

Introduction:

Peter's Anomaly, first described by Albert Peter in 1906, consists of a central corneal opacity related to a malformation of the anterior segment of the eye¹. It's 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), having also 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.

Physiopathology: Peter's 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^{4,5}. 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 and iris and mild to moderate corneal opacity. Type II is characterized by an incomplete separation of the cornea and lens and severe corneal opacity that may involve the entire cornea. 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 Peters' 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 (opacity) 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}.

It is important a binocular vision 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. To prevent amblyopia and provide visual rehabilitation a penetrating keratoplasty (PKP), was recommended⁵. Many children with PKP for Peters' Anomaly Type I can experience good or functional vision in their operated eye. After keratoplasty is very important to improve visual acuity and do amblyopia treatment. Children with glaucoma have a poorer visual prognosis⁹.

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. In congenital strabismus, late surgery will only have esthetic value.

Case-report

Female, 7 years-old with a diagnosis of bilateral Peters' Anomaly, Type I. (Figure 2)

Clinical History

There were no known maternal infections during the pregnancy or during perinatal period.

Bilateral iridectomy performed at 2 months-old. At 1 year-old presents a bilateral low vision for age, alternante esotropia with DVD and latent nystagmus. 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.

Orthoptic's Report

Anomalous head posture (AHP) with elevation of the eyes with the chin depressed (Figure 3).

Visual acuity: (with AHP)

RE (sph +2,50): 2/10 Snellen (Sheridan 6/12)

LE (sph +2,50): 2/10 Snellen (Sheridan 6/9)

Binocular : 2/10 Snellen (Sheridan 6/9)

Ocular Motility:

In dextroversion, limitation of abduction with enlargement of the eyelid left. In levoversion, limitation of the adduction with retraction of the globe. Also a bilateral upshoot in adduction.

(Figure 4) Nystagmus with rapid phase to the right side

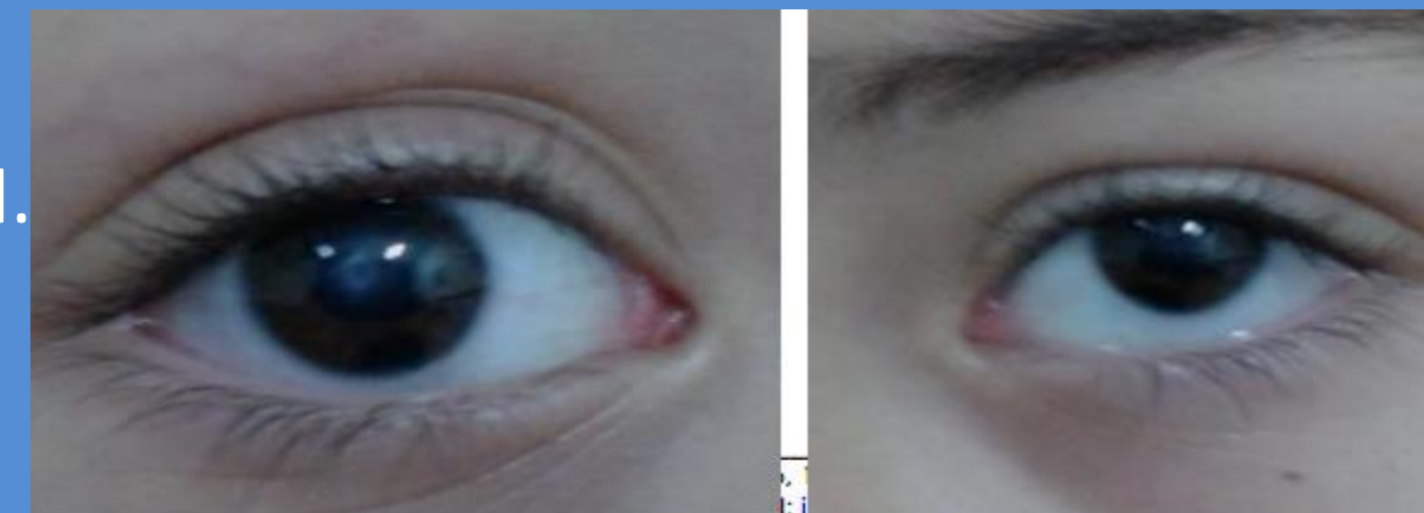


Figure 2. Bilateral corneal opacities

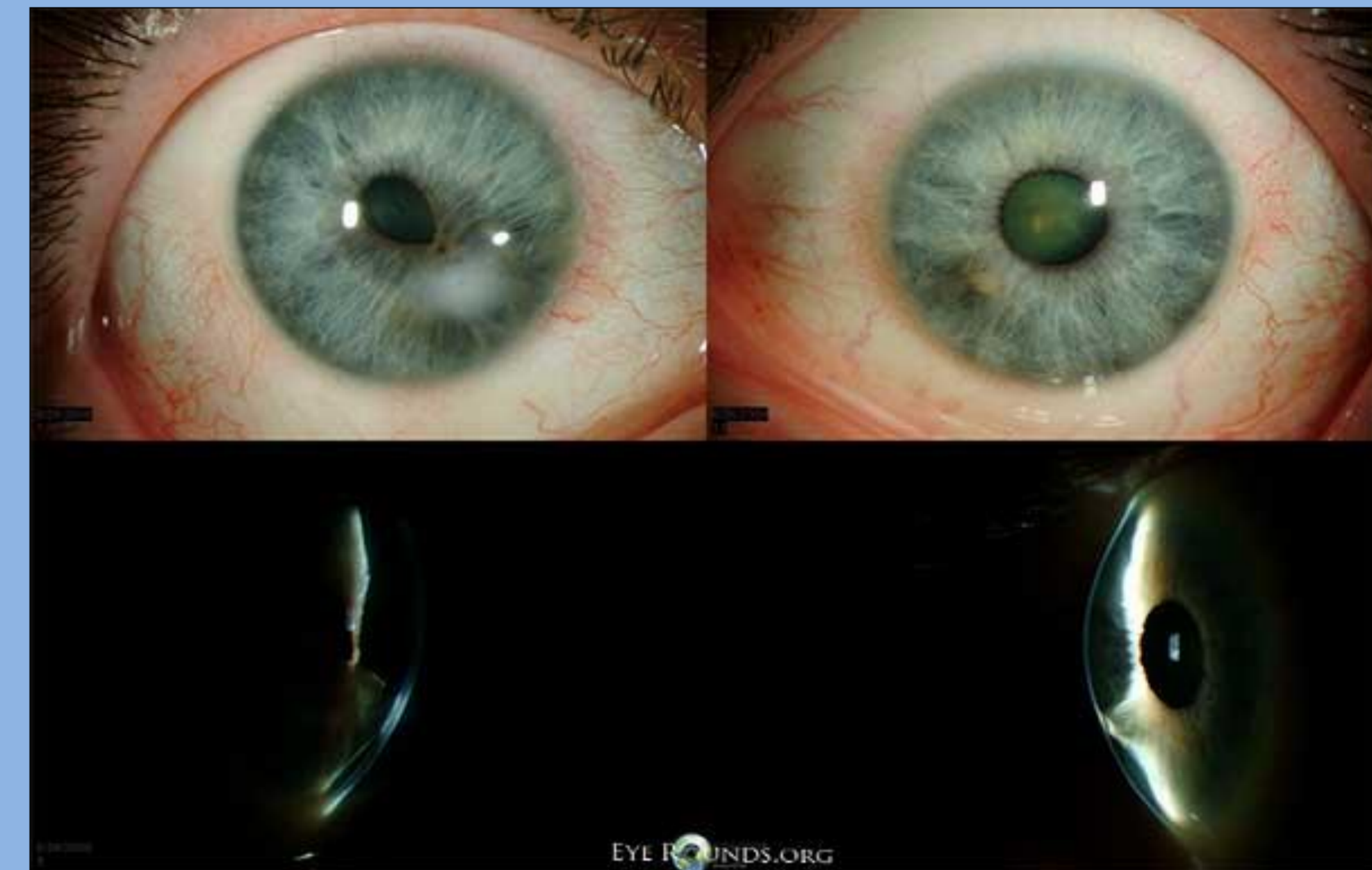


Figure 1. Small bilateral corneal opacities with associated iridocorneal adhesions in a patient with Peters' Anomaly. In top- ocular photography and in bottom slit lamp image. Image source: <http://webeye.ophth.uiowa.edu/eyeforum/cases/187-Peters-Anomaly.htm>



Figure 3. AHP - elevation of the eyes with the chin depressed.

Discussion:

In the present case, the patient presents bilateral Peters' Anomaly, Type I, with alternating esotropia, DVD and bilateral low vision.

The convergent strabismus, is the most prevalent type in patients with Peters' Anomaly², and in this case, presents with motor characteristics of Duane Retraction Syndrome - type I and sensorial status of early onset strabismus.

Peters' Anomaly is the most common indication for penetrating keratoplasty in infants². Growth can reduce the success of the transplant because it increases the risk of rejection. After transplantation, the patient should do systemic immunosuppressants in the early stages and then perform visual stimulation exercises to improve visual acuity^{1,7}, and to improve the success of keratoplasty and the correction of the strabismus.

In the cases of Peter's Anomaly, 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.

In order to reduce the handicap caused by visual impairment and to improve the child's functional vision, technical aids can be adapted.

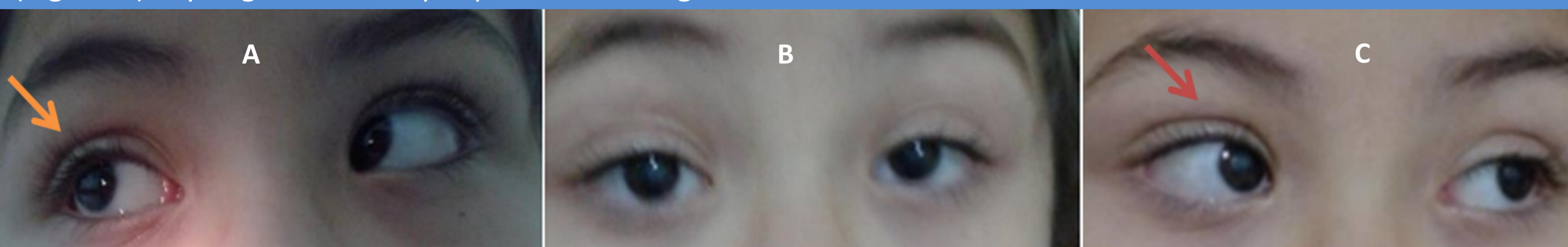


Figure 4. A, in dextroversion limitation of abduction with upshoot. B, primary position, right manifest esodesviation and left hypertropia with right eye fixating. C, limitation of adduction with upshoot and retraction of the left eye. In the images it is possible to identify the presence of a bilateral corneal opacity.

Cover Test (with and without glasses): Alternating esotropia with alternantig hypertropia (DVD)- preferential fixation with LE.

Krimsky: 18^Δ Base-out R/E or E/R 3-6^Δ

Synoptofore: +10^Δ R/E 5^Δ (objective angle). Fusion negative

Binocular vision: Absente.

Stereopsis: Negative (Titmus Stereo teste - Fly test)

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