

Aplasia Cutis Congenita – Plastic reconstruction of scalp and skull defect with three scalp rotation flaps

A. Janssen, MD (1) – N. Vantomme, MD (1) – F. Rogge, MD (2) –
A. Demeester, MD (3) – L. Cornette, MD (4) – J. Dhaese, MD (4)

Department of Neurosurgery (1), Plastic Surgery (2) and Neonatal Intensive Care (4) – AZ Sint-Jan Brugge
Department of Pediatrics – AZ Menen (3)

Abstract

Introduction

Aplasia cutis congenita (ACC) is a rare congenital defect of skin and subcutaneous tissue, more rarely of periosteum, skull and dura. The lesions can involve any location, but most common are scalp defects.

Material and Methods

We report on the successful treatment of a large defect of the scalp with skull involvement in a newborn girl by three advancement flaps with 2 rotation components.

Results

Postoperative course was uneventful. After 3 weeks the skin flaps healed well providing good skin coverage over the defects.

Conclusion

Conservative treatment has been suggested for the possible treatment of ACC. However we think that early surgical intervention is the treatment of choice. In this case operation and postoperative course were uneventful thanks to a good preoperative multidisciplinary assessment.

Case presentation

A 2260g girl was born to a 30-year-old mother gravida 3, para 3, at 38+3 weeks' gestation by vaginal birth. She had an Apgar score of 9/10/10 and a length of 45,5 cm. The baby presented with a 4,5 x 8,7 cm defect along the median line over the great fontanel and two other satellite lesions occipital. All defects include full thickness of the scalp and skull. The dura was completely intact and there was no immediate leak of cerebrospinal fluid. The infant was otherwise healthy.



Initially after birth the defects were covered using occlusive dressings. This kept the wound moist and aseptic. On day 4 reconstruction was scheduled.

Skin was separated from the skull around the defect. A progressive subgaleal dissection was made by the neurosurgeon.

Three skin incisions were made: bilateral frontal incisions and one curved towards posterior. Skin flaps were progressively loosened while preserving the vascular stem. Through turning and moving these skin flaps, the defect can be closed completely. The edges of the skull defect were curetted, enhancing bone growth towards the defect.



The postoperative course was uneventful. After 3 weeks the skin flaps healed well providing good skin coverage over the defects. Further follow-up is necessary to evaluate good and stable functional and cosmetic results. Bony reconstruction was decided to postpone.

Discussion

The best possible treatment of ACC has always been a subject of discussion. Conservative treatment or early surgical intervention have been suggested.[1,2]. We think that early surgical intervention is the treatment of choice.

Conservative treatment	Early surgery
Superficial defects	Large defects and newborn is stable
Spontaneous development of granulation tissue and epithelisation	Local scalp rotation flaps (blood supply from temporal artery) in early days
	Bony defects can be postponed (diminish or heal up spontaneously in most cases)
Humid antibacterial dressings are mandatory	Occlusive dressings before surgery.
Fatal complications are described: meningitis, exsiccation of defect with subsequent rupture of membrane	Risk of arterial flap loss (less if axially vascularized flaps are used)
Electrolyte imbalance caused by scalp defect resulting in hypernatremia, epileptic seizures and apneic episodes	Requires a high level of expertise.

References

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Contact

Dr. Alexander Janssen
Resident
Department of Neurosurgery
AZ-Sint Jan Brugge
Alexander.janssen@azsintjan.be