Split cord malformation

- Effectiveness of conditional cash transfers for uptake and retention in HIV prevention of mother-to-child transmission services in low- and middle-income countries: a systematic review and meta-analysis
- Fetal imaging approach to spinal dysraphism diagnosis
- First-In-Human Application of Human Umbilical Cord-Derived Extracellular Vesicles in Tethered Spinal Cord Release Surgery
- Perinatal palliative care in an infant with exencephaly: Supporting life beyond 3 years of age
- The Predictive Value of Dry Brain Sign for Open Spina Bifida at First Trimester Anomaly Scan: A Case-Control study
- Burden of pediatric neural tube defects at a referral medical center in Tanzania
- Rehabilitation in a child with Chiari II malformation, lumbosacral meningomyelocele, achondroplasia and impaired respiratory regulation - a case report and literature review
- Anencephaly and palatoschisis in 2 newborn puppies

General information

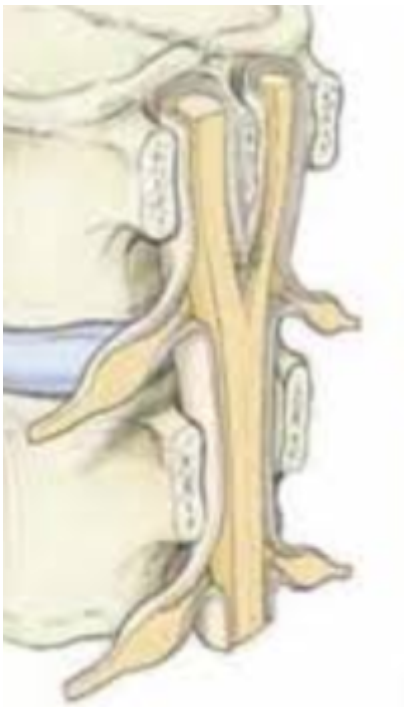
There is no uniformly accepted nomenclature for malformations characterized by duplicate or split spinal cords.

The term split cord malformation (SCM) was first introduced in 1992 by Pang et al., in an attempt to resolve the confusion existing in the pathological definition and the clinical significance of previously existing terminologies in the literature, diastematomyelia and diplomyelia, and the inconsistent usage of these two terms.

Pang et al have proposed the following. The term split cord malformation (SCM) should be used for all double spinal cords, all of which appear to have a common embryologic etiology [1)].

The term split cord malformation (SCM) should be used for all double spinal cords, all of which appear to have a common embryologic etiology.

Split cord malformation (SCM) has a rich history and has intrigued physicians for over 200 years. Many well-known figures from the past such as Hans Chiari and Friedrich Daniel von Recklinghausen, both pathologists, made early postmortem descriptions of SCM. With the advent of MRI, these pathological embryological derailments can now often be detected and appreciated early and during life. Our understanding and ability to treat these congenital malformations as well as the terminology used to describe them have changed over the last several decades [2].



Classification

Split cord malformations (SCMs) are among the rare congenital spinal anomalies. In 1992, Pang et al, proposed the “Unified theory of embryogenesis” and explained the formation of SCM type 1 and 2. This theory has been widely accepted in the neurosurgical literature, backed by several studies. However, there have been reports in the literature that defy both, the classification as well as formation of SCMs, based on the unified theory of embryogenesis.

Pang et al. classified spinal cord duplication anomalies into types I and II. The first is characterized by two hemicords, each contained within its own dural sac, and separated by an osteocartillaginous septum. Type II is defined by two hemicords in the same dural sac, separated by a fibrous septum [3] [4].

Type I split cord malformation

Type 1.5 split cord malformation ? [5].

Type 2 split cord malformation.

Much confusion still exists concerning the pathological definitions and clinical significance of double spinal cord malformations. Traditional terms used to describe the two main forms of these rare malformations, diastematomyelia, and diplomyelia, add to the confusion by their inconsistent usage, ambiguities, and implications of their dissimilar embryogenesis. Based on the detailed radiographic and surgical findings of 39 cases of double cord malformations and the autopsy data on two other cases, this study endorses a new classification for double cord malformations and proposes a unified theory of embryogenesis for all their variant forms and features. The new classification recommends the term split cord malformation (SCM) for all double spinal cords. A Type I SCM consists of two hemicords, each contained within its own dural tube and separated by a dura-sheathed rigid osseocartilaginous median septum. A Type II SCM consists of two hemicords housed in a single dural

tube separated by a nonrigid, fibrous median septum. These two essential features necessary for typing, the state of the dural tube and the nature of the median septum, do not ever overlap between the two main forms and can always be demonstrated by imaging studies so that accurate preoperative typing is always possible. All other associated structures in SCM such as paramedian nerve roots, myelomeningoceles manqué, and centromedian vascular structures frequently do overlap between types and are not reliable typing criteria. The unified theory of embryogenesis proposes that all variant types of SCMs have a common embryogenetic mechanism. Basic to this mechanism is the formation of adhesions between ecto- and endoderm, leading to an accessory neurenteric canal around which condenses an endomesenchymal tract that bisects the developing notochord and causes formation of two hemineural plates. The altered state of the emerging split neural tube and the subsequent ontogenetic fates of the constituent components of the endomesenchymal tract ultimately determine the configuration and orientation of the hemicords, the nature of the median septum, the coexistence of various vascular, lipomatous, neural, and fibrous oddities within the median cleft, the high association with open myelodysplastic and cutaneous lesions, and the seemingly unlikely relationship with fore and midgut anomalies. The multiple facets of this theory are presented in increasing complexity against the background of known embryological facts and theories; the validity of each facet is tested by comparing structures and phenomena predicted by the facet with actual radiographic, surgical, and histopathological findings of these 41 cases of SCM [6].

A new classification system proposed by Mahapatra and Gupta further divides type I SCM into four categories: Ia, bony spur in the center with equally duplicated cord above and below the spur; type Ib, bony spur at the superior pole with no space above and a large duplicated cord below; Ic, bony spur at the lower pole with a large duplicated cord above; and Id, bony spur straddling the bifurcation with no space above or below the spur [7].

Treatment

The risk of neurological deficits developing increases with age; hence, all patients with SCM should be surgically treated prophylactically even if they are asymptomatic [8].

see Type I Split Cord Malformation treatment.

Outcome

SCMs can lead to progressively worsening scoliosis and gait difficulties if left untreated.

Case series

From 1990 to 2014, 37 patients were operated. Five situations lead to the diagnosis (orthopedic disorders (n = 8), orthopedic and neurological disorders (n = 16), pure neurological disorders (n = 5), no symptoms except cutaneous signs (n = 7), antenatal diagnosis (n = 1)). Scoliosis was the most common associated condition. The level of the spur was always under T7 except in one case. There

were more type I (n = 22) than type II (n = 15) SCM.

Patients with preoperative neurological symptoms (n = 21) were improved in 71.4%. Five out of nine patients that had preoperative bladder dysfunction were improved. Eleven patients needed surgical correction of the scoliosis.

For us, the surgical procedure is mandatory even in case of asymptomatic discovery in order to avoid late clinical deterioration. In any case, the filum terminale need to be cut in order to untether completely the spinal cord. In case a surgical correction of a spinal deformity is needed, we recommend a two-stage surgery, for both SCM type. The SCM surgery can stop the evolution of scoliosis and it may just need an orthopedic treatment with a brace [9)].

Over a 16-year period, Mahapatra encountered 300 cases of SCM at AIIMS. Over the same period, more than 1500 cases of NTD were managed. SCM was noticed in 20% of cases with NTD. Skin stigmata were noted in two-third of the cases, and scoliosis and foot deformity were observed in 50% and 48% cases, respectively. Motor and sensory deficits were observed in 80% and 70% cases, respectively. Commonest site affected was lumbar or dorsolumbar (55% and 23%, respectively). In 3% cases, it was cervical in location. Magnetic resonance imaging (MRI) scan revealed a large number of anomalies like lipoma, neuroenteric cyst, thick filum and dermoid or epidermoid cysts. All the patients were surgically treated. In type I, bony spurs were excised, and in type II, bands tethering the cord were released. Associated anomalies were managed in the same sitting. Patients were followed up from 3 months to 3 years.

Overall improvement was noticed in 50% and stabilization in 44% cases and deterioration of neurological status was recorded in 6% cases. However, 50% of those who deteriorated improved to preop status prior to discharge, 7-10 days following surgery.

SCM is rare and not many large series are available. They operated 300 cases and noticed a large number of associated anomalies and also multilevel and multisite splits. Improvement or stabilization was noted in 94% and deterioration in 6% cases. They recommended prophylactic surgery for our asymptomatic patients [10)].

Mahapatra et al. in 2005 reported the first 254 cases of SCM treated surgically during a period of 16 years.

Patients' demographic profiles, imaging studies, operative details, complications, and surgical outcomes were evaluated retrospectively. A new classification based on intraoperative findings is proposed. The mean age of the patients was 7.3 years (female/male 1.5:1). Type I SCM was seen in 156 patients (61.4%) and 98 patients (38.6%) had Type II SCM. Skin stigmata were present in 153 cases (60%); hypertrichosis, being the most common, was seen in 82 cases (32.3%). Asymmetrical lower-limb weakness and sphincter disturbances were present in 173 (68.1%) and 73 (33%) cases, respectively. Of the symptomatic cases, 39% (68 of 173) showed improvement in motor power, 57.9% (33 of 57) experienced sensory improvement, and 27.3% (20 of 73) regained continence. None of the 38 patients in the asymptomatic group had postoperative neurological deterioration. The neurological status was unchanged in 63% of the cases. A new subclassification of Type I SCM is proposed, based on the intraoperative location of a bone spur causing the split, which may have a bearing on surgical dissection and outcome. Based on the authors' experience with 25 cases of Type I SCM, they have classified the disorder into four subtypes: Type Ia, bone spur located in the center with duplicated

cord above and below the spur (12 cases); Type Ib, bone spur at the superior pole with no space above it (four cases); Type Ic, bone spur at the lower pole with large duplicated cord above (three cases); and Type Id, bone spur straddling the bifurcation with no space above or below the spur (six cases). The risk of injury to the hemicords is highest in the Id subtype (four of six patients in this group deteriorated neurologically in the present series, whereas none with subtypes Ia-c worsened).

This is the largest series on SCMs so far reported in the world literature The risk of neurological deficits developing increases with age; hence, all patients with SCM should be surgically treated prophylactically even if they are asymptomatic. This new classification is easy to use and remember and takes into account the use of intraoperative findings that may have a bearing on surgical outcome [11].

Retrospective analysis of 19 cases of SCM, thirteen were grouped under (Pang) type I and 6 in type II. Their ages ranged from 1 month to 9 years (mean 3.5 years). 14 of these were male children. The NOS without neurological signs was detected in 6 cases whereas pure neurological signs without NOS were seen in 8 patients. However, the rest 5 had a mixed picture of NOS and neurological dysfunction. Nine of 19 cases presented with cutaneous stigmata, mainly in the form of a hairy patch. 18 cases had other associated craniospinal anomalies i.e. hydrocephalus, meningomyelocele, syrinx, dermoid, teratoma, etc. Detethering of the cord was done in all cases by the removal of fibrous/bony septum. Associated anomalies were also treated accordingly. Follow up of these cases ranged from 6 months to 6 years. Six cases of NOS group neither showed deterioration nor improvement, and remained static on follow up. However, four of 8 children with neurological signs showed improvement in their motor weakness, and 1 in saddle hypoaesthesia as well as bladder/bowel function. In 5 cases of a mixed group, two had improvement in their weakness and one in hypoaesthesia, but no change was noticed in NOS of this group as well. Hence surgery seemed to be effective, particularly in patients with neurological dysfunction [12].

Proctor and Scott reviewed the results obtained in 16 patients in whom the senior author performed surgery over a 13-year period (average length of follow up almost 8 years).

Presentation, surgical approach, and the outcome are evaluated, and the long-term outcome of neurological status, pain, bowel/bladder disturbance, and spinal deformities are emphasized.

The primary conclusion is that patients with SCM generally tolerate surgery well and experience few complications. Neurological deterioration is rare except in cases in which retethering occurs, (two patients in this series). Although impaired bowel and bladder function was stabilized or improved and pain was reliably relieved postoperatively, preexisting vertebral column deformities usually progressed after surgery and, in most cases, required spinal fusion [13].

In 2000 Forty-eight patients of split cord malformation operated during a six years period were studied clinically and radiologically.

The mean age of symptomatic patients was more than that of asymptomatic ones (6.85 years vs 2.03 years). The dorsolumbar and lumbar regions were most frequently involved and in three cases the cervical spine was affected. Weakness of lower limbs (n=37), muscle atrophy (n=23) and gait disturbance were the most common indicators of motor system involvement. The sensory complaints

were mainly hypoesthesia (n=16), trophic ulcer (n=4) and autoamputation (n=3). Hypertrichiosis was the most common cutaneous marker present alone or in combination with other markers in 21 cases. MRI, done in all cases, correctly established the diagnosis. Additional lesions causing tethering were seen in 50% cases and were simultaneously treated. Associated Chiari malformation was seen in 12%. Of the 42 symptomatic patients, 21 improved, in 17 (40%) the neurological deficits stabilized and 4 showed deterioration. Cerebrospinal fluid fistula occurred in 4 patients and 3 had wound infections. Among the asymptomatic patients none had neurological deterioration postoperatively.

Split cord malformations are rare spinal cord disorders. Complete neural axis should be scanned at the first instance to determine associated lesions. Good results can be expected in about 90% patients with minimal complications [14)].

Thirty-nine patients with split cord malformations (SCM) were studied in detail with respect to their clinical, radiographic, and surgical findings as well as their outcome data. Eight patients were adults and 31 patients were children. According to the classification endorsed by Part I of the SCM study, 19 patients had Type I SCM (6 adults and 13 children), 18 patients had Type II SCM (2 adults and 16 children), and 2 patients had composite SCM with both lesion types situated in tandem. Six SCMs were cervical, 2 were thoracic, and 31 were in the lumbar region. All 8 adults had pain and progressive sensorimotor deficits at diagnosis. Only 16 of the 31 children had symptoms, and among these, 14 had progressive sensorimotor deficits, but only 6 had pain. The difference in the clinical picture between adults and children is similar to that described in the tethered cord syndrome, except for left-right functional discrepancy, which was prominent in 8 children with SCM but rarely seen in tethered cord syndrome due to other causes. Cutaneous manifestations of either occult or open dysraphic states were present in all but 3 patients; hypertrichosis was by far the best predictor of an underlying SCM, being found in 56% in the series. Neurological deterioration in SCM was independent of the lesion type: the Type I:Type II ratio for symptomatic progression was 13:11. It was also independent of the location of the lesion: 67% of patients with cervical SCMs had symptomatic progression versus 64% of patients with thoracolumbar lesions. High-resolution, thin cut, axial computed tomographic myelography using bone algorithms was more sensitive than magnetic resonance imaging in defining the anatomical details of the SCM. Radiographic classifications of the SCM, using the nature of the median septum and the number of dural tubes as criteria, was always possible without ambiguity. However, whereas every Type I bone septum was identified preoperatively, only 5 Type II fibrous septa were revealed by preoperative imaging, even though a fibrous septum and/or other fibroneurovascular bands were found tethering the hemicords in every Type II case at surgery. Complete imaging studies also showed that all lumbar SCMs had low-lying coni and at least one additional tethering lesion besides the split cords, whereas only 1 of 7 cervical and high thoracic SCMs had a low conus and a second tethering lesion. The surgical goal for SCM was release of the tethered hemicords by eliminating the bone spurs, dural sleeves, fibrous septa, or any fibroneurovascular bands (myelomeningoceles manqué) that might be transfixing the split cord. Type I cases were technically more difficult and had a slightly higher surgical morbidity than Type II cases, especially if an oblique bone septum had asymmetrically divided the cord into one larger hemicord and one smaller, hence, very delicate, hemicord [15)].

Case reports

Nazarali et al. reported on two patients who atypically presented with SCM in adulthood and reviewed previous reports [16)].

A rare case of a child with a complex spina bifida with two different levels of split cord malformation (SCM) type 1 and single-level type 2, a nonterminal myelocystocele, coccygeal dermal sinus, bifid fatty filum and hydrocephalus, which substantiates the neurenteric canal theory and have further tried to highlight the importance of complete Magnetic resonance imaging (MRI) screening of the whole spine and brain with SCM to rule out other associated conditions. The patient was admitted with a leaking myelocystocele with bilateral lower limb weakness. MRI of the whole spine with a screening of brain was done. Patient underwent 5 operations in the same sitting- (According to classification given by Mahapatra et al.) removal of SCM type 1a at D7-8; removal of SCM type1c at L2-3; removal of SCM type 2 at D10; repair of nonterminal myelocystocele at D6-D10; low-pressure ventriculoperitoneal shunt on right side with excision of dermal coccygeal sinus; and, excision of bifid fatty filum. The clinic radiological findings in our patient further substantiate the multiple accessory neuroenteric canal theory in the development of a composite type of SCM. The physical and neurological signs of SCM and nonterminal myelocystocele should prompt the neurosurgeon to consider performing the screening MRI of the whole spine with the brain to rule out other composite types of SCM and hydrocephalus [17].

A 78-year-old woman presented for evaluation of back pain, urinary dysfunction, leg weakness and progressive equinovarus foot deformity. She reported that shortly after her birth in 1924, she underwent resection of a subcutaneous 'cyst' in the lower lumbar area. Seven years prior to evaluation at our institution, she had undergone bilateral total knee arthroplasty for osteoarthritis. After the procedure, she began to experience severe low back pain that radiated into her legs. Weakness of the foot inverters, urinary dysfunction and worsening bilateral equinovarus foot deformity developed in the years following the surgery. MRI revealed a split cord malformation with a tethered spinal cord. Because of the patient's age and poor medical condition, her symptoms were managed conservatively. This case demonstrates symptomatic deterioration in an elderly patient with a tethered spinal cord after many years of clinical stability [18].

A 32-year-old man with the adult-onset of impairment of sacral functions with lumbar fibrous diastematomyelia is reported. Surgical release of the spinal cord was followed by improvement of the patient's function [19].

References

[1], [6]
Pang D, Dias MS, Ahab-Barmada M. Split cord malformation: Part I: A unified theory of embryogenesis for double spinal cord malformations. Neurosurgery. 1992 Sep;31(3):451-80. Review. PubMed PMID: 1407428.
[2]
Saker E, Loukas M, Fisahn C, Oskouian RJ, Tubbs RS. Historical Perspective of Split Cord Malformations: A Tale of Two Cords. Pediatr Neurosurg. 2017;52(1):1-5. PubMed PMID: 27806370.
[3]
Pang D, Dias MS, Ahab-Barmada M. Split cord malformation: Part I. A unified theory of embryogenesis for double spinal cord malformations. Neurosurgery 1992;31:451-480.
[4]

Pang D, Dias MS, Ahab-Barmada M. Split cord malformation. Part II: Clinical syndrome. Neurosurgery 1992;31:481-500.

5) Sun M, Tao B, Luo T, Gao G, Shang A. We Are Cautious to Use the Term, 'Split Cord Malformation Type 1.5'. J Korean Neurosurg Soc. 2022 Aug 22. doi: 10.3340/jkns.2022.0058. Epub ahead of print. PMID: 35989187.

7) , 8) , 11) Mahapatra AK, Gupta DK. Split cord malformations: a clinical study of 254 patients and a proposal for a new clinical-imaging classification. J Neurosurg. 2005 Dec;103(6 Suppl):531-6. PubMed PMID: 16383252.

9) Beuriat PA, Di Rocco F, Szathmari A, Mottolese C. Management of split cord malformation in children: the Lyon experience. Childs Nerv Syst. 2018 May;34(5):883-891. doi: 10.1007/s00381-018-3772-3. Epub 2018 Mar 26. Erratum in: Childs Nerv Syst. 2018 May 17;:. Pierre-Aurelien, Beuriat [corrected to Beuriat, Pierre-Aurélien]; Federico, Di Rocco [corrected to Di Rocco, Federico]; Alexandru, Szathmari [corrected to Szathmari, Alexandru]; Carmine, Mottolese [corrected to Mottolese, Carmine]. PubMed PMID: 29582170.

10) Mahapatra AK. Split cord malformation - A study of 300 cases at AIIMS 1990-2006. J Pediatr Neurosci. 2011 Oct;6(Suppl 1):S41-5. doi: 10.4103/1817-1745.85708. PubMed PMID: 22069430; PubMed Central PMCID: PMC3208912.

12) Kumar R, Bansal KK, Chhabra DK. Split cord malformation (scm) in paediatric patients: outcome of 19 cases. Neurol India. 2001 Jun;49(2):128-33. PubMed PMID: 11447430.

13) Proctor MR, Scott RM. Long-term outcome for patients with split cord malformation. Neurosurg Focus. 2001 Jan 15;10(1):e5. PubMed PMID: 16749757.

14) Jindal A, Mahapatra AK. Split cord malformations–a clinical study of 48 cases. Indian Pediatr. 2000 Jun;37(6):603-7. PubMed PMID: 10869139.

15) Pang D. Split cord malformation: Part II: Clinical syndrome. Neurosurgery. 1992 Sep;31(3):481-500. Review. PubMed PMID: 1407429.

16) Nazarali R, Lyon K, Cleveland J, Garrett D Jr. Split cord malformation associated with scoliosis in adults. Proc (Bayl Univ Med Cent). 2019 Mar 27;32(2):274-276. doi: 10.1080/08998280.2019.1573624. eCollection 2019 Apr. Review. PubMed PMID: 31191152; PubMed Central PMCID: PMC6541173.

17) Khandelwal A, Tandon V, Mahapatra AK. An unusual case of 4 level spinal dysraphism: Multiple composite type 1 and type 2 split cord malformation, dorsal myelocystocele and hydrocephalous. J Pediatr Neurosci. 2011 Jan;6(1):58-61. doi: 10.4103/1817-1745.84411. PubMed PMID: 21977092; PubMed Central PMCID: PMC3173919.

18) Pallatroni HF, Ball PA, Duhaime AC. Split cord malformation as a cause of tethered cord syndrome in a 78-Year-old female. Pediatr Neurosurg. 2004 Mar-Apr;40(2):80-3. PubMed PMID: 15292638.

19) Chehrazi B, Haldeman S. Adult onset of tethered spinal cord syndrome due to fibrous diastematomyelia: case report. Neurosurgery. 1985 May;16(5):681-5. PubMed PMID: 3889701.