



Corneal Cross-Linking



**Save Sight
Registries**
Keratoconus



**KERATOCONUS
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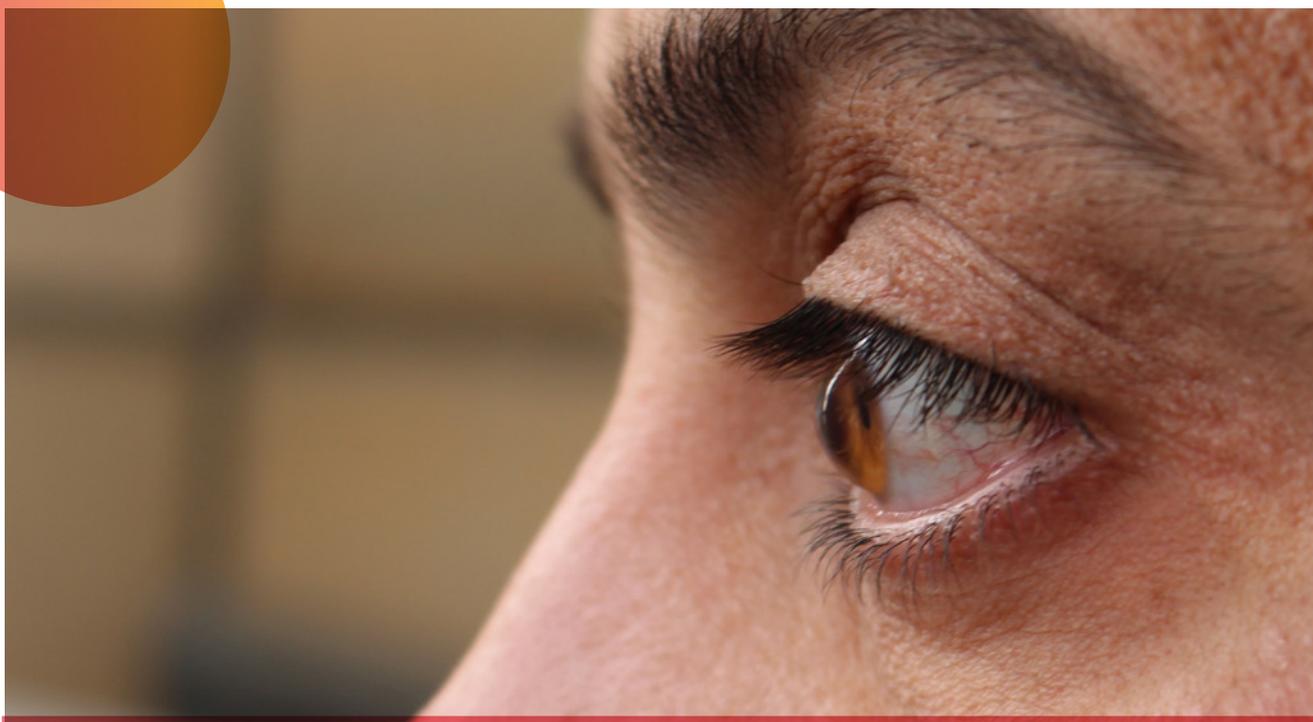
Corneal cross-linking (CXL) is a treatment to slow or halt vision loss caused by the eye disease keratoconus. It is successful in stopping progression in more than 9 out of 10 patients report an improvement in their quality of life. CXL does not cure keratoconus and at best there may be only a minor improvement in vision and corneal shape. After treatment, you will still need to wear glasses or contact lenses if you already do so. Your eye may be very painful for the first 48 hours and will be sore for about one week after the procedure. Although vision is often hazy at first, most patients can resume contact lens wear and return to work after one week. As with all operations, there are risks: CXL is safe, but there is a small chance of mild vision loss. As the treatment is not risk-free, CXL is for patients whose keratoconus is progressing or for patients who are at higher risk for progression such as younger patients.

What is CXL?

Keratoconus is an eye disease that causes the cornea, the clear window on the front of the eye, to thin out and get weaker over time. As keratoconus progresses, the cornea starts to bulge into a cone shape. The cone results in blurred and distorted vision and increased sensitivity to light. Severe vision loss can occur if progression is not treated early.

CXL is a minimally invasive procedure which uses ultraviolet light and vitamin B2 (riboflavin) eye drops to stiffen the cornea. Used together, they cause the fibres within the cornea to cross-link – or bond more tightly to prevent the cornea from bulging more.





Will it work?

CXL is the only treatment currently available that appears to prevent or stop keratoconus from getting worse. Recent evidence from the Save Sight Registry study one year after CXL showed success in stopping keratoconus progression in more than 9 out of 10 patients, with more than 4 out of 10 people also gaining a minor improvement in corneal shape and vision. Longer-term results (up to 10 years) from the Registry suggest a similarly high success rate in preventing keratoconus progression. Vision is slightly better after treatment than before in about 50% of the patients treated with CXL.

Will I need to have both eyes treated?

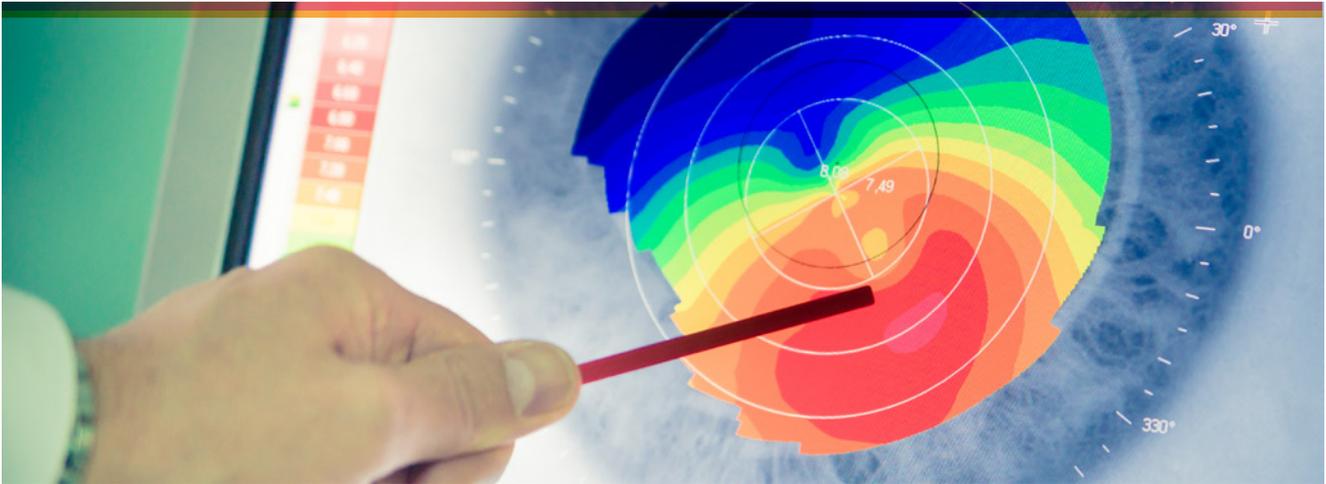
Keratoconus typically affects both eyes. You may therefore need to have CXL on both eyes if the keratoconus is progressing

in each eye., However, it is generally not recommended to treat both eyes with CXL at the same time due to the risks.

Can CXL be repeated?

In cases with continued progression after the primary CXL, a repeat CXL is possible after six months. A repeat CXL is also possible if the effect of the primary CXL diminishes over the years. There is lack of evidence on the effectiveness of the repeated CXL.





What are the risks?

In general, CXL is very safe. But like all operations your eye needs time to heal and problems do occasionally occur. About 3 out of every 100 patients having CXL will have worsening vision due to corneal haze, scarring, surface shape irregularities or infection. Haze generally resolves in the short to medium term. In the uncommon cases where the vision is affected, the vision may improve over time or there are options to improve things such as glasses, hard contact lenses or a corneal transplant. A transplant can be used to replace the cornea entirely; however, this is a much bigger operation with more risks and a long recovery.

In a real-world Save Sight Keratoconus Registry study, none of the patients had new side effects 10 years after the procedure supporting the mid-term safety of CXL.

The ultraviolet light used in the procedure is relatively mild and only enters the cornea, without affecting deeper structures of the eye. Riboflavin, also known as vitamin B2, is a naturally occurring compound found in our diet, such as in cornflakes. The CXL procedure does not cause bleeding of the eye because affected corneas do not have a blood supply.

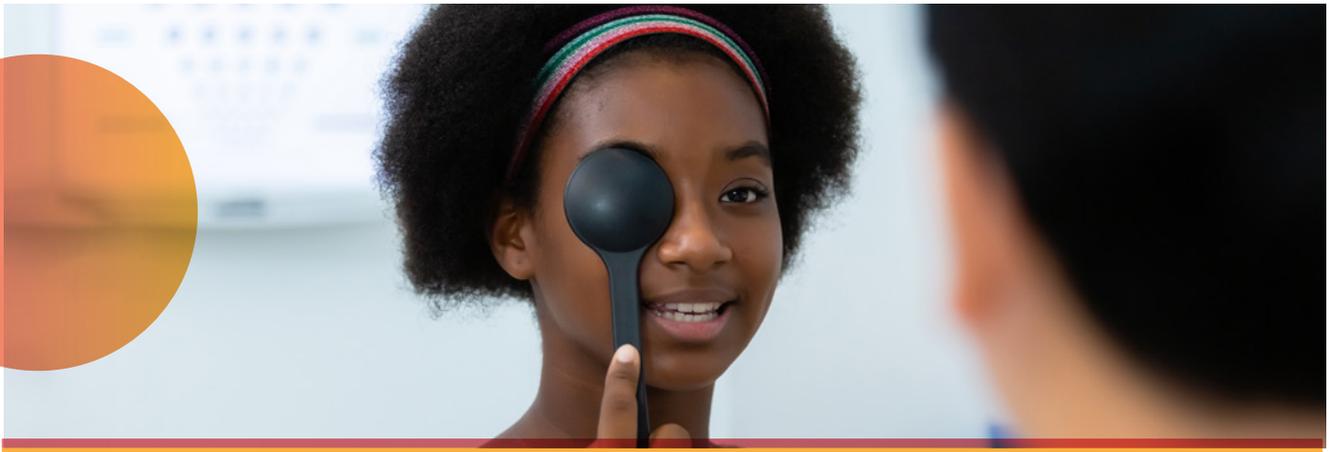
However, there may be very long-term risks to the eye from CXL that have not yet emerged. The Registry will continue to monitor for these risks.

Is it worth the risks?

Deciding to go ahead with an operation is an important decision for any patient or their carers.

Studies on 'epithelium-off' CXL (the traditional and best studied method of cross linking) thus far have shown that CXL is effective at stopping keratoconus progressing. The complications from CXL appear largely minimal and temporary. An important part of weighing up whether CXL is right for you is considering if your keratoconus is progressing and how fast, and what other high-risk factors you have for progression. It is important to discuss this with your doctor to come to a decision.





How will CXL affect my eyes long term?

CXL aims to stabilise keratoconus so in most cases your vision in the long term will remain the same. Corneal shape improves in more than a third of patients after CXL which may lead to a slight improvement in their vision. This small group which gains an improvement in their corneal shape may also require a refit of their hard contact lenses which may then fit more comfortably.

In some patients CXL will cause haze in their cornea, reducing their vision. This is usually at its worst after one month and remains the same until the third month. It then begins to improve over the next year. In some studies, the number of people who get corneal haze after CXL has been found to be as high as 9 out of every 100 patients a year after the procedure. After 5 years, 2 in 100 people may have some corneal haze but it usually doesn't affect vision.

What if I delay or do not have CXL?

If your keratoconus is progressing or you are at high risk of progression without CXL, then your keratoconus will likely progress as it would naturally. If your keratoconus is relatively stable it may be appropriate to monitor you instead of proceeding to CXL and this way you avoid

unnecessary risks from the procedure for little gain. Even if you require CXL in one eye that is progressing, frequently it may be best to just monitor the lesser affected eye for progression.

Data shows that keratoconus is less likely to progress after the age of 26 and often stabilises after the age of 30 years old – especially if you are not rubbing your eyes. If you are in this age bracket, again it may be more appropriate to monitor you instead of performing CXL. If you do not require CXL or choose not to go ahead with it, it is important that you are still monitored regularly by your treating clinician because if your keratoconus advances, this decision may change.

CXL cannot be performed on a very thin cornea so regular reviews will ensure you are able to make up-to-date decisions on whether to have CXL before your cornea thins so much that it is no longer safe to have the procedure.

If your keratoconus is very advanced it may be too late for you to have CXL, but there may be other options such as specialised contact lenses or a corneal transplant. Some patients may not be able to have CXL due to medical conditions, pregnancy, or another eye condition such as herpes simplex keratitis.



Procedure

What are the different types of CXL?

There are many different ways or “protocols”, for doing CXL.

One main variation is how the riboflavin is soaked onto the cornea. The surface skin of the cornea (epithelium) can be removed (epithelium-off) or left on (epithelium-on). The time that ultraviolet light is shone on the eye can also be varied – as accelerated or standard. Accelerated CXL involves the use of a stronger ultraviolet light in a shorter period to achieve the same effect as the Standard CXL. More experimental variations are being used in which other procedures such as laser or corneal ring operations are done at the same time as CXL.

Epithelium-off cross-linking is considered the traditional method. There is sufficient evidence to show that epithelium-off works well to stop keratoconus progression.

Epithelium-on and combined procedures do not have enough studies to back them up yet and more research is required to prove their safety and effectiveness.

Other variations of CXL procedure include adding oxygen to the cornea during treatment, and pulsing the ultraviolet light. Generally, the ultraviolet light is continually

shone to the cornea during CXL. Pulsing the ultraviolet light theoretically may lead to a better photochemical reaction. However, the evidence so far has shown that both pulsed and continuous light CXL are similarly effective. More research is required to prove the benefits of these variations over the standard CXL procedure.



What happens in the 'epithelial-off' treatment

How to prepare?

On the day of the treatment, avoid applying eye makeup, perfume, or after-shave. You may be advised to fast before the procedure. It is important to arrange for someone to accompany you and drive you back home since your vision will be impaired. Make sure to adhere to your doctor's guidelines and instructions diligently.

What happens on the day of surgery?

CXL is performed as a day-case procedure in the operating theatre or consulting rooms, usually under local anaesthetic. Although the procedure takes less than 60 minutes, there is usually some waiting time before treatment and you will also need to stay for a short while afterwards

for checks to make sure that you have everything you need to go home.

During the procedure, you will be asked to lie flat on the treatment table. Anaesthetic drops will be used to numb the surface of your eye before a small clip is placed to keep your eyelids open. The anaesthetic drops may cause a momentary stinging sensation but keep the eye comfortable during the procedure. Following that, the skin surrounding the eye will be cleansed using an iodine solution. Patients are generally awake but may be given sedation before or during the procedure. They do not experience pain or discomfort during the CXL procedure. The surface skin of your eye (epithelium) is removed and riboflavin drops are applied every few minutes for at least 10 minutes. Following this, the ultraviolet light is shone at your eye. A soft contact lens may be placed on your eye at the end of the procedure and acts as a bandage.





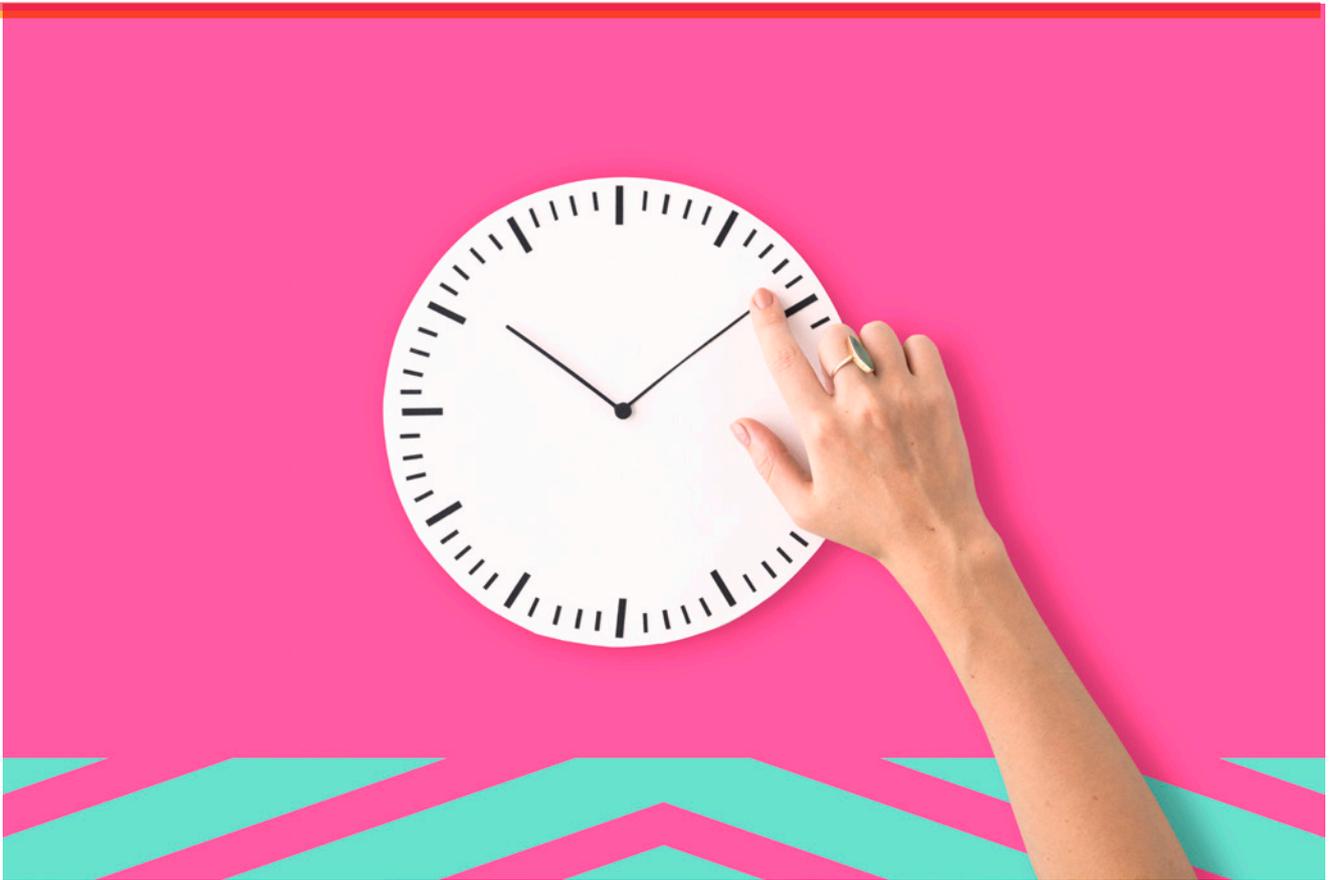
Recovery

What is the pain like?

The anaesthetic drops will wear off later on the day of your procedure, and your eye will be gritty, red and sensitive to light for up to several days. Everyone's experience with CXL is different, with some patients reporting very little discomfort and others describing the first few days as very painful. If your eyes are sensitive to light sunglasses maybe helpful.

Upon discharge, you will be provided with pain relief tablets and a suggested regimen. Additionally, medication to assist sleeping at night may be supplied a few nights





What is it like straight after CXL?

Once the procedure is complete, a plastic eye shield might be placed over the eye. A nurse will provide detailed instructions regarding the aftercare regimen, follow-up appointments, and discharge information.

You will be given eye drops to use after the procedure. The soft 'bandage' contact lens, if placed, will remain in your eye until the surface has healed (usually in under seven days). The surgeon or a team member will remove the contact lens.

If the bandage lens falls out during this time, please throw it away – do not attempt to reinsert it.

Your vision will be quite blurred at first, but will clear gradually over the first few weeks.

It is important to note that following the procedure, you may experience redness

and a gritty or burning sensation in the eye for up to a week, in addition to any discomfort or pain. It is normal for your eyelids to become puffy and your eye to water after the procedure. It is possible for your other eye to also experience watering. These symptoms are expected and will gradually improve within 48 to 72 hours.

When should I immediately contact my doctor?

It is normal to experience fluctuating pain within the first two days after surgery. If you experience increasing pain three or four days after the procedure, increasing redness or yellow-green discharge this could indicate infection and you should seek emergency assessment straight away. Please note that infection is rare, affecting fewer than 1 in every 150 patients.



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What should I do, or not do, after CXL?

It is important to put the eye drops in regularly as prescribed. The eye drops prevent infection and aid in healing and can provide relief for any discomfort you may experience. Wash and shower normally, but try to avoid getting water in your eyes. You may exercise, but should not swim before the surface of your eye has healed. Do not wear any eye make-up for about 2 weeks.

Minimizing ultraviolet exposure by wearing a hat and sunglasses for 6 months after CXL may reduce hazing in the cornea.

It is crucial to refrain from rubbing your eyes following the surgery – forever.

When can I drive again?

We will check your vision in the clinic within about a week after your procedure to confirm if your vision is good enough

to drive. It is normally safe to resume contact lens wear once the eye surface skin layer has healed. This typically happens around the end of the second week after your procedure and your ophthalmologist can advise you on this.

Do I need to take time off?

Yes. You should allow at least one week off from your normal activities while most of the surface healing occurs, or up to two weeks if your job involves a lot of computer work, and the treatment is being done on your best eye. You will be putting eye drops in typically every four hours for the following days.

Day-to-day activities such as watching TV or using a computer will not do any damage to your eye, but you might find it more comfortable to rest with your eyes closed early on.



Cost of CXL

The cost of CXL varies. In Australia, the Medicare rebate is available for the procedure for patients with progressive keratoconus.



CXL in Children

CXL is used less frequently in children and has been studied less than in adults. That being said some studies have performed CXL in children as young as 8 years old and have not found safety of the procedure to be any different from adults. However, how long CXL will last in children and side effects of the procedure are more uncertain than in adults. Nonetheless, several studies have shown CXL to stop progression in a similar way to adults and this can reduce the need for corneal grafting in the long-term.

Please keep in mind that the information provided in this leaflet serves as a general guide. It is important to recognize that each patient is unique, and individual experiences and circumstances may vary. It is always recommended to consult with your healthcare provider for personalized advice and specific information pertaining to your situation.



Save Sight Keratoconus Registry

What is The Save Sight Keratoconus Registry (SSKR)?

The Save Sight Keratoconus Registry is part of the Fight Corneal Blindness! (FCB!) Project. It is a tool that allows you and your doctor to monitor your keratoconus and treatment journey including before and after CXL treatment as well as other treatments. Its innovative system collects data on your vision and eye shape at each clinic visit and stores it securely on a server at the University of Sydney.

A team comprised of clinicians and patients designed the Save Sight Registry and continually update and improve it with input from clinicians, patients and other stakeholders. The system is in use across Australia and internationally.

How will the SSKR help you and your doctor?

The SSKR system can anonymously capture a lifetime record of your eye care. If CXL is needed, it then tracks your response to the treatment to determine if your eye remains stable as the effect of CXL may wear off over the years. It is also valuable for your safety as it can log any side effects that you experience. SSKR is useful for your doctor; it assists them in tracking the progress and results of CXL for their patients, helping them improve the care they deliver. Only your doctor and their health care team can view your records to protect your privacy. The system also helps clinicians understand how keratoconus progresses and how it affects your day-to-day life. With the SSKR clinicians can also benchmark the results of their patients CXL with national and international standards. Understanding keratoconus and evaluating new treatments will help further research into finding a cure for keratoconus. The SSKR has led to over 15 publications in leading national and international journals.

What can you do?

Ask your doctor if you can be a part of the Registry to make sure you are receiving the best care.

For further information,
visit the link or scan the QR code

<https://savesightregistries.org/fight-corneal-blindness>

