

STUDY DESIGN: CASE SERIES OF 19 PATIENTS OPERATED FOR SEVERE SCOLIOSIS AND DIASTEMATOMYELIA

SÉRIE DE CASOS DE 19 PACIENTES OPERADOS PARA ESCOLIOSE SEVERA E DIASTEMATOMIELIA

DISEÑO DEL ESTUDIO: SERIE DE CASOS DE 19 PACIENTES OPERADOS POR ESCOLIOSIS GRAVE Y DIASTEMATOMIELIA

MIKHAIL VITALIEVICH MIKHAYLOVSKIY,¹ JEAN DUBOUSSET,¹ VJACHESLAV VICTOROVICH NOVIKOV,¹ ALEXANDER SERGEEVICH VASYURA,¹ INGA GENNADIEVNA UDALOVA,¹ MIKHAIL ANATOLIEVICH SADOVOI¹

1.Tsiv'yan Novosibirsk Research Institute of Traumatology and Orthopedics, Novosibirsk, Russia.

ABSTRACT

Objective: Diastematomyelia is a rare congenital spine and spinal cord malformation in which the spinal cord is divided into two parts by the osseous or fibrous septum. The incidence of diastematomyelia in patients with the most severe forms of congenital scoliosis is much higher than its general incidence in the population. When performing surgeries to correct scoliotic deformities, the question arises regarding the choice of a strategy for managing the septum. An unambiguous answer to this question does not exist, since the disease is very rare and heterogeneous. The aim was to summarize the data on different surgical strategies for detecting diastematomyelia. **Methods:** Literature review and retrospective analysis of our own clinical data. **Results:** We present our own experience of treating 19 patients with diastematomyelia and severe congenital scoliosis. Posture disorder was corrected in all cases; the septum was removed in none of the cases. Significant correction was achieved for all patients, and no neurological complications were observed in the short- and long-term follow-up. **Conclusions:** Surgical nonremoval of the spur enables compensation to be achieved, without neurological complications either in the immediate postoperative period or in the long-term (more than 2 years) follow-up. **Level of Evidence IV; Case series^h.**

Keywords: Diastematomyelia; Scoliosis; Surgery.

RESUMO

Objetivo: A diastematomielia é uma má formação rara da espinha dorsal e medula congênita, quando a medula espinhal é dividida em duas partes pelo septo ósseo ou fibroso. A incidência de diastematomielia em pacientes com formas mais graves de escoliose congênita é muito maior do que a incidência geral em uma população. Ao realizar cirurgias para corrigir deformidades escolióticas, surge a questão sobre a escolha de uma estratégia para o manuseio do septo. Uma resposta inequívoca à essa questão não existe, pois a doença é muito rara e heterogênea. O objetivo foi resumir os dados sobre diferentes estratégias cirúrgicas para detecção da diastematomielia. **Métodos:** Revisão de literatura e análise retrospectiva de nossos próprios dados clínicos. **Resultados:** Apresentamos nossa própria experiência no tratamento de 19 pacientes com diastematomielia e escoliose congênita grave. Transtorno de postura foi corrigido em todos os casos; em nenhum dos casos o septo foi removido. Correção significativa foi alcançada para todos os pacientes e nenhuma complicação neurológica foi observada a curto e longo prazo de acompanhamento. **Conclusões:** A não remoção cirúrgica do esporão permite obter uma compensação e ter a falta de complicações neurológicas, tanto no período pós-operatório imediato quanto a longo prazo (mais de 2 anos) de acompanhamento. **Nível de Evidência IV; Série de casos^h.**

Descritores: Diastematomielia; Escoliose; Cirurgia.

RESUMEN

Objetivo: La diastematomielia es una malformación congénita rara de la columna vertebral y la médula espinal en la cual la médula espinal se divide en dos partes por el tabique óseo o fibroso. La incidencia de diastematomielia en pacientes con las formas más graves de escoliosis congénita es mucho mayor que su incidencia general en una población. Cuando se realizan cirugías para corregir deformidades escolióticas, surge la pregunta con respecto a la elección de una estrategia para el manejo del tabique. No existe una respuesta inequívoca a esta pregunta, ya que la enfermedad es muy rara y heterogénea. El objetivo fue resumir los datos sobre diferentes estrategias quirúrgicas para la detección de diastematomielia. **Métodos:** Revisión de la literatura y análisis retrospectivo de nuestros propios datos clínicos. **Resultados:** Presentamos nuestra propia experiencia en el tratamiento de 19 pacientes con diastematomielia y escoliosis congénita grave. El trastorno postural fue corregido en todos los casos. El tabique no fue removido en ninguno de los casos. Se logró una corrección significativa en todos los pacientes y no se observaron complicaciones neurológicas en el seguimiento a corto y largo plazo. **Conclusiones:** La no remoción quirúrgica del esporón permite lograr una compensación sin complicaciones neurológicas, ya sea en el postoperatorio inmediato o en el seguimiento a largo plazo (más de 2 años). **Nivel de Evidencia IV; Serie de casos^h.**

Descriptor: Diastematomielia; Escoliosis; Cirugía.

Study conducted at Tsiv'yan Novosibirsk Research Institute of Traumatology and Orthopedics, Novosibirsk, Russia.
Correspondence: Mikhail V. Mikhaylovskiy. Frunze str., 17, Novosibirsk, 630091, Russia. MMikhailovsky@niito.ru

<http://dx.doi.org/10.1590/S1808-185120191801201849>



INTRODUCTION

Diastematomyelia, also known as split cord malformation (SCM), is a congenital anomaly of the spinal cord and spine in which the spinal cord is split into two parts. As opposed to diplomyelia (the so-called true duplication of the spinal cord), SCM is associated with splitting of the spinal cord into two halves by a bone, cartilage, or fibrous septum located in the sagittal or near-sagittal plane. Diastematomyelia is one of the variants of the closed form of spinal dysraphism.¹ SCM was first described by Olivier in 1837.² According to the literature, the rate of SCM in patients with congenital scoliosis is very variable and ranges from 4.9%³ to 41%.⁴

SCM has two aspects: neurosurgical and orthopedic. The neurosurgical aspect is that the risk for the development and progression of neurological symptoms due to traction on the spinal cord by the separating spur increases with the patient's age. The orthopedic aspect involves problems related to full surgical correction of spinal deformities in the presence of the spur separating the spinal cord.

In connection with these aspects, there are various opinions relating to the possible management of patients with diastematomyelia who undergo surgery. There are two dominant viewpoints. One is that the spur should be removed in all cases to avoid the development of severe neurological complications.³⁻⁹ The second suggests a differentiated approach, depending on patient's individual characteristics (e.g. implementation of this manipulation only for the type I (SCM I) anomaly or supposed slight correction of scoliosis).¹⁰⁻¹¹

According to the literature, the risk of neurological complications for resecting the spur is relatively high.^{5,9} However, spur resection is not always beneficial in terms of the extent of the correction. We therefore took the view that the approach of saving the spur during corrective surgery on the spine would be advantageous.

We have the experience of surgical treatment of 19 patients in whom the intraspinal spur was not resected before correction of congenital scoliosis, which had no adverse consequences for the patients.

METHODS

Three hundred fifty eight patients with congenital spinal deformities were treated at the NRITO clinic between 1997 and 2015. In 22 of the patients (6.1%), diastematomyelia was diagnosed during examination. Only 19 of them (13 males and 6 females, with a mean age 11.6 (6–19)) were included in the study group because two patients did not undergo corrective intervention, and one patient had previously been operated on for resection of the intracanal spur. The examination included plain radiographs of the spine in 2 standard upright projections, spondylograms in the lateral bending position, MRI, and MSCT. All the study participants signed an Informed Consent Form.

Scoliosis was diagnosed in 16 patients, and kyphosis was detected in 3 patients. Fourteen patients had bone spur, and 5 patients had fibrous spur. The spur was located in the thoracic spine in eight cases, in the thoracolumbar spine in five cases, and in the lumbar spine in six cases. The spur length varied from 2 to 8 segments and was not longer than three segments in most cases. A concomitant congenital pathology was found in seven patients: syringomyelia (2 cases), joint deformities of the upper and lower extremities (4 cases), Chiari malformation, keeled chest, and kidney duplication.

Preoperatively, 8 patients were neurologically intact, and 11 patients were detected as having neurological symptoms of varying severity: six patients with lower extremity paraparesis or monoparesis (with pelvic dysfunctions in two cases), one patient with pyramidal insufficiency syndrome, and four patients with hypotrophy and hypotension of the lower extremity muscles and lack of Achilles or abdominal reflexes.

The main aim of surgical treatment was to stop the progression of spinal deformity by corrective treatment, and we did not resect the spur in all 19 cases.

Various surgical instrumentations were used to correct the spinal deformity: segmental instrumentation with a hook or hybrid fixation was used in 14 cases; Antares was used in one case; VEPTR

instrumentation was used in four children under 10 years of age (the patients underwent 3, 4, 6, and 7 serial distractions). The spinal cord function was monitored using a wake-up test or evoked somatosensory potentials.

RESULTS

The mean Cobb angle of scoliotic deformities in 16 patients was 82.9° (51–170°). In three patients with kyphotic deformities, the Cobb angle was 10° (a 9-year-old patient with the kyphotic apex in the lumbar spine), 118°, and 160°. In the scoliosis patients, the deformity in the lateral bending position was 66.4° (40–105°), on average, i.e. spine mobility was 19.9%. Details of the characteristics of all patients are listed in Table 1.

In the immediate postoperative period, the mean scoliotic deformity was 59.6° (25–104°), i.e. the correction amounted to 23.3°, or 28.1% of the initial value (table 2). It was not possible to calculate the mean values in patients with kyphotic deformities because control radiography was not conducted in a patient with kyphosis of 118° due to a complication (early abscess). The normal sagittal contour of the lumbar spine was restored in a patient with lumbar kyphosis. Angular kyphosis in the third patient was reduced to 63°. The mean postoperative follow-up period was 32 (24–74) months. At that moment, the mean scoliotic deformity was 67° (31–101°), which means that the total loss of correction was 7.4°. The correction achieved was maintained in patients with angular kyphosis. The patient with lumbar kyphosis developed clinically significant proximal junctional kyphosis (PJK), which required two re-interventions. However, PJK recurred both times. In addition, two early deep abscesses developed, which required removal of an endocorrector in both cases.

No cases of new neurological symptoms or worsening of the preoperative symptoms were observed in the short or long-term postoperative periods.

In our view, our findings on the lack of new neurological symptoms over sufficiently long follow-up periods, in combination with significant correction of deformities, demonstrate that the tactic of saving the spur is an effective and safe approach to minimize risks of neurological symptoms associated with the corrective interventions.

DISCUSSION

Diastematomyelia is a rare disease. However, because of the pathological changes that accompany the disease, the problems associated with the diagnosis and surgical features of SCM are widely discussed in the scientific literature. The issue most actively debated is whether or not the spur should be removed.

We hypothesized that the conservative approach to resection/non-resection of the diastematomyelic spur, in cases without obvious neurological symptoms caused by the spur, would help minimize the risks.

The orthopedic approach to the problem of diastematomyelia is characterized by wide variability. There are three variants of the surgical approach to the patient with progressive congenital scoliosis who is diagnosed with diastematomyelia:

1. Resection of the intracanal spur in all cases, before the scoliosis correction surgery;
2. Resection of the intracanal spur without scoliosis correction;
3. Scoliosis correction without resecting the intracanal spur.

Group I is represented by the following studies

Winter et al.³ observed 27 patients with congenital scoliosis and diastematomyelia. Laminectomy and resection of the septum did not provoke worsening of neurological symptoms in 19 patients, in a 6-month follow-up period. The authors concluded that resection of the spur is important to prevent the progression of neurological deficit; therefore, corrective surgery should be delayed until the septum is removed.

McMaster⁴ diagnosed diastematomyelia in 41 (16%) out of 251 patients with congenital scoliosis. McMaster considered, as

Table 1. Baseline characteristics of patients.

Case number	Age (years)	Type of deformity	Side of deformity	Vertebral anomalies	Spinal cord anomalies	Length of the diastematomyelic bony septum
1	12.9	Scoliosis	D	Complete fusion of the bodies and arches of the T8, T9, T10, and T11 vertebrae. Spina bifida posterior at the 8, T11, T12, L1, L2, and L3 level	No	Complete diastematomyelic bony septum in the spinal canal at the T9-T10-T11 level
2	11.3	Scoliosis	D	Butterfly deformity of the D11 vertebral body; wedge-shaped deformity of the D10 and D12 vertebral bodies; concrescence of the D10-D12 articular processes on the left side, D11-D12 articular processes on the right side, and D10-D12 ribs on the left side; bilateral defect of the interarticular portion of the L4 vertebral arch; Spina bifida posterior at the L2-L5 level	No	Diastematomyelia at the level of lumbar enlargement and cauda equina
3	12.1	Kyphoscoliosis	d	Left wedge-shaped lateral T1 and T3 hemivertebrae; between the T5 and T6, T6 and T7 bodies; extra lateral hemivertebrae, hypoplastic intervertebral discs at the T1-2 - T7-8 levels. Spina bifida posterior of the T7 vertebra	No	A bony septum divides the canal into the right and left halves at the T5-T6 level
4	11.6	Scoliosis	D	Concrescence of the D10-L1 vertebral bodies combined with failure of segmentation. Spina bifida posterior at the D10-S3 level	No	Complete sagittal bony septum (19.4 mm thick in the ventral portion and 4.4 mm thick in the dorsal portion of the spinal cord)
5	13.1	Scoliosis	D	Butterfly deformity of the T7, T10, T12 vertebrae; T9 – right-sided wedge-shaped hemivertebra, spina bifida of T8, L2-S1	No	Widening of the spinal canal at the T12-L2 level; splitting of the lumbar enlargement and the medullary cone
6	9.2	Lordoscoliosis	d	Symmetric butterfly defect of the T3 and T5 vertebrae; nonsymmetric butterfly defect of the T7 and T8 vertebrae; spina bifida of the T1, T6-T10, and L5 vertebrae; spina bifida sacralis totalis	No	Osseous diastematic crest at the T3-T11 level
7	19.6	Lordoscoliosis	D	Butterfly defect of the T3, T4, T6, and T10 vertebrae; spina bifida posterior of the T1, T2, T6, T7, and T9 – T12 vertebrae	No	Sagittal bony septum in the spinal canal at the T6, T7 level
8	11.8	Scoliosis	S	Left-sided posterolateral wedge-shaped defect of the T5 vertebra; butterfly defect of the T7, T9-T11 vertebrae; concrescence of the arches of the T4-T6, T7-T10 vertebrae; multiple synostosis of the ribs on the right and left sides; spina bifida posterior of the C6-T3, T11-L1 vertebrae	No	Bony septum of the spinal canal at the apex of kyphosis (T8-T10) lying in the sagittal plane and separating the spinal canal into the right and left lateral portions
9	14.4	Kyphoscoliosis	d	Hypoplastic T4-5, 10-11, L3-4 vertebral discs; concrescence of spinous processes of the L2- L3 vertebrae, the 2 nd and 3 rd ribs on the left side; spina bifida at the T10-T12, L3-L4 level	No	Bony septum at the T9-T 10 level
10	18	Kyphoscoliosis	d	Hypoplastic T12-L1, L1-2 vertebral discs; concrescence of the posterior portions of vertebrae at the same level	No	Bony septum at the T10-T12 level
11	9.3	Kyphosis		Wedge-shaped posterior defect of the L2 hemivertebra, hypoplastic T12-L2 vertebral discs	No	Splitting of the medullary cone into two halves at the level of the L2 vertebral body
12	5	Scoliosis	D	Butterfly defect of the T3-T8 vertebrae; wedge-shaped hemivertebra between the T2 and T3 vertebrae on the left side; concrescence of the T2-T4, T7-T10 vertebrae at the level of their arches; hypoplastic T3-4, T5-6, and T6-7 intervertebral discs; spina bifida posterior of the T5, T6, T10-T12 vertebrae	No	Bony septum at the T7-T8 level dividing the spinal canal into the right and left halves
13	9	Kyphoscoliosis	d	Butterfly defect of the T11 vertebra with the right half of the vertebral body predominating; the left half of the body is fused with the L1 vertebral body; L2 is the right-sided lateral wedge-shaped hemivertebra; concrescence of the bodies of the T12 and L3 vertebrae. Hypoplastic T12-L1 – L2-3 discs. Spina bifida posterior of the T12 - S2 vertebrae. Left-sided concrescence of the ribs	A CSF-filled cyst (10. mm in diameter and 36.0 mm long) is detected at the level of lumbar enlargement of the spinal cord. Two terminal filaments linked by a commissure at the L4 level are detected caudally.	Partial bony septum at the level of L2 and L3 vertebral bodies (after subtotal resection of the septum) protruding into the lumen by 9 mm, 23 mm long, 12 mm thick
14	8	Kyphoscoliosis	d	Butterfly T8 and T10 vertebra; concrescence of the bodies of the T7-T10 vertebrae; aplasia of left hemiarcs of the T8-T10 vertebrae; L2-L4 vertebral bodies are fused, with their arches separated and a wide diastasis present; concrescence of right hemiarcs at the T12, L1 level; multiple rib abnormalities	Syringomyelic cavity (16 mm x 6.7 mm) with smooth walls at the T9-T10 level; diastematomyelia combined with syringomyelia of every vertebral column starting from the T11-T12 intervertebral disc to the S1 spinal unit.	There is a wide continuous sagittal bony septum in the spinal canal at the L2, L3 level, which separates the dural sac into two unequal halves
15	7	Kyphoscoliosis	s	Left-sided lateral T10 hemivertebra fused with the T11 vertebral body; multiple rib abnormalities on the right side	Syringomyelia at the cervical, thoracic, and lumbar levels. Condition after drainage of the syringomyelic cavity at the C3-5 level. Condition after emptying and draining the cystic tumor in the thoracolumbar spine; trophic lesions of the postoperative cicatrix. Syringomyelic cavities are also visualized in both vertebral columns below the T11 level.	There is a 6 mm thick bony septum in the sagittal plane in the spinal canal at the T11-T12 level
16	12	Scoliosis	D	Concrescence of the T1-2, L5-S1 vertebrae; spina bifida posterior at the T4, L5 level; synostosis of the 1 st -3 rd ribs on the right side	Fixed spinal cord, syringomyelia at the T5-7 level.	Diastematomyelia at the L1, L3 level

Table 2. Main outcomes on patients.

Case number	Cobb angle		Type of surgery	Neurological symptoms prior to surgery	Neurological symptoms after surgery
	preoperative	postoperative			
1	85°	46°	Skeletal traction, correction by segmental spinal instrumentation	No abnormalities	No changes
2	75°	38°	Antares	No abnormalities	No changes
3	100°	76°	Skeletal traction, correction by segmental spinal instrumentation	No abnormalities	No changes
4	68°	65°	Skeletal traction, correction by segmental spinal instrumentation	No abnormalities	No changes
5	70°	46°	Skeletal traction, correction by segmental spinal instrumentation	Mild peripheral paraparesis of the lower limbs without pelvic organ dysfunction	No changes
6	73°	46°	Skeletal traction, correction by segmental spinal instrumentation	No abnormalities	No changes
7	117°	94°	Skeletal traction, correction by segmental spinal instrumentation	No abnormalities	No pathology
8	114°	101°	Correction by segmental spinal instrumentation	Pyramidal insufficiency syndrome	No changes
9	85°	51°	Skeletal traction, correction by segmental spinal instrumentation	No abnormalities	No changes
10	75°	27°	Skeletal traction, correction by segmental spinal instrumentation	No abnormalities	No changes
11	10°	63°	Skeletal traction, correction by segmental spinal instrumentation	No abnormalities	No changes
12	51°	36°	Multi-stage correction using VEPTR instrumentation (5 stages)	No abnormalities	No changes
13	103°	92°	Multi-stage correction using VEPTR instrumentation (4 stages)	No abnormalities	No changes
14	146°	102°	Multi-stage correction using VEPTR instrumentation (the first stage)	Mild combined paraparesis without pelvic organ dysfunction	No changes
15	105°	77°	Multi-stage correction using VEPTR instrumentation (the first stage)	Lumbar myelopathy presenting as flaccid paraparesis and pelvic organ dysfunction	No changes
16	60°	9°	Skeletal traction, correction by segmental spinal instrumentation	Thoracolumbar myelopathy presenting as mild left-sided monoparesis of a lower limb	No changes

indications for surgical treatment in these patients, preventing the progression of neurological disorders and preventing neurological complications. More than 20 patients underwent preventive removal of the spur without changes in neurological status, achieving very significant correction (from 66° to 14°). Only one neurological complication was observed by the author, in a patient with severe deformities who underwent a massive two-stage intervention with resection of the spur. The motor and sensory functions almost completely recovered with time. The author is convinced that the intraspinal anomaly should be removed in any patient with congenital scoliosis, regardless of the neurological status.

Wenpeng Liu et al.¹² followed-up a group of 48 patients. SCM I was detected in 47 of them. Laminectomy and spur resection were performed in all patients before a corrective intervention. The authors did not report the results and complications but recommended a similar approach in all cases of congenital scoliosis and diastematomyelia because, in their opinion, the intraspinal spur might contribute to the progression of congenital scoliosis.

Ayvaz et al.¹³ operated on 32 patients with diastematomyelia and congenital scoliosis. All patients with SCM I (18 people) underwent

resection of the spur before the corrective intervention. Only orthopedic surgery (deformity correction and spinal fusion) was performed in SCM II. Correction of scoliosis amounted to 44–47%. Temporary neurological symptoms were detected in two cases of SCM I; no other complications occurred. The authors recommend using this approach for type I and II diastematomyelia.

Hui et al.¹⁴ demonstrated a similar approach to solve the problem. They operated on 45 patients (15 patients with SCM I and 30 patients with SCM II). All the operations were one-stage surgery, but the spur was resected only in patients with type I anomalies. The deformity was corrected from 73.7° to 33.5° (54.5% of correction). All complications (two cases of mild neurological symptoms and one case of liquorrhea) were corrected. The authors believe that spur resection before correction of scoliosis is indicated for SCM I but not for SCM II.

This differentiated approach to types I and II diastematomyelia associated with congenital scoliosis was also demonstrated by Lifeng Lao et al.¹⁵ They operated on five patients. The spine deformity was decreased from 63° to 30.2° (57.2% of correction). No complications were observed. Two patients had a partial regression

of preoperative neurological symptoms. The authors recommend a single-stage intervention, with resection of the spur only in the case of SCM I.

Sheng-Li Huang et al.¹⁶ reported on 156 patients operated on for resection of the spur. One patient died due to anesthesia complications (anesthetic accident). Liquorrhea developed in 2 cases, transient neurological symptoms occurred in 4 cases, and epidural hematoma occurred in one patient. There were no persistent neurological complications. The authors suggest that surgery be indicated for all patients with SCM I and neurological symptoms or tethered filum, while conservative treatment be indicated for those with SCM II who are asymptomatic for SCM I.

Group II includes the following studies

Pang et al.^{17,18} who suggested the type I and II diastematomyelia classification, depending on the anatomical features, operated on 39 patients and observed an improvement or stabilization of neurological symptoms in 89% of cases.

Ersahin et al.¹⁹ operated on 74 patients. Temporary complications (cerebrospinal fluid fistula, monoparesis,⁶ dysesthesia, wound abscess, urinary retention), which were corrected within 3 weeks, were observed in 17 cases. Persistent complications occurred in three patients (renal failure, spinal arachnoiditis).

Schijman⁸ performed 17 operations and achieved control of negative neurological changes in 13 patients (76.4%). Of 4 patients who were neurologically intact before surgery, the condition remained unchanged in three patients, and symptoms completely disappeared (after temporary worsening) in one patient.

Massive clinical data are presented by a group of neurosurgeons from New Delhi,^{5,20,21} which allowed them to make a significant addition to Pang's classification, by allocating four anatomic subtypes within the SCM I that characterize spatial relationships between the spur and halves of the dural sac. Of 254 patients who underwent spur resection, 68 patients had a partial regression of neurological symptoms; the neurological status remained unchanged in 160 patients; worsening occurred in 15 cases. In addition, 23 patients had liquorrhea, and 6 patients had abscesses. The vast majority of complications were corrected. In a separate paper, the same authors²² analyzed the treatment outcomes in four patients with the dorsal localization of the intraspinal spur. There was one conclusion in all cases: regardless of the symptoms and diastematomyelia type, the spur should be resected.

E.V. Ulrikh,⁹ in his monograph, presented the results of surgical treatment of 87 patients. The intraspinal spur was removed in all cases to prevent aggravation or development of neurological symptoms. An improvement in the neurological status was observed in 15 patients (17.3%); worsening of the neurological status was observed in 16 (18.3%) patients, with two of them developing plegia (with partial or full recovery in six cases); the clinical symptoms remained unchanged in 55 (63.2%) patients. One child died due to productive meningoencephalitis.

Borcek et al.²³ reported 34 operated patients with both types of diastematomyelia. An improvement in neurological status was observed in 16 (47%) patients; worsening of the neurological status was observed in four (11.6%) patients; there were no changes in 14 cases.

Only a few authors have demonstrated an ambiguous surgical approach to prevent progression of the pathological process. Cheng et al.²⁴ analyzed the results of surgery in 112 patients and concluded that the surgery was much more effective in the case of SCM I. Huang et al.¹⁶ operated on 156 patients and concluded that surgery should be performed in

patients with SCM I in the presence of progressive neurological symptoms. Conservative treatment is only required in patients with asymptomatic SCM I and SCM II.

Sinha et al.⁵ reported a large group of patients (n=203). Motor deficit (weakness and atrophy of limb muscles, gait disorders) was observed in 148 (72.9%) patients, sensory disorders (dysesthesia, hypoesthesia, trophic ulcers, and finger autoamputation) were found in 80 (30.4%) patients. Pelvic organ dysfunction was detected in 66 (32.5%) patients; no neurological symptoms were observed in only 27 out of 203 patients. Among the patients with preoperative neurological symptoms, 60 (40.4%) reported improvement in motor function; 33 (41.2%) patients, improvement in sensory function; 20 (30.3%) patients, improvement in pelvic organ function, and 13 (52%) patients, reduced degree of trophic disorders. Fifteen patients reported aggravation of neurological symptoms immediately after the intervention; no recovery was observed in three of these patients.

Group III includes the following studies

Guixing and Jiaming¹⁰ reported that out of 500 patients with congenital scoliosis, 95 had with anomalies of the spinal cord. Surgical treatment did not involve resection of the diastematomyelic spur. Initial neurological symptoms were found in 41 patients, but no postoperative worsening of the symptoms was observed in any case.

Bollini et al.¹¹ believe there is no need to resect the spur if the expected correction of scoliosis is "not so significant".

On the basis of the literature data, we conclude that in patients with congenital scoliosis and diastematomyelia, the risk of neurological complications associated with resection of the bone spur is higher than that associated with corrective treatment of the spinal deformity. In most of the 19 patients of our group, the spinal deformity can be assigned to the neglected category; should be changed to "but in all cases we decided not to perform preparatory resection of the diastematomyelic spur. As the main task of surgical treatment, we aimed to stop the progression of spinal deformity. That is why the above correction was relatively small and slightly exceeded the values of preoperative spine mobility, which was determined using spondylograms in the lateral bending position as 23.3°.

CONCLUSION

There is currently a wide range of surgical approaches for diastematomyelia, with no conventional surgical approach, due to the lack of clinical data and in-depth analysis of the issue. Often, the decisions on the choice of approaches were local, based on the personal experience of the researchers. For this reason, the data obtained are highly heterogeneous and difficult to analyze systematically. We believe that further accumulation of clinical data and the development of a differentiated approach to the choice of a surgical approach are required.

Our study included the relatively small number of cases which were heterogeneous by the nature of spine deformity and by duration and severity of disease. However, the chosen surgical approach demonstrates the potential for application in surgical practice. This is confirmed by our high levels of compensation achieved and the lack of neurological complications, both in the immediate postoperative period and in long-term (more than 2-year) follow-up.

All authors declare no potential conflict of interest related to this article.

CONTRIBUTION OF THE AUTHORS: Each author made significant individual contributions to this manuscript. MM and JD were the main contributors in the drafting of the manuscript. MVM (0000-0002-4847-100X)*, VVN (0000-0002-9130-1081)*, ASV (0000-0002-2473-3140)* and IGU (0000-0001-8914-3451)* performed the surgical procedures, followed up the patients, and gathered clinical data. MVM and MAS (0000-0002-0902-0396)* evaluated the data from the statistical analysis. MVM, JD (0000-0002-4000-5450)*, VVN, ASV, IGU and MAS performed the literature search and review of the manuscript, and contributed to the intellectual concept of the study. *ORCID (Open Researcher and Contributor ID).

REFERENCES

1. Tortori-Donati P, Rossi A, Cama A. Spinal dysraphism: a review of neuroradiological features with embryological correlations and proposal for a new classification. *Neuroradiology*. 2000;42(7):471-91.
2. Ollivier CP. *Triate des maladies de la moelle epiniere*, 3rd ed. Paris: J.B. Bailliere; 1837.
3. Winter RB, Haven JJ, Moe JH, Lagaard SM. Diastematomyelia and congenital spinal deformities. *J Bone Joint Surg Am*. 1974;56(1):27-39.
4. McMaster M. Occult intraspinal anomalies and congenital scoliosis. *J Bone Joint Surg Am*. 1984;66(4):588-601.
5. Sinha S, Agarwal D, Mahapatra AK. Split cord malformations: an experience of 203 cases. *Childs Nerv Syst*. 2006;22(1):3-7.
6. Uzumcugil A, Cil A, Yazici M, Acaroglu E, Alanay A, Akalan N, et al. The efficacy of posterior hemiepiphysiodesis in patients with iatrogenic posterior element deficiency resulting from diastematomyelia excision. *Spine (Phila Pa 1976)*. 2003;28(8):799-805.
7. Hood R, Riseborough EJ, Nehme AM, Micheli LJ, Strand RD, Neuhauser EB. Diastematomyelia and structural spinal deformities. *J Bone Joint Surg Am*. 1980;62(4):520-8.
8. Schijman E. Split spinal cord malformations: report of 22 cases and review of the literature. *Childs Nerv Syst*. 2003;19(2):96-103.
9. Ulrikh E. Split cord syndrome (diastematomyelia). Saint-Petersburg: Synthes Book Ed, 2012.
10. Guixing Q, Jiaming L. A prospective study of one-stage surgery of congenital scoliosis with split cord malformation. *European Spine Journal*. 2011;20(Suppl 4):421.
11. Bollini G, Cottalorda J, Juove J, Labrier C, Choux M. Closed Spinal Dysraphism. *Ann Pediatr (Paris)*. 1983;40(4):197-210.
12. Liu W, Zheng D, Cui S, Zhang C, Liu Y, Jia Y, et al. Characteristics of osseous septum of split cord malformation in patients presenting with scoliosis: a retrospective study of 48 cases. *Pediatr Neurosurg*. 2009;45(5):350-3.
13. Ayvaz M, Akalan N, Yazici M, Alanay A, Acaroglu RE. Is it necessary to operate all split cord malformations before corrective surgery for patients with congenital deformities? *Spine*. 2009;34(22):2413-8.
14. Hui H, Tao HR, Jiang XF, Fan HB, Yan M, Luo ZJ. Safety and efficacy of 1-stage surgical treatment of congenital spine deformity associated with split spinal cord malformation. *Spine*. 2012;37(25):2104-13.
15. Lao L, Zhong G, Li Z, Liu Z. Split spinal cord malformation: report of 5 cases in a single Chinese center and review of the literature. *Pediatr Neurosurg*. 2013;49(2):69-74.
16. Huang SL, He XJ, Wang KZ, Lan BS. Diastematomyelia A 35-Year Experience. *Spine*. 2013;38(6):E344-49.
17. Pang D, Dias MS, Ahab-Barmada M. Split cord malformations: Part I: A unified theory of embryogenesis for double spinal cord malformation. *Neurosurgery*. 1992;31(3):451-80.
18. Pang D. Split Cord Malformations: Part II: Clinical Syndrome. *Neurosurgery*. 1992;31(3):481-500.
19. Ersahin Y, Mutluer S, Kocaman S, Demirtas E. Split spinal cord malformation in children. *J Neurosurg*. 1998;88(1):57-65.
20. Mahapatra AK, Gupta DK. Split cord malformation: a clinical study of 254 patients and a proposal for a new clinical-imaging classification. *J Neurosurg*. 2005;103(6 Suppl):531-6.
21. Gupta D, Mahapatra A. Proposal for a new clinicoradiological classification of type I split-cord malformation: a prospective study of 25 cases. *Pediatr Neurosurg*. 2006;42(6):341-6.
22. Prasad GL, Borkar SA, Satyathee GD, Mahapatra AK. Split cord malformation with dorsally located bony spur: report of four cases and review of literature. *J Pediatr Neurosci*. 2012;7(3):167-70.
23. Borcek AO, Ocal O, Emmez H, Zinnuroglu M, Baykaner MK. Split cord malformation: experience from a tertiary referral center. *Pediatr Neurosurg*. 2012;48(5):291-8.
24. Cheng B, Li FT, Lin L. Diastematomyelia: a retrospective review of 138 patients. *J Bone Joint Surg Br*. 2012;94(3):363-72.