University Journal of Surgery and Surgical Specialties

ISSN 2455-2860 Volume 2 Issue 1 2016

Profile of paediatric patients with split cord malformation

MANORANJITHAKUMARI M
Department of Neuro Surgery,
MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract :
AimTo present a case series of pediatric patients with split cord malformations , and to discuss the clinical presentations, radiological findings and the management.MethodsAll patients with age less than 18 years with clinical or radiological features of split cord malformations who had been admitted during march 2008 to march 2011 at madras institute of neurology were included in this study. 27 patients among the age group ranging from 1yr 3months to 16 yrs age were studied. 12 males and 15 females, with 25 patients having type I(pang classification) and 2 having type 2 split cord malformations. 16 patients had cutaneous stigmata. 11patients had orthopaedic problems. 5 patients had both. Back pain and leg pains were seen in 12 patients.15patients had motor involvement, 12 had sensory involvement and 10 had bladder involvement. 4 patients were without any neurological deficit. 7 patient had split cord at the lumbar level and 7 patient had in the thoracic level, 13 had dorsolumbar region. MRI scan revealed associated dermal sinus tract, epidermoids , lipoma and thickenened filum terminale . Detethering procedure had been done in all patients depending upon the type of tethering. Associated anomalies were also addressed.Follow up neurological examination was done at 3 and 6 months interval.Results18 patients experienced better or stable neurological status, 7 patient did not show any improvement in bladder function, 1 patient had worsening in neurological status 1 patient developed hydrocephalus for which ventriculoperitoneal shunt was done

Conclusion

All cases with simple and recent deficits showed good recovery with early surgery. However complex cases had delayed or less recovery. We recommend early surgery in all patients with split cord malformation before the onset of neurological deficit.

Keyword :

Split cord, split cord malformations, diastematomyelia, diplomyelia, congenital malformations

Profile of paediatric patients with split cord malformation

Introduction :

Overall there is a decreasing trend in neural tube defect (NTD) in the world literature. Since 1950 the incidence has fallen down from 40-50/10000 live births to 3-4/10000 live births. There is no report of exact incidence available from the Indian subcontinent. With the advent of MRI, and MRI being the investigation of choice for patients with scoliosis and dorsolumbar lipoma and with cutaneous stigmata the number of patients referred to us with split cord malformation has increased. In this study we analysed 27 paediatric patients with split cord malformations, clinical presentations, imaging and the management.

CINICAL MATERIAL AND METHODS

Study Design

This study is a retrospective analysis of patients who had been admitted with SCM between march 2008 to march 2011 in Institute of Neurology, Madras Medical College. Clinical profile, neurological findings, radiological finding, surgery and the outcome were analysed. Patients with a demonstrated open neural tube defect were excluded. Only those with at least 6 months of follow-up data were included.

Patient population*:*

Totally 27 patients were taken in to this study. There were 12 male and 15 female patients. Male female ratio was1:1.25.(literature 1:3) Age group was ranging from 1yr 3 months to 16 years and the mean age being 7 years.

Clinical Assessment

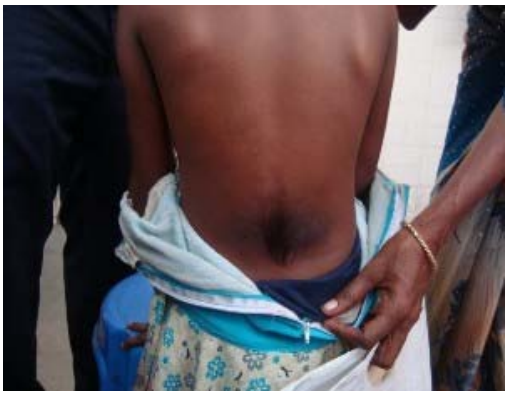
All patients underwent routine physical examination for neurocutaneous stigmata and/or skeletal deformity; they also received a thorough neurological evaluation. Most deficits detected were partial and usually subtle, as well as frequently asymmetric .Neuroimaging studies of the spine consisted of computerized tomography and MR imaging. Split cord malformation is best defined using multiple imaging modalities, because x-ray films, standard and three-dimensional spiral computerized tomography scans, and MR images all play useful roles in defining and following the lesion

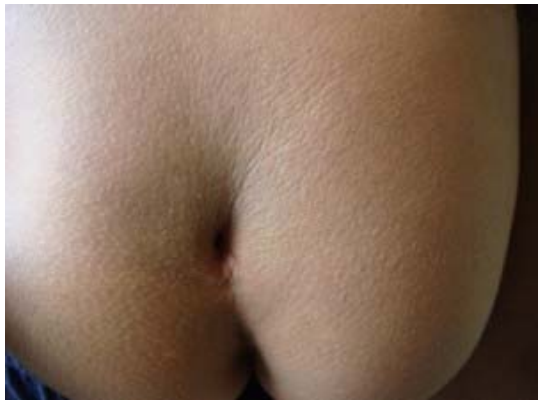
Presenting Signs and Symptoms

Presenting signs and symptoms are summarized in Table 1. Skin stigmata are well recognized and reported in occult spinal dysraphism with SCM Izci et al 2007 [5] highlighted the value of skin marker . In our series the presence of neurocutaneous stigmata was the presenting sign in 16of the 27 patients, the commonest being the hypertrichosis and subcutaneous lipoma . Interestingly, two of these patients had undergone surgery when younger for subcutaneous lipoma with split cord malformation in the lumbosacral region. Both patients presented for neurosurgical evaluation after the onset of back pain later in life and they were found to have fatty filum terminale. Back pain and leg pain were common, being seen in eight patients.Eighteen patients presented with neurological symptoms including leg weakness (15) and/or numbness and dysesthesias(12) and bladder incontinence (10). A skeletal deformity was seen in

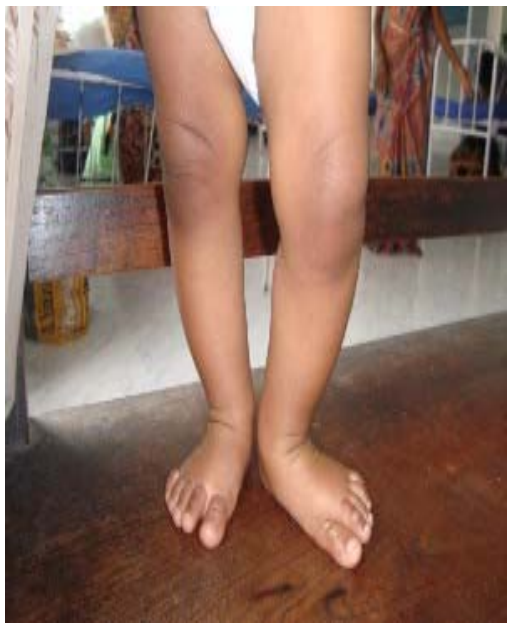
11) patients. Scoliosis , limb length discrepancy, club foot and congenital
dislocation of the hip are the presenting features of our patients with neuro orthopaedic syndrome. Two patients had undergone previous detethering surgery at another institution prior to presentation. Both went on to experience progressive lower-extremity pain and weakness, as well as bladder symptoms. Of note, in these patients secondary pathological entities were detected that had not been explored at the previous surgery (thickened fatty filum in both the patients)
table 1-presenting signs and symptoms:

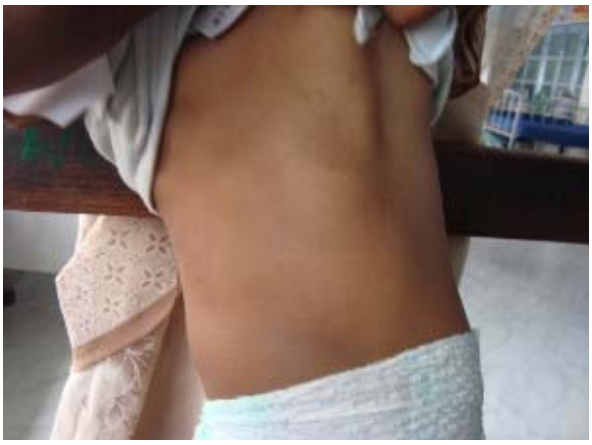
Neuro orthopaedic syndrome	11	40.74%
Scoliosis	9	33.33%
Club foot	7	25.92%
Congenital dislocation of hip	2	7.4%
Limb length discrepency	5	18.51%
Cutaneous stigmata	16	59.26%
Hypertrichosis	12	44.44%
Lipoma	6	22.22%
Dermal sinus tract	4	14.81%
Trophic ulcer	3	11.11%
Other associated systemic anomalies	1	3.70%
Horse shoe kidney	1	3.70%





Hypertrichosis sacral dimpling





Type of SCM

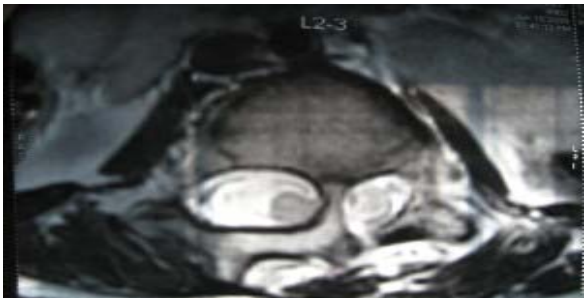
Twenty five of the patients were believed to have Type I SCM (diastematomyelia): a bone spicule and two dural sleeves separating the spinal cord segments were observed. The two remaining patients had Type II SCM (diplomyelia): the spinal cord was longitudinally split by a fibrous band. The SCM in all patients were in the dorsal, dorsolumbar, lumbar or lumbosacral region.92.5% of our patients had ventral bony spur based attached with posterior surface of the vertebral body.7.5% of the patients had dorsal bony spur in which the base of the spur seem to be attached with the lamina. 3 of our patients had multiple vertebral segment splitting by the bony spur, 22 patient had single vertebral level of splitting

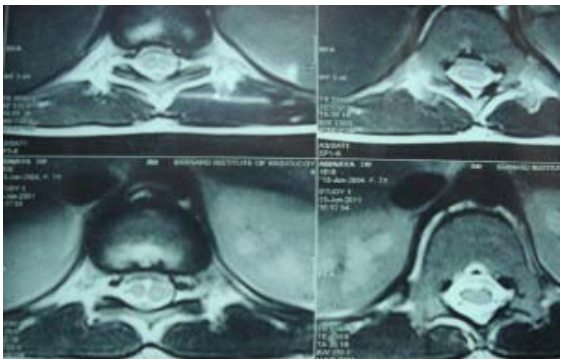
Table 2: Neurological findings

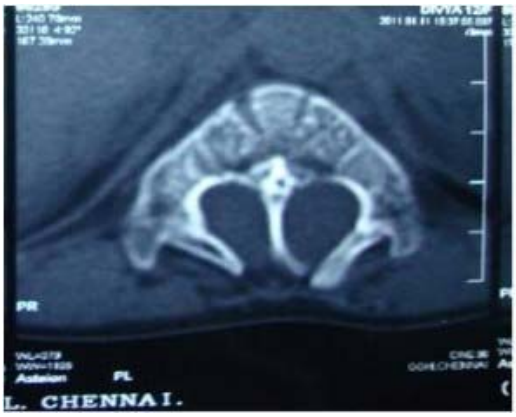
Motor findings	15	55%
Sensory symptoms	12	44.44%
Bladder incontinence	10	37%
Pain	8	30%
No deficit	4	14.8%

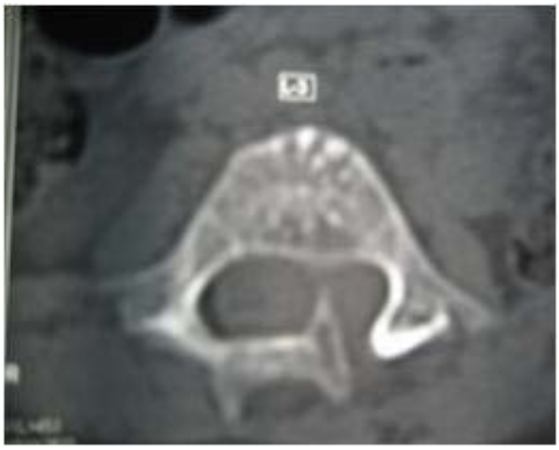
Type I(diastematomyelia)	25	92.59%
TypeII(diplomyelia)	2	7.40%

Level		
Dorsal	7	25.92%
Lumbar	7	25.92%
Dorso lumbar	13	48.18%
Ventral bony spur	23	92.5%
Dorsal bony spur	2	7.5%
Multiple vertebral level of split by bony spur	3	12%
Single vertebral level split by bony spur	22	88%





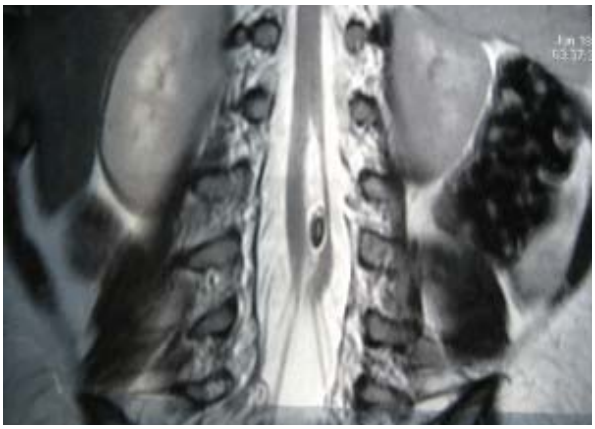


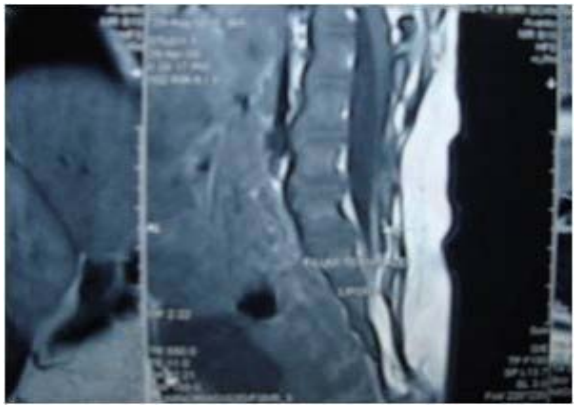


CT- axial Type I ventral spur CT axial Type I- dorsal spur

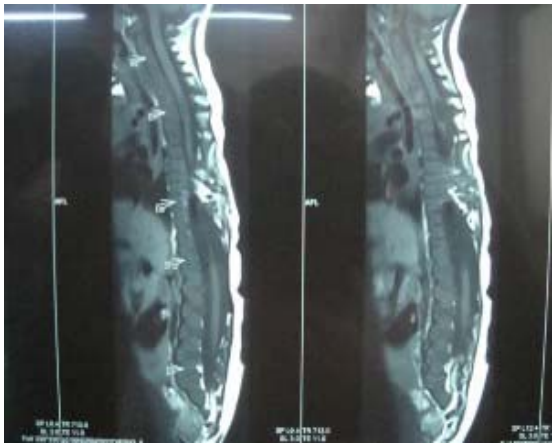
Associated pathological lesions causing tethering: MRI of patients withSCM showed other associated anomalies like thickened filum terminale, myelomeningocle, dermal sinus tract and epidermoids. Thickened filum terminale with low lying conus being the commonest anomaly associated with split cord malformation in our series. The following-table shows the MRI of the patients showing associated lesions

Lipomeningocele	3	11.11%
Dermal sinus tract	4	14.8%
Epidermoid	2	7%
Thickened filum terminale	8	29.6%





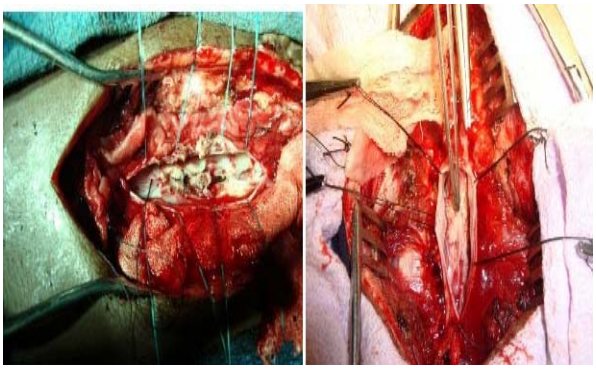
coronal picture - bony spur thickened filum terminale



subcutaneous lipoma

Surgical Treatment:

Prophylactic surgery was done in all patients .The goal of surgery was removal of the fibrous or bone septum, resection of any other local spinal cord attachments causing tethering, and exploration for associated tethering-related anomalies such as dorsal tethering bands or thick filum, lipoma, epidermoid, dermal sinus tract . For patients who had multiple levels of split, laminotomy was performed, and the rest of the patient laminectomy was done. For two patients only the release of the thickened filum terminal was done.Following the detethering prodedure,the two dural tubers were converted in to a single dural tube and the dura was closed posteriorly with or without placement of a patch graft, whereas anterior dural defects were left open. The patients were kept flat postoperatively for an average period of 72 hours and were then allowed to progressively advanced to full activity.



Laminotomy and excision of bony spur	2
Laminectomy and excision of bony spur	21
Laminectomy and excision of fibrous septa	2
Re exploration and release of thickened filum terminale	2
Excision of lipoma	3
Excision of epidermoid	2
Excision of dermal sinus tract	4
Release of thickened filum terminale	6

Surgery-Related Complications

Post operative CSF leak and pseudomeningocele occurred in 3 patients who had been managed conservatively by nursing in prone position and with drugs. One patient developed hydrocephalus for which shunting procedure had been done.

Results :

All the patients were discharged from hospital by 10-15 post operative day. And the follow up neurological examination was done at 3 months 6 months interval.18 patients had improvement in neurological recovery in the form of improvement in bladder function, motor power and relief from dysesthetic or radicular type of pain. 7 patients didnot show any improvement in neurological function.

One patient had developed paraplegia after the surgery done for D7 level SCM. One patient with associated infected epidermoid cyst developed chemical meningitis and hydrocephalus for which ventriculoperitoneal shunt was done. Patients who presented with pain symptoms were likely to experience significant relief after surgery. .

Confounding factors were those cases of progressive scoliosis caused by associated vertebral column abnormalities. Scoliosis at presentation is a clear prognostic indicator for the need for spinal stabilization. Of the 11 patients with signs of spinal curvature at presentation, 5 have required spinal fusion by orthopaedic surgeon

Results:

Improvement	18	66%
No improvement	7	26%
Worsening neurostatus	1	3.7%
Hydrocephalus	1	3.7%

Limitation of the study: 6 months follow up after surgery and regrowth of the bony spur in theis not enough to comment about the rate of retethering se patient. Long term follow up is needed.

Discussion:

Split cord malformation is a form of closed neural tube defect in which the spinal cord is longitudinally split by a fibrous band or a bone spicule. This new nomenclature of SCM was introduced by Pang, et al.[1]in 1992.Diastematomyelia usually refers to a split cord in which the two halves are separated by a bone spicule and contained within separate dural sleeves. In contrast, the term diplomyelia is generally used to describe a conditionof two hemicords within one dural sac, often with two complete sets of nerve roots, separated by a fibrous band. B

Based on the detailed findings in 39 cases and review of embryological features, Pang, et al., proposed a common origin of both malformations: an adhesion between the ectoderm and endoderm leads to an endomesenchymal tract that bisects the spinal cord. If the tract also contains cells of the meninx primitiva, the resultant malformation would be SCM Type I, or diastematomyelia. Otherwise, the formation of a separate dura sleeve and bone septum does not occur, and the malformation is a SCM Type II, or diplomyelia.[2] Both types of SCMs represent lesions that tether the spinal cord during growth and movement.2 Mahapatra and Gupta and Gupta and Mahapatra[3,8] classified type I spur into a-d types depending on the location of spur in between the proximal part and distal part of the split, the space available above and below the spur based on MRI finding. This classification has a direct relation to the surgical approach and risk of postoperative deterioration.As with all TCSs, surgical intervention is usually indicated based on an expected natural history of disease progression in the absence of treatment[9]. It was found that although a minority of the patients with SCM presented with urological signs or symptoms, formal testing demonstrated occult urological abnormalities in 75% of the patients. These urinary abnormalities tended to stabilize or improve after surgical intervention. Unlike other forms of spinal tethering, little is known about the long-term surgery-related outcome of patients with SCMIt is generally established that signs and symptoms in a patients with TCS worsen as they get older.[9] and most pediatric neurosurgeons believe that an infant or young child in whom TCS has been the diagnosed, regardless of origin, should undergo a detethering procedure.[1,4]

Indeed, many of these patients present with signs of spinal cord dysfunction, based either on symptoms, clinical examination,or more sophisticated objective data such as those obtained in urodynamic studies.For certain types of spinal dysraphism the risks of surgery and rates of subsequent retethering, or risk of other complications, are reasonably well established. For instance, a patient with a simple tight terminal filum usually experiences an uneventful postoperative course, and there is little chance of retethering. In contrast, patients with myelomeningoceles or lipomyelomeningoceles can have significant surgeryrelated complications, and the risk of retethering may be as high as 20%, depending on length of the follow-up period. The postoperative course of SCM is less well established. Currently in the literature few long-term outcome studies exist for patients who have undergone treatment of a SCM.4 Neurological outcome in the patients in this series was quite good.

A large number of systemic anomalies are also reported with SCM. Renal and urogenital anomalies, CVS anomalies and anorectal anomalies are not uncommon in patients with SCM.[10,11]

Jindal et al., [11] had reported a patient of SCM who had eventration of the diaphragm. Cases of situs inversus have also been reported. [12] In our series we experienced one case with horse shoe kidney, without any evidence of renal function abnormalities. In most cases the SCM was associated with VB anomalies, and the progression of the spinal deformity was probably unavoidable 7. This theory is supported by other studies in which the authors found that the detethering of a SCM did prevent

neurological complications but did not prevent the neuroorthopedic syndrome.1 With regard to the issues of the spinal column, it is probably fair to compare patients with SCMs and those with other spinal segmentation abnormalities, many of whom will require spinal stabilization early in life. In some cases an external orthosis may be advisable, but frequently it will not be adequate to halt the progression of curvature before skeletal maturity is achieved. In our series 5 of our patients experienced progressive scoliosis and they were referred to ortho spine surgeon for correction of scoliosis.

In contrast to some causes of spinal cord tethering, one important feature of SCM is its frequent association with secondary spinal anomalies. The association of VB anomalies has been discussed, but it is critical to know the high association between SCMs and other spinal cord tethering lesionsbefore surgery. Pang [2]believes that all patients with SCM will harbor a secondary lesion, and other authors contend 50 to 85% of patients will harbor a secondary lesion.[4] In this review, 17 of 25 patients harbored a second lesion. A fatty or tight filum was the most common but lipoma followed by lipoma. The two patients who presented for surgery after undergoing surgery for subcutaneous lipoma split cord malforamtion at another institution were both found to have a second tethering lesion fatty filum terminale that was not initially observed. With the high quality of MR imaging in the current era, these lesions will almost always be visible on preoperative imaging, but the importance of evaluating the distal conus either radiographically or surgically cannot be overstated. In addition, because tethering arachnoid bands extending dorsally and cephalad from the split in the spinal cord to the dura can occur, the existence of these lesions should also be assessed.

CONCLUSIONS

Split cord malformation is rare condition. We have seen about 27 patients in 3 yeras period(2008-2011). MRI is the investigation of choice. A large number of patients had associated other pathologies. In general, the long-term prognosis for patients with SCM is favorable.[4] Patients with SCM may have multiple causes of tethering associated with their congenital anomaly and that these causes need to be carefully examined by the initial evaluating neurosurgeon. Reoperation will almost certainly be required if these secondary lesions, such as fatty filum and dorsal tethering bands, are overlooked. In our series all cases with simple and recent deficits showed good recovery with early surgery. However complex cases had delayed or less recovery. We recommend early surgery in all patients with split cord malformation before the onset of neurological deficit

References:

1.Pang D, Dias MS, Ahab-Barmada M: Split cord malformation Part I: A unified theory of embryogenesis for double spinal cord malformation neurosurgery (31)451-480 1991

2Pang .D – ventral spinal cord tethering in split cord malformation-Neurosurg.Focus / Volume 10 / January, 2001

3Jindal A, Mahapatra AK. Split crd malformation-A study of 48 cases Indian J Paediatr 2003: 37, (603-607)

4.Ersahin Y, Mutluer S, Kocaman S, et al: Split spinal cord malformation in children J Neurosurg 88 – 57-65 1998

4.long term outcome for the patients with split cord malformations- mark r proctor et al- neurosurg focus 2010

5 Izci V, Gonul M, Gonul E. The diagnostic value of skin lesions in split cord malformation . J Clin Neurosci 2007;14:860-3.

6 Agrawal D, Mahapatra AK. Complex split cord malformation associated with situs inversus totalis. J Pediatr Neurosci 2007;2:39-40

7 Gupta DK, Mahapatra AK. Proposal for a new clinico-radiological classification of Type I split cord malformation-A prospective study of cases. Childs Nerv Syst 2007;23:1078

8. Mahapatra AK, Gupta DK. Split cord malformation-A clinical study of 254 patients and a proposal for a new clinical imaging classification Neurosurg 2005;103:531-6

9.Hoffman HJ, Hendrick EB, Humphreys RP: The tethered spinal cord: its protean manifestations, diagnosis, and surgical correc tion **Childs Brain 2:** 145–155, 1976

10.Jindal A, Mahapatra AK. Spinal lipomatus malformation. Indian J Pediatr 2000;67:342-6.

11.Kasliwal M, Mahapatra AK. Surgery for cord lipomas. Indian J Pediatr 2007;74:357-62.

12.Rowley VB, Johnson AJ. Lumbar split cord malformation with lateral hemimyelomeningocele and associated Chiari II malformation and other visceral and osseous anomalies: A case report. J Comput Assist Tomogr 2009;33:923-6