trident appearance. Premature ossification of capital femoral epiphysis is seen. New born with thoracic dysplasia associated with lung dysplasia usually die during the neonatal period due to respiratory failure. In those who survive, chronic renal failure is a common cause of death. Renal histopathology reveals cystic changes later peri-glomerular fibrosis. Cirrhosis liver is also one of the cause of early morbidity. At present there is no biochemical or genetic marker, which could be used for prenatal diagnosis of ATD. However, prenatal ultranographic measurements like TC/ AC (Thoracic circumference/Abdominal circumference) and RCP (Rib Cage Perimeter)/TC helps in diagnosing skeletal dysplasia associated with small thorax.

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Aplasia Cutis Congenita

A 2-day-old female neonate was with a small skin lesion on the scalp. The lesion was 2 × 1 cm area on the right parietal region covered with a thin, partial thickness membrane like covering, with slight fluid accumulation underneath, giving the appearance of a blister (Fig. 1). There was a definite underlying bony defect 1 × 0.5 cm. The child had no neurological defict and the ultrasound examination of the abdomen and echo-cardiographic evaluation were normal. A clinical diagnosis of Aplasia cutis congenital was made and the child was managed conservatively with paraffin gauze dressings (to avoid drying and eschar formation). The child had complete epithelization at two months of age.

Aplasia cutis congenita is a rare disorder associated with a complete or partial absence of an area of skin. Although this may occur anywhere on the body, about 80% of these lesions involve the scalp, usually the vertex. The size of the lesion may vary from a barely perceptible lesion to lesions larger than 10 cm. The smaller lesions are usually partial thickness defects whereas larger defects are likely to be full-thickness with underlying bony defects in 15-30%. They are known to be associated with cardiovascular, neurological, spinal or chromosomal abnormalities and may be familial.

Fig. 1. Head demonstrating a small area with absence of skin, which was covered by a thin parchment like membrane.
The treatment of scalp defect is aimed primarily at prevention of infection and hemorrhage. An infected lesion can rapidly lead to meningitis. Major hemorrhage especially from the superior sagittal sinus is an ever present risk and is usually preceded by warning bleeds. Small defects are most often of partial thickness and observation or wet dressing (to prevent drying or dessication) is an acceptable option in the management of partial thickness defects, as in our case. The treatment of full-thickness lesion with exposed brain is primarily surgical and is usually managed by full-thickness rotation flaps or split skin grafting.

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Editor’s Note

Figure 2 depicts a large area of aplasia cutis on the scalp measuring 10 × 4 × 0.5 cm in another male infant with similar clinical details as the one reported above.

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Fig. 2. A large well demarcated area of scalp with absence of skin and hair.