Retroiridian Pupilloplasty

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Abstract

Objective: To present a novel surgical technique defined as phakic retroiridian pupilloplasty that was performed on a patient with Axenfeld-Rieger syndrome who had no defined pupil.
Surgical technique: Phakic retroiridian pupilloplasty is an original surgical technique involving the creation of a sclerocorneal incision through a peripheral iridotomy, with the surgeon working behind the iris and creating a neopupil with an anterior chamber vitrectome. It requires very few follow-up visits and the patient's recovery is fast. It has been performed on an adult patient with Axenfeld-Rieger syndrome and no complications have been observed after 12 months of the procedure.
Conclusion: The surgical technique described here has proven to be efficient and safe for the formation of a pupil in an adult patient with Axenfeld-Rieger syndrome. This might be of help to improve the patient's visual perception and esthetic appearance, but mainly as an option in pediatric patients to avoid conditions such as strabismus and amblyopia.
Key words: Axenfeld-Rieger syndrome, surgical technique, phakic retroiridian pupilloplasty, amblyopia.

Pupiloplastia retroiridiana

Resumen

Objetivo: Presentar una novedosa técnica quirúrgica definida como pupiloplastia retroiridiana fáquica aplicada en una paciente con síndrome de Axenfeld-Rieger que no tenía una pupila definida.
Técnica quirúrgica: La pupiloplastia retroiridiana fáquica es una técnica quirúrgica original que se

realiza tras una incisión esclero-corneal a través de una iridotomía periférica, trabajando por detrás del iris y realizando la neopupila con un vitréctomo de cámara anterior. No requiere de demasiados controles oftalmológicos y su recuperación es rápida. Se realizó en una paciente adulta con síndrome de Axenfeld-Rieger y no se han observado complicaciones a 12 meses de la cirugía.

Conclusión: Se describe una técnica quirúrgica que resultó eficaz y segura para formar una pupila en una paciente adulta con síndrome de Axenfeld-Rieger. Esto podría ayudar a la mejora de su percepción visual y su aspecto estético, pero principalmente se constituye como una opción en pacientes pediátricos para evitar alteraciones como el estrabismo y la ambliopía.

Palabras clave: síndrome de Axenfeld-Rieger, técnica quirúrgica, pupiloplastia retroiridiana fáquica, ambliopía.

Pupiloplastia retroiridiana

Resumo

Objetivo: Apresentar uma nova técnica cirúrgica definida como pupiloplastia retroiridiana fácica aplicada em um paciente com síndrome de Axenfeld-Rieger que não tinha uma pupila definida.

Técnica cirúrgica: A pupiloplastia retroiridiana fácica é uma técnica cirúrgica original realizada após uma incisão esclero-corneana através de uma iridotomia periférica, trabalhando atrás da íris e realizando a neopupila com uma vitrectomia de câmara anterior. Não requer muitos controles oftalmológicos e a recuperação é rápida. Foi realizada em um paciente adulto com síndrome de Axenfeld-Rieger e nenhuma complicação foi observada 12 meses após a cirurgia.

Conclusão: Descrevemos uma técnica cirúrgica que foi eficaz e segura para formar uma pupila em um paciente adulto com síndrome de Axenfeld-Rieger. Isto poderia ajudar a melhorar sua percepção visual e aparência estética, mas é principalmente uma opção em pacientes pediátricos para evitar distúrbios como estrabismo e ambliopia.

Palavras-chave: Síndrome de Axenfeld-Rieger, técnica cirúrgica, pupiloplastia retroiridiana fácica, ambliopia.

Introducción

Axenfeld-Rieger syndrome (ARS) is one of the so-called iridocorneal dysgenesis syndromes[1], and has been defined since its original description in 1920 as the development of a posterior embryotoxon with iris strands attached to an anteriorly displaced hypertrophic Schwalbe line[2], associated with the universe of congenital iris anomalies such as hypoplasia, corectopia or polycoria[3]. It may also be associated with glaucoma and systemic findings characterized as bone, facial and/or dental defects[4]. ARS is a rare disease affecting the eye bilaterally with an estimated prevalence of 1/200,000 persons with no gender predilection and characterized by autosomal dominant inheritance with complete penetrance of variable expressivity[5].

The aim of this work is to present a surgical technique named phakic retroiridian pupilloplasty in a patient with ARS.

Surgical technique

A brief description of the case. We present a 27-year-old adult patient who came to the ophthalmological consultation referring that she never had a pupil. She had no hereditary family history and she did not use any optical aids. As for her personal history, she mentioned seizures associated with febrile episodes and neonatal jaundice treated with phototherapy. Upon questioning, she reported that an ophthalmology department tried to perform a laser pupilloplasty on her, but due to her iridian thickness, it did not work. At the initial examination, visual acuity was measured, resulting in light vision in the right eye (RE) and 10/10 in the left eye (LE) without correction associated with a mild exotropia. Slit-lamp biomicroscopy revealed the RE with a formed anterior chamber and a transparent cornea that allowed a depigmented and thinned iris to be seen in the place where the pupil should be located. At the level of the temporal peripheral cornea, from 6 to 12 o'clock, there was a significant arched whitish ridge concentric to the lim-

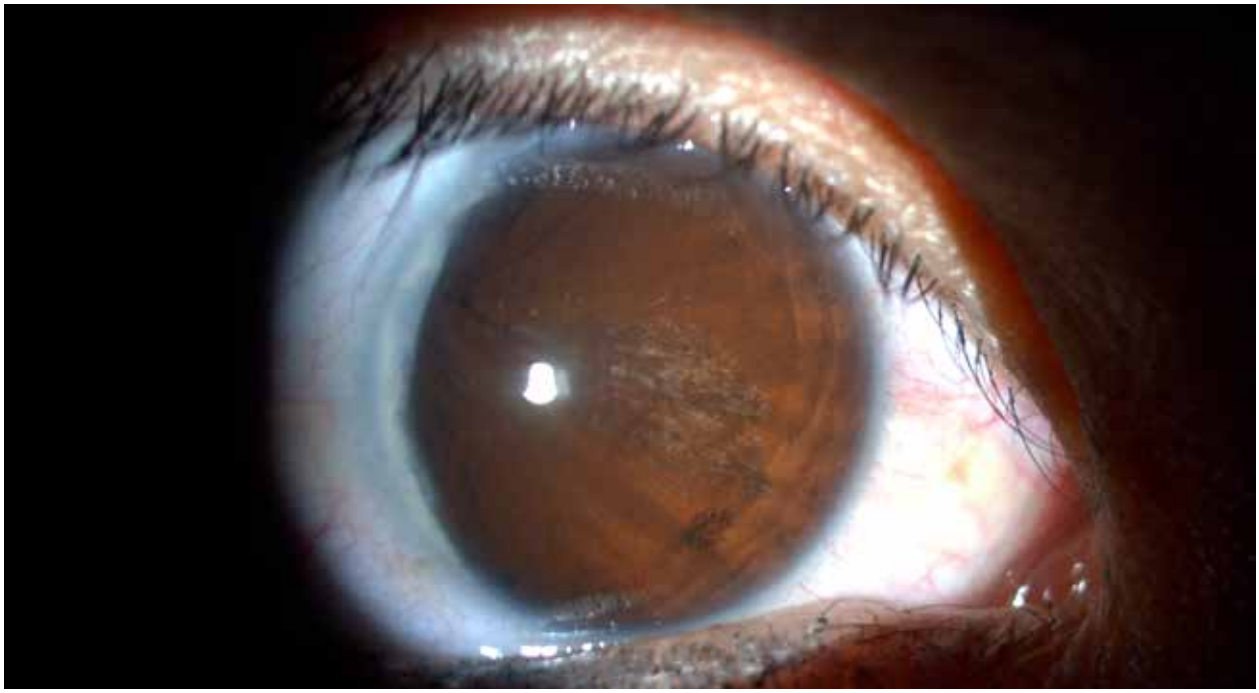

Figure 1. Anterior segment. Pupil occultation associated with a peripheral, whitish, arcuate ridge concentric to the limbus, affecting the entire corneal thickness and corresponding to a posterior embryotoxon.

bus that affected the entire corneal thickness and that would correspond to a posterior embryotoxon associated with anterior synechiae of the peripheral iris, which generated traction with a important displacement that hid the pupil (Fig. 1). Gonioscopy revealed dysgenesis of Schwalbe's line associated with peripheral anterior synechiae that generated iris traction (Fig. 2). Eye pressure was within normal parameters. Once the ophthalmological evaluation was completed, the surgical option was explained to the patient and it was decided to schedule a phakic retroiridian pupiloplasty, a technique that will be described below. In the preoperative period, the use of combined 5% phenylephrine + 0.5% tropicamide topical eye drops was indicated every 15 minutes from 2 hours before, associated with a topical treatment of 0.3% tobramycin four times a day for a period of three days.

Surgery

1. Initial antiseptic measures were carried out by placing 5% iodine solution on the face, ocular surface and cul-de-sacs.

2. Subtenon anesthesia was applied with a curved 19G Stevens cannula, loaded with a mixture of 0.75 ml of 1% lidocaine + 0.75 ml of 0.5% bupivacaine, both without preservatives.

3. The surgeon's chair and the microscope optics were arranged in a temporal position with respect to the patient's position for better handling of the instruments. An incision was then made at the level of the inferior sclero-corneal limbus (IECI) so as not to traumatize the 2.8 mm valved ridge corresponding to the posterior embryotoxon. Comment: it may also be smaller given that during surgery, in addition to the fact that it tends to enlarge due to stretching, smaller diameter instruments (such as the anterior chamber vitrectomy, iris manipulators and cannulae) were used through this incision, avoiding thus the collapse of the anterior chamber during surgery.

4. Abundant 1% lidocaine without preservative in 1:2 dilution of Ringer's lactate was administered into the intracameral space. In addition, an ampoule of 1 ml of adrenaline at 1‰ in 9 ml of lactated Ringer's solution without preservatives was also used in the same space (1:10 dilution). At this point in the surgery, the appearance of a

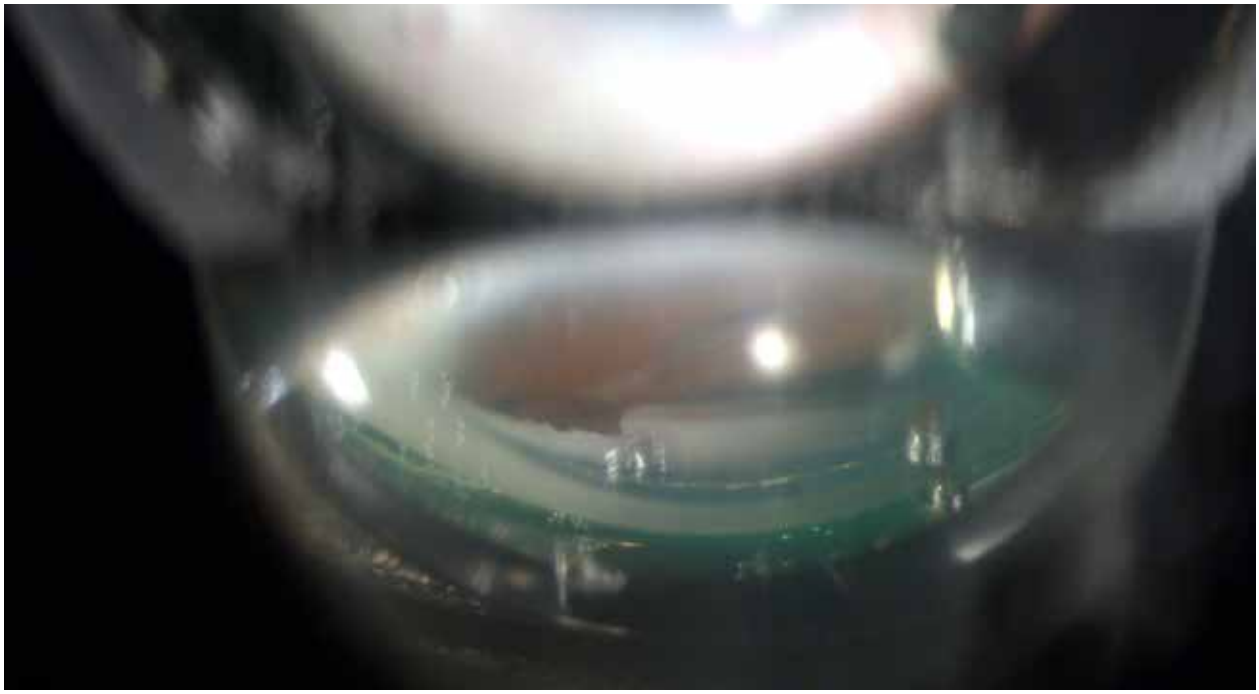

Figure 2. Gonioscopy. Schwalbe's line dysgenesis associated with peripheral anterior synechiae causing iris traction.

very small pupil decentered towards the temporal was observed.

5. Sufficient 2% hydroxypropyl methylcellulose light viscous substance was placed to protect the endothelium, occupying only one third of the anterior chamber in front of the iris.

6. A superior corneal incision (ICS) was made for the management of a 0.8 mm valved second hand with a 15° knife.

7. Through the IECI, an entrance door to the retroiridian space was generated through a small inferior peripheral iridotomy of small diameter with Wescott scissors and a 0.12 mm Serrated Castroviejo forceps.

8. The retroiridian space was shaped by injecting 3% sodium hyaluronate (HS) viscoelastic neoformed peripheral iridectomy to pull it and generate a dome of superior convexity towards the corneal endothelium, occupying two thirds of the anterior chamber without touch the endothelium.

9. The anterior chamber was entered through the previously performed ICS in front of the iris with the continuous irrigation handpiece.

10. A 20 G caliber anterior vitrectomy was used by the IECI, threading through the iridotomy to make a 1.5 mm horizontal path and then tilting upwards until the silhouette of the handpiece was visualized through the iris, to avoid coming into contact with the lens and thus perform a small phakic retroiridian pupilloplasty. The parameters used were: "cut rate of 1000 cpm, rate asp 20 cc/min and a vacuum of 250 mmHg" (Fig. 3).

11. In the surgical technique described, an attempt was made to respect the disposition of the muscular fibers of the iris dilator, taking into account that it was under the mydriatic effects of the drugs used, since under these circumstances the pupil acquires a different position compared to the position of the pupil at body rest. In this way, we worked with the vitrectomy anterior

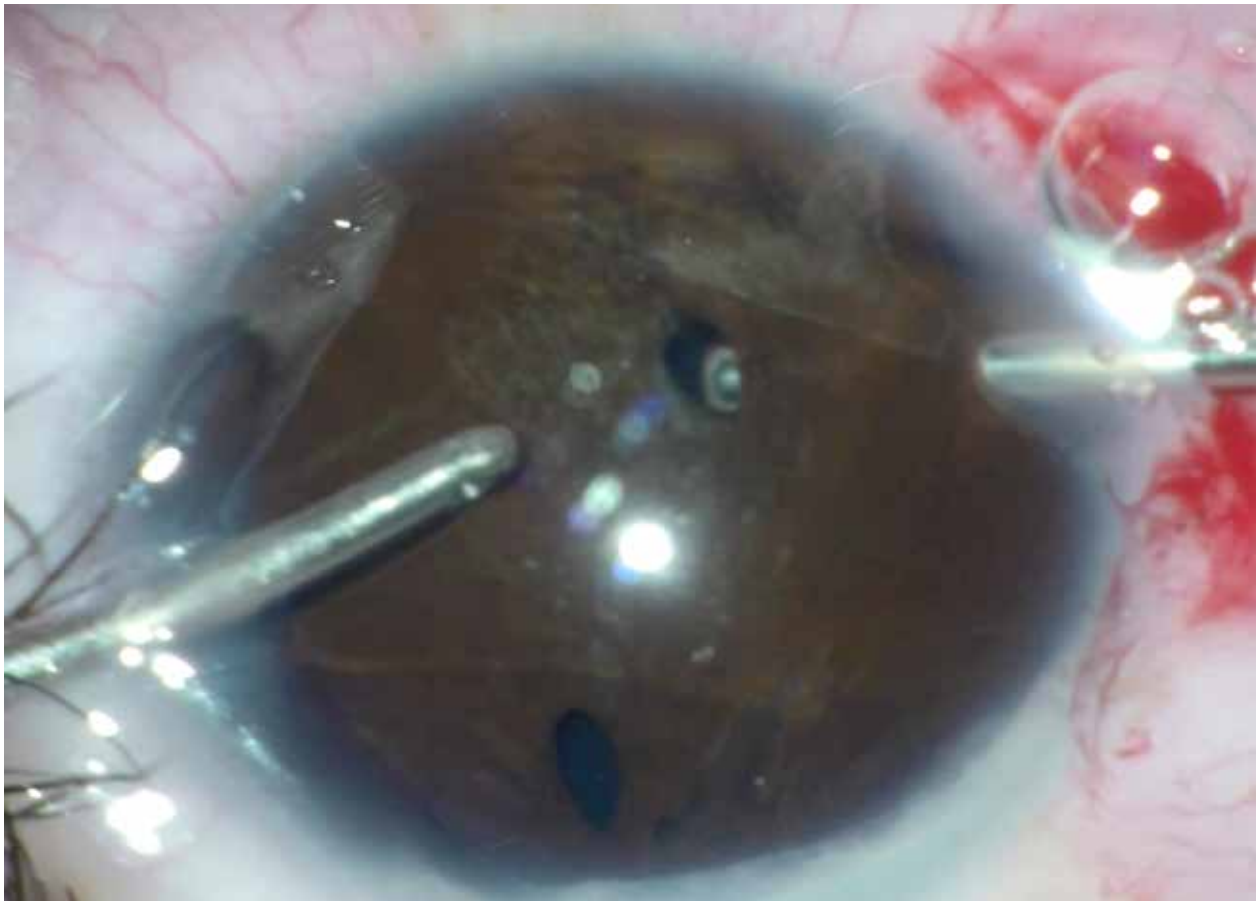

Figure 3. Surgical technique by threading through the inferior iridectomy to perform a small pupilloplasty, drawing a spiral circle that gradually enlarges.

to the iris, drawing an oval figure with a larger vertical diameter (Fig. 4), since at rest it would end up being an oval pupil with a horizontal diameter (Fig. 5).

12. A second time was performed —after placing 3% sodium hyaluronate in the anterior chamber— a bimanual stretching of the neopupil using two Graether "shirt button" type iris manipulators through the upper and lower incisions previously made to achieve a controlled enlargement of the newly formed pupil (Fig. 6).

13. When the iris herniated through the incision, it was relocated in its place using a 2-mm Culler-type spatula.

14. Finally, the viscoelastic was aspirated from the anterior chamber through the neopupil in front of the iris without touching the lens, and a suture was placed in the IECI, given its proximity to the eyelid margin and the risk of infection. In the case where the iridectomy was too large —due to stretching or another reason— it could be sutured.

Likewise, ciprofloxacin 0.3% + dexamethasone 0.1% ointment was placed on the ocular surface.

Post-surgical

A topical treatment of 0.3% ciprofloxacin eye drops plus 0.1% dexamethasone was carried out for a period of 30 days, where the first 15 days were indicated 4 times a day and then twice a day, for another 15 days. The suture was removed at 3 months and controls were performed at 24 hours, 7 days (Fig. 7) and 3 months. Figures 6 and 7 show intra-surgical and late post-surgical images.

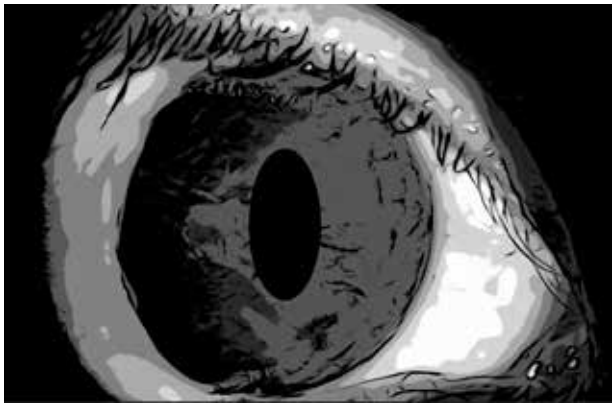

Figure 4. Portrait image of the intraoperative under mydriatics, immediately after pupilloplasty.

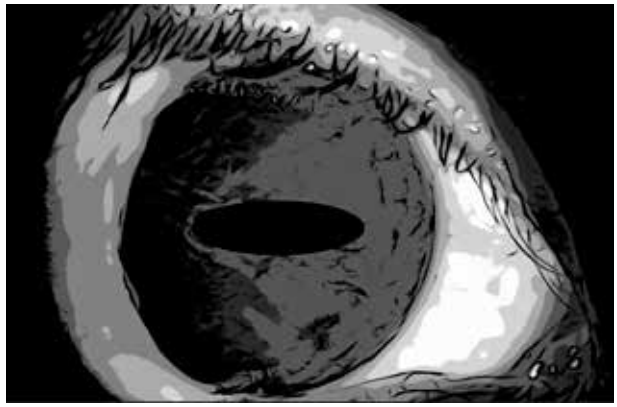

Figure 5. Portrait image of the late postoperative period after pupilloplasty.

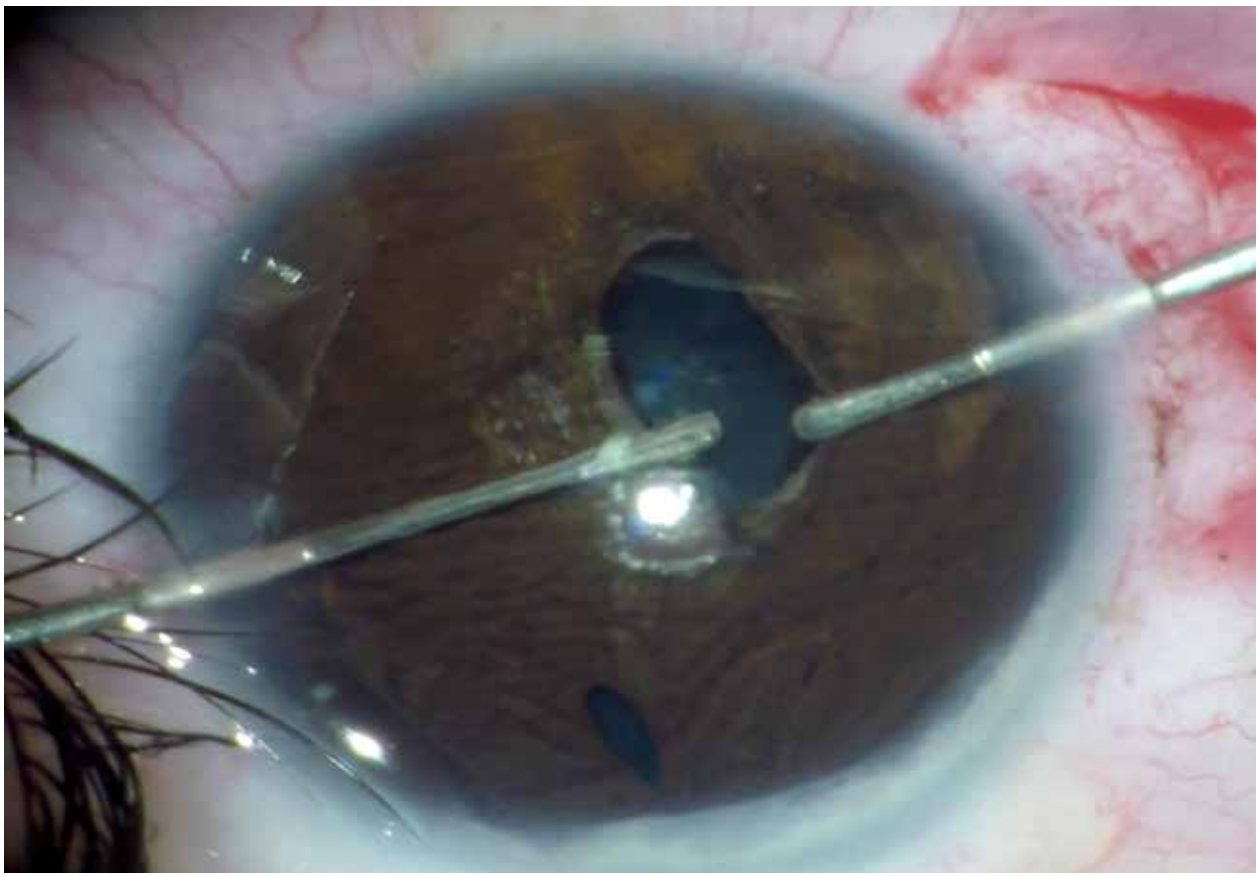

Figure 6. Bimanual stretching of the neopupilla using two Graether "shirt button" iris manipulators.

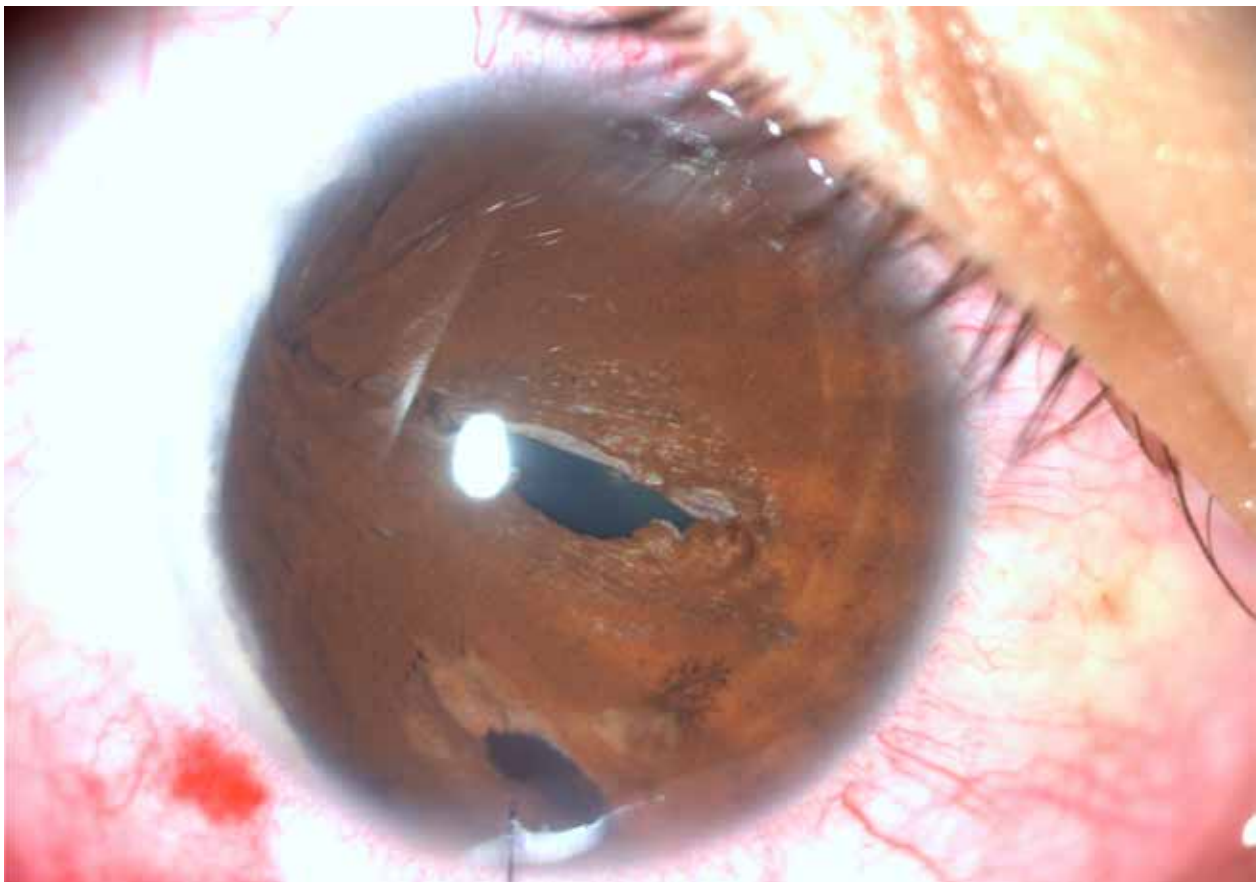

Figure 7. Neoformed pupil 7 days after surgery.

Discussion

There are various causes that can generate iris defects, whether they are congenital (as in the case presented), acquired or traumatic[6]. Pupil alteration in patients with Axenfeld-Rieger syndrome can be difficult to resolve and this situation may affect both functional aspects of the person's visual skills and aesthetic appearance. The type of iris defect can be categorized as: a) complete; b) pigmentary deficiency and c) eye dilation or constriction deficit. The case presented was that of a patient who had a severe dilation deficit associated with a traction with a significant displacement that hid the pupil.

In 1976, the concept of little invasive pupilloplasty was defined, introduced by McCannel when he described a technique in which a long needle was used to pass through two corneal paracentesis to subsequently generate the necessary approximation of the iris, depending on the case[7]. On this basis, the technique underwent modifications and improvements[8-10]. Although there are other options that require the use of prosthetic supplies and devices —which also adds costs—, the current work highlights that an original technique is presented and that is it different from those previously mentioned, and which in turn includes the use of the vitrectome and materials available in any ophthalmology operating room. Although this description is based on a clinical case and expresses the good experience of the authors due to the resolution obtained, it is hoped that other doctors will be able to verify it in a different environment

to really know its precise scope, indications and limitations.

Likewise, it is desired to share this technique with the scientific community, highlighting the importance it would have in pediatric cases, where its performance at an early age could be useful to avoid amblyopia and strabismus due to a lack of the visual stimulus necessary for a correct maturation of the gaze. It should also be considered in adults, since this patient has achieved a recovery of visual acuity, of the visual field, of strabismus, something that is still under follow-up and is expected to be shared in a future long-term report, in addition to the evident improvement of the ocular aesthetics. An important aspect regarding surgery is that it is recommended to aspirate the viscoelastic as much as possible to avoid generating a cataract, not to make very large neopupils so as not to cause severe photophobia, for which it is suggested to respect the embryotoxon due to possible unexpected complications. . Finally, the present technique does not imply greater skill and requires little instrumentation, as previously indicated.

Conclusion

In this work, an innovative technique has been described that allowed to solve a chronic problem of an adult patient with Axenfeld-Rieger syndrome, who was unaware that she had a pupil in her eye, due to tractional displacement and excessive dilation deficit. It is hoped that in the future we will be able to have a series of more cases with a longer follow-up time.

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