SURGICAL TREATMENT OF PATIENTS WITH SCOLIOSIS IN THE 1ST DECADE OF LIFE: A LITERATURE REVIEW

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The paper presents the first part of literature review of the main methods of treatment of patients with infantile and juvenile scoliosis. Particular attention was paid to the results of treatment using various instrumentation with the possibility of stage correction.
Key Words: infantile scoliosis, juvenile scoliosis, surgical treatment.

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History of surgical treatment of patients with infantile and juvenile scoliosis is about 50 years long. According to a SRS consensus statement published in 2015 the concept of "early scoliosis" (Early Onset Scoliosis, EOS) covers deformities identified in patients under the age of 10. This group includes both infantile and juvenile scoliosis of various etiologies (idiopathic, congenital, neuromuscular, syndromic). The choice of method of surgical treatment depends on more than a dozen of factors, the key factors being gender and age of the patient, etiology and severity of the deformity, the rate of its progression, bone maturity, severity of comorbidity [6, 8–10, 36].

The surgeon must select a method that allows achieving optimal correction, avoid frequent re-operations and manipulations of the spine, leading to formation of an artificial bone blocks. This article presents the main approaches, methods and results of staged EOS surgical treatment using different sets of tools. Literature sources were selected from Ovid, Pubmed and Elsevier databases and include publications in reputable periodicals and review monographs with the highest citation index.

It is well known that up to 60 % of the observed growth of the spine and formation of up to 50% of the volume of the thorax occur within the first 10 years of life. Therefore, progression of untreated deformities can lead to irreversible cardiopulmonary and neurological disorders and profound disability and can shorten the lifespan of patients [13, 38]. According to some reports, the risk of such disorders is present when the deformity exceeds 100° [46].

Conservative treatment (regimen, strengthening gymnastics, massage, physiotherapy, swimming) with dynamic observation should be employed already at the initial stages. However, prior to prescribing these treatments it is necessary to conduct specialized examination, including examination by specialists in this particular field, X-rays and MRI of the spine to rule out spinal cord abnormalities [46]. Corset-therapy may be used in case of indications to achieve stabilization and prevent rapid progression. If the conservative treatment proves to be inefficient, a surgery is performed. Currently, the main indication for surgical correction is the severity of deformity with Cobb angle higher than 40° [47, 53].

Over the course of XX century, the main approaches to surgical treatment of progressive scoliosis in children during the first years of their life had been as follows.

In the 1960s, different types of the dorsal and dorsoventral fusion with postoperative immobilization with a plaster corset were used [48]. Dorsal instrumentation without fusion has started to be used at the same time [30]. However, these methods have proved ineffective due to high incidence of complications. Installation of Harrington distractors was accompanied by spontaneous formation of the dorsal bone block and fracture of the construction and support points [35, 39, 58]. Development of hyperlordosis and false joints with the lack of growth of the bone block were observed in the zone of fusion without instrumentation [40].

In the 1970s, epiphysiodesis from transthoracic access to balance growth of the spine and the development of ventral bone block has become widely used method [1, 5, 11, 42]. These interventions were complemented by dorsal fusion with the use of the distractor. The plaster corset and subsequently Milwaukee corset were used for immobilization in the postoperative period. However, in most of the patients the correction failed was accompanied by further progression of the spinal deformity. For the first time,

it was noted that the progression caused by the rotation of the spine around the bone block's axis, and the process was described as a crankshaft phenomenon [23, 50]. This phenomenon has largely defined indications for beginning of surgical treatment and its extent. Prognostic factors for the course of scoliotic spinal deformities were actively researched and some of the features are still relevant to today. [7] The authors have tried to start conservative treatment (corset therapy) as soon as possible to stop the progression of the deformity, and did not put the surgery off until the completion of the growth period [37, 60].

In the 1980s, staged surgical treatment with the final fusion upon reaching adolescence has become a widely used technique [27, 28, 32, 39]. The researchers noted the important role of the dorsal instrumentation (Harrington distractor), the use of which in combination with various types of ventral and dorsal fusion reduced the risk of postoperative progression. However, there were still a high number of complications, including mechanical ones (support points instability, endocorrector fractures) and festering. There were also changes in the frontal balance and sagittal circuit outside the fusion area.

The use of Cotrel – Dubousset dorsal segmental instrumentation in 1990s significantly reduced the risk of mechanical complications and allowed choosing the optimal volume of surgery. The principal approaches to surgical treatment of progressive scoliosis in pre-pubescent children were as follows. The basic approach was a two-stage anterolateral epiphysiodesis and dorsal correction with spine fixation by segmental instrumentation. The method was particularly effective in children at puberty and was supplemented by dorsal fusion that, according to the authors, prevented postoperative progression of the deformity. However, in younger age groups most of the patients showed signs of crankshaft phenomenon. Taking these features into account the advantage of dorsal segmental instrumentation was undeniable, but its continued use was based on the staged corrective actions with the fusion surgery in adolescence. The method of performing isolated anterolateral epiphysiodesis has been justified theoretically, since it allows balancing the growth of bone tissue of the vertebral bodies on the convex and concave sides of the scoliotic arc.

From 2000s to these days new methods of surgical treatment have been actively introduced featuring various multisupport endocorrectors allowing a possibility of staged distraction. The main methods are VEPTR (Vertical Expandable Prosthetic Titanium Rib), a growing rods method, Shilla technique, vertebral stapling, vertebral tethering. The variety of these techniques allow development of more detailed indications for surgical treatment, and, most importantly, timely execution of an effective surgical treatment starting from the age of 6–12 months. However, the principles developed 20 years ago, remains unchanged. In teenage scoliosis, the main task is to perform the optimal correction and stabilization of spinal deformity. In actively growing children, it is necessary to prepare the spine for the final stage of treatment, to enable growth while managing the process itself. The stabilization of the spine is not the best option, as it could restrict its further growth and, consequently, lead to the development of thoracic insufficiency syndrome [3, 4, 17]. The dorsal spinal fusion may also cause uncontrolled progression of spinal deformity and preclude its correction in adolescence. Several methods have recently been proposed to resolve these problems and they have their advantages and disadvantages [34].

Indications and outcomes of using VEPTR instrumentation. Onset and malignant progression of scoliosis in children can lead to early disability. In most cases, conservative treatment of such deformities is limited to a complex of restorative procedures. Traditional methods of surgical treatment (anterior epiphysiodesis and/or dorsal fusion on the convex side of the curvature) do not solve the problem. The issue of the natural course of chest deformation in case of such deformities and the effect of spinal surgery on the progression of a chest deformity and growth of the ribs are poorly studied. According to Emans et al. [26], fusion, performed on five or more thoracic motion segments in children under the age of 5, results in a subsequent decrease in lung function to the 30–79 % from the age norm, and there is an inverse relationship between age at the time of operation and the degree of the severity of subsequent disorders. There is a description of scoliosis caused by a rib block without vertebral anomalies [21].

In 1987 Dr. Campbell [16] of San Antonio (USA) developed a fundamentally new endocorrector intended not only for control of progressive of spinal deformity, but also for normalization of respiratory function of the chest. Originally, a silicon plate was used as prosthetic of the chest wall defect, which was fixed by Steinmann's pins. In 1996, "Synthes" company developed a new design of the plate, and the clinical trials in the US begun; in 2002 the VEPTR instrumentation was first used in Europe (Basel). Later, Campbell suggested expanding thoracoplasty, including the use of VEPTR as both a chest wall prosthesis and a distractor for normalization of the shape and volume of the hemithorax.

The first known publication on of VEPTR outcomes dates back to 2004 [18]. It presents data on 21 patients with congenital scoliosis. The average age at the time of the first observation was 3.3 years; the average duration of follow-up was 4.2 years. Patients who had not been treated earlier displayed a significant growth of the spine: 7.7 mm per year on the concave side of the thoracic region and 8.3 mm on the convex one. 11 patients with non-segmented block displayed 7.3 % of elongation. In three patients who had been operated on previously (spinal fusion), the average elongation of the thoracic spine was 4.6 mm per year on the concave side and 3.7 mm per year on the convex one. Campbell simultaneously has developed the concept of thoracic insufficiency syndrome (TIS), which explains the mechanism of lung injury in patients with deformed chest due to blocking, deformation or absence of ribs. Pathophysiological substantiation of the syndrome has been described in Boffa et al. [14] who

reported a clinical case of a woman with untreated severe spinal deformity, who died of acute heart failure accompanied by gross lung lesions, with lung volume corresponding to that of a 6-years old child and the number of alveolar cells comparable to that of one year-old child. Campbell et al. [17] pointed out that after early epiphysiodesis children with similar pathology of the chest wall can live and maintain close to normal motor activity, but in the late teens on a background of sharply increasing body weight they will inevitably develop respiratory failure whereas pulmonary infection can lead to death as early as the third decade of life. The incidence of latent respiratory failure in young patients with thoracic deformities is unknown. Patients with restrictive lung lesions may be clinically tolerant to them for a long time, but after 40 years many require oxygen support and mortality among them increases sharply. It is believed that the development of lung tissue in children continues until the age of 8 [41]. Correction of spinal deformity and ribs at this age may contribute to normalization of growth of all thorax components including the spine and the lungs. Lung growth is no longer possible in older children. Campbell et al. [17] defined thoracic insufficiency syndrome as inability of the chest to support normal breathing and lung growth. Therefore, the said syndrome consists of two main components.

1. Status of the chest, which prevents normal respiration. The defective chest's inability to support normal secondary breathing is compensated by increase in its frequency (tachypnea) or decrease in motor activity to maintain normal levels of arterial oxygenation. These mechanisms allow a child with mild thoracic insufficiency syndrome to function normally. Worsening of chest deformation leads to further reduction in its mobility and volume, and the expansion of the lung becomes completely dependent on the function of the diaphragm. Respiratory infection exacerbates the situation. As a result, the compensation breaks down and thoracic insufficiency syndrome manifested first in a form of dyspnea and subsequently as inability to exist without oxygen or ventilatory support.

2. Inability to support normal growth of the lungs caused by rigid anomalies of the vertebrae and ribs, defined as gross forms of semimetametric hypoplasia and multiple semimetametric aplasia of the spine [2]. Such deformities limit the growth of the lungs, the volume of which becomes inadequate for normal life already in adolescence. Fatal disorders of form and function of the chest are commonly found in patients with such conditions as Jeune asphyxiating thoracic dystrophy or total shortening of the thoracic spine and hemithorax at spondylocostal dysplasia (Yarkho – Levine syndrome), which are associated with a 33 % mortality rate [45, 49, 57].

Taking into account pathological components of the syndrome, its treatment strategy should be aimed at restoring the volume and function of the chest and preservation of achieved effect throughout the growth period. The growth of the chest is a complicated process, defined by elongation of the thoracic spine and symmetrical increase of the hemithorax due to growth and spatial orientation of the ribs in accordance with the age of the child. The height of the chest is directly related to the growth of the thoracic spine, which normally grows from birth to 5 years of age at a rate of 1.4 cm/year, from 6 to 10 years at the rate of 0.6 cm/year, and from 11 to 15 years at the rate of 1.2 cm/year [22]. The deficit of the thoracic spine extension due to congenital scoliosis can be calculated by dividing its actual height by the expected age norm. The specific relationship between the loss of the hemithorax volume and shortening of the thoracic spine, as well as indirect negative impact on the growth of the lungs, remain unexplored. In any case, gross shortening thoracic spine dramatically reduces the volume of the thorax and therefore the lungs. For example, the thoracic spine in Yarkho – Levin syndrome can actually be represented by one short blocked vertebra that is not more than 1/4 of the normal height. The width and depth of the chest are also very important. In a newborn, the ribs are oriented horizontally, their elongation directly increases the diameter of the chest, whose shape in a horizontal plane approaches a square. At this age, the volume of the chest is 6.7 % from that in adults. By the age of two, the orientation of a child's ribs changes: they bend obliquely downward, and cross section of the chest becomes oval. [43] Too steep slope of the ribs flattens the chest and its sagittal diameter decreases as well as the amount that is reached at the age of 5 years (30 % of adult) and 10 (50 % of adult). During the last third of the skeleton growth period (10 to 16–18 years), the thorax expands more quickly and finally became nearly rectangular in cross-section. Approximately 85 % of lung alveolar cells are formed after birth, and the maximum increase in their number is noted at the age of two [15, 41]. As noted above, this most important process is completed by the age of about 8 years, and after its completion the lungs increase only due to hypertrophy of the alveolar cells.

Therefore, indications for VEPTR have been expanded to allow the use of this technique in different types of infantile and juvenile scoliosis with thoracic deformity in order to avoid spontaneous spinal block.

Indications for surgery: progressive congenital scoliosis in the presence of the abnormal vertebrae at the top of the deformity and at least three blocked ribs on the concave side of the arc; reduction in the hemithorax height by at least 10 % compared with the opposite side; progressive thoracic insufficiency syndrome; age of the patient at least 6 months before the skeletal maturation is complete (the younger the patient, the greater the effect on lung growth); consensus among pediatric orthopedist, pediatric general surgeon and pediatric lung specialist on the need for intervention. Contraindications for surgery: the state of the soft tissue, precluding the firm closure of an endocorrector; the state of bone tissue, precluding the possibility of metal support (as in osteogenesis imperfecta); absence of ribs required for mounting the cranial fixation; the impossibility of repeated anesthesia due to co-morbidi-

ties; active pulmonary infection; disruption of the diaphragm function.

Emans et al. [25] reported the results of surgical treatment of 31 children, 26 of whom had congenital scoliosis with blocked ribs (2 with Yarkho – Levine syndrome), 4 had iatrogenic rib block and thoracogenic scoliosis, 1 had a congenital defect of the chest wall. The average age of patients at the beginning of treatment was 4.2 years, follow up period was 2.6 years, and the average number of distraction stages was 3.5 per patient. Cobb angle could eventually be decreased from 59 to 43°. The initial volume of the lungs (according to CT examinations) was 369 cm^3, after the first surgery it was 394 cm^3, and at the end of the observation period it reached 736 cm^3. There were no fractures of endocorrector, and its displacement occurred in 8 cases and was typically corrected during a staged correction. There were no neurological complications involving CNS. There were two cases of shoulder plexitis on the side of the operation, in which the plexus function was successfully restored. In two cases, ribs separated in the first intervention fused back, which required re-osteotomy of the block. There were two cases of deep festering and two cases of rib fractures. The authors stress that VEPTR need to be implemented in the early period before the deformation become gross. In their analysis of the complications arising from the use of VEPTR technique, Campbell et al. [20] emphasize that the need for multiple repeated interventions inevitably increases the risk of complications. Each new incision is an additional risk of infection or development of bedsores. Another complication-provoking factor is multiple comorbidities. The author of the method, who, naturally, has the largest number of observations, presented his 15-year experience of treatment of complications in children with various forms of juvenile scoliosis, accompanied by deformities of the chest. Overall, 201 children were operated on in Cristus Santa Rosa Children's Hospital (Texas) in 1989–2004. On average, each of them has been subjected to surgery 7 times, with an average follow-up period of 6 years. Infectious complications were observed in 3.3 % of cases, problems with soft tissue in 8.5 %. Mechanical complications were quite common. For example, endocorrector fractures were detected in 6.0 % of patients, and their displacement in 27.0 %. The authors point out that most cases of the displacement of upper or lower end of the endocorrector were asymptomatic and were detected incidentally during the examination prior to the next stage of surgical correction. Displacement occurred slowly; the affected ribs were subjected to complete restructuring and therefore they often could be re-used as a foothold for the fixture.

The author of the method used it not only for congenital deformities, but also for infantile scoliosis of other etiologies [20]. Formation of the rib hump in the absence of rib block leads to severe deformity in which the ribs are twisted around the spinal column, resembling a folded umbrella. In 10 such cases VEPTR was used after multilevel incision of intercostal muscles, which was performed to mobilize the chest wall. Semioval endocorrector or "rib-to-rib" distractor has been used. Horizontalization of the mobilized ribs allowed the author to call this operation "open umbrella". Patients were followed up for 6 years on average. Cobb angle has been reduced from 79 to 51°. The average vital capacity of the lung amounted to 38 % of that expected for the age. Complications: 9 mechanical, 3 festering, 3 bedsores and 2 pneumonias. Campbell et al. [20] suggest that the operation increases the volume of the hemithorax.

Complications are frequent, but respond well to treatment. Emans et al. [24] analyzed the reliability of Dunn - McCarthy iliac hook in33 patients with deformities of various etiologies, 17 of whom had the hook implanted on one side and 16 had the hook implanted bilaterally. The average age of patients at the beginning of treatment was 6.1 years. Ten patients with unilateral implantation of the hook required the revision due to its displacement, which was not observed in case of bilateral implantation. With an average follow-up of 1.2 years there were 1.6 revisions per patient. Usually the hook displacement occurred asymptomatically, it retained certain stability and required reattachment only if it was located close to the *acetabulum*.

Skaggs et al. [52] examined 79 patients from seven different clinics for general changes in nutrition after VEPTR surgery. The aim of the intervention was treatment or prevention of thoracic insufficiency syndrome. Prior to surgery, 62 of the examined patients had body weight below the age norm, and 22 of them displayed marked increase in this indicator after the surgery. Of 17 patients whose weight corresponded to the age norm, 13 showed a marked increase.

Waldhausen et al. [59] presented a retrospective review of the medical histories of 22 children with pulmonary pathology who underwent installation of VEPTR in 2001-2005. Eleven children with ribs concrescence required open drainage of the pleural cavity. Revision surgery was performed in seven cases, due to resorption of bone tissue and implant displacement, and the instrumentation was removed in three cases. There was one case of superficial festering. The authors concluded that the method can reduce the retention of carbon dioxide in the blood in some patients with deformities of the chest and lung diseases.

Song et al. [56] used a slightly different technique in treatment of 14 children with neuromuscular scoliosis. They used one distractor, both ends of which were attached to the spine following the concept of growing rods. The average age of patients at the beginning of treatment was 77 months; the average time of follow up period was 15 months. During this short period of time the patients were subjected at least one staged correction. The average length of the rod implantation was 2.5 hours, of the staged distraction, 30 min. It should be noted that 8 patients in the group had underwent prior surgical treatment with other corrective devices. The average Cobb angle was reduced from 69 to 47°. Complications: 1 fracture of the rod, 2 superficial and 1 deep festering. The authors suggest that the use of VEPTR in patients with spinal deformities without accompanying gross defects of the chest wall demonstrates promising early outcomes.

Schulz et al. [51] retrospectively reviewed the results of treatment of 8 patients. All of them had been diagnosed with infantile idiopathic scoliosis. The average age at the time of operation was 45.8 months; the average scoliotic arc was 84°. During the course of treatment (median of 32 months) the deformity correction rate was 35 %, and the length of the spine increased by 7.1 cm. Three patients had complications of mechanical nature. The authors concluded that the use of VEPTR was safe and effective.

Ramirez et al. [44] reviewed their results of 17 VEPTR implantations and 33 staged surgeries. The initial value of the scoliotic arc was 59°; at the end of treatment on average was 35°. Staged lengthening of the spine and chest has been noted to be associated with certain exacerbation of the thoracic kyphosis, but its value is maintained in the normal range both before and after the operation: 23° (8–45°) and 36° (15–55°), respectively. The ratio of the space available for the lungs, improved in comparison with the preoperative (56–97 %) and postoperative (72–100 %) values. The authors describe a case of scoliosis treatment in a patient with *spina bifida*, who has develope deep festering. The instrumentation was removed; at one of the visits progression of the deformity was up to 100°. In the case of *spina bifida* and meningomyelocoele the authors recommend VEPTR as the only effective method of treatment, provided that two «rib-to-pelvis» rods are installed bilaterally using Eiffel Tower scheme [19]. The authors reported 13% of complications, including 1 deep festering, 4 migrations of pelvic hook, 1 case of broken ribs. Despite this, VEPTR technique is effective in treatment of infantile scoliosis. Smith et al. [54] published a review of their experience in treating 31 children with infantile idiopathic scoliosis. Treatment included restorative procedures, corset therapy and VEPTR. 10 patients who underwent VEPTR procedure had an average baseline value of scoliotic arc of 90° and achieved 33.8 % correction. Akbarnia and Emans [12] published a review of the literature on VEPTR complications. The risk of infectious complications in primary and staged corrections amounted to 1.9 %. The authors also identified a number of other complications: instability of support points, brachial plexus injury, scarring of the chest deformity withribs fusion, sagittal imbalance.

Hasler et al. [31] published the results of a retrospective study of treatment of 23 children with progressive deformities of the spine without rib fusion. The average follow-up was 3.6 (2–5.8) years, age at onset of treatment of 6.5 (1.11–10.5) years, the interval between corrections stages was 6 months. By etiology: 1 infantile idiopathic scoliosis, 11 neuromuscular one, 2, toracogenic ones, 1 Sprengel's disease, 2 cases of hyperkyphosis, 1 with underlying myopathy, 5 syndromic cases. There were 187 surgeries in total; the average number of staged surgeries in the patient was 6.5, of unplanned operations, 15. There were 23 complications (0.13 per surgery): 10 were trophic disorders (decubitus ulcers) of the skin, 5 were displacements of the implant, 2 were broken rods, 6 were infectious complications. The baseline value of scoliotic curve was 68° (11–111°), pelvis skew was 33° (13–60°), T1 vertebra tilt was 29° (5–84°). At the end of the follow up period scoliotic curve value was 54° (0–105°), pelvis skew was 16° (0–42°). The T1 vertebra tilt was unchanged in two patients, but improved in the remaining patients (10–58°). All patients has stable profile in the sagittal plane, two cases of hyperkyphosis with baseline values of 110°/124° improved to 56°/86°. The ratio of space available for the lungs prior to the surgery was less than 90 %, and after the surgery it improved in 9 cases and worsened in 1 case. The authors conclude that VEPTR technique is an alternative to growing rods method, Shilla, with lower number of complications, better control of the sagittal plane and pelvis skew, but with worse correction in the frontal plane. Nevertheless, the use VEPTR avoids the formation of spontaneous spinal blocks, facilitates correction of the deformity at the final stage of treatment. However, if repeated operations and anesthesia are contraindicated, this method can be overlooked in favor of Shilla.

Groenefeld and Hell [29] published a retrospective analysis of data on 1328 spinal radiographs of 57 children operated on using VEPTR procedure. The average age at the time of the initial surgery was 7.7 years, the average duration of follow-up was 29.8 months, and the average number of operations was 5.9. Overall, 24 % (n = 13) patients had spontaneous bone blocks whose development in 92 % of cases were associated with exposure to the instrumentation. In 54 % of cases spontaneous block involved the lumbar spine, in 23 % it involved the ribs and the iliac crest. During the first year of treatment, the formation of bone block was observed in 11 % of children, followed by a similar annual increase in the number of patients. Thus, after 53 months of observations signs of bone blocks formation were observed in 48 % of cases, which was confirmed by strong correlation between the rigidity of the deformation and the number of surgical steps. The authors note that in contrast to the previously published data, the spontaneous formation of bone blocks has been observed more frequently in children with rigid spine deformities and frequent staged corrections and represented a more widespread problem than previously thought.

VEPTR method gained popularity due to its ability to control progression of scoliosis in young children. Most of the children in this group have severe comorbidities, low body mass index and high risk of infectious complications. Typically, implant removal is recommended in case of infectious complications. Smith and Smith [55] published a summary of treatment data for97 patients operated on in 2002–2008, who underwent 678 surgical interventions. There were 19 infectious complications in 16 patients, amounting to 2 % of the number of surgeries. Patients had a variety of diagnoses and low body mass index. In 31 % of cases, the infection has been associated with initial implantation, in 47 % with staged correction, in 5 % with replacement and in 21 % with the revision of the instrumentation. In 13 cases the infections were classified as superficial, in 6 as deep. In 17 of 19 cas-

es, the patients developed wound dehiscence. All patients were treated with irrigation, sanitation of wounds, intravenous antibiotics. The average duration of intravenous therapy was 58 days, followed by oral suppressive therapy an average of 34 (2–126) days. Three patients required more than one debridement. In neither case was the instrumentation removed. These data suggest that an infection that developed after installation of VEPTR instrumentation without fusion procedures can be successfully treated without removing the implant. Nutrition and more careful treatment of soft tissue may be important in reducing the incidence of infection in this patient population.

Hell et al. [33] reported three complications in 15 initial implantations of VEPTR instrumentation and 13 during staged surgeries. They included rib fracture, lumbar hook displacement, formation of bed sores on the skin. Emans et al. [25] encountered 17 complications in 31 primary rib block osteotomies with VEPTR implantation and 110 in staged operations. The authors listed fractures and resorption of ribs, implant migration, deep wound infection, 2 cases of paralysis at the level of the brachial plexus with full recovery among complications.

The analysis of these relatively limited data allows drawing some conclusions. The fundamental novelty of a relatively young technique requires accumulation of experience that is identification of both positive and negative properties of the instrumentation. The duration of follow-up was rather short in all the works. In addition, there is practically no data on completed process when the final stage of the surgical treatment, the dorsal spinal fusion, is performed at the age of completion of skeletal growth. However, it seems that the increase in the volume of the defective hemithorax is an achievable goal. It is possible to control the spinal deformity, even though a decrease in Cobb angle can only be limited, which is understandable given the fact that we are dealing with congenital spine deformities that are rigid in nature. The number of complications is rather high; the majority of them should be regarded as specific for the described treatment. At the same time, most of the complications can be corrected at the next planned phase of treatment without increasing the already considerable number of surgical interventions. The attempt to solve the dual problem (prevention of thoracic insufficiency syndrome and progression of spinal deformity) using VEPTR instrumentation seems quite reasonable in theory, but is fraught with inevitable difficulties in practice. The need to perform numerous staged distractions designed to keep pace with the growth of the patient, causes a lot of problems. However, to date, that method is the only one with the proven efficacy.

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