Surgeons target key challenges in treating adolescent idiopathic scoliosis

Despite years of research, investigators are still uncertain about the etiology of AIS. New genetic studies, however, may shed some light.

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The management of spinal disorders is one of the fastest growing areas of musculoskeleal care, and there has been a recent increase and interest in managing spinal diseases by surgical means. However, there are significant controversies surrounding surgery for the treatment of spinal disorders related to degenerative conditions.

An area that is somewhat less controversial but is also generating a lot of interest is the management of adolescent idiopathic scoliosis. The management of scoliosis by rigid instrumentation is barely 50 years old now, and even though Harrington rods are not used anymore, it is not unusual that an X-ray report will state, "The Harrington rods appear to be in place."

While the genetics of adolescent idiopathic scoliosis have not been revealed or understood completely, there is a tremendous amount of research taking place at centers throughout the country to identify the gene responsible. The advent of classifications designed to minimize the number of vertebral segments fused in the treatment of scoliosis will hopefully improve the long-term back health of adolescent children undergoing treatment for scoliosis.

The thought of being able to limit the number of vertebra stabilized to manage the disorder, the advances in anesthetic management and postoperative pain support, and new implants designed to allow children to undergo stabilization and be virtually brace-free in most cases are quite exciting and appealing.

Because the general orthopedist has rather limited exposure to this disorder, I have convened a virtual roundtable of spinal deformity surgeons from throughout the continental United States.

I presented a list of questions to apprise our readers of the broad scope of the current management of this disorder.

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Moderator

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Alvin H. Crawford, MD: Is there a level of genetic research or clinical research regarding the etiology of scoliosis that you can pass on to other doctors and patients?

Peter O. Newton, MD: Unfortunately, the etiology of adolescent idiopathic scoliosis (AIS) continues to elude us despite years of investigation. There are, however, several facts we know that are beginning to point us closer to the full understanding of the cause(s) of AIS. The genetic link is clear, and several candidate genes remain under investigation. We all have our favorite theory, and mine relates to the relative overgrowth of the anterior vertebral column compared to the posterior elements. This is nicely supported by alterations we see in the sagittal plane of patients with AIS; there is less thoracic kyphosis than normal, and recent work by Jack Cheng's group in Hong Kong showed the different mechanisms of growth between the vertebral bodies (endochondral) and the posterior elements (intramembranous).

It is not hard to imagine a problem in one of several genes that must modulate these different growth pathways being out of sync resulting in anterior column overgrowth with subsequent buckling (lateral curvature and rotation) described by Roaf decades ago.

Lawrence G. Lenke, MD: Although the definitive cause of idiopathic scoliosis is still unknown, there appears to be enough current research regarding genetic contributions emanating from twin studies, genetic-linkage studies, etc., that a genetic transference will soon be delineated. … It is a fairly common scenario to have a skeletally mature patient with a 40° typical right thoracic

idiopathic scoliosis curvature. However, as we are all aware, not all 40° curves "look" the same to the patient, the family or the scoliosis surgeon.

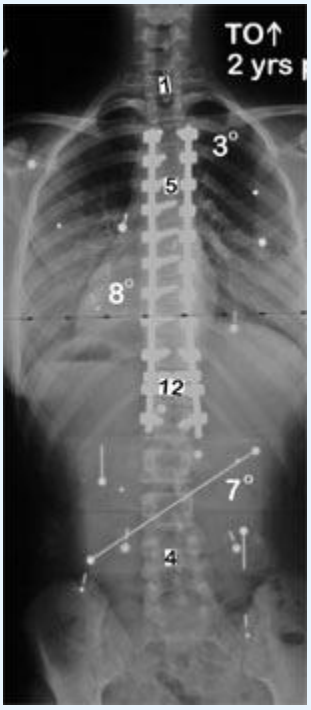
In addition, at this point, it may be difficult to know what the long-term prognosis will be. Many 40° curves seem to stay stagnant, causing little-to-no clinical symptomatology, and should be left alone. However, there certainly are 40° overhang-type thoracic curves that will slowly progress one to two degrees per year and ultimately lead to further deformity, possible pain relating to the spine or chest wall deformation, and ultimately require spinal fusion for curve correction and maintenance of a stable spinal column.

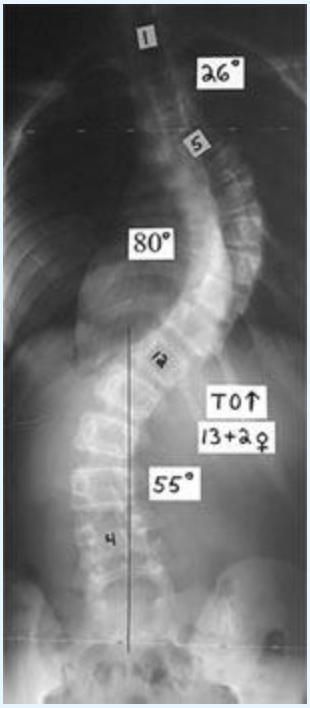
Crawford: What should the doctor tell the parents of a 16-year-old with a 40° right dorsal curve?

Oheneba Boachie-Adjei, MD: Depending on whether it is a boy or girl patient, the recommendation will vary. For a girl patient, it is likely that she has passed her peak growth period, may have had menses and is skeletally mature, as is the case in most girls of this age. For a curve of 40° in the thoracic spine, previous studies have shown that progression into adulthood is minimal without any deterious effect on general function, respiratory function, child bearing or work status. Such a girl can be safely observed into adulthood.

On the other hand, a boy who is 16 years of age with a 40° thoracic curve may still be growing and may benefit from a period of brace application until all signs show cessation of growth.

Daniel J. Sucato, MD, MS: I think the question we need to ask is, "What is the long-term effect of a 40° curve on this patient's overall health?" If a 16-year-old has demonstrated that the growth of his or her spine has stopped — with a thoracic curve of 40° — then the likelihood of progression of the curve is very small and there is no need for surgical treatment from a purely medical standpoint.





At left is a 13-year-old female with an upper thoracic curve of 26°, a thoracic curve of 80°, and a lumbar curve of 55°. She underwent a posterior spinal fusion from T3 to L1. Her two-year postoperative AP shows corrected curves to 3°, 8°, and 7°.

Images: Lenke LG

However, the patient needs to be comfortable with his or her cosmetic appearance, which should be fairly mild for a typical 40° thoracic curve. I inform the patient and family that mild back pain may be more common for them in the future when compared to someone without scoliosis. However, I do not typically provide a curve magnitude at which symptoms will begin to occur, since many patients have variants of body habitus, physical activity, levels, etc., that impact axial back pain from a right thoracic curve.

I also tell families that it is important that adolescents maintain a good regular physical fitness program into their adult life to limit the discomfort that they may feel into the future. This certainly is advice I would give any of our adolescent patients regardless of whether or not they have scoliosis.

Crawford: Should the surgeon measure the X-rays and follow the patient using the Cobb angle system and, if so, does he need to be familiar with a classification system like the Lenke or King classification system?

Randal R. Betz, MD: At a bare minimum, a practicing general orthopedist should measure the worst Cobb angle of all the curves on the X-ray. In addition, an initial full-length lateral X-ray is mandatory to rule out an associated kyphosis (28% of kyphoses present as scoliosis). On the

lateral X-ray, a measurement of 33° or greater from T5 to T11 (Cobb angle) should dictate a further evaluation for kyphosis. This measurement of 33° has been shown to be 100% sensitive but is not necessarily specific.

In addition, a radiograph should be taken every three to four months in a girl who is premenarche and every six months up to skeletal maturity. Monitoring the rib prominence and lumbar rotation via a scoliometer is prudent in order to pick up any increase in cosmetic deformity.

A general practicing orthopedist should make a habit of dictating the exact levels that he or she measures into his or her notes so that sequential x-rays can be taken of the same measured vertebrae. In my experience with referrals, the No. 1 reason for referral for progression has been failure to measure the same landmarks on sequential x-rays.

Lenke: I believe that all orthopedic surgeons should be familiar with measuring the Cobb angle on scoliosis radiographs, which is typically taught during the surgeons' residency training. Obviously, the measurement error for these surgeons will probably be greater than that of scoliosis practitioners; however, it will at least provide a basis for continued follow-up and referral to a scoliosis specialist if a progressive curvature is noted and documented.

I do not believe that general orthopedic surgeons need to memorize the Lenke or King classification systems, but should be familiar with typical idiopathic curve patterns so as to obtain appropriate follow-up and/or MRI evaluations of "atypical" curves such as left thoracic, sharply angulated curves, and those with rapid progression not associated with a documented growth spurt.

Crawford: Is there a clear-cut indication for bracing, and if so, what is it in your hands? What are your criteria?

Newton: I continue to use the Scoliosis Research Society guidelines for bracing (ie, >30° at presentation, >25° with 5° or more of progression, and only for those patients who are still growing).

There seem to be two groups of physicians when it comes to prescribing an orthotic for scoliosis: believers and nonbelievers, and similarly two types of patients when it comes to wearing an orthosis: those who are willing and those who are unwilling. I would put myself in the "believers" group, but I find surprisingly few patients in the "willing to wear" category.

As such, I have many patients who go through the motions with various levels of commitment and success. Every now and then, a dedicated brace wearer reinforces my belief in bracing. Unfortunately, these patients are a rare find. There is a good reason for this: full-time brace wear is extremely difficult to comply with.

Sucato: Bracing is one of the more controversial subjects in AIS. Certainly, there are schools of thought that suggest that there are no randomized control studies that scientifically prove that bracing is effective in preventing curve progression. However, there are very good studies to

support the finding that bracing has at least some beneficial effect on the natural history of AIS in the growing patient.

At our hospital, we have an outstanding orthotics department, led by Don Katz, who has initiated many important studies on bracing, and I have certainly adopted the philosophy that bracing statistically improves the likelihood of preventing curve progression in the skeletally immature patient (Risser 0, 1). It is the only nonoperative treatment method that I know of that has some effectiveness to prevent curve progression.

I typically prescribe a brace when curves I have been following are 25° or greater in a patient who is skeletally immature, and I will brace curves up to 45° when the patient is satisfied with his or her cosmetic appearance and does not want surgery if the curve magnitude is maintained.

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— Oheneba Boachie-Adjei, MD

I think the family and the patient have to understand that bracing is a big investment on everyone's part, and I generally prescribe the brace for 23 hours a day when using the TLSO (can be used in all curve types), and nighttime or Charleston bending bracing, which I use for thoracolumbar/lumbar curves measuring between 25°-35°.

Betz: The problem with bracing is that it does not correct the curve; successful bracing only holds the existing deformity, and waiting for documented progression by X-ray may allow the deformity to worsen, and it will not be correctable with a brace applied six months later.

The [other] problem is that there is really no good Level I evidence to demonstrate if a brace is going to stop a progressive curve. Without some form of genetic or biochemical marker, we don't know which curves are going to progress; therefore, we are obliged to watch for progression. The biggest dilemma with the progression that occurs is, again, an increase in cosmetic deformity that cannot be reversed with the brace. We are really left utilizing a tool for treatment that has no good evidence to know when it works and when it doesn't.

Crawford: What are the indications for surgery, and is there a length of follow-up required showing progression before surgery, or do you draw a line in the sand based on curve angle and skeletal age?

Boachie-Adjei: Surgery should be the last resort for the treatment of AIS. Any adolescent who has properly worn a brace and has progressed beyond 50° and still has growth remaining should be offered a surgical solution. An AIS patient seen for the first time with a curve exceeding 50° should also be counseled regarding surgery.

There is no emergency in performing scoliosis surgery, especially in an adolescent. Therefore, it must be the patient's choice to undergo an elective surgical procedure. The skeletal age (ie, Risser), curve magnitude >50° and menarche status in girls are all criteria to apply in advising surgery for an adolescent patient.

Newton: Defining the indications for surgery is why we have fellowships in scoliosis treatment. The simple rule — "greater than 40° in the immature patients and greater than 50° in mature patients" — is a reasonable starting point for discussion, but the nuances go far beyond these indications.

As we discussed earlier, there is so much more than just the Cobb angle in determining the natural history, and it is the natural history of the untreated condition that must be compared to the expected outcome over time of the surgically treated condition in determining when or not to operate.

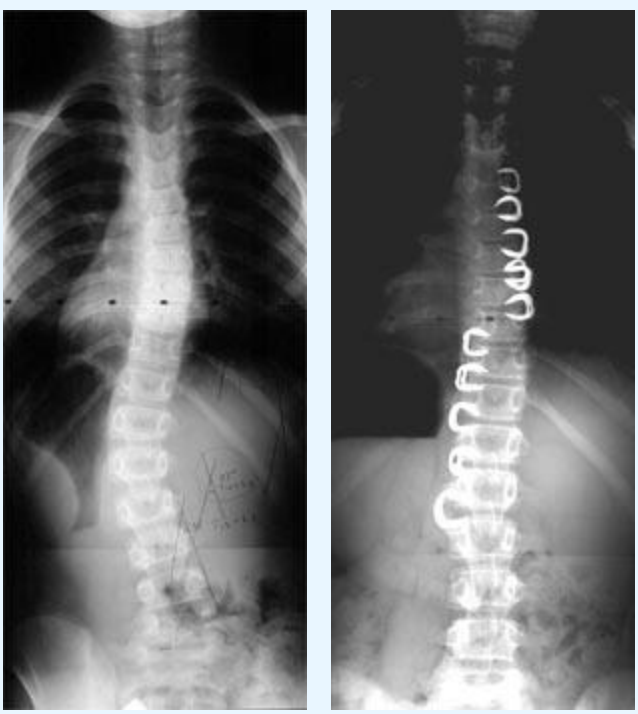
Just as we are lacking data on the untreated condition, we have even less data on those patients who have undergone surgical correction. The Harrington instrumentation was introduced only 40 years ago and the Cotrel-Dubousset method was introduced just 20 years ago.

Given that all of our current methods of scoliosis correction sacrifice motion, there are very likely different long-term "costs" of fusion depending on the levels of fusion, and therefore the indications for fusing should be varied based on those "costs." Although I suspect we all attempt to make such cost benefit analysis as we chose whether or not to recommend surgical correction of each individual patient, we are doing so with very little objective data.

Preventing long-term progression remains the primary indication for surgery; however, the cosmetic indication is another that we frequently dance around. There is little doubt in my mind this is a real factor in decision making for some patients more than others, and one that does deserve consideration.

Crawford: What surgical techniques do you apply (ie, endoscopic, vertebral growth modulators, staples, etc.) and for which indications? What are the current recommended implants? Finally, when are fusion enhancements indicated (ie, When are bone morphogenetic proteins, platelet-derived growth factors and allografts considered viable substitutes for autograft?)"

Lenke: My preferred surgical technique for a 45° to 60° adolescent idiopathic scoliosis curve in a skeletally maturing or mature female is a posterior preferred approach for all curve patterns, except for thoracolumbar and lumbar curves, which we still treat primarily via a short segment anterior dual screw/dual rod instrumentation technique.



At left is an AP photograph of an 8-year-old girl with a progressive idiopathic curve measuring 25° in the thoracic spine and 35° in the lumbar spine. The family chose stapling because the child refused to wear a brace. Follow-up at 2½ years (right) shows correction of the curve to 5° thoracic and 7° lumbar through growth modulation.

Images: Randal R. Betz

We aspire to avoid touching the chest wall in thoracic curves with any type of anterior endoscopic or open release or posterior thoracoplasty procedure. Our goal is to maximize pulmonary function by avoiding any type of chest wall disruption. Pulmonary function decline with aging is very similar to bone density decline with aging; thus, I think it behooves us to maximize children's pulmonary function with scoliosis during surgical treatment, if possible, to ultimately provide them with the best lung function during their later years of life.

I prefer segmental pedicle screw fixation for several reasons, including the fact that it offers maximum secure fixation and a quick return to postoperative activities (most patients are not restricted from any type of early postoperative activities that they are comfortable when performing). It also offers the ability to segmentally derotate thoracic curves to the improve rib prominence, thus avoiding thoracoplasty procedures, which are not performed anymore at our institution for typical scoliosis cases.

I feel that with secure segmental fixation and thorough posterior element decortication, local bone harvested from the base of the spinous and transverse processes and inferior facets supplemented with fresh frozen, non-irradiated allograft bone, if necessary, is adequate in most idiopathic patients. Postoperatively, patients may return to noncontact sports (swimming, soccer,

baseball, golf, etc.) assuming secure segmental pedicle screw fixation is obtained intraoperatively.

Boachie-Adjei: For a patient (Risser 2 classification) with 45°-60° scoliosis of the thoracic spine, most current surgical techniques will provide optimal curve correction and functional outcome. Posterior only approaches with third-generation segmental instrumentation and local bone and allograft without removing bone from the rib or iliac crest will achieve a very successful fusion and clinical outcome. A thoracoscopic approach has a cosmetic appeal at the expense of pulmonary function reduction.

Growth modulation have little role in this case. Since the fusion rate is very high in such patients, fusions using BMP and PDGF are expensive propositions, which add very little to overall successes.

Sucato: Generally, when performing posterior spinal fusion, I use segmental fixation and pedicle screws I have moved toward allograft bone to avoid the morbidity of harvesting autologous iliac crest. Excellent decortication of the spine is more easily performed with newer low-profile implants.

For thoracoscopic anterior spinal fusion and instrumentation, I continue to utilize autologous bone harvested from the iliac crest to provide the best chance for fusion. We have investigated BMP-2 in animal model for thoracic scoliosis fusion using thoracoscopic techniques and demonstrated outstanding results and have preliminary data on some patients using BMP-2 in this clinical situation.

The use of bone graft substitutes will significantly improve the applicability of the thoracoscopic approach in the treatment of AIS in the future.

Betz: Currently, there is an option of vertebral body stapling for curves that are in the bracing or low fusion range. Vertebral body stapling has been shown to be effective with preliminary early results in curves from 20 to 45°.

One significant advantage of stapling over bracing would be the opportunity for curve correction, which does not occur with bracing. In the 80% of patients in which curves have stabilized with stapling, more than 50% of those patients show significant curve improvement (ie, greater than 10°). With follow-up now over five years, we have shown that the procedure is safe and there are no long-term problems or complications apparent at this mid-range follow-up.

Vertebral body stapling has been shown not to be effective in curves greater than 45 or 50°; however, the newest technology on the horizon is that of vertebral body tethering on the convex side of the curve. This involves placing a vertebral body screw in each vertebral body of the curve and tethering them together with flexible, dynamic cable.

For the right thoracic idiopathic curve under 70°, most cases are amenable to a posterior spinal fusion with instrumentation or an anterior thoracoscopic instrumented procedure. While the open anterior instrumented fusion has been shown to be very effective in correcting curves and

treating it in a selective fashion (not fusing to the lumbar spine), pulmonary function declines significantly (it takes two years to recover and may never recover to baseline). While pulmonary function does decline briefly following thoracoscopic anterior instrumentation, it returns more rapidly back to baseline or better.

The use of thoracic pedicle screws has been a huge advance over the last several years. With its ability to control the coronal and sagittal planes, the need for thoracoplasty to correct the cosmetic component of the deformity has dramatically decreased.

Newton: In general, posterior implants can be universally applied for all curve patterns with outstanding results. I have used pedicle screws for all levels of the thoracic and lumbar spine because of improved fixation and transverse plane correction compared to hook-and-wire constructs. For the patient you describe, many combinations of posterior implants can achieve the desired goals, but the use of pedicle screws throughout is my current standard.

There is a subset of patients with moderate right thoracic curves that are candidates for thoracoscopic instrumentation, and this remains a viable minimally invasive option for those unwilling to live with a long posterior scar. This is one of the few procedures, however, that I still harvest a posterior iliac crest bone graft. I also brace these patients for three months postoperatively. These two factors lead some who are thoracoscopic instrumentation candidates to choose posterior instrumentation.

For typical AIS posterior instrumentation, my choice of bone graft, in addition to the local bone (excised facets, spinous processes) is freeze-dried crushed cancellous allograft with 10 cc of demineralized bone powder. One of the advantages of a pedicle screw construct is the availability of all of the lamina for decortication (with a Capner gouge) providing a large continuous surface for fusion.

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