The Corneal Dystrophy Foundation
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Focus on Education – Inspiration – Vision

Visiting Your Doctor

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Introduction

Fuchs' Corneal Dystrophy is an hereditary corneal disease. Once you have been diagnosed with Fuchs' Corneal Dystrophy you'll begin to develop a partnership with your corneal specialist that will last throughout your life. Together you and your doctor will make important treatment decisions, so you should feel comfortable asking questions and talking with your doctor.

You should see an ophthalmologist that specializes in corneal diseases before your symptoms become severe. Not only will this help you develop a better relationship with your doctor, but it will also help your doctor understand how your Fuchs' is progressing over time. If you don't have a corneal specialist yet, you should ask for recommendations from your optician, ophthalmologists, friends, and family. If a doctor's name comes up more than once, he or she is probably a good choice.

You might also join Fuchs' Friends, our on line support and knowledge message board. Open your browser and go to:

http://www.fuchs-dystrophy.com and click on the words, Join Fuchs' Friends in the colored box; fill out the request form completely and click on Submit.

This brochure will first give a brief description of Fuchs' Dystrophy and will describe six common vision tests that doctors may use to figure out your stage of Fuchs'. Understanding these tests will make it easier for you to talk to your doctor and track changes in your vision. Then the brochure will offer some helpful hints to make your appointment with the doctor go smoother. Finally, it will provide a list of suggested questions to ask your doctor.

Fuchs' Dystrophy Primer

Fuchs' Dystrophy affects the cornea of your eye. The cornea is a clear window that allows light to enter the eye. It about a half millimeter thick, about as thick as two business cards. The innermost layer of the cornea contains endothelial cells that form a barrier between the fluids inside the eye and the rest of the cornea.

It is normal for your endothelial cells to die gradually as you age. Patients with Fuchs' Dystrophy have fewer endothelial cells because their cells die at a greater than normal rate. These cells cannot grow back once they die. If you do not have enough endothelial cells to pump fluids away from the cornea, fluids will seep into the corneal tissue. This will alter the shape of the cornea and make your vision cloudy and blurry.

Often the fluids seep into some parts of the cornea more than others and causes the shape of the cornea to be irregular and wavy like a washboard. This waviness can cause astigmatism. Blisters are a rare complication of Fuchs' Dystrophy. If it becomes too swollen with fluids, the outer part of the cornea may develop microscopic blisters. Occasionally these blisters can be larger and may rupture.

If you think you have blisters you should contact your doctor so that he or she can monitor them and provide pain relief. Typically Fuchs' Dystrophy patients have the worst vision in the morning. Sometimes treatments that dry out the cornea, such as concentrated saltwater drops or wafting warm air from a hair dryer across the eyes, can improve vision. If vision becomes worse, a cornea transplant may be done.

Six Vision Benchmark Tests and What They Mean

There are six common vision tests that your doctor may decide to run to see how your Fuchs' Dystrophy is progressing. You do not necessarily need each test to be run in order to have a complete examination and thorough diagnosis. Most patients should consider asking the doctor to tell them their stage of guttata and give them a visual acuity test and a glare test.
The way you perform on these tests may change depending on the time of day that you see the doctor.

Many Fuchs' patients find that their vision is at its worst early in the morning. If you record your test results during every appointment, make sure you include the time of day as well as the date. The significance, procedures, and normal results of six vision tests are described below.

Grade or Stage of Guttata

What the test tells you

The doctor will look at the endothelial cells in your inner cornea. These cells are normally tightly packed and are equal in size. Abnormal endothelial cells produce little bumps on the inner part of the cornea. This causes corneal guttata where the cornea has a “beaten-metal” or “orange peel” appearance. The bumps start off small and over time get larger. Eventually the bumps disrupt the endothelial cells, preventing the cells from pumping fluids away from the cornea. Vision may become “foggy” when fluids seep into the cornea. Your stage of guttata is determined by comparing the size of the endothelial cells to the size of these bumps.

Procedure

The doctor will use an optical microscope called a slit lamp to look inside your eye.

Grades or Stages of Guttata

Dr. JH Krachmer devised a grading scale for Fuchs'. Some doctors say "Grade xx" and others say "Stage xx", but they mean the same. The Krachmer scale is as follows:

Grade 0 indicate no disease, as defined by fewer than 11 central guttae.

Grade 1 represents definitive onset of Fuchs', and is indicated by 12 or more central, nonconfluent guttae in at least one eye.

Grade 2 patients exhibit a zone of confluent central guttae 1 to 2 mm in horizontal width.

Grade 3 is when the area of confluence has expanded to 2 to 5 mm.

Grade 4 is when the area of confluence is greater than 5 mm.

Grade 5 is the same as grade 4, but also edema is exhibited in the corneal stroma and/or epithelium.

Vision Acuity Test

What the test tells you

This test is the most basic test performed during a routine eye examination and tells you how well you can see objects from a distance. If have 20/20 vision you are able to see characters roughly .34 inches or .887 cm high. This is considered to be “nominal” visual acuity. When you have 20/40 vision it means that you need to be 20 feet away to see what a person with nominal visual acuity can see from 40 feet away. Keep in mind that your visual acuity may change depending on what time of day you have the test.

Procedure

The doctor will ask you to read rows of characters that are 20 feet away from you. If the examination room has limited space the doctor will use mirrors to make the characters appear 20 feet away. Many testers will encourage you to guess at characters; we feel that this generates a prescription that may not be useful if you have the test in the PM and have the typical ‘Fuchsie Fuzzies’. Try not to guess or memorize.

Normal Range of Results

"Normal" vision is 20/20. Though some states are more lenient, in most states your corrected vision must be 20/40 or better in at least one eye to pass a driver's licensing test.

Glare Test

What the test tells you

This test will help you understand how your vision is affected under night driving situations. You have disability glare when a light source reduces your visual acuity. This means that even if you have close to 20/20 vision under normal conditions, you can become practically blind at night when faced with the headlights of an oncoming vehicle.

Procedure

The procedure is similar to the vision acuity test except that a light will be directed at your eye as the test is given.

Normal Range

Glare tests are typically measured using the same system as the visual acuity test. Twenty-twenty vision is considered to be "normal." Most states test a driver's vision under normally lit conditions rather than glare conditions.

Corneal Cell Count

What the test tells you

A corneal cell count will tell you how many endothelial cells are present in the inner layer of your cornea. Ophthalmologists do not routinely perform this procedure, but it may be used to evaluate the cornea before cataract surgery.

Procedure

The doctor will look into your eye using a special microscope to see and count individual cells.

Normal Range of Results

A normal cell count for people ages 40-70 is 2,000 to 3,000 cells per mm2 (cells per square mm). When your cell count drops below 500 per mm2 then your cornea might begin to swell and your vision may be blurry.

For intraocular surgery, such as cataract surgery, a cell count of greater than 1,500 cells per mm2 is recommended. The recommended number of cells for intraocular surgery can vary depending on the clinical situation.

Corneal Pressure Test

What the test tells you

The corneal pressure tests measures how much pressure has built up inside the eye. You may also hear it referred to as an intraocular pressure (IOP) measurement. If your cornea becomes harder it can cause pressure to build up behind it. This test is also used to determine whether a person has a risk of glaucoma.

Procedure

It is a quick and painless procedure. One way for a doctor to measure the pressure is to direct a brief puff of air on the eyeball. In other more accurate methods of determining the eye pressure, the doctor uses eye drops to numb your eyes and applies a dye that makes it easier to see the cornea. Then the doctor will briefly touch an instrument to your eye. Make sure you don't hurt your cornea by rubbing your eye before the numbing drops wear off.

Normal Range of Results

The results are measured in millimeters of mercury (mmHg) much like a barometer. Normal readings range from 12 to 21 mmHg.
Keep in mind that your eye pressure will be:
· The greatest when you first wake up
· Increasing as you get older
· Higher if you're a woman
· The most accurate if you avoid drinking alcohol 12 hours before the test or smoking marijuana for at least 24 hours before the test.

Corneal Thickness Test

What the test tells you

The test measures the thickness of your cornea. If the cornea is too thin then it "bows" out and distorts vision further. This "bowing" out of the cornea is called ectasia. The cornea may also be thicker than normal because it is swollen with fluids from the eye.

Procedure

The doctor will use ultrasound to measure your cornea's thickness. The latest Zeiss computerized machine generates a printed graph of the cornea.

Normal Range of Results

The cornea normally becomes thinner as you get older. Your results may vary from the center of the cornea to the outside edges of the cornea.
Normal corneal thickness:

- At 1 year old the cornea will be 0.566 to 0.576 mm thick
- At 50 years old the cornea should be about 0.515 to 0.575 mm thick
- Fuchs' Corneal Dystrophy may cause the cornea to thicken to as much as 0.700 mm.

Though doctors frequently use millimeters (mm) to measure the cornea's thickness, they may use microns instead. The symbol for a micron looks like a lowercase "um." To convert from microns to millimeters take the decimal

point on the micron reading and move it three spots to the left. Now you have millimeters.
For example these are equal:
512.00 um
0.512 mm

Helpful Hints for Your Appointment with the Doctor

As you partner with your doctor to battle Fuchs' Dystrophy it will be important to feel comfortable voicing your concerns and asking questions. The following list offers some suggestions to help you with your appointment:

- Ask questions until you understand your doctor's responses.
- Bring a list of questions that you want to ask the doctor (A good starting point would be the list included at the end of this brochure).
- If you want to discuss a problem with your doctor, bring notes to help you remember your symptoms, their frequency, and their duration.
- As you talk to your doctor take notes or use a tape recorder to help you remember the entire discussion.
- Bring a friend or family member into the doctor's office to take notes for you and to make you feel more comfortable.
- Ask your doctor to write down his or her instructions.
- Keep a journal to track your Fuchs' Dystrophy and cornea transplant.
- Talk to other members of your health care team, such as nurses and pharmacists to gather additional information about Fuchs' Dystrophy.
- Request printed material about Fuchs' Dystrophy or corneal transplants.

Choosing a doctor or surgeon?

Endothelial Keratoplasty (EK) surgery (DLEK or DSAEK) should be considered instead of PK (Penetrating Keratoplasty) in the treatment of endothelial dysfunction such as corneal dystrophy.

The **choice** of an EK surgeon should be based upon their honesty and commitment to you as a patient, and their commitment to making your EK surgery the safest possible experience.

What questions should I ask my doctor or surgeon?

Below is a list of questions to help you get a discussion started with your doctor.

- Have you ever treated a person with Fuchs' Dystrophy?
- What type of corneal transplants have you performed?
    - Penetrating Keratoplastys (PK)?
        - What is your total number of PKs?
        - What is your primary graft failure rate?
        -
    - Partial transplants (EK?
        - What surgical technique do you use for Eks, 3mm or 5mm incision and why?
        - What is your total number of EK cases?
        - What is your dislocation rate (the graft had to be repositioned)?
        - What is your primary graft failure rate?
        - What is your pupillary block rate?
- How often are cornea transplants successful?
- What vision can I expect afterward?
- What complications might occur?
- Does my eye show any signs of glaucoma or cataracts?
- Will any other health conditions I have affect the treatment or progression of Fuchs' (diabetes, high blood pressure, etc.)?
- What tests did you run on me today and what were the results?
- Should I make any lifestyle changes (dietary, quit smoking, etc.)?
- Should I watch for specific symptoms and notify you if they occur?
    - Should I notify you immediately or wait until my next scheduled appointment?
- How might Fuchs' Dystrophy affect my vision now and in the future?
- What should I be expecting to happen, and about when might this occur?
- How will we treat my Fuchs' Dystrophy?
- Can I wear contact lenses?
- Should I ever consider laser surgery? Should my children?
- What is my eye pressure (IOP)?
- Where am I in the progression of this disease?
- Can you guess my timeline for getting a transplant?
- Could you give me the names of any local agencies or organizations that assist people with vision problems?
- What is my cell count?
- What is the cell count of my new cornea?

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Credits

The Corneal Dystrophy Foundation, Fuchs’ Friends Founders, Moderators and all others who have contributed time, money and energy to the creation and continuing success of our two websites and the Fuchs’Friends Group on Yahoo Bulletin Boards wish to sincerely thank Amanda Chase, the original author of this Guide. Her initiative, creativity and skills in developing the Guide have earned her our heartfelt thanks.