deficits, osteoporosis , depression , and significant life stressor
- Complication rate is directly proportional to age

Patient satisfaction/lifestyle priorities

- Because smoking interferes with fusion, patients should stop before their surgery and remain abstinent after surgery
- Adult patients may benefit from a short (1-2 week) in-patient rehabilitation stay after scoliosis surgery
- Children and adolescents are usually able to return to school within 4 to 6 weeks
- Adults may need to stay home from work up to 12 weeks, depending on the type of surgery
- After full healing, patients with a thoracic fusion can usually resume normal activity levels, as the biomechanics of the spine do not significantly change
- After treatment for scoliotic curves at other levels, activity restrictions or avoidance of some types of sports may be required (*eg*, gymnastics
- Female patients can have a normal pregnancy and delivery after a fusion procedure

Consultation

- Consultation with a fellowship-trained spine surgeon (orthopedist or neurosurgeon) is appropriate for children, adolescents, and adults with scoliotic curves who are symptomatic or have curvatures with indications for bracing or surgery
- In general, skeletally immature children should be referred if they have a curve of more than 20°, and those who are skeletally mature should be referred with a curve exceeding 40°
- Patients with respiratory symptoms attributed to scoliotic curves should have pulmonary function testing and may benefit from consultation with a pulmonologist

- If significant respiratory abnormalities are found on spirometry, this may assist in determining need for surgery in patients with significant curves

Follow-up

For scoliotic curves being followed by observation only, repeat standing scoliosis radiographs should be done at appropriate intervals:

- Children with curves of less than 20°—follow up every 6 to 12 months
- Adolescents with curves greater than 20°—repeat every 3 to 4 months
- After skeletal maturity, patients with curves greater than 30°—at yearly intervals for 2 to 3 years, then every 5 years because of potential risk for progression

Complications of untreated scoliosis include:

- Development/worsening of kyphosis
- Restricted cardiopulmonary function
- Back pain

Patient education

Online information for patients

- National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS): Questions and answers about scoliosis in children and adolescents
- The National Scoliosis Foundation : A patient-led nonprofit organization dedicated to helping children, parents, adults, and health-care providers to understand the complexities of spinal deformities such as scoliosis
- KidsHealth.org from the Nemours Foundation: Provides parents and their children with education and advice on scoliosis and other health issues
- Orthoinfo.com from the American Academy of Orthopedic Surgeons: Summary of diagnosis and treatment
- Healthychildren.org from the American Academy of Pediatrics: Information on scoliosis and other pediatric health issues

Resources

Summary of evidence

Evidence

Scoliosis screening programs have a low positive predictive value for identifying scoliotic curves:

- A meta-analysis of 36 retrospective cohort studies of scoliosis screening programs was done to evaluate the clinical effectiveness of school screening for adolescent idiopathic scoliosis. The pooled referral rate (using forward bend testing, angle of trunk rotation, or topography studies) for radiography was 5%. The positive predictive values for predicting curves greater than 10°, greater than 20°, and receiving treatment were 28%, 5.6%, and 2.6%, respectively.[1] *Level of evidence: 2*

In ADS, there is low-quality evidence supporting injection therapy for radicular pain:

- A meta-analysis evaluated the evidence for efficacy and effectiveness of conservative treatments in adult (degenerative) scoliosis. Small case series and individual case studies were included in evaluation of several of the treatment modalities; the number of studies included and total number of patients varied by modality and were not specified for all modalities. There was low-quality evidence from one study supporting injection therapy as conservative treatment for ADS (61 patients, retrospective study). There was no evidence higher than case series to support physical therapy and chiropractic care for ADS. [2] *Level of evidence: 3*

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- A Cochrane meta-analysis included two studies: a prospective cohort study with 286 girls (low-quality evidence that bracing had reduced curve progression at skeletal maturity) and an RCT of 43 girls (low-quality evidence for improved outcomes with a rigid brace*vs*an elastic one in terms of Cobb angle). There were no significant quality-of-life differences in the brace versus control groups in the meta-analysis. The authors concluded that there is very low-quality evidence in favor of using braces and that treatment decisions should be made by multidisciplinary consultation. [4] *Level of evidence: 2*

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Guidelines

The American College of Radiology has produced the following:

- ACR/SPR Practice Guideline for the Performance of Radiography for Scoliosis in Children . Reston, VA: American College of Radiology; 2009

The Society on Scoliosis Orthopaedic and Rehabilitation Treatment has produced the following:

- Negrini S, Grivas TB, Kotwicki T, Rigo M, Zaina F. Guidelines on "Standard of management of idiopathic scoliosis with corrective braces in everyday clinics and in clinical research": SOSORT Consensus 2008 . Scoliosis. 2009;4:2

The U.S. Preventive Services Task Force has produced the following:

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Codes

ICD-9 code

- 737.43 Scoliosis
- 737.30 Scoliosis, acquired
- 754.2 Scoliosis, congenital
- 737.40 Curvature of spine, unspecified
- 737.41 Kyphosis
- 737.42 Lordosis

FAQ

- **Should I screen all school-age patients for scoliosis?**There is some controversy over this issue. Routine screening of asymptomatic adolescents for AIS is not recommended by the U.S. Preventive Services Task Force as of 2004. A 2008 position statement by the American Academy of Orthopaedic Surgeons, the Scoliosis Research Society, the Pediatric Orthopaedic Society of North America, and the American Academy of Pediatrics did not support any recommendation against scoliosis screening. Physicians should understand scoliosis screening methods and screen if there are suggestive findings on physical examination, if it is requested by the patient or a pediatric patient’s parent as part of the physical examination, or if there is a family history of scoliosis
- **How do I perform a scoliosis screening exam, and what constitutes a positive exam?**The Adams forward bend test is a simple maneuver that can help detect coronal imbalance. A scoliometer can also be used to measure the coronal deformity during the Adams forward bend test, but it is less accurate in the lower thoracic and lumbar spine, as the costovertebral attachments in this region do not produce a rib hump. Positive results include a 'rib hump' seen on the Adams forward bend test (correlates with a scoliotic curve greater than 10°) and an angle of trunk rotation (ATR) value greater than 5° on the scoliometer (correlates with a deformity of at least 10°). An ATR of 6° to 10° is used as the cut-point for obtaining x-ray imaging results in higher

positive predictive values. An ATR of 7° is a commonly used cut-point for progressing to further work-up, but the ideal cut-point is not known

- **What are the appropriate imaging modalities if physical exam is suggestive and I want to confirm the diagnosis?**Initially, long cassette (36-inch) posteroanterior and lateral plain films shot at a distance of 72 inches of the standing patient with hips and knees fully extended are the standard postures for examining overall spinal alignment. The Cobb technique can be used to measure the magnitude of the curve. CT and MRI may be useful adjunctive measures but are not usually needed for diagnosis

Current contributors

David M. Ibrahimi, MD , Department of Neurosurgery, University of Maryland Medical Center, Baltimore, Maryland

Christopher M. Maulucci, MD , Department of Neurosurgery, University of Maryland Medical Center, Baltimore, Maryland

Charles Sansur, MD, MHS , Assistant Professor of Neurosurgery, Director of Spine Surgery, University of Maryland Medical Center, Baltimore, Maryland