SURGICAL TREATMENT OF SPINAL PATHOLOGY IN CHILDREN WITH CAUDAL REGRESSION SYNDROME: ANALYSIS OF LONG-TERM OUTCOMES*

S.V. Vissarionov[1, 2], D.N. Kokushin[1], V.V. Murashko[1], S.M. Belyanchikov[1], T.B. Bogatyrev[1]
[1]Turner Scientific Research Institute for Children's Orthopedics, St. Petersburg, Russia
[2]Mechnikov Northwestern State Medical University, St. Petersburg, Russia

Objective. To analyze long-term outcomes of surgical treatment of spinal column pathology in children with caudal regression syndrome.
Material and Methods. The study included 12 patients aged 1.5 to 9 years with caudal regression syndrome. The Renshaw classification was used to determine the type of caudal regression. Surgery involved correction of kyphotic deformity of the spine and elimination of instability, and spinal-pelvic fusion including instrumented fixation of the vertebro-pelvic segment with restoration of the sagittal profile and support ability of the spine and creation of bone block by installing split cortical allografts along the spinal implant.
Results. Children with types III and IV caudal regression syndrome underwent spinal-pelvic fusion, which allowed achieving strong fixation of the vertebro-pelvic segment and ensured its stability at patient's verticalization.
Conclusion. Multi-anchor transpedicular instrumentation in combination with spinal-pelvic fusion with cortical allografts allows eliminating abnormal kyphosis, achieving bone block formation, and retaining the achieved result in the late postoperative period.
Key Words: caudal regression syndrome, sacral agenesis, lumbosacral agenesis, spinal-pelvic instability, surgical treatment, children.

**Vissarionov SV, Kokushin DN, Murashko VV, Belyanchikov SM, Bogatyrev TB. [Surgical Treatment of Spinal Pathology in Children with Caudal Regression Syndrome: Analysis of Long-Term Outcomes]. Hirurgia pozvonocnika. 2015;12(4):36–41. In Russian. DOI: http://dx.doi.org/10.14531/ss2015.4.36-41.*

Caudal regression syndrome is a rare and severe congenital malformation of the vertebral column and spinal cord combined with pathology of the internal organs and lower extremities. According to the clinical-neuroradiological classification proposed by Tortori-Donati et al. [15], the caudal regression syndrome belongs to the group of closed spinal dysraphism without subcutaneous mass. In terms of embryogenesis, this malformation results from disturbed notochord formation during gastrulation [9]. In their article 'Caudal Regression Syndrome' published in 2010 [1], Vissarionov and Kazaryan reviewed the non-Russian papers describing the pathology with special focus placed on the structural features of this syndrome and classification issues.

A number of recent Russian publications have been devoted to surgical treatment of children with caudal regression syndrome [2, 4]. In 2011, Vissarionov et al. [2] thoroughly described the procedure of surgical treatment of spinal pathology in this patient cohort and reported the short-term outcomes of surgical intervention. In 2014, a group of authors from the Novosibirsk Research Institute of Traumatology and Orthopaedics n.a. Ya.L. Tsivyan reported the results of combination surgical treatment of spinal malformation and pathology of the lower extremities in a patient with caudal regression syndrome [4]. The existing literature devoted to treating patients with this pathology typically focuses on surgical treatment outcomes based on individual clinical cases [1, 4–8, 10, 11, 13, 14, 16]. This study continues the previous publications and analyzes the long-term outcomes of surgical treatment of vertebro-pelvic instability in a large group of children with caudal regression syndrome.

The study aims at analyzing the long-term outcomes of surgical treatment of vertebro-pelvic instability in children with caudal regression syndrome.

Material and Methods

The study involved 12 patients (8 boys and 2 girls) with caudal regression syndrome. Patients were 1.5–3 years old; one child was 9 years old.

Clinico-neurological and X-ray examination were employed during the study, including X-ray and multislice spiral computed tomography (MSCT) of the spine and pelvis; MRI of the cra-

niovertebral area, thoracic and lumbar spine.

For all patients, the clinical presentation of the pathology involved kyphotic deformity at the vertebro-pelvic level and barrel-shaped thorax. No distortion of the spinal column in the coronal view was observed. Among seven children with type 4 caudal regression syndrome according to the Renshaw classification [4], five children had skin thinning with hyperemia at the apex of the kyphosis caused by pressure exerted on this region by the caudal spine. All the patients had a shortened intergluteal cleft and hypoplasia of the sacral and gluteal areas. Four patients with type 3 caudal regression had bilateral hip dislocation. Only one child did not have this pathology. Bilateral paralytic clubfoot and tibial muscular hypotrophy were observed for all the patients with type 3 lumbosacral agenesis. The patients with this type of caudal regression were able to maintain vertical position when supported but could not move around independently.

Patients with type 4 caudal regression syndrome had flexion-abduction contractures of hip joints, flexion contractures of knee joints with severe webbing of the skin (pterygium) in the popliteal region and equinovarus foot deformity. These patients had muscle hypotrophy in the proximal and distal regions of the lower extremities. Active movements in the lower extremities were totally absent; passive movement capability was retained within 5–10°.

Neurological examination aimed at detecting motor and sensory disorders of the central nervous system. Pediatric examination made it possible to assess the extent of pathological changes in the internal organs associated with the deformity of the vertebro-pelvic area and concomitant disorders [3].

The neurological status of the patients with type 3 caudal regression syndrome included lower extremity peripheral paraparesis, mostly in the distal regions of the lower extremities. Pain and temperature sensitivity was retained. Pelvic organ dysfunction was revealed. In patients with type 4 caudal regression syndrome, the neurological deficit manifested itself as lower extremity paraplegia, the absence of pain or temperature sensitivity in the lower extremities, and pelvic organ dysfunction.

X-ray examination of the spine and pelvis in the two standard views was carried out for patients in a lying position. It is difficult to accurately quantify the kyphotic deformity of the vertebro-pelvic segment in degrees, since patients with this malformation of the spine and spinal cord have no sacrococcygeal and/or lumbar spinal segments and even the lower thoracic spinal segment in some cases. The method for determining the angle of kyphotic deformity of the vertebro-pelvic segment in children with caudal regression syndrome was designed to reliably assess the 3D position of the spine and pelvic bones and to objectivize surgical treatment outcomes. The method for assessing pathological kyphosis consisted in measuring the angle formed by two intersecting lines: the one running along the posterior surface of the normal vertebral bodies in the caudal spine and the second one running through the anterior surface of the contour of iliac bones (Fig. 1).

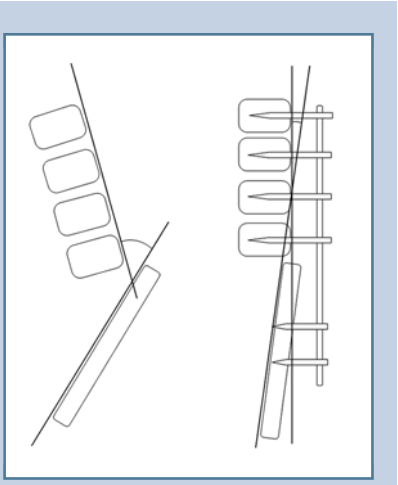

Fig. 1
Method for determining the angle of kyphotic deformity of the vertebro-pelvic segment in children with caudal regression syndrome

According to the X-ray data, the Cobb angle for the kyphotic deformity of the vertebro-pelvic segment determined using the aforedescribed procedure preoperatively varied from 45 to 73° (mean value, 60°) and from 45 to 100° (mean value, 75°) in patients with type 4 and type 3 caudal regression syndrome, respectively.

The anatomical and anthropometric characteristics of bone structures of the deformed vertebrae and the pelvic complex were evaluated using the MSCT data. The resulting data were used to refine the type of caudal regression, the size and shape of the bodies of the caudal spinal segment, and 3D position of pelvic bones as well as to perform preoperative planning and determine the optimal variant, number, type, and size of the supporting elements of transpedicular instrumentation (Fig. 2).

MRI of the spine was performed to detect the presence of intracanal pathology, assess the condition of the spinal cord and its elements. The MRI scans were used to assess the position and type of changes in the spinal cord and determine the level of medullary regression.

To determine the type of caudal regression, we employed the Renshaw classification, which is based on the tactical approach to selecting either the conservative or surgical method for treating patients with this pathology by determining instability of the vertebro-pelvic segment [12].

The results of CT and MRI scans of the spine were used to determine the vertebral and medullary levels in patients with caudal regression syndrome (Table).

According to the data listed in Table, patients with type 3 lumbosacral agenesis had more normal spinal and spinal cord segments compared to patients with type 4 lumbosacral agenesis.

All the patients underwent surgical management that involved correction of the kyphotic spinal deformity, elimination of instability and spinal-pelvis block, which included instrumented fixation of the vertebro-pelvic segment with a multi-anchor dorsal transpedicular instrumentation that allowed restoration of the physiological sagittal pro-

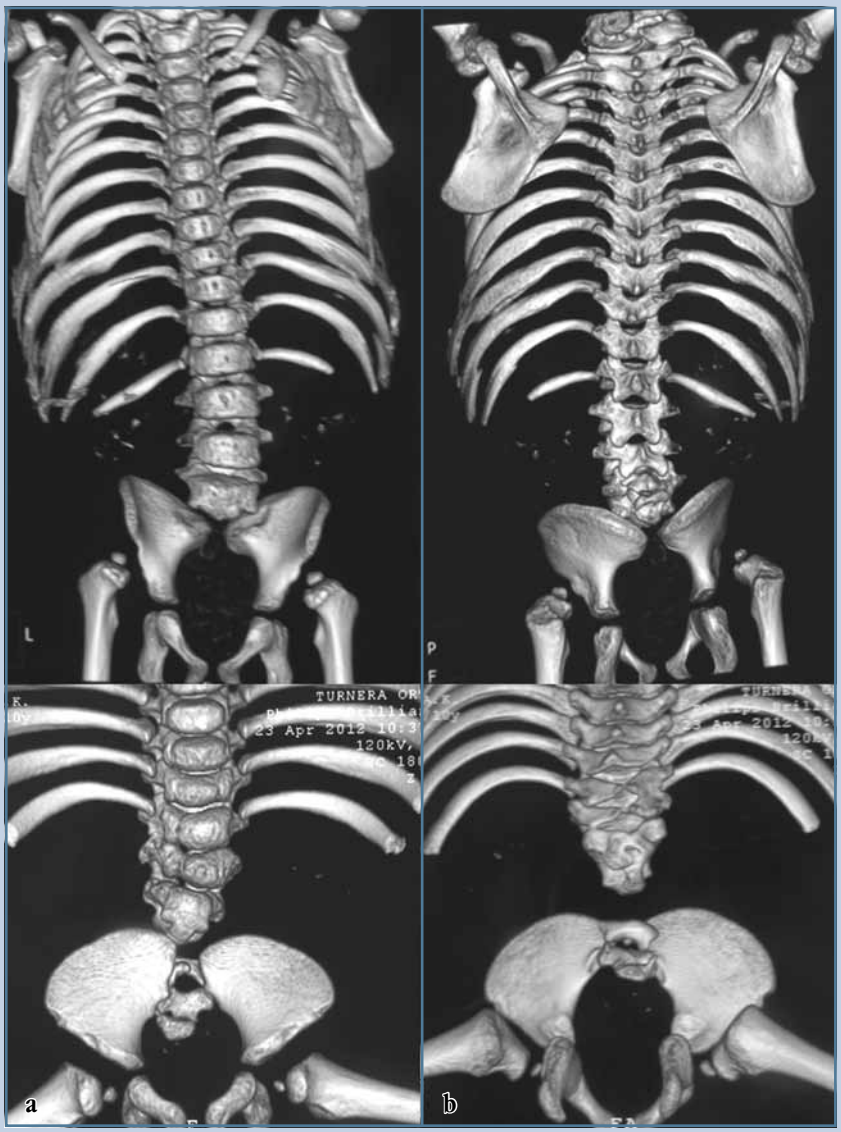

Fig. 2
CT of the spine and pelvis in patients with caudal regression syndrome at admission: **a** – 2-year-old patient E. with type 3 caudal regression syndrome; **b** – 1.5-year-old patient S. with type 4 caudal regression syndrome

Table
Caudal regression

Regression type according to the Renshaw	T8	T9	T10	T11	T12	L1	L2	L3	L4	L5	S1	S2	Total
Vertebral level													
3rd	–	–	–	–	–	–	–	–	4	1	–	–	5
4th	–	1	–	1	1	1	1	1	1	–	–	–	7
Medullary level													
3rd	–	–	1	–	2	1	–	1	–	–	–	–	5
4th	1	3	1	1	1	–	–	–	–	–	–	–	7

file, supporting function of the vertebral column, and formation of fusion by using split cortical allografts along the spinal implant. Transpedicular screws and laminar hooks were installed into caudal vertebral bodies during the surgery; screw and hook support elements of transpedicular instrumentation were used for bone fixation. The choice of support elements to be installed in the caudal spine depended on anatomical and anthropometric characteristics of vertebral bodies, while the type of supporting elements of transpedicular instrumentation used for pelvis stabilization depended on thickness of the cortical laminae and spongy tissue of the iliac bones that is determined based on the CT data. The postoperative period included breathing exercises, massage of the lower and upper extremities, and remedial gymnastics.

Children were examined before surgical management, immediately after surgery, and subsequently 6, 12, and 18 months after the surgery, and then once every year. The duration of postoperative follow-up varied from 2 to 7 years.

Results

Clinical presentation and X-ray examination showed that five patients with caudal regression syndrome had type 3 lumbosacral agenesis according to the Renshaw classification; seven patients had type 4 lumbosacral agenesis.

The pathological kyphosis and instability of the vertebro-pelvic segment were corrected in all the patients after surgical treatment. Three patients with type 3 caudal regression showed improvement of motor activity; they became able to move independently. The same children showed improved function of the pelvic organs as they started to have the urge to urinate or defecate and gained voluntary control over these processes.

X-ray examination showed that the postoperative angle of the vertebro-pelvic segment varied from 28 to 32° (mean value, 29.7°) and 14 to 55° (mean value, 33.2°) in patients with type 3 and 4 lumbosacral agenesis, respectively. The

X-ray and CT data demonstrated that a solid bone block was formed between the caudal spine and pelvic bones in all the patients 2–2.5 years after surgery, which ensured stability at this level (Fig. 3). Transpedicular instrumentation was removed after the surgery in none of the cases.

Patients with type 3 caudal regression syndrome were wearing rigid braces and verticalized on day 3–10. Customized rigid braces that fixed the lower extremities up to the shin level so that the patients were able to sit were manufactured for patients with type 4 caudal regression. The patients were discharged from the hospital and transferred to outpatient care on day 17–21.

Five patients had complications: long healing of surgical wound and destabilization of transpedicular instrumentation. Secondary intention healing of soft tissues at the surgical site was observed in three patients in the early postoperative period, requiring conservative treatment and the use of special bandages. Two patients had a destabilized transpedicular instrumentation element in the area where the supporting elements were installed in pelvic bones. These complications required reoperations aimed at stabilizing supporting elements of transpedicular instrumentation, which had no effect on the final outcome of treatment.

Discussion

According to the literature data, the absence of surgical aid significantly affects the function of the internal organs and life expectancy of patients with caudal regression syndrome [12]. Pathological kyphosis has been corrected, the supporting ability of the vertebro-pelvic segment has been restored, and conditions for growth and development of the spine and internal organs under functionally favorable conditions have been created during surgical treatment in all the patients in our study.

We believe that improvement of motor activity and function of the pelvic organs in patients with type 3 caudal regression syndrome is related to correction of the kyphotic component of the deformity and instability at the level of the vertebro-pelvic segment. This allowed us to provide conditions for patient verticalization, more favorable spine biomechanics in general, and physiological position of the internal organs.

In patients with type 4 caudal regression syndrome, the correction of the deformity of the vertebro-pelvic segment combined with osteoplasty has created conditions for improving the supporting ability of the spine and made it possible for patients to sit and undergo further social rehabilitation.

The complications that emerged during the early postoperative period presenting as long postoperative wound healing can be attributed to severe disruption of soft-tissue trophism caused by the initial neurological deficit in patients with caudal regression syndrome. Destabilization of transpedicular instrumentation was observed in the first patient followed up and in the 9-year-old child. In the former case, the complication was attributed to the fact that it was the first surgical intervention and to the selected variants of supporting elements of transpedicular instrumentation and methods for deformity correction and stabilization of the achieved result. In the latter case, the complication was caused by the severe rigid spine deformity due to the patient's age and by the difficulty of its correction. In both cases, we used hooks as supporting elements of transpedicular instrumentation in pelvic bones. Furthermore, the complications that have emerged resulted from patient noncompliance as well as pronounced hypoplasia and porosity of pelvic bones.

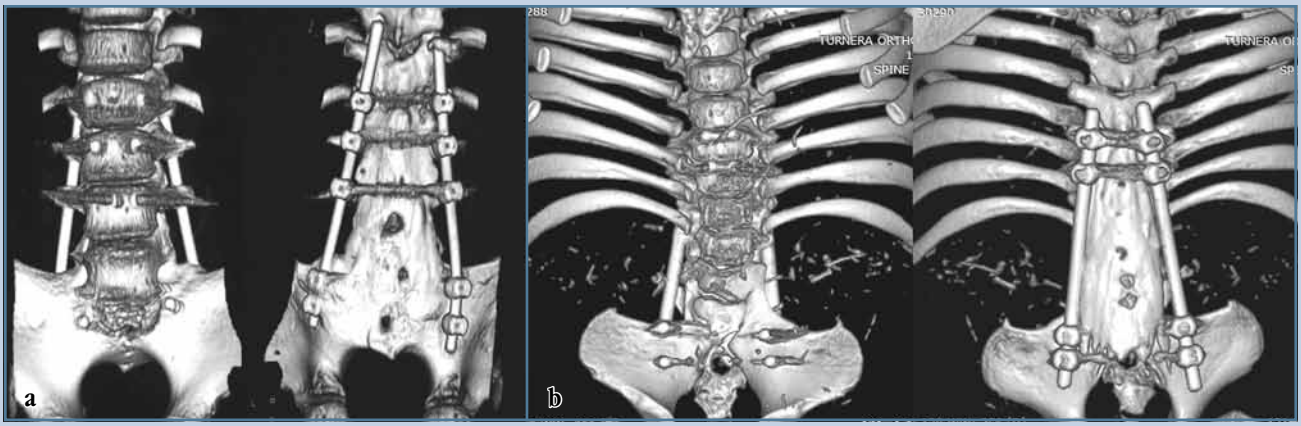

Fig. 3
Postoperative CT scans of the spine and pelvis of patients with caudal regression syndrome: **a** – long-term outcome of surgical treatment of patient E. (6 years old) with type 3 caudal regression 4 years after surgery; **b** – long-term outcome of surgical treatment of patient S. (5 years old) with type 4 caudal regression 3.5 years after surgery

Conclusion

Patients with type 3 and 4 caudal regression are characterized by the presence of kyphosis and instability at the level of vertebro-pelvic segment. We believe that these patients need surgical management at an early age.

Surgical treatment aims at correcting pathological kyphosis and eliminating instability at the level of the vertebro-pelvic segment. The use of a multi-anchor transpedicular instrumentation with screw supporting elements combined with spinal-pelvic fusion using cortical allografts not only allows one to solve the problems described above but also to form the sagittal profile of the vertebral column, ensure its supporting ability, form a bone block at the intervention site, and preserve the result in the long-term postoperative period, thus providing conditions for improving patient's motor activity, enabling his/her verticalization, and possibilities for spine development as children grow, and social adaptation for children.

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Address correspondence to:
Vissarionov Sergey Valentinovich
The Turner Scientific and Research Institute for Children's Orthopaedics,
Parkovaya str., 64–68, Pushkin, Saint-Petersburg, 196603, Russia,
turner01@mail.ru

Sergey Valentinovich Vissarionov, MD, DMSc, The Turner Research Institute for Children's Orthopedics, North-Western State Medical University n.a. I.I. Mechnikov, St. Petersburg; Dmitrii Nikolayevich Kokushin, researcher; Vladislav Valeryevich Murashko, MD; Sergey Mikhailovich Belianchikov, MD, PhD; Timur Bagaudinovich Bogatyrev, MD fellow, The Turner Research Institute for Children's Orthopedics, St. Petersburg, Russia.