Literature review

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Variants of segmental spinal dysgenesis: literature review and our own experience of treatment

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Purpose The purpose of the work was to study the clinical and neuroradiological features of segmental spinal dysgenesis (SSD), congenital anomaly with local hypogenesis of a spinal motion segment and the spinal cord; as well as the selection of an optimal tactics of treating this anomaly. **Methods** The authors reviewed the literature and present seven clinical cases of their own practice. **Results** The severity of clinical deficit correlates with the degree of morphological changes in the spinal cord, the level of dysgenesis, as well as with the presence of combined and concomitant developmental anomalies. Early decompression of neural structures and spine stabilization play an important role in improvement of treatment results but the immature bone structures in infants limit the possibilities of early instrumented fixation. **Conclusion** SSD is a specific developmental anomaly of the spine and spinal cord that features some clinical and radiological peculiarities. This anomaly fills the gap between the infant thoracolumbar kyphosis with segmental subluxation and the syndrome of caudal regression. Early diagnosis and individual approach to determination of an optimal period of surgical treatment and tactics are required in each case.

Keywords: segmental spinal dysgenesis, congenital dislocation of the spine, congenital kyphosis

TERMINOLOGY

Scott R.M. and coauthors [1] first proposed the term *Segmental Spinal Dysgenesis* (SSD) in 1988 and defined the disease criteria: segmental anomaly of spine development by which a spine segment that lies below has a normal structure, the vertebral body at the deformity apex is dysplastic and displaced dorsally thus pressing the spinal cord, its membranes and roots.

Faciszewski T. and coauthors added more details to SSD criteria in 1995 [2]: local stenosis and vertebral canal deformity of the hourglass type, absence of neurocentral union of pedicles, osseous processes, transverse processes at the level of dysgenesis; presence of a bony ring that surrounds the spinal cord which is shifted to posterior; absence of the roots at the stenosis level. The vertebrae below the anomaly have normal structure.

Dubousset J. introduced the term *congenital vertebral displacement* (CVD) prior to R.M. Scott and meant: spine malformation that is characterized by the displacement of the abnormal adjacent vertebrae relative to each other that result in an abrupt spinal angulation and spinal canal narrowing [3]. Literature sources give other terms to the clinical cases described that suit the SSD criteria such as *congenital spinal stenosis*, *congenital duplication of the spine*, *congenital kyphosis*, *congenital dislocated spine*, or *congenital lumbar kypholisthesis*.

Dias M.S. and coauthors [4] proposed to distinguish three groups of anomalies in 1998: CVD, SSD and MSA that differ in severity and manifestation time during the embryonic development. The CVD criteria proposed were dysgenesis of not more than two neighbouring vertebrae with a partial malformation of their posterior elements with the spine and spinal cord above and below the malformation having normal structures while the spinal cord and roots at the malformation level being hypoplastic. Dias's criteria of the SSD were dysgenesis in two to 8 vertebrae associated with the underdevelopment of the posterior structures, spinal canal stenosis, a bony ring round the spinal cord and roots at the stenosis level, relatively

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normal vertebrae above and below the anomaly level, hypogenesis of the spinal cord and roots at the stenosis level, presence of the spinal cord above and below the anomaly level, either almost normal sensomotor function below the disorder level or incomplete neurologic deficit. MSA criteria according to Dias were: segmental or extended agenesis in 3 to 11 vertebrae without an associated lumbosacral agenesis, agenesis of the spinal cord more caudal to the malformation, congenital paraplegia lower the malformation level accompanied by severe limb deformity.

Tortori-Donati P. and coauthors in 1999 [5] concluded that SSD is not a sole disease entity and should be viewed as a morphological continuum in the range from spinal cord hypoplasia with a continuous vertebral column along the anomaly extension at one end to complete spinal cord and vertebral column absence at the other.

Tsirikos A. I. (2010) and Ganesan S. et al. (2015) define such a pathology as infantile developmental thoracolumbar kyphosis with segmental subluxation of the spine that lies at a boarder-line with SSD and is characterized by absent neurologic deficit and kyphosis initially but which appear in the age between six and 18 months [6, 7]. The instrumental study defines an isolated hypoplasia of the body of one of the vertebrae with intact posterior structures. In some cases, progression of the deformity is observed and requires surgical treatment. But frequently, this anomaly can be compensated with age as the spine structures become mature and while treated with bracing.

CLINICAL FINDINGS, DIAGNOSIS AND TREATMENT METHODS

Scott R.M. and coauthors (1988) [1] pointed to the necessity of an early intervention focusing, first of all, on early neural structures decompression. They described their experience of treating three patients with SSD. All underwent posterior and partially anterior decompression along with spondylodesis with an autologous bone graft followed by prolonged fixation in braces.

Faciszewski T. and coauthors (1995) [2] presented 17 patients treated for SSD. Each patient underwent from one to seven operations to achieve the required stabilization of the segment: removal of abnormal vertebral bodies along with anterior spondylodesis with bone grafts, posterior instrumentation spondylodesis. Neurologic status improved in two patients.

Ford E.G. and coauthors (1996) [8] gave a detailed description of treating a female patient with SSD L1 without neurologic symptoms. At the age of five months she underwent removal of the abnormal L1 body from the anterior approach, corporodesis with a rib graft and autologous bone splits. The patient had bracing for eight months postoperatively that was followed by the second intervention of posterior non-instrumented spondylodesis and bracing for two months. The operation achieved a good outcome though there was a slight residual kyphosis that the authors attributed to a partial rib graft osteolysis.

Flynn J.M. and coauthors (1997) [9] presented the treatment outcomes of seven SSD patients. In three of five patients with lower paraparesis, the neurologic deficit was absent initially. Paresis developed during the first two years of their life. The age at surgery was from 4.5 to 92 months. The authors followed such treatment tactics: anterior decompression with removal of abnormal vertebrae and interbody fusion with bone, anterior and posterior non-instrumented fixation. Two cases had instrumentation (one case with a plate at primary surgery and posterior instrumented fixation with a hook system after four years in the second case). One patient died during epistatus in the early postoperative period. Two cases had improved neurologic status, and one with instrumented fixation had transitory paresis progression but then recovered during a year to the postoperative level. Each patient underwent from six to 15 orthopaedic procedures.

Tortori-Donati P.and coauthors (1999) [5] presented 10 patients aged from day one of birth to eight months. Nine had lower paraparesis of different severity, in one it appeared with age. Seven were operated, the rest were planned for surgery. Three patients underwent different variants of arthrodesis. Spinal cord decompression was needed in four cases as adjunct to arthrodesis. In one of these cases, a hemivertebra was removed and three had only posterior decompression. Upon completion of the listed treatments, stabilization without further deformity progression was achieved only in one case. The rest were planned for repeated interventions.

Zarzycki D.and coauthors (2002) [10] described their experience of treating nine patients with CVD type A. Six of them had lower paraparesis. All the patients were operated in the age from 1.8 to 5.5 years. Six of them first had different variants of spondylodesis in situ (anterior, posterior) that was effective only in one case. Therefore, all the patients underwent repeated surgeries that included either a partial or complete vertebroectomy of the abnormal vertebrae along and interbody spondylodesis with titanium cylindrical cages or bone. Patients wore rigid braces for 12 months. After all the stages of treatment undertaken, stabilization was obtained in five cases but the deformity continued progressing in four. Five patients out of six with paresis showed improvement of neurologic deficit after surgery. One that initially did not have neurologic changes developed paraplegia after surgery.

Mushkin A.Yu. and coauthors (2003) [11] presented a case of a 2-year-old patient with SSD in the lower thoracic spine that underwent a shortening vertebrotomy (kyphectomy), corporodesis and posterior instrumentation with two contractors. Satisfactory stabilization was achieved but no improvement in the neurologic status at follow-up examinations.

Ofiram E. and coauthors (2006) [12] described an interesting case of a successful management of a female patient with SSD Th12-L1 that had no neurologic symptoms and was operated at 3 years old with posterior spondylodesis in situ at Th10-L3 level. Post-surgery, immobilization with a hyperextensive brace continued for one year. She was followed up for 50 years, grew up to 146 cm and spontaneous reduction in kyphotic deformity from 75º to 33º happened while she grew, without paresis.

Bristol R.E. and coauthors (2006) [13] treated four SSD infant cases, all of them had lower paraparesis. One patient with cervical spine SSD started to wear neck brace after birth but died from cardiac insufficiency at 8 months' age. Abnormal vertebrae were removed in two newborns and rib grafts applied between the bodies and posterior non-instrumented spondylodesis was performed. The result was poor. The fourth newborn was treated conservatively with rigid bracing; deformity progressed and paresis turned into plegia. At the moment of publication, he had been planned for surgery.

Viehwege E. and coauthors (2009) [14] managed six patients with CVD (according to Dias criteria [4]). All patients had lower limb deformities and pelvic organs disorders. All wore initially rigid braces. Two patients died: one due to a concomitant somatic pathology (no surgery) and the other immediately after the intervention. Removal of abnormal vertebrae from the posterior approach of the VCR type, anterior corporodesis with a rib piece, posterior hook instrumentation with compression were performed in five patients aged from 10 to 34 months. One patient had three operations: posterior instrumental fixation with vertebroectomy as stage 1, anterocorporodesis with a rib graft as stage 2, and the third intervention was due to instrumentation failure. Post-surgery, patients wore braces for 6 months. Stabilization was achieved in five patients that remained alive.

Thus, the main problems of this pathology is a terminological diversity, absent unified opinion on the pathology, necessity of early diagnosis, choice of optimal age to perform surgeries in each case, multidisciplinary assessment of patient's condition, tactics of follow-up till " maturity'" for operation, volume and type of interventions in each case, periods and volume of immobilization after interventions.

METHODS

Seven patients, three boys and four girls, treated surgically by us between 2010 and 2015 were reviewed. Age upon admission ranged from one year to 4 years old.

RESULTS

Brief description of the results (clinical and radiographic findings as well as interventions is given in Tables 1, 2 and 3 (respectively) in which each patient is given a number from 1 to 7.

Table 1

Clinical characteristics of patients

Patient #	Sex/age (years)	Neurologic disorders according to Frenkel	Pelvic organ dysfunction	Associated anomalies
1	f/3	B	present	Lower limbs equinovarus, retardation in cognitive and psychic development, right-side radius aplasia, secondary dystopy of the left kidney, hypoplasia of ear auricle, retardation in psychomotor and physical development
2	m/3	E	none	Left lower limbs equinovarus
3	m/4	C	none	Foot equinovarus
4	f/1	D	present	Hypotrophy of the left lower limb and left foot equinovarus, bilateral ureterohydronephrosis, retardation in psychomotor and physical development
5	f/2	B	present	Left lower limbs equinovarus, finger joints contractures
6	m/1	A	present	Left lower limbs equinovarus
7	f/3	A	present	Left lower limbs equinovarus

Table 2

Spine and spinal cord anomalies

Patient #	Disorder level	Associated anomalies of vertebral development	Dysraphia	Rib anomalies	Deformity degree and laterization of the main curve	Spinal cord condition (MRI and intraoperative findings)
1	Lumbar	Butterfly vertebrae Th1-4	Sacrum	present	90° left	Hypoplasia at SSD level (L1-L3), hydromyelia
2	Lumbar	Butterfly vertebra L2, concrescence of bodies and arches Th2-6	Sacrum	none	60° right	Hypoplasia at SSD level (L3-L4), diastematomyelia
3	Upper lumbar	none	Sacrum	none	60° left	Hypoplasia at SSD level (Th12-L1)
4	Lower thoracic	Anterior atlas arc aplasia, antelisthesis CI, concrescence of some cervical, thoracia , lumbar vertebrae	Lower cervical	present	45° left	Hypoplasia at SSD level (Th11-Th12)
5	Lower lumbar	Butterfly vertebrae Th1-4 with concrescence of arches	Sacrum	present	90°	Hypoplasia at SSD level (L3-L4)
6	Upper lumbar	none	Sacrum	none	25° right	Hypoplasia at SSD level (Th12-L2)
7	Lower thoracic	none	Sacrum	none	80° left	Hypoplasia at SSD level (Th10-Th12)

Table 3

Surgical interventions characteristics

Patient #	Operation	Angle and correction	Complications	Repeated surgeries	Follow-up, years
1	TPF	30°, 50 %	Bedsores in screw area	1. Removal of construct 2. PSO+TPF (system change)	4
2	VSR, mesh, TPF	52°, 87 %	Two screws malposition, moderate paresis in left limb	Reinsertion of two screws	1
3	VSR, mesh, TPF	60°, 100 %	none	none	1
4	VSR, TPF	40°, 89 %	Break of left longitudinal bar	Instrumentation remounting (system change)	3
5	VSR, mesh, TPF	85°, 94 %	Instrumentation failure and risk of screw migration of the lower support block and mesh	Instrumentation remounting (system change)	3 (decompensation due to somatic condition, death after last hospitalization)
6	VSR, mesh, TPF	25°, 100 %	none	none	0.5
7	Bilateral fixation with costal-pelvic distractors VEPPR II and staged distraction up to 6 years	30°, 36 %	Kyphosis progression	1. VSR, UF mesh, TPF	5

Clinical examination

Kyphosis was present in all patients (**Fig. 1**). Three patients had skin stigmata, and namely, angiomas (cases 3 and 4) and a focus of hypertrichosis (case 1). None of the patients had open dysraphia, but all seven had spina bifida occulta at the sacral level. Signs of caudal regression syndrome were absent in all cases. Renal anomalies were found in two cases.

Fig. 1 A girl, 2 years old (case 5) with spinal segmental dysgenesis and severe lower paraparesis

Five patients had severe motor disorders, paraplegia in two cases and lower paraparesis in three, one mild paraparesis, and in one case there was no motor disturbance. Dysfunctions of the pelvic organs were identified in five cases. Equinovarus deformity of the legs was found in all cases.

Radiographic study

Combined anomalies in the development of vertebrae were found in all parts of the spine, only in three patients the defect was isolated. In all cases, the spinal cord was significantly hypoplastic. In one case, hydromyelia was found over the site of stenosis, and in another one type II diastematomyelia. In one patient, the spinal cord ended up above the anomaly site. The most common level of dysgenesis was the thoracolumbar transition, a sharp stenosis of the vertebral canal was observed at SSD site (**Fig. 2**). The rib anomalies were present practically in all patients with a failure of segmentation of the thoracic vertebrae. All patients showed nonconcrescence of the arches of the sacral vertebrae.

Spinal cord condition

The spinal cord was abnormal in all cases. It was confirmed by MRI and intraoperatively. At the level of kyphosis, the spinal cord was hypotrophic in all patients, and absent in one case (**Fig. 3**).

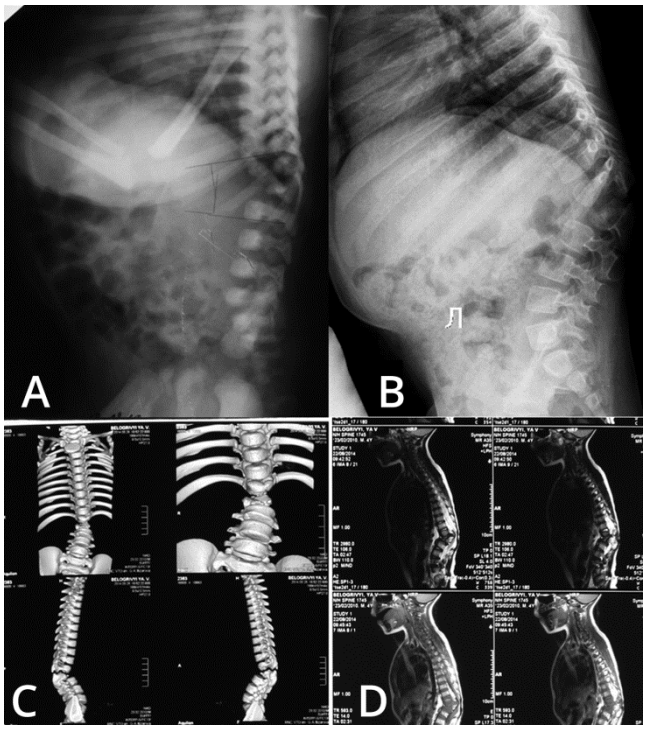

Fig. 2 Radiography, CT and MRI of the patient, 4 years 8 months old (case 3): **A** – lateral radiograph at the age of 1 month: kyphotic deformity, hypoplasia of vertebral bodies of the thoracolumbar junction; **B** – lateral radiograph at the age of 4 years 8 months: deformity progression, dislocation of the spine at the SSD level; **C** – 3D-reconstruction of the spine at the age of 4 years 8 months: hypoplasia of the vertebrae at the level of the thoracolumbar junction with absence of posterior vertebral structures and kyphotic deformity; **D** – sagittal slices of T2-weighted MRI at the age of 4 years 8 months show that the spinal cord ends at the level of the lower thoracic spine, and caudally to SSD the contours of the spinal cord and the dural sac with the cerebrospinal fluid are visualized. At kyphosis apex, there is total stenosis

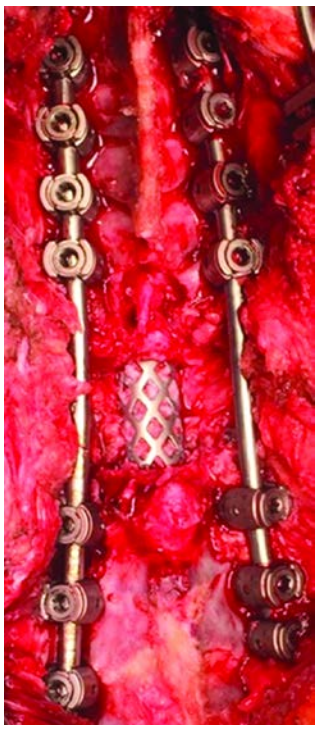

Fig. 3 Intraoperative photo of a 3-year-old patient (case 7) with SSD: spinal cord at the level of deformity is not differentiated

Surgical treatment

Five patients were subjected to extirpation of the rudiments of vertebral bodies with circular decompression of the dural sac at the level of dysgenesis and interbody spondylodesis with a mesh titanium implant and autologous bone chips from the dorsal access as the first stage, and a posterior interbody spondylodesis with a transpedicular system (**Fig. 4**). Three of them needed a re-operation due to instability of the system – system dismantling and its complete change in two cases.

One patient was managed by an isolated transpedicular fixation with the *Synaps* system. But in connection with the development of pressure ulcers in the area of the screw heads (**Fig. 5**), the system was removed and a corrective vertebrotomy followed at the apex of the deformity (Smith-Peterson type), then the transpedicular system *Medtronic Pediatric* was mounted. One patient underwent implantation of the VEPTR II system as the first stage in the age of 3 years, followed by gradual distraction for 3 years. When she was 6 years old, one of the authors of the article performed extirpation of the rudiments of the vertebral bodies at the level of dysgenesis with interbody spondylodesis by a mesh titanium implant and bone chips due to deformity progression along with posterior interbody spondylodesis with a transpedicular system.

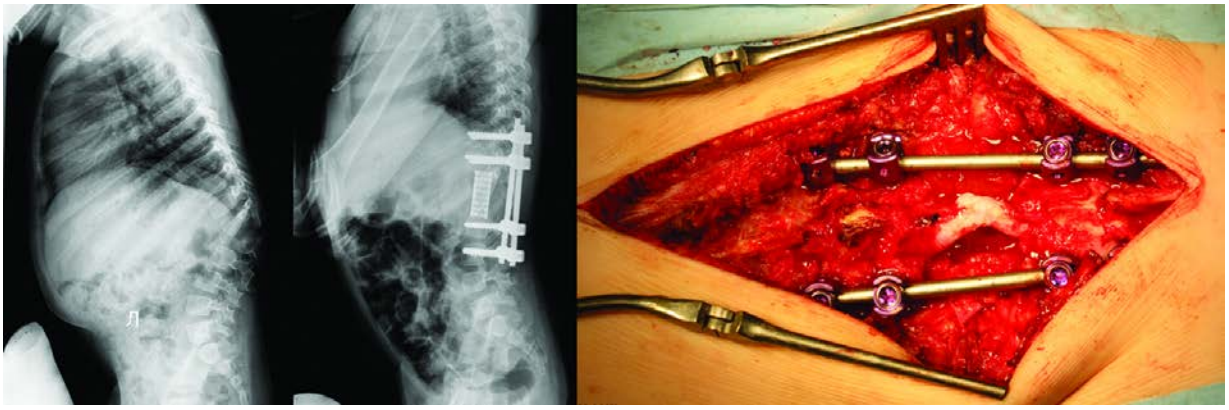

Fig. 4 Patient, 4 years and 8 months old (case 3), with SSD at the level of the thoracolumbar junction: lateral radiographs before and after the operation; intraoperative photo shows the site of the narrowed dural sac, the dura mater is rigid

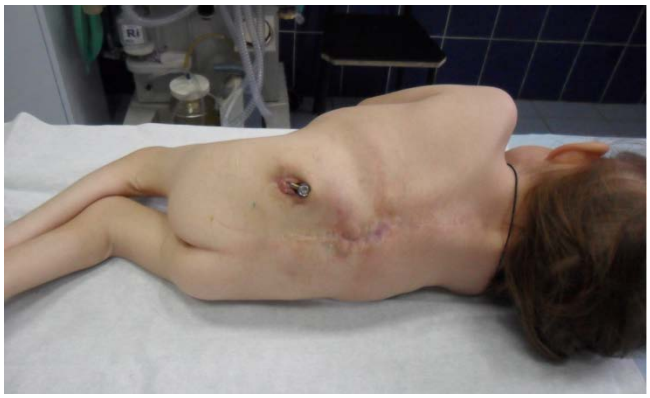

Fig. 5 Patient, 3 years old (case 1) - visualization of a bedsore in the area of the screw head on the left

DISCUSSION

Neuroradiological results

The neuroradiologic picture changes depending on the extent and level of pathology, as well as the presence of concomitant anomalies. Neuroradiological picture of SSD varies from hypoplasia of the vertebral body and posterior structures of one vertebra to the absence of several vertebrae, and always causes considerable instability and kyphosis which, sometimes, may not manifest itself immediately at birth but with the child's growth and motor activity.

MRI of the spine, a method of choice for visualization of the spinal cord, should always be performed in patients with SSD, regardless of the neurological pattern, to exclude the syndrome of the fixed spinal cord which can aggravate the progression of kyphoscoliosis and spinal instability. In some cases, the abnormal region of the spinal

cord may be absent. The segment of the spinal cord lower the SSD level is almost always thickened and unchanged. It is one of the unique signs of SSD; nevertheless, when it comes to the SSD in the lumbosacral area (with the sacrum present below the level of dysgenesis), the distal segment of the spinal cord may be absent due to the fact that the cone of the spinal cord is localized above (case 5). There are very few cases of a combination of open vertebral dysraphies in the literature, but such anomalies as spina bifida occulta, diastematomyelia, lipomas, fixed spinal cord syndrome are often described. Our results showed a close correlation between SSD and closed spinal dysraphy

Clinical correlation

Neurological and clinical findings at birth in children with SSD vary from the absence of any pathological changes and signs to the lower paraplegia (central or peripheral) with severe deformities of the lower extremities, function disorders of the pelvic organs.

Associated deformities of the lower limbs are common and are mainly equinovarus deformities of the legs with muscle contractures of varying severity. Several mechanisms are proposed for the development of neurological symptoms: (1) primary dysplasia of the spinal cord, nerve roots and vessels of the spinal cord at the level of malformation, (2) secondary myelopathy due to compression (chronic in most cases) of the spinal cord, nerve root and vascular structures, (3) combination of primary myelodysplasia and secondary myelopathy.

If a SSD patient does not have paresis, then there is an increased risk for developing a neurological deficit due to mechanical instability of the spine and congenital stenosis of the spinal canal.

Due to the presence of pelvic organs dysfunction in most patients, neurogenic bladder, in particular, they often suffer from chronic urologic infection. In addition, the most common of the accompanying anomalies in these patients are anomalies of kidneys and cardiovascular system. These conditions significantly affect the quality of life of these patients, so these anomalies should be sought in all patients with SSD.

Surgical treatment

There are still disputes about the optimal timing of spondylodesis. Some authors [2, 8] recommend that spondylodesis should be performed only when the diagnosis is confirmed, but they note that it is extremely difficult to achieve stabilization in one stage, especially in infants with immature bone structures. Therefore, multi-stage operations are necessary. Our strategy is to wait for maturation of the bone structures up to at least two years of age, when it is possible to perform a full-fledged fixation of the spine. The optimal tactics of surgical treatment in all SSD cases is circular decompression of the nerve structures with extirpation of the rudiments of the abnormal vertebrae from the dorsal approach, fixation of all three columns; in classical SSD: posterior instrumental fixation by the transpedicular system (as a rule – two or three levels above and below the SSD), corporodesis with a titanium mesh, with moderate compression of the transpedicular system to relieve the tension of the thinned dural sac. In some cases, a non-standard, individual approach is required. The operations must necessarily be performed under neurophysiological monitoring.

Postoperative care

On day 2 or 3 post-surgery, patients were put into a standing position wearing a semi-rigid thoracolumbar brace if they were able to stand or walk before. They were allowed to sit for a while after a week. A week after the operation, it was allowed to sit down for a short time with a gradual expansion of the regime.

CONCLUSIONS

SSD is not a single disease but a group of anomalies, from segmental hypoplasia of the spinal cord with an almost intact spine to a segmental absence of the spinal cord and spine (MSA type). The most optimal treatment for these patients, in our opinion, is “wait and see” tactics up to the age of 2 years, followed by excision of the vertebra rudiments, decompression of neural structures and stabilization of the spine by metal structure, without any dynamic components. It is necessary to remember about the possible concomitant pathology. Its disregard may make the work in the operating room useless, and namely, accompanying spinal cord anomalies, fixed spinal cord syndrome which can lead to spinal deformity progression and myelopathy. Kidney anomalies and infection, develop-

mental anomalies of the heart should be also taken into account.

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