UNUSUAL CONGENITAL ANOMALIES OF NEUROSURGICAL INTEREST IN INFANTS AND CHILDREN*

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This report is based upon some of the rarer congenital anomalies encountered in the Neurosurgery Clinic at the University Hospital during the past few years. No attempt has been made to review the literature on this subject as this has been done so thoroughly by Ingraham and his associates. We wish rather to discuss the practical solution to some of the problems one faces in dealing with these unusual congenital malformations.

Case 1. B. W. was referred on July 5, 1949 to the University Hospital by Dr. R. Atchison of Northville, Michigan, 9 hours after delivery, because of a large congenital defect of the scalp. The pregnancy had been normal, but the description of the delivery is most interesting. Dr. Atchison was told by the nurse that the membranes had ruptured. It was a vertex presentation and on seeing only the shiny scalp defect, it was naturally thought that the membranes were still unruptured. He picked up a hemostat and was about to open the membranes, when fortunately at that moment the patient had a severe labor pain and with this further bulging he realized that it was the child’s head presenting. The remainder of the delivery was normal.

The family history records that a sibling with an entirely similar scalp defect, but no other recognizable congenital defects, died of meningitis at 12 days. This sibling had been seen by us 3 days post partum but the condition was unlike anything we had encountered previously and we deemed the lesion inoperable. The parents were not related to each other and there was no history of a similar lesion in either family.

Examination. A scalp defect, 5X7 cm., extended between the anterior and posterior fontanelles, in which there was no normal skin. The defect was covered only with a shiny, thin transparent membrane beneath which could be seen large venous spaces. It was well demarcated from the normal scalp. Palpation gave the impression of incomplete closure of the skull over the midline at the center of the defect (Fig. 1A). There was also present a deformity of one finger and both feet in that some of the distal phalanges were absent. Several hours after admission, the membrane became dry and lost its sheen.

X-ray films showed the anterior fontanelle to be within normal limits. The posterior fontanelle was difficult to make out, but there was no obvious defect in the calvarium between the two fontanelles as had been suspected on palpation.

Operation. As soon as studies were completed, the scalp defect was excised under local infiltration with metycaene ½ per cent. No abnormality was found in the underlying bone. After mobilization of the scalp bilaterally down to the ears

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and by means of rotation flaps, the entire defect was closed primarily (Fig. 1B). Convalescence was uneventful.

_Histological Examination_ (Dr. E. Cawley). "As the defect is approached, the epidermis becomes thinned and gradually fades out, exposing markedly atrophic corium approximately one-third the thickness of normal corium. There are no sebaceous or sweat glands. A striking feature seen in the atrophic portion is calcification of the hair follicles. Diagnosis: Congenital ectodermal defect of the scalp."

_Comment._ Congenital defects of the scalp are extremely rare, only 76 cases having been described up until 1930. The majority of these were in the same location. A search of the literature revealed no case subjected to surgery. Individuals with large defects have died as a rule of meningitis or hemorrhage from the sagittal sinus or the large veins beneath the defect. It should be stressed that these large congenital scalp defects should be operated upon as an emergency procedure. Procrastination will lead to infection making a plastic procedure most difficult.

**Case 2.** D. C., a 3-month-old white boy, was admitted on Jan. 8, 1947, because of a mass extending from the bridge of his nose (Fig. 2A). This appeared to have increased somewhat in size since birth and fluctuated when the infant cried. The mass, covered by normal skin, was walnut-sized and extended into the palpebral fissure of the right eye and almost into that of the left. A bony defect of the skull was palpable around the periphery of the mass. Neurological findings were entirely normal.

X-rays of the skull showed a spherical soft tissue mass presenting through a defect involving the medial halves of both supraorbital ridges and the inferior frontal area, as well as the expected position of the superior nasal bones.

**Operation.** On Jan. 13, 1947 the sac of the encephalocele was circumscribed superiorly by an incision down to the peristeme. The incision was then carried down to the root of the nose where the scalp and skin of the face were dissected from