LATE SYMPTOMS FROM DIASTEMATOMYELIA

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From the existing information in the literature on autopsy material, it was inevitable that eventually diastematomyelia (cleft or division of the spinal cord) would be diagnosed and operated upon during adult life.

CASE REPORT

E.F. (L#26979) was admitted to the Robert Long Hospital of the Indiana University Medical Center on June 19, 1959, with the chief complaint of pain of increasing severity in her left leg. She was a 43-year-old white housewife with a severe limping gait with the indication that her left leg was short. She had been well until 3½ years before when she noted the onset of pain in the left hip and leg with the result that she began to limp. This gradually progressed in severity, and she consulted physicians who took roentgenograms of her spine.

Fig. 1. Views of lower back. Note that the majority of the hair arises from the right. Marked lordosis is evident.

Fig. 2. (A) There is nonfusion of the neural arches of the 2nd, 3rd, 4th, and 5th lumbar vertebrae. There is fusiform widening of the interpedicular space without evidence of erosion of the pedicles. A midline bony spicule arises from the body of the 3rd lumbar vertebra and is projected over the 2nd intervertebral space. (B) There is narrowing of the anterior posterior diameter of the bodies of the 3rd and 4th lumbar vertebrae. "The bony spicule arising from the posterior surface of the body of the 3rd lumbar vertebra can be well seen" (preoperative note by Dr. Eugene C. Klatte, Radiology Department).
She was told that nothing could be done for she had a deformity with which she had been born. She was provided with a lift for her left shoe. As the pain increased and the gait became worse, she asked for consultation at the Medical Center. She had apparently lived a normal life until the age of 38, having had four pregnancies and four fully grown normal children.

**Examination.** There was marked lordosis with a severe pelvic tilt to the right, tending to pull the left leg up. On measurement, the legs were of equal length and all measurements of girth of the limbs were equal. The feet had normal arches. In the lumbar area there was much pain on pressure surrounding an area of reddish skin, almost entirely to the right of the midline. This area was covered by a heavy growth of long black hair (Fig. 1). There was a deep midline dimple over the sacrum. There was mild compensatory scoliosis with the apex to the right in the lower dorsal region. Upon palpation, there appeared to be bifidity of the last four lumbar and sacral posterior spines, with an unusual bony element in the midline at the level of the 2nd and 3rd lumbar vertebrae. The left leg seemed to be fixed at the hip and the knee showed evidence of contracture. There was pain with straight leg raising on the left and tenderness over the course of the left sciatic nerve on pressure. The remainder of the findings were singularly without major abnormalities. Sensation was intact in all modalities and motion was present in all areas, although weaker on the left. The left ankle jerk was present but not as brisk as on the right.

Roentgenograms taken shortly after disability began revealed the presence of multiple spinal congenital anomalies (Fig. 2) with the suggestion of a midline bony spicule. Films taken at the time of admission showed the same anomalies with evidences of changes in the left hip joint (Fig. 3). There was fusiform widening of the interpedicular space without evidence of erosion of the pedicles.

**Clinical Diagnosis.** Upon review of the information at hand, major consideration was given to intraspinal lipoma, intraspinal meningocele, epidural granuloma, and diastematomyelia. The arguments weighed so heavily toward diastematomyelia that this was indicated as the primary diagnosis when surgical exploration was suggested.

**Operation.** On June 22, 1959, under general endo-

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**Fig. 3.** Note progressive degenerative changes in the left hip from 1955 (A) to 1959 (B).

**Fig. 4.** View of spinal cord as seen at operation. The bony spicule has been sectioned flush with the surface of the cord and is covered with blood clot. The cord is divided into two unequal parts by the spicule.

**Fig. 5.** Drawing based on surgical and roentgenological views of the anomaly.
numb feeling in the right leg, but there were no sensory or motor deficits definable. Function of the bladder was good, and the gait improved immediately, assisted by ultrasound therapy to the left hip and knee. The patient was gainfully employed within 2 months of discharge and her gait has improved considerably in the past year.

**DISCUSSION**

Diastematomyelia is no longer an extremely unusual diagnosis. At the Children's Medical Center in Boston, 22 cases in which operation was performed have been reported. Examination of the records at the Indiana University Medical Center's James Whitcomb Riley Hospital for Children revealed 7 patients operated upon in the past 10 years. All of these patients were children. Of the other reported patients who were operated upon, only 2 were in the adult range, and both of these had suffered severe neurological deficits during growth.

The most important point about a unique case such as this one is not that it is rare just because it is the first reported, but rather that it becomes obvious that congenital anomalies must be considered more carefully in the future in the evaluation of neurological disorders. It would appear that earlier recognition of the basic disorder could have altered the course of the severe changes in the joints. There is an ever-present tendency on the part of physicians to view with disdain the multiple anomalies found in the lumbosacral region. The very frequency of these anomalies discovered in patients without primary complaints referable to the region is cited as the basis for this disdain.

Are there congenital anomalies that should be given primary consideration for attempts at early surgical correction? It would have been difficult indeed to have been confronted with the roentgenograms of this patient at the time when no functional disability was present and to have come to a decision to explore this lesion surgically when symptoms were negligible. Once neurological deficits appeared, would there be any point in attempting surgical correction? The postoperative condition of this patient would speak for the affirmative.

**SUMMARY**

A case of diastematomyelia is presented in which symptoms first occurred late in adult life. Surgical intervention provided almost total relief of symptoms.

I wish to thank the Department of Illustration for devoted efforts in obtaining the reproductions used in this paper.

**REFERENCES**


