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Aplasia cutis congenital type 6 A rare type- a case report

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Abstract
Aplasia cutis congenita is a heterogenous group of disorders where localized absence of skin particularly scalp and associated anomalies were present. It has several types. Most common one is type 1. Here we are presenting a rare case type 6 also known as Bart syndrome. It has good prognosis if proper medical management is done and if not associated with any other anomalies.

Keywords: aplasia cutis congenital, anomalies

Introduction
Aplasia cutis congenita is a disorder of absence of skin. It has classified into 9 types depending upon involvement of organs and associated anomalies. Here we are presenting a case report of type 6.

Case Report
1day old term female child born by SVD to primi mother under non consanguineous marriage, admitted in view of mild respiratory distress and skin abnormalities. On examination there was a absence of skin over whole of left limb and anterior part of neck, bullae were present on the neck, bilateral microtia and dystrophic nails were noted. Maternal history was nothing significant. No history of similar complaints in the family. Distress settled within hours. Appropriate fluid management, aseptic precautions taken, regular wound dressing done and local antibiotics were used. As septic screen positive blood culture taken and started on systemic antibiotics, stopped after blood culture, septic screen were negative. After 20 days gradual epithelialization occurred around the wounds. Baby went home against medical advice due to some personal reasons. All investigations were done to rule out other anomalies in which no abnormality noted. Consent was taken for publication.

Discussion
Aplasia cutis congenita (ACC) is a disorder of localized absence of skin and associated anomalies. This absence of skin most commonly affects the scalp (type-1) but it can occur anywhere in the body [1]. First reported in 1767 by cordon. Incidence is about 2.9 cases/10,000 newborn [2]. In 1986 Frieden classified into 9 groups according to site of involvement and associated anomalies. Type1 is most common about 86%, complete bone defects occur in 20%. Here we are presenting a rare case Type 6 known as Bart syndrome. But presently it is classified in Epidermolysis bullosa (EB). It usually associated with aplasia cutis, EB, microtia and dystrophic nails. It usually associated with AD inheritance but here we are presenting sporadic form [3]. It also associated with systemic involvement like pyloric atresia but our case not associated with any other anomalies conformed by various scans [4].

Pathophysiology: explained by many hypothesis mainly tension induced type of disruption at 10-15 wks where maximum brain growth occur [5]. Another one is premature amniotic band formation and rupture [6]. Mutation involving VII collagen where glycine substituted instead of arginine also explained [7]. Others like intra uterine trauma, vascular compromise, intrauterine infections, medications and mutations in BMS1 and ribosomal guanosine triphosphate. In type 1 where only scalp involvement will occur, type 2 scalp and limb reductions defects, type 3 scalp and epidermal nevus, type 4 associated with spinal dysraphism, type 5 associated with fetus papyraceous, type 7 ACC localized to extremities, type 8 due to teratogens, type 9 associated with trisomy 13 and Adams oliver syndrome.

Diagnosis is mainly clinical and histo-pathological examination and immunofluorescence. In this case histo-pathological examination showed sub epithelial vesicle formation [8]. Mortality is about 20-55% [9] mainly due to infections, dehydration, sinus venous thrombosis and associated anomalies. Conservative management if done properly like proper dressing, wound care and local antibiotics is highly effective [10]. Standard surgical care grafting also available for lesions of size >30 cm^2 [11].

Conclusion
Aplasia cutis congenital is even though looking more severe

disorder regular wound care, fluid management and aseptic precautions leads to good prognosis. No need of extensive surgical involvement if proper medical management is done.

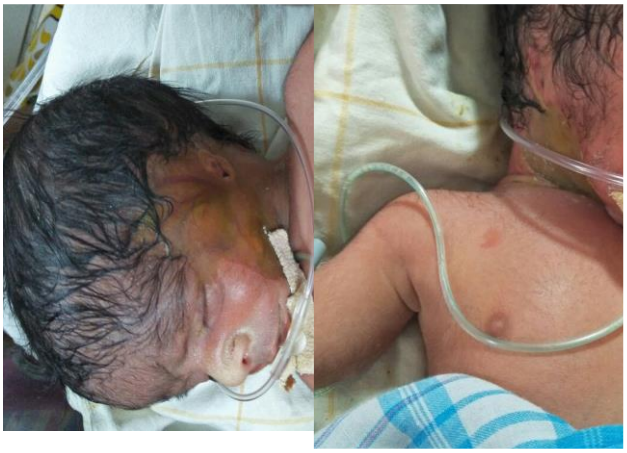


Fig 1: Bilateral microtia

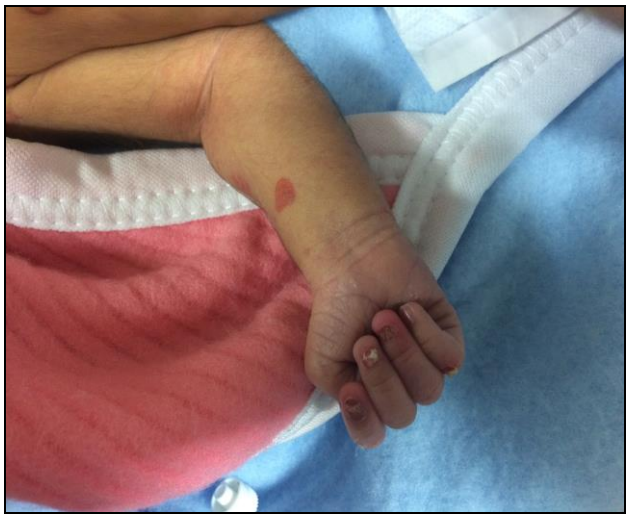


Fig 2: Limb aplasia

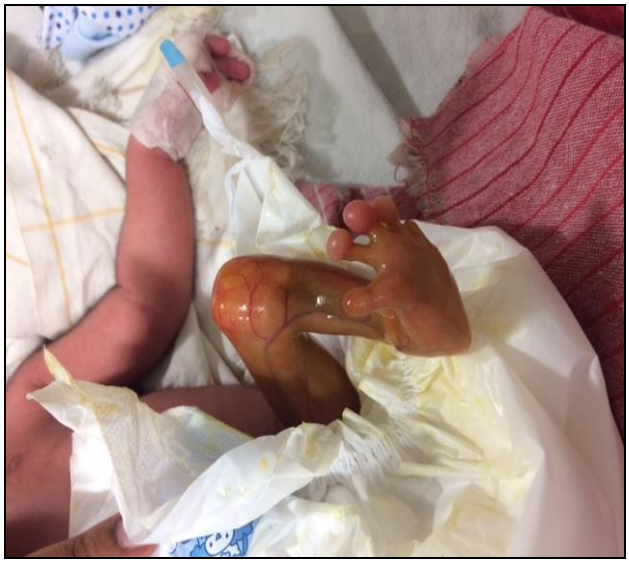
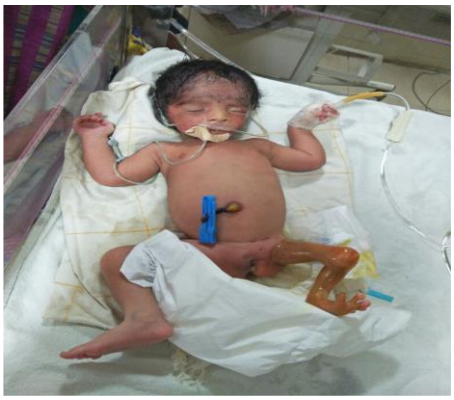


Fig 3: Nail dystrophies



Baby as total

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