CONGENITAL LESIONS OF THE CRANIOSPINAL AXIS*  
REPORT OF FOUR CASES  
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Midline craniospinal anomalies include a variety of congenital malformations such as dermoids, dermal sinuses, lipomas and meningomyeloceles. Basically these represent various expressions of defective closure of the neurul tube together with associated involvement of mesodermal and ectodermal elements. Some of these conditions are rare and present interesting clinical features, diagnostic problems and technical difficulties in their surgical correction. The following case reports illustrate some of these congenital abnormalities.

Diastematomyelia is a rare developmental anomaly in which the spinal cord is split into two lateral halves by a band of either osseous, fibrous or fibro-cartilagenous tissue. The literature reveals association of this lesion with various other abnormalities, cutaneous, skeletal and neurological (Benstead 1953, Ingraham and Matson 1954, Ferret 1958, Seaman and Schwartz 1958, Rao and Dinakar 1970). Diastematomyelia is usually encountered in children, though a few cases have been reported in adults (Seaman and Schwartz 1958, English and Maltby 1967). This condition is more common in the female sex and most of the cases described were below the age of 14 years. Myelography is often diagnostic in which the opaque medium is split into two columns by the septum. Surgical excision of the septum is the standard procedure of treatment.

Dermal sinuses are the result of defective closure of the neural groove with inclusion of the epithelial elements in the developing spinal cord or its coverings. The epithelial tracts may extend to various depths and may expand to form dermoid cysts intraspinaly or they may be associated with other dysraphic manifestations (Rao and Dinakar 1971). The common site is the lumbo-sacral region. Less commonly they occur along the higher levels of the spine. Those which affect the cranium are usually situated in the midline in the occipital region or in the nasofrontal area.

CASE 1. Multiple Anomalies—Diastematomyelia, Dermal Sinuses and Dermoid Cyst. Baby T., a 3-month-old female child, first born to non-consanguinous parents, had a swelling over the occipital region and another over the mid-dorsal region. The swellings had been getting bigger gradually. There was dribbling of urine. On examination there was a cystic swelling over the external occipital protuberance (Figure 1) measuring 3 cm. in diameter with a sinus over its summit. It was not reducible, pulsatile or transilluminant. There was a diffuse swelling over the lower dorsal region in the midline. The skin showed bluish discolouration, hypertricho-
sis and a sinus in the centre from which hair was protruding. There was a dimple on the lower cervical region from which there was a yellowish brown discharge (Figure 1). The child had spastic weakness of both lower limbs and incontinence of urine. Other systems were normal.

Plain X-rays of the spine revealed widening of the lower cervical and lower dorsal spines. There were hemivertebrae at D7 and D8 and scoliosis. There was an oval defect of bone at the inion. A lumbar myelogram using 3 ml. of Myodil (R) and screening showed a filling defect in the centre of the dye column at the level of the hemivertebrae and also at the lower cervical region (Figure 2). When the dye was traced upwards it flowed around the defect in the occipital bone but none entered the swelling.

Under general anaesthesia, the occipital swelling was exposed through a transverse incision. The sinus was traced to a cyst which contained hair and pultaceous material. It was excised. The cervical sinus was traced through the bifid spine intradurally ending in a fibrocartilagenous spur which was excised. The sinus over the dorsal region was similarly dissected and was found to be entering the dura. On opening the dura a cartilagenous spur was found projecting backwards (diastematomyelia) and it was excised. The child did not thrive well after surgery, developed infection in the cervical wound and died on the seventh post-operative day. The child was discharged after a smooth recovery. The child is leading a normal life to date.

Haematoxylin and eosin stained sections revealed keratin and hair follicles and a healthy prior to its onset. The swelling was rapidly increasing in size and a few days before admission involved the left eye-lid also. There was no fever. On examination he was an active child. There was a diffuse swelling above the root of the nose which was cystic, not reducible. The left eye-lid was swollen and there was a small opening with purulent discharge from it. There was a small sinus over the middle of the nose on the dorsal aspect. On further questioning the mother admitted having noticed it from the time of birth without causing any obvious trouble. The pus yielded coagulase negative staphylococci.

Under general anaesthesia, the infected dermoid cyst was excised. The anterior table of the frontal bone was deficient and there was infected granulation tissue over the eroded posterior table. The lower end of the cyst communicated through a small hole at the lower end of the wound near the root of the nose. The wound healed well.

Through a vertical incision encircling the sinus on the dorsum of the nose, the sinus was traced upwards into the frontal region where the main mass was excised previously. The sinus contained cheesy material and hair. The wound was closed and the child was discharged after a smooth recovery. The child is leading a normal life to date.

Case 2. Frontal Dermoid Cyst with a Dermal Sinus. A one-year old boy was admitted with a swelling over the forehead of 2 months' duration. He was...