Growing skull fractures after craniosynostosis repair: risk factors and treatment algorithm

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Abstract: Growing skull fractures (GSFs) are rare complications after severe head injuries in the early childhood and rarely occur after craniosynostosis repair. The aim of this study was to define an algorithm for sufficient treatment for GSF after craniofacial procedures. Literature research was performed to clarify risk factors for GSFs after cranial vault reshaping. Conclusions of the literature and experiences of the authors based on a case of GSF after craniofacial surgery were matched to establish guidelines for successful therapy.

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Introduction

Growing skull fractures (GSFs) have been described as leptomeningeal cyst herniations through a dural rent without evidence of increased intracranial pressure, implicating physiologic growth and brain cerebrospinal fluid pulsations. Synonyms such as leptomeningeal cyst, subdural hygroma, traumatic meningoceles, and cerebrocranial erosion are found in the literature and may even better describe this pathology. GSF is a rare complication affecting 0.6–1.6% of severe head injuries occurring during the first years of life. Given the rapid increase in brain volume during this period, a non-healing bony defect of the skull may lead to the herniation of a dural and arachnoid tear, resulting in the exposure of soft tissues and preventing reossification of the bony defect. Furthermore, the subsequent growth of these pulsatile cranial swellings leads to the enlargement of the skull fracture. The literature mainly mentions accidents, falls, and child abuse as possible causes of GSF, whereas other infrequent causes are poorly described. The aim of this paper was to analyze GSFs after craniosynostosis repair in children to determine whether they necessitate a modification of treatment modality. Guidelines for sufficient treatment of GSFs in patients with craniofacial malformations and craniosynostosis are also discussed.
Methods and Materials

A PubMed database search was performed to review literature on “growing skull fractures” published since 1953. Published reports related to GSF after craniofacial procedures were analyzed to identify the cause of the pathology and conditions associated with a potentially higher risk for leptomeningeal cysts. Common symptoms and radiological findings, as well as differential diagnoses, were reviewed. The authors’ conclusions and experiences with GSFs after cranial vault reshaping were analyzed to define an algorithm for the successful treatment of GSFs after craniosynostosis repair.

Case report

A 16-month-old patient was referred to our clinic. The patient had been diagnosed previously with sagittal and unilateral lambdoidal synostoses and had undergone a strip craniectomy at a different institution at the age of 4 months. Due to the poor esthetic outcome, we performed total cranial vault reshaping at the age of 24 months. No intraoperative problem occurred, and no dural leak was noticed. After an uneventful period, the patient presented a GSF at the age of 30 months.

Results

Using the search term “growing skull fracture” in the PubMed database, we identified 240 relevant articles published since 1953. The addition of the term “craniosynostosis” reduced the sample to only nine items. Table 1 lists the details of identified cases, including the causes and treatments of GSFs. Given the lack of information about treatment outcome provided in these publications, no definitive conclusion could be drawn about this aspect.

The following sections present the results of the clinical and radiological investigations, treatment, and differential diagnosis of an exemplary case of GSF after craniosynostosis repair.
Clinical appearance

Clinical evaluation showed a deep-blue, non-pulsatile, soft swelling (Fig. 1).

Radiological findings

Magnetic resonance imaging (MRI) revealed signs of increased intracranial pressure, such as a leptomeningeal cyst with a subgaleal cerebrospinal fluid (CSF) pad, enlarged Virchow–Robin spaces (suggesting lymphatic diapedesis and compromised lymphatic circulation), a lacunar skull, and a Chiari malformation (Fig. 2).

Intraoperative findings

Intraoperatively, a bony defect was identified (Fig. 3). The entire dural defect must be exposed by a craniectomy because it is usually considerably larger than the bony defect.3

Treatment algorithm

Figure 4 presents guidelines for successful treatment. The treatment strategy presented here was based on the results and authors’ experiences reported in the publications analyzed. Some reports lacked details of treatment after GSF, whereas others described the treatment strategies precisely.

The scalp incision is of major importance to allow sufficient closure after dural repair and cranioplasty; it must be designed to expose the defect completely and allow tensionless closure. Depending of the size of the GSF, most authors found a bicoronal incision to be sufficient.3,4 A scalp rotation flap might be an alternative approach when scalp areas require resection, but this is the authors’ personal opinion. The affected dura must be exposed completely, because exposure of the bony defect alone does not show the size of the dural leak.4 Tissue herniation must be prevented if possible. A watertight, tensionless closure is of great importance. Primary dural closure is insufficient and is impossible in most cases.2,5,6 Most duraplasties employ pericranial or fascia lata grafts. Cranioplasty and vault reshaping should be performed to cover the original dural leak entirely with intact bone.2 Sufficient
scalp closure is essential and may be achieved by scalp rotation to avoid suture placement above the original defect.

As is commonly accepted, ventriculo-peritoneal shunts should be inserted in all patients with signs of increased intracranial pressure.

**Risk factors for GSF**

The following five factors were found to increase the risk for GSFs after craniofacial procedures.

- Signs of increased intracranial pressure or hydrocephaly, such as limited lymph circulation and signs of lymph diapedesis (Fig. 2), ectasia of the dura surrounding the optical nerve (Fig. 2), Chiari malformations with downward displacement of the cerebellar tonsils through the foramen magnum,\(^7\,^8\) and asymmetry of the septum pellucidum (Fig. 5).

- Unilateral or bilateral coronal craniosynostosis due to the displacement of the lateral sphenoid wing and middle cranial fossa.

- Craniofacial syndromes and complex craniostenoses that affect more than one suture. In such cases, increasing intracranial pressure creates impressions on the inner cranium, which makes cranioplasty challenging. The middle cranial fossa and lateral sphenoid wing are displaced, as mentioned above.

- Repeated surgical intervention, which may increase the risk of dural leaks because of scarring and dural adhesions.

  - Endoscopic craniosynostosis repair, which is a minimally invasive procedure that makes the recognition of an intraoperative dural injury more difficult than during open observation.
**Differential diagnosis**

Cephalohematoma

Cephalohematomas may produce symptoms comparable to those of GSFs. However, affected patients usually have a history of trauma, and the swelling is frequently painful. When in doubt, radiological examination shows an intact cranial cortex and the subgaleal hematoma.

Caput succedaneum

Caput succedaneum is a neonatal condition involving serosanguinous, subcutaneous, extraperiosteal fluid collection that may appear similar to a GSF. GSFs have been reported to appear in utero after trauma⁹,¹⁰ or as result of vacuum extraction.¹¹

**Discussion**

Many studies have described GSFs, mentioning falls, vehicular accidents, child abuse, and craniofacial surgical procedures as possible causes. Our literature review identified only nine reports published to date that described GSFs after corrective surgery for craniosynostosis. Esparaza et al.¹² analyzed complications and results in 283 consecutive cases of isolated syndromic craniosynostosis. Postoperatively, 5% of patients (93% of reoperated children) developed dural tears, but no child developed a GSF. Although the initial bony defect may be small after a traumatic GSF, the conditions may differ after cranial vault reshaping. Depending on the type of craniosynostosis or craniofacial syndrome, several bony gaps are left open after craniosynostosis repair. Under such conditions, a GSF may appear after cranial vault reshaping in children with no history of trauma. The question is whether the term “growing skull fracture” describes such cases well. Depending on the ossification of the bony gaps after craniosynostosis repair, herniation of the dura and arachnoid may occur spontaneously without intracranial hypertension. The outward driving force of normal brain growth may be sufficient to cause the herniation of intracranial structures.¹³ Synonyms for GSF that do not suggest a traumatic cause, such as leptomeningeal cyst, subdural hygroma, and cerebrocranial erosion, may be more appropriate in craniofacial cases. In contrast, bagatelle traumas such as falls cannot be ruled out in young children. In
syndromic craniofacial cases affecting the skull base, such as Apert’s, Crouzon’s, Pfeifer’s, and Saethre-Chotzen syndromes, the middle cranial fossa is located much more anteriorly than in non-syndromic craniosynostoses. Furthermore, the lateral sphenoid wings are displaced anteriorly. Under such conditions, the incidence of unrecognized dural lacerations during corrective procedures might be increased because of the limited field of view. By contrast, few reports have described GSFs in the temporal region after craniosynostosis repair. Still, a literature review revealed that 12/19 patients had coronal suture fusion, either isolated or as part of a syndrome. Villarejo et al. reported the development of a leptomeningeal cyst after an iatrogenic fracture of the glenoid fossa during the surgical treatment of an ankylosed temporomandibular joint. In 1998, Yamamoto et al. anticipated the increased reporting of postsurgical GSFs, as cranial vault reshaping techniques became more extensive in the late 1980s. However, a review of the literature showed that few GSF cases have been reported since that time. Contrary to the expectations of Yamamoto et al., minimally invasive procedures may also lead to a GSF. Aryan et al. reported leptomeningeal cyst development after endoscopically assisted craniosynostosis repair in a child with sagittal synostosis. Cohen commented on this report, concluding that endoscopic osteotomy is not risk free. Persing was concerned about the relatively blind nature of cuts made in bone with a curved surface, even with the use of current endoscopic techniques. The potential for cutting the dura is a concern, and the ability to control bleeding is less than optimal. This comment was supported by Steinbok. On the other hand, other authors found no case of GSF after endoscopic procedures in their craniofacial patients. Still, one can summarize that the use of endoscopic procedures should be limited to special indications. Ideally, dural leaks should be recognized intraoperatively and repaired immediately. However, such dural injuries may be small and easily missed. In the authors’ experience, which is in agreement with several publications, the bony defect is smaller than the dural defect in GSFs. Complete exposure of the dural defect is thus necessary to ensure a proper, watertight dural repair. A pericranial flap or a piece of fascia lata may be raised to achieve a sufficient duraplasty. Alloplastic materials should not be used in dural repair. Among the few reported GSFs, four cases developed after the use of alloplastic materials. Herniated brain tissue should not be resected, if possible, to avoid neurological impairment of any kind. Additionally, cranioplasty should be performed to cover the bony skull defect and the original dural leak sufficiently with natural bone (Fig. 6). The need for such a procedure is controversial. Rinehart and Pittman suggested the use of regional
cranietomy and a pericranial dural inlay graft, followed by immediate contour reconstruction with rigid, fixed cranial bone grafts, as the method of choice in GSF repair. Other authors have supported this strategy. Accordinly, we strongly recommend cranioplasty. The bony inlay graft should be fixed with resorbable plates and screws or with resorbable sutures to ensure stable fixation of the graft. Abuzayed et al. described the use of porous polyethylene sheets (MEDPOR®) for bone reconstruction in GSF cases. All cranioplasty methods require sufficient closure of the scalp. The skin incision should be placed to ensure safe closure immediately after cranioplasty. Ideally, no sutures or clamps should be placed immediately over the original defect.

Patients with noticeably limited CSF circulation or increased intracranial pressure are candidates for a ventriculo-peritoneal shunt during cranial vault remodeling procedures. Martinez-Lage et al. recommended the use of dehydrating agents, such as mannitol, to achieve brain shrinkage during craniosynostosis repair, thereby easing separation of the dura from the adjacent bone and preventing accidental dural rupture. Furthermore, these patients should be examined routinely for GSFs during follow-up, preferably until they are about 5 years of age. However, long-term outcomes have rarely been described in cases of GSF after craniosynostosis repair. The patient presented in this study has been followed for 6 years and has shown no sign of recurrence and a satisfactory esthetic outcome, supporting the efficiency of the suggested treatment algorithm.

Conclusion

GSFs are rare complications after early childhood trauma, and GSFs after craniosynostosis repair are even rarer. A successful treatment strategy requires a suitable flap design, total exposure of the dural leak, and sufficient closure of the dura using pericranial or fascia lata grafts. Cranioplasty is strongly recommended to cover the bone and the dural defect, ideally with a bony inlay graft. In patients with signs of increased intracranial pressure, a ventriculo-peritoneal shunt must be inserted.
Conflict of interest

No author has a conflict of interest

References


Figure legends

**Figure 1**: Clinical manifestation of a growing skull fracture as painless, soft swelling

**Figure 2**: Magnetic resonance image of a patient who developed a growing skull fracture after cranial vault reshaping

  a: Leptomeningeal cyst, sagittal view
  b: Ectasia of the perineural sheet of the optical nerve
  c: Enlargement of the Virchow-Robin space

**Figure 3**: Exposure of a growing skull fracture after cranial vault reshaping

**Figure 4**: Treatment algorithm for craniofacial patients with growing skull fractures

**Figure 5**: Displaced septum pellucidum as a symptom of increased intracranial pressure

**Figure 6**: Repair of a growing skull fracture in a young boy

  a: Bony inlay graft raised from an unaffected area of the cranium

  b: Closure of the donor region with an inlay graft

  c: Fixation of the inlays with resorbable sutures

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Fig 1.
Growing skull fracture after craniosynostosis repair

Signs for increased intracranial pressure?

Yes

No

VPS

Scalp rotation flap

Exposure of entire bony defect

Cranietomy

Exposure of entire dura leak

No resection of herniated tissues

Watertight closure of dural defect

Cranioplasty

Scalp closure