# Aplasia Cutis Cerebri With Partial Acrania—Total Reconstruction in a Severe Case and Review of the Literature

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***Background:*** Aplasia cutis cerebri with partial acrania is a rare anomaly that can involve the epidermis, dermis, and subcutaneous tissues of the scalp with significant bone defect.

***Methods:*** The authors present the first successful report of providing one-stage complete cover in a case of aplasia cutis cerebri with major skull defect by using local rotational flaps. Also presented is a review of literature.

***Results:*** The newborn had a receding forehead and a large scalp defect from frontal to occipital bone measuring 10 × 8 $cm^2$ involving full thickness of cranium centrally, thereby exposing the dura. Local rotation flaps were raised from the scalp to cover the 10- × 8-$cm^2$ defect. The edges of the defect were showing granulation tissue with ingrowth of epithelium. Partial-thickness skin grafts were used to cover the raw area left at the nape of the neck after raising the rotation flaps. The child was discharged on the 19th postoperative day.

***Conclusions:*** Aplasia cutis cerebri with partial acrania, as in our case, has a high mortality rate secondary to infection or to hemorrhage from ulceration of the sagittal sinus. The successful outcome of our one-stage local rotational scalp flap technique provided complete cover to this major scalp defect. This technique will definitely improve the management of this complex disorder keeping the morbidity to the minimum.


INDEX WORDS: Aplasia cutis, acrania, skin flaps, neonate, calvarium.

APLASIA CUTIS CONGENITA is a rare condition characterized by the total congenital absence of all skin layers, most commonly on the scalp. There is a noninflammatory, well-demarcated defect of skin and subcutaneous tissue and, more rarely, periosteum, skull, and dura. The more severe forms predispose to multiple potentially life-threatening complications including massive hemorrhage, secondary infection, and meningitis.[1] We present a case of successful reconstruction of aplasia cutis cerebri involving more than 75% of scalp with partial acrania.

## CASE REPORT

A full-term girl weighing 2 kg was born to a primigravida, 28-year-old mother by elective lower segment cesarean section with face presentation. The baby had dysmorphic facies, receding forehead, and a large scalp defect (Fig 1a) from frontal to occipital bone measuring 10 × 8 $cm^2$ involving full thickness of cranium centrally, more toward the left parietal area, thereby exposing the dura. There was no cerebrospinal fluid leakage, but there was herniation of pia arachnoid through a wall defect (1 × 0.75 $cm^2$) in the dura mater. There was no abnormal vasculature or any other grossly evident abnormal feature. There was no significant family history. The neonate was admitted to neonatal surgical intensive care unit at day one of life, stabilized and the scalp defect covered with saline-soaked gauze. On investigation, computed tomography (CT) scan of the head showed congenital skull vault defect bilaterally in the parietal regions. The left-sided defect was larger, and on the right the lesion was irregular, enhancing and extra axial in nature. Echocardiography essentially was normal except for a small patent foramen ovale with left to right shunt. The child underwent surgery on day 5 of life. Local rotation flaps were raised from the scalp to cover the 10- × 8-$cm^2$ defect (Fig 1b). The edges of the defect were showing granulation tissue with ingrowth of epithelium. Partial-thickness skin grafts were used to cover the raw area left at the nape of the neck after raising the rotation flaps. The postoperative period was uneventful, only a small portion (1 × 1 $cm^2$) of graft was lost over the occipital region, thereby leading to over 95% graft take-up. The residual raw area healed by secondary intention. The child was discharged on the 19th postoperative day. After 3 years of follow-up the child has a full-thickness hairy scalp (Fig 2); fundus examination shows a normal disc and macula, and she follows direction of light normally.

## DISCUSSION

Aplasia cutis congenita is a rare condition characterized by the congenital absence of epidermis, dermis, and, in some cases, subcutaneous tissues. It was first described by Cordon in 1767, and more than 500 cases have been reported since,[2] with an estimated incidence of 1 in 10,000 births.[3] Although the lesions may occur on any body surface, localised agenesis of the scalp is the most frequent pattern (aplasia cutis cerebri). In approximately 20% of the cases, an underlying bone defect also is found. The most frequent occurrence is sporadic, but

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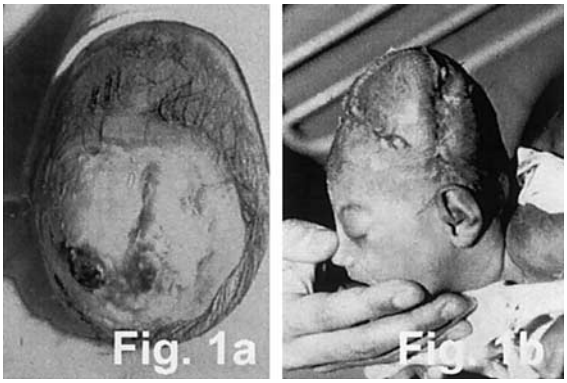


**Fig 1. (a) Aplasia cutis cerebri and partial acrania with exposed dura in the newborn. (b) One month after covering the defect by rotational flaps.**

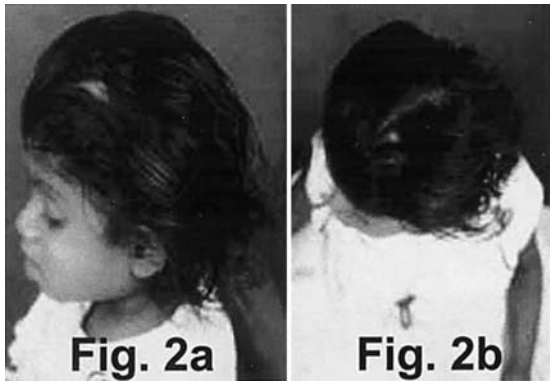


**Fig 2. At age 3 years the scalp has full hair-bearing skin.**

autosomal recessive and dominant inheritance have also been described.[4-6] The etiology is uncertain but suggested hypotheses include ectodermal arrest during embryogenesis, vascular anomalies in the fetus, intrauterine trauma, amniotic bands, and hereditary factors.[4,6,7] Examination of placenta and the umbilical cord may aid in the diagnosis. It is important to recognize this as a sporadic intrauterine event unrelated to birth trauma or forceps delivery.

Frieden has classified aplasia cutis into 9 types.[5] Although treatment of less-severe forms is relatively straightforward, the management of more severe forms remains challenging, with the mortality rate from massive hemorrhage and infection in the latter reported as 20% to 55%.[8] Conservative, operative, or combined approaches have been attempted. Conservative approaches involve regular dressing to keep the defect moist, prevent desiccation, and allow spontaneous epithelialization. The operative treatments include primary closure, skin grafting,[9] local scalp flaps with or without tissue expansion,[10] and cranial vault reconstruction using split rib grafts and free latissimus dorsi muscle flap[11] (microvascular surgery).

Reports include saline dressings, continuous saline drips,[12] betadine solution,[13] bacitracin ointment,[14] and silver sulphadiazine dressings.[15] Although conservative treatment may appear at the onset to be the preferred management option because of its simplicity, reports have shown some serious complications during conservative management of large scalp defects like the one encountered by us. These included biochemical abnormality and hyponatremia secondary to sodium loss from the scalp defect leading to epileptic seizures and apneic episodes. Other complications also are know such as hyperkalemia from silver surfadiazine cream, repeated hemorrhage requiring emergency surgery to control the blood loss, unexpected poor local flap viability when transferred immediately or after a delayed procedure, and prolonged hospitalization up to a total of 9 months to achieve complete wound cover.[16] Surprisingly, the experience with local scalp flap reconstruction as a primary treatment is variable. Several investigators experience[16,17,12] that local skin flaps are unreliable in this condition. They attribute this either to abnormal vascularity of the skin adjacent to these lesions (seen in few of their patients)[16] or to the previous granulation tissue, which has epithelialized in utero—devoid of the axial vascular pattern. All previous attempts to provide primary cover in such major defects have failed.[12,16,17] The success of rotation flaps covering the large area of $10 \times 8$ cm$^2$ and involving 75% of scalp with partial acrania in our case convincingly allays these apprehensions. It also has negated the possible complications of wound infection, biochemical disturbance, and massive hemorrhage and reduced the hospital stay and cost to the minimum. Surgery provided cover to the exposed dura by local rotational skin flaps at day 5 of life. At age 3 years, the scalp has full hair-bearing skin. A stable reconstruction of the cranial vault is proposed with periosteal onlay technique for neoosteogenesis as described by us for craniosynostosis.[18]

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