Ventriculus Terminalis of the Conus Medullaris: MR Findings in Children

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PURPOSE: To determine the appearance of the ventriculus terminalis of the conus medullaris and the frequency with which it is seen on MR. **METHODS:** Four hundred eighteen normal spine MR examinations were reviewed. **RESULTS:** Eleven (2.6%) of 418 children, all younger than 5 years of age, demonstrated an ovoid, nonenhancing, smooth dilation of the central canal within the conus medullaris. Averaged volume of the oblate spheroid was 0.18 cm^3. Fluid within the cavity behaved as cerebrospinal fluid on all pulse sequences, and there was no abnormal signal in the cord tissue surrounding the cavity. **CONCLUSION:** Asymptomatic localized dilatation of the ventriculus terminalis is a normal developmental phenomenon that can be seen on MR.

Index terms: Spinal cord, anatomy; Spinal cord, conus medullaris; Spinal cord, magnetic resonance; Pediatric neuroradiology

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The ventriculus terminalis, also known as the "fifth ventricle" (1), is a small ependyma-lined cavity in the conus medullaris, which is usually in continuity with the central canal of the rostral spinal cord (1, 2). Until recently, this normal cavity was of interest only to pathologists (1), but high-resolution magnetic resonance (MR) imaging now shows the ventriculus terminalis. This normal structure must be distinguished from syringohydromyelia and intramedullary cystic tumors.

We retrospectively analyzed all spinal MR imaging performed at the Children's Hospital of Philadelphia over the past 3 years to determine the frequency of visibility of the ventriculus terminalis, the age distribution in our pediatric population, and features that might assist in differentiating this normal structure from acquired cystic lesions of the conus.

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Methods and Materials

Four hundred eighteen children with normal spine MR findings were studied at The Children's Hospital of Philadelphia between November 1990 and January 1994. All patients with intrinsic diseases of the spinal cord, such as tumors, myelitis, congenital anomalies, and traumatic injuries, were excluded from this series. These MR examinations were reviewed retrospectively.

The reasons for referral for MR imaging included: cranial tumors with question of spinal tumor dissemination, 29% (121 children); sacral dimple or pigmented skin lesion of the lower back, 27% (113 children); anorectal and genitourinary anomalies, 10% (43 children); phakomatoses, 5% (21 children); scoliosis, 5% (21 children); back pain, 3% (12 children), trauma or hemorrhage, 2% (8 children); and miscellaneous, 19% (80 children).

Of the 418 children reviewed, 185 were female and 233 were male (ratio, 0.8:1.0). The age range was from 5 days to 20 years, with 47% younger than 5 years and 53% between 5 years and 20 years of age.

All imaging was performed on a Magnetom Sp 63 1.5-T unit (Siemens, Erlangen, Germany). Multiple sequences (600–800/15 and 2500–3000/25–90/1 [repetition time/echo time/excitations]) and scan planes (axial, coronal, and sagittal) were used. All had axial and sagittal imaging, but not all had coronal imaging. Some patients received gadopentetate dimeglumine (0.1 to 0.2 mg/kg; maximum, 20 mg).

The central cavity consistent with the ventriculus terminalis was measured in three dimensions (length in the sagittal or coronal plane and the anteroposterior and transverse diameters). The cavity was more readily identifiable

in the axial and coronal planes. Measurements were performed by a variety of methods, which included using a light box and calipers, using the section thickness and number of sections over which the lesion was seen, and finally using the MR system software on the MR viewing console.

Results

Eleven children with isolated localized dilatation of the ventriculus terminalis in the conus were identified. This represented 2.6% (11 of 418) of our study population. Eight children had one MR image, two had two MR images (9 months and 18 months, 5 days and 20 months), and one had three MR images (5, 9, and 21 months). None had a history of trauma symptoms to suggest spinal disease, or evidence of Chiari malformations, spinal dysraphism, or other cord disease.

Of these 11 children, the female-to-male ratio was 1.2:1.0. The average age of the children at the initial MR was 15.8 months (age range, 5 days to 4 years 6 months). All children (11 of 11) were younger than 5 years of age. In addition, 8 (72.7%) of 11 were younger than 1 year of age. The clinical indications for their MR studies follow.

Indication	No. of Children
Cranial tumor with question of spinal tumor dissemination	1 (9.1%)
Sacral dimple/pigmented skin lesion lower back	5 (45.4%)
Anorectal anomalies, genitourinary anomalies	3 (27.3%)
Phakomatoses	. . .
Scoliosis	. . .
Back pain	. . .
Trauma, hemorrhage	1 (9.1%)
Miscellaneous	1 (9.1%)

The MR findings were similar in all 11 children. The conus was normally located between T-12 and L-2 and was normal in size. The ventriculus terminalis was ovoid, smooth walled, and had no internal septae (Fig 1). The dilatation was localized to the conus medullaris and appeared on imaging to be continuous with a nondilated central canal superiorly. There was no abnormal signal in or thinning of the surrounding conus. The intracystic fluid followed cerebrospinal fluid (CSF) characteristics (being low signal on T1-weighted, intermediate on proton density–weighted, and high on T2-weighted sequences) (Figs 1 and 2A and B). There was no abnormal enhancement of the wall or the adjacent conus medullaris in the 8 (72.7%) of 11 given gadopentetate dimeglumine (Fig 3).

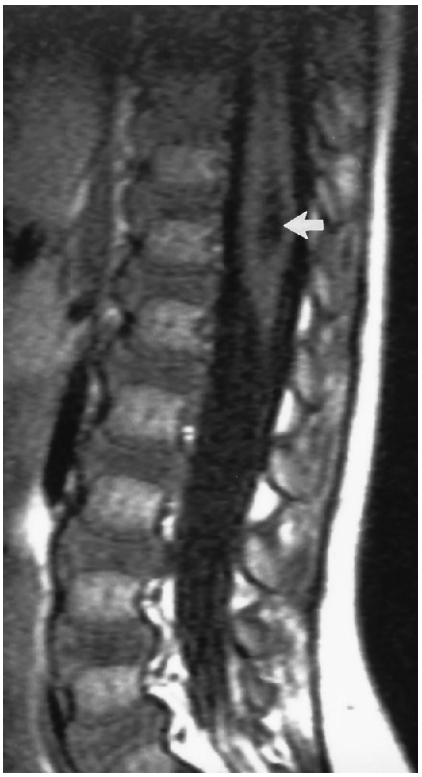


Fig 1. Eleven-month-old girl. Sagittal T1-weighted (600/15) image of the distal spinal cord shows the ovoid ventriculus terminalis (*arrow*) as a distal continuation of a more rostral central canal.

The average measurements of the ventriculus terminalis are: length, 22 mm (range, 15 to 30 mm); anteroposterior diameter, 4.1 mm (range, 1.5 to 6 mm); and transverse diameter, 4.2 mm (range, 1.5 to 6 mm).

An estimate of the volume was made assuming the ventriculus terminalis was an oblate spheroid ($4/3\ \pi \times R/2 \times R/2 \times R/2$, where R is the diameter in each of the three dimensions). The average volume was 0.18 cm^3. In three children who had follow-up scans, there was no significant change in size or signal characteristics of the terminal ventricle (maximum time of follow-up was 21 months).

Discussion

Although Stilling described the ventriculus terminalis in 1859, Krause, in 1875, identified it as a true ventricle lined by ciliated ependymal cells and named it the "fifth ventricle." He identified it in all adults but noted that it is relatively

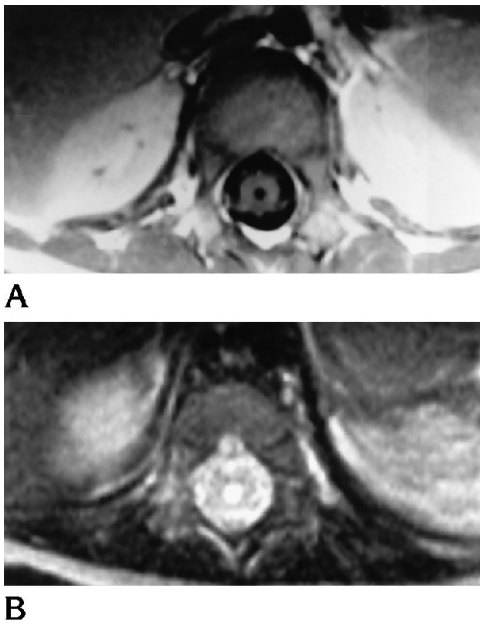


Fig 2. Two-year-10-month-old boy. *A*, Axial T1-weighted (800/14) image at L-1 shows the ventriculus terminalis with low signal consistent with CSF.

B, Axial T2-weighted (2500/80) image shows the ventriculus terminalis with high signal consistent with CSF.

larger in children and the elderly than in middle age (1).

Kernohan's studies, in 1924, provided the most comprehensive evaluation of the growth and development of the "fifth ventricle" in the fetus, children, and the adult population (1). He showed that the ventriculus terminalis has formed by the ninth week of gestation (22-mm fetal length) but that it does not attain its maximum dimension until after 2.5 years of life.

The spinal cord is formed embryologically in two stages: neurulation and canalization and retrogressive differentiation. Neurulation, which begins at 3 weeks of embryonic age (1.5-mm embryo length), involves flexion and closure of the neural plate to form the neural tube. This flexion and closure does not occur along the entire length of the neural plate simultaneously. First the neural folds meet and fuse together at the level of the third to fourth somite when the embryo is at the sixth to seventh somite stage and progresses in a sequential fashion or "wave" cephalad and caudad. The craniocephalad closure occurs by 23 days' gestation, ending at the anterior neuropore (site of the primitive lamina terminalis). The caudad closure is complete around 26 days, ending at the posterior neuropore. This is thought to be between the T-10 and L-4 vertebral segments (3), although the exact level is uncertain. Neurulation, which is completed by about 4 weeks, forms most of the spinal cord.

After neurulation, at about 4.5 weeks' gestation, the caudal end of the neural tube and the notochord blend to become an aggregate of undifferentiated cells called the *caudal cell mass*. Small vacuoles develop within this cell mass, coalesce (canalize), and eventually form an ependyma-lined tube that usually fuses with the more rostral central canal. This wide canal lined by ependyma in the distal conus is the ventriculus terminalis (Fig 4). Finally, during retrogressive differentiation, a major portion of the distal cord involutes to become a fibrous glioependymal strand, the filum terminale (4) (Fig 5). As with all developmental processes, there is potential for a wide range of variation on this main theme. Accessory lateral and dorsolateral canals, which may or may not communicate with the central canal, are commonly seen in embryos, a phenomenon referred to as "forking." This occurs because vacuoles form at many sites and coalesce (link up) variably. Lendon and Emery (2) showed that 10% of healthy adults have major forking of the canal in the conus, and 35% have minor forking.

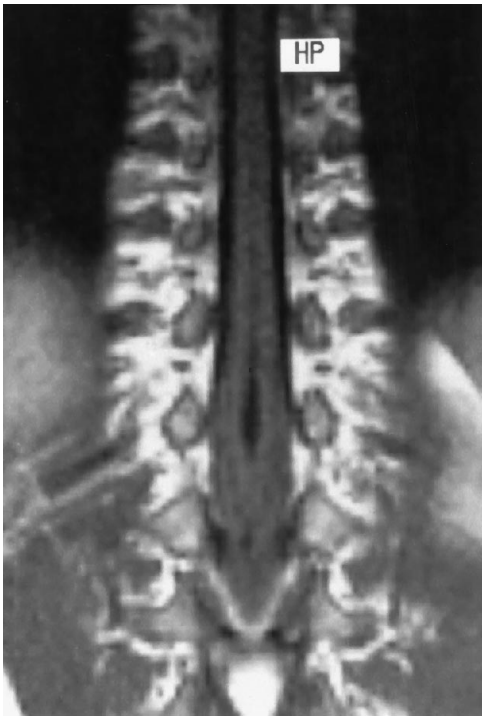
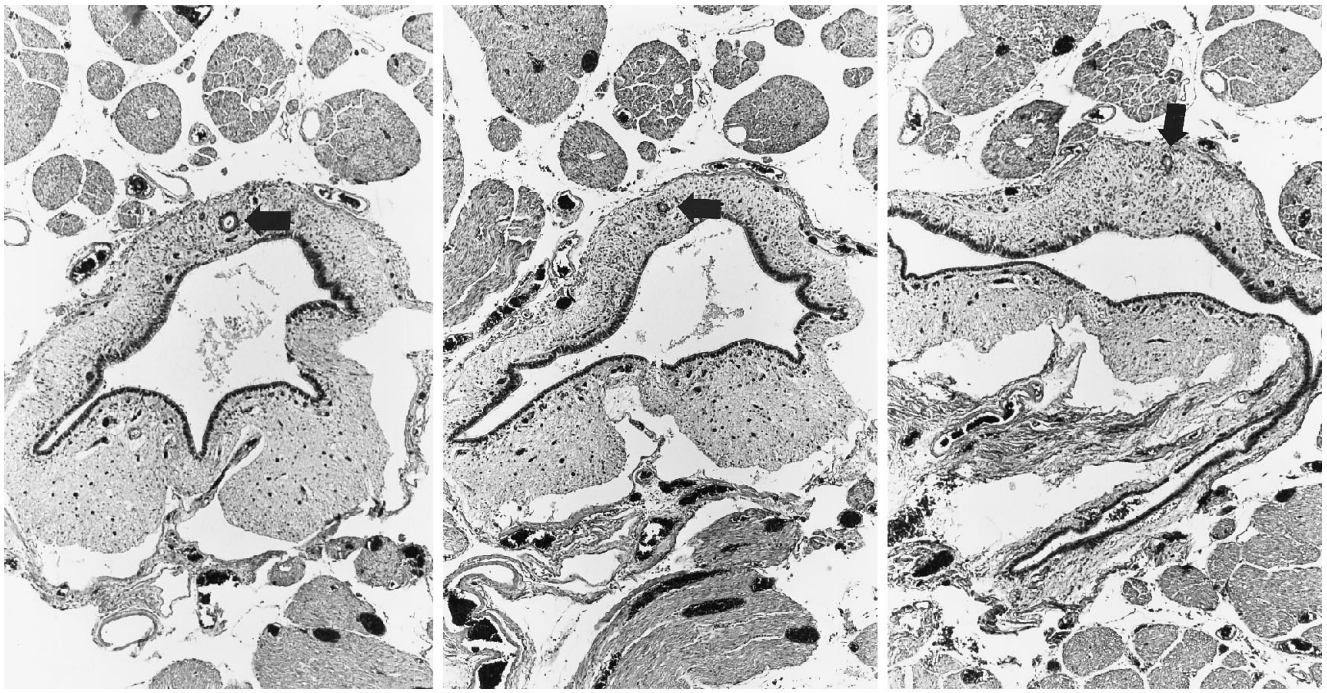


Fig 3. Eighteen-month-old boy. Coronal T1-weighted (600/15) postgadolinium image of the lower spinal cord shows the ventriculus terminalis with no enhancement.



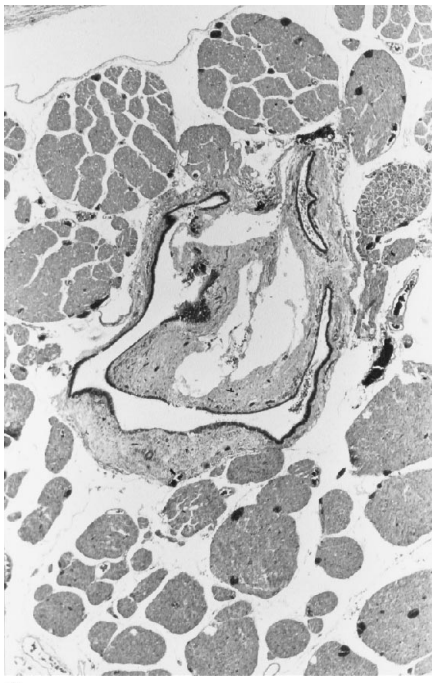


Fig 4. Four levels of ventriculus terminalis from a 6-month-old infant.

A, Most rostral level of ventriculus terminalis in conus medullaris. Note the well-preserved cilia on the ependymal cells. Note the irregular contour and small round dorsal accessory canal (*arrow*) (magnification, × 40).

B, Level slightly caudal to that in *A*. Note the changed configuration and persistant accessory canal (*arrow*) (magnification, × 40).

C, Level caudal to *B* shows striking enlargement and a serpiginous course of ventriculus terminalis. The dorsal accessory canal persists (*arrow*) (magnification, × 40).

D, Most caudal level of ventriculus terminalis shows a large cavity with an irregular contour. Note the satellite cavity in the left inferior region. This would probably be shown to be continuous with the larger cavity if serial sections were done. A tiny dorsal accessory canal is again seen (magnification, × 20).

The mean measurements of the terminal ventricle in the 11 children whom we studied were 22 × 4.1 × 4.2 mm with an estimated volume of 0.18 cm^3. These measurements were larger than those observed by Kernohan in his postmortem study, specifically 15 × 0.5 × 1.5 mm (1). In view of the fact that the ventriculus terminalis is always identifiable on postmortem examination but was seen in only 11 (2.6%) of 418 in this study and was not seen in anyone older than 5 years of age, it is likely that it must achieve a specific size to allow MR visibility. Necropsy studies have established that the ventriculus terminalis is smallest in middle age and

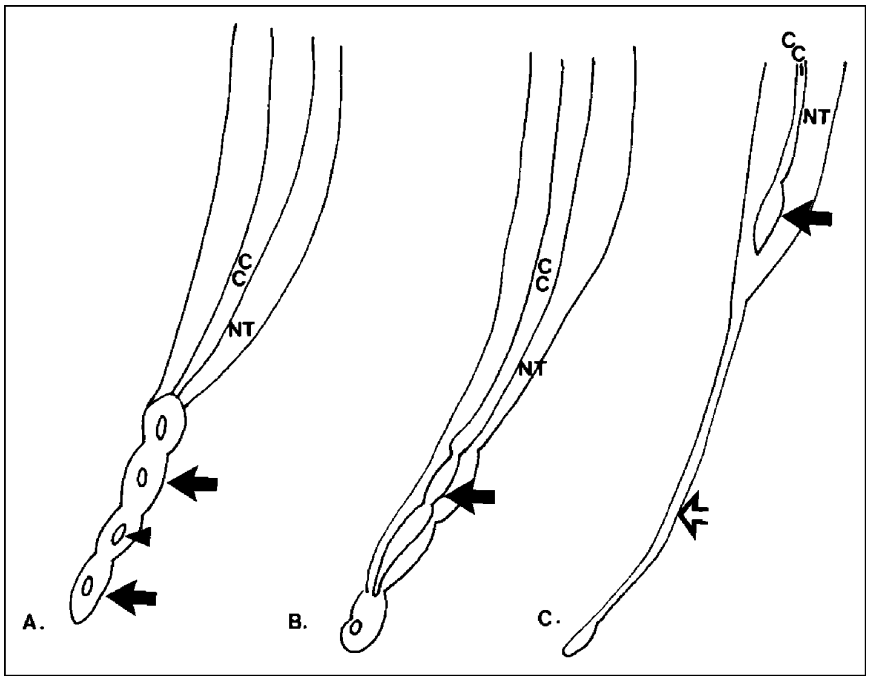


Fig 5. Embryologic development of the distal spinal cord. *CC* indicates central canal and *NT*, neural tube.

A, Vacuolization. Small vacuoles (*arrowhead*) form in the caudal cell mass (*arrows*).

B, Canalization. The small vacuoles fuse or coalesce (*arrow*) to form a dilated ependymal lined tube, which usually merges with the more rostral central canal.

C, Retrogressive differentiation. The dilated ependyma-lined canal becomes the ventriculus terminalis (*arrow*). There is a regression of the distal portion of the caudal cell mass to form the filum terminale. The filum terminale often contains residual nests of ependymal cells (*open arrow*).

largest in early childhood and in the elderly (1, 5, 6). Sigal et al (7) reported four adults who were thought to have cystic dilatation of the ventriculus terminalis, and Nassar et al (8) reported a similar finding in an adolescent.

Nassar et al (8) described a 16-year-old boy who had a progressive scoliosis but was neurologically intact. Two days after spinal fusion lower limb weakness and spasticity developed. Follow-up surgery found marked cystic dilatation of the ventriculus terminalis, which was confirmed histopathologically. After decompression his symptoms resolved.

Sigal et al (7) described four female patients, ages 35 to 65 years, who presented with nonspecific low back pain, sciatica, and bladder dysfunction. Their cystic cavities were also localized to the conus, were nonenhancing and smooth walled, and followed CSF signal characteristics on MR. Surgery and histopathologic analysis in two cases confirmed an ependyma-lined cavity with no evidence of malignancy. The clinical symptoms were unchanged after surgery.

These cystic lesions measured between 25 and 40 mm craniocaudally and 17 and 25 mm transversely with the cyst having a significant mass effect, thinning the neural tissue in the conus to 2 mm or less.

These lesions were significantly larger than those found in our series, were symptomatic compared with our asymptomatic population, and had a mass effect associated with thinning of the neural tissue that was not observed in our 11 children. In addition, one had a Chiari 1 malformation, whereas none of our children had spinal anomalies. Interestingly, surgical relief of the Chiari malformation had no effect on the cyst size in the conus.

Sigal et al (7) postulated that the cystic dilatation in their patients may represent one end of the spectrum of developmental variants at which there is no connection between the ventriculus terminalis and the central canal and/or because CSF fluid dynamics and physiologic changes precipitate enlargement of the ventriculus terminalis into a cyst. Given the size of the cavities and the thinning of the cord tissue by

the cyst, these seem to represent a pathologic change leading to obstruction of the ventriculus terminalis in the adult. In the short-term follow-up (a maximum of 21 months), no significant change was observed in three infants in our series in whom second and third studies were done.

Poser (1956) reviewed more than 200 cases of syringomyelia and found that 12.6% extended into the lumbosacral region from more rostral cord levels, whereas only 5 were restricted to the lumbosacral segments (8). Moreover, there is a high incidence of associated defects with syringohydromyelia, most commonly spina bifida occulta, pes cavus, and syndactylism (9). None of these features was present in the children in whom the ventriculus terminalis was seen. Where there is spina bifida with tethering of the spinal cord and/or the presence of a dorsal dermal sinus tract, then the possibility of a congenital lesion such as an epidermoid would have to be considered.

Conclusion

Asymptomatic localized dilatation of the ventriculus terminalis is a normal developmental phenomenon and should be distinguished from syringohydromyelia and cystic intramedullary tumors. The diagnosis of ventriculus terminalis should be considered if a nonenhancing, ovoid, nonseptated cystic structure localized to a normally positioned conus is seen on MR imaging in an asymptomatic child younger than 5 years. Follow-up imaging would be indicated if clinical symptoms develop. Present evidence suggests that visibility of the ventriculus terminalis on MR imaging in a small number of children younger than 5 years of age simply represents the MR-visible part of the spectrum of a normal developmental process.

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