What the time has taught about Hyperteleorbitism

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Introduction: Hypertelorbitism is a clinical feature that may be present in various types of pathologies and in various degrees of manifestation. Observing the evolution of techniques over time and understanding the specific characteristics of each case demonstrates the difficulty of establishing treatment protocols.

Material and method: Retrospective analysis of over 150 cases of hypertelorbitism operated on over 44 years by the main author. The increased interorbital distance had varied etiologies: dysplasia, meningoencephaloceles, and dysostosis. Ages ranged from 3 months to 38 years.

Results: Retrusion of the facial middle third throughout growth in congenital hypertelorbitism was present in both dysplasia and dysostosis groups. In cases of early treatment with Tessier's glasses osteotomy there was zygomaticmaxillary atrophy with trapping of tooth germs. The cases of grade III and IV Hyperteleorbitism, with or without meningoencephalocele, which were treated without concomitant skullcap expansion, evolved to endocranial hypertension and required further treatment. Retrospective evaluation of their CT scans revealed craniosynostosis of one or more sutures, associated with predominant dysplasia. Patients in all groups treated after adolescence showed stable late postoperative results, as well as those treated in childhood by the technique of facial bipartition.

Discussion: Hypertelorbitism is a clinical feature that may be present in various types of pathologies and in varying degrees of intensity. Many treatment options have been proposed over the past 50 years. The opportunity to watch and experience the evolution of these techniques made it possible to choose the most appropriate treatment for each patient's clinical characteristics.

Conclusion: Due to the variability of manifestation of hypertelorbitism, the massification of procedures and the establishment of treatment protocols are quite limited.

Keywords: Hyperteleorbitism, orbital hypertelorism, frontonasal dysplasia, frontoethmoidal meningoencephalocele, rare facial clefts.

INTRODUCTION

In a hospital, the surgical center is one of the most Distances greater than 25mm between adult orbits are called Hyperteleorbitism. (1) This clinical characteristic may be present in both congenital (Dysplasias and Dysostoses (2)) and acquired pathologies (trauma, tumours, mucoceles). As the acquired pathologies do not compromise all the orbital walls, in practice we will have true orbital hypertelorism only in congenital form (3), so we will focus only on it.

In dysplasias, the orbits are distant due to changes in the fusion mechanism of embryonic processes, which can be fissures or hyperplasia with mesodermal effusions. Dysplasias classified by Paul Tessier as 14, 13, 12 and 11 (4) are the ones that can manifest as hypertelorbitism (figure 1).

Usually the nose is deformed, due to the continuity of the dysplastic defect in the middle third of the face, since the dysplasias 14 , 13,12 and 11 continue with dysplasias 0, 1, 2 and 3. In this group it is common to find central bone defects, with agenesis of the nasal bones (dysplasias 0-14) which suggests the needing of bone grafting during surgical treatment procedure, in addition to the nasal reconstruction itself

The predominance of the Shh factor of the HedgeHog family (neuroectoderm cytokines) between the third and fourth week of intrauterine life determines the suppression of Pax6 (of this same family of cytokines) which is responsible for the medial migration of the optic process (5). Thereby, not only the nose suffers an evolutionary stop, but also the floor of the anterior cranial fossa, which no longer

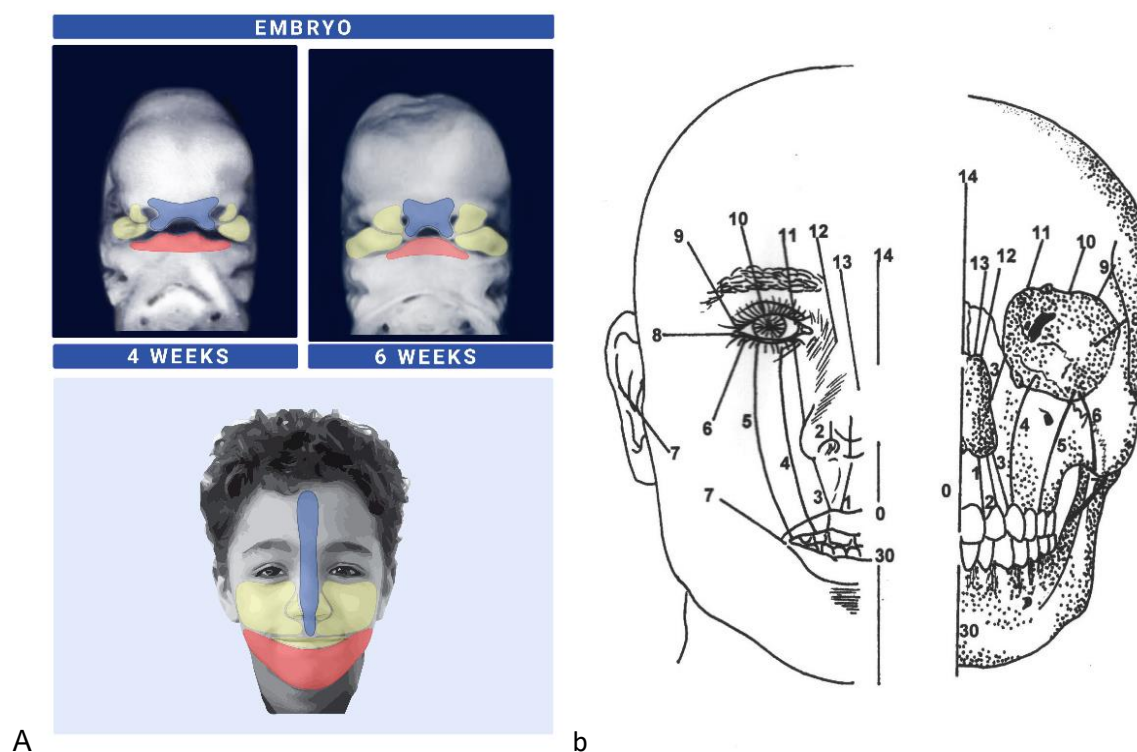**What the time has taught about Hyperteleorbitism**

Figure 1- a- Embryonic areas involved in the onset of hyperteleorbitism, in blue; b- Paul Tessier's Classification of Fissures/Dysplasias**.**

adequately contains the brain tissue in formation. That is why the presence of frontoethmoidal meningoencephalocele is common in frontonasal dysplasias.

In Dysostoses, the underlying pathology is the premature fusion of bony sutures of the skull and face, caused by altered sutural osteogenic factors. In the dura mater attached to stenotic sutures cell proliferation is decreased and there is an increase in osteogenic cytokines (TGF-Beta1 and TGF-Beta2) and extracellular matrix molecules (collagen type I and II, osteocalcin and alkaline phosphatase). (6)

Premature fusion of some sutures prevents adequate expansion of the skull, and in order to sure the growing brain does not suffer, the patent sutures grow beyond what would be anatomically expected. When the fronto-sphenoethmoidal sutural complex is affected, there will be craniofacial suture stenosis, with bone growth restriction of both the skull and the face (figure 2). Beside that the metopic suture and ethmoidal sutures are normal; they will grow compensatory trying to offer the brain the space that is being denied by the stenosis of other sutures. Thereby, hypertelorbitism will have been installed.

In this group, the nose is spared from deformity and bone continuity is preserved in the nasal region, which can serve
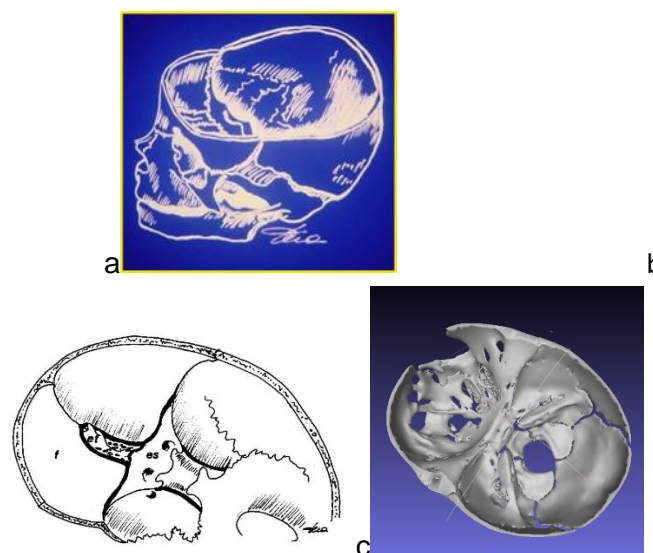

Figure 2- Frontosphenoethmoidal Sutural Complex, affected in craniofacial stenosis and in some craniosynostosis.

as a three-dimensional reference for surgical mobilization of the orbits.

Although didactically the etiologies can be well distinguished between Dysplasias and Disostoses, in a single case may be several specific features of the two etiologic mechanisms.

The etiological and clinical presentation differences between congenital hypertelorbitism already points to the need for a thorough understanding of each clinical manifestation so that the global treatment of the affected patient can be adequate as possible. Based on the experience accumulated in 44 years of care for congenital craniofacial malformations, we will bring relevant considerations to the approach of the cases.

MATERIAL AND METHOD

This paper performs a retrospective analysis of congenital hypertelorbitism cases performed over 44 years by the main author. Ages ranged from 3 months to 38 years. Historical aspects and the evolution of the techniques were discussed considering the author's experience, highlighting points of anatomical and functional interest, as well as technical details improved during that time.

RESULTS AND DISCUSSION

The treatments performed in the group of patients with dysplasia could be grouped according to the table 1:

Table 1 – Frontonasal dysplasia treatment options

Among patients with Hyperteleorbitism in craniofacial stenosis, treatments were associated with advancement of the middle third of the face, varying between those that were accompanied by frontal advancement (in Monobloc) or not, as shown in the table 2

Table 2 – Craniofacial dysostosis treatment options

Techniques of intracranial approach in Dysplasias:

Frontonasal Dysplasias with the presence of meningoencephalocele:

When the fusion defect of embryonic process is sufficiently intense to the point of brain tissue extravasation beyond the skullcap, three basic reasonings are necessary:

-In the bone defect through which the meningoencephalocele is extruded, there is no osteogenic matrix, and any bone graft that corrects this defect must be composed with the parietal periosteum inserted into the bone table so that it can supply osteocytes to revitalize the graft.

-When a portion of brain tissue overflow from the cranial vault, not only the overflowed tissue has a low level of functionality, but also the brain remaining in the cranial vault brings limited viability, with structural agenesis in addition to large and deformed ventricles (figure 3). In this scenario, it is reckless to resect and discard the extruded brain tissue. And if this tissue is simply reduced into the skull, it will create endocranial hypertension. Therefore, it is mandatory to expand the skullcap to a volume proportional to the expected increase at meningoencephalocele reduction. The cases operated on before learning this maxim all evolved with endocranial hypertension, and required complementary cranial decompression surgery. Currently, all cases in which meningoencephaloceles are reduced or in which the floor of the anterior fossa is raised to medialize the orbits (Tessier grade III or IV hyperteleorbitisms) receive proportional expansions of the skullcap with dynamic osteotomies: or sutural activation with springs(7)(8) or expansion with helical osteotomies named Nautilus (9).
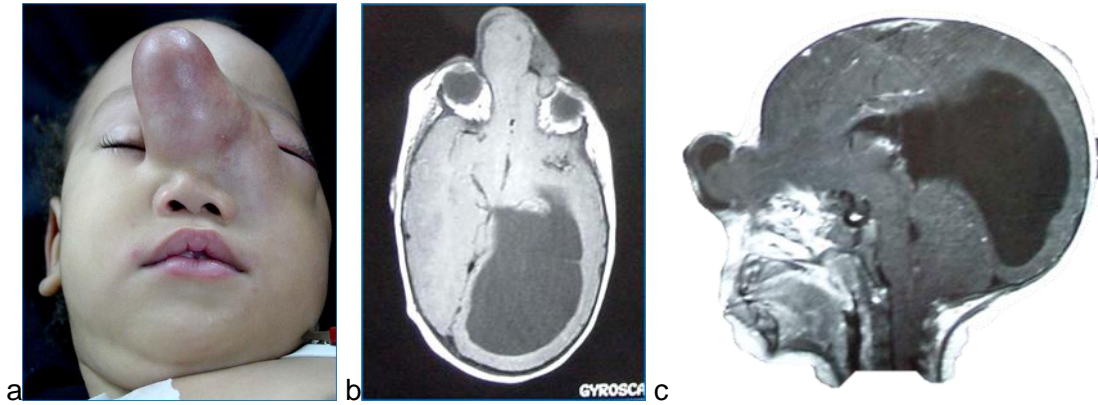
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Figure 3- a- Hyperteleorbitism with transethmoidal meningoencephalocele; b- axial tomographic section; c- sagittal tomographic section.
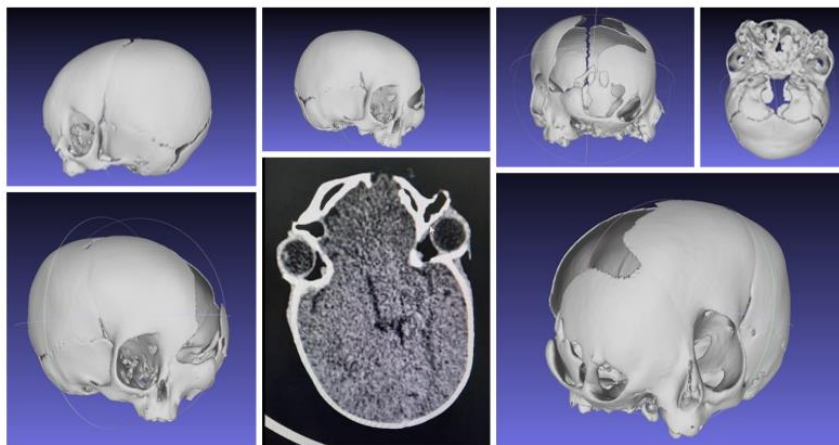

Figure 4 - Hyperteleorbitism with great dystopia of the brain in the ethmoidal area, showing bilateral coronal and squamous craniosynostosis.

-This finding of rebound endocranial hypertension to meningoencephalocele reduction led to a more careful retrospective study of skulls with frontonasal dysplasia, making it possible to find premature fusions of one or more sutures in many of the cases that were considered to have only dysplasia, not dysostosis (Figure 4). It remains to be known whether these synostosis are primary, due to alteration of the osteogenic matrix, or secondary, in response of the emptying of the cranial cavity by meningoencephalocele. Whatever its nature, the adaptation of the intracranial space to the reduction of meningoencephalocele must be done. And the choice of dynamic osteotomies in which the dural envelope is expanded by mobilizing the bone to which it is inserted seems to be the most adequate for this expansion purpose

Techniques for correction of hyperteleorbitism using an intracranial approach:

The first technique described for the correction of hyperteleorbitism was orbital box osteotomy by Paul Tessier (10), which for many years remained the only treatment option in all craniofacial surgery centers (Figure 5).
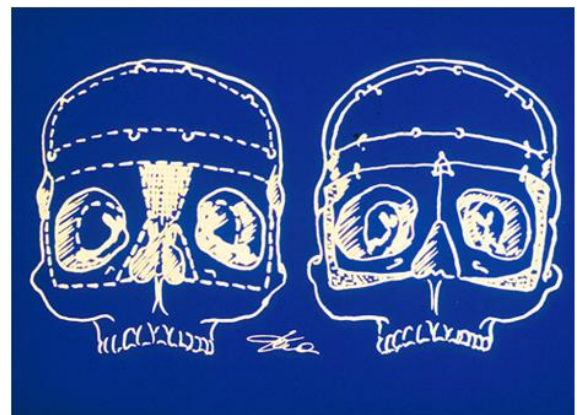

Figure 5- Schematic of the orbital box technique or Paul Tessier glasses for the treatment of Hyperteleorbitism.
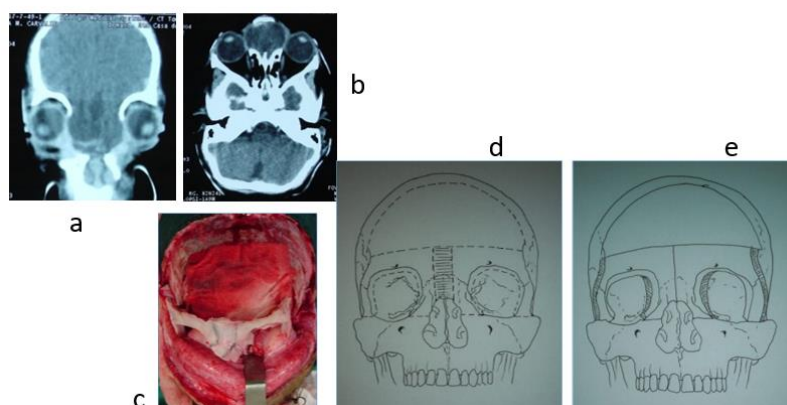
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Figure 6:-Hyperteleorbitism in tomographic section (a) coronal and (b) axial; c- intraoperative view of the central ostectomy demarcation; d- scheme of inverted "U" orbital osteotomies; e- orbital walls mobilized medially.

Figure 7- Psillakis at all T-Delta technique: a- tracing of osteotomies and preoperative; b- mobilized and postoperative orbital walls.

Our older patients, who were operated on using this technique, presented two constant characteristics in the late postoperative period: large maxillozygomatic hypoplasia, corresponding to the area of the inferior horizontal osteotomy of the orbital box, where the maxillary sinuses did not develop; and trapping of dental germs in the orbital floor throughout the entire period of growth and adulthood, compromising not only the masticatory function (due to the absence of dental parts) but mainly the airway, due to maxillary retrusion and deformation of the maxillary sinuses. This finding led us to mobilize the orbital walls in an inverted U shape, leaving the orbital floor intact, which acts as a horizontal plane for the medial sliding of the orbits (Figure 6)

The radical resection of the interorbital bone tissue, to allow medialization of the orbits, also led to two problems: the first, the need to replace the original nose and glabella bones with bone grafts, with all the aesthetic and viability limitations that this entails. And the second, that the void left by the central bone resection made the medialization of the orbits extremely difficult to make an arcuate plane that accompanied the convexity of the forehead: the orbits were medialized in a straight, coronal line, giving an unwanted appearance to the face.

We then started to maintain a fixed central axis, whenever there was bone tissue in the glabella region (11), which was not possible only in frontoethmoidal meningoencephaloceles. This central axis, in addition to providing a very anatomical nasal shape, allowed the mobilization of the orbital walls with the central reference anteriorly, thus respecting the natural curve of the frontal convexity and offering an aesthetically adequate result (Figure 7). The natural evolution of this technique was to save the orbital floor, medializing only the other 3 walls (Figure 8).
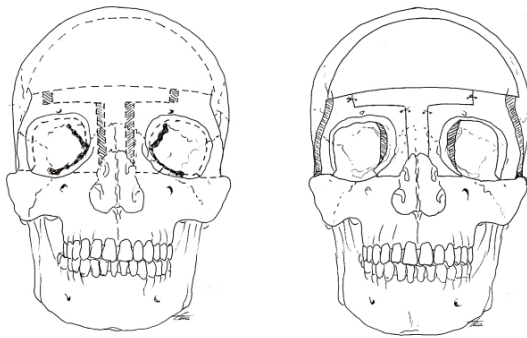

Figure 8- Modification of the T-Delta technique maintaining the nasal floor intact.

Since the first years of treatment for hyperteleorbitism, one of the many concerns with the results was the relapse during growth: the progressive aeration of the ethmoid sinuses caused the orbits to move away and the patient was already hyperteloric in adolescence. The answer to this came with Van Der Meulen's facial bipartition (12), proposing to mobilize the entire morphofunctional units, keeping the maxillary tooth germs intact. When there is no meningoencephalocele, most frontonasal dysplasias lead to midline shortening and upper arch atresia, and this bilateral crossbite directs the masticatory force vectors obliquely and laterally, tending to progressively move away the orbits (Fig 9). The rotation of the hemifaces medially displaces the vector of masticatory forces, which, when propagating in a more vertical axis, helping to maintain the result of the orbits approach during growth.

Extracranial approach techniques in Dysplasias:

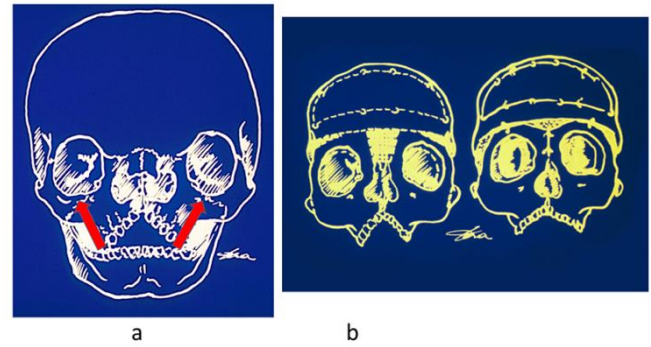In the same way as Tessier's glasses received the described modifications over time, Van Der Meulen's facial

Figure 9 - a- superior arch atresia in frontonasal dysplasia, stimulating orbital separation through the distribution of masticatory forces; b- Intracranial facial splitting technique.

bipartition technique also received improvements. The first of then was the extracranial approach, noting that, like the floor, the orbital roof also offers a horizontal sliding plane, not needing to be mobilized for the approximation of the orbits. In the absence of meningoencephalocele, a craniofacial disjunction was performed (Le Fort III) without craniotomy and the hemifaces were bipartite in the central line, where a "V" with palatal fulcrum was removed to centripetal rotate the hemifaces (Figure 10).

In patients who had bone continuity of the midline, we started to maintain an intact central bone axis (as in the modification made to Tessier's glasses), to avoid grafting and to maintain the spatial reference of rotation of the hemifaces, and not medialize them in coronal plane (Figure 11). This sum of modifications was called Double V Extracranial Facial Bipartition(13). (Figure 12).

Extended Rhinoplasty

This technique is suitable for the treatment of grade I and II hypertelorbitism, where the orbital enlargement caused by the medialization of only one orbital wall (the medial) will not be intense enough to cause enophthalmos. Whenever there is intact bone tissue in the nasal dorsum, this will be used as a central axis to accommodate the medial walls that will slide behind this axis, creating a good nasal contour. The direct Z-shaped approach in the midline of the face dispenses the coronal approach, and makes the surgery very non-invasive, in addition to allowing the elongation of the nose, which is usually short. The craniotomy holes in the epitrochlear area allow safe intraorbital osteotomies to be performed (Figure 13).

Intracranial approach techniques in craniofacial stenosis hyperteleorbitism:

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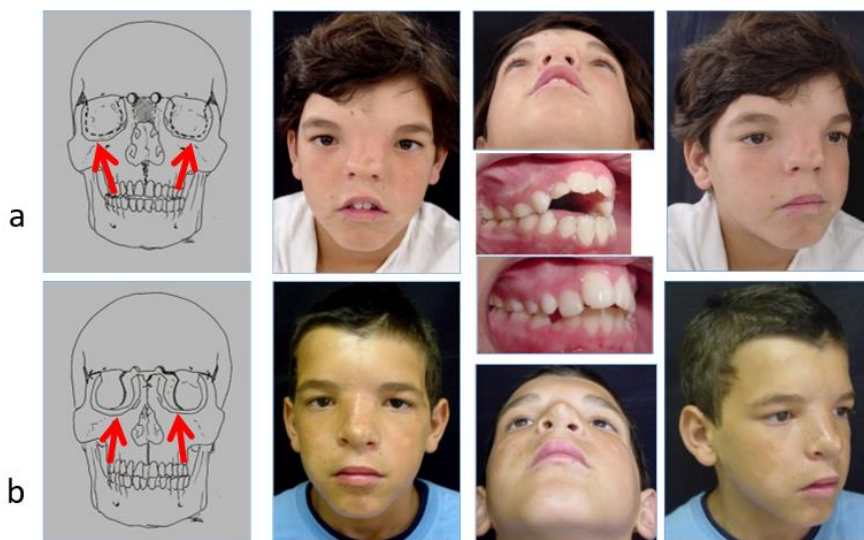**What the time has taught about Hyperteleorbitism**

Figure 10 - Extracranial Van Der Meulen facial bipartition. A- preoperative; b- postoperative.
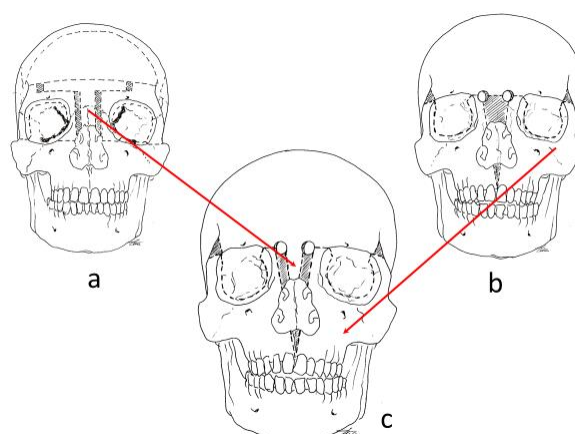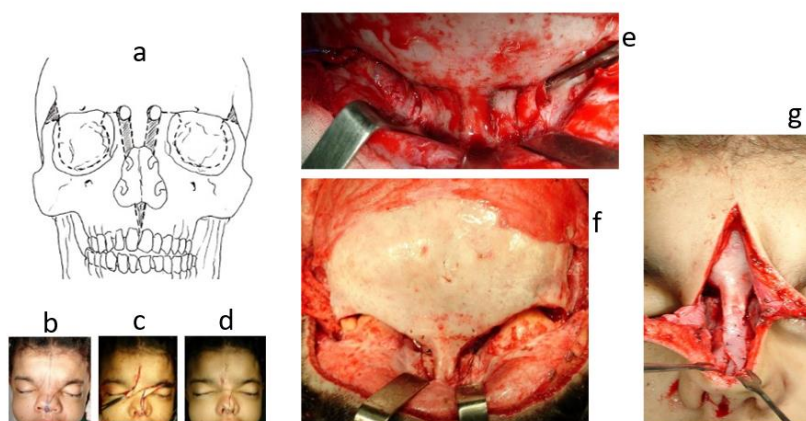

Figure 11- Taking advantage of the idea of maintaining the central axis fixed in the T-Delta technique (a) and Van Der Meulen's extracranial facial bipartition approach (b) to treat hypertelorbitism using the Double V technique.

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Figure 12 - Double V technique for the treatment of Hyperteleorbitism; a- schema of osteotomies; b- demarcation of the nasal incision in Z; c- crossing the flaps in Z; d- Z closure with nose lengthening; e- intraoperatively demarcation of the ostectomy areas laterally to the fixed central axis; f- mobilization of the medial walls of the orbits after resection of the bone bands and their fixation with steel wire; g- cartilage graft on the distal nasal dorsum, continuing to the fixed bone central axis.

Figure 13- Extracranial Extended Rhinoplasty for Grade I or II hypertelorbitism; a- demarcation of the open-rhinoplasty approach in Z-plasty, exposure of rudimentary nasal structures and crossing of the Z-shaped flaps; b- preoperative tomography and technique scheme; c- pre and postoperative.

Associated with fronto-facial advancement in monobloc

The proposal of advancing the forehead together with the middle third meets both the need for cranial expansion and maxillary advancement to expand the airways and orbits in patients with dysostosis. In the presence of hyperteleorbitism, when performing the Le Fort III craniofacial disjunction, the facial bipartition will be performed with the medial rotation of the orbits, maintaining the central bone axis intact. At the time when cranial remodelling was performed using a back table technique, transforming the frontal bone into a graft, facial splitting with advancement was performed using an intracranial approach. Based on Lauritzen's (7) proposal for the direct expansion of the osteogenic matrix through forces applied by springs on the margins of liberating osteotomies, the frontal cap was no longer removed from the surgical field to provide access to the disjunction, and the facial bipartition started to be done extracranially. Certainly the craniotomy holes in the epitrochlear and sphenoid wing areas allow the safe performance of all osteotomies, but from this modification onwards, we have greater freedom regarding the age of cranial expansions and greater stability of results.

The remodelling of the osteogenic matrix, keeping the frontal bone alive and vascularized by the inserted dura mater, has provided regularity and good contour of the forehead in the long term, contrary to what we observed in back table operated cases, where the grafted bones undergo major deformations during growth(14).

Considering that the hypertelorbitism found in craniofacial suture stenosis is not usually intense, being always between grades I and II, the preferred treatment option is Extended Rhinoplasty, associated with craniofacial disjunction with advancement of the middle third. With or without associated frontal advancement (monobloc), but always keeping the central bone axis preserved, which guarantees the perfect contour of the nasal dorsum (Figure 14). In this group of patients with dysostosis, the choice of Extended Rhinoplasty to treat hypertelorbitism is even more suitable than in the dysplasia group, as the mobilization of a single orbital wall offers a very welcome expansion in diameter to treat the exorbitism that it is always present, in a greater or lesser degree.

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CONCLUSION

The opportunity to treat hyperteleorbitism over a period as long as 44 years allowed us a huge contact with a wide range of malformations, and mainly to watch and experience the evolution of treatment proposals. The lessons that time has brought lead us to conclude that hyperteleorbitism does not fit protocols or algorithms, and each case must be studied and planned individually, taking into account its particularities.

DISCLOSURES

Ethical approval

This study was performed in line with the principles of the Declaration of Helsinki. Approval was granted by the local Ethics Committee

Consent to participate

The patient gave consent to use his information and images for research proposes.

Consent for publication

The patient gave consent to use his information and images for publication.

Conflict of interest

The authors declare no conflicts of interest with respect to the content, authorship, and/or publication of this article.

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