CUTIS APLASIA: PERIOPERATIVE MANAGEMENT AND CASE REPORT

By Steven M. Levine, MD, Derek D. Reformat, MD, and Charles H. Thorne, MD

Abstract Aplasia cutis congenita, a rare congenital disorder involving defects of some or all of the layers of the cranium, is associated with potential life-threatening complications. Although treatment involves both nonsurgical and surgical techniques, the importance of perioperative management cannot be overstressed. A multidisciplinary team, including personnel from nursing, neonatology, pediatrics, radiology, neurosurgery, and plastic surgery services, diagnosed aplasia cutis congenita and planned local wound care, surgical correction, and prevention of potentially life-threatening complications in a 1-day-old boy with a 6x5-cm full-thickness scalp defect. (American Journal of Critical Care. 2012;21(3):215,212-214)

Aplasia cutis congenita (ACC), also termed cutis aplasia, is a rare congenital abnormality involving defects of some or all of the layers of the skin; underlying subcutaneous tissue and, less commonly, underlying periosteum; and bone. Most often the lesions occur on the scalp. The majority are solitary lesions on the parietal surface or vertex, but defects also occur on the trunk and limbs. Of lesions on the scalp, 20% can involve the cranium, exposing the underlying dura membrane.

First described in 1767 by Cordon, ACC is a rare physical finding; only 500 cases had been reported by 2009. The diagnosis is made on the basis of findings on physical examination. Patients typically have a hairless, smooth skin defect covered with atrophic tissue or dark-colored eschar. The estimated incidence of ACC is 1 per 10,000 live births.

Historically, the cause was thought to be related to skin avulsion caused by amniotic bands or trauma during birth. However, the etiology remains unclear; both genetic and environmental causes have been implicated. Vascular abnormalities, including disruption of vascular blood supply, arrest of midline embryological development, defects in closure of the neural tube, and syphilis, have at one time or another been attributed as the cause.

Although ACC has been reported to occur within families or among twins, the mode of inheritance...
is complex (either autosomal recessive or autosomal dominant), with inconsistent levels of expression and penetrance.\(^7\) In addition, several chromosomal disorders, including trisomy 13 and 4p deletion (Wolf-Hirschhorn syndrome), have been associated with ACC.\(^2\) In our case, the infant with ACC had a concomitant chromosome 1p duplication.

Finally, ACC can be associated with various other morphological abnormalities or malformations. In 1986, Frieden\(^1\) proposed a classification of ACC based on the location of the lesion and associated abnormalities. In addition, ACC is considered a major criterion for the diagnosis of Adams-Oliver syndrome in patients with terminal transverse limb defects.\(^9\) ACC can also be associated with other abnormalities, including cleft lip and palate, tracheoesophageal fistula, patent ductus arteriosus, pyloric stenosis, dermal hypoplasia, epidermolysis bullosa, and renal abnormalities.\(^2,10\)

### Treatment Options

The natural course of ACC varies, depending on the size of the defect and the involvement of underlying tissues. Small lesions on the scalp are typical, and in many of these instances, the defects decrease in size via secondary epithelialization from surrounding healthy tissue. Currently, conservative management, including observation and local wound care (eg, antimicrobial barrier dressing containing silver nanocrystals [Acticoat; Smith & Nephew, London, England], silver sulfadiazine cream, bacitracin, 3% bismuth tribromophenate in a special petrolatum blend on fine mesh gauze [Xeroform, artificial skin; Covidien, Mansfield, Massachusetts], continuous saline infusions) with daily gauze dressing changes is recommended only if lesions are small (up to 2 cm in diameter).\(^11,12\) If larger full-thickness defects are left exposed, the underlying dura eventually desiccates, forming a black eschar, which can create undue tension on both the dura and the surrounding healthy skin. The most feared complications of eschar formation are sagittal sinus thrombosis or life-threatening hemorrhage if the dura overlying the sinus is disrupted (if the defect is midline or at a fontanelle) or leakage of cerebrospinal fluid and meningoecephalitis (if the dura overlying the subarachnoid space is compromised).

Although local wound care with daily dressing changes to avoid desiccation of the defect and formation of eschar prevents the risks associated with surgery, such care does not prevent sagittal sinus hemorrhage.\(^13\) The specific risk of hemorrhage from the sagittal sinus when conservative management is used is unknown, but a 50% mortality rate for all cases of cutis aplasia has been reported.\(^13\) Irons and Olson\(^13\) reported a case in which a sagittal sinus hemorrhage was successfully treated by covering the hemorrhage for 1 week with a pack fastened to the surrounding scalp via mattress sutures.

In some instances, observation with or without local wound care is not considered aggressive enough to avoid massive hemorrhage or infection. In order to prevent eschar formation or excise preexisting eschars greater than 2 cm in diameter, definitive surgical management is indicated. Treatment options include debridement of the eschar (if already formed) to reveal the underlying dura and replacement of the defect with either autologous or alloplastic tissue. Examples of allogenic or alloplastic materials previously described to cover scalp defects in ACC include acellular dermal graft (AlloDerm; LifeCell Corporation, Branchburg, New Jersey), gelatin foam, cellulose, and polyurethane foam.\(^11,12\) Autologous options include split-thickness skin grafts (taken from the patient’s thigh, buttock, or adjacent nonaffected scalp), cultured epithelial autografts, bicipital or rotational scalp flaps, and cranioplasty.\(^1,2,5,13,14\)

### Perioperative Management

After diagnosis of ACC, which should be suspected at the time of the first physical examination if not detected prenatally, the infant should be transferred to the neonatal intensive care unit (NICU). The purpose of immediate transfer is to underscore the severity of possible complications, not an urgent need for higher acuity care. The surgical team, usually the plastic surgery or the neurological surgery service, should be consulted when the presumptive diagnosis is made. In our institution, infants with ACC are routinely cared for jointly by both services. Imaging studies including magnetic resonance imaging may be used to evaluate underlying brain parenchyma and to investigate other comorbid intracranial abnormalities. Noncontrast computed tomography may be used to confirm the diagnosis (ie, absence of part of the cranium), and is helpful for presurgical planning via 3-dimensional reconstructions.

Next a therapeutic plan is developed that may or may not involve surgery. However, the plan will always involve temporizing local wound care as described earlier. In order to prevent desiccation, a regular schedule of wound care should be maintained.
We find that “q” shift wound care is the least prone to error, regardless of whether care is every 8 hours or every 12 hours. An important caveat pertains to infants undergoing simultaneous phototherapy. The additional heat increases the risk of desiccation, and doubling dressing changes during scheduled phototherapy is recommended.

Local wound care can be provided by the NICU nursing staff, the NICU medical staff, or the surgical staff. Regardless, responsibility should be clearly delineated to prevent a potentially devastating complication due to an assumption that another service is providing the care. Whichever service takes responsibility for the dressing changes, as with any potentially complicated wound, the practitioner must understand the relevant anatomy of the wound and surrounding structures and assess the infant for changes in the wound bed (ie, evidence of infection, desiccation, exposure of dura, or bleeding).

Because of the low incidence of ACC, health care providers are usually unfamiliar with its management. This likelihood, combined with the potentially devastating complications of ACC, necessitates seamless interdisciplinary communication. All too often, surgical staff treating patients in the NICU discuss case details with the medical staff but not the nursing staff. Commonly, members of the surgical staff will want to maintain their own schedule of local wound care; however, in our experience, the NICU nursing staff are in the ideal position to perform the agreed-upon local wound care. Members of the nursing staff already maintain a schedule of continuous patient assessments and are highly conscious of aseptic technique. Further, having a single dedicated service responsible for delivering the local wound care aids in assessing the need to increase the frequency of the care if early desiccation occurs.

In order to prepare for the complication of a sagittal sinus hemorrhage and to circumvent exsanguination, patients with ACC have blood typed and cross-matched for 4 units of blood on standby at all times. If marked bleeding occurs, pressure should be applied at the source of the hemorrhaging, the surgical team should be notified along with the operating room, and the blood bank should make the 4 units available.

**Case Report**

A 2730-g boy was delivered by a 23-year-old mother, gravida 1 para 0, by normal spontaneous vaginal delivery at 39 weeks and 6 days of gestation. At the age of 1 day, the infant was admitted to the NICU because of a documented duplication of chromosome 1 and other morphological abnormalities. The infant had a 6x5-cm full-thickness scalp defect involving both the skin and calvaria over the sagittal sinus from the posterior fontanelle to the anterior fontanelle (see Figure). Overlying the entire irregularly shaped lesion was a black eschar. Magnetic resonance imaging and computed tomography showed the absence of both skin and bone over dura in the region of the sagittal sinus, and the diagnosis of ACC was confirmed. In addition to this abnormality, the infant had malformed low-set ears, micrognathia, widened spacing between the second and third toes bilaterally, a white spot on the lateral side of the right eye, 2 sacral dimples, a single umbilical artery, and an undescended right testicle.

Initial management included local wound care by the NICU nursing staff with bacitracin and artificial skin to prevent further desiccation of the eschar. At 6 days of age, the patient was taken to the operating room for neurosurgery to remove the eschar directly overlying the exposed dura over the sagittal sinus. The surgery was completed without complications, and the infant had immediate placement of a 0.03-cm-thick (0.012-inch-thick) acellular dermal graft sutured to the adjacent intact skin with 4-0

Figure  A 6x5-cm defect of the cranium and skin with overlying eschar in a newborn boy with face pointed to top of photograph (left), intraoperative removal of eschar (left center) at age 6 days, removal of excess alloderm (not shown), and placement of split-thickness skin graft at age 13 days (right center). Note skin graft donor site along the right temporal scalp. Scalp appearance at age 10 months (right).
chronic catgut sutures. The wound was then dressed with artificial skin covered with bacitracin ointment. The dressings were changed twice daily by NICU staff. On the 13th day of life, the infant was returned to the operating room for removal of any excess allograft that had not taken. Sufficient granulation tissue was present overlying the dura; therefore, a 0.02-cm-thick (0.008-inch-thick) split-thickness skin graft was obtained from the patient’s right temporal cranium and was secured in place with 4-0 chromic catgut sutures. The wound was then dressed with artificial skin covered with bacitracin ointment; the dressing remained over the skin graft for 5 days. Removal of this dressing on the 18th day of life revealed 100% graft take, and no further care was required.

**Conclusion**

ACC is a rare, but potentially devastating disease. The treatment algorithm for the management of ACC involves a multidisciplinary approach that includes both nonsurgical and surgical interventions. Surgical management, when indicated, is relatively straightforward. However, the importance of perioperative care cannot be overstressed. Transfer to a monitored setting, continual and diligent local wound care to maintain a moist wound bed, and availability of blood products (as well as a surgical team if necessary), are the tenets of appropriate management of ACC. Although any of the teams involved can deliver proper wound care, the NICU team, including clerical and nursing staff, neonatologists, and pediatricians, should be present for monitoring after birth and medical optimization before and after surgery, if required. The neurological surgical team should be available for the removal of any eschar, and the plastic surgery service should be accessible for coverage of the defect. With this multidisciplinary approach and clear communication, complications can be minimized and outcomes optimized.

**FINANCIAL DISCLOSURES**

None reported.

---

**REFERENCES**


---

**eLetters**

Now that you’ve read the article, create or contribute to an online discussion on this topic. Visit [www.ajcconline.org](http://www.ajcconline.org) and click “Respond to This Article” in either the full-text or PDF view of the article.
**Cutis Aplasia: Perioperative Management and Case Report**

Steven M. Levine, Derek D. Reformat and Charles H. Thorne

Am J Crit Care 2012;21 212-215 10.4037/ajcc2012904

©2012 American Association of Critical-Care Nurses
Published online http://ajcc.aacnjournals.org/

Personal use only. For copyright permission information:
http://ajcc.aacnjournals.org/cgi/external_ref?link_type=PERMISSIONDIRECT