Early-onset scoliosis – Current treatment

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Abstract

Early-onset scoliosis, which appears before the age of 10, can be due to congenital vertebral anomalies, neuromuscular diseases, scoliosis-associated syndromes, or idiopathic causes. It can have serious consequences for lung development and significantly reduce the life expectancy compared to adolescent scoliosis. Extended posterior fusion must be avoided to prevent the crankshaft phenomenon, uneven growth of the trunk and especially restrictive lung disease. Conservative (non-surgical) treatment is used first. If this fails, fusionless surgery can be performed to delay the final fusion procedure until the patient is older. The gold standard delaying surgical treatment is the implantation of growing rods as described by Moe and colleagues in the mid-1980s. These rods, which are lengthened during short surgical procedures, can avoid the progression of scoliosis and may bring more benefits. The technique of growing rods is based on the principle that preferentially attach to the ribs to specifically address chest wall and spine dysplasia. The second category of surgical devices consists of rods used to guide spinal growth that do not require repeated surgical procedures. The third type of fusionless surgical treatment involves slowing the growth of the scoliosis convexity to help reduce the Cobb angle. The indications are constantly changing. Improvements in surgical techniques and greater surgeon experience may help to reduce the number of complications and make this lengthy treatment acceptable to patients and their family. Long-term effects of surgery on the Cobb angle have not been compared to those involving conservative “delaying” treatments. Because the latter has fewer complications associated with it than surgery, it should be the first-line treatment for most cases of early-onset scoliosis.

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1. Introduction and definitions

In some cases, scoliosis that appears after puberty progresses – despite conservative treatment – into deformities that are more significant as the patient is younger (Fig. 1) and in most cases will require spinal fusion. Their progression at the same time as the development of pulmonary alveoli can seriously alter respiratory function. During this period, spinal fusion must be avoided so as to prevent this restrictive lung disease from getting worse due to thoracic growth arrest [1]. This contradiction between the need to correct the scoliosis and the impossibility of performing fusion has led to severe progressive scoliosis that occurs before 10 years of age being defined as “early-onset scoliosis” (EOS), with the goal of bringing specific answers to the therapeutic challenges brought on by these deformities.

The progression mechanism for EOS varies depending on its form and etiology: congenital anomalies, neuromuscular diseases, scoliosis-associated syndromes (neurofibromatosis), or idiopathic causes. Congenital malformation conditions such as asphyxiating thoracic dysplasia (Jeune’s Syndrome) and spondylolocostal dysostosis (Jarcho-Levin Syndrome) require a specific and very challenging treatment.

The main treatment options for EOS will be described in this review.

Although conservative treatment, by definition, does not stem the progression of these deformities, the available surgical alternatives can lead to many complications and have not been shown to be better in terms of the final outcome once growth is finished. Conservative treatment is well-known and has been used for a long time. It will only be touched on briefly in this work, but

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it remains the first-line treatment, pending the age of arthrodesis, because of its favorable benefit/risk ratio.

2. Spinal and pulmonary growth and thoracic insufficiency syndrome

The lungs grow in a non-linear manner over time (Fig. 2); the alveolar–capillary proliferation reaches its peak between 0 and 2 years of age and ends near 8 years of age; the volume of the bronchial tree increases as the child grows [2]. Any deformity and loss of flexibility in the vertebra–rib–sternum complex triggered by the progression of scoliosis alters the dynamic capacity of the respiratory system and negatively affects the development of the alveoli in terms of their number and volume [3]. This restrictive lung disease can evolve into pulmonary arterial hypertension, which itself is responsible for right heart failure or pulmonary heart disease in adults that can be life threatening early on [3,4].

The relationship between growth and respiratory function was described at length by Dimeglio and colleagues [5,6] and underlined by the work of Karol et al. [7], who showed a direct correlation between the results of respiratory function tests and the height of the T1-T12 segment measured on skeletally mature patients who had been operated on as children for congenital scoliosis. The vital capacity was reduced more than 50% when spinal fusion was performed over more than 60% of the thoracic spine before 8 years of age.

Congenital and dystrophic scoliosis patients have reduced thoracic compliance, which explains why the respiratory tolerance to deformity is lower than in patients with idiopathic scoliosis for the same Cobb angle [8]. Respiratory problems are also at the forefront of neuromuscular scoliosis with respiratory failure that is proportional to the Cobb angle and to trunk collapse as evidenced by the T1-T12 distance.

Evaluating lung function is often difficult in children under 7–8 years of age. Emans [9] has shown that the width of the pelvis and thorax are correlated to the theoretical height of the thoracic spine, which makes it easier to monitor the theoretical thorax size during growth (Fig. 3). In addition to performing a pulmonary function test, regular monitoring of the T1-T12 height (Table 1) is a good proxy for the seriousness of the situation and the effects of

From [10] with permission.
treatment. This distance must be greater than 20 cm at skeletal maturity to avoid severe restrictive lung disease [7].

These concepts of restrictive lung disease and scoliosis have recently been named “thoracic insufficiency syndrome” by Campbell and Smith [10] (Fig. 4). This condition occurs when the thorax no longer allows normal breathing or harmonious lung growth. This description has brought the significant respiratory problems associated with early-onset scoliosis back to the forefront.

Although surgical expansion thoracoplasty has been shown to increase the chest volume [11], others have shown reduced lung function due to a less compliant thoracic cage because of the stiffening cause by the thoracic instrumentation [12,13]. Thus reducing the Cobb angle does not always result in better respiratory function.

Table 1
Change in the T1–T12, L1–L5 and T1–S1 distances during growth [5].

<table>
<thead>
<tr>
<th></th>
<th>Birth</th>
<th>0 to 5 years</th>
<th>5 years</th>
<th>5 to 10 years</th>
<th>10 years</th>
<th>During puberty</th>
<th>At maturity</th>
</tr>
</thead>
<tbody>
<tr>
<td>T1–T12</td>
<td>11–12 cm</td>
<td>+1.3 cm/year</td>
<td>18–19 cm</td>
<td>+0.7 cm/year</td>
<td>22 cm</td>
<td>+1.1 cm/year</td>
<td>26–28 cm</td>
</tr>
<tr>
<td>L1–L5</td>
<td>7 cm</td>
<td>+0.7 cm/year</td>
<td>10.5 cm</td>
<td>+0.4 cm/year</td>
<td>12.5 cm</td>
<td>+0.7 cm/year</td>
<td>16 cm</td>
</tr>
<tr>
<td>T1–S1</td>
<td>20 cm</td>
<td>+2 cm/year</td>
<td>30 cm</td>
<td>+1 cm/year</td>
<td>35 cm</td>
<td>+1.8 cm/year</td>
<td>43–45 cm</td>
</tr>
</tbody>
</table>

The distance is measured from the superior endplate to the inferior endplate of the vertebrae in question.

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Fig. 3. Estimate of the theoretical thoracic spine height by measuring the thorax and pelvis width.

From [9] with permission.

Fig. 4. Thoracic insufficiency syndrome.

Fig. 5. EDF casting under anesthesia using a Cotrel frame; very light traction is applied.

3. Treatment methods

3.1. Conservative treatment

Conservative (non-surgical) treatment and its limitations are well-known when it comes to neuromuscular and congenital
scoliosis. For the former, the treatment has little effect on the progression of the scoliosis but is often needed to help the patient maintain a satisfactory seated position in cases of significant hypotonia and maintain effective chest expansion. The Gancher brace [14] remains the preferred treatment starting at a very young age.

Although these braces do not stop the progression of the spinal congenital deformity, they can be useful for controlling the counter-curves that can develop by themselves.

For syndrome-associated and idiopathic scoliosis, conservative treatment is still relevant. The time consecrated to it should be proportional to the prognosis of EOS, particularly for infantile scoliosis. The Milwaukee brace has long been considered the gold standard treatment for young patients, but in our eyes, it does not effectively stabilize many scoliosis conditions and it is poorly tolerated by patients. It is now possible to use more effective braces right away, namely an adjustable multi-shell brace, which has sufficient modularity to adapt to growth and not impede rib cage development.

Use of a plaster cast [15,16] is still the best way to gradually correct progressive infantile scoliosis. This treatment helps to interrupt the vicious circle of self-aggravating scoliosis (Hueber-Volkmann law) and brings the spinal column into a mechanical environment that can be conducive to spontaneous correction (Frost law). Casts can be made under general anesthesia (Fig. 5) so that the child is fully relaxed; no traction is used so as to avoid neurological complications. Premedication can be used to avoid anesthesia-related complications but requires effective distraction methods to reduce the stress that such a procedure can cause to a child. These procedures require that an MRI be performed beforehand to eliminate any abnormalities in the central nervous system. The casts typically have windows and are changed every 2 months until the best possible correction has been obtained (Fig. 6). This intensive treatment requires buy-in from a dedicated team, the child and the child’s family to avoid progression that would require surgery in the medium term.

3.2. Surgical methods

Surgical methods can be divided into three broad categories [17]: growing rods that apply distraction force to the spine and/or ribs, guided growth systems that keep the spine in its reduced position without restricting its growth, and compression-based systems that apply compressive force to the convexity of the curve to inhibit its growth.

3.2.1. Growing rods

The technique first described by Moe and colleagues [18] consisted of placing rods in the concavity of the curve (Fig. 7) and exposing the spine only at the ends of the construct and then making with first correction by distraction during the implantation. This requires regular additional surgical procedures to lengthen the rod (Fig. 8) so as to maintain the result obtained during the index surgery and to follow the growth of the spine, which is evaluated by measuring the T1-S1 distance and comparing it to the Dimeglia growth curves (Table 1). Growing rods are now considered the gold standard thanks to the work of Akbarnia and others [17,24].
Fig. 9. Lengthening of a Magec® rod in the office; the amount of lengthening is controlled externally.

The rods can now be lengthened in a non-invasive manner (Fig. 9) through a magnetic mechanism [19–21]. This increases the frequency and progressiveness of lengthening, thereby reducing the risk associated with this surgery and increasing its tolerance and effectiveness. The recently developed Magec® system [21] (Fig. 10) is currently being evaluated [22,23]; its preliminary results are more promising than those of the Phenix® system [20], which was plagued by lock-up of its internal mechanism. However, the follow-up is still very short and the device’s technology does not guard against the risk of gradual stiffening of the spine between lengthening session and the possibility that the magnetic distraction force will not be able to overcome the scoliosis-related stiffness after one or two years of use.

Growing rods satisfy their goal of stabilizing the curve and the growth of the T1-S1 segment [24] but are also fraught with complications [25–28]. The frequency of these complications varies from one study to another, but increases linearly with the number of procedures performed [25]. The percentage of unscheduled surgical procedures needed to treat these complications is a good indication of their frequency and severity.

3.2.1.1. Complications.

3.2.1.1. Mechanical complications. Breakage of the rod (Fig. 11) occurs in at least 15% of cases; it mainly occurs near the connection points or an area where the rod is greatly bent, especially in cases of hyperkyphosis; the rod is more likely to break if its diameter is too small and only one rod is used [28].

Dislodging of the implants occurs at the upper end of the construct in 95% of cases (Fig. 12). It is more likely to occur in hyperkyphosis and can be prevented by using solid fixation over two or three vertebrae that is reinforced with local bone graft application. Screw application seems to result in the most solid proximal
anchoring [29] but the neurological complications can be horrendous if the screw fixation fails [30]. This complication has only been observed in constructs secured by a single screw. There is currently no consensus as to the ideal superior anchoring method. Most surgical teams anchor the inferior end of the rod by screwing it into two adjacent vertebrae. The anchoring points must be located such that the rod will be as parallel as possible to the support vertebrae at that level, so as to reduce the stresses and prevent induction of a fixed angle, which will not allow progressive correction with each distraction (Fig. 13). The inferior end is typically placed on a stable vertebra; preoperative radiographs of the spine under traction can be used to better define this level.

There is no evidence that a brace will help to reduce the risk of rod breakage or anchor failure.

3.2.1.1.2. Deterioration of spinal balance. The tension placed on the two ends of the rods at the junction between a rigid and flexible segment causes unwanted deviations of the spinal column, which progress with each lengthening session. The most common one is junctional kyphosis of the upper end of the construct (Fig. 14). This can be prevented by using a construct with good coverage and extended fixation with descending upper hooks [31], using screws [25] instead of hooks, which have an excessively large posterior moment arm, and by careful preoperative planning which allows the instrumentation to be placed on the most balanced part of the spine, and to be secured proximally relative to the top of the kyphosis. No study has specifically evaluated the repercussions of surgical treatment for early-onset scoliosis on the sagittal spinal balance.

Using dual growing rods as recommended by Akbarnia et al. [24] helps to reduce the mechanical complication rate, however it increases the number of revision procedures for subcutaneous impingement and the risk of spontaneous spinal fusion due to the stiffness of the construct.

Placing fusionless instrumentation can create a crankshaft effect if lengthening is not carried out often enough or to a sufficient degree [32]. The ideal time frame for lengthening is every 6 months [33], but often the amount of lengthening that can be accomplished is reduced as more procedures are done due to ankylosis of the spine [34].

3.2.1.1.3. Infection. Mackenzie et al. [35] reported a 6.7% infection rate in their patients, with 69% of cases requiring surgical revision. The frequency of infections increases as more surgical procedures are performed [25]. The rods must be placed under the muscle layers to avoid subcutaneous impingement, which is most common in young, hypotrophic patients. In our opinion, it is better to make one long incision (Fig. 15) to achieve the optimal rod position than to make multiple small incisions at the anchoring points and inserting the rod blind. One patient has died due to intra-thoracic false trajectory [36].

3.2.1.1.4. Neurological complications. No neurological complications have been reported during rod lengthening. Intra-operative monitoring has mainly been recommended during the implantation or changing of rods or instrumentation [37]. Although there are no recommendations for systematic monitoring during lengthening procedures, there is agreement on the need to be vigilant and
screen at-risk patients by systematically performing a spinal cord MRI at the start of the treatment.

3.2.1.5. Psychological and social complications. Acaroglu et al. [32] report that the total duration of hospitalization was 101 days between the first surgical procedure and the final fusion, with an average of 4.6 lengthening sessions being performed by patient. These results provide evidence of the burden of scoliosis care and the possible psychological and social consequences for the patients and their families [38].

3.2.1.2. Intercostal and costovertebral distractors. These are growing rods with one or two support points on the ribs. The most well-known device is the vertical expandable prosthetic titanium rib (VEPTR). This is a growing rod with two telescopic parts that are spread apart gradually at each surgical lengthening procedure (Fig. 16). It was designed by Campbell to treat thoracic insufficiency syndrome related to congenital chest wall and spine deformities [11] such as unilateral thoracic hypoplasia due to missing ribs or synostosis (VATER syndrome) or bilateral thoracic hypoplasia (Jeune's and Jarcho-Levin Syndromes) [39]. The VEPTR is positioned using a specialized anchoring system around the ribs bordering the deformity to maintain the synostosis resection area open and increase the thoracic volume. The VEPTR has been shown to have the ability to correct any associated spinal deformities in parallel, which has broadened its indications for use.

The numerous complications reported for this device [39] are mainly related to its size and the fragile nature of the patients targeted for this treatment. Along with the complications associated with standard growing rods, there are more infections, local muscle atrophy and brachial plexus compression related to migration of the fixation on the first rib. Its ability to improve respiratory function has been called into question [12,13]. The alternative is to construct spine–rib or rib–rib assemblies with standard rods and implant, which are less bulky, by using specific rib anchoring systems available from certain manufacturers.

3.2.1.3. Final fusion after growing rods. This procedure has not yet been standardized, but it is performed when sufficient growth has occurred or if the complications are too frequent or severe to continue using the growing rods. This procedure is challenging because of stiffening of the vertebral column and presence of autofusion areas in the spine away from the anchoring points [40], which requires posterior osteotomy procedures. Some authors

![Fig. 16. A. Chest wall and spine dysplasia (coll. P. Violas). B. After VEPTR lengthening sessions (coll. P. Violas).](image)

![Fig. 17. Shilla technique. The rods are secured to the apex of the deformity, which is fused. The vertebrae at the boundaries of the scoliosis migrate during growth because of special screw heads that slide freely along the rod.](image)
have removed the growing rods without performing a fusion and have noted stiffness in the area bridged by the instrumentation [41].

3.2.2. Growth guidance systems
These are surgical devices that are positioned along the vertebral column to guide its straightening. They allow passive distraction during growth and stretching movements without the need for a surgical procedure. The growth guidance Shilla technique described by McCarthy et al. [42] (Fig. 17) and the modified Luque Trolley technique [43] both use vertebral fixation implants that slide freely along the rod to allow them to migrate gradually. These implants require a sufficient number of anchoring points on the spine and can be at the origin of fusion, which can be voluntary at the tip of the scoliosis in the Shilla procedure [42] or involuntary despite an extraperiosteal approach in the Luque Trolley technique [44]. Medium term results have only been reported by the inventors of these techniques on a small number of patients, but in the two studies, many fewer surgeries were needed in comparison to the use of growing rods.

These devices seem to be particularly relevant for cases of scoliosis having enough flexibility to allow sufficient reduction during the instrumentation. Independent evaluation of these systems must be carried out with longer follow-up before they can be used on a wider scale.

Metallosis has been observed with these devices, so this potential complication must be evaluated and taken into consideration [45].

3.2.3. Convexity compression devices
These techniques consist of slowing down the growth of the curvature convexity using the same principle as epiphysiodesis, but with implants that in theory avoid the need for definitive asymmetric spinal fusion.

Shape memory staples that are positioned through a small incision to compress and bridge the growth plates so as to reversibly block growth, have only been shown effective for curvatures under 35° [46]; their rigidity leads to fears of stiffening of the instrumented area [47], which must be long enough to be effective (Fig. 18).

Stretching a tether over the convexity [48] likely contributes to greater mobility and as a consequence, a lower risk of spontaneous fusion. The only published case report was in a patient with 40° Cobb angle who was operated at 8.5 years of age; the Cobb angle was 25° immediately after the tethering surgery and was 6° after 4 years of follow-up (Fig. 19) with mobility that was partially maintained [49]. For this patient, the tether prevented growth of the convexity. Its use before 8 years of age is not recommended because there is no evidence in a larger population of patients that spinal growth returns to normal once the tether is cut. There are no reports of this technique being used in cases of more severe scoliosis.

These devices have proven efficacy but their relevance and safety, in comparison to well-conducted conservative treatment and for cases of scoliosis with unknown progression, still needs to be demonstrated.
4. Indications

Knowledge of lung growth and the respiratory effects of early fusion have now led to spinal fusion being contraindicated before 8 years of age and preferably before 10 years of age. The gold standard “delaying” surgical technique, used by numerous teams, is the growing rod. Many variations of this technique exist, which are the result of diverse experience with more than 10 years of follow-up and a large number of publications describing the treatment outcomes.

The decision to carry out surgical treatment is a difficult one to make given the complications and the quality of the final outcome. Growing rods actually provide better control over the Cobb angle during maturation, but there is no statistical proof that the final angle after fusion is better for operated patients than non-operated ones. Conservative treatment does not cause the significant stiffness that is caused by repeated surgical procedures, the fibrosis induced by the implanted devices and the rigid spinal immobilization required by these different devices.

One must accept mediocre results during the conservative treatment period to obtain a result, once growth ends, which is comparable to the one obtained after a long, difficult delaying surgical treatment.

In most cases of syndrome-associated or idiopathic scoliosis, conservative treatment will result in less than 100° curvature at 10 years of age.

When the decision is made to carry out surgical treatment, it is not based on precise, reproducible criteria, but based on a set of arguments where the surgeon’s experience, etiology and progression of the scoliosis, and the psychological context and motivation of the patient and family will converge onto the same strategy.

Surgical treatment is debatable beyond 9 years of age as it is possible, except in rare cases, to wait one or two years to perform the final fusion at about 10 years of age with a multi-segment construct using pedicle screws at the tip of the deformity to avoid the risk of crankshaft phenomenon if the Y cartilage is still open.

For congenital scoliosis and severe chest wall and spine dysplasia, surgery is often unavoidable. Convex epiphysiodoses require a dual approach and are only effective when they are performed very early on over a sufficiently long span, which can hinder thorax growth. For these reasons, their indications are rare.

Most of the time, we prefer performing an osteotomy or vertebral resection at the tip of the deformity while avoiding an extended fusion. These procedures can be combined [50] or replaced by vertebral and/or rib distraction, which seems to stimulate growth of congenitally deformed areas [11,51]. However it seems risky to rely on a single distraction device to effectively expand a deformed block of bone.

Neuromuscular scoliosis cases should be treated conservatively while waiting for the spinal fusion procedure. When braces are no longer bearable, implantation of growing rods or growth guidance systems, which fit the pathophysiological mechanism for hypotonic scoliosis, can be considered to improve the tolerance of the brace, which can be less restrictive and especially ensure a role in the maintenance of the head and overall balance.

This surgical alternative must be carefully thought-out in patients with a fragile respiratory status [52] and must not be used solely to avoid the use of a brace. Use of dual rods is particularly indicated to address the bone fragility and muscle weakness in these patients.

The scoliosis associated with certain progressive neurological diseases can be surgically treated early on to avoid the treatment impasse of deformities that may be inoperable in patients who have become too fragile.

The Cobb angle and average age for implantation of growing rods are 80° and 5.7 years, respectively, in published studies [24,25,34] with minimum and maximum values of 32° to 147° and 1.4 to 9.5 years.

Growth guidance systems are theoretically an interesting alternative to growing rods, but the currently available instrumentation is highly invasive and has not been evaluated by a sufficient number of non-inventor teams to be used regularly. Although the newer techniques of convexity growth stoppage are attractive, they cannot be part of the current treatment arsenal for early-onset scoliosis because they have only been shown effective in patients with smaller-angle scoliosis and there is no medium term data on the reversible nature of the growth stoppage and the stiffness induced by the instrumentation.

Use of cranial halo: the halo is a good method to prepare the placement of a growing rod or guide, so as to position the implant on a vertebral column that has been corrected to the best possible degree, with the goal of reducing the stresses on the rod and the risk of mechanical complications.

5. Conclusion

Knowledge of the interactions between scoliosis and lung growth has led to a better understanding of the consequences of extended spinal fusion performed before 8 years of age, and has led to the emergence of fusionless surgical treatments as an alternative to conservative treatment when the latter is unable to curb the progression of early-onset scoliosis.

Many technical innovations, tempered by the need to treat scoliosis cases with difficult prognosis and by the technical and strategic challenges represented by fusionless scoliosis correction, have been used in recent years, sometimes overly so. Growing rods are now widely used despite this treatment’s inherent complications, which can exceed 100% with some devices. However, no study has shown this technique to be superior in terms of the Cobb angle after final fusion when compared to the same fusion carried out after a delaying conservative treatment.

It is important to remember that most of the devices are still being evaluated and are being used without marketing approval for the indication, which makes the surgeon fully responsible for the implantation.

In our eyes, conservative treatment has a major role in the treatment of early-onset scoliosis. Conservative treatment is not considered a failure when the Cobb angle continues to increase. It remains the treatment that provides the best compromise between tolerance and effectiveness.

The heterogeneity of patients and short follow-up for surgical interventions can largely explain the subjectivity of the surgical decisions, which are not very reproducible [53]. The coming years will probably allow us to better define the indications for early surgical treatment by taking the Cobb angle at the end of growth into consideration instead of the change in Cobb angle during the treatment.

The chosen treatment must be matched to the day-to-day life of patients and their families after they have been thoroughly and clearly informed.

Disclosure of interest

The author declares that he has no conflicts of interest concerning this article.

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