Surgical management of early-onset scoliosis: indications and currently available techniques

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Abstract

Early-onset scoliosis (EOS) is a heterogeneous group of spinal deformities affecting children under the age of 10 years of which the aetiology, natural history and treatment options vary considerably. In progressive EOS, treatment is based on exhausting conservative measures (casting or bracing) to halt curve progression while allowing for continuous growth of the spine and chest development. Early spinal fusion leads to loss of longitudinal spinal growth and restriction of cardiopulmonary function. In rapidly progressive curves that have failed conservative treatment a range of 'growth-friendly' surgical techniques have been developed to control curve deterioration. The indications and characteristics of distraction-based or compression-based methods, growth guidance and promising new techniques are discussed according to aetiology of EOS. Definitive spinal fusion remains reserved for patients ideally towards the end of their spinal growth and for short-segment treatment in congenital scoliosis.

Keywords early onset scoliosis; future techniques; growing rods; growth guidance; growth-friendly surgery; magnetically controlled growing rods

Introduction

Early onset scoliosis (EOS) is defined as a scoliotic deformity developing in a growing child before the age of 10 years and accounts for 10% of all paediatric scoliosis cases. EOS comprises a heterogeneous group of spinal deformities whose aetiology, natural history and treatment varies widely. The Growing Spine Study Group and the Children's Spine Study Group classify EOS according to aetiology, coronal and sagittal curve severity.[1]

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In the Classification for Early-onset Scoliosis (C-EOS), aetiology is grouped into:

- neuromuscular scoliosis
- congenital/structural scoliosis, including scoliosis related to chest wall abnormalities
- syndromic scoliosis
- idiopathic scoliosis.

Curve severity is classified according to maximum coronal Cobb angle (<20°, 20–50°, 51–90° and >90°) and maximum sagittal Cobb angle for kyphosis (<20°, 20–50° and >50°). The progression modifier differentiates the change in curve progression (<10°, 10–20° and >20°), as expressed per year.

Treatment of severe scoliosis in the skeletally mature adolescent spine mostly involves spinal fusion which results in loss of spinal motion and remaining longitudinal growth. Treatment of severe scoliosis in the young child presents unique challenges given the remaining growth of the spine, chest and cardiopulmonary system. In addition, these children often have associated complex cognitive, functional or medical co-morbidities. Treatment of EOS aims to hold progression or correct the deformity, preserve spinal growth, maximize the thoracic volume to avoid cardiopulmonary compromise and optimize health-related quality of life. Due to notable progress in the proposed surgical treatment options for previously lethal conditions, it is a rapidly evolving area in spinal care. Currently, non-operative methods (corrective casting and bracing) are used to avoid or delay the need for spinal surgery and should be considered the mainstay of treatment. However, for rapidly progressive curves that require surgical stabilization, innovative 'growth-friendly' corrective techniques are utilized.

According to the classification by Skaggs et al.[2] the proposed surgical techniques in the management of EOS can be divided into:

- ***'Growth-friendly' surgical methods***
  - **Distraction-based methods** (traditional growing rods, magnetically controlled growing rods, vertical expandable prosthetic titanium rib-VEPTR), also known as growth preservation/stimulation techniques (Figure 1)
  - **Compression-based methods** (vertebral staples or vertebral tethering techniques), also known as growth modulating techniques (Figure 2)
  - **Growth guidance** (modern Luque trolley, Shilla technique) (Figure 3)
  - **Hybrid** 'growth-friendly' methods, such as convex growth arrest + concave distraction, or distraction-aided growth guidance (Figures 4-6).
- ***Definitive spinal fusion***
- ***Combinations*** of limited spinal fusion with 'growth-friendly' methods

In this overview, we present the current indications and available surgical treatment options for the management of EOS, structured by aetiology. Table 1 summarizes the currently available techniques for growth-friendly surgery of different types of EOS. It is important to note that in a recent survey of 20 worldwide experts in this field on six representative EOS cases, no consensus could be reached, demonstrating the considerable practice variation.[3] We are not elaborating on the conservative management of EOS as the principles of non-operative treatment and options available have been discussed in a previous review article.[4]

Idiopathic EOS

Background

Infantile (0–3 years) and juvenile (4–9 years) idiopathic EOS represents only 19% of all idiopathic scoliosis. While the natural history is often favourable with 80–90% of infantile curves spontaneously improving or resolving with growth, curves with a rib-vertebra-angle-difference (RVAD) exceeding 20° and initial Cobb angle greater than 30° are at high risk for progression and have an indication for corrective casting or bracing.[5] Juveniles with idiopathic scoliosis exceeding 20° have significant risk of curve deterioration during remaining spinal growth and therefore will also benefit from brace treatment.

Indications for surgery

Surgical treatment in idiopathic EOS is indicated in curves exceeding 60° at infantile or juvenile age, when conservative treatment has failed to stop progression.[6] To guide the decision on necessity and timing of 'growth-friendly' surgery, an assessment of the peak growth velocity, remaining spinal growth and risk of curve progression is required. It should be noted that each year of delay of instigation of traditional growing rod surgery, reduces the total complication rate by 13%.[7] During 'growth-friendly' treatment into early adolescence, radiological measures of skeletal maturity can be combined with physiological factors (age, menarche, Tanner staging) to provide information on remaining growth. A method of directly assessing the stage of spinal growth is currently not available when reviewing spinal radiographs. The most viable radiological measures of skeletal maturity that can be used in EOS patients are the ossification of the iliac apophysis (Risser stages 0–5), triradiate cartilage (open or closed) and proximal humeral ossification (Proximal Humeral Ossification System) on postero-anterior full-spine radiographs or the digital ossification on hand radiographs (Sanders Simplified Skeletal Maturity System).[8,9]

Treatment options

Surgical options to address idiopathic EOS include distraction-based or compression-based methods, guided-growth, hybrid techniques and definitive spinal fusion.

Distraction-based methods

Traditional growing rods (TGR) – the patient and their family should be informed of the prolonged treatment programme requiring repetitive surgeries and the high complication rates. The original Harrington rod was introduced in 1962 as a distraction system (one distraction rod with a proximal and distal hook anchor) without fusion.[10] Nowadays, traditional growing rods are the most commonly used spinal instrumentation technique in the growing spine to treat idiopathic, neuromuscular and syndromic EOS.

A TGR construct is composed of a proximal and distal vertebral anchor connected by two rods and a telescopic or parallel connector. Lengthening of the posterior construct requires repetitive surgery (which is planned usually every 6 months) with distraction of the rods through the connectors. Growing rods can

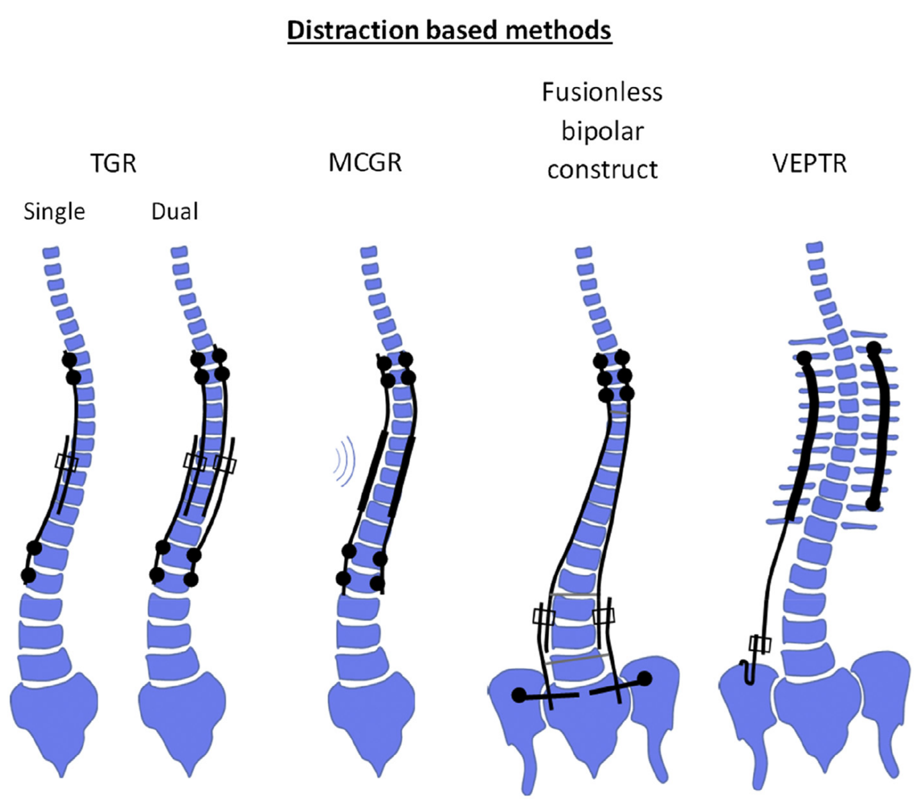

Figure 1 Schematic representation of distraction-based techniques. TGR, traditional growing rods; MCGR, magnetically controlled growing rods; VEPTR, vertical expandable prosthetic titanium rib.

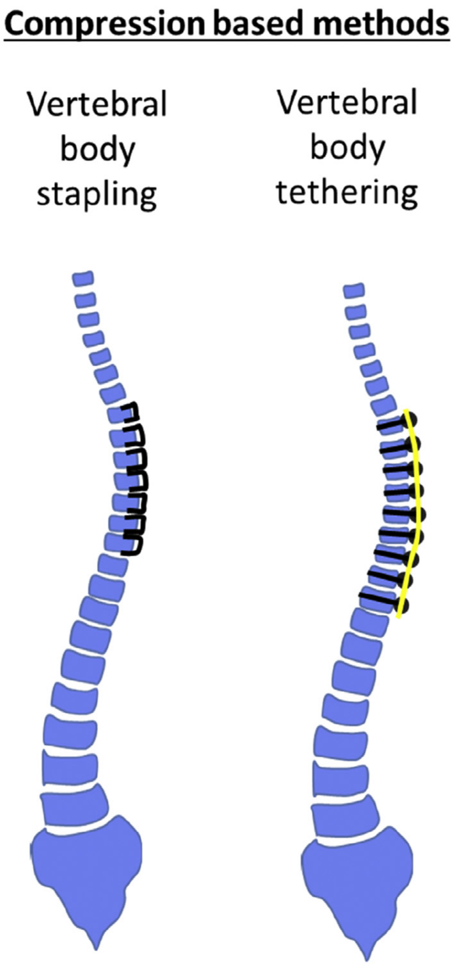

Figure 2 Schematic representation of compression-based techniques.

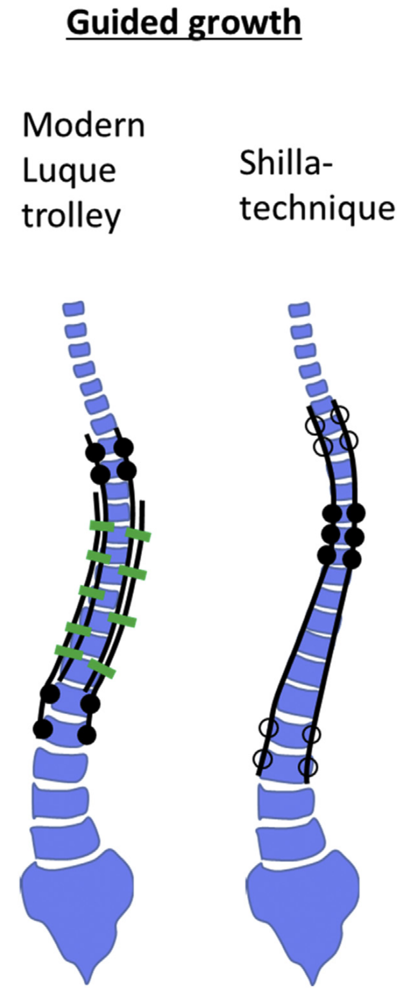

Figure 3 Schematic representation of growth guidance techniques.

be utilized as a single (unilateral/concave) or dual (bilateral) rod system, and with hooks or screws for proximal anchorage. While the single construct is less prominent under the skin, the dual rod system is generally more stable. Pedicle screws or hooks can be used for the proximal anchor with the screws providing increased stability.[11] However, the use of pedicle hooks as cephalad fixation point creates a less constrained construct that allows spontaneous proximal spinal growth between the lengthening procedures. For distal anchorage pedicle screws are used to produce a strong foundation. Bone graft is placed across the cephalad and caudal anchors of the growing rod construct to increase stability and reduce the risk of dislodgement, especially during repeat lengthenings. The initial distraction should be moderate to reduce the risk of failure of the vertebral anchors or neurological complications. While neuromonitoring should be used for the initial distraction, its use may be avoided in lengthening procedures.[12]

Growing rods in children with idiopathic EOS can provide correction of the coronal and sagittal deformity and allow for spinal growth. Following the Hueter-Volkmann principle, repetitive distraction through the rods can stimulate longitudinal spinal growth. Survivorship modelling has shown that after implantation at the age of 6, at least 10 lengthening procedures are required.[13] Wijdicks et al.[11] showed in a recent systematic review, that a considerable portion of the reported spinal growth during growing rod treatment is the result of the initial and final surgical correction and not due to the growth-friendly implant. The average spinal growth achieved by initial correction only was 3.9 cm (based on 34 studies), by the repetitive lengthenings 0.3 cm/year between T1–T12 (15 studies) and the spinal growth achieved by the final fusion was on average 2.3 cm (based on four studies).

The overall complication rate of repetitive lengthenings, however, is high. In the largest cohort (n = 140) so far, the total complication rate was 55% with a mean of 2.2 complications per patient.[7] The most common complications were: proximal anchor failure, rod breakage and infections. Another problem of traditional growing rods is spontaneous fusion of the non-instrumented segments. At the initial surgery, no subperiostal exposure is performed and care is taken to tunnel the rods intramuscularly without exposure of the posterior elements of the uninstrumented segments. Still, spontaneous fusion of the spine has been reported to occur in 80% of patients after five lengthening procedures.[14] Therefore, it is recommended to delay initial surgery by continuing casting or bracing as long as reasonably possible. Stiffening of the spine across the instrumented levels of the growing rod results in reduced ability to

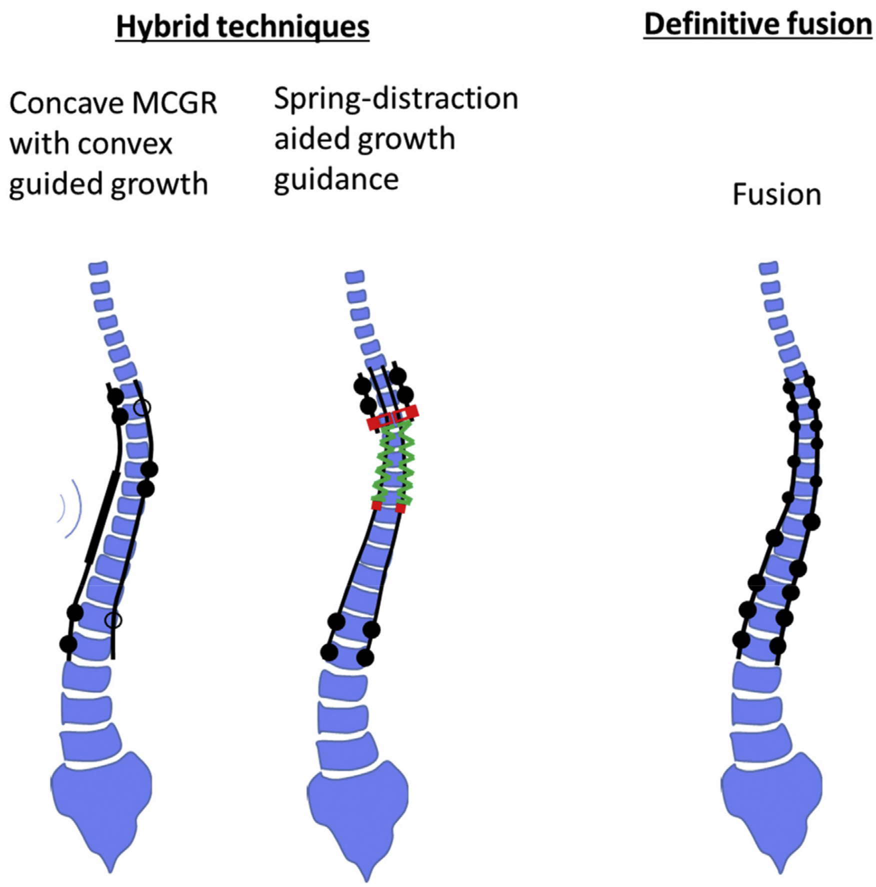

Figure 4 Schematic representation of hybrid ‘growth-friendly’ techniques and definitive spinal fusion. MCGR, magnetically controlled growing rods.

lengthen the rods and therefore correct the scoliosis after the initial lengthening; the so called ‘law of diminishing returns’.

Other issues encountered with the use of growing rods are the kyphogenic effect produced during lengthening of the posterior vertebral column which precipitates the development of proximal junctional kyphosis and occasionally proximal junctional failure. Finally, the growing rod technique cannot control vertebral rotation, and this results in exacerbation of chest torsion and the rib cage deformity over time.

Magnetically controlled growing rods (MCGR) — the indications and technique of MCGR is similar to growing rods. Instead of repetitive surgeries for lengthening of the construct the telescopic connector can be lengthened or shortened transcutaneously with an external remote control. The only currently available system is MAGEC X (NuVasive, Inc., San Diego, CA, United States). In contrast to traditional growing rods, lengthenings are performed in the outpatient clinic, without the need for anaesthesia or analgesia. MRI is no longer contraindicated in patients with MCGR.

One would expect that this technique provides fewer complications and psychological burden to the patients related to repeat lengthening procedures and serial hospital admissions. MCGR can provide correction of the coronal deformity and allow for spinal growth. Sagittal bending of the rod is difficult since the area of the actuator is straight and cannot be contoured. The complication rates and spontaneous fusion of the spine during resulting growth are similar to traditional growing rods as outlined previously.[15] Even though the overall number of lengthenings is higher, the spinal length achieved during resulting growth is similar compared to traditional growing rods.[16]

In 2014, the National Institute of Health and Care Excellence in the UK produced recommendations on the use of MCGR as a growth friendly treatment option for children with EOS suggesting that there is lack of quality or quantity of evidence to draw firm conclusions regarding the clinical efficacy of MAGEC rods compared with conventional growing rods.[17] Thakar et al.[13] performed a systematic review on the outcomes and complications associated with MCGR compared to traditional growing rods.[15] Fifteen studies on 196 patients showed that at 2.5 years, follow-up, the scoliosis improved from 65° to 35°, and T1−T12 spinal length increased by 1.4 cm/year. The total complication rate was 45%. The most common complications were unplanned revision surgery (33%), implant failure (11.7%) and rod breakage or foundation failure (10.6%). There is nowadays a significant number of studies reporting implant failure of distraction or component breakage in association with adjacent soft tissue metallosis, which led to modifications of the implant design. Until the failure of the distraction mechanism of MAGEC is completely addressed, EOS may be more safely treated by TGRs.[18]

Compression-based methods: aim to arrest vertebral growth of the convexity of the scoliotic curvature and allow growth on the

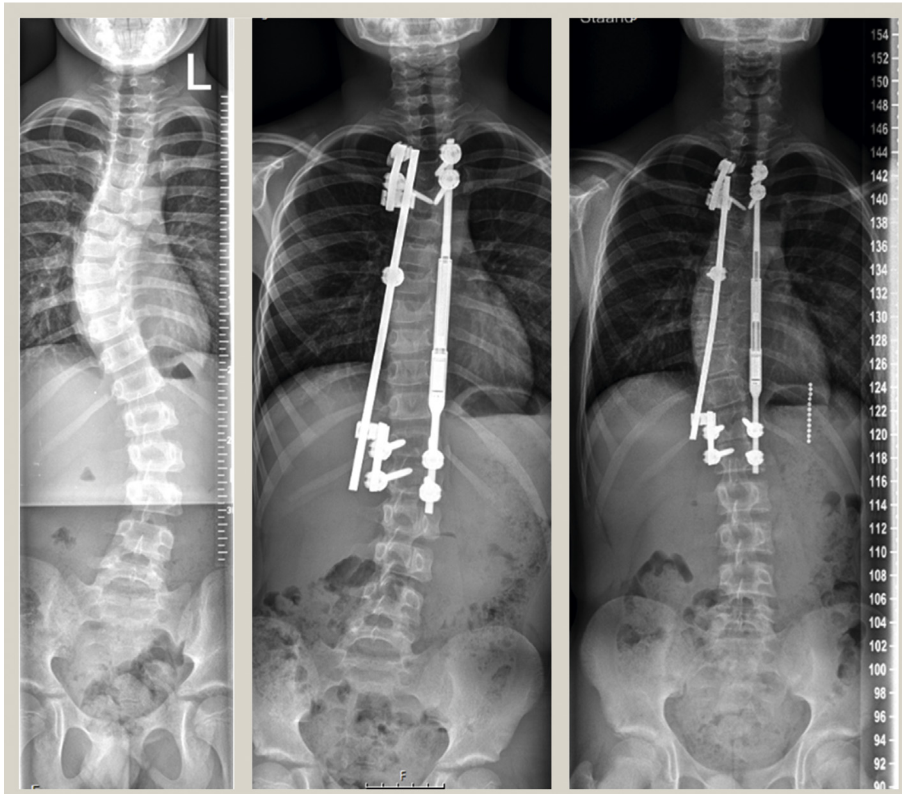

Figure 5 Example of a patient with an early onset idiopathic scoliosis treated with a hybrid technique (concave magnetically controlled growing rods with convex guided growth) which controlled the coronal deformity and preserved growth of the spine.

concave side by a hemi-epiphysiodesis effect, based on the Hueter-Volkmann principle. With these techniques, spinal growth is modulated, not guided. Convex growth arrest is an old concept of hemi-epiphysiodesis, with mixed reported results.[19] Currently, compression-based methods, such as anterior vertebral body stapling and tethering (AVBT), are promising techniques that could be used for skeletally immature patients with primarily flexible thoracic idiopathic curves of moderate severity. These techniques may not be as effective in non-idiopathic scoliosis.[20] Although the preliminary results of AVBT in skeletally immature patients over 8 years of age are encouraging, there is still limited long-term evidence and no evidence on the use of this technique in the younger population.[21,22] Theologis et al.[22] reported on 3.5 year follow-up of 12 patients with moderate idiopathic EOS treated with AVBT. Their mean scoliosis angle improved from 33^o to 23^o at follow-up; 3 patients (25%) had pulmonary complications.

The major attraction of AVBT is the possibility to achieve gradual curve correction by harnessing remaining concave vertebral growth in the presence of a convex tether, without the need for final fusion surgery. There have been major complications associated with the procedure which has been often wrongly portrayed as minimally invasive. In addition, there is no evidence on the results of this technique beyond skeletal maturity. Recently, Newton et al.[23] compared AVBT to definitive fusion, and showed that scoliosis correction can be better maintained by definitive fusion with less revision procedures and similar SRS-22r health-related quality of life outcomes.

Guided growth: the first self-growing rod system for scoliosis was the Luque trolley. The idea is that the coronal deformity can be controlled by connecting multiple vertebrae with anchors that can slide on the rods and preserve spinal growth. The Luque technique used segmental sublaminar wires but has been associated with high rates of spontaneous fusion resulting in limited subsequent vertebral growth. The concept of guided growth has been reintroduced by Ouellet et al.[24] in 2011. As compared to the Luque technique, the modern Luque trolley uses gliding achors of polyethylene that can slide along overlapping polished titanium rods. While preliminary results have been reported on 5 patients, further results of long-term clinical studies are needed to assess the effectiveness of this technique. Mehdian et al.[23] reported recently their mid-term results using a modified version of the original Luque trolley self-growing technique with pedicle screw fixation over the proximal and distal end foundations of the construct and the use of segmental sublaminar wires in between across overlapping bilateral rods. They demonstrated that this can be an effective technique for the treatment of EOS in non-ambulatory hypotonic children with an underlying myopathic condition. However, they noted a high rate of rod breakage occurring at the thoracolumbar junction in patients with an idiopathic or cerebral palsy EOS which limits the use of this technique.

Another method of guided growth is the Shilla technique, which connects the apex, as well as the proximal and distal ends of the scoliotic spine by rods that can glide in the proximal and distal vertebral anchors. In this way, the technique aims to control the apical segments by local derotation and fusion, while

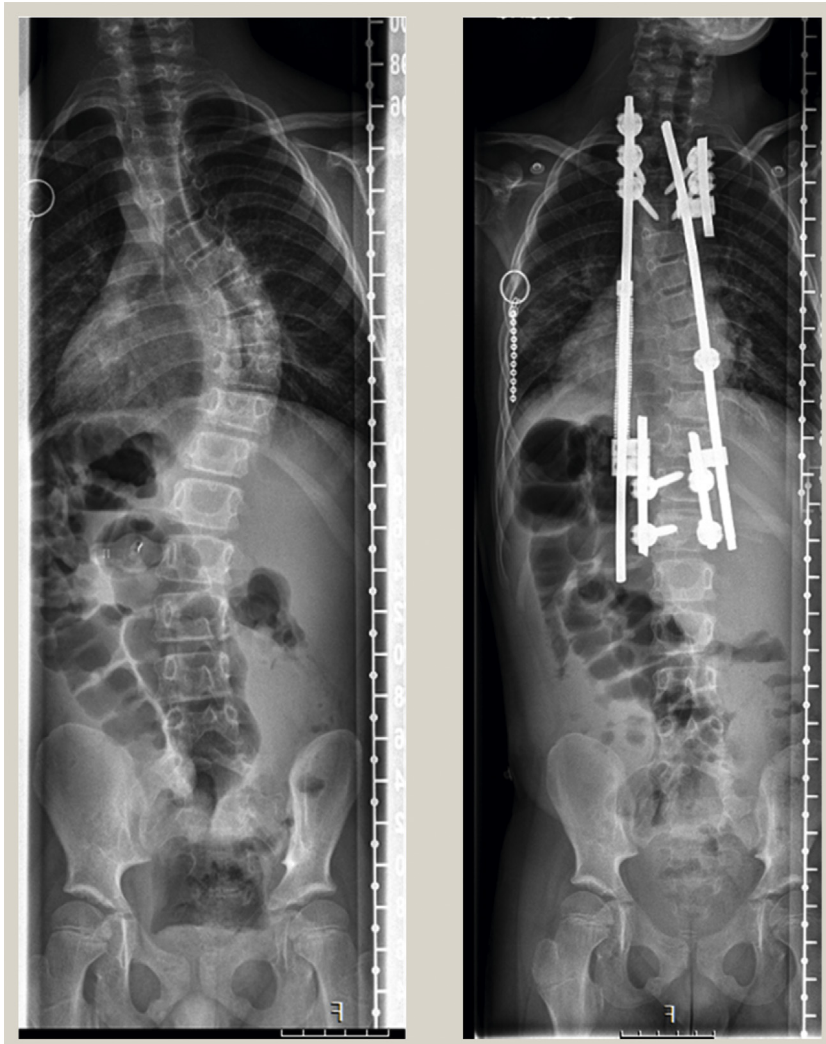

Figure 6 Example of a patient with an early onset idiopathic thoracic scoliosis who underwent a hybrid technique (spring distraction aided growth guidance).

allowing for proximal and distal spinal growth. In 2013, McCarthy et al.[25] showed that the Shilla technique required fewer reoperations (3 versus 7) but resulted in reduced spinal growth (0.7 cm/year) and lesser scoliosis correction with higher complication rates compared to growing rods. Therefore, its use has become limited to selective cases which need apical control.

Hybrid methods: recently, Wijdicks et al.[26] reported on the use of a combination of concave distraction-based technique (MCGR) with convex growth guidance and apical control (Shilla technique) in 18 EOS patients (6 idiopathic) with 3-year follow-up. The scoliosis angle improved from 65° preoperative to 38° at final follow-up and the post-implantation spinal growth rate averaged 10 mm/year at latest follow-up, which is similar to the physiological growth rates reported by Dimeglio.[24] There were 13 implant-related complications occurring in 6 patients and 4 non-surgical complications. Therefore, the hybrid method of unilateral MCGR and contralateral growth guidance is able to correct the deformity and maintain spinal growth comparable to the bilateral MCGR technique and is probably more cost-effective.

Spring-distraction aided growth guidance: there is increasing interest in growth modulating techniques, since current growth friendly systems have serious disadvantages (repetitive lengthenings, unplanned procedures, spontaneous fusion), especially at long-term follow-up. Recently, the spring-distraction aided growth guidance concept (Spring Distraction System) has been introduced as a promising growth-friendly technique; this works with integration of a continuous distraction force exerted by a spring with a sliding technique, without the need for repetitive surgeries.[25,26] In more detail, the implant is similar to dual traditional growing rods, except for that it includes compressed helical springs positioned around the rods and that the parallel rod connector is left dynamic to allow for longitudinal spinal growth. Lengthening is achieved by active continuous distraction through the compressed springs using the inherent spinal growth, while the rods can slide through the parallel connectors. A recent report on 24 patients with EOS (5 idiopathic, 7 congenital, 3 syndromic, 9 neuromuscular) and 2-year postoperative follow-up indicated that the concept of spring distraction can be feasible as an alternative to present growing spine

Treatment options for early onset scoliosis according to aetiology

	Idiopathic	Syndromic	Congenital	Neuromuscular
Distraction-based techniques	• TGR • MCGR	• TGR • MCGR • Fusionless bipolar construct	• TGR • MCGR • Chest wall deformities: vertical expandable prosthetic titanium rib (VEPTR)	• TGR • MCGR • Fusionless bipolar construct
Compression-based techniques	• Vertebral body stapling • Anterior vertebral body tethering		Hemi-epiphysiodesis	
Guided growth	• Modern Luque trolley • Shilla technique	• Modern Luque trolley • Shilla technique		• Modern Luque trolley • Shilla technique
Hybrid	• Concave MCGR with convex guided growth • Spring-distraction aided growth guidance	• Concave MCGR with convex guided growth • Spring-distraction aided growth guidance	Spring-distraction aided growth guidance	Spring-distraction aided growth guidance
Fusion	Definitive fusion	Definitive fusion	• Hemivertebra resection • In situ fusion • Definitive fusion	Definitive fusion

MCGR, magnetically controlled growing rods; TGR, traditional growing rods.

Table 1

solutions without the need for repeat lengthening.[25,26] Scoliosis correction and growth was maintained at follow-up; however, the rate of reoperations was high (17 of 24 patients) which indicates that this technique requires further improvement. Further longer-term investigations into the effectiveness, risk of revision surgery and complications are expected.

Definitive fusion: spinal fusion will result in loss of spinal motion and remaining longitudinal growth of the involved segments. In general, short segment fusions are reserved for local curative procedures to control congenital deformities and long fusions are considered the bail out for failed conservative and growth-friendly treatment methods. Fusion surgery should be delayed as long as possible to avoid shortening of the thoracic spine and chest, and thus cardiopulmonary compromise or thoracic insufficiency syndrome. If spinal fusion is performed to treat EOS in young children, it traditionally requires a combined anterior and posterior procedure. Posterior fusion alone can result in anterior overgrowth with worsening rotational deformity, also known as the crankshaft phenomenon. The use of segmental posterior instrumentation, especially with pedicle screws can provide better control across all three vertebral columns and this may obviate the need for anterior surgery.

Syndromic EOS

Indications for surgery

Treatment of syndromic EOS tends to follow the principles applied in children with an idiopathic or a neuromuscular scoliosis depending on the presence or absence of significant neurodisability associated with hypotonia, hypertonia or epilepsy. Specifically, in patients with disorders associated with joint hyperlaxity (e.g. Marfan, Ehlers—Danlos) bracing is generally ineffective to control the deformity. Children with a short trunk (e.g. achondroplasia) are not candidates for brace treatment because of their body size, whereas patients with osteogenesis imperfecta have greater risk of failure of ‘growth-friendly implants’ due to inherent bony weakness. Because of the high heterogeneity of presentation of syndromes associated with EOS, decisions on the risk-benefit of growth-friendly surgery in this population should be made on an individual patient basis.

Treatment options

Distraction-based methods, guided-growth, hybrid techniques and definitive spinal fusion.

Congenital/structural scoliosis

Background

The natural history of congenital scoliosis depends on the anatomical nature of the vertebral abnormality, the number of anomalies and their location across the spine (congenital defects in transitional areas of the spine are likely to produce a progressive deformity), as well as the remaining growth potential. The anticipated prognosis of congenital scoliosis based on these factors will determine the timing and type of surgical treatment. The congenital anomaly can be a failure of vertebral formation (most commonly a hemivertebra), failure of vertebral segmentation (most commonly a unilateral unsegmented bar) or both (mixed pattern). The risk of progression is highest in the presence of a unilateral unsegmented bar with contralateral hemivertebra at the same level. A structural compensatory scoliosis often develops at the levels above or below the congenital curve as an attempt of the spine to achieve global balance and this can be rapidly progressive (occasionally at a rate greater than that of the congenital scoliosis). Bracing cannot affect the congenital scoliosis but is indicated in the early treatment of the structural compensatory curve.

Indications for surgery

Surgical treatment is recommended for progressive curves or if the type of congenital anomalies predict a high risk for rapid curve deterioration.

Treatment options

Surgical options to address the congenital anomalies include limited *in situ* or corrective fusion (with or without hemivertebra resection), convex growth arrest (hemi-epiphysiodesis) and growth-friendly techniques. Thoracic insufficiency syndrome is the primary indication for expansion thoracoplasty (opening-wedge thoracostomy) using the vertical expandable prosthetic titanium rib (VEPTR).

A *failure of vertebral formation* due to a fully or semi-segmented hemivertebra produces asymmetrical growth of the spine and often a progressive scoliosis. Depending on the patient's age, this can be addressed by a convex growth arrest (hemi-epiphysiodesis) or a hemivertebra resection followed by a limited posterior fusion. The convex growth arrest procedure can be used primarily in the treatment of a congenital thoracic scoliosis due to a hemivertebra in children under the age of 5 years with a curve less than 50°. The technique has been initially described as a combined anterior and posterior growth arrest procedure. However, in the senior author's (AIT) experience this can be performed through an isolated anterior convex approach and involves removal of two discs and the adjacent end plates above and two below the hemivertebra combined with autologous rib strut bone grafting. This technique stops the deforming convex force that causes the curve and allows for continuous concave vertebral growth that can gradually improve the scoliosis during the remaining spinal development. The convex hemi-epiphysiodesis can be combined with the use of a growing rod that can control any residual deformity and stimulate concave vertebral growth to further correct the scoliosis.

In more severe curves, or when the hemivertebra is located in the lumbar or lumbosacral area below the conus medullaris, hemivertebra resection is indicated in patients with progressive deformities. This is a definitive treatment that allows for immediate spinal realignment and eliminates the cause of the deformity. The technique is best performed at the age of 2 years, as the hemivertebra can be easily identified and the spine is ossified to allow the use of instrumentation. A posterior hemivertebra resection is the technique of choice and this involves excision of the contralateral disc, as well as the end plates above and below the hemivertebra to allow curve correction and achieve fusion. In older patients with stiff and complex congenital deformities, an instrumented correction and fusion involving longer segments above and below the hemivertebra can produce correction through the adjacent to the anomaly levels. In the presence of severe congenital kyphosis or kyphoscoliosis (especially if this produces neurological compromise) there may be need for an additional posterior vertebral column resection followed by an instrumented fusion.

Surgical treatment of scoliosis due to the presence of a unilateral unsegmented bar with or without contralateral hemivertebra at the same level is indicated even for small curves as there is anticipated rapid progression with further growth of the spine. If the scoliosis is diagnosed early, it is in the cervicothoracic or upper thoracic spine and the residual deformity is acceptable it can be treated with an *in situ* posterior fusion with the use of bone graft. If the scoliosis affects the mid to low thoracic or the thoracolumbar spine an anterior convex fusion can halt curve progression by producing a block segment that has no growth potential. The technique is no different to the anterior convex hemi-epiphysiodesis described above. However, the difference is that in the presence of a unilateral unsegmented bar there is no remaining concave growth to partly correct the curve and therefore any additional improvement of adjacent level deformity can be achieved by the use of a growing rod construct with serial lengthenings.

A structural compensatory curve affecting adjacent levels to the congenital scoliosis that fails bracing can be treated in early age with TGR and MCGR. If the patient presents late with a rigid structural compensatory deformity this will have to be included in the instrumented correction and fusion of the congenital scoliosis.

Chest wall abnormalities

Growing rods are not an effective treatment method for patients with a primary chest wall abnormality the most common of which is congenital fusion of two or more ribs occurring usually on the concavity of a thoracic or thoracolumbar scoliosis adjacent to a unilateral unsegmented bar.[27] Structural restriction of thoracic and pulmonary growth can lead to thoracic insufficiency syndrome, causing inability of the thorax to support normal respiration and lung growth. This condition is associated with a high mortality in untreated EOS. Thoracic insufficiency syndrome is the primary indication for expansion thoracoplasty by using the vertical expandable prosthetic titanium rib (VEPTR) in immature patients.[28] Similar to traditional growing rods, this rib-based device expands the chest and thereby the space available to the lung by repetitive lengthening procedures. A recent review of the French VEPTR series (54 patients) reported unacceptably high complication rates of 137% per patient.[29]

Neuromuscular EOS

Background

The aetiology of neuromuscular EOS varies widely. The underlying neurological or muscular disease can lead to altered muscle tone, intraspinal anomalies, sensory impairment, hip dislocations, and pelvic obliquity. The more severe the degree of existing neurodisability, the higher the risk of progression and the severity of the spinal deformity. Severe neuromuscular scoliosis can lead to problems with sitting balance, hygiene and nursing care or cause cardiopulmonary compromise; the spinal deformity is a significant factor in decreased health related quality of life of these patients. Children with a lesser degree of neurological involvement, such as hemiplegic cerebral palsy or Friedrich's ataxia, can develop a spinal deformity that resembles idiopathic scoliosis, without major involvement of the pelvis. Decision making on the treatment of neuromuscular scoliosis in patients with complex co-morbidities and disabilities should be tailored to the individual needs of each patient. The presence of cardiorespiratory compromise will determine patient's life expectancy and guide treatment options.

Observation is recommended for children with small, flexible neuromuscular curves that do not cause functional impairment. Wheelchair modifications and seating adaptations can be effective for wheelchair bound patients to maintain a balanced posture and avoid pressure areas. Moulded seats may be contraindicated in growing children with neuromuscular scoliosis because their shape is constantly changing. The benefit of bracing for halting progression of neuromuscular curves is limited and this is mostly used as a temporary measure for postural support. In progressive neuromuscular EOS, treatment is based on maximum conservative measures during continuous growth, with the aim to maintain growth of the spine and development of the chest.

Indications for surgery

The potential benefits and risks of surgery before completion of growth must be carefully discussed with the multidisciplinary teams followed by shared decision-making with the caregivers. Surgery is generally indicated for severe deformities that produce back pain or costo-pelvic impingement, major functional or pulmonary impairment or challenges for nursing care due to the patient's poor posture. Scoliosis surgery can improve cardiopulmonary function, sitting balance, physical appearance and quality of life. However, in this medically vulnerable population, spinal surgery is considered a major undertaking with high associated risks. Techniques that require multiple repeat surgeries can have substantial morbidity and mortality in these patients. The natural history and specific morbidities of the underlying condition are essential to consider during treatment planning; for example, patients with Duchenne muscular dystrophy or congenital myopathies develop progressive cardiomyopathy and may not be able to tolerate scoliosis surgery at skeletal maturity. In those patients, the decision for scoliosis correction should be made early as at a younger age surgery is less complicated and safer.

Contraindications for surgery

Severe neuromuscular EOS is often associated with major comorbidities that may limit the treatment options and preclude spinal surgery. In addition, neuromuscular scoliosis patients can have extreme rigid deformities, with limited ability for correction. In patients in whom limited correction is expected, the risk of surgery may outweigh any potential benefits.

Preoperative work-up

A multidisciplinary assessment is critical as it will provide a comprehensive estimate of the risks of scoliosis surgery in the context of the patient's condition and existing medical comorbidities. The patients need to undergo a thorough anaesthetic, respiratory, cardiac and neurology review. It is vital that the patient's nutritional status is optimized before surgery and that the gastroenterology/dietetic teams are aware of the presence of gastroesophageal reflux, feeding disorders and gastrointestinal dysmotility. A clear plan should be made on postoperative enteral and/or parenteral feeding. Metabolic assessment may be warranted for patients with osteopenia or osteomalacia. A joint multidisciplinary decision can be made on the risk-benefit ratio and timing of surgery.

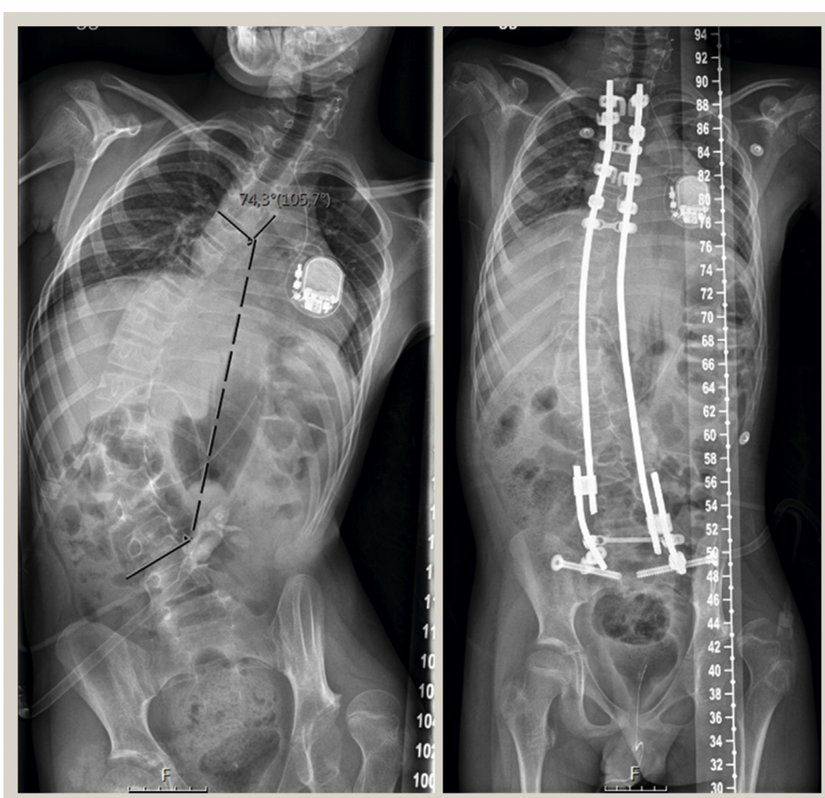

Figure 7 Example of a patient with early onset neuromuscular scoliosis who underwent a bipolar fusionless technique with good control of the coronal deformity and associated pelvic obliquity.

Treatment options

Surgical options to address neuromuscular EOS include previously described distraction-based techniques (TGR and MCGR). In neuromuscular scoliosis, growth friendly treatment options also include a fusionless bipolar construct and guided growth such as the modern versions of the Luque trolley technique.

If the underlying condition does not preclude repetitive lengthening procedures, a TGR can be successfully used in this subgroup of neuromuscular patients. There is limited literature on the use of growing rods, TGR or MCGR, in neuromuscular scoliosis. Chandran et al.[30] described 50% scoliosis improvement in 11 children with SMA type 1 and 2 without surgical complications or unplanned reoperations. Yoon et al.[31] reported a positive effect of MCGR on pulmonary function in 6 young neuromuscular scoliosis patients. McElroy et al.[32] recorded a 30% rate of deep-wound infection among 27 cerebral palsy patients after TGR procedures. Hanna et al.[33] described 12 SMA patients who underwent TGR lengthening without the need for a final fusion at completion of treatment.

Miladi et al.[34] introduced the concept of a fusionless bipolar system for neuromuscular patients (Figure 7). Similar to a TGR construct, it consists of a proximal and distal anchor connected by rods and parallel connectors. The concept is to perform a less invasive, fusionless surgery with the use of a solid construct that connects the anchor at the upper thoracic spine to a bilateral pelvic anchor. For end of construct fixation, the technique uses specific ilio-sacral screws distally and supralaminar and pedicle hooks proximally. This instrumentation can be used for repetitive lengthenings but it can also be left as a definitive construct in low-demand or wheelchair bound patients. Among 100 patients (5−21 years old) with neuromuscular scoliosis, the authors reported a mean scoliosis correction from 89° to 33° and a correction of pelvic obliquity by 17°. Seventy-two patients underwent one or two lengthening procedures during growth, while in the remaining 28 children the initial implant placement was their only surgery. There were 69 complications occurring in 26 of the 100 patients.

Conclusion

The treatment of EOS can be very challenging and must be tailored around the specific considerations of the type of deformity, the underlying aetiology, as well as the presence of medical co-morbidities. 'Growth friendly' surgery by means of growth stimulation or growth guidance is the preferred treatment modality in children with EOS. These techniques can be seen primarily as a temporizing measure in a growing skeleton in order to delay the need for a definitive spinal fusion for a later age while preserving spinal and chest development. ◆

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