

Surgical Treatment of Early Onset Scoliosis Probable causes of undesirable results

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ABSTRACT

AIM: Early Onset Scoliosis (EOS) is in many cases a surgical challenge. These deformities are present during the child's most active physical development and may raise several issues, especially cardio-respiratory problems which will irreversibly affect the quality of life. The aim of EOS treatment is to ensure a proper lung development and to control the kyphoscoliotic deformity without fusion. The purpose of the study represents the analysis of postoperative evolution in a series of operated EOS patients made us aware of possible therapeutic related issues. We consider the importance to highlight them, in order to carefully choose the surgical treatment in EOS.

MATERIAL AND METHODS: The study represents a retrospective clinical and imagistic analysis of operated EOS patients. A series of

52 EOS patients, operated by different treatment methods (2003-2014) has been analyzed. The series included congenital, idiopathic infantile and one syndromic scoliosis. Surgical procedures consisted in hemivertebral resection concomitant with instrumentation, the use of VEPTR or Shilla devices or combinations of these. The follow-up period ranged 4-15 years. The patients were investigated by clinical and imagistic exams. 15 patients with poor clinical and radiological results after surgery were identified (worsening of the curve, extended fusion with a short, stiff spine, impaired trunk imbalance and/or pseudarthrosis).

RESULTS: The possible identified therapeutic issues included: the use of surgical techniques which resulted in extended posterior fusion, the choice of an possibly poor surgical technique, improper or faulty applied implants.

CONCLUSIONS: Complications and possible postoperative evolutions related to the chosen therapeutic decision should be very thorough taken into account before any procedure. A poor strategy treatment of these deformities decreases quality of life. The prognosis is variable, depending on the cause inducing the deformity. That's why the treatment should be individualized for each EOS patient. An improper treatment may produce real disasters or cuts short any therapeutic chance, maybe the only one available, in order to ensure the best life quality obtainable.

Key words: Early Onset Scoliosis; Congenital scoliosis; Idiopathic infantile scoliosis; Hemivertebra

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INTRODUCTION

Early onset scoliosis (EOS) represents a group of spinal deformities identified before the age of 8-10 years^[1], according to the Spine Research Society. This category includes the following entities:

Idiopathic type scoliosis, with no obvious cause, being itself

divided into *idiopathic infantile*, present in a child younger than 3 years of age, and *juvenile*, in children aged 4 to 10 years of age. Statistics show that about 20% of these cases present with medullar malformations. That's why all cases should undergo a thorough examination, including CT-scans and MRI. The measurement of the Metha angle and the RVAD (*rib-vertebra angle difference*) are very important in establishing the prognosis of the evolutive potential of the deformity^[2,3].

Congenital scoliosis, even if rare, represents a challenge to the spinal surgeon. The progressive potential is variable, related to the malformation type. The most aggressive progression of the scoliotic curve is due to fully segmented hemivertebrae associated to an opposite non-segmented bony bar, followed by segmented hemivertebrae^[4]. These patients may present with associated occult spinal and neurologic malformations, including spinal dysraphism, in about 30% of the cases. That's why it is mandatory to perform a detailed examination of the patient, including proper X-rays, CT-scans and MRI exams^[5].

The main problem in approaching to EOS patients is the progressive potential of the deformity which has repercussions in the development and growth of the vertebral column and on the cardio-respiratory function. The presence of thoracic insufficiency syndrome, due to lack or improper treatment, represents a major risk factor for physical impairment by pulmonary and, consecutively, cardiac dysfunctions with premature death^[6]. The main goals of EOS treatment are represented by stopping the curve progression, deformity correction, ensuring a near to normal lung development and function, in order to allow the best possible quality of life during childhood to adulthood^[7]. These goals will not be achieved, unless the spinal fusion by surgery is a minimal one, ensuring mobility and growth potential of the vertebral column and thoracic cage. The therapeutic arsenal is based on observation, conservative and surgical treatment which has to be adapted and applied at the right moment and in a proper manner to achieve the mentioned goals. Any wrong decision in the therapeutic approach, especially the surgical one, will complicate the patient's evolution. Even more, it may annihilate any other proper treatment option in order to benefit of a close to normal development.

METHODS

We retrospectively analyzed a series of 52 patients with operated EOS. The follow-up period of these patients ranged 4 to 15 years, surgery being performed between 2003-2014. We identified 15 patients with unsatisfactory clinical and radiological results (worsening of the curve, extended fusion with a short, rigid and/or crooked spine, impaired trunk imbalance and/or pseudarthrosis). 37 cases (including congenital scoliosis, idiopathic infantile and juvenile scoliosis and syndromic scoliosis) presented with fair and good results, according to results presented in the specialty literature. These cases were operated respecting strict indications of approach and spinal device. The series of 15 EOS patients with poor results highlighted a number of possible surgical treatment issues grouped as following: extended spinal fusions, possible improper spinal implants related to the specific pathology, possible improper surgical techniques or combinations of these. The types of operated EOS with poor results were represented by congenital scoliosis (12 cases), infantile idiopathic scoliosis (2 cases) and syndromic scoliosis (1 case). All cases were operated up to the age of 6 years. The applied techniques consisted in hemivertebra resection by a posterior or anterior approach concomitant with spinal instrumentation, VEPTR

and SHILLA technique.

RESULTS

The probable noticed cause of treatment failure in the surgical treatment of infantile idiopathic scoliosis consisted in a premature extensive spinal fusion with consecutive crankshaft phenomenon under a non-expandable spinal implant (Figures 1 and 2). In congenital scoliosis it consisted in an incomplete or total failure of hemivertebra resection, followed by ineffective instrumentation, in the use of VEPTR with unusual device positioning, in the failure of optimal distraction and in extended spinal fusion under an unusually placed device (Figure 3). In syndromic scoliosis it consisted in the a probably improper use of a VEPTR device to correct a dystrophic curve in von Recklinghausen disease, which worsened the short-arch kyphoscoliosis after consecutive distractions and destabilized the vertebral column due to the appearance of a vertebral pseudarthrosis with consecutive neurological impairment. The evolution of these spinal deformities as a result of a possible improper surgical treatment was an unwanted one, with consecutive complications, despite initially the approach seemed to be an efficient one. The complications consisted invariably in the appearance of stiff and severe curves due to premature spinal fusion on extensive spinal segments and to the lack of spinal growth up to near normal parameters in long-segment instrumentations. The patients required or will need for sure a change in therapeutic strategy (being in observation for other surgical procedures), as a desperate measure to correct the spinal deformities and/or enforce the growth and development of the spine in order to combat the thoracic insufficiency syndrome. These "repairing" strategies consisted in implant conversions: repositioning of VEPTR devices in an usual position,

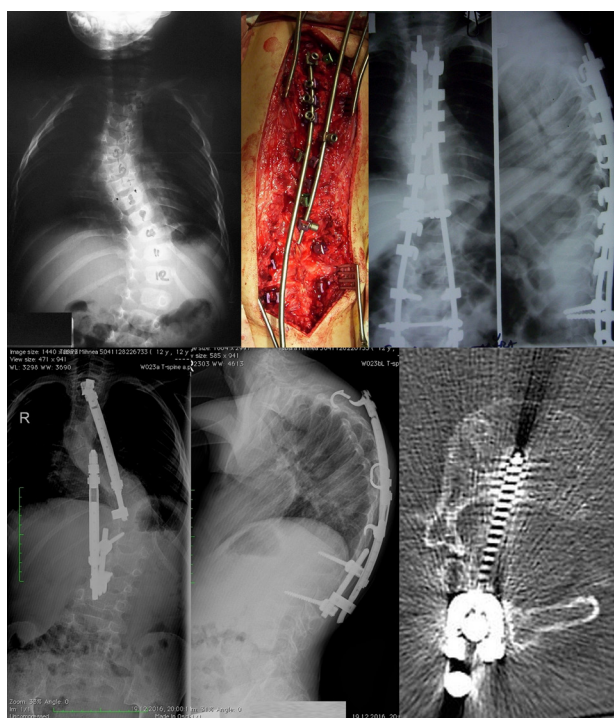


Figure 1 A case of infantile idiopathic scoliosis operated at the age of 1 year and 8 months, with extensive subperiosteal exposure and a non-expandable spinal implant. The patient developed crankshaft phenomenon and the next tried solution was hardware removal and the use of 2 VEPTR devices which failed too, probably due to premature and extensive fusion and unusual placement. The lower pedicular screw was placed in the spinal canal, resulting in pain located to the thoracic wall, too.

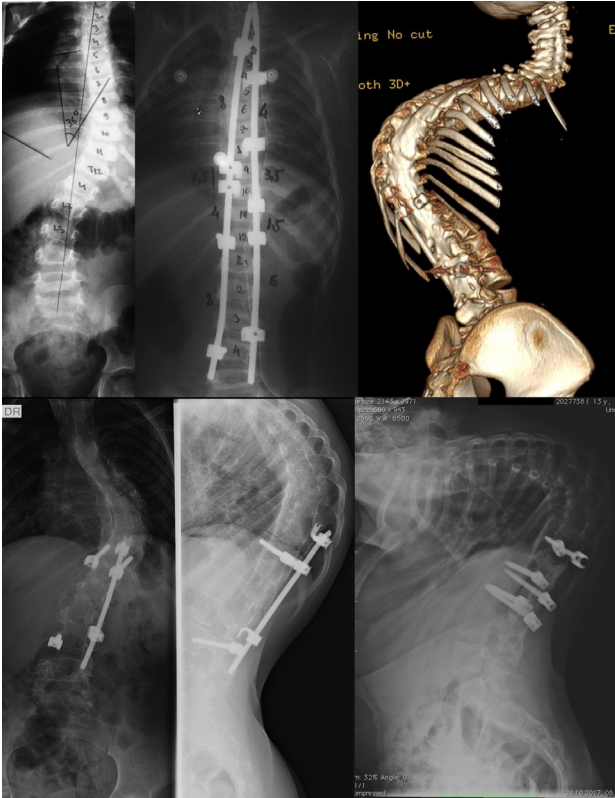


Figure 2 A case of infantile idiopathic scoliosis operated at the age of 3 years, with extensive subperiosteal exposure and a non-expandable spinal implant. The patient developed severe kyphoscoliosis and after complete hardware removal and a further surgical procedure, the patient developed neurological complications. Further surgical treatment has been abandoned.



Figure 3 Unusual VEPTR placement, with the upper module inserted in the spinal canal, intended to act as a *vertebra-vertebra* construct (up), and failed hemivertebra resection with spinal instrumentation failure (down). The results following such complications led to severe and stiff scoliotic curves and trunk imbalance.

implant removal and exchange with an adequate one to theoretically allow at least the deformity to be corrected and in the resumption of more efficient surgical techniques (Figure 4). The SHILLA device in case of congenital scoliosis presented with pulled out upper screws and trunk imbalance due to uncertain indication, rod misshape and unusual screw placement. It required SHILLA removal and hemivertebra resection, followed by short posterior fusion (Figure 5).

The further evolution after changing the therapeutic strategy proved to be a difficult one, with modest and difficult-to-obtain fair results in all cases, when compared to the results of actual standard treated cases. Unfortunately, a further therapy of a part of the analyzed cases proved either to delay or even make impossible an efficient treatment, in order to remedy the initially obtained results.

CONCLUSIONS

As it is already known in the specialty literature, the choice and applying of a poorly therapeutic strategy without careful attention related to the spinal deformity and neglecting the fact that the spine in children requires further growth and development, may lead to disastrous results. These unwanted results are consecutive to poor surgical techniques, to inadequate spinal implants and, even worse, to combination of these, with major impact on normal development and quality of life. Not a few times, the spinal surgeon has “*just one bullet*

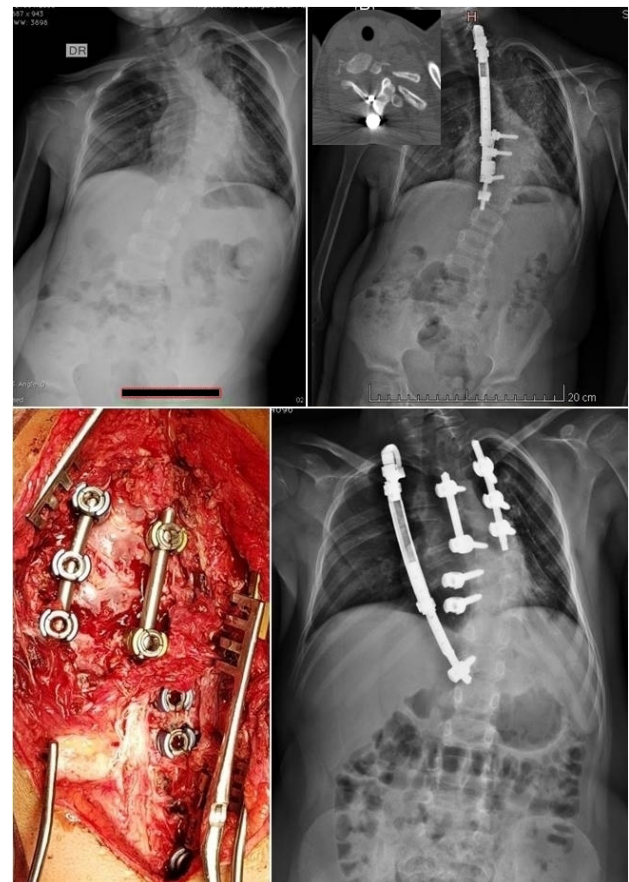


Figure 4 A case of congenital scoliosis with unusual placement of VEPTR with the upper module placed intentionally in the spinal canal, intended to act as a *vertebra-vertebra* construct. Distraction proved to be ineffective and the construct had to be replaced in a standard position with hemivertebra resection and a short fusion. The aim of the second surgical treatment is a desperate try to allow spinal and thoracic growth with progressive curve correction until final fusion.

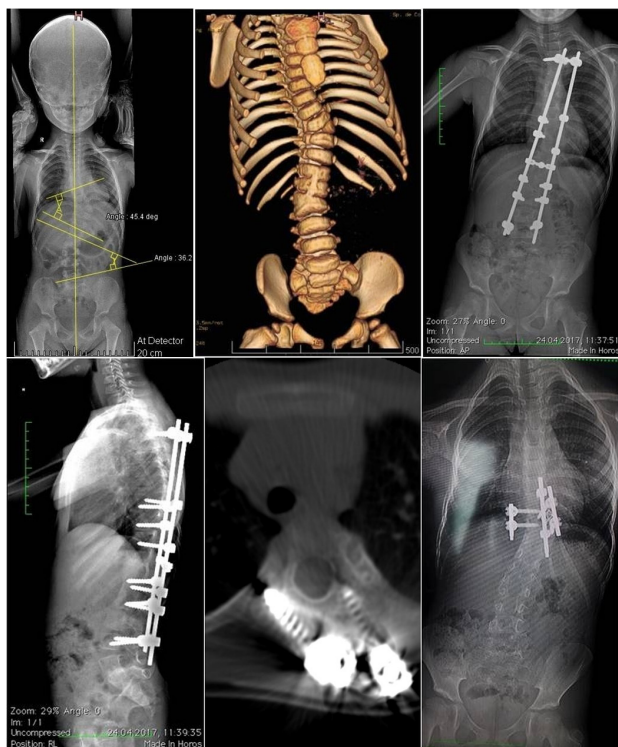


Figure 5 Congenital scoliosis with 2 hemivertebrae operated at the age of 2 years with failed hemivertebra resection and unusual SHILLA technique planning. The upper screws pulled out and the spine presented spatial imbalance which required device removal, hemivertebra resection and a short fusion spinal implant.

to shoot” and missing the target may cease any chance of a desired surgical result in these patients. The conservative *delaying strategy* in the treatment of infantile idiopathic scoliosis, until the loss of control of the deformity, is an important goal of the treatment. Surgery should become a priority when losing curve control. Guided growth and distraction based devices represent the actual best options when surgery is mandatory in these patients^[8]. Congenital scoliosis with a high progressive potential should be surgically approached at a young age, starting with 1 year of age, in order to stop curve progression and prevent structural changes of the vertebral column. Surgical results obtained in the cases with an older age are consecutively modest. Hemivertebra resection by a posterior only approach represents the *golden standard* nowadays, ensuring a good correction, mobility preservation and development of the spine^[9,10]. The VEPTR device proved its’ limited and strict indications consecutive to the long enough obtained experience^[11]. The SHILLA device is on the way to prove its’ efficiency and limits, presenting a standardized principle

of the surgical technique^[12]. That’s why a proper surgical technique, the compliance to indications, adapted to each case and the use of proper spinal implants related to specific pathology ensures chance of success in the treatment of this delicate pathology.

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