Advances in the Management and Treatment of Adolescent Idiopathic Scoliosis

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Overview

• Introduction to adolescent idiopathic scoliosis (AIS)
• The clinical evaluation process for AIS
  – Initial presentation and screening
  – Physical examination
  – Imaging studies
• Available treatment options for AIS
• Factors that affect the choice of therapy
• Genetic testing to predict curve progression—the ScoliScore AIS Prognostic Test
• The role of external independent medical review in determining medical necessity for genetic testing for AIS progression
Adolescent Idiopathic Scoliosis: An Overview

- AIS is the most common type of scoliosis
- Underlying cause remains unknown
- Affects children between the age of 10 and young adulthood
  - AIS is most commonly diagnosed between the ages of 10 and 12 years
- Parents may first notice it, or it may be discovered during school screenings or at pediatric visits
- Once scoliosis is suspected, patients are referred to orthopedic specialists
  - Patient is evaluated to determine the severity of the patient’s curvature
Natural History of AIS

- AIS is not typically associated with significant morbidity
- Larger scoliotic curves or curves with significant rotation may cause significant cosmetic concerns
- Progressive deterioration of the scoliotic curve may result if scoliosis is left untreated
- Large scoliotic curves can lead to:
  - Diminished lung capacity
  - Development of restrictive lung disease
Factors That Affect Curve Progression in AIS

- **Sex**
  - Girls are more likely than boys to have curves that progress to a point that treatment is ultimately required

- **Age at menarche (girls)**

- **Remaining growth potential**

- **Magnitude of the curve at the time of diagnosis**
Role of Genetics in AIS

• Population studies have shown that:
  – Scoliosis runs within families
  – There is a higher prevalence of scoliosis among relatives of patients with scoliosis than within the general population

• More recent family studies have shown that AIS is a polygenic disorder with multiple patterns of inheritance

• The identification of genetic markers related to AIS curve progression has led to the development of the ScoliScore AIS Prognostic Test

Initial Presentation for AIS

• AIS does not usually cause any pain, neurological dysfunction, or respiratory problems
• Most patients initially present due to perceived deformity
• The patient, a family member, school nurse, or primary care physician may perceive asymmetry about the patient’s shoulders, waist, or rib cage
Screening for AIS: The Adams Forward Bend Test

- Often used by pediatricians and in initial school screenings
- The child bends forward dangling the arms, with the feet together and knees straight
- In a child with scoliosis, the examiner may observe an imbalanced rib cage or other deformities along the back
- A scoliometer, which measures distortions of the torso, is often used in conjunction with the Adams forward bend test
Physical Examination

• Baseline assessment of posture and body contour
• Assessment of:
  – Lower (and often upper) extremity reflexes
  – Abdominal reflex patterns
  – Hamstring tightness
• Screening for ataxia and/or poor balance or proprioception (i.e., Romberg test)
• Measurement of leg length
Determining the Magnitude of the Scoliotic Curve: The Cobb Angle

- Derived from a standard posteroanterior standing x-ray of the spine
  - A line is drawn perpendicular to the top of the superior vertebrae of the scoliotic curve
  - A similar perpendicular line is drawn along the bottom of the inferior vertebrae

Imaging Studies for AIS: Recommendations of the AAOS, SRS, POSNA, and AAP

- Adolescents without significant spinal deformity who are referred to a specialist for evaluation often do not require x-rays
- Avoid the inappropriate use of x-rays
- If x-rays are needed, physicians should take necessary precautions to limit the patient’s exposure to radiation
- Routine magnetic resonance imaging (MRI) evaluation of all patients with AIS is not recommended

AAOS=American Academy of Orthopaedic Surgeons; SRS=Scoliosis Research Society; POSNA=Pediatric Orthopaedic Society of North America; AAP=American Academy of Pediatrics

Factors That Affect the Choice of Therapy for AIS

- Type and degree of curve
- Child’s age
- Number of remaining growth years until the child reaches skeletal maturity
Nonsurgical Treatment Options for AIS

• Observation
  – An appropriate option when the curve is mild (<20 degrees) or if the child is near skeletal maturity
  – The curve should be re-checked every 3 to 6 months to see that it is not progressively worsening

• Bracing
  – The goal is to prevent scoliotic curves from getting worse
  – Can be effective if the child is still growing and has a spinal curvature between 25 and 45 degrees
  – There are several types of braces, mostly the underarm type
Surgery for AIS

- Generally consists of a spinal fusion and instrumentation
  - An implant is attached to the vertebrae of the most curved portion of the spine, along with bone graft for that portion of the spine to heal solid
  - Can be performed in the anterior spine, posterior spine, or both
- May be recommended if the curve is >45 degrees and the child is still growing
- If the patient has reached skeletal maturity, surgery may still be recommended for curves >50 to 55 degrees
Genetic Testing to Predict Curve Progression: The ScoliScore AIS Prognostic Test

- A multi-marker genetic test that provides an assessment of the likelihood of spinal curve progression for patients diagnosed with AIS
- Analyzes DNA extracted from a saliva sample
  - Compares genetic markers that have been linked in clinical trials to the risk of spinal curve progression
  - The more genetic markers there are, the higher the score, indicating high risk of curve progression
ScoliScore:
Earlier Intervention May Improve Outcomes

• Assigns a numerical value of the likelihood of AIS curve progression that is specific to each patient
  – Facilitates earlier identification of appropriate treatment plans for individual patients when used in conjunction with other diagnostic information

• Potentially reduces:
  – Office visits
  – Exposure to diagnostic radiation
  – Unnecessary treatments
  – Psychological distress
ScoliScore: Health Plan Coverage

- Due to lack of studies, many health plans consider ScoliScore experimental and investigational
- Most health plans do not cover ScoliScore
  - Evidence about the usefulness of the ScoliScore test is extremely immature, thus, by any measure, must be considered investigational. To date, only a single validation study, along with multiple editorials, have been published. The test is not covered.
  - The ScoliScore AIS Prognostic Test is considered experimental and investigational and is therefore not covered.
The Role of External Independent Review in Determining Medical Necessity for Genetic Testing for AIS Progression

- An independent medical review looks at whether or not a specific therapy or procedure is medically necessary.
- Although many health plans do not cover genetic testing to predict curve progression in AIS, ongoing and future studies may lead to widespread acceptance of the procedure.
- Independent review organizations (IROs) provide ready access to specialists, which healthcare plans may lack internally.
  - Allows for unbiased and timely determinations of whether the requested procedure falls under the medical necessity guidelines.
  - Board-certified physician specialists who work with IROs keep up-to-date with the latest medical research literature and the latest standard of care, staying on top of continually evolving technologies and procedures as they are studied more extensively and potentially accepted into clinical guidelines.
Conclusions

• Successful treatment of AIS remains a challenge for orthopedic surgeons, who must continually monitor patients in order to effectively time treatment according to the progression of the scoliotic curve

• The continual evolution of diagnostic and treatment approaches to AIS has led to increased emphasis on the importance of early detection and management of AIS

• New technologies and procedures, such as ScoliScore, facilitate earlier diagnosis and identification of deformities by providing patient-specific information that allows physicians to select more appropriate treatment interventions

• Results of ongoing and future studies may help to establish genetic testing as a standard of care for patients with AIS
Questions and Answers

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