

REVIEW ARTICLE

Chinese expert consensus on diagnosis and management of split cord malformation

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ABSTRACT

Split cord malformation (SCM) is a neural tube defect that the spinal cord is longitudinally separated into two hemicords with individual functions, which causes severe spinal cord impairment and sensorimotor deficit. As a kind of myelodysplasia and a special type of tethered cord syndrome, SCM is not widely understood, and common issues in the diagnosis and treatment of SCM should be clarified. In this paper, the Chinese Split Cord Malformation Working Group made a consensus for SCM on embryopathogenesis and types, clinical presentations, neuroimaging assessment, indications and principle of the surgery, surgical techniques and nuances, and prognosis and follow up.

1 Introduction

Split cord malformation (SCM) is a neural tube defect that the spinal cord is longitudinally separated into two hemicords with individual functions, which causes severe spinal cord impairment and sensorimotor deficit. French pathologist Ollivier d' Angers initiated "diastematomyelia" from the Greek "diastema" and "myelos", meaning "cleft" and "spinal cord" to denote the malformation in 1837 [1, 2]. Some authors named the malformation as "split notochord

syndrome" [3] for its ontogenetic mechanism. Inconsistent definitions of the malformation increased the vagueness and confusions. Therefore, the Chinese Split Cord Malformation Working Group adopted the term "split cord malformation", which was named by Dachling Pang in 1992, for standardization, and reached a global consensus [4].

Split cord malformation is a kind of myelodysplasia and a special type of tethered cord syndrome, accounting for 4%–9% of all congenital spinal cord malformations. The deficit

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occurs more commonly in lower thoracic and lumbar segments than in upper thoracic segments, while rarely in cervical and lumbosacral spine [5–7].

A considerable number of neurosurgeons and spinal surgeons lack sufficient understanding of the occurrence, clinical characteristics and treatment of the disease, which leads to misdiagnosis and mistreatment. After collective discussion, the Chinese Split Cord Malformation Working Group has reached the following consensus on the diagnosis and treatment of SCM. The participants are listed in the end of this consensus (Appendix 1).

2 Embryopathogenesis and classification

2.1 Embryopathogenesis

The pathogenesis of SCM remains controversial. Unified theory of embryogenesis [4] firstly proposed by Pang et al. contends that all SCMs originate from one basic ontogenetic error occurring around the time when the primitive neurenteric canal closes, during the 3rd to the 4th week of embryogenesis. Maternal risk factor for SCMs may include: folate deficiency, in utero exposure to certain viruses or chemical compounds, maternal diabetes mellitus and genetic disorders, etc. The aforementioned risk factors may contribute to an abnormal mid-line adhesion or fistula between the ectoderm and endoderm, which splits the notochord and neural plate. The consequential notochord dehiscence induces neuroschisis and mesenchyme condensation in the vicinity of the fistula. The aggregated mesenchymal cells differentiate into osseo-cartilaginous, fibrous tissues and vascular structures, then bisect the spinal canal and spinal cord.

Three pathophysiological elements lead to clinical presentations: 1. Asymmetrical develop-

ments of the hemicords, which often result in a tenuous and dysplastic hemicord on one side; 2. Shearing and tethering effect on the spinal cord from the bisecting elements. Interior and posterior fibrous bands in the split and aberrant nerves originating from the interior aspect of the hemicord may contribute to the tethering effect as well [8, 9]; 3. Traction injury of the terminal conus medullaris due to mild-to-severe low-lying conus medullaris and thickened filum terminale.

2.2 Classification

Postulated by Pang et al. [1], SCMs can be categorized into two types. Type I SCM accounts for 40% of all SCMs and is a condition in which two hemicords, each with individual central canal and pia mater, inhabit their own dural sacs and are transfixed by a dural-sheathed rigid, bony median septum. Type II SCM accounts for 60% of all SCMs and indicates that two hemicords share a single dural sac despite separated by a nonrigid fibrous septum.

Pang Classification has catered for clinical requirements and became a commonly recognized method. There are reports that cannot be rigorously validated by Pang classification, albeit it is rare [10–12].

3 Clinical presentations

Split cord malformation, occurring more frequently in females than in males [13], usually has an insidious onset with slow progression. Initial hospital visit is often made before adolescent age.

3.1 Cutaneous lesions

Split cord malformation has many related cutaneous lesions including hypertrichosis, hyperpigmentation, dimples, pilonidal sinus, hemangiomas and subcutaneous lipoma [14, 15].

Cutaneous lesions are essential for an early diagnosis and assessment of SCM as primary reasons for SCM patients' hospital visits.

3.2 Lower extremities

Leg problems are the second earliest and most frequent symptoms associated with SCM following dermatological lesions. A pivotal element pertinent to the symptom is the very existence of the asymmetrical development of the hemicords [8]. A thinner maldeveloped hemicord compared to the better developed side often promises ipsilateral neural deficit such as atrophy of the leg and foot, ankle deformity, pain and numbness. While in the case of two equally divided hemicords, leg symptoms can be slight and unnoticeable.

3.3 Scoliosis

Scoliosis can be detected in 60% of all SCM patients, especially among those who have asymmetrically developed hemicords [16–19], and even be the only symptom in some cases [8, 20]. Scoliosis can be compensation to the tilted pelvis attributed to atrophy of the ipsilateral leg controlled by a maldeveloped hemicord, or a result of myodynamia imbalance surrounding the column, which is secondary to tethering-derived neural deficit. Vertebral body deformity, often seen in SCM patients, is another possible factor contributing to the development of scoliosis.

3.4 Lower back and leg pain

For SCM patients with symmetric hemicords, lower back pain is usually the main complaint without obvious lower extremity change or scoliosis. Pain can be either unilateral or bilateral, although the latter is more common. Traction from the septum damages the proximal cord and causes syringomyelia regardless of the symmetry of two hemicords. Syringomyelia can exist anywhere in the cord, from site above SCM

to either side of hemicords, which causes unilateral or bilateral lower back and leg pain.

3.5 Sphincter dysfunction

Bladder and bowel dysfunction is a less common symptom among SCM patients. It is usually not severe and primarily affects older children and adults. Sphincter dysfunction includes constipation or inactive bowel movement, frequent and urgent micturition, and enuresis, etc. The location of the septum is pivotal to the sphincter dysfunction. A higher located septum means relatively less significant low-lying conus, which promises less possibility of sphincter dysfunction with a later onset and slighter severity.

Each aforementioned symptom can happen alone or simultaneously in one patient. Children are more likely to have deformity, leg weakness and sphincter dysfunction and adults tend to present with lower back and leg pain. But some SCM Type II patients could be asymptomatic with only radiographic evidence [21]. Misdiagnosis and missed diagnosis are what we saw quite often in the triage of patients who were referred to orthopedic care. Unnecessary orthopedic procedures were performed based on a misdiagnosis of talipes equinovarus or idiopathic scoliosis. Delayed medical consultation would result in more neurological impairments and more orthopedic deformities.

4 Neuroimaging assessment

4.1 Magnetic resonance imaging

Besides reflecting associated vertebra anomalies noted ahead, sagittal images can help to locate the septum accurately. Microseptum and fibrous band at split point, which are always subtle and undistinguishable, should be recognized carefully in these images as tethering factors. The

axial images covering whole lesions can directly show the hemicords, two dural sacs, septum-pointed direction and abnormal fila terminale. Coronal images can reveal the relationship between septum and the convergence point of two hemicords. A full spine magnetic resonance imaging (MRI) is the best to present all coexisting lesions [12, 14, 16, 22].

4.2 Computed tomography

Computed tomography (CT) is valuable for studying bone structure, and the high-resolution CT can even catch a minuscule bony spur [15]. Three-dimension (3D) reconstruction CT plays a significant role in presenting spinal deformities including laminospinous defect, abnormal bifurcation of spinous process, hemivertebra and bifida vertebra. Especially, it provides 3D images of bony septum for clinicians to plan a surgery.

4.3 X-ray

X-ray scan can assist to show enostosis and spinal deformities such as scoliosis, kyphosis, hemivertebra, butterfly vertebra, Klippel-Feil syndrome and spina bifida. However, it lacks high resolution on bony ridge at the mid-line of spinal canal. With high sensibility in detecting and accuracy in typing, associated neuroimaging information is vital for clinical diagnosis and surgical plans.

4.4 Ultrasound testing

Ultrasound testing is the most favorable method in prenatal screening for its convenience and non-invasive nature. Detected by ultrasound testing [23], the lesion lying in the midline of two partly widened vertebral posterior ossification centers is a special characteristic of SCM, which may be furthered confirmed by fetal MRI. Ultrasound testing can also help to assess tethering severity and monitor post-operative

remission among the children under 12 months old by the pulsation of conus medullaris.

Electrophysiological tests, urogenital sonography, post-void residual volume (PVR) and urodynamic testing are optional and can serve in assessing surgical outcomes.

5 Indications and principle of the surgery

Surgery is the only effective way to remove abnormal anatomic structures tethering spinal cord in SCM.

5.1 Indications

5.1.1 Type I SCM

Surgical timing and indications of Type I SCM are similar to the ones of spina bifida occulta with tethered cord [7, 24]. Once diagnosed, infants and children should accept immediate surgical intervention [25, 26]. For adults, if there is no symptom or the condition is stable in recent years, the operation is unnecessary, and the condition can be observed continuously; however, if there is lumbago and leg pain or the condition tends to worsen, the operation should be performed to relieve the pain, prevent or delay the further aggravation of nerve damage caused by tethered cord.

5.1.2 Type II SCM

The surgical indications of Type II SCM are controversial. Some authors preferred an aggressive intervention regardless of symptoms and fibrous bands [13, 26], while others suggested a conservative monitoring for progression [21]. Lan et al. argued that the patients of Type II SCM may benefit few from surgery for lacking of symptoms or fibrous bands [9]. Shang et al. proposed that congenitally asymmetric hemicords cause most neural deficits in Type II SCM patients, whose growth hardly deteriorate

conditions unless cord tethering is involved, and surgery should only be considered with a progressive disease [27]. On the contrary, Pang et al. recommended Type II SCM patients to accept surgeries, because tethering factors surely existing in many cases will weaken neural functions constantly and surgery may protect them against deterioration [28].

Chinese Split Cord Malformation Working Group has reached a consensus that close monitoring on disease progression should be the initial option for children, while surgery is necessary if there exists: (1) symptoms resulted from tethered cord; or (2) evidence of fibrous bands, syringomyelia or low-lying conus medullaris on MRI [29]. In our practice, we performed surgeries on Type II SCM patients based on the same indications with Type I and identified fibrous bands-tethered cord in almost all of the procedure.

5.2 Principle of the Surgery

Remove any anatomic abnormality tethering spinal cord including osteocartilaginous tissues, fibrous septum and fixed connective tissues. Pay more attention to integrity of intervertebral facets for higher stability and less complications.

6 Surgical techniques and nuances

6.1 Bony septum

Precise localization of bony septum is crucial in SCM treatment, while it could be laborious on asymmetrical Type I patients, especially with scoliosis. For less bleeding and accidental injuries, it is beneficial for surgeons to review neuroimages preoperatively and correlate imaging manifestations to anatomical landmarks intraoperatively. Elder age, sturdy septa base and narrow spinal canal challenge neurosurgeons. Unossified septa will turn to be ossified

after toddler hood, which leads to a difficult extirpation. Impeded by ventrally originating blood supply, any manipulation without clear identification of spinal cord may bring about a catastrophe, massive epidural bleeding within a narrow surgical field. What we recommend, to preserve the spinal cord, is that an early durotomy of the sacs flanking the septum should be performed after rongeurizing the septum surface. Then, resect the residuals meticulously. Next, cut the vessels, aberrant nerves and fibrous bands between the interior cord and septum by electrocoagulation and gently pull the patty-covered cord outwards to avoid subarachnoid hemorrhage. Blood irrigation to the septum and adhesion will be cut off once a clear demarcation is dissected off. Also, once identified, supplying vessels of septum should be cut off. Commonly, the septa grow from the anteroinferior to the posterosuperior with a sturdy base, which require a resection following this longitudinal deviation, especially for the tightly squeezed cord. Microsurgical techniques and delicate instruments such as mini Friedman bone rongeurs, 3 mm high speed burr and ultrasonic bone scalpel will facilitate the extirpation of the septa. To circumvent further injury, little residual of the septa base is acceptable if the spinal cord is released from the oppression of septa. Covering rough bony bed with bone wax can achieve hemostasis and avoid air embolism and cord-shearing injury. Suture is unnecessary because ventral dura is so clung to the vertebra that CSF hardly leaks out. However, the dorsal dura needs a water-tight closure. Notably, insufficient resection of connective tissues (such as meninges in the split and fibrous bands), or unopened dura, can result in an incomplete detethering of the cord [7], which we should avoid.

6.2 Type II SCM

Type II SCM tethered by taut fibrous bands or septa requires a surgical performance. Many septa in Type II SCMs are too tenuous to appreciate in MRI and sometimes a surgical exploration could be inconclusive with unexperienced surgeons. However, a septum splitting and tethering the cord is identifiable in all Type I SCMs and most Type II SCMs in the lower end of the split, where the two hemicords rejoin. Fibrous bands from the posteroinferior to the anterosuperior terminate at the inferior pole of the split where surgery incision should be oriented. Dimples in the skin is the cutaneous end of the fibrous band indicating the deep end and an excellent mark to make incision. For patients without skin dimples, surgeons should carefully look for the defects in spine processes or laminae at the inferior end of the split where fibrous bands underneath bone defects can be easily detected in the epidural space. CT and MRI images are indispensable to localize the penetrating point of fibrous band on dura. Pang et al. reported that ventral tethering could occur in about 21% of Type II SCMs [28].

6.3 Aberrant nerves

Aberrant nerves originate from inner sides of hemicords and terminate at the dura on the surface of osseous septum or dorsal dura. Once aberrant nerves, serving as adhesive and tethering factors [8], are confirmed non-functional by neuro-electrophysiology monitoring, rhizotomy would be rational and necessary.

6.4 Filum terminale

Transection of the fila terminale is unnecessary without sphincter dysfunction or low-lying conus medullaris. However, severe low-lying conus medullaris, especially combined with thick fila terminale and lipoma, is a solid indicator of fila terminale transection regardless

of the symptoms. Some authors suggested fila terminale transection on all SCM pediatric patients [30, 31] instead of a delayed surgery and close monitoring on asymptomatic or stable adult patients, while the surgery should be performed if condition deteriorates. Extirpation of the septa should be performed prior to filum terminale transection [32]. Retraction of the spinal cord after filum terminale transection can cover the whole view of septum and make the resection more challenging. Filum terminale transection and spectrum transection should be performed simultaneously only the former may exacerbate the symptoms. In a rare case, two hemicords will not rejoin and a condition called "duplicate fila terminale" occurs. Although no splitting septum is between two filum terminale, the transection of both fila terminale is required to fully release the tethering of spinal cord.

6.5 Accompanying spinal cord pathology

If the accompanying pathologies coexist in the symptom progression or are potentially deleterious in the future, they can be removed at one stage with detethering procedures, especially within the same operation field with the septa. Most common comorbidities to SCM are congenital masses such as neuroenteric cyst, spinal bifida and other types of tethered cord syndrome [33].

6.6 Scoliosis

Scoliosis should be corrected after SCM surgery. Scoliosis correction before Type I SCM surgery could lead to spinal cord retraction in pediatric patients [16]. Some spine surgeons proposed a single stage surgery of both SCM (especially Type II) and scoliosis [34–36]. However, we recommend a second stage scoliosis correction following Type I SCM surgery to reduce the surgical risks. SCM extirpation may arrest the progression of preexisting scoliosis in non

Chiari II pediatric patients with a Cobb angle $< 30^\circ$. Scoliosis correction is needless if the scoliosis stabilizes or improves after SCM extirpation. A single-staged surgery can be performed at surgeons' discretion on Type II SCM patients with only filum terminale tethering but no fibrous bands.

6.7 Intraoperative neuroelectrophysiology monitoring

Intraoperative neuro-electrophysiology monitoring (IONM) can ensure the safety of surgical practice and is highly recommended. Constant EMG monitoring can detect nerve irritation in real time, and intraoperative electrostimulation is suggested to distinguish the functional nerves from tethered bands and non-functional nerves. Alteration in latency and amplitude of somatosensory evoked potential (SEP) and motor evoked potential (MEP) are essential real-time parameters on examining sensorimotor pathway integrity.

7 Prognosis and follow up

Recommended early surgical intervention promises a satisfactory recovery, especially for patients presenting with mild symptoms or recent onset of symptom. However, those with severe long-term symptoms improve indistinctively after SCM extirpation [37]. Pain can be remarkably relieved, and other symptoms including leg weakness, sphincter dysfunction, scoliosis and leg deformity may be partially diminished or just remain unchanged. Importantly, surgery can only contribute to detethering the cord and preventing further injury, but cannot reverse the existed malformation like leg atrophy due to asymmetrical hemicord. But delayed progression after surgery can also benefit young patients. Regular postoperative follow-up should include

MRI scan. The first-time postoperative follow-up needs to include the assessment on the extent of the septum extirpation, patency of the subarachnoid space and the extent of cord detethering, etc. Routine follow-ups should assess potential risk of retethering, the relapse of syringomyelia or congenital tumors, which rely on not only MRI but also the combination of neuroimaging, history, symptoms and IONM [38]. Of note, leg deformities may be more prominent as children grow up. It can be the result of retethering or asymmetrical hemicord development, which can be distinguished by routine EMG and SEP test. Symptoms in retethering patients will deteriorate while it remained unchanged in patients with asymmetrical hemicords. A whole spine X-ray is required for SCM patients with scoliosis to assess the progression and necessity for scoliotic surgical intervention.

Conflict of interests

The authors declare that they have no conflict of interests.

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Appendix 1 Participants of consensus on the diagnosis and treatment of SCM from Chinese Split Cord Malformation Working Group
(In alphabetical order of expert's family name)

1	Nan Bo	Shanghai Children's Medical Center
2	Qian Chen	Shenzhen Children's Hospital
3	Ruoping Chen	Xinhua Hospital Affiliated to School of Medicine, Shanghai Jiaotong University
4	Shuo Gu	Hainan Women and Children's Medical Center
5	Xiaosheng He	Xijing Hospital of Air-force Military Medical University
6	Wen Jin	Shanxi Children's Hospital
7	Cuicui Li	The 7th Medical Center of PLA General Hospital
8	Fangcheng Li	Guangzhou Women and Children's Medical Center
9	Hao Li	Fudan University Children's Hospital
10	Fuyun Liu	Zhengzhou University the Third Hospital
11	Wei Liu	Beijing Tiantan Hospital, Capital Medical University
12	Lin Qi	Children's Hospital Affiliated to Zhengzhou University
13	Xiang Qi	Beijing Children's Hospital, Capital Medical University
14	Aijia Shang	The First Medical Center of PLA General Hospital
15	Zhipeng Shen	The Children's Hospital , Zhejiang University
16	Hangyu Shi	Xi'an Children's Hospital
17	Benzhang Tao	The First Medical Center of PLA General Hospital
18	Feng Wan	Tongji Hospital Affiliated to Huazhong University of Science & Technology
19	Guangyu Wang	Qilu Children's Hospital of Shandong University
20	Jvlei Wang	Tangdu Hospital of Air-force Military Medical University
21	Linlin Wang	Institute of Reproductive and Child Health, Peking University Health Science Center
22	Shuihua Wu	Hunan Children's Hospital
23	Bo Xiu	The 7th Medical Center of PLA General Hospital
24	Zhi Yuan	The Second Hospital of Lanzhou University
25	Xuan Zhai	The Children's Hospital, Chongqing Medical University
26	Rui Zhang	Shandong Provincial Hospital Affiliated to Shandong First Medical University

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