OPEN ACCESS
Saudi Journal of Medicine
Abbreviated Key Title: Saudi J Med
ISSN 2518-3389 (Print) |ISSN 2518-3397 (Online)
Scholars Middle East Publishers, Dubai, United Arab Emirates
Journal homepage: https://saudijournals.com/journal/sjm/home

Review Article

Hypertelorism

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| **Received:** 25.04.2019 | **Accepted:** 04.05.2019 | **Published:** 11.05.2019
DOI: 10.36348/sjm.2019.v04i05.001

Abstract

The term orbital hypertelorism denotes “widely apart orbits.” This may also be associated with the abnormal orientation of the orbits in terms of the vertical position (dystopia). This deformity may be seen unilaterally or bilaterally. They may present as symmetric or asymmetric and can manifest in a variety of craniofacial conditions. The treatment is primarily carried out for aesthetic reasons. This paper emphasizes on the current understanding of this condition.
Keywords: hypertelorism, widely apart orbits.

INTRODUCTION

Greg in 1924 coined the term “ocular hypertelorism” to signify widely placed eyes [1]. He considered the interpupillary distance (IPD) as a means to record its presence. Tessier emphasized that IPD could be increased because of exophoria alone without any bony abnormality of the orbit. Hence, he proposed the term “orbital hypertelorism” (ORH), wherein there is true lateralisation of the whole of the bony orbit in a way that the distance between the lateral canthus and auditory meatus is shortened [2].

Orbital hypertelorism is not a disease in itself but the manifestation of a craniofacial deformity that may be present in a variety of craniofacial conditions. The orbits are placed wide apart resulting in an abnormal appearance with ‘broad nose’. It may be present in conditions such as a craniofacial dysplasia, encephaloceles and craniosynostosis syndromes. The increased inner intercanthal distance (ICD) can also occur following trauma or tumours in the naso-orbital region; however there is no change in the position of the lateral wall of the orbit. This ‘pseudo-hypertelorism’ is referred to as ‘telecanthus’ [3].

Orbital hypertelorism can be recognized at about 28 mm embryo stage when a defective development of the nasal capsule leads to freezing of the future fronto-naso-orbito-ethmoid complex thereby preventing the movement of the orbit towards midline [4]. Another suggested possibility could be a deficient latero-medial movement of the orbit [5]. Some studies show that early ossification of the lesser wings of sphemoids leads to arrest of orbits in the foetal position. If the frontonasal prominence remains in its embryonic position, the optic placodes cannot move towards the midline resulting in orbital hypertelorism [6].

Management

The main purpose of surgical intervention in hypertelorism is to bring the two orbits closer together, correct any orbital dystopia, narrow the nasal dorsum and create a normal nose with adequate projection and correct any soft tissue blemishes such as excess skin over the nose, and any nasal clefts or displaced eyebrows [3].

In order to provide a better social well-being to the patient it is always proposed to operate as early in life as possible. However, it is not advocated to operate in children who are less than 5 years of age as the osteotomy lines during the surgical intervention go through the inferior orbital rim, and hence it is likely to injure the un-erupted tooth roots with resultant maxillary growth disturbances. In addition, the bone stock may not be good for holding the osteotomies together with fixation, thereby predisposing to relapse of the deformity. Hence, the literature suggests delaying

the correction of orbital hypertelorism to about 5-7 years of age. The results of surgery performed in adults are more reliable, stable, and the operation is technically much simpler than in a child [3].

The corrective surgery is indicated primarily for cosmetic reasons. Paul Tessier in late 1960's developed a combined intra-cranial and extra-cranial technique for the correction of the deformity. He demonstrated that it was possible to move the orbits in three dimensions without adversely affecting the vision. He suggested that this was possible because of the inherent laxity of the optic nerve in the socket. He also demonstrated that large areas of the craniofacial skeleton can be devascularised and these would still survive completely provided healthy lining and cover is present [7, 8].

The techniques of orbital hypertelorism correction have been further refined by Converse van der Meulen *et al.,* Ortiz-Monasterio *et al.,* and Marchac *et al*., [9-12]. They advocated visualizing the bony orbit into two parts — the outer square shaped box containing the globe and the inner cone shaped part housing the optic nerve. If these parts can be separated, the outer box can be moved in three dimensions without adversely affecting the vision.

Frontofacial surgery includes those operative techniques that separate the intact orbits from the anterior skull base to move the orbits in relation to each other or as a combined unit with the maxilla in relation to the basicranium. The orbits may be translocated independently (orbital "box" osteotomy), or advanced as a "monobloc" with the maxilla and upper dental arch [13].

Box Osteotomy

The classic orbital box osteotomy involves an en-bloc movement of the orbits medially into the space created by resection of abnormally wide nasal and ethmoidal bones. The procedure is best performed by a combined intracranial and extracranial approach [3]. Orbital box osteotomy procedure is generally chosen whenever the dental occlusion is normal.

The Box Osteotomy Movement of the orbits (without the maxilla) as orbital box osteotomies is commonly undertaken for medial and/or vertical translocation of the orbit, behind the equator of the globe, so the consequent globe, canthal, and palpebra movements result in greater midfacial symmetry. The orbits are thus moved in relation to each other and separately from the maxilla [13]. Since all the walls of the orbit are moving as one single unit, the orbital volume remains unchanged. Tessier advocated keeping a supraorbital bar to stabilize and guide the mobilized orbits [7].

Orbital box osteotomies are valuable for the correction of symmetric or asymmetric hypertelorism in a variety of craniofacial syndromes, for the correction of vertical orbital dystopia, or in conjunction with segmental cuts for orbital expansion in micro-orbitism / microphthalmos syndromes [13]. Use of cranial bone grafts after relocation of the orbits adds to stability of the results achieved on the operating Table.

Facial Bipartition

It is the surgery of choice in clinical situations where the dental occlusion is angulated as in cases of Apert's syndrome or some craniofacial clefts. It involves medialisation of the two hemi faces. A medial resection of the nasal dorsum is performed in a "V" shaped form, and the palatal bone is split in the midline. The lateral cuts go through the zygomatic arch and pterygomaxillary junction. The ensuing rotation allows correction of the narrow maxillary arch and also widens the nasal fossae and improves breathing. It would also result in a change of the orbital axis. This procedure is very useful for severe deformities. Facial bipartition would also increase the orbital volume and may improve the proptotic eyes seen in many of these cases [3].

In facial bipartition cases, no frontal bar is preserved; however in box osteotomy the frontal bar is preserved to guide the medial movement of the orbit. This prevents any inadvertent rotational deformity. In facial bipartition since facial rotation is needed, the frontal bar is not kept in order to permit unhindered facial rotation.

CANTHOPEXY

The medial canthi need to be identified and anchored. These can be fixed using trans-nasal wires that fix both the canthi. A two-hole titanium plate is used for this purpose. The plate is secured to the thick nasal bone, and the lower hole is kept at the level of lacrimal crest. The ipsilateral canthus is securely fixed this hole using steel wires [14].

The removal of ethmoids and the nasal septum exposes the extradural space to the nasal cavity and can be a source of ascending infection. A vascularized flap such as fronto-galeal flap as described by Jackson *et al.* is interposed between the two cavities to prevent infection [15]. Majority of these patients will need augmentation of the dorsum and this is best accomplished by using split cranial bone graft as a cantilever.

Correction of hypertelorism would necessitate a combined intra-cranial and extra-cranial approach. The ethmoid sinuses are opened up and there is a danger of ascending infection from the nasal passages. The use of fronto-galeal flap can effectively seal this passage and prevent infection [15]. A seriously threatening complication could be of vision loss.

However, with a careful approach this complication can be avoided.

Hypertelorism can occur as isolated conditions, or may be associated with facial clefting, encephalocoele, sinonasal pathology, or trauma [16]. Conditions in which there is underdevelopment of nasal projection may give an appearance or illusion of hypertelorism like in Larsen syndrome and Binder syndrome [17].

In such cases, nasal reconstruction giving adequate nasal projection will correct the apparent deformity, and may give a perceptual improvement in cases of mild hypertelorism where major surgery is not indicated. However, for most significant orbital malformations, some orbital osteotomy will be required [18, 19]. Numerous orbital osteotomies have been described, which may involve movement of one, two, three, or four orbital walls, either unilaterally or bilaterally depending on the severity and nature of the deformity [20, 21].

Of great significance in orbital osteotomies is the management of the medial canthal ligament. It is better to leave it attached to its bony base and great care must be taken in subperiosteal dissection to avoid encroaching on the area of the medical canthus. If detachment of the medial canthus is unavoidable or occurs inadvertently, then careful transnasal medial canthopexy is necessary, with bone grafting required in order to re-position the canthus and give it a secure attachment. However, results of medial canthopexy can be disappointing, and is best avoided by maintaining the attachment of the medial canthal ligament intact. Bone grafting is always required in orbital osteotomies to fill bony gaps left by the orbital movements and to ensure postoperative stability. Subsequent secondary procedures are sometimes required for the management of associated facial deformity or postoperative ocular problems [16, 17].

CONCLUSION

Orbital hypertelorism can occur in a variety of situations such as craniofacial dysplasia, craniofacial clefts and some craniosynostosis syndromes. It may or may not be associated with narrowing of the maxillary arch and dental malocclusions. The indication for surgery is for cosmetic reasons and to establish asocial well-being to the patient. The correction can be performed using a combined intracranial and extracranial approach. The orbits can be mobilised into the medial position after removing part of the enlarged nasal and ethmoid bones by using either ‘box osteotomy’ or ‘facial bipartition’ technique. The morbidity and mortality in these cases is negligible if the orbital hypertelorism correction is approached in a systematic manner following proper surgical protocols.

REFERENCES

1. Greig, D. M. (1924). Hypertelorism: A hitherto undifferentiated congenital cranio-facial deformity. *Edinburgh Medical Journal*, *31*(10), 560.
2. Tessier, P. (1972). Orbital hypertelorism: I. Successive surgical attempts. Material and methods. Causes and mechanisms. *Scandinavian journal of plastic and reconstructive surgery*, *6*(2), 135-155.
3. Sharma, R. K. (2014). Hypertelorism. *Indian journal of plastic surgery: official publication of the Association of Plastic Surgeons of India*, *47*(3), 284-292.
4. Vermeij-Keers, C., Mazzola, R. F., der Meulen Van, J. C., & Strickler, M. (1983). Cerebro-craniofacial and craniofacial malformations: an embryological analysis. *The Cleft palate journal*, *20*(2), 128-145.
5. Poelmann, R. E., & Vermeij-Keers, C. (1976). Cell degeneration in the mouse embryo: a prerequisite for normal development. *Progress in Differentiation Research*, 93-102.
6. Patten, B. M. (1968). 3rd ed. New York: McGraw Hill; Human Embryology.
7. Tessier, P., Guiot, G., Rougerie, J., Delbet, J. P., & Pastoriza, J. (1967, June). Cranio-naso-orbito-facial osteotomies. Hypertelorism. In *Annales de chirurgie plastique*, 12(2), 103-118.
8. Tessier, P. (1967, December). Total facial osteotomy. Crouzon's syndrome, Apert's syndrome: oxycephaly, scaphocephaly, turricephaly. In *Annales de chirurgie plastique*, 12, (4), 273-286.
9. Converse, J. M., Ransohoff, J., Mathews, E. S., Smith, B., & Molenaar, A. (1970). Ocular hypertelorism and pseudohypertelorism advances in surgical treatment. *Plastic and reconstructive surgery*, *45*(1), 1-13.
10. Van der Meulen, J. C. (1979). Medial faciotomy. *British journal of plastic surgery*, *32*(4), 339-342.
11. Ortiz-Monasterio, F., & Carrillo, A. (1978). Advancement of the orbits and the midface in one piece, combined with frontal repositioning, for the correction of Crouzon's deformities. *Plastic and reconstructive surgery*, *61*(4), 507-516.
12. Marchac, D., Renier, D., & Arnaud, E. (1997). Infrafrontal correction of teleorbitism. In *Proceedings from the Seventh Meeting of the ISCFS. Santa Fe: Menduzzi*, 173-175.
13. Britto, J. A., Greig, A., Abela, C., Hearst, D., Dunaway, D. J., & Evans, R. D. (2014, August). Frontofacial surgery in children and adolescents: techniques, indications, outcomes. In *Seminars in plastic surgery* (Vol. 28, No. 03, pp. 121-129). Thieme Medical Publishers.
14. Sharma, R. K., Makkar, S. S., & Nanda, V. (2005). Simple innovation for medial canthal tendon fixation. *Plastic and reconstructive surgery*, *116*(7), 2046-2048.

15. Jackson, I. T., Adham, M. N., & Marsh, W. R. (1986). Use of the galeal frontalis myofascial flap in craniofacial surgery. *Plastic and reconstructive surgery*, *77*(6), 905-910.
16. Lin, K. Y., Ogle, R. C., & Jane, J. A. (2002). *Craniofacial surgery: science and surgical technique*. WB Saunders Company, 332-344.
17. Richardson, D., & Thiruchelvam, J. K. (2006). Craniofacial surgery for orbital malformations. *Eye*, *20*(10), 1224-1227.
18. Tessier, P., Guiot, G., & Derome, P. (1973). Orbital hypertelorism: II. Definite treatment of orbital hypertelorism (OR. H.) by craniofacial or by extracranial osteotomies. *Scandinavian journal of plastic and reconstructive surgery*, *7*(1), 39-58.
19. Ortiz-Monasterio, F., & Molina, F. (1994). Orbital hypertelorism. *Clinics in plastic surgery*, *21*(4), 599-612.
20. Persing, J. A., Jane, J. A., Park, T. S., Edgerton, M. T., & Delashaw, J. B. (1990). Floating C-shaped orbital osteotomy for orbital rim advancement in craniosynostosis: preliminary report. *Journal of neurosurgery*, *72*(1), 22-26.
21. Psillakis, J. M., Zanini, S. A., Godoy, R., & Cardim, V. L. N. (1981). Orbital hypertelorism: modification of the craniofacial osteotomy line. *Journal of maxillofacial surgery*, *9*(1): 10-14.