Abstract:
Background Diastematomyelia involving more than one level is rare that too involving the cervical region and it has to be corrected at the earliest to avoid the neurological symptoms occurrence and also to avoid the worsening of the symptoms. Case presentation We report two rare cases of diastematomyelia or split cord malformation, one with more than one level (cervical and lumbar) split cord by bony spur (type1at lumbar and type2 at cervical) and another case of split cord due to fibrous septum (type2). 12 year old boy, a known case of spinal dysraphism underwent myelomeningocele correction at newborn presented with fluid discharging sinus in the cervical segment in midline posterior aspect and imaging showed cervical and lumbar split cord due to bony septum. Another case of 9 year old female child presented with bladder dysfunction, imaging showed lumbar split cord by the fibrous band. Conclusion craniospinal MRI remains the choice, as this will not only delineate cord anatomy, but also screen the entire neuraxis for other associated cranial and spinal anomalies. In order to avoid irreversible damage to the neural tissue, surgical intervention must be instituted as soon as diagnosis is made.

Keyword:
Split cord malformation, spinal dysraphism, tethered cord syndrome

INTRODUCTION:
Diastematomyelia is a variety of spinal dysraphism in which there is a congenital splitting of some part of spinal cord or rarely more than one level involvement. Historically the term diastematomyelia (greek for “clefted medulla”)
and diplomyelia (Greek for “doubled medulla”) were used to describe the developmental anomalies now grouped together as split cord malformation. The sine qua non of these developmental anomalies are the presence of longitudinal cleft within the spinal cord across the one or more vertebral segments. Type 1 malformations (formerly diastematomyelia) are characterized by bony septum that cleaves the spinal cord in the sagittal plane and duplicated thecal sac. Type 2 malformations (formerly diplomyelia) are characterized by cleft cord within the single dural sac often tethered by a fibrous midline septum to the adjacent dura. Not all SCMs fit these descriptions precisely, but the classification scheme provides a framework for the surgical approach to these conditions. A unified theory attributing both type I and II SCM to a single error in embryogenesis: adhesion between the ectoderm and endoderm leading to a persistent neuroenteric canal. As with other forms of occult spinal dysraphism, SCM is often heralded by characteristic cutaneous and orthopedic findings. Mechanical tethering of the spinal cord at the level of cleft or because of the presence of other associated dysraphic lesions commonly leads to a recognizable constellation of neurological and urologic symptoms. Early detection and surgical intervention may prevent or interrupt deterioration.

CASE REPORT

Case 1

**Image 1 clinical picture showing cervical sinus tract**
12 year old boy, known case of spinal dysraphism underwent meningo-myelocele correction in newborn (lumbosacral region) presented with fluid discharging sinus (image 1) in posterior cervical segment in midline for six months duration. No history of features suggestive of meningitis. No history suggestive of limb weakness, sensory deficit or sphincter disturbances. On examination there is hypertrichosis in the lumbar region with irregular scar. Spino-motor bulk was normal and no evidence of LMN and UMN lesion.

**Image 2 clinical picture showing hypertrichosis in the lumbar region with scar**
MRI revealed sinus tract in the cervical segment in posterior midline passing obliquely upwards from C4 and connected to the thecal sac at the level of C2 (image 3) with split cord at C2 (image 4) due to fibrous band. The lumbar region showed there is split of the cord due to bony spur at the level of L2 and L3 with tethering of the cord (image 5). Computed tomography of brain was normal and there is no evidence of hydrocephalus (image 6).
image3: MRI of Cervical region sagittal view in T1WI image shows posterior cervical dermal sinus attached to dura

image4: T1WI axial view of cervical cord at C2 showing split
image 5: T2WI Axial view of lumbar spine showing split cord by bony spur

image6: Computed tomography of Brain shows no evidence of hydrocephalus

image7: Intraoperative picture showing cervical sinus tract

1st SURGERY:
On prone position the cervical sinus tract dissected carefully all around up to C2 lamina and dura opened with tract which continued dorsally, fibrous tract within the cord left undisturbed (image7). Postoperative period was uneventful. Patient advised second surgery for lumbar correction at the earliest, but he didn't turned up for surgery for two months.
After three months patient developed right lower limb weakness in the form of tripping of toes and knee buckling, also developed sphincter dysfunction in the form of difficulty in initiation of micturition. On prone position by midline lumbar incision, L3 Laminectomy done carefully from periphery to centre where the cord split by bony spur. L4 lamina also removed for neural decompression. The bony spur excised by bony rounger carefully with minimal retraction of the cord after separating the thecal sac attachment from the bony spur (image 8). Cord moved above after exision of bony septum (image 9). Dura opened and cord detethered. Dural repair was done to make single tube. Postoperatively weakness and bladder control improved slowly and patient developed CSF leak, managed conservatively.

**Image 9: Intraoperative picture showing cord after removal of bony spur**

**2nd SURGERY**

After three months patient developed right lower limb weakness in the form of tripping of toes and knee buckling, also developed sphincter dysfunction in the form of difficulty in initiation of micturition. On prone position by midline lumbar incision, L3 Laminectomy done carefully from periphery to centre where the cord split by bony spur. L4 lamina also removed for neural decompression. The bony spur excised by bony rounger carefully with minimal retraction of the cord after separating the thecal sac attachment from the bony spur (image 8). Cord moved above after exision of bony septum (image 9). Dura opened and cord detethered. Dural repair was done to make single tube. Postoperatively weakness and bladder control improved slowly and patient developed CSF leak, managed conservatively.
Nine year old female child presented with difficulty in initiation of micturition of three months duration. No history suggestive of lower limb weakness and sensory disturbances.

**Image12: MRI Sagittal view of Lumbosacral region**

**Image13: MRI Axial view of lumbar-sacral region shows split cord**

MRI revealed syrinx at the level of L2-L4 and split cord at the level of L4-L5 and L5-S1 with tethered cord and dural sinus tract at the level of L5 (images 12 and 13).

**Image14: Intraoperative picture shows fibrous band split the cord**

SURGERY:

On prone position by midline skin incision, L4, L5 and S1 laminectomy done. Dura opened, the fibrous band causing split cord excised. Duroplasty was done. Postoperative period became uneventful and patient regained bladder function slowly (image 14).

**Discussion:**

Diastematomyelia is a rare congenital anomaly that constitute one third of cases of spinal dysraphism. The incidence of spina bifida with split cord malformation and Meningomyelecele was reported in first case. Surprisingly Diastematomyelia is predominantly seen in females (9-12). Here we reported one case with type 2 diastematomyelia. Skin manifestations are very common in case of split cord malformation. Hypertrichosis was the most frequent cutaneous marker. In split cord malformation patients have Myelomeningocele sac as a superficial manifestation. Therefore, it should be kept in mind that the patients with overt spina bifida may harbor underlying Split cord malformation in significant proportion of cases and that should be given equal importance in overall management of spinal dysraphism patients. Other skin lesion noticed in patients with split cord malformation include capillaryhaemangioma, nevus, skin dimple and subcutaneous mass. As the incidence of skin marker is very high, the presence of which should alert the physician of a possibility of underlying occult spina bifida, particularly in Split Cord Malformation. As Pang has suggested that skin lesions such as hypertrichosis, capillary haemangioma and dural sinus tract represent minor aberration in the development of surface ectoderm that is due to the adverse influence of a dorsal endomesenchmal tract, but these aberrations may be overshadowed by chaotic changes in the ectoderm in case of an associated myelomeningocele. The leg length discrepancy was observed in first patient. James and Lassman and Guthkel discussed the orthopedic and neurological syndromes in children with diastematomyelia. Asymmetrical paresis of lower limb is common in patients with Split cord malformation. First child had asymmetrical lower limb weakness, where as children manifested as symmetrical weakness also reported.
Sphincter dysfunction has been variously reported in cases of split cord malformation. Bladder dysfunction had been present in second female child. It was based on clinical assessment only. It has been seen that neurological deterioration was more marked in patients with Split cord malformation who did not undergo early surgery. This deterioration was thought to have been caused by tethering of the cord, secondary to the continued growth of the spinal canal in respect to spinal cord. However the basic pathophysiology responsible for neurological deterioration remains, an intermittent, but chronic, repetitive ischemia to progressive ischemia resulting into spinal cord dysfunction in tethered cord syndrome, who underwent surgery for meningocele at birth without adequate investigation were having progressive neurological manifestation and after resurgery for Split cord malformation, their deficit became stabilized. Therefore we may infer from these that risk of having neurological deficits increases with age.Ultrasonography has been used to diagnose Split cord malformation in the prenatal period. CT myelography and MR imaging are needed for delineating the type of Split cord malformation and for showing the associated lesions. CT myelography is superior to MR imaging in defining the type of Split cord malformation. MRI has the added advantage of being noninvasive and very useful in defining the precise anatomy of the cord, nerve root and dural sac. Patient had at least one associated lesion, responsible for other causes of cord tethering. The reported spinal lesions in association with Split cord malformation are Meningomyelocele, meningocele, lipomeningomyelocele, limited dorsal myelischisis, teratoma, neuroenteric cyst, lipoma, dermal sinus tract, dermoid, epidermoid, AVM, epidural venous angioma, arachnoid cyst and thick filum terminale. A type I Split cord malformation was more frequently associated with a myelomeningocele than a type II Split cord malformation and the meningo-myelocele is usually situated below the bony septum. Aneuropathological deterioration is rarely seen after surgery and use of microsurgical technique and high speed drill are very helpful in avoiding neural injury. Though the pertinent data is not available for CSF leak from the operative site, but it remains one of the most distressing problem in the postoperative period. First child had postoperative CSF leak which has managed conservatively. Deaths are generally unrelated to the surgery of the Split cord malformation. The goal of intervention should be to detether cord and prevent neurological and orthopedic deterioration in the long run. The majority of the patients with Split cord malformation will be either stable or better following surgery as was noticed, however some will continue to worse in these cases screening should performed to find out lesion as a cause for progressive deterioration. Intradural endoscopic exploration helpful in type2 split cord malformation to inspect the cord all around and allowing entire length and extend of split cord and syrinx without doing long segment laminotomy.

**Conclusion:**
Split cord malformation cases may have association with Meningomyelocele in significant number of patients hence CT myelography is better in diagnosing the type of cord malformation, however craniospinal MRI remains the choice, as this will not only delineate cord anatomy, but also screen the entire neuraxis for other associated cranial and spinal anomalies. In order to avoid irreversible damage to the neural tissue, surgical intervention must be instituted as soon as diagnosis is made.
References:
1. Russell NA, Benoit BG, Joaquin AJ, al Fayez N. Adult diatematomyelia Can J Ne-
rol sci 1994.21:72-74

2 Korsvik HE, Keller MS, Sonography of occult dysraphism in neonates and in-
fants with MR imaging correlation. Radi-
ographics 1992, 12:297-306

3 Reigel DH, Rotenstein D. Spinabifida In: Paediatric neurosurgery of the American Association of neurological surgeon(ed). Paediatric Neurosurgery (3rd ed). Philadel-
phia: WB saunders,1994,pp 51-76

4 Pang D, Dias MS, Ahab-BarmadaM. Split cord malformation Part I A unified theory of Embrogenesis for double Spinal cord mal-

5. Kumar R, Bansal KK, Chhabra DK. Oc-
currence of split cord malformation in Men-
ingomyelocele:complex spinabifida. Pedi-


7 Kumar R, Singh SN. Spinal dysraphism-