Clinical & Refractive Optometry is pleased to present this continuing education (CE) article by Dr. Ron Melton and Dr. Randall Thomas entitled **Fuchs' Endothelial Corneal Dystrophy.** In order to obtain 2-hours of COPE-approved CE credit, please refer to page 178 for complete instructions.

Fuchs' Endothelial Corneal Dystrophy

Ron Melton, OD; Randall Thomas, OD

Subjective

A 52-year-old Caucasian female presents with a recent history of blurred vision and photophobia, especially in the left eye (Fig. 1). Her vision seems to be worse in the early morning and improves later in the day. She had an eye examination seven years ago and was given a prescription for bifocals. She is taking ibuprofen for arthritis and has no known drug allergies. There is a family history of a grandmother having poor vision in her 60s. The cause for this poor vision was unknown.

Fig. 1 This 52-year-old female presented with a recent history of blurred vision and photophobia, especially in the left eye. Note that there is no significant injection in either eye

Objective

- Visual acuity: OD 6/7.5 (20/25); OS 6/12 (20/40) (pinholed no improvement)
- Gross observations: normal OU
- Lids: normal OU
- Conjunctiva: white and quiet OU
- Cornea: OS shows central endothelial guttata with 2+ secondary stromal edema and folds in Descemet's membrane (Fig. 2). Indirect view adjacent to the slit-lamp light beam shows a stippling pattern created by the corneal edema (Fig. 3). The optic section is thickened centrally. OD shows central endothelial guttata with 1+ stromal edema
- Anterior chamber: clear with no cells/flare OU
- Tension by applanation: 17/16 mmHg at 8:30 a.m.
- Internal: 0.3 cup-to-disc ratio with maculae and retinal blood vasculature normal OU

Assessment

Fuchs' corneal endothelial dystrophy OS greater than OD

Plan

- Educate patient regarding this eye disease and the need for long-term follow-up
- Prescribe sodium chloride 5% ophthalmic drops q.i.d. OU and sodium chloride 5% ophthalmic ointment at bedtime over the next month and follow-up at that time

Follow-up one month

- The patient stated that she has noticed a significant improvement in vision and less sensitivity to lights
- Visual acuity was OD 6/7.5 (20/25) and OS 6/9 (20/30)
- The corneas showed less edema. In particular, the cornea OS revealed the classic beaten metal appearance of the confluent central endothelial guttata associated with increased pigmentation that is found in Fuchs' dystrophy (Figs. 4, 5)
- The patient was given instructions to continue the sodium chloride 5% ointment at bedtime and use non-toxic artificial tears three to four times daily. She was to schedule a complete eye examination in one to two months

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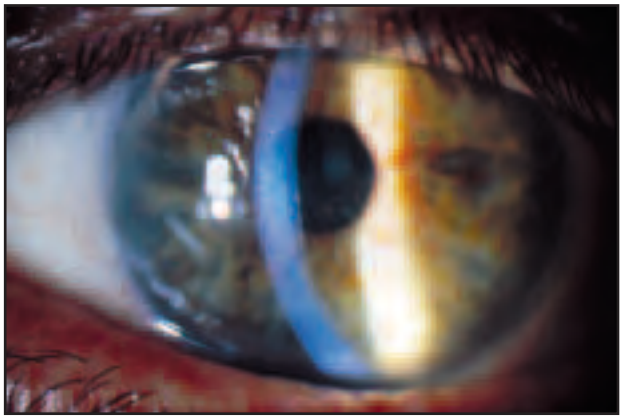

Fig. 2 The left cornea shows central endothelial guttata with secondary stromal edema and folds in Descemet's membrane.

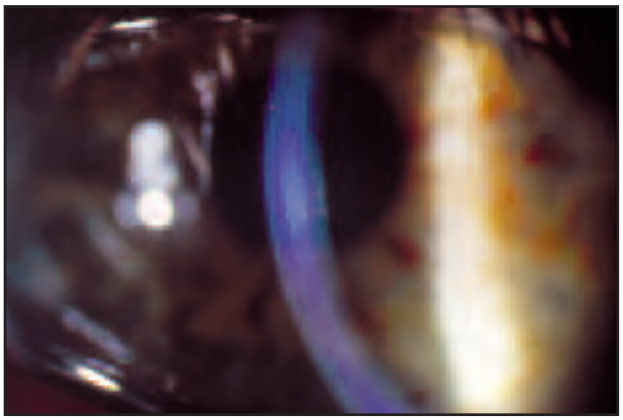

Fig. 3 Under higher magnification, the indirect view adjacent to the slit-lamp beam shows a stippling pattern created by the corneal edema.

Comments: In Fuchs' corneal endothelial dystrophy, clinical findings will vary depending on the severity of the disease. The first evidence of this dystrophy is centrally located corneal guttata that spreads toward the periphery. Coalescence of the endothelial guttata centrally associated with increased pigmentation produces a classic beaten metal appearance. This appearance was noted at the one month follow-up with this patient. There is a thickening of Descemet's membrane with subsequent folds and stromal edema. With further decompensation of the endothelium, epithelial edema develops, causing microcystic edema and epithelial bullae. These epithelial bullae may rupture in the later stages of the disease. During this process, the cornea may double in its normal thickness.

Fuchs' dystrophy can be a challenging eye disease to manage because of the way it affects vision. As with this case, patients present with complaints of blurred vision, expecting to update their glasses and see perfectly. Giving them instructions to use a thick ophthalmic ointment and drops that might sting is not what they want to hear. It is therefore critical that we educate these patients of the importance of long-term compliance with recommended therapy.

Some experts advocate the use of a hair dryer to supplement dehydrating the cornea when patients show considerable edema and are moderately symptomatic. Because of compliance issues and the variability of operation of hair dryers, recommend this if the sodium chloride drops and ointment are not helping after a month or two.

Fortunately, most patients can be managed with medical therapy. In advanced cases, patients may develop microcystic corneal edema and epithelial bullae. A therapeutic soft bandage lens may be tried to help control the pain associated with more severe cases. If vision is compromised to an unacceptable level, patients have an excellent prognosis with a penetrating keratoplasty.

The progression of Fuchs' dystrophy is typically slow, which allows for management by medical therapy alone. Patients should be advised that refraction results will vary dramatically, depending on the amount of active corneal edema present.

General Observations

- Bilateral, often asymmetric, autosomal dominant corneal endothelial dystrophy
- Slow chronic disease that generally progresses over many years
- Higher prevalence in women over age 50
- Vision may fluctuate markedly from day to day

Etiology

- Unknown, keratoconus, glaucoma, female hormones, trauma, inflammation, viruses, etc.

Diagnosis

- Pleomorphic endothelial cells synthesize abnormal basement membrane, leading to epithelial and stromal edema
- Central endothelial excrescences (guttata) are the hallmark clinical sign, and can lead to endothelial cell decompensation and corneal hydration (Fuchs' dystrophy)
- Advanced cases can lead to opaque, hydrated corneas

Clinical Stages

- Stage I
  - central corneal guttata
  - asymptomatic
- Stage II
  - stage I and corneal edema

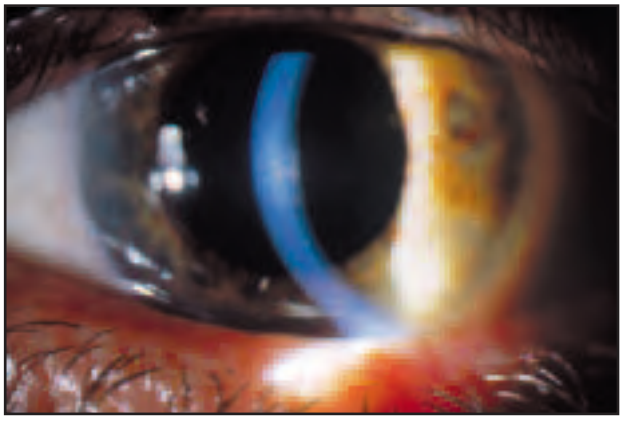

Fig. 4 After using sodium chloride 5% ophthalmic drops and ointment for one month, the left cornea shows less edema. The optic section remains thickened centrally, which is characteristic of Fuchs' corneal endothelial dystrophy.

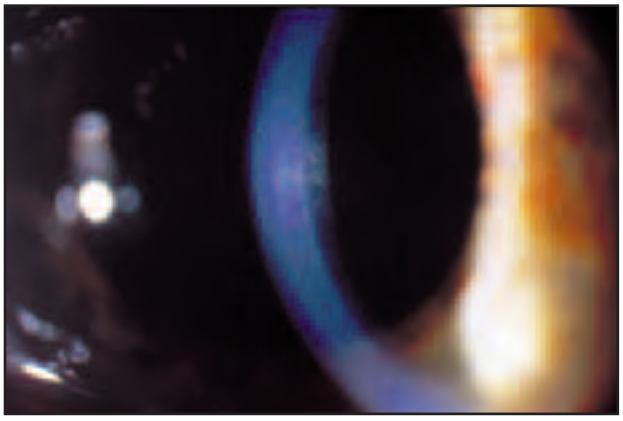

Fig. 5 The left cornea reveals the classic beaten metal appearance of the confluent central endothelial guttata associated with increased pigmentation that is found in Fuchs' dystrophy.

- painless decrease in vision with photophobia
- worse on awakening

- Stage III
  - epithelial bullae and pain
- Stage IV
  - subepithelial scarring with severe vision loss

Differential Diagnosis

- Other congenital hereditary endothelial dystrophies, Hassall-Henle warts, pseudophakic bullous keratopathy, Chandler's syndrome, central herpetic disciform keratitis, etc.

Treatment

- Sodium chloride 5% ophthalmic solution four to six times daily and/or sodium chloride 5% ophthalmic ointment at bedtime if patient is very symptomatic
- Corneal evaporation using a hair dryer at low heat, held at arm's length three to four times daily has been advocated, although it is difficult to do
- If moderate pain, use a therapeutic soft bandage
- Lowering the intraocular pressure may temporarily help the edema
- If vision is compromised enough, penetrating keratoplasty is highly successful (only necessary in a small percentage of patients)
- Excimer laser phototherapeutic keratectomy has not been shown to help

Disclaimer: Not every detail of every case is discussed, rather the key clinical findings are described. For example, if nothing is said about the corneal status, you should assume that the cornea is normal, etc. When vision is recorded, it should be assumed to be best corrected or pinholed. Regarding therapy, we show how we treated the particular case. Given that medicine is an art, as well as a science, therapy will — and often does — vary with each unique patient presentation depending on severity, known drug allergies, prior treatment, response to therapy, etc.

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QUESTIONNAIRE

Fuchs' Endothelial Corneal Dystrophy

Ron Melton, OD; Randall Thomas, OD

1. Which of the following signs/symptoms were present on initial presentation?
   - ❑ Blurred vision
   - ❑ Photophobia
   - ❑ Greater vision impairment in early morning
   - ❑ All of the above

2. Which of the following findings **DOES NOT** describe this patient's case?
   - ❑ Conjunctiva: red OU
   - ❑ Lids: normal OU
   - ❑ Cornea OS: endothelial guttata with 2+ secondary stromal edema
   - ❑ Anterior chamber: clear with no cells/flare OU

3. Which of the following statements regarding treatment is **FALSE**?
   - ❑ Sodium chloride 5% ophthalmic drops q.i.d. OU are recommended
   - ❑ Once effective treatment has been administered, long-term follow-up is not necessary
   - ❑ Patient education is an important part of an effective treatment plan
   - ❑ Sodium chloride 5% ophthalmic ointment at bedtime for 1 month is recommended

4. Which of the following tests/examinations is essential in this type of case?
   - ❑ Slit-lamp examination
   - ❑ IOP measurement at initial visit
   - ❑ View of fundus, documenting ONH appearance and any fundus abnormalities seen
   - ❑ All of the above

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5. Identify the **FALSE** statement regarding this case:
   - ❑ Stromal edema is characteristic of this condition
   - ❑ Thickening of Descemet's membrane is characteristic of this condition
   - ❑ Epithelial edema is characteristic of this condition
   - ❑ None of the above

6. Which of the following is the **FIRST** evidence of dystrophy?
   - ❑ Centrally located corneal guttata spreading toward the periphery
   - ❑ Coalescence of the endothelial guttata centrally associated
   - ❑ Doubling of the corneal thickness
   - ❑ Development of epithelial bullae

7. Which of the following statements does **NOT** describe the patient's condition at follow-up one month?
   - ❑ Less edema in the left cornea, following use of sodium chloride 5% as the sole treatment
   - ❑ Central thickening to optic section of left cornea
   - ❑ Visual acuity OD 6/7.5 (20/25) and OS 6/9 (20/30)
   - ❑ Increased pigmentation typical of Fuchs' dystrophy

8. Fuchs' dystrophy is difficult to treat medically because:
   - ❑ Patients frequently do not present until far into the disease process
   - ❑ Patients are reluctant to report the condition because of the fear of vision loss
   - ❑ Patients often dismiss their symptoms, assuming they will quickly disappear
   - ❑ Patients may find use of a thick ophthalmic ointment inconvenient and somewhat painful

9. Which of the following statements about Fuchs' dystrophy is **TRUE**?
   - ❑ Most patients can be managed with medical therapy
   - ❑ A basic objective of treatment is to dehydrate the cornea in the face of edema
   - ❑ When severe, the condition can be quite painful
   - ❑ All of the above

10. Which of the following is **NOT** characteristic of the condition?
    - ❑ Fluctuating vision from day-to-day
    - ❑ Progression over many years
    - ❑ Asymmetric corneal endothelial dystrophy
    - ❑ Higher prevalence in men over age 50

11. Which of the following may help alleviate the pain of severe cases?
    - ❑ A therapeutic soft bandage lens
    - ❑ Acetylsalicylic acid 325 mg t.i.d.
    - ❑ Hydration through frequent use of saline solution in the affected eye
    - ❑ A non-steroidal anti-inflammatory, such as naproxen sodium 500 mg b.i.d.

12. Identify the **FALSE** statement about Fuchs' dystrophy:
    - ❑ In younger patients, it progresses slowly; in patients over 50, progression is more rapid
    - ❑ Penetrating keratoplasty is reserved for treating the condition in its early stages only
    - ❑ Permanent vision loss is a near-certainty
    - ❑ All of the above

13. Which of the following describes the possible etiology of Fuch's dystrophy?
    - ❑ Keratoconus
    - ❑ Glaucoma
    - ❑ Trauma
    - ❑ All of the above

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14. Which of the following statements regarding diagnosis of Fuchs' dystrophy is **FALSE**?
    - ❑ Central endothelial excrescences (guttata) are the hallmark clinical sign of advanced cases
    - ❑ Guttata are more typically seen in younger patients with the condition
    - ❑ Guttata can lead to endothelial cell decompensation, however, this is unlikely
    - ❑ All of the above

15. Identify the **INACCURATE** statement regarding clinical stages of the condition:
    - ❑ Pain is progressive, typically initiating in Stage I
    - ❑ Stage II loss of visual acuity is painless
    - ❑ Epithelial bullae and pain usually occur in Stage III
    - ❑ Severe loss of visual acuity is characteristic of Stage IV

16. Which of the following relates to differential diagnosis of Fuchs' dystrophy?
    - ❑ Chandler's syndrome
    - ❑ Central herpetic disciform keratitis
    - ❑ Pseudophakic bullous keratopathy
    - ❑ All of the above

17. All of the following statements regarding treatment are true, **EXCEPT**:
    - ❑ Use of a therapeutic soft bandage is recommended at Stage I
    - ❑ Lowering IOP may temporarily lessen edema
    - ❑ Excimer laser phototherapeutic keratectomy has not been proven helpful
    - ❑ Corneal evaporation using a hair dryer at low heat may be recommended

18. Which of the following statements is **FALSE**?
    - ❑ Corneal edema typically causes a stippling pattern
    - ❑ Sodium chloride 5% ophthalmic drops, combined with ointment, may diminish corneal edema
    - ❑ Central endothelial guttata may cause a beaten metal appearance
    - ❑ None of the above

19. All of the following statements are true, **EXCEPT**:
    - ❑ When properly treated early in the disease process, refraction results are predictable, patient-to-patient
    - ❑ Refraction results may vary from patient-to-patient, depending on the amount of active corneal edema present
    - ❑ Treatment efficacy depends largely on long-term patient compliance to therapy
    - ❑ If vision loss reaches an unacceptable level, penetrating keratoplasty may be recommended

20. Identify the statement that describes the treatment plan in this case:
    - ❑ At one month, non-toxic artificial tears three to four times daily
    - ❑ At one month, further follow-up consisting of a complete eye examination in one to two months
    - ❑ Continuation of sodium chloride 5% ointment at bedtime
    - ❑ All of the above