

# Split cord malformations: A two years experience at AIIMS

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## ABSTRACT

**Background:** Over a 2-year period, 2008-2009, a total of 53 cases of split cord malformation (SCM) were treated at the All India Institute of Medical Sciences (AIIMS). This study is a retrospective analysis of clinical features, radiological findings, and surgical outcome of these patients.

**Materials and Methods:** During this period, 53 cases of SCM were treated at AIIMS. They constitute around 27% of all spinal dysraphism surgeries performed at the department of Neurosurgery, AIIMS; as 200 cases of spinal dysraphism were operated during the study period. The data was obtained from case files, operation notes, discharge summaries, and follow-up files.

**Observations:** There were 30 cases of SCM type I and 23 cases of type II SCM. Seven patients were adult above 18 years of age. Except 7 patients, remaining 46 were symptomatic. Bony deformity of spine was recorded in 24 patients; of them, 19 had scoliosis and 4 had kyphosis. Deformity of foot was recorded in 10 patients. Thirteen patients had hypertrichosis, while four had dermal sinus. Magnetic resonance imaging (MRI) was performed in all patients. MRI revealed syringomyelia in 14 patients; however, only one patient had associated Chiari malformation. Six patients had meningocele. Intra-operative; thick filum was noticed in 10 cases and in another 9 cases, there was filum lipoma. Dermoid was encountered in 4 patients, one patient had epidermoid tumor. Site of split was thoracic in 22, followed by lumbar region in 21 patients. Only 3 patients had split in cervical spinal cord. Seven patients had two separate splits at two different levels. Two patients had posteriorly located bony spur. All patients underwent surgery. Seven patients, those who had no neurological deficits pre-op, remained unchanged post-op. Amongst the 46 patients who had preoperative neurological deficits, eight had neurological deterioration post-op; five had deterioration in motor power and three had urinary problem. Five of these patients had type Id split, 2 had type Ic split, and one had type Ib split. However, among 8 patients who deteriorated post-op, four improved to preoperative status by the time of discharge. Thus, 4/53(7%) patients had long-term deficits, all with type Id split. Follow-up data was available for 36 patients (68%) and mean follow-up period was 12 months (range 6-24 months). Follow-up MRI revealed decrease in syringomyelic cavity in 6 of the 14 patients (44%) who had syringomyelia on preoperative MRI scans.

**Conclusion:** Overall, SCM is an uncommon condition. In all cases of progressive scoliosis, MRI must be carried out. We subjected all asymptomatic patients to surgery and none developed post-op deterioration. Overall post-op neurological deterioration was noticed in 15% patients, of which 8% had transient post-operative deterioration. The new Type I SCM subclassification system proposed by Mahapatra and Gupta is found to have a significant prognostic value in assessing post-operative neurological deterioration in patients with type I SCM.

**Key words:** Spinal dysraphism, split cord malformation, unified theory

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## Introduction

Split cord malformations (SCMs) are relatively rare forms of occult spinal dysraphism and tethered spinal cord syndrome. The majority of these cases present in early childhood with neurocutaneous stigmata being an early presenting feature. SCM constitutes about one third cases of spinal dysraphism, where spinal cord is divided over a portion of its length into two equal or unequal halves.<sup>[1]</sup> SCM are of two types, type I consists of two hemicords, each contained within its own dural sheath and separated by a median bony spur, and type II consists of two hemicord housed in a single dural tube separated by a fibrous median septum.<sup>[2]</sup> Mahapatra and

Gupta<sup>[3]</sup> proposed a new sub-classification for type I SCM's, wherein they divided type-1 SCM's into four types based on the intra-operative location of the bony spur causing the split. The bony septum usually originates from the vertebral body and proceeds posteriorly causing splitting of the cord. However, rarely there can be type I SCM with posteriorly located bony spur i.e. bony spur proceeding in a dorsoventral direction, base being attached to inner surface of lamina.<sup>[4,5]</sup> The authors present their experience of managing SCM cases in this article.

## Materials and Methods

It was a retrospective study carried out at the department of Neurosurgery, All India Institute of Medical Sciences, New Delhi, India. The study period was January 2008 – December 2009. During this period, 53 cases of SCM were treated at AIIMS. They constitute around 27% of all spinal dysraphism treated at our center, as 200 cases of spinal dysraphism were operated during the study period. The data was obtained from case files, operation notes, discharge summaries, and follow-up files. We used the new classification system proposed by Mahapatra and Gupta to subclassify patients with type I SCM.<sup>[3]</sup>

## Observations

There were 30 cases of SCM type I and 23 cases of type II SCM. Out of 30 patients of type I SCM; 15 had type Ia split, 5 had type Ib split, 4 had type Ic split and 6 patients had type Id SCM. Seven patients were adult above 18 years of age. Except 7 patients, remaining 46 were symptomatic. Bony deformity of spine was recorded in 24 patients; of them, 19 had scoliosis and 4 had kyphosis. Deformity of foot was recorded in 10 patients. Clinical examination revealed hypertrichosis in 13 patients, while 4 had dermal sinus. Magnetic resonance imaging (MRI) was performed in all patients. MRI revealed syringomyelia in 14 patients; however, only one patient had associated Chiari malformation. Retrospectively reviewing the MRI findings, it was noticed that syringomyelia was present in all cases of type Id split (6 patients), 3 cases of type Ic split, 4 cases of type Ib split and only one case of type Ia split. Six patients had meningocele. Site of split was thoracic in 22, followed by lumbar region in 21 patients. Only 3 patients had split in cervical spinal cord. Seven patients had two separate splits at two different levels. Two patients had posteriorly located bony spur with its base attached to inner surface of lamina.

All patients underwent surgery. The site of split was explored first and after adequately removing the spur and other tethering elements at the site of split, filum detethering was carried out in case of low lying tethered cord. The general principles were as follows:

We used a posterior midline approach centered over the spur. Laminotomy was performed at least one level above and one below the level of the spur. However, in case of posteriorly located bony spur, laminotomy/laminoplasty was avoided due

to high risk of cord laceration. Instead in such cases, lamina is gradually drilled to reach the base of the bony spur. The bony spur was gradually drilled of using a diamond drill. The dura was opened in a conventional linear manner with a gentle curve encircling the spur. The dural incision was extended about two levels above and below the level of lesion so that there remained sufficient dura for closure. Rest of the tethering elements like arachnoid bands and fibrous septa were released. Syringostomy/Syringosubarachnoid shunting was not performed in any of the cases. The dura was mobilized both vertically and in a lateral direction and watertight dural closure was performed using 5-0 vicryl suture.

Intra-operative, thick filum was noticed in ten cases and in another nine cases, there was filum lipoma. Dermoid was encountered in four patients and one had epidermoid tumor. Seven patients, those who had no neurological deficits pre-op, remained unchanged post-op. Amongst the 46 patients who had preoperative neurological deficits, eight had neurological deterioration post-op; five had deterioration in motor power and three had urinary problem. Five of these patients had type Id split, two had type Ic split, and one had type Ib split. However, among eight patients who deteriorated post-op, four improved to preoperative status by the time of discharge. Thus, 4/53 (7%) patients had long-term deficits, all with type Id split. [Table 1] gives an insight into the relationship of subtype of type I SCM with preoperative findings and post-operative outcome.

## Discussion

SCM is an uncommon congenital anomaly in which a segment of the spinal cord is divided into two parts by a fibrous or rigid bony spur.

These two hemicords may be separated by a bony/osseocartilagenous spur and be contained in separate dural sheaths (SCM type I) or they may be separated by a fibrous spur and contained in a single dural sheath (SCM type II).<sup>[2]</sup> The authors have used the new classification system proposed by Mahapatra and Gupta<sup>[3]</sup> to subclassify patients with type I SCM and found it to have a significant prognostic value. According to this new classification, type I SCM has been subdivided as follows:

- Type Ia. Bony spur in the center with equally duplicated cord above and below the spur

**Table 1: Subtype of SCM type I and its relationship to pre-operative findings and post-operative outcome**

SCM I subtype	Syringomyelia on pre-operative MRI pictures (n=14)	Post-operative deterioration (n=8)
Ia	1/14	0/8
Ib	4/14	1/8
Ic	3/14	2/8
Id	6/14	5/8

SCM – Split cord malformation; MRI – Magnetic resonance imaging

- Type Ib. Bony spur at the superior pole with no space above and a large duplicated cord lower down
- Type Ic. Bony spur at the lower pole with a large duplicated cord above
- Type Id. Bony spur straddling the bifurcation with no space above or below the spur.

Many hypotheses have been postulated for the genesis of these malformations. Based on observations made in chick embryos, Herren,<sup>[6]</sup> in 1940, postulated that SCM, then referred to as diplomyelia, resulted from an exaggerated folding of the neural plate. Gardner<sup>[7]</sup> hypothesized that the hydromyelic distension of the neural tube and its secondary rupture both on its ventral and dorsal aspect would constitute two neural tubes. Then the fibrous tissue of mesodermal origin that penetrates the space between the two neural tubes would constitute the fibrous or bony spur. Most widely accepted theory about embryogenesis of SCM was originally proposed by Bremer<sup>[8]</sup> and subsequently modified by Pang *et al.*,<sup>[2]</sup> as 'Unified theory of embryogenesis.'

Bremer<sup>[8]</sup> presented a theory in 1952 taking into account a dorsal intestinal fistula. From the archenteron that gives rise to the gut, a diverticulum develops that, upon expansion, separates the notochord and the neural plate into two parts. If this diverticulum opens at the skin level, it gives rise to the dorsal enteric fistula that is an open form of split notochord syndrome. If the endodermal elements disappear totally, there remains a fibrous or osseous septum between the two hemicords.

Pang *et al.*,<sup>[2]</sup> stated unified theory that is based on the concept of an endomesenchymatous tract and applies to both cases with a bone septum and associated vertebral malformation (SCM Type I) and cases where there is only a fibrous tract between the two hemicords and no associated vertebral malformation (SCM Type II). The endomesenchymal tract that determines SCM Type II is only constituted by meninx primitiva that does not contain precursors of bone cells. According to the theory, these lesions were divided into two types depending on the type of the midline mesenchymal derivative and the dural investment of the hemicords. SCM-I is characterized by the presence of double dural sacs, rigid extradural bony/cartilaginous spur leading into symmetrical or asymmetrical division of the cord. In SCM-II, there is a single dural sac with a non rigid fibrous spur and symmetrical division of the cord. SCM-II are slightly more common than SCM-I, constituting around 50-60% of SCMs.<sup>[2,9]</sup>

SCMs can present with a myriad of clinical manifestations. These can range from asymptomatic ones to pain, gait disturbance, motor or sensory deficits, and autonomic dysfunction.<sup>[3,10-13]</sup> In the current series, except 7 patients, remaining 46 were symptomatic. Thirteen patients had hypertrichosis, while four had dermal sinus. Bony deformity of spine was recorded in 24 patients; of them, 19 had scoliosis and

5 had kyphosis. Deformity of foot was recorded in 10 patients.

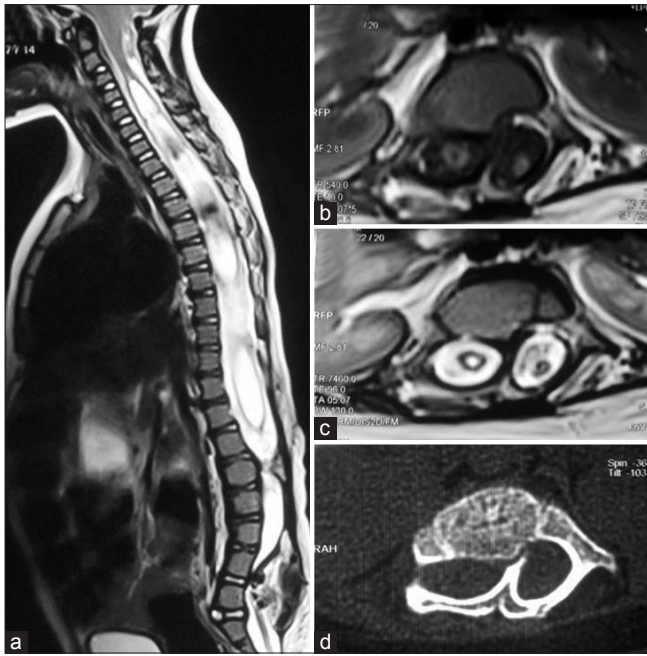
Magnetic resonance imaging is the diagnostic modality of choice and imaging of whole spine is essential to rule out other associated anomalies. CT scan is complimentary to MRI and is helpful in evaluating the nature of the spur and associated vertebral body anomalies [Figures 1 and 2]. In our series, MRI was performed in all patients. MRI revealed syringomyelia in 14 patients; however, only 1 patient had associated Chiari malformation. Six patients had meningocele. Intra-operative, thick filum was noticed in 10 cases, and in another 9 cases, there was filum lipoma. Dermoid was encountered in four patients and only one patient had epidermoid tumor. Double split was noticed in seven patients. Site of split was thoracic in 22, followed by lumbar region in 21 patients. Only three patients had split in cervical spinal cord. Seven patients had two separate splits at two different levels. Two patients had posteriorly located bony spur.

Although the unified theory of Pang *et al.*<sup>[2]</sup> has resolved the pathogenetic classification and clinical significance of these conditions, this theory does not explain the unusual variant such as dorsally located bony spur and complex SCM i.e. those associated with lipoma, dermoid, etc. In our study, two patients had type I SCM with bony spur which was situated dorsally and running in a dorsoventral direction. Chandra *et al.*,<sup>[5]</sup> in their reported case of dorsal bony spur in a lumbar SCM hypothesized two mechanisms for the occurrence of



**Figure 1:** (a-d) Sagittal, axial T1W, axial T2W MRI images and axial CT image of a representative case showing SCM type I with classic bony spur at L1 vertebral level causing asymmetric division of spinal cord into two separate hemicords with separate dural sheaths





**Figure 2:** (a-d) Sagittal, axial T1W, axial T2W MRI images and axial CT image of a representative case showing SCM type I with dorsally located bony spur at D12 vertebral level. Also note low lying cord and long segment syrinx

this rare variant: 1) passage of an abnormal cell population dorsally with subsequent loss of contact with the ventrally situated cell population and 2) migration of cells around the hemicords and subsequent passage between them in a dorso-ventral direction. Furthermore, Katoh *et al.*,<sup>[14]</sup> reported a case of SCM-II without a fibrous spur which again is not explainable by the unified theory. Hence, although a hypothesis has been proposed for this uncommon variant by some authors, further embryological studies are necessary to validate such proposals.

Surgery is the treatment of choice wherein the bony spur should be excised microsurgically. Low lying conus should also be addressed along with the excision of bony spur.<sup>[3,10]</sup> In all our cases which were associated with low lying conus, exploration and excision of the bony spur was followed by filum detethering in the same stage using standard microneurosurgical principles. All patients in this series underwent surgery. Seven patients, those who had no neurological deficits pre-op, remained unchanged post-op. Amongst the 46 patients who had pre-operative neurological deficits, 8 had neurological deterioration post-op; 5 had deterioration in motor power and 3 had urinary problem. Five of these patients had type Id split, two had type IC split, and one had type Ib split. However, among eight patients who deteriorated post-op, four improved to preoperative status by the time of discharge. Thus, 4/53 (7%) patients had long-term deficits, all with type Id split. Thus, the new subclassification system proposed by Mahapatra and Gupta is found to have a significant prognostic value

in assessing post-operative neurological deterioration in patients with type I SCM. We retrospectively correlated the intraoperative finding of type of split with pre-operative MRI finding of syringomyelia. It was noticed that syringomyelia was present in all cases of type Id split (6 patients), three cases of type Ic split, four cases of type Ib split, and only one case of type Ia split. Six of the fourteen patients (44%) had improvement in syringomyelic cavities after surgery at a mean follow-up period of one year.

The operative steps can be summarized as follows: 1) Midline posterior approach; 2) laminectomy is preferred and should be done at least one level above and one below the level of the spur. Laminoplasty should never be performed in cases with posteriorly located bony spur as it has very high risk of cord laceration. The bony spur needs to be handled with utmost care and should be gradually drilled of using a diamond drill; 3) The dura is opened in a conventional linear manner with a gentle curve encircling the spur. The dural incision is extended about two levels above and below the level of lesion so that there remains sufficient dura for closure; 4) The initial work on the bony spur is done by a pneumatic drill and the thinned out portion removed with Kerrison's punch; 5) The dura is mobilized both vertically and in a lateral direction. Watertight dural closure is of prime importance to avoid post operative cerebrospinal fluid leak. Dura is closed primarily with 5-0 vicryl suture and duraplasty is performed when primary dural closure is difficult; and 6) Rest of the wound is closed in layers.

## Conclusion

Split cord malformations are uncommon, complex conditions of spinal dysraphism. Dorsally situated bony spur is a very rare entity. In all cases of progressive scoliosis, MRI must be carried out. MR imaging is the diagnostic modality of choice complemented by CT scan. Other associated spinal anomalies should always be looked for, especially a low lying conus and syringomyelia. We subjected all asymptomatic patients to surgery and none of them developed post-op deterioration. Overall post-op neurological deterioration was noticed in 15% patients, of which 8% had transient post-operative deterioration. Surgical excision of the spur with detethering of filum, in cases of low lying conus, is the treatment of choice. The new Type I SCM subclassification system proposed by Mahapatra and Gupta proved to be of significant prognostic value in assessing post-operative neurological deterioration in patients with type I SCM.

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