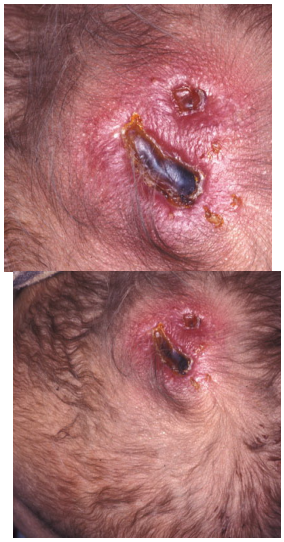

Congenital absence of skin

Congenital absence of skin is a rare condition which was first reported in

1826. Until 1960 only 213 cases hadbeen reported in the literature. It usually

presents in the new-born as localized areas of skin deficiency, most

commonly involving the scalp. The aetiology of the condition is

unknown, but various theories havebeen 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. Underlying defects of the skull bone are common and may be

large.The mortality in cases with scalp de¬fects is about 20%, the main cause of death

being menin¬gitis.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 pre¬ference be covered with a scalp flap,