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MINI REVIEW

Spinal deformity in children

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ABSTRACT

The treatment of early onset scoliosis is a challenge because of the need to correct the deformity of the spine without using the traditional systems that require spinal fusion at this age. The conservative treatment of early onset scoliosis with brace is usually not effective but can help to postpone the surgical treatment at a time closer to skeletal maturation. The surgical procedure includes systems that do not fuse the spine but allow for further growth. These systems can be (a) the growing rods, (b) the guided rods and (c) compressive-based. These systems are a good option for the treatment of childhood scoliosis, but as they are not «protected» by a bone spine fusion they are subject to ongoing stress and forces. As a result, there is a higher failure rate of materials compared to the traditional fusion systems.

KEY WORDS: early onset scoliosis; thoracic insufficiency syndrome; growing brace for scoliosis; growing rods; guided rods; compressive-based system

Introduction

Scoliosis is a three-dimensional deformity of the spine in which there is a shift of the vertebrae, and abnormal curves that are mainly at the side of the spine. This deformity can be observed in children before puberty. In some cases, the scoliosis is progressive, and the deformation of the rib cage can affect the development of pulmonary alveoli. During this period there is a need to protect the rib cage from deformation and lessen the effects on the pulmonary parenchyma. On the other hand, we can't correct the deformity with spinal fusion and suspend its development because that will limit the growth of the rib cage, causing obstructive pulmonary disease. The inability to interfere in scoliosis developed in young children can lead to serious scoliosis with serious co-morbid obstructive pulmonary disease. For all the above reasons these deformities are a distinct clinical entity termed «early onset" scoliosis and appears before the age of 10[1]. The way the scoliosis is going to develop depends on the etiology (congenital, neuromuscular, syndromic, i.e. neurofibromatosis, and idiopathic). Some syndromic scoliosis with congenital anomalies such as asphyxiating thoracic dysplasia (Jeune's Syndrome) and the vertebra-rib dysostosis (Jarcho-Levin-Syndrome) will take a very special and demanding treatment.

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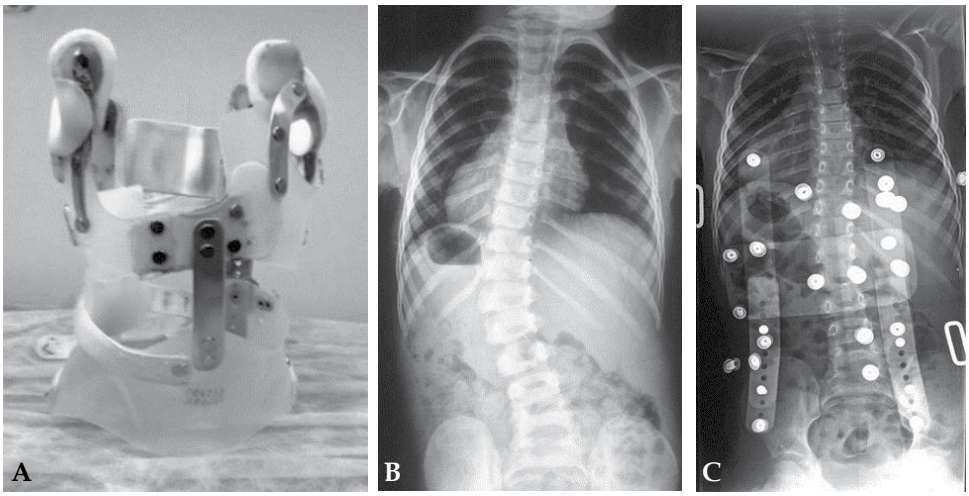

Figure 1. *Treatment with a growing brace (A). The initial scoliosis of a girl of 1 years old with left thoracolumbar curve was 33° (B) and after 1 year of bracing that improved at 12° (C).*

Thoracic insufficiency syndrome

The goal of the treatment of early scoliosis is to stop the deterioration of scoliosis into severe deformation and to prevent the syndrome of thoracic insufficiency[2]. The number of pulmonary alveoli grows rapidly until the age of 2 years, and then continues to increase at a slower rate until the age of 8 years. After this age the cells increases in size but not in number[3]. Therefore, any deformation or reduction in the elasticity of the thoracic cage affects the development of the respiratory parenchyma and therefore the respiratory function. The restriction of the lung parenchyma causes arterial pulmonary hypertension that leads to life-threatening diseases, i.e. pulmonary heart, heart failure[4]. This pathological condition is called syndrome of thoracic insufficiency. It seems that there is a direct correlation of respiratory function with the level of the T1-T12 vertebrae, in children who were subjected to spinal fusion due to congenital scoliosis. In these children a decrease of the pulmonary vital capacity of 50% was observed after spinal fusion. In fact, in children younger than 8 years of age who were subjected to spinal fusion, the vital capacity decreased by 60%. These studies revealed that the height of T1-T12 must be greater than 20 cm in order to avoid the syndrome of thoracic insufficiency[5]. Therefore, in these ages spinal fusion is prohibitive because it reduces the vital capacity of the lungs. The spinal fusion, also, decreases the elasticity of the thoracic cage, which acts negatively on the respiratory function[6,7]. In addition, in children of this age, with a great expectancy of growth, treatment with posterior instrumented spinal fusion can cause the crankshaft phenomenon. This is a new rotation deformity that develops after spinal fusion, due to the growth of the body of the vertebra, which cannot be restrained by the posterior instrumented spinal fusion. Last contraindication for spinal fusion in early scoliosis is the asymmetry of the torso-limb that can be caused by the spinal fusion.

Treatment

Conservative treatment

The conservative treatment has poor results in terms of evolution of childhood scoliosis because it can't stop the progression of scoliosis. However, it is a useful tool because a) it can help the patient with hypotonia stay in the sitting position, b) it can control the secondary curves in patients with congenital scoliosis and c) it saves us some time before deciding upon a more active intervention[1]. For the conservative treatment brace of the spine, thermoplastic seat, back props, and casts of the spine can be used. In addition, special physical therapy programs such as SCHROTH, SEAS, and SCOLISMART can be helpful. The brace of the spine can be hard or soft. The hard brace is the most widely used and documented over time. The

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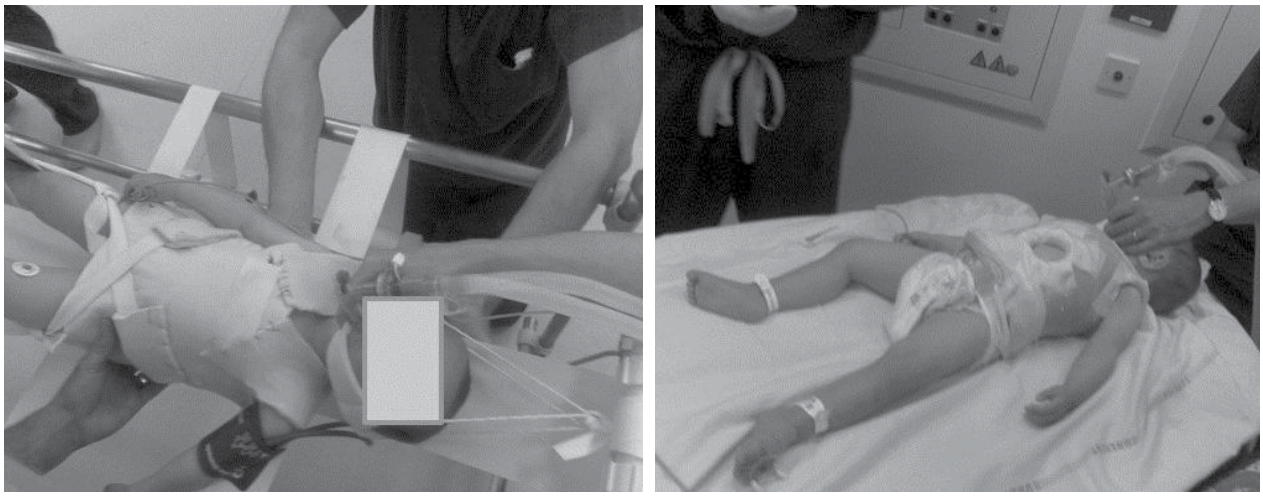

Figure 2. *Procedure of serial casting for early onset scoliosis.*

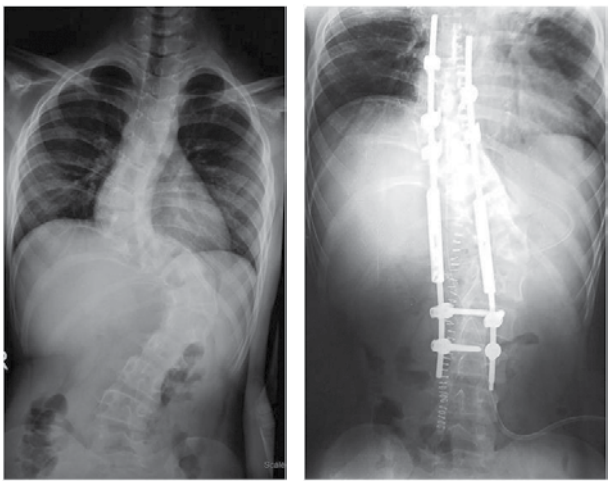

Figure 3. *A young girl of 10 years with neurofibromatosis treated with growing rods.*

most common type is the Boston and the Cheneau brace. Attempts have been made to treat the scoliosis with soft braces (i.e. Spinecor, Scolismart), but they did not prove their effectiveness. The traditional hard scoliosis braces can have some good results in idiopathic scoliosis and in some syndromic scoliosis with characteristics of idiopathic scoliosis. On the contrary, the hard brace has poor results in neuromuscular and congenital scoliosis. A brace for scoliosis, applied to a child less than 3 years old will need to be changed every 4 months due to the development of the spine. In adolescence the brace needs to be changed every 1 ½ years. This increases the costs of bracing during childhood. For this reason, in our clinic we have designed and used a type Boston or Cheneau growing brace which consists of two parts. The first part is applied to the pelvis, and the second part is applied to the torso and connects with the first part with three metal plates (**Fig. 1**). Using these plates, we can increase the brace's height every 4 months. We can repeat this increase in height 2-3 times, in order to keep the brace for 1 to 1 ½ years. These braces have been used since 2012 in 4 kids with success. In case that the scoliosis cannot be controlled by a brace, the next step is the use of a serial spinal cast. In this case, a plaster cast is placed to the child under general anesthesia. The child is placed at a type Cotrel-Dupusett table of traction and a plaster cast is applied. Special care is needed not to squeeze the viscera with the cast by creating windows of relief (**Fig. 2**). This cast needs to be changed every 2 months. The complications that can be observed with conservative treatment are pressure of the abdominal viscera, reduction in the glomerular filtration rate, a slight decrease in the volume of the lungs, tubular thorax, flattening of the lumbar spine and psycho-social disorders.

Surgical treatment

When the conservative treatment is unsuccessful surgical treatment is needed. Surgical treatment at this age includes systems of correction of the spine with-

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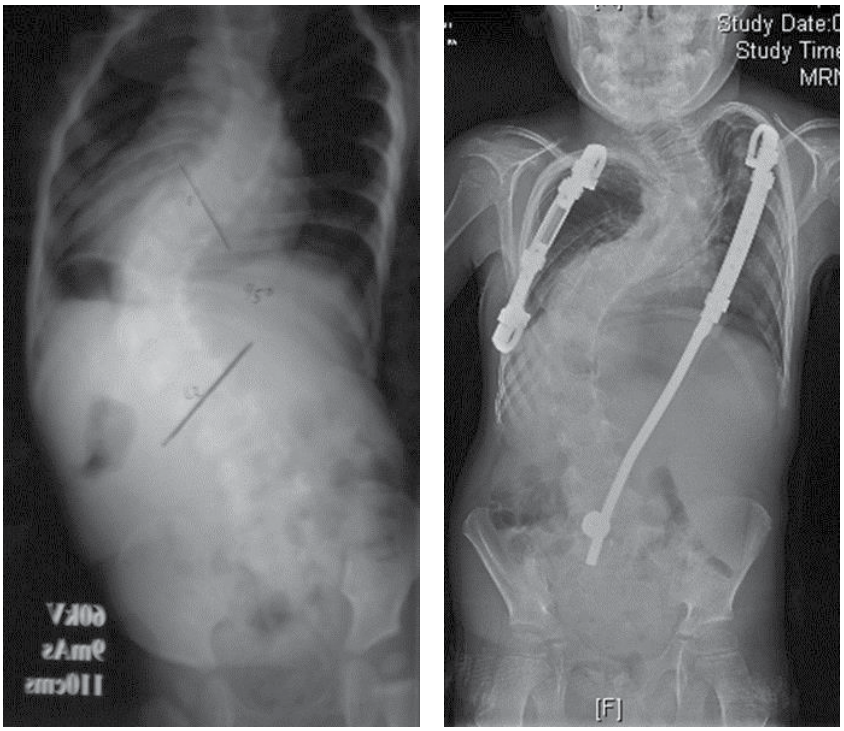

Figure 4. *A young boy of 5 years old treated with the VEPTR system.*

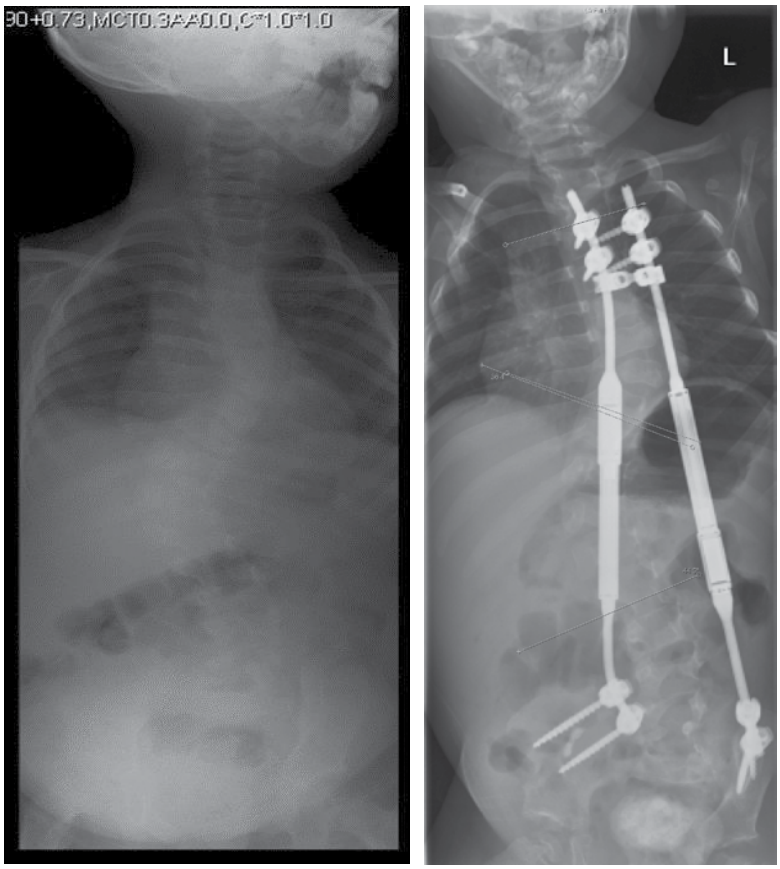

Figure 5. *A young boy of 3 ½ years old with Stickler syndrome treated with MAGEC system (courtesy by Colin Nnadi).*

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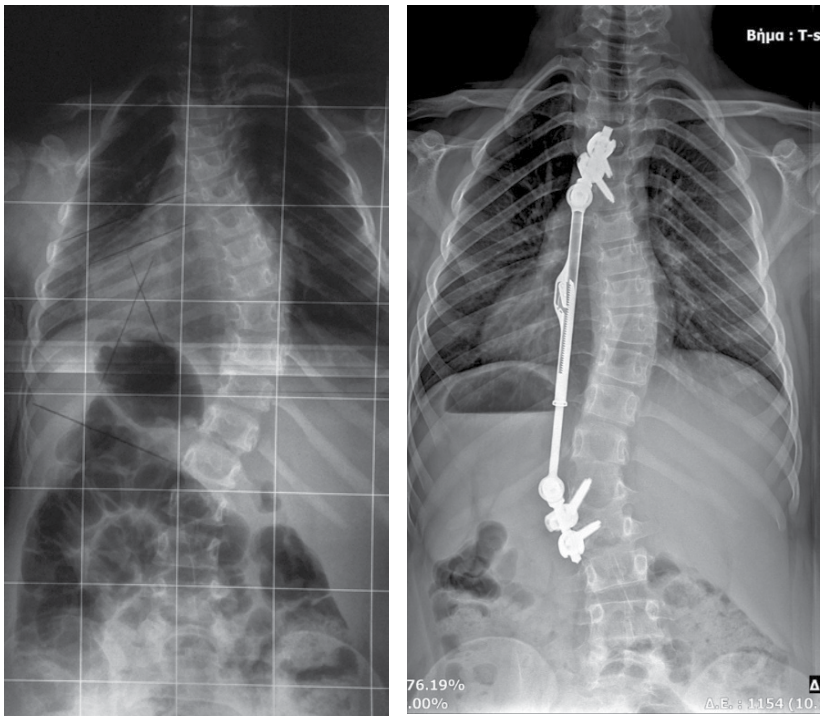

Figure 6. *A girl of 8 years old treated with Apifix EOS system.*

out spinal fusion. These systems can be divided into three categories: systems with growing rods, systems with guided rods and compression-based systems[1,9].

The traditional growing rods belong to the growing rods of the first category of systems, that need to be distracted, manually, every 6 months under general anaesthesia. This system is placed by posterior approach by pedicle screws at two vertebrae above and two below the scoliosis. The rods have a mechanism that allows lengthening (**Fig. 3**). The fixation of the rod to the vertebrae, other than screws, can be done with hooks, sublaminar wires (or straps), or with rib rings. The most powerful support is achieved with the pedicle screws. Each support system has its biomechanical peculiarities and it is useful to have alternative options in case it is not possible to place pedicle screw or because we need to preserve the elasticity of the correction system that we use. These traditional growing rods are the «gold standard» of surgical treatment of childhood scoliosis. The system VEPTR (Vertical Expandable Prosthetic Rib) also belongs to the first category of systems with growing mechanisms[8]. This system, basically, is a mechanism of stretching of the ribs. This system is fixed to the upper thoracic cage with rib rings and to the lower part of the scoliosis either with rib rings, pedicle screws or pelvic hooks (**Fig. 4**). The lengthening needs to be made manually every 4-6 months under general anesthesia. This system has limited effectiveness in the correction of scoliosis, but it is a good choice for patients with rib fusion and a restrictive chest cage (i.e. Jeune's Syndrome). In order to avoid recurrent surgeries in children, a new system, the MAGEC (Magnetic Expansion Control), has been designed. This is placed with pedicle screws like the classic growing rods, but the MAGEC rods have a magnetic mechanism that allows lengthening with a remote control at the outpatient department[10,11,12] (**Fig. 5**). Similarly, the APIFIX-EOS system was designed consisting of a self-growing rod that is placed in the concave side of the scoliosis by pedicle screws above and below the curve (**Fig. 6**)[13,14]. The lengthening of this rod is made mechanically with the movement of the torso and with special physical therapy. Also, a similar system, EURO V2, designed by Lotfi Miladi, is placed with special sacroiliac screws and

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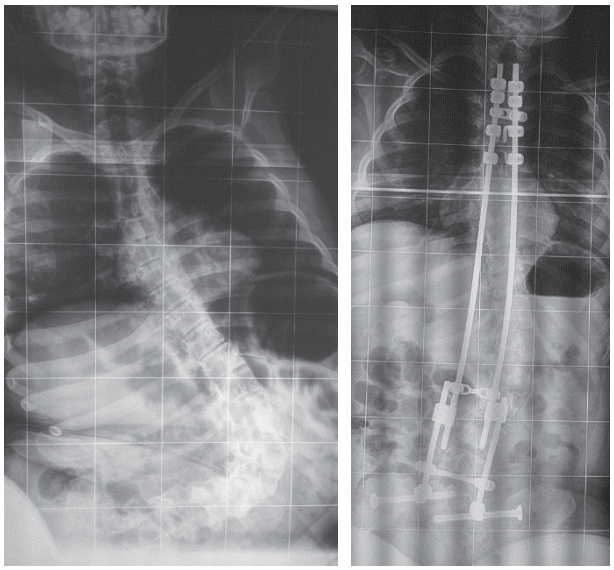

Figure 7. *A girl of 10 years old treated with EURO V2 system.*

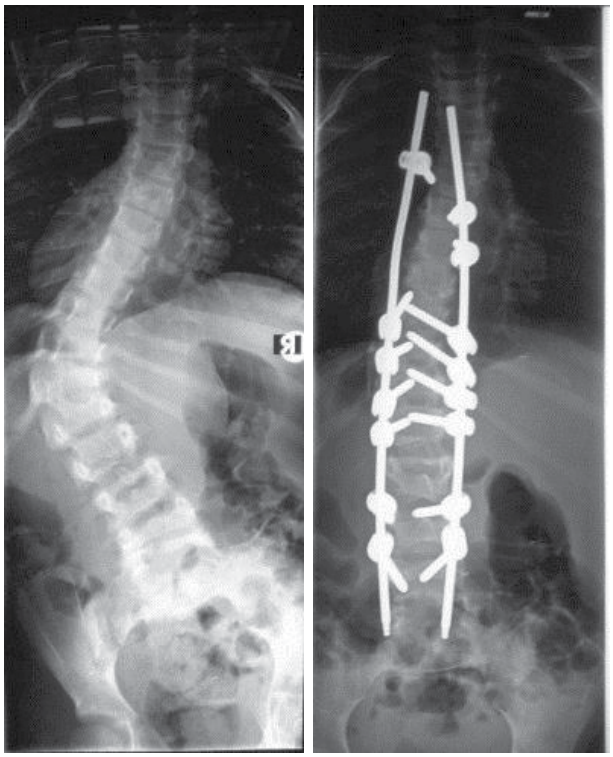

Figure 8. *A young boy with neurofibromatosis of 6 years old treated with Shilla system.*

4 hooks at the upper part of scoliosis. The rods have a mechanism that allows the lengthening following the growing of the spine (**Fig. 7**).

In the second category includes the guided rods. The Luque-Trolley was the first system of this class and consists of two rods which are fixed in the spine with sublaminar wires. The wires can slip on the rod following the growing of the spine. This system is low cost and used since the 1970s with satisfactory results. However, the extensive subperiostal approach that needs to be done to place it can lead to early spontaneous spinal fusion. Later systems such as the Shilla[15,19] and the Modern Trolley are placed with a minimal approach. The system of Shilla has special pedicle screws that slide on the rod, like the sublaminar wires (**Fig. 8**), while the Modern Trolley is consists of pedicle screw with a head from polyester that are connected to and slip on the rods.

The third category includes the compression-based systems: Vertebral Body Stapling[20] and Vertebra Body Tethering[21]. These systems affect the growth of the spine by hemiepiphysiodesis in a similar way as in the treatment of varus or valgus knee. The hemiepiphysiodesis includes the curved part of the scoliosis in order to stop the growing of the convex side of the spine and to leaves the concave side free to grow correcting the scoliosis. The hemiepiphysiodesis of the vertebrae can be made with buckles (stapling) or dynamic fusion systems (tethering) which are placed by anterior approach (open or thoracoscopic). These systems modify the growing of the vertebra and use the viscoelastic properties of the intervertebral disc to correct the scoliosis. Therefore, these systems are used also for adults.

The materials of all these systems implemented at children without a spinal fusion undergo continuous forces because they are not protected by a spinal fusion. For this reason, these systems are prone to complications of failure of materials such as fracture of the rod (15%) and failure of the upper support (up to 95%)[16,17,18]. Breakage of the pin of the magnetic mechanism has been observed with the use of the magnetic rod (MAGEC). With the compression systems subcorrection or overcorrection of scoliosis has been observed, because it is difficult to calculate the correct age to use these systems.

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Also, breakage of the connective belt between the screws has been observed with these systems. The surgical revision of these systems is particularly difficult due to the anterior approach. Metallosis of growth-guided systems has been observed due to friction of materials. Another complication that may occur is wound infection (6.7%, of which 67% will need surgical debridement). Other complications include early spontaneous spinal fusion (that may occur due to the surgical approach), and junctional kyphosis (that may occur due to the sagittal imbalance and due to the increased «hardness» of the stabilized part of the instrumented spine). Although the new systems do not require regular revisions per semester, they may often need non-scheduled re-operations but, these are fewer compared to these needed with the classical system. We always need to bear in mind is the psycho-social consequences of the repeated operations to the children.

Conclusion

The treatment of early onset scoliosis is a challenge because of the need to correct the deformity of the spine avoiding the spinal fusion with the traditional systems. The new systems offer new options to treat children with scoliosis and avoid the spinal fusion. Still the perfect system has not been invented. Every spine surgeon has to choose the system he is going to use to each individual child, and hence choose the complications he will encounter. A

Disclosure of interest

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

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