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THE OFFICIAL NEWSMAGAZINE OF THE AMERICAN ACADEMY OF PEDIATRICS

# AAP News

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Richard M. Schwend and Brian A. Shaw

*AAP News* 2014;35;13

DOI: 10.1542/aapnews.2014359-13

The online version of this article, along with updated information and services, is located on the  
World Wide Web at:

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American Academy of Pediatrics

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## Focus On Subspecialties

### Early onset scoliosis an under-recognized but serious condition

by **Richard M. Schwend, M.D., FAAP**, and  
**Brian A. Shaw, M.D., FAAOS, FAAP**

Early onset scoliosis (EOS) is a complex and heterogeneous group of disorders in children under 5 years of age. The actual prevalence is unknown since it involves a spectrum of conditions from infantile resolving scoliosis to severe progressive deformity.

A subset of children with EOS may have what Robert Campbell, M.D., FAAP, termed thoracic insufficiency syndrome (TIS), the inability of the thorax to support normal lung function (*J Bone Joint Surg Am.* 2003;85-A(3):399-408). Pediatricians may be the first to detect a child with a deformity or may be asked to assist in perioperative management due to the presence of multiple comorbidities. Although the prevalence of EOS is lower than for adolescent or juvenile idiopathic scoliosis, mortality is higher than the two combined.

EOS and thoracic insufficiency syndrome are mostly a restrictive disease but may have a component of obstruction. The thorax becomes stiff with reduced intercostal motion and more dependent on the diaphragm. However, the diaphragm also becomes weaker and less efficient due to its malposition and malrotation that occurs in EOS. Since lung function normally declines in the mid-30s, the full effect of the pulmonary insufficiency may not be appreciated during childhood.

Michael G. Vitale, M.D., FAAP, has developed a validated classification for early onset scoliosis called the C-EOS. Key variables in the system include: etiology (idiopathic, neuromuscular, syndromal, congenital), Cobb angle of the major curve, amount of kyphosis and curve progression in degrees/year. Health effects and outcomes depend on the classification, underlying medical conditions and deformity effects on the spine or thorax.

Dr. Vitale also developed a questionnaire (EOS-Q) that reflects many important aspects of health: lung function, sleep, play, activities of daily living, and physical and psychological function. Whereas quality of life scores are low in EOS and TIS, children who had thoracic spine fusions before 5 years of age have markedly decreased scores and pulmonary function tests.

There has been a greater awareness of diagnostic radiation exposure in young children, especially in the child with EOS who may receive



Photos courtesy of Richard M. Schwend, M.D., FAAP

This 15-month-old boy (left) by age 6 years had progressed to a 90-degree left scoliosis that diminished his thoracic volume and pulmonary function.

multiple lifetime exposures, including spine computed tomography (CT) scans. As low as reasonably achievable (ALARA) radiation exposure is the standard for children. New low-dose imaging technology adapted by many pediatric spine programs can obtain simultaneous anterior-posterior, lateral and reconstructed 3D whole spine images at about seven times lower dose. Magnetic resonance imaging delivers no radiation and is replacing CT scanning for spine and lung imaging, although it requires anesthesia in the young child.

Traditional treatment involved observation, prolonged bracing or early spine fusion. However, a spine fusion before age 5 years, especially if fused in the upper thoracic spine, can lead to a short chest with iatrogenic thoracic insufficiency. To avoid early surgery, "growth friendly surgery" became popular, requiring periodic lengthening surgical procedures with spine implants. In the 1990s, the first expandable rib-based device approved by the Food and Drug Administration (FDA) for children (the vertically expanding prosthetic titanium rib or VEPTR) was developed.

Current treatment involves the medical home, specialists and parents through shared decision-making and support groups. Treatments that prevent deformity while increasing lung and chest volume and

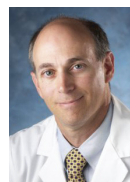
motion are ideal. Casting the young child has become popular, similar in principle to the Ponseti Method for the clubfoot. Traction is used for more severe deformity in the young child, prior to surgical treatment. Spine or chest implants that distract the spine or chest may provide greater chest volume, but also may create a stiffer chest and do not restore lung function. Newer implants that do not require repeat anesthesia and surgery include magnetic distraction rods and anterior vertebral body tethers that provide guided growth.

Challenges with current implant technology include a lengthy FDA approval process, relatively few cases to gain collective experience, and a lack of financial and other incentives for industry to test and develop devices for children. The “off label” use of existing spine

implant devices is appropriate and is supported by the Academy for treatment of spine and chest deformities; however, many implants designed for adults are too large or inappropriate for children. Several research study groups have been created to improve the care of children with EOS.



**Dr. Schwend**



**Dr. Shaw**

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