SEPTEMBER 2011
VOL. 11, ISSUE 5

IU Health Physicians

ADVANCED praxis

A JOURNAL OF CURRENT TRENDS IN MEDICINE FROM IU HEALTH PHYSICIANS, A PARTNERSHIP OF IU SCHOOL OF MEDICINE AND INDIANA UNIVERSITY HEALTH

CASE MANAGEMENT
Progressive Infantile Scoliosis

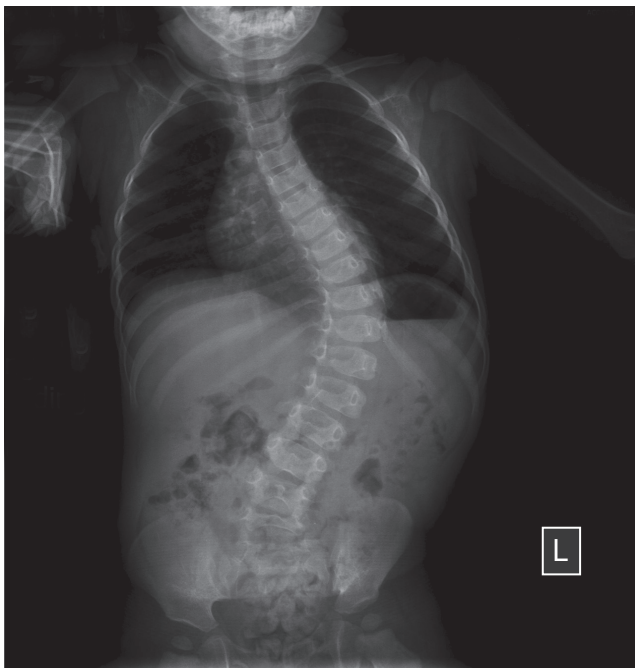

Figure 1. Patient at presentation (age 19 months)
Curvature of the thoracic spine, shown on an AP radiograph, is >60 degrees.

A 19-month old male with a spinal deformity detected by his parents and confirmed by his pediatrician is referred to Shyam Kishan, MD for evaluation. Clinical presentation suggests a syndromic disorder; a standing anteroposterior (AP) radiograph demonstrates a thoracic curve of more than 60 degrees *(Figure 1)*. MRI fails to reveal an underlying spinal cord abnormality. The child is diagnosed with progressive infantile scoliosis associated with an unknown syndrome.

What is IU Health Physicians?

IU Health Physicians brings together Indiana University School of Medicine faculty physicians, IU Health-affiliated physicians and private practice physicians to form the fastest-growing, most complete, multi-specialty practice group in Indiana. This unique partnership gives our highly skilled doctors access to innovative treatments using the latest research and technology.

Our goal is to provide seamless patient care through an integrated and coordinated delivery health system. It will also improve access for patients and referring physicians, provide an excellent environment for world-class scientific research, and further our academic commitment to tomorrow's healthcare professionals.

Infantile scoliosis is traditionally defined as a lateral curvature of the spine with concordant vertebral rotation diagnosed within the first three years of life. Eighty percent of cases are idiopathic, and the remainder are associated with known and unknown syndromes, congenital abnormalities, and neurologic disorders. Male infants are affected more frequently than females, and most curves are left-sided, occurring in the thoracic area, although double curves involving both thoracic and lumbar vertebrae are also seen.

Ninety percent of children with infantile scoliosis experience spontaneous resolution of the spinal curvature. The other 10 percent of cases, primarily youngsters with a rib-vertebral angle difference greater than 20 degrees (measured on a supine AP radiograph),[1] progress to a severe, disfiguring, and disabling condition by the age of two or three years and experience relentless progression of scoliosis until skeletal maturity.

"Left untreated, progressive infantile scoliosis is one of the few potentially fatal noncancerous orthopaedic disorders," reports Dr. Shyam Kishan, associate professor of pediatric orthopaedic surgery at the Indiana University School of Medicine. "Mortality is linked to a type of pulmonary failure, termed thoracic insufficiency syndrome, that results from poor lung growth and development caused by the restricted volume of the hemithorax and the restricted motion of the involved ribs."[2,3]

Treatment Options

Spinal Fusion

Definitive posterior spinal fusion is an effective treatment option for teenagers with scoliosis. However, its use in children younger than 10 years of age exacerbates the spinal deformity by restricting growth of the posterior spine while allowing the anterior spine to continue growing. The end result can be severe respiratory compromise and a short trunk.

> "The end result can be severe respiratory compromise and a short trunk."

"The formula for determining the impact of spinal fusion on trunk growth is the number of vertebrae fused times the number of years of growth remaining times 0.8 mm," Dr. Kishan explains. "Fusing 10 vertebrae in a six-year-old boy, who normally has approximately 10 years of growth remaining, will decrease final height by about eight centimeters."

Growing Rods

Expandable surgical instrumentation systems have been developed in an attempt to restore spinal alignment while permitting spine and chest growth.[4] Single or dual growing rods are inserted under the skin and attached to the spine above and below the curve with hooks or screws (used off-label in children). The child returns about every six to nine months for an outpatient procedure to have the rods lengthened by approximately one centimeter to keep pace with growth.

> "Moreover, once a family commits to this path, there is no turning back, as nonsurgical strategies are no longer an option."

"Growing rods can control progressive infantile scoliosis and delay definitive fusion, but they rarely cure the spinal curvature," Dr. Kishan notes. "The insertion procedure is challenging to perform and is associated with a moderate to high rate of complications that include rod fracture and loosening, hook displacement, and infection. Moreover, once a family commits to this path, there is no turning back, as nonsurgical strategies are no longer an option."

Bracing

Bracing is another technique that is primarily used to slow the inevitable progression of scoliosis and allow the child to attain sufficient growth for good pulmonary function before definitive surgery is performed. A variety of braces are available, and new ones must be fabricated every 12 to 18 months to accommodate growth. Braces are usually intended to be worn full-time and removed only for bathing.

> "Braces are usually intended to be worn full-time and removed only for bathing."

Casting

Serial casting for the treatment of progressive infantile scoliosis was first described more than 50 years ago. However, two recent prospective studies conducted in the United Kingdom[5] and the United States[2] (in which Dr. Kishan participated) suggest that casting is poised to become the future of progressive infantile scoliosis management. These studies, which collectively enrolled more than 200 children, showed that derotational cast correction cures some young children. In older and/or syndromic patients, casting was found to reduce the curve size, improve chest and body shape, and delay surgery.

"Casting has fewer and less serious potential complications….Another important feature of casting is that is does not burn any bridges."

"Casting harnesses the rapid growth of early childhood to straighten the spine through the continuous application of an external force," Dr. Kishan explains. "During the first year of life, the mean growth rate is almost twice that occurring during the adolescent growth spurt—approximately 20 to 25 cm versus 12 to 14 cm. Growth in the second year of life matches the peak at adolescence."

Compared with growing rods, casting has fewer and less serious potential complications that include skin irritation and breakdown (more common in humid climates) and patient intolerance of the cast. Appropriate patient selection together with correct casting techniques can avoid rib deformities and difficulties eating and sitting.

"Another important feature of casting is that it does not burn any bridges," says Dr. Kishan. "If scoliosis worsens during casting, the cast can be removed, and growing rods placed or spinal fusion performed."

The parents elect to proceed with casting, and the first cast is placed when the child is 22-months of age *(Figure 2)*.

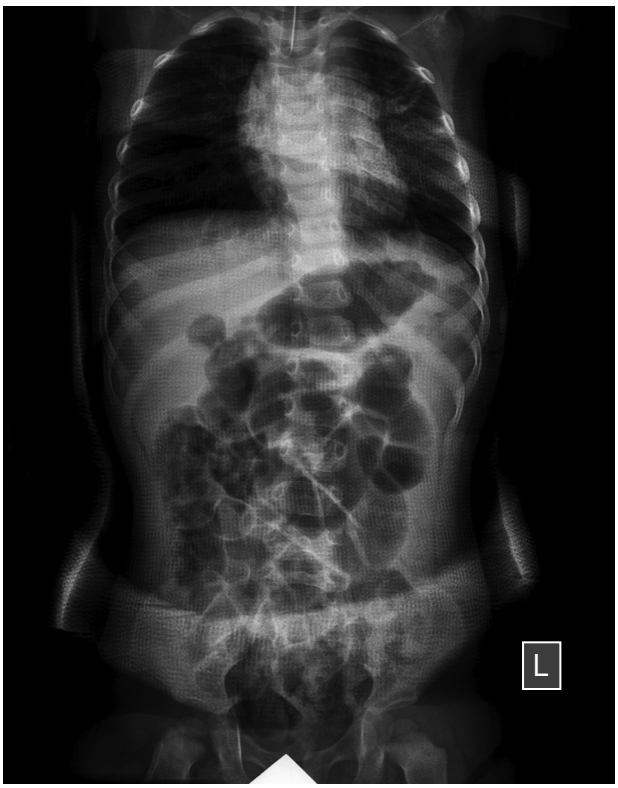

Figure 2. Patient after first casting (age 22 months)
The supine AP radiograph shows significant correction of the curvature and some derotation, as evidenced by the position of the pedicles.

Casting Technique

Casting is performed in the operating room under general anesthesia. The procedure takes approximately one hour and is performed on an outpatient basis. A special table is used to support the child in traction while leaving the body free for cast application. Head halter and pelvic traction provide stabilization and narrow the body *(Figure 3)*.

"Plaster or fiberglass are used to construct the cast," Dr. Kishan explains. "Posteriorly rotated ribs are rotated anteriorly to create a more normal chest configuration, with counter rotation applied through the pelvic mold and upper torso. An anterior window is created to relieve chest and abdominal pressure while preventing the lower ribs from rotating *(Figure 4)*. A back window is cut to allow the depressed concave ribs and spine to move posteriorly."

Normal activity is encouraged during casting, and Dr. Kishan reports that casted infants learn to crawl, walk, and even climb stairs without apparent difficulty.

Serial Casting and Bracing

Casts are generally changed every three to four months. The improved shape of the spine is assessed clinically and from AP radiographs obtained before and after each cast change.

"The cast is worn for about 12 to 18 months, or until supine X-rays without the cast show a curvature of less than 10 degrees," says Dr. Kishan. "At this point, the patient is fitted with an underarm brace that is worn for another year. Good compliance with brace-wearing is critical to avoid relapse and allow normal growth to resolve any residual curve. After the brace is discarded, patients are followed with yearly X-rays unless the parents report changes in spine shape."

Children who require casting for extended time periods may be given cast holidays. Although this practice allows them to enjoy activities such as swimming, Dr. Kishan cautions that there is a risk that scoliosis will progress during the holiday.

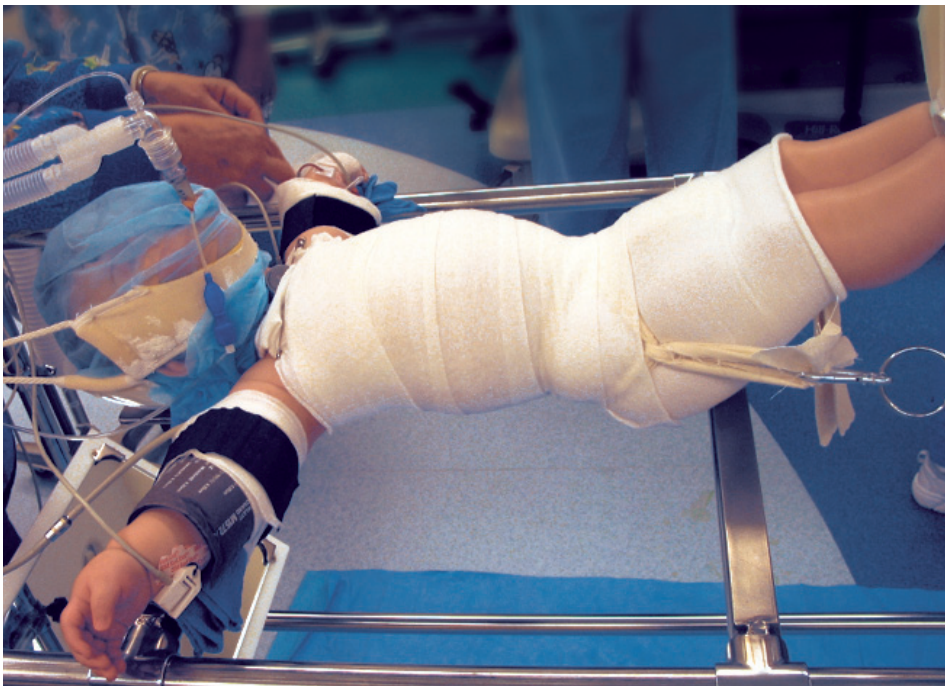

Figure 3. Casting technique
The patient is positioned on the table, with traction applied to the head halter and pelvis.

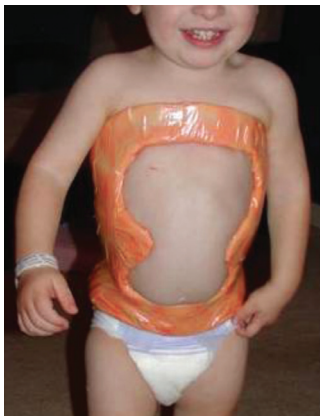

Figure 4. Patient in a cast
The anterior window relieves cast pressure on the chest and abdomen while preventing rotation of the lower ribs.

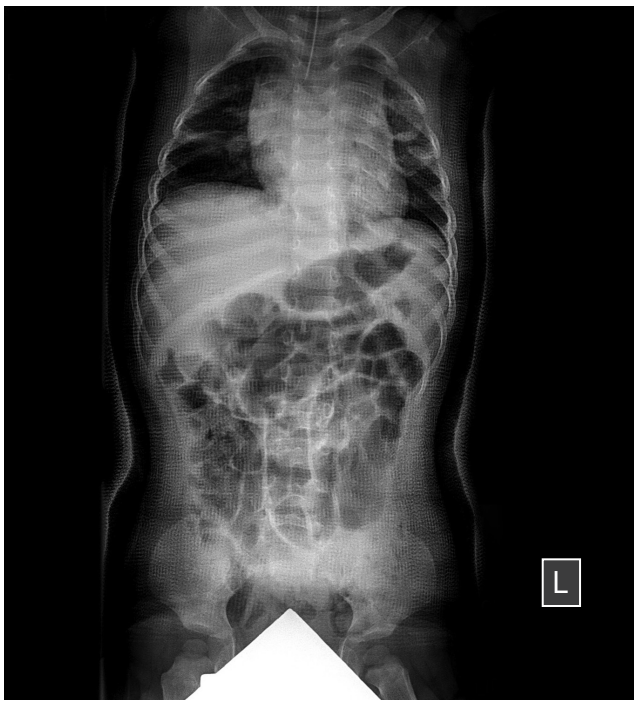

Figure 5. Patient at second casting (age 25 months)
The supine AP radiograph shows further improvement in both curve correction and spine derotation.

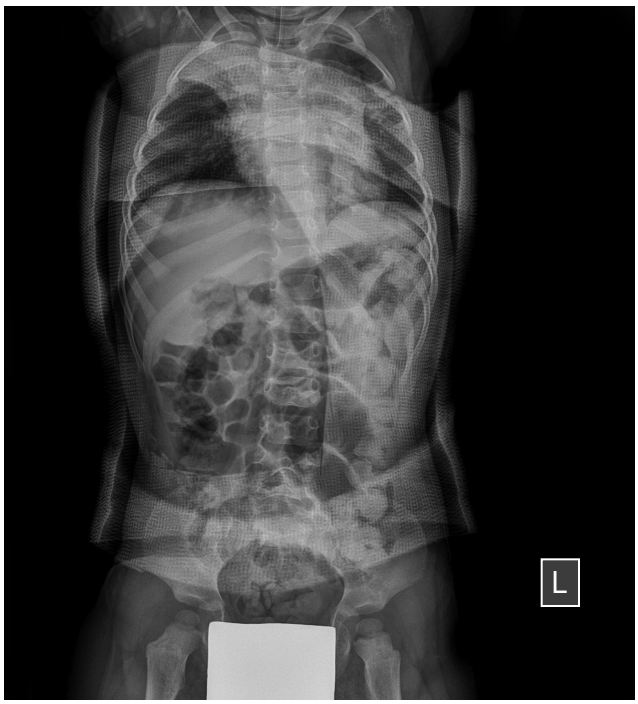

Figure 6. Patient at third casting (age 27 months)
When the patient is in a cast, the curve is almost fully corrected, and the focus shifts to spine derotation. Large windows cut in the cast allow continuing correction as the child grows.

The child receives another cast at age 25 months and again at age 27 months *(Figures 5 and 6)*. The casts are very well tolerated and do not limit activity. After the third cast is removed at age 30 months—eight months after initiation of serial casting—the curve has decreased to less than 10 degrees *(Figure 7)*. He is fitted with a custom-molded underarm thoracolumbar sacral orthosis (removable body jacket) brace that he is to wear except when sleeping for one year.

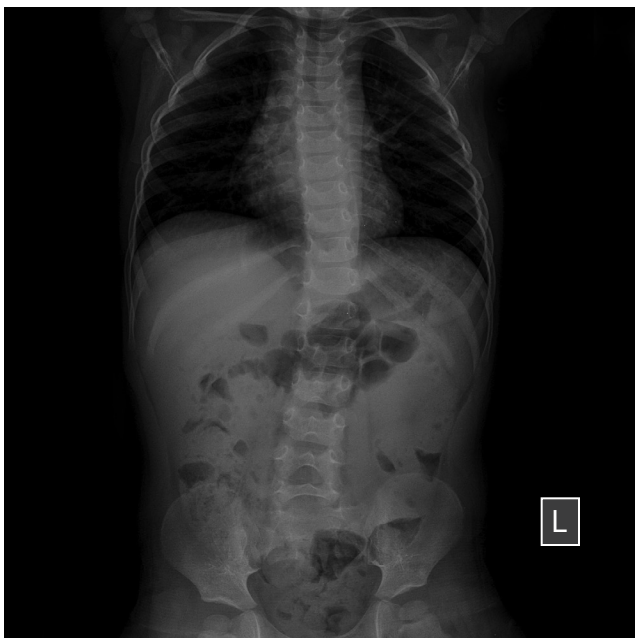

Figure 7. Patient after removal of final cast (age 30 months)
The supine AP radiograph shows that thoracic curve has decreased to <10 degrees, allowing the cast to be removed and a brace substituted.

A standing AP radiograph taken at the time of final brace removal at almost four years of age shows full correction of scoliosis *(Figure 8.)*

Prognosis

The best results for serial casting are obtained in children under two years of age who have good muscle tone and idiopathic thoracic curves measuring less than 60 degrees.[2,5] Children with known and unknown syndromes have the poorest outcomes.

"The younger the child at the initiation of casting, the better the results, underscoring the importance of early diagnosis and treatment," Dr. Kishan emphasizes. "Children under one year of age typically have a very good prognosis, whereas outcomes are not as good for those older than five years. Moreover, a delay of even a few weeks between detection and treatment is sufficient for some syndromic curves to progress to a point where reversal is no longer possible.

"Anecdotal experience indicates that casting improves or cures scoliosis in 25 percent of patients, and that another 25 percent deteriorate and require surgical treatment," he continues. "The remaining 50 percent of patients are in various stages of treatment."

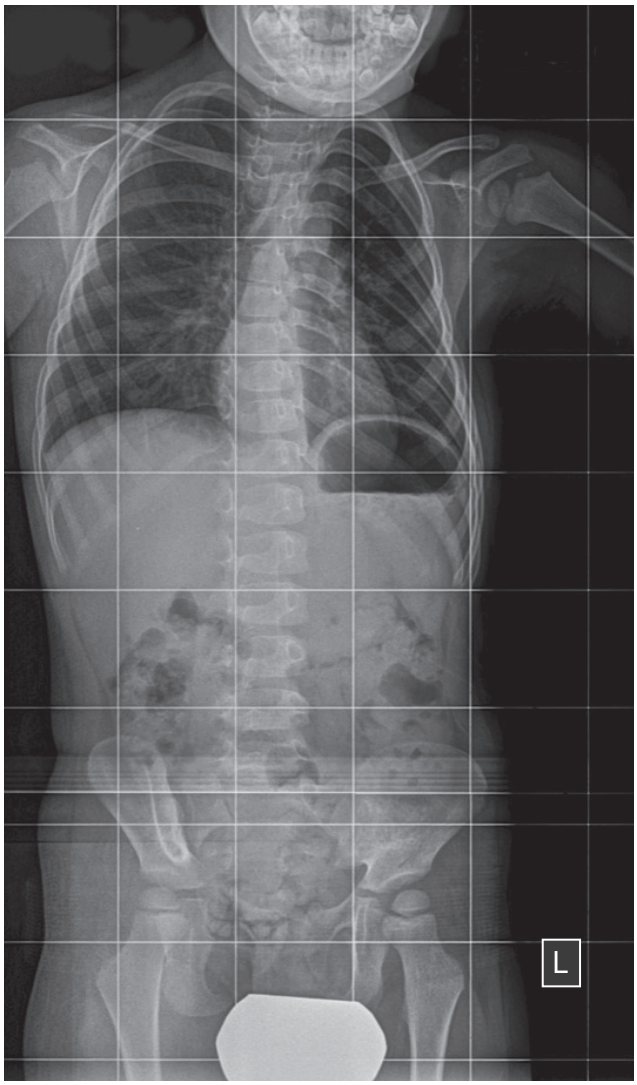

Figure 8. Patient at brace removal (age ~4 years)
A standing AP radiograph shows complete and sustained correction of the thoracic curve.

"Anecdotal experience indicates that casting improves or cures scoliosis in 25 percent of patients, and that another 25 percent deteriorate and require surgical treatment. The remaining 50 percent of patients are in various stages of treatment."

After three years of follow-up, the child continues to show full correction of scoliosis despite having two negative prognostic factors: an unknown syndrome and a curve exceeding 60 degrees. Long-term follow-up is needed to determine if the curve will recur, but available data suggests that casting achieves a permanent correction.[5]

Shyam Kishan, MD

Associate Professor, Department of Orthopaedic Surgery
Indiana University School of Medicine, Riley Hospital for Children

skishan@iupui.edu

Dr. Kishan received his medical degree from the Jawaharlal Institute of Postgraduate Medical Education and Research, Madras University, India and completed a residency and fellowship in pediatric orthopaedics and scoliosis at San Diego (CA) Children's Hospital. Prior to his appointment at Riley Hospital for Children, Dr. Kishan was an assistant professor of orthopaedic surgery at Loma Linda (CA) University Medical Center, where he established a center for early-onset scoliosis casting. His clinical interests, in addition to scoliosis, include pediatric limb deformity, complex hip disorders, and trauma. Growth plate cartilage modulation and spinal cord perfusion are among his research interests.

Dr. Kishan is certified by the American Board of Orthopaedic Surgery and is a fellow of the American Academy of Orthopaedic Surgeons and the Pediatric Orthopaedic Society of North America. He is the author of numerous journal articles and textbook chapters and has lectured in the United States and abroad.

References

1. Mehta MH. The rib-vertebra angle in the early diagnosis between resolving and progressive infantile scoliosis. J Bone Joint Surg Br 1972;54:230-43.
2. Sanders JO, D'Astous J, Fitzgerald M, Khoury JG, Kishan S, Sturm PF. Derotational casting for progressive infantile scoliosis. J Pediatr Orthop 2009;29:581-7.
3. Campbell RM, Jr., Smith MD, Mayes TC, et al. The characteristics of thoracic insufficiency syndrome associated with fused ribs and congenital scoliosis. J Bone Joint Surg Am 2003;85-A:399-408.
4. Thompson GH, Akbarnia BA, Campbell RM, Jr. Growing rod techniques in early-onset scoliosis. J Pediatr Orthop 2007;27:354-61.
5. Mehta MH. Growth as a corrective force in the early treatment of progressive infantile scoliosis. J Bone Joint Surg Br 2005;87:1237-47.