Iridocorneal Endothelial Syndrome

- abnormal corneal endothelium that is responsible for variable degrees of iris atrophy, secondary angle-closure glaucoma in association with characteristic peripheral anterior synechiae (PAS), and corneal edema.
- Three clinical variations have been described:
  - Iris nevus (Cogan-Reese) syndrome;
  - Chandler's syndrome;
  - Essential (progressive) iris atrophy.
- ICE -- Yanoff suggested the term iridocorneal endothelial in 1979
- unilateral
- subclinical irregularities of the corneal endothelium commonly noted in the fellow eye.
- 20-50 years of age
- more often in women.
- No consistent association has been established with any other ocular or systemic disorder, and familial cases are very rare.
- 50% Chandler's syndrome; the other two 25% of all cases.
- Glaucoma occurs in approximately 50% more severe in patients who have the progressive iris atrophy and Cogan-Reese variations, as opposed to those who have Chandler's syndrome.
- The degree of angle closure does not always correlate to the elevation in intraocular pressure (IOP), since some angles may be closed functionally by the endothelial membrane without the occurrence of synechial closure.
- present with complaints of pain, decreased vision, and an abnormal iris appearance.
- The reduced vision and pain are secondary to corneal edema and/or secondary angle closure, which may occur later in the disease.
- Patients frequently note a mild blur of vision in the morning hours as the result of lid closure and mild corneal edema that occurs during sleep.
- They also may relate the improvement of vision throughout the day, as the cornea dehydrates with exposure to the air.
- Microcystic corneal edema may be present without elevated IOP, especially in Chandler's syndrome.
- In the advanced stages of the syndrome, symptoms of blurred vision and pain may persist throughout the day.
- Patients also may present with the complaint of an irregular shape or position of their pupil (corectopia), or describe a dark spot in the eye, which may represent hole formation (pseudopolycoria) or stromal atrophy of the iris.
Progressive (Essential) Iris Atrophy
- severe iris atrophy that results in heterochromia, marked corectopia, ectropion uveae, and pseudopolycoria (hole formation). Hole formation is the hallmark finding of

Cogan-Reese Syndrome
- The iris atrophy tends to be variable and less severe. Tan, pedunculated nodules may appear on the anterior iris surface. The entire spectrum of corneal and other iris defects may occur in this variant.

Chandler's Syndrome
- This variation shows minimal or no iris stromal atrophy, and mild corectopia may occur. The corneal edema and angle findings predominate and are typical
IN ALL fine, hammered-silver appearance of the posterior cornea, similar to the guttata seen in Fuchs' corneal endothelial dystrophy, is noted -- this results from the abnormal endothelial cells posterior to normal Descemet's membrane.

Researchers, using electron microscopy, have shown this endothelial layer to vary in thickness from a single layer to multiple layers.

Also evident is that, within the same eye, the endothelial cell layer may be of different thickness in different areas.

The evidence of filopodial cytoplasmic processes and cytoplasmic actin filaments implies that the endothelial cells are able to migrate.

The morphology of the endothelium suggests a widespread state of high metabolic activity.

Corneal edema is secondary to these marked endothelial abnormalities.

The anterior chamber angle may show high peripheral anterior synechiae (PAS) that extend beyond Schwalbe's line.

Such PAS are caused by the contraction of this endothelial cell layer and surrounding collagenous, fibrillar tissue, which are continuous and extend from the peripheral cornea over the trabecular meshwork and iris.

An angle-closure glaucoma results as these PAS contract and close the angle.

The pupil is drawn toward the sector that has the most prominent PAS.

Secondary glaucoma with an open angle also may occur because of endothelial membrane that covers the trabecular meshwork without evidence of synechial formation.

The extension of the endothelial cell layer over portions of the anterior iris surface from the anterior chamber angle contracts, which distorts and pulls the iris toward itself.

Hole formation occurs opposite the location of the abnormal endothelial cell layer secondary to the contracture.

Hole formation may be associated with ischemia of the iris, as suggested by fluorescein angiography.
In Cogan-Reese syndrome the pigmented, pedunculated nodules seen are composed of underlying iris stroma pinched off by abnormal cellular membrane.

A viral cause has been postulated for the mechanism of the ICE syndrome.

Epstein-Barr and Herpes simplex viruses have been found serologically in ICE patients.

This theory was postulated after lymphocytes were seen on the corneal endothelium of an ICE patient, which indicated the presence of chronic inflammation.

The diagnosis of ICE syndrome must be considered in younger patients who have unilateral angle-closure glaucoma; it is confirmed by specular microscopy.

Corneal edema and secondary glaucoma are the major concerns to be addressed.

Corneal edema often may be controlled using hypotonic saline solutions and, when elevated, the reduction of intraocular pressure (IOP) may help to lessen corneal edema.

Elevated IOP that occurs with secondary glaucoma often may be controlled medically using aqueous suppressants.

Miotics often are ineffective and the role of latanoprost remains uncertain.

If the IOP remains uncontrolled, filtration surgery may be indicated, although late failures have been reported secondary to fistular endothelialization.

These fistulas may be reopened successfully when the endothelial cell membrane is cut using the YAG laser.

Success rates of initial trabeculectomy operations at 1 and 3 years were 64% and 36%.

Second and third operations at 1-year intervals were both 58%.

Seton procedures are indicated for cases refractory to the above treatments.