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Case Report

Two-Step Iridocorneal Endothelial Syndrome Management: Endocapsular Intraocular Lens and Artificial Iris Followed by Descemet's Stripping Automated Endothelial Keratoplasty

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Keywords
Iridocorneal endothelial syndrome · Descemet stripping endothelial keratoplasty · Corneal edema · Artificial iris · Case report

Abstract
A 54-year-old female presented with complaints of glare and progressive visual loss OS with a corrected distance visual acuity (CDVA) OS of 20/100. The patient had grade 1 corneal edema with a "beaten bronze" appearance on specularly reflected light, pseudopolycoria, and a nuclear sclerotic cataract. The diagnosis of nuclear cataract and progressive iris atrophy iridocorneal endothelial (ICE) syndrome was made, and the patient underwent uneventful phacoemulsification with capsular bag placement of an AcrySof SA60AT intraocular lens combined with pseudopolycoria repair using an endocapsular Model A REPER artificial iris. Six months later, the patient was submitted to a Descemet's stripping automated endothelial keratoplasty (DSAEK) procedure, and 6 months after that the CDVA was 20/32 with no corneal edema and normal intraocular pressure. This two-step surgical approach, combining phacoemulsification and endocapsular foldable iris prosthesis placement followed by DSAEK, may be considered a promising option to successfully treat progressive iris atrophy ICE syndrome patients.

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Introduction

Iridocorneal endothelial syndrome (ICE) is a predominantly unilateral disorder of the corneal endothelium characterized by proliferative and structural abnormalities of the corneal endothelium, progressive obstruction of the iridocorneal angle and iris anomalies such as atrophy, corectopia, and polycoria [1]. Patients present with varying degrees of visual dysfunction arising from corneal decompensation, iris anomalies, and secondary glaucoma [2].

Management of ICE syndrome is aimed at controlling IOP, treating corneal hypotransparency, and limiting unwanted optical phenomena arising from iris defects [3]. In ICE syndrome, penetrating keratoplasty (PK) and Descemet's stripping automated endothelial keratoplasty (DSAEK) have similar graft survival rates; however, when compared to other corneal conditions submitted to keratoplasty, both demonstrate a higher endothelial cell loss rate, shorter graft survival, and higher rejection rates. Several factors explain the worst graft survival rates in ICE syndrome: higher incidence of glaucoma and glaucoma surgical interventions, abnormal anatomy of the anterior chamber and iris, and shallower anterior chamber depth [4–6]. In ICE syndrome, iris tissue is typically friable and poses a challenge to suture repair and, as such, reconstruction is often only possible with artificial iris implantation [7, 8].

The herein case report describes the surgical management of an ICE syndrome, progressive iris atrophy type, patient with cataract, pseudopolycoria, and corneal decompensation using a two-step approach: phacoemulsification surgery with placement of an AcrySof SA60AT intraocular lens (IOL) and a foldable endocapsular artificial iris (REPER Model A, Ophtec) followed by a DSAEK procedure, 6 months later. The CARE Checklist has been completed by the authors for this case report and is attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000534277).

Case Report

A 54-year-old female was referred to our department with complaints of glare and progressive loss of visual acuity from her left eye (OS) over a 3-year period. There were no associated systemic symptoms or previous relevant ophthalmic history. On examination, the patient's corrected distance visual acuity (CDVA) was 20/20 OR and 20/100 OS with a refractive error of −6.00 D and −5.25 D on the right and left eye, respectively. Intraocular pressure (IOP) was normal in both eyes.

Slit lamp examination of the left eye revealed a hazy cornea with a "beaten bronze" appearance when viewed in specularly reflected light, pseudopolycoria, with 3 peripheric holes at 2, 7, and 9 h (Fig. 1a); and a nuclear sclerotic cataract. Gonioscopy OS showed several areas of stromal iris atrophy, along with iris holes extending to the iris root and peripheral synechiae at various locations. Left eye fundoscopy was normal, including the disc which had sharply demarcated borders, a cup-to-disk ratio of 0.4, and no signs of glaucomatous damage. The complete right eye observation was unremarkable.

Automatic endothelial specular microscopy OR revealed a normal endothelium with 2,700 cell/mm^2, while the OS revealed rounded, dark ICE cells with light borders (dark/light reversal), pathognomonic of ICE syndrome [9–11] (Fig. 2a). Cell count using automatic endothelial specular microscopy in OS was not possible. Anterior chamber depth was 3.18 mm^3, the anterior chamber angle measured 30.9°, and pachymetry was 545 μm. OCT measured retinal nerve fiber layer thickness revealed a global thickness of 115 μm (Fig. 3). A diagnosis of nuclear cataract, polycoria, and corneal hypotransparency in the context of a progressive iris atrophy – ICE syndrome was made, and a two-step surgical procedure was planned.

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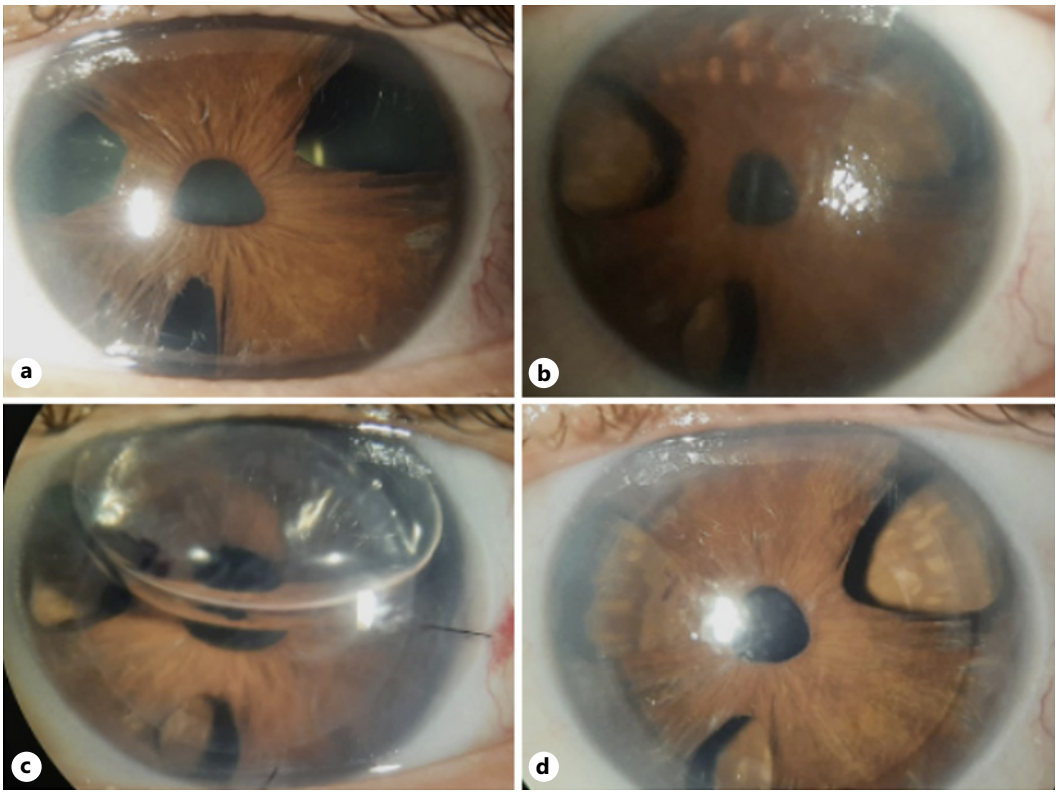

Fig. 1. Slit lamp photography. **a** OS with a nuclear cataract and progressive iris atrophy iridocorneal endothelial (ICE) syndrome. Grade 1 corneal edema and three atrophic peripheric iris holes are seen. **b** Grade 3 corneal edema 6 months post-cataract surgery and artificial iris placement. **c** Day one post-Descemet's stripping automated endothelial keratoplasty (DSAEK) surgery, showing a well-positioned graft and the residual air bubble. **d** Six months post-DSAEK with a clear cornea and the implanted iris prosthesis and intraocular lens (IOL) in their correct placement.

First, the patient underwent uneventful phacoemulsification with capsular bag IOL placement combined with pseudopolycoria repair using an endocapsular artificial iris. Surgery was carefully planned, and special attention was dedicated to the capsulorrhexis that was performed as large as possible to facilitate the introduction of the artificial iris within the capsular bag. The artificial iris was carefully folded in a roll and loaded into an IOL injector. Nevertheless, the iris prosthesis was larger than the capsulorrhexis, which made it difficult to introduce it directly into the capsular bag, so the artificial iris was cut radially and the two extremities were overlapped to allow placement inside the bag with the help of a forceps and lens hook. This maneuver maintained centration of the artificial iridial pupil. Postoperative treatment regimen included topical moxifloxacin, dexamethasone, and bromfenac in a tapering fashion (4 weeks), as well as hypertonic saline solution and aqueous suppressant agents in an attempt to mitigate possible postsurgical corneal decompensation which the patient was made aware of beforehand. At the 6-month follow-up appointment, a centered artificial iris and a correctly placed pseudophakic IOL were observed, together with grade 3 corneal edema; no visual acuity improvement was appreciated (Fig. 1b). The patient was then subjected to a DSAEK procedure: an ultra-thin endothelial lenticule (85 μm centrally) with 8.5 mm diameter was prepared and implanted in the eye after endothelium scraping using the Busin-glide technique.

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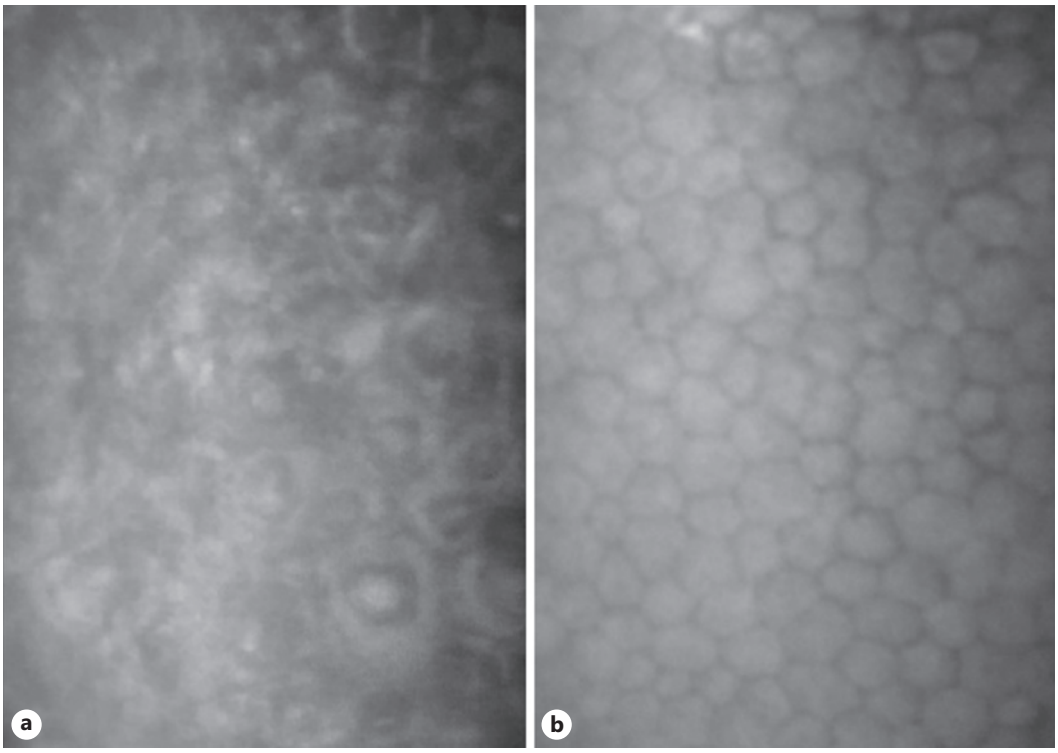

Fig. 2. Specular microscopy. **a** Corneal endothelium showing typical findings of ICE syndrome. **b** Normal endothelium morphology, with 1,640 cell/mm^2 count, 6 months after DSAEK.

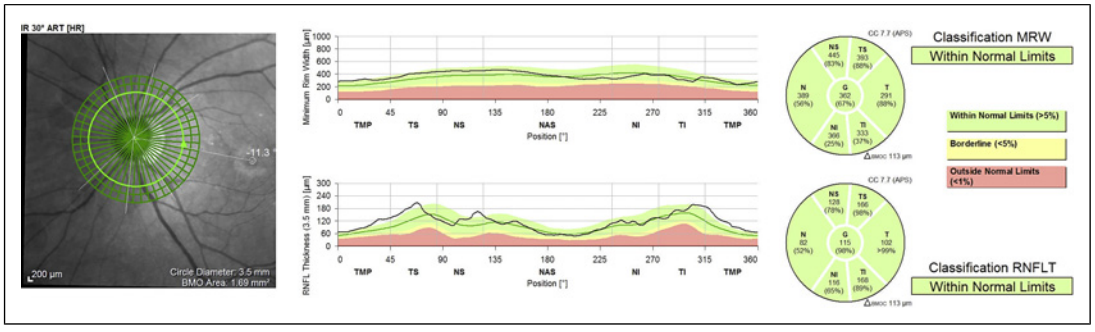

Fig. 3. OS SPECTRALIS SD-OCT retinal nerve fiber layer thickness (RNFLT) and minimum rim width (MRW) analysis showing no signs of glaucomatous damage, with RNFLT and MRW within the normal range of the normative values.

The anterior chamber was filled with air for lenticule attachment and the patient was instructed to lie in supine position for 24 h. At postoperative day 1, the lenticule was attached, the anterior chamber was formed and IOP was normal (Fig. 1c). The patient was treated with topical corticosteroid (tapered monthly), topical ofloxacin (2 weeks), and topical aqueous suppressant drops (4 weeks).

Six months post-DSAEK, CDVA OS improved to 20/32 (with −2.00D sph) and IOP was 13 mm Hg. Biomicroscopy revealed a clear cornea and a successfully attached endothelial graft. The implanted iris prosthesis and IOL were correctly positioned and centered, all iris

holes were covered, and the pupil presented only with slight corectopia (Fig. 1d, 3). Endothelial microscopy revealed a DSAEK with normal endothelium morphology and a 1,640 cell/mm^2 central count (Fig. 2b). Anterior chamber depth was 3,490 mm^3 and the anterior chamber angle predictably widened after the surgeries and measured 45.9°. In Figure 4, the anterior segment OCT examination (Anterion, Heidelberg, Germany) illustrates all structures mentioned in the case properly positioned and centered on the visual axis.

Discussion

The present case describes a novel surgical approach for the treatment of iris abnormalities and endothelium decompensation in progressive iris atrophy ICE syndrome by combining pseudophakic endocapsular iris prosthesis implantation followed by a DSAEK procedure. Patients with unilateral iris defects, as seen in progressive iris atrophy ICE syndrome, often complain of photophobia and glare and suffer from cosmetic disfigurement and optical abnormalities that result in serious visual impairment [8]. A permanent surgical alternative is the reconstruction with an artificial iris implant [7]. In this case, an endocapsular REPER Model A artificial iris was used. This model consists of a foldable acrylic artificial iris that can be selected from 300 existing designs to match the fellow eye's iris or be made custom. This resulted in aesthetically pleasing results and greatly improved vision with the correction of pseudopolycoria. In our opinion, the progressive nature of the disease advises the use of an artificial iris device implantation to repair the esthetic and visual defect in lieu of iris suture; the extensive manipulation of iris tissue could further induce uveal inflammation and progressive additional iris atrophy, IOP increase, and secondary glaucoma.

In this clinical report, 6 months after cataract surgery and artificial iris placement, there was a worsening of corneal edema possibly resulting from surgical decompensation of an already diseased endothelium. There is paucity of literature in regards to keratoplasty techniques in ICE syndrome. PK has been proposed as an option for surgical treatment of secondary corneal edema with varying degrees of success [12]. With the improvements in lamellar corneal transplants, DSAEK presents as an attractive alternative to more invasive PK. Due to the infrequency of ICE syndrome and the scarce amount of literature available, there is no demonstrated superiority of either DSAEK or PK in the management of these patients [12]. Nonetheless, DSAEK has been proven to successfully treat corneal edema and visual loss in ICE syndrome with rapid and better visual rehabilitation and reduced astigmatic effect [13, 14], but performing lamellar endothelial transplant in eyes with ICE is challenging due to synechiae formation, severe iris atrophy, and anterior displacement of the lens-iris diaphragm, that can make lenticule insertion and air tamponade more challenging [13, 15]. These hurdles are partially, if not completely, overcome by iris diaphragm reconstruction in a first surgical stage, that allows for a safer barrier between the anterior and posterior chambers, facilitating the lamellar endothelial transplant attachment.

In our case, no signs of glaucomatous damage were observed during the follow-up period. Although gonioscopy revealed peripheral synechiae at various locations, IOP was consistently normal at repeat measurements, retinal nerve fiber layer thickness was stable, and fundus observations, including optic disc, were consistently within normal limits during the follow-up visits. These findings, led us to conclude that there was no immediate need for glaucoma surgery. However, the authors acknowledge that long-term follow-up is crucial for assessing the potential development of glaucoma in ICE syndrome patients, especially in the present case, as it may also impact the longevity of the endothelial transplant. We plan to continue monitoring the patient closely to detect any future changes in IOP or optic nerve damage, and will remain vigilant for the need of glaucoma interventions if warranted.

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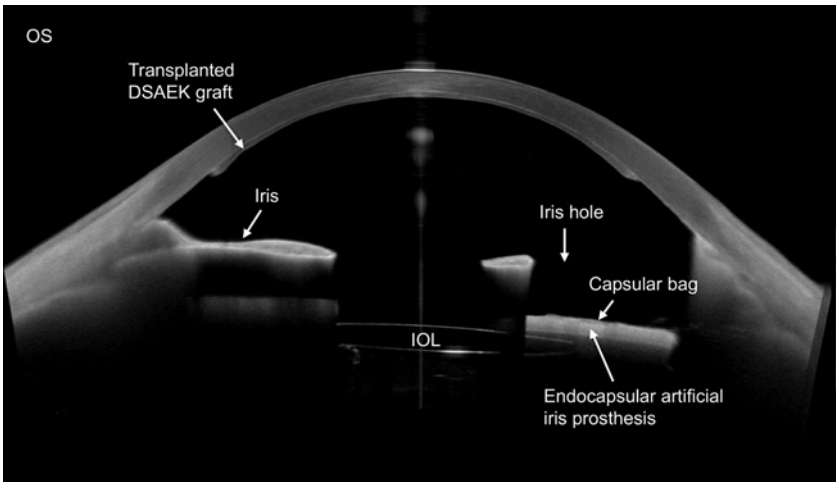

Fig. 4. Labeled anterior segment OCT image of OS post phacoemulsification with IOL placement, endocapsular artificial iris placement, and DSAEK surgery. DSAEK, Descemet's stripping with endothelial keratoplasty.

In addition, even though at the time of submission the patients' CDVA was greatly improved, long-term graft survival in these cases is uncertain as the potential for glaucoma with raised IOP in this patient population seems to have an impact on graft survival rates [12]. Nonetheless, the authors feel that DSAEK is still a better choice than PK in the present case. The sequential approach shown in this case report possibly allows for a greater success rate in an otherwise complicated lamellar endothelial transplant procedure in ICE patients with iris deformities presenting as surgical obstacles for successful graft placement.

Although this sequential surgical approach seems promising, it should be pointed out that this is a single case report and that these findings need to be confirmed by other studies ideally of a prospective nature.

Statement of Ethics

This retrospective review of patient data did not require ethical approval in accordance with local/national guidelines. This published case report complies with the guidelines for human studies in accordance with the World Medical Association Declaration of Helsinki. The patient provided written informed consent for publication, including publication of images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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No funding or grant support was received for publication of this clinical case.

Author Contributions

All authors attest that they meet the current ICMJE criteria for authorship. João Pinheiro-Costa and Manuel Falcão were the attending physicians overseeing the care of the patient described in the case, aided in the diagnosis, had direct patient contact, were a principal surgeons, were in charge of patient follow-up, made substantial contributions to the conception of the report and the acquisition, analysis, and interpretation of data, participated in the critical revision for important intellectual content, gave the final approval of the version to be published, and agreed to be accountable for the appropriate investigation regarding the accuracy and integrity of the report. Inês Coelho-Costa wrote the largest share of the report, made substantial contributions to the conception of the report and the acquisition, analysis, and interpretation of data, participated in the critical revision for important intellectual content, gave the final approval of the version to be published, and agreed to be accountable for the appropriate investigation regarding the accuracy and integrity of the report. Tiago Monteiro had direct patient contact, aided in the diagnosis, was a principal surgeon, was in charge of patient follow-up, made substantial contributions to the conception of the report and the acquisition, analysis, and interpretation of data, participated in the critical revision for important intellectual content, gave the final approval of the version to be published, and agreed to be accountable for the appropriate investigation regarding the accuracy and integrity of the report. Fernando Falcão-Reis made substantial contributions to the conception of the report and the acquisition, analysis, and interpretation of data, participated in the critical revision for important intellectual content, gave the final approval of the version to be published, and agreed to be accountable for the appropriate investigation regarding the accuracy and integrity of the report.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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