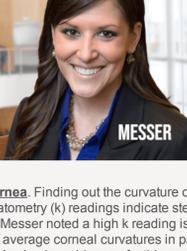


# March 2023

# Update



## Time to Celebrate Your Eye Doctor



Subscribers to *NKCF Update* can send a special **THANK YOU** to their favorite doctor before March 30. Share the story of how your optometrist or ophthalmologist makes a difference in your life. NKCF will send your doctor a special message of appreciation on your behalf. And check the next *Update* for a list of all doctors nominated and our choice for Top Doc 2023. Since NKCF began this National Doctors' Day celebration in 2017, 400 eyecare professionals from 45 states have been recognized by their patients for the care they provide.

[My Nomination for Top Doc](#)

## The Difficulty with Diagnosing KC

**Dr. Brooke Messer OD**, a keratoconus expert at Vance Thompson Vision in West Fargo, North Dakota lectured at the recent Global Specialty Lens Symposium in Las Vegas. She reminded eye care professionals there are several myths and misunderstandings about keratoconus, and offered evidence why this disease is so often misdiagnosed. While doctors may create a mental checklist of clues to the disease, she cautioned that not all signs are present for all patients. Here are some of the reasons why keratoconus may not be so easy for your doctor to diagnose:



- You don't need to have a steep cornea.** Finding out the curvature of a cornea gives doctors insight into the quality of a patient's vision. Higher keratometry (k) readings indicate steeper corneas; anything above k=48 is suspected keratoconus. However, Dr. Messer noted a high k reading is not essential to be diagnosed with keratoconus, and showed examples of average corneal curvatures in patients that had KC.
- Your cornea doesn't need to be thin.** Again, evidence of a thin cornea is a way some doctors discover KC. Normal corneas have an average thickness of about 550µm, anything less than 500µm may indicate a corneal irregularity or keratoconus. Messer noted there are numerous individuals with documented KC whose corneal thickness falls in the normal range.
- Both eyes need not be affected.** While keratoconus is considered a bilateral (both eyes) disease, it often has an asymmetrical presentation. One eye may show absolutely no signs of disease, while the fellow eye has irregularities. Messer cautioned that keratoconus could be the diagnosis even if it appears to be unilateral, and that doctors need to monitor both eyes at every eye exam.
- You don't age out of keratoconus.** While most patients are diagnosed in their teens or twenties, there are individuals who develop symptoms of KC later in life. Messer reported people in their 30s, 40s, or even 50s may learn they have keratoconus for the first time or experience new onset of disease progression. She advised doctors to continue to think about keratoconus as a possible diagnosis for patients with unexplained vision findings well into middle-age.
- You can have perfect vision and still have keratoconus.** Another myth is that patients with keratoconus can't achieve 20/20 reading an eye chart. On a simple vision screen, some patients appear to have perfect vision, but they may complain of excessive glare or trouble driving at night. In these cases, doctors are encouraged to think about keratoconus and to go beyond a simple vision test.

Dr. Messer's presentation showed that no single data point can guarantee a patient has keratoconus. She described how some indicators of the disease can be absent; a patient may have a combination of normal and abnormal test results. It underscores why this condition is often misdiagnosed, and why doctors need to gather all the information they can about the cornea strength, structure, and vision to make a definite diagnosis.

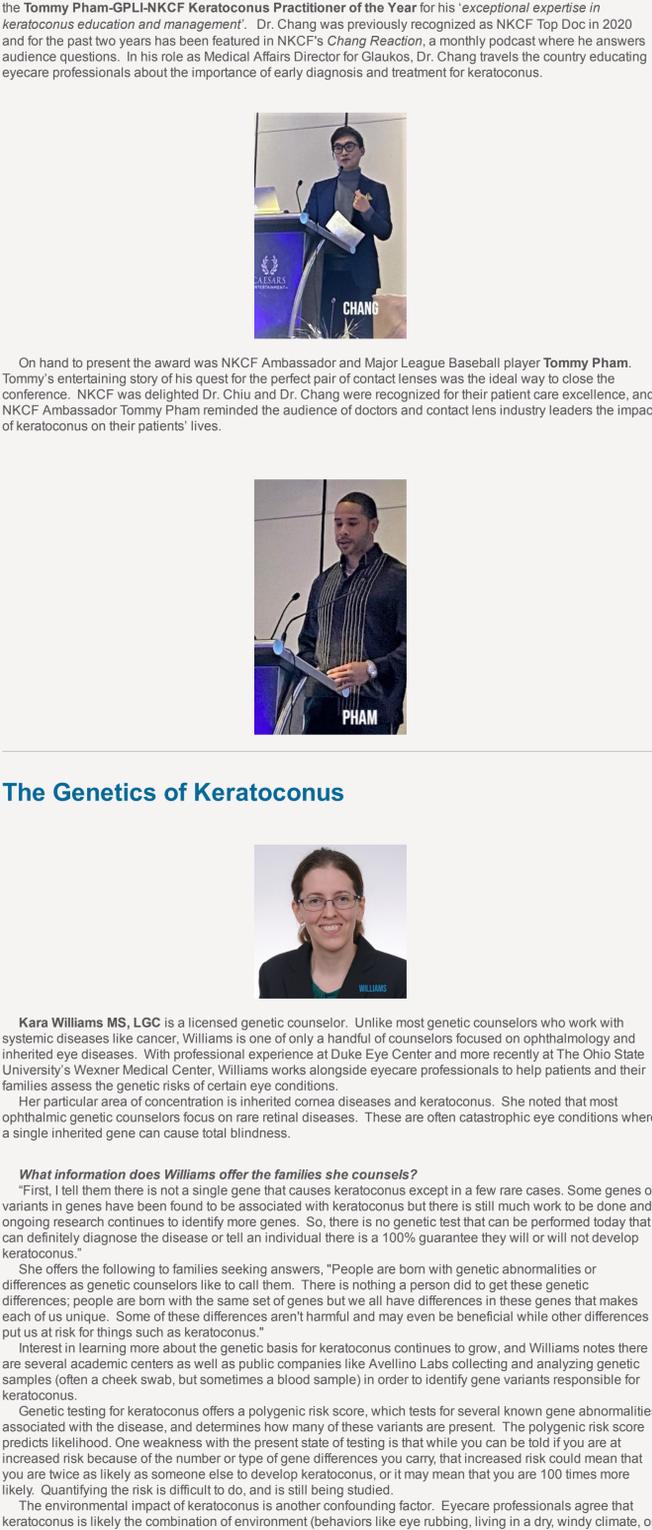
## How Long Does CXL Last?

One frequent question from individuals considering corneal crosslinking (CXL) is **"How long does it last?"**. The procedure that is recommended to halt or prevent corneal thinning and bulging and accompanying changes to vision was first described in 2003. Doctors at Carl Gustav Carus Hospital in Dresden, Germany reported that a combination of ultraviolet light and riboflavin eyedrops stopped keratoconus progression. **Fifteen-year results of the initial cohort of patients was recently published**, this represents the longest time period that individuals who undergo CXL have been monitored, and the results are informative.



Patients who had mild, moderate, or severe keratoconus were crosslinked between 2001 and 2006 using the Dresden Protocol, the epi-off method that is the basis for the FDA-approved treatment in the US. The mean age of these patients at time of treatment was 26 years old. Three years after treatment, the doctors reported that CXL was largely effective, with 92.5% success in stopping disease progression.

Now, fifteen years after treatment, 42 crosslinked eyes were examined and retested. The vision for 28 of 42 eyes (67%) was stable or better than 15 years previously. Corneal thickness and corneal steepness were also improved over the baseline results.



Six eyes (14%) required retreatment. **Retreatment is a repeat of the CXL protocol with ultraviolet light and riboflavin drops.** One eye was retreated nine months after the initial CXL. The remaining five began to show signs of increased corneal steepening after many years. One patient was retreated 7 years after the original CXL, two patients at year 11, and two at year 12. All patients who experienced treatment failure had severe keratoconus or suffered from allergies or inflammatory disease.

In cases where a second treatment was necessary, disease progression was halted. No patients required a corneal transplant or a third CXL treatment. No serious complications resulting from crosslinking were reported. Mild corneal scarring was detected in 36% of the patients (15 of 42 eyes), but these scars did not affect their vision.

These results should be useful to patients considering CXL for progressive keratoconus. In the majority of cases, the Dresden protocol results in stable cornea curvature and stable vision. However, **the procedure is not a 'one and done' for everyone.** Doctors should continue to monitor patients for changes that suggest a retreatment is recommended. This is especially true for patients with severe keratoconus or who continue to rub their eyes because of allergies or habit.

Reference: Corneal crosslinking of life stable and progressive 'early-stage' keratoconus patients. Steinberg J, Bassmann N, et al. Acta Ophthalmol. 99:e196-e201, 2021. Am J Ophthalmol, Jan 31, Online ahead of print, 2023

## Crosslinking Improves Quality of Life for Individuals with Progressive KC

Corneal crosslinking (CXL) results in stopping or slowing changes to the corneal curvature and stabilizing vision changes. In addition to the physical changes, are there psychological or emotional changes? It appears quality of life (QoL) improves for patients who undergo CXL.

In a 2021 study, doctors surveyed the quality of life of their patients with keratoconus. The doctors found that patients with mild, stable disease had responses very similar to the responses of study subjects with normal vision. When keratoconus progression was identified, QoL scores fell, even if the vision changes were mild. The authors suggest **the psychological burden of keratoconus is greater than many suspect** and patients struggle contemplating futures that may include increased dependency on others, or difficulty performing common tasks. The authors recommend limiting the time before CXL is performed to minimize decreased QoL.

A more recent study looked at patient reported quality of life before and after crosslinking. The survey tool used was the Keratoconus Outcomes Research Questionnaire (KORQ) which is designed to specifically capture activity limitations and symptoms that impact individuals with keratoconus. Seeing in the distance was the activity limitation most often reported, and the most common symptom reported was trouble with vision when tired.

A group of 39 patients within this study completed the KORQ survey, underwent crosslinking and then repeated the survey more than six months after the procedure. This gave researchers a rare before-and-after look at the impact of CXL on quality of life as reported by patients themselves. There were improvements in all categories. The authors noted the improvement in QoL scores were most noticeable for patients with severe keratoconus. "Although the primary aim of corneal crosslinking is to halt progression, it may improve patient reported outcomes, particularly the visual functional, symptoms and emotional status in people with keratoconus."

Reference: Quality of life in stable and progressive 'early-stage' keratoconus patients. Steinberg J, Bassmann N, et al. Acta Ophthalmol. 99:e196-e201, 2021. Crosslinking Improves the Quality of Life of People with Keratoconus: A Cross-Sectional and Longitudinal Study from the Save Sight Keratoconus Registry, Kandil H, Chen JY, et al. Cornea, Online ahead of print, PMID: 36729643, 2022.

**NKCF wants to hear from you! Share your keratoconus journey and help to inspire others. Each month we tell a personal story from one of our subscribers. To tell your story in a future Update, click [here](#).**



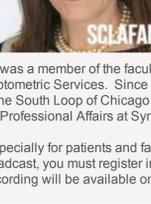
## Rising Stars of Keratoconus

NKCF was well-represented at the Rising Stars reception that closed the **Global Specialty Lens Symposium**. This event caps a three-day conference that brings together international experts, and focuses on improvements in contact lenses. Out of a handful of awards, two of the top awards were made to doctors with close ties to NKCF.

**Dr. Gloria Chiu OD** has been the moderator for the NKCF Evening Webinar series for the past two years. An Associate Professor of Clinical Ophthalmology at USC / Keck School of Medicine in Los Angeles, Dr. Chiu is a prolific author and in-demand lecturer, as well as a busy clinician. Dr. Chiu was awarded the **Practitioner of the Year**, awarded *for outstanding professional expertise in fitting customized contact lenses to benefit patients with improved vision and corneal health, and to advance the contact lens field*.



Another top award was made to **Dr. Clark Chang OD** of Wills Eye Hospital in Philadelphia. He was named the **Tommy Pham-GPLI-NKCF Keratoconus Practitioner of the Year** for his *exceptional expertise in keratoconus education and management*. Dr. Chang was previously recognized as NKCF Top Doc in 2020 and for the past two years has been featured in NKCF's *Change Reaction*, a monthly podcast where he answers audience questions. In his role as Medical Affairs Director for Glaukos, Dr. Chang travels the country educating eyecare professionals about the importance of early diagnosis and treatment for keratoconus.



On hand to present the award was NKCF Ambassador and Major League Baseball player **Tommy Pham**. Tommy's entertaining story of his quest for the perfect pair of contact lenses was their ideal way to close the conference. NKCF was delighted Dr. Chiu and Dr. Chang were recognized for their patient care excellence, and NKCF Ambassador Tommy Pham reminded the audience of doctors and contact lens industry leaders the impact of keratoconus on their patients' lives.



## The Genetics of Keratoconus



**Kara Williams MS, LGC** is a licensed genetic counselor. Unlike most genetic counselors who work with patients associated with the disease, Williams is one of only a handful of counselors focused on ophthalmology and inherited eye diseases. With professional experience at Duke Eye Center and more recently at The Ohio State University's Wexner Medical Center, Williams works alongside eyecare professionals to help patients and their families assess the genetic risks of certain eye conditions.

Her particular area of concentration is inherited cornea diseases and keratoconus. She noted that most ophthalmic genetic counselors focus on rare retinal diseases. These are often catastrophic eye conditions where a single inherited gene can cause total blindness.

**What information does Williams offer the families she counsels?**  
"First, I tell them there is not a single gene that causes keratoconus except in a few rare cases. Some genes or variants in genes have been found to be associated with keratoconus but there is still much work to be done and ongoing research continues to identify more genes. So, there is no genetic test that can be performed today that can definitively diagnose the disease or tell an individual there is a 100% guarantee they will or will not develop keratoconus."

She offers the following to families seeking answers, "People are born with genetic abnormalities or differences as genetic counselors like to call them. There is nothing a person did to get these genetic differences; people are born with the same set of genes but we all have differences in these genes that makes each of us at risk. Some of these differences aren't harmful and may even be beneficial while other differences put us at risk for things such as keratoconus."

There is learning more about the genetic basis for keratoconus continues to grow, and Williams notes there are several academic centers as well as public companies like Avelino Labs collecting and analyzing genetic samples (often a cheek swab, but sometimes a blood sample) in order to identify gene variants responsible for keratoconus.

Genetic testing for keratoconus offers a polygenic risk score, which tests for several known gene abnormalities associated with the disease, and determines how many of these variants are present. The polygenic risk score predicts likelihood. One weakness with the present state of testing is that while you are at risk if you are at increased risk because of the number or type of gene differences you carry, that increased risk could mean that you are twice as likely as someone else to develop keratoconus, or it may mean that you are 100 times more likely. Quantifying the risk is difficult to do, and is still being studied.

The environmental impact of keratoconus is another confounding factor. Eyecare professionals agree that keratoconus is likely the combination of environment (behaviors like eye rubbing, living in a dry, windy climate, or poorly fitting contact lenses that irritate the cornea) and a genetic predisposition for a weak cornea. Williams notes that the percentage of each needed to trigger keratoconus is unknown. "It is possible that someone with a small genetic risk can have multiple environmental factors that pushes them to develop keratoconus. We know there are others who have very few environmental influences, but still develop the disease. It is the chicken and the egg with keratoconus: **what causes this disease in certain people and not in others?**"

When Williams is providing counseling for individuals or families, she learns what high-risk they are seeking. "I ask families to think about what genetic testing will offer them. If you are told you have a high-risk score for keratoconus, will you be relieved to have a likely diagnosis, or will you feel guilty if you have a low-risk score while other family members have a high-risk score? Many families tell me they are seeking genetic testing for peace of mind, but that does not always occur."



She added, "Genetic test results for inherited eye disease are not always conclusive. In the best of cases, they may influence a decision on treatment choice, or motivate other family members to be screened for the condition. The undeniable benefit of genetic testing for keratoconus is, the more samples that are studied, the more our understanding of keratoconus increases, and this builds evidence that will benefit future patients."

**Genetic counselors collect detailed family medical histories and assess the risk of certain inherited diseases. To learn more, visit the National Society of Genetic Counselors ([nsgc.org](#))**

## Clearly KC is coming!



This month NKCF will launch a new podcast devoted to keratoconus. Our host is Dr. Melissa Barnett OD, principal ophthalmologist at Univ. of California, Davis, a leading authority on keratoconus and a long-time friend of NKCF. She'll be talking with patients and families affected by KC, as well as medical professionals and industry leaders. The goal of Clearly KC is to share information to help patients manage their keratoconus, seek out the best hybrid contact lenses, and live a full and productive life.

You'll find **Clearly KC with Dr. Melissa Barnett** on the usual platforms like Apple or Google podcasts, Stitcher or Spotify. Follow and Subscribe and you'll be alerted when new episodes drop.



Eye care professionals interested in learning more about keratoconus are invited to attend **Keratoconus: Front to Back and Everything in Between** on April 22 & 23 in Scottsdale, Arizona. The event will offer continuing education credits for ophthalmologists and optometrists and is sponsored by the International Keratoconus Academy, a professional partner of NKCF. For information or to register, click [here](#).

NKCF will host a roundtable at the upcoming ASCRS (American Society for Cataract and Refractive Surgeons) meeting in San Diego. Cornea specialists who perform crosslinking and who manage keratoconus in people with Down syndrome (DS) will exchange best practice tips for special needs patients. Individuals with DS have a higher incidence of KC than the general population; there is an ongoing need to educate parents, caregivers and primary care providers about the importance of screening for KC in this population. The **NKCF Task Force on Down Syndrome** was established in 2022 and is led by **Dr. Ann Ostrovsky MD** of New York University.



## Your EyeCare Team

Recent surveys of eye doctors explore referral patterns for their patients with keratoconus. Optometrists in different countries reported different referral models, largely dependent on their training and the diagnostic equipment available in their practices. In some places, a consultation referral is made to another doctor to confirm a diagnosis, or for specialized treatment like crosslinking. Sometimes, primary care eye doctors manage their patients' vision and eye health but refer to another doctor more experienced in fitting complex contact lenses.

It raised the question of what kind of interprofessional collaborations exists in the US. How often and under what circumstances are optometrists and ophthalmologists working on a team for their patients with keratoconus?

We've created a short survey to find out who you consider your primary eye doctor, and whether or not your doctor works with other eyecare providers who may have different training or expertise. Not all patients realize that the doctor who performs crosslinking or cataract surgery is a medical doctor and is likely not to be the same doctor you would visit for contact lens fitting.

Take our quick survey and let us know who is on your team. We'll summarize the results in the next newsletter. One lucky reader will receive a gift card for completing the survey.

[Take our 2-Minute Survey](#)

## Webinar Rewind: Keratoconus in Children



**Dr. Kathryn Hatch MD**, Assistant Professor of Ophthalmology at Harvard's Massachusetts Eye & Ear Infirmary was the NKCF Evening Webinar speaker on January 10. Her presentation, **Keratoconus in Children** offered an overview of the condition as it impacts the pediatric population.

She explained why keratoconus, when detected in young patients is concerning: "Our cornea tissue is extremely elastic when we are young and is more susceptible to changes. As the body ages, the tissue gets stiffer, and these changes are slower and less dramatic. **Keratoconus that develops at a young age is usually aggressive with more significant changes in a shorter period.** Ideally, we want to detect the condition as early as possible. We like to treat keratoconus before it become symptomatic with vision loss."

While no one is born with keratoconus, the disease has been diagnosed in pre-teen children. Dr. Hatch noted that while certain families having multiple members with keratoconus, most cases are sporadic and only a single individual in a family is affected.

She warned of the impact of eye rubbing. In response to an audience question, Dr. Hatch said that stopping this habit takes a team effort and that she enlists parents and other caregivers to remind the child not to rub his or her eyes. She spoke of the importance of treating any underlying causes for itchy eyes like allergies, and sometimes recommends wearing glasses or goggles as a barrier to eye rubbing.



In cases where keratoconus is suspected, Dr. Hatch may have the child return as frequently as every 6-8 weeks to look for evidence of progression. She will recommend CXL as soon as she has evidence the disease is progressing. Dr. Hatch has performed successful CXL on many patients under the age of 14; she cautioned that it is important to contact your insurance carrier in advance to find out their policy on reimbursement for crosslinking children. To view the **Keratoconus in Children** presentation, click [here](#).

## Evening Webinar Preview: Hybrid Contact Lenses for KC with Dr. Louise Sclafani OD

**MARCH THIS!**

Hybrids are not just cars that run on both electricity and gasoline. The term implies mixing two things to get something new. Hybrid contact lenses have been around for a while and take the good vision that comes from a GP (gas permeable or hard) lens in the center of the lens, surrounded by a soft contact lens. For many, the result is clear vision without discomfort.

Our guest speaker is the leading authority on hybrid lenses for keratoconus and irregular corneas. **Dr. Louise Sclafani OD** is a graduate of the Illinois College of Optometry and a fellow of the American Academy of Optometry.



For more than twenty years Dr. Sclafani was a member of the faculty at the University of Chicago where she was Associate Professor and Director of Optometric Services. Since 2017, Dr. Sclafani has split her time between a private contact lens practice in the South Loop of Chicago (SoLo Eyes), teaching at the Illinois Eye Institute, and her role as Vice President for Professional Affairs at SynerEyes, the division of CooperVision, that makes hybrid contact lenses.

Dr. Sclafani created this presentation especially for patients and families who want to learn more about hybrid contact lens options. To attend the live broadcast, you must register in advance. If you are not available to hear Dr. Sclafani's talk on **March 14**, a video recording will be available on the NKCF website.



## Join NKCF for the Evening Webinar Series - Second Tuesday at 5pm PDT / 8 pm EDT

Register for Live Events or visit [nkcf.org](http://nkcf.org) for past recordings

- September 13, 2022 - Brian Chou OD, San Diego, CA
  - Straightening out Keratoconus Misinformation**
- November 8, 2022 - Jack Parker MD, Birmingham, AL
  - International Perspective on Keratoconus Surgery (World KC Day Lecture)**
  - January 10, 2023 - Kathryn Hatch MD, Waltham, MA
    - Keratoconus in Children**
    - March 14, 2023 - Louise Sclafani OD, Chicago, IL**
      - Hybrid Contact Lenses for Keratoconus**
        - May 9, 2023 - Chantelle Mundy OD, Columbus, OH
          - Risk Factors for Keratoconus**
        - July 11, 2023 - John Gelles OD, Steven Greenstein MD & Peter Hersh MD, Teaneck, NJ
          - Addressing Keratoconus Vision Needs Throughout Life**

## New King of Queens!



**Tommy Pham**, keratoconus patient and Ambassador for NKCF signed a one-year contract with the New York Mets in January. Tommy has been a long-time supporter of this organization and has raised public awareness by sharing his ongoing keratoconus journey with the media. He's talked about frustrations and triumphs with other patients and eyecare professionals on an NKCF webinar ([Watch here](#)). Everyone who knows someone living with keratoconus cheers for Tommy's continued success and looks forward to watching him in the outfield this season. Follow him @tphamlv @mets.

## Garg Family Challenge

On World KC Day 2022, **Dr. Sam Garg MD** and his family established a challenge grant, matching donations made at year's end by optometrists and ophthalmologists. Dr. Garg is Professor of Ophthalmology at UC-Irvine and medical director for NKCF. He is also the chief medical editor of *EyeWorld*, a popular publication for cornea surgeons. Dr. Garg's challenge resulted in increased support for NKCF. A big **Thank You** to doctors who made contributions eligible for the Garg Family Challenge.

**Dr. Brian Chou, OD** - San Diego, CA

**Dr. Michael Greenwood MD** - Fargo, ND

**Dr. Stephen Kurtin OD** - Addison, TX

**Dr. Stephanie Pisano OD** - Columbus, OH



GARG

GARG