The Corneal Dystrophy Foundation

"Focus on: Education • Inspiration • Vision"

Visiting Your Doctor

By: Robert Bellizzi

LEGAL DISCLAIMER

The information in this booklet is for educational purposes only. It should never be used for diagnostic or treatment purposes. If you have questions regarding a medical condition, always seek the advice of your physician or other qualified health professional. Our booklets and brochures provide a brief overview of rare diseases. For more specific information, we encourage you to contact your personal physician.

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"To give hope through knowledge and community"

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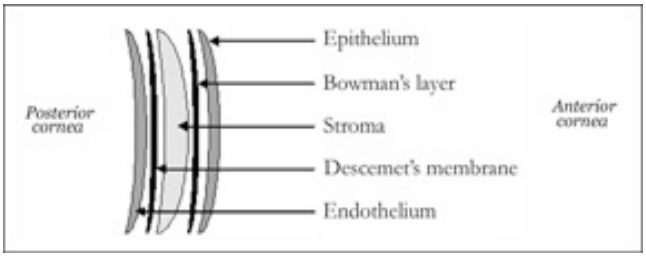


[i]Diagram of the Cornea (Front)

Contents

Introduction

Fuchs' Corneal Dystrophy (FCD[ii]) is a hereditary corneal disease. Once you have been diagnosed with FCD you will 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. It is important that you should feel comfortable asking questions and discussing your condition with the 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 someone you should interview. You might also join "Fuchs' Friends", our on line community. Open your browser and go to http://www.cornealdystrophyfoundation.org and click on the words, "To Join Fuchs' Friends" in the colored box; fill out the request form completely and click on Submit.

This brochure is intended to give you:

A brief description of FCD.

A description of 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.

Helpful hints to make your appointment with the doctor go more smoothly.

A list of suggested questions to ask your doctor.

Fuchs' Corneal Dystrophy Primer

FCD affects the cornea of your eye. The cornea is a clear window that allows light to enter the eye. It is about a half millimeter thick (1/254th of one inch), 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 corneal endothelial cells to die gradually as you age. FCD patients' cells die at a greater than normal rate. These cells cannot grow back once they die. Without enough endothelial cells to pump fluids away from the cornea, fluids collect in the corneal tissue altering the shape of the cornea and making vision cloudy and blurry.

Often the fluids collect in some parts of the cornea more than others and this causes the shape of the cornea to be irregular and wavy like a washboard. This waviness can cause astigmatism. If the cornea becomes too swollen with fluids, the outer part of the cornea (epithelium) may develop microscopic blisters. Blisters are a rare complication of FCD. Occasionally these blisters may rupture causing a sharp, stabbing pain. This usually happens after you have been asleep and you first open your eyes. If you think you have blisters you should contact your doctor so that he or she can monitor them and provide pain relief.

Typically FCD patients have the worst vision in the morning. Some treatments that dry out the cornea, such as concentrated saltwater drops or wafting warm air from a hair dryer[iii] across the eyes, can temporarily reduce the symptoms of FCD and improve vision. If vision becomes worse, a cornea transplant is the only way to correct the problem.

Six Vision Benchmark Tests and What They Mean

There are six vision tests, which your doctor may decide to run:

To confirm a diagnosis of FCD or

To monitor how your FCD is progressing.

You do not necessarily need each test to be run in order to have a complete examination and thorough diagnosis. The results of 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 Fuchs' Dystrophy

What the test tells you

The doctor will look at the endothelial cells in your inner cornea by using a slit lamp. 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 FCD 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 cornea. **Grading Scale**

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

Grade 0 indicates 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 you have 20/20 vision you are able to see characters roughly .34 inches or .887 cm high at 20 feet. 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 the test is done.

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 morning “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.[iv]

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. 20/20 vision is considered to be "normal." Most states test a driver's vision under normally lit conditions rather than glare conditions.

Endothelial Cell Count Test[v]

What the test tells you

An endothelial 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.

Note: Many doctors consider Corneal Thickness a better indicator of stage of Fuchs' Dystrophy than an Endothelial Cell Count.

Normal Range of Results

A normal cell count for people ages 40-70 is 2,000 to 3,000 cells/mm^2 (cells per square millimeter). A cell count of 500/mm^2 is considered extremely low, often resulting in the swelling of the cornea and blurry vision.

For intraocular surgery, such as cataract surgery, a minimum cell count of greater than 1,500 cells/mm^2 is recommended. The recommended number of cells for intraocular surgery can vary depending on the clinical situation.

Intraocular Pressure Test

What the test tells you

An intraocular pressure test measures how much pressure has built up inside the eye. This test is also used to determine whether a person is at risk for glaucoma[vi].

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 test will be:

The highest when you first wake up

Increasing as you get older

Higher if you're a woman

Will be more accurate if you avoid drinking alcohol for at least 12 hours and smoking marijuana for at least 24 hours before the test.

Corneal Thickness Test

What the test tells you

This test measures the thickness of your cornea. If the cornea is too thin 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 or the Zeiss computerized OCT machine which 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

FCD 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."Ц" is the symbol for microns. 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, 512.00 ц = 0.512 mm

Helpful Hints for Your Appointment with the Doctor

As you partner with your doctor to battle FCD 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:

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.

If you are uncomfortable adjusting your body to fit a machine, the machine or chair is not properly set up for you. This can cause back and other pain. Simply ask the machine operator if they would adjust the machine so you are in a more comfortable position and work with them till you are comfortable. This is not an imposition on the doctor or the staff and will allow more consistently true readings.

Ask questions until you understand your doctor's responses.

Bring a friend or family member into the doctor's office to take notes for you and to make you feel more comfortable, or take notes or use a tape recorder to help you remember the entire discussion.

Ask your doctor to write down his or her instructions if you don't have someone else to take notes.

Keep a journal to track your FCD and cornea transplant.

Talk to other members of your health care team, such as nurses and pharmacists to gather additional information about FCD.

Request printed material about FCD or corneal transplants.

Talking to Your Corneal Specialist or Doctor[vii]

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

Have you ever treated a person with FCD?

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 life-style changes(dietary, quit smoking, driving, etc.)?

What symptoms should I watch for?.

Should I notify you immediately or wait until my next scheduled appointment?

How might FCD affect my vision now and in the future?

How will we treat my FCD?

Can I wear contact lenses?

What about laser surgery? Is it safe for me, my children?

What is my eye pressure (IOP)?

Where am I in the progression of this disease?

What do you see as my timeline for getting a transplant?

What local agencies or organizations assist people like me with vision problems?

Do I need to know my cell count?

What do I need to do to handle this condition?

Since this disease is typically inherited what should I tell my children and when should they be tested?

Choosing a Doctor or Surgeon

In a jointly authored letter on January 29, 2009 to a major insurance supplier, Anthem, the American Academy of Ophthalmology, The Eye Bank Association of America and the Cornea Society all stated that the preferred surgery for Fuchs' Endothelial Dystrophies is [viii]DSAEK or DSEK which are in a class called Endokeratoplasty (EK)[ix] rather than Penetrating Keratoplasty (PK)[x].

Dr. Mark Terry[xi], MD, FACS, an originator of EK in the US, stated “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.” He suggested the following questions would be helpful to ask the surgeon to better understand his/her experience with EK surgery.

Questions to ask a Physician.

Do you perform Partial transplants (DSAEK or DMEK)?

What surgical procedures do you use for these surgeries?

What is the total number of EK procedures you have performed?

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?

Credits

The Corneal Dystrophy Foundation wishes to thank Amanda Chase, the original author of this Guide. Her initiative, creativity and skills in developing the Guide have earned her our heartfelt thanks. We also thank the Corneal Dystrophy Foundation Fuchs' Friends' Community for their comments and suggestions.

Endnotes

[i] Diagram from "Endokeratoplasty for Decompensated Corneal Endothelium" Used by permission of the Australian Government

[ii] The Corneal Dystrophy Foundation http://www.cornealdystrophyfoundation.org/html/cd_defined.html

[iii] http://www.permanente.net/homepage/kaiser/pdf/60649.pdf Kaiser/Permanente Fuchs' Dystrophy Overview 6/23/2009

[iv] http://sdhawan.com/glare.pdf Dhawan, Dr. Sanjay. "Glare." September, 2002

[v] Laing, Ronald A., Ph.D. "Clinical Specular Microscopy: In Vivo Findings of Specular Microscopy."

[vi] WebMDHealth. "Tonometry." October 2008.

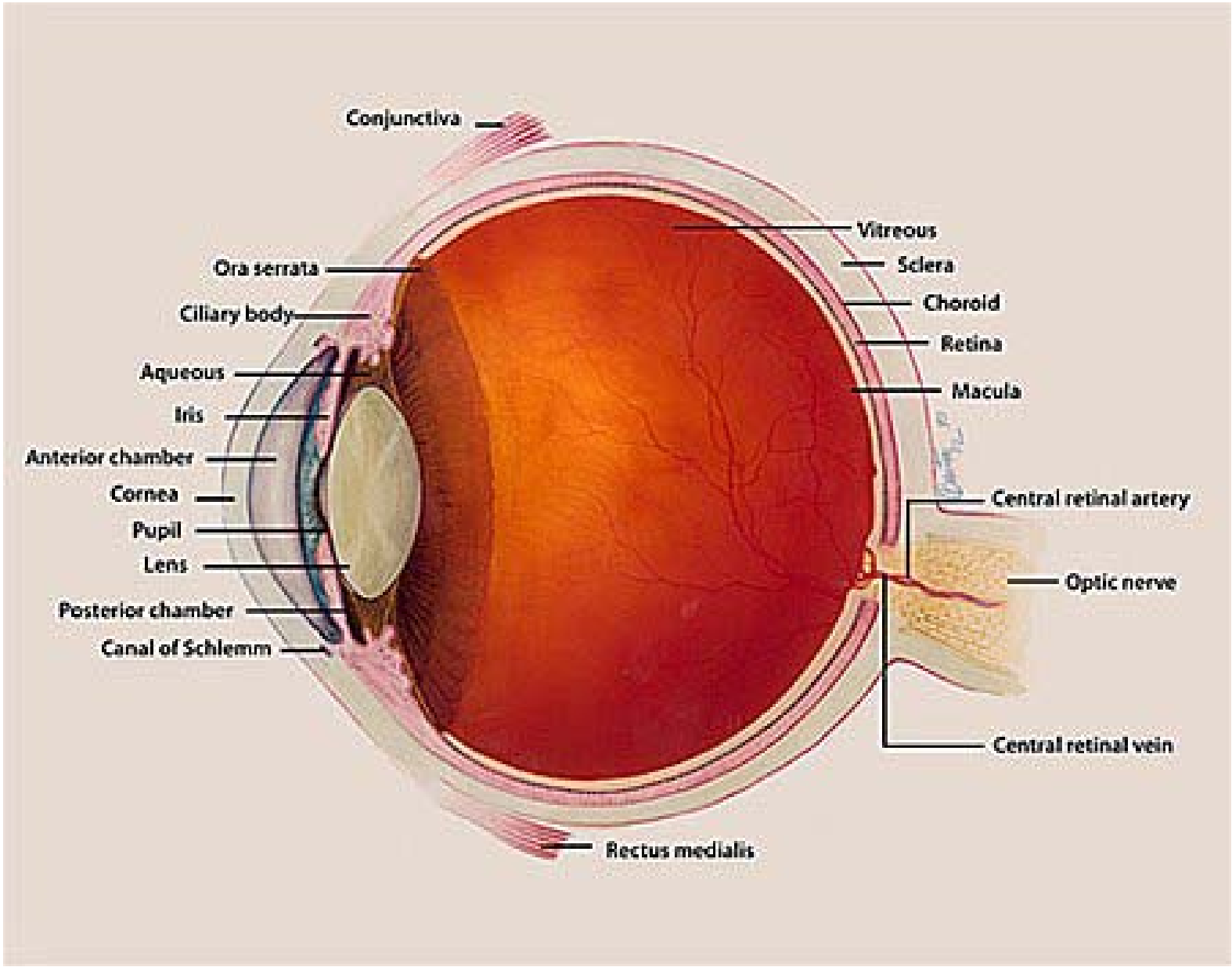
[vii] National Eye Institute. "Talking to Your Doctor." August 2002.

[viii] DSAEK or DSEK (Endokeratoplasty or EK) Unlike a PK, the DSAEK replaces only Descemet's Layer, the Endothelium and a small part of the Stroma. This is done through a small incision at the edge of the cornea without disturbing the remaining front layers of it. After this material is removed a 'graft' from a donor eye consisting of the same layers is folded, inserted through the same incision and made to open up and press on the rear of the recipient's remaining cornea by injection of an air bubble. The patient usually must lie prone for some time, dependent on the surgeon, and reduce activity for a prescribed time while the air bubble dissipates and, in a very high percentage of cases (~85%), the graft remains in place. If the graft 'dislocates' and doesn't stick to the recipient's cornea, it is usually repositioned and 'rebubbled' to keep it in place for another few days.

[ix] Endokeratoplasty - surgical procedures which remove and replace the corneal Endothelium without replacing most of the Cornea. DSAEK and DSEK are procedures of this type.

[x] PK (Penetrating Keratoplasty) The original corneal transplant; all layers of the cornea are removed from approximately a 9mm area of the recipient's cornea. A similar section (button) is removed from a donor's eye and anchored in place with anchor stitches and a running stitch around the periphery. The PK was the only choice of procedures for corneal transplants until approximately 2003.

[xi] "Endothelial Keratoplasty": History, Techniques, and Results Terry, Dr. Mark A.,MD,FACS, Director, Corneal Services, Devers Eye Institute, Portland OR Corneal Dystrophy Symposium, July 2007.


Conjunctiva
Vitreous
Ora serrata
Sclera
Ciliary body
Choroid
Retina
Aqueous
Macula
Iris
Anterior chamber
Central retinal artery
Cornea
Pupil
Optic nerve
Lens
Posterior chamber
Canal of Schlemm
Central retinal vein
Rectus medialis

To join our Fuchs' Friends point your browser to the main page of our website:

www.cornealdystrophyfoundation.org and Click on the colored box in the upper right corner that says, "Join our online support group. Click here."

Follow the instructions, fill out the application completely and click on the SUBMIT button. A moderator will process your application and issue an invitation within a day or two.

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