

Surgical Technique in Pediatric Craniofacial Surgery

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The surgical correction of non-syndromic and syndromic craniosynostoses (CS) presents several challenges mainly related to the complexity of skull reconstruction, along with the risk of blood loss. Moreover, the surgical team has the commitment to achieve pleasant cosmetic results while minimizing morbimortality. Additionally, several strategies and instruments have been developed to handle this concern, such as technical surgery with minimal bleeding and piezo surgery and technique to obtain more bone autografts and minimize bone healing disturbances. This study was designed to present such techniques in a comprehensive revision of literature and describe them in a step-by-step fashion according to the current state of the art and the experience of pediatric neurosurgery in a single Brazilian institution.

Keywords: craniosynostosis, pediatric neurosurgery, craniofacial surgery, surgical technique

INTRODUCTION

In a hospital, the surgical center is one of the most Craniosynostosis (CS) is a congenital disease caused by the early fusion of one or more sutures of the skull vault and could be classified as non-syndromic if no genetic abnormalities are detected, with an incidence of 1 in 2,000 live births [1]. Additionally, in syndromic CS, facial deformities are associated with genetic mutations, such as Crouzon and Apert syndromes, with an overall incidence of 1 in 30,000 live births [2–4]. Both conditions are treated by a multidisciplinary team, which performs pediatric craniofacial surgery (PCS) [5].

Regarding pathophysiology, CS presents with chronic intracranial hypertension and, in some instances, orbital compression, and both disorders could present in variable degrees, leading to late neuropsychological compromise and ophthalmological trouble [6–15] (Fig. 1). Moreover, the dysmorphic shapes of the skull and face contribute to patients suffering bullying during school ages and negatively

impact social adjustment [16]. Therefore, the surgical team should be committed to relieving intracranial and orbital hypertension and achieving a more pleasant craniofacial shape as much as possible [5, 17].

The surgical technique is affected by the increased risk of bleeding mainly due to enlarged emissary veins caused by chronic intracranial hypertension; the risk of injury to soft tissues, such as the dura mater and periorbital, during osteotomies; and the risk of bone healing disturbances after reconstruction [18–23]. Furthermore, some types of CS, especially the syndromic and non-syndromic forms, are complicated by multiple sutures and Chiari malformation, which necessitates suboccipital decompression, which impacts the surgical technique [24, 25]. Even though endoscopic techniques have gained popularity in the last decade [26], the open technique remains the most used worldwide, and considering the general principles of PCS, several strategies have been developed to minimize these risks, such as blood loss control, the use of the piezo surgery



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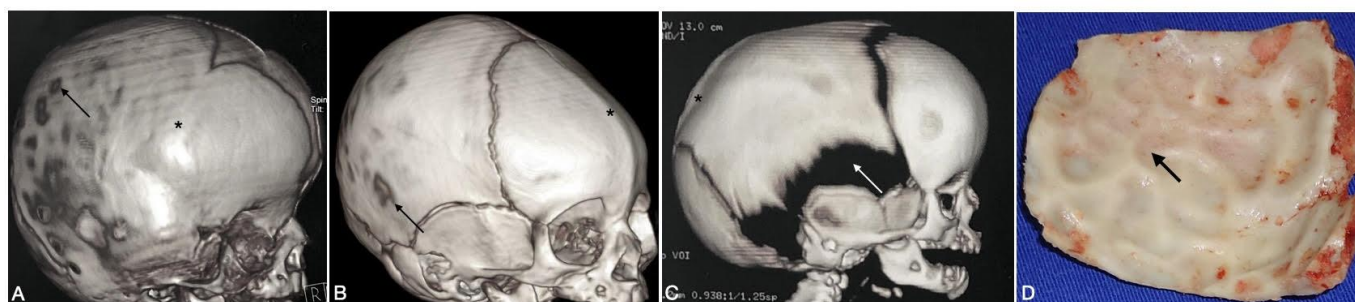


Figure 1 - The radiological and intraoperative signs of intracranial hypertension in craniostenosis. Radiological sign is characterized by copper beaten skull (thin black arrow), which is seen on computed tomography with 3D reconstruction in a syndromic brachycephaly in a patient with apert syndrome (A) and in a non-syndromic trigonocephaly (B). Enlargement of the squamosal suture (thin white arrow) as a compensatory sign of intracranial hypertension in a patient with non-syndromic scaphocephaly (C). The inner face of the cranial vault of a patient with Crouzon syndrome is markedly compressed by the brain gyri as an intraoperative expression of copper beaten skull (thick black arrow). The early fused suture is represented by *.



Figure 2- Skin strategies to improve the cosmetic aspect. The incision is launched above the ears, and minimal hair shaving is recommended, marking the incision before surgery (left). This incision position (white arrow) provides pleasant results in the postoperative phase (right).

during osteotomies on the orbit, and technical nuances during the reconstructive phase.

Therefore, this study was designed to provide the current information regarding technical nuances of PCS, serving as a guide to help young neurosurgeons use the best approach in a step-by-step fashion during skin incision and skull vault exposition, and explain the rationale for osteotomy, bone reconstruction, and avoidance of pit falls and complications.

GENERAL PRINCIPLES OF THE TECHNICAL SURGERY

Skin opening

The appropriate management of the skin during PCS is paramount to warrant an appropriate healing and avoid complications, and as the first choice to accomplish this goal, the minimal removal of hair and making the incision above

the ears is optimal [27]. Although the open approach of PCS requires large incisions, thus increasing the risk of blood loss compared with the endoscopic approach, surgeons can control the blood loss if they follow an adequate technique [28, 29]. Moreover, the manner and position of the skin incision in the head should consider the following factors: (1) ability to expose the entire skull vault of interest, (2) availability to be used in a second time, and (3) the ability to be less obvious as possible. To our knowledge, the biparietal incision with gentle curves provides adequate healing and less likelihood of ischemia, and care must be taken to avoid making an incision anterior to the tragus where surgical scars are usually apparent. Otherwise, the incision starts posteriorly or superiorly to the helix (Fig. 2).

Following the aforementioned intent of minimizing blood loss, a skin incision using low-intensity electrocautery

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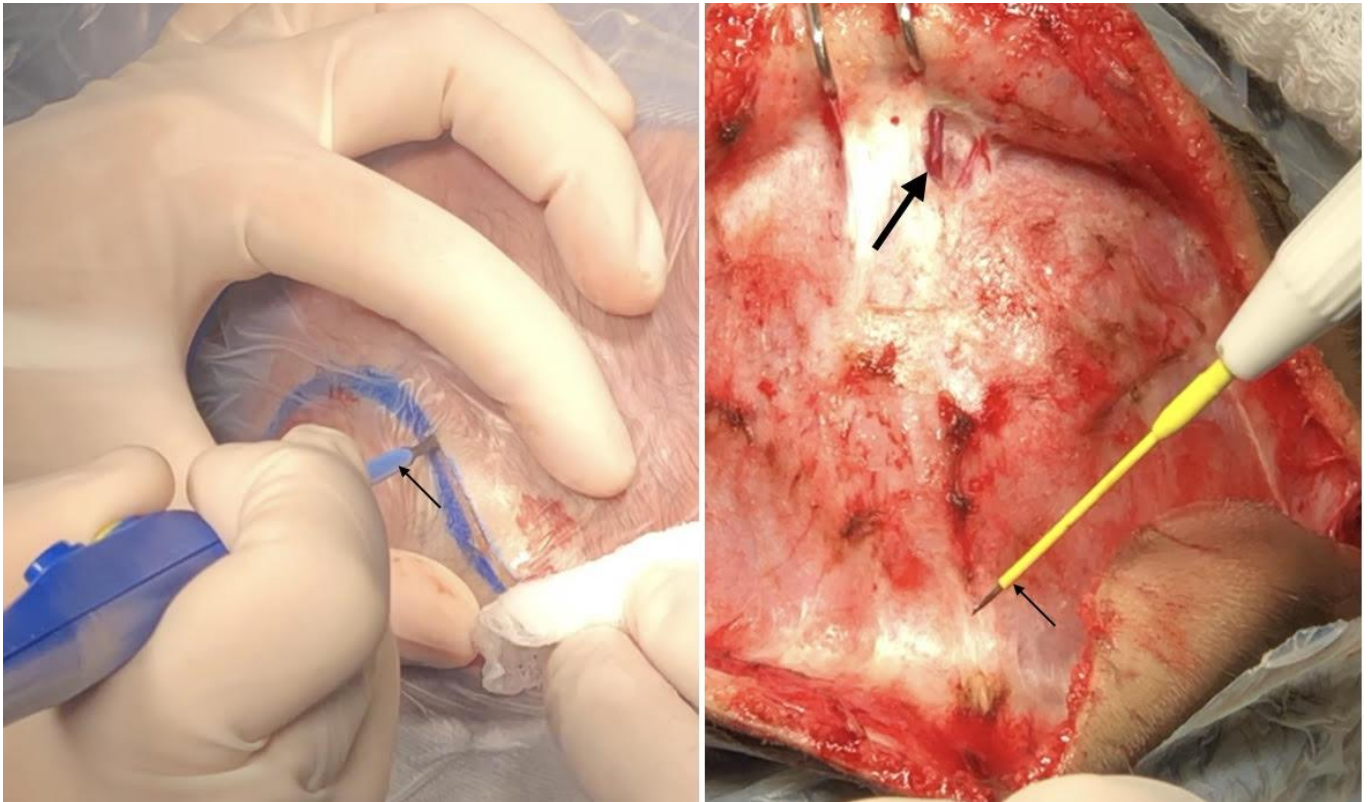


Figure 3 - The strategy to minimize blood loss during skin opening consists of using electrocautery on a cut mode (thin black arrow) during the opening of the epidermal and dermal layers (Left) or during the separation of the galea from the pericranium (right). The preservation of the pericranium during dissection allows the surgeon to see the enlarged emissary veins (thick black arrow) and its coagulation on the approach.

provides an adequate exposition of the galea. Ideally, this step includes the bipolar opening of the epidermal and dermal layers using scissors, allowing the hemostatic control of the emissary vein, consequently avoiding unnecessary galeal bleeding (Fig. 3).

Wood et al. [30] have examined the advantages of electrocautery over the use of a scalpel in making skin incisions, and despite the alleged advantages of electrocautery, such as shorter surgical time and lesser blood loss and postoperative pain, no statistically significant difference was found. Moreover, the decision to use this device is reserved to the preference of the neurosurgeon.

Skull vault exposition and craniotomy

An adequate exposition should display the zone of interest in the skull for reconstruction with less blood loss as much as possible. To minimize bleeding from the emissary veins, not removing the pericranium during this step is recommended (Fig. 3).

Demarcations on the cranial vault using methylene blue serves as a plan to warrant the most symmetrical shape after craniotomies [23]. Additionally, the anterior open fontanel could be dissected, and the epidural space should be assessed, which could help the craniotomy (Fig. 4) [31]. Following the principle of bleeding control, the ideal sequence of burr holes and osteotomies is performing those with less risk of bleeding first and then those with a high risk of perforations near the superior sagittal sinus last. Furthermore, the dura is more attached on the cranial vault near the compromised structures, sagittal sinus, and Wormian bones, and the neurosurgeon should detach it from the lateral portion to midline to avoid damage to the sagittal sinus and seek to feel the bone using gentle dissectors to minimize dural tears [32]. Bone hemostasis is commonly made using bone wax, and despite its efficacy, care should be taken in using it in excess as it can increase the risk of impairing bone healing [33]. To our knowledge, all bone fragments were collected during craniotomy and used in the reconstruction as an autograft.

This step is difficult as it depends on the level of intracranial hypertension, and the neurosurgeon should be cautious in cases in which there are more than one

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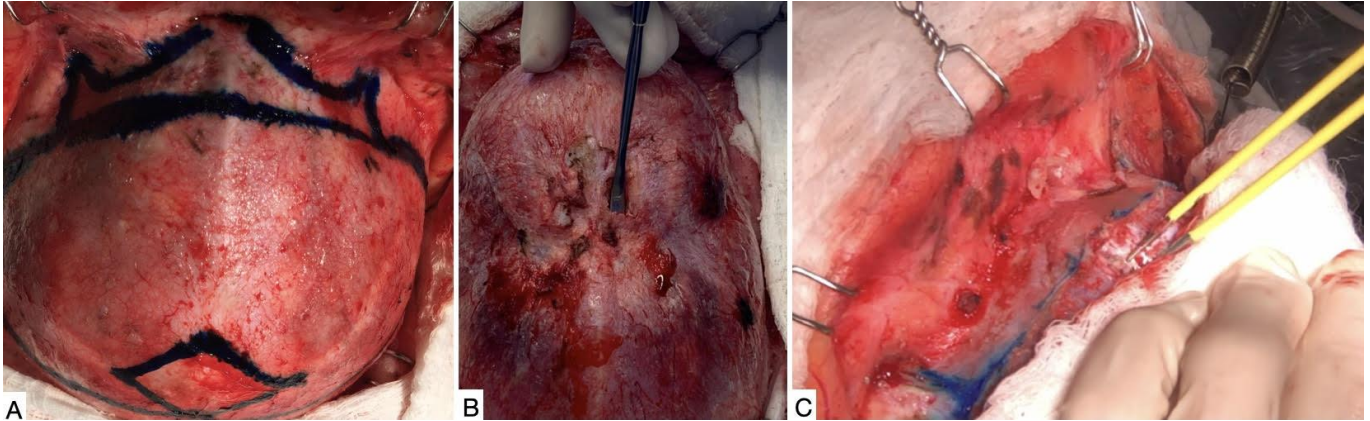


Figure 4 - Strategies during craniotomy. Marking the skull vault using methylene blue in a trigonocephaly (A). Dissection of a natural bone hole due to chronic intracranial hypertension using a dissector facilitates the access to the extradural space and craniotomy in an unusual scaphocephaly (B). After the removal of a bone flap, the dura is gently coagulated using bipolar forceps (C).

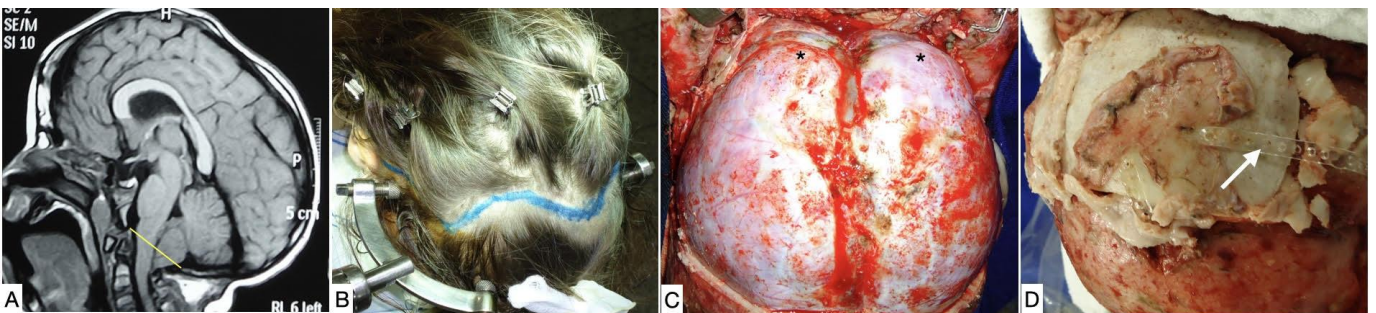


Figure 5 - A patient with Crouzon syndrome had progressed with Chiari malformation (A) and intractable headaches. The patient was placed in the ventral decubitus position, with the head fixed using three pins (B). An approach including suboccipital craniectomy(*) and occipital reconstruction was indicated using absorbable plates and screws (thick white plate) (C and D). Yellow line displays McRae's line.

structures affected, such as oxycephaly and syndromic CS, where more bleeding is expected.

Another concern is related to the deformity pattern presented by the patient, and in some cases, the surgeon could use the holes on the skull due to intracranial hypertension and make the craniotomy safer (Fig. 4).

This step ends with the exposition of the dura mater, which is covered using wet gauze to avoid bleeding.

Chiari malformation is especially associated with syndromic CS and oxycephaly [24, 34]. Moreover, an association was observed between early closure with a lambdoid suture and intra-occipital and petro-occipital synchondrosis, which could be evaluated with impairment of the posterior fossa. In those cases, posterior fossa craniectomy could be associated with occipital reconstruction [25, 35] (Fig. 5).

Orbitotomy

During the frontal orbital advancement, disconnecting the bandeau and orbits from the temporal bone and skull base is important. This step presents some risks, such as augmented bleeding and injury to the periorbita and orbits

and bradycardia due to orbit compression by the surgeon. Using a dissector, the periorbita should be separated from the orbital roof, and care should be taken to avoid damage to the supraorbital nerve and the surrounding vessels. In some instances, the fracture of the floor of the supraorbital foramen using a chisel or piezo sonic warrants the protection of the supraorbital nerve and surrounding vessels [36]. Furthermore, the anesthesiologist should consider the risk of bradycardia during this step.

Usually, the upper limit of the bandeau (last 1 cm from the orbital rim) and the technique of lateral disconnection from the temporal bone differ among authors. Some advocated to incorporate the temporal squamous into the bandeau. However, to our knowledge, due to the risk of atrophy of the temporal muscle, avoiding the detachment of this muscle from its insertion and making the disconnection near the fronto-zygomatic suture are recommended (Figs. 6 and 7). Thus, although the classical technique “tongue-in-groove” is largely accepted [37], the use of a straight absorbable plate connecting the lateral segment of the bandeau could replace temporal osteotomy [38].

Given that the piezo sonic device generates micro-vibrations on its tip and cuts the bone with a relatively gentle movements, some advantages are observed in its use during

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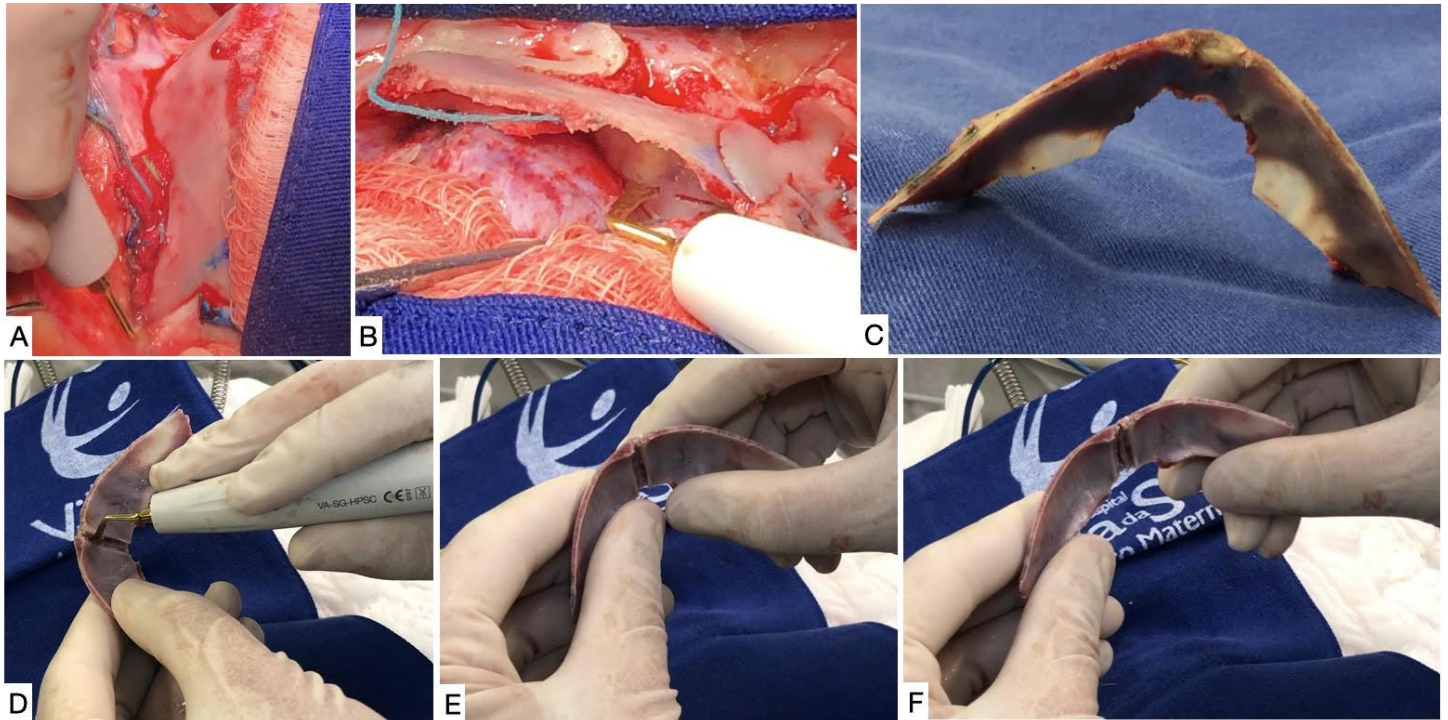


Figure 6 - Orbitotomy and reshaping of the orbits. Osteotomies using the piezo surgery and disconnection from the lateral (A) and orbital roofs (B). The angulation of the orbits in the trigonocephaly (C) is corrected by performing a semi-incision using a piezo sonic device, causing a greenstick fracture (D, E, and F).

this phase over other devices: (1) can perform precise cutting in the bones with minimal bleeding; (2) no need to use force to fracture the skull base and harm the orbits; (3) less risk of injury to soft tissues, such as the periorbita and dura. Currently, some pieces of evidence have been published and support that the advantages of the piezo sonic device are beyond the ability to perform osteotomies; the use of such a device could also allow the surgeon to remodulate the orbits and achieve a more pleasant shape in the anterior coronal CS and trigonocephaly [36, 39–45] (Fig. 6).

Craniofacial reconstruction

Bone reconstruction should follow the following principles: (1) symmetry; (2) appropriately securing the cranial vault; (3) and minimizing problems of ossification. To achieve those goals, it is paramount for the surgeon to assume a position in front of the patient's midline during bone reconstruction and be helped by two auxiliary in positioning the bone flap to avoid displacements and prevent bone spicules.

In the literature, there are several descriptions of materials used for bone fixation during PCS, such as wires [46], resorbable bone stitches [13], titanium plates and screws [47], and resorbable plates and screws [6, 38, 39]. To our knowledge, resorbable plates and screws present many advantages, such as moldability, less risk of skin rupture, no

skull growth interference, and no incorporation of the dura in cases of reoperation. In contrast, handling these materials more appropriately is necessary to avoid an increase in the cost and complications.

After bone fixation, many gaps are created due to the advancement and bone expansion, and to minimize the lack of ossification, several strategies have been reported in the literature, such as autograft [48], bone cement [49], demineralized bone matrix [49], split calvarial bone grafting [50], and autologous stem cell regeneration [51]. In our pediatric neurosurgery service, obtaining a particulate autograft, performing several semi-perforations on the inner face of the bone flap, and covering all bone gaps with fibrin glue are preferred. In exceedingly special situations, the split bone technique using piezo surgery could be used to cover other bone defects [50] (Fig. 7).

CONCLUSIONS

The surgical approach for craniofacial disorders fundamentally depends on the team experience, and the learning curve is mainstream to achieve better results. Although the technical nuances could vary among pediatric neurosurgery services, the general principles of bleed control and commitment to relieving intracranial hypertension should be followed.

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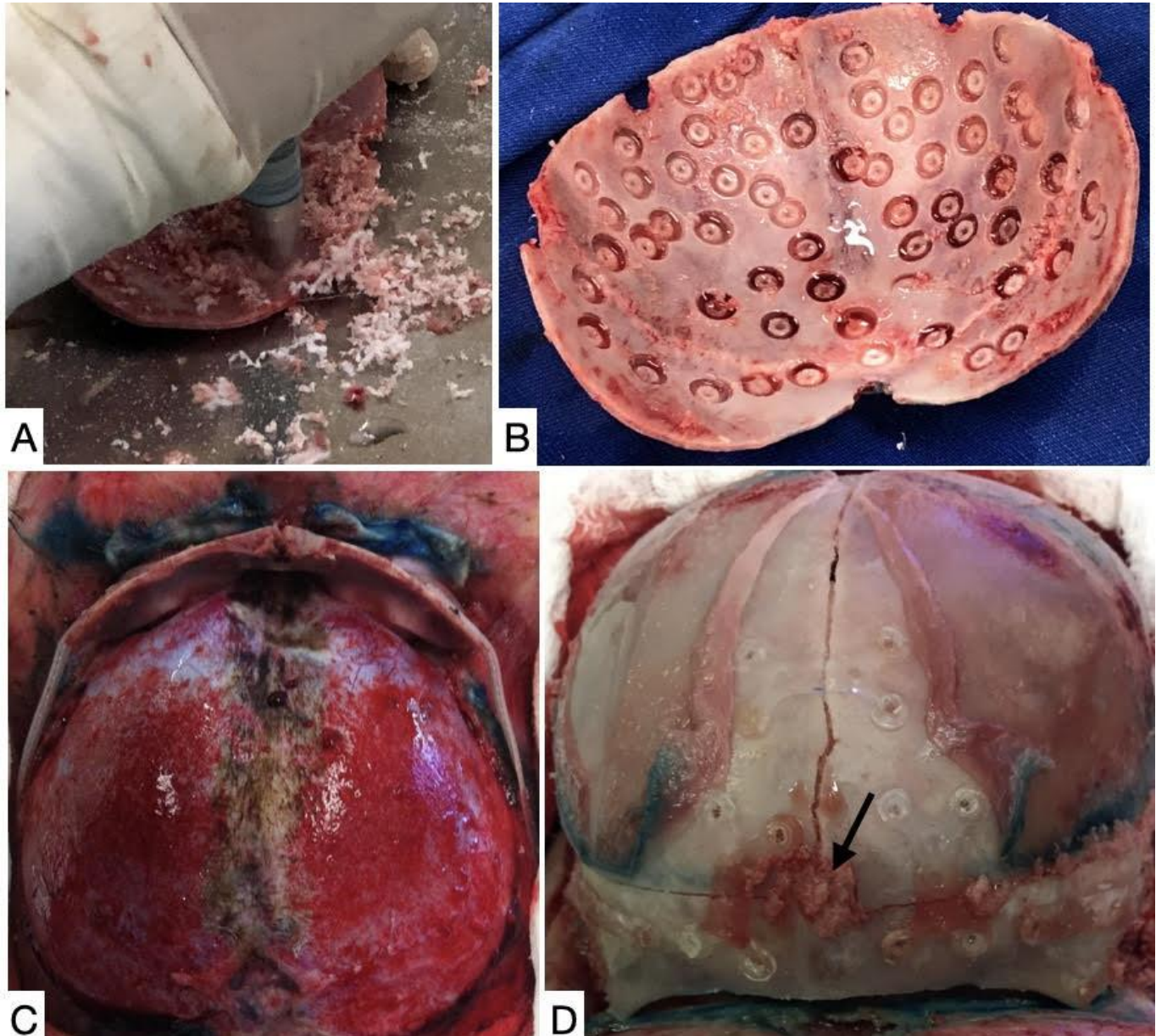


Figure 7- Auto bone graft is obtained through semi-burr holes on the inner face of the skull vault (A and B), and after the orbits (C) and bone flap were fixed using absorbable plates and screws, the gaps were filled with bone fragments (arrow) (D).

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