Surgical Correction of Spinopelvic Instability in Children With Caudal Regression Syndrome

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Abstract

Study Design: Retrospective cohort.

Objective: To analyze the outcome of surgical correction of children with caudal regression syndrome.

Methods: The study included 12 patients aged 1.5 to 9 years with caudal regression syndrome. In order to determine the type of caudal regression, the Renshaw Classification was used. The surgery included correction and stabilization of the kyphotic deformity at the unstable lumbosacral region, with reconstruction of the sagittal balance using a bony block constructed from allograft. Short- and long-term outcomes were evaluated. The study was approved by the local institutional review board.

Results: Children with types III and IV caudal regression syndrome underwent spinal-pelvic fusion, with 100% fusion rate, which allows sufficient stabilization of the lumbopelvic segment permitting patient mobilization and standing in type III patients. There were 5 complications needing additional care.

Conclusion: Multilevel pedicular screw fixation in combination with spinopelvic fusion with cortical allografts allows reconstruction of the sagittal alignment with solid bony fusion improving the quality of life for these patients.

Keywords
caudal regression syndrome, sacral agenesis, lumbosacral agenesis, spinopelvic instability, surgical treatment, children

Introduction

Caudal regression syndrome is a rare congenital malformation of the spine and caudal spinal cord combined with a pathology of the visceral organs and lower extremities. According to the clinical and neuroradiological classification of Tortori-Donati et al,1 caudal regression syndrome pertains to a group of closed forms of spinal dysraphism without subcutaneous mass. In the context of embryogenesis, this defect is a result of a failure of notochord formation occurring at the stage of gastrulation.2,3

Renshaw et al4 divide the condition into 4 groups (Figure 1): type I is defined as a partial or total unilateral sacral agenesis. Type II has a partial sacral agenesis with a bilateral symmetrical defect, a normal or hypoplastic sacral vertebra, and an articulation between the ilia and the first sacral vertebra. In type III, there is a variable lumbar and total sacral agenesis, with the ilia articulating with the sides of the lowest vertebra present; and in type IV, a variable lumbar and total sacral agenesis exist, with the caudal endplate of the lowest vertebra resting above either fused ilia or an iliac amphiarthrosis.

Recently, a number of studies focusing on the surgical treatment of children with caudal regression syndrome were presented.5,6 The existing literature on the treatment of patients with this pathology considers the results of the surgical treatment of individual cases with short-duration follow-up.5-15

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The objective of this article is to analyze the long-term results of surgical treatment of spinopelvic instability in children with caudal regression syndrome.

**Materials and Methods**

Twelve patients (8 boys and 4 girls) with caudal regression syndrome were surgically corrected and retrospectively reviewed in the study (Table 1). Surgeries were performed between 2008 and 2014 at a single referral center.

Clinical and radiological examinations (spine and pelvis X-rays, computed tomography [CT] scans, and magnetic resonance imaging (MRI) examinations of the cervical, thoracic, and lumbar spine) were reviewed.

Patient pathology was described according to the Renshaw Classification (Figure 1), which divides patients’ pathology according to the level of spinopelvic segment instability and aims to assist in treatment planning between conservative and surgical treatment of patients with this pathology.4

Neurological examination of the patients is aimed to detect any motor and sensory disorders that may vary from deformity to deformity. The pediatric physical examination evaluates the nature of pathological changes in visceral organs associated with the deformity of spinopelvic segment and concurrent conditions.16

Spine and pelvis X-rays are performed in 2 standard projections in the supine position. Kyphosis was measured through the Cobb angle formed by the intersection of 2 lines: one passing through the posterior surface of the lowest remaining vertebrae and other through the anterior surface of the pelvis ilium (Figure 2).

Anatomical and anthropometric features of bony structures of the deformed vertebrae and pelvic complex were assessed according to CT findings. The type of caudal regression, size and shape of caudal vertebral bodies, and spatial position of the pelvis were specified, and preoperative planning was done with the choice of optimal instrumentation design and the number and sizes of anchoring elements (Figure 3).

Intracanal pathology and the condition of the spinal cord and its components were identified on the spine MRI scans, which

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**Table 1. Vertebral Level of the Caudal Regression.**

| Renshaw Type of Regression | T9 | T10 | T11 | T12 | L1 | L2 | L3 | L4 | L5 | S1 | S2 | Total |
|-----------------------------|----|-----|-----|-----|----|----|----|----|----|----|----|-------|
| Type III                    | —  | —   | —   | —   | —  | —  | 4  | —  | —  | —  | —  | 5     |
| Type IV                     | —  | —   | —   | —   | —  | —  | —  | —  | —  | —  | —  | 7     |

*Figure 1. Renshaw sacral agenesis classification. (A) type I; (B) type II; (C) type III; (D) type IV.*

*Table 1. Vertebral Level of the Caudal Regression.*

Vissarionov et al
allowed assessing the location and nature of changes in the spinal cord and detecting the level of medullary regression.

Based on the results of spine CT and MRI, the affected vertebral and conus medullaris levels were determined in patients with caudal regression syndrome (Tables 1 and 2).

Children were examined before surgery, immediately after surgery, and at 6, 12, and 18 months after surgical treatment, and thereafter once a year. Postoperative follow-up period ranged from 2 to 7 years (see Figure 4).

**Surgical Approach (Figure 5)**

Surgical approach in all patients included a corrective osteotomy of the kyphotic deformity of the spine combined with an S fixation in order to eliminate the instability. This was performed with a posterior based spinopelvic fusion restoring the physiological sagittal profile. The newly generated support for the spine is based on the rods with the placement of split cortical allografts along the spinal implants allowing solid bony fusion (Figure 5C). Pedicle screws or laminar hooks were inserted into vertebral bodies of the caudal segments of the spine and the pelvis. The selection of the instrumentation inserted into the caudal spine segments was dependent on the anatomical features of vertebral bodies. The instrumentation used to stabilize the pelvis was based on the thickness of cortical plates and iliac spongy tissue as determined by CT measurement.

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**Figure 2.** Method for measuring the magnitude of kyphotic deformity of spinopelvic segment in children with caudal regression syndrome.

**Figure 3.** Admission spine and pelvis computed tomography scans in patients with caudal regression syndrome on admission: (a) Patient E, 2-year-old child with type III caudal regression syndrome; (b) Patient S, 1.5-year-old child with type IV caudal regression syndrome.

**Table 2.** Medullary Level of the Caudal Regression.

| Renshaw Type of Regression | T8 | T9 | T10 | T11 | T12 | T13 | L1 | L2 | Total |
|----------------------------|----|----|-----|-----|-----|-----|-----|-----|-------|
| Type III                   | —  | —  | 1   | —   | 2   | 1   | —   | 1   | 5     |
| Type IV                    | 1  | 3  | 1   | 1   | 1   | —   | —   | —   | 7     |
Patients with type III caudal regression were mobilized at the 3rd to 10th days after surgery into a rigid brace. Patients with type IV were protected with custom-made rigid braces with a pantaloon extension allowing sitting. Patients were discharged after surgery to outpatient treatment at the 17th to 21st days after surgery.

The postoperative rehabilitation program included respiratory exercises, massage of lower and upper extremities, and restoration exercises such as balanced sitting, standing, and ambulation with the braces.

**Results**

Twelve children were included in the study. The age of children operated ranged between 1.5 and 3 years, and only one child was 9 years old.

**Clinical and Neurological Status**

Clinical presentation of the disease included kyphosis at the level of the spinopelvic segment in all patients. The thorax was barrel-shaped. No scoliosis was present. Five of the 7 children having Renshaw type IV caudal regression suffered from skin thinning with hyperemia over the apex of kyphosis due to the pressure caused by caudal part of the spine. All patients had shallow intergluteal cleft and hypoplasia of the sacrum and gluteal region.

Concerning the associated pathology in the lower extremities, 4 patients with type III caudal regression had bilateral hip dislocations. Bilateral paralytic clubfeet and leg muscle hypotrophy was observed in all cases of type III lumbosacral agenesis. Patients with this type of caudal regression retained the ability to stay upright with a support but could not move independently.
Patients with type IV caudal regression had flexion-abduction contractures of the hip, flexion contractures of the knee with severe skin pterygia of popliteal areas, and equinus foot deformity. These patients had muscle hypotrophy of the proximal and distal parts of lower extremities. Active movements in the lower extremities was completely absent, and the passive range of motion remained within 5° to 10° at all joints. Neurological status of patients with type III caudal regression included peripheral lower extremity paraparesis, mainly in the distal lower extremities. Pain and thermal senses were preserved. Examination revealed bowel and bladder dysfunction. Neurologic deficit in type IV caudal regression was manifested by lower extremity paraplegia, absence of pain and thermal sense in lower extremities, and bowel and bladder dysfunction.

**Imaging**

According to findings of X-ray examination, the mean preoperative kyphotic angle of spinopelvic segment as defined by the above-mentioned method was 60° (range = 45° to 73°) in patients with type III caudal regression and 75° (range = 45° to 100°) in patients with type IV caudal regression.

Tables 1 and 2 show that patients with type III lumbosacral agenesis had greater number of intact segments of the spine and spinal cord as compared with those with type IV.

**Surgical Results**

The performed surgical treatment corrected abnormal kyphosis and prevented spinopelvic instability. Three patients with type III caudal regression showed an improvement in motor function manifesting as the ability to ambulate independently. These children also improved the bowel and bladder function in the form of independent urge and control of urination and defecation.

Postoperative X-rays showed mean angle of spinopelvic segment to be 29.7° (range = 28° to 32°) in patients with type III lumbosacral agenesis and 33.2° (range = 14° to 55°) in patients with type IV lumbosacral agenesis. According to the CT scans, a solid bone fusion was formed in the area of surgical intervention between the caudal segment of the spine and pelvis in all patients at 2 to 2.5 years after surgery, providing stability at this level (Figure 4). None of the patients underwent the removal of spinal implant after surgery.

Five patients had complications after surgery. Delayed wound healing was observed in 3 patients in the early postoperative period (25%), leading to healing by secondary healing with the use of special wound dressings. In 2 patients (16.6%), loosening of the pelvic implants was observed. These patients underwent revision surgery to improve pelvic anchorage. At latest follow-up all fixations were stable without loss of correction. Neurological status that was gained in surgery was not lost and the children remained ambulators. None of the complications affected the long-term outcome of the patients.

**Discussion**

Caudal regression syndrome is a grave congenital defect, and the absence of surgical care and correction of the deformity and stabilization of the spine has a significant impact on the function of the visceral organs and shortens the life expectancy of the patients. The surgical treatment in caudal regression syndrome is difficult with a high complication rate. Surgery needs to reduce kyphosis and allow a balance of sitting and standing positions with balanced sagittal profile. In this series, all patients presented showed elimination of pathological kyphosis, restoration of support ability of spinopelvic segment, and creation of functionally favorable conditions for the growth and development of the spine and visceral organs.

The improvement of motor activity and bowel and bladder function in patients with type III caudal regression was, in our opinion, due to elimination of the kyphotic component of deformity and instability at the level of the spinopelvic segment. This enabled the verticalization of patients, more favorable biomechanics of the spine, and physiological arrangement of visceral organs.

In patients with type IV caudal regression, the performed correction of deformity of spinopelvic segment combined with bone grafting created the conditions for support ability of the spine and provided an opportunity for physiological sitting and further social rehabilitation.

It should be noted that the exact numerical measurement of the kyphosis of spinopelvic segment is challenging since this congenital malformation of the spine and spinal cord implies the absence of sacroccygeal joint and/or lumbar spine, and in some cases of the lower thoracic spine. To reliably assess the spatial position of the spine and pelvis and objectify the results of surgical treatment, the method for measuring kyphosis of the spinopelvic segment in children with caudal regression syndrome was developed by the authors and used here.

There is little data regarding the optimal correction of this rare condition. Older techniques did not use modern instrumentation and lacked full correction of the deformity. Freland et al described a case series of 6 patients that used vascular rib grafts with complete correction of the deformity. This technique requires harvesting of the ribs and additional surgery is not needed in the technique described here. Fusion rates in both series are high; however, the complication rate was higher in their series, reaching 7 revision procedures in 4 patients (66% complication rate) versus 5 complications in the series presented here (41.6% complication rate).

Complications that occurred in the early postoperative period in the form of delayed healing of surgical wounds can be attributed to severe impaired trophism of soft tissues caused by the initial neurological deficit in patients with caudal regression syndrome. Destabilization of the implant was observed in the first patient under our supervision, and in a child aged 9 years. In the first case, the complication was explained by the learning curve needed with inferior selection of appropriate anchoring elements and methods of deformity correction and maintaining the achieved results. In the second case, a
complication was associated with severe rigid deformity of the spine due to the patient’s age and with challenges in its correction. Anchoring elements in both cases were hooks inserted into pelvic bones.

The limitations of the study are its retrospective nature and small number of patients; however, it is one of the largest series on this pathology documented.

**Conclusion**

Patients with types III and IV caudal regression syndrome are characterized by the presence of kyphosis and instability at the level of spinopelvic segment. In our view, such patients should undergo surgical treatment early in life.

Surgical treatment is aimed at eliminating abnormal kyphosis and instability at the level of spinopelvic segment. Application of metal implant with multiple anchoring elements in combination with spinopelvic fusion with cortical allografts allows not only to solve the problem described above but also to form the sagittal profile of the spine, create its support ability, achieve a bone block in the area of intervention, and maintain the achieved result in the late postoperative period. All this enables the improvement of motor activity and verticalization of patients, development of the spine in the process of growth, and social adaptation of the children.

**Declaration of Conflicting Interests**

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**References**

1. Tortori-Donati P, Rossi A, Biancheri R, Cama A. Congenital malformations of the spine and spinal cord. In: Tortori-Donati P, Rossi A. *Pediatric Neuroradiology*. Berlin, Germany: Springer-Verlag; 2005:1551-1608.
2. Vissarionov SV, Kazaryan IV. Caudal regression syndrome [in Russian]. *Hir Pozvonoc*. 2010;(2):50-55.
3. Harlow CL, Partington MD, Thieme GA. Lumbosacral agenesis: clinical characteristics, imaging, and embryogenesis. *Pediatr Neurosurg*. 1995;23:140-147.
4. Renshaw TS. Sacral agenesis. *J Bone Joint Surg Am*. 1978;60:373-383.
5. Vissarionov SV, Kazaryan IV, Belyanchikov SM. Treatment of patients with caudal regression syndrome [in Russian]. *Hir Pozvonoc*. 2011;(3):56-59.
6. Semyonov AL, Ryzhikov DV, Mikhailovsky MV, Vasyura AS. Result of comprehensive surgical treatment of a patient with caudal regression syndrome [in Russian]. *Hir Pozvonoc*. 2014;(4):106-111.
7. Cama A, Palmieri A, Capra V, Piatelli GL, Ravegnani M, Fondelli P. Multidisciplinary management of caudal regression syndrome (26 cases). *Eur J Pediatr Surg*. 1996;6(suppl 1):44-45.
8. Dal Monte A, Andrisano A, Capanna R. The surgical treatment of lumbo-sacral coccygeal agenesis. *Ital J Orthop Traumatol*. 1979;5:259-266.
9. Dumont CE, Damsin JP, Forin V, Carlizio H. Lumbosacral agenesis. Three cases of reconstruction using Cotrel-Dubousset or L-rod instrumentation. *Spine (Phila Pa 1976)*. 1993;18:1229-1235.
10. Guidera KJ, Raney E, Ogden JA, Highhouse M, Habal M. Caudal regression: a review of seven cases, including the mermaid syndrome. *J Pediatr Orthop*. 1991;11:743-747.
11. Perry J, Bonnett CA, Hoffer MM. Vertebral pelvic fusions in the rehabilitation of patients with sacral agenesis. *J Bone Joint Surg Am*. 1970;52:288-294.
12. Phillips WA, Cooperman DR, Lindquist TC, Sullivan RC, Millar EA. Orthopaedic management of lumbosacral agenesis. Long-term follow-up. *J Bone Joint Surg Am*. 1982;64:1282-1294.
13. Rieger MA, Hall JE, Dalury DF. Spinal fusion in a patient with lumbosacral agenesis. *Spine (Phila Pa 1976)*. 1990;15:1382-1384.
14. Singh SK, Singh RD, Sharma A. Caudal regression syndrome—case report and review of literature. *Pediatr Surg Int*. 2005;21:578-581. doi:10.1007/s00383-005-1451-4.
15. Winter RB. Congenital absence of the lumbar spine and sacrum: one-stage reconstruction with subsequent two-stage spine lengthening. *J Pediatr Orthop*. 1991;11:666-670.
16. Vissarionov SV, Kokushin DN, Bogatyrev TB. Malformations of the internal organs and systems in children with asymptomatic spinal dysraphism [in Russian]. *Pediatr Traumatol Orthop Reconstr Surg*. 2015;3:5-9.
17. Ferland CE, Sardar ZM, Abduljabbar F, Arlet V, Ouellet JA. Bilateral vascularized rib grafts to promote spinopelvic fixation in patients with sacral agenesis and spinopelvic dissociation: a new surgical technique. *Spine J*. 2015;15:2583-2592.