SURGICAL TREATMENT OF A PATIENT WITH CONGENITAL DEFORMITY OF THE SPINE, THE THORACIC AND LUMBAR PEDICLE APLASIA, AND SPINAL COMPRESSION SYNDROME

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The paper presents a case study of surgical treatment of a 14-year-old patient with multiple skeletal malformations, congenital kyphoscoliosis of the thoracic spine, non-fusion of vertebral arches and vertebral bodies from the C5 to the sacrum (bodies are separated from arches with slit-like defects along the whole length), and spinal compression syndrome.
Key Words: pedicle aplasia, congenital kyphoscoliosis, spinal stenosis, stereolytographic full-scale model, surgical treatment.

Please cite this paper as: *Kuleshov AA, Vetrile MS, Lisyansky IN, Makarov SN, Sokolova TV. Surgical treatment of a patient with congenital deformity of the spine, the thoracic and lumbar pedicle aplasia, and spinal compression syndrome. Hir. Pozvonoc. 2016;13(3):41–48. In Russian.*
DOI: http://dx.doi.org/10.14531/ss2016.3.41-48.

Surgical treatment of severe congenital spinal deformities remains an urgent problem of modern orthopedics. According to the State Statistics Committee, there were a total of 28,202,500 children aged 0 to 17 in the Russian Federation in 2006. The average incidence of scoliosis in pediatric population (9 %) allows calculating that 2.53 million children and adolescents suffer from scoliotic deformity of the spine. Assuming that the proportion of congenital scoliosis in pediatric scoliosis is 2.5 %, the absolute number of patients suffering from this pathology in Russia can be estimated as 63,250 people. [6] There are well developed methods of surgical treatment of vertebrae formation disorders and pure congenital kyphoses [5, 7, 8, 18]. A single hemivertebra can be extirpated with simultaneous correction of deformity and fixation of the spine over a limited section (2–3 segments). Modern instrumentation allows for fixation either by fixing an implant at vertebral arcs or by introduction of pedicle screws to support the structure. The optimal strategy in case of kyphosis is wedge vertebrectomy with multisupport endocorrectors. Currently, the method of staged correcting distraction of the spine and thoracic volume correction using VEPTR instrumentation is employed [2, 9, 10].

We present a clinical observation of surgical treatment of a patient with multiple skeletal malformations, congenital kyphoscoliosis of the thoracic spine, non-fusion of vertebral arches and vertebral bodies from the C5 to the sacrum (bodies are separated from arches with slit-like defects along the whole length), and spinal compression syndrome.

Patient N., 14 years of age, was admitted to the department with complaints of spine deformity, weakness in the lower limbs, and dysfunction of pelvic organs. The deformation of the spine was identified at birth. The patient received conservative treatment (physical therapy, massage) and was monitored by a neurologist at the place of residence. In 2006, at the age of 9 he was examined in the CITO and diagnosed with severe spinal deformity of congenital nature. Clinically, at that time, dysfunction of pelvic organs of peripheral type manifested as true incontinence and involuntary defecation. There was no paresis and sensory disturbances. The child was monitored and received conservative treatment at the place of residence. The parents have noted gradual progression of the deformity. In 2011, he developed spastic contractions in the muscles of the lower limbs, required support for walking, and weakness and spasticity in the legs gradually increased; in 2012, one week before the admission to the CITO he lost the ability to walk.

Neuroorthopedic status on admission: clinical symptoms of kyphoscoliosis of the thoracic and lumbar spine and chest deformity. Paravertebral muscles are relaxed, painless. The spinal deformity is not corrected by traction of the head. Neri, Dejerine and Lasegue symptoms are negative. Motor functions: upper limbs, no pareses, tendon reflexes were brisk and symmetrical; lower limbs, lower spastic paraparesis, iliopsoas muscles 3 points on the right, 4 points on the left, quadriceps muscle 3 points on the right, 5 points on the left; extensors of feet and fingers 2 points on the right, 4 points on the left; flexors of the lower leg 3 points on the right and left, medium and small

gluteal muscles 2 points, large, 1 point. Knee-jerk reaction of medium strength, Achilles reflexes cannot be engaged due to spasticity and forced position of the feet in flexion. Abnormal Babinski reflex on both sides; Preserved top and middle left abdominal reflexes. Equinovarus position of the feet. If the legs are bent at the hip and knee joints, the feet move to physiological position of 90°; conductor hypoesthesia (hypoalgesia) from T7–T8 level. Impaired function of pelvic organs of incontinence type. According to his mother, the child tries to move around a room over short distances, in upright position, using both arms for support with the weight on the forefeet.

Conclusion: thoracic myelopathy, lower spastic profound paraparesis with dysfunction of the pelvic organs (Frankel C; ASIA: motor, 81 points, needle prick, 84 points, touch, 84 points).

The following special methods of investigation were used: X-ray, CT, CT myelography, MRI, angiography.

CT of the spine: severe malformation of the cervical, thoracic and lumbar spine with rough kyphoscoliosis at the thoracic spine level; aplasia of C2 vertebra arch with C2–C3 bone block. From level of C5 on the left and C6 on the right, non-fusion of vertebral bodies and arches to the sacrum (bodies are separated from arches with slit-like defects along the whole length, Fig. 1); there are also multiple butterfly-like vertebrae at different levels; T5 vertebral body is hypoplastic, wedge deformed, and synostotic with T4 and Th6 bodies; kyphotic deformation of the thoracic spine with apex at T5 (kyphosis angle of 90°); visible spinal stenosis at the level of T5–T6 vertebrae and to a lesser extent at the level of T12; the lumbar vertebral arches are close together, bodies of the lumbar vertebrae are dislocated towards pelvis with underlying hyperlordosis; hypoplasia of sacral vertebral bodies and aplasia of the lateral masses of the sacrum (pelvic cavity dramatically reduced in volume); there are also the thoracic malformation, aplasia of dorsal section of ribs IV–VI on the left, ribs V and VI on the right and complete aplasia of rib I on the left and ribs I and II on the right (Fig. 1).

CT myelography: contrast agent can be traced at all examined levels, dural sac is deformed at the level of the narrowing of the spinal canal (T5–T6 and T12 vertebrae), the spinal cord is thinned at these levels; maximum narrowing of the spinal canal at the T5–T6 level up to 80 % (Fig. 2).

MRI: a series of tomograms reveals changes in the axis of the thoracic and lumbar spine; at the studied levels the endplates of the vertebral bodies and the articular surfaces of the intervertebral joints have irregular contours, are moderately compressed and notably deformed; at the middle thoracic and upper lumbar spine vertebral bodies and posterior elements are malformed, the spinal canal is narrowed with pronounced deformation; sacrum is partially present; the intervertebral discs and vertebral body at the studied level have reduced height and signal intensity; the spinal cord has pronounced deformation due to compression by the deformed vertebral bodies at the middle thoracic and upper lumbar levels, which also cause the narrowing of the spinal canal, with no signs of CSF.

Abdominal aortography, spinal arteriography (Fig. 3), excretory urography: the thoracic and abdominal sections of the aorta are identified on a series of angiograms, passable throughout; no deformation of the aorta, visceral abdominal aorta branches are arranged in a typical manner and are unchanged; great anastomotic artery is visible orig-

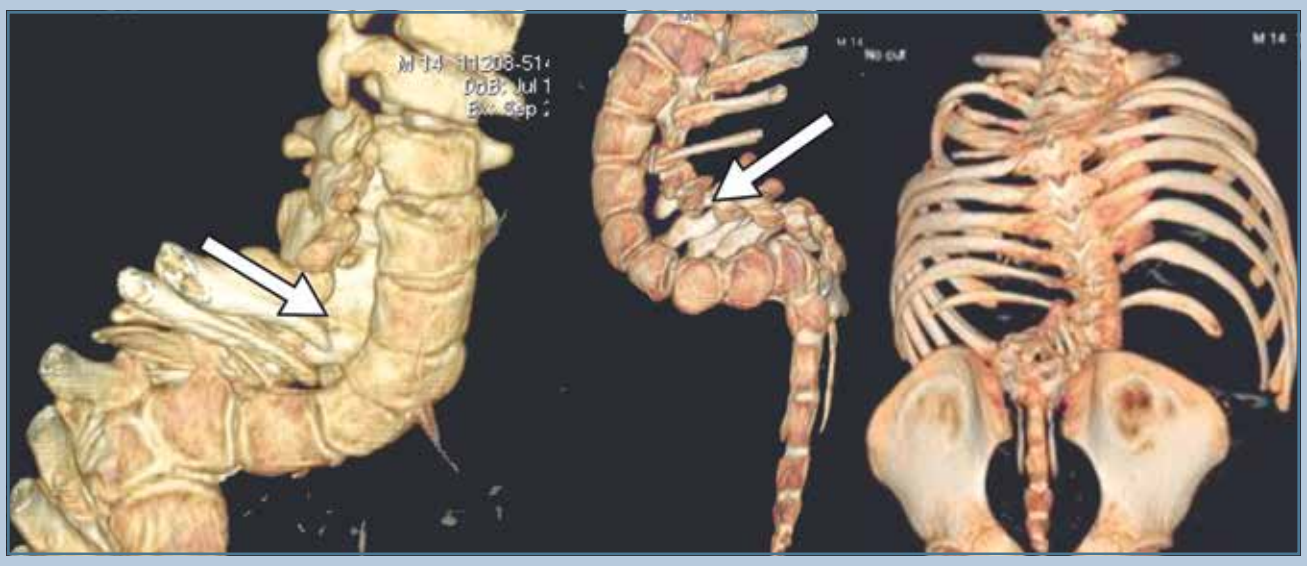

Fig. 1
CT of the Patient N., 14 years old, prior to the surgery: the arrows indicate the absence of arc roots in the thoracic and lumbar spine

inating from the left intercostal T11 artery; great anastomotic artery runs into the anterior spinal artery at T9 level; ascending and descending sections of the anterior spinal artery are not visible; the spinal cord cone is not contrasted; renal pelvis system is unchanged; ureters are passable throughout.

Spirography: disruption of ventilator function of the lungs of restrictive type.

Given the complexity of the malformation, the severity of the spinal deformity, the presence of gross neurological symptoms, a CT-based full-scale stereolithographic model of the patient's spine has been produced to better assess the situation and choose the optimal approach to surgery (Fig. 4). A particular feature of this model is its exact match to the anatomy of the patient's deformed spine. Analysis of the clinical presentation, the data of the specialized examination, and the direct visualization of the spine using the stereolithographic model led to the following conclusions: 1) a surgical intervention is indicated to eliminate the neurological disorders; 2) to achieve this goal it is necessary to perform spinal decompression with spinal stabilization (ideally also with the correction of the spinal deformity).

The examination of the stereolithographic model of the spine and its comparison to the commonly used implants revealed futility of using dorsal fixation systems, namely the complete ineffectiveness of hook systems and highly questionable feasibility of using pedicle screws. After weighing all pros and cons, we decided to perform surgery within the following scope: anterior decompression and anterior fixation of the spine by resection of vertebral bodies at the apex of the deformity with decompression of the spinal cord, anterior spinal fusion using a graft from the patient's own rib and ventral fixation with metal construction (Fig. 5).

Course of the surgery. Right transthoracic access through the bed of rib IV. Marked kyphosis with the apex at the level of T4–T5–T6 vertebrae. The vertebral bodies are deformed. Resection of T4–T5–T6 vertebrae bodies, decompression of the dural sac along the entire length of the kyphosis. Screws are introduced into the frontal plane of T3 and T6 vertebral bodies, a rod is placed on the screws and fixed with nuts. The rib autograft is laid on the lateral surface of the vertebral bodies is tied to the metal construction using Dacron. The postoperative course was uneventful; the patient was kept in the intensive care unit for 24 hours for observation, and then transferred to the department. CT control: full decompression of the spinal canal (Fig. 6).

Examination by a neurologist on Day 1 after the surgery: lower spastic paraparesis is present with unchanged severity, conduction hypoesthesia (hypoalgesia) from T7–T8 level. Conclusion: no negative dynamics in the early postoperative period. Neurological status 20 days after the date of the surgery: clear positive dynamics in neurologic status in a form of increase in the lower limb muscle strength to 5 points, but pronounced spastic hypertonicity with varus positioning of the feet. The patient can overcome spasticity on one's own.

Pronounced postural and protective pathological reflexes with redistribution of spastic hypertonia. Conduction hypoesthesia (hypoalgesia) from T7–T8 level on the left, with hypertension on the

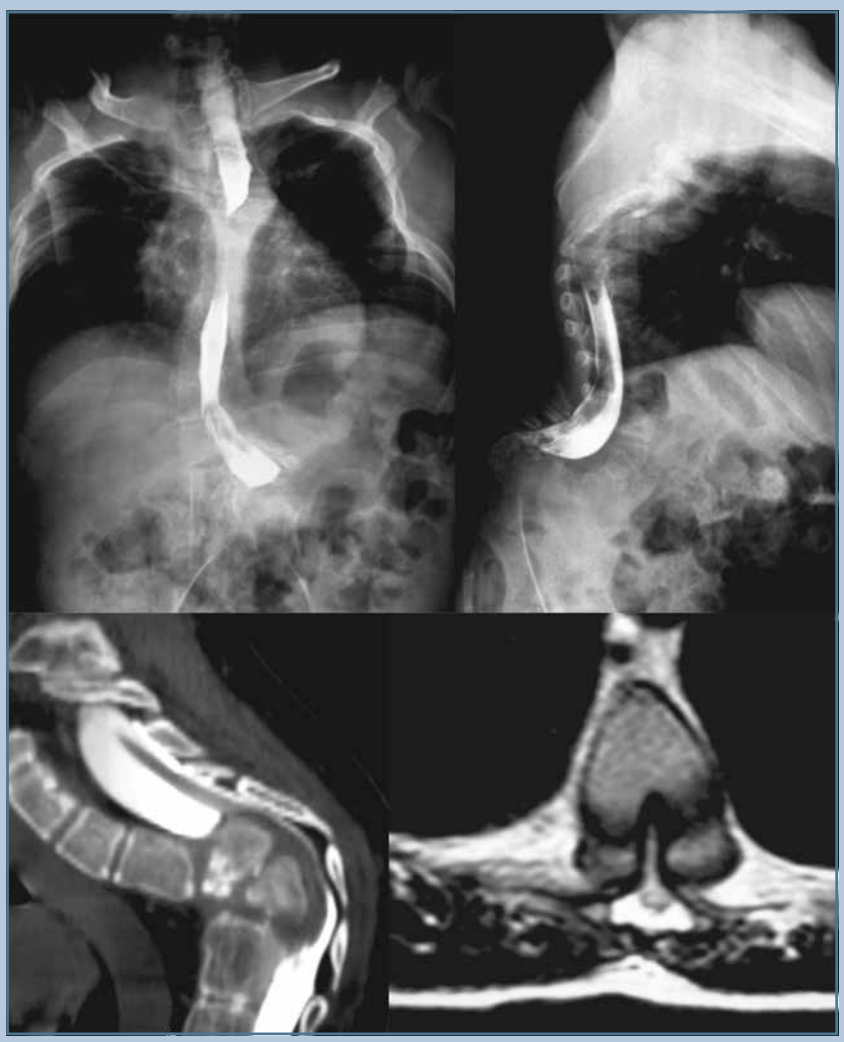

Fig. 2
Myelography and CT myelography of the Patient N., 14 years old, prior to the surgery

right (Frankel D; ASIA: motor, 90 points, needle prick, 84 points, touch, 84 points). Leningrad-type corset was made for the patient. Upright in the corset.

The patient was followed up on an outpatient basis and arrived to control examination 6 months after the surgery. Positive dynamics was observed. The patient moves independently, without additional support. Recovery of muscle strength in the limbs to 5 points (Frankel E; ASIA: motor, 100 points, needle prick, 112 points, touch, 112 points; see Table). The metal construction was stable based on clinical and X-ray data; CT reveals signs of bone block formation. Normal function of the pelvic organs.

Three years from the date of the surgery, clinical and radiological examination reveal no deterioration.

Discussion

There is rather limited number of reports on arch root anomalies in the literature; as a rule, they are represented by descriptions of individual clinical cases. The first arch root aplasia was described by Hadley in 1946 [12] in the cervical spine. In 1986, Lederman and Kaufman [14] gave a description of aplasia and hypoplasia of the roots of the thoracic vertebrae arches. In 1997, Villas and Barrios [19] reported a case of a 23-year-old man with congenital kyphoscoliosis, who had no posterior elements (arches and arches roots) at L2 vertebra. The patient suffered from pain in the spine and the progression of the deformity. Two-stage surgical intervention was performed (dorsal correction and fixation, anterior L1–L4 interbody fusion).

In 2000, Rauzzino et al. [16] reported a case of a 14-year-old boy with bilateral aplasia of the arch roots from T4 to T8 vertebrae, who suffered from pain and lower spastic paraparesis. The examination revealed spinal stenosis at the apex of the kyphosis. The spinal deformity was stabilized using the posterior multisupport tools in combination with the posterior spondylosyndesis. There were no improvements in the neurological status in the postoperative period. In 2008, Geoffray et al. [11] examined a 9-month-old child with congenital kyphoscoliosis, who had no arc roots on both sides between T2 and T9 vertebrae. There were no neurological symptoms. Surgical treatment of the patient was not performed.

In 2010, Ishida et al. [13] reported a case of a 59-year-old patient with congenital type 2 kyphosis, who developed

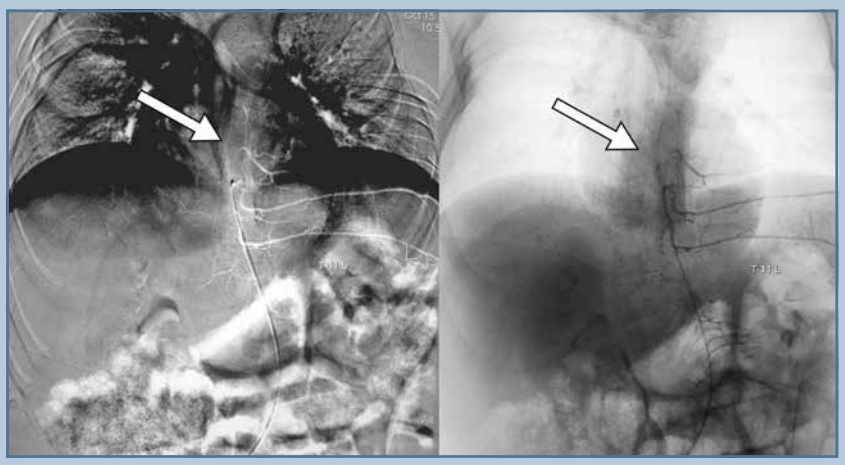

Fig. 3
Spinal arteriography of the Patient N., 14 years old: the arrows indicated the great anastomotic artery

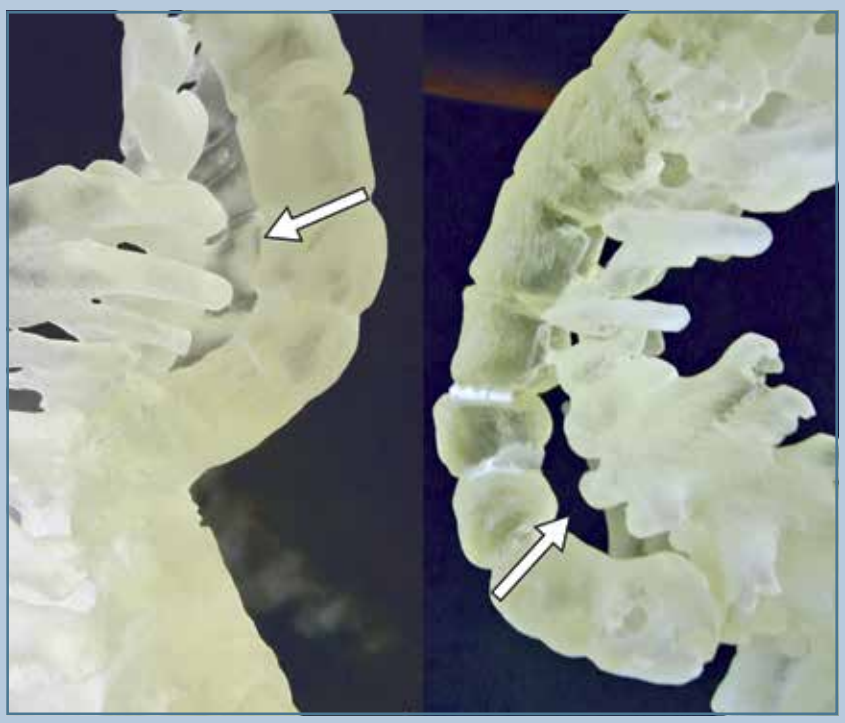

Fig. 4
Stereolithographic model of the Patient N.'s spine, 14 years old: the arrow indicate the absence of arches roots in the thoracic and lumbar spine

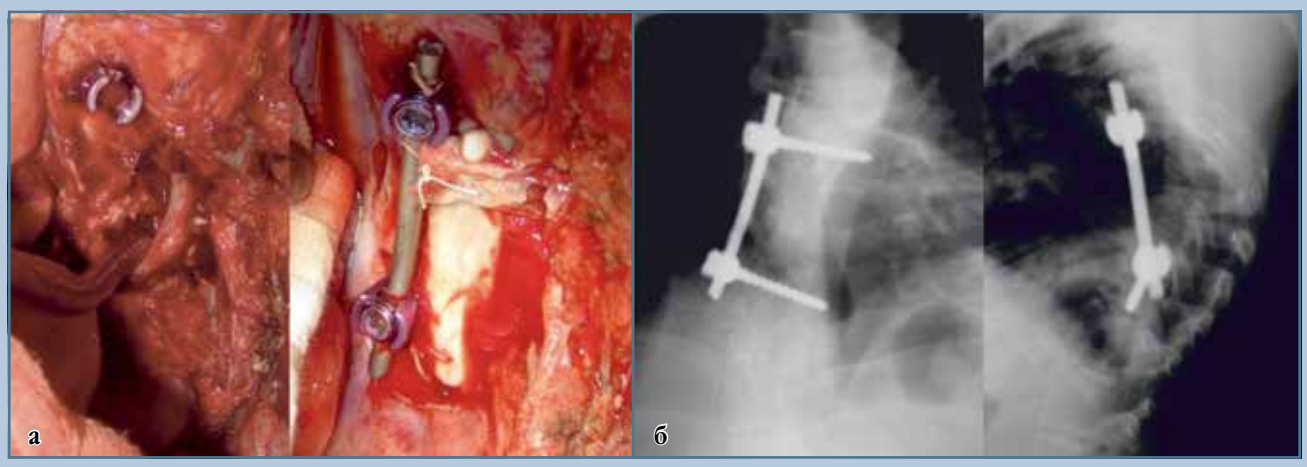

Fig. 5
Intraoperative snapshot with spinal cord decompression and installed metal construction with rib autograf **(a)** and radiographs **(b)** of the Patient N., 14 years old, after surgery

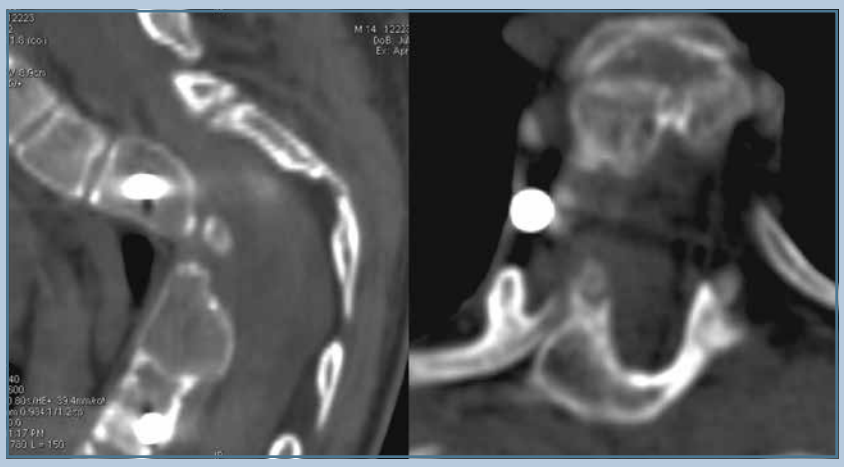

Fig. 6
CT of the Patient N., 14 years old, after surgery: visible spinal decompression

spinal stenosis at the level of T10–T11 with myelopathy, lower paraplegia and pelvic organs dysfunction. The patient underwent a surgical intervention: costotransversectomy, laminectomy at the T10–T11 level with the spinal cord decompression and dorsal fixation of the spine with a metal structure along T7–L2. Neurological symptoms regressed after the surgical intervention. In 2013, Patel et al. [15] considered congenital hypoplasia of the arc roots of the lumbar vertebrae as a rare cause of spondylolisthesis in children. In 2015, S.V. Vissarionov et al. [1] described a case of a 2-year-old patient with a congenital deformity of the thoracic spine with multilevel bilateral aplasia of the arc roots of the thoracic and lumbar vertebrae. The spinal deformity was stabilized using posterior multisupport tools under the control of the active 3D-CT- navigation. Stable correction improved the scoliotic deformity of the spine by 30 % and kyphosis by 50 %.

In our case, the patient had an extremely complex congenital malformation of the spine and the ribs. We were unable to find any description of such skeletal abnormalities in the literature. I would like to remark on its features once again: multiple anomalies of ribs (aplasia of the dorsal sections of ribs IV–VI on the left, ribs V and VI on the right and full aplasia rib I on the left and rib II on the right); aplasia of the arc roots in the cervical, thoracic and lumbar spine; hypoplasia of the T5 vertebral body with the formation of the thoracic kyphosis and spinal canal stenosis; complex deformity of the sacrum, which does not fit into the 5 known Cama types of caudal regression, hypoplasia of the sacral vertebrae bodies and aplasia of the lateral masses of the sacrum [17]; a decrease in the volume of the pelvic cavity.

The development of neurological symptoms in this patient was due to stenosis of the spinal canal at the apex of kyphosis. The kyphotic deformations of the vertebral bodies are always dynamically unstable, and the rate and the extent of kyphosis progression are inversely related to the extent of the development of anterior and middle vertebral column. The risk of neurological complications in congenital kyphosis is high and directly related to the type of deformity and the degree of spinal

Table
The dynamics of neurological disorders during the treatment

Scales	Before surgery	After surgery	
		After 20 days	After 6 months
Frankel	C	D	E
ASIA, points			
– Motor	81	90	100
– Needle pick	84	84	112
– Touch	84	84	112

canal stenosis. The average incidence of vertebral myelopathy in kyphosis is 21.1 %. In case of vertebral bodies agenesis, the incidence of neurological complications is greater than 66.0 % (the degree of the spinal canal stenosis is 2/3 or more of their proper values), in case of hypogenesis it amounts to 33.0 %, and in case of hypoplasia to 20.0 %. Combined kyphosogenic deformities (type III kyphoses) are accompanied by myelopathy in 30.8 % of cases [3, 4].

The use of three-dimensional full-scale models of the spine is becoming more widely used by spinal surgeons who deal with extremely complex deformities of the spine of congenital or another etiology [20]. 3D-models are constructed based on data from high-resolution CT, which provides the exact anatomic matching to the bone structures. There are no Russian articles on the use of three-dimensional plastic models in spine surgery. According to foreign publications, the most common approach to manufacture the model is stereolithographic prototyping method, which was used in this case. The availability of a full-scale plastic model of the deformed spine provides an invaluable advantage in comprehending and understanding the nature of spinal deformity due to the excellent visualization, even when compared with the three-dimensional CT image, and tactile information. In addition, it made it possible to conduct a stimulated operation, namely establish the scope and nature of the required bone resection for the purpose of decompression of neural structures, and planning and carrying out corrective osteotomies, etc. [21]. Equally important is the ability to assess the effectiveness of various metal fixators. We appreciated the advantages of using a volume model of the spine in this clinical case.

The clinical presentation of the complex congenital spine anomaly with spinal compression syndrome, the data of visualization techniques and assessment of the deformity using the stereolithographic model defined the choice of a method for surgical treatment. We ruled out posterolateral decompression of the spinal canal, since its implementation was associated with extremely high risk of exacerbation of the existing neurological symptoms. The deformity correction using dorsal instruments was also ruled out due to extremely high risk of destabilization of the metal construction in the postoperative period. This made us decide in favor of anterior decompression and anterior stabilization.

Conclusion

The surgical intervention helped to restore the spinal cord function, achieve regression of neurological symptoms and return the patient to active life. Even though the spinal deformity was not eliminated, in this particular case the removal of spinal stenosis and fixing the anterior spinal column were of paramount importance and were successfully achieved. The atypical congenital anomalies of the spine require case-by-case approach to the selection of method for surgical correction, which first of all takes into account the clinical presentation and anatomical features of the patient. Stereolithographic 3D model allows qualitative assessment of deformation and the selection of the best option for surgery.

References

1. **Vissarionov SV, Murashko VV, Murashko TV, Kokushin DN, Belyanchikov SM.** Surgical treatment of patients with congenital deformities in multilevel bilateral thoracic and lumbar pedicle aplasia. Hir. Pozvonoc. 2015;12(3):19–27. In Russian. DOI: http://dx.doi.org/10.14531/ss2015.3.19-27.
2. **Mikhailovsky MV, Ulrikh EV, Suzdalov VA, Dolotin DN, Ryabykh SO, Lebedeva MN.** VEPTR instrumentation in the surgery for infantile and juvenile scoliosis: first experience in Russia. Hir. Pozvonoc. 2010;(3):31–41. In Russia. DOI: http://dx.doi.org/10.14531/ss2010.3.31-41.
3. **Mushkin AYu.** Surgical correction of non-systemic angular kyphosis in children: Abstract of the MDSc Thesis. St.Petersburg, 2000. In Russian.
4. **Ulrikh EV, Mushkin AYu.** Vertebrology in Terms, Numbers, and Figures. St. Petersburg, 2004. In Russian.
5. **Ulrikh EV, Mushkin AYu.** Surgical Treatment of Malformations of the Spine in Children. St. Petersburg, 2007. In Russian.
6. **Ulrikh EV, Mushkin AY, Gubin AV.** Congenital spine deformities in children: epidemiological prognosis and management. Hir. Pozvonoc. 2009;(2):55–61. In Russian. DOI: http://dx.doi.org/10.14531/ss2009.2.55-61.
7. **Bergoin M, Bollini G, Taibi L, Cohen G.** Excision of hemivertebrae in children with congenital scoliosis. Ital J Orthop Traumatol. 1986;12:179–184.
8. **Bollini G, Bergoin M, Labriet C, Jouve, JL, Cottalorda J.** Hemivertebrae excision and fusion in children aged less than five years. J Pediatr Orthop B. 1992;1:95–101.
9. **Campbell RM Jr, Hell-Vocke AK.** Growth of the thoracic spine in congenital scoliosis after expansion thoracoplasty. J Bone Joint Surg Am. 2003;85:409–420.
10. **Campbell RM Jr, Smith MD, Hell-Vocke AK.** Expansion thoracoplasty: the surgical technique of opening-wedge thoracostomy. Surgical technique. J Bone Joint Surg Am. 2004;86 Suppl 1:51–64.
11. **Geoffray A, Clement JL, Leloutre B, Albertario M, Bechard-Sevette N.** A case of severe congenital kyphoscoliosis secondary to multiple bilateral thoracic pedicle aplasia. Pediatr Radiol. 2008;38:780–782. DOI: 10.1007/s00247-008-0790-4.
12. **Hadley LA.** Congenital absence of pedicle from the cervical vertebra. Am J Roentgenol Radium Ther. 1946;55:193–197.
13. **Ishida K, Aota Y, Uesugi M, Shinoda K, Mizuma K, Saito T.** Late onset of thoracic myelopathy with type 2 congenital deformity: a case report. The Open Spine Jornal. 2010;2:21–23.
14. **Lederman HM, Kaufman RA.** Congenital absence and hypoplasia of pedicles in the thoracic spine. Skeletal Radiol. 1986;15:219–223. DOI: 10.1007/BF00354064.
15. **Patel AJ, Vadivelu S, Desai SK, Jea A.** Congenital hypoplasia of aplasia of the lumbosacral pedicle as an unusual cause of spondylolisthesis in the pediatric age group. J Neurosurg Pediatr. 2013;11:717–721. DOI: 10.3171/2013.3.PEDS12579.
16. **Rauzzino MJ, Shaffrey CI, Bartkowski H, Nockels R, Wiggins G.** Multilevel thoracic pedicle aplasia causing congenital thoracic kyphosis: case report. Neurosurgery. 2000;46:988–990.
17. **Renshaw TS.** Sacral agenesis. J Bone Joint Surg Am. 1978;60:373–383.
18. **Ruf M, Harms J.** Halbwirbelresektion bei kongenitaler Skoliose. Operat Orthop Traumatol. 2004;16:205–220.
19. **Villas C, Barrios RH.** Congenital absence of the pedicles and the neural arch of L2. Eur Spine J. 1997;6:354–356. DOI: 10.1007/BF01142686.
20. **Yamazaki M, Okawa A, Fujiyoshi T, Kawabe J, Yamauchi T, Furuya T, Takaso M, Koda M.** Simulated surgery for a patient with neurofibromatosis type-1 who had severe cervicothoracic kyphoscoliosis and an anomalous vertebral artery. Spine. 2010;35:E368–E373. DOI: 10.1097/BRS.0b013e3181c42559.
21. **Yang JC, Ma XY, Lin J, Wu ZH, Zhang K, Yin QS.** Personalised modified osteotomy using computer-aided design-rapid prototyping to correct thoracic deformities. Int Orthop. 2011;35:1827–1832. DOI: 10.1007/s00264-010-1155-9.

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Received 15.03.2016

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