

Conservative Healing of an 11 × 9-cm Aplasia Cutis Congenita of the Scalp with Bone Defect

Victoria Fröjd¹ Giovanni Maltese¹ Lars Kölby¹ Peter Tarnow¹

¹The Craniofacial Unit, Department of Plastic and Reconstructive Surgery, Institute of Clinical Sciences, Sahlgrenska Academy at the University of Gothenburg, Sahlgrenska University Hospital, Gothenburg, Sweden

Address for correspondence Peter Tarnow, MD, PhD, Department of Plastic Surgery, Sahlgrenska University Hospital, SE-41345 Gothenburg, Sweden (e-mail: peter.tarnow@vgregion.se).

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Abstract

Objectives Aplasia cutis congenita is a rare congenital condition, and it is difficult to find scientific support for optimal treatment strategies. In addition, these may vary due to defect size, tissue layers involved, contemporary malformations, and the physiologic status of the affected child.

Clinical Presentation This case report describes complete skin coverage in 20 weeks and uneventful healing of a large 11 × 9-cm defect of the vertex, involving both skin and skull bone, using conservative treatment. To prevent infection and promote healing, the defect was kept moist and covered at all times, and it was treated with surgical debridement when necessary. For infection control, ionized silver-coated dressings were used in addition to prophylactic antibiotics over the first 3.5 weeks. Follow-up was 2 years.

Keywords

- ▶ scalp defect
- ▶ cranial defect
- ▶ aplasia cutis congenita

Conclusion Surgical treatment is usually preferred for larger aplasia cutis congenita defects, but it is accompanied with potential risks and will exacerbate secondary reconstruction of alopecia or skull bone defects. This case shows that even very complex defects may be treated conservatively.

Introduction

How best to treat rare congenital diseases such as aplasia cutis congenita (ACC) is still being debated. The best source would certainly be prospective randomized studies, but these cannot be performed due to the limited number of cases. The number of newborns affected with ACC is estimated to be 1 to 3 in 10,000, and the within-group variation is large. Exposure to teratogens during the fetal period¹ or genetic predisposal (with mainly autosomal dominant inheritance) may play a role in the development of ACC. In Adams-Oliver syndrome, scalp ACC is most often found together with terminal transverse limb defects.^{2,3} However, the pathogenesis is often unknown, and the condition could possibly be caused by early fetal vascular disruptions or incomplete closure of the ectodermal surface of the neural tube.

Different etiologies have been linked to specific subgroups of ACC that were classified by Frieden⁴ and by Sybert.⁵ The subgroups reflect the ability of ACC to affect the vertex, the body, or the limbs, and its possible association with cerebral disorders and malformation syndromes. A review by Demmel (1975)⁶ included 474 patients and showed that 85.8% of single defects affected only the scalp, which would be classified as group 1 by both Frieden⁴ and Sybert.⁵ In group 1 ACC, the skull bone and the dura are affected in ~ 20 to 38% of larger defects.⁶ Thus the inner meninges and the sagittal sinus may be exposed. Fatal complications may be due to exposure of vital structures, large wound area, and possibly vascular hemorrhage and infections. The mortality rate for ACC has been reported to be ~ 20%, with a higher risk when the condition is associated with multiple congenital anomalies.⁶

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Because of the severe complications mentioned and also the fragile state of newborns, optimal treatment strategies would be of great value. The treatment strategies that are currently possible are conservative treatment, surgical treatment, or the two combined. In addition, initial treatment can be separated from late reconstructive treatments. Conservative treatment is a current trend for management in the neonatal period.⁷ This strategy includes various dressings—adhering or nonadhering—and with or without antibacterial properties such as silver coating,^{8–10} and allografts.¹¹ If the dura is exposed, the wound should be kept moist and covered at all times to avoid risk of eschar formation and drying, which may lead to rupture of the dura.¹² Local or pedicled scalp flaps (with or without primary closure) or skin grafts for soft tissue coverage are the most commonly reported surgical techniques.¹³ Skin expansion, free flaps, and bone grafts may also be performed initially or as delayed cranioplastic treatments. Due to the variation in size, the tissue layers involved, position of the defect, and concurrent medical conditions, considerations must be made on an individual basis for all patients.

Case Report

Anamnesis and Status before Treatment

A boy was born at 38 gestational weeks to healthy parents with a healthy 3-year-old son. At delivery, the midwife noticed a skull defect, and the child was moved directly to the neonatal department. There he was examined by a dermatologist and diagnosed with ACC (►Fig. 1). Magnetic resonance imaging 4 days after birth revealed a complete lack of skull bone down to the dura mater and a small area of cerebral ischemic changes. The ischemic area was also seen with computed tomography at 19 days.

The boy's grandfather on the mother's side was born with an alopecic scar on the vertex of his head. Physiologic status in general—such as circulation, neurology, natural functions, and birthweight (3,910 g)—was normal. His Apgar scores were 9/9/9. Eye examinations revealed moderate bleeding

on both retinas, probably from the time of birth. In addition, cutis marmorata (which his brother had at the same age and has been associated with ACC^{7,14}) prompted examination by a cardiologist who did not find any heart or circulatory abnormalities.

Conservative Treatment Strategy

The defect was kept moist and covered with an Aquacel Ag Hydrofiber (ConvaTec Inc., Skillman, New Jersey, United States) dressing soaked in saline and covered with Tegaderm (3M, St. Paul, Minnesota, United States). The dressing was changed daily. The edges of the wound were cleaned with chlorhexidine solution every second day.

The boy underwent surgical revision of the defect site under general anesthesia on day 7 and also at the age of 21 days. Initially, the anterior part of the defect showed a clean nonirritated wound with signs of healing. The posterior part of the defect contained a superficial hematoma covered by a thin membrane. After incision, an old hematoma was revealed and removed, together with thrombotized veins that were cut and coagulated to prevent further hemorrhage and infection. Large areas showed signs of initial bone formation, and there were no signs of infection at either time.

At 38 days after birth, the defect measured 9 × 6 cm and showed fine granulation (►Fig. 2). The boy was discharged from the hospital with continued outpatient dressing changes and weekly control visits to the plastic surgery department.

Follow-up

At the age of 14 weeks, the size of the defect was 4.5 × 2.5 cm. The bone defect had also decreased in size but still remained larger than the skin defect. At the age of 20 weeks, the wound had healed with a small crust remaining, and lubrication without dressings was recommended (►Fig. 3)

At 5.5 months of age, hydrocephalus was suspected because the circumference of the head had increased to +3 standard deviation along with increased tonus over non-bone-covered parts. The boy was treated with ventriculocisternostomy at the



Fig. 1 Six-day-old boy before revision.



Fig. 2 Six weeks postnatally, showing ongoing healing by secondary intention.



Fig. 3 At 4.5 months, showing complete secondary healing of the aplasia cutis congenita defect.

age of 8 months. The incision was made in a skin area separate from the newly healed area. At all times, the boy showed normal psychomotor development.

At 2 years of age, the patient was still showing normal development. The bone defect was further reduced to 45×60 mm. Reconstruction of alopecia and the residual skull bone defect will be performed at a later age (**► Fig. 4**).

Antibiotics, Infection Control, and Medical Considerations

The boy was put on prophylactic antibiotics during the first 3.5 weeks. At first, isoxazolyl penicillin and aminoglycoside were administered, but 5 days after birth the antibiotic regime was changed to a cephalosporin and ampicillin due to increased body temperature and elevated C-reactive protein. At 10 days of age, isoxazolyl penicillin was administered, but again during a period of higher body temperature, the antibiotics were changed to amoxicillin with clavulanic acid.



Fig. 4 Patient at 2 years. Reconstruction of the alopecia is planned at a later age.

Thereafter, antibiotics were administered only on suspicion of infection. At the age of 3 months, the boy was put on antibiotics due to the appearance of small furuncles surrounding the wound.

Wound cultures from the day before the first surgical revision and from the time of surgery later revealed growth of enterococcus and a mixed bacterial flora. At the age of 3 weeks, the boy was anemic with hemoglobin 82 g/L, so iron supplementation and a blood transfusion were given.

Discussion

The patient presented here underwent successful conservative treatment of a large 9×11 -cm ACC on the scalp including the skull bone without critical complications. There have been reports of severe complications following both conservative and surgical treatment of scalp and skull defects.^{15,16} Even so, the present case is a valuable addition to the literature because there was uneventful and complete skin healing and substantial bone healing of a large defect. The other option would have been surgical treatment and early definitive closure, as suggested in the literature.¹⁷⁻¹⁹ Furthermore, in accordance with the present case, conservative treatment has been presented as the first choice for initial treatment^{10-13,20} even for large defects of the scalp vertex (6×4 cm, 8.5×9 cm, and 8×12 cm), involving both skin and calvarial bone.²¹⁻²³ Complications of conservative treatment (e.g., ruptured dura and exposure of the sagittal sinus, leakage of cerebrospinal fluid, and massive hemorrhage)¹³ represent imperative indications for surgical intervention. Donor site morbidity and flap necrosis²² are the two main drawbacks of surgery. The increased risk of flap necrosis or poor adaptation may be due to compromised skin around ACC defects, with reduced elastic fibers, thinned epidermis, and an intermediate zone of malformed dermal appendages.²⁴

Whether or not bone healing capacity can be compromised with skin grafts that may adhere to the underlying brain tissue has been debated.^{12,17} Follow-up with computer tomography of a girl treated conservatively with an originally 6×8 -cm defect of the skull with intact dura showed that the defect was almost closed after 1 year and almost fully healed at the age of 5 years.²⁵ Radiographic follow-up of a boy with a vertex defect originally 4×4 cm including skull bone and dura mater who was treated surgically with a full-thickness local flap showed osseous reconstitution of the calvarial defect after 4 years.²⁶ In any case, bone healing appears likely following either conservative or surgical strategies with skin grafts.

When used, conservative treatment must include prevention of infections, and it must optimize conditions for healing. The dressings used for the present case were chosen to maintain a moist and antibacterial environment. The dressings used, containing ionized silver, have shown antibacterial qualities *in vitro*²⁷ and faster wound healing of excision wounds than iodoform dressings.²⁸

To summarize, although flap coverage is advocated in the literature for larger ACC, conservative treatment can result in complete skin coverage, reduced bone defects, and uneventful

healing. When indicated, surgical debridement may reduce the risk of hemorrhage and infection. Close infection control along with antibacterial dressings and initial prophylactic antibiotics may be of equal importance to avoid infections. Secondary healing of large ACC avoids flap complications and should make secondary reconstruction easier due to the surrounding tissues being untouched.

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