ORBITAL HYPERTELORISM COMPARATIVE STUDY OF SURGICAL RECONSTRUCTIVE PROCEDURES

Thesis

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بِسْمِ اللَّهِ الرَّحْمَنِ الرَّحِيمِ

قَالُوا سبحانك لَا علْم لَنا إِلا ما علمتنا إِنك أَنت الْعليم الْحكيم

صدق الله العظيم

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ABSTRACT

Congenital hypertelorism is part of a complex malformation produced by incomplete medial migration of the lateral elements of the upper and the midface. *Orbital hypertelorism* signifies an increased distance between both medial sides and lateral sides of the orbits. According to the interorbital distance: first degree, 30 to 34 mm; second degree, greater than 34 mm with normal shape and orientation of the orbits; third degree, greater than 40 mm. Bony measurements on plain radiographs or computed tomographic (CT) scans accurately document bony interorbital distance (BIOD) and correlate with direct intraoperative measurements. In the present study 10 patients are grouped according to the BIOD and operated upon by two techniques, the combined intra-extra cranial approach with total orbital mobilization and the intracranial approach with hemiorbital mobilization. Hemiorbital mobilization proved to be effective in moderate to severe orbital hypertelorism. In severe cases with exaggerated cranial width total orbital mobilization is preferred to correct the facial deformity. Both techniques proved to be safe and effective with stable results. The Hemiorbital mobilization proved to be effective in moderate to severe cases with less operative time and without any major complications except minor infection in one case.

Key words: orbital hypertelorism, bony interorbital distance, frontonasal dysplasia, intracranial approach, subcranial approach, Hemiorbital mobilization, orbital osteotomies.

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INTRODUCTION

"Ocular" hypertelorism was a term employed by ***Greig (1924)*** in his classic paper to describe two cases of congenital facial deformity with a "great breadth between the eyes." The term has been confusing, however, for a variety of reasons ***(McCarthy et al., 1990a).*** Orbital hypertelorism is defined as a lateralization of the total orbit. This condition is associated almost always with congenital abnormalities ***(Zihni et al., 2007)***.

Congenital hypertelorism is part of a complex malformation produced by incomplete medial migration of the lateral elements of the upper and the midface. The shape of the orbits may be altered as well as their position relative to the sagittal plane. The width of the eithmoid is increased, and the nose is broad and short; medial and paramedial clefts are frequently observed. These may vary from a bifid tip to a complete separation of the two halves or even absence of the nose. The vertical dimension of the face is always decreased, and anterior open bite is common. These features differentiate this malformation from the frontonasal meningoencephalocele, which was previously described as the "long noses" **(Ortiz Monasterio et al., 1990).**

Interpupillary distance can be misleadingly increased by the exotropia, which is so common in these patients. Furthermore, an illusion of hypertelorism results when there is a lateral displacement of the medial canthi (telecanthus), as occurs in a variety of deformities, such as

Waardenburg's syndrome, canthus inversus, or blepharophimosis. Patients with these deformities also show epicanthal folds and overlapping of the caruncles and the medial portion of the sclera. The patient appears hyperteloric because of the medial canthal soft tissue deformities, and yet the radiographic interorbital distance is normal. Orbital hypertelorism is, therefore, the preferred term and can be simply defined as an abnormally wide distance between the orbits ***(McCarthy et al., 1990a).***

Craniofacial surgery consists of techniques involving the cranial as well as the facial skeletal structures. It includes those surgical procedures employed to correct complex congenital and acquired deformities involving the cranium, orbits, facial bones, and jaws. Surgical osteotomies and advancement or recession of the bones are accomplished either through a combined intra-extracranial approach or through a subcranial (extracranial) approach alone. The definitive surgical correction of craniofacial deformities initiated by ***Tessier and associates (1967)*** is one of the major advances in modern reconstructive plastic surgery.

The goal of surgical correction of ocular hypertelorism is to normalize the appearance of the face by reducing the abnormally large interocular distance. Tessier's revolutionary surgical correction of orbital hypertelorism, by means of four-wall intra/extracranial osteotomies of the ventral two-thirds of the orbit followed by medial, and at times ventral with or without cephalad/caudad translocation, presumes that the orbital contents move proportionately to the movement of the osseous orbit.

Numerous technical modifications of ***Tessier's (1972)*** "useful orbit" operation have been published subsequently ***(Panchal et al., 1999).***

The combined preoperative accurate soft tissue and bone planning is primordial for good aesthetic results ***(Lin & Ogle, 2002).***

Aim of the Work:

As a prospective study, the early functional and aesthetic results by using different surgical techniques employed in correction of orbital hypertelorism will be compared.

Definition of hypertelorism (What is Hypertelorism?)

Hypertelorism refers to the widened distance between bilateral structures. *Orbital hypertelorism* means lateralization of the entire orbital structure. Hypertelorbitism is a synonym preferred by ***Dr. Paul Tessier (1972)*** to describe this condition, which presents within the context of many congenital craniofacial malformations.

Hypertelorism is not a syndrome but a physical finding in many craniofacial anomalies. It is *etiologically heterogeneous* (i.e., there are different causes) and *pathogenetically heterogeneous* (i.e., different etiologies can result in similar features). Descriptive classifications must suffice until there are precise molecular explanations for the various hyperteloric phenotypes. Anatomic classifications are based on physical characteristics, whereas morphogenic classifications consider embryology and suggest pathogenesis. The diagnostic categories used in this analysis integrated both anatomic and morphogenic systems of classification.

The word *hypertelorism* is used to describe increased interorbital distance, a condition that is causally and pathogenically heterogeneous. Because *not all wide-set eyes are the same*, accurate terminology and nosology are critical to understanding and management. *Orbital hypertelorism* signifies an increased distance between both medial sides and lateral sides of the orbits. *Interorbital hypertelorism* denotes increased distance only between the inner orbital walls ***(Tan & Mulliken, 1997).***

Hypertelorism has come to denote excessively wide spacing of the eyes in the craniofacial anomaly literature over the past 3 decades. The Greek roots, from which hypertelorism derives, literally mean "excessive apartness" and can be applied to any bilateral structure. Thus, an abnormally increased distance between the eyes is properly referred to as ocular hypertelorism. That term, ocular hypertelorism, was coined by ***Greig in 1924*** to describe his postmortem findings on the skull of Mary McDougal, whose in vivo craniofacial dysmorphology had previously been noted to resemble that of a fetus by ***Thompson***. Ocular hypertelorism remained the preferred term for description of widely spaced eyes until the mid-20th century when ***Tessier (1972)*** began to study intensively craniofacial anomalies and devise novel operations for their reconstruction ***(Panchal et al., 1999).***

Correction of hypertelorbitism is frequently only one aspect of a complex, multistage surgical treatment strategy. Until surgical techniques were developed in the 1960s, patients who suffered from severe physical abnormalities encountered a profound handicap in their ability to achieve normal psychosocial development, regardless of their intellectual or physical condition. Now, improvements in treatment allow for a more normal appearance, even in extreme cases. These advances allow for an earlier acceptance into society, especially for children who might otherwise be shunned by family and community members.

The precise means by which the condition is measured and defined distinguish hypertelorbitism from *telecanthus* or *pseudohypertelorbitism.* The latter terms are used when the distance between the medial canthi is

increased. Because the soft tissue distance between the globes or interpupillary distance is variable, hypertelorbitism must be assessed by bony landmarks, including the interorbital and outerorbital distance. The interorbital distance was originally defined specifically as that between the two dacryons on a radiograph taken in the direct posteroanterior projection. The distance between the lateral orbital rims at the level of the dacryons constitutes the outerorbital distance. Hypertelorbitism is strictly defined as both measurements being more than 2 standard deviations (SD) from the normal mean.

In adults, ***Tessier (1972)*** classified hypertelorbitism into three degrees of severity based on the interorbital distance (IOD): first degree—30- to 34-mm IOD; second degree—34- to 40-mm IOD; and third degree—greater than 40-mm IOD. ***Van der Meulen and colleagues*** distinguished the term *interorbital* hypertelorism as interorbital distance more than 2 SD from the normal mean, matched for age and gender. Others report strictly on the basis of deviation from the mean to accommodate for normal variation. Within this classification, first degree = 2 to 4 SD from the mean; second degree = 4.1 to 8 SD from the mean; and third degree = more than 8 SD from the mean. Reporting 90 cases, ***Tan and Mulliken (1997)*** distinctly differentiate between hypertelorbitism and interhypertelorbitism for the purpose of describing and categorizing the many conditions with which hypertelorbitism is associated.

Because the "normal" position of the orbits is difficult to pinpoint relative to the course of craniofacial growth and development, the