Ref: Ro J Neurol. 2024;23(2)
DOI: 10.37897/RJN.2024.2.7

Autograft cranioplasty for the correction of a severe frontal skull defect due to hypocalvaria: A case report and review in management

Hana Ranu Herjuna[1], Muhammad Arifin Parenrengi[2], Wihasto Suryaningtyas[2]

[1]Neurosurgery Department "Dr. Soetomo" General Hospital/Faculty of Medicine, Airlangga University, Surabaya, Indonesia
[2]Neuropediatric Division, Neurosurgery Department, "Dr. Soetomo" General Hospital/ Faculty of Medicine, Airlangga University, Surabaya, Indonesia

ABSTRACT

Objective. Here, the authors describe their institutional experience in managing a patient with severe frontal skull defect due to hypocalvaria with brain herniation.

Material and methods. The study was conducted in the Neurosurgery Department, "Dr. Soetomo" Academic General Hospital, Surabaya, Indonesia. The neonates had a congenital anomaly, with skull defect in frontal skull duo to hypocalvaria. Defect closure with autograft cranioplasty using skull bone was performed in surgery.

Outcome. The surgery was performed with autograft cranioplasty using the temporal bone on the left region. We performed decompression of the left temporal area and insertion of a ventriculo-peritoneal shunt at the right keen point to reduce intracranial pressure. Surgical treatment of patient with severe skull defect had to be done.

Conclusion. Autograft with skull cranioplasty is biocompatible, as it is more easily harvested and with less infection and reaction risks.

Keywords: cranioplasty, hypocalvaria, autograft

INTRODUCTION

Hypocalvaria is a rare congenital anomaly in neonates. It is characterized by the partial absence of flat bones of the skull. Hypocalvaria is a post-neurulation defect, although some argue that it occurs because of defective primary neurulation resulting in the non-closure of the anterior neuropore [1]. The bones of the membranous neurocranium are present, but are hypoplastic. This post-neurulation defect has been attributed to an abnormal migration of mesenchymal tissue at the beginning of the fourth week of embryonic development [1,2]. This congenital anomaly is usually associated with a variable spectrum of cerebral malformations [3]. The congenital defects of scalp bones are rarely reported and surgical treatment for correcting the skull defect is rarely discussed in the literature to date. In this case report we perform a skull reconstruction by bone grafting and cranioplasty, highlighting the importance of initial management and supportive care in patients with calvaria.

CASE PRESENTATION

A new-born baby male was referred to our hospital, because of multiple congenital anomalies such as bilateral anophthalmia, labiognatopalatoschizis, microtia and an unusual lesion in the frontal region with a skull defect. He was born at 41 weeks of gestation (unexplained preterm onset of labor) with a birth weight of 2,600 g and an uncomplicated antenatal course. The baby cried at birth and didn't need resuscitation. There was no history of consumption of drugs during the antenatal period. He was born in a first gravid mother. In this baby, a skull defect was found in the frontal region with a size of 9 cm × 8.3 cm × 4cm, his skull was palpable over the frontal

Corresponding author:
Muhammad Arifin Parenrengi
E-mail: muhammad.arifin@fk.unair.ac.id

Article History:
Received: 18 April 2024
Accepted: 3 June 2024

FIGURE 1. Clinical photograph showing size mass (9 cm x 8,3 cm x 4cm), bilateral anophthalmia and labiognatopalatoschizis

region, translumination is negative and no discharge.

Surgical approach was performed while the patient was under the influence of general anesthesia. The surgical procedure was performed when the patient was 7 days old. The utilization of a MRI procedure serves to meticulously identify any irregularities and intricacies within the brain's structures, without exerting any influence or modification upon the subsequent procedures and interventions undertaken for the benefit of this patient. Prior to cerebrospinal fluid diversion and reducing intracranial pressure, the patient was positioned supine with a planned coronal incision, and a ventriculoperitoneal shunt was performed before autograft cranioplasty. The ventriculoperitoneal shunt was surgically implanted at the precise anatomical location known as Keen's point on the right side, following a trajectory that precisely aligns with the right lateral ventricle. Incision was made, and the periosteum was incised with a rasparatorium to locate the frontal calvarial defect's border. Then, 9 × 7 centimeters of autograft bone were harvested from the left calvarial bone. The reparative procedure for calvarial herniation entails the meticulous execution of left temporal bone decompression. Following the meticulous decompression of the left temporal and parietal bones, accompanied by the precise insertion of the ventricular shunt, the herniated brain in the frontal region was skillfully and effectively reduced, culminating in the successful execution of the cranioplasty reconstruction procedure. We reconstruct the bone graft to close the frontal defect and then secure it with silk 2.0 thread. After autograft cranioplasty, the defect in the frontal region of the skull can be adequately reconstructed, and then the skin is sutured layer by layer.

DISCUSSION

This study reports a case of a neonate with hypocalvaria characterized by a large skull defect in the frontal region. We performed cranioplasty of the autograft taken from the temporal bone of the patient. The option of using a split calvaria from the temporal bone is actually very good, but in this case the temporal bone in the neonate is still very thin, so the risk of absorption from the calvaria is very high. Cranial vault defects and congenital defects of scalp bones are rarely reported. Hypocalvaria is an extremely rare condition and it has been sparingly reported in medical literature with only a handful of case reports. Its causation is poorly understood and no concrete etiology has been described. In some studies, hypocalvaria is associated with intrauterine exposure to an Angiotensin Receptor Blocker (ARB) [4,5].

The cause of hypocalvaria is still largely unknown, but it is believed to be a result of a developmental defect during embryonic growth. This sug-

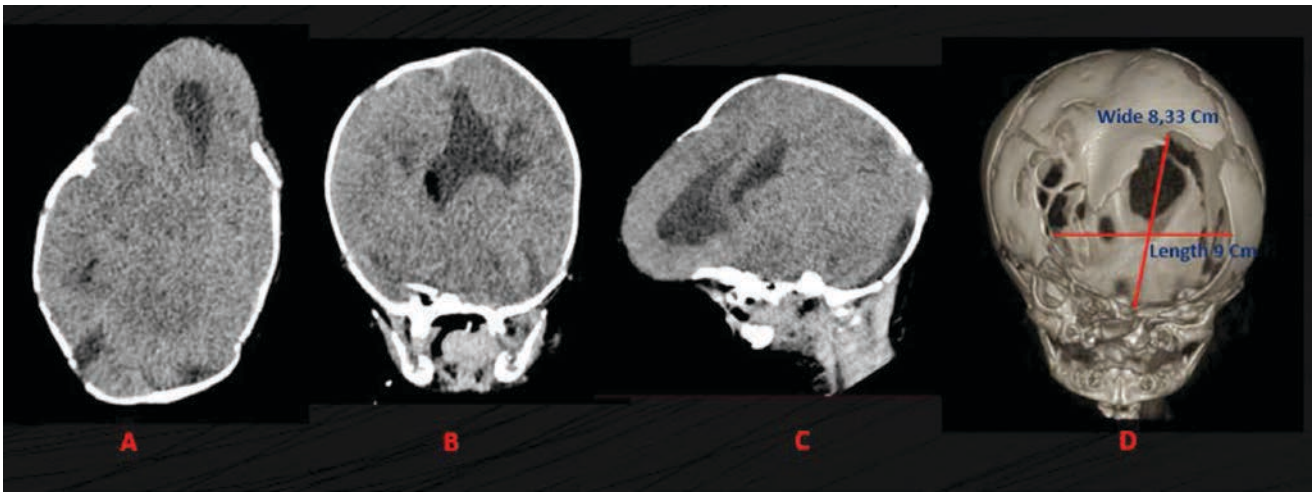

FIGURE 2. Brain CT scan (A) and (C) showed skull bone defect in frontal region. (B) Agenesis corpus callosum and absence of septum pellucidum. (D) Brain CT 3-dimension showing severe skull defect

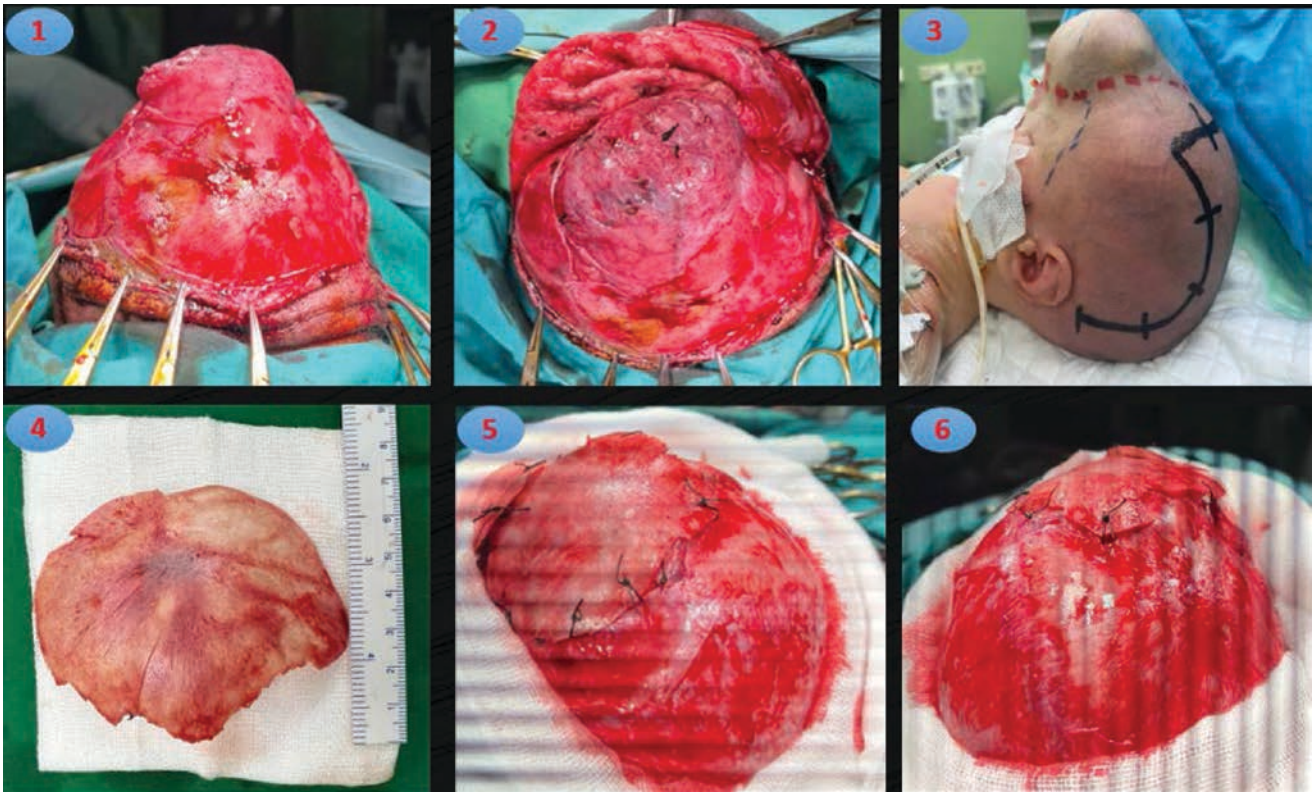

FIGURE 3. Intra-operative images showing. (1) and (2) skull bone defect and brain herniation from calvaria, (3) and (4) Skull bone graft from left frontotemporoparietal calvaria (Size 9 cm × 7 cm), (5) and (6) autograft cranioplasty was performed to close the skull bone defect

gests that it is caused by a disrupted movement of the mesenchyme (which forms muscle and bone) alongside a normally positioned embryonic ectoderm (which develops into skin and scalp) in the 4th week of gestation when the cranial end of the neural tube closes. Consequently, the brain is left covered only with an intact layer of skin with absence of calvarium [6]. Hypocalvaria is typically not linked to any chromosomal abnormalities, and most cases occur randomly. In cases related to amniotic band, early amnion rupture is the triggering event. The amnion can sometimes coil tightly, causing the fetal head to become trapped. This leads to acalvaria due to the incorrect migration of the membranous neurocranium. For other cases of exposure to angiotensin converting enzyme (ACE) inhibitors or ARBs during the second and third trimesters of pregnancy, complications such as oligohydramnios, renal failure, neonatal hypotension, bony deformity, and skull ossification defects have been reported [4,5,7]. Hypocalvaria is different with fronto-ethmoidal (anterior) encephalocele. Anterior encephalocel is a congenital malformation where intracranial con-

tents protrude through a defect in the skull at the junction of the frontal and ethmoidal bones. One commonly accepted theory suggests that the development of fronto-ethmoidal encephalocele may be linked to a disruption in the closure of the rostral neuropore towards the end of the third week of intrauterine life. This disruption causes long-lasting adhesions between the neuroectoderm and surface ectoderm where the nasal fields eventually close [8].

Congenital defects of the scalp and skull present a challenge for care providers because of a combination of their rarity and the magnitude of potential morbidity. Recent advancements in autogenous and alloplastic cranioplasty and scalp reconstruction techniques argue for a comprehensive consideration of this problem [9].

Cranioplasty involves surgically repairing a defect in the calvaria. Several materials have been utilized for the purpose of repairing cranial defects. An ideal material for cranioplasty should possess qualities such as being radiolucent, resistant to infections, non-conductive to heat or cold, and resistant to biomechanical processes. Autograft bone grafts are highly favored due to their ability to minimize the introduction of foreign materials into the body. Additionally, the bone flap can be easily accepted by the host and seamlessly integrated back into the skull. It is more favorable in pediatric patients, as the child's original skull material will naturally reintegrate as they grow older [10,11]. In addition the patient shall undergo periodic clinical evaluations on a monthly basis, followed by a cerebral computed tomography (CT), MRI examination at the 6-month mark, and subsequently at the 1-year milestone post the primary intervention to show an adequate bone ossification.

Another method for skull reconstruction involves using autogenous calvaria and employing an expansion technique. This procedure involves making multiple interdigitating osteotomies to expand the cranium. In infants and neonates, the calvaria is carefully opened to fully utilize the available donor bone, as it can be challenging to separate. With this technique, smaller cranial defects are generated, increasing the likelihood of spontaneous re-ossification [12,13].

Limitations

We have not yet carried out the head MRI. This MRI procedure would be better if it was carried out so that we could evaluate and analyze the structure better. CT scan and MRI of the head will be carried out 3 and 6 months after the procedure as an evaluation.

CONCLUSION

Neurosurgeons are highly concerned about cases of hypocalvaria accompanied by severe brain herniation, as they require urgent intervention. Early intervention can help reduce the risk of future complications related to compression of the subarachnoid space and damage to the brain parenchyma caused by the calvarial bone at the herniation site. Autograft cranioplasty is a practical choice due to the compatibility of autografts with skull cranioplasty, their ease of harvesting, and their reduced risk of infection and adverse reactions. It is essential to closely monitor the patient's progress through clinical observation and regular imaging at 3 and 6 months after the surgery. This allows us to assess the development of the graft and identify any potential risks of bone resorption.

Conflicts of interest: none declared
Financial support: none declared

REFERENCES

1. Anne RP, Angurana S, Suthar R, Saxena A, Sundaram V. Aplasia cutis congenita with hypocalvaria in a neonate. *Indian J Pediatr Dermatol.* 2020; 21:337. doi: 10.4103/ijpd.IJPD_39_19.
2. Faqeih E, Patay Z, Rahbeeni Z, Murtada J, Al Shail E, Hall JG. Localized acalvaria with craniosynostosis. *Clin Dysmorphol.* 2008 Jul;17(3):165-168. doi: 10.1097/MCD.0b013e3282f4a10e. PMID: 18541961.
3. Ri S., Shin J, Je B-K, Choi BM. Hypocalvaria of Newborn Infant: Intrauterine Exposure to an Angiotensin Receptor Blocker. *Perinatology*. 2017; 28:103. doi: 10.14734/PN.2017.28.3.103.
4. Lallemant M, Prévost S, Nobili F, Riethmuller D, Ramanah R, Seronde MF, Mottet N. Prenatal hypocalvaria after prolonged intrauterine exposure to angiotensin II receptor antagonists. *J Renin Angiotensin Aldosterone Syst.* 2018 Oct-Dec;19(4):1470320318810940. doi: 10.1177/1470320318810940. PMID: 30394825; PMCID: PMC6243420.
5. Maman You Espérance B, Landry K, Martial Olivier M, Didier E, Sanni B. Bone Defect of the Cranial Vault: Difficulty of the Diagnostic about a Case, and Revew of Literature. *Open J Mod Neurosurg.* 2023; 13:33–40. doi: 10.4236/ojmn.2023.131005
6. Evans C, Marton T, Rutter S, Anumba DO, Whitby EH, Cohen MC. Cranial vault defects: the description of three cases that illustrate a spectrum of anomalies. *Pediatr Dev Pathol.* 2009 Mar-Apr;12(2):96-102. doi: 10.2350/08-02-0415.1. Epub 2008 Apr 28. PMID: 18442303.
7. Mohammadi F, Azari A, Nikparto N, Ziaei H. Reconstruction of the Occipital and Parietal Congenital Defect with 3D Custom-Made Titanium Prosthesis: A Case Report with Four and a Half Years of Follow-Up and a Brief Review of Literature. *Case Rep Dent.* 2021 Oct 20;2021:7027701. doi: 10.1155/2021/7027701. PMID: 34721909; PMCID: PMC8550828.
8. Koko AM, Ismail NJ, Shehu B, Lasseini A. Surgical Outcome of Anterior Encephalocele: Report of Two Cases and Review of Literature. *J Surg Rehabil.* 2020;1–3. doi: 10.31487/j.JSR.2019.01.08.
9. Feroze AH, Walmsley GG, Choudhri O, Lorenz HP, Grant GA, Edwards MS. Evolution of cranioplasty techniques in neurosurgery: historical review, pediatric considerations, and current trends. *J Neurosurg.* 2015 Oct;123(4): 1098-107. doi: 10.3171/2014.11.JNS14622. Epub 2015 Feb 20. PMID: 25699411.

10. Salam AA, Ibbett I, Thani N. Paediatric cranioplasty: A review. *Interdiscip Neurosurg Adv Tech Case Manag.* 2018; 13: 59–65. doi: 10.1016/j.inat.2018.03.004.
11. Aaron S, Alexander M, Moorthy RK, Mani S, Mathew V, Patil AK, et al. Decompressive craniectomy in cerebral venous thrombosis: a single centre experience. *J Neurol Neurosurg Psychiatry.* 2013 Sep;84(9):995-1000. doi: 10.1136/jnnp-2012-303356. Epub 2013 Apr 16. PMID: 23591554.
12. Shah AM, Jung H, Skirboll S. Materials used in cranioplasty: a history and analysis. *Neurosurg Focus.* 2014 Apr;36(4):E19. doi: 10.3171/2014.2.FOCUS13561. PMID: 24684331.
13. Goldstein JA, Paliga JT, Bartlett SP. Cranioplasty: indications and advances. *Curr Opin Otolaryngol Head Neck Surg.* 2013 Aug;21(4):400-9. doi: 10.1097/MOO.0b013e328363003e. PMID: 23770828.