Peters'Anomaly – Strabismus and Amblyopia: a case report

I. M. Poças[1], P. M. Lino[2]
[1] *Departamento de Ciências e Tecnologias de Reabilitação, Escola Superior de Tecnologia da Saúde de Lisboa (ESTeSL), Instituto Politécnico de Lisboa, Lisboa, Portugal,* [2] *Departamento de Oftalmologia, Hospital Cuf Cascais, Portugal*

ABSTRACT: *Purpose.*Peters' Anomaly is a rare congenital corneal opacity related to a malformation of the anterior segment of the eye, causing severe amblyopia. It can be identified isolated or in association with other ocular or systemic abnormalities. The etiology of Peters' Anomaly remains uncertain, but the most likely causes are related to genetic, infectious, traumatic and toxic factors. A range of possible treatment strategies exists, though the effectiveness of each of them depends on how the disease occurs and whether it is identified in early or advanced stages - the earlier the diagnosis, the higher the possibility of a successful intervention. This work reports a case of bilateral Peter's Anomaly, type I, in a 7-years-old girl with amblyopia, horizontal strabismus also had dissociated vertical deviation, and ocular movements is compatible with a bilateral Duane Syndrome, type I.

KEYWORDS: Peters' Anomaly, strabismus, amblyopia, corneal opacity, keratoplasty

1 INTRODUCTION

Peters' Anomaly, first described by Albert Peter in 1906, consists of a central corneal opacity related to a malformation of the anterior segment of the eye[1]. It is a disease in a constellation of diseases that causes corneal opacity, iridocorneal adhesions due to dysgenesis of the anterior segment during development. Peters' Anomaly can cause devastating corneal opacity in an infant leading to severe amblyopia. It frequently occurs with associated strabismus, usually convergent sensorial type, also had dissociated vertical deviation (DVD) [2].
The exact prevalence of Peters' Anomaly is unknown. This condition is one of a group of disorders known as congenital corneal opacities (Figure 1), which affect three to six individuals per 100,0003.
Pathophysiology: Peters' Anomaly is a rare dramatic finding at birth, manifests in utero during the first trimester of pregnancy (10-16 weeks of gestation), and can be associated with other systemic malformations[4]. It is classified in two types, which are distinguished by their signs and symptoms. Peters' Anomaly Type I, is characterized by an incomplete separation of the cornea adhesion. Keratolenticular adhesion is absent in this type. Type II is characterized by an incomplete separation of the cornea and lens and severe corneal opacity that may involve the entire cornea. It presents with keratolenticular adhesion. Type II is more associated with systemic alterations and tends to be bilateral [6,7].
During development of the eyes, the elements of the anterior segment form separate structures. However, in Peter's Anomaly, development of the anterior segment is abnormal, leading to incomplete separation of the cornea from the iris or the lens. As a result, the cornea is cloudy

(opaque), which causes blurred vision. The opaque area of the cornea varies in size and intensity from a small, faint streak to a large, white cloudy area that covers the front surface of the eye. Additionally, the location of the opacity varies; the cloudiness may be at the center of the cornea or off-center. Large, centrally located opacities tend to cause poorer vision than smaller, off-center ones [7,8]. Complications include amblyopia and decreased vision or blindness from glaucoma. In bilateral Peters 'Anomaly little is known about the development of strabismus and amblyopia. Marked asymmetry in the development of the visual system, in severe unilateral cases, would be expected to produce amblyopia and sensory strabismus, but in bilateral Peters 'Anomaly the incidence of strabismus and the occurrence of amblyopia are unknown [3].
It is important a binocular vison evaluation in order to identify, qualify and quantify the type of ocular deviation, characterize the real and potential binocular single vision and the amblyopia. The motor and sensorial tests must be appropriate to the case in question, in particular, visual acuity and fixation. The treatment involves a corneal transplant which is often complicated due to the young age of the affected patient[9].To prevent amblyopia and provide visual rehabilitation penetrating keratoplasty (PKP), was recommended[5]. Many children with PKP for Peters' Anomaly Type I can experience good or functional vision in their operated eye. After PKP is very important to improve visual acuity and treat the amblyopia.

2 CASE REPORT

A 7-years-old girl with a diagnosis of bilateral Peters' Anomaly, Type I. There was history of bilateral iridectomy performed at 2-months-old and no known maternal infections during the pregnancy or during perinatal period.
At 14-month-old the girl presented a bilateral low vision acuity for age (Teller acuity cards):
RE (sph +4,00): 20/710 (too low for the age)
LE (sph +3,50): 20/260 (lower limit for the age)
Binocular: 20/190 (lower limit for the age)
At the moment of examination there was an alternant esotropia, as well as an alternating hipertropia and latent nystagmus. The girl also presented a limitation of the abduction of the right eye. Corneal opacity circunstrited. Ocular fundus examination with indirect ophtalmoscopy, under sedation, was normal. The maculae were normal-looking, pink optic discs with defined edges without increased digging of the optic nerve, and Goldmann ocular pressure in both eyes, was 6.0 mmHg.
The patient is currently waiting for corneal transplantation. Ophthalmic examination maintains the initials characteristics.

Orthoptics Report
Bilateral corneal opacities, central in the right eye and paracentral in the left eye (Figure 1).

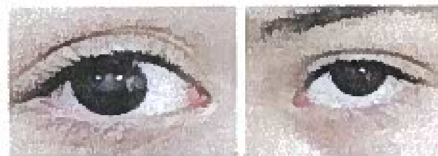

Fig. 1 - Bilateral corneal opacities

Anomalous head posture (AHP) with elevation of the eyes with the chin depressed (Figure 2).

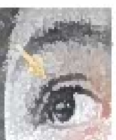

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Fig. 2 AHP - Elevation of the eyes with the chin depressed.

Visual acuity: (with AHP)
RE (sph +2.50): 2/10 (using the illiterate E Snellen)
6/12 (Sheridan cards)
LE (sph +2.50): 2/10 (using the illiterate E Snellen)
6/9 (Sheridan cards)
Binocular: 2/10 (using the illiterate E Snellen)
6/9 (Sheridan cards)
Ocular Movements: In dextroversion, limitation of the abduction with enlargement of the left eyelid. In levoversion, limitation of the adduction with retraction of the globe, with narrowing of the palpebral fissure. Also a bilateral upshoot in adduction (Figure 3).
Nystagmus with rapid phase to the right side
Cover Test (with and without glasses): Alternating esotropia with alternating hipertropia, preferential fixation with LE.
Krimsky: 18$^{\Delta}$ Base-out R/E or L/R 3-6$^{\Delta}$

Fig. 2 Ocular movements - horizontal versions: A. in dextroversion limitation of abduction with upshot. B. primary position, left esotropia and left hipertropia, with right eye fixating.
C. limitation of adduction with upshoot and retraction of the left eye. In the images it is possible to identify the presence of bilateral corneal opacities.

Synoptofore: +10° R/E 5$^{\Delta}$ (objective angle). Fusion negative
Stereopsis: Negative (Titmus Stereo teste) Binocular vision: Absent.
To improve visual acuity, the amblyopia was treated with part-time patching, for six hours, alternating eyes.

3 DISCUSSION

Visual deprivation secondary to Peter's Anomaly resulst in sensory deprivation amblyopia. This child presents a moderate visual loss.

Najjar and Christiansen[2], claim the convergent strabismus is the most prevalent type in patients with Peters' Anomaly, and is present in this case. Oculomotor status and eye movements are compatible with bilateral Duane syndrome, co-existing with Peters' Anomaly, type I.

The treatment of strabismus in cases of Peters' Anomaly follows the general rules of treatment of concomitant strabismus. The first step should be the best optical correction possible. The surgical proposal must be made after achieving visual acuities between the two eyes.

Peters' Anomaly is the most common indication for penetrating keratoplasty in infants[2]. Growth can reduce the success of the transplant because it increases the risk of rejection. After transplantation, the patient should receive systemic immunosuppressant in the early stages and then perform visual stimulation exercises to improve visual acuity [1,7], and also to improve the success of keratoplasty and the correction of the strabismus. In this cases, it is important the multidisciplinary evaluation in the ophthalmology area (orthoptists – squint evaluation and orthoptic rehabilitation, ophthalmologists of various sub-specificities - strabismus, cornea, glaucoma) to improve treatment success[6]. Children with Peters' Anomaly require special educational needs depending on the visual acuity. In order to reduce the handicap caused by visual impairment and to improve the child's functional vision, technical aids can be adapted. A low vision specialist should evaluate these children. Patients may need special equipment (loupes binocular, other low vision aids) depending on the visual potential [4].

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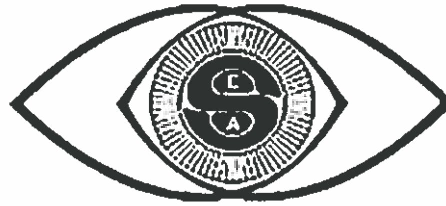

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Daniela Eleonora Cioplean

Ophthalmology Clinic OFTAPRO
Bucharest, Romania