ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

www.surgicalneurologyint.com

Surgical Neurology International

Editor-in-Chief: Nancy E. Epstein, MD, Professor of Clinical Neurosurgery, School of Medicine, State U. of NY at Stony Brook.

SNI: General Neurosurgery

Editor
Eric Nussbaum, MD
National Brain Aneurysm and Tumor Center, Twin Cities, MN, USA

SNI
Open Access

Case Report

Isolated hypertelorism: Late surgical correction using the box osteotomy technique

Bruna Cavalcante de Sousa[1], Pedro Henrique Costa Ferreira-Pinto[1], Domênica Baroni Coelho de Oliveira Ferreira[1], Eduardo Pantoja Bastos[2], Marcio Lima Leal Arnaut Junior[2], Bruno Santos de Barros Dias[2], Thiago Schneider[2], Valéria Claro[2], Henrique Pessoa Ladvocat Cintra[2], Maud Parise[1], Eduardo Mendes Correa[1], Thaina Zanon Cruz[1], Wellerson Novaes da Silva[1], Flavio Nigri[1]

[1]Neurosurgery, Department of Surgical Specialties, Pedro Ernesto University Hospital, Universidade do Estado do Rio de Janeiro, [2]Treatment of Craniofacial Anomalies Center, Pedro Ernesto University Hospital, Universidade do Estado do Rio de Janeiro, Rio de Janeiro, Brazil.

E-mail: *Bruna Cavalcante de Sousa - brunasousa2702@gmail.com; Pedro Henrique Costa Ferreira-Pinto - pedrohcfp@gmail.com; Domênica Baroni Coelho de Oliveira Ferreira - baronidomenica@gmail.com; Eduardo Pantoja Bastos - edupantoja12@gmail.com; Marcio Lima Leal Arnaut Junior - arnautcraniofacial@gmail.com; Bruno Santos de Barros Dias - brunosbdias@yahoo.com.br; Thiago Schneider - thiagocirurgia@gmail.com; Valéria Claro - valeria_claro@hotmail.com; Henrique Pessoa Ladvocat Cintra - dr.henriquecintra@gmail.com; Maud Parise - maud.parise@gmail.com; Eduardo Mendes Correa - drcorreaneuro@gmail.com; Thaina Zanon Cruz - thaina.zanoon@hotmail.com; Wellerson Novaes da Silva - wellersonnovaess@gmail.com; Flavio Nigri - flavionigri@gmail.com

***Corresponding author:**
Bruna Cavalcante de Sousa, Neurosurgery, Department of Surgical Specialties, Pedro Ernesto University Hospital, Universidade do Estado do Rio de Janeiro, Rio de Janeiro, Brazil.
brunasousa2702@gmail.com

Received: 28 December 2023
Accepted: 03 April 2024
Published: 26 April 2024

DOI
10.25259/SNI_1029_2023

Quick Response Code:

ABSTRACT

Background: Orbital hypertelorism is a rare congenital condition caused by craniofacial malformations. It consists of complete orbital lateralization, characterized by an increase in distance (above the 95th percentile) of the inner canthal (ICD), outer canthal, and interpupillary distances. It can be approached surgically, and the main techniques are box osteotomy and facial bipartition. The surgical procedure is usually performed before the age of 8. We describe here two patients who underwent late surgical correction using the box osteotomy technique.

Case Description: Patient 1: A 13-year-old female presenting isolated hypertelorism with 5 cm ICD and left eye amblyopia. Patient 2: A 15-year-old female with orbital hypertelorism, 4.6 cm ICD, and nasal deformity. Both patients underwent orbital translocation surgery and had no neurological disorders.

Conclusion: The article reports two cases of isolated hypertelorism treated late with the box osteotomy technique. Both surgeries were successful, with no postoperative complications. It appears that it is possible to obtain good surgical results even in patients who have not been able to undergo surgery previously.

Keywords: Box osteotomy, Hypertelorism, Late correction, Orbital translocation

INTRODUCTION

Orbital hypertelorism, also known as hypertelorism, is a congenital condition first described in 1924.[6] It consists of a complete orbital lateralization [2] characterized by an increase in distance (above the 95th percentile) of three anthropometric measurements: Inner canthal (ICD), outer canthal (OCD), and interpupillary (IPD) distances.[18,19] It can be isolated or associated with other craniofacial malformations, such as meningoencephalocele, frontonasal dysplasia, and some other genetic syndromes (e.g., Apert and Crouzon).[8,17]

This congenital anomaly can be addressed surgically, and the two main techniques are box osteotomy and facial bipartition.[8,17] The maxillary width and shape define the best method.[8,9] Box osteotomy is usually indicated for a maxilla with normal shape and size, while facial bipartition is preferred for a narrow maxilla with a high palate or V-shaped arch.[9] Both surgical techniques are generally performed before the age of 8.[9,17] In this article, we describe two successful late surgical corrections by the box osteotomy technique.

CLINICAL PRESENTATION

Case 1

A 13-year-old female [Figures 1 and 2] with isolated hypertelorism and amblyopia of the left eye. On physical examination, there was an ICD of 5 cm. Cranial computed tomography (CT) scan demonstrated isolated hypertelorism [Figure 2]. An orbital translocation and a secondary palatoplasty were performed without complications. The pre and postoperative ICD and OCD [Figure 3] were obtained from imaging studies using the RadiAnt DICOM Viewer Software (Medixant, Poznan, Poland).[11]

Case 2

A 15-year-old female [Figures 4 and 5] with isolated hypertelorism and normal vision. Physical examination revealed isolated hypertelorism with an ICD of 4.6 cm and nasal deformity. Cranial CT scan demonstrated isolated hypertelorism [Figure 5]. An orbital translocation and a secondary palatoplasty were performed without complications. The pre and postoperative ICD and OCD [Figure 6] were obtained from imaging studies using the RadiAnt DICOM Viewer Software (Medixant, Poznan, Poland).[11]

Operative technique

The box osteotomy was performed in both cases. This technique can be divided into eight steps: (1) patient positioning, airway, and corneal protection; (2) incision and exposure; (3) frontal craniotomy; (4) initial facial osteotomy; (5) final osteotomy and mobilization; (6) skeletal fixation; (7) medial canthal reinforcement and nasal reconstruction; and (8) closure.[7]

The procedure begins by applying ophthalmic ointment and suturing the upper and lower eyelids horizontally to protect the patient's cornea. A zigzag-shaped coronal incision is made in the temporal region, followed by a coronal incision on the scalp below the galea. Posteriorly, a subperiosteal dissection is carried out on the supraorbital edges, and the supraorbital nerves are released. The surgeons then make an incision into the zygomatic bone and maxillary buccal, which extends from the medial side of the first molar to the contralateral side. The periorbita is released circumferentially along the roof to 1 cm anterior to the optic nerve superiorly, laterally, medially, and the lateral one-third of the floor, exposing the bilateral zygoma, the arches, the superior orbital rims, the superior surface of the nasal dorsum, the medial wall of the orbit, and the lateral third of the infraorbital rim.[7]

Once the necessary incisions are made and structures exposed, the neurosurgeons perform a frontal craniotomy by creating burr holes in the pterional and upper frontal regions [Figures 7a and b]. It is crucial to protect the midline's superior sagittal sinus, in addition to preventing bleeding

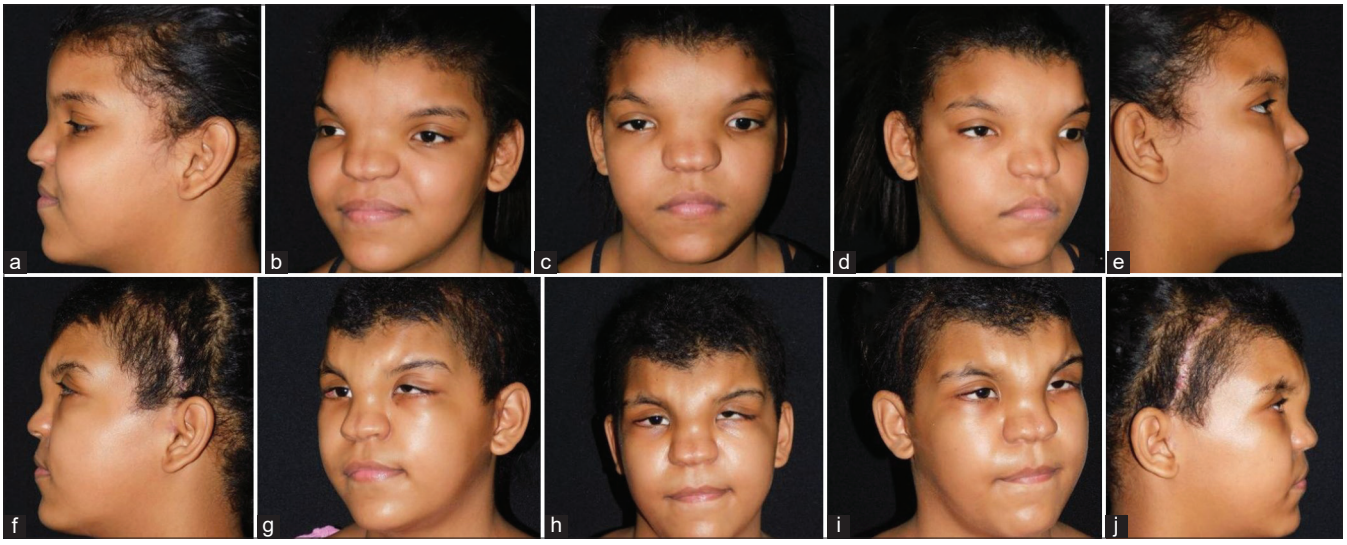

Figure 1: (a-e) Pre- and (f-j) postoperative photos of patient 1. A 13-year-old female with isolated hypertelorism and amblyopia of the left eye.

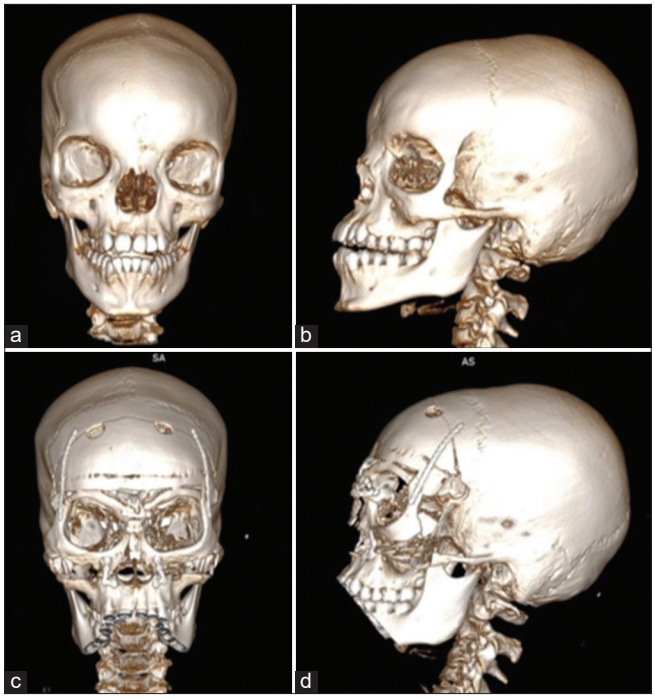

Figure 2: (a-b) Pre- and (c-d) 6-day postoperative cranial CT scan of patient 1.

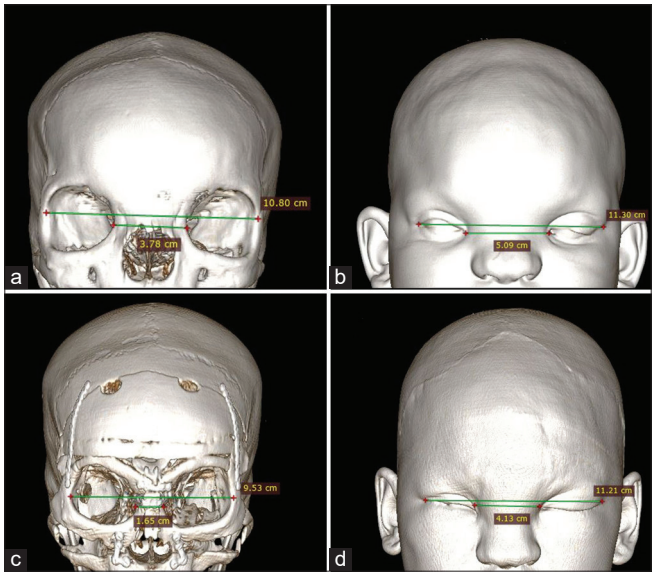

Figure 3: (a-b) Pre- and (c-d) postoperative cranial CT scan with ICD and OCD measures of patient 1. (b and d) Images were simulated by the RadiAnt DICOM Viewer Software (Medixant, Poznan, Poland) to evaluate ICD and OCD through the skin.

and cerebral venous infarction. In case of dural tears, they are repaired using 4.0 Prolene sutures. Afterwards, the frontal bone flap is removed, which provides access to the orbital roof, cribriform plates medially, and the greater wing of sphenoids laterally [Figures 7c-e]. A frontal bandeau is created, leaving approximately 1 cm between the frontal bone flap and the osteotomies of the supraorbital rim [Figure 7f]. This helps with the fixation and positioning of the orbits and frontal bone flaps. After that, the brain is retracted to expose the anterior edge of the crista galli, and osteotomies are performed transcranially on the supraorbital and the upper third of the medial orbits. Finally, the osteotomies of the medial orbital, medial two-thirds of the orbital floor, the zygoma, and the maxilla are completed.[7]

A V-shaped osteotomy is made in the midline, extending anterior to the crista galli. Depending on the desired translocation of the orbit, its shape and symmetry may vary. To move the orbits closer to each other, the frontal bone, ethmoids, midline nasal dorsum, and superior part of the bony septum are removed. The mobilization of the orbit consists of medial translocation and superior rotation of the lateral orbital rims, allowing hypertelorism correction. Fixation is done using titanium plates and screws. Since the surgery can result in excess skin, it is commonly necessary to remove some skin along with bone resection.[7] It is important to note that neurosurgeons play a crucial role in this surgery, avoiding high/bifrontal retraction and protecting the dura mater and nerve tissue from injuries.

In the past, some bone structures, such as the ethmoidal labyrinth, nasal bones, and cribriform plate, were removed during box osteotomy. However, it was discovered that this step generally impairs olfactory function due to the anatomical association between the olfactory nerve and the cribriform plate. As a result, it is no longer recommended.[8]

DISCUSSION

Embryology and physiopathology

The development of the axial skeleton occurs at the end of the 4th week by osteogenesis. The cranium ossification is divided into intramembranous and endochondral bone formation, whose common pathway is the mesenchymal cells.[12] The skullcap and viscerocranium (squamous temporal, maxillary, and zygomatic bones) are originated by intramembranous ossification, whose mesenchyme is directly transformed into bone tissue.[12,16] The skull base and the craniocervical junction are formed by endochondral ossification; in this sense, mesenchyme from neural crest and mesoderm/somites are turned into cartilage before bone tissue mineralization.[3] The anterior part of the brain is developed into frontonasal prominence (FNP) from neural crest cells (both intramembranous and endochondral ossification). The FNP contains two prominences: medial and lateral. The first one gives rise to the nasal septum, midline of the nose, philtrum, premaxilla, and four incisors, whereas the second forms the nasal turbinates and alae of the nose. Congenital errors in the signaling and migration of these cells can lead to genetic malformations, such as orbital hypertelorism.[15]

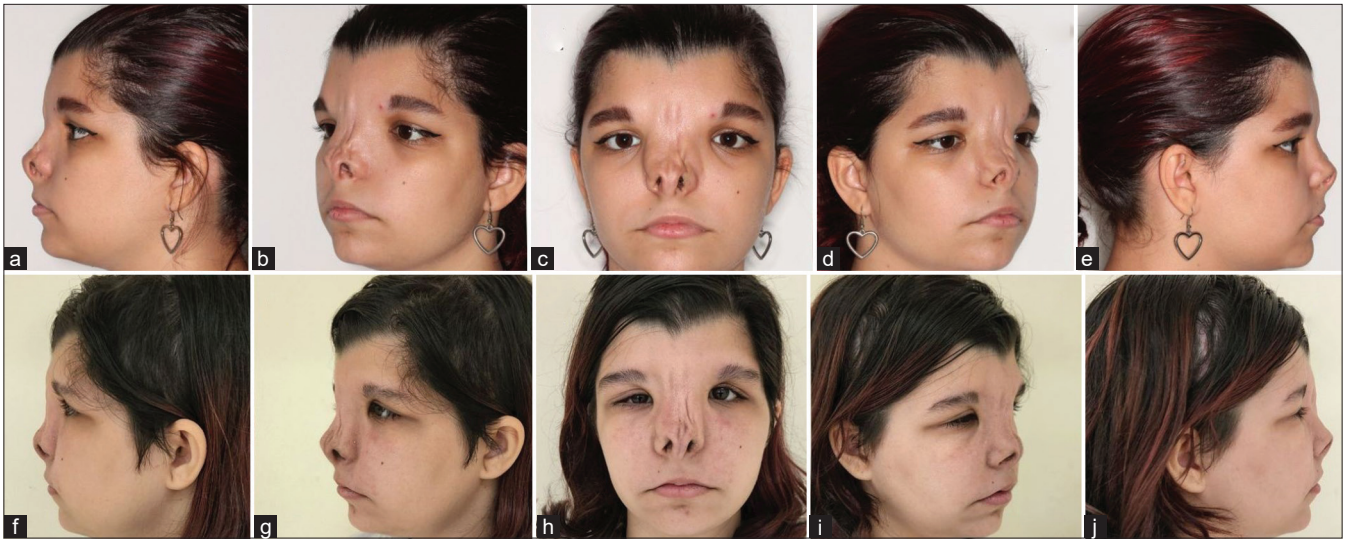

Figure 4: (a-e) Pre- and (f-j) postoperative photos of patient 2. A 15-year-old female with hypertelorism. No other health conditions were associated.

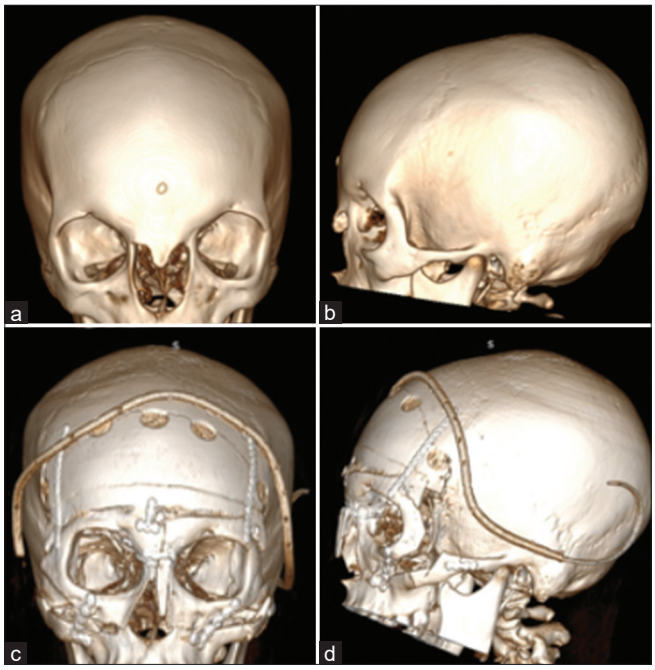

Figure 5: (a-b) Pre- and (c-d) 1-day postoperative cranial CT scan of patient 2.

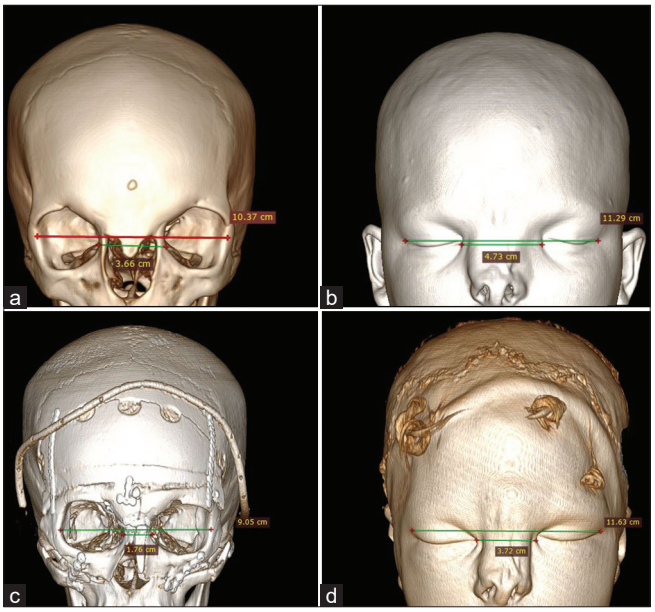

Figure 6: (a-b) Pre- and (c-d) postoperative cranial CT scan with ICD and OCD measures of patient 2. (b and d) Images were simulated by the RadiAnt DICOM Viewer Software (Medixant, Poznan, Poland) to evaluate ICD and OCD through the skin.

There is no common physiopathological pathway that explains its occurrence.[2] Some possibilities are found in the literature:[2,17] (1) fixation of the orbits in the fetal position due to early ossification of the lesser wings of the sphenoid; (2) inappropriate development of the nasal capsule with filling its usual space by the primitive brain vesicle; (3) disorders in cranial base formation; (4) action of teratogenic agents causing the death of midline mesenchyme cells in the frontonasal process and neuroepithelium; and (5) medication inducing embryonic damage to the blood vessels of the FNPs. Due to its heterogeneity, the surgical plan becomes specific to each patient.

Group classifications

Patients with hypertelorism can be classified by age group or orbital bone measurements. The first classification is divided into early (from 0 to 7 years old) and late (8 years old or older). It is used to determine the influence of age on surgical

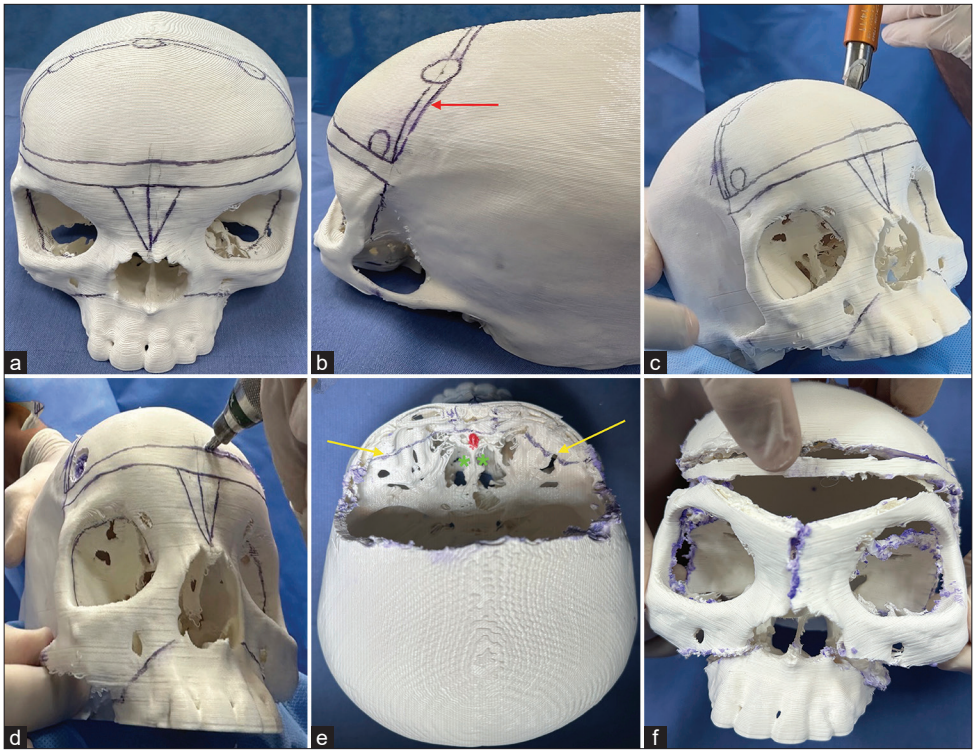

Figure 7: (a-f) The box osteotomy technique was performed on a 3D printer model – polylactic acid. A high-quality computed tomography scan of patient 1 was used as a model. (a) The 3D skull was painted with a dermatological surgical pencil. The frontal craniotomy and a frontal bandeau of 1 cm were maintained between the frontal bone flap and the orbital osteotomies. A V-shaped osteotomy was performed in the midline. The shape and symmetry vary depending on the desired orbital translocation. (b) Craniotomy burr holes were made in the pterional and upper frontal regions. The red arrow represents the trajectory of the craniotomy. (c) Using a special drill, the neurosurgeon drilled holes in the skull to expose the dura. It is important to protect the superior sagittal sinus in the midline. (d) The craniotome connected each burr hole. (e) The frontal bone flap and bandeau were removed, providing access to the orbital roof, greater wing of sphenoids laterally, and medially cribriform plates. Gentle brain retraction was necessary to avoid injury to the crista galli (red color) and cribriform plate (green asterisk) and preserve olfactory function. Yellow arrows demonstrate orbital roofs and midline osteotomies. (f) Final appearance after the box osteotomy technique, showing that both orbits were adequately translocated.

results. The ideal age for operating is widely discussed in the literature and differs between some authors. Marchac *et al.*[9] asserts that school age (around 5 years old) is appropriate, with small chances of sequelae in facial growth. In addition, good postsurgical results have been evidenced, especially in adults who underwent the procedure in childhood. Sharma[17] states that the surgery should be performed after 5–7 years old due to the lower chances of maxillary growth disorders. In adults, the procedure is safer, more stable, and simpler compared to children. Raposo-Amaral *et al.*,[14] in agreement, affirm that the surgery should not be performed before 8 years old due to the greater chance of recurrence compared to older patients – who already have a mature facial skeleton. Some authors agree that hypertelorism must be corrected before school age to avoid psychological and body image problems.[9,17]

Raposo-Amaral *et al.*[14] presented 10 craniofrontonasal dysplasia cases with hypertelorism and late presentation (13.4 ± 7.68 years old) that underwent surgical correction. All patients did not have intracranial hypertension and early surgical intervention during childhood. Postsurgical complications were limited to infection (two patients), cerebrospinal fluid (CSF) leak (one patient), severe edema (one patient), and bone necrosis (one patient), and the patients had good outcomes. The authors agreed that delaying the correction of hypertelorism can guarantee better results, such as definitive correction and avoiding neurosurgical reapproach.

We consider that the best period for surgical correction is before the age of 8. The early correction can improve or prevent the progression of serious neurological disorders

(e.g., amblyopia) and promote better esthetic and psychological outcomes. However, some patients cannot be evaluated in the correct period, and early surgery is not an option. This occurs for several reasons, such as parental knowledge, demographic barriers, and administrative problems in public health (e.g., lack of reference centers/professionals resulting in long wait times for appointments). For them, late surgery can be a reasonable option with good results.

The second classification includes the ICD, measured between the anterior lacrimal crests. It can be classified as mild (30 mm < bony interorbital distance ≤34 mm), moderate (34 mm < bony interorbital distance ≤40 mm), and severe (bony interorbital distance >40 mm).[19] Not only does it help classify the severity, but it also allows surgeons to visualize future outcomes.[14] Our patients were classified as severe presentation because their ICDs were 50 mm and 46 mm.

Surgical techniques and complications

There are several techniques described in the literature: orbital box osteotomy, monobloc frontofacial advancement by distraction, frontofacial bipartition advancement by distraction, and facial bipartition.[1,4,5] Nonetheless, the most common are box osteotomy and facial bipartition.[8,9,13,14,17]

Some authors[9,14,17] consider facial bipartition a good option for mixed dentition and/or dental arch deformities due to the shape of the maxilla and angulation of the palate. Otherwise, box osteotomy is better for permanent dental arch to avoid the tooth buds,[9] as well as centralizing the orbits and correcting the vertical orbital dystopia.[17] Considering that our patients presented isolated hypertelorism and no other significant bone malformation, box osteotomy was the technique performed.

The postoperative complications include infections, CSF leakage, hyposmia or anosmia, oculomotor disorders, epiphora, enophthalmos, obstruction of nasal passages,[13] and, less commonly, death.[9] The gustatory function can also be affected during the hypertelorism correction, causing dysgeusia, mainly due to possible impairment of the olfactory nerve. This taste perception is typically associated with hyposmia or anosmia due to its close relationship with the ability to smell.[10] In our patients, we preserved the olfactory nerve, conserving the crista galli area.

Some soft-tissue deformities are generally expected after correction of hypertelorism, such as bifid nose, eyelid ptosis, epicanthal fold, deviated septum, and excess skin in the midline, which can be corrected later by additional surgeries.[14] It is also worth mentioning that the medial canthus fold can create a false impression of residual hypertelorism, even if the orbital correction was successful, as described by Tessier.[14,19] Our patients are still waiting for additional necessary soft-tissue procedures.

CONCLUSION

Isolated hypertelorism is a genetic condition that must be corrected to avoid neurological deficits and psychological disturbance. Although some authors suggest better surgical results before 8 years old, our present study describes two late corrections with good outcomes.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Fundação de Amparo à Pesquisa do Estado do Rio de Janeiro (FAPERJ) and Center of High Complexity Neurosurgery Intern Patients (NIPNAC).

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation:

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

REFERENCES

1. Britto JA, Greig A, Abela C, Hearst D, Dunaway DJ, Evans RD. Frontofacial surgery in children and adolescents: Techniques, indications, outcomes. Semin Plast Surg 2014;28:121-9.
2. Cohen MM Jr., Richieri-Costa A, Guion-Almeida ML, Saavedra D. Hypertelorism: Interorbital growth, measurements, and pathogenetic considerations. Int J Oral Maxillofac Surg 1995;24:387-95.
3. Di Ieva A, Bruner E, Haider T, Rodella LF, Lee JM, Cusimano MD, *et al*. Skull base embryology: A multidisciplinary review. Childs Nerv Syst 2014;30:991-1000.
4. Ferreira Junior TA, Fontoura RR, Marques do Nascimento L, Alcântara MT, Capuchinho-Júnior GA, Alonso N, *et al*. Frontofacial monobloc advancement with internal distraction: Surgical technique and osteotomy guide. Oper Neurosurg (Hagerstown) 2022;23:e33-41.
5. Greig AV, Britto JA, Abela C, Witherow H, Richards R, Evans RD, *et al*. Correcting the typical Apert face: Combining bipartition with monobloc distraction. Plast Reconstr Surg 2013;131:219e-30.
6. Greig DM. Hypertelorism: A Hitherto undifferentiated

congenital cranio-facial deformity. Edinb Med J 1924;31: 560-93.
7. Kademani D, Tiwana P. Atlas of oral and maxillofacial surgery. 1st ed. Netherlands: Elsevier; 2016. p. 487-97.
8. Laure B, Batut C, Benouhagrem A, Joly A, Travers N, Listrat A, *et al*. Addressing hypertelorism: Indications and techniques. Neurochirurgie 2019;65:286-94.
9. Marchac D, Sati S, Renier D, Deschamps-Braly J, Marchac A. Hypertelorism correction: What happens with growth? Evaluation of a series of 95 surgical cases. Plast Reconstr Surg 2012;129:713-27.
10. McCarthy J. Formal problems in Semitic phonology and morphology. Unpublished Ph.D. Dissertation, Cambridge, MA: MIT Press; 1979.
11. Medixant. RadiAnt DICOM viewer. Version 2021; 2021. Available from: https://www.radiantviewer.com [Last accessed on 2023 Dec 21].
12. Moore KL, Persaud TV, Torchia MG. The developing human clinically oriented embryology. 10th ed. Netherlands: Elsevier; 2015.
13. Patel SY, Ghali GE. Orbital hypertelorism. Atlas Oral Maxillofac Surg Clin North Am 2022;30:101-12.
14. Raposo-Amaral CE, Resende G, Denadai R, Ghizoni E, Raposo-Amaral CA. Craniofrontonasal dysplasia: Hypertelorism correction in late presenting patients. Childs Nerv Syst 2021;37:2873-8.
15. Roth DM, Bayona F, Baddam P, Graf D. Craniofacial development: Neural crest in molecular embryology. Head Neck Pathol 2021;15:1-15.
16. Setiawati R, Rahardjo P. Bone development and growth. In: Osteogenesis and bone regeneration. London: IntechOpen; 2019.
17. Sharma RK. Hypertelorism. Indian J Plast Surg 2014;47: 284-92.
18. Sirkek B, Sood G. Hypertelorism. In: StatPearls. Treasure Island, FL: StatPearls Publishing; 2023. Available from: https://www.ncbi.nlm.nih.gov/books/NBK560705 [Last accessed on 2023 Jul 24].
19. Tessier P. Experiences in the treatment of orbital hypertelorism. Plast Reconstr Surg 1974;53:1-18.

How to cite this article: Sousa BC, Ferreira-Pinto PHC, Ferreira DBCO, Bastos EP, Arnaut Junior MLL, Dias BSB, *et al*. Isolated Hypertelorism: Late surgical correction using the box osteotomy technique. Surg Neurol Int. 2024;15:145. doi: 10.25259/SNI_1029_2023