

# Iridocorneal Endothelial Syndrome

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Iridocorneal endothelial (ICE) syndrome encompasses a spectrum of disorders characterised by corneal endothelial abnormalities, progressive formation of peripheral anterior synechiae (PAS) without pupillary block and iris defects.<sup>1</sup> It is traditionally subdivided into Chandler's syndrome, progressive iris atrophy, and iris nevus (Cogan-Reese) syndrome. In their purest form, the 3 syndromes can be easily distinguished but there is often considerable overlap in the clinical features. The cause of ICE syndrome is unknown. A membrane grows from abnormal corneal endothelium across the angle onto the iris surface and contracts, causing the formation of PAS and iris abnormalities.<sup>1</sup>

The condition is uncommon and typically affects one eye of young to middle-aged women. Patients often present with a distorted pupil in a

previously normal eye. The iris may show ectropion uvulae and full-thickness iris holes, and there are often varying degrees of iris atrophy. The posterior corneal surface has a characteristic beaten metal appearance and specular microscopy may reveal the distinctive ICE cells, which are pathognomonic of ICE syndrome. The clinically uninvolved contralateral eye may also have sub-clinical endothelial abnormalities.

The course of the disease is progressive and there is no means of arresting it. Serious sequelae of ICE syndrome are glaucoma and corneal decompensation. Glaucoma occurs in about 50% of patients with ICE syndrome,<sup>2,3</sup> especially in the variants of ICE syndrome in which abnormal ICE cells involve the entire posterior corneal surface. Glaucoma is caused by secondary synechial angle closure as a result of contraction of the abnormal tissue in the angles.

IOP may be controlled initially with medical therapy but medical treatment is generally ineffective.<sup>2,4</sup> Management of glaucoma due to ICE syndrome is primarily surgical. A wide range of surgical procedures to control the IOP in ICE syndrome have been described, with variable but often limited success.<sup>2,4-6</sup> In one large series of 66 patients with ICE syndrome, the success rate of conventional first trabeculectomies at 1 year and 5 years were only 60% and 21%, respectively.<sup>2</sup> The high failure rate is related to the progressive nature of the disease with endothelialisation occurring over a filtration fistula or within the bleb itself. In unresponsive eyes, the use of a glaucoma drainage implant should be considered.

Glaucoma secondary to ICE syndrome is often initially attributed to other causes. In all patients with unilateral glaucoma, particularly if they are young, ICE syndrome should be considered as a possible diagnosis and its signs, which may be subtle, should be sought.



## References

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Figure. Iridocorneal endothelial syndrome.

