Long-term outcome for patients with split cord malformation

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Object. Split cord malformations (SCMs) are relatively rare forms of occult spinal dysraphism (OSD) and tethered spinal cord syndrome. The majority of these cases present in early childhood, with neurocutaneous stigmata being an early presenting feature. Prophylactic detethering surgery is advocated by most neurosurgeons due to the risk of neurological deterioration over time caused by patient growth and activity. However, unlike other forms of OSD, the course of SCM progression after surgery is not well understood, and little has been published about long-term follow-up results. In this study the authors review 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).

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

Conclusions. 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 post-operatively, preexisting vertebral column deformities usually progressed after surgery and, in most cases, required spinal fusion.

KEY WORDS • split cord malformation • spinal dysraphism • tethered cord syndrome • spina bifida occulta • diastematomyelia • diplomyelia

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.,^{16,17} in 1992, to eliminate confusion created by the use of the terms diastematomyelia and diplomyelia. 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 condition of two hemicords within one dural sac, often with two complete sets of nerve roots, separated by a fibrous band. Unfortunately, inconsistencies in the use of these terms have created substantial confusion on this subject in the literature. 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 dural

sleeve and bone septum does not occur, and the malformation is a SCM Type II, or diplomyelia.^{17,20}

Both types of SCMs represent lesions that tether the spinal cord during growth and movement.¹⁷ As with all TCSs, surgical intervention is usually indicated based on an expected natural history of disease progression in the absence of treatment.^{9,22} In a previous review we examined the pre- and postoperative urinary function in patients with SCM by performing formal urodynamic studies.²⁰ 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 SCM. In this series we examine the long-term outcome of patients with SCM who have undergone surgical detethering.

CINICAL MATERIAL AND METHODS

Study Design

This study is a retrospective analysis of patients who underwent surgical repair of SCM between 1987 and

Abbreviations used in this paper: MR = magnetic resonance; OSD = occult spinal dysraphism; SCM = split cord malformation; TCS = tethered cord syndrome; VB = vertebral body.

1997; all operations were conducted by the senior author. Hospital and office records patients were reviewed. Patients with a demonstrated open neural tube defect were excluded. Only those with at least 3 years of follow-up data were included. In our previous review of urinary abnormalities in patients with SCM, 15 patients were reviewed, but the present patient population reported here is different. Only six patients from the previous study were eligible for inclusion in this study, because our criteria required that currently all patients be surgically treated by the senior author, thus allowing for a more homogenous patient population with regard to surgical technique and outcome data.

Patient Population

The mean overall patient age at surgery was 11 years (median 5 years, range 1–44 years). Only three patients presented outside of the pediatric age group, all with symptoms of adult-onset TCS. Their respective ages of 20, 40, and 44 years may be higher than expected for such a group. The mean age of the patients presenting in the pediatric age group alone was 5.2 years and the median age was 3 years. There were six males and 10 females, with a 1:1.7 ratio. In the literature, the male/female ratio is generally 1:2 to 3.

Clinical Assessment

All patients underwent routine physical examination for neurocutaneous stigmata and/or skeletal deformity; they also received a thorough neurological evaluation, which was performed by the senior author in all patients. Most deficits detected were partial and usually subtle, as well as frequently asymmetric, with no predisposition found for one leg over the other. In six patients pre- and postoperative urodynamic studies were performed (this data is reviewed in a prior publication^{2,20}). 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.^{7,21}

Surgical Treatment

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, which can be seen in the majority of patients.^{4,11,16} Following the detethering procedure, 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 levels.

RESULTS

Patient Population and Follow Up

Using the aforementioned criteria, 16 patients were eligible for inclusion in this study. In the same time period

the senior author treated seven additional patients with SCM associated with myelomeningocele. In the literature it has been estimated that 6% of patients with myelomeningocele also have SCM, the majority of which are near the neural placode.¹⁰ The mean follow-up period was 7.6 years (range 3–14 years).

Type of SCM

Eleven 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 five remaining patients had Type II SCM (diplomyelia): the spinal cord was longitudinally split by a fibrous band. Of note, the SCM in all patients was in the lumbar or lumbosacral region.

Presenting Signs and Symptoms

Presenting signs and symptoms are summarized in Table 1. The presence of neurocutaneous stigmata was the presenting sign in seven of the 16 patients. Interestingly, two of these patients had undergone surgery when younger for cosmetic purposes: a hairy patch was removed in one and a lipoma in the other. Both patients presented for neurosurgical evaluation after the onset of back pain later in life. Back pain and leg pain were common, being seen in eight patients. Eight patients presented with neurological symptoms including leg weakness and/or numbness and dysesthesias. A skeletal deformity was seen in six patients. Three patients presented with isolated scoliosis, two with leg-length asymmetry, and one with scoliosis and leg asymmetry. Bowel and bladder symptoms, alone or combined, were present in eight patients. One girl presented with failure to thrive and a history of vomiting, which proved to be secondary to severe constipation from neurogenic bowel dysfunction. After an abdominal x-ray film revealed widened pedicles at L-3, an MR image of the spine was ordered, which led to the diagnosis of SCM.

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 (lipoma in one patient and fatty filum in the other).

TABLE 1
Presenting signs and symptoms in 16 patients with SCM

Sign or Symptom	No. of Patients
neurocutaneous stigmata	
hypertrichosis	5
lipoma	2
neurological deficit	
motor weakness	7
sensory loss	3
leg/back pain	8
bowel/bladder dysfunction	8
skeletal deformity	
leg or foot abnormality	3
scoliosis	4
hip dislocation	1

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Few patients in this series presented with a solitary finding. With the exception of one patient with a cutaneous lipoma and a second patient with scoliosis, all other patients were believed to have multiple signs or symptoms at presentation.

Tandem Pathological Lesions

In 10 of 16 patients a second lesion was discovered in addition to the SCM, and one patient harbored two associated lesions. The most common second lesion was a fatty filum (six patients). Lipomas were found in four patients. An isolated syrinx was observed in one patient. In the patient with two additional lesions, one was a lipoma and the other was a Chiari malformation with syringomyelia that required decompressive surgery.

Surgery-Related Complications

There was only one early surgery-related complication: one patient developed a cerebrospinal fluid leak that required wound reexploration on postoperative Day 10. No source for the leak was found, and the patient recovered uneventfully with bedrest. This patient went on to make an excellent recovery with complete resolution of preoperative symptoms.

Late surgery-related complications were relatively rare as well. Two patients have required a second operation for retethering. One of the patients experienced symptomatic back and leg pain and required reexploration 4 years after her first surgery. Of note, this patient harbored a complex intradural lipoma, which makes the case unique in this series. At the second surgery, dense arachnoid adhesions were found and surgically released. Now 6 years post-surgery, the patient is once again experiencing worsening back pain. A third exploration for tethering is being considered, although the source of the pain is unclear; she also has progressive scoliosis, which will require spinal fusion. One other patient in the series in whom surgery was previously performed by the senior author in 1979, presented 8 years later (during the study period) with tethered cord syndrome that required reoperation. She has done well since the second operation, which was 13 years ago.

At this point, three of the patients have required spinal fusion for progressive scoliosis after the initial detethering surgery. This is not necessarily a failure of the surgery and can be related to progression of the preexisting scoliosis caused by underlying VB anomalies. A fourth patient is experiencing progressive L1–2 kyphosis 11 years after surgery. She has no clinical or MR imaging evidence of retethering, but at age 13 will probably require spinal fusion. Another patient, mentioned above because of the need for a second detethering procedure, may also need spinal fusion. All patients with subsequent deformity of the spine had initially presented as infants or toddlers for their surgery, and this complication was not seen in the adult-onset patients in whom the skeleton was mature.

DISCUSSION

It is generally established that signs and symptoms in patients with TCS worsen as they get older.^{9,16,22} 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.²⁰ 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 surgery-related 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.^{4,11}

Neurological outcome in the patients in this series was quite good. Many patients (eight of 16) presented with subtle lower-extremity dysfunction, which seemed to stabilize or improve after surgery. Fifteen of the 16 patients experienced stabilization or improvement in their neurological function postoperatively. In only one patient did progressive neurological dysfunction develop. However, in addition to SCM she also harbored a complex lipoma. This girl had undergone surgery at 9 months of age, and she returned at age 5 years with progressive lower-extremity weakness. Urodynamic studies confirmed progression of spinal cord dysfunction, and at reoperation dense arachnoid scarring was found. Six years postoperatively she is neurologically stable but having back pain, which may be primarily caused by progressive spinal curvature. This low rate of progression of neurological deficit (one patient) compares favorably with other tethering lesions. This rate is significantly below that seen with lipomyelomeningoceles, another form of occult spinal dysraphism, which at our institution has a retethering rate of approximately 20%.

Patients who presented with pain symptoms were likely to experience significant relief after surgery. This result seems to be in accordance with other reported outcomes.¹³ Confounding factors were those cases of progressive scoliosis caused by associated vertebral column abnormalities, as well as the case of complex lipoma in which retethering occurred.

Scoliosis at presentation is a clear prognostic indicator for the need for spinal stabilization. Of the four patients with signs of spinal curvature at presentation, three have required spinal fusion. The fourth patient, currently aged 13 years, is experiencing progressive L1–2 kyphosis and may require a stabilization procedure. In most cases the SCM was associated with VB anomalies, and the progression of the spinal deformity was probably unavoidable. 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 a study examining the progression of scoliosis in children with myelomeningocele, the authors also found a poor correlation between detethering surgery and the subsequent need for stabilization. In cases in which significant spinal curvature ($> 40^\circ$) was demonstrated before surgical detethering surgery, 86% of the patients subsequently required spinal stabilization.¹⁹

In a previously published study²⁰ of patients with SCM, we evaluated the disparity between the subtle urinary symptoms at presentation and the actual objective degree of urinary dysfunction. Changes in the urinary tract system were seen in approximately 75% of patients. This degree of urinary tract dysfunction tended to stabilize or improve after surgery.²⁰ Based on our previous study, it is advocated that pre- and postoperative urodynamic testing be performed in patients with SCM. This is in agreement with recommendations reported by Perez, et al.,¹⁸ who examined 27 patients with SCM, in 11 of whom urodynamic studies had been conducted (only three had pre- and postoperative studies). Only those 14 patients with urological complaints were sent for urological evaluation; the most common complaint was urge incontinence. No patient in this group required long-term catheterization, although chronic anticholinergic therapy was needed in some. Because of the prevalence of urodynamic problems in this patient group (50 to 60% in their study), they concluded that all patients with SCM should undergo formal urological evaluation.¹⁸ The prevalence of urinary tract dysfunction in patients with SCM seems to be similar to other patients with TCS. It is well established in TCS that urodynamic testing can reliably demonstrate early changes in spinal cord function indicative of tethering seen before surgery.^{3,5,6,8,12,14,15,18} Furthermore, it is an excellent way to monitor postoperative outcome, and it is a sensitive index of the possibility of retethering. Several reports on the postoperative improvement of urodynamic symptoms and bladder function also exist.^{5,6,8}

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 lesions before surgery. Pang¹⁶ 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.^{4,11} In this review, 11 (69%) of 16 patients harbored a second lesion. A fatty or tight filum was the most common (six patients), but lipoma was also common (four patients). The two patients who presented for surgery after undergoing detethering surgery at another institution were both found to have a second tethering lesion 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. The exact frequency of this finding cannot be determined with certainty in our own operative series, but it appears to be

a frequently observed source of tethering in these patients as well.

CONCLUSIONS

In general, the long-term prognosis for patients with SCM is favorable.^{4,11} It is also clear from our study that 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. However, a lower rate of retethering is expected if the spinal cord is adequately released at the initial operation. Because we were surprised to find one patient with a Chiari Type I malformation and extensive syringomyelia in association with her SCM, our neuroradiologists therefore now screen all suspected SCM patients by conducting sagittal cranial and spine MR imaging as part of their standard workup. The embryological and causal relationship between these disorders is not clear. We have been struck by the number of patients who eventually required surgery for progressive scoliosis. Most of these patients displayed no progressing neurological deficits, and it is presumed that the bone anomalies themselves, perhaps accentuated by previous bone removal, led to this outcome. We have not investigated the role of segmental fusion at the time of initial SCM repair, although it may be one way of preventing this late-onset complication.

References

1. Andar UB, Harkness WF, Hayward RD: Split cord malformations of the lumbar region. A model for the neurosurgical management of all types of 'occult' spinal dysraphism? **Pediatr Neurosurg** 26:17–24, 1997
2. Bauer SB: Neurogenic vesical dysfunction in children, in Walsh PC, Retik AB, Stamey TA, et al (eds): **Campbell's Urology**. Philadelphia: WB Saunders, 1992, pp 1634–1668
3. Dator DP, Hatchett L, Dyro FM, et al: Urodynamic dysfunction in walking myelodysplastic children. **J Urol** 148:362–365, 1992
4. Ersahin Y, Mutluer S, Kocaman S, et al: Split spinal cord malformations in children. **J Neurosurg** 88:57–65, 1998
5. Flanagan RC, Russell DP, Walsh JW: Urologic aspects of tethered cord. **Urology** 33:80–82, 1989
6. Gross AJ, Michael T, Godeman MF, et al: Urological findings in patients with neurosurgically treated tethered spinal cord. **J Urol** 149:1510–1511, 1993
7. Harwood-Nash DC, McHugh K: Diastematomyelia in 172 children: the impact of modern neuroradiology. **Pediatr Neurosurg** 16:247–251, 1991
8. Hellstrom WJG, Edwards MSB, Kogan BA: Urological aspects of the tethered cord syndrome. **J Urol** 135:317–320, 1986
9. Hoffman HJ, Hendrick EB, Humphreys RP: The tethered spinal cord: its protean manifestations, diagnosis, and surgical correction. **Childs Brain** 2:145–155, 1976
10. Iskandar BJ, McLaughlin C, Oakes WJ: Split cord malformations in myelomeningocele patients. **Br J Neurosurg** 14: 200–203, 2000
11. Jindal A, Mahapatra AK: Split cord malformations—a clinical study of 48 cases. **Indian Pediatr** 37:603–607, 2000
12. Johnston LB, Borzyskowski M: Bladder dysfunction and neu-

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- rological disability at presentation in closed spina bifida. **Arch Dis Child** **79**:33–38, 1998
13. Kaminker R, Fabry J, Midha R, et al: Split cord malformation with diastematomyelia presenting as neurogenic claudication in an adult: a case report. **Spine** **25**:2269–2271, 2000
 14. Mevorach RA, Bogaert GA, Baskin LS, et al: Lower urinary tract function in ambulatory children with spina bifida. **Br J Urol** **77**:593–596, 1996
 15. Palmer LS, Richards I, Kaplan WE: Subclinical changes in bladder function in children presenting with nonurological symptoms of the tethered cord syndrome. **J Urol** **159**:231–234, 1998
 16. Pang D: Split cord malformation: Part II: Clinical syndrome. **Neurosurgery** **31**:481–500, 1992
 17. Pang D, Dias MS, Ahab-Barmada M: Split cord malformation: Part I: A unified theory of embryogenesis for double spinal cord malformations. **Neurosurgery** **31**:451–480, 1992
 18. Perez LM, Barnes N, MacDiarmid SA, et al: Urologic dysfunction in patients with diastematomyelia. **J Urol** **149**:1503–1505, 1993
 19. Pierz K, Banta J, Thomson J, et al: The effect of tethered cord release on scoliosis in myelomeningocele. **J Pediatr Orthop** **20**:362–365, 2000
 20. Proctor MR, Bauer SB, Scott RM, et al: The effect of surgery for split spinal cord malformation on neurologic and urologic function. **Pediatr Neurosurg** **32**:13–19, 2000
 21. Skalej M, Duffner F, Stefanou A, et al: 3D spiral CT imaging of bone anomalies in a case of diastematomyelia. **Eur J Radiol** **29**:262–265, 1999
 22. Yamada, S, Zinke DE, Sanders D: Pathophysiology of "tethered cord syndrome." **J Neurosurg** **54**:494–503, 1981

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