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Case Report

Surgical Technique for Severe Microphthalmic Orbit Reconstruction: Case Report

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Abstract

Purpose: To report a case of severe bilateral microphthalmos treated with a unilateral orbital reconstruction procedure.

Case Report: This is the case report of a 4-year old patient with bilateral microphthalmos who showed upper face underdevelopment with left palpebral and orbital structures more affected. She underwent a unilateral left orbital reconstruction procedure to achieve proper volume for an 18mm bio ceramic implant and achieved optimal postoperative cosmetic results.

Conclusion: This case shows that orbital osteotomies are indicated to provide upper face symmetry and allow the fitting of an orbital implant big enough to stimulate orbital development not needing to be replaced over time and providing optimal postoperative cosmetic results.

Keywords: Microphthalmia; Microphthalmic Orbit; Orbit Reconstruction; Orbital Osteotomy

Abbreviations

CT: Computed Tomography; MRI: Magnetic Resonance Imaging

Introduction

Microphthalmia indicates a small eye within the orbit due to poor development or regression of the optic vesicle. Severe microphthalmia can be difficult to distinguish from clinical anophthalmia in routine ophthalmic practice. Clinically it seems reasonable to use the term microphthalmia for an eye with axial length two standard deviations below the age-adjusted mean; this typically correlates to a corneal diameter less than 10 mm, and an axial length less than 20 mm for adult eyes [1].

The prevalence of anophthalmia/microphthalmia has been estimated at 0.2 - 3.0 per 10,000 births [2-4]. It is reported in 3.2 - 11.2% of blind children. Both, anophthalmia and microphthalmia may occur in isolation or as part of a syndrome, in one-third of cases. These conditions have complex etiology with chromosomal, monogenic and environmental causes identified. The diagnosis is usually based upon clinical findings and imaging.

Once a diagnosis has been established, both ocular and systemic imaging tests (ultrasound, computed tomography [CT], and magnetic resonance imaging [MRI]) should be performed to rule out neurological, renal, cardiac, or other associations [2,4]. Because of the wide phenotypic spectrum of the condition, it is vital to assess these patients with multi-disciplinary teams.

Microphthalmia can be classified as mild, moderate or severe according to clinical and biometrical characteristics [5]. Treatment is directed towards maximizing existing vision and improving cosmesis through simultaneous stimulation of both soft tissue and bony orbital growth. In severe cases, treatment is usually started early to maximize the overall development of these children. The aim is to enlarge the palpebral fissure, conjunctival culde-sac and orbit using stepwise methodical approaches to avoid facial asymmetry [3-6].

There are few reports describing orbital osteotomies for the management of severe bilateral microphthalmia. Orbital osteotomies are only indicated in the more severe cases, or for the ones presenting late in the course of the disease [7,8].

Case Presentation

At 6 months old, patient showed a bilateral partial cryptophthalmos; upper and lower conjunctival fornices 3/4 mm in both eyes and palpebral fissure of 12/11 mm and clear facial asymmetry with undergrowth of upper face. No light perception was documented in both eyes, and no other systemic or genetic abnormalities were documented.

Initially, expansible catheters were implanted in both orbits. Progressive increases in volume were made allowing a precise control of conjunctival socket expansion and a correct prosthesis fitting that lasted for several years.

Orbital development was followed until the patient was 4 years old and showed with upper face underdevelopment presenting with left palpebral and orbital structures more affected (Figure 1). In order to achieve proper volume for a bio ceramic implant (18 mm) further orbital space was required. Therefore, the patient underwent a unilateral left orbital reconstruction procedure: Lateral canthotomy and cantholysis plus soft tissue dissection allowed a wide access to the zygomatic-frontal rim. An automatic saw was used in order to perform a squared lateral orbitotomy getting a free movable lateral wall with squared margins. Previous tomographic orbital images allowed to accurately calculate safe osteotomy margins. Lateral wall was displaced laterally as the 18 mm implant was located in the empty cavity with a perfect fitting (Figure 2A-2C). Postoperative cosmetic results can be seen in figure 3A and 3B.

Figure 1: Preoperative photograph. Upper face underdevelopment with left palpebral and orbital structures more affected.

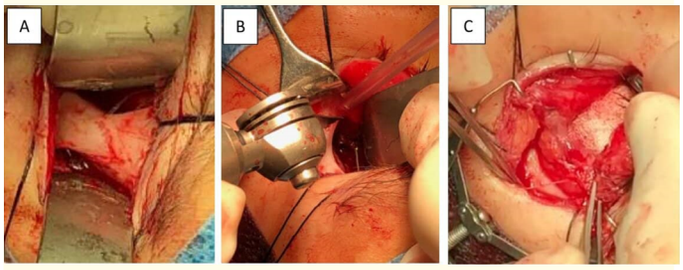

Figure 2: Orbital reconstructive procedure. A. Exposure of lateral orbital wall. B. Squared lateral orbitotomy with automatic saw. C. 18 mm implant displacing lateral wall.

Figure 3: Postoperative results. A. Postoperative result at 24 hours showing adequate expansion of the left orbit. B. Postoperative result at 8 weeks. Upper face symmetry with optimal cosmetic appearance.

Discussion

Mild/moderate microphthalmia is generally managed conservatively with insertion of a conformer and periodically increasing its size. Several techniques have been proposed for the management of severe microphthalmia and anophthalmia. Therapy aims to maximize existing vision and enhance cosmetic appearance rather than improving sight. Most surgeons believe in the concept of "active conservative treatment" in which early external orbital conformers are used for socket expansion followed by subsequent orbital surgery, when the child becomes 3 or 4 years old [9].

Reconstructive strategies rely upon the simultaneous management of both soft tissue hypoplasia and asymmetric bone growth using a stepwise approach. Treatment is usually started within weeks of life using conformers to enlarge the palpebral fissure, conjunctival culde- sac and orbit [5]. Endo-orbital volume replacement using implants of progressively increasing size can be used to stimulate expansion of the developing bony orbit. Volume replacement using implants and expanders can also be supplemented by the use of dermis-fat grafts [7,8,10].

Orbital osteotomies are indicated in more severe cases. Early osteotomies have been mostly unsuccessful and frequently lead to retardation of orbital growth and overlying soft tissue contracture. This is why orbito-cranial advancement surgery with osteotomies and cranial bone grafts is reserved for severe cases or for the ones presenting late in the course of the disease [9,10]. It is important to mention that the main objectives when performing an osteotomy are to provide upper face symmetry and allow the fitting of an orbital implant big enough not to be replaced over time.

In the presented case, lateral orbitotomy was needed to increase the orbit volume and obtain a wider orbital margin allowing for a correct receptor size implant. With smaller implants suboptimal functional and esthetic results would be obtained. Secondary socket reconstruction with mucous graft or dermal-fat graft will provide a perfect host for a normal size adult prosthesis.

Conclusion

This case shows that treatment in severe microphthalmos without existing vision is directed towards avoiding facial asymmetry and improving cosmesis, orbital osteotomies are indicated to provide upper face symmetry and allow the fitting of an orbital implant big enough not to be replaced over time. Based in our experience, this surgical approach results in optimal esthetic results.

Conflict of Interest

No conflict of interest exists for any author.

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