DUPLICATION OF THE SPINAL CORD (DIPLOMYELIA)
AN ACCOUNT OF A CLINICAL EXAMPLE WITH A CONSIDERATION OF OTHER REPORTS

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On Dec. 3, 1946, a 14-year-old girl (B. P., R. I. H. 414450) was admitted to the Neurosurgical Department of the Rhode Island Hospital, complaining of pain and numbness in the right leg and foot, with disability in walking. Her family history was not remarkable. She had had an enlarged thymus, scurvy, and cyclic vomiting as an infant. At 10 years of age she began to limp and walked with the right foot turned in.

Examination showed a small, short right foot with high arch and persistent inversion, with atrophy and general weakness of the muscles of the entire right leg; there was anesthesia to light touch on all the toes on the right, with hypesthesia to touch and pinprick up to the knee, and normal temperature sense. Reflex findings were normal except for an absent knee jerk and a sluggish ankle jerk on the right. Roentgenograms of the skull and right foot were normal; the spine showed flattening and deformity of the 10th, 11th and 12th dorsal bodies, with narrowing of the interspaces, partial absence of the 11th and 12th dorsal neural arches, and complete absence of the 1st lumbar arch. Lumbar puncture findings were normal except for a protein content of 62 mg. per cent.

Pantopaque myelography (Fig. 1) was done by Dr. Thomas McOsker; and Dr. Lawrence Martineau, the roentgenologist, reported a split in the oil column at the level of the congenital defect, with a large irregular defect at the 10th dorsal body without complete obstruction; he felt that this appearance was consistent with a tumor mass of undetermined origin, but that it might

be associated with or be a result of congenital deformity. Dr. Ernest Burrows of the Neurological Department then saw the child in consultation; he believed that she was suffering from a developmental anomaly of the spine and cord, but that she was entitled to exploration to rule out the presence of tumor.

Operation. On Jan. 16, 1947, an incision was made centering on the 10th dorsal spine, and the spines and laminae from the 9th to the 12th dorsal vertebrae were exposed. The spine of the 9th appeared to be normal; that of the 10th was composed of two fused bulbous masses; and the neural arches of the 11th and 12th were incompletely closed. The spinous process and laminae of the 9th vertebra were removed, and the fused mass representing the spinous process of the 10th was then carefully rongeured off. As this mass was being removed it became evident that there was a ventral projection of bone impinging on the dura and its contents. The incompletely fused arches of the 11th and 12th vertebrae were therefore removed to give better exposure, and the removal of the spine and laminae of the 10th was then completed.

The ventral bony projection, which had at first been thought to be somewhat in the nature of a stalactite, was found to pierce the membranes and the cord and to be firmly attached to the underlying vertebral body. The dura was opened above the bony spur and this exposed an apparently normal cord which divided to pass around the bony partition (Fig. 2). Opening of the dura below the obstruction showed that the split in the cord continued, with obvious separation of the two parts, for at least 2 inches. At this point the parts of the cord came to lie in apposition but did not ap-

Fig. 2. Drawing of the condition found at operation after removal of the spines and laminae, with the membranes opened.
pear to fuse into a single cord as far as they could be seen. At the point of its attachment to the spinous process the bony partition was thick, about ½ inch across, but it narrowed down like the keel of a yacht as it approached the vertebral body. It was about ⅓ inch in length along the canal. Around this bony partition, the divided cord was enclosed in two separate tubes of dura and arachnoid (Fig. 3); below the obstruction the divided cord had a single dural and arachnoid covering with separate pial envelopes. It was found possible to remove the bony partition without injury to the nerve elements by rocking it off as one would remove a tooth. The dural envelopes were then retracted and the jagged end of the partition was removed with rongeurs. The dura was closed with interrupted silk stitches and the wound was closed in the usual manner.

Postoperative Course. The girl made a very good recovery. Dr. Burrows was asked to re-examine the child to see if he could discover neurological signs associated with the division of the cord. His findings were essentially the same as those previously recorded and he concluded: "One might expect dissociation of sensation, but there is none. There is probably duplication of the tracts in the cord."

The patient was discharged on her 25th postoperative day. At this time her pain had disappeared, she could move her toes and foot better, and her anesthesia had lessened to a considerable degree.

Addendum

Since the preparation of this paper the girl was examined again on Jan. 13, 1949, 2 years after the operation. She then said that she had no pain and was not troubled by any sense of numbness in the leg. She walked well without any limping. The atrophy of the right leg muscles had disappeared to a large extent, their size and strength being almost equal to that of the corresponding muscles in the other leg. The tendency to inversion of the foot had gone, the height of the arch was less, and the foot itself was almost as large as the one on the left. The knee jerk was still ab-
sent and the ankle jerk was still sluggish. The hypesthesia had disappeared, and the anesthesia of the toes was limited to the three outer ones. Contemplated orthopedic corrective operation will probably not be needed.

PREVIOUS REPORTS

As neither my associates nor I had ever met a condition similar to this, we were at a loss for an explanation or even for a name until one of my interns, Dr. Thomas Mathieu, came across the article by Marr and Uihlein. This led us to a fairly extensive literature on the subject, which I shall now review briefly.

In an interesting pathological study of the artefacts of the nervous system, Van Gieson, in 1892, described a series of congenital deformities of the cord associated with monstrosities and incompatible with extrauterine life, as follows:

(1) Amyelia—complete absence of the cord associated with absence of the brain.
(2) Atelomyelia—partial development with various sections of the cord missing, usually associated with absence of the brain.
(3) Diastematomyelia—a portion or the whole of the cord split into lateral halves, in which some fusion may occur.
(4) Diplomyelia—true duplication of the cord found in various kinds of double monsters.

Steizer in an inaugural dissertation in 1895 reported 35 cases of cord duplication and added 1 of his own. I have been unable to consult this dissertation, but quote from Bruce, McDonald and Pirie.

In 1906, Bruce, McDonald and Pirie reported a case of partial doubling of the spinal cord. This occurred in a woman 50 years of age who had no evidence of such an abnormality during life, but at postmortem examination was found to show division of the cord in the lumbar region with reunion below.

Zalewska-Ploska, in 1913, reviewed the literature and added 2 cases of her own. The first patient was a 50-year-old woman with spina bifida occulta and hypertrichosis, without symptoms referable to this condition. At postmortem examination she was found to have doubling of the cord at the 2nd lumbar segment. The second patient was an 8-month-old girl with subcutaneous myelomeningocele and doubling of the cord. In very beautiful plates, the author shows that there were two complete cords in the lumbar region, rotated at 90° to each other, each with a central canal and a ventral fissure. In this instance, the condition was found at operation, but the patient did not survive.

Weil and Mathews, in 1935, reported the case of a 5-month-old child, with meningocele and spina bifida, who died of an ascending urinary infection. Postmortem examination showed that the cervical cord was normal. The middorsal region showed hydromyelia, and the lower dorsal cord was
divided completely. Sections showed mirror images turned at an angle of 90° with the anterior fissures facing each other. There were two separate anterior horns in each cord, but the posterior horns were united into one at the middle of the lateral circumference of each cord. There were also elevated cylindrical ependymal cells in each central canal.

In 1936, Hamby reported doubling of the cord in a young woman of 20. In this case, there was a history of spina bifida in the female members of the family. The patient had pain in the lumbosacral area radiating to the right hip and leg, with stiffness of the right leg, and there was a profuse growth of hair over a dimple in the lumbar area. The right knee jerk was slightly exaggerated, there was anesthesia in the saddle area, and a band of hypesthesia down the backs of both legs. X-rays showed distortion of the laminae of the 3rd, 4th, and 5th lumbar vertebrae and most of the sacrum with a failure of fusion in the midline, and congenital fusion of the bodies of the 4th and 5th lumbar vertebrae. At operation, a hair cyst was found underlying the dimple, with a pedicle extending down to a bony spur attached to the vertebral body, the cord being completely separated around this spur. The patient made a good recovery from the operation.

Lichenstein, in 1940, made an interesting and quite exhaustive study of spinal dysraphism, in the course of which he reported 1 instance of cord doubling. In this case there was hydromyelia of the thoracolumbar segments, separation of the central canal into two main branches, and disastasis of the lower cord into two completely separated portions with a common dural envelope. There were secondary diverticulum-like outpouchings of the central canal with multiple atypical central canal formations, and fusion of the subependymal gray substance into irregular areas of so-called primary ependymal gliosis. He felt that this was diastematomyelia and not a true duplication, and he came to the conclusion that all previously reported cases were only pseudoduplications, and that no true doubling had ever been demonstrated.

Later in 1940 Herren and Edwards wrote an excellent review of the reports up to that time. They studied all available records and selected 42 cases as having true duplication of the cord, adding 1 more report of a patient on whom they had operated. The condition was found to be variously named as doubling, duplication, reduplication, bifurcation, and diastematomyelia, but they preferred to call it diplomyelia. It occurred about equally in the sexes, from fetal life to 76 years of age, but only 3 of the cases were found in adolescent or adult life. More than 2 of the instances were associated with spina bifida, usually occult. In practically all cases where there was associated meningocele or myelomeningocele death occurred at an early age, probably from infection. The spinal canal was found to show deformity varying from moderate dilatation to partial doubling; this latter condition was found in 3 of the cases in which there was a midline cone-shaped osseous or osseochondromatous process with its base attached to or continuous with the dorsal aspect of one or several vertebral bodies and with its apex directed into the
lumen of the spinal canal. This process was usually single but there were occasionally two or more; in some instances, a band of connective tissue, sometimes containing cartilage, took the place of the bony spur. Defects in the vertebral bodies, consisting of underdevelopment or absence of several bony segments, occurred in 10 per cent of the cases. The extent of duplication was usually limited to about 10 spinal segments in the lower dorsal or lumbar region, only 2 cases being found above the middorsal area. In all instances the cords were lateral to each other, and each appeared to have rotated through an angle approaching 90°, so that the ventral gray columns of the two cords faced each other and the ventral fissures lay in about the same frontal plane. In several cases, each cord was associated with four primary nerve roots; a well-formed pair of dorsal and ventral roots lying laterally and a rudimentary pair lying medially, these latter roots being usually associated with underdeveloped gray columns. Where doubling occurred, it usually continued down through the sacral segments, but in a few instances there was fusion below, with the re-establishment of a single well-formed cord. There was occasionally hydromelia in the cord above the division. Abnormal development of the brain was definitely an exception to the usual finding. The most common associated deformity was club foot; there was 1 instance of double ureter and kidneys. No specific clinical features were found to be common to the cases studied.

These authors felt that this condition cannot be explained by arrested development, as there is no normal embryonic stage in which the cord is double. They considered it a primary alteration of the cord, with the bony changes secondary, as the cord is laid down long before the bone. They pointed out that, in the chick embryo, as the lateral extremities of the neural plate approach the midline dorsally they may be directed ventrally for a short distance. Usually they retreat dorsally, fuse, and unite to complete the tube. If they should continue ventrally, they might fuse in such a way as to form two medullary tubes which might go on to form two separate cords. If the distance between these cords were small, the undifferentiated mesenchyme might develop into pia mater; if it were greater, arachnoid, dura, connective tissue, cartilage, or bone could form in this area. After careful study, they came to believe that true doubling of the cord represents an incipient form of twinning.

The patient on whom they operated was a 23-year-old woman with a left club foot which had ulcerated many times. The muscles of this leg were atrophic, and there were no knee or ankle jerks. There was fusion of the bodies from the 8th to the 11th dorsal vertebrae and there were bifid spines from this point to the 3rd lumbar vertebra. At operation, it was found that the arch of the 11th dorsal vertebra was inverted so that it bisected the cord, and was attached to the underlying body. This partition was removed and the patient made good recovery, to die of another disease about 5 months later. Postmortem examination showed division of the cord from the 7th dorsal level down, with a common dural envelope except at the region of the
bony partition. In the area where the dural tubes were doubled, there was a dentate ligament from the medial aspect of each cord. The pathological findings in the cords were similar to those which they had previously described.

Marr and Uihlein,5 in 1944, reported operating on a 12-year-old girl with upper thoracic scoliosis and kyphosis, club feet, and spastic gait. The left lower extremity was hypersensitive to cold, and there was numbness of the left great toe. The left leg was weak, the toes on this side could not be dorsiflexed, and there was some incontinence and retention of urine. X-rays showed fusion of the 3rd and 4th ribs posteriorly on the right, a 2nd lumbar hemivertebra, fusion of the bodies of the 3rd, 4th and 5th lumbar vertebrae, lumbar scoliosis and kyphosis, and sacral spina bifida. Pantopaque studies showed a partial block to the passage of oil at the level of the 3rd lumbar vertebra. The protein in the spinal fluid was 30 mg. per cent. At operation a narrow bone spicule was found extending from the body to the arch of the 3rd lumbar vertebra. At this point the cord was completely divided into two parts for a distance of about 4 cm. The doubled cord had a common covering of dura. The bone spicule was removed, and the patient made a good recovery, being discharged in 15 days.

In 1946, Maxwell and Bucy6 reported a clinical case of diastematomyelia. This occurred in a child of 9 months who had had a dimple in the lumbar region since birth, but who was otherwise apparently normal, with no motor or sphincter disturbances. X-ray examinations showed anomalous development of the lumbar vertebrae, with a calcareous shadow between the 3rd and 4th vertebrae. At operation a bony spur was found extending dorsally from the body of the 3rd lumbar vertebra. The cord was completely separated into two portions at this point and each part had an individual envelope of dura. The spur was removed and the child made a good recovery. Four months later the child was re-examined; walking and talking were then normal, and the only abnormal neurological finding was a suggestive left Babinski response. The authors followed Lichtenstein in feeling that these cases represent a splitting of the cord into two lateral halves, rather than the formation of two relatively complete cords.

SUMMARY

Duplication of the spinal cord, variously called doubling, reduplication, bifurcation, diastematomyelia, and diplomyelia, may occur in varying degrees. There may be an abortive attempt at doubling which results in partial separation. Entire division into parallel structures over a short or relatively long distance may occur, and the resulting cords may be incomplete, or each may contain practically all the elements of a normal spinal cord. They may be separated by pia only, or there may be arachnoid, dura, connective tissue, cartilage, or bone intervening. In the most marked examples there may even be doubling of the lower spinal column. Some observers4,6 have felt that only pseudoduplication ever occurs; it does not seem possible to agree with this
contention in view of reports showing two practically complete cords, each with its full complement of nerve roots. The most reasonable explanation of the condition seems to be that offered by Herren and Edwards: that it is a primary alteration of the cord, with secondary bone changes, and that it represents an incipient form of twinning.

Available published reports, covering 46 cases, are reviewed. In most of the patients the condition was found at postmortem examination, and in many instances it was not suspected during life. A recent clinical example from the author's own experience is reported in some detail. This account brings the total number of cases to 47. In 6 instances the condition was found at operation, and 5 of these patients survived.

REFERENCES