AUTOFUSION IN SURGERY FOR EARLY ONSET SCOLIOSIS: LITERATURE REVIEW AND ANALYSIS OF OWN DATA

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The literature review considers the pathophysiology of autofusion, its frequency, and the consequences of the spontaneous bone block development in growing children, as well as the necessity of final fusion using segmental instrumentation and bone grafting in surgery for early onset scoliosis. The article presents the authors' own material on the surgical treatment of 131 patients with early onset scoliosis of various etiologies using VEPTR instrumentation, of which 84 patients completed the cycle of multi-stage treatment. During stage distractions and final fusion, the presence of spontaneous bone blocks of various localizations was ascertained. At the points of distraction rod fixation, the signs of autofusion were noted in 100 % of cases. There was not a single case of posterior vertebral autofusion along the apical and periapical zones of the main curve. In 21 patients, 22 complications were detected that required repeated intervention after the final fusion. The presented experience shows that the final stage of surgical treatment of patients with early onset scoliosis should include removal of VEPTR rods, correction of the deformity with segmental instrumentation and spinal fusion with local autobone along the entire length of the curvature.

Key Words: early onset scoliosis; autofusion; surgical treatment.

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Historical background

First experience. Until the second half of the twentieth century, treatment of early onset scoliosis consisted of observation with examinations and periodic radiography. Surgery was recommended in case of progression and included two options: immobilization with an orthotic and spinal fusion. However, spinal fusion was tended to be performed after the pubertal growth spurt. Yet small children with severe progressive deformities could not be treated with orthotics, and fusion was known to limit the growth of the spine and lungs. The first use of the Harrington distraction rod without bone grafting was performed in 1962. The primary focus of the surgery was to provide correction of the spinal deformity while maintaining its development until the onset of growth spurt. Harrington used a distractor without fusion, expecting the block would form later, before the growth spurt. He believed that scoliosis in children under 10 could be treated using only a distractor. He implanted a distraction rod and performed subperiosteal vertebral separation as in spine fusion. Harrington pointed out that this technique results in the formation of a "partial" block or cicatrization [1].

Somewhat later, Marchetti and Faldini [2] described a special technique using distal and proximal anchor points by forming a bone block of two vertebrae at each end of the instrumented area (the "end fusion" technique). The spine was exposed in a subperiosteal manner, and supplemental correction was reached periodically with a distractor.

In 1979, Moe et al. [3] implanted the Harrington rod subcutaneously in small children, while only the hook installation sites were exposed. In this paper, Moe et al. briefly mention autofusion in a series of 20 children, nine of whom underwent posterior spinal fusion as the final stage of treatment. Four cases revealed autofusion, but none at the apex of the deformity, but only at its ends.

In 1982, Luque [4] used the Harrington distractor with sublaminar wires in children under 8. Subperiosteal separation was performed only on the concave side. The spontaneous block was formed only in 6 out of 47 patients. Eberle [5] revealed the same outcome in 7 patients out of 19 operated on along 5–8 segments.

The problem of autofusion in the posterior spine began to emerge almost from the beginning of the development of the surgical treatment of early onset scoliosis. Nowadays, its main aspect is to decide whether a posterior fusion is appropriate as the final stage of treatment, i.e., the so-called final spinal fusion. This issue has grown in interest during the past few years.

Definitions and classification. The term "autofusion" refers to the presence of bone blocking masses at the levels of the spinal column where spinal fusion has not been intentionally performed [6]. This definition does not include the areas of ends of the corrective instrumentation where fusion may or may not have been planned. On visual inspection, the autofusion area is a bone layer that is not very different from the result of posterior fusion.

In the fundamental publication by Menapace et al. [7], who studied in detail a group of 28 patients operated on using traditional growing rods (TGR) and magnetically controlled growing rods (MCGR), it was found that if two adjacent vertebrae move together as a unit and/or are connected by a bone bridge, such a segment is in the autofusion state. In contrast, if two adjacent vertebrae move independently of each other, such a segment is reported as autofusion-free one. The assessment was performed independently by two experienced spine surgeons. According to the data obtained, Menapace et al. [7] proposed an estimate of the extent of autofusion obtained by dividing the number of blocked segments by the total number of segments in the altered section of the spinal column: grade I – 0 to 25 %, II – 25 % to 50 %, III – 50 % to 75 %, IV – 75 % to 100 %, and V – 100 %. Herewith, grades I–II are considered low, and grades III–V are considered high.

The authors have also studied practically crucial issue of risk factors for the development of autofusion. Out of the total list of 22 items, they were able, based on detailed statistical analysis, to identify both risk factors and so-called protective factors, in the presence of which the possibility of developing autofusion is considered to be reduced.

Risk factors include the age of the first growing rod implantation being under 8 years, unplanned surgeries during staged distractions, and the Cobb angle of the main curve after the first distraction being over 30°. *Protective factors* include preoperative spine length (T1–S1) greater than 30 cm and primary MCGR implantation (in a number of patients, primary implanted TGRs were replaced by MCGRs during treatment).

According to Cahill et al. [8], autofusion may be a biological reaction to immobilization of a non-completed growth of the spine. Another risk factor may be long (over 10 months) intervals between distractions.

In 2014, Zivkovic et al. [9] regarded the development of autofusion as a largely benign biological response to the patient's routine motor performance in the presence of a polyaxial fixed implant. The features of the biomechanics of the trunk, spine, and thorax provide an explanation for the different incidence of autofusion development under VEPTR application conditions. The authors have proposed a classification of autofusion depending on the localization of the newly formed bone masses:

• type I – at the implant anchor points (ribs, semi-arches, iliac crest);

• type II – along the longitudinal axis of the distraction rod, including above the ribs, in the lumbar spine;

• type III – re-ossification in the area of separation of congenital rib blocks.

The authors have presented the treatment outcomes of 65 patients; 42 (65 %) of them had autofusion: 60 of type I, 54 of type II, 5 of type III.

The first record of autofusion development refers, surprisingly, to the cervical spine. In 1975, O'Brien [10] described this condition as a complication of halo-pelvic traction. A little later, Dove [11] presented a detailed description of autofusion case series. All these findings concerned patients who underwent halo-pelvic traction for severe spinal deformities of various origins (tuberculous kyphosis, idiopathic scoliosis), and in all cases bone blocks were formed in the cervical spine. Five cases from a group of 83 people treated with a halo-gravity traction device were included. The mean age of halo-treatment onset was 13.6 years; its mean duration was 10.4 months. Both anterior and posterior vertebral regions could be involved in the area of autofusion (Fig. 1). The authors failed to formulate the reason for autofusion development, even though they did not exclude that when using a large corrective force (more than 60 % of body weight), the sites of ligament attachment to the bone are injured with local hemorrhage and subsequent bone tissue formation.

The pathophysiology of autofusion has not been adequately studied. Chalmers et al. [12] were apparently the first to analyze the possibility of bone development in soft tissues (pancreas, liver, kidneys). They placed up there bone grafts treated with hydrochloric acid as an inducing substance. The tissues of the above organs suppressed osteogenesis, in contrast to muscle and fascia. The authors concluded that three conditions are essential for bone induction in soft tissues: an inducing substance, osteoprogenitor cells, and an environment that supports the osteogenesis process. The ossification of body tissues or its absence may depend on the balance of osteogenic and osteogenesis-suppressing factors acting both locally and systemically. However, the question remains far from being resolved.

Contemporary authors are inclined to believe that the formation of autofusion may be the result of a number of factors: immobilization; local damage to the paraspinal muscles, periosteum, and soft tissues; direct contact between metal and spine; and, finally, the inherent ability of woven bone to fast fracture union. Bosch et al. [13] suggested that osteoprogenitor cells exist in skeletal muscle and confirm indirectly the relationship between muscle and bone tissue precursors. It is still unclear which cells in muscle tissue are responsible for osteogenesis.

In 2005, Martinez et al. [14] suggested the influence of periosteal injury on the formation of autofusion. They described osteotylus formation after induced stress fracture of the radius without fragment displacement with a periosteal defect in mice in their study. The response of the fractured bone depended on the severity of periosteum injury induced by mechanical stress. Still, regulation mechanism of this process remained incompletely understood. While discussing this study, Groenefeld and Hell [15] emphasize that when using VEPTR, periosteal injury is associated with surgical procedures, the flexibility of the graft, its migration, and the increasing pressure of the metal on the bone at their junctions. The same authors note a considerable correlation between the development of autofusion and the extent of scoliotic deformity correction during the first surgery: children with more rigid spinal deformities and less scoliotic deformity correction are more likely to develop autofusion.

In 2020, Huber et al. [16] studied gene expression at the site of tissue damage (comparable to tissue damage in early onset scoliosis surgery). Using single-cell RNA sequencing, early increased activation of gene expression of cell adhesion regulation and extracellular matrix-receptor interactions resulting in bone or cartilage development were demonstrated.

The autofusion formation in patients with early onset scoliosis has been reported not only in the spinal region when it comes to expansion thoracoplasty. Betz et al. [17] showed no significant differences in the incidence of autofusion between two groups of idiopathic scoliosis patients who underwent posterior fusion with or without grafts. A half of a group of 91 patients were treated with posterior instrumentation. The lack of fusion did not matter – the block developed in all operated patients. Therefore, the suggestion is that autofusion may be a common physiological response to immobilization of the immature spine. Fisk et al. [18] emphasize that the reason for autofusion development remains unknown; however, they believe that repeated distractions may cause microhemorrhages with subsequent osteogenesis.

Frequency of autofusion formation. This point attracts special attention of everyone but the number of reliable data is very limited (Table 1). In an attempt to show that autofusion is formed very often, many authors refer to the paper by Cahill et al. [8], according to whom this complication occurs in 89 % of cases. We accumulated information suggested that the frequency of autofusion formation is considerably lower, yet it is highly variable. Autofusion was found in 139 (48.1 %) of 289 patients with early onset scoliosis of various origin who underwent final spinal fusion after staged distractions. We did not consider the data of Fisk et al. [18] and Sestero, Perra [19] since it is a short, specially selected series, we have included it in Table 1, as these data proved interesting in its own right.

Consequences of autofusion development in growing children. The fact that the formation of autofusion limits the growth of the vertebral column and prevents the correction of scoliotic deformity using staged distractions has been known since the use of growth-friendly surgery in children with early onset scoliosis. It is autofusion that is regarded as the reason for the "law of diminishing returns". In 2011, Sankar et al. [24] described this phenomenon. The core of this phenomenon is summarized in the following statement: "In the staged treatment of early onset scoliosis, each subsequent distraction is less effective than the previous one." According to Sankar et al., the first distraction provides almost 50 % correction, and subsequent distractions generally increase the corrective effect insignificantly. The same trend was found when measuring the length of the spine (T1–S1). According to the authors [24], "a possible explanation for the reduced corrective effect could be the progressive rigidity of the immature

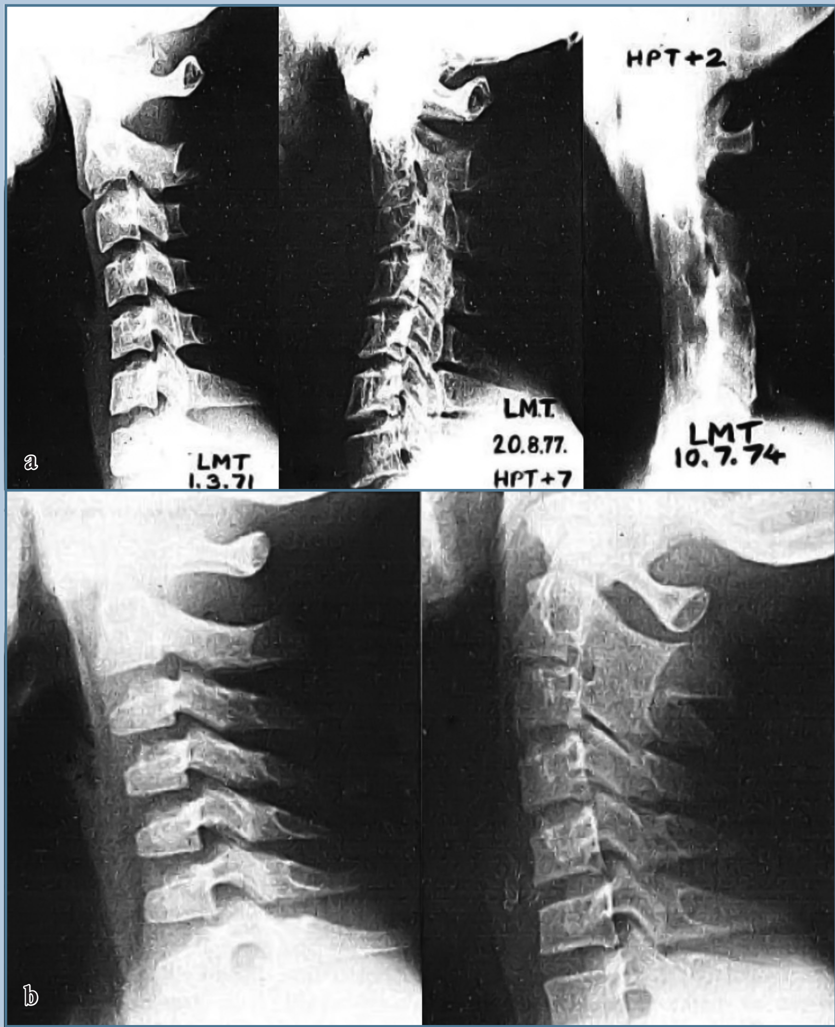

Fig. 1
X-ray images and CT scans of the cervical spine after the stage of halo-pelvic traction in a patient with severe scoliotic deformity of the spine **(a)**: CT scan shows signs of autofusion of the cervical vertebral bodies [10]; X-rayy of the cervical spine after the stage of halo-pelvic traction **(b)**: signs of autofusion of the posterior regions of the C2 and C3 vertebrae [11]

vertebral column, resulting from the presence of instrumentation or even the development of autofusion." We have not become more aware of autofusion over the past 40 years.

Noordeen et al. [25] studied intraoperatively applied distraction force (60 surgeries in 26 patients) and found that it sharply increases during the fifth distraction compared to the previous one. Moreover, the linear magnitude of elongation progressively decreases during each distraction, reaching a minimum (8 mm or less) during the same fifth distraction.

The effect of autofusion on the body length is not well understood. Despite the Sankar rule, the growth throughout T1–S1 is preserved at a normal level during stage treatment [24]. This phenomenon may be attributed to the biological activity of the bone masses responding to distraction forces during staged lengthening.

Autofusion and magnetically controlled growing rods. It was initially assumed that MCGR technology does not predispose to the formation of bone blocks in the distractor bed area for two reasons. The first one is fewer surgeries, i. e. minimizing injury to the paravertebral muscles and bony structures of the spine. The second one is increase in the number of distractions with less elongation, thereby promoting the maintenance of a prolonged distraction force. In 2017, Gardner et al. [26] showed, and rather convincingly, that the Sankar rule does not work when using magnetically controlled growing rods, at least not as well as with traditional distracting rods. 53 magnetically controlled growing rods were implanted in 28 patients; the mean number of elongations was 10 over two years. Meanwhile, no significant difference was observed in the achieved elongation sections of the system. In contrast, Hatem et al. [6] stated the existence of three articles reported that the use of MCGR was followed by recorded formation of autofusion. The first paper was about patients with Ehlers – Danlos syndrome, the second one – with Prader – Willi syndrome, and the third one – with cerebral palsy cases were discussed. It is still unclear what the cause is (failure to elongate, block formation) and what is primary. Gilday et al. [27] revealed a correlation between the amount of distraction and the distance from the skin to the magnetic actuator — 2.1 % elongation per 1 mm of tissue depth. Another possible cause of failure when using MCGR is slippage. Cheung et al. [28] defined it as the inability of the internal magnet to make a full turn, followed by stopping and returning to the initial position. The risk factor is the distance from the outer magnet to the actuator and the increased distance between the inner magnets. The patient described by Cheung et al. had a growth spurt; therefore, the distance between the controller and the actuator increased rapidly.

How can we limit the development of autofusion? There are two growing rod systems designed to reduce the risk of autofusion development. One of them is semiconstrained growing rods (SCGRs). Their difference is the ability to perform axial rotational displacements of one component relative to the other. Bouthors et al. [29] studied the treatment outcomes of 28 patients and stated that final spinal fusion gave an additional correction of 20.3°, and the increase in the T1–S1 distance amounted to 31.7 mm, i.e., the obtained data implicitly indicated a minimal level of autofusion formation. The number of complications was low — 0.096 per 1 patient per year (according to the literature [30], this number reaches 0.32 per 1 patient per year when using traditional rods). The second system is the so-called minimally invasive bipolar technique proposed by Miladi [31]. The technique is based on gradual correction of the deformity using the viscoelastic properties of the torso tissues. The telescopic structure overlaps the deformity and maintains tension between its ends. Proximal anchor: two laminar-pedicular pairs on both sides over 4–5 segments. The distal anchor is formed from pedicle screws — 2–3 on each side. The anchors are connected firmly with one or two rods and with minimal soft tissue injury to reduce the risk of scarring and autofusion. Initial outcomes are promising.

Autofusion and final spinal fusion. A number of authors with considerable clinical experience have expressed their opinion on final spinal fusion.

Rinsky et al. [20] reported nine operated children (mean age 8.5 years) treated without final spinal fusion. After 28 months, there was a 32 % loss of achieved correction with three broken rods, spinal growth of 0.8 cm instead of 2.1 cm in the normal range. There was no evidence of autofusion; late fusion was complicated by pronounced fibrosis.

Cahill et al. [8] noted a 44 % (48.7 to 24.4) correction of the main curve during the final spinal fusion. Akbarnia et al. [32] reported only 24 %. Meanwhile, in patients treated in adolescence for severe idiopathic scoliosis and not subjected to stage distraction, the correction is 60–70 % of the initial curve [33, 34].

Flynn et al. [22] performed final spinal fusion in 79 of 92 operated patients. The mean patient age at the time of surgery was 12.4 years. The correction obtained during final spinal fusion was minimal (less than 20 %) in 18 % of patients, moderate (21–50 %) in 48 %, and significant (greater than 50 %) in 15 %. Among 58 patients who underwent final spinal fusion (with the entire folder of documents), 47 (81 %) had areas of autofusion, and the spine was rigid or completely immobile. 22 patients underwent vertebrotomy, and seven patients underwent thoracoplasty. Final spinal fusion is performed in most patients, but usually the correction achieved during this procedure is less than 50 % of the Cobb angle at the end of the distraction phase.

Jain et al. [35] provided the surgical outcomes of 167 patients; final spinal fusion was performed in 137 of them, and the distraction rods were not removed in the remaining patients. There were no considerable differences in the results of distraction sessions between these groups. The authors conclude that patients with early onset scoliosis and signs of skeletal maturation with satisfactory correction of body shape and growth, with minimal effect of the last distraction and in the absence of implant-related complications (IRC), can avoid final spinal fusion. If spinal dis-

Table 1
Frequency of autofusion formation in patients operated on for early onset scoliosis of various origin

Study	Patients, n	Spinal deformity origin	Corrective instrumentation	Age at the first distraction, years	Cobb angle before treatment, degrees	Cobb angle before final spinal fusion, degrees	Cobb angle after final spinal fusion, degrees	Number of final spinal fusions	Number of cases of autofusion, n
Rinsky et al. [20]	9	–	SSI	8.5	–	–	–	9	0
Moe et al. [3]	20	–	Harrington	–	–	–	–	–	4
Luque [4]	47	Paralytic scoliosis	Luque	–	–	–	–	–	6
Eberle [5]	19	–	Harrington	–	–	–	–	–	7
Mardjetko et al. [21]	9	Various	Luque	9.0	51.0	27.0	51.0	9	9
Fisk et al. [18]	3 (series)	Various	Harrington	12.0 2.7 5.4	73.0 55.0 75.0	45.0 89.0	45.0	–	1 1 1
Sestero, Perra [19]	2 (series, siblings)	Central core disease	Segmental	14.0 17.0	187.0 108.0	75.0 64.0	–	–	1 1
Cahill et al. [8]	9	–	GR	4.8	72.6	48.7	24.4	9	8
Flynn et al. [22]	58	Various	GR	12.4	–	–	–	58	47
Lattig et al. [23]	5	Various	VEPTR	10.0–15.0	96.0	74.8	50.0	5	4

traction is possible within less than 1 cm at the last surgery, this is an indication of autofusion, which allows spinal fusion to be unnecessary. However, autofusion may be incomplete, the bone mass may be thin and precarious, and the block may be unendurable.

Sawyer et al. [36] noted that the correction of scoliosis (17 %) and maximal kyphosis (15 %) during final spinal fusion is rather less than expected in typical cases of early onset scoliosis, for example, in patients who have not undergone prior surgery.

Kocyigit et al. [37] presented the outcomes of two different types of final surgery: implants were removed in patients with stable radiographic findings, but instrumented spinal fusion was not performed; in the second group, where the correction was insignificant, the growing rods were removed and then posterior spinal fusion was performed with segmental instrumentation. 9 of the 10 patients in the first group showed significant progression of the deformity after removal of the growing rods. This treatment mode was eliminated from further practice for ethical reasons, since it does not give grounds to expect a reliable autofusion. The authors emphasize that such a protocol is impractical.

Ahuja et al. [38], in a review including data from 11 studies, found no significant differences between the outcomes of two surgical approaches: preservation of distraction rods, removal of distraction rods + posterior spinal fusion using segmental instrumentation. The authors did not find differences between the groups in such important parameters as Cobb angle, spine height (T1–T12, T1–S1) at the beginning of the staged distraction period and after completion of treatment. Significant differences were found only in the number of revision surgeries: they were performed more often in patients with final spinal fusion performed than in patients with preserved distraction rods.

Autofusion and unintended surgeries. Although small, correction is achieved through a complex and time-consuming surgery that may include osteotomy of the formed autofusion. The frequency of osteotomy can be as high as 24 [20] and even 30 % [39]. According to Flinn et al. [22], an additional anterior release of the vertebral column was required in 13 % of cases.

Moreover, the presence of autofusion unavoidably changes the anatomy and points required for placement of multiple pedicle screws. This lengthens the surgery duration and increases the risk of mistakes and complications. Du et al. [40] analyzed the risk factors for revision surgery after final spinal fusion in 167 patients treated with traditional rods. The total number of such surgeries was 32 (19 %). It turned out that patients who required revision surgery after final spinal fusion were treated with staged distraction longer than usual, and the number of levels covered by distraction rods and treatment duration were directly associated with the need for revision surgery after final spinal fusion.

The number of complications after final spinal fusion can be quite significant, which does not always qualify it as a final surgery. Poe-Kochert et al. [41] found 30 complications (1.5 complications per patient) in 20 out of 100 patients with two-year long-term outcomes of multi-stage treatment and final spinal fusion. 57 additional surgeries were required. Parents of patients should be reminded that "final" does not always mean "last". Sawyer et al. [36] reported revision surgeries in 24 % of cases: implant removal, restoration of implant integrity, infection, or junctional kyphosis. Murphy et al. [42] performed revision surgeries in 22 % of cases with a follow-up period of up to five years.

Studer et al. [43] presented the treatment outcomes of 34 patients, 17 of whom underwent final spinal fusion. The initial magnitude of the scoliotic curve in these patients was 73°. The final surgery corrected the deformity by 14 %, but a considerable loss of correction was further noted. The total number of complications in 34 patients was 65 (on mean 1.9 complications per a patient); patients who underwent final spinal fusion had complications in 41 % of cases (17 cases in 7 patients) and required 6 repeated surgeries. These authors, as well as a number of others, concluded that final spinal fusion is not always the last surgery.

The paper by Menapace et al. [7] is the most comprehensive study of the problem of autofusion in surgery for early onset scoliosis. The most important conclusion of the authors is that autofusion is not a clear obstacle to performing final spinal fusion. It is possible to achieve elongation of the torso and maintain correction even with extended autofusion. Generally, there is no consensus on the necessity of this surgery.

Own data

The vertical expandable prosthetic titanium rib (VEPTR) instrumentation has been used in the clinic of pediatric and adolescent spine surgery at the Novosibirsk Research Institute of Traumatology and Orthopaedics n.a. Ya.L. Tsivyan since 2008 in the treatment of early onset scoliosis. This technique was first developed in 1987 by the American surgeon Campbell [44]. We are experienced in the surgical treatment of 131 patients with early onset scoliosis of various origins, 84 of whom completed the cycle of multi-stage treatment. 892 staged distractions (including primary placement) and 84 final surgeries were performed. Therefore, the mean number of staged distractions per a patient was 6.8.

84 patients (37 boys, 47 girls) underwent final spinal fusion. The mean age of the patient at the first distraction was 4.5 ± 2.1 years, and it was 14.4 ± 2.1 years at final spinal fusion; the mean postoperative follow-up period was 6.2 ± 1.8 years.

According to origin features, the patients were subdivided as follows: with congenital scoliosis – 33, with idiopathic scoliosis – 27, with syndromic scoliosis – 22, and with neuromuscular scoliosis – 2.

The technique of the final surgery in 74 cases was the replacement of VEPTR instrumentation with modern segmental instrumentation (Legacy, Esculap, ExPed, NITEC) combined with posterior spinal fusion with local autograft. The following surgeries were performed in the remaining 10 cases: posterior spinal fusion without VEPTR removal – 5, VEPTR removal because of self-correction of the deformity – 2, VEPTR replacement with traditional growing rods – 2, revision of the bone block – 1.

The changes over time of the main radiometric parameters describing the state of the deformed spine are given in Table 2.

Outcomes of VEPTR application. The initial value of the main curve was rather severe on mean (93.2°), but was reduced by more than 35° during the first distraction that is almost 40 % of the initial value. Later, at the distraction stage, it was possible to preserve most of the achieved effect, with a loss of only 17°. During final surgery, the deformity was reduced by almost 40 % and subsequently almost completely preserved – the loss of correction in the long term after the surgery was only 3°. Meanwhile, the value of thoracic kyphosis and lumbar lordosis remained within normal parameters at all stages of treatment that corresponded to the positive changes of frontal imbalance. The length of the thoracic and lumbar spine during treatment increased at a rate corresponding to the normal development of the spinal column.

We have constantly noted (both during stage distractions and during final spinal fusion) the presence of autofusion of various localizations, and all findings are within the classification of autofusion proposed by Zivkovic et al. [9] in 2014. At the points of fixation of distraction rods (type I — upper thoracic ribs, lumbar vertebral semi-arches, iliac crests), signs of autofusion were detected in 100 % of cases. In the region of cranial anchor, blocks of adjacent ribs were often formed and could be used as new anchor points for distractor (Fig. 2). Autofusion in the lumbar spine was noted in the area of laminar hook placement and cranially, where the distractor rod was closely adjacent to the posterior regions of the spine (Fig. 3). In the case of anchoring to the iliac crest, metal subsidence into the bone depth with small osteophytes around this area was always recorded.

As for type II autofusion (blocks along the distraction rod length), the pattern was quite different. We have never observed posterior vertebral blocks along the apical and periapical zones of the main scoliotic curve. This circumstance is what we tend to explain the considerable (29°) correction of the main curve during final spinal fusion. Spontaneous fusion of ribs located under the body of the distractor was also not found in any case. Type III autofusion (rib fusion after dissection of congenital blocks) was noted in all cases, but these cases are few among our patients.

22 complications were identified in 21 patients requiring repeated surgery after final spinal fusion. This consisted of 14 instrumentation remounting surgeries (rod fractures, junctional kyphosis, spinal imbalance, lack of anchor points), 4 surgeries for surgical site infection, and 2 anterior spinal fusions, including one anterior decompression of the spinal cord in case of continued progression of the curve associated with neurofibromatosis, 1 hematoma debridement, and 1 duraplasty for liquorrhea.

Discussion

Surgical treatment of early onset scoliosis is a challenging issue in spine surgery. Severe progressive spinal deformity (frequently combined with multiple comorbidities) dramatically changes the life of children and their families, requiring long-term multi-stage surgical treatment. Despite considerable collective experience and many publications, the problem is far from being resolved.

One of its significant aspects is the development of vertebral and rib autofusion in the area of placement of instrumentation system. According to the literature, currently the nature of this phenomenon remains unclear, and no reliable techniques for prevention have been developed. Different types of autofusion probably have an ambiguous effect on the outcome of multi-stage surgeries. In the course of stage distractions, their efficiency progressively decreases (the Sankar rule), and it drops to almost zero after 5-6 distractions. The achieved correction can be only partially preserved until the skeletal formation is complete; therefore, performing final spinal fusion, as the author of the technique intended, seems to be completely justified and reasonable. Nevertheless, in practice, many surgeons prefer the classic option of retaining the VEPTR distractors without replacing them with segmental instrumentation or removing them without replacing with other instrumentation. In the first part of this paper, we tried to show that this discrepancy in strategy is very common and that there is no consensus.

Our own experience shows that the processes of autofusion formation are controversial. We have never observed autofusion of the vertebrae in the region of the main scoliotic curve, both its apex and compensatory regions. We can explain this mismatch with the data of other authors only by one circumstance. We considered it essential to follow Dr. Campbell's recommendations for VEPTR distractor placement [44] with the strictest care in all cases. We have never exposed the area of the main scoliotic curve, even minimally. The rod was placed subcutaneously and fixed to the upper and lower anchors: cranially to the ribs, caudally to the lumbar vertebrae, or caudally to the iliac crest. Due to the severity of the scoliotic curve, the rod was always placed lateral to the spine – above the ribs of the concave side of the curvature. We have seen similar observations in the literature only once – the aforementioned study by Moe et al. [3], who found the lack of autofusion in all four patients treated by them.

The dynamic analysis of spinal deformity magnitude showed (Table 2) that the deformity increased by a mean of 17° (30%) between the first distraction and final spinal fusion, and the correction reached 30° (29%) as a result of the last treatment. We consider these data to indicate high mobility of the main curve, throughout which autofusion is not observed (Fig. 4). We assume that this fact proves that the final stage of surgical treatment of patients with early onset scoliosis should include removal of VEPTR rods, deformity correction with segmental instrumentation, and spinal fusion with local autograft along the entire curvature. This is indirectly

Table 2

Changes over time of radiometric parameters of spinal deformity in patients who received a full course of multi-stage treatment for early onset scoliosis

Parameter	Before the first distraction	After the first distraction	Before final spinal fusion	After final spinal fusion	At the end of follow-up
Main curve, degrees	93.20 ± 14.80	56.9 ± 10.00* (correction – 38.9 %)	73.8 ± 11.6 (increase – 29.7 %)	44.8 ± 11.1 (correction – 39.3 %)	47.8 ± 14.1** (increase – 6.7 %)
Counter-curve, degrees	42.80 ± 16.00	31.8 ± 12.8* (correction – 25.7 %)	38.3 ± 11.8 (increase – 15.2 %)	36.4 ± 6.8 (correction – 5.1 %)	32.4 ± 8.4** (correction – 9.9 %)
Thoracic kyphosis, degrees	41.10 ± 11.90	35.6 ± 10.4* (correction – 13.4 %)	60.5 ± 10.6 (increase – 32.0 %)	34.0 ± 9.3 (correction – 43.8 %)	24.5 ± 8.5** (correction – 59.5 %)
Lumbar lordosis, degrees	49.50 ± 4.90	48.6 ± 9.0* (correction – 1.9 %)	41.3 ± 13.4 (correction – 16.6 %)	45.5 ± 8.5 (correction – 9.9 %)	38.4 ± 5.1** (correction – 6.6 %)
Pelvic tilt, degrees	8.67 ± 5.20	4.3 ± 2.2*	4.2 ± 1.9	3. 3 ± 2.2	4.4 ± 2.7**
Frontal imbalance, mm	38.20 ± 12.50	24.0 ± 18.4*	32.3 ± 17.4	32.0 ± 9.6	18.0 ± 4.7**
T1–S1 length, mm	270.00 ± 18.40	293.3 ± 22.5*	325.5 ± 17.2	389.5 ± 11.8	392.5 ± 10.6** (total growth – 122.5 mm)

* Statistically significant differences at $p < 0.05$ compared to the initial value;

** statistically significant differences at $p < 0.05$ compared to the value after surgery.

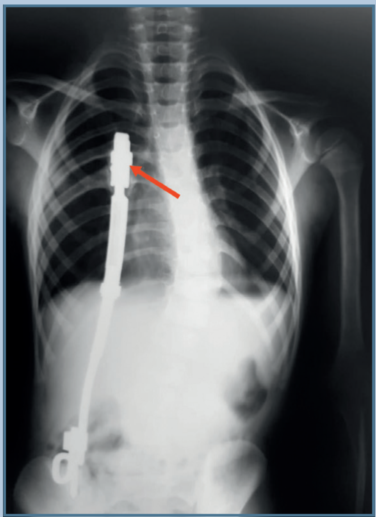

Fig. 2
Osteophytes surrounding the implant are visible in the area of the rib anchor

Fig. 3
Intraoperative image of posterior regions of lumbar vertebrae during final spinal fusion: the semi-arches and spinous processes of the T12–L2 vertebrae are fused together into a jointless bone structure

proved by the changes of such indicators as trunk balance and the length of the thoracic and lumbar spine (Table 2). The above-mentioned published data show that the number of complications (and, accordingly, unplanned surgeries) is generally not higher than that reported by other authors.

Conclusion

The main conclusion that we consider necessary to make is that in children with progressive early onset scoliosis of various origins, the multi-stage surgical treatment should include replacement of distraction rods with segmental instrumentation and autologous bone grafting in spinal fusion. As far as we can determine, our data is one of the largest,

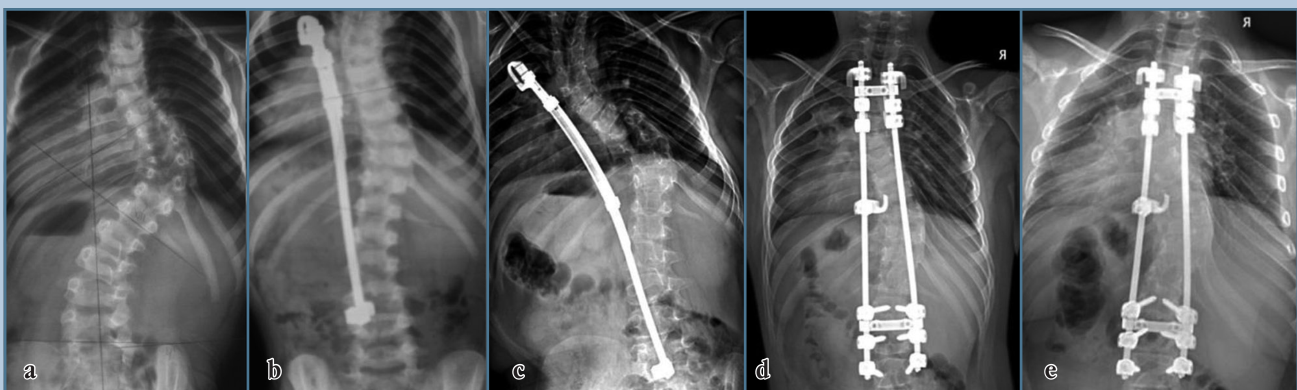

Fig. 4
X-ray images of a 7-year-old patient with idiopathic right-sided thoracic scoliosis (71°): **a** – before surgery; **b** – after correction with VEPTR rod (rib-spine), the deformity was reduced to 35°; **c** – before final spinal fusion, the spinal deformity increased to 61°, frontal imbalance of the trunk developed; **d** – after final spinal fusion, the spinal deformity was reduced to 37°, the trunk imbalance was eliminated; **e** – 4 years after surgery, the deformity progressed by 2°, the trunk balance is not disturbed

but regardless of this fact, new studies are still required that can reliably justify one surgical strategy or another for the treatment of patients with early onset scoliosis.

The drawback of this paper is the lack of data concerning the quality of life of patients undergoing multi-stage surgical treatment for progressive spinal deformities of the first decade of life. We have such information and plan to devote a separate publication to it.

The study had no sponsors. The authors declare that they have no conflict of interest.
The study was approved by the local ethics committee of the institution. All authors contributed significantly to the research and preparation of the article, read and approved the final version before publication.

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