Clinical Practice & Referral Guideline: Scoliosis

*The recommendations in the below guideline do not indicate an exclusive course of treatment. The guidelines intent is to build a consensus of care in the pediatric market and provide a framework for clinical decision making.*

PURPOSE: To assist the clinician in the evaluation of scoliosis and determine when referral to Orthopedics is warranted.

DEFINITION: Scoliosis is defined as a lateral curvature of the spine that is usually accompanied by rotation. Scoliosis is not a diagnosis but a description of a structural alteration that occurs in a variety of conditions.

By convention, >10° of curvature (as measured by the Cobb angle) defines a scoliosis. Curves ≤10° are referred to as mild spinal asymmetry and usually are of no long term clinical significance.

THREE BROAD CATEGORIES OF SCOLIOSIS:

Neuromuscular:
- Occurs in patients with neurologic or musculoskeletal problems such as cerebral palsy, myelomeningocele, or muscular dystrophy
- Is the result of muscle imbalance and lack of trunk control
- May be structural or nonstructural (which has no rotational component)

Congenital:
- Results from asymmetry in the vertebrae secondary to congenital anomalies (hemivertebrae, failure of segmentation)
- Usually manifests before adolescence

Idiopathic:
- There is no definite etiology
- Three subcategories based on age at presentation
  - Infantile: 0-3 years
  - Juvenile: 4-10 years
  - Adolescent: ≥11 years

Adolescent idiopathic scoliosis (AIS) is the most common form of idiopathic scoliosis accounting for 80-85% of cases. The prevalence of AIS is approximately 3%, but only 10% of those require treatment. Females and males are affected equally for small curves. For curves over 30°, the ratio increases to 10:1 (F: M). The natural history of untreated AIS is one of stabilization or progression of the curve. Untreated curves **progress in**
approximately 2/3 of skeletally immature patients before they reach skeletal maturity. Scoliosis can continue to progress after skeletal maturity in untreated patients with curves >30°, but the highest risk of progression is in curves >50°. Most patients with untreated AIS have little, if any, functional limitation or pain in adulthood.

Presentation:
- Incidental finding on physical exam
- Complaints related to the deformity (asymmetry of the shoulders, hips, breasts, etc.)
- Noted by school nurse during screening at school
- Restrictive lung disease (curves >80° usually presenting before adolescence--rare)

Evaluation:
- **Objectives:**
  - Identify an underlying etiology
  - Assess the magnitude of the curve and need for x-rays
  - Determine the risk of progression
- **History:**
  - Determine if an underlying etiology exists and risk for progression (based upon estimation of remaining progression of linear growth)
  - When was the deformity first noted?
  - What is the rate of progression? (rapid progression suggests a non-idiopathic etiology)
  - Is there associated back pain, muscle weakness, bowel or bladder problems, headache, or neck pain?
  - Is there shortness of breath or difficulty breathing?
  - Has the pubertal growth spurt begun?
  - Has menarche occurred? (pubertal growth spurt usually occurs just before menarche, linear growth completed 24 months after menarche)
  - Is there a family history? (AIS tends to run in families)
  - Is there a history of lower limb fracture, joint infection, or arthritis? (which may result in leg length discrepancy)
- **Physical Exam:**
  - Measure height and plot on growth curve to estimate remaining growth potential
  - Assess Tanner stage
  - Examine the skin for café-au-lait spots and axillary freckling (neurofibromatosis); vascular, hypopigmented or a patch of hair overlying the spine (spinal dysraphism); dimpling in the lumbosacral area (intraspinal tumor)
  - Assess for leg length discrepancy
  - Examine for excessive skin or joint laxity (Ehlers-Danlos or Marfans)
  - Examine feet for high arches and hammer toes (neuromuscular disease, e.g. Friedreich ataxia)
  - Scoliosis exam:
    - Simple inspection for asymmetry of shoulders, waist, etc.
    - Adams’ Forward Bend test
    - If asymmetry is noted, measurement with a scoliometer if available
• **Radiographic Evaluation:**
  - Required to confirm the diagnosis of scoliosis, determine the type and severity, and evaluate skeletal maturity
  - Indications for x-ray include:
    - Scoliometer reading $\geq 7^\circ$
    - Clinically evident scoliosis on exam
    - Thoracic or lumbar asymmetry on exam of skeletally immature children with a family history of scoliosis
    - Monitoring progression in previously diagnosed AIS
  - Standing full length PA (PA views minimize radiation to the breasts and thyroid) and lateral views of the spine **exposed on a single long film cassette**
  - Determination of the **Cobb angle** (measurement of degree of curvature in the coronal plane on the xray)
  - Determination of the **Risser sign** (visual grading of the degree to which the iliac apophysis has undergone ossification and fusion): the lower the grade, the more growth remaining and the greater the risk of curve progression
  - MRI may be indicated in cases suggestive of intraspinal pathology and in children with a left-sided thoracic curve, boys with a curve that demonstrates any degree of progression, and all children with a curve that rapidly progresses.

**Risk for Progression:**
- Unfortunately, it is impossible to predict with complete accuracy which curves will progress and which will not.
- Risk is increased 3-10 times in girls compared to boys
- Curves of 20-29° more likely to progress by $> 5^\circ$ than curves of 5-19°
- Patients younger than 12 years have 3 times the risk of progression
- 68% of girls post menarche have no further progression

**REFERRAL:**
- Refer all prepubescent children with a **Cobb angle** $> 10^\circ$ and all cases of congenital or neuromuscular scoliosis to orthopaedics
- Refer all adolescent patients with **Cobb angle** $> 20^\circ$
- Refer all skeletally mature patients with **Cobb angle** $> 30^\circ$

**Treatment:**
- Goal of treatment of AIS is a curve with a Cobb angle of $30^\circ$ or less at skeletal maturity
- Options for treatment include observation, bracing and surgery
- Choice of therapy depends upon the degree of curvature and potential for further growth.
  - **Observation:** Adolescents with Cobb angles $< 20^\circ$ may be followed clinically and/or radiographically every 6 months by their primary care provider, if the provider feels comfortable doing so. **Curves can progress 1° per month during the adolescent**
growth spurt. If the Cobb angle has progressed by ≥ 5°, treatment may be indicated and a referral made.

- **Bracing:** Bracing does not correct curvature that is present at the time of diagnosis, but may prevent curve progression and reduce the need for surgery.
  - Indicated only for skeletally immature patients (Risser sign 0-2), and:
  - Cobb angle is 20-40°, and:
  - Curve has progressed ≥ 5°
  - **Contraindications:** skeletal maturity, Cobb angle > 40°, obesity, inability to comply

- **Surgery:** Primary goal of surgical treatment of AIS is prevention of curve progression through spinal fusion; partial curve correction is also frequently achieved as well for better cosmesis.
  - Indicated in progressive curves with Cobb angle ≥ 45°
  - Most common surgical procedure is a posterior spinal fusion with instrumentation and bone grafting.
  - Contemporary implants are segmental, which consists of a variety of hooks, screws, and wires that can be used to attach the rods to the spine at multiple vertebrae, which affords the surgeon greater control over positioning and rotation of the spine.

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**References:**
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