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DIASTEMATOMYELIA

Report of two Cases Submitted to Laminectomy

By

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Diastematomyelia is a rare congenital anomaly, associated with spina bifida. The term was first used in 1892 to describe a split in the spinal cord resulting from experimental interference with frogs' ova (3). Because of the publication of a number of articles in recent years (1–14), it now represents a well-known condition; however, to our knowledge no operated cases have previously been reported from Scandinavia.

Diastematomyelia simply means a bifid state of the spinal cord. The cord may be split by fibrous or bony tissue that extends from the vertebral body towards the lamina in the sagittal direction or a similarly localized area may be divided longitudinally without associated interposed tissue. In the latter cases aberrant tissue or even neoplasms may rarely interpose. In the former case the two parts of the cord have a dural sheath of their own (11).

When a condition of complete doubling of the spinal cord exists in a localized sector it ought to be termed diplomyelia. This represents a different entity than the above described abnormity, diastematomyelia, which is in contrast only splitting or division of the spinal cord. However, many authors use the terms synonymously.

The majority of cases with diastematomyelia manifest no clinical signs. The pathological mechanism is considered to be prevention of the normal ascent of the spinal cord within the vertebral column. Lateral pressure from the splitting structure has also been suggested as an additional factor. These signs are mainly manifested by neurological deficits in the lower limbs, bladder and bowel, and often appear early in life, supporting the hypothesis that they result from traction due to the different growth rates of the cord and the spinal column. Still there are cases described in which the first symptoms do not appear until

adult life. In the group with a common dural sheath, operation has frequently revealed aberrant posterior nerve roots passing from the bifurcation of the cord to or through the dura to a neural arch. At transection it could be seen that they exerted a traction effect. Whether or not this traction per se can result in neurological deficits is poorly understood (11).

The clinical signs are not specific for diatematomyelia since they may also occur in different types of spinal dysraphism, either alone or combined with diatematomyelia. The most frequent and earliest manifestation is usually retarded unilateral growth of one leg and foot often combined with a varus deformity. There may also be sensory loss, trophic ulceration and functional disability of the bowel and bladder. Even cases with paraplegia are reported.

When there are clinical signs of spinal dysraphism, a radiological investigation of the spine is indicated. Plain X-ray films in cases with diastematomyelia demonstrate widening of the spinal canal corresponding to the cleft bulbous widened cord and the extent of the spina bifida (which is most often occult) and sometimes additional anomalies like hemi- and fused vertebrae. If a sagittally directed bone septum is shown the diagnosis is definite. However, in cases with no interposition or with a fibrous septum not sufficiently calcified to show on radiographs, which is often the case in young children, myelography is needed for the diagnosis. This investigation is of further pre-operative interest as it gives information as to the site and extent of the anomaly. Gas-myelography is the method of choice. It provides the most accurate anatomic delineation of the entire anomaly, and it does not include the risk of arachnoiditis since the gas is quickly and totally absorbed. The latter advantage is especially worth considering when young growing patients are to be investigated, where impairment of the cord ascent could result.

Cases with diastematomyelia and neurological signs of spinal dysraphism require surgical correction of the anomaly. Because of the progressive character of the neurological deficits with the increasing risk of irreversible damage to the cord, this treatment should be as early as possible. Operative results showing some degree of improvement in two thirds of the patients with spinal dysraphism have been reported (10).

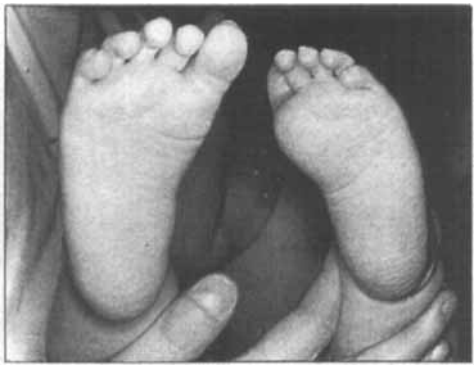


Fig. 1.
Clearly shows the underdevelopment of the left foot.

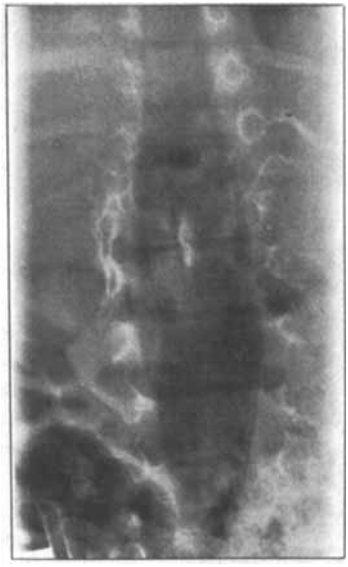
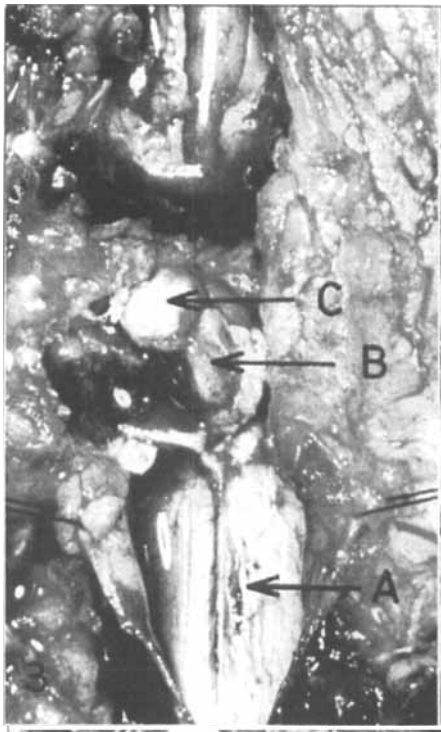


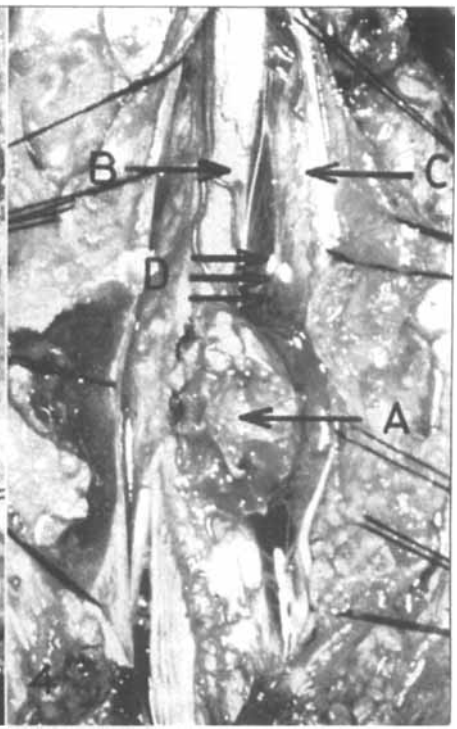
Fig. 2.
Gasmyelography demonstrates the bulbous widening of the distal spinal cord through which the bony spur is traversing.

CASE REPORTS

Case 1: 1½ year old girl born with a spina bifida occulta associated with a large hairy mole over the lumbar region. There was no family history of congenital anomalies. At several months of age it was observed that the left foot was smaller than the right but there was no history of abnormal sphincter control.

Physical examination at the age of 1½ years revealed the child would not walk





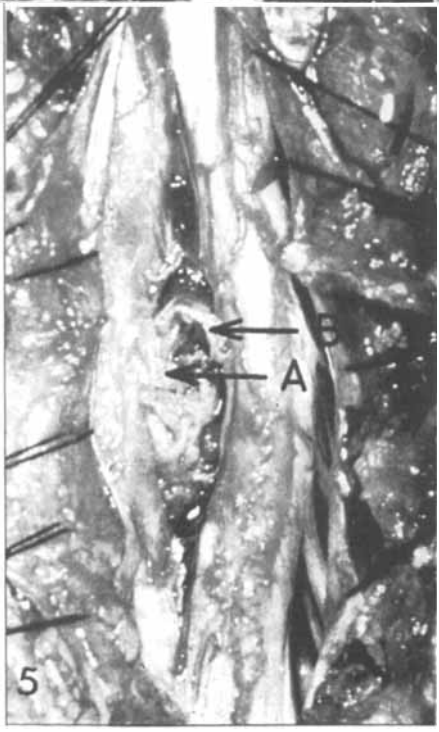


Fig. 3.
Laminectomy performed.
Arrow a) unopened dural sac.
„ b) bony spur.
„ c) dermoid cyst within bony spur.

Fig. 4.
Dural sac opened.
Arrow a) bony spur.
„ b) right half of spinal cord.
„ c) left side spinal cord.
„ d) adhesions.

Fig. 5.
Bone spur removed with its dural sheath.
Arrow a) remnant of bone spur.
„ b) dural sheath.

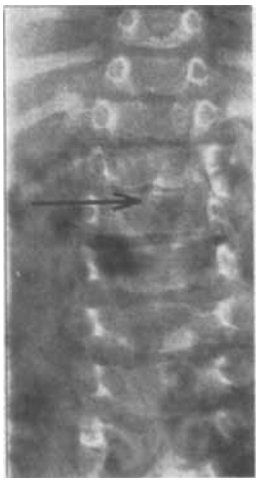


Fig. 6.
Shows widening of the spinal canal in the lower dorsal and lumbar region associated with a number of vertebral anomalies. The bony spur from Th 12 is indicated by the arrow.

or stand, the left lower extremity was smaller and the foot 1 cm shorter than the opposite side (Fig. 1). The left achilles reflex was absent. There was no paresis but slight equino-varus deformity of the left foot was noted.

Roentgen examination (Fig. 2) showed multiple malformations in the thoracolumbar region with hemivertebrae at T 11 and T 12. Bifid lamina at T 10 – L 2 associated with widening of the spinal canal in lower dorsal and lumbar region were present. Gasmyelography revealed localized spindleshaped widening of the caudal spinal cord with an elongated central defect extending from lower part of Th 9 to L 2. A bony spur extending from the lower posterior body to the lamina of T 12 and traversing the middle portion of the defect could also be identified. Intravenous pyelography was normal.

Laminectomy revealed that the bony spur contained a small dermoid cyst associated with a sinus tract extending to the skin (Fig. 3). The bony spur was enclosed in a dural sheath which was adherent to the adjacent spinal cord (Fig. 4). Numerous adjacent adhesions were incised and the bony spur together with its dural covering resected (Fig. 5).

Postoperative course was uneventful.

Case 2. A nine month old male child was noted at birth to have a spina bifida occulta. There was no family history of congenital malformations.

Physical examination showed slight, unilateral underdevelopment of the left lower extremity and moderate bilateral equino-varus deformity. Neurological examination was normal.

Roentgen examination revealed localized widening of the spinal canal in the lower dorsal and upper lumbar region associated with several bifid laminae (Fig. 6). Gasmyelography showed localized spindle-shaped widening of the caudal spinal

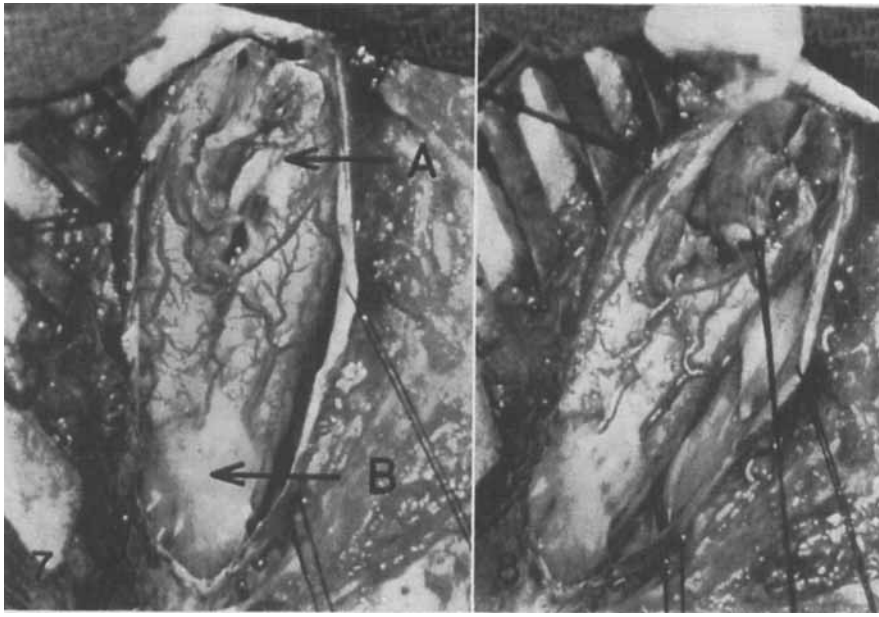


Fig. 7.
Case 2: Dural sac opened.
Arrow a) bonespur with dural sheath.
„ b) lipoma in conus.

Fig. 8.
Bone spur partly removed, adhesions severed.

Fig. 9.
Bone spur removed demonstrating the defect in the spinal cord.

cord with an elongated central defect which was traversed by a bone spur arising from T 12.

Laminectomy showed a bone spur similar to the previous case which was again enclosed in a dural sheath and associated with numerous adhesions to the adjacent cord (Figs. 7, 8, 9). There was an incidental finding of a small lipoma in the conus medullaris. The adhesions were incised and the bony spur and dural sheath resected. No attempt was made to remove the lipoma.

Postoperative course was uneventful.

SUMMARY

A short review of the main features and the treatment of diastematomyelia is presented and two cases submitted to laminectomy reported. The advantage of gasmyelography because of its superior delineation of the entire cord lesion and freedom from complicating arachnoiditis is emphasized.

RESUME

Un court aperçu des principales caractéristiques et du traitement de la diastematomyélie est présenté et deux cas soumis à une laminectomie sont rapportés. L'avantage de la myélographic gazeuse est souligné en raison de sa délinéation supérieure de la lésion entière et il est souligné qu'elle ne produit pas de complications de nature de l'arachnoïdite.

ZUSAMMENFASSUNG

Eine kurze Übersicht der Hauptzüge und der Behandlung der Diastematomyelia wird gegeben und über zwei Fälle, die einer Laminektomie unterworfen wurden, wird berichtet. Der Vorteil der Gasmyelographie wegen ihrer überlegenen Aufzeichnung der gesamten Strangbeschädigung und wegen des Nichtauftretens einer komplizierenden Arachoiditis wird hervorgehaben.

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