Terminology

61

Ying Li, Ron El-Hawary, Behrooz A. Akbarnia, and Tricia St. Hilaire

Contents

Key Points

- EOS presents before the age of 10 years (9 years of age or younger), regardless of etiology.
- The classification for early-onset scoliosis (C-EOS) should be used to describe the etiology of EOS.
- The term "growth friendly" is used to classify implants and techniques used for the treatment of EOS that allow for continued growth of the spine.
- A "graduate" is a patient who has undergone any surgical program to treat EOS, has reached skeletal maturity, and does not have a planned surgical intervention for EOS in the future.

Y. Li (✉)
Department of Orthopaedic Surgery, C.S. Mott Children's Hospital, Michigan Medicine, Ann Arbor, MI, USA
e-mail: yingyuli@med.umich.edu

R. El-Hawary
Paediatric Orthopaedics, School of Biomedical Engineering, IWK Health Centre, Dalhousie University, Halifax, NS, Canada

B. A. Akbarnia
Department of Orthopaedic Surgery, University of California, San Diego, San Diego, CA, USA

T. St. Hilaire
Pediatric Spine Foundation, Valley Forge, PA, USA

61.1 Introduction

The treatment of young children with scoliosis has steadily evolved over the past 15 years. The term early-onset scoliosis (EOS) has become widely used in the medical literature

B. A. Akbarnia et al. (eds.), *The Growing Spine*, https://doi.org/10.1007/978-3-030-84393-9_61

as an increasing number of research efforts have focused on this condition. In 2013, members of the Children's Spine Study Group (CSSG) and Growing Spine Study Group (GSSG), now known collectively as the Pediatric Spine Study Group (PSSG), formed a terminology subcommittee with the goal of creating consensus on the definition of EOS, the terminology used for the classification of EOS, and the terminology used for the treatment of EOS [1]. Use of the same nomenclature in all research related to EOS will hopefully increase the consistency and validity of these studies and allow for more reliable interpretation of study findings.

The terminology subcommittee obtained consensus on defining EOS as "scoliosis that is present before the age of 10 years (9 years of age or younger), regardless of etiology." [1] This definition has been approved by the PSSG, the Growing Spine Committee of the Scoliosis Research Society, and the Pediatric Orthopaedic Society of North America. In addition, the terminology subcommittee recommended consistent use of the classification for early-onset scoliosis (C-EOS) to describe the etiology of EOS [1]. The four etiologies of scoliosis in the C-EOS are congenital/structural, neuromuscular, syndromic, and idiopathic [2]. The Growing Spine Committee of the Scoliosis Research Society also published an Early Onset Scoliosis Consensus Statement in 2015 that describes the diagnostic categories of EOS. In this consensus statement, thoracogenic scoliosis, defined as EOS resulting from multiple congenital rib fusions or changes in the chest wall following thoracic surgery, is listed as a fifth diagnostic category for EOS [3]. This category of scoliosis is included in the congenital/structural etiology in the C-EOS. Lastly, the terminology subcommittee supported the term "growth friendly" to classify implants and techniques used for the treatment of scoliosis that allow for continued growth of the spine [1]. The three categories of growth-friendly spine implants are based on the correction forces that the implants exert on the spine and include distraction-based, compression-based, and guided growth systems [4].

This chapter discusses terminology that is commonly used in relation to EOS. We encourage the use of this terminology in clinical practice as well as in research related to EOS.

61.2 Terminology

61.2.1 Early-Onset Scoliosis (EOS)

Scoliosis that develops before the age of 10 years (9 years of age or younger), regardless of etiology [1, 3].

61.2.2 Congenital/Structural EOS

Scoliosis that develops as a result of a structural abnormality or asymmetry of the spine and/or thoracic cavity (Table 61.1) [2, 3].

61.2.3 Neuromuscular EOS

Scoliosis that develops secondary to a neuromuscular condition of high or low tone (Table 61.1) [2, 3].

61.2.4 Syndromic EOS

Scoliosis that develops as a result of a syndrome with a known or possible association with scoliosis (Table 61.1) [2, 3].

61.2.5 Idiopathic EOS

Scoliosis with no clear associated cause [2, 3].

61.2.6 Thoracic Insufficiency Syndrome (TIS)

The inability of the thorax to support normal respiration or lung growth [5].

61.2.7 Thoracic Height

The vertical length between two parallel lines drawn from the center of the superior endplate of T1 and the center of the inferior endplate of T12 measured on an upright anteroposterior spinal radiograph.

61.2.8 Spine Height

The vertical length between two parallel lines drawn from the center of the superior endplate of T1 and the center of the superior endplate of S1 measured on an upright anteroposterior spinal radiograph.

61.2.9 6-Minute Walk Test (6MWT)

This is a simple test that is routinely used to assess exercise capacity in patients with chronic pulmonary disease and restricted respiratory capacity. It has been used as an objective assessment of cardiorespiratory function in adolescents

Table 61.1 Etiologies of scoliosis

Congenital/structural	Neuromuscular	Syndromic
Hemivertebrae	Flaccid spinal cord injury	Spinal dysraphism
Fused ribs	Spinal muscular atrophy	Ehlers–Danlos (and other connective tissue disorders)
Thoracogenic	Muscular dystrophy	Prader–Willi syndrome
Iatrogenic (post-thoracotomy)	Spina bifida	Marfan syndrome
Tumor (before or after resection)	Low tone cerebral palsy	Achondroplasia
Amniotic band syndrome	Friedreich's ataxia	Arthrogryposis
Hemihypertrophy	Familial dysautonomia	Diastrophic dysplasia
Neurofibromatosis (dysplastic type)	Syringomyelia	Ellis–van Creveld syndrome
Congenital diaphragmatic hernia	Charcot–Marie–tooth syndrome	Neurofibromatosis
Congenital heart defect (status post repair)	CHARGE (coloboma of eye, heart anomaly, choanal atresia, retardation, and genital and ear anomalies) syndrome	Osteogenesis imperfecta
Proteus syndrome	Spastic cerebral palsy	Spondyloepiphyseal dysplasia
Jeune syndrome	Spastic spinal cord injury	Down syndrome
Constrictive chest wall syndrome	Rett syndrome	Goldenhar syndrome
Jarcho–Levin syndrome		Klippel–Feil syndrome
Spondylothoracic dysplasia		
Spondylocostal dysplasia		
VACTERL (vertebral, anal, cardiac, tracheal, esophageal, renal, and limb abnormalities)		

Source: Based on data from Ref. [2]

with idiopathic scoliosis [6]. Standard pulmonary function tests can be difficult to perform in children with EOS due to their young age and inability to cooperate. The 6MWT can be easily performed in a clinic setting and age-matched normal values exist for comparison [7]. Patients are asked to walk down a 30 meter straight hall as fast as possible for 6 minutes and the distance walked is recorded. Vital signs, including blood pressure, heart rate, respiratory rate, and peripheral oxygen saturation, can also be monitored [6].

61.2.10 24-Item Early-Onset Scoliosis Questionnaire (EOSQ-24)

A validated patient-reported outcome measure that evaluates the health-related quality of life of patients with EOS and the burden on their caregivers. The questionnaire is completed by the caregiver and age-matched normative values exist [8].

61.2.11 Mehta Casts

Cotrel and Morel described the elongation–derotation–flexion (EDF) casting technique for scoliosis in 1964 [9]. Mehta popularized the technique when she published the results of a prospective study that demonstrated that serial EDF casting led to curve resolution in patients with moderate infantile scoliosis if treatment was started at a young age, and casting resulted in curve improvement in older children with more severe curves [10].

61.2.12 Risser Casts

Described by Risser for the correction of scoliosis in 1955. Differs from the EDF casting technique in that a lateral force is applied to the ribs at the apex of the curve with less focus on derotation of the spine [11].

61.2.13 Distraction-Based Systems

Correct spinal deformities by mechanically applying a distractive force on the spine. The proximal and distal anchors can attach to the ribs (rib-based), spine (spine-based), ribs and spine (hybrid), or pelvis. Examples are the Vertical Expandable Prosthetic Titanium Rib (VEPTR) (Fig. 61.1), traditional growing rods (TGRs) (Fig. 61.2), and magnetically controlled growing rods (MCGRs) (Fig. 61.3) [4].

61.2.13.1 VEPTR

Originally developed to treat TIS resulting from severe progressive congenital thoracic scoliosis with fused ribs. At the time of VEPTR insertion, an opening wedge thoracostomy is performed to expand the hypoplastic hemithorax and increase thoracic volume. Serial lengthening of the VEPTR devices allows for continued increase in the height of the thoracic spine [12].

61.2.13.2 TGR

The goal of the growing rod technique is to control the spinal deformity while allowing for continued growth of the spine.

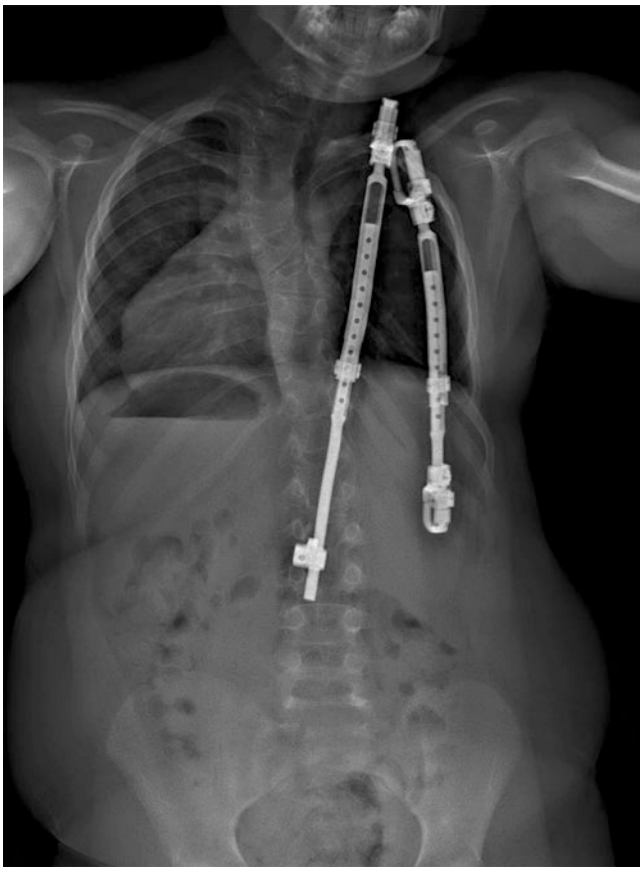

Fig. 61.1 A child with congenital scoliosis treated with rib-to-rib and rib-to-spine Vertical Expandable Prosthetic Titanium Ribs (VEPTRs)

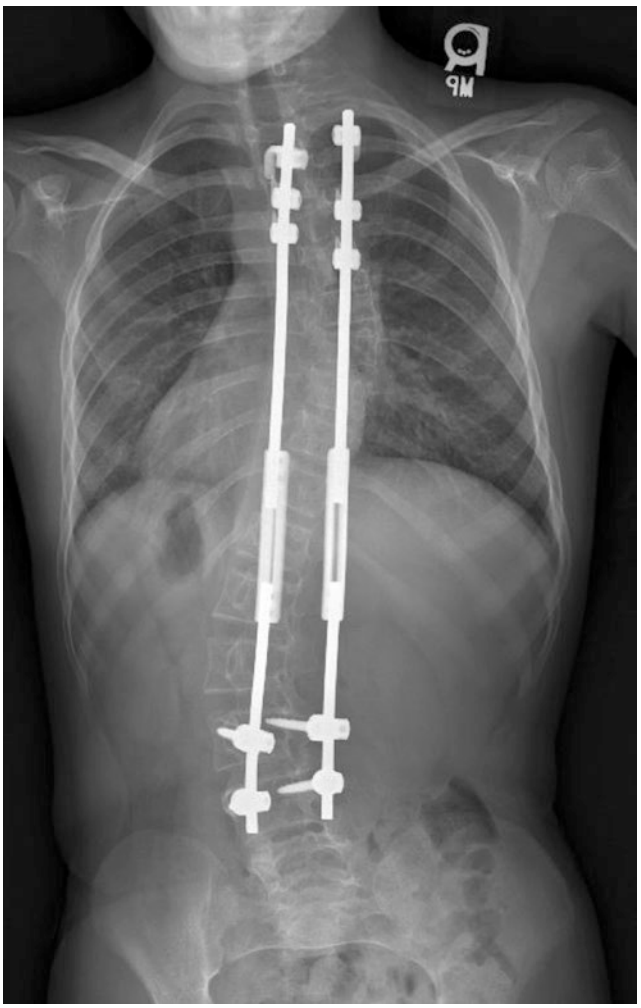

Fig. 61.2 A child with neurofibromatosis treated with traditional growing rods (TGRs)

Anchors are placed and a limited fusion is performed at the location of the proximal and distal foundations. A foundation is defined as an assembly of at least two anchors and rods that are stable and strong enough to accept corrective loads and resist deforming loads without dislodgment of the anchor or plastic deformation of the rods [13]. The remainder of the spine between the proximal and distal anchors is left unexposed in order to prevent unwanted fusion.

61.2.13.3 MCGRs

These are available as standard and offset rods. In a standard rod, the magnet is within the distal portion of the actuator when the "cephalad" arrow is pointing cephalad and the rod telescopes at the proximal end of the actuator. In an offset rod, the magnet is within the proximal portion of the actuator when the "cephalad" arrow is pointing cephalad and the rod telescopes at the distal end of the actuator [14]. TGR are surgically lengthened every 6 months [15]. MCGRs are lengthened noninvasively, typically in an outpatient setting without the need for sedation, using an external remote controller (ERC) [16]. The frequency of MCGR lengthening varies according to surgeon preference but usually ranges from 6 weeks to 4 months.

61.2.14 Compression-Based Systems

Correct spinal deformities by applying a compressive force to the convex side of the curve. Curve correction occurs as a result of mechanically applying a compressive force on the spine at the time of implant insertion and growth modulation of the vertebral endplates over time. An example is vertebral body tethering (Fig. 61.4) [4].

Fig. 61.3 A child with lipomeningocele treated with magnetically controlled growing rods (MCGR)

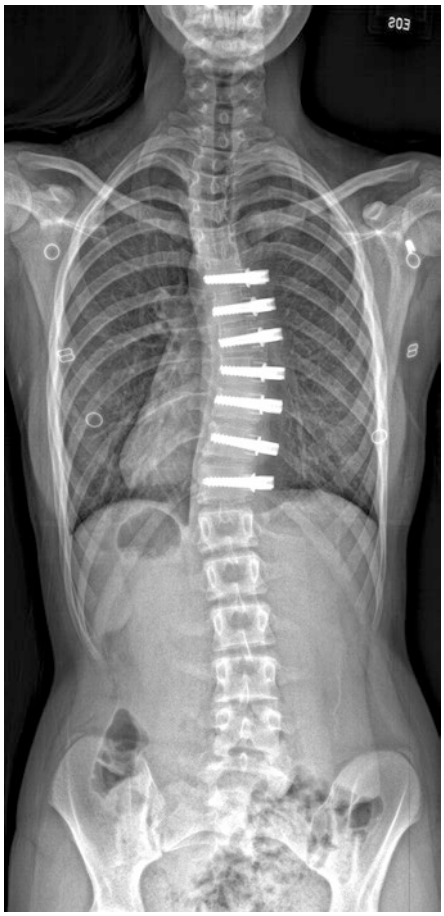

Fig. 61.4 An adolescent with idiopathic scoliosis treated with the vertebral body tether (VBT). Courtesy of Firoz Miyanji, MD

61.2.15 Guided Growth Systems

Correct spinal deformities by anchoring the apical vertebrae to rods and mechanically applying a translational force at the time of implant insertion. The majority of the other anchors are loosely attached to the rods, which allows the spine to grow longitudinally as the anchors slide over the rods. Examples are the Luque Trolley and Shilla (Fig. 61.5) [4].

61.2.15.1 Shilla Procedure

Developed as an alternative to the growing rod technique. While the main principle is still to control the spinal deformity while allowing for continued growth of the spine, in contrast to growing rods, a fusion is performed at the apex of the curve where the deformity is the most pronounced and the remainder of the spine is left unexposed. Rods that are fixed to the apical screws are placed into extraperiosteally inserted sliding pedicle screws proximally and distally, with the goal of guiding the growth of the spine into a more normal alignment without the need for repeat surgeries [17].

61.2.16 Graduate

A patient who has undergone any surgical program to treat EOS, has reached skeletal maturity, and does not have a planned surgical intervention for EOS in the future [18].

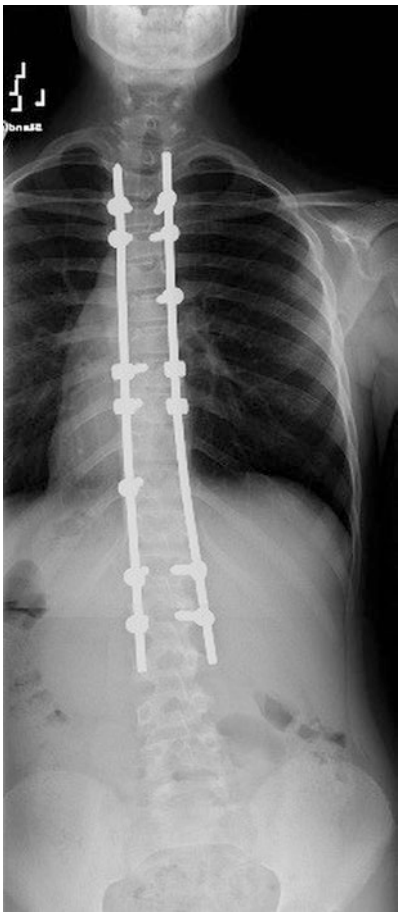

Fig. 61.5 A child with seizures and idiopathic scoliosis treated with Shilla. Courtesy of Scott Luhmann, MD

61.3 Conclusion

As treatment options for EOS become more widely available and an increasing number of studies are conducted on this subject, it is important to use consistent terminology in both clinical practice and research related to EOS. We encourage the use of the terminology presented in this chapter.

References

1. El-Hawary R, Akbarnia BA. Early onset scoliosis – time for consensus. Spine Deform. 2015;3(2):105–6.
2. Williams BA, Matsumoto H, McCalla DJ, Akbarnia BA, Blakemore LC, Betz RR, et al. Development and initial validation of the Classification of Early-Onset Scoliosis (C-EOS). J Bone Joint Surg Am. 2014;96(16):1359–67.
3. Skaggs DL, Guillaume T, El-Hawary R, Emans JB, Mendelow MJ, Smith JS, et al. Early onset scoliosis consensus statement. Spine Deform. 2015;3(2):107.
4. Skaggs DL, Akbarnia BA, Flynn JM, Myung KS, Sponseller PD, Vitale MG, et al. A classification of growth friendly spine implants. J Pediatr Orthop. 2014;34(3):260–74.
5. Campbell RM Jr, Smith MD, Mayes TC, Mangos JA, Willey-Courand DB, Kose N, et al. The characteristics of thoracic insufficiency syndrome associated with fused ribs and congenital scoliosis. J Bone Joint Surg Am. 2003;85(3):399–408.
6. Alves VL, Avanzi O. Objective assessment of the cardiorespiratory function of adolescents with idiopathic scoliosis through the six-minute walk test. Spine (Phila Pa 1976). 2009;34(25):E926–9.
7. Lammers AE, Hislop AA, Flynn Y, Haworth SG. The 6-minute walk test: normal values for children of 4–11 years of age. Arch Dis Child. 2008;93(6):464–8.
8. Matsumoto H, Williams B, Park HY, Yoshimachi JY, Roye BD, Roye DP Jr, et al. The final 24-item early onset scoliosis questionnaires (EOSQ-24): validity, reliability and responsiveness. J Pediatr Orthop. 2018;38(3):144–51.
9. Cotrel Y, Morel G. The elongation-derotation-flexion technic in the correction of scoliosis. Rev Chir Orthop Reparatrice Appar Mot. 1964;50:59–75.
10. Mehta MH. Growth as a corrective force in the early treatment of progressive infantile scoliosis. J Bone Joint Surg Br. 2005;87(9):1237–47.
11. Risser JC. The application of body casts for the correction of scoliosis. Instr Course Lect. 1955;12:255–9.
12. Campbell RM Jr, Smith MD, Mayes TC, Mangos JA, Willey-Courand DB, Kose N, et al. The effect of opening wedge thoracostomy on thoracic insufficiency syndrome associated with fused ribs and congenital scoliosis. J Bone Joint Surg Am. 2004;86(8):1659–74.
13. Yazici M, Asher MA, Hardacker JW. The safety and efficacy of Isola-Galveston instrumentation and arthrodesis in the treatment of neuromuscular spinal deformities. J Bone Joint Surg Am. 2000;82(4):524–43.
14. MAGEC technique guide. NuVasive Specialized Orthopedics Inc.; 2016.
15. Akbarnia BA, Marks DS, Boachie-Adjei O, Thompson AG, Asher MA. Dual growing rod technique for the treatment of progressive early-onset scoliosis: a multicenter study. Spine (Phila Pa 1976). 2005;30(17 Suppl):S46–57.
16. Cheung KM, Cheung JP, Samartzis D, Mak KC, Wong YW, Cheung WY, et al. Magnetically controlled growing rods for severe spinal curvature in young children: a prospective case series. Lancet. 2012;379(9830):1967–74.
17. McCarthy RE, Luhmann S, Lenke L, McCullough FL. The Shilla growth guidance technique for early-onset spinal deformities at 2-year follow-up: a preliminary report. J Pediatr Orthop. 2014;34(1):1–7.
18. Hardesty CK, Murphy RF, Pawelek J, Glotzbecker MP, Hosseini P, Johnston C, et al. Defining an early onset scoliosis "graduate" – the Pediatric Spine Study Group experience. Spine Deformity [Submitted, 2020].