DIASTEMATOMYELIA

BY

C. C. MICHAEL JAMES and L. P. LASSMAN

From the Departments of Orthopaedic Surgery and Neurological Surgery, Newcastle upon Tyne General Hospital

(RECEIVED FOR PUBLICATION MAY 12, 1958)

Diastematomyelia is a rare condition and in our opinion is tending to be recognized more frequently by the radiologist, but its clinical significance does not appear to have been stressed. The purpose of this communication is to describe a case of this syndrome and to emphasize the importance of surgical treatment.

The term diastematomyelia is used to designate a congenital condition in which the spinal cord is divided longitudinally into two portions over some part of its extent. According to Cameron (1957) the term diastematomyelia was originally used by Hertwig in 1892 to describe a split in the spinal cord which occurred as a result of experimental interference with frogs' ova. Diastematomyelia becomes of clinical importance when there is a septum across the neural canal passing antero-posteriorly between the two portions of the spinal cord. The septum may be of bone, fibrous tissue, fibro-cartilage, or a mixture of these; it transfixes the spinal cord and divides partially or completely the neural canal antero-posteriorly in the midline. It most commonly affects the lower thoracic and lumbar regions but it has been described in the thoracic region as high as T.V.2 (Cowie, 1951). A similar septum may be found, in the absence of diastematomyelia between the nerves of the cauda equina, some of which may be adherent to the septum and diverted from their normal course. It has been found to be associated with other congenital anomalies of the vertebral column, particularly spina bifida, often of the spinal cord as in myelocoele and occasionally with dermoid cysts (Matson, Woods, Campbell and Ingraham, 1950).

Some authors prefer the term diplomyelia for this condition because they regard it as the result of an attempt at twinning (Herren and Edwards, 1940; Benstead, 1953) but Cameron (1957) suggests that it is a failure of development associated with the delayed closure of the human equivalent of the blastopore. Cameron also, with Kapsenberg and Lookeren (1949), regards the presence of a septum as being due to the inductive influence of the split

spinal cord upon the closely associated mutual development of dura and bone column, the septum being an abortive attempt at the formation of a midline neural arch by inversion of the laminae.

A septum is liable to fix the spinal cord so that traction is exerted on it as the vertebral column grows in length relatively faster than the cord, a process usually described as 'ascent' or 'migration' of the cord. The effect of transfixion becomes more marked with growth and, if it is severe and unrelieved, paralysis is likely to ensue owing to tension upon the cord itself, upon the nerve roots, and possibly owing to reduction of blood supply by traction on the blood vessels. A septum, therefore, is likely to be the cause of a progressive neurological deficit in the lower limbs, disparity of size of the lower limbs, incontinence, paresis, trophic changes and even paralysis.

The condition is usually recognized in childhood but has also been reported in adults in their twenties. The presenting symptoms are usually progressive weakening or failure to develop strength in a lower limb, or deterioration of gait. Occasionally the parents have brought the child because they were anxious about the presence of a hairy patch or naevus on the back and a radiograph has demonstrated diastematomyelia (Matson *et al.*, 1950).

Case Report

A girl, aged 10 years, attended an orthopaedic clinic complaining of discomfort in the right fourth toe in relation to the plantar aspect of the metatarso-phalangeal joint. No obvious cause for this was found but the appearance of the lower limbs attracted attention. The legs were of equal size and the feet normal except that the right foot was slightly smaller than the left. The skin of the legs and feet was livid and the circulation poor although it was a warm day. The whole appearance resembled that commonly seen in children with a myelomeningocele. Her mother said the girl frequently suffered from chilblains and that she always had 'very poor circulation in her legs', which went blue and mottled very easily; they never went 'dead' or painful. There was no history of bladder disturbance.

On examination of the back there was no abnormality evident except an excess of sparse hair over the lumbosacral area. The right plantar reflex was equivocal on that day but on the next examination four weeks later was found to be definitely flexor. There was no weakness in the lower limbs. She was small for her age; her three sibs, all younger, were of normal height. On admission to hospital two months later her condition on examination was unchanged but it was not possible to elicit her right plantar reflex: there was no demonstrable loss of sensation. Diagnosis was confirmed by radiological appearance and by myelography.

Plain radiographs showed spina bifida of L.V. 3, 4 and 5, and S.1. There was also a dense bony opacity situated in the midline and apparently extending from the spinous process of L.V.2 to the posterior aspect of the body of L.V.3 (Figs. 1 and 2), which was confirmed by tomography as being a midline bone spur. On myelography, the myeloid column divided about 1 cm. below the bony spur and united again above the spur. A narrow, negative midline shadow was present for about 1.5 cm. above the spur (Figs. 3 and 4).

Operation. The spinous processes were removed, starting two segments lower than the expected site of the lesion. On removal of the spine of L.V.2 a bone pillar was displayed transfixing the cord. The pillar was extradural and closely invested by the dura; it was firmly attached posteriorly at the peak of the neural arch. The pillar at its narrowest was oval in section, 4 mm. in length (cranial-caudal) and 3 mm. wide. Its anterior attachment to the body of L.V.3 was not firm and was presumably fibrous.

The dura was opened, showing a division of the spinal cord into two equal parts, the split extending cranially from the pillar for 1.5 cm. All the roots were lateral to the cord. The split did not extend caudally from the pillar. There was a thin fibrous band ascending to a

higher level within the dura and above the bony pillar; the band was divided and the bone removed, piecemeal and with difficulty, through the very small hiatus in the cord since it was not possible to rotate the cord without dividing the nerve roots.

FIGS. 3 and 4.—Myelography demonstrates the separation of the myeloid above the spur by the fibrous membrane (Fig. 3) and below the spur by its base. (The spur is ringed in Fig. 3.)

The cord was free at the end of operation; it had not ascended when released. The conus medullaris was situated 1½ cm. below the pillar. Closure was normal.

The post-operative course was uneventful; for a few hours the patient had pain in the left buttock. About three months later she complained of pain in her left great toe, but this rapidly subsided.

Five months after operation, her legs were of normal appearance as far as her circulation was concerned; she had no pain but clinically there was slight weakness of the left flexor hallucis longus.

Nine months after operation, there was no abnormality to be found. The circulation was improved, but was still not completely normal, and she had had no chilblains.

Discussion

The preliminary radiographical examinations had quite clearly demonstrated what was actually found at operation. The curious 'narrow, negative midline shadow' seen in the myelogram was found to be due to the thin fibrous band within the dura lying in the midline cleft in the spinal cord. It was attached to the dura internally, both at the cranial edge of the bone pillar and posteriorly. It was not possible at operation to determine clearly if the fibrous band was also attached to the dura overlying the posterior aspect of the vertebral body. The myeloid column divided about 1 cm. below the bony spur because of the obliquity of the spur whose anterior attachment was more caudal than its posterior attachment.

FIGS. 1 and 2.—Survey views of the lumbar spine showing the bone spur lying in the sagittal plane.

As Matson *et al.* (1950) have pointed out, the septum passing antero-posteriorly across the spinal canal may not be sufficiently calcified in young children to show on a radiograph. If the presence of a septum is suspected, repeat examinations will be necessary later on, or, as our findings showed, the condition can be demonstrated by myelography and unnecessary delay in making a diagnosis thereby avoided. Also there are two other radiographical pointers which can indicate the possibility of diastematomyelia: increased inter-pedicular distance and spina bifida. Herren and Edwards (1940) found that the spinal canal is widened where there is congenital abnormality of the cord, and both Neuhauser, Wittenborg and Dehlinger (1950) and Cowie (1951) have pointed out that increased inter-pedicular distance without pedicular erosion is a sign of congenital spinal cord abnormality and is present in cases of diastematomyelia. If the views of Kapsenberg and Lookeren (1949) and Cameron (1957) on developmental anomalies of the spinal cord are accepted, it seems to be impossible for diastematomyelia, with or without a septum, to occur without a very close relationship to spina bifida.

The decision to operate was based on the knowledge that the effects of traction on the cord were likely to be progressive. There was no reason to delay by watching to find out if indeed there would be progressive deterioration of circulation and muscle tone in the lower limbs, particularly since these effects may not appear until the age of 23 years; they may be rapid and irreversible. It is these facts which make it important to make the diagnosis early and to act to correct the anomaly. In this patient, the conus medullaris of the spinal cord was lying at a lower level (lower border of L.V.4) than is normally found at the time of birth, a state which was not demonstrated pre-operatively, and its retention at this level had so far produced remarkably little neurological result.

It is not possible to explain satisfactorily the mildness of the neurological effects of the lesion. They were not in themselves causing any disability but were noticed quite by chance during a clinical examination for another complaint. The circulatory deficiency had been present for many years. There are, however, various factors to be considered, the age at which there is the greatest differential rate of growth of bone column and spinal cord, the point of fixation of the cord and the firmness with which the cord is tethered.

There does not appear to be any information about the levels of the conus in the years between birth and completion of growth, and it is not known

whether the bone column and spinal cord maintain the same rate of growth relative to each other throughout the growing period or whether, for example, they grow equally for a period and then the bone column grows much faster. Since the bone column and spinal cord are developed in close association with each other in embryo it is likely that they are closely associated throughout the post-natal growing period and it is reasonable to argue from what is known about the growth of the skeleton. The period of greatest increase in height is around puberty and it is likely to be at this time that the 'ascent' of the cord is most rapid. The dura and arachnoid normally grow at almost the same rate as the bone but keep a definite relationship with the spinal cord in that the nerve roots in the adult have only a short course within the main dural sheath before acquiring their own individual sheaths. The dura therefore is itself not normally under any great traction force and actively lengthens as the bone column grows, just as do the spinal nerves (Streeter, 1919). The spinal cord also grows, but more slowly than the bone; its length at the end of the growth period is greater than at birth. If for any reason the bone age of the individual is less than normal, the ascent of the cord will be equally delayed. In the present case the bone age was normal and the child had not attained puberty. Since there is relatively less ascent of the spinal cord at the cranial end than at the caudal end of the vertebral canal, as shown by the obliquity of the nerve roots in their course to the neural foramina, fixation caudally is more likely to produce neurological signs. In the present case the point of fixation of the cord was relatively low in the bone column and relatively high in the spinal cord but the cord was not immobilized. The bone septum was situated in a division of the spinal cord of about 1.5 cm. length and was at the caudal end of the division; when the septum was removed, the cord did not alter its position. This suggests that the spinal cord had already been able to ascend to a small extent but would in a short time have been prevented from ascending further except at the expense of the elasticity of the cord tissues with resultant pressure effects and increased neurological signs.

As already mentioned, this child was small for her age. Using the crown-rump measurement, but not as accurately as Provis and Ellis (1955), our case was 64.9 cm. long in the stem which corresponds with the average of 64.5 cm. and 66.9 cm. found in Edinburgh girls at mean ages of 6.54 and 7.49 years respectively. Her bone age as estimated by radiographs of her elbows and wrists was the same as her true age so that the delay in development of

neurological signs cannot be ascribed to late bone growth.

Summary

Diastematomyelia is a congenital division of the spinal cord in its length. A case is described where there was an associated bone spur passing across the spinal canal in the lumbar region between the two halves of the cord.

The operation findings are described and discussed in relation to the clinical signs and to the radiological appearances. Preliminary tomography and myelography clearly demonstrated the anatomical state but the clinical signs bore no relation to the abnormal position of the spinal cord, fixed as it was in the vertebral canal by a bone septum, with the conus medullaris at a level below that normally found at birth. The various factors associated with this abnormality are discussed in detail in an attempt

to explain the paucity of neurological signs in this case.

The need for early diagnosis and preventive surgery in cases of diastematomyelia transfixd by a septum is stressed.

Our grateful thanks are due to Dr. Sheila Cameron, radiologist, and Dr. Gordon Gryspeerdt, neuro-radiologist, Newcastle General Hospital, for their assistance in the diagnosis of this case.

REFERENCES

- Benstead, J. G. (1953). *J. Path. Bact.*, **66**, 553.
 Cameron, A. H. (1957). *Ibid.*, **73**, 195 and 213.
 Cowie, T. N. (1951). *Brit. J. Radiol.*, **24**, 156.
 Herren, R. Y. and Edwards, J. E. (1940). *Arch. Path. (Chicago)*, **30**, 1203.
 Kapsenberg, J. G. and Lookeren, J. A. van (1949). *Acta Anat. (Basel)*, **7**, 366.
 Matson, D. D., Woods, R. P., Campbell, J. B. and Ingraham, F. D. (1950). *Pediatrics*, **6**, 98.
 Neuhauser, E. B. D., Wittenborg, M. H. and Dehlinger, K. (1950). *Radiology*, **54**, 659.
 Provis, H. S. and Ellis, R. W. B. (1955). *Arch. Dis. Childh.*, **30**, 328.
 Streeter, G. L. (1919). *Amer. J. Anat.*, **25**, 1.