Early-onset scoliosis: clinical presentation, assessment and treatment options

M Zaki B Choudhury

Athanasios I Tsirikos

David S Marks

Abstract

Early-onset scoliosis (EOS) is an 'umbrella' term defining scoliosis presenting before the age of 10 years. It reflects a constellation of conditions, which are challenging to treat. EOS is subdivided aetiologically into: idiopathic, congenital, thoracogenic, neuromuscular and syndromic. Each group has unique issues to address. The cornerstone of treating EOS is the facilitation of optimal conditions for lung development, thoracic growth and spinal movement. As a proxy marker, thoracic height contributes significantly to lung volume. One must adopt a holistic approach with consideration of the impact of multiple treatment interventions in early life on overall development. Treatment is guided by the principles above. In certain patients, bracing or serial casting treatment can restore spinal parameters to normal with no late residual. In other cases, such as neuromuscular and syndromic, operative interventions are often necessary. Surgical options range from primary fusion, to growth sparing implants that are periodically extended to allow spinal lengthening and thereby thoracic volume increase. 'Growing rods' have evolved over time—some require multiple surgeries, whilst others rely on a guided growth principle. A further recent development is the externally lengthened magnetic 'growing rod'. This review addresses the underlying conditions, assessment and treatment of patients with EOS.

Keywords bracing; early-onset scoliosis; EOS; growing rods; serial casting; thoracic insufficiency

M Zaki B Choudhury *BA(Hons) FRCS(Tr&Orth) DipSEM(UK&I) Spinal Fellow, Scottish National Spine Deformity Centre, Royal Hospital for Sick Children, Edinburgh, UK. Conflicts of interest: none declared.*

Athanasios I Tsirikos *MD FRCS PhD Consultant Orthopaedic and Spine Surgeon, Scottish National Spine Deformity Centre, Royal Hospital for Sick Children, Edinburgh, UK. Conflicts of interest: educational/teaching commitments with DePuy/Synthes Spine.*

David S Marks *FRCS FRCS(Orth) Consultant Orthopaedic Spine Surgeon, Royal Orthopaedic Hospital and The Birmingham Children's Hospital, Birmingham, UK. Conflicts of interest: consultancy agreement with DePuy/Synthes Spine; Speaker bureau: DePuy/Synthes Spine, Stryker Spine, Medtronic, Nuvasive. Royalties: DePuy/Synthes Spine.*

Introduction

Early-onset scoliosis (EOS) is defined by the Scoliosis Research Society (SRS) Growing Spine Study Group as 'spine deformity that is present before 10 years of age'.[1] It is an uncommon condition, which can be life limiting with a complex group of underlying diagnoses. As a result multiple different methods of treatment are used, in part due to the heterogenous nature of causes.

Physiological importance

EOS is clinically extremely significant due to the potentially devastating consequences on thoracic growth and lung development. Pehrsson et al.[2] undertook a review of the natural history and reported early deaths and respiratory failure in patients with untreated scoliosis. Post-mortem studies have demonstrated not only small but also hypoplastic lungs. In the seminal work on spinal growth DiMeglio[3] demonstrated the most rapid period of spinal growth occurs in the first 5 years of life. In this period the spine increases 50% of its length, but reaches 95% of adult canal diameter. Between 5 years and 10 years old spinal growth continues at a slower rate but this is a critical period of alveolar development, both in terms of number and functional complexity. Thoracic spine height averages 11 cm at birth, 18 cm at 5 years age and 22 cm at 10 years. Consider then that in the context of early fusion in early-onset scoliosis in a 5-year-old child this could lead to a 12.5 cm loss of spinal growth, leading to significantly curtailed pulmonary development.[4] The cornerstone of treating this particular age group therefore is facilitation of lung development, thoracic growth and, if possible, preservation of movement. Spinal shape is regarded as a proxy marker of lung function with thoracic height contributing significantly to lung volume though in a non-linear way. The effects of spinal fusion prior to near maximal lung maturity at 10 years of age have been shown, with patients having a mean forced vital capacity (FVC) 41% of normal at maturity. This compared with 68% of normal FVC in patients having spinal fusion older than 10 years.

It is worth noting however that the relationship is often not quite so simple, with interventions sometimes leading to loss of chest wall compliance and paradoxically less improvement in lung function. Campbell's[5] work focused on the analysis of lung function in the context of fused ribs and congenital scoliosis. This can create a three-dimensional (3D) thoracic deformity with adverse effects on thoracic growth and function termed 'thoracic insufficiency syndrome' (TIS), describing the inability of the thorax to support normal respiration or lung growth.

This review shall be structured around the SRS consensus document but will seek to expand each aspect to provide a global overview. The sub-groupings of EOS are considered in Table 1.

Prognosis

When discussing prognosis there are two factors to consider: firstly the curve prognosis and secondly the overall morbidity and potential mortality to the patient (Table 2). As the diagnostic categories show this is a very diverse group of patients. As such in some of the diagnostic subgroups the incidence of significant cardiac and respiratory co-morbidities is much

Please cite this article in press as: Choudhury MZB, et al., Early-onset scoliosis: clinical presentation, assessment and treatment options, Orthopaedics and Trauma (2017), https://doi.org/10.1016/j.mporth.2017.09.006

Diagnostic categories of early-onset scoliosis (EOS)[1]

Idiopathic	Congenital	Thoracogenic	Syndromic	Neuromuscular
• No apparent cause or related underlying aetiology • Infantile idiopathic — a subgroup which develop scoliosis in infants and children less than 3 years old • Juvenile idiopathic age 3—10 years	• Vertebrae develop incorrectly in uterus • Sometimes associated with cardiac and renal abnormalities, e.g VATER, VACTERL • Evaluation may include studies of heart and kidneys	• Multiple congenital rib fusions as seen in spondylocostal or spondylothoracic dysostosis • May have congenital vertebral anomalies • May also be considered congenital scoliosis • Changes in the chest wall following thoracic surgery which may function as a tether which promotes change in the shape of the spine	• Syndromes, such as Marfan's, Ehlers—Danlos and other connective tissue disorders • Neurofibromatosis • Prader—Willi • Numerous bone dysplasias may be associated with EOS	• Can develop in children with neuromuscular disorders • Spinal muscular atrophy (SMA) • Cerebral palsy • Spina bifida • Brain or spinal cord injury

Table 1

higher. Overall consideration must be given to the prognosis if the condition is left untreated as discussed above, but the fundamental assumption is that some form of treatment is better than observation alone. In patients with idiopathic scoliosis there are a number of features to suggest good prognosis if treated conservatively (i.e with casting or bracing). The early work identified that idiopathic EOS greater than 35° is likely to progress. In many children under 2 years old with infantile idiopathic curves less than 35°, scoliosis may resolve without treatment. Furthermore Mehta[6] observed the rib vertebral angle difference (RVAD) as a predictor of curve progression in infantile idiopathic scoliosis (Figure 1). This measurement is performed by drawing a line perpendicular to the endplate of the most translated apical vertebra and a line down the midpoint of the concave and convex rib at this level. The angle created on the convexity is subtracted from the opposite concave angle to measure the RVAD. Mehta found scoliosis resolution in 90% of children with RVAD less than 20°. Mehta also described the rib head phase as an adjunct to RVAD in determining curve progression. This is a rotational measure defined by the relationship of the rib head and vertebral body at the apex with overlapping of the vertebral body and the rib head predicting curve progression.

Assessment of EOS

A detailed history and examination should be undertaken at initial clinical presentation. It is possible that what is labelled as 'EOS' is actually a missed congenital scoliosis. A detailed birth history can reveal complications suggestive of periods of difficult or obstructed labour, and thus potential for a diagnosis of cerebral palsy, albeit perhaps the more subtle end of the spectrum. Similarly a thorough perinatal history can reveal issues with growth and intrauterine development suggestive of VATER/

Factors affecting prognosis

Idiopathic	Congenital	Thoracogenic	Syndromic	Neuromuscular
Note: consider role of MRI in curve >20° or rapid progression — higher incidence of syrinx and Chiari malformation	Vertebrae develop incorrectly in utero Sometimes associated with cardiac and renal abnormalities, e.g VATER, VACTERL Evaluation may include studies of heart and kidneys	Can be considered either: 'cause' — as in the case of thoracic insufficiency syndrome, with rib malformations and issues arising from ventilatory inadequacy, or: 'effect' — Changes following thoracic/cardiac surgery in early infancy which may function as an external tether. Note also potential ongoing thoracic/cardiac issues	Severity of connective tissue disorders Neurofibromatosis with associated complications: optic glioma, dural ectasia, soft tissue neurofibromata, bone quality Prader—Willi: behavioural issues and compliance with certain forms of treatment may limit their effectiveness	Related to severity of underlying condition, can have bearing on ability to recover from anaesthesia. Can also limit expected postoperative function

Table 2

Please cite this article in press as: Choudhury MZB, et al., Early-onset scoliosis: clinical presentation, assessment and treatment options, Orthopaedics and Trauma (2017), https://doi.org/10.1016/j.mporth.2017.09.006