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FOCUS SESSION

Craniocerebral disproportion after decompressive craniectomy in infants: The hidden enemy of cranial repair?

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Abstract

Introduction Cranioplasty aims at restoring the physiological integrity and volume of the skull. Any disproportion between the intracranial content and the volume of the container may favor the occurrence of complications. A classification of volume mismatches is proposed. A negative mismatch, consisting of intracranial content minor to skull volume, is well represented by the sinking flap. On the other side, a positive mismatch, consisting of intracranial content higher than skull volume, usually depends on CSF collection or hydrocephalus once the brain edema is regressed. Though, in children, this condition may result from physiological brain growth after decompressive craniectomy. Treatment algorithm based on this classification is presented.

Illustrative case A 1-year-old boy with a severe traumatic brain injury underwent right decompressive craniectomy, evacuation of subdural hematoma, and dural expansion at another institution. After failure of autologous bone-assisted cranioplasty for infection, a helmet was recommended in order to postpone the cranial repair. Patient was admitted to our institution 3 years later. CT scan showed brain herniation through the cranial defect, associated to a condition of acquired craniocerebral disproportion, due to the condition of "open skull". Augmented hydroxyapatite cranioplasty (CustomBone, Finceramica, Faenza, Italy) was performed in order to manage this rare condition of positive volume mismatch. Subsequent course was uneventful and no complication was recorded at 30-month follow-up.

Conclusions This illustrative case highlights the possible occurrence of a positive structural mismatch between the skull and the intracranial content after decompressive craniectomy, thus configuring a condition of acquired craniocerebral disproportion, aside of other brain or CSF complications. We firstly recognize this condition in the literature and propose it as a possible factor affecting the outcome of cranioplasty in infants and young children.

Keywords Cranial growth · Cranial repair · Cranioplasty · Craniocerebral disproportion · Pediatric head injury · Personalized medicine

Introduction

Cranioplasty aims at reconstructing the structural anatomy of the skull, for aesthetical and protective functions. Furthermore, cranial repair should aim to restore the physiological volume of the skull, since this factor is essential for the compliance of the intracranial compartment and the functional consequences related to this as the perfusion of the brain.

Any mismatch between the intracranial content and the volume of the container may favor the occurrence of complications.

Provisional results of the present study have been presented at the European Society for Pediatric Neurosurgery (ESPN) Consensus Conference, held in Paris, 28th of February–1st of March 2019.

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On these grounds, a classification of volume mismatches is proposed and a treatment algorithm based on this classification is presented.

Volume mismatch classification and treatment

A negative volume mismatch consists of intracranial content volume minor than skull volume. This condition is easily identified, as the skin flap is sinking. It may be related to several factors that are isolated or coexisting. On these grounds, it may result from severe brain atrophy, an impaired perfusion due to the decompressive craniectomy that is well known as sinking flap syndrome, or the presence of a CSF shunt. This condition has been identified as a negative prognostic factor that is related to postoperative epidural collection and impairment of cerebral autoregulation, which could be eventually related to sudden death after cranioplasty [1]. Therefore, surgical planning should contemplate the correction of modifiable factors, such as upgrading of shunt valve if present, and additional measures to address also the non-modifiable factors. Indeed, the placement of epidural drainage could help the expansion of the cerebral parenchyma. However, closed vacuum suction should be avoided in the postoperative period, since it could be responsible of sudden shifts in intracranial pressure with subsequent impairment of autoregulation [2, 3] (Fig. 1).

On other side, a positive volume mismatch consists of intracranial content volume higher than skull volume. This condition is fairly recognized when the skin flap is full and tense, but could be also more subtle and insidious. The absence of a correct surgical strategy could result in the impossibility to close the skull, due to intracranial hypertension and subsequent bradycardia during the surgical procedure, in the most severe cases. In less severe cases, cranial repair could be completed but the outcome of cranioplasty may be negatively affected, with higher risk of mechanical complications, such as instability or dislocation and fracture of the prosthesis. Factors contributing to this mismatch may be residual brain edema, requiring to delay the cranial repair, or impaired CSF circulation, which could be responsible of hydrocephalus or subdural collections. In case of hydrocephalus, the best management option would be to place a perioperative external ventricular drainage at the time of cranial repair. If hydrocephalus persists after cranioplasty, then CSF shunting is performed. This strategy allows to avoid the concomitant placement of permanent extrathecal CSF shunting device and cranioplasty, which has been associated to a higher risk of complications and to avoid the placement of permanent CSF shunting device in ventriculomegaly related to decompressive craniectomy without overt hydrocephalus [4]. Similarly, subdural collection may be drained at the time of cranial repair and an internal subdural shunting device is reserved to cases with recurrent collections after cranioplasty.

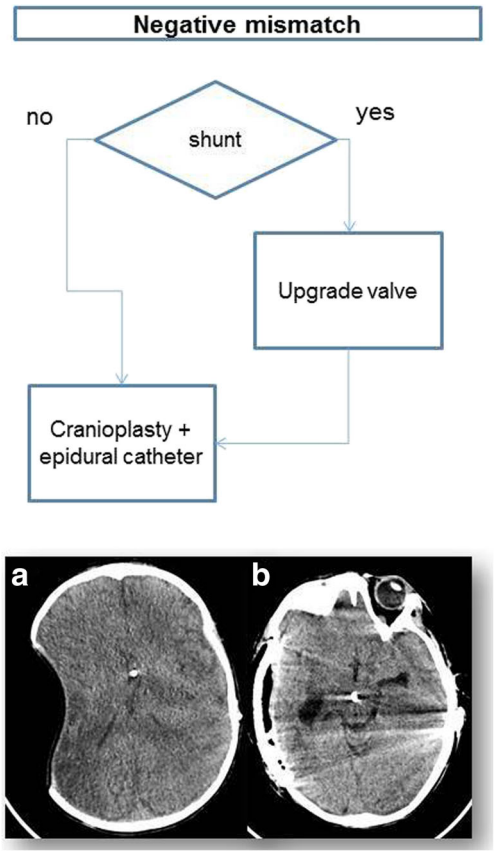

Fig. 1 Management algorithm of negative volume mismatch in cranial repair. Right hemispheric craniectomy with severely sinking flap (A), repaired with upgrade of the shunt valve and epidural catheter at the time of cranioplasty (B) (see text for further details)

However, in infants, a positive volume mismatch may be more subtle and insidious, occurring aside of any brain and CSF complications. In fact, it may be a consequence of cranial decompression and arrested growth of the skull. We firstly recognize this condition in the literature and propose acquired craniocerebral disproportion (ACCD) as a possible factor negatively affecting the outcome of cranioplasty, if not identified and properly addressed during cranial repair (Fig. 2).

Indeed, age has emerged as one of the main factors affecting the outcome of cranioplasty [4–6]. Several factors, directly related to the age of the patient, have been involved. In this context, the role of the growing brain has been included but poorly defined.

The growth of the skull directly depends on the growth of the brain. Another critical factor linking the growth of the content to the growth of the container is the immature dura mater. The growing fracture is a peculiar post-traumatic complication in children highlighting what happens when the integrity of the dura mater is violated, with the vector of force of the growing brain redirected to the locus of minor resistance of the dura mater and skull.

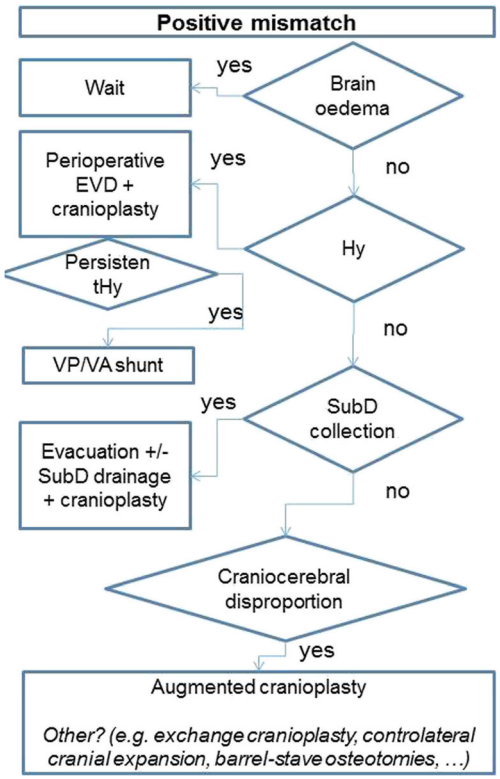

Fig. 2 Management algorithm of positive volume mismatch in cranial repair (see text for further details)

After decompressive craniectomy, a similar condition occurs with violation of both the integrity of the dura mater and of the skull. In this condition, the large post-surgical skull defect totally dampens the force of the growing brain. Subsequently, the remaining skull stops its growth, thus causing a condition of ACCD in the long term. This hypothesis is largely confirmed by the illustrative case.

Obviously, this is an extreme case due to the young age of the patient at decompression and the extremely long delay to the cranial repair. However, we may speculate that, in variable proportion, this condition could occur after every decompressive craniectomy in children under 7 years of age, thus when the skull is still growing. Thus, the degree of ACCD may directly depend on the age of the patient at decompressive craniectomy and the delay occurring between the decompressive craniectomy and the cranial repair.

Additionally, the preservation of the brain parenchyma with its potential of growth is necessary to cause ACCD. In fact, a concomitant sufferance of the brain with loss of parenchymal volume may reduce the effect of the mechanism. This could eventually explain why this condition has been underlooked in the literature so far.

Thus, a thorough knowledge of the mechanism of growth of the brain and skull is required (see previous paper in this focus session for further details), and a careful evaluation of the degree of ACCD should be performed when a cranioplasty is planned in infants and children.

Radiological presentation shares some signs that have been already described for craniocerebral disproportion complicating CSF shunting [7], though the discontinuity of the skull makes the picture more subtle. Thus, particular attention should be paid to the brain herniation through the defect, in the absence of brain edema or hydrocephalus, and the shift of the midline towards the site of the cranial defect with effacement of the cisterns and the subarachnoid spaces on the unaffected site.

Management of this condition requires accurate surgical planning. Intraoperatively, expansion osteotomies (i.e., barrel stave) borrowed from the experience in craniosynostosis surgery may help the cranial repair in less severe cases (Fig. 3). In most severe cases, contralateral expansion of the skull could be considered. As an alternative option, "augmented" cranioplasty may be performed.

In conclusion, this is the first report describing ACCD after cranial decompression in children, and the prevalence of this condition should be further investigated, as well as the impact on the outcome of the cranioplasty.

Illustrative case

A 1-year-old boy with a severe traumatic brain injury underwent right decompressive craniectomy, evacuation of subdural hematoma, and dural expansion at another institution. Post-operative

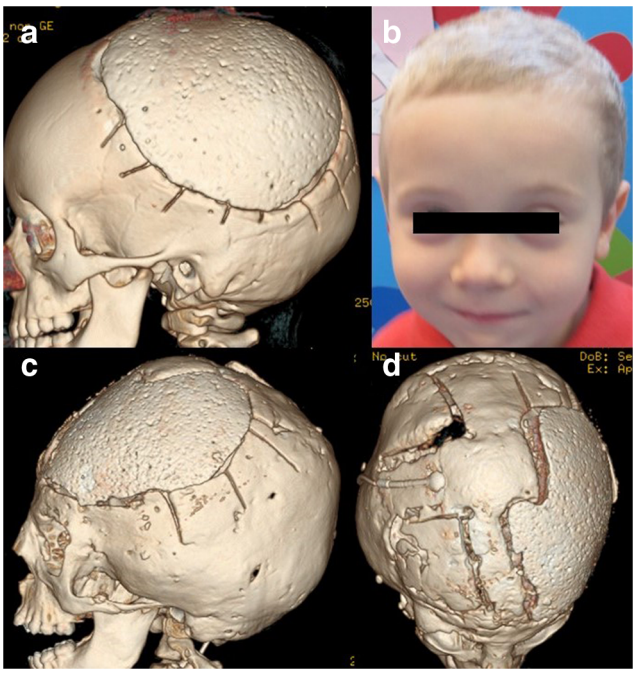

Fig. 3 Five-year-old boy with clinical history of repeated failures of cranioplasty. Successful cranioplasty was obtained with custom-made hydroxyapatite prosthesis and barrel-stave osteotomies at the receiving bone site to manage a mild craniocerebral disproportion (**a**, **b**). Cranioplasty required contralateral cranial expansion to succeed in a 3-year-old child, due to a more severe condition of craniocerebral disproportion (**c**, **d**) (see text for further details)

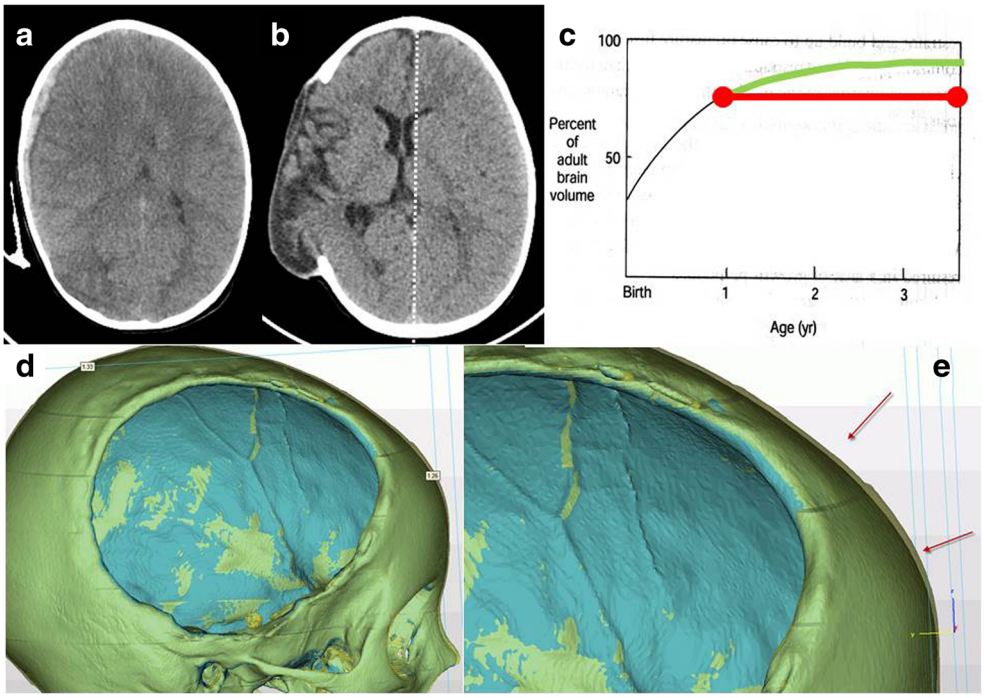

Fig. 4 Illustrative case of ACCD after decompressive craniectomy. **a** Right acute subdural hematoma. **b** CT scan 3 years after cranial decompression. **c** Diagram showing the expected physiological growth of the skull (green line) compared with the present absent growth (red line). The absence of any growth of the skull after decompressive craniectomy is confirmed by overlapping CT scan at the time to cranial decompression to CT scan at 4 years of age (**d**, **e**), with resulting severe craniocerebral disproportion

course was marked by full neurological recovery, and autologous bone-assisted cranioplasty was performed 1 month later at the same institution. Unfortunately, the second procedure was complicated by infection that was treated with the removal of the bone flap and antibiotic. Thereafter, a helmet was recommended in order to postpone the cranial repair.

Patient was admitted to our institution 3 years later. After removing the protective helmet, severe swelling of the right hemispheric surgical flap was evident. CT scan confirmed brain herniation through the cranial defect, associated to a condition of ACCD. Superimposing the CT scan performed at the time of traumatic brain injury with the present CT scan confirmed the absent growth of the skull, due to the condition of "open skull" (Fig. 4).

Expansive hydroxyapatite cranioplasty (CustomBone, Finceramica, Faenza, Italy) was planned. At surgery, ventriculostomy was performed and, after reductive duraplasty, cranial repair was achieved with success. Post-operative weaning

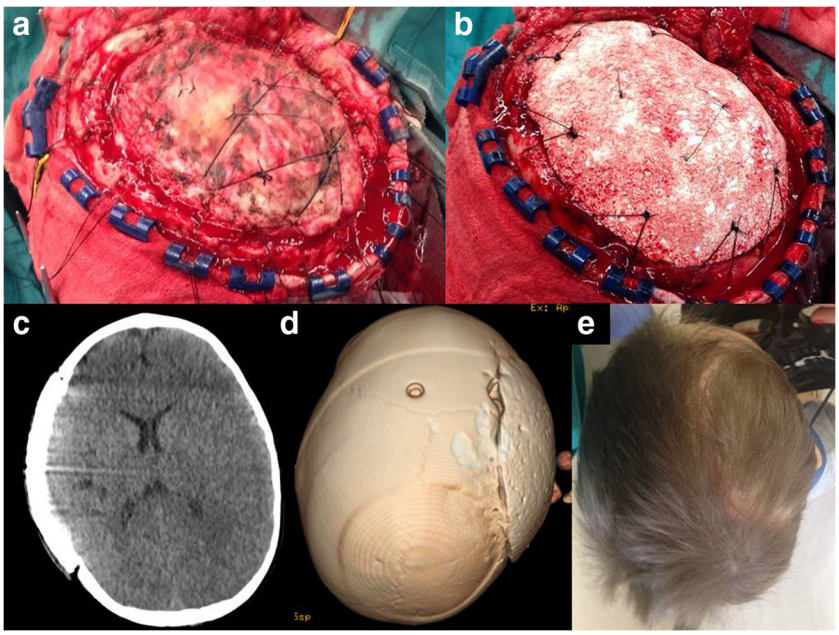

Fig. 5 Steps of cranial repair, namely reduction duraplasty (**a**) and augmented cranioplasty (**b**). Post-operative CT scan confirming the different curvature of the prosthesis compared with the unaffected side (**c**, **d**), without significant aesthetical impact (**e**)

from CSF subtraction allowed to remove the ventricular catheter. Subsequent course was uneventful and no complication was recorded at 30-month follow-up (Fig. 5).

Conclusions

Cranioplasty is not technically demanding but intellectually demanding, in particular in the pediatric population. Volume mismatch between the intracranial content and the skull should be carefully recognized and properly addressed, in order to prevent complications of cranial repair. On these grounds, craniocerebral disproportion may occur in infants and toddlers after decompressive craniectomy, aside other brain or CSF complications. It is strongly related to the age of patient at decompressive craniectomy, delay of cranial repair, and status of the brain parenchyma. It may negatively affect the outcome of cranioplasty and, therefore, it should be managed with different surgical options according to the variable severity of this condition.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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