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## Original research article

# Surgical management of 142 cases of split cord malformations associated with osseous divide



AND NEUROSURGERY

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#### ABSTRACT

*Objectives*: To investigate the key surgical points in treating split cord malformations associated with osseous divide and scoliosis (SCM-OD-S).

Materials and methods: The surgical options and methods of a total of 142 SCM-OD-S cases were retrospectively analyzed, and the surgical precautions and imaging diagnosis were also discussed.

Results: The 142 patients were performed osseous divide resection plus dural sac molding, which achieved good results and no serious complication such as spinal cord and nerve injury occurred; certain symptoms such as urination-defection disorders, muscle strength subsidence, Pes Cavus, and toe movement disorder in partial patients achieved various degrees of relief, and it also created good conditions for next-step treatment against scoliosis.

Conclusions: The diagnosis of SCM-OD mainly depended on imaging inspection, routine magnetic resonance imaging (MRI) combined with computed tomography (CT) 3D reconstruction, which can comprehensively evaluate the types and features of diastematomyelia as well as other concomitant diseases. SCM alone needed no treatment, but surgery will be the only means of treating SCM-OD. Intraoperatively removing osseous divide step-by-step, as well as carefully freeing the spinal cord and remodeling the dural sac, can lay good foundations for relieving tethered cord, improving neurological symptoms, and further scoliosis orthomorphia, thus particularly exhibiting importance for the growth and development of adolescents.

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#### 1. Introduction

Split cord malformations (SCM) is a congenital spinal cord malformation characterized by segmental osseous divide of spinal cord tissue in the sagittal plane, is more common in children and adolescents, and mainly occurs in the thoracolumbar segment, as well as often being accompanied by such deformities as back skin abnormalities, spina bifida, myelocele (meningocele), Tethered cord, or scoliosis, among which scoliosis is the most common [1,2]. The biggest threat of

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SCM comes from osseous divide caused rigid segmentation of spinal cord, which makes the patients suffer from sustained spinal cord traction and oppression after birth, followed by spinal cord growth restriction and gradual aggravation of clinical symptoms. Surgery is still the only treatment method, especially for the patients with split cord malformations associated with scoliosis (SCM-S). Initial-stage surgery can remove osseous divide and artificially remodel the dural sac, which not only clears the way for late-stage orthomorphia but also can avoid post-orthopedic traction injury of spinal cord tissue, so it is particularly important for adolescents in their growing period [3].

In recent years, with the development of imaging inspection methods, the understanding of this disease has achieved great progresses. Meanwhile, the application of micro-surgery and the further development of the concept of minimally invasive treatment provide important guarantees for improving surgical outcomes and reducing postoperative complications. However, due to lacking the orientation signs of the nervous system in early stages, as well as lacking comprehensive and systemic laboratory inspections, clinical misdiagnosis and missed-diagnosis are common. At the same time, the causes and nerve damage mechanisms of SCM still lack detailed understanding, and the current treatment opinions about whether SCM should be treated earlier than scoliosis orthomorphia are still controversial [4,5]. Furthermore, surgical methods toward split cord malformations associated with osseous divide (SCM-OD) are diverse and difficult. Therefore, clinical classification, diagnosis, clinical pathological features of SCM, as well as diagnosis, treatment, and prognosis of SCM-OD, still need further standardization and improvements.

Our department has treated a total of 3850 patients with congenital scoliosis from April 2000 to May 2014, including 189 SCM cases, among who 142 cases are formed by SCM combined with intra-spinal OD. Therefore, we performed this retrospective clinical study, hoping to provide theoretical basis and practical experience for the surgical purposes, indications, strategies, postoperative complication prevention, and longterm surgical efficacy evaluation of such disease based on our advantages of having sufficient cases, as well as complete clinical data and follow-up information.

#### 2. Materials and methods

#### 2.1. Clinical data

Patient selection: This study had collected a total of 142 patients underwent SCM-OD resection in the department of neurosurgery in our hospital between April 2000 and May 2014. All the patients had complete imaging and clinical follow-up data, including 28 males and 114 females (M:F = 1:4), aging 3–36 years, with the median age as 13.4 years. The intraspinal crests in all the patients were in the thoracolumbar segment, mainly 1–2 cm in length; furthermore, 5 patients had two simultaneous discontinuous crest. At the same time, such data as the nerve function improvement and long-term scoliosis improvement degree after OD resection were completely recorded (Table 1).

Clinical symptoms: Among the 142 patients, 97 patients were combined with low spinal cord and filum terminale

# Table 1 – Clinic pathologic features of 142 patients with split cord malformations.

Clinical case characteristics	Cases (%)
Age	
<14	108 (76.1)
≥14	34 (23.9)
Gender	
Male	28 (19.7)
Female	114 (80.3)
Longitudinal interval number	
≤1	137 (96.5)
>1	5 (3.5)
Longitudinal interval position	
Cervical vertebra	0 (0)
Thoracic vertebra	90 (63.4)
Lumbar vertebra	52 (36.6)
Sacral vertebrae	0 (0)
Longitudinal interval length	
≤1 section	136 (95.8)
>2 sections	6 (4.2)
Combined lesion	
None	7 (4.9)
Thickening of low spinal cord	97 (68.3)
Spinal fracture/Meningocele	62 (43.7)
Syringomyelia	56 (39.4)
Sacral cyst	41 (28.9)
Intraspinal tumor	14 (9.9)
Nerve injury symptoms	
No	35 (24.6)
Yes	107 (75.4)

thickening, 62 patients were combined with segmental syringomyelia and congenital intraspinal tumor (including bronchogenic cysts, intestinal cysts, or lipoma), 70 patients were combined with unilateral lower limb muscle atrophy, muscle weakness, or foot deformity, partial patients were combined with mild urination or defecation disorder, and 42 patients were combined with thoracic deformity and moderate – severe pulmonary dysfunction.

#### 2.2. Surgical indications

OD can cause the Tethered cord syndrome, such as urination or defecation disorder, bi-lower extremity weakness, or talipes equinus. Because the patients are in the growth and development period, so compression of OD toward the spinal cord will limit the activity of the spinal cord, thus producing the Tethered cord syndrome toward the children in their growth and development period. Although no symptom may appear, scoliosis dose exist and needs orthopedic surgery, which may longitudinally extend the spinal column and cord.

#### 2.3. Surgical methods

Preoperative localization: CT-guided positioning was mainly used; each patient was placed in the prone or lateral side on the CT bed (with the convex side of scoliosis downwards when in the lateral position). According to the cursor position, the needle was guided to stab the spinous process or the spinal lamina if the spinous process was absent, certain amount of methylthioninium chloride was injected onto the surface of the positioned spinous process to complete the preoperative localization. Anesthesia: Intubation under general anesthesia was selected.

Position: Each patient was generally placed in the prone or lateral position, with OD vertical to the operating bed so as to facilitate intraoperative operations.

Resection of spinous process and spinal lamina: Expose the OD as well as the upper and lower spinal canals based on the center of preoperatively localized point. According to the size, hardness and direction difference of OD, appropriate rongeurs, needle holders, and arc bone chisels were used to resect OD, or high-speed grinder can also be used, during which period, wide brain spatula can be used to block the dura mater and the spinal cord so as to prevent the spinal cord injury caused by the sliding of the grinder.

Dural sac molding: The edge of ventral SDM was intermittently sutured, the dorsal SDM was performed continuous suture so as to make the furcal SDM form one cavity; the patients accompanied with tethered cord or filum terminale thickening were performed filum terminale cut off microscopically to lift tethered cord. The patients coexisting syringomyelia or congenital intra-spinal tumor or cyst were surgically treated accordingly.

#### 2.4. Neuroelectrophysiological monitoring

Partial patients combined with complex spinal deformities were continuously monitored the free electromyography (EMG) of lower limb muscles, as well as intermittently monitoring the somatosensory evoked potential (SEP) and motor evoked potential (MEP) of lower limbs. In the process of spinal cord dissection, suspicious adhesive nerve tissues were actively stimulated so as to judge whether spinal cord and nerve injury occurred during the operation, and to avoid injuring important nerves such as the pudendal nerve.

#### Results

#### 3.1. General shape and distribution of OD

Through recording the anterior vertebral segments of each bone crest and rotating their angles, the standard sagittal plane of the crest was obtained for the analysis. All the OD cases were located in the thoracolumbar segment, including 20 cases in the upper thoracic segment (14.1%), 33 cases in the middle thoracic segment (23.2%), 37 cases in the lower thoracic segment (26.1%), 39 cases in the upper lumbar segment (27.5%), and 13 cases in the lower lumbar segment (9.1%). The sizes and shapes of OD varied. 136 cases had the length of the crest less than 1 segment (95.8%), while 6 cases had the length more than 1 segment (4.2%), among who the longest case involved 4 segments. There were 27 cases of bifurcated crest (19.0%), 155 cases of non-bifurcated crest, 67 column cases, 29 lamellar cases, 10 acicular cases, and 9 irregular cases.

#### 3.2. Surgical efficacy

The 42 patients with intraoperative neuroelectrophysiological monitoring showed no significant change of SEP or SEP + MEP. The 97 patients combined with Tethered cord or filum

terminale thickening were performed simultaneous filum terminale incision, among who 3 patients exhibited lateral lower limb muscle strength decrease, while recovered 3 months. Some patients appeared numbness or pain on the surgery side, while more alleviated within 1-2 weeks. The preoperative stool dry symptoms in 28 cases were relieved, and the 12 patients with poor lateral toe motions and difficulty in wearing shoes exhibited significantly improved fine motor functions. 117 patients were followed up for 11-80 months with a median follow-up period as 35 months. No new neurological damage appeared during follow-up. All the 142 patients were performed postoperative CT review and confirmed the complete resection of OD, therefore, the compression on the spinal cord was relieved. 109 patients were performed scoliosis correction 2-6 months later, and the correction degrees were greater than 30°.

#### 4. Discussion

SCM was firstly discovered and named by Ollivie in 1837, which is caused by the compression and fixation of the spinal cord by bony, fibrous, or cartilaginous intervals, and because the growth rate of spinal bone is faster than that of the spinal cord, so the rising of the spinal cord is blocked by the intervals and pulled by the filum terminale, thus causing a series of neurological disorders and deformities [6,7]. The pathogenesis of SCM is still unclear. Some scholars believe that its onset has autosomal recessive hereditary; Sepulveda et al. [8] once reported cases with SCM among direct relatives, and believed that such exogenous factors in pregnancy as radioactive radiation and viral infection may also cause SCM. Presently, most scholars more agree with the theory proposed by Pang et al. [9] that SCM originates from abnormal developments of neural tube closure in the embryonic period, which forms one endodermal canal with split notochord and nerves, thus leading to the appearance of two neural tube.

According to the shape of the dural tube and its relationship with the spinal cord, scholars divide SCM into dual-and singletube type, the former type refers to that the dural tube independently splits into two sub-tubes and develop along the spinal cord, while the latter refers to that the dural tube in lesions expands and exhibits one single tube, in which two split spinal cords are contained. Patients with dual-tube type, especially those with scoliosis, exhibit the spinal cord as asymmetric division, and the development differences in the both sides of spinal cord are huge. The spinal cord also exhibits obvious tension. Therefore, the duab-tube type is the focus of surgical treatment of SCM. All the patients in this group belonged to the dual-tube type. In addition, OD not always lies in the middle of the spinal canal, and the segmentation of the spinal canal can be asymmetric, so the spinal cord in the narrow side is also smaller, which can also cause semi-spinal canal stenosis. All the patients in this study with transient unilateral lower limb paralysis had lateral spinal canal stenosis.

SCM is commonly seen in children, and female patients account for about 70% of all cases; the preferred site is in the thoracolumbar spine, consistent with the results of this study. The main clinical manifestations include dorsal skin wrinkles and pigmentation, asymmetric development of lower limbs accompanied by pes areuatus, equinus foot, or toe atrophy, as well as pain below the SCM plane; certain neurological symptoms may include unilateral lower limb muscle weakness, muscle atrophy, numbness, sensory loss. and sphincter dysfunction below the lesion level [10,11]. Meanwhile, since SCM affects embryonic developments, so the occurrence of scoliosis has certain correlations, and it is reported that about 60–79% SCM patients are combined with scoliosis. Master [12] retrospectively analyzed 251 patients with scoliosis, and found 18.3% of the patients had SCM. According to the cases in our hospital, SCM accounted for about 15% of the total scoliosis cases, those with SCM-OD accounted for about 9% of the total scoliosis cases, and the patients with two or multiple OD accounted for about 2% of all the OD patients.

In recent years, with the extensive developments of scoliosis correction and imaging techniques, the detection rate of scoliosis combined with SCM has been increased significantly. Ultrasound is not the preferred detection means against SCM, it exhibits obvious advantages in the early diagnosis of SCM combined with other spinal cord malformations in fetal stage, and prenatal ultrasound can detect abnormalities early [13]. Normal X-ray is mainly used to display spinal cord malformations and can well display intraspinal OD, which mainly appears as spinal canal expansion, intervertebral space widening, or semi-vertebral malformations. Mahapatra [14] reported that the positive rate of X-ray examination in revealing iliac crest was 57%. However, X-ray examination also has obvious insufficiency in diagnosing SCM, especially when the angle of spinal twisting and rotation is very large, it will be difficult to accurately locate and diagnose SCM. Furthermore, normal X-ray shows insufficiently high positive diagnostic rate against cartilaginous or fibrous divide, and cannot find associated intraspinal abnormalities. With the applications of coronal and sagittal 3D reconstruction and CT thin-slice scanning, the positive detection rate of OD is close to 100%, which cannot only clearly show the SCM sites but also fully visualize the shape and traveling trend of the bony intervals in SCM; meanwhile, it can distinguish cases with cartilaginous, fibrous, or even no interval. MRI can clearly show the nature, type, and length of SCM, and shows significant advantages in determining tethered cord than CT, so it is one effective non-invasive examination method against SCM, and has unique advantages in displaying syringomyelia and determining the nature of associated lesions [15,16]. We believe the diagnosis of SCM mainly depends on imaging techniques, and different types of examination means have their own advantages and disadvantages in displaying the type, vertebral structure, and other associated malformations of SCM. Therefore, we propose the combination of normal spinal MRI scan plus whole spine 3D CT reconstruction, which can comprehensively evaluate the type, interval properties, and associated diseases of SCM when in conjunction with such auxiliary means as EMG, SEP, or urodynamics (Fig. 1).



Fig. 1 – Applications of spinal MRI scanning combined with whole spine 3D CT reconstruction against SCM-OD-S, which not only can fully visualize the site, shape, and traveling trends of OD but also has unique advantages in displaying syringomyelia and determining other associated spinal cord lesions.

Surgery is currently the only effective treatment method against SCM. As for the dual-tube type of SCM, OD is normally entrapped in SCM, and therefore becomes the direct cause of tethered cord and pulling impairment. Therefore, surgeries should be carried out as early as possible, especially for children in their development stage, which can help to prevent secondary damages caused by the growth of spinal cord or daily exercises [17]. There still exist controversies about the surgical treatment method against the single-tube type of SCM, and no consensus on the surgical treatment of SCM-OD currently. Some scholars believe that as for the patients with dual-tube SCM with scoliosis, if no preoperative Tethered cord syndrome appears and the imaging shows the upper and lower sites of OD exist split spinal cord, the intraspinal crest needs not to be dealt with before dealing with scoliosis [18]. However, presently, most scholars believe that scoliosis itself will lead to nerve damage, and spinal correction surgery will tract and extend the spine, so it will inevitably stretch the spinal cord and make it shift. Therefore, once the patients with scoliosis are diagnosed the combination of OD and crest formation, they should be surgically removed SCM, followed by scoliosis correction 2-6 months later [19,20].

OD generally locates in the protruded or adjacent site of scoliosis. During our study, intraoperative observation revealed that the lower part of SCM in most patients was in the same plane as the bottom of OD, while the upper end of SCM was much higher than that of OD; meanwhile, the OD bottom will be very close to the compression on the dural sac. Thus, it can be implied that during the development process, due to faster spinal development than the spinal cord which eventually results in longer spinal column than the spinal cord, OD will fix the spinal cord like a nail, thus resulting in tethered cord symptom of the spinal cord. If SCM combined with scoliosis can be early detected, as well as OD can be resected as soon as possible and the tethered spinal cord can be removed, it will prevent the further development of nerve damage and may also reduce the increase of the curvature of scoliosis. In this study, the patient with the longest OD involves four vertebrae, and that with the shortest OD is only like needle, so accurate preoperative positioning is of great importance, which can avoid intraoperative difficulties in finding OD and cutting off excess spinous process and vertebral lamina. The patients with postoperative transient unilateral lower limb paralysis all have lateral spinal stenosis, which causes the operations to be very difficult. So, preoperative CT localization must be accurate, and the intraoperative body position should be determined according to the OD direction, which should make OD perpendicular to the operating table as much as possible. During the laminectomy, vertebral plate forceps selected should be as thin as possible, and particular attention should be paid so as to avoid damaging the spinal cord in the narrower side. During resecting OD, the integrity of spinal dura mater should be tried to keep intact. The base of OD is the vertebral cancellous bone and gradually widens at the part near the vertebra, which sometimes may exhibit more bleeding when chiseling OD, so bone wax or hemostatic sponge should be prepared for timely hemostasis. Furthermore, OD not always lies in the middle of spinal canal, and the division of spinal canal can be

asymmetric, in which the narrow side has corresponding finer spinal cord, thus causing semi-spinal stenosis. After removed the crest, the dural mater of the bilateral longitudinal SCM should be both cut open so as to relieve the membrane compression on the spinal cord. The dural sac should be cut open spindly, the resultant dura mater should be retained as much as possible, and the ventral and north side of spinal dural mater should be sutured, respectively, so as to make the two semi-spinal cords located in the same dural cavity; if suturing the ventral side has difficulties, interrupted suture or non-suture is also acceptable. Because scoliosis commonly occurs in children at school age, and these children often develop poorly and frailly, especially some patients may have poor lung functions, the preoperative assessment must be accurate. Under normal circumstances, SCM and scoliosis should be carried out in different stages so as to ensure the safety.

Intraoperative application of neurophysiological monitoring significantly improved the surgical safety of complex scoliosis combined with SCM. Continuous free EMG monitoring can prompt real-time interference degree of surgical operations toward important nerves, thus setting up an invisible safety red line for the surgeons. By monitoring the latency and amplitude changes of SEP and MEP, the entire sensory and motor access can be real-time determined the fluency, especially in the surgical fields with severe adhesion or complex deformity, as well as those with un-determinable tissue, active stimulations by one nerve stimulator can determine the importance of the tissue, thus providing the greatest protective degree toward such important functions as defecation or sexual function. Combined with preoperative electrophysiological findings, patients' prognosis can be initially determine during surgery. However, due to complex intraoperative monitoring environments, which cannot provide good quantitative comparison, it is also the only option for intraoperative monitoring.

This study is a retrospective study toward the data collected, and it still needs large-sample, prospective, and randomized controlled trials for further investigating the classification of SCM combined with OD, as well as the surgical importance and risk prevention points in different types and the correlations between different types of SCM-OD and scoliosis. Secondly, the follow-up time is not long enough, and some patients are still in the growth and development period, so the follow-up needs to be continued to understand the long-term results.

#### **Ethical approval**

This study was conducted in accordance with the declaration of Helsinki. This study was conducted with approval from the Ethics Committee of the 306th Hospital of PLA. Written informed consent was obtained from all participants.

#### **Conflicts of interest**

None declared.

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