We present two cases of thoracic myelocystocele diagnosed prenatally by sonography. In one of the fetuses, the myelocystocele was associated with a separate lumbar myelomeningocele.

### CASE 1
A 43 year old gravida 3 para 2 woman with a clinically normal pregnancy had a serum triple screen test at 16 weeks’ gestation. The triple screen test measures levels of HCG, AFP, and UE3. The test demonstrated an increased probability of Down syndrome (that is, low levels of maternal serum AFP, elevated levels of HCG, and low levels of UE3). A fetal sonogram was then performed at 18 weeks’ gestation to assess for fetal anomalies associated with Down syndrome. No such abnormalities were found, but a mass was observed along the posterior aspect of the fetal thoracic spine. Note that the maternal serum AFP test (a component of the triple screen test) showed low levels, whereas an open neural tube defect is associated with an elevated serum AFP level.

The mass arose from the upper thoracic area and had a cyst-within-a-cyst appearance (Fig. 1). No dysraphic abnormality of the posterior spinal elements was seen. Within the posterior fossa, compression of the cerebellum (banana sign) and obliteration of the cisterna magna were noted (Fig. 1). The lateral ventricles were normal in size. A subsequent amniocentesis revealed normal AFP and acetylcholinesterase levels and normal 46,XX karyotype. The woman opted for a therapeutic abortion, which was performed at 23 weeks’ gestation. Sonography, MR imaging, and CT of the fetal specimen confirmed that the central cyst communicated with the spinal cord through a narrow channel (Fig. 1). The shallow posterior cranial fossa was also demonstrated. The cyst was skin-covered. Pathologic examination confirmed the existence of the aforementioned morphologic abnormalities related to the spinal cord (Fig. 1).
Figure 1  Thoracic myelocystocele. A, Coronal fetal sonogram of myelocystocele at 19 weeks’ gestation demonstrates cyst-within-a-cyst arising from the upper thoracic spine area. r, Rib arising from lower thoracic spine; s, spinal ossification center, lower thoracic spine; short arrows, inner cyst wall; long arrows, outer cyst wall. B, Transaxial sonogram of posterior fossa and myelocystocele at 19 weeks’ gestation demonstrates compression of the cerebellum (arrowheads) into a banana shape. The cyst-within-a-cyst lies along the posterior aspect of the occiput. Short arrows, inner cyst wall; long arrows, outer cyst wall. C, Fetal specimen at 23 weeks’ gestation. Lateral view demonstrates the attachment of the skin-covered cyst to the upper thoracic area. D, Transverse sonogram through the myelocystocele of fetal specimen at 23 weeks’ gestation demonstrates the cyst-within-a-cyst appearance. A small hypoechoic tract (arrows) courses from the cystic mass toward the spinal cord (sc). Note the hyperechoic dot in the position of the central canal of the spinal cord. This dot may arise from the central canal of the spinal cord. E, Transverse CT scan of fetal specimen at 23 weeks’ gestation, in the same plane as sonogram of D demonstrates contrast material (cm), which was injected into the cyst. A faint contrast-enhanced tract (continued)
(long arrows) courses to the spinal cord (short arrows). The white dot in the center of the spinal cord is probably contrast material in the central canal of the spinal cord. F, Sagittal MR image of fetal specimen through the cervical and upper thoracic spine at 23 weeks’ gestation demonstrates the collapsed cyst mass (C) along the posterior aspect of the fetal spine. Note the thin tract (arrows) between the cyst and the spinal cord (sc). G, Lateral view of whole specimen mount of brain, spinal cord, and myelocystocele corresponds to sagittal MR image of F. The collapsed cystic mass is tethered to the posterior aspect of the spinal cord. H, Histologic examination of the spinal cord and myelocystocele tract, transverse section, demonstrates an abnormal tract (short arrows) through the posterior aspect of the spinal cord, communicating with the central canal (c) of the spinal cord. w, Wall of myelocystocele; sp, posterior spinal element; long arrows, tract extending into posterior soft tissue; curved arrow, skin surface.
CASE 2

An obstetric sonogram was performed on a 25 year old gravida 5 para 4 woman at 27 weeks' gestation to assess fetal size and anatomy. There was no family history of congenital anomalies and no history of maternal exposure to medications. The sonogram demonstrated two cystic masses, one posterior to the upper thoracic spine and one posterior to the lumbar spine (Fig. 2). The posterior spinal elements showed splaying at the thoracic and lumbar levels. Mild enlargement of the left lateral ventricle and evidence of a small cisterna magna also were noted.

Labor was induced at 38 weeks' gestation and a 2855 g female infant with Apgar scores of 9 and 9 at 1 and 5 min, respectively, was delivered. The infant appeared vigorous. She had good lower limb tone, but the ankle reflex and rectal tone were decreased. The upper thoracic mass was skin-covered, but the thoracolumbar lesion was not.

At 1 day of age, CT of the head demonstrated a dilated left lateral ventricle. The posterior fossa was shallow, suggesting Chiari II malformation. Sonography demonstrated a cyst-within-a-cyst appearance in the thoracic mass; the central cyst was connected to the spinal canal cord via a thin channel (Fig. 2). The MR image demonstrated these findings as well (Fig. 2). Radiographs of the thoracic spine showed segmentation anomalies and splaying of the posterior elements in the thoracic and lumbar areas. The thoracic and lumbar lesions were repaired without immediate complications. Improvement of lower limb function was noted immediately postoperatively. The infant subsequently required a ventriculoperitoneal shunt for hydrocephalus at 3 weeks of age. Ultrasonography of the hips subsequently demonstrated a subluxed left hip with dysplastic left acetabulum. At 7 months of age, a cutaneous vesicostomy was required for management of neurogenic bladder. At 11 months of age, cognitive development was appropriate for age. Motor function in the lower extremities was poor, and the infant was not expected to walk.

DISCUSSION

Myelocystocele is an uncommon form of spinal dysraphism that may occur at any level of the spine.1 Few sonographic descriptions of thoracic myelocystoceles have been published.2 Cervical myelocystocele has been diagnosed in utero with sonography and MR imaging.3

In a myelocystocele, the inner cyst is connected via a thin stalk to the central canal of the spinal cord through a defect in the posterior spinal cord (Fig. 3). The inner cyst is lined by the same ependymal cells that line the spinal cord canal. Often the spinal cord canal is dilated cranial and caudal to the defect (i.e., hydromyelia). The inner sac or cyst is surrounded by a meningocele sac that is contiguous with the meninges surrounding the spinal cord. The space between these two sacs may be filled with CSF and is continuous with the subarachnoid space. The meningeal layer is covered by skin. Because the soft tissue defects associated with myelocystocele are small and midline or posterior, the patient may have no visible spine bifida defect in the ossified posterior spinal elements. In case 1, no splaying of the posterior spinal elements was noted (Fig. 1E), but the fetus in case 2 did have splaying (Fig. 2A).

The cyst-within-a-cyst appearance on prenatal sonography should suggest a myelocystocele even if careful examination of the ossified neural arches fails to identify spinal dysraphism or a spina bifida defect. The communication between the cyst and the spinal cord canal may be through a tiny midline pinhole canal that cannot be easily detected. A thin stalk between the inner cyst and the spinal cord may be detected if the scan plane is perpendicular to the fetal spine and perpendicular to the stalk. It is important to examine the rest of the spine for possible associated defects and especially important to examine the fetal head for signs of Chiari II malformation (obliterated cisterna magna, deformed cerebellum [banana sign], enlarged cerebral lateral ventricles, and concavity of the frontal bones [lemon sign]). The presence of a Chiari II malformation would imply a poorer prognosis than its absence would.3,6

Myelocystoceles differ significantly from the more common “terminal myelocystoceles.”4,7 Terminal myelocystocele is closely related to severe anomalies of the trunk and tail bud. These defects include anal atresia, omphalocele, cloacal exstrophy, spinal anomalies, and partial sacral agenesis. Terminal myelocystoceles are not associated with Chiari II malformation. In a terminal myelocystocele that is located in the lumbosacral region, sonographic findings include splaying of the posterior spinal elements, a protruding mass, extension of echogenic material (fat) from the spinal canal into the mass, normal fetal head with normal-sized ventricles, normal posterior fossa, and no evidence of a deformed skull.7

Steinbok8 hypothesized that myelocystocele and meningocele are closely related and caused by the same underlying developmental abnormality. At 24 to 26 days of gestational age, a failure of fusion of the posterior neural tube occurs, and at this level the cutaneous ectoderm does not separate from the neural tube. The overlying tissues develop normally except for a small area in the midline traversed by the neural tissue, or neural stalk, which extends from the dorsal aspect of the spinal cord to the skin. As CSF forms in the subarachnoid space, the CSF surrounding the neural stalk forms the meningocele. If there is significant hydromyelia which is continuous
Figure 2 Thoracic myelocystocele and lumbar myelomeningocele. A, Transverse sonogram in utero at 27 weeks' gestation demonstrates a complex cystic mass (white arrows) arising from the posterior aspect of the thoracic spine area. Note the splaying of the posterior spinal elements. Black arrows, Pedicles of a thoracic vertebra. B, Transverse sonogram 1 day after birth through posterior thoracic mass demonstrates the myelocystocele as a cyst-within-a-cyst. The inner cyst (I) communicates through a narrow channel (arrows) within the dilated central canal (S) of the spinal cord. O, Fluid in outer cyst. C, Transverse MR image day after birth through the posterior thoracic mass demonstrates the inner cyst (I) communicating through a narrow channel (arrows) within the dilated central canal (S) of the spinal cord. This is the same scan plane as the sonogram in B. D, Sagittal MR image 1 day after birth demonstrates the thoracic myelocystocele (M) and the lumbar myelomeningocele. s, Spinal cord elements extending into the myelomeningocele sac.
with the center of the neural stalk, a localized diverticulum of the central canal may form and herniate through the neural stalk, forming a myelocystocele. If no hydromyelia is present, the neural stalk may regress, resulting in meningocele.

Sonography and MR imaging are both useful in the postnatal evaluation of suspected myelocystoceles. Although MR imaging can provide excellent anatomic detail, the requirement for sedation and the thermal instability of the neonate may make the procedure impractical.\(^2\) The portability of equipment and the high resolution of ultrasonography make it an excellent screening procedure. Altman and Altman\(^9\) state that MR imaging is the preferred method for spinal dysraphism postnatally, with the caveat that their study did not address the use of ultrasonography.

The operative management consists of removal of the cystic mass and untethering the spinal cord, which requires an intradural exploration. Cutting the myelocystocele stalk close to its attachment at the end of the spinal cord often provides adequate decompression of the hydromyelia.\(^8\)

The prognosis depends largely on the degree of myelodysplasia, which almost all of the infants with myelocystoceles have to some degree. A young infant may show intact neurologic function postoperatively, but as the child ages, neurologic deficits usually become apparent. Some patients with myelocystoceles are reported to have normal neurologic function at long-term follow-up.\(^6\) The prognosis for infants with myelocystocele is poorer than for those with simple meningocele, as infants with simple meningocele often remain normal neurologically.

Our second case demonstrated two separate spinal abnormalities within the same patient, which is a very rare occurrence. Doran and Guthkelch\(^10\) reported only one case of multiple lesions (thoracic and lumbar spine meningoceles) in a series of 308 patients. Separate thoracic meningocele and lumbar sac lipomyelomeningocele also have been reported.\(^11\) The prognosis for these rare patients has not been addressed previously in the literature. Patients with myelomenigoceles may have a better neurologic outcome if delivered by cesarean section, but this is controversial.

REFERENCES