CONGENITAL ABSENCE OF SKIN (APLASIA CUTIS CONGENITA)

A CASE REPORT

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Congenital absence of skin is a rare condition which was first reported in 1826. Until 1960 only 213 cases had been reported in the literature. It usually presents in the newborn as localized areas of skin deficiency, most commonly involving the scalp.

The aetiology of the condition is unknown, but various theories have been postulated. One of the earlier theories was that of the amniotic adhesion theory. Intra-uterine trauma because of pressure was also postulated.

There is no doubt that hereditary factors play a role because familial histories have been obtained.

In those cases where microscopical studies of the lesions were performed, it was found that there was an absence of sebaceous glands, hair papillae and sweat glands in the affected area. There was also an absence of elastic fibres in the corium of the affected area and to a certain extent in the surrounding areas. The condition was therefore compared with epidermolysis bullosa hereditaria, in which there is also believed to be a deficiency of elastic fibres in the corium.

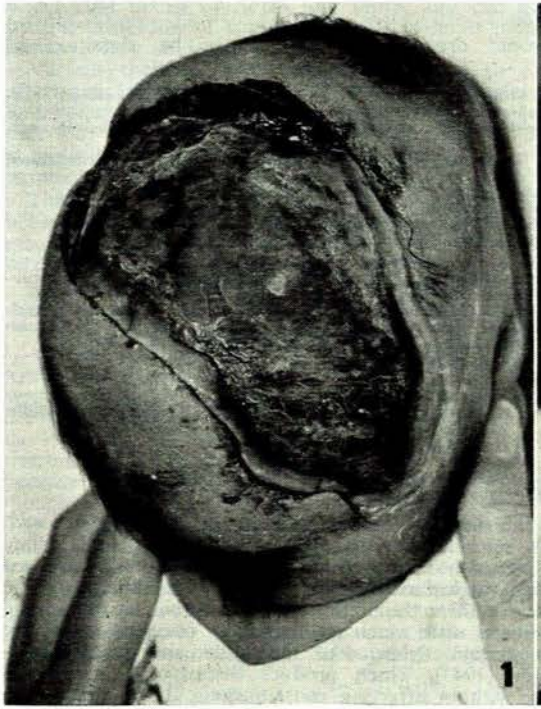

Fig. 1. See text.

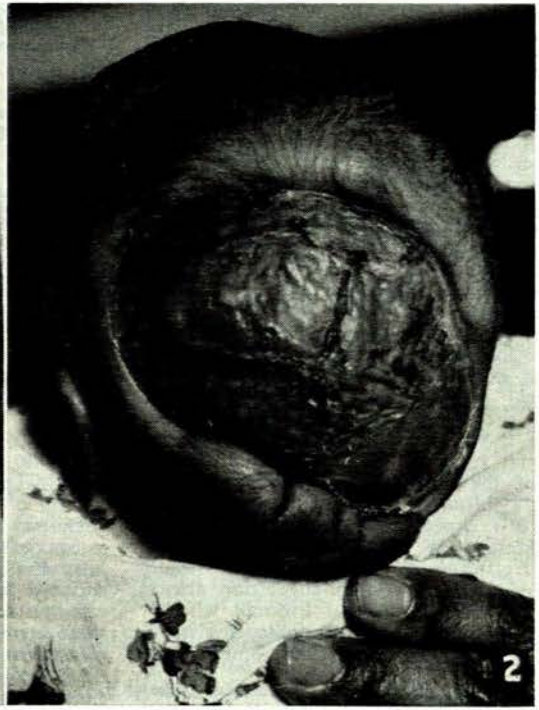

Fig. 2. See text.

Underlying bone defects of the skull are common and may be large.

The mortality in cases with scalp defects is about 20%, the main cause of death being meningitis.

The treatment in cases where only the scalp is deficient consists of a Thiersch graft and later, if necessary, rotation of scalp flaps. If there is a large defect of the underlying bone the defect should by preference be covered with a scalp flap,

and reconstruction of the bone should be considered at a later date.

CASE REPORT

The patient, a Coloured male child, was admitted to hospital on 4 December 1961 at the age of 8 days, with a history that the scalp over the vertex was seen to be very dark. The child was otherwise normal at birth, but became slightly jaundiced after 24 hours.

This was the first child and the pregnancy and labour were normal. The mother was in labour for about 24 hours and was not attended by a doctor or midwife. There was no family history of skin abnormalities.

On examination the patient was deeply jaundiced. His weight on admission was 5 lb. 12¼ oz.

There was an area over the occipital region about 4 inches x 3 inches which was covered by a dry, adherent, black scab. The surrounding skin was covered by very few hairs in comparison with the rest of the scalp (Fig. 1).

The total serum-bilirubin level on admission was 21 mg. per 100 ml. Poor growth of *E. coli* only, sensitive to 'chloromycetin', streptomycin and 'terramycin', was obtained from a blood culture and a pus swab from the affected area. The Wassermann and Berger tests were negative.

Skull X-ray on 15 December showed wide separation of the suture lines with some thinning of the parietal bones posteriorly and superiorly.

The patient was treated with intravenous fluids and chloromycetin. The local area on the scalp was treated by exposure on the same lines as a burn.

On 15 December the patient developed irregular twitchings of the hands and feet, which subsided again spontaneously. No other signs to suggest meningitis developed.

By the end of December the thick black scab had separated on its own and an area covered by fresh granulation tissue was left.

A Thiersch graft was applied to the scalp on 3 January 1962. The take of the skin graft was 100% and the scalp was healed 10 days after grafting (Fig. 2). The grafted area will be revised and adjusted again at a later date.

The patient was discharged three weeks after grafting.

I wish to express my thanks to Dr. J. Mostert, Medical Superintendent of the Red Cross War Memorial Children's Hospital, for his permission to publish this case, and Prof. J. H. Louw and Dr. David Davies for their advice and encouragement.

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