Split spinal cord malformations in children

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The authors reviewed and analyzed information on 74 patients with split spinal cord malformations (SSCMs) treated between January 1, 1980 and December 31, 1996 at their institution with the aim of defining and classifying the malformations according to the method of Pang, et al.

Computerized tomography myelography was superior to other radiological tools in defining the type of SSCM. There were 46 girls (62%) and 28 boys (38%) ranging in age from less than 1 day to 12 years (mean 33.08 months). The mean age (43.2 months) of the patients who exhibited neurological deficits and orthopedic deformities was significantly older than those (8.2 months) without deficits (p = 0.003). Fifty-two patients had a single Type I and 18 patients a single Type II SSCM; four patients had composite SSCMs. Sixty-two patients had at least one associated spinal lesion that could lead to spinal cord tethering. After surgery, the majority of the patients remained stable and clinical improvement was observed in 18 patients.

The classification of SSCMs proposed by Pang, et al., will eliminate the current chaos in terminology. In all SSCMs, either a rigid or a fibrous septum was found to transfix the spinal cord. There was at least one unrelated lesion that caused tethering of the spinal cord in 85% of the patients. The risk of neurological deficits resulting from SSCMs increases with the age of the patient; therefore, all patients should be surgically treated when diagnosed, especially before the development of orthopedic and neurological manifestations.

Key Words * diastematomyelia * diplomyelia * spinal dysraphism * split spinal cord malformation * tethered spinal cord * children

Diastematomyelia and diplomyelia are congenital spinal anomalies in which the spinal cord is longitudinally split by a rigid or fibrous septum, respectively. However, the terms "diastematomyelia" and "diplomyelia" have been used interchangeably.[14,17,19,20,28,34] Pang and colleagues[24,26] have suggested that these confusing and misleading terms should be abandoned and have proposed an alternative classification to deal with all double spinal cord malformations. They also have proposed a unified theory that explains the embryogenetic mechanisms of all variants of split spinal cord malformations (SSCMs). All SSCMs originate from one basic error that occurs at approximately the time at which the primitive neurenteric canal closes. This basic error is the formation of an accessory neurenteric canal between the yolk sac and amnion, which is subsequently invested with mesenchyme to form an endomesenchymal tract that splits the notochord and neural plate. These authors have defined
two types of SSCMs. A Type I SSCM consists of two hemicords, each contained within its own dural tube and separated by a dura-sheathed rigid median septum. A Type II SSCM consists of two hemicords housed in a single dural tube separated by a nonrigid, fibrous median septum.

With this classification in mind, we have reviewed and analyzed 74 cases of SSCMs treated at our institution and present our findings.

CLINICAL MATERIAL AND METHODS

Patient Population

The cases of 74 patients with SSCMs who were surgically treated between January 1, 1980 and December 31, 1996 were reviewed. Patient age and gender, symptoms and signs, radiological and operative findings, complications, associated anomalies, outcome, and pathological specimens were evaluated retrospectively in patients treated more than 3 years ago and prospectively in patients treated in the past 3 years. When diagnosed, all SSCMs (diastematomyelia or diplomyelia) were surgically treated, even if the patient was neurologically intact.

Surgical Procedures

In patients who had Type I SSCMs, laminectomy was performed around the attachment of the rigid median septum and the septum was dissected subperiosteally from its dural sleeve within the dural cleft. The septum was removed by using a rongeur or a high-speed microdrill. After removal of the septum, the dura was opened on both sides of the dural cleft. Fibrous bands or paramedian dorsal roots from the medial aspects of the hemicords, which were adherent to the dural sleeve, were sectioned and the dural sleeve was resected. Only posterior dural closure was performed.

In patients with Type II SSCMs, laminectomy was performed near or at the caudal end of the split segment. The dura was opened in the midline. The fibrous septum and/or paramedian dorsal roots were excised. Any associated spinal lesions that might lead to spinal tethering, such as a thick filum terminale, dermal sinus tract, or lipoma, were also surgically treated in both types of SSCMs. In the past 3 years, the filum terminale was explored even if magnetic resonance (MR) imaging did not show the presence of a thick filum.

Split spinal cord malformations were found and treated in some patients who underwent surgical repair of a myelomeningocele shortly after birth. These patients did have postoperative radiological workup. Follow-up evaluation of all patients ranged from 2 months to 12 years (mean 30.4 months). Fisher's exact test, chi-square, and a t-test were used in statistical analyses.

RESULTS

Patient Age and Gender

Patients ranged in age from less than 1 day to 12 years (mean 33.08 months). There were 46 girls (62%) and 28 boys (38%). The female predominance in these cases was more remarkable in Type I SSCMs than in Type II SSCMs. Girls comprised 66% of the patients with a Type I SSCM and 50% of those with Type II SSCM; however, this difference was not statistically significant (p = 0.268). The mean age of patients with neurological deficits and orthopedic deformities (43.2 months) was significantly older than those without deficits (8.2 months) (p = 0.003).
Symptoms and Signs

The presenting symptoms can be summarized as skin lesions, spina bifida aperta, scoliosis or kyphoscoliosis, sphincter disturbance, foot deformities and weakness, and/or atrophy in the lower extremities (Table 1).

<table>
<thead>
<tr>
<th>Symptoms &amp; Signs</th>
<th>No. of Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>skin stigmata</td>
<td>41 (54)</td>
</tr>
<tr>
<td>orthopedic deformities of foot</td>
<td>22 (30)</td>
</tr>
<tr>
<td>spina bifida aperta (myelomeningocele, hemimyelomeningocele, meningocoele)</td>
<td>29 (9)</td>
</tr>
<tr>
<td>weakness in lower extremities</td>
<td>20 (27)</td>
</tr>
<tr>
<td>scoliosis</td>
<td>12 (16)</td>
</tr>
<tr>
<td>bladder &amp; bowel disturbances</td>
<td>7 (9)</td>
</tr>
<tr>
<td>short &amp;/or thin leg</td>
<td>4 (5)</td>
</tr>
<tr>
<td>back pain</td>
<td>1 (1)</td>
</tr>
<tr>
<td>physical &amp; neurological signs</td>
<td></td>
</tr>
<tr>
<td>skin findings</td>
<td></td>
</tr>
<tr>
<td>hypertrichosis</td>
<td>32 (43)</td>
</tr>
<tr>
<td>capillary hemangioma</td>
<td>9 (12)</td>
</tr>
<tr>
<td>hyperpigmentation</td>
<td>7 (9)</td>
</tr>
<tr>
<td>dimple</td>
<td>5 (7)</td>
</tr>
<tr>
<td>subcutaneous lipoma</td>
<td>4 (5)</td>
</tr>
<tr>
<td>orthopedic deformities</td>
<td></td>
</tr>
<tr>
<td>scoliosis, kyphoscoliosis</td>
<td>22 (30)</td>
</tr>
<tr>
<td>unilat leg atrophy</td>
<td>15 (20)</td>
</tr>
<tr>
<td>pes echinovarus</td>
<td>10 (14)</td>
</tr>
<tr>
<td>pes cavus</td>
<td>6 (8)</td>
</tr>
<tr>
<td>pes valgus</td>
<td>3 (4)</td>
</tr>
<tr>
<td>pes calcaneovarus</td>
<td>2 (3)</td>
</tr>
<tr>
<td>trophic ulcers in foot &amp; or spontaneous</td>
<td>3 (4)</td>
</tr>
<tr>
<td>amputation of toes</td>
<td></td>
</tr>
<tr>
<td>neurological findings</td>
<td></td>
</tr>
<tr>
<td>paraparesis</td>
<td>19 (26)</td>
</tr>
<tr>
<td>unilat leg paresis</td>
<td>15 (20)</td>
</tr>
<tr>
<td>bladder &amp; bowel dysfunction</td>
<td>14 (19)</td>
</tr>
</tbody>
</table>

The duration of symptoms ranged from birth to 12 years (mean 21.7 months). The duration of symptoms was longer in the patients with neurological deficits and orthopedic deformities (mean 28.3 months) than in those without deficits (mean 8.2 months). This difference was found to be statistically significant (p = 0.01). Twenty-nine patients presented with spina bifida aperta (myelomeningocele, hemimyelomeningocele, or meningocoele); six other patients who had been born with spina bifida aperta underwent surgery for myelomeningocele or meningocoele before admission to our department. Hypertrichosis was the most common skin finding and was present in 32 patients. Capillary hemangioma, hyperpigmentation, and dimple were other skin stigmata. Some patients had more than one skin lesion and 36 patients (49%) had no skin lesions. Eleven of 35 patients with spina bifida aperta exhibited skin lesions, although 27 of 39 patients without spina bifida aperta exhibited these lesions (p = 0.004). Twenty-three patients presented with orthopedic deformities of their lower extremities. These orthopedic deformities included: pes echinovarus (10 patients), pes cavus (six patients), pes valgus (three patients), pes calcaneovarus (two patients), short leg (three patients), and trophic ulcers and/or
spontaneous amputation of toes (three patients). Some patients had more than one deformity. Scoliosis was the sole presenting symptom in six children. Paraparesis was present in 19 patients; however, 15 of these patients had a myelomeningocele. Unilateral leg paresis and atrophy were detected in 15 patients and bladder and/or bowel incontinence was present in 15 patients (Table 1). The proportion of patients with neurological deficits or orthopedic deformities did not significantly vary with the type and level of SSCMs.

### TABLE 2

<table>
<thead>
<tr>
<th>Findings</th>
<th>No. of Patients</th>
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<tbody>
<tr>
<td>bifid lamina</td>
<td>70</td>
</tr>
<tr>
<td>widened interpediculate distance</td>
<td>63</td>
</tr>
<tr>
<td>bony median septum</td>
<td>32</td>
</tr>
<tr>
<td>scoliosis</td>
<td>28</td>
</tr>
<tr>
<td>bifid vertebra</td>
<td>20</td>
</tr>
<tr>
<td>blocked vertebra</td>
<td>17</td>
</tr>
<tr>
<td>hypertrophic lamina</td>
<td>7</td>
</tr>
<tr>
<td>hemi vertebra</td>
<td>7</td>
</tr>
<tr>
<td>kyphoscoliosis</td>
<td>6</td>
</tr>
<tr>
<td>narrowed intervertebral disc space</td>
<td>5</td>
</tr>
<tr>
<td>accessory lamina</td>
<td>2</td>
</tr>
<tr>
<td>sacral agenesis</td>
<td>2</td>
</tr>
<tr>
<td>hemilamina</td>
<td>1</td>
</tr>
<tr>
<td>fused ribs</td>
<td>1</td>
</tr>
</tbody>
</table>

**Radiological Findings**

Plain x-ray films of the spine were obtained in 70 patients. Magnetic resonance images, computerized tomography (CT) scans, myelograms, and CT myelograms were available in 38, 10, 29, and 15 patients, respectively. Plain x-ray films disclosed a number of deformities of the spine (Table 2). All patients had a bifid lamina, which was often located at a different level; a widened interpediculate distance at the level of SSCM was present in 63 patients. Of 51 patients with a Type I SSCM for whom a plain x-ray film was available, 32 showed a bone spur (Fig. 1 left).
Fig. 1. Left: Plain x-ray film showing a bone spur at L-2. Right: Coronal MR image showing a Type II SSCM with hydromyelia. Note the presence of a split filum terminale.

A low conus medullaris was detected on all but one MR image. Besides splitting of the spinal cord and other dysraphic lesions, such as myelomeningocele, meningocele, and lipoma, MR images also revealed a thick filum terminale in 11 patients, hydromyelia in 10, split vertebra in six, and rigid median septum in five patients (Fig. 1 right).

Computerized tomography could be used to diagnose only Type I SSCMs, because it was not possible to evaluate the intradural structures by means of plain CT scans. However, CT myelography was superior to other radiological tools in defining the type of SSCM (Fig. 2). The correlation between radiological and operative findings was evaluated in terms of the type of SSCMs. The false-positive rates were 17% on MR imaging, 3% on myelography, and 0% on CT myelography. However, these differences were not statistically significant (p = 0.071). In one patient with composite SSCMs, MR imaging did not show a second Type II SSCM at the S-2 region in addition to the first Type I SSCM at T-8. The second SSCM was found when we performed exploratory surgery for a thick filum terminale because the conus medullaris was very low on MR imaging.
We also analyzed the presence of a thick filum terminale on MR imaging and at surgery. Both MR and operative findings were the same in 10 patients. False-positive and false-negative findings on MR imaging were present in one and 10 patients, respectively. Sometimes, it was very difficult to identify the filum terminale and the level of the conus medullaris, particularly on MR images obtained in children with severe scoliosis.

**Length of the Split Segment**

The length of the split segment of the spinal cord was longer in patients in whom a diagnosis was made after they reached 1 year of age (mean 3.05 vertebral segments) than in those in whom it was made before they reached 1 year of age (mean 2.55 vertebral segments). The split length was not significantly different in both types of SSCMs.

**Operative Findings**

Fifty-two patients had a single Type I and 18 a single Type II SSCM; four patients had composite SSCMs. Both hemicords were equal in size in 51 patients. An osseocartilaginous septum was detected in Type I SSCMs and a fibrous septum in Type II SSCMs. Fibrous bands and/or paramedian dorsal roots from the medial aspects of the hemicords to the dural sleeve were present and cut in all cases of Type I SSCMs. All Type II SSCMs had a fibrous median septum and/or paramedian dorsal roots that tethered the spinal cord, particularly at the caudal end of the split. We detected ventral paramedian roots in only three cases. In 21 patients, one hemicord was smaller than the other. The hemicords reunited after splitting in 63 patients, but did not reunite in 11 patients. In patients with an asymmetric involvement in the lower limbs, the smaller hemicords were almost always associated with an atrophic lower limb on the same side (\( p = 0.005 \)). However, some atrophic legs were associated with symmetrical hemicords. In four patients, composite SSCMs were present. Two Type I SSCMs located in tandem were detected at T-12 and L-1 in one patient. Another patient had a Type I SSCM at T-8 and a Type II SSCM at S-2. The two SSCMs were separated by a normal spinal cord. An unusual type of composite SSCM was observed in a 3-month-old boy.[5] At the T-11 level, the patient seemed to have a single dura dorsally. The dermal sinus tract penetrated the dorsal dura and was connected to a fibrous median septum, attaching the median aspects of the both hemicords. A small bony septum, which had a conical shape and was covered with a dural sheath, was also noted ventrally. This composite SSCM had a bony septum and two dural
tubes located ventrally, as well as a fibrous septum and a single dural tube located dorsally at the same level. Another patient had the same composite type of SSCM associated with a dorsal lipoma, a dermoid cyst, and a dermal sinus tract (Figs. 3 and 4).

Fig. 3. a: Axial MR images showing two hemicords and a dorsal lipoma and cyst. b: Computerized tomography scan. Note the bone spur. c: Coronal and sagittal MR images showing a cyst and a lipoma.
Fig. 4. a: Schematic drawing of surgical findings showing a Type I SSCM located ventrally and a Type II SSCM dorsally. Note a dorsal lipoma and a dermoid cyst associated with a dermal sinus tract. b-d: Photomicrographs. H & E, original magnification 20. b: Mature adipose tissue. c: Cyst lining of stratified squamous epithelium is filled with concentric layers of keratinous material. Skin adnexa are also present. d: Dermal sinus tract is lined by granulation tissue.

**Level of SSCMs and Conus Medullaris**

There were no cervical SSCMs in this series. The upper level at which SSCMs were found was at T-3 and the lowest was at S-2. The majority of SSCMs were located in the lumbar region (Fig. 5 upper). The conus medullaris was located below L-2 in all but one patient (Fig. 5 lower).
Sixty-two patients had at least one associated spinal lesion that could lead to spinal cord tethering (Table 3). However, of 12 patients with no associated spinal lesion, two had a composite type of SSCM; that is, more than one SSCM at different levels. Therefore, 85% of the patients had more than one spinal lesion. A thick filum terminale was the most common associated spinal lesion and was seen in 30 patients. Myelomeningocele and meningocele followed the thick filum terminale in frequency. Myelomeningocele was often associated with a Type I SSCM. Of 24 cases of SSCM with associated myelomeningocele or hemimyelomeningocele, only one was a Type II SSCM. However the predominance of a Type I SSCM in the other patients without myelomeningocele was less noteworthy (p = 0.019). The bony septum was caudal to the neuroplacode in two patients and rostral in 22 patients with myelomeningocele. Myelomeningocele manqué, dermal sinus tract, lipomyelomeningocele, teratoma, dermoid cyst, and intradural arachnoid cyst constituted the other associated spinal lesions. Occipital encephalocele, craniosynostosis, and dural arteriovenous malformation of the posterior fossa were encountered in three patients with SSCMs. Hydrocephalus in cases of a myelomeningocele associated with an SSCM was not
Complications of Treatment

Transient postoperative complications were seen in 17 patients. All of these complications improved within 3 weeks. The transient complications consisted of cerebrospinal fluid (CSF) fistula, transient leg paresis, wound infection, urinary retention, subcutaneous CSF collection, and dysesthesia in the legs (Table 4). Permanent complications consisted of renal failure due to a neuropathic bladder in one patient and spinal arachnoiditis caused by the contrast material used in myelography in two patients. Those patients with spinal arachnoiditis also had paraparesis.

Pathological Findings

Pathological examinations of the specimens from the median septa revealed fetal renal tissue, tubular epithelia, lymphoid tissue,[5] dermoid cyst, muscle tissue, ganglion, and blood vessels in addition to bone and cartilage in Type I SSCMs and fibrous tissue in Type II SSCMs (Fig. 6). One of the patients with a Type I SSCM at L-2 had an intradural teratoma located at the cervicothoracic region. Endodermal, ectodermal, and mesenchymal structures detected within the median septa support the theory of endomesenchymal tract and ectoendodermal adhesion.
Patient Outcome

There were two deaths in this group of patients. A patient who had a myelomeningocele and an SSCM was found dead in his bed, presumably as a result of apnea. Another infant died of ventriculitis. Of five patients whose conditions deteriorated during the long-term follow up, two had spinal arachnoiditis, and three had scoliosis. The majority of the patients remained stable and clinical improvement was observed in 18 patients.

DISCUSSION

Split spinal cord malformations are uncommon congenital anomalies that are seen primarily in females.[3,9,10,15-17,19,22,24,25,36] The female predominance was not marked in cases of Type II SSCMs reviewed in this series. Skin lesions are very common in cases of SSCMs. Hypertrichosis was the most frequent skin manifestation, as has been reported in many series.[3,9,15-17,19,22,24,25,31,36] It is significantly more common in SSCMs than in other types of dysraphism. Other skin lesions found in patients with SSCMs include capillary hemangioma, nevus, skin dimple, and subcutaneous mass.[3,9,10,15-17,19,22-25,36] No skin lesions were found in 36 patients (49%) in this series. More than half of our patients had skin lesions that should warn the physician of the possibility of an occult spinal dysraphism, particularly SSCM. As Pang[24] has indicated, we also found that the incidence of skin lesions in patients with myelomeningocele was significantly less than in those without myelomeningocele. He suggested that skin lesions such as hypertrichosis, capillary hemangiomas, and dermal sinus tracts represent minor aberrations in the development of surface ectoderm that result from the adverse influence of a dorsal endomesenchymal tract, but that these aberrations might be largely overshadowed by chaotic changes in the surface ectoderm occasioned by the unneurulated neural plate in the case of an associated myelomeningocele.[24]

Scoliosis is not uncommon in patients with SSCM.[3,9,15-17,19-24,36] In 1974, Winter, et al.,[36]
reported a 4.9% incidence of diastematomyelia associated with congenital scoliosis. In 1984, McMaster[21] determined that diastematomyelia was the most common anomaly (16.3% of patients) in patients with congenital scoliosis. It is likely that the reported incidence of SSCMs will be higher as noninvasive and sophisticated radiological tools become more widely used. Scoliosis is usually seen in 30 to 60% of patients with SSCM.[3,10,11,15,16,19,23,24] However, Miller and associates[22] reported a 79% incidence of congenital scoliosis in their patients with diastematomyelia. Hilal and colleagues[15] reported that the risk of scoliosis increases significantly in older children with diastematomyelia. James and Lassman[18] and Guthkelch[11] determined the orthopedic and neurological syndromes in children with diastematomyelia. The orthopedic syndromes consist of foot deformities and dwarving of one leg with muscle atrophy in addition to spine deformity.

Asymmetrical paresis of the lower limbs is not rare in SSCMs.[11,17,19,22,24,26,36] In our series, most children with myelomeningocele had paraparesis. Sphincter dysfunction has been described in cases of SSCMs.[3,10,12,17,19,23,24] Bladder and bowel dysfunction had been present since birth in seven patients in our series. In the series described by Pang,[24] all adult patients had either new or worsening bladder symptoms, whereas none of 16 symptomatic children had deterioration of bladder function. Although pain is a dominant feature in adult patients, only one of our patients complained of back pain.[27,31]

Unlike Miller, et al.,[22] we found no significant association between the level of the lesion and its neurological manifestations. Miller, et al., noted incontinence of the bowel in only five patients who had a diastematomyelia in the lumbar spine. In serial follow-up examinations, neurological deterioration was described in patients with SSCM who did not undergo surgery. This deterioration was considered to have been caused by tethering of the spinal cord secondary to growth of the spinal canal.[22] We also found that both the patient's age and the duration of symptoms in patients with deficits were significantly greater than in those without deficits. Therefore, we may conclude from these data that the risk of having neurological deficits increases with age. In asymptomatic adult patients, symptoms and neurological deficits may arise following trauma or strenuous exercise.[7,31]

The bone deformities seen on plain x-ray films obtained in patients with SSCMs are many: scoliosis, bifid lamina, widened interpediculate distance, hemivertebra, bifid vertebra, fused vertebra, and narrowing of the intervertebral disc space have all been observed.[3,13,15-17,19,23,24,36] Of 70 patients in whom a plain x-ray film was obtained, all had a bifid lamina, which was often located at a different level, and 63 patients had a widened interpediculate distance at the level of the SSCM. The spinous process is prominent in the area of the bone spur, especially if there is an intersegmental fusion of adjacent laminae.[15] The bone spur can be identified on plain x-ray films in patients with a Type I SSCM. In infants, ultrasonography has been used in diagnosing SSCMs, even in the prenatal period.[8,35,37] Computerized tomography myelography and MR imaging are needed for delineating the type of SSCMs and for showing the associated lesions. Computerized tomography myelography is superior to MR imaging in defining the type of SSCMs.[13,23,24] Magnetic resonance imaging has the advantage of being noninvasive and of displaying hydromyelia and defining the content of a lesion such as a neurenteric or dermoid cyst.

Because the majority of patients had at least one tethering lesion unrelated to the SSCM, the whole spinal neuraxis should be included in the screening MR image.[24] In one of the patients in this series who had composite SSCMs, MR imaging did not show the second Type II SSCM at the S-2 region.
In all types of SSCMs, a rigid or fibrous septum and the fibrous bands and/or dorsal paramedian roots transfixing the spinal cord were observed at surgery. The fibrous septa are not usually seen on CT myelograms and MR images. Pang[24] was able to show the fibrous septa in only five of 18 cases of Type II SSCM preoperatively. A smaller hemicord was almost always associated with an atrophic leg on the same side in cases with asymmetrical hemicords. However, all asymmetrical legs were not associated with a smaller hemicord. In two of our cases with hemimyelomeningoceles, the hemineural placode was segmental and smaller than the other hemicord and the child had a discrepancy between the lower limbs. An oblique septum has been shown to divide the neural plaque into a large and a small hemicord, the latter corresponding to the side of poor function.[24] In the small hemicords, it has been pathologically shown that the gray horns were rudimentary and the white funiculi were disorganized.[4,29] The hemicords are usually reunited after splitting, but rarely do they not rejoin.[13,15] In 11 of our patients, the hemicords did not rejoin after splitting.

Composite SSCMs are very rare.[10,13,19,24] In our series, one patient with composite SSCMs had a Type I SSCM at T-8 and a Type II SSCM at S-2. Those two SSCMs were separated by normal spinal cord. This results from two separate loci of ectoendodermal adhesions and endomesenchymal tracts, according to Pang.[25] Another patient had two Type I SSCMs in tandem at T-12 and L-1. Two patients with composite SSCMs had a Type I SSCM located ventrally and a Type II SSCM dorsally at the same level. In both patients, this composite type of SSCM was associated with a dermal sinus tract. The upper level of septa was at T-3 and the lowest was at S-2. The majority of septa were localized at the lumbar region, as in other series.[3,9,10,13,15-17,19,22-24,31,36] Cervical and sacral locations are extremely rare. The conus medullaris is usually located caudal to L-2.[13,15,17,23,24,26] In this series, all but one patient had a conus below L-2. The length of the split segment was greater in patients diagnosed after reaching 1 year of age than in patients diagnosed before reaching 1 year of age, as Pang[24] has indicated. The mean split length is also greater in Type I than in Type II SSCMs. Pang[24] has hypothesized that the neural tube is firmly transfixed by a rigid derivative of the Type I endomesenchymal tract and that its subsequent ascent results in a long cleavage in the spinal cord. However, the difference between the means of split lengths in both types of SSCMs was not statistically significant in our series. Eighty-five percent of our patients had at least one associated lesion. A thick filum terminale was the most common associated lesion and was seen in 30 patients. Some cases in which the filum terminale has not been divided may show neurological deterioration due to the tethered cord syndrome.[12] Hemimyelomeningocele and myelomeningocele were present in three and 21 patients, respectively. The reported spinal lesions in association with SSCMs are a thick filum terminale, myelomeningocele, meningocele, lipomyelomeningocele, limited dorsal myeloschisis, teratoma, neurenteric cyst, lipoma, dermal sinus tract, dermoid cyst, epidermoid cyst, arteriovenous malformation, epidural venous angioma, and arachnoid cyst.[1-3,10,12,13,17,19,22-24,26,31,36] The incidence of SSCMs in patients with a myelomeningocele has been reported to be 78% by Emery and Lendon.[4] We did not find such a high rate of SSCM in our patients with a myelomeningocele, but they were not routinely screened for SSCM until 3 years ago. The rate of SSCMs in patients with spinal dysraphism was 5% in the series reported by Pang.[24] We have screened all patients with spinal dysraphism for SSCMs for 3 years and 13% of our patients with a myelomeningocele had an SSCM. A Type I SSCM was more commonly associated with a myelomeningocele than a Type II SSCM. The myelomeningocele is usually located below the bony septum.[4,17,21,23,24] Pang found at least one unrelated tethering lesion in all lumbosacral and lower thoracic SSCMs and in a much smaller number of cervical SSCMs. This researcher has suggested that the entire neuraxis should be studied radiographically and, if a second
Pathological examinations of specimens of median septa disclosed interesting results. Fetal renal tissue, lymphoid tissue, and tubular epithelia have been found within the septa in two cases of SSCM that were previously reported.[5] Blood vessels, muscle tissue, and dermoid cysts were microscopically present in the septa in other cases, as in the series reported by Pang, et al.[26] Pluripotential cells of the endomesenchymal tract could develop into a variety of tissues. Cases of SSCM associated with Wilms' tumor[6] and ectopic renal tissues have been reported.[7,32] Tibbs, et al.,[33] reported five cases of midline hamartomas masquerading as myelomeningoceles or teratomas. These findings may implicate the accessory neurenteric canal, a basic error involved in the development of all SSCMs. Ganglion cells within the fibrous septum were also detected.[26,30]

An immediate neurological deterioration in patients is rarely seen after surgery.[9,24,25] A microneurosurgical technique and a high-speed drill are helpful in avoiding spinal cord injury. All complications such as leg paresis, CSF fistula, urinary retention, and dysesthesia seen in the early postoperative period were conservatively managed and improved within 3 weeks. Deaths are generally unrelated to the surgery of the SSCM. One of the patients in the series of Goldberg and associates[9] died of meningitis that developed in the presence of a CSF fistula. The aim of surgery should be to untether the spinal cord and prevent neurological and orthopedic deterioration. The majority of patients with SSCMs will be either stable or better following surgery; however, some will continue to worsen.[3,10,12,17,22,24,31,36] Screening should be obtained for another lesion in those cases found to deteriorate.

**CONCLUSIONS**

In reviewing and analyzing the records of 74 cases with SSCMs, we reached the following conclusions.

1) The classification of SSCMs proposed by Pang, et al.,[26] will eliminate the current chaos in terminology. The terms "Type I" and "Type II" SSCMs should be used instead of diastematomyelia and diplomyelia, respectively.

2) Magnetic resonance imaging is the radiological tool of choice for screening for SSCMs. However, CT myelography is superior to MR imaging in defining the type of SSCM that has been located.

3) In all SSCMs reviewed in this series, either a rigid or a fibrous septum was found to transfix the spinal cord. In addition, at least one unrelated lesion was found to tether the spinal cord in 85% of the patients.

4) The risk of neurological deficits in cases of SSCMs increases with the age of the patient. Therefore, all SSCMs should be surgically treated when diagnosed, especially before the development of orthopedic and neurological manifestations.

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**Dedication**

This paper is dedicated to Professor Erdem Tunçbay, our teacher and the founder of the Department of Neurosurgery at Ege University.
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