PAEDIATRIC DERMATOLOGY

APLASIA CUTIS: SYNDROMIC VARIANTS

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Background: Frieden has described nine different types of Aplasia cutis congenita based on clinical presentation. They are also, syndromic and non-syndromic, of which the latter has the most common presentation and the former has an incidence of 1 -3 in 10,000. We herein present two unique syndromic variants of Aplasia cutis.

Observation: Aplasia syndromic children with Type-5 and amniotic bands, and a Type 6.
Case 1: A day old male, born to consanguineous parents, at full term normally with no complications except aplasia. Soon, child was in ICU with cardiovascular distress. History revealed a papyraceous fetus. Skin examination showed necrosis, amniotic bands with aplasia. Child expired within 48 hours. A post mortem was refused by the parents.
Case2: A 6-hour old boy, born of a second-degree consanguinity, at term by caesarean, had aplasia. The mother, had earlier lost 2 children immediately at birth with similar skin lesions. On systemic examination, ear, cardiac and renal involvement was noted. At 28 hours, the child had a bulla and biopsies from both areas confirmed Aplasia cutis and subepidermal bullae respectively and diagnosed as Bart's syndrome. A post mortem was refused by the parents.

Key message: Recognising the importance of papyraceous fetus, undergoing routine advised scans in pregnancy, Aplasia syndromic overlaps and genetic counselling prior to conception in previous bad obstetric history.