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NF2 Breakout

Ocular Manifestations of Neurofibromatosis: Relief for Dry Eyes

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D. Sclafani, a 1989 alumnus of ICO, began her career at the University of Chicago in 1993 where she is an Associate Professor of Surgery and Director of Optometric Services. Her main interests include contact lenses, corneal disease, eye trauma, dry eye and corneal surgery co-management. She was awarded the status of Diplomate by the American Academy of Optometry in the Cornea and Contact Lens and Refractive Technology section and is a frequent lecturer on these topics. She is the 1998 recipient of the Illinois Young Optometrist of the Year, earned the Residents excellence in Teaching Award in 1995 and the Outstanding Lecturer Award in 2001 from the University of Chicago, the 2002 Roger Kame Contact Lens Award, the 2004 Excellence in Education Award from the Illinois College of Optometry, 2009 ICO Alumnus of the Year and was invited to be a Distinguished Practitioner in the National Academies of Practice. She is a frequent contributing author for Review of Contact Lenses, Contact lens Spectrum, and Primary Care Optometry News. In 2008 she received the Illinois Optometrist of the Year and was selected by Review of Optometry as one of the Top 10 Females at the Forefront of Optometry. She has served on the Illinois State Board, is a Past-President of the Illinois Optometric Association, and Past Chair of the Cornea and Contact Lens Section for the AOA. She is on the professional advisory panels and speakers bureau for several drug and contact lens manufacturers and has served as one of team eye doctors for the NHL Chicago Blackhawks. She resides in Chicago with her husband Jeff McClimans (Optos, North America) and son, Liam. A "hockey mom" who enjoys travel, entertaining, wine tasting, triathlons and sailing.

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>> We're very pleased to have **Dr. Sclafani** here from the University of Chicago.

>> It's an honor to be here. Very humbling to be here because I am not a neurofibromatosis expert. However my patient population does include a lot of patients that have NF. So I'm very humbled to be around experts such as yourself presenting today. And I'm learning a lot from speakers as well as from patients. So it's nice to be here.

A little bit of a disclosure. I work at the University of Chicago. I've been there for over 20 years. By the way, I am extremely impressed with your typing skills. And I'll try not to use my Chicago-ese where I speak a little too fast but I'll try to get you out on time for lunch because I am what's keeping you between lunch.

But I am a consultant for a number of contact lens companies, a number of dry eye companies. And so I am privy to a lot of their research, a lot of the things and ability to use this technology on patients.

I'm also not a marathon runner. I have competed in some triathlons, sprint triathlons. I'm embarrass to say I don't have the courage to do it but I am proud about the people who do. It's wonderful.

There are a lot of different manifestations of Neurofibromatosis and whether the classification of its NF 1 or NF 2. A lot of the topics have been discussed earlier. We're aware of the café au lait spots and the Neurofibromatoses, the axillary and inguinal freckling, the optic nerve gliomas, Lisch Nodules and optic lesions. These are some of the diagnostic criteria to determine if one has **NF 1**. I thought I would spend time on the ocular manifestations because some of these might be the first diagnosis, the first symptom that a patient might experience.

So let's talk a little about Lisch nodules. When we examine this under the microscope, we examine the color in here, we might notice that the patient might have Iris freckling or pigmentation freckling. It is not diagnostic of NF 1 but over 90% of patients who do have Neurofibromatosis will present with these Lisch nodules.

These are very specific. These are discrete nodules. They're usually bilateral. They're very elevated. They're domed shaped, smooth and velvety. It's different than this type of pigmentation which is a little bit more sporadic and doesn't have that gelatinous kind of look to it.

And again it doesn't have any correlation with the severity or the prognosis of the disease, but it is one of the diagnostic criteria for NF 1 and it's present in about 90 percent of patients over 6 years old and will increase in numbers throughout their childhood.

Now, I was just in the other room listening to Dr. Tonsgard talk about optic nerve gliomas, they are extremely rare, however, they are present in NF 1 as you know. Up to 15% of patients who have NF 1 will present with optic nerve gliomas. 25 to 50% of optic nerve gliomas are associated with NF 1.

Now, symptoms may be no symptoms at all or it may cause severe vision loss.

The vision loss is due to the compression of these lesions on the optic nerve. And it is dependent on where they are present.

So when we look at this MR, you can see that this is the optic nerve here and there's a very high translucity of the optic nerve here representing a glioma, an optic nerve glioma. Now, pushing on the optic nerve can cause optic nerve disease, resulting in loss of vision. But it can also cause proptosis or pushing forward of the globe of the eye. So the eye will look like it's protruding out of it. Can also cause increased intercranial pressure. This causes less changes in vision, changes in your visual field or your peripheral vision as well as headaches.

60%, though, of patients who have MR findings or who present with optic nerve gliomas will present very early in life. And those patients, if it's found on an MRI, might not necessarily have visual symptoms. So visual symptoms don't always correlate with the presence of these.

In fact, if they are present more anterior, they're anterior to the chiasm occurs or the crossing over of the optic nerve, if it presents anterior to it, many of those patients are asymptomatic. And so treatment of those might not be necessary. In fact, treatment of any optic nerve glioma's controversial and there's not an accepted treatment for it.

75% of patients who will have an optic nerve glioma and NF will present in less than 10 years of age. It's very rare for them to occur later or for them to progress, as well.

If they are on the chiasm or post chiasmal, those will be aggressive and those patients will definitely need some form of treatment to reduce the risk of vision loss. That treatment might be surgical, although this is a very tenuous area for surgery and can cause lots of secondary problems. Radiation therapy or even chemotherapy for that treatment.

Now, glaucoma's also something that can occur in Neurofibromatosis. And this isn't a classic glaucoma that we see in our non-neurofibromatosis patients. This glaucoma that might occur earlier in life probably due to the presence of plexiform Neurofibromatoses. These large masses cause a little bit too much pressure onto the globe and that can increase the pressure. That's one of the thoughts that might be the cause of the glaucoma. Another is that the part of the drainage system called the trabecular mesh work of the eye may have a membrane called Barkan's membrane. And Barkan's membrane is prominent in congestive glaucomas. So glaucomas that occur in Neurofibromatosis, if it's associated with the Neurofibromatosis, will occur earlier in life usually in infancy. However, this is a very, very rare manifestation of Neurofibromatosis.

Again, it may be a result of this pressure on the globe. And patients who have glaucoma very early in life, because the eyeball is very flexible in terms of its ability to grow or to change in shape, those eyes with congenital glaucoma will appear larger. So the entire eye is larger. The cornea is larger and the effects are obviously much more devastating. These patients generally don't have a very good visual prognosis.

I think about 20% of patients who have NF 1 will have some type of osseous malformation. And the ocular manifestation would be affecting the sphenoid bone. So the sphenoid bone is part of the structure that maintains the globe in its socket. And this is a socket that has a present Sphenoid bone. And this is one that the sphenoid bone is obviously missing. It could be displastic or it could be missing, as it is in this case, or it could be partial or total. And the problem with this is that now we have some area for either brain matter to protrude through or a globe to protrude backwards. This may occur with or without Neurofibromatosis in the area. So it may not have anything to do with the little tumors.

The patients will have this associated defect or pulsating defect which we call, pulsating exophthalmus, it means the eyes are protruding in. If you look at this young man, you can see the eyeball is either smaller or larger. This is the affected eye. He has this proptosis. You look at him through a slit lamp you'll see that that eye is pulsating forwards and backwards where the other one is just in the normal position. And that has to do with the normal pulsing that reflects cerebral pulse that is more apparent on that side.

I have to tell you when I was in optometry school 28 years ago, the first patient that I saw with Neurofibromatosis happened to be a very good friend of mine. When you're in medical school, you have to examine each other. And I was examining my friend with a slit lamp and I noticed this pulsating proptosis. And nobody knew she had Neurofibromatosis. And she was in a state of not denial but she didn't talk to anybody about it. And it was the first open discussion that we were able to have. And so I was very honored that we had that discussion. And she's a good friend of mine to this day even though we live in different states.

So those are some of the diagnostic criteria for NF 1.

For **NF 2,** obviously we know that one of the basic criteria is the presence of bilateral acoustic neuroma. We will talk about that in just a minute. But I wanted to talk about a couple more ocular manifestations of NF 2 and one being cataracts. Now we're very familiar with cataracts. If we're all lucky enough to live long enough to develop cataracts, we're all going to have cataract surgery. I'm sure a couple of you in the room have had cataract surgery or are contemplating it. We look at the front of the eye. We see the normal lens is found behind the Iris here. And as we mature, that lens starts to change. Our vision will change with it. You can see a clouding of that cataract here, of that natural lens and a clouding of the vision and it's very gradual. Cataracts can progress and one can have cataracts for 15, 20 years without needing any cataract surgery. The reason why it progresses slowly is because there are different types of cataracts. This is the most classic type of cataract called nuclear sclerosis. It's a yellowing of your natural lens. It doesn't cause cloudiness, per se, but it does change your eye glass prescription. So one gets what we call either second sight or more near sighted. So your eye doctor might say your prescription's changing and it kind of reflects the fact that you have this cataract. This is not the cataract we're talking about. This is age related. You can see there's a whole bunch of others. This is pseudo exfoliation syndrome the front surface of the lens starts to shave off a little bit. This is a sun burst cataract or a cortical spoking cataract. This is a Christmas tree cataract. If you look at this patient you'll see beautiful different colors, reds, greens and blues in there. Then we also have the cataract which causes protrusion or ectasia, or pointiness, in that cataract.

The type of cataracts that our patients with Neurofibromatosis 2 get are pseudo posterior subcapsular cataract. So instead of getting yellow or having these little spokes of opacities, we get these very large opacities. And they're in the posterior back surface of the lens. And those tend to affect your central or your near vision first and then progress to distance.

This is a young man who was diagnosed at about seven years old, saw him at University of Iowa recently. This is his posterior subcapsular cataract. The good news about cataracts, though, is they are very, very treatable as you know. That natural lens is removed. Then artificial lens has been put in place. Relatively outpatient procedure under local anesthesia, most patients fare very well. There are complications but very rare. This is a different kind of implant. In the past we would correct you for one specific distance. Do you

want to eliminate the need to use glasses for distance or close? It's called monovision. That's one type of cataract surgery. But there's been a new wave of using multifocal IOLs, or multifocal intraocular lenses. These are like bifocals in your glasses where you have to look down these are concentric and contained within the pupil. And these implants are for patients who are younger cataract patients, meaning patients who have never had to wear bifocals. So under 40 years old. They're often very good candidates for these. Or patients who have only one cataract, maybe the other eye is not affected. And so that other eye is going to have normal functioning throughout its life. It's going to have the ability to focus up close and far away. This is a good option for the other eye that has that cataract implant placed in there because this cataract lens will help you up close and far away.

Acoustic neuromas are probably one of the biggest concerns that we have with our patients with NF 2. Obviously you know that it is the criteria for NF 2. And it is a Schwannoma. It's a very slow growing tumor which manifests itself on the sheath surrounding cranial nerve 8 and affects the inner ear and it represents 8% of all primary brain tumors. Patients who suffer from this may be asymptomatic. They may suffer from balance issues or obviously hearing loss. So the spectrum is quite large.

In general acoustic neuromas affect people between the ages of 30 and 60 and women more often. It's a malfunction on the chromosome 22 T. I'm not going to go into Merlin cells because I know you heard that this morning and that is not my area of expertise. But as you can see here, this patient has if we look at where the acoustic neuroma is, there's a lot of nerves around it, we have the facial nerve, the vestibular and of course the cochlear nerve. Because of that, it has so many manifestations, vision as well as hearing loss. The sporadic form, 95% of them are acoustic neuromas. And as I mentioned occur in these decades. And unilateral ones can be associated with NF 1. However, once we see that it's bilateral, it's almost 100% associated with NF 2.

And treatment for it obviously is dependent on its severity and how it's affecting one's functional life. In general, surgical treatment can be used. 95% of the tumor can be eradicated. Some of it is left so as not to affect the surrounding nerves and maybe radiation therapy might be used in conjunction with it. But one of the problems, the complications that we see affects the eyes and affects the tear function.

Damage to cranial nerve 7 affects both the motor functioning of the face. And so patient will have loss of motor function, drooping as you can see. This patient here had bilateral neuromas and she had the nerve 7 palsy. You can also use the secretory, secretions of lacrimal gland. It is located right here. The lacrimal gland is responsible for a certain production of our tears which we'll talk about. Cranial nerve 8 leading to deafness or balance loss. And cranial nerve 5 the trigeminal nerve can lead to sensory loss or facial numbness.

And as you probably are aware, about 5%, 5 out of 100 will have a recurrence of these types of tumors, neurofibromas.

So you saw her face droop. I want you to look a little bit closer at the lid drooping. This lower portion here is supposed to have very good apposition to the globe or to the eye. And the reason why is it will help in the spreading of tears and it helps in the production of tears.

As I mentioned, we have lacrimal gland located in this area here that secretes a large majority of the tears, but we have glands also associated along the lid margin that secrete tears that help prevent the evaporation. These are called meibomian glands when they have poor contact to the eye, we will have reduction in secretion and reduction in the spread of the tears. And this is a very extreme example of a patient who has the ectropion, as we call it, of the lower lid. Very little apposition of this lid to the globe.

And as a result of these, this poor apposition, one could get some punctate erosion. You can liken this to scratching on the skin. If one takes a finger and scratches their skin and you see the cells coming off or little dry patches, that's what's going on on the cornea. And it's excessive exfoliation. Our cornea constantly regenerates just like our skin does. If this occurs at an abnormal rate or if you're getting too many scratches on the cornea you will have an appearance like this. We call this staining the cornea. And at some point it can become very excessive and that staining can coalesce and one could get a corneal abrasion. And corneal abrasions are extremely painful and puts us at risk for infection and of course our vision gets impaired with that.

So leading to dry eye symptoms that one can have, that's those punctate erosions that I just showed you cause patients to have a foreign body sensation, a gritty feeling. And you can imagine if you're looking through a pair of glasses that have scratches onto, just imagine if you're looking through a cornea that has scratches on it. It impairs your vision.

And so there's a lot of different treatments that we use for dry eye depending on the patient's symptoms and depending upon the severity of it.

We'll go through a couple of these things. But starting out with just wearing glasses in general can help protect us from environmental factors. We'll go through it in a minute with all the different steps here.

There was a study that asked about quality of life in dry eye. And people who have severe to moderate dry eye liken it to having angina. So it is something that affects your daily life, as you probably are aware. People would gladly give up four years of their life to be symptom free of their dry eye according to this study by Schiff that published in 2003.

So there's lots of different ways to treat dry eye. One is you can restore the tear film meaning give you more tears. One is to maintain the tear film, meaning making the tears that you have more functional, of better quality. And the other is surrounding the whole premise of dry eye is what causes dry eye? Some believe that dry eye causes inflammation. And others believe inflammation cause dry eye. But we find it's a very vicious circle.

So when you look at somebody or you have the symptoms of dry eye, you're having symptoms of inflammation. You have cells that just aren't happy. When you think of all the diagnostic criteria for inflammation, heat, redness, all that. That's what people are experiencing on their eye.

So this is a classic picture of what we experience in our normal environment and some of the things that we can change. Treating dry eye is not always just treating the eye. We've got to treat our lifestyle and our environment first. In this picture, by the way, this is a coding expert. You can kind of sympathize with her job, doing a lot of billing and coding. But people who work behind a computer are exposed often to forced air

or air conditioning or heating units, first of all, that might be coming right into their eyes. So repositioning that is very important.

But working on a computer and working at it the wrong distance is a problem. A lot of times patients, first of all, you could imagine that when you work on a computer, you don't blink. And we know that. In fact, your blink rate is reduced by 50% throughout the day when you stare at a computer all day. And blinking is very essential for the spread of tears. You can have as many tears as you want, but if they're not spread properly, you're going to have symptoms.

The other thing is when people look at a computer, quite often they're looking at it straight on. Or sometimes even up. And that's the wrong position. First of all looking up is bad for your neck so that's not a good thing. But you should really lower your computer level to about 20 degrees. And when you do that, you have less exposure of your globe, of your eye ball because your lid will cover more of the cornea and again help spread the tears. So looking down and of course not having air forced at you.

Obviously certain climates will be more. You will have more risk of having dry eye. Smoking is a risk factor. Menopausal women, higher risk factor for dry eye. Certain foods. We used to say that caffeine contributed to dry eye, however there are some new studies that show that it might actually stimulate tear production. So the word is still out on that.

Obviously some outdoor activities. Refractive surgery can transiently cause more dry eye, meaning patients in their post op period might experience more dry eye because when we use some of the instrumentation on the eye to keep it open, we can sometimes destroy or kill off some of the goblet cells. These are cells that are on the eye ball itself. Cataract surgery as well will do that. But these can be restored over time, too.

I often wonder with eyes how things change with technology, what we're going to have in the future and if that will be more helpful for us with dry eye or not. I think of my son who is 10 years old and he's already suffering computer vision syndrome as we call it here.

So environmental as I mentioned, keeping the air out and all that, but vitamin supplements have been also helpful. We're very much aware of omega 3s. How helpful they are for anti inflammatory effects. The recommended dosage of omega 3 is 1,000 milligrams BID. Most of the literature is based on flaxseed oil. We know fish oil in its natural form is very helpful, as well. But for a dry eye, we lean a little more towards flaxseed. Both of them, though, and you can either get that in the form of your foods, salmon and your fishes or you can get it in tear supplements or vitamin supplements. Tears hydrate. Theratears, there are a lot out there that are produced that have the recommended dosage in there. They have the basis of having anti inflammatory effects, we have a good idea the heart effects they have on the heart but also they may help with the oily layer. So again the oily layer are the most external layer of the tears. We have three layers of our tears. One is right attached to the cornea. Then we have a very liquidly, watery level right in the middle. That's produced by the lacrimal gland as well as some other glands. And that middle layer is the layer that often gets overproduced. You know, some patients who have excessive dry eye actually have watery eyes. They'll come in with these very watery, teary eyes have and that's because that middle layer is having more of a reflex excessive tearing and the reason why is the exterior layer, the oily layer, that layer that prevents evaporation is not sufficient. It's either not being spread properly or it's not appropriate.

And the reason why it might not be appropriate is because, again, maybe you don't have the lid touching the eye or you may have something called blepharitis or meibomianitis. This is kind of like dandruff of the eye lashes. That's how I explain it to my patients. Because when we look at them, they are glands that are very much congested rather than secreting a nice oily layer or olive oil, you could appreciate the olive oil, our Italian friend over there. So we're supposed to secrete olive oil but instead these patients are secreting Crisco. It does not really spread the tears very well. So using some of these vitamin supplements and using warm compresses on the lid to help melt those oils, so melt that Crisco to olive oil will help with the dry eye, as well.

So you have probably gone to your eye doctor and they say yeah go get some eye drops. Put some eye drops in your eye and everything will be okay. I listen to my good colleague friends, my retina specialists and I hear them say that all the time and I want to just hit myself on the head because there are different lubricants. And I'm not going to get into too many details. One of my colleagues actually just hands this to a patient. And I'm laughing because whoever typed it up put "perserved" instead of preserved. They repeated it down here. She used to hand this out. And I could understand handing something out, but when you go to the store and you see all these different lubricants out there, you're not really sure which one to get. I can't really tell you without examining you what layer you need to replace but that's what you need to talk to your eye doctor about. Because certain lubricants replace different layers.

Here's a hotline called the dry eye talk. It's a website that patients can commiserate on online.

These are some of the newest brands, Sustained Balance and Systane Balance. This is for the lipid layer, this is for all the layers. This is also for the lipid layer, Refresh Advance. Blink Tears is made by Abbott labs. It has something called sodium hyperonic or HA in it. It's known to have visceral elastic properties. It may not mean much to you. But everybody who jumps up and down or bends their knees knows that when you do that, you are using your synovial joints. This is the same substance in those synovial joints that cause the joints to rub freely rather than crickety, crickety. This compound is also found in this lubricating drop. When you put the lid on your eye and the lids rub up against it, it has that lubriciousness. It rubs and is much more smooth and much more comfortable for the patient. It also has anti inflammatory factors in there, as well.

There's something called autologous serum doctor drops. This is the foo foo version. Once one finds that the over the counter lubricating drops don't work any longer or they're using them too often this, and we may have to move into something else.

First of all if you're using preserved lubricating drops, you need to be cautious. Some of these preservatives are toxic to the eye themselves and can cause problems. And some other drops we use, if you have glaucoma, those have preservatives in it, that can cause some dry eye. If you have allergies, some of the by mouth allergy medications, especially Benadryl can cause some drying of the eye. So although we're trying to treat eye symptoms, they can cause dry eye. So if you're using something that has preservatives in it, sometimes they are transient. Meaning the drop goes on the eye, once they combine with some of the enzymes on the tears, the preservative goes away. But estimates we need to use non-preserved. So if you're using it more than four times a day, we recommend patients to use nonpreserved lubricants. And these are available in single use form.

Now, if those lubricants don't work, there is one prescribed lubricant called Fresh Coach that was on my slide. I'm not going to talk about it. But autologous serum drops are custom made drops utilizing your own blood

serum. So blood is drawn. Your own blood is drawn and it's spun down and it's mixed with some non-preserved artificial tears. And the benefit of using it is that it has your own immune response in it. It has your own co-factors for healing. And it has growth factors and some enzymes that are good against inflammation. And these are often reserved for our most severe patients. This can be done at various hospitals or compounding agencies. And it does require a couple draws per year. And the serum, you could keep this autologous serum for about four or five months. Because it is not preserved, it needs to be kept cold. There is some risk of obviously contamination. So it does have to be done in a very sterile environment. That's something that's involved.

Now, it's important that, again, I know a lot of times our inclination is just to go to our drugstore and pull out any lubricant that there is. And I think it is important to pick the right one. I mean, I have patients who come in, I see somebody who is using lubricants right now. I see some patients coming in using a lot of Murine or things that say 'Get the red out." And the things that get the red out are the last things you want to use. And the reason why is it can cause constriction of the vessel, vasoconstriction of the vessels, which is great if you're going to go for a job interview or have a picture taken or something where you want to look nice. But if you used it every day, at some point your blood vessels are going to kind of poop out and they're not going to be helped. This drop actually causes your eyes to get redder and redder. It's called rebound hyperemia. So when a patient comes in and they're on that, the first thing I say to them is let's just take you off of this cold turkey. And it's tough for them but that's what you need to do. And then you need to get into some of these other lubricants. So like I say, people come with the craziest thing. This is red eye. I tell them that's the wrong way to use it. You should always use it that way.

[Laughter]

Most people will feel a lot better if they do that.

Finally there's another lubricant that is not a lubricant. It's called Restasis. Many of you may be familiar with Restasis. Restasis came out about eight years ago and it totally revolutionized the way we thought about dry eye. Before, dry eye was just a lack of tears. But now we know it's inflammation. And so Restasis is an immunomodulator. It's actually used in patients who have transplants and other systemic issues. But it's used to reduce inflammation on the eye. And it's a very low concentration, .05%. And it's used twice daily. Patients will not have any absorption systemically. It does get absorbed by the lacrimal gland and that has been shown to secrete more after use of Restasis. It does restore some of those goblet cells. Those are the cells on the eye ball that also secrete tears that we talked about. And reduces inflammatory that are found around the eye. So it is an anti inflammatory in this case, but it does not get absorbed inside the eye ball itself. In fact we can't find it when they remove part of the aqueous humor inside the eye. You cannot get it into the eye ball or end organs. The biggest complication or side effect that people might experience is some stinging when you put it in. But I tell my patients to keep it in the refrigerator. And they'll feel the coolness, instead.

Other methods of reducing inflammation is by the use of oral Doxycycline. Doxycycline as you know is an antibiotic. However when used in very small dosages, 50 milligrams BID, it's now an anti inflammatory. It has to be used on a long time period and then tapered off.

Another is topical Azithromycin. It's in the form for the eye is called AzaSite. We put that on our lid margin and that can help with those glands that we talked about, those meibomian glands. It can help with the secretions. It can make the glands create the right kind of tear.

So, after we treat with lubricants and we treat the environment and maybe we put the patient on anti inflammatories, another way is to maintain the tears that are on there. And this is called punctal plugs. Punctal plugs. We used to use these first before we knew that it was so much inflammation. But in the eye, there are two puncta, one on the top and one on the bottom. And if we close the hole or the drainage system so that the tears are retained on the eye instead of going down the drain, that may be of some help for patients. That used to be the mainstay. And I think it's a very good treatment for short term. Eventually a lot of patients will change their baseline. There's like a feedback loop, a neurological loop that the eye finally says after a while, do you know what? I don't need to be making so many tears. And so it just slows down its production of tears. And Steve Loofelder, famous dry eye expert, demonstrated that a couple times. But these are very easy to put in in a practice. These are what we call temps because we could put them in eye temporary, because we could put them in and take them out.

This is the drainage. This is the puncta here and here. We close off the drainage system so the tears stay on the eye. But if we find that the patients have spillage of their tears, too many tears flowing outside the eye then we can easily take them out. So you could talk to your doctor about that, too.

Unfortunately some patients who get these terrible corneal abrasions from this might need some more severe treatment. This is called a tarsorric (spellling?) view where we suture the lids together. Get that abrasion to heal and hopefully eventually get to take these off. If you have exposure issues in the middle of the night, meaning you can't close your lid and you have nocturnal lagophthalmos. There's certain things that you could have the patient close their eye. You put this tape on and it will keep the lid closed at bedtime. That's very helpful. Also, you know those little things that you put on that say "do not disturb" when you're on a plane or whatever so you can sleep. Those are really wonderful, too, for patients who sleep with their eyes open a little bit. My son has a little bit of exposure. So we put these on occasionally and it keeps moisture inside there and a little less exposure to the air.

Now, our patients who suffer from acoustic neuroma and who have surgery and end up with ***peripheral*** nerve 7 will have lack of thalamus. We asked the patient to close her lid and this is as far as she can close it. This is the extreme she would have. Treatment might include a treatment of a gold weight in this area. This is pretreatment. And post treatment. The gold weight will drop the lid. And so then she'll have full closure. Obviously we're using the muscles in this area to get that to drop down like that.

Patients who have this exposure interiorly can have plastic inserts. This patient here has a plastic insert here to get less of this exposure in that area there. This is a patient, I call bling for the eye because gold weights. Unfortunately hers is getting exposed. That was removed and our surgeons put a new one in. And this is it here. It's much more natural. Now I want to show you this again. She's got some exposure here. She does have a contact lens on her eye. We'll talk about her in a minute. That's when she closes her eye, how nicely it goes down.

So the other manifestations, as I mentioned we have the dry eye. And the dry eye can be due to these neurofibromas that occur along lid margin and reduce the spread of tears. If you look right here you see little

bumps in this area here, these little dots represent that Crisco oil coming out instead of olive oil coming out. She's got them all along this top region here. So they affect the spreading of the tears. But they also put pressure on the eye. And that pressure on the eye ball causes changes on the cornea which we call astigmatism. You're very familiar with what astigmatism is, right? Now some people have normal astigmatism, meaning their eye ball instead of being shaped like a basketball, it might be shaped like a football. But you can see that. And you probably heard your doctor say that. But you know that those two curves are 90 degrees apart, right, like a football.

People who have mechanical pressure on their eye like this may have astigmatism that is not so regular. It's much more irregular. And so their astigmatism looks like this. Remember a relief map. These are the valleys and these are the peaks. This is very irregular. And this our patient who has bilateral acoustic neuroma and she had a very irregular astigmatism and that was due not from these gold weights it was due to exposure because when the eye is exposed or if it has any surgical procedures, you can have this irregularity. I believe in her case she had so much exposure, she ended up needing a corneal transplant. And that corneal transplant has these irregularities involved in it. Now, you're going to see she has a contact lens on it. This is a rigid contact lens which we'll talk about now. The rigid contact lens fills in the gaps where we have all the peaks and valleys and corrects the astigmatism and also serves as a tear reservoir. Before one has those topographical changes or mild topographical one in order to have an abrasion we need to treat, not going away, rather than putting a Band Aid on, we put a soft contact lens on. Soft contact lenses could be used to treat abrasions or treat dry eye or they can be used to treat those little superficial cuts that we see. And this patient has that contact lens on her eye here. And what it does is it makes her feel better. It helps it heal that abrasion. And it gives her a more uniform optical surface so that she can see better. So soft contact lenses are one form of bandaged contact lenses to help with this.

Now, some patients end up having a disease called keratoconus. Keratoconus is an ectasia of the cornea. What that means is it bulges out. The reason the cornea bulges out is because it's thin. Imagine that you have a balloon that you blow up and the rubber tissue is thinner, it's going bulge. That's what keratoconus is. You can see extreme how bulging it is here. This represents that bulge. And so if you have a big bulge here, you got to correct it. And glasses don't correct it. So you'd have to put a contact lens over this whole area to fill in the gaps where there is lots of tears. So there's an association of kerato conus with Neurofibromatosis. And she's telling me more about NF because of watching her throughout her years and she had both keratoconus as well as Neurofibromatosis. She's got NF 1. And this is a lot about keratoconus. I'll skip that. More specifically with NF, there was a study review by Y. Rabinowitz at Cedar Sinai medical center. They wanted to see what it was. I showed you a long list there of associated systemic disease and Neurofibromatosis being one of them. And he looked at 300 keratoconus patients and only two of them had NF. Even though everybody associates it with NF, only 2 had it. So that represents .6%. Another had Down Syndrome, another .6%. But 99% of the patient who is had keratoconus had no associated systemic disease at all. So there is enough of an association that we link them together.

So why do they get keratoconus? They do have a very, very specific genetic makeup. It's not found on the NF chromosomes, though. We have the keratoconus along the lids that might irritate the lid or patients might do excessive rubbing. Patients who rub their eyes too much either because they have lid abnormalities or severe dry eye or allergies, they are more likely to develop keratoconus. In fact 40% of the keratoconus population, 40% of them have lid allergies. So it definitely has to do with rubbing.

So what were the symptoms of corneal astigmatism, regular stigmatism or keratoconus? Patients will have blurred vision, distortion, double vision in one eye. So not binocular, meaning neurological. It's basically in one eye. Cover an eye and they see a couple of you. They may have sensitivity to light. Have halos. But the thing I see in my practice is this. Patient comes to me with what we call a bag of glasses. That means they've been to lots of different doctors, they tried different glasses and nothing works and that's because their disease is in progress. And they're not very happy about that.

These patients will get iron depositions on their cornea, they'll get folds on their cornea. They'll get this swirl like pattern. This is epidire deposition. These are folds in the cornea here. They'll get scars. And at the point where one gets scars because of ectasia is the point at which we might have to consider corneal transplant surgery. What do we do before that? Sometimes they get breaks in their cornea that causes fluid to flow in. These patients if you have them look down, they have this angulation of their lid representing that cornea. You shine the light on one end and it focuses on another one. These are fun party tricks you can do.

So how do we treat these patients with keratoconus? I'm going to say keratoconus but I'm also going to say patients who have extreme dry eye, as well. So patient who have keratoconus were initially fit with these small diameter contact lenses that fit within the cornea. Eventually we went larger. Because we found that patients, if we go larger, we can get away from the cornea and it's a little healthier. I want you to see. See where all that yellow is? That represents where tears are. Eventually we went larger. This is the whole thought process behind treating dry eyes with these special lenses because these are called sclerolenses. Sclerolenses are designed to not rest on the cornea buttress then on the sclera and create a tear bank underneath the lens to provide moisture for the cornea. These were a gift to me from a patient. Her little girl had I remember I was treating her but her grandfather was an optometrist who fit the first contact lenses in the 60s. Now if any of you wear contact lenses, soft contact lenses are about 14 millimeters. These are 23 millimeters. And these are hard contact lenses. When she gave to it me as a gift, I got all excited because first of all I saw these but first of all I thought it was jewelry because she gave to it me in this beautiful leather box. I was kind of honored to have those in my office still.

As you can see we fit these sclerolenses and some of the problems we had were lots of bubbles and deposits on them but sclerolens versus come a long way. And these Sclerolenses designs as I mentioned before they rest here on the cornea and they vault over the cornea or the diseased tissue. Another example. Vaulting over the diseased tissue. And again another one. Fluorescence filling in. Little bit of touch on the diseased tissue but it rests over it. I'll go right to this one here. So this is an image showing here's the corneal tissue. Here's the lens. And you could see a very large .33 millimeter gap where tears fill in. And in that gap is that reservoir to help our dry eyed patients. And it's very helpful. There's clinics around, if you're in the Midwest, there's clinics around here that could fit you in these Sclerolenses if you're suffering with these. Some of these are custom made specifically to your topography and some are more stock fitting but it does require specialized fitter to do these.

And this is a patient who has a lens from another, scar tissue where the lid and the cornea kind of attached to each other and this lens, we put a little notch in it. And it fit over that, helped that patient quite a bit.

This is called Clearkone. It has a different lens. It has a hard center. And that hard center helps with the topographical problems but it's got a soft skirt so people who don't want to wear full hard contact lenses

because maybe they just don't like the way it feels or they're concerned about the handling, they benefit from these clear cone lenses. And again hard center, soft skirt. Very helpful for patients. And then finally the KeraSoft lens was recently launched. This was a soft lens for patients who have corneal irregularities. It's like in its fifth year of development. And this is another lens that can promise to give a lot of help for our patients who have dry eye and topographical changes with it.

Finally if our patients have severe dry eye because of this exposure of their lid issues or if they develop keratoconus and they end up with scar tissue where they can't see through that cornea any longer, they may be a candidate for corneal transplantation surgery. And for this type of treatment, what we do is the host tissue is cut out about 8 millimeters and a donor tissue is placed on there. It's sutured in. I will tell you that cornea transplants are very, very common procedure done in the United States. About 40,000 are done a year. There's not a long wait time for corneal transplants. Other countries, it's totally different. Much different. Other countries are experimental; they're doing a lot of nonhuman donors. They're using plastic graphs instead. In the United States those plastic graphs are generally reserved for keratoprosthesis. Those are generally reserved for failed corneal transplantation. But this is a very successful procedure in this country.

These are ring segments. These are plastic inserts put in one's corneal tissue to change that topography so they don't have so much irregular astigmatism. And this is also done on keratoconus.

Finally the newest procedure is something called cross linking. The purpose of cross linking is to make that cornea harder. We make it harder so it doesn't bulge so much. And ultraviolet A light combined with riboflavin is applied onto the cornea for about 30 minutes. And basically what it does is it ages your eye because as we mature, our cornea does get stiffer. So with regards to the corneal progression, it doesn't progress as we mature. It's much more of a younger disease.

So this is a procedure that's been getting a lot more attention in the United States. It's not FDA approved but there is a site in Chicago if you're interested in that. And the risks are very minimal.

So, with that, any questions? I know I kind of went a little fast on some of those topics. But be glad to answer any questions.

>> When you were talking about optic gliomas, it was specifically limited to NF1. Do you find that pretty much in NF 2?

>> DR. SCLAFANI: Not as often. I know that it could be in either form.

>> Because that was my son's initial diagnosis. Glioma and people wondered about that.

>> DR. SCLAFANI: Yes?

>> My son has NF 1. And as a child, he had several surgeries in his eyelid because he had a tumor in there. And he has the problem with the eye. It's dry constantly. It's red. We use the P tears the regular eye drops just don't do anything for him. With the redness and that, they said he doesn't make tears. He doesn't make a lot of tears. Has he been on Cyclosporine. They haven't done anything for him.

>> DR. SCLAFANI: Cyclosporine as I mentioned can increase tear production. That's one option. But you want to use that in combination with punctual occlusion can maintain what he does have. Punctal occlusion. Depending upon the the tarsorrhaphy I showed is pretty dramatic. That's not a permanent solution. For a temporary solution that's fine. But for a more permanent one, you could have a tarsorrhaphy done in the corner of the lid so you get less exposure, as well. Again does he have any closure of his lid?

>> Yes, he does. It just stays open slightly at the bottom. So it's constantly being exposed to the elements and that. But he doesn't have very much vision in it, in the eye itself. It's kind of distorted the eyeball itself. It's kind of distorted. Do you think that possibly that Contact lens?

>> DR. SCLAFANI: If he has topographical, the astigmatism, it will give it a more normal appearance. Sometimes if patients don't have enough of their own tears to fill in, then artificial tears are put in there and that reservoir is kept in there.

>> Thank you.

>> I was just wondering my daughter had Schwannoma removed and one of the things she has, no facial nerve paralysis but she does have dry eye. Is that ever temporary and it will come back?

>> DR. SCLAFANI: I'm not certain. How long ago is it? Can some of the nerves regenerate you're asking?

>> Right.

>> DR. SCLAFANI: Some of them can. Depends on if it's the main vessel or if it's more peripheral vessels.

>> DR. WELLING: One of the facial nerves goes to the gland. If that particular nerve is damaged, it may or may not come back. If it's been quite a while and still dry, then it's probably because that nerve was damaged.

>> DR. SCLAFANI: The lacrimal gland, yeah. Thank you.

[Applause.]