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Addressing hypertelorism: Indications and techniques

B. Laure^{a,c,d,*}, C. Batut^{a,d}, A. Benouhagrem^{a,d}, A. Joly^{a,c,d}, N. Travers^{b,c}, A. Listrat^{b,c},
A. Pare^{a,c,d}

^a Department of pediatric maxillofacial surgery and craniofacial surgery, Clocheville hospital, 37044 Tours, France

^b Department of pediatric neurosurgery, Clocheville hospital, 37044 Tours, France

^c Reference center for rare craniofacial malformations, Clocheville hospital, 37044 Tours, France

^d François Rabelais university of medicine, Tours university hospital, 2, bis boulevard Tonnellé, 37000 Tours, France

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ABSTRACT

From its first descriptions in the early 1920s to today's use of cutting guides via computer-assisted surgery, surgical techniques to address hypertelorism have progressed. The present article aims to provide historical background and an overview of the development of surgical techniques during the late 20th century and in recent years. First, a historical overview identifies the most important surgical advances leading to the present state of the art. Each major surgical innovation is described, to explain the changes in this surgical field, according to the type of approach. Then, a precise description of today's most recent practices is provided, with particular emphasis on the spectacular advances deriving from computer-assisted surgery. A thorough description of the use of cutting guides throughout the surgical phase is given.

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1. Introduction

The term “ocular hypertelorism” was first mentioned in 1924 in Greig's article on what he then called “an undifferentiated congenital craniofacial deformity” [1]. It was meant to describe the increase in space between the two eyes in two of his patients. However, this description may lead to misinterpretation, insofar as the distance between the eyes does not necessarily correspond to orbit position. Hypertelorism can be simulated without bone anomalies in the case of convergent strabismus or primary or post-traumatic canthal dystopia (epicanthus, telecanthus). The interpupillary and intercanthal distances measured on clinical examination thus do not match orbital reality [2].

Tessier (1967) first introduced the notions of “orbital hypertelorism” and “hyperteleorbitism” [3], with the purpose of avoiding confusion. Orbital hypertelorism should refer solely to the enlargement of distance between the two orbits due to skeleton anomaly in the context of a craniofacial malformation. Tessier defined hypertelorism as a craniofacial malformation that associates an enlarged ethmoid related to the nasal root and an increase in distance between the frontal process of the maxilla, medial canthi and eyes [4].

Greig viewed hypertelorism as an isolated idiopathic anomaly, whereas Tessier highlighted the fact that they are mostly linked to multiple cranial, facial or craniofacial malformations [3,4]. Orbital hypertelorism is hence often associated with:

- cranial or craniofacial malformation: meningoencephalocelis, craniofacial dysostosis, frontonasal dysplasia, or craniofrontonasal dysplasia;
- facial malformation: maxillary and nasal malformation, or median maxillopalatine cleft.

The inter-orbital distance (IOD) between two dacryons, in other words the junction between the maxillary, frontal and lachrymal bones, enables diagnosis of 3 types of orbital hypertelorism: 30 mm < IOD < 34 mm (1st degree), 34 mm < IOD < 40 mm (2nd degree) et IOD > 40 mm (3rd degree) [5]. IOD can be easily measured on CT scan after locating the dacryons.

2. Some background on the history of surgical correction of orbital hypertelorism

Since the mid-1950s, many surgeons have attempted to address this anomaly via naso-orbital skin or bone resection [6–9]. However, results were disappointing (Fig. 1). During the 1960s, Tessier developed extra-cephalic and infra-ethmoidal correction techniques for hypertelorism, with various adaptations, known as the 3 B techniques: B1, B2 and B3 (Fig. 2). These intra-cranial

* Corresponding author. Department of pediatric maxillofacial surgery and craniofacial surgery, Clocheville hospital, 49 boulevard Beranger, 37044 Tours, France.
E-mail addresses: b.laure@chu-tours.fr, boris.laure@wanadoo.fr (B. Laure).

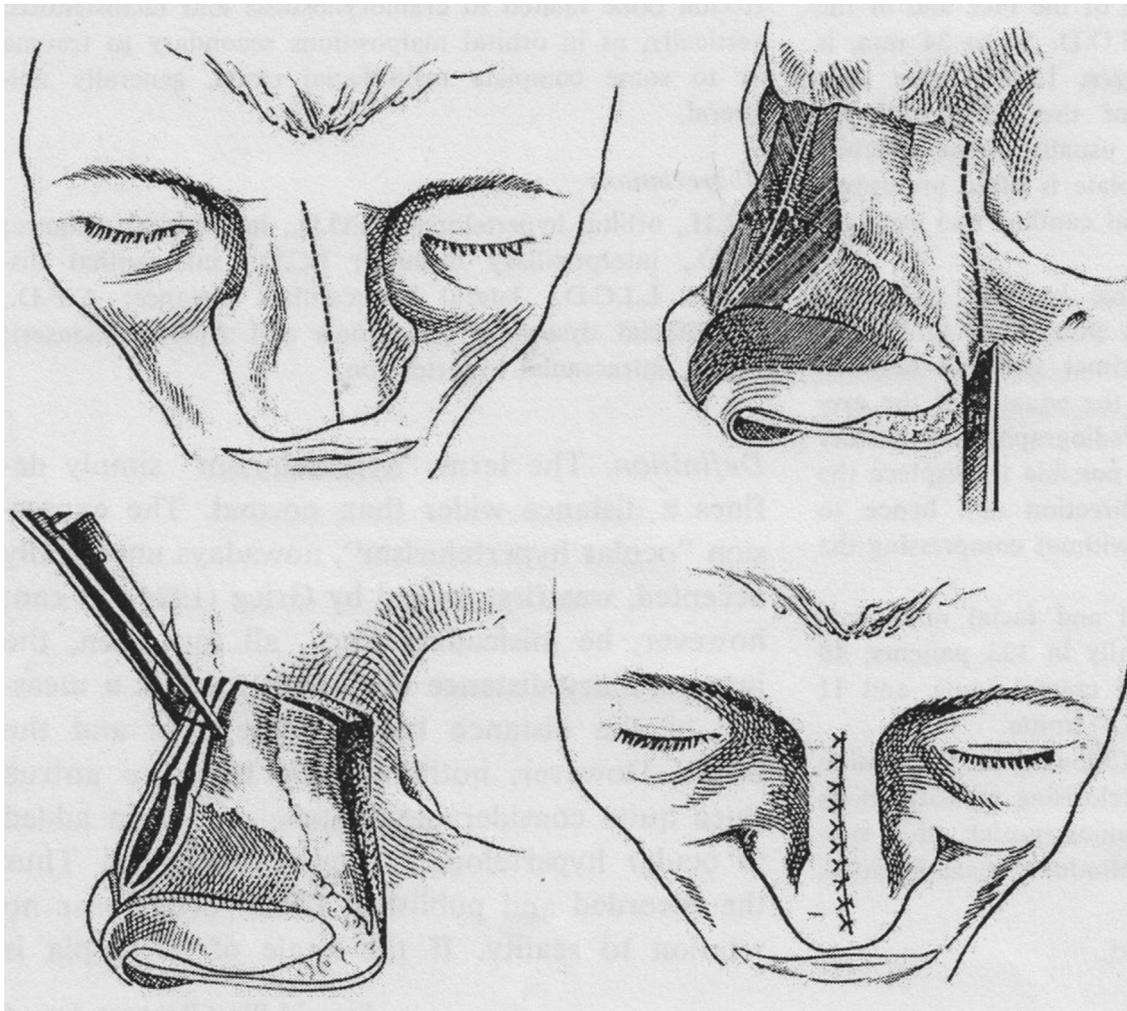


Fig. 1. 1st degree orbital hypertelorism (Courtesy of the Collection Tessier by the French Association of Facial Surgeons, AFCF).

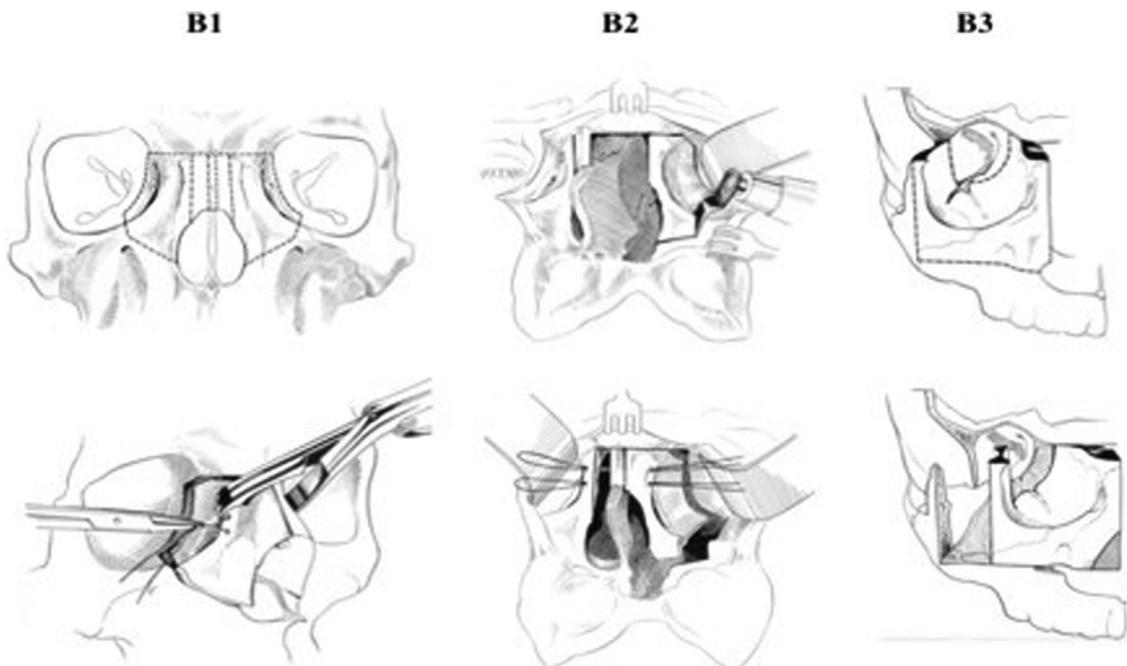


Fig. 2. B Procedures: extra-cranial techniques of orbital hypertelorism correction (Courtesy of the Collection Tessier by the French Association of Facial Surgeons, AFCF).



Fig. 3. Mrs. L., Tessier's first case of orbital hypertelorism (Courtesy of the Collection Tessier by the French Association of Facial Surgeons, AFCF).

techniques are, however, considered insufficient and not well adapted to orbital hypertelorism, and no surgical technique showed real efficacy at that time.

In 1967, Tessier finally came up with a new surgical proposal, consisting in inter-orbital distance reduction with a 2-step intra-cranial procedure [3]. This innovation was the result of team-work between Tessier and some neurosurgeons, especially Guiot, in the

Foch Hospital (Paris), that led to the merging of the two specialties [10] in what became the beginning of craniofacial surgery.

To address the first case of extreme hypertelorism (Fig. 3), Tessier developed three new intra-cranial techniques of correction of orbital hypertelorism, called the A procedures (Fig. 4). Until 1968, they were implemented following a 6-month period of skull-base sealing by skin graft. The first step was intended to limit the risk of meningitis while the second step consisted in inter-orbital distance reduction. This is how Tessier inspired a whole generation of surgeons who, in turn, contributed to the development of craniofacial surgery. The box osteotomy technique first introduced by Tessier progressed during the late 20th Century, enabling 1-step correction of orbital hypertelorism, while conserving surrounding structures, and notably the olfactory roots [11].

In 1976, Van der Meulen [12] described the facial bipartition technique that enables 1-step correction of the orbital anomaly and of the maxillary malformations that can be associated (Fig. 5). It also enables correction of orbital hypertelorism associated with vertical and transversal maxillary deficiency inducing open bite.

Inter-orbital distance reduction by box osteotomy and facial bipartition is still the gold-standard procedure for moderate to severe hypertelorism. The choice of technique depends on possible associated maxillary deficit.

3. Surgical techniques

3.1. Indications

The two major surgical procedures to address hypertelorism are box osteotomy and facial bipartition. The choice between the two is dictated by maxillary width: when it is normal, box osteotomy is indicated or, when it is short, facial bipartition. Soft-tissue

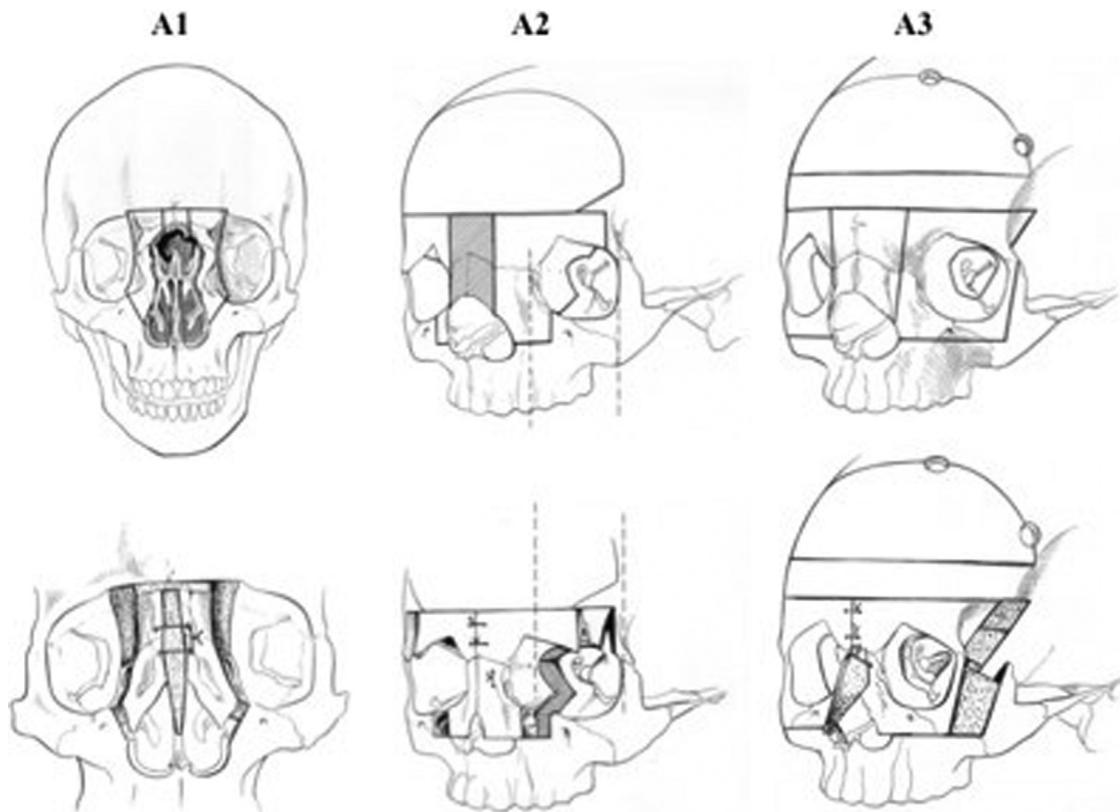


Fig. 4. A Procedures: intra-cranial techniques for orbital hypertelorism correction (Courtesy of the Collection Tessier by the French Association of Facial Surgeons, AFCF).

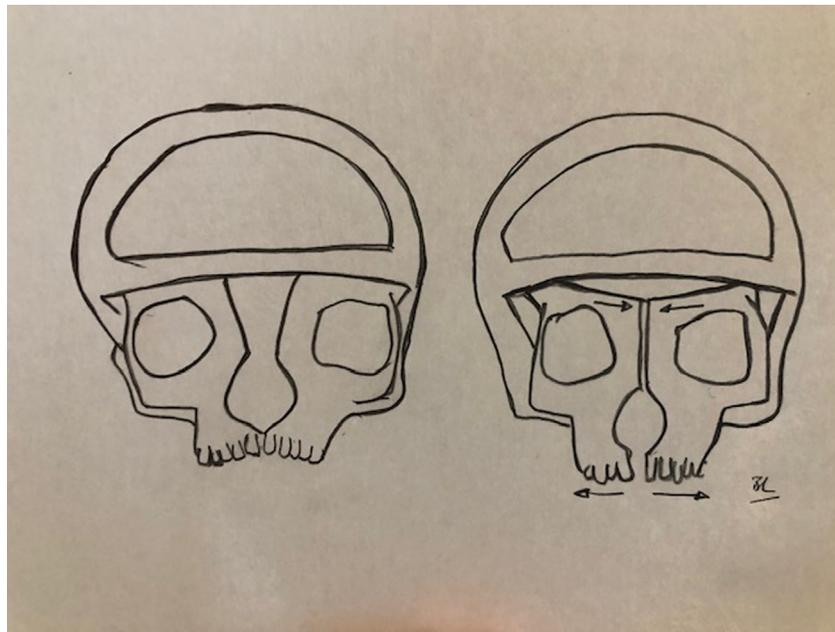


Fig. 5. Facial bipartition procedure.

management is more difficult, and depends on the classification of the malformation.

3.2. Box osteotomy

Deriving from techniques developed in the late 20th Century, today's surgical procedures for moderate to severe hypertelorism are as follows: the approach is coronal, allowing subperiosteal dissection down to the zygoma, followed by circumferential bilateral orbital dissection. The temporal muscles are removed from the temporal fossae, in order to access the lateral orbital walls. Then a frontal craniotomy is performed to access the skull-base. A supra-orbital arch needs to be conserved, in order to fix the future orbital osteotomies. Osteotomies of the 4 orbital walls and lower horizontal osteotomy from the piriform aperture to and then through the zygoma allow displacement of the useful orbit supporting the eyeball during the various transverse or vertical bone movements. The osteotomy line is just below the inferior orbital foramen, preserving the infra-orbital nerve. These actions leave the posterior part of the zygoma and the zygomatic arch in place.

Monobloc osteotomy of the two orbits is then conducted, with central bone resection of the nasofrontal region, allowing the eyes to move closer to one another (Fig. 4). The nasal bones, ethmoidal labyrinth and cribriform plate used to be removed, but this is no longer the case, since it tended to impair olfactory function. Along with the bone resection, skin resection is sometimes required, particularly in case of bifid nose or in the more frequent case of excess skin resulting from the orbital correction. Finally, bone grafts are required to fill the defects usually left by orbital correction. More specifically, bone grafts are necessary around the zygomas and lateral sides of the orbits, to prevent potential enophthalmos.

There are some variants of this generic procedure.

One: box osteotomies are performed by central resection when the nasal dorsum has major defects and needs to be replaced. With this technique, the nasal dorsum is removed, leaving a supra-orbital arch in place. The orbits are then moved as close together as necessary, and the nose is finally reconstructed with a bone graft (Fig. 4).

Two: box osteotomies can be performed by paracentral resection, with an arch left in place when removing the paracentral bone elements (Fig. 6). The orbits are then corrected and the olfactory

nerve can be more easily spared than with the central approach. Fig. 7 shows the case of a young boy with hypertelorism before surgery, and Fig. 8 shows the outcome 1 month after surgery on a paracentral approach.

Three: box osteotomies can be performed without conserving the supra-orbital arch. Marchac [13] described a technique that involves lateral spurs created by frontal bandeau removal. These spurs allow orbital correction along their edges, which act as markers for good stabilization and fixation (Fig. 9). Bone graft is required to fill the space left on the orbit sides, and to reconstruct the nasal dorsum.

Four: in case of orbito-zygomatic asymmetry, more specifically in the sagittal plan, it is possible to modify the osteotomy so as to reposition the whole zygoma and thus restore symmetry (Fig. 10).

3.3. Facial bipartition

Van Der Meulen first introduced this technique in 1976 [12,14]. It was called “median faciotomy” and differed from Tessier's original technique in that the infra-orbital osteotomy was performed differently, separating the orbit and the maxillary. The osteotomy line was replaced by a pterygomaxillary and an intermaxillary disjunction. This technique allowed rotation of each hemi-face, which in turn allowed appropriate orbital correction and simultaneous transverse widening of the maxillary (Fig. 6). Two triangular defects were left below the frontal bone on each temporal side, which needed filling by bone grafts. This technique was used with patients with a specific transverse maxillary arch defect.

3.4. Limitations of the technique – complications – how to avoid them – and how to assess results

These techniques should be reserved for children and parents with strong motivation. The most important question is age at correction of hypertelorism and possible relapse. Some teams, such as Raposo-Amaral's, recommend > 8 years' age at correction, because otherwise the relapse rate is significantly higher [15]. Other teams, such as Marchac's, perform bone surgery earlier [13].

The most usual complications are cerebrospinal fluid leakage and associated infection. This can be resolved by lumbar

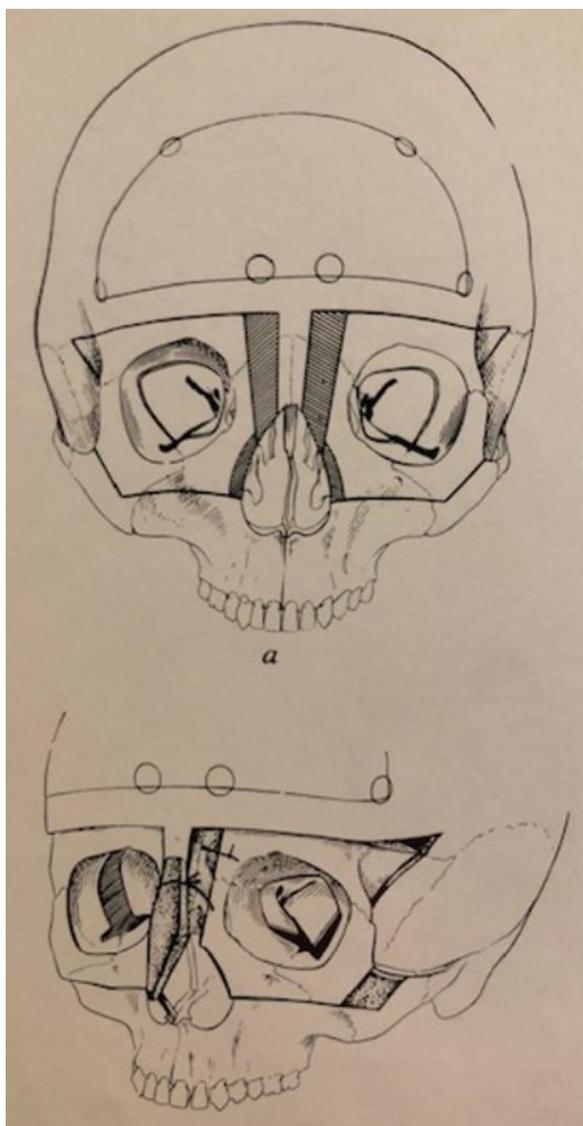


Fig. 6. Box osteotomy with paracentral resection.

puncture or a lumbar drain. Rare cases of blindness or death have been reported. Nevertheless, it is mandatory to collaborate with a strabologist/ophthalmologist, as shortening the two orbits can induce oculomotor disorder.

Bone assessment is straightforward on CT. Soft-tissue assessment is more difficult; the Whitaker classification on standardized facial photographs is useful [16].

3.5. Soft-tissue management

Soft-tissue management is the greatest challenge in the correction of hypertelorism.

The three major corrections concern the midline, with excess soft tissue after bone shortening, canthi positioning, and nasal correction. As Paul Tessier so rightly said, “The correction of hypertelorism is surgery of the nose”.

For excess midline soft tissue, direct resection can be performed with or without Z plasty. Kawamoto described the “K-stitch technique”, reducing soft tissue without scar [17].

For canthi repositioning, it is essential not to use subperiosteal dissection of the inner canthus. Sometimes transnasal canthopexy with wire is necessary, with or without epicanthal-fold Z plasty

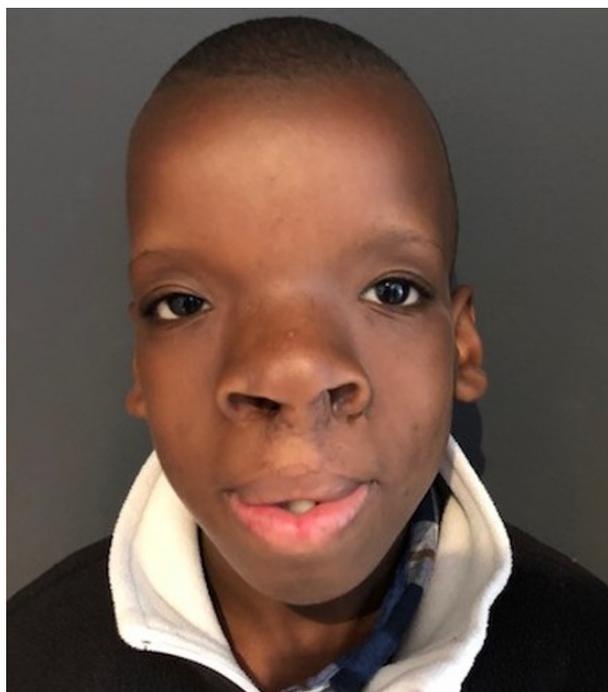


Fig. 7. Hypertelorism, before surgery.

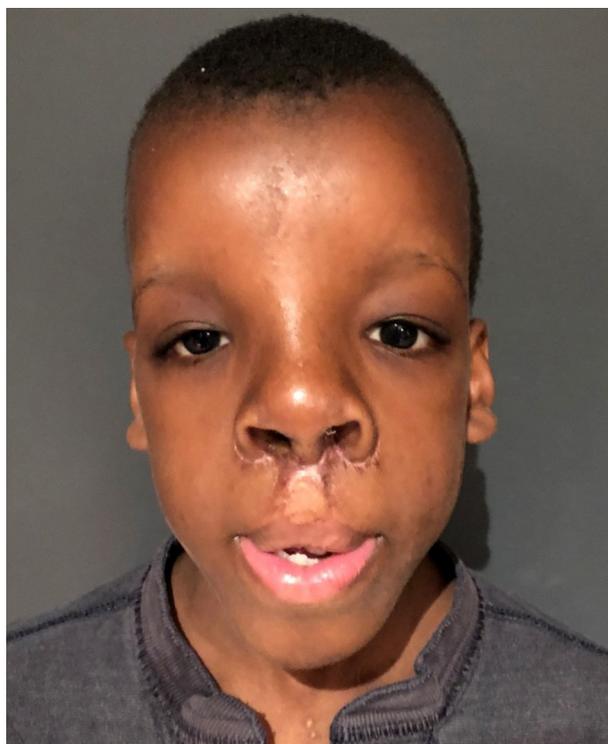


Fig. 8. Hypertelorism, one month after surgery with paracentral approach.

[18]. We systematically perform lateral canthopexy via a coronal approach.

For the nose, the soft-tissue surgical approach is at the surgeon's discretion. In frontonasal dysplasia, open rhinoplasty with bifid nasal tip correction is performed. We systematically reconstruct the nasal dorsum with a calvarial bone graft; but other sites are also available (ilium, ribs).

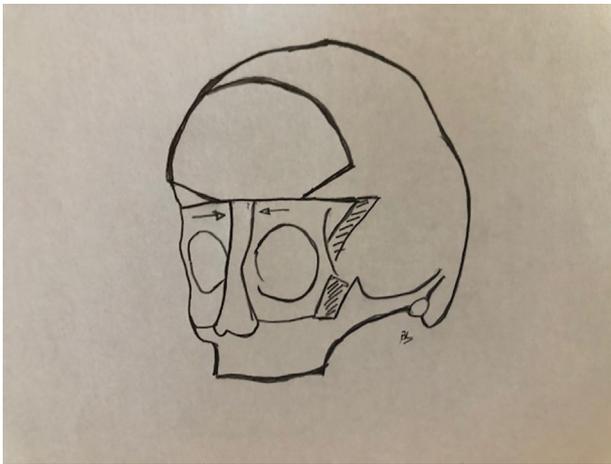


Fig. 9. Box osteotomy with central resection and lateral spurs.

In case of wide craniofacial cleft, a forehead or scalp flap is mandatory to reconstruct the nasal soft-tissue defect.

4. Modernization of surgical techniques

Surgical procedures for orbital hypertelorism have undergone little change since the second half of the 20th Century. This is all the more paradoxical as more recent techniques are increasingly effective: osteosynthesis plates, 3D printing, computer-assisted surgical planning.

These new technologies appeared in the 1980s, when an anatomical model with detailed geometric characteristics was constructed from CT-scans. This technology was used to produce rough physical models of the head and face. However, the complexity required for detailed anatomical models could not be replicated. More effective techniques, such as stereolithographic models, enabled 3D printing of solid cranial models that could be used in various indications in craniomaxillofacial surgery: for instance, to guide implant manufacturing or to plan frontonasal dysplasia surgery.

In turn, stereolithography became a precise and pragmatic tool for planning hypertelorism correction, as it enables any malformation to be observed from various angles. Therefore, the surgical team has a more tangible approach, with good prediction of otherwise unexpected events that could occur during surgery. Sailer et al.

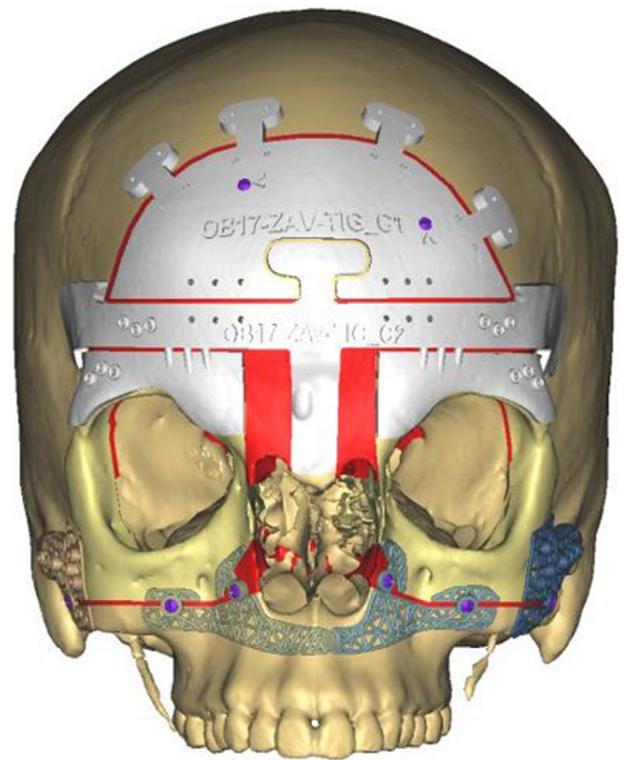


Fig. 11. Presurgical plan of box osteotomies: polyamide and titanium cutting guides.

were the first to use a stereolithographic model in surgical simulation for facial bipartition, in the 1990s [19]. Before the advent of this new technology, hypertelorism correction could only be simulated on anteroposterior cephalograms by geometrical planning, which was less accurate [20].

Subsequently, computer-assisted surgery (CAS) became a prominent tool in surgical planning, and its use has been widening to more craniofacial applications in recent years [21,22]. However, very few teams have used CAS planning and cutting guides in hypertelorism [23,24]. There are several reasons for this. The first is that few pathologies are indicated for such use of CAS, and the second is that many surgical teams have not yet been trained in full extensive use of all of the possibilities offered by CAS. Our own team has been implementing use of cutting guides for

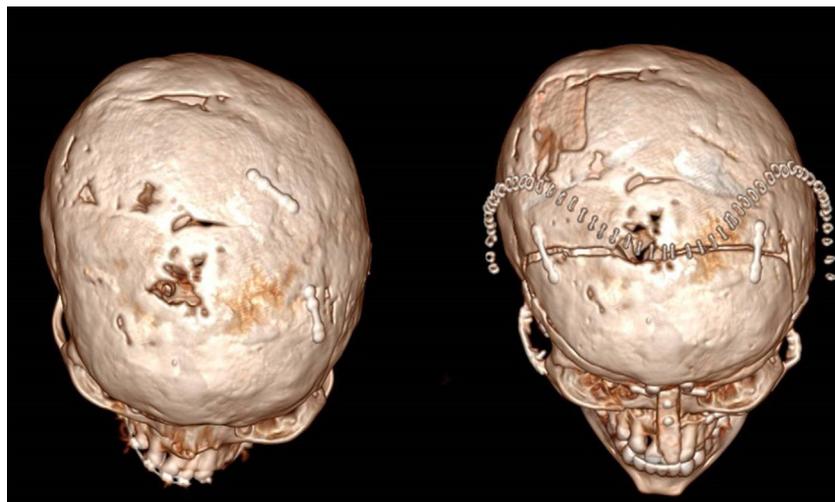


Fig. 10. Box osteotomy with whole zygoma repositioning: upper view of 3D CT scan with asymmetry in the sagittal plan (pre- and post-operative).



Fig. 12. Virtual image of the hypertelorism reconstructed via DICOM based on patient's CT-scans.

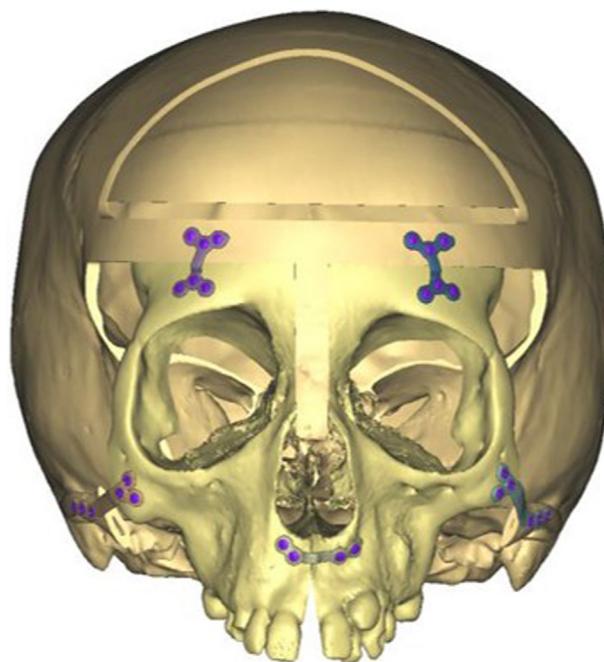


Fig. 14. Simulation of osteotomies repositioning.

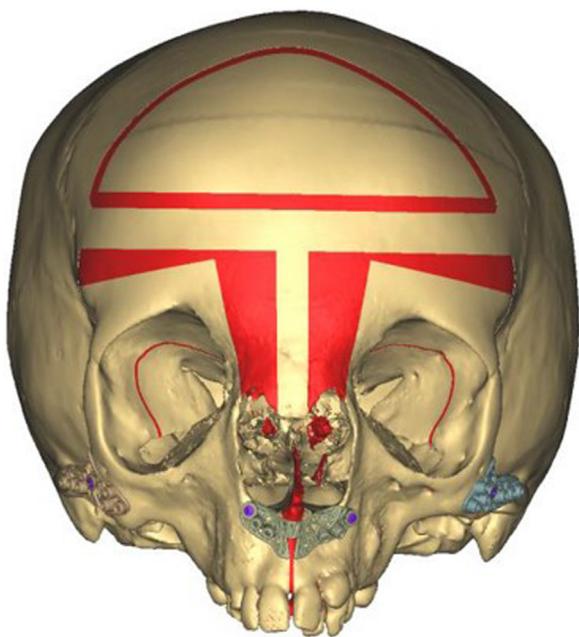


Fig. 13. Simulation of facial bipartition osteotomies.

hypertelorism surgery for about 4 years, with satisfactory results in terms of cutting precision and of 3D repositioning of osteotomies [25].

There are the three stages that our team usually goes through in such surgeries.

Stage 1 is the presurgical step during which the surgeon works hand-in-hand with the clinical engineer, using CMF Planner[®] 3.0 software (Materialise, Leuven, Belgium). First, the surgeon transfers all the appropriate CT data to the clinical engineer in DICOM format. Then, the engineer designs the osteotomy delineation following the surgeon's instructions. Osteotomies vary depending on the shape of the hypertelorism (symmetrical, asymmetrical) and on the nature of the osteotomy (box osteotomy, facial bipartition, osteotomy with both zygomas or not, etc.). The clinical engineer

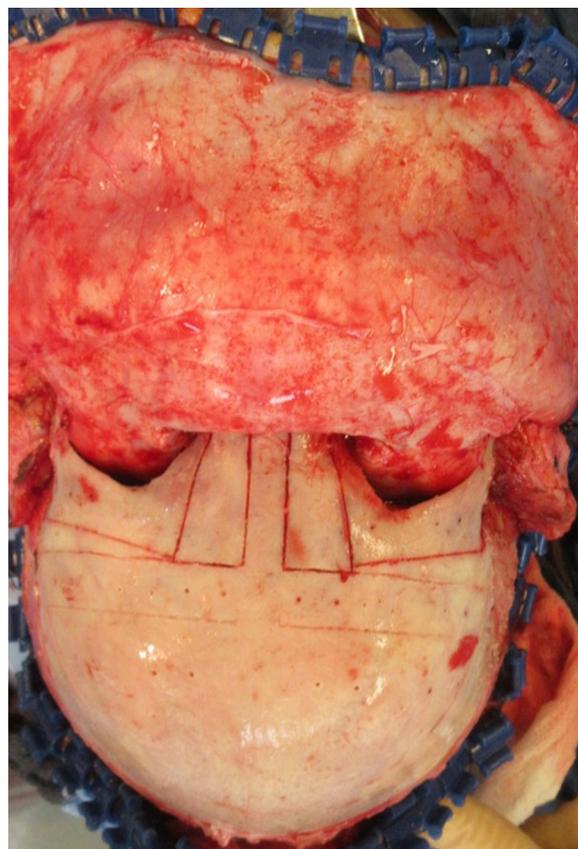


Fig. 15. Intraoperative marking after the use of the cutting guide.

can then simulate various hypotheses, which enables the surgeon to decide upon the most appropriate option and to visualize the expected outcome of surgery (Fig. 11). More specifically, the inter-orbital distance can be defined with very good precision for final outcome, which enables the surgeon to know ahead of surgery

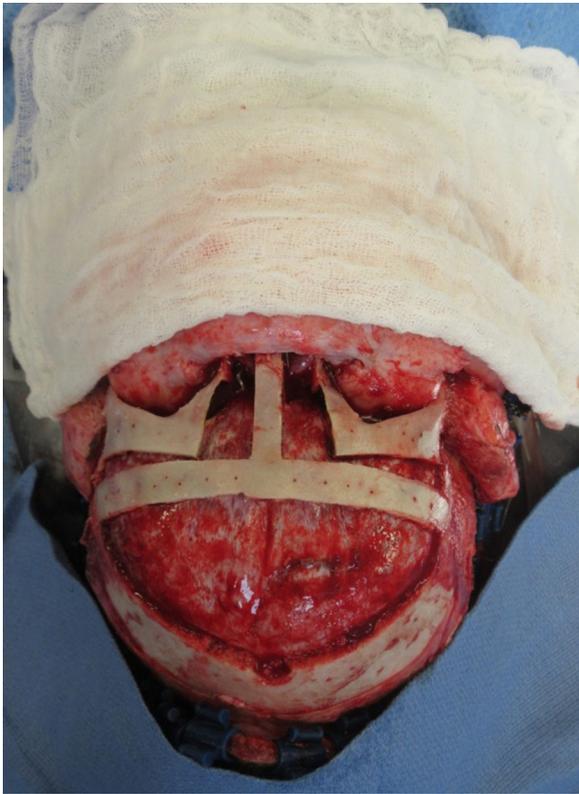


Fig. 16. Intraoperative osteotomies.

what objective is to be achieved. **Figs. 12–14** provide an example of simulated images used in a case of hypertelorism managed by our team. We performed facial bipartition to shorten the inter-orbital distance and to increase maxillary width.

Once the surgeon has validated Stage 1, it is time to design the cutting guides and repositioning guides (Stage 2), using 3-matic®-11.0 software (Materialise, Leuven, Belgium). Again with the help of the clinical engineer, the first task is to calculate very precise cutting guides that will be used in the first step of surgery. In the particular case of hypertelorism, two types of cutting guide need to be designed. One is a large white polyamide guide delineate the forehead, paracentral and supra-orbital resections and

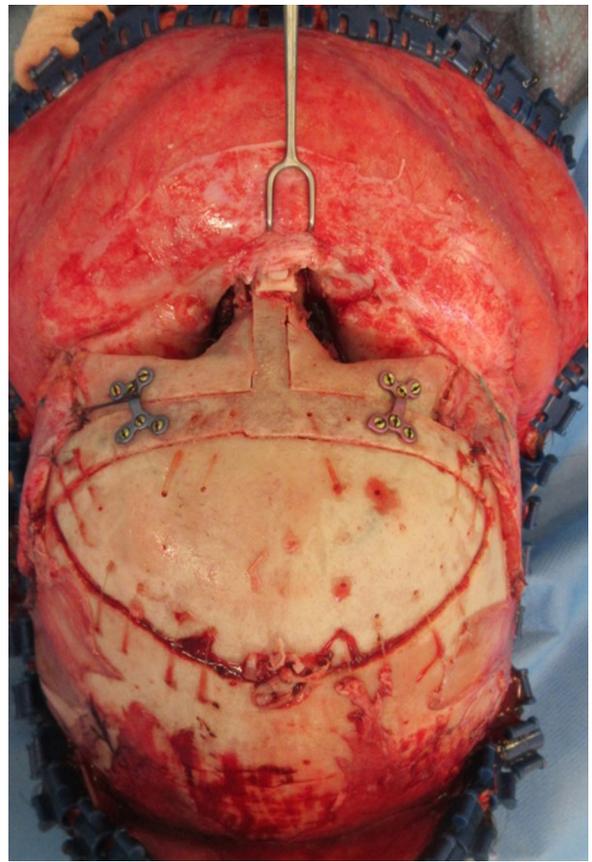


Fig. 17. Perfect fit of the osteotomies after using the titanium repositioning and osteosynthesis plate.

landmark pre-drilling for the future positioning of the titanium repositioning plates. The second cutting guide is made of titanium, and is designed to landmark the future cutting in the maxillary and zygoma regions, and for pre-drilling ahead of future repositioning.

Stage 3 is in the operating room. First, the surgeon needs to position the cutting guide, and then the exact contours of the cutting areas are marked with the burr. After marking, the cutting guide is removed, enabling the marking to be clearly seen (**Fig. 15**). Next, osteotomies are performed following the exact marking, which

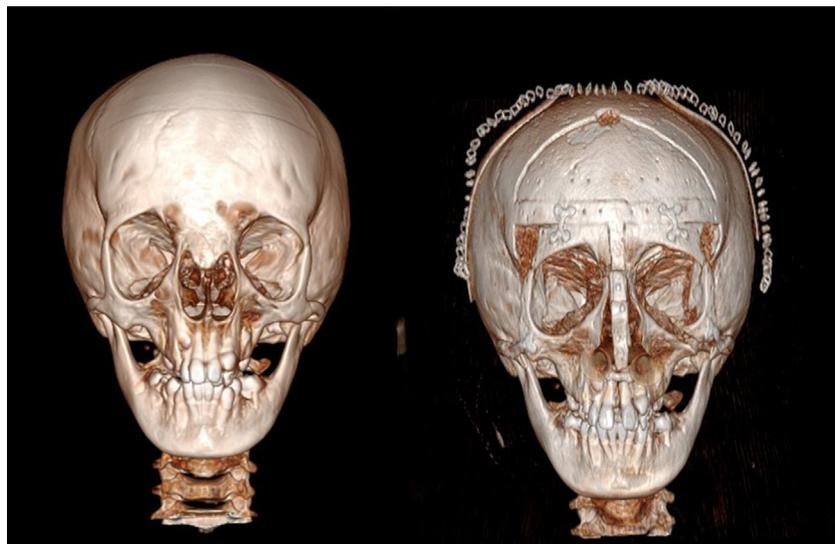


Fig. 18. Pre- and post-operative 3D CT-scans of facial bipartition.

leads to very precise cutting (Fig. 16). Finally, the osteotomies are implemented as planned ahead of surgery thanks to the use of repositioning guides. Fig. 17 displays the outcome in a case of hypertelorism. The fit was very accurate and the titanium plates were perfectly in place, thanks to the customized titanium repositioning plates used for both osteotomy positioning and internal fixation. Finally, pre- and post-operative CT-scans provide a good view of the surgical precision (Fig. 18).

Overall, the surgical tool introduced here does not radically change the surgical skills required in such cases. It only enables better surgical planning and a better focus during surgery on the essential technical gestures. The greatest benefit of this technology is that repositioning is much more precise and effective than with the traditional technique, as 3D positioning is possible with the repositioning guide. Without this, the surgeon may have difficulty in perfectly reconstructing and reassembling the various parts of the skull. To conclude, this is a more time-saving and effective approach than previously available.

Disclosure of interest

The authors have not supplied their declaration of competing interest.

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