



# Ex-PRESS shunt combined with ab-interno peripheral iridectomy: A new surgical procedure for iridocorneal endothelial syndrome

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## ABSTRACT

**Purpose:** Iridocorneal endothelial (ICE) syndrome may cause refractory glaucoma due to progressive synechial closure or membrane formation at the anterior chamber angle. Filtration surgeries are often required but are associated with a higher rate of surgical failure or complications than other types of glaucoma. Herein, we report a new and effective surgical procedure for glaucoma secondary to ICE syndrome: Ex-PRESS shunt combined with ab-interno peripheral iridectomy using a small-gauge vitreous cutter.

**Methods:** Three patients with ICE syndrome who underwent surgery were included. Intraoperatively, an ab-interno peripheral iridectomy was performed using a small-gauge vitreous cutter through a corneal incision in the superior-nasal or superior-temporal quadrants to create space for the insertion of Ex-PRESS shunt free from the iris tissue. The shunt was inserted under the scleral flap. The first patient underwent combined cataract surgery, whereas patients 2 (pseudophakia) and 3 (phakia) underwent Ex-PRESS alone.

**Results:** No intraoperative complications were observed. The intraocular pressure remained stable until the final postoperative visits at approximately 7, 4, and 1 year in Cases 1, 2, and 3, respectively. Case 2, with mild preoperative corneal edema due to graft failure in Descemet's stripping automated endothelial keratoplasty (DSAEK), underwent reDSAEK 6 months postoperatively.

**Conclusions and importance:** Ex-PRESS shunt combined with ab-interno peripheral iridectomy using a small-gauge vitreous cutter may be a safe and effective surgical procedure in patients with ICE syndrome, regardless of the lens status.

## 1. Introduction

Iridocorneal endothelial (ICE) syndrome is a unilateral, non-hereditary, and progressive disease that is classified into Chandler syndrome, Cogan-Reese syndrome, and progressive iris atrophy. It causes refractory glaucoma because of abnormal corneal endothelial cells, which are responsible for peripheral anterior synechia (PAS), pupillary deviation, iris atrophy, and iris nodules, by producing membrane-like materials on the surface of the anterior chamber angle and iris.<sup>1-3</sup> Although trabeculectomy<sup>4-8</sup> and tube shunt surgery<sup>9,10</sup> are indicated as surgical treatments for ICE syndrome, surgical outcomes are relatively poor, and postoperative complications are common. The Ex-PRESS shunt, which does not require peripheral iridectomy, has been used in some patients with the ICE syndrome.<sup>11-13</sup> However, it is difficult to insert a shunt in the presence of high PAS. Ab-interno iridectomy with a vitreous cutter has been reported to be useful for treating malignant

glaucoma,<sup>14</sup> iris biopsy,<sup>15</sup> and iris incarceration after deep sclerectomy.<sup>16</sup> A 25- or 27-gauge vitreous cutter is considered less invasive than a larger gauge cutter. Herein, we report an Ex-PRESS shunt combined with ab-interno peripheral iridectomy using a small-gauge vitreous cutter as a new and effective surgical technique for ICE syndrome.

## 2. Materials and methods

This study was approved by the Medical Ethics Committee of Kanazawa University Hospital and followed the tenets of the Declaration of Helsinki. Informed consent was obtained from all patients. A total of 3 patients with ICE syndrome who underwent an Ex-PRESS shunt combined with ab-interno peripheral iridectomy using a small-gauge vitreous cutter were included in this study. Medical records were collected and analyzed.

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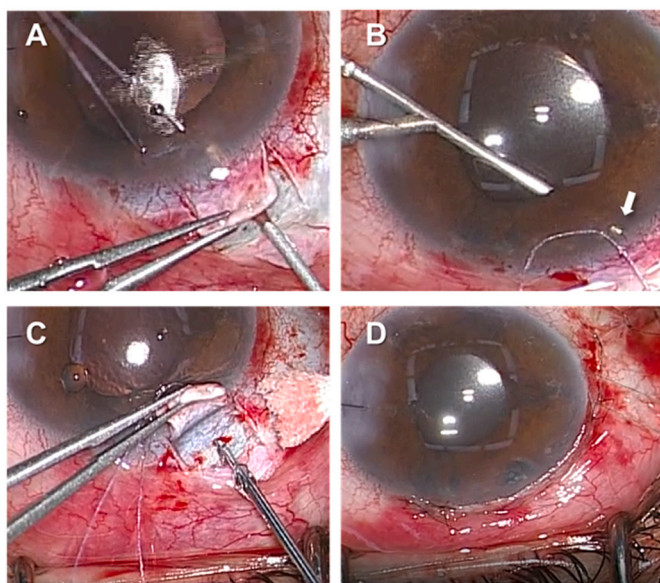
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## 2.1. Surgical Technique

After sub-tenon anesthesia, a fornix-based conjunctival incision was made in the superior-nasal or superior-temporal quadrants, followed by the creation of a 3.5 × 3.5 mm square half-thickness scleral flap. Mitomycin C (0.4 mg/ml)-soaked sponges were applied to the sub-Tenon's space for 4 min and then washed with saline. In Case 1, cataract surgery using a temporal clear corneal incision was performed. Miosis was induced by an intracameral injection of acetylcholine chloride. An incision with a 25-gauge needle confirming the presence of iris tissue interfering with the needle path was made under the scleral flap to prepare for Ex-PRESS implantation (model P50, Alcon Laboratories, Fort Worth, TX, USA) in the anterior chamber (AC) angle (Fig. 1A). Subsequently, peripheral iridectomy was performed at the center of the scleral flap using a 25- or 27-gauge vitreous cutter (cutting rate, 2500 cpm; aspiration, 300 mmHg) through a corneal incision (Fig. 1B). An AC maintainer was used for irrigation. Ex-PRESS shunt was then inserted under the scleral flap in a regular fashion (Fig. 1C). The scleral flap was sutured using 10-0 nylon sutures. The conjunctival flap was closed at the limbus using 10-0 nylon sutures (Fig. 1D).

## 3. Results

The clinical courses of the three patients are shown in Table 1. Preoperatively, two eyes were diagnosed with progressive iris atrophy and one with Cogan-Reese syndrome (Fig. 2A–C). In all cases, the posterior trabecular meshwork at the superior angle was invisible gonioscopically owing to a broad high PAS. The patient with Cogan-Reese syndrome (Case 2) with a history of filtration surgery and Descemet's stripping automated endothelial keratoplasty (DSAEK) was pseudophakic and had poor visual acuity due to bullous keratopathy. Intraoperatively, there were no complications, such as marked bleeding or lens injury in the phakic eye. An Ex-PRESS shunt was implanted without difficulty after iridectomy. Postoperatively, intraocular pressure (IOP) remained controlled without medication until the last visit, although two patients required bleb needling during the follow-up period (Fig. 2D–I). Case 2 underwent reDSAEK six months postoperatively because of bullous keratopathy that persisted from the preoperative period. The corneal endothelial cell density was reduced by 31% (4.6%/year) in Case 1 and remained stable in Case 3.



**Fig. 1.** Surgical procedure  
The tip of vitreous cutter was visible through the iridectomy window (Arrow in B).

**Table 1**

Clinical course of three patients with ICE syndrome.

|   | Case 1                   | Case 2   | Case 3                   |
|---|--------------------------|--|--------------------------|
| Age at Ex-PRESS surgery (years)           | 51                       | 51   | 23                       |
| Sex                                       | Male                     | Female   | Female                   |
| Affected eye                              | Left                     | Left   | Right                    |
| Diagnosis                                 | Progressive iris atrophy | Cogan-Reese syndrome                                       | Progressive iris atrophy |
| Surgeries before Ex-PRESS surgery         | None                     | Trabeculectomy, bleb revision with cataract surgery, DSAEK | None                     |
| Preoperative VA (logMAR)                  | 0                        | 1.30   | 0.52                     |
| Preoperative IOP (mmHg)                   | 37                       | 24   | 45                       |
| Preoperative number of medications        | 6                        | 4  | 5                        |
| Preoperative ECD (cells/mm <sup>2</sup> ) | 1430                     | 745  | 851                      |
| Postoperative additional procedures       | Bleb needling (3Y 2 M)   | reDSAEK (6 M)  | Bleb needling (6 M)      |
| Most recent VA (logMAR)                   | 0                        | 0.15   | 0                        |
| Most recent IOP (mmHg)                    | 6                        | 9  | 10                       |
| Most recent number of medications         | 0                        | 0  | 0                        |
| Most recent ECD (cells/mm <sup>2</sup> )  | 983                      | 625  | 1055                     |
| Follow-up periods after Ex-PRESS surgery  | 6Y 10 M                  | 4Y   | 1Y 4 M                   |

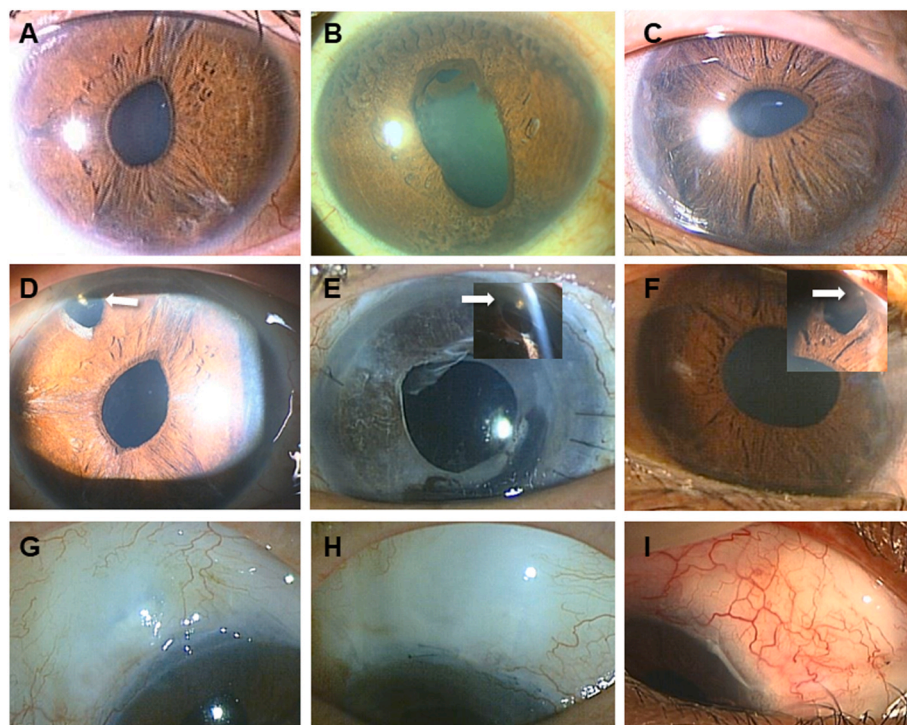
ICE = Iridocorneal endothelial, VA = visual acuity, logMAR = logarithm of the Minimum Angle of Resolution, IOP = intraocular pressure, ECD = corneal endothelial cell density, DSAEK = Descemet's stripping automated endothelial keratoplasty, Y = years, M = months.

## 4. Discussion

ICE syndrome causes refractory glaucoma, which often requires multiple glaucoma surgeries, including trabeculectomy and tube shunt surgeries.<sup>4–10</sup> Success rates for trabeculectomy have been reported as 60–79% at 1 year<sup>1,2,4,6</sup> and 21–33% at 5 years,<sup>1,2,6</sup> and for tube shunt surgery as 71–94% at 1 year and 53–73% at 3–5 years postoperatively.<sup>6,9,10</sup> Aggressive scarring at the filtration site due to young age<sup>1</sup> and the proliferation of abnormal corneal endothelial cells that close the trabeculectomy window may be the reasons for poor IOP control after trabeculectomy in ICE syndrome.<sup>5</sup>

Furthermore, the membrane of proliferating endothelial cells (i.e., the ICE membrane) or the iris may block the tube ostium, resulting in failure of tube shunt surgery.<sup>5,6</sup> Recently, a case series of four eyes with a median follow-up of 14 months showed that the insertion of a nonvalved glaucoma drainage device in the ciliary sulcus was useful for ICE syndrome.<sup>17</sup> Tube insertion in the sulcus was not affected by high PAS scores, and the tube ostium may be less likely to be blocked than AC insertion. However, this procedure is not suitable for clear phakic eyes because of the risk of cataract development.

There have been three reports of four patients with ICE syndrome who underwent an Ex-PRESS shunt.<sup>11–13</sup> Ex-PRESS shunt surgeries have advantages over tube shunt surgeries, including low postoperative inflammation and the absence of plate-related complications such as diplopia. In the reported cases, an Ex-PRESS shunt was inserted at the site without significant PAS. Despite the fact that one patient required bleb revision one month postoperatively, the final IOPs were 9 and 14 mmHg at 6 months, 15 mmHg at 15 months, and 15 mmHg at 3 years, with or without antiglaucoma medication.<sup>11–13</sup> Conversely, our three patients had a broad and high PAS at the superior angle, which



**Fig. 2.** Clinical appearance of three patients at presentation and at the most recent visit

A–C, At presentation; D–I, at the final visit; A, D, G = Case 1; B, E, H = Case 2; C, F, I = Case 3. Arrows indicate the tip of the Ex-PRESS shunt visible at the iridectomy sites. Well-formed filtration blebs in each case (G–I).

prevented the insertion of the Ex-PRESS shunt in front of the iris without the shunt being positioned close to the corneal endothelium. Owing to the iris-free space created by the ab-interno iridectomy, we successfully implanted Ex-PRESS shunt at the optimal site (i.e., the superior-nasal or superior-temporal quadrants) in a regular fashion. In addition, the iris-free space, which remained unchanged until the last follow-up visit, may have contributed to preventing closure of the tip of the Ex-PRESS shunt by the ICE membrane or the iris. We used a 25- or 27-gauge vitreous cutter for the iridectomy. These small-gauge cutters are less invasive and allow us to finely resect tissues, which was useful even in the phakic eye of Case 3, with no cataract formation postoperatively. Furthermore, the postoperative IOPs in our three cases remained low without antiglaucoma medication during their follow-up period which ranged from 1 to 7 years, indicating the usefulness of our procedure with an Ex-PRESS shunt for ICE syndrome.

The corneal endothelial cell density decreased by 31% in Case 1. However, the rate of cell loss (4.6%/year) is comparable to that after trabeculectomy in patients with primary open angle glaucoma.<sup>18,19</sup> The corneal decompensation is one of common features in the ICE syndrome, which was seen in Case 2. Although the corneal endothelial cell density remained stable in Case 3, long-term follow-up in more cases is necessary to determine whether our surgical procedure is safer to the corneal endothelial cells than other filtration surgeries.

The IOP at the last follow-up visit was  $\leq 10$  mmHg in all cases. We continued topical administration of betamethasone to suppress bleb scarring in all patients; this may have contributed to the favorable IOP outcome. However, Case 1 required bleb needling approximately 3 years postoperatively. It should be noted that the problem of bleb scarring derived from the young age of the patient and the postsurgical inflammation in ICE syndrome cannot be solved by our technique.

## 5. Conclusions

Regardless of lens status, Ex-PRESS shunt combined with ab-interno peripheral iridectomy using a small-gauge vitreous cutter may be a safe

and effective surgical procedure in patients with ICE syndrome. Given the limited data with three cases in the current study, more studies and longer follow-ups are needed.

## Patient consent

The patients consented to publication of the case in writing. This report does not contain any personal information that could lead to the identification of the patient.

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## Declaration of competing interest

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