Kansas City

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Interventions for Progressive Scoliosis

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Next Review: 11/2014

Policy

Blue Cross and Blue Shield of Kansas City (Blue KC) will provide coverage for interventions for progressive scoliosis when it is determined to be medically necessary because the criteria shown below are met.

When Policy Topic is covered

A cervical-thoracic-lumbar-sacral or thoracic-lumbar-sacral orthosis may be considered **medically necessary** for the treatment of scoliosis in juvenile and adolescent patients at high-risk of progression which meets the following criteria:

- Idiopathic spinal curve angle between 25 and 40 degrees; AND
- Spinal growth has not been completed (Risser grade 0-3; no more than 1 year post-menarche in females)

  OR
- Idiopathic spinal curve angle greater than 20 degrees; AND
- There is documented increase in the curve angle; AND
- At least 2 years growth remain (Risser grade 0 or 1; pre-menarche in females)

When Policy Topic is not covered

Use of an orthosis for the treatment of scoliosis that does not meet the criteria above is considered **investigational**.

Vertebral body stapling for the treatment of scoliosis is considered **investigational**.

Considerations

This policy does not address conventional surgery for scoliosis in patients with curve angles measuring 45 degrees or more. Brace treatment for idiopathic scoliosis is usually recommended for juveniles and adolescents with curves measuring between 25 and 40 degrees who have not completed spinal growth, with maturity defined as Risser 4, or 2 years post-menarche for girls. (1, 2) Bracing may also be recommended for curves greater than 20 degrees in a patient who has a rapidly progressing curve with more than 2 years of growth remaining.

- A cervical-thoracic-lumbar-sacral orthosis is primarily prescribed for patients with thoracic apices above T7 for control of upper thoracic sagittal deformities and for other spinal deformities not amenable to treatment with lower-profile designs.
- A low profile, rigid thoracic-lumbar-sacral orthosis worn full-time (18-23 hours per day) through skeletal maturity is used for most idiopathic curve patterns with a thoracic curve apex at or below T7 (the majority of idiopathic curves).
- Night time bracing systems are more effective in patients with isolated flexible thoracolumbar and lumbar curves than in double curves; they may also be indicated in patients who are noncompliant with a full-time wear program, patients in whom other types of orthotic management had failed, and patients nearing skeletal maturity who may not require full-time wear.

Description of Procedure or Service

Orthotic bracing attempts to slow curve progression and reduce the need for fusion surgery in patients with progressive scoliosis. Recently, fusionless surgical procedures (e.g., vertebral body stapling and implantation of vertical titanium growing rods) have been evaluated as alternatives to bracing to slow or correct curve progression in pediatric patients with scoliosis.

Background
Scoliosis is an abnormal lateral and rotational curvature of the vertebral column. Treatment of scoliosis currently depends on 3 factors: the cause of the condition (idiopathic, congenital, or secondary), the severity of the condition (degrees of curve), and the growth of the patient remaining at the time of presentation. Children who have vertebral curves measuring between 25 and 40 degrees with at least 2 years of growth remaining are considered to be at high-risk of curve progression. Genetic markers to evaluate risk of progression are also being evaluated. Since severe deformity may lead to compromised respiratory function and is associated with back pain in adulthood, surgical intervention with spinal fusion is typically recommended for curves that progress to 45 degrees or more.

Bracing is used in an attempt to reduce the need for spinal fusion by slowing or preventing further progression of the curve during rapid growth. Commonly used brace designs include the Milwaukee, Wilmington, Boston, Charleston, and Providence orthoses. The longest clinical experience is with the Milwaukee cervical-thoracic-lumbar-sacral orthosis (CTLSO). Thoracic-lumbar-sacral orthoses (TLSO), such as the Wilmington and Boston braces, are intended to improve tolerability and compliance for extended (more than 18-hour) wear and are composed of lighter-weight plastics with a low-profile (underarm) design. The design of the nighttime Charleston and Providence braces is based on the theory that increased corrective forces will reduce the needed wear time (i.e., daytime), thereby lessening social anxiety and improving compliance. Braces that are more flexible than TLSOs or nighttime braces, such as the SpineCor, are also being evaluated. The SpineCor is composed of a thermoplastic pelvic base with stabilizing and corrective bands across the upper body.

Fusionless surgical procedures, such as vertebral body stapling, are being evaluated as an alternative to bracing. It is hoped that fusionless procedures improve the curve as well as prevent its progression, maintain spine mobility following correction, and provide an effective treatment option for patients who are non-compliant or refuse to wear a brace. In the current stapling procedure, nickel-titanium alloy staples with shape memory are applied to the convex (outer) side of the curve. The shape memory allows the prongs to be straight when cooled and clamp down into the bone when the staple returns to body temperature. The goal of vertebral stapling is to unilaterally reduce the rate of spine growth, thus allowing the other side to “catch up”. The memory shape staple was tested in a goat model of scoliosis for safety and efficacy prior to its use in humans. The vertical expandable prosthetic titanium rib (VEPTR), described in more detail in policy number 7.01.110, is also being explored for treatment of infantile and juvenile scoliosis that has advanced beyond 45 degrees. Use of the VEPTR requires expansion surgery every 4-6 months as growth occurs and may be replaced as needed.

Regulatory Status
Some of the braces used for the treatment of scoliosis are considered Class I devices by the U.S. Food and Drug Administration (FDA). Examples include the Boston scoliosis brace and the SpineCor Scoliosis System.

Staples, using a shape memory nickel-titanium alloy, have 510(k) clearance from the FDA for a variety of indications for bone fixation. For example, Nitinol staples (Sofamor Danek, Memphis Tenn.) are indicated for fixation with spinal systems. Other memory shape staples that have 510(k) clearance for bone fixation include the OSStaple™ and the reVERTO™. Vertebral body stapling in scoliosis is considered off-label use.

Rationale

This policy was created in 2010 and updated periodically with searches of the MEDLINE database. The most recent search was performed though April 2, 2013.

The Scoliosis Research Society (SRS) Committee on Bracing and Nonoperative Management provided evidence-based recommendations in 2005 for bracing studies to facilitate comparison of brace trials. (3) The first study to use the SRS criteria concluded that a brace should prevent progression in 70% of patients to be considered effective. (4) The SRS evidence review and recommendations may also aid in the evaluation of fusionless surgical treatments for scoliosis progression in children.

The SRS review of the natural history of scoliosis indicates that skeletally immature patients and patients with larger curves (between 20 and 29 degrees) are significantly more likely to have more than 5 degrees curve progression. (3) Success from brace treatment is most frequently defined as progression of less than 5 degrees before skeletal maturity, although alternative definitions in the literature may include progression of less than 10 degrees before skeletal maturity or preventing the curve from reaching the threshold for surgical intervention. Surgery is usually recommended when the curve magnitude exceeds 45–50 degrees (before or at skeletal maturity), although many patients will not undergo surgery at this point. Based on this information, the SRS provided the following recommendations for brace studies on adolescent idiopathic scoliosis (AIS):

- Optimal inclusion criteria for AIS studies are patients 10 years or older, Risser sign 0 to 2, initial curve magnitude of 25 to 40 degrees, and no prior treatment at the initiation of brace treatment. (Risser sign is defined by the amount of calcification present in the iliac apophysis and measures remaining spinal growth by progressive anterolateral to posteromedial ossification. Immature patients will have 0% to 25% ossification [Risser grade 0 or 1], while 100% ossification [Risser grade 5] indicates maturity with no spinal growth remaining. Children may progress from a Risser grade 1 to grade 5 over a brief, e.g., 2-year, period.)
- Assessment of brace effectiveness should include the percentage of patients with less than 5 or greater than 6 degrees of progression at maturity, curves exceeding 45 degrees at maturity, and progression resulting in surgery or the recommendation for surgery.
- Intent-to-treat analysis should be performed, regardless of compliance. Efficacy analysis should also be considered, in which noncompliant patients are excluded in the analysis.
- A minimum of 2 years' follow-up beyond skeletal maturity should be obtained for each patient who was "successfully" treated with a brace. Skeletal maturity is considered achieved when less than 1-cm change in standing height has occurred on measurements made on 2 consecutive visits 6 months apart, when Risser 4 is present, or in females, when the patient is 2 years post-menarche.

Bracing

Dolan and Weinstein published a systematic review of observation and bracing in AIS in 2007. (5) Selection criteria for inclusion were: study of patients with AIS (diagnosed at or after the age of 8 years), who met the following indications for brace initiation (primary angle between 20 and 45 degrees, age younger than 15 years, Risser 0 to 2) and had follow-up to at least skeletal maturity. Fifteen studies of bracing alone were included in the analysis, with a range for surgery of 1% to 43% and a pooled rate of 23%. Three studies were included for observation alone, with a pooled rate of 22% for surgery and a range of 13% to 38%. Both the study population (school screening program) and indications for surgery were unusual in the observational studies. This meta-analysis is limited by the heterogeneity of the study results and illustrates the difficulty in evaluating bracing efficacy when study populations include patients who would not progress without a brace. The authors considered their recommendation (no clear advantage of bracing) to have a grade of D, due to "troublingly inconsistent or inconclusive studies of any level."

Nighttime Brace. Using the new SRS criteria, Janicki and colleagues reported outcomes from a database of patients with AIS who had used a thoracic-lumbar-sacral orthosis (TLSO) or a nighttime orthosis. (4) Retrospective analysis identified 160 patients treated orthotically for idiopathic scoliosis between 1992 and 2004. Patients with incomplete follow-up were phoned and asked to return if needed. From the cohort of 160 patients, 83 met the SRS inclusion criteria and had complete data. Due to poor outcomes with the TLSO, which the investigators suspected were predominantly due to a lack of compliance, practice had been changed from using a TLSO to recommending a nighttime orthosis. Thus, the 48 patients treated with a TLSO and 35 treated with a nighttime orthosis were not concurrent. For patients with an initial curve between 25-40 degrees and treated with a TLSO, 85% progressed

greater than 5 degrees, 56% progressed to greater than 45 degrees, and 79% progressed to surgery. With the nighttime orthosis, 69% progressed greater than 5 degrees, 45% progressed to greater than 45 degrees, and 60% progressed to surgery. Thus, only 21% in the TLSO group and 40% in the nighttime orthosis group were considered to have had successful orthotic management. Subgroup analysis showed little benefit of either brace type in patients with an initial curve between 36 and 40 degrees, with 86% of the TLSO group and 91% of the nighttime orthosis group progressing to surgery.

Micro-computer Controlled Brace. Lou et al. published a pilot study that compared the smart brace with a standard rigid brace in 12 patients with scoliosis. (6) Compliance with the microcomputer-controlled brace in the first year of bracing (2 years of total bracing) was similar in the 2 groups. The smart brace was associated with greater pad pressure and improved outcomes. None of the patients in the smart brace group had a significant change in their curves (a Cobb angle change of less than 5 degrees), whereas 2 of 6 patients in the standard TLSO group had a significant change in Cobb angle (7 and 20 degrees) over the 3 years of the study.

Flexible Brace. In 2008, Wong and colleagues reported a prospective study of clinical efficacy and acceptance of rigid or flexible spinal bracing in 43 patients with moderate adolescent scoliosis. (7) Female patients with a Cobb angle between 20 and 30 degrees, apical vertebra below T5, age between 10 and 14 years, and Risser sign equal to or less than 2 were randomized to the flexible SpineCor orthosis or a rigid underarm brace. The subjects were requested to wear the brace 23 hours a day, with one hour for bathing and physical exercises. Follow-up visits took place after the first month of intervention and then every 3 months. Acceptance of the brace was measured with a 16-question visual analog scale (VAS) assessing pain, skin irritation, and daily activities. The patients were followed until completion of skeletal growth or removal from the study because of curve progression greater than 5 degrees. At the end of a 45-month study period, a significantly lower percentage of the subjects (68%) in the flexible brace group did not show curve progression compared to nearly all (95%) of subjects in the rigid brace group. Patients' acceptance of the two orthoses was similar. Although the rigid brace caused significantly more problems with heat (85% vs. 27%, respectively), as well as difficulties with donning and doffing, the patients using the elastic braces had difficulties with toileting.

Plewka et al. reported the efficacy of the SpineCor brace (n=45) compared with physiotherapy and observation (n=45) in children with scoliosis. (8) The control group comprised children who qualified for brace treatment but whose parents did not agree to treatment or in whom the treatment was not possible due to social reasons. Baseline measures of the 2 groups were similar. After 2 years of treatment, the patients treated with the SpineCor brace showed significant improvements in clinical parameters. There was no significant difference in measurements between baseline and follow-up in control patients. Changes in Cobb angle over the course of the study were not reported.

Vertebral Body Stapling

A total of 5 publications on vertebral stapling for scoliosis have been identified; all are from Betz and colleagues. (9-13) The 2010 and 2011 publications reported on 29 patients with juvenile or adolescent idiopathic scoliosis who met the study inclusion criteria (out of a database of 93 patients). The reasons for excluding 69% of the patients from the database were not specifically described but included a change in the type of staple in 2002. Included in the report were patients with idiopathic scoliosis, a coronal curve magnitude of 20 to 45 degrees, Risser 0 or 1, staples with tines proportional to staple size (beginning in 2002), and a minimum 2-year follow-up. One patient from the series was lost to follow-up after 1 year, leaving 28 patients (96%) in the analysis. The average age at the time of stapling was 9.4 years (range, 4 to 13 years), with an average follow-up of 3.2 years (range 2 to 5.3 years). Only the thoracic curve was stapled in 13 patients, both thoracic and lumbar curves were stapled in 13 patients, and only the lumbar curve was stapled in 2 patients. For thoracic curves greater than 35 degrees at baseline, 75% progressed to greater than 50 degrees (threshold for recommending spinal fusion). The authors now use additional treatments such as growing rods or nighttime braces for curves that are greater than 35 degrees at baseline or that cannot be corrected to less than 20 degrees on first standing radiograph. For thoracic curves less than 35 degrees at baseline, 6% of patients progressed to greater than 50 degrees (threshold for surgery), 22% progressed between 5 and 50 degrees, and

78% had no change. Notably, 8 curves in 7 patients improved from baseline, and one curve in a 6-year-old (25 degrees) reversed direction, leading the authors to recommend waiting until the child is 8-years old or until the curve has exceeded 30 degrees in a younger child. Betz et al. concluded that the results should be considered preliminary, as follow-up to skeletal maturity will be needed for definitive results. Additional studies from other centers are also needed. The authors comment that their approach is almost always to recommend bracing first, and offer stapling only if the child/adolescent has a difficult time wearing the brace. (12)

Ongoing Clinical Trials

Higher quality evidence on bracing is expected from a National Institutes of Health (NIH) -sponsored multicenter randomized trial of bracing in patients with adolescent idiopathic scoliosis (BrAIST) versus watchful waiting (available online at: ClinicalTrials.gov; NCT00448448). Enrollment criteria include: skeletally immature (Risser grade 0 to 2); pre-menarchal or post-menarchal by no more than 1 year; primary angle between 20 and 40 degrees; curve apex caudal to T7; and no previous surgical or orthotic treatment for AIS. Participation in the study will last until a participant reaches skeletal maturity or his or her spinal curve progresses to 50 degrees, after which usual care will continue. The trial began in 2007 with an estimated enrollment of 500 patients; completion of the trial is expected in August 2014.

Summary

Bracing has been considered the only available option to prevent curve progression in juvenile or adolescent idiopathic scoliosis, although efficacy has not been consistently demonstrated when compared with watchful waiting. The inconclusive evidence may be due, in part, to lack of compliance in this population, as well as variability in inclusion criteria and definitions of success in case series. The quality of evidence on bracing is expected to be improved with a new randomized controlled trial on bracing versus watchful waiting. Based on the currently available evidence of efficacy in some patients, lack of alternative treatment options, professional society recommendations, and potential to prevent the need for a more invasive procedure, bracing may be considered medically necessary for the treatment of scoliosis in patients with high-risk of curve progression. Curves have a high-risk of progression when they measure 25 degrees or more and spinal growth has not been completed, or when a 20-degree- curve is progressively worsening and at least 2 years of growth remain.

Over the past decade, investigators have been evaluating use of memory shape staples for preventing curve progression in juvenile and adolescent scoliosis. The most recent paper on vertebral stapling suggests that for many patients with curves between 20 and 35 degrees, vertebral stapling may maintain or even improve the curve, thus reducing the rate of subsequent spinal fusion and providing a potential option for the treatment of idiopathic scoliosis. The evidence to date, which consists of only 5 publications with limited follow-up from a single center that developed the technique, is insufficient to permit conclusions concerning the effect of this procedure on health outcomes. Additional studies from other centers, with a larger number of total subjects and longer follow-up, are needed to evaluate the safety and efficacy of this surgical procedure. Therefore, vertebral body stapling for scoliosis is considered investigational.

Practice Guidelines and Position Statements

The Scoliosis Research Society (SRS) states that the treatment of adolescent idiopathic scoliosis falls into three main categories (observation, bracing, and surgery) and is based on the risk of curve progression. (14) In general, AIS curves progress in two ways: first, during the rapid growth period of the patient, and second, into adulthood if the curves are relatively large. Since scoliosis gets larger during rapid growth, the potential for growth is evaluated taking into consideration the patient's age, the status of whether females have had their first menstrual period, as well as radiographic parameters. The Risser grading system rates a child's skeletal maturity on a scale of 0 to 5. Patients who are Risser 0 and 1 are growing rapidly, while patients who are 4 and 5 have stopped growing.

- Observation is generally for patients whose curves are less than 25 degrees who are still growing, or for curves less than 50 degrees in patients who have completed their growth.
- Bracing is for patients with curves that measure between 25 and 40 degrees during their growth phase. The goal of the brace is to prevent the curve from getting bigger.

- Surgical treatment is used for patients whose curves are greater than 45 degrees while still growing or greater than 50 degrees when growth has stopped. The goal of surgical treatment is two-fold: First, to prevent curve progression and, secondly, to obtain some curve correction. Implants are used to correct the spine and hold the spine in the corrected position until the spine segments which have been operated on are fused as one bone.
- Alternative treatments to prevent curve progression or prevent further curve progression, such as chiropractic medicine, physical therapy, yoga, etc., have not demonstrated any scientific value in the treatment of scoliosis.

Information updated in 2010 from the American Academy of Orthopaedic Surgeons (AAOS) indicates that the type of treatment required for idiopathic scoliosis in children and adolescents depends on the kind and degree of the curve, the child's age, and the number of remaining growth years until the child reaches skeletal maturity. (15)

- Observation is appropriate when the curve is mild (less than 20 degrees) or if the child is near skeletal maturity.
- The goal of bracing is to prevent scoliotic curves from worsening. Bracing 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, most being the underarm type.
- Surgery may be recommended if the curve is more than 45 degrees and the child is still growing. If the patient has reached skeletal maturity, surgery may still be recommended for scoliotic curves that exceed 50 to 55 degrees. An implant made up of rods, hooks, screws, and/or wires is used to straighten the spine. Bone graft from the bone bank, or from the patient's hip region, is also used to help the operated portion of the spine heal solid.
- At present, the main research focus in idiopathic scoliosis is investigation into genetic factors as a cause of scoliosis.

The National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS) in 2012 indicates that many children who are sent to the doctor by a school scoliosis screening program have very mild spinal curves that do not need treatment. (16) When treatment is needed, an orthopedic spine specialist will suggest the best treatment for each patient based on the patient's age, how much more he or she is likely to grow, the degree and pattern of the curve, and the type of scoliosis.

- Observation may be advised if the patient is still growing (is skeletally immature) and the curve is mild.
- Doctors may advise patients to wear a brace to stop a curve from getting any worse in patients who are still growing with moderate spinal curvature. As a child nears the end of growth, the indications for bracing will depend on how the curve affects the child’s appearance, whether the curve is getting worse, and the size of the curve.
- Surgery may be advised to correct a curve or stop it from worsening when the patient is still growing, has a curve that is more than 45 degrees, and has a curve that is worsening.

NIAMS also stated that studies of the following treatments have not demonstrated prevention of curve progression or worsening:

- Chiropractic manipulation
- Electrical stimulation
- Dietary supplements
- Exercise

Medicare National Coverage
Not applicable

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Billing Coding/Physician Documentation Information

There is no specific CPT code for the insertion of vertebral body staples or vertical expandable titanium prosthetic ribs. The procedure would most likely be reported with the unlisted code 22899.

Additional Policy Key Words

N/A

Policy Implementation/Update Information

11/1/10	New policy; may be considered medically necessary.
11/1/11	No policy statement changes.
11/1/12	Material on VEPTR [vertical expandable prosthetic titanium rib] moved to policy 7.01.110
11/1/13	No policy statement changes.
