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Diastematomyelia in Children: Metrizamide and CT Metrizamide Myelography

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Diastematomyelia is an uncommon dysraphic lesion of the spine which has been diagnosed more frequently since the advent of newer diagnostic methods. A series of 21 cases was examined using metrizamide myelography and computed tomographic metrizamide myelography (CTMM) over a 3-year period. These examinations, in addition to plain radiographs of the spine, have demonstrated certain features of diastematomyelia hitherto unreported. A bony or cartilaginous spur was an uncommon finding, occurring in only six cases. Also, the split spinal cord was found within an unsplit dural sac in 15 cases. Coexistent tethering of the spinal cord, even in the absence of a spur, was present in 16 of the 21 cases. CTMM proved superior to metrizamide myelography in demonstrating the spinal cord anomalies; plain films and CT are complementary in showing the bony anomalies. The radiographic investigative protocol of diastematomyelia includes plain anteroposterior and lateral spine films, metrizamide myelography, and CTMM. Conventional tomography and plain CT are unnecessary; improved density resolution and availability of computed radiographic anteroposterior and lateral scout views will further reduce the need for plain films and intrathecal injection of contrast medium.

Computed tomography (CT), metrizamide myelography (MM), and computed tomographic metrizamide myelography (CTMM) have, in recent years, added a new dimension to the diagnosis of dysraphic lesions of the spine [1–3]. An uncommon condition in this group of anomalies is diastematomyelia, which is anatomically characterized by a longitudinal splitting of the spinal cord at one or more vertebral levels, usually in the lower thoracic and upper lumbar area, sometimes associated with a bony, cartilaginous, or fibrous spur lying within the spinal canal and protruding through the dural sac into the spinal cord. This is distinct from diplomyelia which is a true duplication of the spinal cord with two discrete dural sacs and two pairs of anterior and posterior nerve roots, and which is very rarely diagnosed during life.

Numerous dysplastic bony anomalies are known to be associated with the neural malformation of diastematomyelia including extensive spina bifida, fusion of neural arches, hemivertebrae, fusion of vertebral bodies, and increased interpediculate distance. These are usually appreciated in part or in whole on plain films of the spine.

The exact *etiology* of diastematomyelia is controversial and several theories have been proposed to explain the abnormal embryogenesis [4–6]. These include: (1) a failure of organization and differentiation of the neural tube from the primitive neuroectoderm in the third or fourth week of gestation, leading to persistence of abnormal mesodermal cells within the neural tube [4, 5] and (2) a persistent accessory neurenteric canal or dorsal intestinal fistula [6]. According to the latter theory, partial obliteration of the fistula may lead to such congenital anomalies as spina bifida, myelomeningocele, hemivertebra, butterfly vertebra, disordered vertebral segmentation, paravertebral cysts, and intraspinal bony spur and diastematomyelia.

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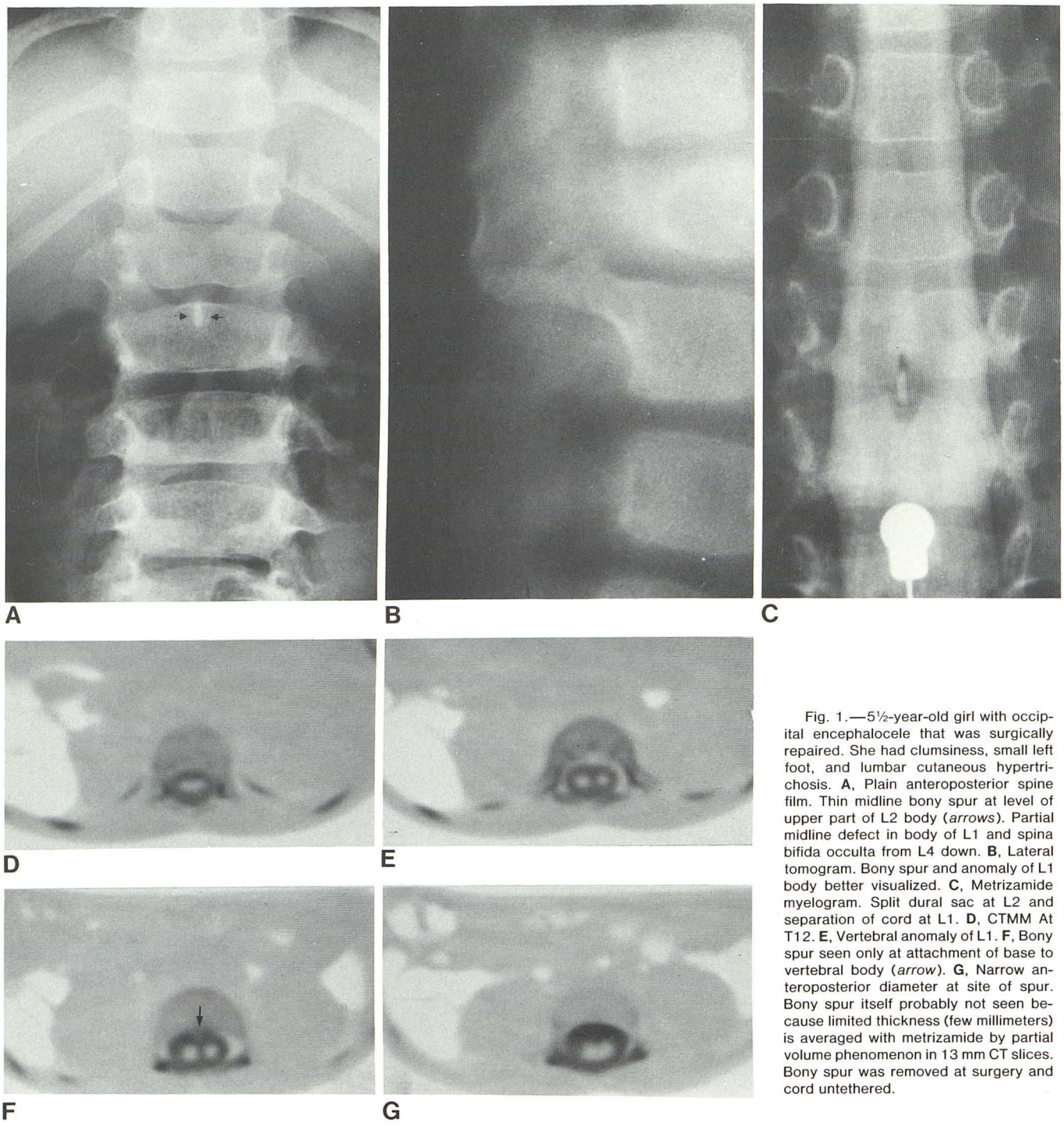


Fig. 1.—5½-year-old girl with occipital encephalocele that was surgically repaired. She had clumsiness, small left foot, and lumbar cutaneous hypertrichosis. **A**, Plain anteroposterior spine film. Thin midline bony spur at level of upper part of L2 body (*arrows*). Partial midline defect in body of L1 and spina bifida occulta from L4 down. **B**, Lateral tomogram. Bony spur and anomaly of L1 body better visualized. **C**, Metrizamide myelogram. Split dural sac at L2 and separation of cord at L1. **D**, CTMM At T12. **E**, Vertebral anomaly of L1. **F**, Bony spur seen only at attachment of base to vertebral body (*arrow*). **G**, Narrow anteroposterior diameter at site of spur. Bony spur itself probably not seen because limited thickness (few millimeters) is averaged with metrizamide by partial volume phenomenon in 13 mm CT slices. Bony spur was removed at surgery and cord untethered.

Clinically, the symptomatology is not specific and does not differ from that seen in other forms of spinal dysraphism such as tethered conus or lipomeningocele [4, 7–9]. The severity of symptoms and signs varies greatly, ranging from completely silent lesions (diastematomyelia is an occasional serendipitous finding in routine scoliosis surveys [10]) to gait disturbance with mild atrophy or weakness of one or both lower extremities, absent deep tendon reflexes, progressive paraparesis, and bladder or bowel dysfunction. Cutaneous abnormalities on the back such as hypertrichosis, pigmented nevus, dimple, or lipoma may be associated and scoliosis is often present. In all large reported series there is a marked predominance of females. This condition is usually diagnosed in children but adult cases have been reported [11, 12].

The *standard radiographic diagnosis* depends on the

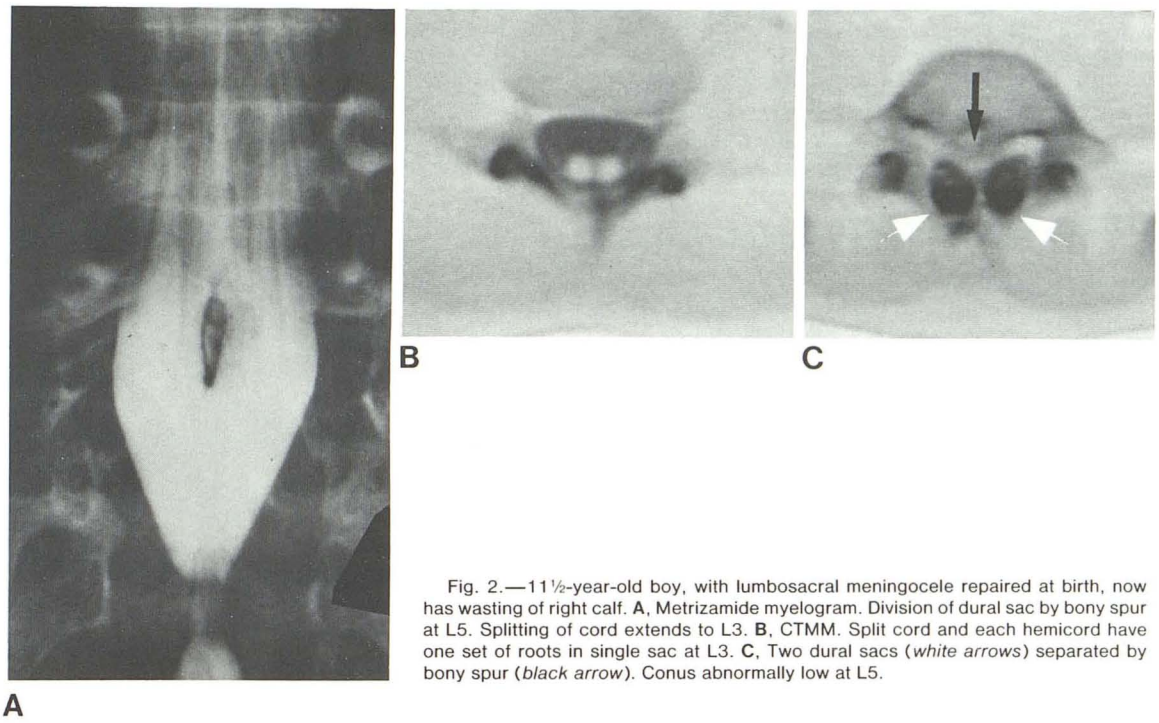


Fig. 2.—11½-year-old boy, with lumbosacral meningocele repaired at birth, now has wasting of right calf. **A**, Metrizamide myelogram. Division of dural sac by bony spur at L5. Splitting of cord extends to L3. **B**, CTMM. Split cord and each hemicord have one set of roots in single sac at L3. **C**, Two dural sacs (*white arrows*) separated by bony spur (*black arrow*). Conus abnormally low at L5.

demonstration of the bony spur when present and the appreciation of the different associated features of spinal dysraphism such as spina bifida occulta, block vertebrae, widened interpediculate distance, hemivertebrae, and scoliosis [12–14] when the spur is absent. Oil myelography may show a defect in the opaque column, usually midline and located near the lower end of the intermedullary cleft. In the presence of a bony septum the dural sac is usually divided into two more or less equal halves. The two separate "hemicords" may be recognized as independent filling defects within the contrast column [14]. Air myelography is much less reliable in making the diagnosis [15].

The neurosurgical management of diastematomyelia has been discussed by several authors [4, 5, 16, 17] and long-term follow-up after surgery has been reported in a large number of cases [9]. Surgery has been used prophylactically to forestall the development of neurologic deficits in asymptomatic patients and to prevent clinical deterioration in those patients who have neurologic signs and symptoms at the time of diagnosis. Keim and Greene [10] recommended that patients undergoing surgery for scoliosis should first have a myelogram and if a diastematomyelia is shown, the tethering bone spike or fibrous band should be removed first. The usual surgical procedure involves excision of the bony spur at its base and closure of the dorsal defect in the dura.

Computed tomography provides a new method of radiologic investigation of the spine; the spinal cord is better demonstrated after intrathecal injection of metrizamide (CTMM) [18]. The CT features of diastematomyelia were first described by Weinstein et al. [19] who reported two cases. Tadmor et al. [20] and Wolpert et al. [2] each reported a case. Lohkamp et al. [21] reported nine cases, eight of whom had a bony septum. In none of the previously reported cases was intrathecal contrast medium used; the CT examination was directed toward demonstrating the bony spur. In nine children with diastematomyelia who were part of a larger group of patients with various dysraphic lesions of the spine, Resjo et al. [1] found that CTMM was more sensitive than conventional radiography and myelography in demonstrating both the bony spur and the splitting of the spinal cord.

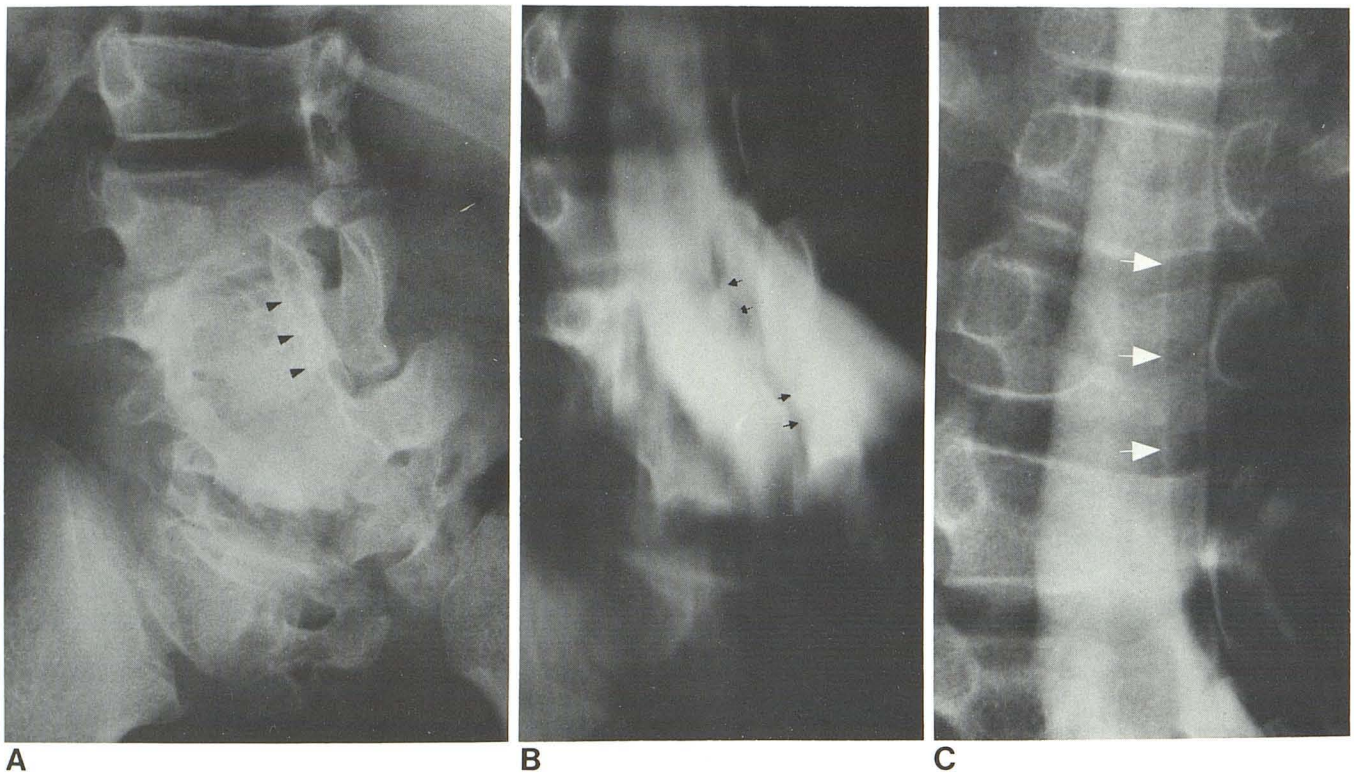
The aim of this paper is to present the characteristic CT findings of diastematomyelia in children, to assess the relative merits of this and other radiologic methods in the investigation of this condition, and to establish an updated protocol for diagnostic workup.

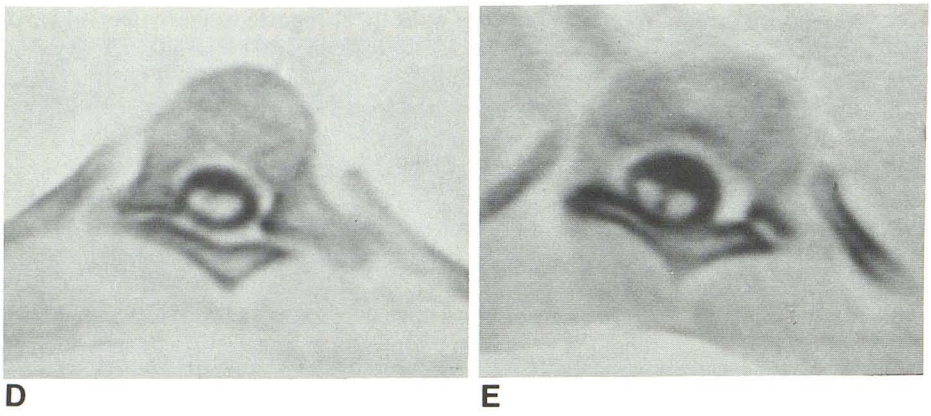
Subjects and Methods

During a 3-year period, 21 patients admitted to the Hospital for Sick Children with diastematomyelia were examined by CT. In 20 of these, plain films of the spine and conventional metrizamide myelography, usually under general anesthesia, were performed before CTMM. One case with a bony spur had CT examination of the spine without metrizamide and in one patient CTMM was preceded by a plain CT examination. CT was performed with an Ohio-Nuclear Delta 50 scanner. A 13 mm collimator was used and a scan field diameter as small as possible was chosen.

Results

There were 13 girls and eight boys; mean age was 7½ years (range, 2 months to 18 years).





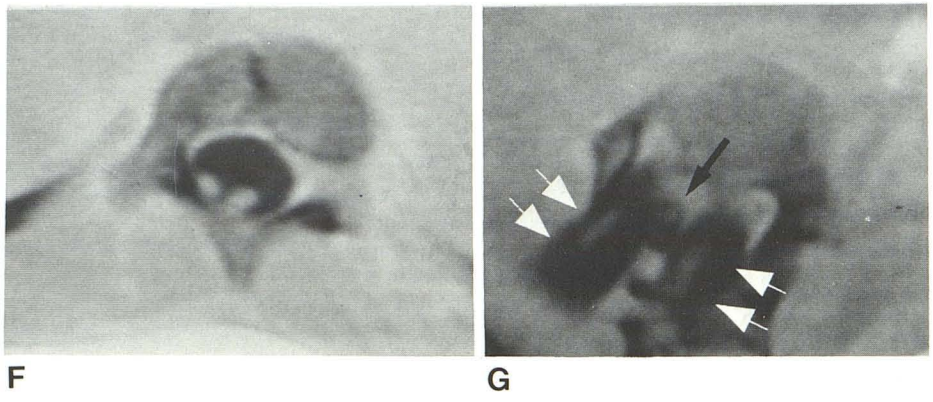


Fig. 3.—10-year-old boy with right foot deformity, left foot weakness, pressure sore on left ankle, neurogenic bladder. **A**, Plain film suggests bony spur (*arrowheads*) poorly recognized due to extensive dysraphic changes. **B**, Metrizamide myelogram. Split dural sac. Despite tomography, lesion not completely demonstrated (*arrows*). **C**, Splitting of cord extends at higher than lumbar levels within single sac, shown by metrizamide between two hemicords (*arrows*). **D**, CTMM. Cord divided from T10. **E**, T12. **F**, Lumbar conglomerate. **G**, Lumbar bony spur divides dural sac into two independent parts, each containing neural tissue. Bony spur (*black arrow*); dural sacs with hemicords (*white arrows*).

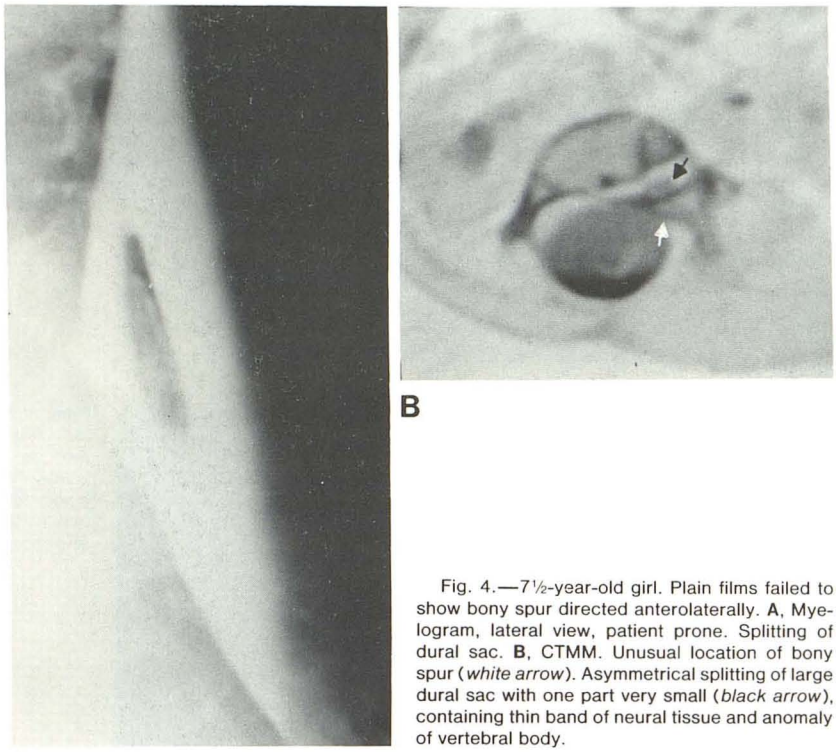


Fig. 4.—7½-year-old girl. Plain films failed to show bony spur directed anterolaterally. **A**, Myelogram, lateral view, patient prone. Splitting of dural sac. **B**, CTMM. Unusual location of bony spur (*white arrow*). Asymmetrical splitting of large dural sac with one part very small (*black arrow*), containing thin band of neural tissue and anomaly of vertebral body.

Clinical Findings

Clinical findings comprised gait disturbance and foot or leg deformity in 18 patients, poor bladder control in 12, and scoliosis in 13. Five patients had surgery at a younger age: repair of myelomeningocele in four and of an occipital encephalocele in the other.

Standard Radiography

A *bony spur* was demonstrated in only six patients. It was clearly seen on plain films of the spine in four cases (figs. 1 and 2), while in two others it was difficult to recognize due to associated complex anomalies of the vertebrae or to its lateral location (figs. 3 and 4). It was in the lumbar area in all cases. Increased interpediculate distance was present in 19 patients, fusion of the vertebrae in 12, scoliosis in 13, and various degrees of spina bifida were seen in all patients. None were normal.

Metrizamide Myelography

Metrizamide myelography in 20 patients showed the splitting of the cord with varying degrees of reliability. Visualization was excellent in three cases (all with a bony spur) (figs. 1–3), good in four (fig. 4), and fair in three (fig. 5). In the other 10 patients demonstration of the split cord was poor or absent although the myelogram was always abnormal, showing an enlarged thecal sac, a tethered conus, or a thickened filum terminale. In 16 cases metrizamide myelography showed associated anomalies such as tethered conus, thickened filum, meningocele, or intraspinal lipoma. Tomography was performed at the time of the myelogram whenever the findings were equivocal and in a few cases the cord abnormality was better demonstrated (fig. 5).

Computed Tomographic Metrizamide Myelography

Demonstration of the split cord with computed tomographic metrizamide myelography (CTMM) was excellent in 15 cases, good in four, and fair in one. In this respect, CTMM was far superior to metrizamide myelography. The bony spur was clearly demonstrated in the two cases in which its presence was questionable on plain films (figs. 3 and 4), but was poorly seen in one case in which the plain films were unequivocal (fig. 1). *Duplication of the dural sac* at the level of the spur was seen in all five cases, both sacs containing part of the divided neural tissue; however, the splitting of the cord (figs. 2 and 3) extended for varying distances above and below the spur within the dural sac.

All the other 15 patients (i.e., those with no bony or cartilaginous spur) had a simple separation of the cord into two parts *within a single dural sac* for varying distances from a minimum of one to a maximum of 10 vertebral levels (figs. 5–7). The highest level at which a split cord was demonstrated was T4; the thoracic cord was involved in six cases, the lumbosacral area in the rest. In half of our cases the two parts of the divided cord were equal and symmetrical (figs. 1–3); in the other half they were asymmetrical (figs. 4–7). In one case the bony spur was eccentric, emerging

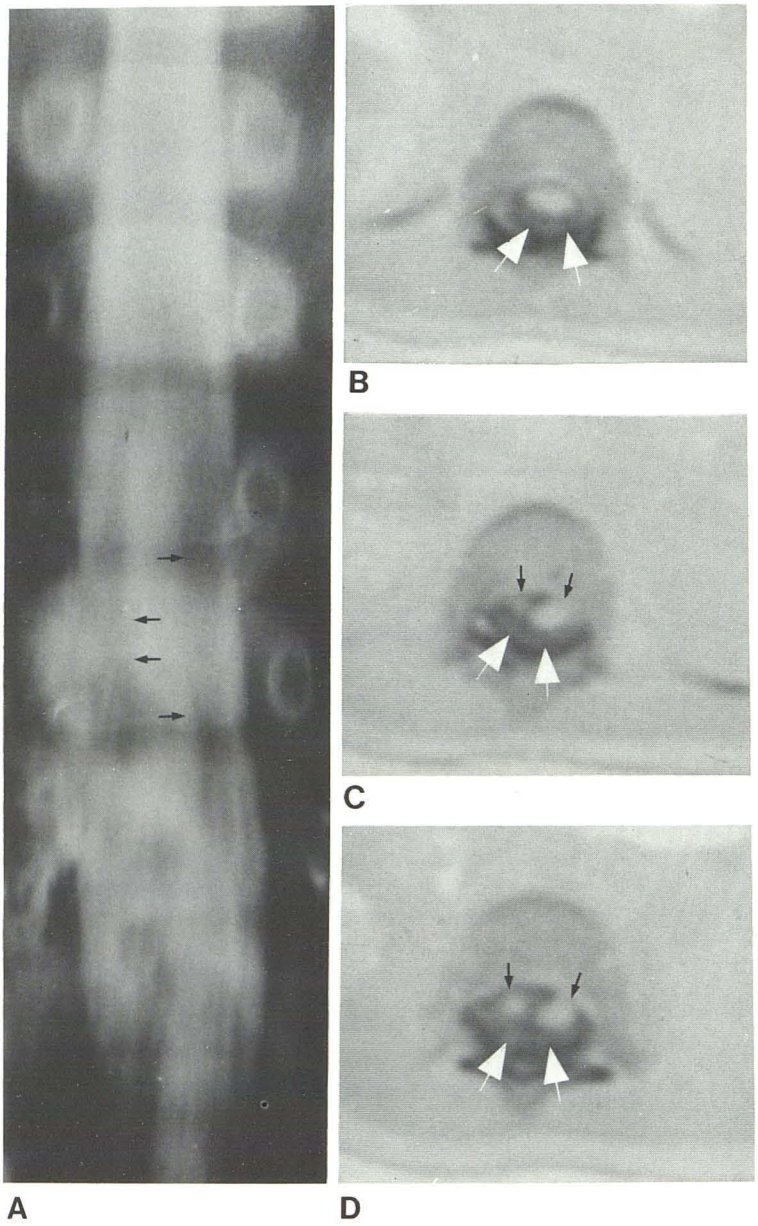


Fig. 5.—21-month-old boy with right varus deformity and small subcutaneous lumbosacral mass. **A**, Myelogram with tomography. Splitting of low cord in lumbar area (*arrows*). Irregular filling defects in opaque column. **B–D**, CTMM at T12–L2. Splitting of cord clearer (*black arrows*). Intradural bandlike structure extensively covers dorsal part of cord (*white arrows*), found at surgery to be diffuse intraspinal lipomatous tissue. Conus was tethered.

from the region of the pedicle and dividing the dural sac into two very asymmetrical parts (fig. 4). The splitting of the neural tissue in this case involved only the nerve roots of the cauda equina. In two cases only the conus was split. These CTMM findings were confirmed in all 15 cases on whom subsequent surgery was performed, either to remove the bony spur and/or release the tethered cord.

CT in the axial plane also showed the dysraphic anomalies of the vertebral bodies to great advantage with especially good demonstration of those anomalies of fusion involving the ossification centers of the bodies. Splitting of the neural arches and irregularities of the bony spinal canal were also nicely outlined. Narrowed or absent disc spaces with bony fusion of adjacent vertebral bodies however were better demonstrated by plain films of the spine. Axial CT showed to best advantage absolute narrowing of the anteroposterior diameter of the spinal canal (figs. 1 and 6) [14, 19], in most cases at the level of the dysraphism only, except in the rare case of severe vertebral disorganization (figs. 3 and 4).

Discussion

Recognition of diastematomyelia in children must be radiological since the neurologic signs, such as asymmetry of the lower limbs, limping, urinary incontinence, and scoliosis, are not specific and are common to other types of spinal dysraphism.

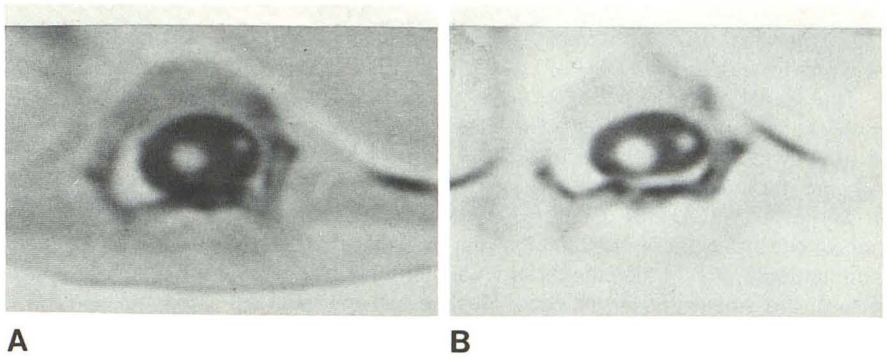


Fig. 6.—2-year-old girl with right lower limb smaller than left and recent incontinence of urine after initial period of bladder control. CTMM clearly shows asymmetrical split cord with single dural sac, without bony spur. Right hemicord smaller than left. Metrizamide myelography demonstration of split cord was poor. Reduced anteroposterior diameter of vertebral bodies and dysplastic neural arches with spina bifida occulta at T10–T12 levels. Tethered conus was released at surgery.

A B

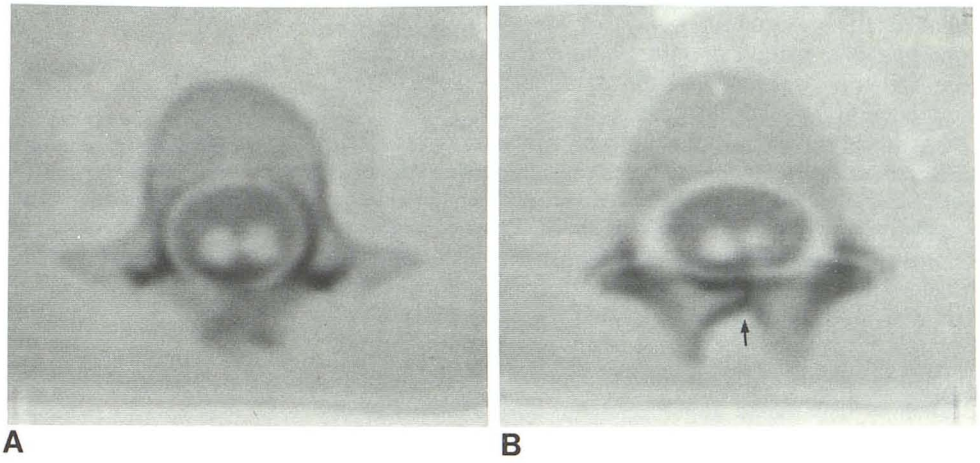


Fig. 7.—4½-year-old girl with small right leg and absent right ankle jerk. Fair myelographic demonstration of split cord. **A**, CTMM. Slightly asymmetrical cord splitting at L2 is clear. **B**, Spina bifida occulta (*arrow*) at L3. Patient had surgery for untethering of low fixed cord.

A B

Very few cases were diagnosed in vivo before 1950 and diastematomyelia was usually discovered incidentally at autopsy. Increasing radiologic interest aroused clinical awareness of the association between vertebral anomalies (such as widened interpediculate distance and dysraphic vertebrae) and splitting of the cord, even in the absence of a bony spur. From 1935 to 1967 the Mayo Clinic reported only 10 new cases of diastematomyelia diagnosed antemortem and treated surgically [8]. At our institution, 14 cases of diastematomyelia were recorded from 1950 to 1962 [16]. The diagnosis was made at autopsy in seven; in the other seven the diagnosis was clinical and radiologic, and confirmed at operation in six.

The advent of new and more sophisticated neuroradiologic techniques and the wider use of myelography has led to the more frequent diagnosis of this disease. The recent introduction of the water-soluble contrast medium, metrizamide, and its use in association with CT (CTMM) has proved to be highly effective for the diagnosis of spinal cord lesions. We have shown [1] that CTMM in spinal dysraphism provided an accurate delineation of the other complex bony abnormalities, associated intra- and paraspinal masses and their relation to the cord, as well as excellent visualization of the normal or abnormal cord itself. In the diagnosis of diastematomyelia, CTMM is superior to both metrizamide and pantopaque myelography in that it provides a complete picture of all aspects of this anomaly throughout the entire spectrum of its manifestations.

During a 3-year period, we detected 21 cases with a split spinal cord. The dramatically increased number of cases detected in this short period is probably the most striking evidence of the improvement brought about by CT in the diagnosis of congenital lesions of the spine. The most interesting result of our work was the high incidence of split spinal cord within a single dural sac, without any spur, a finding not previously reported. The discovery of the split cord was a surprise in some patients who were investigated because of other manifestations of spinal dysraphism. Another unexpected finding in our series, not sufficiently stressed in the literature, is the frequent association of diastematomyelia with other cord anomalies such as tethered cord and filum and low conus, which were present in over three-fourths of our patients.

It is now clear that the presence of a body spur is relatively uncommon in cases of split cord, having been found in less than one-third of our patients. When present, it usually transfixes the dural sac, which is duplicated over a variable number of vertebral segments. Contrary to some earlier reports the bony spur is not necessarily midline; it may arise eccentrically from one side of the spinal canal. Further, the dural sac may be split at such a level that only the nerve roots of the cauda equina are involved while the cord (be it

normally situated or abnormally tethered by a thick filum) remains unaffected.

The spinal cord divides with equal frequency into symmetrical or asymmetrical parts. However, the type of splitting is not related to the pattern of clinical signs or symptoms. Asymmetric splitting may be associated with bilateral signs in the lower limbs, and unilateral leg signs do not always occur on the side of the smaller part of the cord. The affected side, that is the smaller or weak leg, may be on the side of the smaller or larger cord. Neither can the level or extent of the cord separation be predicted from the severity or type of clinical symptoms. Thus, it is obvious that the symptoms in diastematomyelia are not necessarily related to the bony spur within the spinal cord or to the traction effect resulting therefrom, but rather to an intrinsic myelodysplasia. Microscopic or ultrastructural changes within the cord or nerve roots may be the determining factor in the production of the clinical signs.

It has long been known that bony anomalies such as spina bifida are present in all patients with diastematomyelia. While the CT demonstration of the vertebral bony anomalies is good, plain films and tomograms of the spine are usually superior in revealing the full complexity of dysraphic changes particularly of the vertebral bodies, such as fused vertebrae or hemivertebrae. With newer generation CT scanners the facility for sagittal and coronal reconstruction of images will undoubtedly further improve the CT demonstration of disordered anatomy in the bony spine, but at present plain spine films remain indispensable.

Although CTMM has proved superior to metrizamide myelography in demonstrating the abnormalities of the spinal cord in diastematomyelia, particularly the site, extent, and asymmetry of the split cord, the two examinations must remain complementary, the myelogram providing accurate localization of the lesions to be demonstrated by CTMM. Conventional pluridirectional tomography after metrizamide myelography is unnecessary if CTMM is planned, and constitutes excessive and unnecessary irradiation to the patient. Plain CT examination of the spine without metrizamide is at present inappropriate in the investigation of diastematomyelia since the current scanners do not yet provide sufficient resolution of the soft-tissue densities within the spinal canal.

Moreover, while the plain CT examination may demonstrate a bony spur in some cases, we have shown that the presence of a split cord without a bony spur is common, which may not be appreciated without CTMM. We do not believe that the rare occasion on which a bony spur is hidden at CTMM by the surrounding metrizamide due to the partial volume phenomenon (fig. 1) is sufficient justification for routine use of CT examinations *without* metrizamide. This situation is easily avoided if, in doubtful cases, a further CT slice is obtained at the appropriate level, on the day after the metrizamide has cleared from the cerebrospinal fluid.

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