If you're living with cystinosis, you should know:

THE EYES HAVE IT

LOOK INSIDE ▸

to learn how to recognize different levels of corneal crystal accumulation due to cystinosis

Please see Important Safety Information about CYSTARAN® (cysteamine ophthalmic solution) 0.44% on page 2 of this brochure.

cystaran®
(cysteamine ophthalmic solution) 0.44%

What is CYSTARAN®?

CYSTARAN (cysteamine ophthalmic solution) 0.44% is an eyedrop medication used to treat cystine crystal accumulation in the corneas of patients who have cystinosis.

What is the most important safety information I should know about CYSTARAN?

- To help prevent contamination of the dropper tip and eyedrop medication, try to make sure that CYSTARAN is dropped directly onto the eye without touching it. Try not to touch the eyelids or surrounding areas with the dropper tip of the bottle when you are using CYSTARAN. Keep the bottle tightly closed when not in use.
- CYSTARAN contains an ingredient called benzalkonium chloride which can be absorbed by soft contact lenses. Remove contact lenses before using CYSTARAN eyedrops and wait at least 15 minutes before reinserting them.
- CYSTARAN should only be used as an eyedrop medication.

What are the side effects of CYSTARAN?

- The most common side effects of CYSTARAN, which have occurred in at least 10% of people using the medication, were sensitivity to light, eye redness, eye pain and irritation, and headache.

The risk information provided here is not comprehensive. To learn more, talk to your healthcare provider or pharmacist about CYSTARAN. The full FDA-approved product labeling can be found at www.cystaran.com.

You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.fda.gov/medwatch or call FDA at 1-800-FDA-1088.

WHAT TO EXPECT DURING A SLIT LAMP EXAM

- The initial test for corneal crystals will probably involve a slit lamp microscope (shown above), which is a bright light with a microscope that's used with most eye exams
- The slit lamp microscope can show whether you have crystals in your corneas and help the ophthalmologist give an estimate of how densely they are packed.
- They may use a CCCS, which stands for **corneal cystine crystal score**, to describe how densely packed the crystals are.*
- A CCCS goes from 0 (no crystals) to 3 (heavily packed with crystals) in increments of 0.25

* The CCCS scoring system was developed as a qualitative measure by the researchers at NIH for the study that supported theapproval of CYSTARAN

In the United States, around 600 children and adults have cystinosis. **All of them will develop corneal crystals**.

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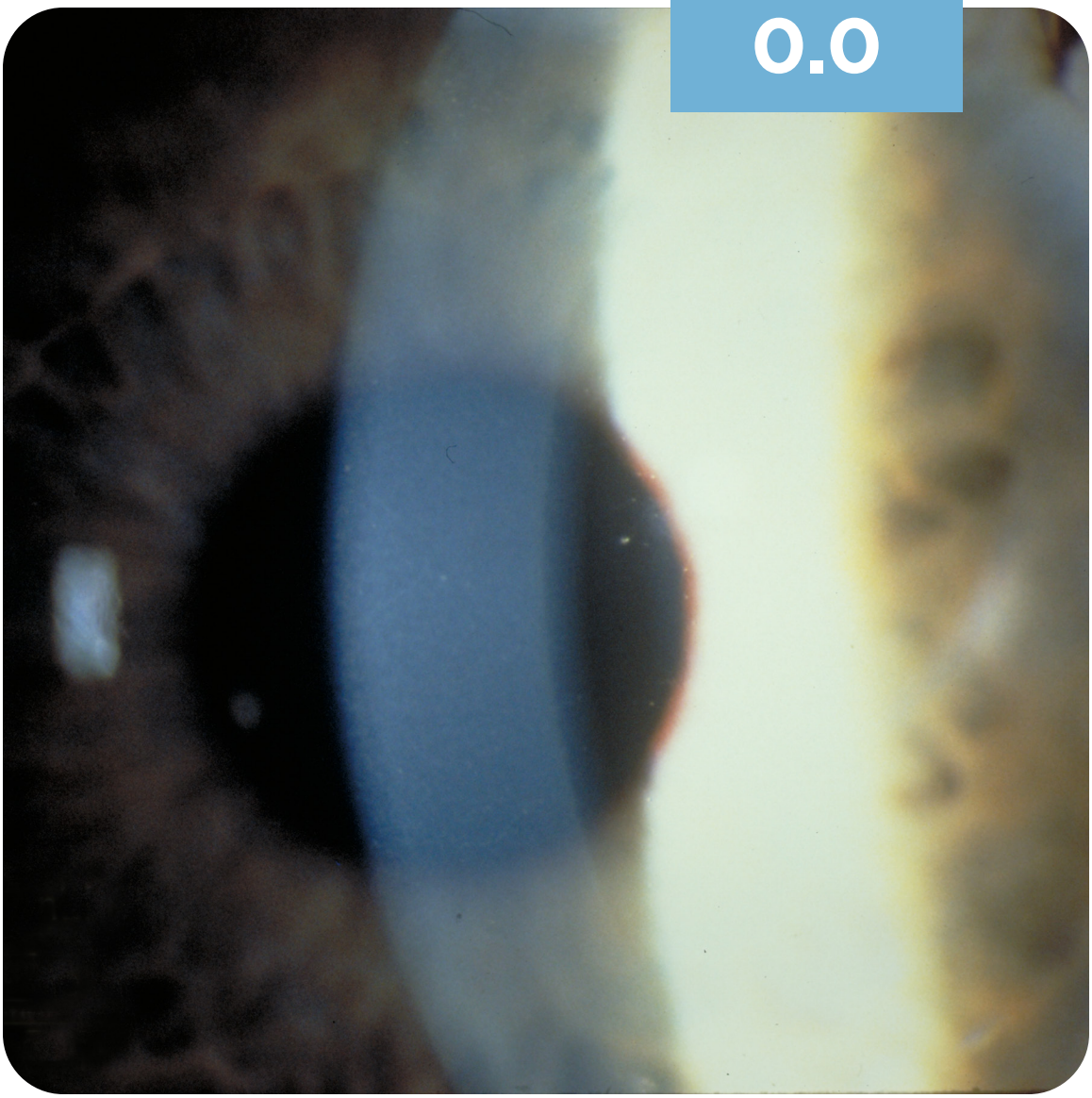

The slit lamp microscope shines a focused sheet of light in the eye to illuminate and magnify the anatomy of the eye. There are no crystals present in this eye.*

* Photos provided are examples only and are not intended to be a uniform representation of all cystinosis patients.

Doctors can detect crystals **as early as 6 months in some cystinosis patients and after 16 months in all patients.**

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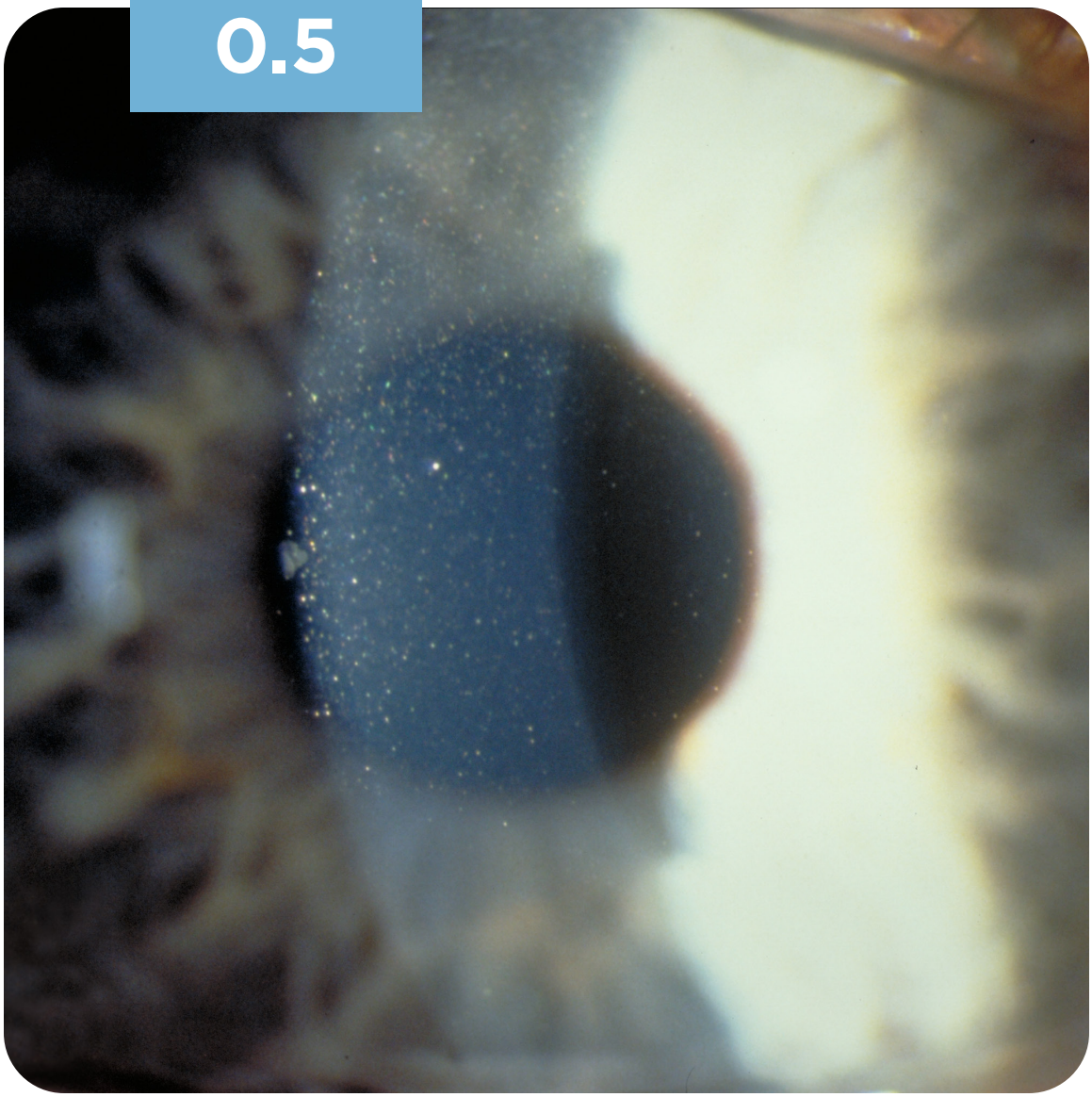
0.5



There are faint signs of corneal crystal accumulation visible in this eye with a CCCS of 0.5. Crystals appear as bright dots when the light from the slit lamp shines on them.*

* Photos provided are examples only and are not intended to be a uniform representation of all cystinosis patients.

Early care from an ophthalmologist (an eye specialist) can help cystinosis patients avoid eye problems that may impact their quality of life.

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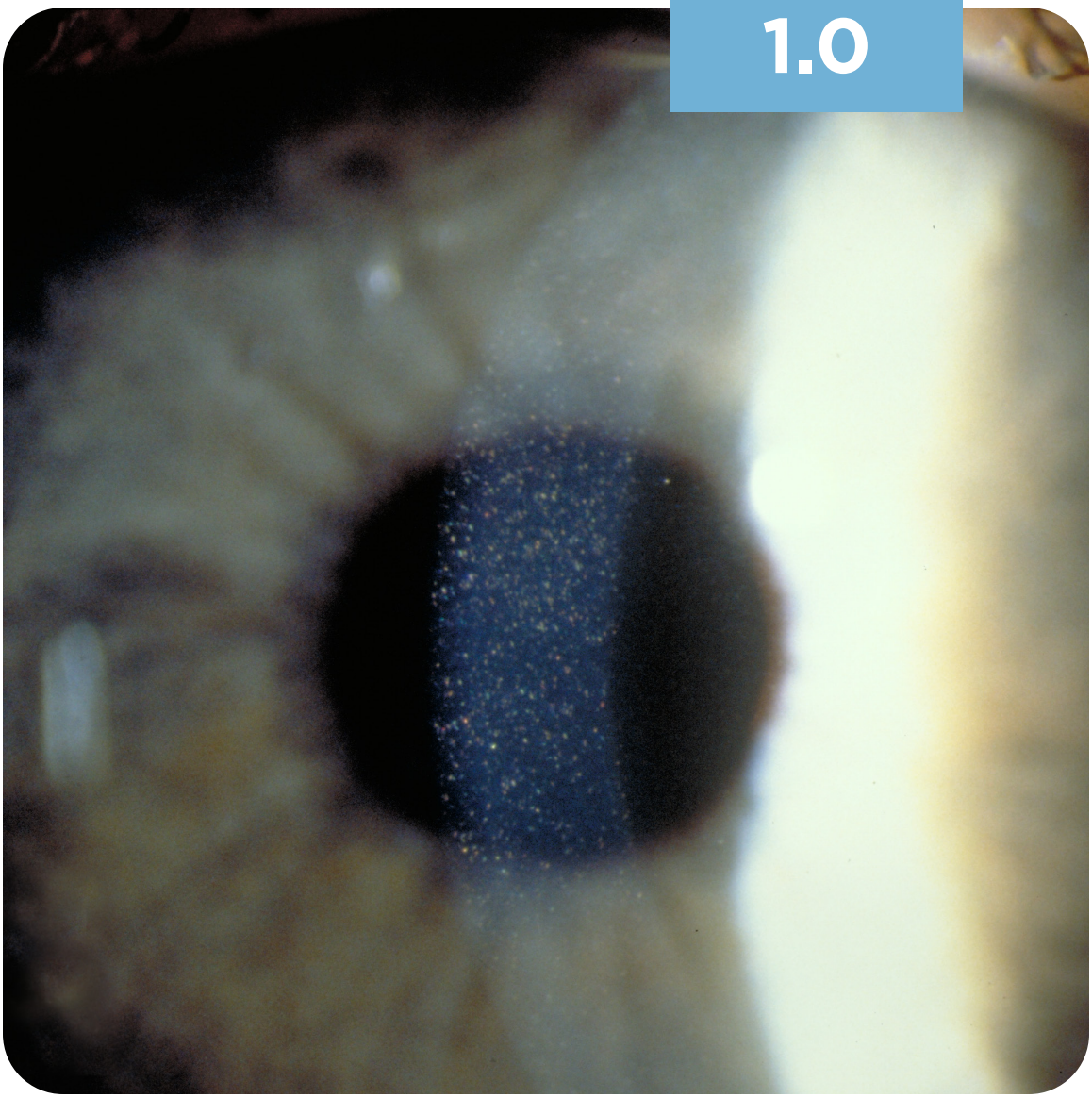

This is what an eye with a CCCS of 1.0 looks like. The crystals are spread throughout the cornea, but can only be seen where the light from the slit lamp falls on them.*

* Photos provided are examples only and are not intended to be a uniform representation of all cystinosis patients.

Cystine crystals are formed when lots of cystine (an amino acid) begins to accumulate in one place. The structures that form when cystine accumulates are called crystals because of the specific way the molecules arrange themselves.

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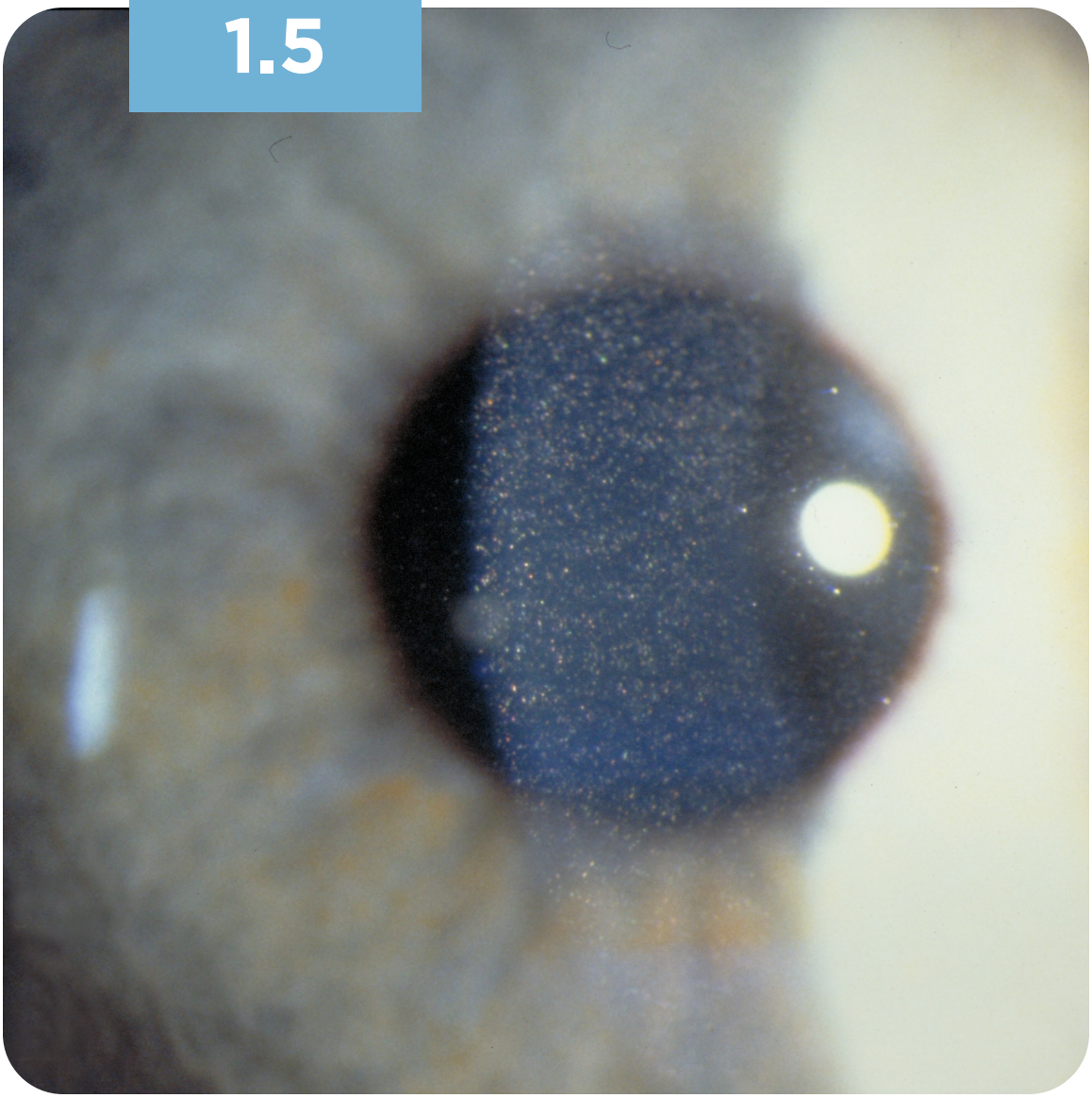
1.5



This eye with a CCCS of 1.5 shows signs of moderate corneal crystal accumulation.*

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Sensitivity to light from crystal accumulation in the corneas may be seen in toddlers but usually starts during late childhood or early adolescence (8-12 years).

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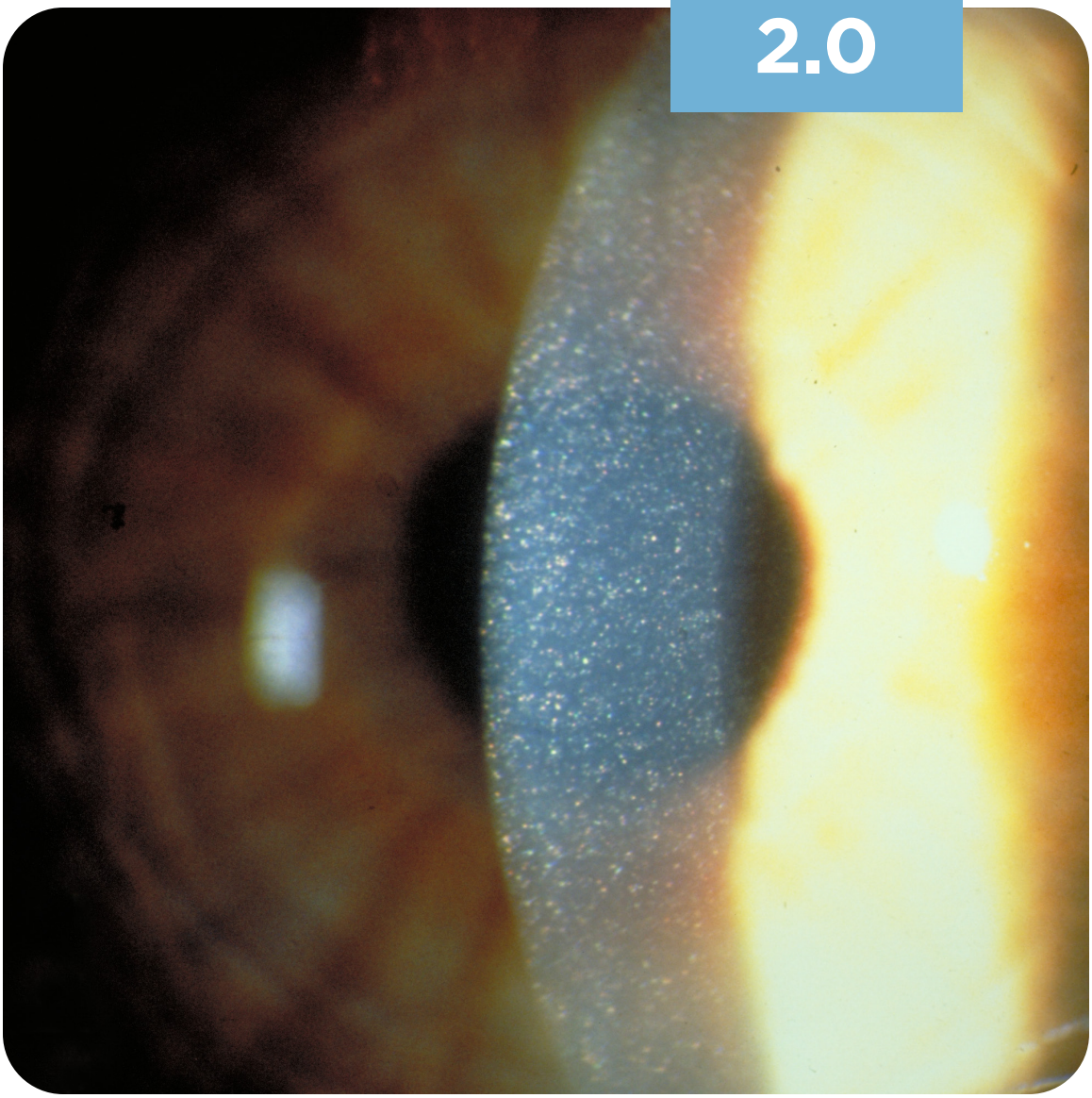

The bright, large specks that you can see in this eye with a CCCS of 2.0 are areas of heavy cystine crystal accumulation.*

* Photos provided are examples only and are not intended to be a uniform representation of all cystinosis patients.

Oral cystinosis medication can't reach the cornea because there is no blood supply to the cornea to deliver the drug.

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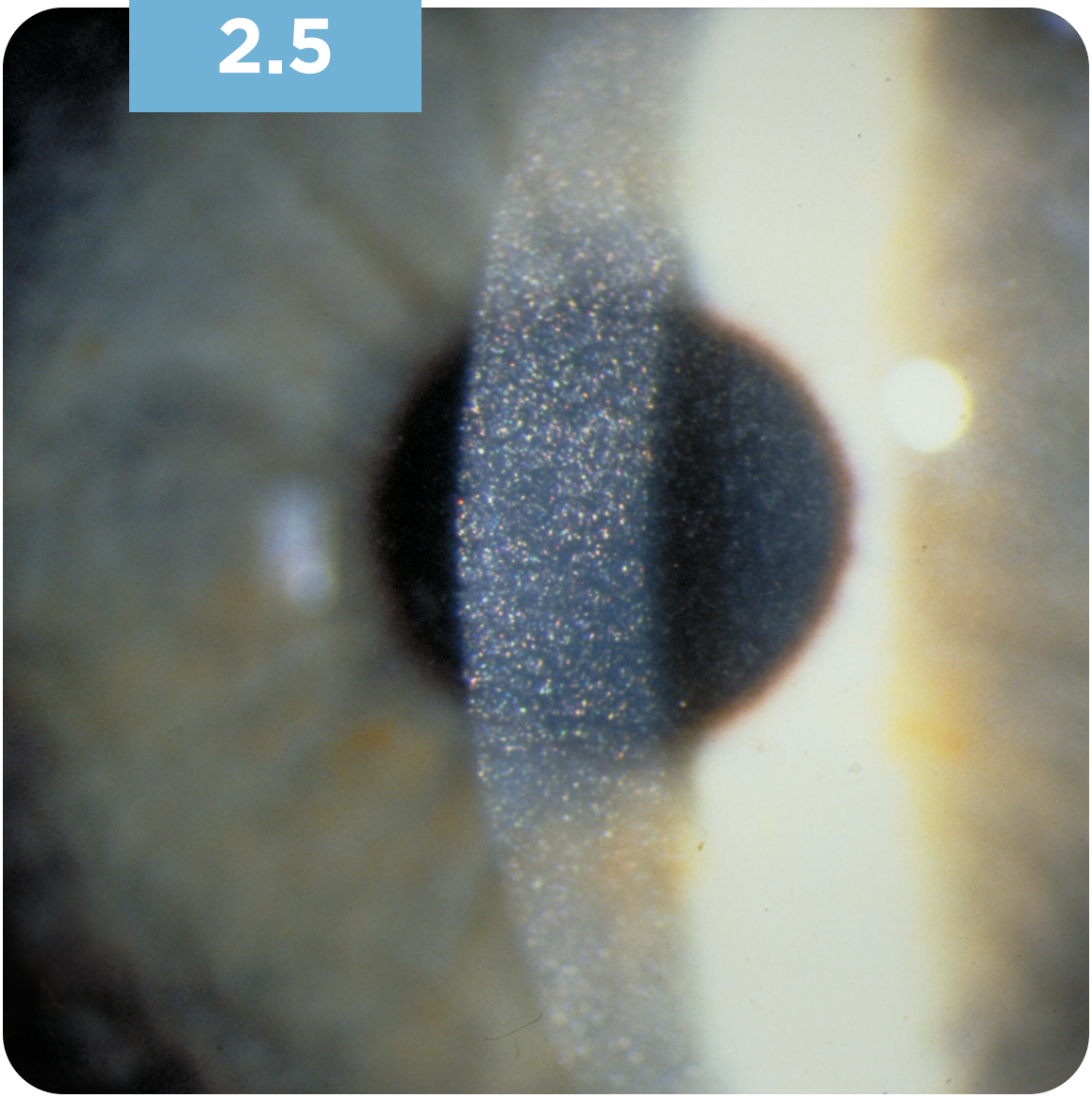
2.5



An eye with a CCCS of 2.5, like this one, will have almost no areas that appear free of cystine crystal accumulation.*

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In one study, no cystinosis patients older than 15 who had not previously received eye drop therapy were assigned a CCCS score of less than 2.5 during their initial examination.

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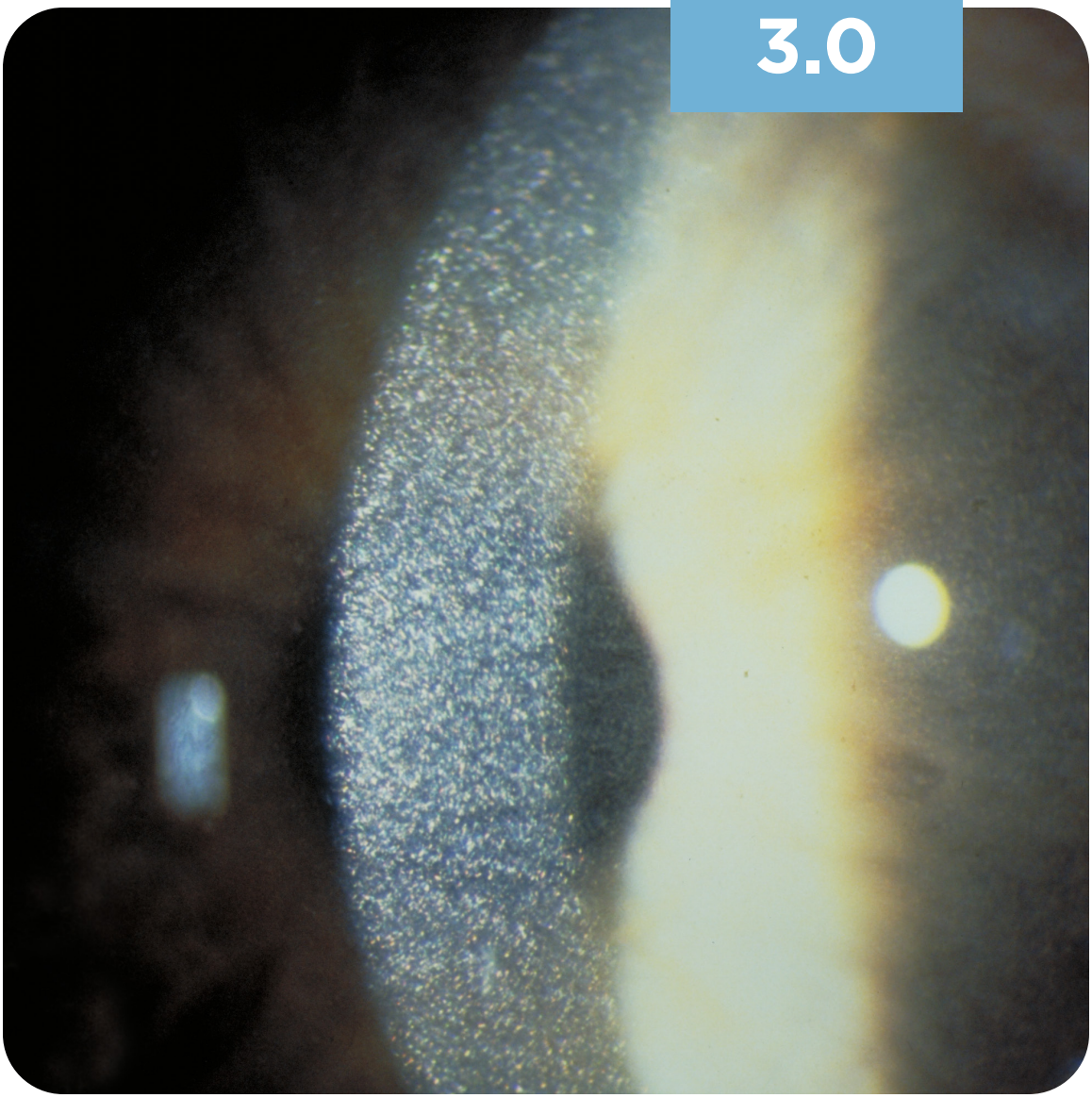

This eye has a CCCS of 3.0 (the highest possible score), showing dense cystine crystal accumulation throughout the cornea.*

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It's never too late to start using an eyedrop therapy, as patients of **all ages** may be able to reduce their crystal buildup with proper treatment.

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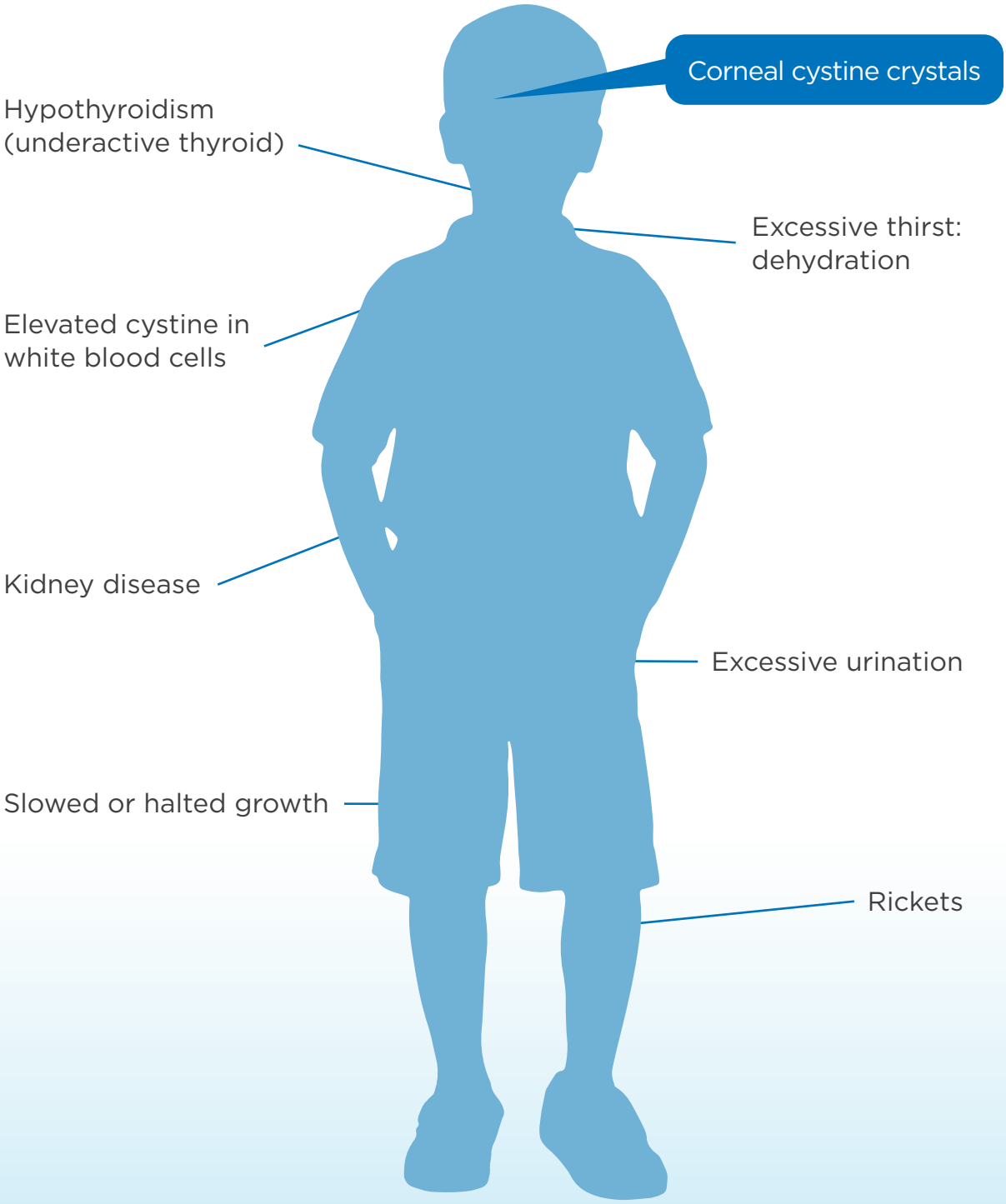
SYMPTOMS OF CYSTINOSIS

Cystinosis can affect many different parts of the body. The most common and severe type of cystinosis initially manifests as a type of kidney disease (Fanconi's Syndrome) as early as 6 months of age.

Early Signs and Symptoms of Cystinosis Include:



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SYMPTOMS OF CORNEAL CRYSTAL ACCUMULATION

Common symptoms of corneal crystal accumulation include:

- Sensitivity to light (photophobia)
- Constant squinting and muscle spasms in the eyelids (blepharospasm)
- Persistent (chronic) red eye
- Feeling like something is “in your eye” (foreign body sensation)
- Eye pain

If you are experiencing any of these symptoms, be sure to let your ophthalmologist know right away.

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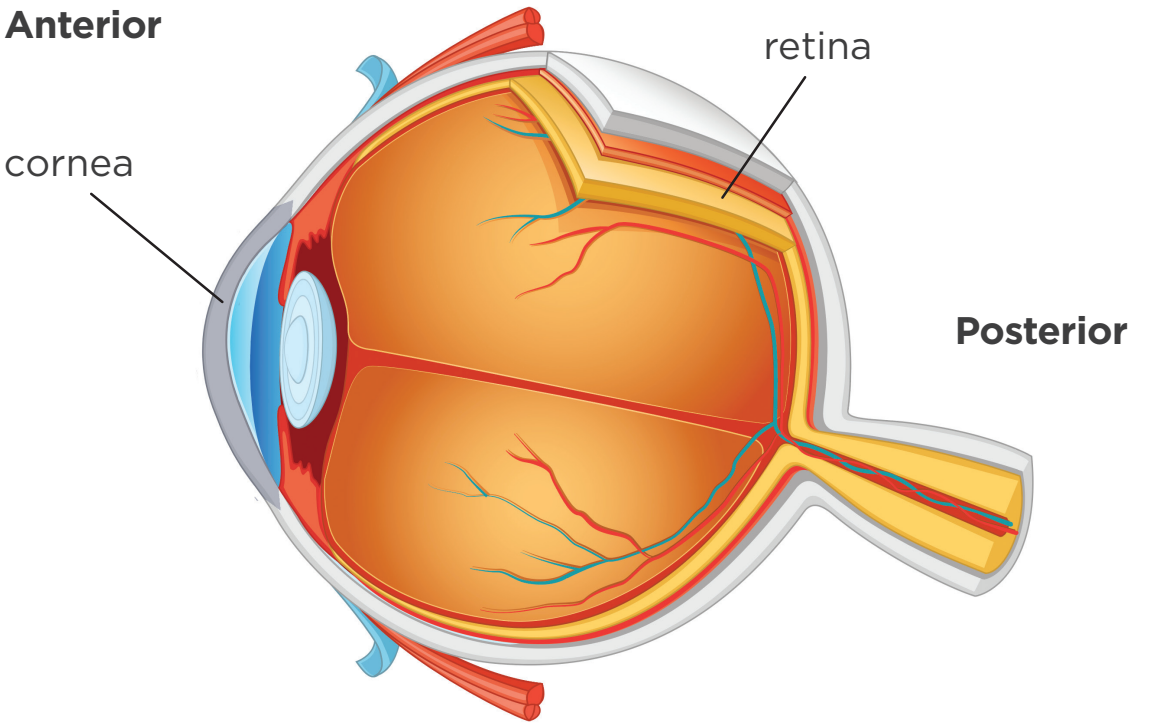
SUPPORTING EYE HEALTH WITH A CYSTINOSIS DIAGNOSIS



The cornea is located in the anterior (front) segment of the eye. The retina is located in the posterior (back) segment.

Anterior (Front 1/3 of the Eye)

- The cornea, which is the clear front surface of the eye, is responsible for refracting (bending) and focusing light that enters the eye to be processed by the retina.

Posterior (Back 1/3 of Eye)

- The retina, which is located at the back of the eye on the inside, contains special cells that process light and color and send those signals along to the brain.

While oral cystinosis medication may help treat crystals in some parts of the eye, it can't reach the cornea because there is no blood supply to the cornea to deliver the drug. **To fight corneal crystal accumulation, you also need a therapy that goes directly on the eye, such as an eyedrop.**

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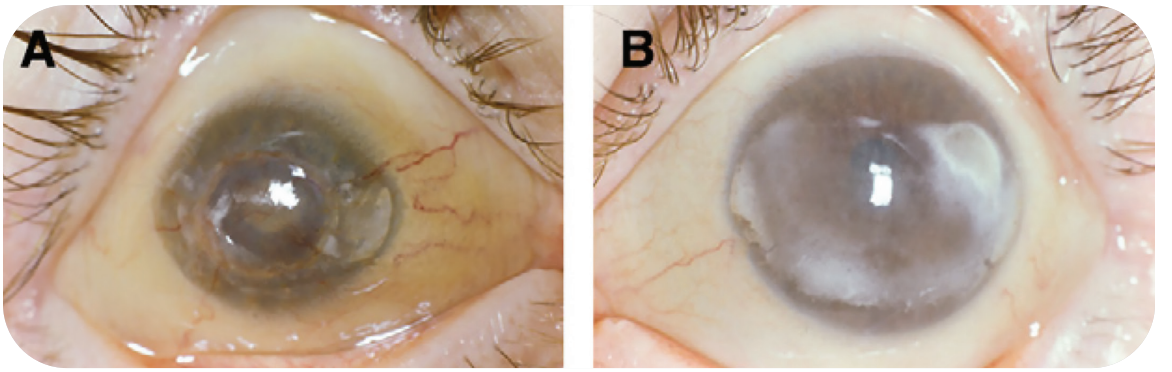
THE IMPORTANCE OF LIFELONG TREATMENT

Treatment for corneal crystal accumulation should be **lifelong**, since corneal crystals continue to accumulate into adulthood.



Slit lamp photograph of right eye *(A)* and left eye *(B)*. Calcification is present in both corneas. Severe corneal neovascularization is seen in the right cornea. Prolonged heavy accumulation of cystine crystals may lead to inflammation, which can then lead to factors that cause abnormal blood vessels to grow in the cornea.

Anterior Complications

- When corneal crystals are left to accumulate, they can lead to potentially vision-impairing corneal scarring.
- Buildup can also lead to chronic red eye, corneal scrapes or injury, inflammation, and even glaucoma.
- Corneal crystal accumulation is treatable with cysteamine eye drop medication.

Posterior Complications

- Cystinosis can cause declines in vision because of accumulation in the retina:

- If the retina is damaged enough, significant visual impairment is possible.
- Taking oral cysteamine medication as directed may help prevent posterior segment issues, including retinal problems.

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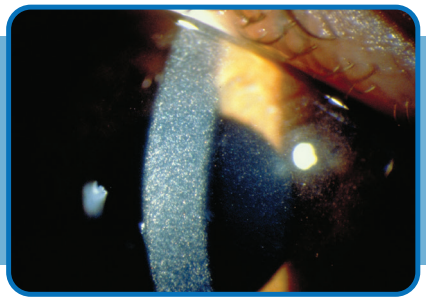
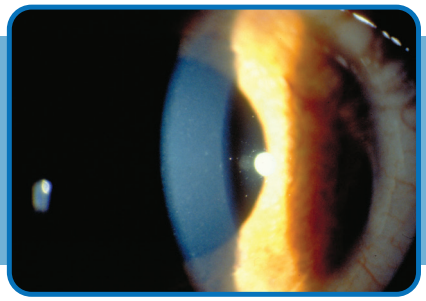
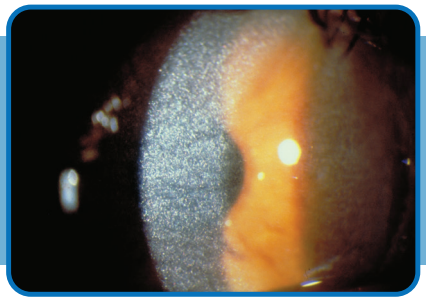
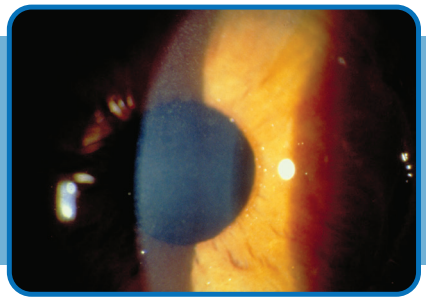
SEE EXAMPLES OF TREATMENT WITH CYSTARAN® (cysteamine ophthalmic solution) 0.44%

Age at Start of Treatment:	BEFORE	After 15 Months of Treatment
5-year-old patient		
25-year-old patient		

** Corneal slit-lamp photographs of patients treated with CYSTARAN. These examples represent patients who responded to treatment and in subsequent follow up appointments. Duration of therapy varied from 8 – 41 months. Response may vary from patient to patient*

Corneal crystals accumulate if CYSTARAN is discontinued.

CYSTARAN IS ONLY AVAILABLE FROM ALLIANCERX WALGREENS PHARMACY.

Your medication will be shipped directly to your home.

To order, call 1-877-534-962.

Friendly associates exclusively available to help CYSTARAN patients will be happy to assist you Mon–Fri, 8AM–7PM EST.

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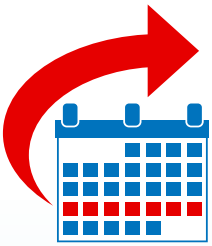
HOW TO TAKE CYSTARAN® (cysteamine ophthalmic solution) 0.44%

Always wash your hands thoroughly with soap and water before administering eye drops

Instill one drop of CYSTARAN in each eye, **every waking hour**

Do not touch dropper tip to any surface, as this may contaminate the solution



Discard after 1 week of use

There may be medication left in the bottle; however, the bottle must be discarded by the patient because the **medication is only stable for 1 week after thawing**

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Summary of Information about CYSTARAN® (cysteamine ophthalmic solution) 0.44%

What is CYSTARAN used for?

CYSTARAN (cysteamine ophthalmic solution) 0.44% is an eyedrop medication used to treat cystine crystal accumulation in the corneas of patients who have cystinosis.

What warnings should I know about CYSTARAN?

To help prevent contamination of the dropper tip and eyedrop medication, try to make sure that CYSTARAN is dropped directly onto the eye without touching it. Try not to touch the eyelids or surrounding areas with the dropper tip of the bottle when you are using CYSTARAN. Keep the bottle tightly closed when not in use.

CYSTARAN contains an ingredient called benzalkonium chloride which can be absorbed by soft contact lenses. Remove contact lenses before using CYSTARAN eyedrops and wait at least 15 minutes before reinserting them.

CYSTARAN should only be used as an eyedrop medication.

What are the side effects of CYSTARAN?

In at least 10% of patients, CYSTARAN causes sensitivity to light, eye redness, eye pain and irritation, and headache.

How do I take CYSTARAN?

Once every waking hour, instill one drop of CYSTARAN into each eye. Do not let the CYSTARAN dropper tip touch your eyelid or the surrounding area.

How do I store CYSTARAN?

1. Store CYSTARAN bottles in the freezer in the original carton.
2. Each week, remove one new bottle from the freezer.
3. Allow the bottle to thaw completely (about 24 hours) before using.
4. After the bottle is thawed, record the discard date on the bottle label. The discard date is 7 days (1 week) from the day the bottle is thawed.
5. Store the thawed bottle at no greater than room temperature for up to 1 week. No refrigeration required after thawing. **Do not refreeze the bottle.**
6. At the end of 1 week, throw away the entire bottle even if there is still medication inside. The medication is only stable for 1 week after thawing.

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