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Meningocele Manqué: Radiologic Findings with Clinical Correlation

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Purpose: To determine whether meningocele manqué can be detected by neuroimaging techniques in dysraphic patients. **Methods:** We reviewed the records and imaging studies of 16 patients with surgically proved meningocele manqué seen at our institution between 1989 and 1990. Both CT and MR imaging techniques were used. CT of the spine was performed immediately following contrast myelography. **Results:** Nine of 16 patients (CT, four; and MR, five) showed evidence of meningocele manqué which corresponded to intraoperative findings. Fourteen of 16 patients were found to have diastematomyelia, eight with medium septum and six without a septum. Associated findings included syrinx (six), lipoma (five), dermoid cyst (one), and neuroenteric cyst (one). After completing this review, we were able to prospectively diagnose dorsal bands in two new patients; these bands were confirmed at surgery. **Conclusion:** Dorsal bands can be detected in dysraphic patients with CT or MR using operative findings as a road map.

Index terms: Diastematomyelia; Spina bifida; Spinal cord, tethered cord; Spinal cord, computed tomography; Spinal cord, magnetic resonance

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Meningocele manqué (MM) is the name given to single or multiple dorsal bands composed of fibrotic or atretic neural tissue found in dysraphic patients; they are discovered incidentally during surgical exploration for occult spinal dysraphism. James and Lassman (1, 2) encountered many examples of this structural abnormality named "meningocele manqué" to designate a type of meningocele that failed to develop, as opposed to one that developed completely and then became atretic. The word "manqué" is defined in the Shorter Oxford English Dictionary as "that which might have been but is not." While these "bands" are most frequently found at the site of clinical tethering, either by filum terminale or by associated dysraphic states including diastematomyelia, dermoid cysts, and neuroenteric cysts, MM can exist at some distance from the obvious clinical tether site. Its clinical importance stems largely from the possibility that such a dorsal band might be missed at the time of surgery with

persistence of the tethering syndrome. The orthopedic, radiologic, and neurosurgical aspects of MM are described in detail by James and Lassman (1, 2). The purpose of our review was to identify the basic anomaly from documented examples at surgery, and then, review the neuroimaging studies to see if the documented MMs could be detected.

Subjects and Methods

Between July 1989 and February 1990, 16 patients (seven males and nine females) between the ages of 16 days and 38 years underwent surgical exploration for suspected occult spinal dysraphism and, among other anomalies, were found to have manifestations of MM. Median age was 12 years; mean age was 11 years. The detailed operative reports were studied and correlated with the imaging studies that were reviewed retrospectively.

Ten patients had computed tomography (CT) scans. All CT studies were performed immediately following myelography which—in the past few years—has been performed at our institution using iopamidol for intrathecal contrast.

Nine patients had MR studies performed on a 1.5 T superconducting magnet. The spin-echo pulse sequence utilized was 500/20/4 (TR/TE/excitations). Axial slice thickness was 3 or 4 mm with an intergap measurement of 1 mm. This was the standard protocol used when searching for developmental anomalies and additional sequences were only obtained if an intradural lesion was also noted. This occurred in one patient with an intradural

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extramedullary lesion (which was not of fat intensity) for which additional sequences 2000/30 and 2000/80 were obtained. We specifically looked for linear filling defects on the postmyelo CT scans and thin soft-tissue bands on the magnetic resonance (MR) scans in the axial projection. When such soft-tissue linear bands were detected, we attempted to separate these from abortive midline septas.

Three patients had both CT and MR studies. Myelograms were not reviewed in this study; however, the findings on plain films were noted.

After the review was completed, we were able to diagnose dural bands in two new patients; the bands were confirmed at surgery.

Results

Clinical

Table 1 lists the presenting signs and symptoms in our group of patients with surgically proven MM. There are no specific signs or symptoms that could serve as an indicator of MM, since these findings could be present in any dysraphic condition that causes spinal cord tethering.

Imaging

All of the patients' plain films of the spine showed lumbosacral osseous abnormalities. These consisted of posterior element defects

(most common), segmentation anomalies, widened interpediculate distance, and even sacral agenesis in two patients. These findings on plain film raise the possibility of occult spinal dysraphism.

MM could be suggested on axial CT images with intrathecal contrast in four of 10 patients. The most common finding was a thin linear filling defect, extending from the spinal cord or hemicord to the dorsal dura. In one patient, there was a dorsal band from a hemicord to the dorsal dura, and a proximal cauda equina nerve root was adjacent to the dura. At surgery, an adhesion was found between the nerve root and the dorsal dura (Fig. 1). One of the youngest patients in our series was a 16-day-old girl who was noted to have less spontaneous movement of the left leg and bilateral absence of deep tendon reflexes at the knees, as well as hypertrichosis at the lumbosacral region. CT imaging revealed a thin left-sided posterolateral intradural filling defect, which was a surgically proven dorsal band at the upper lumbar level (Fig. 2).

On axial T1-weighted MR images, the dorsal bands appeared as small, thin linear structures of equal signal intensity to the spinal cord, and extended from the dorsal aspect of the spinal cord or hemicord to the dorsal dura (Figs. 3 and 4). While nine of 16 underwent MR imaging, only five of the nine studies showed findings suggestive of MM. Three patients had both CT and MR studies and two and three patients showed evidence of MM on MR only, and the other patient showed evidence only on CT.

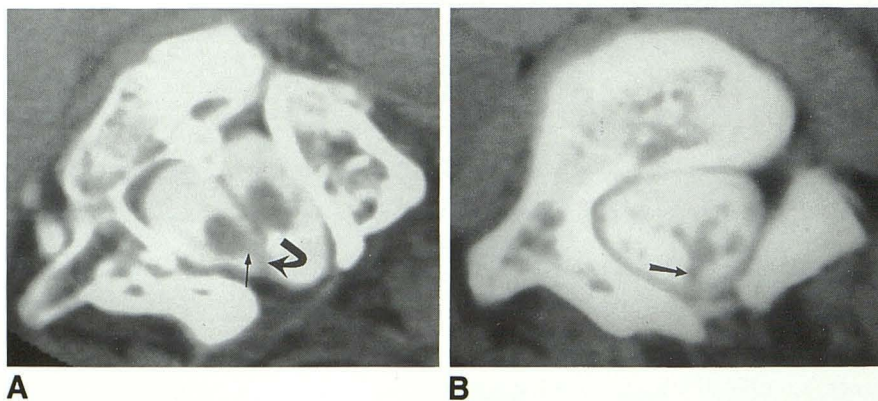
Associated abnormalities either observed radiologically or seen at the time of surgery are listed in Table 2. In one patient, there were two areas of diastematomyelia with osseous septums (mid-thoracic and lower lumbar) and another patient

TABLE 1: Presenting signs and symptoms in MM patients

Sign/Symptom	No. of Patients (n = 16)
Cutaneous stigmata	9
Progressive lower extremity neurologic deficits	8
Lower extremity atrophy	4
Foot deformities	4
Urinary and fecal inconti- nence	2
Sexual impotence	1

Fig. 1. A, Axial CT section at T12 level immediately following the myelogram in a 16-year-old girl shows diastematomyelia with a fibrous septum and a dorsal band (arrow) extending from the right hemicord to the fibrous septum (curved arrow) at the level of a laminal defect.

B, Axial CT section at L2 level in the same patient shows an aberrant dorsal nerve root on patients right (arrow) extending to the dorsal dura. Surgery verified the dorsal band.



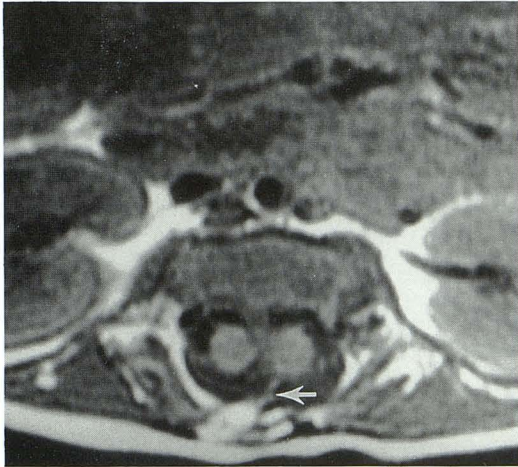


Fig. 2. This axial T1-weighted MR image at the L1 level from a 2-month-old girl shows a dorsal band (arrow) extending from the left hemicord to a pathologically proven dermoid cyst. Note that this dorsal band originates from dorsal medial surface of the hemicord, classical for the dorsal bands seen with diastematomyelia in our patients. However, a dermal sinus terminating in the left hemicord could not be ruled out on neuroimaging alone.

had three areas of diastematomyelia with osseous septums (one thoracic and two lumbar). There was one patient with the conus located at the first lumbar level and a normal filum as determined by sagittal and axial T1-weighted MR images who, at surgery, was found to have intradural dorsal bands, causing some tension on the conus. These bands were not seen on MR nor were there any other abnormalities detected. MR was able to detect multiple coexisting abnormalities in two patients (Fig. 4).

Surgical

Cutaneous stigmata (one in particular that was tender to touch) and the neuroimaging studies led to surgical exploration. The majority of the young patients had a suggestive sign or symptom and the older patients presented with progressive sensory or motor neurologic deficit. At surgery, 10 patients had dorsal bands (atretic dorsal nerve

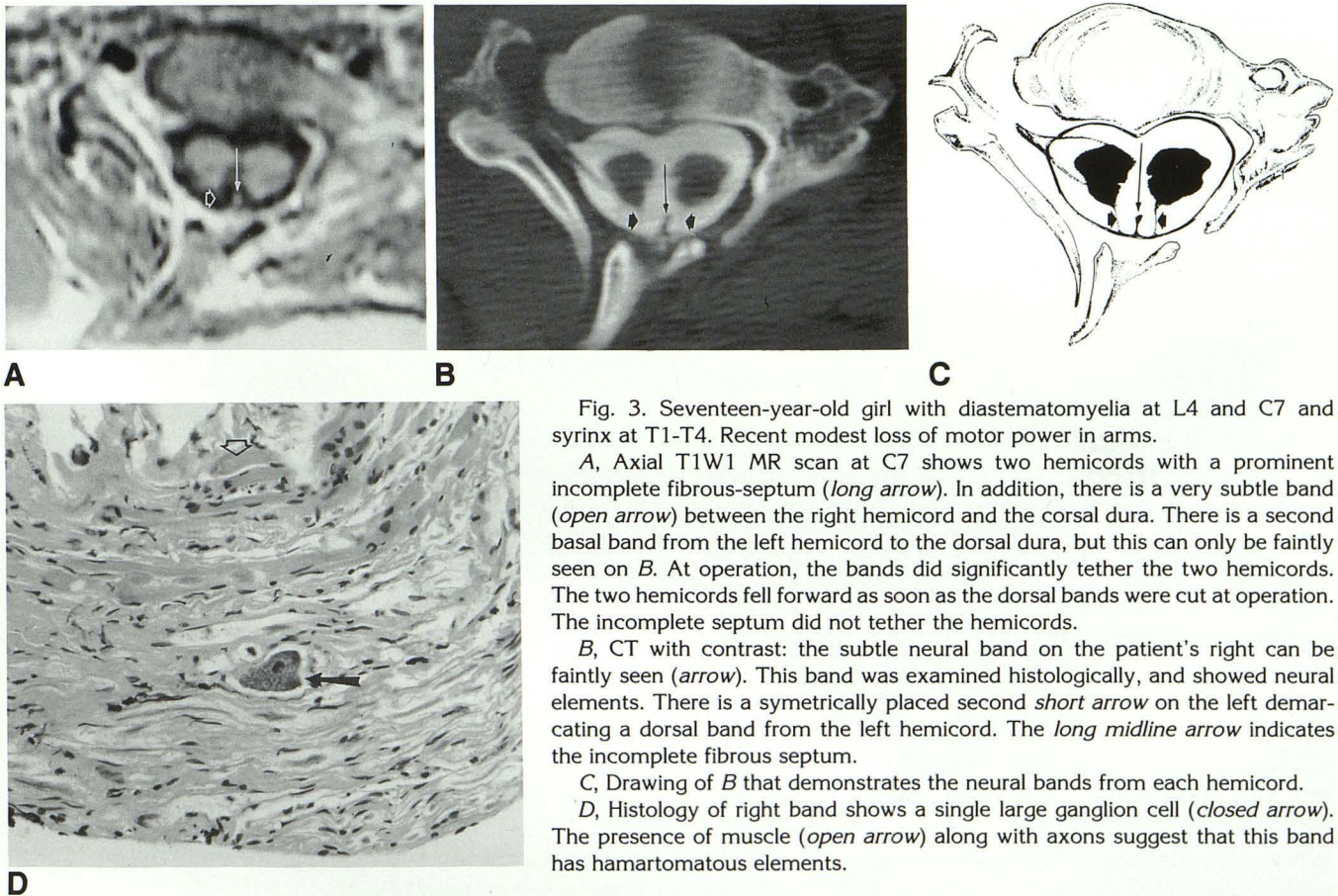


Fig. 3. Seventeen-year-old girl with diastematomyelia at L4 and C7 and syrinx at T1-T4. Recent modest loss of motor power in arms.

A, Axial T1W1 MR scan at C7 shows two hemicords with a prominent incomplete fibrous-septum (long arrow). In addition, there is a very subtle band (open arrow) between the right hemicord and the dorsal dura. There is a second basal band from the left hemicord to the dorsal dura, but this can only be faintly seen on B. At operation, the bands did significantly tether the two hemicords. The two hemicords fell forward as soon as the dorsal bands were cut at operation. The incomplete septum did not tether the hemicords.

B, CT with contrast: the subtle neural band on the patient's right can be faintly seen (arrow). This band was examined histologically, and showed neural elements. There is a symmetrically placed second short arrow on the left demarcating a dorsal band from the left hemicord. The long midline arrow indicates the incomplete fibrous-septum.

C, Drawing of B that demonstrates the neural bands from each hemicord.

D, Histology of right band shows a single large ganglion cell (closed arrow). The presence of muscle (open arrow) along with axons suggest that this band has hamartomatous elements.

Fig. 4. *A*, This axial T1-weighted image at the L2 level is from a 38-year-old man who presented with increasing sexual impotence and lower lumbar hypertrichosis. There is a small band (*arrowhead*) extending from the right hemicord to the dura. The neuroenteric cyst seen on *B* was not visualized on this image as it is at a higher level. The thecal sac is also capacious.

B, T1-weighted MR sagittal image from the same patient reveals an intradural lesion (*arrow*) at L3 levels of less signal intensity than the cord but of greater signal than the cerebrospinal fluid. This was a neuroenteric cyst. There is a small high intensity lipoma on the dorsal cord just caudal to the cyst, and a much larger lipoma in the dorsal epidural space L1-L3.

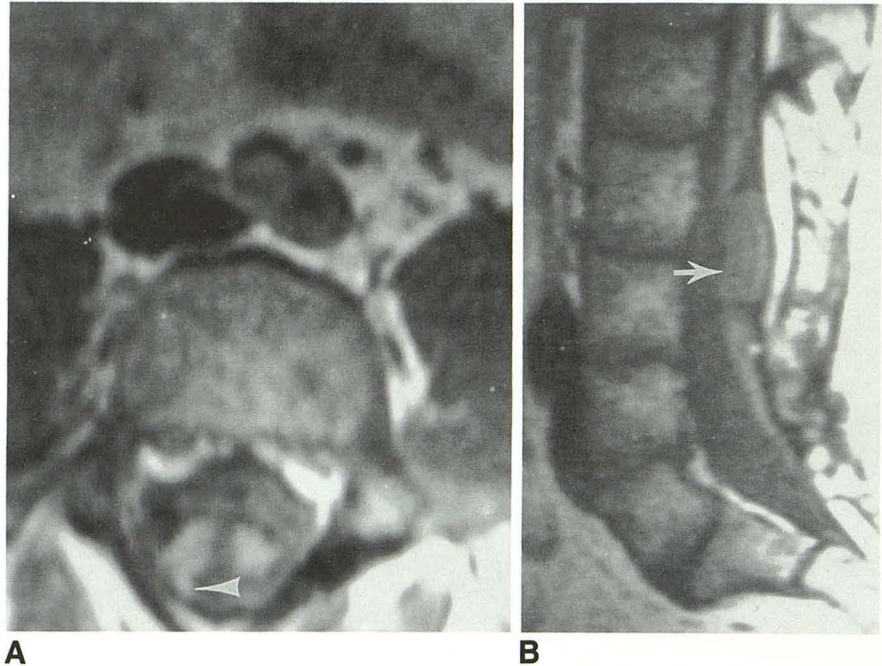


TABLE 2: Associated abnormalities seen with MM

Finding	No. of Patients (n = 16)
Diastematomyelia	14
without septum	6
with septum	8
Tethered cord	16
Syrinx	6
Lipoma	5
Neurenteric cyst	1
Dermoid cyst	1

roots or fibrotic bands) that extended from the dorsal aspect of the spinal cord to the dorsal dura (Figs. 3, 5, and 6). In six additional patients, these intradural bands pierced the dura and extended to the underside of the lamina or to the fibrotic tissue at the laminal defect. At the time of surgery, after surgical resection of these nonfunctioning atretic neural or fibrotic bands, the spinal cord fell to a more ventral relaxed position in the canal in four patients. Thus far, these patients preoperative signs and symptoms have not progressed.

After this review was completed, we were able to detect significant dorsal bands prospectively in two new patients. The first was a 17-year-old girl with diastematomyelia at C7 with recent modest loss of motor power in the upper extremities. While the diastematomyelia was not associated with a bony or fibrous septum, the cord was

tethered by dorsal bands on MR and CT images. These bands proved to be tethering the cord posteriorly to the dura at operation (Fig. 3). They were the only cause of tethering in this patient; after surgery, motor power returned to the upper extremities. The second patient was a 2-day old boy with an obvious meningocele at L5-S1. Dorsal bands extended from the terminal cord anteriorly through the dorsal meningocele defect to the posterior dura of the meningocele (see Fig. 5).

Discussion

As described by James and Lassman (1), the basic anomalies of MM are the so-called dorsal bands, which extend from the dorsal aspect of the spinal cord or a hemicord to the dorsal dura. These bands may either be entirely fibrotic or contain atretic nerve roots that usually originate from the medial aspect of the hemicords and course dorsally. The bands may penetrate the dura of an associated bony median septum or may simply exit dorsally; they occasionally pierce the dura and terminate on the undersurface of the laminae. At times, the bands may exit the dura and form tense bands to a midline cutaneous defect that may be quite tender to touch. Cutaneous stigmata may or may not be present. These bands seem roughly to fit into the type II split

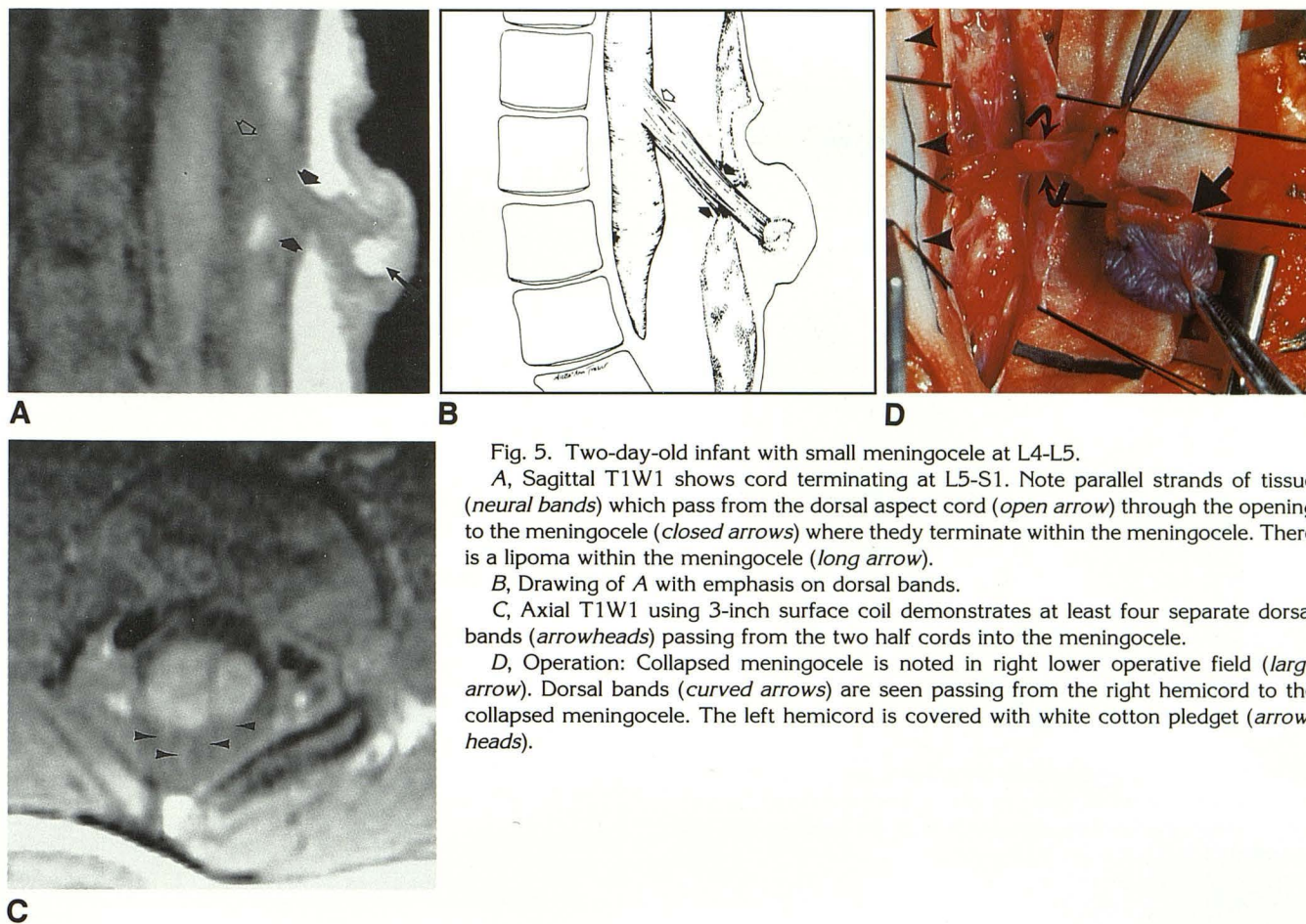


Fig. 5. Two-day-old infant with small meningocele at L4-L5.

A, Sagittal T1W1 shows cord terminating at L5-S1. Note parallel strands of tissue (neural bands) which pass from the dorsal aspect cord (open arrow) through the opening to the meningocele (closed arrows) where they terminate within the meningocele. There is a lipoma within the meningocele (long arrow).

B, Drawing of A with emphasis on dorsal bands.

C, Axial T1W1 using 3-inch surface coil demonstrates at least four separate dorsal bands (arrowheads) passing from the two half cords into the meningocele.

D, Operation: Collapsed meningocele is noted in right lower operative field (large arrow). Dorsal bands (curved arrows) are seen passing from the right hemicord to the collapsed meningocele. The left hemicord is covered with white cotton pledget (arrowheads).

cord malformation of Pang (3). When biopsied in this series, the tissue corresponded to roots that had been under tension for sometime and appeared to have no target tissue.

It was not the purpose of this retrospective analysis to compare CT and MR; we simply wished to determine if these dorsal bands could be observed using the operative findings as a road map. We feel this was successful, but there were limitations. For example, some of the earlier CT studies were performed on older scanners with poorer spatial resolution. CT studies should be performed only after water soluble contrast medium is injected intrathecally. Varying surface coils, fields of view, and slice thicknesses were used on MR studies. Perhaps with standardization and use of the most optimal resolution parameters (including axial gradient echo images) the bands may be better detected.

James and Lassman (1) found MM in 45 out of 200 patients who underwent surgical exploration for occult spinal dysraphism. Of the 45

patients with MM, there were 28 patients with associated diastematomyelia (18 without a septum and 10 with septum). In our studies, we found 14 of 16 patients with associated diastematomyelia (six without a septum and eight with a septum). It should be emphasized that MM is a common association with diastematomyelia (40% in our series); when surgically exploring or imaging patients with a known diastematomyelia, every attempt should be made to detect these bands. The bands are even more important in the patient without a median septum. In this situation, the clinician might be persuaded not to offer surgical intervention to a patient with appropriate symptomatology, because no tethering band could be demonstrated (Fig. 3). Based on our experience, there is a likelihood of finding functionally significant medial bands in patients with diastematomyelia without a median septum to warrant operative intervention if clinical symptoms match the level of the cord separation. In this series, despite our current high quality im-

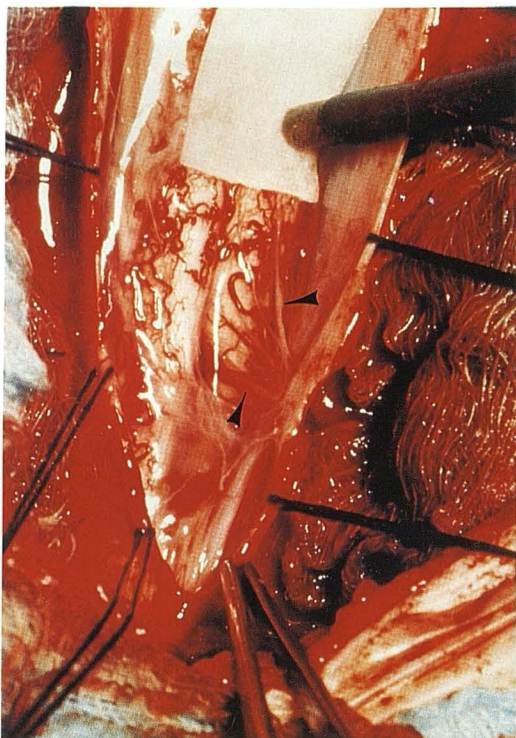


Fig. 6. Intraoperative photograph taken from the patient's left side with the patient in prone position. There are bands (arrowheads) extending from the dorsal aspect of the spinal cord to the dorsal dura.

aging techniques, a significant number of patients with clinical deterioration from the MM did not have these bands appreciated either prospectively or retrospectively. In future cases, gradient echo MR imaging will be used to improve the conspicuity of these bands.

There is very little information available in the radiologic literature concerning MM. Usually, there is a brief reference made to fibrous bands when discussing the topic of causes of spinal cord tethering (3, 4). We feel that this clinical entity can frequently be imaged with CT or MR. There are articles that now favor MR as the procedure

of choice for screening patients with spinal dysraphism (5, 6). If findings on MR studies are equivocal, then CT scanning following intrathecal administration of contrast should then be considered. One certain advantage of MR would be the ability to detect associated conditions, such as diastematomyelia with concurrent syringohydromyelia, or to help characterize a mass lesion, such as, in our case, of an intradural neuroenteric cyst (7, 8).

In summary, we emphasize the importance of these tethering bands. Although these bands usually occur at the site of diastematomyelia (with or without a median septum), they may occur with other forms of dysraphism (tethered filum) as well.

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