

# Trigonocephaly: Case Report, Review of Literature and a Technical Note

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## Abstract

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**BACKGROUND:** Premature fusion of the metopic suture results in a type of craniosynostosis known as trigonocephaly. The treatment of trigonocephaly is surgical and is likely to remain so. Surgical methods and techniques for correction of craniosynostosis-related skull deformities have evolved, and a single best procedure is yet to be presented.

**CASE REPORT:** Here we present a technical remark in a case of open cranial vault reconstruction.

**CONCLUSION:** Although the literature, in general, prefers barrel stave (radial) frontal bone osteotomies, a technique with longitudinal frontal bone osteotomies were performed, without fixation of the bony flaps, frontal bone or supraorbital arch, with a quite satisfactory result.

## Introduction

Trigonocephaly is one of the many types of craniosynostosis. Craniosynostosis represents the premature fusion of calvarial sutures. The incidence of craniosynostosis is estimated at approximately 1 in 2000-2500 live births [1], [2], [3]. It has traditionally been classified as syndromic and non-syndromic based on phenotypic features. The syndromic type of craniosynostosis, which represents fewer than 5% of all cases of craniosynostosis, is usually associated with a positive family history of craniosynostosis, fusion of multiple sutures and other abnormalities (skeletal most often), whereas the non-syndromic type, which is the more common variant, often has single-suture synostosis without other abnormalities. It is generally assumed that non-syndromic craniosynostosis (single suture synostosis) have no genetic abnormalities, but recent advances have

shown that almost all craniosynostosis have genetic background [4]. Single suture synostosis is more commonly observed. There is a slight male predominance with a ratio of 2:1. [2], [3]. The metopic synostosis, which this case is about, has a frequency of about 4-10%, just behind the sagittal suture synostosis, accounting for 53-60% and coronal synostosis accounting for 17-29%. Accounting for less than 2% the lambdoid suture synostosis is the least common. The case we are presenting is about a 6-month-old infant, previously diagnosed with trigonocephaly. Presented here is the case with a special note regarding the technical management (operative technique), as the frontal bone was managed differently from the operative techniques proposed in most of the literature where barrel stave (radial) osteotomies are mostly used. Also, the bone flaps were left loose, as no fixating plates or other fixating techniques were used.

## Case Report

This case report describes a case of trigonocephaly in a 6-month-old infant, diagnosed at the age of two months. No previous treatment is undertaken. The cranial perimeter is 37 cm, weight is 8.5 kg. The infant is asymptomatic, despite the visible head deformity.

A preoperative CT scan was obtained, demonstrating metopic suture synostosis, noted as a mid-forehead ridge, hypotelorism, flattening of the frontal bones, anterior displacement of the coronal sutures, compensatory bulging of the parieto-occipital region and temporal narrowing (Figure 1, Figure 2).

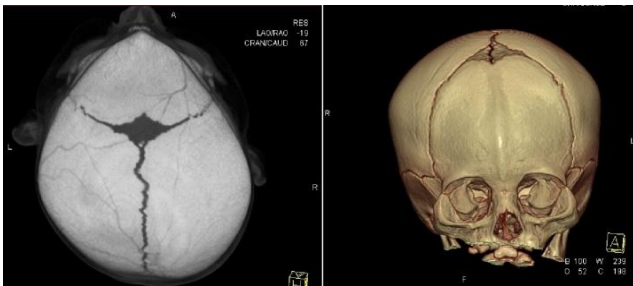


Figure 1: 3D reconstructed CT scan. Note the metopic suture synostosis, seen as a mid-forehead ridge, hypotelorism, flattening of the frontal bones, anterior displacement of the coronal sutures, compensatory bulging of the parieto-occipital region and temporal narrowing

After intubation and introduction into general anaesthesia, the patient, 6 month old male infant, in this case, was placed in supine position with the head stabilised on a horseshoe headrest. Next, the operative field was prepped with antiseptic betadine solution and sterile single-use drapes were used for isolation.

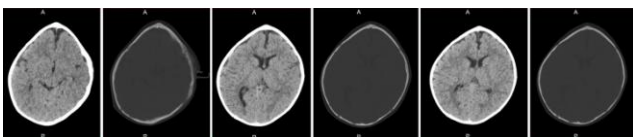


Figure 2: Axial cross section head CT scan: note the triangular forehead, prominent midline sagittal ridge and shortening of the anterior cranial fossa, with compensatory bulging of the parieto-occipital region and temporal narrowing

A bicoronal sinusoid (zigzag) incision was made; subperiosteal skin flap was created and elevated. It was secured in place by three fish hooks. After the exposition of the bone structure of the scalp, frontal and parietal bones were identified, midline frontal bone ridge (metopic suture), and also anterior fontanel. The skin flap was elevated just below the glabella and the nasofrontal suture enough to expose the supraorbital ridge. Next, using an Olivecrona dissector, the dura was easily separated from the tabula interna of the midline of the frontal and parietal bones at the level of the anterior fontanel, just above

the superior sagittal sinus. Kerrison rongeur was used to perform suturectomy at the level of the coronal suture, starting from major fontanel. Using high-speed pneumatic saw (Medtronic Midas Rex system) the initial osteotomy was extended to the level of the sphenofrontal and zygomaticofrontal suture, then turning toward midline just above the supraorbital arch (about 1 cm above) to form a frontal bone flap. The bone flap was left fixed at the midline level just above the nasion. Kerrison rongeur was used to safely complete the frontal bone craniotomy in one piece, to avoid injury to the dura or the anterior part of the superior sagittal sinus. Next, using an Olivecrona dissector the periorbita was dissected from the orbital bone, the point is the protection of the periorbital tissues from the drill and exposition of the bony supraorbital arch for additional drilling. The same was performed using a high-speed drill (Medtronic Midas Rex pneumatic high-speed drill), used for drilling of the ridge at the level of nasofrontal suture, superomedial and superior orbital wall, extending to the previously addressed zygomaticofrontal suture. Intracranial drilling was performed just anterior to the cribriform plate and the crista gali. The bony supraorbital arch was removed in one piece, thus safely opening the orbits and exposing the periorbita (Figure 3).

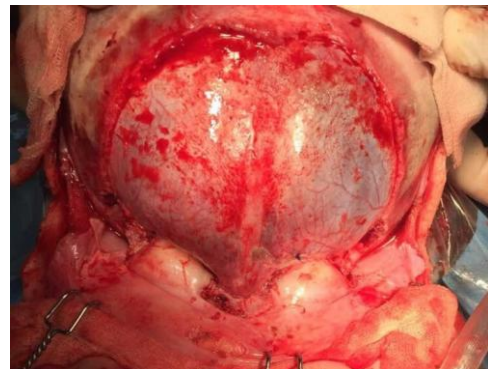


Figure 3: Intraoperative look just after frontal bone craniotomy, supraorbital arch osteotomy. Exposition of the dura underneath and the orbital tissues

The frontal bone, initially removed in one piece, was cut down the middle longitudinally through the metopic suture using a high speed saw, to the level just above the glabella, leaving the glabella as a separate piece, thus creating two halves of the prematurely fused frontal bone, plus the glabella. Each half was additionally cut down the middle (longitudinally), again using high speed saw, thus creating four free bone flaps from the frontal bone. Following, the previously removed supraorbital ridge was addressed.

Two vertical osteotomies were made from each side of the glabella, creating three different parts of the supraorbital ridge. Next, two barrel stave (radial) osteotomies approximately 1 cm wide were cut into the parietal and temporal bone from each side

to facilitate reshaping to match the widened frontal bones, using a Kerrison rongeur. In the next step, the bones were arranged — first, the bony supraorbital ridge, next to the bony glabella and then the four frontal bone flaps. The bone fragments were left loose; they were not fixed in place, as no fixating plates, or other fixating techniques, were used (Figure 4).

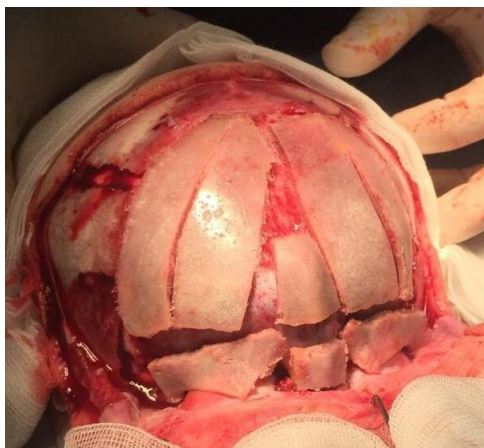


Figure 4: Arrangement of the bone flaps. The bone fragments were left loose, as no fixating plates or other fixating techniques, were used

The previously elevated periosteum, undissected from the skin flap was sutured to the rest of the periosteum using rapidly resorbable polyfilament suturing material, thus covering all of the free bone flaps/bone fragments. The skin was sutured in single vertical mattress sutures using a monofilament nonresorbable suturing material. No epicranial/subperiosteal drainage was used.



Figure 5: Follow up at 10 days postop, just before suture removal

The operating time was 3 hours 45 minutes, and the estimated blood loss was less than 50 ml. The procedure was well tolerated by the patient, and there were no postoperative complications. There was periorcular and facial oedema, most severely expressed on the 2<sup>nd</sup> postoperative day. The patient was discharged on the 3<sup>rd</sup> postoperative day. At follow up visit at ten days from the operation, the incision was well healed, the facial and mid-forehead oedema was in resolution, and no other complications were noted.

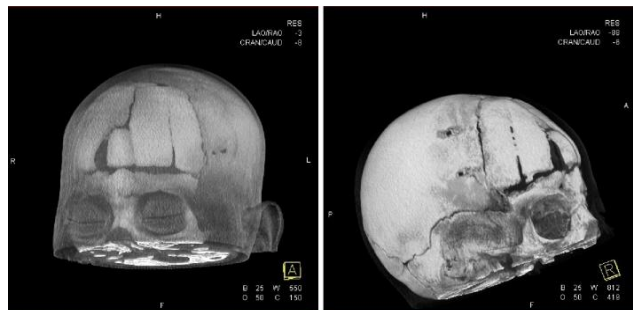


Figure 6: Post-operative 3D reconstructed head CT at 3 months follow up visit

A postoperative CT scan at three months follow up was also obtained demonstrating a satisfactory anterior cranial base decompression with no displacement of the bone flaps (Figure 6, Figure 7).

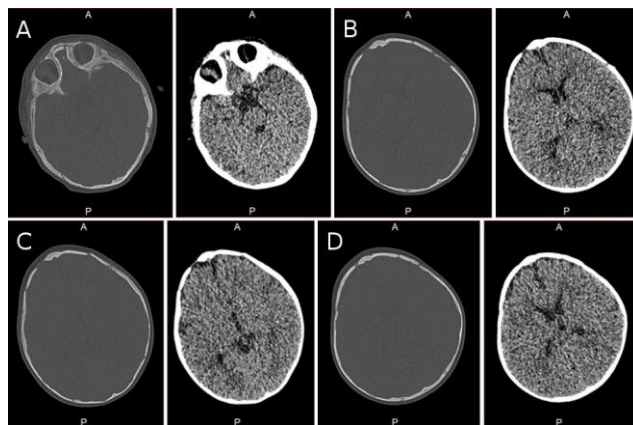


Figure 7: Postoperative head CT scan at 3 months follow up. Note the anterior cranial base decompression

## Discussion

Trigenocephaly is the result of premature fusion of the metopic suture, resulting from restricted lateral growth of the frontal bones, leading to characteristic appearance of “keel forehead”, posterior displacement of the superolateral orbital rims, hypotelorism, flattening of the frontal bones, anterior displacement of the coronal sutures, compensatory

bulging of the parieto-occipital region and temporal narrowing. This results in a triangular forehead, prominent midline sagittal ridge and shortening of the anterior cranial fossa [5]. Normal closure of the metopic suture is expected to occur by nine-month, although normal closure can occur as early as 3 months of age, and yet not all children with prematurely closed metopic suture develop trigonocephaly [6]. Interestingly, trigonocephaly has the highest rate of associated cognitive impairment among the single suture synostosis [7]. The diagnosis can be made based on clinical appearance, although radiographic imaging is often used to rule out an associated intracranial anomaly. Skull x-rays or craniogram is often obtained when a diagnosis is suspected, with minimal clinical value, compared with detailed physical examination of an experienced clinician. Three-dimensional computed tomography (CT) provides a comprehensive view of the suture as well as the overall head shape. However, if a diagnosis of synostosis is suspected and concern for brain pathology exists the use of ultrasound (US), or magnetic resonance (MR) imaging should be considered as a safer alternative to CT. The treatment of trigonocephaly and craniosynostosis remains surgical and is likely to remain so. In general, the two main indications for surgical treatment of any craniosynostosis including trigonocephaly include correction of the skull deformity for aesthetic and psychosocial purposes and ensuring there is adequate space for normal brain growth [8].

The aesthetic deformity associated with craniosynostosis alone is considered sufficient to justify the treatment, the point being the social and psychological impact on the affected children. Increased intracranial pressure (ICP) is an absolute indication for surgical repair. Surgical methods and also techniques for correction of craniosynostosis-related skull deformities have evolved. However, available data have yet to demonstrate a single best procedure for the treatment of synostosis. At the moment the surgical techniques available allow performing an open cranial vault reconstruction or minimally invasive reconstruction using endoscopic techniques. The timing of the operation should also be considered, as the procedures in infants tend to be less invasive. Open cranial vault remodelling procedures are often delayed until 6 to 12 month, mainly because the observation of an increased incidence of revision surgery in patients operated on before the age of 6 months. The endoscopic techniques are generally best performed by 3 to 6 month of age. In our patient, an otherwise a healthy 6-month-old infant, the open technique for cranial vault reconstruction was used. A technical remark in the management in this specific case is the osteotomies performed on the frontal bone and also the remodelling of the supraorbital arch, which,

traditionally, is not often addressed. Although the literature, in general, prefers barrel stave (radial) frontal bone osteotomies, another technique was used in this case. As previously noted, longitudinal frontal bone osteotomies were performed, and the result is quite satisfactory. Also, none of the bone flaps was fixed, frontal bone or supraorbital arch. The postoperative head CT scan at three months postop demonstrates an acceptable anterior cranial base decompression. However, this type of cranial vault reconstruction, as reasoned by the author, may only be used in trigonocephaly, as the approach itself only allows for anterior cranial base decompression. Other types of craniosynostosis require a different exposition, perhaps through the same skin incision, and also different sutures to be addressed.

In conclusion, this modified approach for anterior cranial base decompression in metopic suture synostosis is technically simple and appears to be associated with no greater morbidity than the traditional approach. Potential advantages include reduced blood loss; less postoperative pain decreased the length of stay. Surgical indications are identical as for any other approach. However, due to the satisfying outcome, this technique may become a surgical option of choice in patient's metopic suture synostosis.

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