Concurrent hydromyelia and diastematomyelia.

A E Schlesinger, T P Naidich and R M Quencer

Concurrent Hydromyelia and Diastematomyelia

Alan E. Schlesinger[1]
Thomas P. Naidich[1]
Robert M. Quencer[2]

Hydromyelia was discovered in six of 13 patients with diastematomyelia. In one patient, hydromyelia affected only the segments of spinal cord above the diastematomyelia. In five patients, hydromyelia extended downward from the single cord into one or both hemicords. Because hydromyelia and diastematomyelia occur simultaneously, because they may produce very similar clinical changes, and because simultaneous or sequential surgical correction of both conditions may be necessary to achieve the best clinical result, the possibility of hydromyelia should be evaluated specifically in each patient demonstrated to have diastematomyelia.

Diastematomyelia signifies sagittal clefting of the spinal cord, conus medullaris, and/or filum terminale into two, sometimes asymmetric hemicords [1–8]. There are two major types of diastematomyelia [1]. In the first, both hemicords are enveloped in a common subarachnoid space by a single arachnoid-dural sheath. In the second, a portion of each hemicord is separately encased by one of the paired arachnoid-dural sheaths. The two types have a nearly equal frequency.

Hydromyelia signifies dilatation of the central canal of the spinal cord, with or without persistent communication with the fourth ventricle [6–21]. Both hydromyelia and diastematomyelia may present clinically with similar scolioses, sensory-motor deficits, and orthopedic deformities of the feet.

Current use of sonography, water-soluble contrast myelography, computed tomography (CT), and magnetic resonance imaging (MRI) has led to increased awareness of the frequency and morphologic features of hydromyelia [6, 17, 20, 21] and diastematomyelia [1, 3, 4, 6] in patients with scoliosis and dysraphism. Isolated instances of concurrent hydromyelia and diastematomyelia have been reported [2, 6–8]. This report addresses the frequency of such concurrence and the need to seek hydromyelia specifically in all patients demonstrated to have diastematomyelia.

Materials and Methods

The myelograms and metrizamide CT myelograms of 13 patients with diastematomyelia studied during the 5-year period from January 1980 through December 1984 were reviewed. Metrizamide myelography and 4–6 hr delayed metrizamide CT scans of the spine from these patients were analyzed for evidence of hydromyelia. Unequivocal, substantial dilatation of the central canal of the spinal cord was taken to represent hydromyelia. Subtle increase in central cord density on delayed CT myelography was not considered sufficient evidence of hydromyelia for the purposes of this study.

Results

Hydromyelia was demonstrated in six of 13 consecutive patients with diastematomyelia for an incidence of 46% in unselected cases. The occurrence of
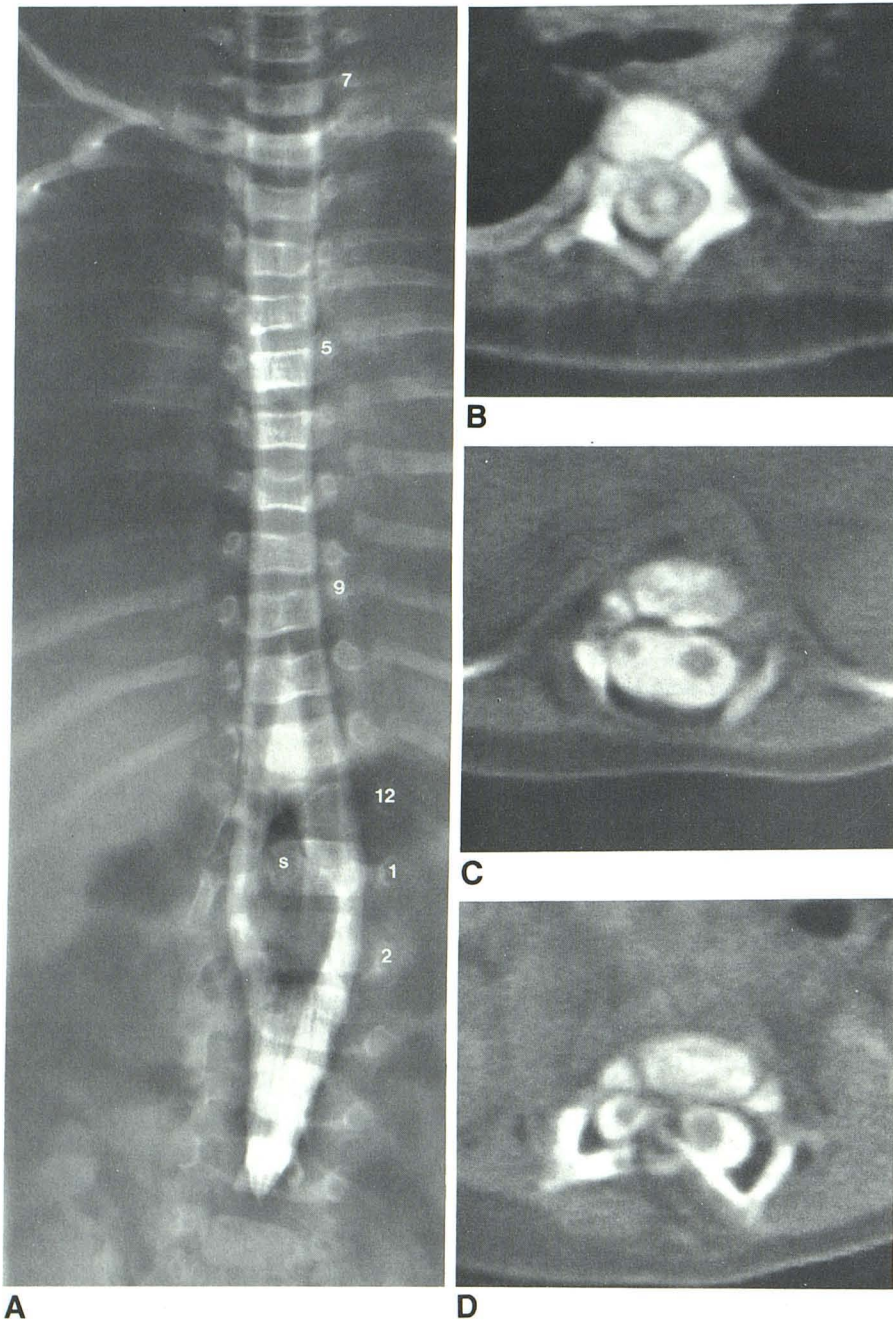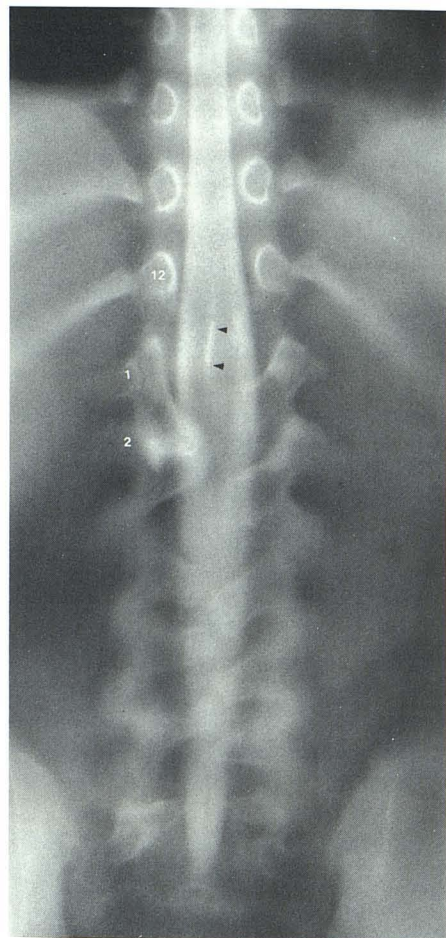
Fig. 1.—Concurrent diastematomyelia and hydromyelia in 5-month-old girl with lumbar nevus pilosus and palpable, skin-covered spina bifida. Neurologic examination was normal. **A**, Frontal metrizamide myelogram reveals tethered cord, dilatation of spinal cord at C7–T5, diastematomyelia with asymmetric hemicords at T9–L2, separation of arachnoid and dura into two tubes (one about each hemicord at T12–L2), bone spur (S) between dural tubes at L1, and reunion of hemicords and dural tubes at L2, below bone spur. **B**, Immediate postmyelogram CT section through thoracic cord. Opacification of large central canal signifies hydromyelia. **C**, Axial CT section at T11 shows spina bifida, lateral position of asymmetric hemicords, and absence of any definite hydromyelia at level of hemicords. **D**, Axial CT section at T12–L1 shows separate arachnoid-dural sheaths about each hemicord, infolding of laminae and part of bone spur between arachnoid-dural sheaths. Cord tethering was released surgically. Hydromyelia is currently being followed.

hydromyelia appeared to be independent of the type and site of diastematomyelia. The six patients with concurrent hydromyelia and diastematomyelia ranged in age from 1 month to 13 years, while the whole group of 13 patients ranged in age from 1 month to 34 years. Five of the six patients with hydromyelia were female. In two patients with hydromyelia both hemicords were enclosed together in a single arachnoid-dural sheath. In the other four patients the hemicords were enclosed in separate arachnoid-dural sheaths.

In one patient, the hydromyelic cavity was present only cephalad to the diastematomyelia (Fig. 1). It did not extend caudally into the hemicords. In this patient each hemicord had a separate arachnoid-dural covering. In five patients the hydromyelia extended caudally, in continuity, from the uncleft cord above into one or both hemicords below. In one of these five the hydromyelia extended as low as the partly reunited cord at the caudal end of the diastematomyelia (Fig. 2). Two of the five patients with hydromyelia of one or both hemicords had diastematomyelia with a single arachnoid-dural sheath (Fig. 3), and three had double arachnoid-dural sheaths.
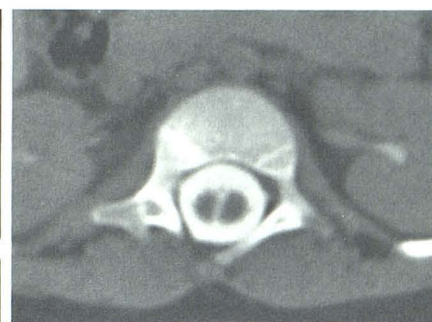
The other seven patients with diastematomyelia but no hydromyelia were all female and ranged in age from 2 to 34
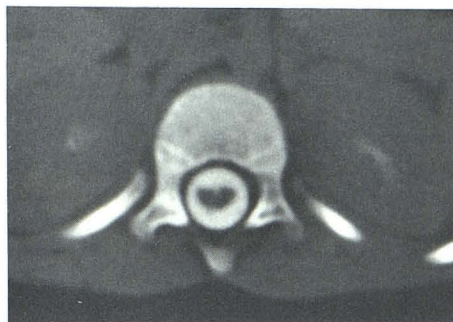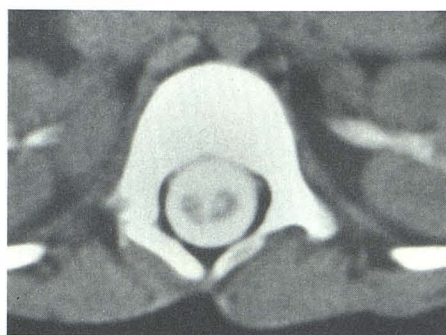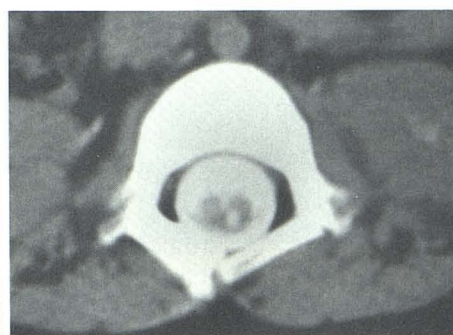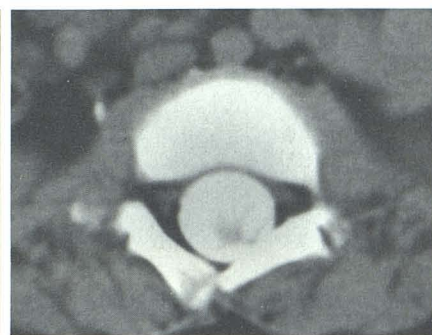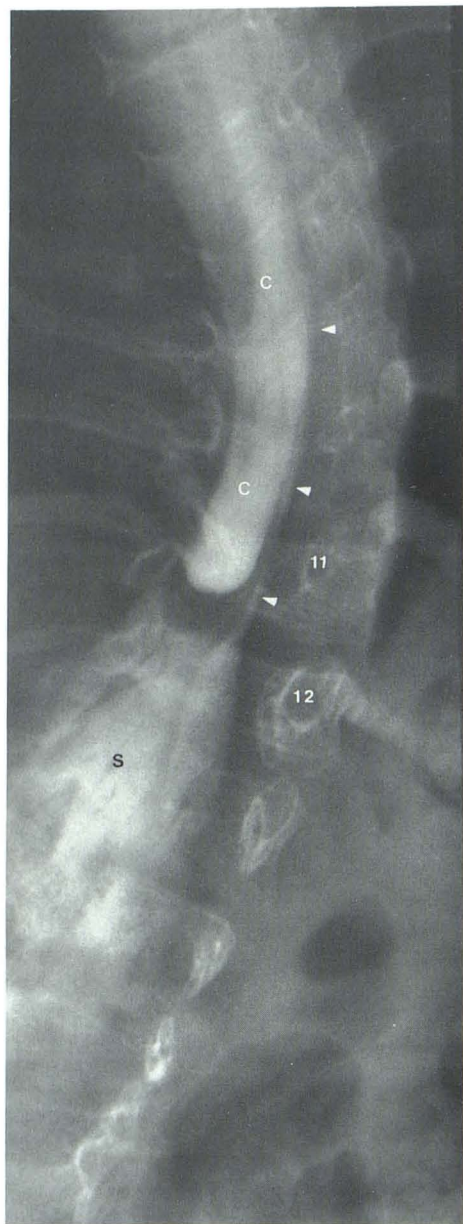
Fig. 2.—Concurrent diastematomyelia and hydromyelia in 4½-year-old girl with nevus pilosus and recent leg weakness. **A,** Frontal pluridirectional tomogram from metrizamide myelogram reveals spina bifida, intersegmental fusion of right L1–L2 laminae, and linear contrast material within obliquely oriented diastematomyelia (*arrowheads*) extending from T12 to L1. **B** and **C,** Axial CT sections immediately after myelography show diastematomyelia with single arachnoid-dural sheath. No hydromyelia is evident. **D–F,** Repeat series of axial CT sections 6 hr later. Dense opacification of enlarged central canal of cord and definite extension of hydromyelia into left hemicord, down to caudal extent of cleft where hemicords begin to reunite. Surgery has not yet been performed.

years. Three had diastematomyelia with a single arachnoid-dural tube and four, double arachnoid dural tubes.

Eight patients had surgery. Six of eight patients underwent subsequent release of cord tethering by resection of the bone spur (four patients) and/or section of a short, thick filum terminale (six patients). Two of these same eight patients had laser fenestration of the hydromyelic cord and placement of a hydromyelia-peritoneal shunt. In one patient with a definite but small hydromyelic cavity, it was decided not to shunt the cavity at the time of surgery. The other five patients are currently being followed clinically with plans to correct the diastematomyelia and/or the hydromyelia surgically in the future.

There was no clinical or radiologic evidence of hydrocephalus in the seven patients who did not have hydromyelia. Two of the six patients with hydromyelia had been shunted for hydrocephalus in the past, but manifested no present evidence of hydrocephalus or shunt malfunction. Both of these had an associated myelomeningocele and were the only two with myelomeningocele in the series.
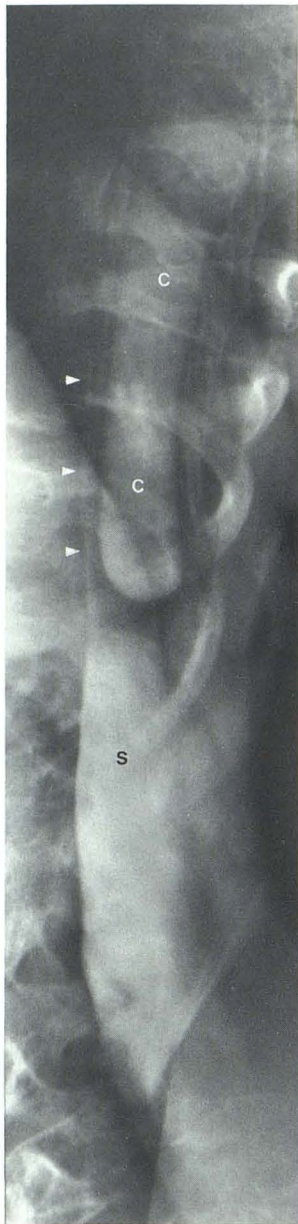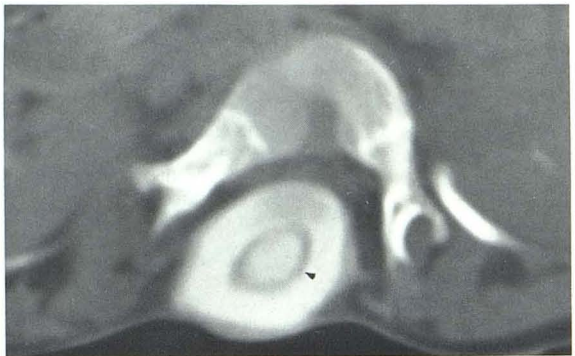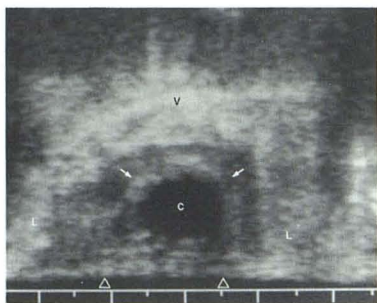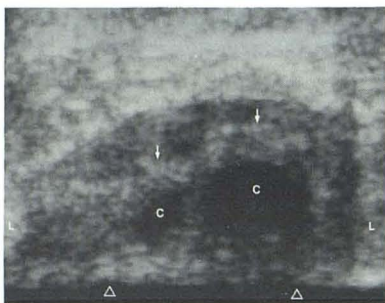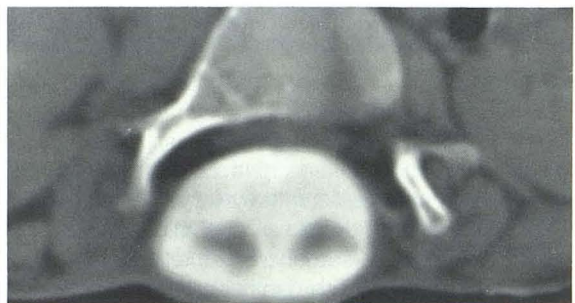
Fig. 3.—Concurrent diastematomyelia and hydromyelia in 5-year-old Brazilian girl with untreated, spontaneously epithelialized thoracolumbar myelomeningocele, shunted hydrocephalus, and progressive severe scoliosis. Initial low lumbar puncture documented complete block to cephalic flow of contrast material at T12. Needle placed at T11–T12 entered a separate space from which clear CSF flowed spontaneously and into which contrast material passed easily. Frontal (A) and lateral (B) radiographs show contrast material within hugely distended central canal (c) of cord, contrast material within lumbar subarachnoid space (S), passage of subarachnoid contrast material (arrowheads) cephalad to T12 after needle decompression of hydromyelia, outline of greatly thinned thoracic cord by intra- and extramedullary contrast material, and poor definition of any cord tissue caudal to T12. C–F, Immediately after myelography, serial axial CT scans obtained at T11–T12 (C), T12 (D), T12–L1 (E), and L2 (F) show spina bifida with very thin skin cover, gross hydromyelia with thin cord, diastematomyelia with single arachnoid-dural sheath, and extension of hydromyelia partway into each hemicord. G and H, Computed axial sonograms obtained percutaneously before surgery with 5.0 MHz linear-array transducer (Acuson). Transverse images oriented with ventral at top conform with CT sections. G, Just cephalic to diastematomyelia. Thin skin (arrowheads), widely bifid laminae (L), sagitally short vertebral body (v), thin, echoic cord (arrows), and huge, anechoic hydromyelia (c) H, More caudal transverse section. Wider spina bifida, bifurcation of cord into two hemicords (arrows) and extension of hydromyelia (c) into each hemicord. Surgery confirmed both hydromyelia and diastematomyelia.

Discussion

In patients with spinal dysraphism and diastematomyelia, progressive scoliosis and increasing sensory-motor deficits may result from hydromyelia alone, from cord tethering alone, or from the combination of the two [4, 5, 12, 17, 19]. Correct identification of both lesions may then permit simultaneous or sequential surgery for either or both cause(s) of clinical difficulty and may help to explain partial response to therapy when only one lesion is repaired.

In some cases, the concurrence of hydromyelia and diastematomyelia may reflect a common underlying anomaly. In patients with myelomeningocele, for example (Fig. 3), Cameron [7] and Emery and Lendon [8] found hydromyelia in 29%–77% and diastematomyelia in 31%–46%. The diastematomyelia affected the cord above the placode in 31%, the placode itself in 25% and the cord below the placode in 25% of cases [8]. In the present series, two of the six patients with concurrent pathology had myelomeningocele (33%).

Hydrocephalus may play a significant role in the production of the hydromyelia. In patients with myelomeningocele, compensated hydrocephalus, extensive communicating hydromyelia and progressive paraparesis, Batnitzky et al. [11] and Hall et al. [12, 15, 17] demonstrated flow of a radionuclide tracer from the dilated lateral ventricles into a dilated central canal of the cord. Similarly, Emery and Lendon [8] observed hydromyelia above or at the placode, but not below it, presumably because escape of cerebrospinal fluid (CSF) from the open placode prevented dilatation of the central canal of the cord below.

Myelomeningocele and hydrocephalus cannot be the only determinants, however. In our series, four of the six patients with concurrent pathology manifested neither myelomeningocele nor hydrocephalus (Figs. 1 and 2). The other two patients were born with myelomeningoceles and had been shunted for hydrocephalus in the past, but had no present evidence of either hydrocephalus or shunt malfunction. Kasantikul et al. [18] observed that the central canal of the spinal cord remained closed in 94% of patients with hydrocephalus. In only 6% was the central canal patent, and in those it resembled the central canal normally observed in patients less than 20 years of age [18]. In cases without myelodysplasia or hydrocephalus, therefore, the concurrence of hydromyelia and diastematomyelia may reflect a separate derangement in embryogenesis, perhaps impaired permeability of the rhombic roof, as proposed by Gardner [9, 10].

REFERENCES

1. Naidich TP, Harwood-Nash DC. Diastematomyelia: hemicord and meningeal sheaths; single and double arachnoid and dural tubes. *AJNR* **1983**;4:633–636
2. Herren RY, Edwards JE. Diplomyelia (duplication of the spinal cord). *Arch Pathol Lab Med* **1940**;30:1203–1214
3. Fernbach SK, Naidich TP, McLone DG, Leestma JE. Computed tomography of primary intrathecal Wilms tumor with diastematomyelia. *J Comput Assist Tomogr* **1984**;8:523–528
4. Hendrick EB, Hoffman HJ, Humphreys RP. The tethered spinal cord. *Clin Neurosurg* **1982**;30:457–463
5. Yamada S, Zinhke DE, Sanders D. Pathophysiology of "tethered cord syndrome." *J Neurosurg* **1981**;54:494–503
6. Han JS, Benson JE, Kaufman B, et al. Demonstration of diastematomyelia and associated abnormalities with MR imaging. *AJNR* **1985**;6:215–219
7. Cameron AH. Arnold-Chiari and other neuro-anatomical malformations associated with spina bifida. *J Pathol* **1957**;73:195–211
8. Emery JL, Lendon RG. The local cord lesion in neurospinal dysraphism (meningomyelocele). *J Pathol* **1973**;110:83–96
9. Gardner WJ. Hydrodynamic mechanism of syringomyelia: its relation to myelocele. *J Neurol Neurosurg Psychiatry* **1965**;28:247–259
10. Gardner WJ. *The dysraphic states*. Amsterdam: Exerpta Medica, **1973**:15–36
11. Batnitzky S, Hall PV, Lindseth RE, Wellman HN. Meningomyelocele and syringomyelia: some radiological aspects. *Radiology* **1976**;120:351–357
12. Hall PV, Campbell RL, Kalsbeck JE. Meningomyelocele and progressive hydromyelia: progressive paresis in myelodysplasia. *J Neurosurg* **1975**;43:457–463
13. Hall PV, Muller J, Campbell RL. Experimental hydrosyringomyelia, ischemic myelopathy, and syringomyelia. *J Neurosurg* **1975**;43:464–470
14. Hall PV, Kalsbeck JE, Wellman HN, Campbell RL, Lewis S. Radioisotope evaluation of experimental hydrosyringomyelia. *J Neurosurg* **1976**;45:181–187
15. Hall PV, Kalsbeck JE, Wellman HN, Batnitzky S, Campbell RL, Lewis S. Clinical radioisotope investigations in hydrosyringomyelia and myelodysplasia. *J Neurosurg* **1976**;45:188–194
16. Hall P, Godersky J, Muller J, Campbell R, Kalsbeck J. A study of experimental syringomyelia. *Neurosurgery* **1977**;1:41–47
17. Hall PV, Lindseth RE, Campbell RL, Kalsbeck JE. Myelodysplasia and developmental scoliosis: a manifestation of syringomyelia. *Spine* **1976**;1:48–56
18. Kasantikul V, Netsky MG, James AE. Relation of age and cerebral ventricle size to central canal in man: morphological analysis. *J Neurosurg* **1979**;51:85–93
19. McLone DG. Results of treatment of children born with a myelomeningocele. *Clin Neurosurg* **1982**;30:407–412
20. Han JS, Kaufman B, Yousef SJ, et al. NMR imaging of the spine. *AJNR* **1983**;4:1151–1159, *AJR* **1983**;141:1137–1145
21. Modic MT, Weinstein MA, Pavlicek W, Boumphrey F, Starnes D, Duchesneau PM. Magnetic resonance imaging of the cervical spine: technical and clinical observations. *AJNR* **1984**;5:15–22, *AJR* **1983**;141:1129–1136