



Aplasia Cutis Congénita

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Image illustrates a newborn with Aplasia Cutis Congenita

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Aplasia cutis congenita (ACC) comprises a heterogeneous group of disorders whose main feature is the focal absence of skin.^{1,2} It is a rare entity that affects both sexes equally and its etiology is still unclear.³ In most cases it occurs in the scalp, although other areas of the body may also be involved. Other congenital malformations have been reported in appearance with ACC, of which the limb defects appear to be a specific association.^{1,2}

The diagnosis is mainly clinical and histology is not used routinely. Several skin defects of varying size can be clinically observed. The characteristics of the lesions vary within a wide spectrum, ranging from fragile skin, scabs, pseudoampullar elements, and ulcers covered with false membranes to real atrophic scars.³

A classification based on different genetic entities has been suggested. ACC type I is limited to the scalp. Type II involves the body or scalp; type II A involves defects of the body or limbs. Type III is limited to the scalp or limbs. Type IV is associated with epidermolysis bullosa, and type IVA is Bart Syndrome. Type V is a non-scalp aplasia cutis with papyraceous fetus; type V lesions are due to skin necrosis, usually of bilateral symmetry, non

inflammatory, well circumscribed and vary in size from 0.5 to 10cm^{1,2}

In most patients, the lesions evolve from the re-epithelization with atrophic or hypertrophic scarring sequelae and alopecia totalis, in a range of days or months. The possible medical or surgical therapy is related to the extent and depth of the lesion.³

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