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VEPTR INSTRUMENTATION IN THE SURGERY FOR INFANTILE AND JUVENILE SCOLIOSIS: FIRST EXPERIENCE IN RUSSIA*

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Objective. To analyze results of new method of treatment for progressive infantile and juvenile scoliosis of different etiology using VEPTR instrumentation.

Material and Methods. Twenty-two patients aged 1.5-8.7 years were operated on, most of them for idiopathic and congenital scoliosis. The mean value of primary scoliotic curve was 66.3°. Besides routine investigations all patients underwent MRI and MSCT of the spine, as well as thorough examination by a neurologist, pediatrician, and specialized doctors. The arrangement of endocorrector depended on etiology and type of the spine deformity.

Results. The first operation reduced the primary scoliotic curve angle to 47.9°. Correction was 18.4° (27.8 %). Before the first stage distraction in 12 patients, the mean deformity angle was 65.5° , and after expansion -54.0° . Only two patients underwent more than two stage distractions. Complications were noted in two cases. Despite the large number of comorbidities, all patients showed tolerance to the performed intervention.

Conclusion. The principal novelty of the method of treatment for progressive infantile and juvenile scoliosis requires accumulation of experience and revelation of advantages and drawbacks of the VEPTR instrumentation.

Key Words: infantile scoliosis, juvenile scoliosis, VEPTR instrumentation, osteotomy of costal fusion, staged distractions.

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The most challenging problem in spinal surgery is treatment for progressive infantile and juvenile scoliosis of different etiologies [5]. Spinal deformities are detected early (in case of congenital scoliosis, almost within the first days and weeks of life) and rapidly progress, resulting in body ugliness and often leading to the thoracic insufficiency syndrome [1]. At the age when the locomotor system becomes mature, such patients in fact become incurable.

Conservative treatment for progressive scoliosis in children is considered to be completely unfavorable. Zeller [14] has attempted to use plaster jackets in such patients until the age when the fusion surgery becomes feasible, but the results are unknown. Multiple variants of the instrumental correction without fusion performed during the first years of child's life followed by staged distractions were also unsuccessful [8-11].

In 1987, Dr. Campbell in San Antonio (Texas, USA) developed a novel endocorrector aimed at controlling the progressive spine deformity, as well as normalizing the respiratory function of the rib cage [2]. A silicon plate for prosthesis of the thoracic wall defect, fixed by Steinmann pins was originally used. In 1996, Synthes company developed a new design of the plate; clinical trials started in the USA. In 2002, VEPTR (vertical expandable prosthesis titanium rib) instrumentation was used in Europe (Basel, Germany) for the first time.

The first surgeries using the VEPTR instrumentation in Russia were conducted on the 4th of April, 2008, by Prof. Rudiger Krauspe from the D sseldorf University (one of the authors of this paper was trained in the D sseldorf University Hospital). In autumn 2008, surgeons of the Clinic of Children Vertebrology of Novosibirsk Research Institute of Traumatology and Orthopedics

started performing surgeries using the VEPTR. In early 2009, the first two surgeries were performed in St. Petersburg Children City Hospital № 1. The resulting experience is rather sparse; however, given the novelty of the method that is not familiar to Russian physicians, we decided to provide and analyze the first results. The data obtained at two hospitals are provided; multi-centered studies are of special importance in evidencebased medicine.

The objective was to analyze the results of applying the new method of treatment for progressive infantile and juvenile scoliosis of different etiology using VEPTR instrumentation.

Material and Methods

Since April 2008, 23 children (14 girls and 9 boys) have been operated on at two hospitals. The median age of patients when the treatment was started

was 5.4 years (range: 1.6–8.7 years). Distribution of patients over etiology was as follows (Table 1): idiopathic scoliosis – 7, congenital scoliosis (including patients with Jarcho–Levin syndrome) – 14, Kartagener syndrome – 1, neurofibromatosis – 1. Twenty-one children had accompanying diseases; 14 children were operated on earlier; many of them were operated on several times (4 – due to spine deformities) (Table 2).

An examination was performed before surgery, including routine methods, MRI and MSCT of the spine, a comprehensive inspection of a patient by a neurologist, pediatrician and specialized physicians. Due to the age of children, the respiratory function was examined only in several patients.

The endocorrector was arranged depending on the etiology and type of spine deformity (Table 1): rib-to-rib, rib-to-spine – 8 patients, rib-to-spine – 11 patients, rib-to-pelvis (1 or 2 rods) – 4 patients (Fig. 1). In 9 cases, only primary correction of the deformity of the rib cage and spine was performed, in 12 – one staged distraction, in two more cases – two and three-staged distractions (one for each patient). The mean interval between the treatment stages was 8.2 (4–12) months.

Indications for surgery included:

- progressive congenital scoliosis, when a vertebrae are present at the deformity

apex and more than three fused ribs on the concaved side of the arch;

- decrease in the height of the hemithorax by at least 10 % as compared to the opposite side;
- progressive thoracic insufficiency syndrome;
- patient's age is at least 6 months before the skeleton becomes mature; the intervention will affect the lung growth stronger in younger patients;
- agreement between the decisions of pediatric orthopedist, surgeon, and pulmonologist regarding the need for surgery

Contraindications for surgery:

			nts opera					
Patients	Gender	Age at the start of treatment	Number of stages, n	Interval between stages, mns	Etiology of scoliosis	Arrangement type	Height, cm	Weight, kg
4.01		4.70	0	2.10			04.0.400.7	455 450
1 st	m	1.58	2	6;12	Congenital	Rib-to-rib, rib-to-spine	91.0-100.5	13.7-15.9
2 nd	m	4.25	3	6;7;7	Congenital	Rib-to-rib, rib-to-spine	100.0-110.0	15.4-18.4
3rd	f	5.75	1	8	Congenital	Rib-to-rib, rib-to-spine	107.0-113.0	18.4-19.1
4 th	m	5.00	1	12	Congenital	Rib-to-rib, rib-to-spine	94.0-103.0	15.5-17.85
5 th	m	3.17	1	12	Congenital	Rib-to-rib, rib-to-spine	89.0-95.5	13.6-15.2
6 th	f	5.00	1	12	Congenital	Rib-to-spine	109.5-117.0	16.2-17.6
7 th	f	3.92	1	12	Congenital	Rib-to-spine	90.0-92.5	13.9-14.5
8 th	m	7.33	1	12	Idiopathic juvenile	Rib-to-spine	105.0-110.0	16.2-17.9
9 th	f	6.42	1	10	Kartagener syndrome	Rib-to-pelvis	105.5-116.0	19.7-21.0
10 th	m	2.83	1	8	Congenital	Rib-to-spine	90.0-92.0	12.0-13.4
$11^{\rm th}$	f	4.75	1	8	Idiopathic	Rib-to-spine	104.0-108.0	15.3-16.2
12 th	f	7.42	1	4	Idiopathic	Rib-to-spine (right) shifted to rib-to-pelvis (subsequently resected), rib-to-pelvis (left)	111.5—117.0	17.1-17.0
$13^{\rm th}$	f	5.75	0	_	Idiopathic	Rib-to-spine (resected)	101.0-102.0	13.7-13.7
$14^{\rm th}$	f	7.00	0	_	Jarcho-Levin syndrome	Rib-to-rib, rib-to-spine	116.0-117.5	17.6-17.2
$15^{\rm th}$	f	7.67	0	-	Idiopathic	Rib-to-spine	128.0-130.0	20.0-20.1
$16^{\rm th}$	m	1.58	0	_	Jarcho-Levin syndrome	Rib-to-spine	-	12.3-12.0
17 th	m	7.25	0	_	Neurofibromatosis	Rib-to-rib, rib-to-spine	110.0-111.5	16.4-16.1
18 th	f	6.00	0	_	Idiopathic	Rib-to-pelvis, rib-to-pelvis	111.0-115.5	18.0-17.9
19 th	f	8.67	0	_	Idiopathic	Rib-to-pelvis	124.5-125.5	21.1-21.2
20 th	m	6.67	0	_	Congenital	Rib-to-pelvis, rib-to-pelvis	96.0-100.0	13.1-14.4
21st	f	4.83	0	_	Congenital	Rib-to-spine	-	_
22 nd	f	3.00	1	12	Congenital	Rib-to-rib, rib-to-spine	84.5-92.0	_
23 rd	f	2.00	1	12	Congenital	Rib-to-spine	83.5-87.5	_

Table 2

Comorbidities and earlier interventions in patients operated on

Dotient	Compubilities and equipmentings
Patients	Comorbidities and earlier interventions
1 st	Congenital heart disease: ventricular septal defect with unchanged hemodynamics; abnormality of chordal apparatus of the left ventricle;
1	grade 0 chronic cardiac insufficiency
2 nd	Operated on for spine deformity: right-sided posterior fusion of T3—T12; interbody epiphysiodesis of T6—T10 with autologous costal bone
ζ	powder
3 rd	–
4 th	Multi-stage surgical correction of double-sided clubfoot
5 th	A plastics of inquinal hernia
6 th	Thoracic diastematomyelia, chronic adenoiditis, remission, grade II adenoids
7 th	Thoracic diasternatomyena, chrome adenoiditis, femission, grade 11 adenoids
8 th	Chromosomal pathology; radical correction of the Fallot's tetralogy; infundibulectomy of the right ventricular outflow tract; healing of
O.	the ventricular septal defect; lung commissurotomy; transannular patch repair of the outflow tract of the main lung artery; 0 grade chronic
	cardiac insufficiency; moderate mental retardation; bilateral inquinal testis; dwarfism; subcerebellar arachnoid cyst; arachnoid cyst of the
	left temporal pole; outer hydrocephaly in the left-sided frontal-temporal lobe; inner asymmetric non-obturational hydrocephalus
9 th	Kartagener syndrome of the autosomal recessive type; Arnold—Chiari malformation type I; functional shortening of the left lower limb by
9	3.5 cm; condition after transsection of the terminal thread of the spinal cord; hypertensive retinal angiopathy
10 th	3.3 cm, condition after transsection of the terminal thread of the spinal cord, hypertensive retinal angiopathy
10 th	Abnormality at the craniovertebral junction; decompression of the posterior cranial fossa; excision of cicatrices; resection of the C1 posterior
11	arch
12 th	Equinovarus deformity of left foot; intestinal dysbiosis
12 13 th	Mild hypermetropia; descending atrophy of the optic nerve, physical retardation; subclinical hypothyreosis; mental retardation of complex
10	genesis; cyst in the right cerebellar hemisphere
$14^{ m th}$	Balancing fusion at the convex side of the deformity at the T5–L2 level, extirpation of the posterolateral hemivertebra between T10–T11,
	implanting of the contractor, anterior fusion, local posterior fusion, contractor removal, posterior fusion
15 th	Surgical treatment of the hernia of the left cupula of the diaphragm, plastic repair with a nylon endoprotesis; surgery for adhesive intestinal
	obstruction; congenital cardiac anomaly, open aortic channel; ventricular septal defect
16 th	Several surgeries for congenital ankyloproctia (colostomy, proctoplastics, abdominal commissures)
17 th	Inherited monogenic pathology; type I neurofibromatosis, a family form; AD inheritance; diffuse osteoporosis
18 th	Functional shortening of the right lower limb by 4 cm
19 th	Congenital lung malformation; surgery for diaphragmatic hernia
20^{th}	Revision of the posterior sections of the thoracic and lumbar spine in attempt to correct the spine deformity, dural sac damaged at L3—
	L5 levels, liquorrhea stopped; a week after the surgery the patient fell down on buttocks opening the liquorrheal fistula; reoperation for
	stopping the liquorrhea, mounting of the lumbar drainage
21st	Surgery for the anterior meningocele of T3 and fixed spinal cord syndrome - osteoplastic laminectomy of T2-T5 was performed,
	meningocele resection, plastic repair of the hernial orifice at the T3 level
22 nd	Congenital malformation of the cervical, thoracic, and sacrococcygeal spine; thoracic hypoplasia; esophageal atresia with lower
	tracheoesophageal fistula; condition after the direct anastomosis of the esophagus; cloaca, the post-plastics condition, double colostomy;
	doubling of a single right kidney; neurogenic urocyst; secondary chronic pyelonephritis; ventricular septal defect; patent foramen ovale;
	hypotrophy of the right lower limb
23 rd	Incomplete doubling of the single left kidney, dislocation of the right hip joint; hypotrophy of the right lower limb

- condition of the soft tissues does not allow one to safely close the endocorrector;
- condition of the bone tissue does not allow one to anchor the metal construct (as in patients with osteogenesis imperfecta);
- lack of ribs required to mount the cranial claw;
- accompanying diseases prevent repeated anesthesia;
 - active respiratory infection;
 - diaphragm dysfunction.

Opening-wedge thoracostomy in patients with scoliosis and fused ribs. The patient lies in the lateral position (corresponding to the convex side of the deformity). The induced somatosensory

evoked potentials of the spinal cord from the upper and lower limbs are monitored. Shoulders are retracted by no more than 90°. Skin incision is started 4 cm caudally from the upper medial corner of the scapula, continued in parallel to its inner edge, and subsequently continued ventrally as an L-shaped incision (Fig. 2). Next, mm. trapezius, romboideus, latissi-

mus dorsi are dissected along the skin incision with an electrocautery blade. Soft tissues between the scapula and the rib cage are separated with blunt instruments and the scapula is pulled up. The attachment site of mm. scalenus med. et dors. together with the neurovascular bundle localized more ventrally is identified. Paraspinal muscles are shifted medially to the apices of spinous processes so as not to damage the costal periosteum

a b c

Fig. 1 Types of arrangement of the VEPTR endocorrector (data provided by Synthes company): \mathbf{a} – rib-to-rib; \mathbf{b} – rib-to-spine; \mathbf{c} – rib-to-pelvis

and not to expose the dorsal elements of the vertebrae, which may cause the formation of a bone fusion.

The place for anchoring the cranial costal claw is subsequently prepared (Fig. 3). It should localize within the cranial part of the scoliotic curve. More cranial arrangement may result in the development of compensatory counter-bending without correction of the main curve. The rib intended for mounting the claw should be at least 1 cm thick. Otherwise, the claw is mounted at the two adjacent ribs. The first rib is never used due to the possible shifting of the endocorrector, which can injure the brachiplex. The claw is arranged as close as possible to the transverse processes; both halves of the claw are implanted and blocked against each other.

The next stage is osteotomy of the costal block (Fig. 4). The level should be determined clinically and confirmed by X-ray. Fibrous tissues are located ventrally or dorsally and follow the osteotomy line. They are dissected with an electrocautery blade so that the pleura remained undamaged. An expander is placed between the ribs; the intercostal space is carefully widened. A narrow elevator, which is used to detach the periosteum or pleura from the bone fusion along the line of supposed osteotomy, is inserted into this space. Osteotomy is performed with Kerrison forceps or other tools (high-speed surgical drill, osteotome device) in ventral direction. The intercostal interval is gradually widened, the lower-lying pleura is shifted with a wet swab in cranial and caudal directions. The dorsal costal mass is dissected subperiostally with forceps under visual control, leaving 5 mm to the vertebral body and leaving transverse processes intact. The last section of the costal fusion is separated with a curved curette in the direction away from the spine so as not to injure the contents of the spinal canal. Anomalous vessels are left intact to preserve the circulation in the spinal cord. Bone bleeding is stopped using wax. After finishing the

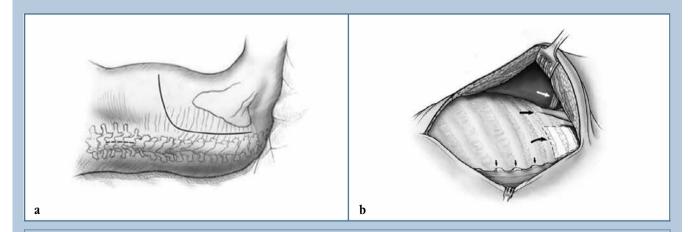


Fig. 2
Surgical technique of open wedging thoracostomy in patients with scoliosis and fused ribs (data provided by Synthes company):

a – patient's position and lines of skin incisions (solid line – rib-to-rib distractor, dashed line – mounting of the laminar hook); b – scapula is uplifted, the area of dissection of mm. scalenus is shown, the neurovascular bundle localizes ventrally (white arrow)

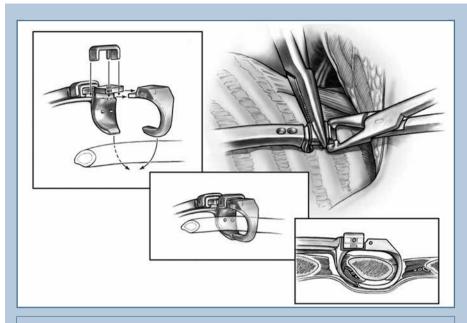


Fig. 3
Assembly, installation and blocking of the costal claw (data provided by Synthes company)

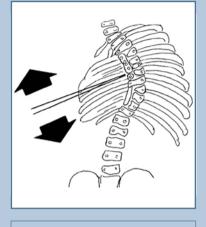


Fig. 4
Dissection of the osteochondral block from the apices of transverse processes: the resulting defect is gradually extended (arrows) with a bone spreader (data provided by Synthes company)

osteotomy, the intercostal interval can be widened to make the heights of the left and right hemithoraces equal (Fig. 5). When the cranial rib forming the intercostal interval is arranged horizontally, the osteotomy stage is considered to be finished. A long costal retractor is inserted in the resulting defect to maintain the correction achieved. In this position, the necessary distance (in cm) between the upper and lower claws is determined; the retractor is subsequently removed to let tissues relax. The place for implantation of the lower costal claw is prepared and the retractor is subsequently returned to its place. Elements of the lower claw are implanted in the same manner as that for the upper claw; both claws are subsequently connected with the distractor body and fixed with special brackets in the position of the achieved correction of the hemithorax. Small pleural ruptures (1.5–2.0 cm in size) are not a problem. If the lesions are more significant, the defect should be covered with absorbable Surgisis sheet that is sutured to the pleura edges.

Hybrid endocorrector in patients with thoracolumbar scoliosis. First, the upper costal claw is implanted; next, a

place for lower anchorage is prepared using another incision. A laminar hook is typically placed at the arch of the lower neutral vertebra (L1, L2 or L3). A 4 cm long skin incision length is made 1 cm laterally from the spinous process of this vertebra. Muscles are detached using an electrocautery blade, the yellow ligament is resected, and a supralaminar hook is mounted to the corresponding hemilamina. The distractor of the proper length is chosen. The extender of the distractor is fixed to the cranial claw and blocked with a special bracket. Keeping the costal retractor in the position of desired widening of the intercostal interval, the plain part of the distractor is inserted in the gap of the extender. The pivot part of the distractor is cut with forceps 2 cm distally from the laminar hook and is slightly bent in accordance with the lumbar lordosis. The distractor is then relocated from the upper wound to the lower one through the paraspinal muscles. Its pivot part goes through the hook opening for the desired length; the plain part is subsequently connected with the extender of the distractor. Still keeping the costal retractor in its position, a correcting force is applied to the

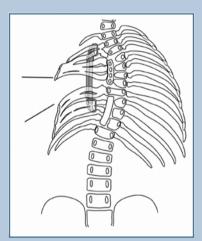


Fig. 5
The upper rib, included in the defect formation, acquires a horizontal position (i.e., increasing the hemithoracic height) can considered reasonable; without removing the spreader, the rib-to-rib endocorrector is implanted to stabilize the resulting hemithorax correction (data provided by Synthes company)

distractor to achieve tension of the soft tissue. For this purpose a special distracting tool and a temporal anchorage at the

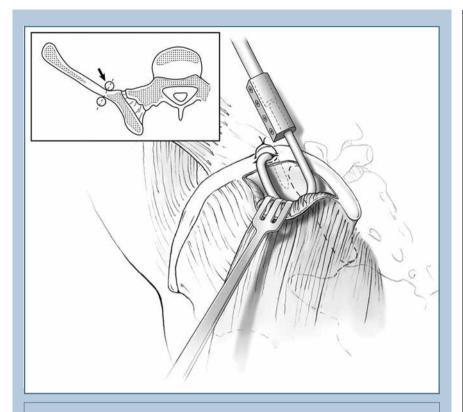


Fig. 6
Implanting the Dunn–McCarthy pelvic hook in the dorsal third of the iliac bone (data provided by Synthes company)

pivot part of the distractor are used. The rib-to-rib endocorrector is then implanted and the cranial claw is formed at the same ribs as in the case of the hybrid distractor. Caudal claw is mounted on a stable rib (not lower than the 10th rib). After the rib-to-rib endocorrector was blocked, the retractor is removed and additional distraction is performed for the hybrid endocorrector. The screw nut at the laminar hook is tightened. Autologous bone graft harvested by edge resection of ribs that are not used for endocorrector anchorage is placed around the hook. The purpose is to form the onelevel fusion and to reinforce the anchorage point of the hook.

In patients with severe thoracolumbar deformities or in cases when posterior vertebral elements are absent due to the myelomeningocele, a Dunn–McCarthy hook is placed to the iliac crest between its medium and dorsal thirds (Fig. 6). Apophysis of the iliac bone is incised,

and the hook is inserted in the resulting opening, maintaining the integrity of apophysis.

Wound suturing and postoperative follow-up. Muscles are carefully sutured layer-wise. The lower edge of the scapula is fixed to the chest wall with absorbable threads to reduce the inside pressure to the suture line. The wound is drained. After skin suturing, a control X-ray survey in two projections is performed. Some patients may need intubation and lung ventilation for up to three days. Wound drainage is removed when the flow is less than 20-25 ml/day, thoracic drainage is removed when the flow is less than 1 ml/kg per day. The patients should stay in bed for at least 3-5 days. No external immobilization is required. The full motion activity is allowed 6 weeks after hospital discharge, but parents need to be vigilant to prevent possible injuries that may result in breakage of the anchoring ribs and shifting of the endocorrector. Another important issue is preventing pressure sore and damage to the integrity of soft tissues above the endocorrector. These complications often occur due to the low muscle volume and thin skin in children.

Staged distractions and endocorrector replacement. Repeated staged distractions are performed every 6 months. Patient's position on the operating table is the same as at the main stage. The localization of the blocking bracket is exposed through a 3 cm incision. The bracket is removed; the distractor is extended using a special tool by 5–10 mm and placed at the new position.

In a typical case (congenital scoliosis with costal fusion), the purpose of intervention is to balance the growth of the left and right sides of the rib cage and to indirectly correct the scoliotic deformity of the spine. All the components of three-dimensional deformity of the rib cage are fixed without fusion, which suppresses the spine growth [3].

In children under 18 months, it is usually sufficient to use a single rib-to-rib distractor. In older patients, if the width of the spinal canal is sufficient and the laminar hook can be implanted, the hybrid endocorrector is used (rib-to-rib + rib-to-spine) to ensure better correction of the deformed hemithorax and to stabilize any scoliotic curve spreading to the thoracolumbar junction. Thus, the preoperational CT is mandatory. This surgical strategy is based on an assumed correlation between the lung and rib cage growth, so correction of the rib cage growth along with correction of the spine deformity will provide the largest possible lung volume by the age of patient's skeleton becomes mature.

Results

The mean primary scoliotic curve in all patients was 66.3° ($34-104^{\circ}$), and after the first surgery it decreased to 47.9° ($15-82^{\circ}$). The correction was 18.4° (27.8°). Before the first staged distraction, the mean deformity in 12 patients was 65.4° ($32-98^{\circ}$), while after the distraction it was 54.0° ($17-77^{\circ}$). It was infeasible to calculate the mean values

 $Table \ 3 \\ The \ dynamics \ of the \ main \ scoliotic \ curve \ in \ patients \ operated \ on \ at \ the \ different \ treatment \ stages, \ deg.$

Patients	Main	Staged	Staged	Staged	
	surgery	correction I	correction II	correction III	
1 st	52-34	41-38	49-43	-	
2 nd	89-80	89-71	88-73	75-63	
$3^{\rm rd}$	66-66	78-77	-	-	
4^{th}	74-59	68-69	-	-	
$5^{ m th}$	63-55	64-60	-	-	
6^{th}	51-36	49-42	-	-	
7^{th}	53-34	57-51	-	-	
8 th	71-40	60-40	-	-	
9^{th}	106-66	83-71	-	-	
$10^{\rm th}$	40-25	54-35	-	_	
$11^{\rm th}$	123-77	98-81	_	-	
12^{th}	99-60	72-74	_	-	
13 th	67-30	-	_	-	
14^{th}	49-41	-	_	-	
15^{th}	54-50	-	_	-	
16 th	69-58	-	-	-	
$17^{\rm th}$	79-63	-	_	-	
18 th	65-44	-	_	-	
19 th	55-26	-	-	-	
$20^{\rm th}$	45-32	-	-	-	
21st	87-82	-	_	_	
22 nd	35-28	-	_	-	
$23^{\rm rd}$	34-15	_	_	_	

in patients with more than one staged distractions because it was performed only in two patients. The dynamics of the mean curve in all patients are shown in Table 3.

The two most abundant subgroups of patients (the ones with idiopathic or congenital scoliosis) were analyzed separately. In 7 children with idiopathic scoliosis, the initial distortion was 76.3° ($54-123^{\circ}$); after the first surgery it was 46.7° ($26-77^{\circ}$); correction was 29.6° (38.8 %). The Cobb angle before the first staged distraction in 4 patients was 82.8° ($60-98^{\circ}$), after -65.0° ($40-81^{\circ}$).

In 14 patients with congenital deformities the following results were obtained: initial Cobb angle 61.5° (34–106°), after the first surgery – 46.1° (15–80°), correction was 15.0° (24.4 %). Before the first staged distraction in 9 patients the value of the scoliotic curve was 56.7° (32–89°); after the distraction it was 49.0° (17–77°).

Thoracic kyphosis in the total group changed in the following manner: originally – 37.9°, after the first surgery – 30.6°, before the first staged distraction – 45.1°, after the first distraction – 38.6°. For lumbar lordosis, these values were 53.6°; 39.5°; 53.0°; 51.7°, respectively. Postoperative external immobilization was not used. Children started standing up three or four days after the first intervention and the next day after staged distraction.

The mean patient's height in the standing position before the treatment was 101.9 (range: 83–128) cm, after the first staged distraction it was 108.7 (range: 87–130) cm. Patient's weight was 15.9 (range: 12–21) kg and 16.7 (range: 12–21) kg, respectively.

Complications were observed in 2 patients: in one girl, the endocorrector had to be removed because of early suppuration; in another one, a shift of the laminar hook was detected; the pelvic

hook was remounted and the second rib-to-pelvis distractor was implanted. Ten days later, a shift of the first (right) pelvic hook with a hematic abscess was detected. The right rod was removed.

Discussion

The first known article focused on VEPTR was published in 2004 [3]. A total of 21 patients with congenital scoliosis were described. The mean age at the first examination was 3.3 years; the mean duration of postoperative follow-up was 4.2 years. Significant growth of the thoracic spine was found in patients who were not treated: 7.7 mm at the concave side and 8.3 mm at the convex side. In 11 patients, the unsegmented bar increased by 7.3 %. In 3 patients who were operated on earlier (fusion), the mean growth of the thoracic spine was 4.6 mm at the concave side and 3.7 mm at the convex side.

In 2005, Emans et al. [6] reported the surgery outcomes in 31 children: 26 patients with congenital scoliosis with costal block (two of those had the Jarcho - Levin syndrome), 4 patients with iatrogenic costal block and thoracogenic scoliosis, 1 patient with a congenital defect of the chest wall. The mean age at treatment start was 4.2 years, the duration of follow-up was 2.6 years, the mean number of staged distractions was 3.5 per patient. The Cobb angle was decreased from 59 to 43°. The original lung volume (according to the CT data) was 369 cm³, after the first surgery it was 394 cm³, and at the end of the follow-up period it was 736 cm³. No endocorrector fractures were observed, but the endocorrectors were displaced in 8 cases. Shifting was fixed in a typical procedure during the staged distraction. No neurologic complications involving the CNS were detected. Two cases of brachial plexitis were found at the operated side, where the functions of the plexus were restored. In two cases, the ribs detached in the first intervention were fused and repeated osteotomy was needed. Two cases of deep infection and two cases of rib fractures were observed. The authors emphasized the importance

of using the VEPTR in the early period, before the deformity becomes severe.

Campbell et al. [4], while analyzing the complications of using the VEPTR, mentioned that the need for multiple repeated interventions inevitably increases the risk of possible complications. Each new incision is a risk of an infection or development of a pressure sore. Multiple comorbidities are another risk factor for complications. The author of this method has provided his 15-year experience of treating children with different juvenile scolioses accompanied with rib cage deformities. A total of 201 children were operated on at the Cristus Santa Rosa Children's Hospital (Texas. USA) in 1989-2004. On average, each child underwent seven surgeries with the mean follow-up period of 6 years. Infectious complications were detected in 3.3 % children; the complications were associated with soft tissues in 8.5 %. Mechanical complications were also frequent. Thus, fracture of an endocorrector was found in 6.0% of patients, shifting – in 27.0 %. The authors noted that shifting of either the upper or the lower end of the endocorrector was in most cases asymptomatic and was detected occasionally before the next stage of surgical correction. Displacement was slow and the damaged ribs were totally rearranged so they often could be used as capture anchorage points. The author of the method used it both in patients with congenital deformities and in patients with infantile scoliosis of etiologies other than congenital [4]. Development of the costal hump without a costal fusion causes a severe deformity when ribs are twisted around the spine like a closed umbrella. In 10 such cases, VEPTR was used after the multi-level dissection of intercostal muscles to increase chest wall mobility. Either a semi-elliptical endocorrector or the rib-to-rib distractor was used. The author called this operation "an open umbrella" due to the horizontalization of the mobilized ribs. The mean follow-up period was 6 years. The mean Cobb angle decreased from 79 to 51°. The mean vital capacity was 38 % of that expected for this age. The following complications were observed: 9 - mechanical, 3 – infection, 3 – pressure sores, and 2 pneumonia cases. Campbell et al. suppose that the surgery increases the hemithoracic volume. Complications are frequent but cane be efficiently treated.

Emans et al. [7] have analyzed the safety of the Dunn-McCarthy iliac hook in 33 patients with deformities of different etiology. In 17 patients, the hook was implanted on one side, in 16 patients it was implanted bilaterally. The mean age of patients at the start of treatment was 6.1 years. In 10 patients with one-side installation of the hook, a revision was needed due to its migration, no migration was detected when the hook was implanted bilaterally. With the mean follow-up period of 1.2 year, 1.6 revisions per patient were needed. The migration of the hook is usually asymptomatic; it maintains reasonable stability and needs to be reinstalled only when localized close to acetabulum.

Skaggs et al. [12] examined 79 patients in 7 different institutions as they searched for changes in general nutrition after surgeries using the VEPTR. The purpose of the intervention was to treat or prevent the development of the thoracic insufficiency syndrome. Before surgery body weight was lesser than normal in 62 patients; 22 patients showed a significant increase in this value after the surgical treatment. Among 17 patients with normal weight, 13 exhibited a significant increase in body weight.

Song et al. [13] used a modified technique to treat 14 children with neuromuscular scoliosis. They used one distractor with both ends attached to the spine as "growing rods". The mean age of patients at the start of treatment was 77 months, the mean follow-up period was 15 months. Over this small period of time, at least one staged correction was performed. The mean duration of rod implanting and staged distraction was 2.5 h and 30 min, respectively. Noteworthy, 8 patients from this group were previously operated on using other correcting instrumentation. The mean Cobb angle was reduced from 69 to 47°. Complications were as follows: one fracture of the rod, two cases of superficial and one case of deep infection. The authors presumed that the use of VEPTR has demonstrated early promising results in patients with severe spine deformities without accompanying defects of the chest wall.

The analysis of these sparse data allows one to draw several conclusions. The novelty of the method requires data accumulation to reveal positive as well as negative features of the instrumentation. The follow-up periods were short in all the studies. There is a lack of data on completed treatment when the final stage (dorsal fusion) was performed at the age of skeleton maturation. However, it appears that an increase in the volume of abnormal hemithorax is an attainable purpose. The spine deformity can be controlled, despite the low decrease in the Cobb angle, which can be attributed to the rigidity of congenital spine deformities. The number of complications is quite high, and most of them appear to be specific for the method described. Meanwhile, many complications can be treated during the planned treatment stage, without increasing the number of interventions required.

Small children with severe progressive deformities of the spine are not a rarity. We follow over 200 patients with this deformity, and the extremely high cost of the implant toolkits is the only limiting factor of our surgical activity. The problem should be considered as extremely urgent due to the high risks of disability caused by this pathology.

An attempt to solve the ambiguous problem (prevention of the thoracic insufficiency syndrome and progression of the spine deformity) using the VEPTR instrumentation is theoretically reasonable but has several practical limitations. It is rather difficult to perform multiple staged distractions as a patient grows up. Nevertheless, no alternative method is currently known, while children cannot wait.

Conclusions

1. The modern potential of anesthesiologic protection allows one to perform rather invasive interventions in patients with severe and diverse comorbidities.

- 2. Surgical treatment of infantile and juvenile scoliosis should be started at the age when prognosis of fast progression becomes undoubtable. A long follow-up and X-ray evidence on deformity progression will lead to secondary structural and functional losses, which are not easily corrected.
- 3. The dynamics of the Cobb angle suggests that scoliotic deformity progression can be prevented in most cases and a certain correction can also be achieved. Notably, scoliosis in this group of patients possesses a high progression potential, which is evident from rapid

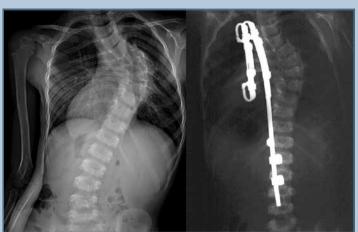
increase in the Cobb angle between the stages of surgical treatment. Besides, the sagittal contour of the thoracic and lumbar spine preserves its normal parameters.

4. The VEPTR instrumentation was originally designed to treat patients with congenital vertebral and costal abnormalities (Fig. 7). It can also be used for patients with scoliosis of different etiology, but requires individual arrangement of the endocorrector. In our opinion, the use of the rib-to-pelvis bilateral distractor is reasonable in patients with idiopathic scoliosis, especially the ones affecting the lumbar or thoracolumbar

spine (Fig. 8). Complications resulting from this arrangement are caused only by technical errors.

We do not report the results of numerous methods that can provide important data on patients' condition during the treatment stages. These data will probably be described in the near future.

We intend to continue our multicenter study by elaborating the unified protocol in compliance with the requirements of the evidence-based medicine. Colleagues are welcome to cooperate in this research.



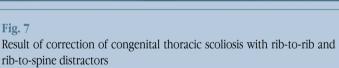




Fig. 8
Result of correction of idiopathic thoracolumbar scoliosis with rib-to-pelvis and rib-to-pelvis distractors

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