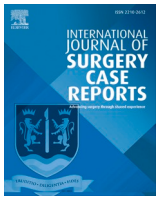
International Journal of Surgery Case Reports 126 (2025) 110717

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr



Case report

Surgical pupil reconstruction for congenital idiopathic acorea with lens transparency saving. Case report

Nadiia Bobrova *, Tetiana Romanova, Olga Dovhan

Pediatric Ophthalmic Pathology Department, SI "The Filatov Institute of Eye Diseases and Tissue Therapy of the NAMS of Ukraine", 49/51 Frantsuzkyi Bulvar, Odesa 65015, Ukraine

ARTICLE INFO

Keywords:
Acorea
Iridectomy
Pupilloplasty
Angle-closure glaucoma

ABSTRACT

Introduction: Idiopathic congenital acorea is extremely rare. There are several techniques for preserving the clear lens during pupil formation.

Case presentation: The complete pupil absence caused acute glaucoma attack 2 weeks after birth, which was controlled in a 1-month-old child by peripheral iridectomy and disconnecting iris-corneal synechias.

Six years later pupilloplasty was performed. Under the iris was found a dense white membrane tightly fused with the iris but separated from the lens anterior capsule during preliminary blind viscodissection. Introducing viscoelastic into the pinhole allowed forming a round pupil 3.0 mm in diameter with vitreal scissors and vitrectomy and maintain the anterior capsule intactness and lens transparency. The artificial pupil exhibited a good cosmetic effect. Low visual acuity (1.7 logMAR) can be explained by a long deprivation period, resulted in deep amblyopia and axial myopia.

Clinical discussion: Previously, artificial pupil formation have been performed with partial, not complete, pupil absence because the eyes had an anterior chamber. Fluid and IOP levels remained within normal limits as the flow between the chambers occurred. This is confirmed by detecting slit-shaped pupils hidden in the iris folds using mydriatics. The presence of pinpoint or slit-shaped pupils allowed light to enter the eyes and residual form vision.

Conclusion: Congenital acorea causes acute glaucoma attack due to a lack of fluid outflow between the anterior and posterior chambers. The differential diagnosis between microcoria and acorea is clinically challenging. Preserving lens transparency in pupilloplasty for acorea was possible by gentle viscodesection technique and vitreal instruments.

1. Introduction

Acorea, the total absence of the pupil, is an extremely rare congenital anomaly. To the best of our knowledge, there have been no previous reports on idiopathic congenital acorea and lens-saving techniques for artificial pupil formation. In this report we present the case of surgical pupil reconstruction for congenital idiopathic acorea with lens transparency saving. The case report has been reported in line with the SCARE criteria [1].

2. Presentation of case

A 1-month-old patient was urgently hospitalized. The patient had no pupil aperture in the right eye, which was complicated by acute secondary glaucoma attack. The child was born on time from the first pregnancy. Family history was not complicated. The pupil's absence in the right eye was detected in the first days after birth. The left eye was normal. The child became disturbed and refused to take the mother's breast two weeks after birth.

The examination was performed under general anesthesia. Biomicroscopy of the right eye showed a slightly swollen cornea, absent anterior chamber, and absent puppilary aperture. Ultrasound biometry measured axial length (AL) of 19.4 and 18.0 mm in the right and left eyes, respectively. Ultrasound scanning revealed the posterior segment without changes. The keratometry data were 45.0^{D} and 45.5^{D} in the right and left eyes, respectively. Applanation tonometry showed intraocular pressure (IOP) of 36.0 and 21.0 mmHg in the right and left eyes, respectively.

* Corresponding author.
E-mail address: filatovbobrova@gmail.com (N. Bobrova).

https://doi.org/10.1016/j.ijscr.2024.110717
Received 23 October 2024; Received in revised form 4 December 2024; Accepted 7 December 2024
Available online 10 December 2024

To eliminate an acute attack of secondary glaucoma in the right eye, the patient underwent peripheral iridectomy with anterior synechiolysis. A large amount of intraocular fluid was released during iris iridectomy. Anterior synechiae lysis was complicated with insignificant hemorrhage. The child became calm after surgery. IOP was normalized.

A repeat visit happened six years later. The 6-year-old child had a cosmetic problem. The examination showed calm eyes, transparent corneas, and medium-depth anterior chambers in both eyes. The pupil in the right eye was absent (Fig. 1), whereas that in the left eye was centrally located and of normal round shape and mobility. There was no fundus reflex in the right eye, whereas the fundus in the left eye was within normal limits.

The IOP level was within normal limits, 16.0 and 20.0 mmHg, in the right and left eyes, respectively (applanation tonometry). The ocular hydrodynamic indices were: true IOP (Po) = 12.0, outflow facility coefficient (C) = 0.24, Becker's coefficient (Po/C) = 51, and rate of aqueous formation (F) = 0.54 in the right eye and Po = 16.0, C = 0.21, Po/C = 76, and F = 1.23 in the left eye.

Examination under general anesthesia of the right eye revealed that the cornea was transparent. The corneal diameters were 10.0 and 11.0 mm in the right and left eyes, respectively. Examination of the right eye revealed the anterior chamber of a middle depth, the violated iris drawing, and the pupil absent (Fig. 2). The peripheral coloboma appeared at 11 o'clock. The reflex of the ocular fundus was absent. Ultrasoud biometry revealed asymmetry with AL of 25.7 mm and 22.4 mm in the right and left eyes, respectively. Ultrasound scanning of the right eye revealed the anterior chamber of medium depth with the anterior chamber angle of about 45°. The total pupil aperture was absent. Single dot-fiber patterns of low echogenicity were detected in the lens. The retina was attached. Outside the optic disc, posterior staphyloma with a depth of 1.7 mm and a length of 9.0 was detected (Fig. 3). Keratometry showed 42.25^{D} and 44.0^{D} in the right and left eyes, respectively. Gonioscopy of the right eye revealed that mesodermal tissue partially covered the angle structures and multiple goniosynechia were present.

Visual acuity (VA) was light perception and 0.0 logMAR in the right and left eyes, respectively. The left eye was healthy. IOL was calculated using the SRK formula as 16.4D and 23.3D in the right and left eyes, respectively.

The patient was diagnosed with idiopathic acorea, operated on secondary glaucoma attack, deep deprivation amblyopia, and axial myopia in the right eye.

Since ultrasound examination showed no visible signs of cataract, surgery was performed to form an artificial pupil and preserve the clear lens.

2.1. Surgery procedure

A limbus paracentesis was made above the peripheral iridectomy, which had previously been formed at 11 o'clock. Viscoelastic material was inserted into the anterior and posterior chamber through the iris peripheral iridectomy, and viscodissection of the iris attached to the lens

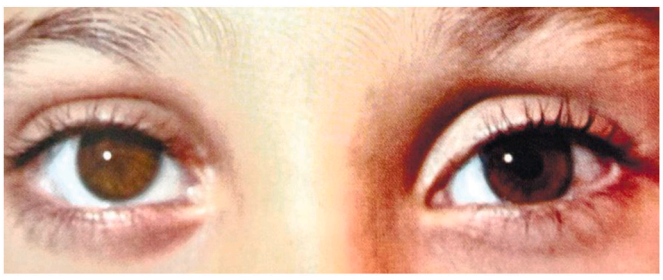


Fig. 1. A 6-year-old girl diagnosed with congenital ideopathic acorea of the right eye.

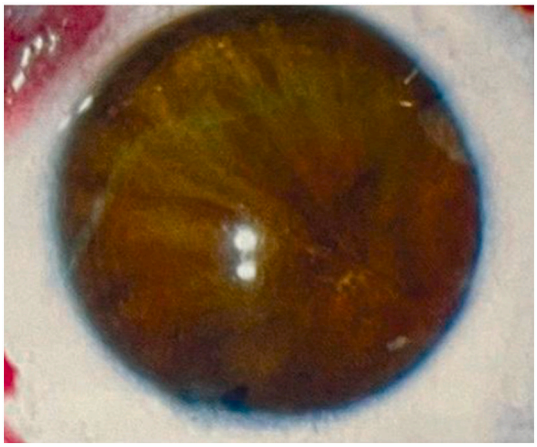


Fig. 2. Congenital idiopathic acorea.

started (Fig. 4a). That was effective at the periphery. In the center, where the pupil was meant to be located, strong adhesions were noted between the lumped iris and the underlying lens. By making certain efforts and gradually moving toward the center, it was possible to break the invisible central adhesions and increase the iris central zone (Fig. 4b). Second limbus paracentesis was performed at 1 o'clock and an attempt was made to create a hole in the iris in the pupil area. The hole was formed very small because of a thick white tissue that could be seen under the iris. This thick film was destroyed using only a knife leaning on a spatula, which was pushed to the center through peripheral iridectomy (Fig. 4c). Only then was it possible to see the surface of the lens, which appeared to be transparent (Fig. 4d). Collect scissors were inserted into the opened iris and film to create guide incisions in both directions (Fig. 4e, f). Continuing to form the pupil, a round hole was made with a vitreotome on a dispersive viscoelastic (Fig. 4g, h); no irrigation fluid was injected. An artificial round pupil was formed in the center of the iris and through it; the intact lens was clearly visualized (Fig. 4i). Minor bleeding occurred from the iris vessels.

2.2. Results

The postoperative period was peaceful. 9 days post-operatively, the newly formed pupil acquired little-bit vertical ovalisation, apparently due to the preserved iris dilator fibers in the inferior outer quadrant (Fig. 5). In the fundus of the eye, the optic disk was pale, the contours were clear, the macular area was smooth, and reflexes were not differentiated. The vessels were evenly narrowed. Skiascopic refraction was -7.0^{D}. VA in the right eye improved to 1.7 logMAR owing to severe amblyopia. The cosmetic result was exceptional, and lens transparency was preserved (Fig. 6).

At 2 months after surgery, the examinations revealed the calm eye with the transparent cornea and the medium depth anterior chamber. The artificially formed pupil was located centrally with horizontal and vertical diameters of 3 and 4 mm, respectively, i.e. the pupil preserved its shape. The crystalline lens remained clear. The ocular fundus had changes as previously. Skiascopic refraction was -7.0^{D}.

2-mm pinhole visual acuity test showed no improvement in vision, from which we can conclude that the shape of the formed pupil does not influence visual acuity. IOP fluctuated within the normal range when hypotensive drops were used.

Further follow-up was not possible due to the emigration of the child's family.

3. Discussion

Acorea is a rare congenital anomaly that appears as complete absence of the pupil. It develops when the mesodermal tissue of the iris

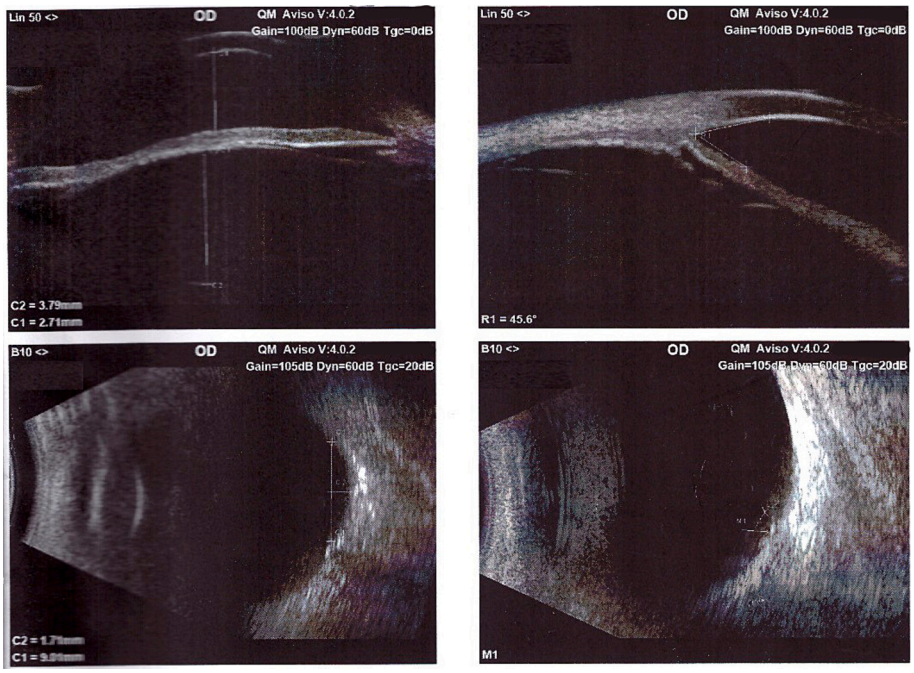


Fig. 3. Ultrasound scanning of the right eye showing the anterior chamber of medium depth with the anterior chamber angle of about 45°. The total pupil aperture is absent. Single dot-fiber patterns of low echogenicity are detected in the lens. The retina is attached. Outside the optic disc, posterior staphyloma with a depth of 1.7 mm and a length of 9.0 is observed.

fails to regress during embryogenesis and can be hereditary or sporadic [2].

Kondo et al. [3] described Familial acorea, combined with microphthalmos, cataracts, and glaucoma, in five affected family members in three generations: the paternal grandmother was blind, had subatrophy with corneal opasification, vitreus degeneration and retinal detachment; four patients had congenital microcoria and acorea. All affected patients had microphthalmic and cataracts in both eyes. The lenses were cloudy. Surgical reconstruction included pupil formation and cataract removal. Improvement in vision after surgery was noted in all cases. No pathology was detected on the fundus.

Our case can be considered idiopathic given that the family history was not complicated and the defect was unilateral with a healthy paired eye with high vision. There were both differences and similarities in the Kondo familial acorea. Thus, in our case, AL at one month of age was slightly greater than that of the healthy left eye, 19.4 and 18.0 + 0.2 mm, respectively, and the difference in AL at the age of six years increased to 25.7 vs. 22.36 mm, respectively. In addition, there was a weakly expressed microcornea; at the age of 6 years, the corneal diameter of the right eye was 10.0 mm, compared with 11.0 mm in the left eye. The observed patient exhibited a clear lens, compared to cataracts in Kondo's cases [3]. The common feature was angle-closure secondary glaucoma, which was managed by performing peripheral iridectomy at one month of age.

Several descriptions of isolated cases of sporadic acorea are available in the literature, including two patients from India: a 13-year-old girl [4] and a young man [5], and a case of binocular acorea developing in a 62-year-old leprosy patient [6]. Through a detailed clinical analysis of the cases they described, the authors came to an understanding that there was not acorea but microcoria when the pupillary aperture existed, although of tiny size. It was detected in response to mydriatic drops or during surgery. Thus, after injecting a viscoelastic substance into the anterior chamber and smoothing out the iris folds, a hidden pinpoint or slit-like pupil was observed.

It is very difficult to make a differential diagnosis between acorea, when the pupil is completely absent, microcoria, when the pupil is too small, and hidden microcoria, when the pupil is hidden by iris folds.

The principal distinguishing feature between microcoria and acorea may be the presence of aqueous humor in the anterior chamber, because if there is no flow of aqueous humor from the posterior to the anterior chamber, iris bombe and an acute attack of angle-closure secondary glaucoma occur. This condition was present in our case of a 1-month-old child, which was managed by peripheral iridectomy. An uncured acute attack of glaucoma progresses to chronic and then to absolute glaucoma, which apparently was the case in the Kondo reports [4], as well as in the case of a 61-year-old woman diagnosed with Sturge-Weber syndrome and acorea with absolute glaucoma described by Rahhal-Ortuño et al. [7]. In the other clinical cases described as “acorea” [4–6], there were microcoria because fluid produced by the ciliary body entered from the posterior to the anterior chamber through the micropupils.

The second clinical sign of microcoria can obviously be considered the presence of object vision and the absence of pronounced deprivation amblyopia because light penetrating into the eye, even through a reduced pupil, contributes to retinal maturation. In our case, due to the long interval between the first and second visits, deep amblyopia had formed, and vision remained low after pupil formation and clear lens saving.

It should be noted that optical coherence tomography (OCT) of the anterior segment is not indicative of a differential diagnosis of microcoria and acorea because its resolution capacity does not allow the capture of micropupils covered by iris folds.

An ultimate distinction may be provided by a genome study of familial microcoria; disease locus has been mapped on chromosome 13q 31-32 [8]. It should be noted, however, that genetic studies have been conducted on hereditary forms, whereas data on sporadic forms are not available.

There are several surgical techniques for pupil deformities in previous studies. Xiage et al. have reported pupilloplasty by radiofrequency diathermy (RFD) [9], which was developed by Kloti for anterior lens capsulotomy [10] and further developed by Gassman et al. [11]. No cases of congenital acorea were reported in this study. Four patients were aphakic and developed secondary severe pupillary fibrous membranes after trauma, previous surgeries, and uveitis, and only one child had congenital microcoria with ectopia. According to the authors, the

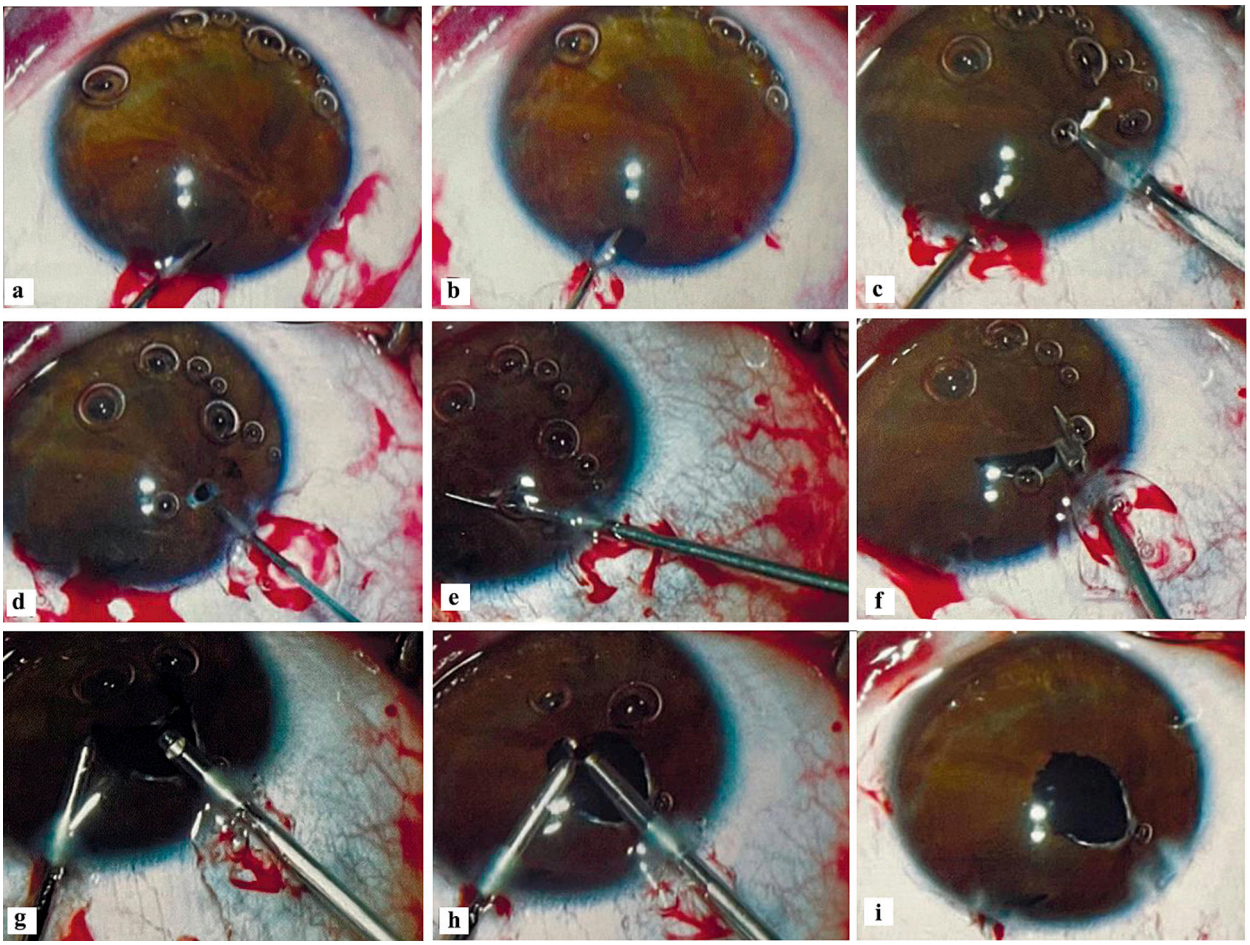


Fig. 4. Intraoperative photographs of the right eye: a, viscodissection of the iris from the anterior lens capsule through peripheral iridectomy; b, additional viscodissection in the upper half; c, membranotomy on the spatula in the area of the supposed pupil; d, additional injection of viscoelastic into the formed opening; e and f, expansion of the formed opening with collet scissors in both directions; g, condition after two iridotomies; h, final pupil formation with vitreotome; i, formed pupil with preservation of lens transparency.

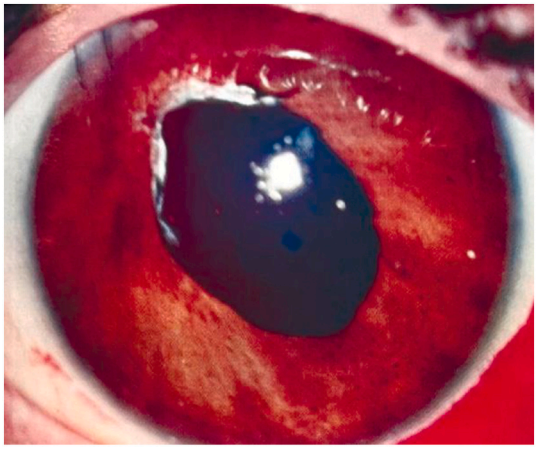


Fig. 5. Day 9 after artificial pupil formation. The crystalline lens is transparent. The newly formed pupil is ovalized. A dense white membrane can be seen under the iris in the upper half.

appearance of air bubbles during surgery obscured the surgical field and made manipulation difficult. In addition, scissors and tweezers were required to dissect the dense membranes. However, the main risk associated with this technique is apparently the use of high t° (approximately 160 °C) in close proximity to the intact lens, which may provoke overheating and consequent opacification.

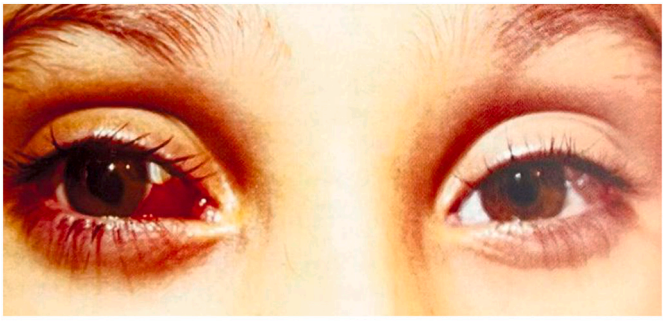


Fig. 6. The right eye after surgical formation of the pupil with preservation of the crystalline lens transparency. Good cosmetic results with preserved clear lens are observed.

It should also be noted that RFD was not developed for surgery on the iris, which has a different structure from the anterior capsule of the lens. The authors have described that RFD, on the first attempt, cannot always dissect the iris with fibrous membranes, and repeated resection is required, which may result in warming of the anterior chamber media with subsequent damage to the corneal endothelium. Moreover, the insufficient cutting ability of RFD forces authors to use scissors. In addition, RFD cannot completely eliminate bleeding from the iris vessels

during pupil formation. Thermal exposure to the iris tissue causes iridocyclitis and posterior synechiae, which may deform the newly formed pupil. Therefore, RFD can be used in combination with other cutting instruments (scissors, knife, vitreotome) for pupil shaping in case of secondary pupil deformities in the presence of aphakia, pseudophakia, uveitis, and cataracts [9–11]. However, it is hardly advisable to use RFD in congenital microcoria and acorea due to possible damage to the transparent lens, a risk that many authors [5,6], including us, have reported on.

A rare case of pupil dilation in microcoria has been reported by Hao et al. using iris scissors and hook after injecting a viscoelastic substance into the anterior and posterior chamber with preservation of a clear lens [6]. It should be noted that neither Xiage nor Hao detected a dense white membrane under the iris, which was identified in our surgical case [6,9].

However, Robb reported seven cases of microcoria and dyscoria with dense pupillary iris-crystalline membranes. During surgical enlargement of pupil size, Robb found a clear lens [12], which was also observed in our patient with acorea. Similar membranes have been previously described in 2 children with congenital pupillary - iris - lens membrane, which were unilateral and sporadic. These membranes, according to the author, differ from the usual pupillary membranes, which are fused together in the iris or attached by one side to the lens [13].

We used a newly created previously-formed peripheral iris coloboma through which we entered the retroiridian space and performed viscodissection of the iris from the lens; however, we detected a membrane fused to the iris on the pigment epithelium side. A similar approach was proposed by Alza14; however, the authors did not find any membrane when entering the retroiridian space through the already existing intrinsically displaced micropupil.

The dense white membrane we detected under the iris, constricting the upper half of the iris into rough folds and forming an acorea, was not visible on the iris surface. Additionally, it separated from the anterior capsule of the lens during vigorous viscodissection, which we performed "blindly" through a peripheral iridectomy. The membrane we detected is probably the persisting remnants of the unresolved tunica vasculosa lentis, which, when its regression failed, provoked the formation of acorea and caused complete blockage of the anterior chamber.

Thus, a novel advancement of this case is the presence of a dense white membrane under the iris, which was discovered during artificial pupil formation in congenital acorea. However, the presence of such a membrane has been previously described in microcoria [12,13], all available cases of pupil formation, when the pupil is absent, have not revealed such a membrane under the iris [4–7].

Delicate layer-by-layer dissection of the iris first and the revealed membrane then on the surface of the spatula allowed the preservation of the intactness of the lens anterior capsule and, therefore, lens transparency and accommodation, which is so important in childhood. One-stage opening of the iris and the membrane could cause injury to the crystalline lens, as it was performed blindly in the pupil's absence. The formation of an artificial pupil in acorea is recommended in early childhood to prevent amblyopia.

However, the surgical procedure can have certain limitations and complications both during and after surgery. Among the surgical risks of pupilloplasty is cataract development. Lens opacification may occur after any pupilloplasty because of irrigating fluid in the anterior chamber. Then, complications can occur when the anterior capsule is touched and damaged by cutting instruments (knife, scissors, vitreotome) due to insufficient care or lack of experience of a young surgeon. In addition, when using RFD for pupil formation, the increased temperature of the tip working on the iris can cause lens opacity because the shortest distance from the iris to the lens is in the pupil area, where the lens has the most significant curvature. When RFD needs to be reused, there is a risk of corneal endothelial damage due to overheating of the anterior chamber media [9].

Among the postoperative complications is the risk of severe iridocyclitis when RFD is thermally applied to the iris, which may lead to posterior synechiae and loss of round pupil shape [9]. In addition, secondary glaucoma in the postoperative period may develop in the second decade of life in the presence of unoperated congenital anterior chamber angle pathology combined with congenital acorea. Moreover, surgical intervention can only accelerate the development of secondary glaucoma [14].

The use of sufficient amount of viscoelastic material, minimal incisions, and collet instruments reduce the traumatic nature of the operation, and the risks of lens trauma and its subsequent opacity. Compared with pupilloplasty by radiofrequency diathermy, this approach reduces the incidence of iris inflammation and the risk of secondary glaucoma.

4. Conclusion

A unique case of congenital idiopathic acorea resulting from disruption of iris embryogenesis is presented. The complete absence of the pupillary aperture caused an acute attack of angle-closure secondary glaucoma two weeks after birth, which was controlled in a 1-month-old child by performing peripheral iridectomy and disconnecting the anterior iris-corneal synechiae.

During pupillary pinhole formation, a dense white membrane was found under the iris. The membrane was tightly fused with the iris, but separated from the anterior capsule of the crystalline lens during preliminary viscodissection. Dissection of the membrane and introduction of viscoelastic into the formed pinhole allowed the formation of a round pupil 3.0 mm in diameter with vitreal scissors and vitreotome and to maintain the intactness of the anterior capsule and transparency of the crystalline lens.

The artificial pupil exhibited a good cosmetic effect. Low visual acuity which can be explained by a long deprivation period that resulted in deep amblyopia and high axial myopia.

CRediT authorship contribution statement

BN: Concept, formal analysis, research, methodology, writing, reviewing and editing. RT: Formal analysis, data curation, research, writing, reviewing and editing. DO: Data curation, research, writing, reviewing and editing. All authors have read and approved the final manuscript.

Consent

Written informed consent was obtained from the patient's parents for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethical approval

All procedures were performed in accordance with the tenets of the Declaration of Helsinki and approved by the Ethics Committee.

Guarantor

Nadiia Bobrova.

Research registration number

N/A.

Funding

No funding or grant support.

Declaration of competing interest

None.

References

[1] C. Sohrabi, G. Mathew, N. Maria, A. Kerwan, T. Franchi, R.A. Agha, The SCARE 2023 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, Int. J. Surg. Lond. Engl. 109 (5) (2023) 1136.

[2] S. Duke-Elder, System of Ophthalmology, 1st ed., Henry Kimpton, London, 1964.

[3] H. Kondo, T. Tahira, K. Yamamoto, et al., Familial acorea, microphthalmia and cataract syndrome, Br. J. Ophthalmol. 97 (9) (2013) 1155–1160.

[4] P. Priyambada, Rajesh V. Prabu, et al., A rare case of acorea: congenital absence of pupil, J. Clin. Ophthalmol. Res. 9 (2) (2021) 85.

[5] S. Ramasubramanian, P.D. Majumder, Acorea: a rare congenital anomaly, Indian J. Ophthalmol. 66 (3) (2018) 450.

[6] L. Hao, Z. Ma, C. Song, et al., A rare case of congenital pupillary abnormality: a case report, BMC Ophthalmol. 22 (1) (2022) 201.

[7] M. Rahhal-Ortuño, M. Díaz-Llopis, L. Alonso-Muñoz, et al., An unusual case of acoria in Sturge–Weber syndrome, Eur. J. Ophthalmol. 30 (1) (2020) NP5–NP6, https://doi.org/10.1177/1120672118804795.

[8] C. Angée, B. Nedelec, E. Erjavec, et al., Congenital microcoria: clinical features and molecular genetics, Genes (Basel) 12 (5) (2021) 624.

[9] W. Xiang, X. Zhong, H. Chen, et al., Pupilloplasty by radiofrequency diathermy, Acta Ophthalmol. 97 (3) (2019) e479–e481.

[10] R. Klöti, Bipolar-Nassfeld-Diathermie in der Mikrochirurgie [Bipolar wet-field diathermy in microsurgery], Klin. Monatsbl. Augenheilkd. 184 (5) (May 1984) 442–444 (German).

[11] F. Gassmann, B. Schimmelpfennig, R. Klöti, Anterior capsulotomy by means of bipolar radio-frequency endodiathermy, J Cataract Refract Surg 14 (6) (Nov 1988) 673–676.

[12] R.M. Robb, Fibrous congenital iris membranes with pupillary distortion, Trans. Am. Ophthalmol. Soc. 99 (2001) 45–50 (discussion 50-1).

[13] G.W. Cibis, D.S. Walton, Congenital pupillary-iris-lens membrane with goniodysgenesis, J. AAPOS 8 (4) (2004) 378–383.

[14] A.G. Alza, Case report: congenital pseudoacorea in an ocular Axenfeld-Rieger syndrome: what is it? J. Clin. Cases Rep. 6 (4) (Oct 2023) 173–186, https://doi.org/10.46619/joccr.2023.6-1145 (Epub 2023 Mar 28).