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Clinical and Specular Microscopic Manifestations of Iridocorneal Endothelial Syndrome

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Purpose: To investigate the correlation between the clinical pictures and the specular microscopic findings in patients with iridocorneal endothelial (ICE) syndrome.

Methods: The records of 15 patients with ICE syndrome who presented at the National Taiwan University Hospital between 1993 and 1996 were examined. The medical history, clinical pictures of the cornea, iris and anterior chamber angle, intraocular pressure, specular microscopic findings, and the correlation between clinical and specular microscopic findings were assessed.

Results: Endothelial changes in specular micrographs were found in all the patients, even in those patients with minimal angle involvement by peripheral anterior synechiae. Corneal decompensation resulting in corneal edema and bullae formation was the main cause of visual impairment. Neither ICE grading nor endothelial cell density correlated with corneal edema or intraocular pressure, but they correlated with the angle involvement in ICE syndrome. The intraocular pressure was difficult to control in 8 of these patients, even after treatment with anti-glaucoma agents and trabeculectomy, especially in the patients with Cogan-Reese syndrome.

Conclusion: Although specular microscopy provides an invaluable method for the diagnosis of ICE syndrome, it is not a reliable tool for predicting prognosis. Close follow-up of intraocular pressure and early detection of glaucoma are important steps to preserve visual functions in patients with ICE syndrome. **Jpn J Ophthalmol 2001;45:281–287** © 2001 Japanese Ophthalmological Society

Key Words: Clinical pictures, corneal endothelium, glaucoma, iridocorneal endothelial syndrome, specular microscopy.

Introduction

The iridocorneal endothelial (ICE) syndrome, which includes progressive essential iris atrophy, Chandler's syndrome, and the iris nevus (Cogan-Reese) syndrome, is characterized by abnormalities of the cornea, anterior chamber angle, and iris.[1,2] Some studies have shown the abnormalities in the corneal endothelium as a characteristic hammered-silver appearance by slit-lamp biomicroscopy and an image reversal pattern of the endothelium by specular microscopy.[3–7] Previous histopathological studies found that the endothelial cells underwent epithelial changes including desmosomal junctions, surface microvilli, and increased intracytoplasmic filaments.[3,4,7] These endothelial changes can lead to corneal edema and growth of a membrane onto the iris. Contraction of the membrane may cause peripheral anterior synechiae with secondary glaucoma and various changes in the iris.[8–10] Ultrastructural studies of cases with advanced corneal edema show scant, abnormal corneal endothelial cells lining a thickened, multilayered Descemet's membrane.[3–7]

The prognosis of ICE syndrome is dependent on the severity of corneal involvement and the presence of secondary glaucoma.[1] Because specular microscopy can visualize corneal endothelial cells in vivo, it can be used to confirm the clinical diagnosis of ICE syndrome and to determine the involvement of ICE cells in the corneal endothelium.[11–16] However, its value in predicting the prognosis of ICE syndrome is not determined. Whether the degrees of corneal in-

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volvement by ICE cells correlate with corneal edema and intraocular pressure (IOP) is an issue worth investigating. In this study, we correlated the clinical pictures of ICE syndrome with the findings of specular microscopy.

Materials and Methods

At the National Taiwan University Hospital, 15 patients were diagnosed clinically and confirmed by specular microscopy to have ICE syndrome, ie, presence of ICE cells, from 1993 through 1996. There were 11 cases of Chandler's syndrome, 1 of progressive essential iris atrophy and 3 of iris-nevus (Cogan-Reese) syndrome. All 15 patients included in this study shared one or more of the common features of ICE syndrome: corneal abnormalities, broad peripheral anterior synechiae, and iris abnormalities. There were 3 men and 12 women whose average age was 54 years (range. 32–72 years of age). The clinical diagnosis followed the diagnostic criteria proposed by Shields[1] including abnormal endothelial cells with variable degree of corneal edema, secondary glaucoma due to progressive peripheral anterior synechiae, and iris changes. Visual acuity, findings of slit-lamp examination of the anterior segment, visual fields, IOP measured by applanation tonometry, findings of gonioscopic examination, and fundus examination were reviewed retrospectively in the patient records. A mean defect greater than 5 dB in Octopus automated perimetry (Octopus 500 or 2000R, program 32/34 or G1) was considered to be a significant visual field defect.

To evaluate the endothelial changes in the affected corneas, a Konan SP-5500 wide-field specular microscope (Konan, Hyogo) was used to obtain the endothelial picture. The specular micrographs were examined for both the affected and unaffected eyes. Each cornea was scanned at three areas: superior, middle, and inferior parts of the corneal endothelial layer. The corneal scanning was recorded by a video recorder system and analyzed by Bambi Video Analysis System (Bio-Optics, Arlington, MA, USA) for cell density counting. Two methods were used to count endothelial density: (1) a fixed-frame method for endothelium with more clearly defined cell mosaic or smaller cells; and (2) a variable-frame method for endothelium with poorly defined cell mosaic or larger, irregular cells. The mean endothelial density was obtained by averaging upper, middle, and lower readings of corneal endothelial density. Using double-blind conditions, specular micrographs were reviewed by two corneal specialists; then the characteristics of ICE cells common to the affected endothelia were described and recorded.

For the morphological changes in endothelial tissue, the Hirst grading system from 1 to 3 was adopted; and the Sherrard ICE tissue classification system was also used for the affected corneas.[11,12] In the Hirst grading system, grade 1 (minimally affected cornea) showed early changes, including rounding of the endothelial cells, ie, loss of hexagonal shape, increased percentage of pentagonal cells, and greater granularity of individual cells. Increased polymorphism with kite-shaped cells and enlargement of the blackout areas within the cells were noted in grade 2 (moderately affected corneas). In grade 3 (markedly affected cornea), the endothelium layer was no longer recognizable as a mosaic of cells; the blackout areas within cells abutted each other.[11]

Another classification proposed by Sherrard divided ICE tissues into four types: (1) disseminated ICE, in which the endothelium appears normal but it is difficult to focus on the individual cells and ICE cells are scattered throughout; (2) total ICE, in which the entire corneal endothelium is replaced with ICE tissue; (3) subtotal ICE (+), where a variable portion of the endothelium is replaced and the surviving endothelium is composed of very small cells; and (4) subtotal ICE (−), where only a portion of the endothelium bears ICE tissue but the surviving endothelium is composed of enlarged cells.[12] Endothelial cell densities were calculated by the following method: (1) if the ICE cell grading was grade 1 or grade 2, the mean endothelial density was an average of the endothelial cell densities of both the surviving cells and the ICE cells; (2) if the ICE cell grading was grade 3, only the large or small surviving cells were chosen for the endothelial cell density calculation because the involved endothelial layer was no longer recognizable as a mosaic of cells.

The correlation between specular findings (endothelial cell density, ICE cell grading) and IOP, cornea edema, or visual acuity was calculated by Pearson's correlation coefficient, and a significant correlation was designated as a $P < .05$.

Results

Symptoms and Ocular Findings

The clinical data of all 15 patients are summarized in Table 1 and Table 2. In 11 of the 15 patients, visual disturbance was the initial symptom due to corneal edema, cataract, or glaucomatous optic nerve damage. The other 4 patients had nonspecific complaints during the first visit. A finely hammered-sil-

Table 1. Clinical Data in PAtients with Iridocorneal Endothelial Syndrome

Patient No.	Age/Sex	Diagnosis	Initial Symptoms	Initial VA (od/os)	IOP (mm Hg) (od/os)	Corneal Findings	Iris and Pupil Findings	Angle Involved (%)
1	58/F	Chandler's (OD)	BV	0.3/0.8	28/18	HS, bullae edema	Mild corectopia and ectropion uveae	50
2	72/F	Chandler's (OD)	BV	0.2/0.3	14/12	HS, edema	Mild corectopia and ectropion uveae	25
3	44/F	Chandler's (OU)	BV	1.0/0.3	12/12	HS, guttae edema	Mild corectopia and ectropion uveae	50
4	52/F	Chandler's (OD)	BV	0.6/1.0	26/18	mild opacity, HS	Mild corectopia	75
5	59/F	Chandler's (OS)	BV	0.8/0.3	9/39	edema, DM folding, KP, HS	Mild corectopia	50
6	61/F	Chandler's (OS)	BV	0.7/0.5	12/13	mild opacity, KP, HS	Mild corectopia and iridodialysis	25
7	57/F	Chandler's (OD)	tearing	0.8/0.8	14/12	guttae, HS	Mild corectopia	25
8	65/F	Chandler's (OD)	FS	0.5/0.5	42/16	guttae, HS	No abnormailty	10
9	53/F	Chandler's (OS)	none	0.9/0.7	12/15	HS	Mild corectopia and ectropion uveae	25
10	60/M	Chandler's (OD)	BV FS	0.5/1.2	17/13	HS	Mild iris atrophy	75
11	62/M	Chandler's (OS)	BV	0.1/0.2	14/16	HS, Bullae, edema	Mild corectopia	75
12	50/F	Cogan-Reese (OD)	BV	0.3/1.0	30/15	HS, DM thickening	Iris nevi	25
13	32/F	Cogan-Reese (OS)	BV	0.9/0.3	15/40	HS, guttae edema	Iris nevi	50
14	33/F	Cogan-Reese (OS)	FS	1.0/0.9	14/26	mild opacity, HS	Iris nevi corectopia	25
15	43/M	Essential Iris Atrophy (OS)	BV	1.2/0.2	21/17	Bullae, edema, KP, HS	Iris atrophy corectopia	95

OD: right eye, OS: left eye, OU: both eyes, VA: visual acuity, BV: blurred vision, FS: fullness sensation, KP: keratic precipitates, DM: Descemet's membrane, VF: visual field, PKP: penetrating keratoplasty, HS: hammered-silver appearance, IOP: intraocular pressure at referral.

ver appearance on all or part of the posterior corneal surface examined using the slit-lamp biomicroscope with specularly reflected light was present in all 15 patients; of these, 7 had corneal edema. Three patients (patients 1, 11, and 15) had corneal bullae formation, and received penetrating keratoplasty for vision improvement and pain relief.

Iris abnormalities were found in 14 of the 15 patients. The abnormalities included subtle stromal atrophy (as in Chandler's syndrome), full thickness iris stromal atrophy and/or stretch holes (as in essential iris atrophy), and formation of iris nodules on the anterior iris surface (as in iris nevus syndrome). Peripheral anterior synechiae occurred in every patient and was often accompanied by corectopia and ectropion uveae except in patient 8, who had minimal angle involvement and almost no iris abnormality. Nevertheless, patient 8 had a hammered-silver appearance on the corneal endothelium and the presence of ICE cells. Patient 15 had the most severe iris changes, including iris hole formation and atrophy, which were compatible with the diagnosis of essential iris atrophy. Clusters of pigmented nodules were found over the surface of the iris in the 3 patients (patients 12,13, and 14) with iris-nevus (Cogan-Reese) syndrome.

Elevated IOP was found in 7 of the 15 patients at the first visit, and glaucoma medications were administered to 10 patients. Of these, 6 had significant visual field defects. Patient 11 underwent a trabecu-

Table 2. Treatment Data on Iridocorneal Endothelial Syndrome

Patient No.	Treatment	Visual Field Defects
1	2% Pilocarpine, PKP	None
2	None	None
3	2% Carteolol	None
4	0.5% Timolol	None
5	2% Pilocarpine, 0.5% timolol	(+)
6	None	Not done
7	None	Not done
8	2% Pilocarpine, 0.5% timolol	None
9	None	None
10	2% Pilocarpine	None
11	PKP, TRA	(+)
12	0.5% Timolol, TRA, LI, TRP	(+)
13	TRA, 0.5% betaxolol, 2% pilocarpine	(+)
14	TRA, 2% pilocarpine, 0.5% betaxolol	(+)
15	PKP, 0.25% timolol	(+)

PKP: penetrating keratoplasty, TRA: trabeculectomy, LI: laser iridotomy, TRP: laser trabeculoplasty, none: no significant visual field defect, +: with significant visual field defect, not done: VF test was not performed.

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letomy before referral. During the follow-up examinations (from 14 months to 56 months, mean = 42 months), IOP was within normal limits without medication in 4 patients (patients 2, 6, 7, and 9). Two patients (patients 3 and 10) had satisfactory IOP control with anti-glaucoma therapy. We could not control IOP in the other 8 patients despite medical and laser therapy. Three patients (patients 12, 13, and 14) with Cogan-Reese syndrome required trabeculectomy to control the IOP. In spite of surgery, the IOP was still elevated in these 3 patients. The gonioscopic examination revealed angle occlusion and peripheral anterior synechiae, which was estimated at 10–95% in all these 15 patients. However, elevated IOP was noted in only 8 of them, excluding the 3 patients (patients 3, 10, and 11) whose IOP was within normal limits at the first visit due to previous medical or surgical therapy for glaucoma. In the 3 patients with Cogan-Reese syndrome, the extent of angle involvement by peripheral anterior synechiae ranged from 25% to 50% (25%, 50%, and 25% for patients 12, 13, and 14), which was somewhat proportional to the elevated IOP (30, 40, and 26 mm Hg, respectively). However, in the other 5 patients with elevated IOP, the extent of angle involvement was not in proportion to their IOP.

Specular Microscopic Findings of ICE Syndrome

Iridocorneal endothelial cells appeared in all patients and the diagnoses were confirmed using a specular microscope. Under the specular microscope, the ICE cell was characterized by a dark area with a central spot of light and often with a peripheral bright zone (Figure 1). Sometimes dark cell margins were visible and cells varied in size. Clear endothelial visualization was obtained and showed varying degrees of endothelial abnormalities, except in patient 5, who had severe corneal edema that interfered with the visualization of endothelial images and ICE cell grading. In 5 patients, poor identification of cellular margins prevented accurate calculation of the mean cell density (patients 2, 5, 7, 8, and 9) (Table 3).

In the Hirst classification, there were 4 grade 3 patients, 6 grade 2 and 4 patients below grade 2. In the 4 patients with grade 3 ICE cells, elevated IOP was found in 1 patient, and, in 2 patients, the angle involvement ranged from 10% to 25% and corneal edema appeared. In patient 3, the density of surviving endothelial cells was within normal limits or even higher, but ICE cells were grade 3 and corneal edema was noted. Of 10 patients who had an ICE grading below grade 3, elevated IOP at referral was found in 7 patients, and the angle involvement ranged from 25% to 95%. Corneal edema was noted in 5 patients.

Figure 1. Specular micrograph of endothelium of patient 3 showing iridocorneal endothelial cells (arrow) that were characterized by dark area with central spot of light and peripheral bright zone.

According to the Sherrard classification of ICE types, one patient was the disseminated type, 4 patients were total, 3 patients were subtotal (+) and 5 patients were subtotal (−). Of these, 1 patient of the total type, 1 patient of the subtotal (+) type, 2 pa-

Table 3. Specular Microscopic Findings and Corneal Endothelial Function in Patients with Iridocorneal Endothelial (ICE) Syndrome

Patient No.	Endothelial Cell Density (mm^2)	ICE Cell Grading	Corneal Edema	ICE Tissue Typing
1	1450	2	+	subtotal(−)
2	N/A	3	+	disseminated
3	3246/4173	3	+	subtotal (+)
4	1157	2	−	total
5	N/A	N/A	+	N/A
6	729	1–2	−	subtotal (−)
7	N/A	3	−	subtotal (+)
8	N/A	3	−	total
9	N/A	2	−	subtotal (+)
10	960	1–2	−	subtotal (−)
11	761	2	+	subtotal (−)
12	2500	2	−	total
13	1780	2	+	total
14	1831	1–2	−	subtotal (−)
15	366	1	+	N/A

N/A: not available due to poor identification of cellular margins, +: presence of corneal edema, −: indicates absence of corneal edema.

Table 4. Pearson Correlation Coefficient Between Clinical Pictures and Specular Microscopic Findings in Patients with Iridocorneal Endothelial Syndrome

	IOP	VA	VF	Angle Involvement	Corneal Edema
ECD	−0.100	0.321	0.09	−0.844	−0.041
P value	.335	.335	.982	.02	.893
Grading	0.044	0.309	−0.434	−0.684	0.129
P value	.88	.283	.182	.007	.723

ECD: endothelial cell density, IOP: intraocular pressure, VA: visual acuity, VF: visual field.

tients of the subtotal (−) type and 1 patient of the disseminated type had corneal edema. Clinically significant visual field defects were noted in every type of patient at referral. The other 2 patients (patients 5 and 15) could not be classified due to the presence of severe corneal edema that interfered with the analysis of all the specular micrographs.

Pearson correlation coefficients were calculated between the specular microscopic findings (endothelial cell density, ICE cells grading) and IOP, corneal edema, visual field defect or visual acuity. We found that there was no correlation between them (Table 4). The Pearson coefficients were also calculated for the correlation between grading and endothelial density, and angle involvement (Table 4). Interestingly, we found that angle involvement correlated negatively to the endothelial cell grading and cell density.

Unilateral or Bilateral ICE Syndrome

All 15 patients, except patient 3, had unilateral ocular disease. In patient 3, corneal edema, corectopia, ectropion uvea, and peripheral anterior synechiae were noted by slit-lamp biomicroscopy in both eyes. Specular microscopic examination revealed grade 3 ICE cells and subtotal ICE (+) tissues in both eyes. However, the degree of corneal edema differed greatly between the 2 eyes. The left cornea was edematous (Figure 2A) while the right cornea was relatively clear except for early band keratopathy at the 3 o'clock and 9 o'clock positions of the peripheral cornea (Figure 2B). The difference in corneal clarity resulted in different visual acuity (1.0 for the right eye, 0.3 for the left eye) in this patient. No family history of corneal endothelial disease was noted in the family of patient 3.

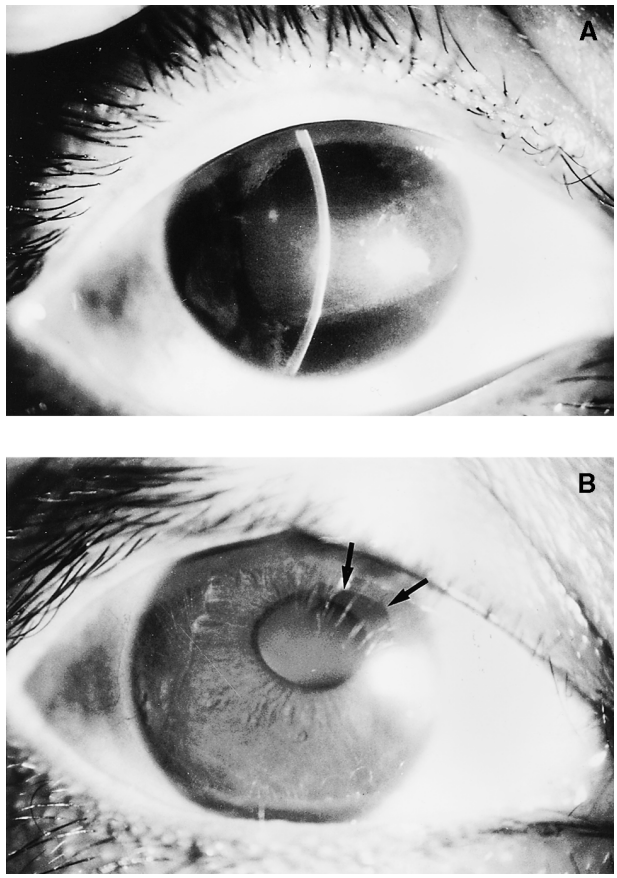

Figure 2. Patient 3. (**A**) Biomicroscopic photography of left eye showing corneal edema with subepithelial fibrosis involving visual axis, ectropion uveae, and corectopia. (**B**) Biomicroscopic photography of right eye showing ectropion uveae at nasal-upper quadrant (arrows), and band keratopathy at 3 o'clock and 9 o'clock positions of peripheral cornea. Central cornea was relatively clear.

Discussion

Diagnosis of ICE syndrome was based on abnormalities in the corneal endothelium, distortion of the pupil with ectropion uvea, thickening of the iris stroma with increased pigmentation, iris atrophy, peripheral anterior synechiae, glaucoma, and unilaterality of disease.[1,2] However, the clinical pictures may also be consistent with the diagnosis of other endothelial disorders, eg, posterior polymorphous dystrophy, Fuchs' dystrophy and mesodermal dysgenesis.[16–19] It is sometimes difficult to definitely diagnose ICE syndrome solely by slit-lamp biomicroscopy. Specular microscopy provides an invaluable method for the differential diagnosis of difficult cases.[16–19] For instance, iridocorneal adhesion, stromal iris atrophy and ectropion uvea have also been reported in posterior polymorphous dystrophy, making differential diagnosis difficult. In posterior polymorphous dystrophy, the endothelium appears as dark rings with distinct, often scalloped edges surrounding a

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lighter mottled center which can be differentiated from ICE cells using specular microscopy. In Fuchs' dystrophy, the guttae under a specular microscope appear as black holes in the endothelial mosaic, which is also quite distinct from ICE cells. As the corneal guttae become more extensive, dark areas are found almost completely throughout the involved endothelium.

Poor vision in patients with ICE syndrome might be related to corneal edema, glaucomatous optic nerve damage, cataract formation, or a combination of these factors.[1,2,20,21] Of the 15 patients included in this retrospective study, 11 patients had visual disturbance as the initial manifestation at the time of diagnosis. In 7 of our 15 patients, corneal edema or opacity was found on the first visit to our hospital. Corneal decompensation resulting in corneal edema and opacity was the major cause of vision impairment in patients with late ICE syndrome.

Chandler's syndrome occurred the most frequently in our patients with ICE syndrome (11/15), which is consistent with most of the previous studies.[1–4] However, Teekhasaenee and Ritch[27] reported that Cogan-Reese syndrome was the most common form in Asian patients while Chandler's syndrome was more common in white patients. They included 60 patients with ICE syndrome in their study, among them 3 patients initially diagnosed as having Chandler's syndrome and 1 as having progressive essential iris atrophy progressing to Cogan-Reese syndrome during the follow-up period up to 12 years. In our patients, the follow-up period ranged from 14 months to 56 months. There is a possibility that some of our patients who were diagnosed as having Chandler's syndrome may progress to Cogan-Reese syndrome in the future. As to the sexual difference in patients with ICE syndrome, we found that women composed the majority of the patient group (80%), in support of the description by Sherrard that "the typical patient is a woman".[12]

Both the Hirst and the Sherrard classifications of ICE cell grading were adopted in this study: We could not find any distinct correlation between the ICE grading and the occurrence of corneal edema. According to the present study, the classification of ICE grading was not a reliable tool for predicting prognosis. We also could not find a correlation between endothelial cell density and endothelial function in ICE syndrome; this is contrary to the findings in most corneal diseases. Because the endothelial cell density counted in ICE syndrome included both the surviving cells and the ICE cells, the endothelial cell density could not reflect the true endothelial function. The ICE cells were not normal corneal endothelial cells and did not work normally even when they were within the normal range of endothelial count.[3,4,9,19]

Significantly elevated IOP was found in 8 of our patients and the control of IOP was poor. There was no distinct correlation between the degree of angle involvement by peripheral anterior synechiae and the occurrence of glaucoma, which is in accordance with a previous study.[11] This kind of secondary glaucoma was refractory to medical treatment and the success rate of surgery was 60% 1 year after surgery and 21% 5 years after the first operation, as described previously.[20–22] In the present study, the IOP was much more difficult to control in the cases of Cogan-Reese syndrome when compared with other patients with ICE syndrome. The IOP was poorly controlled in our 3 cases with Cogan-Reese syndrome, in spite of anti-glaucoma medications and trabeculectomy. This result was in accordance with the findings of Wilson and Shields[23] that the control of IOP was more unsatisfactory in Cogan-Reese syndrome. The reason for this finding is difficult to explain due to the incomplete occlusion of the angle in these affected eyes. We suspect that the fine architecture of the trabecular meshwork in Cogan-Reese syndrome patients might be distorted by ICE cells or membrane microscopically, even in the absence of gross angle occlusion.

Although ICE syndrome is mainly a unilateral disease, bilateral ICE syndrome (progressive bilateral essential iris atrophy and Chandler's syndrome) has been reported by other authors.[24,25] A case of bilateral Chandler's syndrome (patient 3) was found in our study using specular microscopic examination. Slit-lamp examination revealed a bilateral beaten-metal appearance in the endothelium, microcystic corneal edema, corectopia, and ectropion uvea. Gonioscopy revealed bilateral peripheral anterior synechiae. Intraocular pressure in both eyes remained normal despite the extensive angle involvement. Similar specular micrographs were observed in both eyes, including grade 3 of ICE cell grading and subtotal ICE (+) tissue type. In bilateral ICE syndrome, specular microscopy is invaluable. The only way to differentiate bilateral ICE syndrome from other endothelial disorders is by using specular microscopy.

In conclusion, we analyzed the records of 15 patients and correlated the clinical findings of ICE syndrome with specular microscopic evaluations. Specular microscopy is a good tool for visualizing endothelial abnormalities directly, and for assisting in differential diagnosis. Corneal decompensation

and secondary glaucoma are the major causes of decreasing visual acuity in these patients. In Cogan-Reese syndrome, visual prognosis was worse due to poor control of IOP. The classification of ICE syndrome by the Hirst and Sherrard systems are not reliable methods for predicting prognosis. Because damage to visual function is not proportional to the extent of angle involvement in patients with ICE syndrome, we suggest that close follow-up of intraocular pressure and early detection of glaucoma are important to preserve visual function in patients with ICE syndrome.

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