Early-onset scoliosis: a narrative review

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- Early-onset scoliosis (EOS) is defined as any spinal deformity that is present before 10 years old, regardless of etiology.
- Deformity must be evaluated based on the intercorrelation between the lungs, spine, and thorax.
- Curvatures of early-onset have increased risk of progression, cardiorespiratory problems, and increased morbidity and mortality.
- Progression of the deformity may produce thoracic insufficiency syndrome, where a distorted thorax is unable to support normal respiratory function or lung growth.
- Management and treatment of EOS should pursue a holistic approach in which the psychological impact and quality of life of the patient are also taken into consideration.
- Growth-friendly surgical techniques have not met the initial expectations of correcting scoliotic deformity, promoting thoracic growth, and improving pulmonary function.

Introduction

Early-onset Scoliosis (EOS) is a term experts adopted to refer to pathologies of the growing spine that manifest and progress in different forms. Historically, its challenging nature has made it difficult to introduce a comprehensive definition that brings consensus in diagnosis and management. Initially, the term EOS was used by Ponseti and Friedman in 1950 to describe idiopathic scoliosis in patients less than 10 years old ([1]). They determined that those patients carried a worse prognosis when compared to older individuals. Subsequently, in 1954, James subdivided idiopathic scoliosis based on three periods of age at onset: infantile (≤3 years), juvenile (4–9 years), and adolescent (10 years to maturity) ([2]). However, in 1985, Dickson pronounced against the juvenile idiopathic scoliosis classification as those patients may correspond to the infantile subgroup ([3]). Years later, Dickson proposed that idiopathic scoliosis should be divided into early-onset (0–5 years) and late-onset (>5 years) ([4]). Despite the adoption of this definition in recent literature, it has been recognized that 'the treatment principles for children between the ages of 5 and 10 years more closely resemble those used for children under the age of 5 years than they do for children over the age of 10 years' ([5]). Consequently, the Growing Spine Study Group (GSSG) and the Children Spine Study Group (CSSG) defined EOS as any spinal deformity that is present before the age of 10, regardless of the etiology ([5], [6]).

Etiologies

EOS includes a varied set of patients with complex pathologies that affect their spinal curvature. In 2014, a group led by Vitale established a definition for EOS that considered etiology besides the age of onset. Their four categorical etiologies were idiopathic, neuromuscular, congenital, and syndromic ([6]).

Idiopathic refers to those deformities with no causal agents or associations with other diseases ([6], [7]). Their cause is unknown and presumably multifactorial, diagnosed when clinical and radiologic findings do not provide a definitive explanation ([7], [8], [9], [10]).

Neuromuscular deformities ensue due to an underlying neuropathic or myopathic disease that results in muscle tone imbalance without previous congenital or structural abnormality ([6], [7], [11]).

Congenital (i.e. structural) deformities consist of defects during vertebral development. Formation defects, segmentation, or mixed events lead to an asymmetric growth of the spine or thoracic cavity ([7], [12]).

Syndromic deformities are caused by clinically defined patterns (i.e. syndromes) associated with scoliosis and are

not directly attributed to congenital or neuromuscular causes (6, 7).

Classification

Classification systems allow characterization of a problem, suggest a potential prognosis, and offer guidance in determining the optimal treatment approach for a particular condition (13). Until recently, there had not been a standardized classification for EOS. Variability among treatment methods (14, 15) and the need for consistency in EOS research and literature prompted the development of a classificatory system (6).

It has been long established that age represents an important aspect in the progression and treatment of the EOS population. Nevertheless, as age alone did not provide adequate guidance for treatment, etiologies were included as part of the decision-making process. The standardized method contains a continuous classification prefix for age, three core variables (etiology, major curve, and kyphosis), and an optional modifier for curve progression (6). This classification system was based on the expertise of 15 experienced surgeons demonstrating a high level of agreement, consistency, and reliability in clinical and academic settings (16). However, the classification system has not yet been widely used in EOS literature, considering that it is relatively new. To some extent, its variables and cutoffs are based on expert opinion; therefore, it still requires further studies to validate its content (17). In 2020, Dragsted et al. found that the designed classification system had a substantial agreement for etiology. The reliability for major curve angle was excellent but somewhat lower and accurate for kyphosis and annual curve progression (17). They suggest a revision to the annual curve progression since its limit of agreement was larger than the 10° per year increments initially proposed by the classification (17). As new studies surge, further modifications could be expected to the classification, increasing its validation, reliability, and acceptance in literature to allow orthopedists to guide their EOS management.

Natural history – the importance of 10 years of age in the EOS cutoff

It is well known that in patients that EOS progresses, the earlier the onset, the worse the final curvature and its prognosis (18, 19). Progressive curves usually cause significant deformity, cardiopulmonary deterioration, and life-shortening (2, 20). In 1955, Scott et al. found that EOS progression is expected to occur at a rate of 5° per year (19). Thus, a child with an initial curve of 30° would be expected to have a major curve of approximately 100° at

around 14 years (19). Once the deformity progresses over 70°, if left untreated, it may progress to more than 100° worsening its prognosis (2). Pehrsson et al. found that patients with early-onset had a higher risk of developing severe scoliosis with increased morbidity and mortality compared to adolescent scoliosis (20). The increased mortality is explained by the structural changes that put constraints on the thoracic cavity leading to restrictive lung disease, cardiovascular complications, and respiratory failure (19, 20, 21, 22). Consequently, EOS needs to be assessed considering the intercorrelation between lung, spine, and thorax development to understand its negative impact on cardiopulmonary function.

Lung development is a crucial consideration in the history and progression of EOS. The most rapid growth of alveoli occurs during the first 8 years of life when they increase exponentially (23, 24) (Fig. 1). This finding contributed to establishing the cutoff for EOS to be 10 years instead of 5 years old. Limitations in the anatomic boundaries of the thorax reduce the volume early in life, negatively affecting the size of the lungs at maturity (25). Lung hypoplasia, where alveoli and arteries are few, has been observed in those patients in which scoliosis has
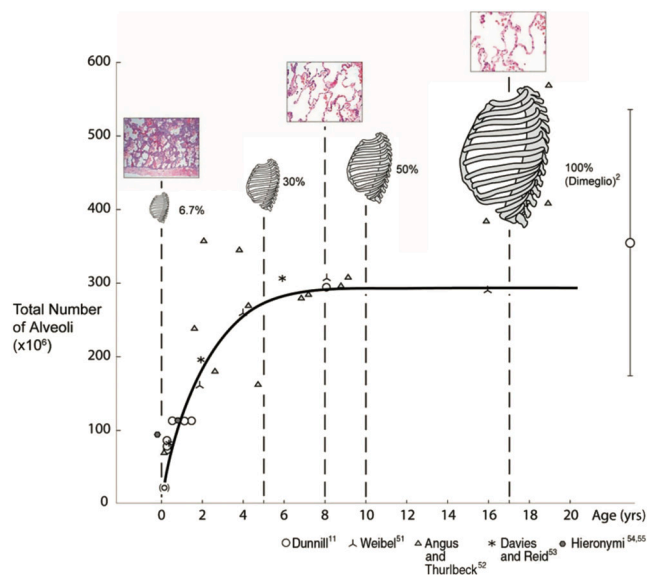
Figure 1

Graph shows the lung growth curve based on alveolar cell multiplication as a function of age. This summarizes several small autopsy series. The regression line was calculated by Dunnhill, (23) and the solid vertical line is the mean and range of the number of alveolar cells at maturity as reported by Angus and Thurlbeck . The synchronous thoracic volume increase is labeled as a percent of adult volume . Histographic sections of alveoli at the various stages of development are also shown. (Reproduced, with permission, from Campbell RM Jr, Smith MD. Thoracic insufficiency syndrome and exotic scoliosis. J Bone Joint Surg Am. 2007 Feb;89 Suppl 1:108-22; with permission from Wolters Kluwer Health, Inc.)

become prominent during this critical period of lung growth, often including emphysematous changes (26).

Spine growth is characterized by a synchronized growth that influences standing and sitting height, thoracic cage shape, volume and circumference, and finally, lung development (27). Measurement of the sitting height provides an indirect reflection of spinal growth and correlates strictly with trunk height (28). It occurs in a predetermined pattern of acceleration and deceleration events. Sitting height approximates 66% of the final sitting height from birth to 5 years, growing from 35 to 62 cm (27, 28, 29). Between 5 and 10 years, it increases by approximately 12 cm, while puberty is characterized by an additional 12 cm (27). In children with severe spinal deformities, loss of sitting height is related to the severity of the deformation (27).

Assessment of the T1-S1 segment becomes more relevant since many spinal deformities originate in this segment. The T1-S1 segment grows approximately 10 cm during the first 5 years of age (>2 cm/year), about 5 cm between 5 and 10 years (0.9 cm/year), and approximately 10 cm during puberty (1.8 cm/year). Thus, the TI-S1 segment's most rapid and greater total growth period occurs before 10 years of age (27, 29, 30).

As the spinal deformity progresses, it is crucial to understand that the thoracic cage growth is also modified. The thoracic spine provides the vertical component of the thoracic volume, whereas the rib cage contributes to the width and depth (31). Thoracic cage volume at birth is approximately 6% of its final size; at age 5, it reaches 30%, and by 10 years, it reaches 50% (27, 32). At birth, thoracic spine height measures 12 cm; at 5 years, it measures 18 and 27 cm at skeletal maturity (27, 30).

Karol et al. found a direct association between thoracic height and pulmonary function. The shorter the thoracic spine, the smaller the forced vital capacity (FVC) and the greater the likelihood of restrictive pulmonary disease (33). She found that to avoid severe restrictive lung disease (FVC <50%), thoracic height should measure greater than 22 cm at skeletal maturity (30, 33). Since we do not have the thoracic height necessary to avoid this respiratory disturbance at 5 years old, it became another reason to establish the cutoff for EOS to less than 10 years instead of 5 years old.

EOS – a common cause of thoracic insufficiency syndrome

It is appreciated that significant growth and development of the spine and thorax occur in a synchronized pattern early in life. Therefore, EOS produces structural changes in the multiple interrelated growth processes during a critical developmental period. Campbell et al. stated that a common denominator in patients with EOS is a small,

stiff, distorted thorax that could not provide volume for lung growth and thus functioned poorly. He called this manifestation thoracic insufficiency syndrome (TIS), defined as the inability of the thorax to support normal respiratory function or lung growth (25). Likewise, a deformed rigid chest cage causes lung and respiratory pathologies that can lead to a scoliotic heart, defined as right ventricular hypertrophy with or without congestive heart failure secondary to the increased pulmonary vascular resistance (22). As the spinal deformity progresses in EOS, it eventually interferes with lung development and cardiac function, leading to a potentially cor pulmonale and lethal TIS (27).

The diagnosis of TIS requires a multidisciplinary approach. As initially stated by Campbell (25, 31), assessment of the thoracic deformity and its progression is based on a comprehensive evaluation that includes physical examination, radiographs, CT, pulmonary function studies, and other tests of respiratory function. Attempts to recognize a specific finding to diagnose TIS have been documented (33, 34, 35). Through multiple studies, Ramirez et al. (35, 36), using thoracic and lumbar radiograph parameters, whole spine CTs, echocardiograms, arterial blood gases, and pulmonary function tests, did not identify a specific feature that could help diagnose primary and secondary TIS in EOS patients. Therefore, its diagnosis remains a combination of clinical, radiological, and respiratory function evaluations.

Psychologic burden and quality of life

EOS has significant physical and physiological implications, but in the same way, the psychological impact on the patient must be considered, which ultimately affects their quality of life. By itself, the natural progression of the physical deformity has esthetical consequences that influence self-esteem and other psychological aspects (37). In addition, these patients often need lifestyle adjustments to cope with disease limitations (37). When we consider those elements that add to the multiple medical visits, hospitalizations, comorbidities, and repetitive surgical interventions, it is easy to visualize their psychological impact (37). It has been shown that depression and anxiety are more prevalent in patients with EOS, along with dysfunctional areas of daily living (38). Moreover, those who develop TIS have reported lower quality of life scores than those with chronic conditions such as cardiac disease or malignancy (39).

A correlation between recurrent surgery during childhood and significant psychological pathology has been demonstrated (40). Due to the nature of the disease and its modern treatment modalities (i.e. growth-friendly procedures), EOS patients often undergo repetitive surgeries exposing them to the harmful effects of anesthesia

(41, 42), hospitalizations, surgical complications, lengthening procedures, and follow-ups that cumulatively produce chronic stress (40, 43). In one of the first reports about the psychological impact of growth-friendly surgery in EOS patients, John Flynn from Puerto Rico and colleagues found that 58% of individuals with repeated surgeries exhibited psychological dysfunction with trends toward positive correlations with depression, anxiety, and anger management (40). Similarly, Matsumoto *et al.* found an association with aggression, rule-breaking, and conduct problems. Those at risk of developing these lasting psychological problems were surgically intervened earlier in life, more significantly than undergoing multiple interventions (44). This supports the notion that delaying surgery is beneficial in limiting complications and allowing the body to mature while decreasing the negative psychological impact (44). However, dependence on the health care system, multiple trips to medical facilities, and disruption of daily routines contribute to the associated stress and psychological dysfunction (43).

Based on this information, we conclude that EOS is not just an orthopedic problem with cardiopulmonary implications. A multidisciplinary approach considering these patients' psychological impacts and needs is essential to provide the best possible outcomes in treatment. Access to mental health services should be prioritized as part of a holistic approach toward patient care, treatment outcomes, and quality of life.

Overview of treatment alternatives

Early spinal fusion was the treatment of choice for EOS, supported by the notion that a short straight spine was better than a progressing curve (45, 46, 47). Recent studies have demonstrated the importance of spine and chest growth to maximize thoracic volume and pulmonary function; thus, a shift toward growth-friendly treatments has emerged (25, 33). Growth-friendly alternatives comprise both non-surgical and surgical techniques. Non-surgical techniques include serial casting and bracing. Surgical procedures required the development of growth-friendly implants not previously known. In 2014, Skaggs *et al.* classified those growth-friendly surgical techniques as distraction-based, compression-based, and guided growth methods (48). Distraction-based implants include Traditional Growing Rods (TGR), Vertical Expandable Prosthetic Titanium Rib (VEPTR), and Magnetically Controlled Growing Rod (MCGR). Compression-based implants are Vertebral Body Stapling (VBS) and Vertebral Body Tethers (VBT). Guided growth systems include the Luque Trolley and Shilla techniques (48).

The 21st century has brought multiple advances in EOS, including an eruption of many of the treatment alternatives mentioned above. Nonetheless, other techniques used since many decades ago, such as Vertebral Column Resection (VCR) and Convex Hemiepiphysiodesis, still hold relevance and important value in specific circumstances and etiologies. It is important to note that the use of each treatment alternative will be based on the patient's characteristics, the surgeon's expertise, and equipment availability.

Conservative management

Casting

The use of casting for spinal curve correction has been described as early as 1863 by Bradford *et al.* (49) in a detailed description using plaster jackets for correction through mechanical and external forces (49). More recently, while the methods vary, the long-abandoned Mehta casting has increased recognition. Mehta *et al.* stated that growth in EOS could be harnessed through casting to straighten progressive curves that would otherwise advance into severe deformities (50). The technique carefully applies and molds a plaster jacket on a Cotrel frame with a head halter and pelvic traction. This method, also known as elongation-derotation-flexion casting, provides a three-dimensional correction force that counteracts the scoliotic deformity. Casts are then changed every 2–3 months with a clinical and radiological assessment to evaluate progression, followed by observation or bracing (50, 51, 52). In 2005, Mehta *et al.*, in a prospective study, catalyzed the resurgence of serial casting as a treatment alternative by reporting a 100% curve correction in 94 children if they were referred early in the disease and had a moderate curve (50). For those referred at greater than 2 years with greater curve progression, the curve could be reduced but not reversed (50). Risks and complication rates were low, most related to skin breakdown and irritation, musculoskeletal discomfort, and cast intolerance (53, 54). It is important to note that the non-idiopathic population has not shown the same promising results of significantly reducing the morphologic deformity; nevertheless, it has delayed surgical treatment (50, 55).

Bracing

The effectiveness of bracing in adolescent idiopathic scoliosis has been encouraging, as it is currently recommended in patients with curves from 25 to 40° (8, 56). However, the effectiveness of bracing in the EOS population is still under debate (57, 58, 59, 60). Recent studies by Thometz *et al.* (58, 60) showed favorable results. They developed an elongation-bending-derotation brace to apply the same force as the serial casting technique without many drawbacks. Bracing has demonstrated not only stabilizing and decreasing curvature progression but also correction in up to 44% of patients. Also, a decrease in

progression of the curve of 5° or less was observed in 67% of patients with a median Cobb angle reduction of 15° ([58], [59], [60]). Bracing might be more convenient than casting because it can be removed as desired, but casting provides a continuous corrective force. While bracing might be an effective treatment, further studies are needed to compare bracing to casting in this population ([61]).

Surgical management

Distraction-based systems

Distraction-based systems correct the spinal deformity by using anchors as a mechanical distractive force exerted on the spine segments, ribs, or pelvis ([48], [62]).

TGR

TGR has been the most applied technique for treating EOS ([61], [63]). The usage of rods as a distraction system to correct scoliosis instead of spinal fusion was first described by Harrington in 1962 ([64]). The technique was later modified to a 'subcutaneous rod' by Moe *et al.* ([65]) with unsatisfactory results and high complication rates ([65], [66], [67]). Akbarnia *et al.* ([68]) demonstrated better results implementing the dual growing rod technique instead of a single rod. However, TGR requires repetitive surgical lengthening contributing to the complications associated with this technique ([69], [70]). Multiple reports have shown a high rate of complications, with up to 58% of patients having at least one complication ([68], [70], [71], [72]). Implant failure, surgical site infections, and wound healing problems are common complications of this procedure, while exposure to repeated anesthesia remains an important concern ([61], [63]).

The literature demonstrates TGR's effectiveness in deformity correction and spinal growth, yet further studies are needed to assess its efficacy in pulmonary improvement and health-related quality of life ([43]).

VEPTR

The VEPTR system consists of a titanium alloy longitudinal rib distraction device ([73]). It was first described by Campbell *et al.* ([73], [74]) to treat TIS associated with congenital scoliosis and fused ribs. The ribs are used as anchor points in addition to spine or pelvis anchor points at the base of the construct to address the three-dimensional thoracic deformity with or without expansion thoracoplasty ([48], [62], [73], [75], [76]) ([Fig. 2]). Initial studies showed correction of Cobb angles, increment in the space available for the lungs, and total spine growth ([74], [77], [78]).

On the other hand, throughout the history of this instrumentation, subsequent studies have not been able to support the initial findings. For example, objective measures
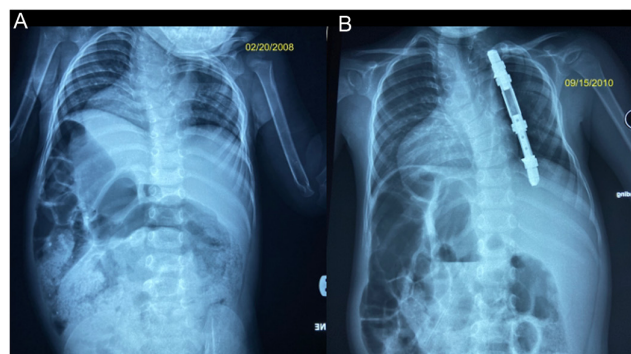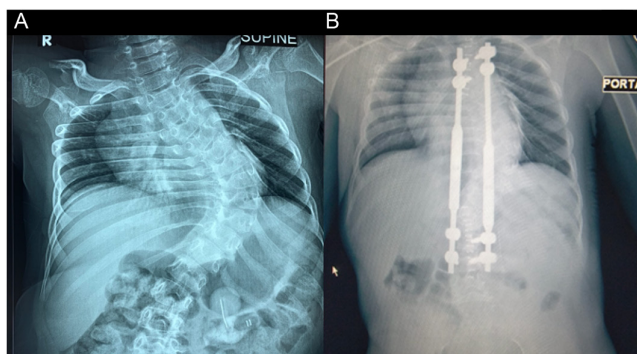

Figure 2

VACTERL syndrome patient's radiographs Pre (A) and Post (B) Vertical Expandable Prosthetic Titanium Rib (VEPTR) Implantation.

of pulmonary function have not shown the expected improvements, and further studies are required to better assess the benefits in this area ([62], [79], [80], [81]). Other studies have not demonstrated an adequate correction of Cobb angle nor growth improvement. More recently, a study of 15 years by Ramirez *et al.* ([82]) presented at the American Pediatric Academy confirmed that respiratory function did not improve, growth of the spine was moderate, and Cobb angle correction was not as expected. Lastly, complication rates have been significant, with failure of proximal fixation being the most common ([77], [80], [83], [84], [85]). Campbell *et al.* ([74]) reported a 163% of complications in patients with congenital scoliosis, Hasler *et al.* ([77]) a 100% in patients with non-congenital scoliosis, and Ramirez *et al.* ([86]) up to 73.1% in neuromuscular scoliotic patients. Thus, the benefits of the VEPTR have been questioned, and when considered, the consensus is to be cautious and use a multidisciplinary approach ([62], [81]).

MCGR

MCGR consists of a distraction-based implant designed to minimize the problems associated with TGR ([48], [87]) ([Fig. 3]). The concept first introduced by Takaso *et al.* ([88]) found MCGR to be an alternative treatment with comparable results to TGR without the expected complications ([89], [90]). Akbarnia *et al.* showed that MCGR had similar results to TGR in major curve correction and comparable spinal and thoracic height ([89]). MCGR was proposed to reduce the number of planned surgical interventions by avoiding repeated open lengthening procedures, thus decreasing the rate of complications ([89], [91], [92]). Nevertheless, unplanned surgical revisions because of complications remained a concern ([89], [91]). While there is evidence that MCGR reduces the risk of deep wound infections and surgical site infections ([91], [93], [94], [95]), a recent study by Teoh *et al.* ([95]) suggests that the likelihood of having metalwork problems in MCGR is 4.67 times the risk of

Figure 3

Syndromic early-onset scoliosis (EOS) patient's radiographs Pre (A) and Post (B) Magnetically Controlled Growing Rod (MCGR) Implantation.

conventional growing rods. They also found a higher complication rate than conventional growing rods. In 2018, a systematic review of 15 MCGR studies demonstrated that despite achieving curve correction, growth complication rates were reported to be 44.5%, with 33% unplanned revision rates (96). Regarding the psychological effects of multiple surgeries on patient mental health, Aslan et al. (61, 97) found no improvements with MCGR. The noninvasiveness of the procedure does not appear to provide the expected advantages in the psychological and health-related quality of life compared to TGR (43, 97).

Compression-based implants

Compression-based implants (i.e. VBS and VBT) correct the spinal deformity by modulating growth through the application of a compressive force to the convex side of the spinal curve inhibiting its growth while it allows the development of the concave side of the spine. The technique follows the Hueter-Volkmann principle that proposes that physeal growth can be retarded through a compressive mechanical force and promoted by decreasing mechanical load (48, 98).

VBS

VBS involves placing metal staples to induce an asymmetric growth plate inhibition to correct the spinal deformity (87, 99). Clinical results of VBS were presented as early as 1954 but had poor outcomes at the time (100). More recently, Betz et al., in a 2-year follow-up study, showed successful results in correcting thoracic curves less than 35°s. Still, other alternatives were recommended for those with a deformity greater than 35° (101). Traditionally, immature moderate curves have been treated with bracing; however, whenever bracing is no longer an option, VBS can be considered (56). Current indications for VBS are a diagnosis of idiopathic scoliosis, Risser stage 0–2, coronal

deformity of 25–40° , and bracing failure (102). In 2018, Cahill et al. demonstrated that VBS effectively prevented progression and fusion in thoracic and lumbar curves with mean Cobb angles of 29.5 and 31.1°respectively (103).

VBT

VBT represents the most recent compression-based implant first described by Crawford et al. in 2010 (104). It consists of thoracoscopically placing anterior vertebral body screw anchors with a tightened flexible tether between them (104, 105) (Fig. 4). In 2015, Samdani et al. (106) reported thoracic curve correction in 32 patients from 42.8 to 21.0° on the first erect radiograph and 17.9° at the latest follow-up. In 2020, Hoernschemeyer et al. (107) presented the results of a study in patients with skeletal immaturity in which VBT achieved a 74% success rate in attaining curve magnitudes less than 30° at skeletal maturity.

At the moment, recommendations for the usage of these techniques are for patients around 9–10 years with growth remaining (10). Many of the concerns associated with compression-based implants are related to overcorrection of the curve typical in immature patients, surgical approach complications such as pulmonary problems, bowel injuries, implant failure, lack of long-term results, and studies discrepancies in rates of complications or adverse events (105, 106). Considering that VBS and VBT are recent techniques for treating spinal deformity, the literature is limited. Further studies are required to
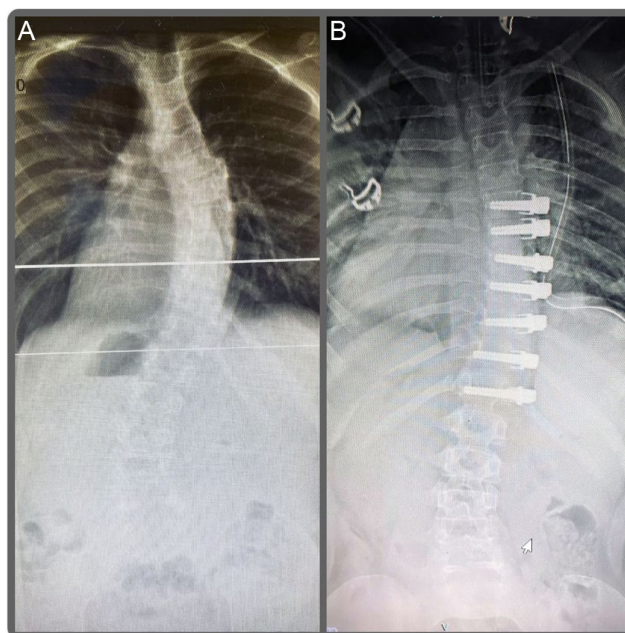
Figure 4

Idiopathic early-onset scoliosis (EOS) patient's radiographs Pre (A) and Post (B) Vertebral Body Tether (VBT) Implantation.

assess its effectiveness, safety profile, and age at which patients will benefit the most.

Guided growth systems

Guided growth systems correct the spinal curvature by using implants that anchor, fixed and non-fixed, multiple vertebrae to rods to direct the growth of the spine along with the spinal implant (48). The main benefits of this technique are that it avoids common concerns in distraction-based systems such as infections, hardware failure, repeated surgical lengthening, or exposure to anesthesia (48, 62).

Luque trolley technique

Luque trolley technique uses sublaminar wires to fix rods segmentally to the spine with an approach that limits subperiosteal dissection to avoid unintentional spinal fusion (8). However, this technique is not routinely used due to documented spontaneous spinal fusion, limited spinal growth, and control of the spinal deformity (108, 109). More recently, a Modern Luque Trolley system has been proposed, with Ouellet *et al.* publishing a 5-year retrospective study of five patients whose correction of the primary curve was from 60 to 21° and maintained after a 2-year follow-up (110). Concerns regarding spinal fusion may remain, but the study reported fewer implant failures when compared to the original trolley results (110).

Shilla technique

The Shilla technique developed by McCarthy and colleagues was first reported in 2014 (111). As a relatively new procedure, it maintains the same growth principles of guided growth systems by fixing dual rods with pedicle screws to the curve's apex with proximal and distal gliding screws placed with minimal subperiosteal dissection to avoid spontaneous fusion at those segments (48). Current indications include a failed course of bracing and coronal curves greater than 50° (111). In 2015, McCarthy *et al.* presented the results of 40 patients with a minimum of 5 years of follow-up in which preoperative curves were 69° on average, 25° following index procedures, and 38.4° at the most recent follow-up or prior to definitive spinal fusion (112). In 2017, Luhmann *et al.* (113) radiographically compared the outcomes of the Shilla technique and TGRs. They found that the curves with preoperative mean values of 61 and 65° between the two groups at the latest follow-up had corrected to 27 and 29°, respectively. Growth of the T1-T12 segment increased 4.6 cm for the Shilla technique and 5.2 cm for the TGR. They eventually concluded that both methods were compared favorably with radiographic outcomes for growth, curve correction, and complications; however, one distinguished difference was the threefold decrease in overall surgeries with the Shilla technique (113). While there is an advantage in overall surgeries, complication rates remain a concern as it is with MCGR and TGR. They have been reported as high as 73% requiring return to the operating room due to wound infections, spinal alignment, or implant issues (112).

Other alternatives

Vertebral column resection

Vertebrectomy for severe scoliosis was first described by MacLennan in 1922 (114). The technique has improved since, with VCR currently consisting of a three-column circumferential osteotomy involving the vertebral body, intervening disks, pedicles, and all the dorsal elements to create a segmental defect with sufficient instability to require provisional instrumentation (115, 116, 117). Current indications include short angular deformities when other methods are not technically possible, typical of congenital scoliosis (CS) or congenital kyphosis at an early age (63, 115). The most common pathology for CS is hemivertebrae, with a known poor prognosis (118). Hemivertebrae resection has become the gold standard for CS caused by hemivertebrae with great results in terms of curve correction (61, 119). Traditionally, it was performed through an anterior-posterior approach, but due to extensive operative times, significant blood loss, and high complication rates, more recently, a tendency toward a posterolateral approach has been observed (115, 116, 119). Despite improvement in operation times and blood loss with a posterolateral approach, concerns remain about its technical difficulty, blood loss, and high complication rates, especially neurologic complications. The technique, when considered, should be performed prudently and by an experienced surgical team (116).

Convex hemiepiphysiodesis

Convex hemiepiphysiodesis, also known as convex growth arrest, used to be one of the most used techniques in the treatment of congenital scoliosis in children (120, 121). The technique consists of arresting spinal growth on the convex side of the curve, allowing the concave side to grow and slowly straighten the spine. Commonly used for multilevel congenital deformities, it was considered a safe, simple, and efficacious technique but with an unpredictable method of guiding and modulating spinal growth (120, 122). Primarily used for congenital scoliosis, the technique was ineffective for other types of EOS (62, 123).

Future directions

EOS treatment currently emphasizes the management of the spinal and thoracic cage deformity, improvement of cardiopulmonary dysfunction, psychological impact, and

health-related quality of life parameters (25, 44, 124, 125). Clear advances have been made in understanding EOS's natural history, progression, and long-term consequences. In the same way, during the past few decades, technological innovations have also increased the treatment alternatives for EOS patients (8, 48). However, despite progress, EOS remains a treatment challenge, evidenced by the lack of consensus between the experts (126). Significant expansion of treatment options has outpaced evidence-based literature, contributing to the uncertainty in management (127). Also, besides the rapid treatment expansion, there are multiple obstacles to retrieve high-level evidence, including the small and heterogeneous patient population, necessary long periods of follow-ups, and difficulties in assessing pulmonary outcomes in this population (10, 127). While there is evidence of treatments' effectiveness in curvature correction and spinal growth, concerns remain about these techniques' effects on improving pulmonary function (25, 128). It has been demonstrated that traditional radiographic studies do not adequately reflect outcomes in EOS respiratory function (10, 128). The development of imaging techniques that provide three-dimensional dynamic measurements along with other modalities to assess pulmonary outcomes in treated children holds promise in improving the assessment of the spine–thorax pulmonary relationship (10, 127, 128). Improvements in these areas will provide a better understanding of how to avoid the ultimate manifestation of TIS that leads progressive scoliotic patients to cardiopulmonary compromise (25).

Continued efforts to improve these concerns allow for an optimistic approach toward EOS management in the future. The necessity of a standard and efficient approach for EOS research has been recognized. Therefore, multicenter study groups such as the GSSG, CSSG, and Pediatric Spine Study Group have been established to increase research consistency and reliability (10, 127).

Lastly, tools such as the 24-item Early-Onset Scoliosis Questionnaire developed by Matsumoto *et al.* will provide a high-quality, consistent assessment of the impact of each treatment technique on the health-related quality of life parameters and its psychologic effects (124).

Conclusion

EOS refers to pathologies of the growing spine with multiple etiologies and manifestations. Treatment should be focused on correcting spinal and thoracic cage deformity, improving respiratory function and health-related quality of life parameters. Future efforts should be guided toward technological and treatment refinement, improve modalities for pulmonary function assessment, high-level clinical research, and improve patients' psychologic health and quality of life. While it is

a condition that we have abundant knowledge, there is no definitive solution in terms of treatment.

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