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Diastematomyelia: Hemicord and Meningeal Sheaths; Single and Double Arachnoid and Dural Tubes

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Metrizamide computed tomographic myelography demonstrates well: partial- and full-thickness diastematomyelia; the presence, shape, and (a)symmetry of the two hemicords; focal persistence of two anterior spinal arteries, one supplying each hemicord; the single or double subarachnoid space(s), arachnoid tubes, and dural tubes which encompass the hemicords; the dural septum formed by the medial walls of the two dural tubes; the dural cleft situated between the two walls of the dural septum; the bone spur within the dural cleft; and the presence of any tethering dorsal fibrous bands or aberrant dorsal nerve roots.

Diastematomyelia is a form of spinal dysraphism characterized by partial or complete sagittal clefting of one or more segments of the spinal cord, conus medullaris, and/or filum terminale. The affected segments may be contiguous or anatomically distant from each other. The sagittal clefting usually produces two variably (a)symmetric hemicords, each of which contains one central canal, one dorsal horn that gives origin to the ipsilateral segmental dorsal nerve root, and one ventral horn that gives origin to the ipsilateral segmental ventral nerve root (fig. 1) [1]. By definition, the term *diastematomyelia* (literally *diastema* = cleft, *myelos* = cord) designates the clefting of the spinal cord *irrespective* of the presence or absence of a fibrous partition or bony spur interposed between the hemicords [1].

Diplomyelia is a more or less perfect duplication of the spinal cord, which produces two true spinal cords, each of which contains one central canal, two dorsal horns, two ventral horns, and four segmental nerve roots at each level [1]. To our knowledge, diplomyelia occurs only in patients with true duplication of the spinal column. No instance of true diplomyelia has been reported to occur within a single or cleft spinal column [1], although the lesions observed in some patients more nearly approximate diplomyelia than diastematomyelia.

Weinstein et al. [2], Wolpert et al. [3], Tadmor et al. [4], Lohkamp et al. [5], Resjo et al. [6], Scotti et al. [7], Arredondo et al. [8], and Pang and Wilberger [9] have provided the initial descriptions of the computed tomographic (CT) appearance of diastematomyelia. The present report attempts to extend their work and to shift the emphasis of CT observation from the bony canal and spur to the spinal cord and the meninges.

Materials and Methods

Metrizamide CT myelography was performed according to the protocol previously described [10]. The case histories, routine myelograms, and metrizamide CT myelograms of 46 patients with diastematomyelia seen at the Children's Memorial Hospital, Chicago, and the Hospital for Sick Children, Toronto, in the period from 1976 to 1982 were reviewed and correlated with operative findings and prior descriptions of the anatomic derangements of this condition to elucidate the nature of the CT findings displayed.

Results and Discussion

Medullary Cleft and Hemicord

In diastematomyelia, the medullary cleft is usually directed sagittally and extends through the full thickness of the spinal cord to form two adjacent hemicords (figs. 1–5). Partial sagittal clefting of the cord is typically observed as a transitional state at the cranial and caudal extremes of the main cleft (figs. 1, 2, and 4C) [1, 11]. Less often, the clefting is limited solely to the ventral or dorsal parts of the cord, conditions designated partial ventral diastematomyelia and partial dorsal diastematomyelia, respectively [12–14]. The cleft is frequently focal and of variable length (two to 15 vertebral segments or 1.0–9.5 cm) [14]. The two hemicords most often (91%) reunite into a single, reconstituted cord below the cleft (fig. 4C) [15]. When the cleft arises in the distal cord or extends far caudally, one may observe a bifid filum alone or a bifid conus medullaris with a single or bifid filum terminale [15]. Very low clefts may affect only the roots of the cauda equina [15]. Hilal et al. [15] have demonstrated that the cleft is situated at a locus between T9 and S1 in 85% of cases. It is exclusively thoracic in 20.6%, thoracolumbar in 17.6%, and exclusively lumbar in 61.8% [15]. The conus medullaris is typically low in position and lies below L2 in 76% of cases [15].

The two hemicords are each smaller than the unsplit cord above the cleft [8]. The two hemicords are usually (70%) nearly equal in size (figs. 1–5), but have dissimilar numbers of cells and neurons [8, 11, 14]. Frequently, the ventral median sulcus of the unsplit cord extends progressively more dorsally and progressively to each side to split the cord and to indent the medial surfaces of the two hemicords, so that the two hemicords falsely appear to have rotated

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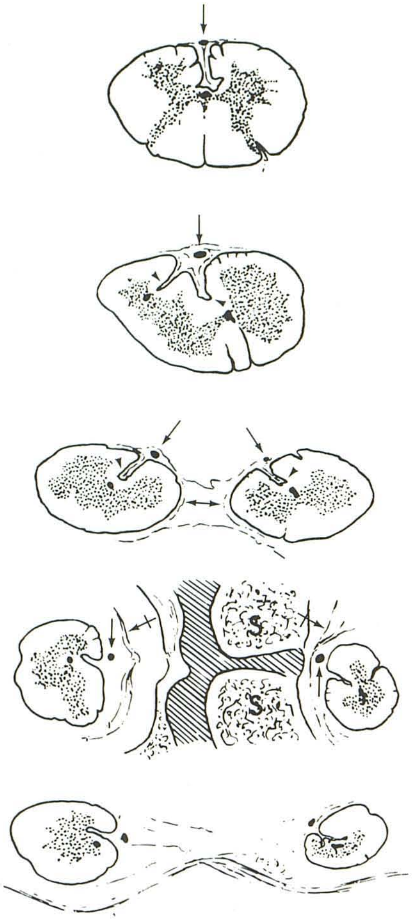

Fig. 1.—Anatomic relationships in diastematomyelia. Ventral is toward top. **Section 1,** Normal cervical cord. Single central canal (*black dot in cord*), single anterior and posterior horns on each side (*stippling*), deep ventral median sulcus, and single anterior spinal artery (*arrow*). Cord has normal pial investment. **Section 2,** Lower cervical cord. Dorsolateral invagination and beginning bifurcation of ventral median sulcus (*arrowheads*) (partial ventral diastematomyelia); partial duplication of anterior spinal artery (*arrow*); formation of two, well separated central canals (*two black dots in cord*); and beginning separation of gray matter (*stippling*) into two parts, each part containing one ventral and one dorsal horn. Pial investment follows deepening ventral sulcus (cf. figs. 2A and 2B). **Section 3,** Progressive dorsolateral invagination of ventral sulcus (*arrowheads*) has caused complete sagittal diastematomyelia with formation of two adjacent, nearly symmetric thoracic hemicords, each containing one central canal (*black dot in each hemicord*). Anterior spinal artery (*arrows*) is duplicated. Each hemicord has its own pial investment (*double-headed arrow*). At this level, both hemicords are contained together in single arachnoid tube and single coaxial dural tube. Hemicords and meninges remain in this form for length of medullary cleft in patients with diastematomyelia and single arachnoid and single dural tubes. **Section 4,** In patients with double arachnoid and double dural tubes, the two hemicords and anterior spinal arteries (*uncrossed arrows*) usually separate more widely. Arachnoid and dura (*crossed arrows*) also become duplicated, forming separate arachnoid and dural tube about each hemicord. Dural cleft, which lies between medial walls of two dural tubes, is usually occupied by fibrous partition or osteocartilaginous bone spur (S). Note cartilage-filled spaces (*hatch lines*) between ossification centers. **Section 5,** The two hemicords may remain separate in separate dural and arachnoid tubes, may remain separate in reformed single dural–single arachnoid tube, or may reunite to form nearly normal spinal cord within nearly normal meninges. (Reprinted from [1].)

their ventral surfaces toward the midline (figs. 1 and 3A) [1]. The gray matter within the hemicords is typically disorganized, but generally forms one dorsal column and one ventral column (which drapes over and appears to be split by the dorsolateral end of the ventral median sulcus) (fig. 1) [1].

Each hemicord typically gives rise to the ipsilateral segmental dorsal and ventral nerve roots (figs. 2C and 3A) [1, 8, 11]. Infrequently, one hemicord may give rise to three roots—the ipsilateral roots plus the contralateral ventral root, for example—while the other hemicord gives rise to only one root, the ipsilateral dorsal nerve root. In other instances, accessory nerve roots may be observed.

The nerve roots, which should have decussated in the region of the diastematomyelia, may cross the medullary cleft in scattered commissural bands which bridge the intermedullary space [14]. Rarely, such commissural fibers pass directly through an intervening bone spur. Alternatively, the decussating fibers may form thickened bundles of nerves at the cranial or caudal ends of the medullary cleft [14].

The paired primordia of the anterior spinal artery form in the second to third weeks of embryonic life, and then migrate medially and fuse into the single anterior spinal artery in the sixth week to fourth month of embryonic life [16]. In patients with diastematomyelia, the paired primordia may fail to fuse segmentally and may persist as paired anterior spinal arteries, each supplying the ipsilateral hemicord (figs. 1 and 3) [1].

Meningeal Sheaths and Bone Spur

In patients with diastematomyelia, each hemicord typically has its own pial lining (fig. 1) [1, 14]. A single, loose pial membrane envelops both hemicords in one pial sheath at the cranial and caudal extremes of the medullary cleft [1]. The arachnoid and dural tubes exhibit variable relations to the two hemicords [1, 14]. The precise arrangements of the arachnoid and dural tubes define two distinct groups of patients with diastematomyelia [14]:

Group A: diastematomyelia with coaxially arranged single arachnoid and single dural tubes. In 50% of patients with diastematomyelia, the two hemicords lie side by side within a single subarachnoid space and are enveloped together in a single arachnoid tube and a single coaxial dural tube (figs. 2 and 3) [14]. That is, in this group, the hemicords and the pial linings are doubled, while the arachnoid tube and dural tube remain single. The 50% of patients with this form of diastematomyelia have no fibrous septum or bone spur extending through the medullary cleft [11, 14]. Rarely, a plug of fibrofatty tissue may lie between and adhere to the dorsal portions of the two hemicords [14].

Group B: diastematomyelia with double arachnoid tubes and double dural tubes. In the other 50% of patients with diastematomyelia, the arachnoid and dural tubes are also doubled. They form a pair of separate and parallel coaxial arachnoid-dural tubes, one surrounding each of the two hemicords (figs. 1 and 4) [1, 14]. Each hemicord then lies in a separate subarachnoid space and a separate coaxial arachnoid-dural tube (fig. 4B). The double arachnoid and double dural tubes merge into each other above, and often below, the cleft to reform single coaxial arachnoid and dural tubes around the yet-unsplit and the reunited cord (figs. 4A and 4C) [1, 11, 14]. In patients with diastematomyelia and double arachnoid-double dural tubes, the medial walls of the two dural tubes form a double-layered dural septum within the medullary cleft [11]. The space between the medial walls of the two dural tubes may be designated the dural cleft. The cleft in the cord (i.e., the diastematomyelia) is necessarily longer than the cleft in the interposed dural septum by millimeters to centimeters, so the medullary cleft extends into the single arachnoid-single dural tube cranial and, to a lesser extent, caudal to the double arachnoid–double dural tubes (figs. 4A and 4C).

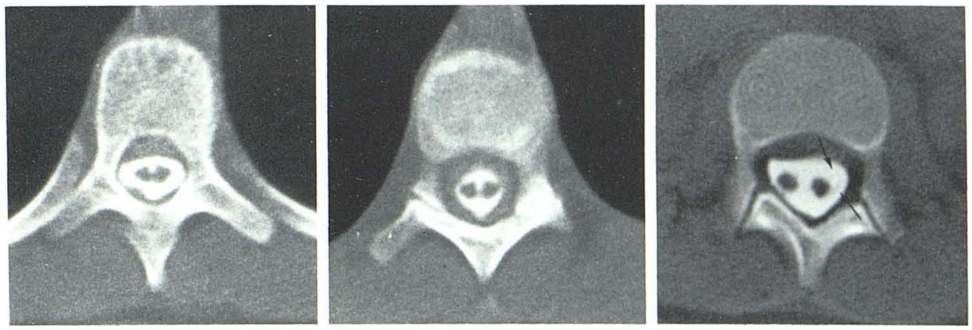

Fig. 2.—Diastematomyelia with single arachnoid tube and single dural tube. Metrizamide CT myelogram in 10-year-old girl with normal spinal cord immediately cranial to these images. **A** and **B**, Sections at cephalic end of medullary cleft show progressive deepening of ventral median sulcus (partial ventral diastematomyelia). Dorsal halves of cord remain united in midline. **C**, At greatest extent of clefting, two nearly symmetric hemicords have separated widely but remain within single subarachnoid space, single arachnoid tube, and single dural tube. Each hemicord gives off ipsilateral dorsal and ventral nerve roots (*arrows*). These hemicords did not reunite distally.

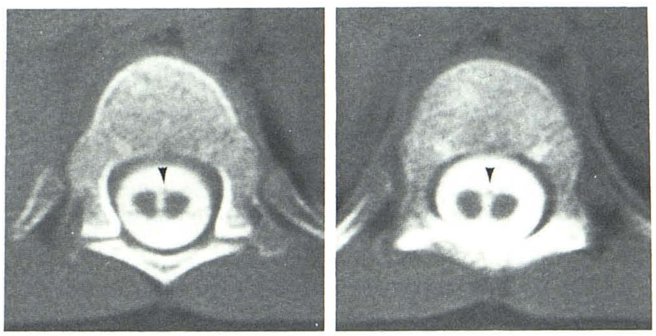

Fig. 3.—Diastematomyelia with single arachnoid tube and single dural tube. Metrizamide CT myelogram in 5-year-old girl. **A**, Low thoracic CT section. Small strand of tissue joins dorsal portions of two otherwise separated hemicords. Such thin strands at extreme end of cleft may be residuum of unsplit (or reunited) cord or may be decussating commissural fibers. The two hemicords are nearly symmetric, give off ipsilateral segmental dorsal and ventral nerve roots, and show indentations on medial surfaces that represent dorsolateral ends of invaginated ventral median sulcus (cf. fig. 1, sections 2 and 3). Single anterior spinal artery (*arrowhead*) just visible at ventral extent of cleft. **B**, Next lower section. Complete separation of two hemicords, bifurcation of anterior spinal artery (*arrowhead*) to supply each hemicord, and envelopment of both hemicords in single subarachnoid space, single arachnoid tube, and single dural tube. These hemicords reunited distal to this level before resplitting still more caudally.

Group B patients with diastematomyelia, double arachnoid tubes, and double dural tubes nearly always have a fibrous partition or osteocartilaginous spur that traverses the dural cleft between and external to the medial walls of the two dural tubes (figs. 1 and 4) [14]. This partition or spur is often oriented sagittally and usually attaches to the vertebral body and lamina by a synchondrosis (fig. 4B) [1]. These synchondroses often appear on CT as radiolucent junctions between the spur and its attachments or between portions of the spur which have formed as separate ossification centers [11]. At the cranial and caudal extremes of the spur, the spur typically extends only partway across the sagittal dimension of the spinal canal (fig. 4). In some cases, including those with only partial dorsal or partial ventral diastematomyelia, even the greatest sagittal dimension of the spur extends only partway across the canal [12–14]. The exact orientation of the spur is variable. In different patients, the spur: (1) may lie in the midsagittal plane of the body and divide the spinal canal into two approximately equal hemicanals; (2) may lie along the spinal midline but be rotated into an oblique plane because of concomitant scoliosis (fig. 4B); or (3) may lie in a plane oblique to the spinal midline and divide the spinal canal into two distinctly unequal hemicanals [11–14]. In (3), the obliquely oriented spur often attaches to a pedicle or an extremely everted lamina.

Aberrant Dorsal Roots, Aberrant Dorsal Ganglia, and Tethering Dorsal Fibrous Bands

Aberrant dorsal nerve roots are frequently detected in patients with diastematomyelia [13, 14]. Aberrant dorsal root ganglia may lie within the dorsal subarachnoid space [14]. In both types of diastematomyelia—those with single and those with double arachnoid-dural tubes—the hemicords may be tethered by one or more of these aberrant dorsal nerve roots or by anomalous dorsal fibrous bands (perhaps representing the residuum of closed meningocele stalks) (fig. 5) [13, 14]. These aberrant roots and bands typically arise from the cranial or caudal ends of the diastematomyelia or from the crossing commissural bands and extend to the dura or through the dura to the bony neural arch [14]. In patients with diastematomyelia and single arachnoid-dural tubes, James and Lassman [14] observed aberrant bands or dorsal roots extending to the dura in 27% of cases and extending through the dura to the neural arch in 45% of cases. Kennedy [17] reported similar "lumbosacral bands" in 23% of patients with diastematomyelia and cauda equina bands in 8% of patients with diastematomyelia. When large, such bands may mimic the appearance of hemicords [13]. Adhesions formed between the hemicords and the arachnoid, the arachnoid and dura, or the two medial walls of dura across the dural cleft may also tether the cord to the meninges and canal wall.

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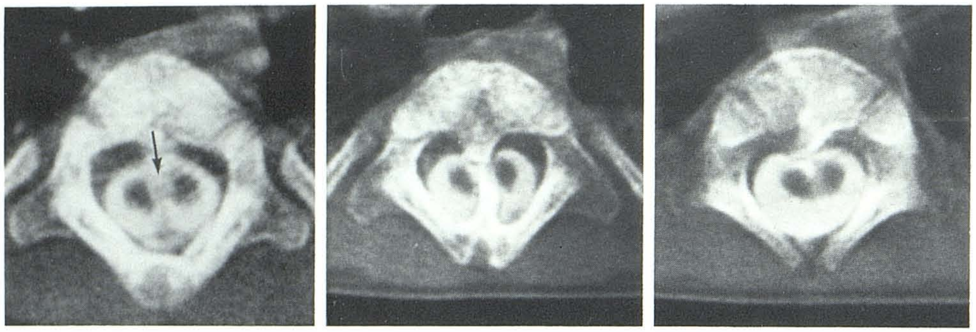

A B C

Fig. 4.—Diastematomyelia with transitions from single to double arachnoid and dural tubes. Metrizamide CT myelogram in 2-year-old boy. CT section immediately cranial to **A** showed single subarachnoid space containing single cord with unusually deep ventral median sulcus. **A,** Upper transition zone. CT section just above T9 demonstrates diastematomyelia with two nearly symmetric hemicords separated by faint, midline, linear lucency (*arrow*) representing extreme upper end of medial walls of double arachnoid and double dural tubes where they merge into single tubes. Metrizamide between two hemicords lies in part of intermedullary cleft extending superior to dural cleft. Thickening of ventral surface of bony canal indicates uppermost aspect of bone spur. **B,** At T9. Each hemicord is now contained in its own arachnoid and dural tube. Bone spur traverses dural cleft between medial walls of two dural tubes. Linear lucency across bone spur is synchondrosis between parts of spur formed as separate ossification centers. **C,** Lower transition section at T10. The two hemicords in **A** and **B** have joined to form reunited cord with deep ventral median sulcus. Reunited cord lies in single subarachnoid space, single arachnoid tube, and single dural tube. Caudal end of bone spur invaginates ventral surface of subarachnoid space at level below dural cleft.

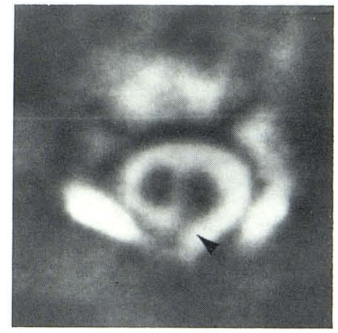

Fig. 5.—Diastematomyelia with tethering band. Metrizamide CT myelography in 6-year-old girl. Linear lucency represents tethering fibrous band (*arrowhead*) which passes from dorsal aspect of one hemicord to extraarachnoid space. Subarachnoid space appears tented by band.

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