

Treating Scoliosis in Neuromuscular Conditions

by John E. Lonstein, M.D.

Idiopathic scoliosis is the most common type of scoliosis (a lateral spinal curvature). It affects about 4 percent of the population (80 – 85 percent of people with scoliosis). The etiology of idiopathic scoliosis is as yet unknown. This type of curvature more commonly needs treatment in girls and often runs in families. It often is discovered in adolescence by screening programs and routine annual screenings by pediatricians or family practitioners.

Treatment for idiopathic scoliosis is bracing for curves between 20 and 45 degrees. Beyond 45 degrees, braces are ineffective, and correcting the curve requires surgery.

The second most common type of scoliosis treated is neuromuscular scoliosis, the topic of this article. Children with neuromuscular scoliosis have multisystem involvement due to the underlying neuromuscular disease, and the age of onset is much younger than in idiopathic scoliosis.

Types of Neuromuscular Scoliosis

Neuromuscular scoliosis is classified into two causes: neuropathic (involving the nerves) and myopathic (involving the muscles). Neuropathic diseases can involve the upper- or lower-motor neuron. Upper-motor neuron diseases include cerebral palsy, spinal-cord trauma and tumors, and spinal muscular atrophy. Lower-motor lesions include polio. Patients with spina bifida often have a combination of upper- and lower-motor neuron lesions.

Myopathic lesions include arthrogryposis (joint contractures) and muscular dystrophy. Despite the differences, the common factor in these conditions is an inability to provide muscular support to the spinal column (as seen in spina bifida or muscular dystrophy) or an imbalance of the muscular control of the spine (as seen in cerebral palsy).

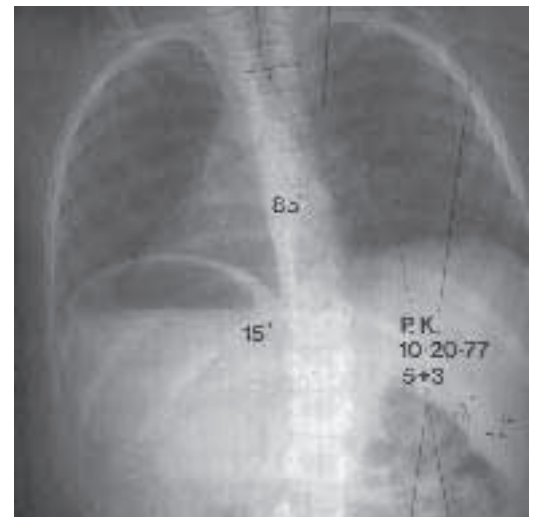
At-Risk Population for Neuromuscular Scoliosis

In general, children with neuromuscular diseases are either: *spastic* (very tight muscles), with poor brain control of the muscle, or *flaccid* (weak muscle control), such as in a flaccid child with a spinal cord injury.

The prevalence of scoliosis in neuromuscular conditions varies. Children who are paralyzed before age 10 have a 100-percent chance of developing neuromuscular scoliosis, as do children who have spina bifida with a high level of paralysis. Patients with spastic quadriplegia (cerebral palsy) have a 60- to 70-percent chance of developing neuromuscular scoliosis, whereas a child with hemiplegic cerebral palsy has only a 5-percent chance of developing scoliosis.

The three illustrations in this article show the progression of neuromuscular scoliosis from initial diagnosis through surgical fusion.

Pictured here is the spine of a 5-year-old with spastic cerebral palsy. The child has a 15-degree curve and was placed in a sitting support.



Age of onset obviously varies with etiology. For example, spinal muscular atrophy produces deformity at a younger age than does muscular dystrophy.

An additional difference in neuromuscular scoliosis is that, in these children, scoliosis doesn't appear alone — other systems are involved. The children can have skin breakdown due to insensate skin, hip and knee contractures, mental retardation, seizures, vision and hearing loss, and pulmonary problems.

Presenting Symptoms

The diagnosis of a neuromuscular condition is made when a child is quite young. The scoliosis is detected on a routine

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checkup of the child, or the parent or caretaker notes the curve because of a change in body position or change in ability to sit.

Evaluating Neuromuscular Scoliosis

Once the curve is present, gravity and a child's posture provide the impetus for worsening deformity. As the curve progresses, it changes the way the child sits, often leading to frequent wheelchair (if one is used) modifications to keep the person well-supported and to prevent pressure sores and other problems. Although a properly fitted chair can help support the spine, it cannot stop the progression of the curve.

A progressive or large curve can affect a child's pulmonary function by leading to collapse of the torso and elevation of the diaphragm, which reduce pulmonary space. This can manifest itself in recurrent pneumonias. This effect is more important when there is already pulmonary compromise due to intercostal paralysis (as seen in muscular dystrophy or high-level paralysis).

In addition, scoliosis affects a child's ability to function. The curve increase is accompanied by pelvic obliquity. Because of changes in the child's sitting position, repeated repositioning is necessary to maintain a good sitting posture. In addition, a child with some sitting balance will use one or both hands for support, which reduces hand function — probably making a child functionally quadriplegic.

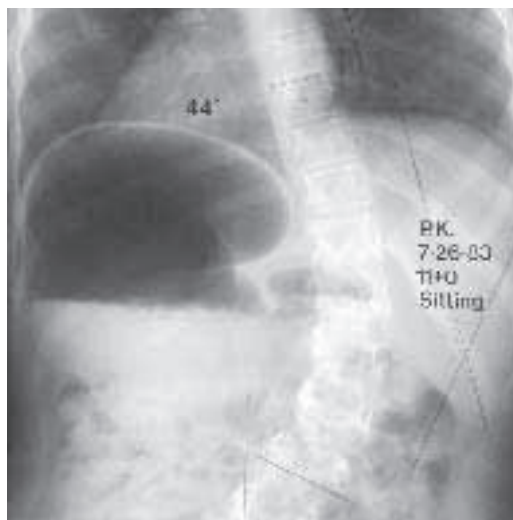
Treatment for Neuromuscular Scoliosis

Because many of these conditions are related to poor muscle control of the spine, surgery is the ultimate treatment. Surgery enables patients to sit straight and tall, improving existing hand function and making feeding and transportation easier. Surgery also can correct large curves, which affect pulmonary function. In such cases, surgery can help lessen occurrences of pneumonia and diminish swallowing and aspiration problems.

Bracing

Non-operative treatment consists of either seating support or use of a brace. Seating supports are used in young children who haven't developed sufficient balance for sitting. In such cases, we aren't treating the curve; we're helping children sit properly. As a child becomes taller and heavier, the spine becomes more unstable and the curve increases. Surgery is usually indicated at the onset of the adolescent growth spurt.

In some children with flaccid spines, a brace is used to support the spine externally when they are upright. The brace is a body brace or thoraco-lumbo-sacral orthosis (TLSO). With these children, a brace can help control the curve while they're growing. Although curve control is common in these children during the juvenile years, it fails at the onset of the adolescent growth spurt, when surgery is commonly performed.

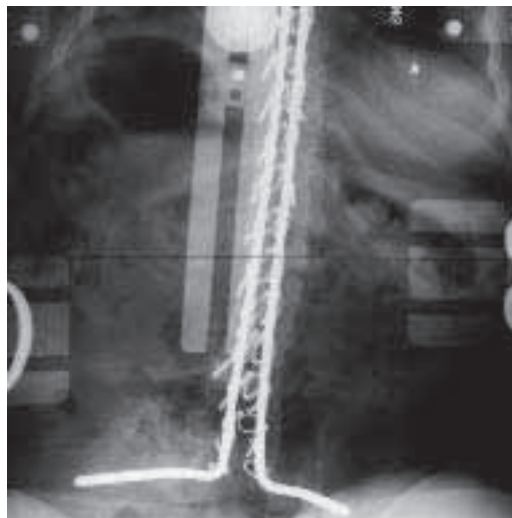


At the onset of the child's growth spurt, the curve increased to 44 degrees.

Surgery

Surgery is recommended for patients who present with a very large curve or when the curve increases in a child in a seating support or TLSO. Surgery involves a spinal fusion, which gives some curve correction, stabilizes the curve, and prevents the curve from increasing. In addition, the aim of surgery is to balance the spine by making the torso upright and balanced over a level pelvis.

Surgery to treat neuromuscular scoliosis falls into two groups. In ambulatory patients, the pelvis is generally level and the curve or curves present are treated as they are with idiopathic scoliosis. In non-ambulatory children, the fusion tends to be a long one — from the upper thoracic area to the sacrum. Fusion to the sacrum is necessary to correct the pelvic obliquity and to connect the spine to a stable base — the pelvis — for a stable sitting position. In addition, implants consisting of rods with hooks, screws and wires are used to correct the spinal deformity and hold the spine in the corrected position while the fusion becomes solid. Wires at



The child was treated with a posterior fusion and instrumentation. This sitting view shows a balanced spine with a solid fusion.

every level have proven invaluable in these children, because the bone is osteoporotic and the segmental fixation distributes the forces over many fixation points.

In the pre-operative assessment, certain areas should receive special attention in these children. The nutritional status should be assessed with blood values, and the protein status and other nutritional levels should be evaluated. In addition, in cerebral palsy patients with swallowing and regurgitation problems, swallowing studies are performed. Corrective measures include Nissen fundal plication, insertion of a gastrostomy tube, and nutritional support when necessary.

In children with seizures, certain seizure medications (such as Depekane) can affect blood coagulation, so pre-operative clotting and bleeding studies are performed. If an effect is found, the seizure medication is changed. In children with pulmonary compromise, a pulmonologist should evaluate the child. In general, such children require post-operative care in a pediatric intensive care unit.

The surgery can be performed using a posterior approach or a combined anterior and posterior approach. When two procedures are necessary, they are generally performed under the same anesthetic. Staged procedures are used if the anterior operation is long, or if the child is not hemodynamically stable after the anterior procedure.

The anterior approach, with the excision of the discs, helps obtain correction in less flexible curves. In addition, bone added to the disc spaces helps achieve a solid fusion. The anterior approach is thus used in cases which show loss of flexibility on a traction radiograph, where the pelvis is not level and/or the torso cannot be balanced over the pelvis. The anterior approach also is added in cases where there is poor bone stock posteriorly for a fusion (as found in spina bifida or myelodysplasia). Cadaver bone is used as a bone graft when the fusion is long and there is insufficient local and iliac bone.

In general, these children spend five to seven days in the hospital. They do not require external immobilization in a brace unless the internal fixation is poor. They are returned to their pre-operative functional status, with children in wheelchairs requiring wheelchair modifications due to their altered sitting posture. Functionally, it has been found that there is improved pulmonary status, with a marked reduction in incidences of pneumonia, as well as improved sitting ability and easier feeding and transportation.

Summary

Scoliosis is common in neuromuscular deformities. It often occurs when a child is young. The prevalence varies with the different types of neuromuscular scoliosis. Because early detection is important, these children need to be checked annually for developing spinal deformities. Treatment is generally surgical, and a long fusion is usually necessary. The fusion has been shown to be beneficial to these children.

Author Profile

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John E. Lonstein, M.D., is an orthopaedic surgeon at Gillette Children's Specialty Healthcare. He also practices at the Twin Cities Spine Center in Minneapolis. He graduated from the University of Witwatersrand Medical School in Johannesburg, South Africa, and completed his surgical residency at Boston University Medical Center. He completed his orthopaedic residency at the University of Minnesota.

Lonstein, who has been on staff at Gillette since 1973, served as chief of staff in 1987-1988. He currently heads our Spinal Orthopaedics Program and sees patients with scoliosis and other spinal deformities.

Lonstein is certified by the American Board of Orthopaedic Surgeons and the Minnesota State Board of Examiners. His professional associations include the Academic Orthopaedic Society, Scoliosis Research Society, Academy of Cerebral Palsy and Developmental Medicine, Pediatric Orthopaedic Society of North America, Societe Internationale de Chirurgie Orthopedique et de Traumatologie (SICOT), North American Spine Society, American Orthopaedic Association, and Academic Orthopaedic Society.

Volume 11 Number 3
May/June 2002

A Pediatric Perspective focuses on specialized topics in pediatrics, orthopaedics, neurology and rehabilitation medicine.

Please send your questions or comments to:

A Pediatric Perspective
Marketing Communications
200 East University Avenue
St. Paul, MN 55101
651-229-1744

Editor-in-Chief.....Steven Koop, M.D.
Editor.....Beverly Smith-Patterson
Designer.....Kim Goodness
Photographer.....Anna Bittner

06-02SEXTON7600GG



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New at Gillette Evren Akin, M.D.



Gillette Children's Specialty Healthcare is pleased to welcome pediatric rheumatologist Evren Akin, M.D. She joins Richard Vehe, M.D., pediatric rheumatologist, at Gillette, seeing clinic patients with juvenile arthritis and related conditions. She'll also be an adjunct faculty member at the University of Minnesota School of Medicine and an active member of the University's department of pediatrics.

Akin received her medical degree from the Medical Faculty of Istanbul University in Turkey. She completed her internship and residency in pediatrics at Massachusetts General Hospital at Harvard Medical School in Boston, Mass. Akin was a postdoctoral fellow in pediatric rheumatology at Tufts University's Floating Hospital for Children in Boston. She also was a research fellow at Beth Israel Hospital at Harvard Medical School.

Most recently, Akin was assistant professor of pediatrics at Tufts University's School of Medicine in Boston, Mass. She has served as a staff pediatrician with the division of pediatric rheumatology and as a research associate in the division of rheumatology at New England Medical Center. Her particular research interest is treatment-resistant Lyme arthritis.

Akin is available for patient appointments at Gillette's main clinic in St. Paul. To refer a patient, please call Scheduling at 651-229-3944 or 800-719-4040.

Referral Information

Gillette accepts referrals from physicians, community professionals and outside agencies. Contact the admitting manager at the number listed below. Physicians who are on staff may admit patients through our Admitting department from 7 a.m. to 4:30 p.m. Physicians who are not on staff should contact the admitting manager.

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