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DIASTEMATOMYELIA: A TREATABLE LESION IN INFANCY AND CHILDHOOD WITH CASE REPORT

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It is our purpose to call attention to the tell-tale clinical and radiologic features of the above-named entity since, as expressed by Matson et al,¹ "it constitutes good preventive medicine to carry out surgical treatment of diastematomyelia associated with spina bifida occulta at any time the diagnosis is made during infancy and early childhood." Early diagnosis and treatment is imperative, for what constitutes sound and successful prophylactic surgery on a patient of that age becomes more of an academic surgical exercise on the adult patient in whom full disability has long since been present.

Diastematomyelia, by definition, means any fissuring of the cord, regardless of cause, extent, or internal appearance.² Other terms, actually more specific, but used synonymously, include: diplomyelia, doubling, duplication, and pseudo-duplication of the cord—all conveying Herren and Edwards' idea of form fruste twinning as the underlying pathogenesis. Common usage in the growing list of articles since 1950 on this subject favors the designation "diastematomyelia" in the sense of: congenital malformation manifested by longitudinal splitting of the cord or cauda over any number of segments into perfect or near perfect halves. The commonest site of involvement is in the thoracic or lumbosacral cord or cauda and usually below T₇.

Symptoms and signs (v. i.) apparently cannot be attributed to the "doubling" per se (Herren and Edwards), but rather to fixation of the cord or cauda by the usually-present midline septum (bone, cartilage, bone and cartilage, or fibrocartilage) which, at the level of the split, is attached anteriorly to one or more vertebral bodies and posteriorly to the dura; as a result of this fixation there is progressive torsion and stress on the immobilized cord and roots as the faster-growing vertebrae and their foramina move away from the original site. It is this fact—namely, the differential growth of cord and spine during infancy, childhood, and adolescence—that makes early recognition and treatment of this disorder "good preventive medicine." Cure should not be promised, but predictable progressive defect can be prevented.¹ This is, of course, of special significance to the pediatricians or general practitioners who first see these young patients, and then to the radiologists, orthopedists, neurologists, or neurosurgeons to whom they may be referred.

Now, the diagnostic clues of which all concerned should be aware include under headings as listed:^{3,4}

a) History—

1. Difficulty in walking or in learning to walk;
2. Regression in urinary control or reappearance of enuresis once stopped;
3. Increasing scoliosis or clubbed foot.

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b) Physical Examination—

1. Various midline cutaneous defects in region of underlying neural abnormality—tufts of hair, skin dimples, port-wine stain or cutaneous angioma, poorly circumscribed masses of subcutaneous fat;
2. Scoliosis and foot deformities;
3. Mixed upper and lower motor neurone signs in the lower extremities;
4. Variable sensory changes in the lower extremities.

c) X-ray—

1. Widening of neural canal over several segments (increased interpedicular distance) without erosion of pedicles;
2. Midline osseous spicule at the level of the widened neural canal;
3. Division of pantopaque (myelography) into two columns, outlining the cleft in intraspinal contents;
4. Other congenital osseous vertebral defects, including spina bifida occulta, fused or small vertebral bodies, hemivertebrae, incomplete laminal arches, etc.

The listed clues from history and physical should suggest to the physician the possibility of diastematomyelia, which is then readily confirmed by the pathognomonic radiologic signs on plain films of the area and myelography.

The very dire late sequelae of this lesion that can be avoided by prompt diagnosis and surgical treatment while the patient is still a child include: variable degrees of monoparesis or paraparesis or even monoplegia or paraplegia, atonic sphincters, hydrocephalus, or a full-blown Arnold-Chiari syndrome.⁵

We now add a case report from our recent experience, as an example of adult morbidity, an inevitable sequel of this congenital abnormality when untreated in childhood.

CASE PRESENTATION

B. C., a 21 year old white female, was admitted to the Neurological service at Henry Ford Hospital via the Emergency Room April 11, 1955. Every year since age 14 the patient had had one or two acute attacks of severe low back pain and headache concurrently, along with frequent mild low back and leg pains in the interim. The present illness began following a tooth extraction about three weeks prior to admission, with acutely increased pain in the low back and anterior aspect of the legs on walking. This subsided with penicillin therapy, but recurred two weeks prior to admission, when the patient developed sore throat, generalized headache, chills and fever. Local physician made a diagnosis of "tonsillitis" and treated the patient successfully with penicillin and some sulfonamide for three days. Then three days prior to admission there was again increased pain in the back and legs which continued until the time of admission; the day of admission the patient developed a severe occipital headache in addition to the back and leg pain, had a fever of 100°, and complained of general malaise and vomiting.

The neurological history was otherwise negative except for vaguely described "black-out spells" occasionally since age 15—all had occurred in settings of tension and, from description, were very suggestive of hysterical episodes.

Belatedly, we obtained another very informative and interesting bit of history—

viz., the patient's mother had had spina bifida occulta with a teratoma (probably in the region of the cauda).

Investigations during previous episodes similar to the patient's present illness included: 1) X-ray diagnosis of dorsolumbar spina bifida occulta and posterior fusion of the 5th and 6th ribs on the right; 2) an attempted but unsuccessful lumbar puncture at age 15 as a "polio suspect"; 3) a myelogram at another local hospital (v. i.).

General physical examination on admission was negative except for the patient's obvious acute and severe discomfort, due to head pain and low back pain. Temperature 99.8; pulse 100; respirations 20; blood pressure 116/72. Back pain was so severe as to prohibit active and passive movement and local back "tenderness" was so great as to prevent palpation and lumbar puncture. The patient vomited after our checking her Romberg and gait. The neck was resistant, not rigid. The Kernig maneuver elicited right posterior thigh pain when the right leg was elevated. There was marked lumbar lordosis with a dimple over the spina bifida occulta and there was slightly swollen, boggy tissue over the low back (Figure 1 & 2), with dorsolumbar scoliosis (Figure 3), convex to the left. There was pes vavus and equinovarus deformity of the left foot (Figure 4). There was no atrophy. There was weakness (-2 to -3) of all the left leg muscles and (-1) of the right quadriceps and hamstrings. Stretch reflexes in the legs were normal except for the right ankle jerk, which was barely obtainable. The plantar responses were downgoing on the right, definitely upgoing on the left (positive Babinski). There was diminished vibratory sense in all toes of the left foot, no other sensory abnormality. The rest of the neurological examination was normal.

Routine laboratory studies, including C.B.C., S.T.S. and urinalysis, were all within normal limits. Chest x-ray showed "fusion of the posterior portions of the 5th and 6th ribs on the right, which arise from a common point" (Figure 5). Spine films showed "narrowing between T₅ and T₆, with non-visualization of the pedicles on the right side; this is the area of common origin of the right 5th and 6th ribs. Spina bifida of D₁₁ and D₁₂ and of all the lumbar vertebrae, (Figure 6) with odd configurations posteriorly. Narrowing between D₁₁ and D₁₂." X-ray summary: "Extensive congenital anomalies of rib cage, thoracic and lumbar spine." Films of the November, 1953 myelogram at Grace Hospital (courtesy of Dr. Gurdjian) were obtained (Figure 7), and the Grace Hospital report: "Myelographic examination demonstrated no abnormality in the cervical area. In the lumbar area the column divides into two at the level of L₃ and reunites into a single column at D₁₂. This is highly suspicious of an abnormal segmentation of subarachnoid space in the upper lumbar area into two distinct columns which subsequently reunite. No evidence of obstruction, and dye flowed freely throughout subarachnoid space." (This clinched the diagnosis of diastematomyelia) (Figure 7).

Without anything more than supportive therapy the patient became symptom-free within three or four days. Lumbar puncture had not been done because of the exquisite low back pain and tenderness. We felt the clinical picture was consistent with long-standing lumbosacral myelo-radiculopathy secondary to diastematomyelia.

This patient's picture, then, is illustrative of and in keeping with a fairly typical diastematomyelia when the adult stage is reached—the history of chronic low back



Figure 1
Dimple over spina bifida

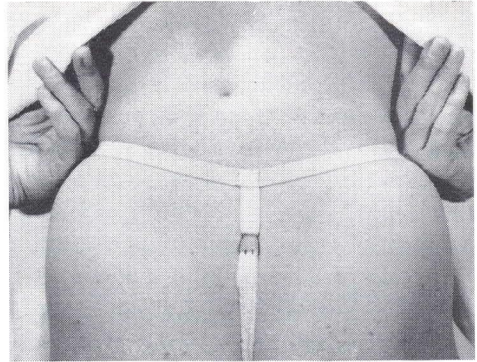


Figure 2
Boggy swelling over lower back

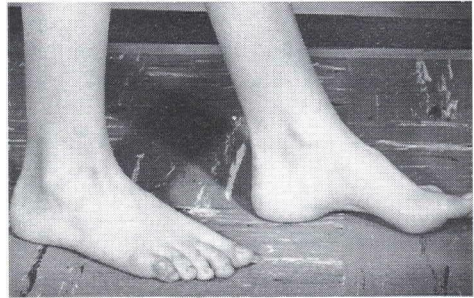


Figure 4
Pes cavus and equinovarus deformity
of the left foot

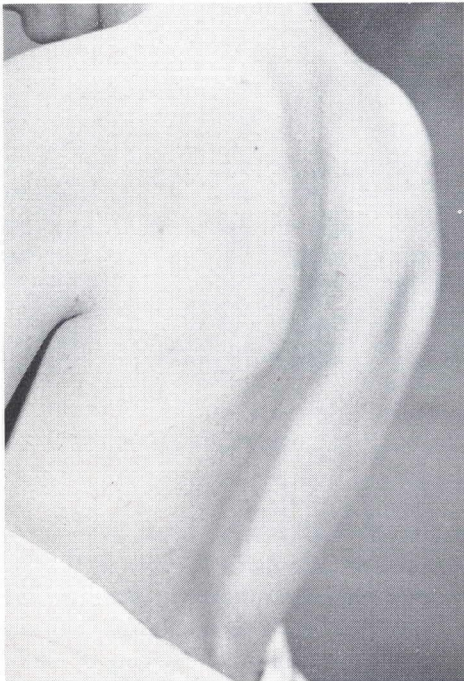


Figure 3
Dorsolumbar scoliosis

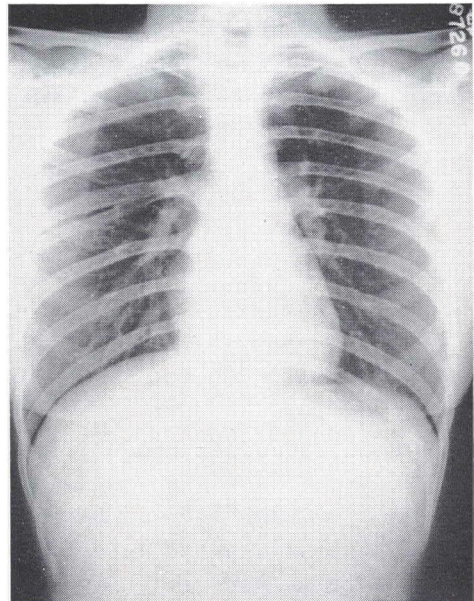


Figure 5
Fusion of 5th and 6th ribs on right side

pain and leg pain with recurrent acute episodes of head pain and low back pain; the lumbar lordosis and scoliosis and left foot deformity, the midline dimple over the level of spina bifida occulta and the poorly circumscribed mass of subcutaneous fat over the low back; mixed upper and lower motor neurone signs in lower extremities with minimal sensory change; the radiologic clues of a widened neural canal, a midline bony spicule, extensive spina bifida occulta and other congenital abnormalities, including posterior fusion of the right 5th and 6th ribs. We can not be sure if the recurrent acute headache and neck rigidity are related to recurrent subarachnoid hemorrhage secondary to an independent intraspinal tumor or to transient, incomplete blockage of the subarachnoid and cisternal pathway at the foramen magnum, as would occur more permanently in the Arnold-Chiari syndrome.

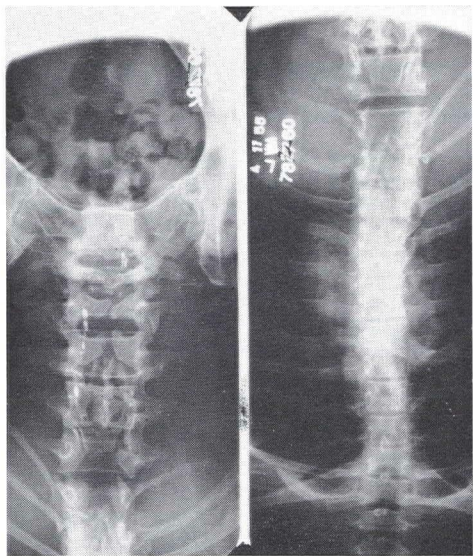


Figure 6
Spina bifida of 11th and 12th
dorsal vertebrae

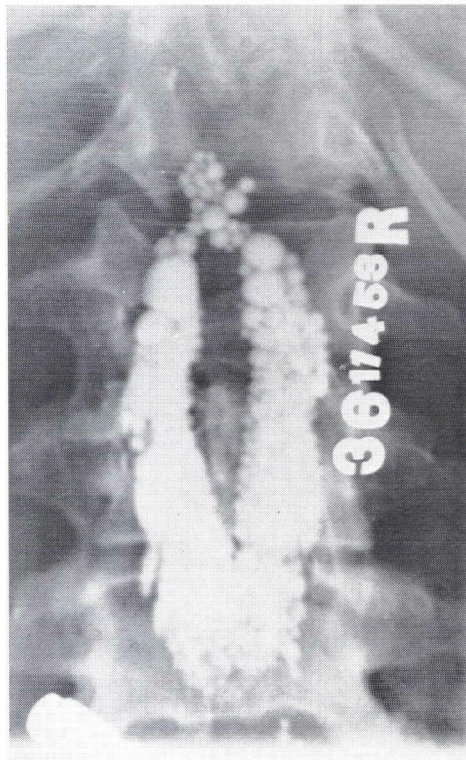


Figure 7
Lumbar myelogram

SUMMARY

Diastematomyelia, a congenital malformation of the cord or cauda manifested by splitting over a varying number of segments and transfixing of the intraspinal contents by a midline spicule, leads to progressive neurologic deficit in the legs and sphincters and ultimately to the Arnold-Chiari syndrome and hydrocephalus; it is not rare. Most importantly, it is treatable when recognized early in infancy or early childhood, and diagnosis is made easy by very definite x-ray signs, as well as definite leads from history and physical. These clues have been listed.

The most important factor in the diagnosis of diastematomyelia is awareness of the possibility of the lesion.

An illustrative case report is given.

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