Terminal and nonterminal myelocystoceles

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Object. The purpose of this study was to report the author’s experience with 14 cases of terminal and nine cases of nonterminal myelocystoceles and to highlight the differences between these two groups in regard to the embryological origins, clinical presentation, operative findings, results on neuroimaging studies, and prognosis for these lesions.

Methods. This is a retrospective analysis of 14 cases of terminal and nine of nonterminal myelocystoceles treated between January 1998 and January 2006. All patients underwent neurological examination, plain x-ray films of the spine, computed tomography scans of the brain, and magnetic resonance (MR) imaging of the spine. In seven of these cases, MR imaging included three-dimensional constructive interference in steady-state sequences, and in four cases MR myelography was also done. Follow-up duration ranged from 3 months to 4 years.

Results. All of the patients with terminal myelocystoceles presented with swelling in the low back and varying degrees of neurological deficits, except four who had normal results on neurological tests. The MR images revealed classic features of terminal myelocystoceles in all patients. In each case, patients underwent excision of the meningocele sac and drainage of the syringocele with untethering of the spinal cord. During the last follow-up visit, there was no change in the neurological status of these children. In the nonterminal myelocystocele group, one lesion was cervical, six were thoracic, and two were lumbar lesions. All except one patient presented without neurological deficits; that patient had paraplegia with incontinence. Admission MR images revealed Rossi Type I nonterminal myelocystocele in six and Rossi Type II nonterminal myelocystocele in three patients. Children with Type I lesions underwent excision of the fibroneurovascular stalk and excision of the meningocele sac, whereas those with Type II lesions underwent drainage of the syringocele, untethering of the cord, and excision of the meningocele sac. There was no change in the neurological status postoperatively. During the follow-up period no patient in either group presented with retethering.

Conclusions. Myelocystoceles, both terminal and nonterminal, are different from other skin-covered masses in the back. A proper imaging evaluation is required to differentiate myelocystoceles from other skin-covered masses in this area, because the surgical treatment and prognosis are different for this subset of patients with occult spinal dysraphism. Terminal myelocystoceles are different from nonterminal ones embryologically, clinically, radiologically, surgically, and prognostically. These differences are discussed. (DOI: 10.3171/PED-07/08/087)

Key Words • magnetic resonance myelography • meningocele • myelocystocele • pediatric neurosurgery • spina bifida • spinal dysraphism • tethered spinal cord • three-dimensional constructive interference in steady-state imaging

Terminal myelocystoceles constitute approximately 5% of skin-covered lumbosacral masses and are characterized pathologically by the following features: 1) a skin-covered lumbosacral spina bifida; 2) an arachnoid-lined meningocele that is directly continuous with the spinal subarachnoid space; and 3) a low-lying hydromyelic spinal cord that traverses the meningocele and then expands into a large terminal cyst.10 This terminal cyst does not communicate with the subarachnoid space and is lined by ependyma and dysplastic glia. Terminal myelocystoceles are common in patients with cloacal exstrophy.4,7,11,16

Nonterminal myelocystocele is a very rare type of closed spinal dysraphism characterized by a skin-covered mass in the midline posteriorly, a narrow posterior spina bifida, a CSF-filled cyst, an expanded dural sheath (meningocele), and varying amounts of dorsal fat that are continuous with the subcutaneous fat of the back and skin.22 Nonterminal myelocystoceles are much less common than terminal myelocystoceles and are frequently misdiagnosed as meningoceles. Rossi and colleagues22 have classified nonterminal myelocystoceles into two types, as follows: Type I, a forme fruste type (myelocystocele manqué), in which the meningocele sac is traversed by a fibroneurovascular stalk that is attached to the dome of the sac; and Type II, a complete form in which there is a focal hydromyelia that has displaced the posterior wall of the spinal cord into the meningocele sac. The aim of the present study is to compare the clinical, radiological, surgical, and prognostic differences between terminal and nonterminal myelocystoceles treated by a single surgeon during a period of 8 years.
Clinical Material and Methods

This was a retrospective analysis of all cases of myelocystoceles treated by a single surgeon between January 1998 and January 2006. This study included two patients reported earlier. All patients underwent standard neurological evaluation, plain x-ray films of the spine, computed tomography scans of the brain (if MR imaging had not included brain screening), and MR imaging of the spine. The MR evaluation included standard sequences; MR myelography (TR 2800 msec, TE 1110 msec) and 3D-CISS imaging (TR 12 msec, TE 6 msec) sequences were performed in patients who underwent MR evaluation at our institution, whereas this was not available for patients referred from outside institutions who had undergone MR imaging at the referring hospital.

Patient Population

Fourteen patients with terminal myelocystoceles were treated during the study period. The clinical, neuroimaging, and outcome data in these patients are summarized in Table 1. Briefly, their ages ranged from newborn to 7 years, and there were 11 girls and three boys. During the same study period, nine cases of nonterminal myelocystoceles were treated (Table 2). The ages of these patients ranged from 10 days to 3 months, and all were girls. All patients underwent excision of the sac with untethering of the spinal cord. Neurological outcome during the last follow-up evaluation was recorded.

Results

Terminal Myelocystoceles

There was no positive prenatal history of myelocystoceles in this group of children. All of them presented with a swelling in the low back; the size of the swellings ranged from 3 to 15 cm in diameter. In nine of the patients the swelling had normal skin at the base and the sides, whereas the dome of the swelling was covered by dystrophic skin. None of the sacs leaked CSF or showed signs of impending CSF leakage. Ten patients exhibited varying degrees of neurological deficits, whereas four had none. Among the children in whom neurological evaluations yielded normal results, the four patients’ ages were 7 and 4 years, 1 month, and 19 days. Associated abnormalities included aqueductal stenosis in four patients and tonsillar ectopia and CM-II in two each. No patient in the study group had OEIS complex or vertebral, anal, tracheoesophageal, renal, and radial anomalies syndrome.

Surgery. All patients underwent excision with untethering; the sac was dissected up to its neck. The first lamina immediately rostral to the defect was removed. The dura mater was opened from the normal area and then the incision was extended to the dome of the meningocele sac. Arachnoidal adhesions and meningocele manqué were encountered in most of the cases, although the degree of adhesions varied from case to case. Nerve roots were often found passing anteriorly from the myelocystocele. The distended cord was detached from the sac, which often resulted in the

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age, Sex</th>
<th>Location</th>
<th>Neuro Status</th>
<th>Associated Anomalies</th>
<th>Neuroimaging Findings</th>
<th>Outcome (last FU)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>7 yrs, M</td>
<td>lumbosacral</td>
<td>normal</td>
<td>none</td>
<td>conus at L5–S1, lipomyelocele</td>
<td>normal (4 yrs)</td>
</tr>
<tr>
<td>2</td>
<td>1 yr, M</td>
<td>lumbosacral</td>
<td>incontinence</td>
<td>aqueductal stenosis</td>
<td>conus at L5–S1, myelocystocele</td>
<td>incontinence improved (18 mos)</td>
</tr>
<tr>
<td>3</td>
<td>4 yrs, M</td>
<td>lumbosacral</td>
<td>normal</td>
<td>tonsillar ectopia</td>
<td>conus at L5–S1, myelocystocele</td>
<td>normal (6 mos)</td>
</tr>
<tr>
<td>4</td>
<td>5 mos, F</td>
<td>lumbosacral</td>
<td>incontinence, CTEV</td>
<td>none</td>
<td>conus at L5–S1, myelocystocele</td>
<td>status quo (1 yr)</td>
</tr>
<tr>
<td>5</td>
<td>1 mo, F</td>
<td>lumbosacral</td>
<td>normal</td>
<td>none</td>
<td>conus at L5–S1, myelocystocele</td>
<td>normal (6 mos)</td>
</tr>
<tr>
<td>6</td>
<td>19 days, F</td>
<td>lumbosacral</td>
<td>incontinence, bilateral CTEV</td>
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<td>conus at L5–S1, myelocystocele</td>
<td>normal (1 yr)</td>
</tr>
<tr>
<td>7</td>
<td>3 days, F</td>
<td>lumbosacral</td>
<td>normal</td>
<td>aqueductal stenosis</td>
<td>conus at L5–S1, myelocystocele</td>
<td>status quo (6 mos)</td>
</tr>
<tr>
<td>8</td>
<td>18 days, F</td>
<td>lumbosacral</td>
<td>incontinence, lower limb weakness</td>
<td>hydrocephalus, CM-II</td>
<td>conus at L5–S1, myelocystocele</td>
<td>status quo (5 mos)</td>
</tr>
<tr>
<td>9</td>
<td>5 days, F</td>
<td>lumbosacral</td>
<td>incontinence, lower limb weakness</td>
<td>CTEV</td>
<td>conus at L5–S1, myelocystocele</td>
<td>status quo (3 mos)</td>
</tr>
<tr>
<td>10</td>
<td>6 mos, F</td>
<td>lumbosacral</td>
<td>incontinence</td>
<td>tonsillar ectopia</td>
<td>conus at S-1</td>
<td>status quo (4 mos)</td>
</tr>
<tr>
<td>11</td>
<td>2 yrs, F</td>
<td>lumbosacral</td>
<td>incontinence</td>
<td>none</td>
<td>conus at L-4</td>
<td>status quo (1 yr)</td>
</tr>
<tr>
<td>12</td>
<td>4 days, F</td>
<td>lumbosacral</td>
<td>incontinence, CTEV</td>
<td>none</td>
<td>conus at L-4</td>
<td>status quo (3 mos)</td>
</tr>
<tr>
<td>13</td>
<td>3 wks, F</td>
<td>lumbosacral</td>
<td>CTEV</td>
<td>aqueductal stenosis</td>
<td>conus at L-4</td>
<td>status quo (5 mos)</td>
</tr>
<tr>
<td>14</td>
<td>1 mo, F</td>
<td>lumbosacral</td>
<td>lower limb weakness, incontinence</td>
<td>aqueductal stenosis</td>
<td>conus at L5–S1</td>
<td>status quo (4 mos)</td>
</tr>
</tbody>
</table>

* Treatment consisted of excision of the sac with untethering of the spinal cord in all patients. Abbreviations: CTEV = congenital talipes equinovarus; FU = follow-up; neuro = neurological.
Terminal and nonterminal myelocystoceles

<table>
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<tr>
<th>Case No.</th>
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<th>Neuroimaging Findings</th>
<th>Surgery (intraop findings)</th>
<th>Outcome (last FU)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>3 mos, F</td>
<td>lower thoracic</td>
<td>paraplegia; incontinence</td>
<td>aqueductal stenosis; CM-II</td>
<td>hydromyelic dilation of SC from lower spinal cord; hydromyelic SC expanded into funnel-shaped structure within meningocele sac</td>
<td>excision &amp; repair w/ untethering (Rossi Type II NT myelocystocele)</td>
<td>no change in neuro status (5 mos)</td>
</tr>
<tr>
<td>2</td>
<td>2 mos, F</td>
<td>upper thoracic</td>
<td>normal</td>
<td>split cord malformation</td>
<td>split cord malformation w/ hydromyelia of the lt hemicord, which communicated through a stalk-like structure to the dorsal meningocele</td>
<td>excision of meningocele, sectioning of the CSF-containing stalk; excision of septum between hemicords (Rossi Type I NT myelocystocele)</td>
<td>no change in neuro status (8 mos)</td>
</tr>
<tr>
<td>3</td>
<td>45 days, F</td>
<td>upper thoracic</td>
<td>normal</td>
<td>none</td>
<td>upper thoracic meningocele; extension of hydromyelic SC into meningocele sac</td>
<td>excision of sac; untethering (Rossi Type II NT myelocystocele)</td>
<td>normal (7 mos)</td>
</tr>
<tr>
<td>4</td>
<td>2 mos, F</td>
<td>upper thoracic</td>
<td>normal</td>
<td>none</td>
<td>upper thoracic meningocele; extension of hydromyelic SC into meningocele sac</td>
<td>excision of sac; untethering (Rossi Type II NT myelocystocele)</td>
<td>normal (13 mos)</td>
</tr>
<tr>
<td>5</td>
<td>18 days, F</td>
<td>lower thoracic</td>
<td>normal</td>
<td>none</td>
<td>lower thoracic meningocele w/ stalk extending into the sac; conus at L-4</td>
<td>excision of sac &amp; untethering (Rossi Type I NT myelocystocele)</td>
<td>normal (3 mos)</td>
</tr>
<tr>
<td>6</td>
<td>10 days, F</td>
<td>lower thoracic</td>
<td>normal</td>
<td>none</td>
<td>lower thoracic meningocele w/ stalk extending from dorsal SC into meningocele sac; conus at L-4</td>
<td>excision &amp; untethering</td>
<td>normal (18 mos)</td>
</tr>
<tr>
<td>7</td>
<td>18 days, F</td>
<td>cervical</td>
<td>normal</td>
<td>none</td>
<td>cervical meningocele w/ a stalklike structure extending from dorsal surface of the SC into the meningocele &amp; attached to the sac dome</td>
<td>excision &amp; untethering (Rossi Type I NT myelocystocele)</td>
<td>normal (3 mos)</td>
</tr>
<tr>
<td>8</td>
<td>3 mos, F</td>
<td>lumbosacral</td>
<td>normal</td>
<td>none</td>
<td>lumbosacral meningocele; SC tethered at sacral level; a fibroneurovenous stalk extended from dorsal aspect of low-lying SC to the sac dome (Rossi Type I NT myelocystocele)</td>
<td>excision of fibroneurovenous stalk; untethering of SC by filum terminale sectioning; excision &amp; repair of meningocele sac</td>
<td>normal (6 mos)</td>
</tr>
<tr>
<td>9</td>
<td>10 days, F</td>
<td>lumbosacral</td>
<td>normal</td>
<td>none</td>
<td>lumbosacral meningocele; SC tethered at sacral level; a fibroneurovenous stalk extended from dorsal aspect of the SC to the meningocele sac dome</td>
<td>excision of fibroneurovenous stalk; untethering of SC by filum terminale sectioning; excision &amp; repair of sac</td>
<td>normal (3 mos)</td>
</tr>
</tbody>
</table>

*NT = nonterminal; SC = spinal cord.

The egress of CSF from the dilated central canal (Figs. 1C, 2D, and 2E). The amount of CSF egress varied from case to case. After the outflow of CSF, the trumpetlike spinal cord collapsed. No attempt was made to reconstruct the conus by using pial sutures as has been done in some of the earlier studies; I believe that leaving the central canal open permits free communication of the dilated central canal with the subarachnoid space, and that this may reduce the chances of

**Fig. 1.** A: Sagittal T1-weighted MR image demonstrating a dilated central canal in a low-lying spinal cord herniating into a meningocele sac. B: Axial T2-weighted MR image demonstrating the dilated central canal surrounded by the meningocele sac. C: Intraoperative photograph showing the trumpetlike flaring of the hydromyelic cord (double arrows) and the meningocele sac (single arrow).
recurrence of syringocele. In one patient a teratoma was encountered in the sac and was excised. Five of these children required plastic surgery repair in the form of rotation flaps to cover the defect left behind by sac excision because these children had significant areas of dystrophic skin, which precluded tension-free closure of the skin. Postoperatively, patients were maintained in the prone position with the foot end of the bed elevated until the sutures were removed.

Postoperatively, CSF leaks were encountered in five patients. All CSF leaks were treated conservatively by placing the patient prone, elevating the foot end of the bed, and applying compressive dressings. None of the children with CSF leaks had untreated hydrocephalus. Three children experienced flap necrosis. There was no change in the neurological deficits in any of these patients during the follow-up period. Six of the 14 patients were old enough during the last follow-up visit for their ambulatory status to be assessed, and all six were found to be ambulatory without support. No child with preoperative incontinence achieved complete sphincter control. No patient has presented with retethering so far.

Nonterminal Myelocystoceles
All the children presented with swelling in the back; the base and sides of the swelling were covered by normal skin, whereas varying extents of the dome of the swelling were covered by dystrophic skin. None of the sacs leaked CSF. Except for one child who presented with paraplegia,
all the others were neurologically normal. The paraplegic child also had macrocephaly due to aqueductal stenosis.

Neuroimaging. Plain x-ray films of the spine were obtained in all children, and MR examinations of the spine and brain were performed as well. Three of these children also underwent 3D-CISS imaging and MR myelography. The MR imaging revealed Rossi Type I nonterminal myelocystoceles in six patients and Rossi Type II nonterminal myelocystoceles in the remaining three (Figs. 4A–D and 5A and B).18 The 3D-CISS images and MR myelograms were very useful in the evaluation of the nature of these rare malformations (Figs. 4E, 5C, and 6). Associated anomalies included split cord malformation in one patient with a thoracic myelocystocele, and a low-lying conus due to a thickened filum terminale in two patients with lumbosacral nonterminal myelocystoceles. The patient with the split cord malformation had a hydromyelic hemicord that communicated with the dorsal meningocele.18

Surgery. The surgical procedure varied with the type of nonterminal myelocystocele. In the three patients with Type II lesions, the meningocele sac was opened and the dilated cord with syringocele was detached from the sac, the syringocele was drained, and the meningocele sac was excised (Fig. 4F and G). In Type I nonterminal myelocystoceles, the sac was opened and the neurofibrovascular stalk extending from the spinal cord to the dome of the sac was cut close to the cord surface, and the sac was then excised (Fig. 5D). In the only patient with associated split cord malformation, excision of the septum was then done. In the two patients who had lumbosacral nonterminal myelocystoceles with a low-lying conus due to a thickened filum terminale, the filum terminale was identified and sectioned. There was no change in the neurological status in this group of patients. In one patient with Type I nonterminal myelocystocele and associated split cord malformation, a transient CSF leak developed, which subsided with conservative treatment. There was no change in the neurological status during the follow-up period, which ranged from 3 to 18 months. No patient with nonterminal myelocystocele has presented with retethering so far.

Discussion

The term “myelocystocele” was originally used by Lassman et al.13 to designate a relatively severe form of spina bifida cystica and lumbar hydromyelia in which the dilation of the central canal is so excessive that the cyst may project through the laminar defect and occupy the major portion of the sac.16 These authors used the term “myelocystomeningocele” to designate a relatively milder form of spina bifida cystica and lumbar hydromyelia in which only the thinned and stretched dorsal wall of the cyst will enter the main protrusion of the meninges to a limited extent.

Myelocystoceles can occur at any level of the spine.16 They are classified into terminal and nonterminal lesions depending on their location and postsurgical pathological findings.1,2,7–9,22–27 The term “nonterminal myelocystocele” was introduced recently in the literature by Rossi and colleagues.22 Nevertheless, the existence of this entity has been recognized for many years, although by different names.1,2,8,9,24–27 Doran and Guthkelch14 analyzed a series of 308 patients with spina bifida cystica and noted the existence of a lesion accompanied by hydromyelia of the cord and a cystic lesion at the level of the bone defect at the cervical or thoracic level, which they called a “syringocele.”24–26 Even though...
Barson\textsuperscript{2} had reported similar lesions earlier, it was Friede\textsuperscript{9} who used the term “myelocystoceles” to refer to these lesions in the cervical and thoracic regions. Subsequently, several authors have indicated that many so-called cervical and thoracic meningoceles are in fact myelocystoceles.\textsuperscript{1,14,19,20,24–27} The true incidence of this abnormality is not known because many of the lesions are erroneously diagnosed as meningoceles. There has been a spike in the reports of non-terminal myelocystoceles since the advent of MR imaging.\textsuperscript{1,18,22,24–26}

The following discussion highlights the differences between these two entities in embryological features, clinical presentation, results on neuroimaging studies, surgical findings, and prognosis. The differences between terminal and nonterminal myelocystoceles are summarized in Table 3.

Embryological Differences

**Terminal Myelocystoceles.** These lesions develop from abnormalities of the caudal cell mass (Fig. 7). McLone and Naidich\textsuperscript{16} proposed the following theory for the mechanisms involved in the pathogenesis of terminal myelocystoceles: 1) for unknown reasons, CSF is unable to exit from the early neural tube; 2) this CSF vents into the terminal ventricle after canalization of the tail bud occurs; 3) the terminal ventricle dilates; 4) the expanding terminal ventricle bulges into and disrupts the dorsal mesenchyme but not the superficial ectoderm; 5) as a result, the posterior elements of the spine fail to develop (spina bifida), but the lesion remains deep beneath an intact skin cover; 6) as the terminal ventricle balloons into a cyst, it distends the arachnoid lining of the distal spinal cord, forming a meningocele; 7) the bulk of the cyst prevents the ascent of the cord, producing a tethered cord; 8) after formation of arachnoid, progressive distension of the distalmost cord causes it to bulge caudally below the end of the meningocele into the extrarachnoid space, where it is covered by fat; 9) the cyst also bulges cephalically to expand the distal cord; and 10) disruption of the caudal motor segments produces symptoms that may be present at birth or may appear later and be progressive. The intimate association of the caudal cell mass to the progenitors of the hindgut and urogenital system is responsible for the frequent occurrence of abnormalities of these systems in patients with terminal myelocystoceles.

**Nonterminal Myelocystoceles.** The embryology of non-terminal myelocystoceles can be explained by the limited dorsal myeloschisis theory of Pang and Dias\textsuperscript{19} (Fig. 8). In limited dorsal myeloschisis, neurulation proceeds normally in all areas except in the area of the abnormality, where final fusion of the opposed neural folds does not occur. Basic configuration of the neural tube has been achieved, however, except for a thin slip in the dorsal midline. Here, disjunction between cutaneous ectoderm and neuroectoderm never

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**Fig. 4.** Images demonstrating a Type II nonterminal myelocystocele. A: Sagittal T1-weighted MR image demonstrating a portion of the spinal cord with the dilated central canal herniating into the meningocele sac. B: Sagittal T2-weighted image demonstrating the presence of CSF in the portion of the cord herniating into the meningocele sac. C: Axial T1-weighted image demonstrating a portion of the cord with the central canal herniating into the meningocele sac. D: Axial T2-weighted image demonstrating a portion of the CSF-containing cord within the meningocele sac. E: A 3D-CISS image clearly delineating the nature of the lesion in the Type II nonterminal myelocystocele. F: Intraoperative photograph showing the hydromyelic cord (arrow) attached to the dome of the meningocele sac. G: Intraoperative photograph showing the untethered cord with the probe in the opening of the syringocele cavity.
truly occurs, but the midline gap between the converging cutaneous ectoderm and dorsal sclerotomes from the opposite sides of the embryo remains very narrow. Further development of the full-thickness dorsal myofascial tissues (except for the narrow slip in the midline) progressively sets the integument farther away from the neural tube, which ultimately retains its intraspinal location. A dorsal median stalk exists between the neural tube and the cutaneous ectoderm. The CSF from the central canal dissects through the median stalk and ultimately distends into a sac to form the nonterminal myelocystocele. The attachment of the dorsal median stalk tethers the cord posteriorly.

Even though this theory of limited dorsal myeloschisis was originally proposed by Pang and Dias, for the pathogenesis of what they termed “cervical myelomeningoceles,” as pointed out by McComb and Parent, some of the cases described by Pang and Dias are in reality cervical myelocystoceles, and therefore this theory is applicable to all nonterminal myelocystoceles. By using 3D-CISS imaging we have shown that the fibroneurovascular stalk in nonterminal myelocystoceles occasionally contains a bleb of CSF (Fig. 5C), thereby proving the limited dorsal myeloschisis theory of Pang and Dias.

Clinical Differences

Terminal Myelocystoceles. Patients with terminal myelocystoceles usually present with a lumbosacral mass at birth; the size of the mass is variable. The skin overlying the mass may be normal or may exhibit a hemangioma, nevus, or hypertrichosis. In our series, however, nine of 14 patients had dystrophic skin covering the dome of the sac. Even though patients with terminal myelocystoceles usually present with lumbosacral masses, recently Cartmill et al. reported on a patient who did not have a lumbosacral mass, and in whom this led to a delay in the diagnosis. Early reports of patients with this condition mentioned the absence of neurological deficits at birth as a prominent feature. With experience, however, it is now more evident that many of these children present with a neurological defect at birth.

The presence of neurological deficits at birth is understandable because, as pointed out by McLone and Naidich, in their theories on the pathogenesis of terminal myelocystoceles, the distension of the terminal ventricle might disrupt the developing caudal motor segments. Sim et al. have reported on a patient with this condition who presented with saddle anesthesia and incontinence. All four children with terminal myelocystoceles reported by Cohen had neurological deficits, as did 47% of patients reported by Gupta and Mahapatra and five of the eight patients reported by James and Lubinsky. In the present study, 10 of the 14 children had varying degrees of neurological deficits.

Associated Anomalies. In early reports of terminal myelocystoceles, a disproportionately high incidence of abdominal wall defects and OEIS complex was noted in children with these lesions. With the widespread availability of MR imaging, however, recent reports do not indicate such
a high association,\textsuperscript{10,12} with the exception of the report by James and Lubinsky.\textsuperscript{11} In the present study, no patient had associated OEIS complex. The importance of coexisting cloacal exstrophy in the outcome of patients with this condition will be discussed subsequently. The incidence of CM-II in patients with myelocystoceles has been the subject of much discussion.\textsuperscript{15,17} Midrio et al.\textsuperscript{17} reported on a patient in whom a prenatal diagnosis of terminal myelocystocele was made on the basis of the absence of CM-II on prenatal ultrasound and MR imaging studies, and based on negative alpha fetoprotein levels in the amniotic fluid. Midrio et al. depended on the absence of CM-II to diagnose terminal myelocystocele and thus avoided prenatal surgery. As pointed out by McLone,\textsuperscript{15} and as shown by the present study, in which two of the 14 patients had CM-II, the incidence of CM-II is low when compared with open forms of spinal dysraphism, but its mere presence does not exclude the diagnosis of terminal myelocystocele. This gains importance in the era of prenatal surgery, when proper prenatal diagnosis is necessary to differentiate myelomeningocele from terminal myelocystocele.

Nonterminal Myelocystoceles. Nonterminal myelocystoceles have frequently been reported in the cervical, cervicothoracic, and thoracic regions,\textsuperscript{1,10,24–27} but rarely in the lumbar region.\textsuperscript{22} Among the six patients with nonterminal myelocystoceles reported by Rossi et al.,\textsuperscript{22} only one had a lesion with a lumbar location, three had lesions in the cervical area, and two had lesions in the thoracic region. However, in our series there were six patients with thoracic, two with lumbar, and one with cervical myelocystoceles. As was common in the series of Rossi et al., most of these patients presented with masses that had good skin coverage over the base and sides, but with dystrophic epithelium over the dome of the swelling.

The clinical features of nonterminal myelocystoceles are quite variable. In their recent series of nonterminal myelocystoceles, Rossi et al.\textsuperscript{22} reported on six patients, all of whom were neurologically normal at presentation. Nevertheless, earlier reports of this condition indicate that some of these patients have neurological deficits at presentation. Steinbok and Cochrane\textsuperscript{26} presented a series of patients with cervicothoracic midline cutaneous mass lesions. Among these patients, there were three with myelocystoceles, two of whom had neurological deficits at presentation. In the present study also, among the nine patients with nonterminal myelocystoceles, one patient presented with paraplegia and incontinence. Thus, when compared with terminal myelocystoceles, a higher proportion of patients with nonterminal lesions are neurologically better preserved at presentation.

Associated Anomalies. In the series of cervical myelocystoceles reported by Steinbok and Cochrane,\textsuperscript{26} all three patients had associated hydrocephalus. In a series of six patients with nonterminal myelocystoceles published by Rossi et al.,\textsuperscript{22} five of the patients had associated malformations as follows: three with CM-II and one each with a neurenteric cyst, hydrocephalus, and filar lipoma. In the present series, two had thickened filum terminale, one had a split cord malformation, and one had aqueductal stenosis and a CM-II. Thus, associated anomalies are not uncommon in nonterminal myelocystoceles. Nevertheless, in all patients with nonterminal myelocystoceles reported to date, associated anomalies were confined to the nervous system, unlike terminal myelocystoceles, for which there is a disproportionately high association with abdominal wall defects.

**Neuroimaging Differences**

**Terminal Myelocystoceles.** The neuroimaging features of terminal myelocystoceles have been described adequately in earlier studies.\textsuperscript{5,7,8,6,21} In the present series, all patients had the classic features of terminal myelocystoceles, which are characterized by low-lying hydromyelic cord herniating

<table>
<thead>
<tr>
<th>Feature</th>
<th>Terminal Myelocystocele</th>
<th>NT Myelocystocele</th>
</tr>
</thead>
<tbody>
<tr>
<td>embryology</td>
<td>abnormality of caudal cell mass (abnormality of secondary neurulation)</td>
<td>limited dorsal myeloschisis (abnormality of primary neurulation)</td>
</tr>
<tr>
<td>neuro deficits</td>
<td>common</td>
<td>rare</td>
</tr>
<tr>
<td>associated anomalies</td>
<td>common</td>
<td>rare</td>
</tr>
<tr>
<td>radiology findings</td>
<td>homogeneous</td>
<td>two types</td>
</tr>
<tr>
<td>op</td>
<td>drainage of syringocele, unthatching of the SC, &amp; excision of meningocele sac</td>
<td>varies w/ the type of NT myelocystocele</td>
</tr>
<tr>
<td>prognosis</td>
<td>less favorable</td>
<td>more favorable</td>
</tr>
</tbody>
</table>
Terminal and nonterminal myelocystoceles

**Nonterminal Myelocystoceles.** Recently, Rossi and coauthors described the neuroimaging findings in patients with nonterminal myelocystoceles. They divided nonterminal myelocystoceles into two types depending on the neuroimaging findings, as follows: Type I, which is a forme fruste myelocystocele in which the meningocele sac is traversed by a fibroneurovascular stalk extending from the spinal cord to the dome of the meningocele sac (this forme fruste type was also termed “myelocystocele manqué”); and Type 2, which is a complete form in which the major component of the CSF-filled cyst is a focal hydromyelia that has displaced into the meningocele. In their series, they had three Type I and three Type II nonterminal myelocystoceles. In my series, however, there were six patients with Type I nonterminal myelocystoceles and three with Type II nonterminal myelocystoceles. In both types, the cord appears tented posteriorly at the site of the abnormality. This differentiation of nonterminal myelocystoceles into two types based on neuroimaging results is of surgical importance, as will be discussed subsequently. My experience has shown that MR myelography and 3D-CISS sequences are very helpful in delineating the nature of the underlying entity in this rare form of occult spinal dysraphism. I recommend inclusion of 3D-CISS sequences and MR myelography in the routine evaluation of all skin-covered masses in the back. In this study, it has been shown for the first time on 3D-CISS imaging the presence of CSF loculation within the fibroneurovascular stalk of nonterminal myelocystoceles.

**Surgical Differences**

**Terminal Myelocystoceles.** Surgery for terminal myelocystoceles differs from surgery for meningoceles due to the fact that the spinal cord herniates into the meningocele sac to which it is attached. As a consequence, it is mandatory to expose the lamina immediately proximal to the defect and remove it. The dural opening should start from the normal dura mater and then be extended toward the meningocele. This will prevent inadvertent cord injury, which might occur if the sac were to be opened directly at the dome of the meningocele. In most cases, the trumpetlike flaring of the hydromyelic cord was visible on opening of the sac, with the nerve roots passing back into the spinal canal from the cord. In addition, arachnoidal adhesions and meningocele manqué were frequently encountered. After detaching the hydromyelic cord from the sac, there was egress of CSF from the central canal. The amount of CSF outflow was variable and was not profuse. In all cases, the fluid was clear and colorless, unlike in certain reports in which it was found to be xanthochromic or even purulent. Many earlier authors have recommended closing the decompressed hydromyelic conus with pial sutures to reconstruct the spinal cord. In this series this was not done because I believe that leaving the central canal open allows free communication between the dilated central canal and the subarachnoid space, and thereby theoretically reduces the chance of recurrence of syringocele, even though such a recurrence has not been reported to date.

**Nonterminal Myelocystoceles.** Surgical techniques in non-terminal myelocystoceles...
Terminal myelocystoceles are dependent on the recognition of the two subtypes of these entities. In Type I, in which there is a fibroneurovascular stalk extending from the spinal cord to the dome of the meningocele sac, the surgical technique consists of untethering the cord by sectioning the stalk flush with the cord and then excising the meningocele sac. Failure to untether the cord by this method will lead to delayed deterioration, as reported by Pang and Dias. On the other hand, in Type II nonterminal myelocystoceles, the procedure consists of opening the meningocele sac, untethering the cord from its attachment to the sac, and opening the syringocele to drain the CSF, followed by excision and repair of the meningocele sac. The cord with the collapsed syringocele is easily contained within the spinal canal because of the existence of the posterior spina bifida. Failure to untether the cord and/or to drain the syringocele might lead to progressive neurological deterioration. Pang and Dias reported on nine patients with “cervical myelomeningoceles,” six of whom underwent simple sac excision without untethering of the cord. Five of the six patients exhibited late neurological deterioration after a mean follow-up duration of 3.6 years. A second surgical procedure to untether the cord resulted in neurological improvement. This shows the importance of untethering the cord.

**Prognostic Differences**

**Outcome in Terminal Myelocystoceles.** The outcome in children with terminal myelocystoceles is variable. Initial studies of terminal myelocystoceles reported a favorable outcome. McLone and Naidich reported that patients with terminal myelocystoceles were born without neurological deficits. However, subsequent reports have shown that many of these children had disabling neurological deficits. Choi and McComb reported on nine patients with a mean follow-up duration of 63 months. In their series, all patients had associated OEIS complex, and all had sphincter disturbances and were significantly neurologically impaired. Cohen reported on five patients with cloacal exstrophy, four of whom had terminal myelocystocele, and the fifth had lipo(myelomeningocele. All four children with terminal myelo-
Terminal and nonterminal myelocystoceles had significant neurological deficits, and after surgery one did not ambulate, two ambulated independently, and one ambulated with orthoses. Recently, James and Lubinsky reported on a unique series of patients with terminal myelocystoceles with and without abdominal wall defects. Among the eight patients in their series, four had abdominal wall defects and four did not. Those without abdominal wall defects were able to ambulate with the aid of orthotic devices, whereas those with associated abdominal wall defects were not able to ambulate independently. Recently, Gupta and Mahapatra reported on 17 patients with terminal myelocystoceles, of whom eight experienced varying degrees of neurological deficits, including two patients with complete paraplegia.

In the present study, 10 of the 14 patients had neurological deficits. The neurological status remained stable in all patients during the follow-up period. From my experience and based on a review of the literature, it is evident that patients with terminal myelocystoceles do not constitute a single homogeneous group. Those with associated cloacal exstrophy generally have severe neurological deficits, requiring multiple surgical procedures, and their prognosis is less favorable. On the other hand, in those without cloacal exstrophy, the outcome is directly related to the neurological status at presentation; those who are well preserved neurologically improve and maintain their improvement, in contrast to those with neurological deficits at presentation whose improvement is less favorable. Surprisingly, in all the series reported to date, including the present study, there was no report of retethering.

Outcome in Nonterminal Myelocystoceles. The outcome in these entities is generally more favorable than the outcome in terminal myelocystoceles. In this series, eight of the nine children with a nonterminal myelocystocele presented without neurological deficits, and they maintained the same neurological status during the follow-up period. In comparison, 10 of the 14 patients with terminal myelocystoceles presented with neurological deficits, and their neurological status did not improve significantly during the follow-up period. In the series published by Rossi et al., 22 follow-up assessments were available for four of the six patients, and in all of these there was no evidence of neurological deterioration. Overall, the outcome in nonterminal myelocystoceles is likely to be better than in terminal myelocystoceles, probably because nonterminal myelocystoceles are not associated with abnormalities in other systems (such as cloacal exstrophy), which contribute significantly to the overall handicap of these patients. 11

Conclusions

Terminal and nonterminal myelocystoceles differ from one another embryologically, clinically, radiologically, surgically, and prognostically. With the widespread availability of MR imaging, many cases of terminal and nonterminal myelocystoceles are likely to be recognized. A proper understanding of these rarer subtypes of occult spinal dysraphism is necessary for providing optimal care to these patients.

References


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