

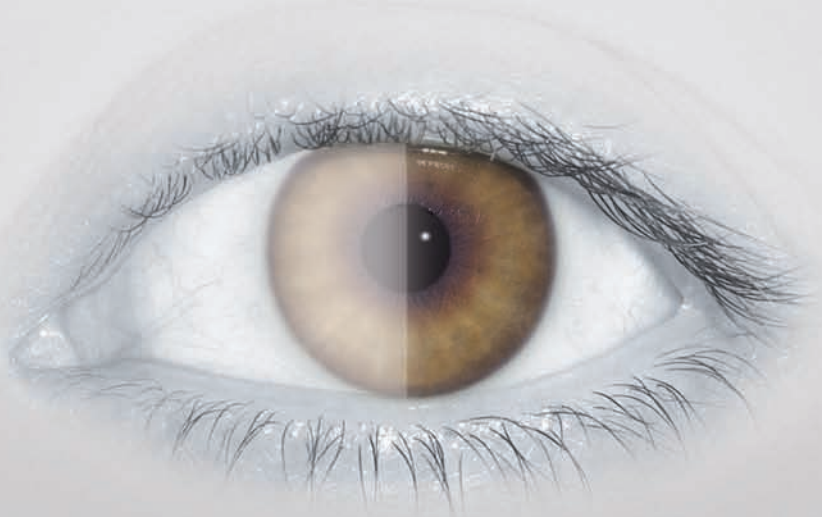


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Understanding Keratoconus

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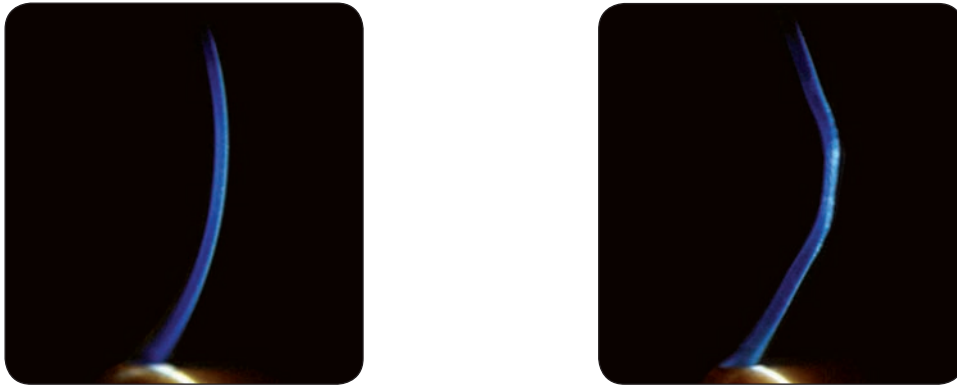
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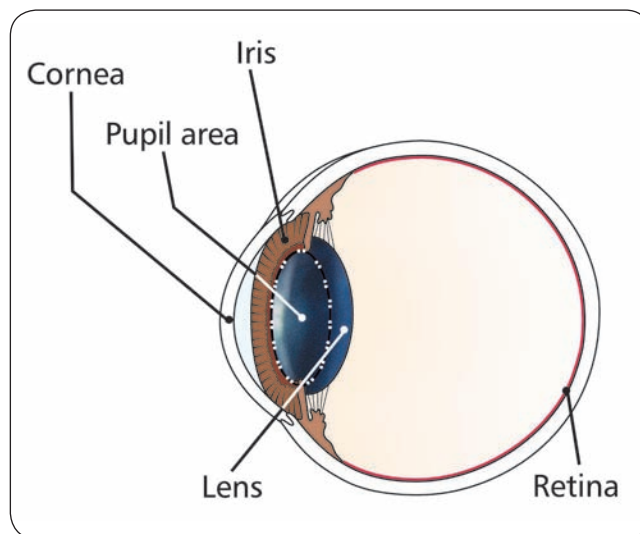
What is keratoconus?

Keratoconus is a disorder of the cornea - the transparent 'front window' of the eye

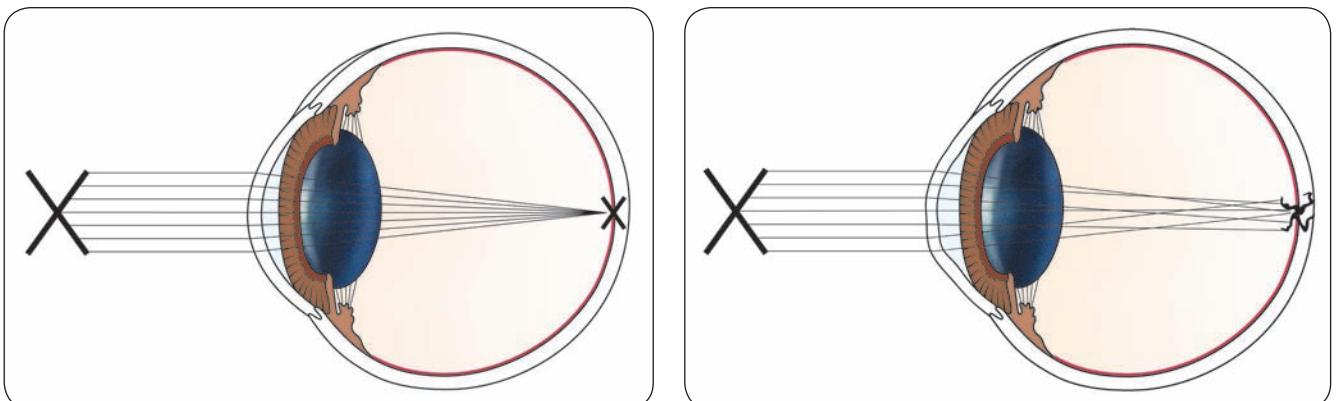
The cornea focuses light so any abnormality of its shape will reduce vision. Normally it has a smoothly curved surface like a tennis ball. In keratoconus the cornea is distorted by a protrusion shaped like a microscopic volcano: as it is only fractions of a mm high it cannot be felt or seen, except in severe cases.



Normal cornea profile (left) is smoothly curved. Keratoconic cornea (right) is distorted by a volcano-like protrusion



Structure of normal eye



Normal cornea (left) focuses accurately onto retina. Keratoconic cornea (right) blurs image ✗

Symptoms

The main symptom is blurred vision

Keratoconus usually begins in the late teens to early 30s and affects men and women. It is significant for 1:2000 people but mild cases occur up to ten times more often. One or both eyes may be affected. It may begin in one eye and later involve the other.

The main symptom is blurred vision even in glasses. The glasses prescription increases with time and usually shows high levels of myopia (short sight) and astigmatism. Some patients have glare driving at night or from sunshine.

If the distortion of the cornea worsens, so will vision. This happens in some patients but in many the condition stabilises. It is not possible to predict whether keratoconus will be progressive or at what rate it will worsen but in general the younger the age and the more severe the disorder when it begins, the more rapidly it is likely to deteriorate.

In advanced cases 'hydrops' can occur: this is the sudden onset of discomfort, redness and reduced vision caused by a split in the inside of the cornea. It usually clears after a few weeks but may leave permanent scarring. A 'proud nebula' is a small white scar at the tip of the keratoconic protrusion: it may worsen vision and glare and decrease contact lens comfort.

Cause

In most cases the cause is unknown

Keratoconus is not usually inherited, although there are exceptions, and is not lifestyle-related, although persistent heavy eye rubbing may be a risk factor. It is slightly more frequent in severe asthma or eczema and patients with chromosomal disorders such as Down's syndrome quite often have keratoconus.

Diagnosis

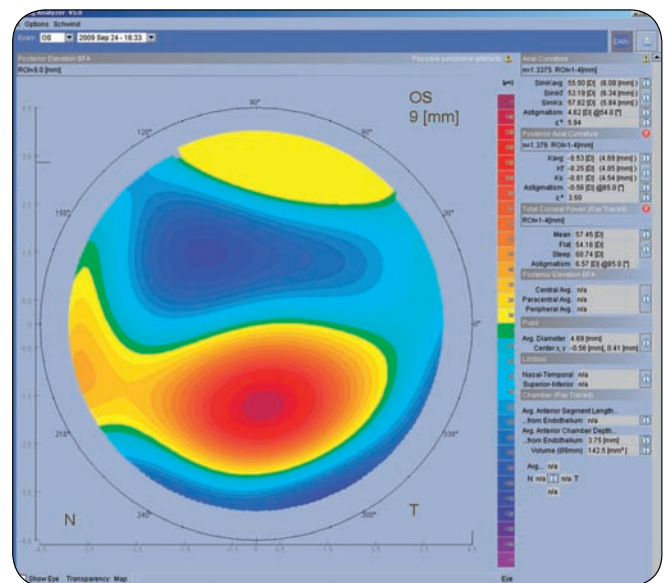
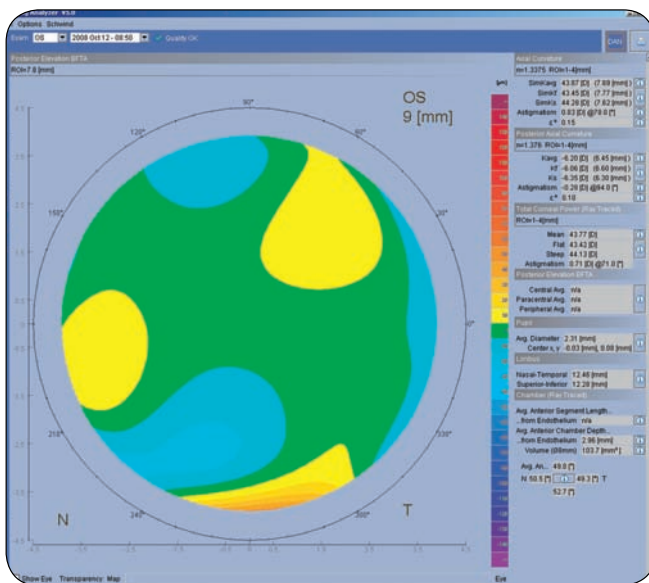
Diagnostic equipment includes a cornea topographer

The protrusion on the cornea may be seen with a slit lamp microscope. Retinoscopes and keratometers are used to measure eyes for glasses and contact lenses: in keratoconus they show abnormal readings.

The cornea topographer is the most important diagnostic device: it makes a map of the cornea surface and accurately measures the keratoconic protrusion. I use the most advanced topographer currently available - the Ziemer Galilei.



The Galilei topographer



Topography of normal cornea (left) shows mostly green and yellow colour in the centre. Contrast with a keratoconic cornea (right) where the volcano-like protrusion shows as a red colour

Treatments

New technologies have improved the treatment of keratoconus

Until recently, patients have worn contact lenses unless they needed a cornea transplant - there was no choice and no way of stopping keratoconus from getting worse.

Fortunately several important new treatments are now available. Here is my approach -

| KERATOCONUS TREATMENT | |
|--|---|
| <p>Mild-to-moderate keratoconus Vision acceptable in gas permeable contact lens Keratoconus not getting worse</p> <p>➔</p> <p>Gas permeable contact lens wear</p> <p>➔</p> <p>Acceptable vision</p> | |
| <p>Keratoconus getting worse</p> <p>➔</p> <p>Collagen cross linking</p> <p>➔</p> <p>Keratoconus stable</p> | |
| <p>Mild-to-moderate keratoconus Vision not acceptable in gas permeable contact lens</p> <p>➔</p> <p>Laser and collagen cross linking</p> <p>➔</p> <p>Improves vision</p> | <p>➔</p> <p>Further improvement required</p> <p>➔</p> <p>Phakic intraocular lens</p> <p>➔</p> <p>Improves vision</p> |
| <p>Moderate-plus keratoconus Vision not acceptable in gas permeable contact lens</p> <p>➔</p> <p>Intacs implants</p> <p>➔</p> <p>Improves vision</p> | <p>➔</p> <p>Further improvement required</p> <p>➔</p> <p>Laser and collagen cross linking</p> <p>➔</p> <p>Improves vision</p> |
| <p>Proud nebula present Glare and discomfort in gas permeable contact lenses</p> <p>➔</p> <p>Remove</p> <p>➔</p> <p>Improves symptoms</p> | |
| <p>Advanced keratoconus Other treatments not effective Inner layer of cornea normal</p> <p>➔</p> <p>Partial cornea transplant</p> <p>➔</p> <p>Improves vision Reduced risk of transplant rejection</p> | |
| <p>Advanced keratoconus Other treatments not effective All cornea abnormal</p> <p>➔</p> <p>Full thickness cornea transplant</p> <p>➔</p> <p>Improves vision</p> | |

Contact lenses

The standard treatment

Contact lenses do not change the keratoconus, but cover the protrusion on the cornea so neutralising it. Gas permeable (rigid) contact lenses are usually needed but sometimes special types of soft lenses work.

Normal vision is restored to all except advanced cases. Lifelong wear is necessary. Unfortunately it may be difficult to fit a contact lens over the protrusion and wearing it may be uncomfortable.

Collagen cross linking

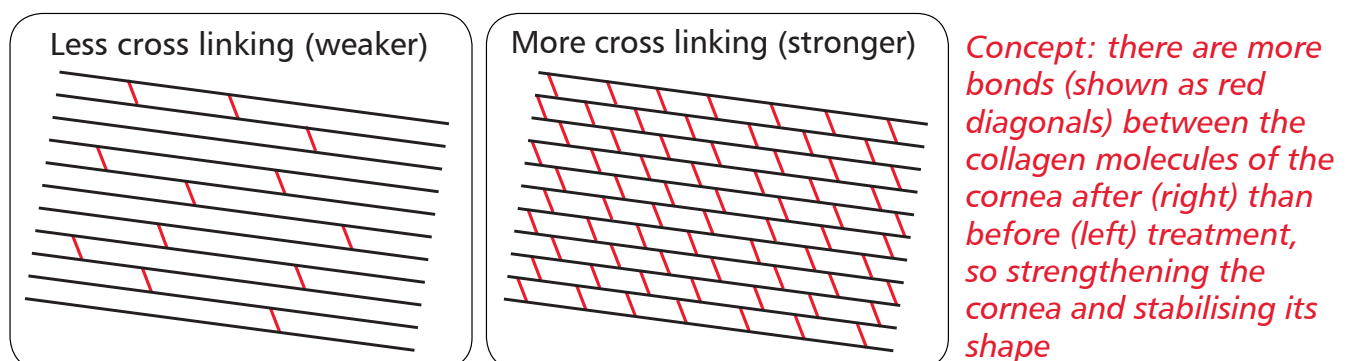
Stops keratoconus from getting worse

What is it? – Collagen cross linking is an important new treatment that *stops keratoconus from getting worse*. As it does not reduce keratoconus that is already present, it follows that it is best done at an early stage. Most patients need only one treatment per eye, but it may be repeated if necessary. It is done under local anaesthetic (eye drops) in outpatients and does not require hospital admission.

How may benefit be maximised? – After collagen cross linking has been done to stabilise keratoconus, vision may be improved by laser, Intacs implants and phakic intraocular lenses.

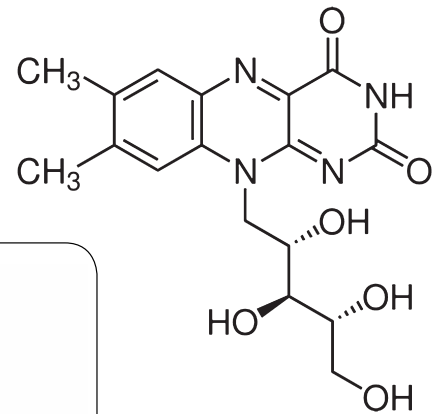
How long has it been available? – The technique was developed in 1998 in Germany.

How does it work? – The cornea is composed of collagen. In keratoconus the collagen is weak, for unknown reasons. Cross linking increases the strength of the collagen in keratoconic corneas by 3–4 times. Collagen fibres, previously separate, are bonded together ('cross linked') by a chemical reaction. This is done by applying vitamin B₂ (Riboflavin) as an eye drop and then exposing the cornea to ultra-violet light from a medical device intended for this function. I use the Vega Cross Linker.



What are the risks? – Collagen cross linking has an excellent safety record. Two relevant regions of the eye are 1) the inner lining ('endothelium') of the cornea. To protect this layer the cornea must be at least 400 microns thick. Although many keratoconic corneas are thinner than this, there is a simple technique to temporarily swell the cornea just before treatment. 2) the retina at the back of the eye. This is not affected by the ultraviolet light because the Vitamin B₂ in the cornea absorbs it, preventing onward transmission to the retina and because cross linking devices such as the Vega Cross Linker are designed to focus the light on the cornea only.

*Riboflavin (vitamin B₂):
chemical composition*



The Vega cross linker

Laser

A high-tech new treatment to improve vision

Laser may now be used to treat keratoconus: this is an important development. The technique is similar to the Lasek laser done for short sight etc, but uses a cornea topographer to guide the laser. In medical jargon this is 'topography-driven treatment'. The laser reduces the keratoconic protrusion, significantly improving vision. When controlled by a topographer only a few microns of cornea need be removed, greatly enhancing safety. The technique is effective for mild-to-moderate keratoconus. To stabilise the cornea after laser, collagen cross linking is done. Only the most sophisticated lasers, such as the Wavelight Concerto I use, are capable of this treatment.



The Wavelight Concerto laser

Intacs implants

An established treatment for more advanced keratoconus

Intacs are two small pieces of plastic inserted into a channel created in the cornea. They can't be felt and require no maintenance. They are normally invisible except on close inspection when they look like a contact lens on the eye.

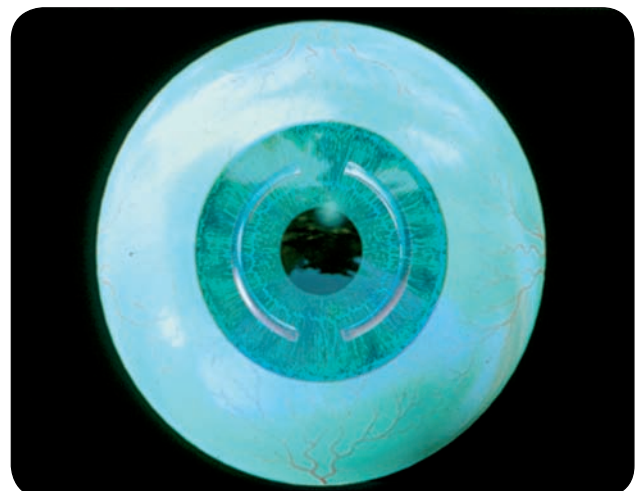
The implants work by evening out the keratoconic protrusion, so improving vision. The technique works best for moderate-plus keratoconus. Improvement varies between patients and may be enhanced by laser and phakic intraocular lenses.

In 2000 I was one of the first surgeons in the UK to use Intacs, initially for patients who were short sighted but whose eyes were otherwise normal. I have treated keratoconus with them since 2002 and was an expert advisor to the National Institute for Clinical Excellence (NICE), which has endorsed their use. They are also approved by the Food and Drug Authority (FDA) in the USA.

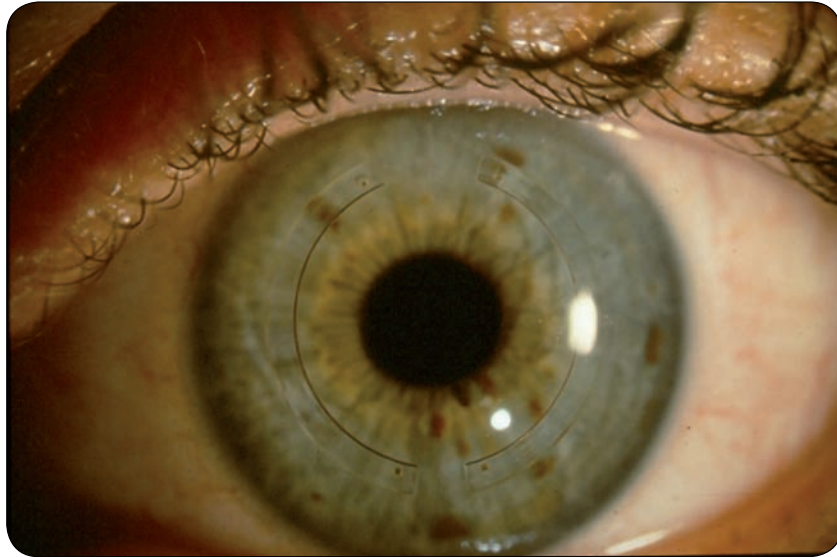
An advantage of Intacs are that they are reversible as the implants can be removed (by the doctor) and the cornea returns to its previous state. No cornea is removed during insertion ie: the technique is 'additive' rather than 'subtractive'. Reversibility may be helpful when new technologies for treating keratoconus become available.



Two Intacs implants rest on a fingertip



Intacs after insertion (simulation)

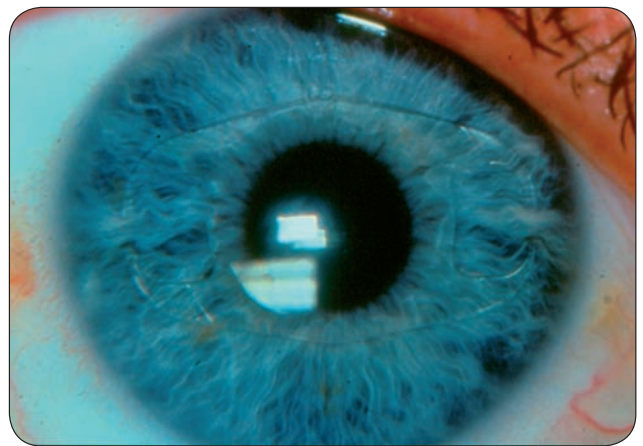
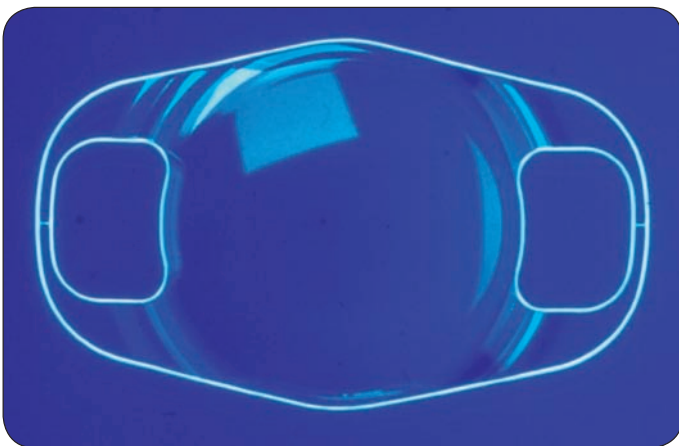


I gave this patient Intacs

Phakic intraocular lens

An effective treatment after cornea shape has been normalised

This is a miniature focusing lens, inserted via a keyhole incision. It is held permanently in place on the iris (coloured part) of the eye by special attachments. It works together with the eye's natural lens which is not removed. In a normal cornea it is capable of correcting even the most severe short sight and astigmatism. In keratoconus it is most effective after collagen cross linking, laser or Intacs have been done to stabilise the cornea and normalise its shape as much as possible.



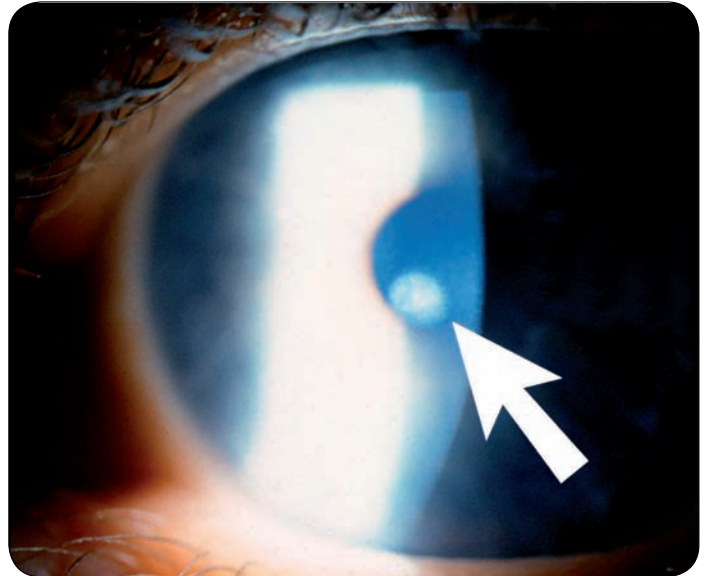
A phakic intraocular lens (left) is implanted eye and attached to the iris (right). It is invisible under normal circumstances

Proud nebulae

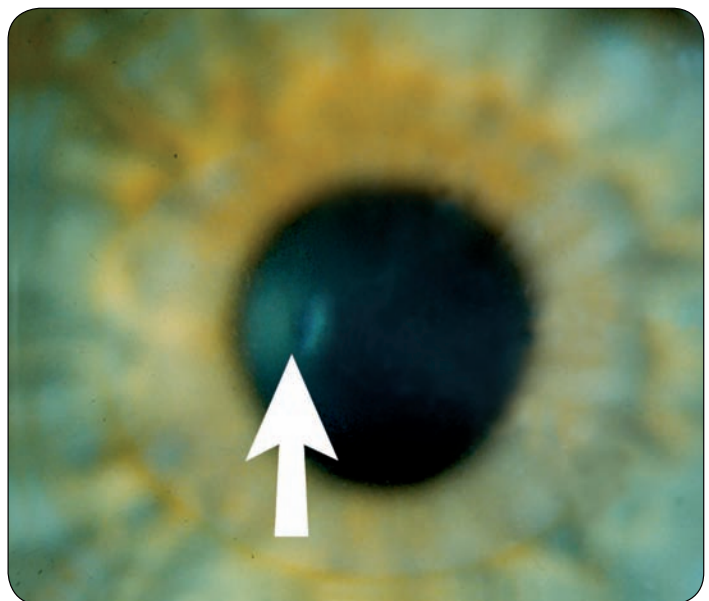
Removal improves vision and comfort

These can usually be removed by a simple procedure, improving vision and contact lens comfort and reducing glare.

A proud nebula is seen as a white scar on this keratoconic cornea.



Keratoconic cornea after removal of proud nebula. Only a faint impression of the scar remains (arrow)



Partial cornea transplantation

Sophisticated transplantation with a low risk of rejection, for advanced keratoconus

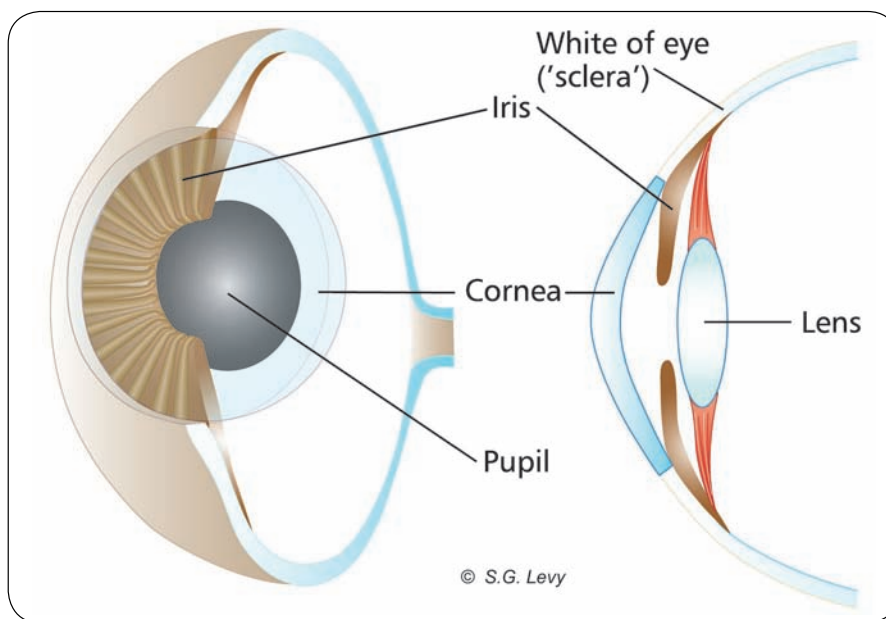
This new method saves some of the patient's own cornea and only replaces the outer layer. This is ideal in keratoconus because usually only the outer layer of the cornea (the 'stroma') is abnormal whilst the inner layer (the 'endothelium') is healthy. Preserving the patient's healthy endothelium is important as it greatly reduces the risk of transplant rejection.

The technical name for this procedure is 'deep anterior lamellar keratoplasty'.

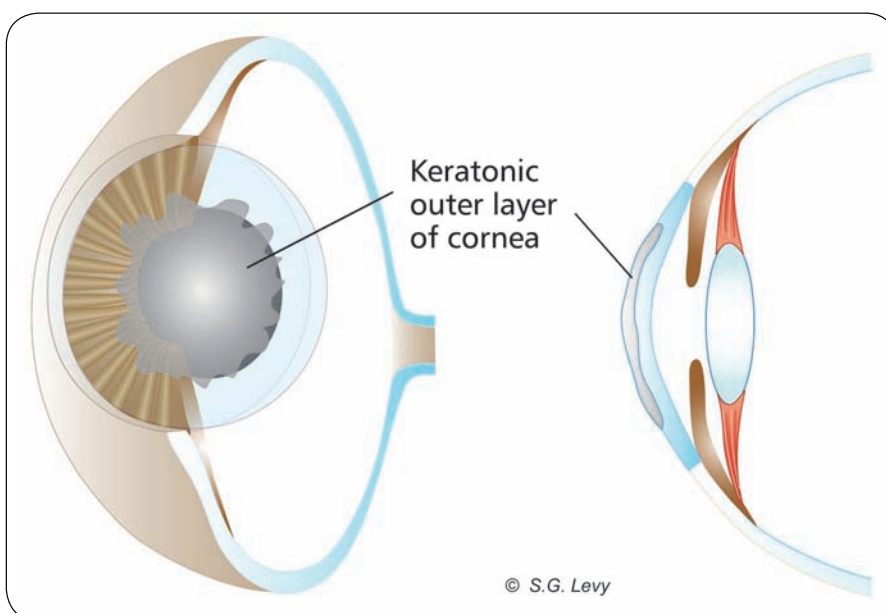
I always perform partial transplantation when possible. However if the keratoconus is too advanced, for example the cornea is severely scarred, full thickness transplantation is done instead.

Gas permeable contact lens wear may be necessary after the surgery.

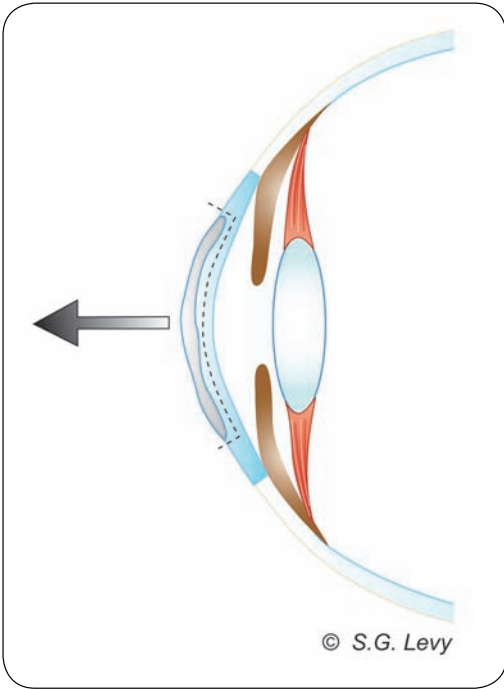
These illustrations of partial cornea transplantation (deep anterior lamellar keratoplasty) are a general guide, not an exact description.



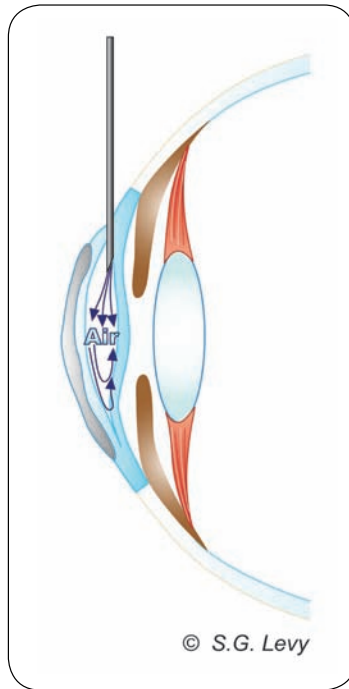
In health, the cornea is perfectly clear



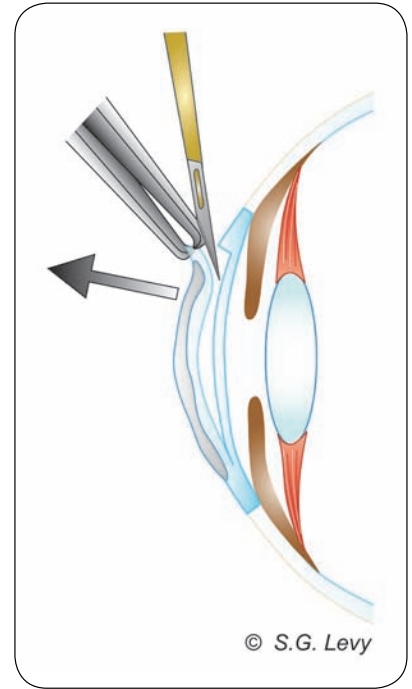
The technique is suitable when keratoconus involves only the outer layer of the cornea



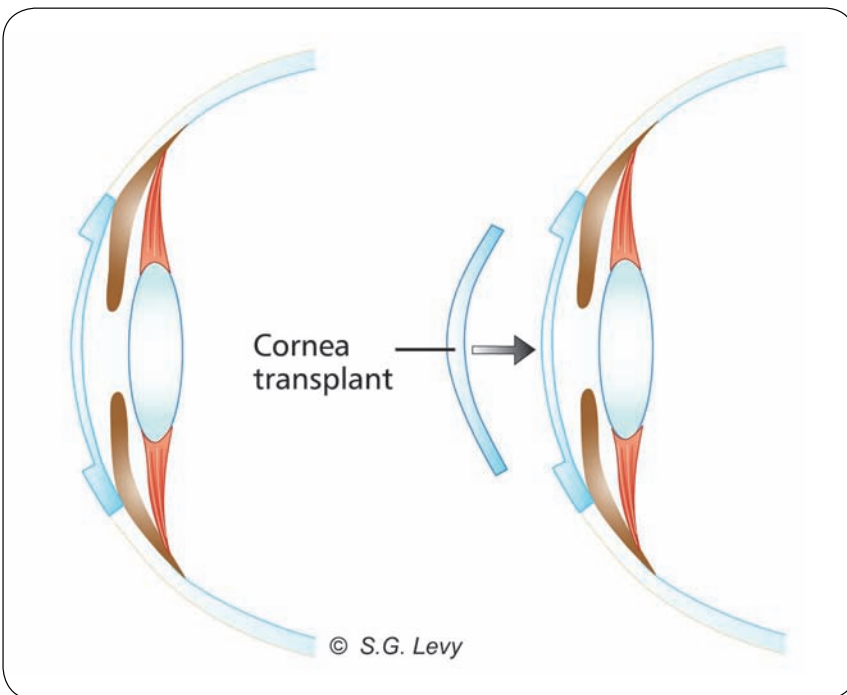
The keratoconic outer layer of cornea will be removed and replaced



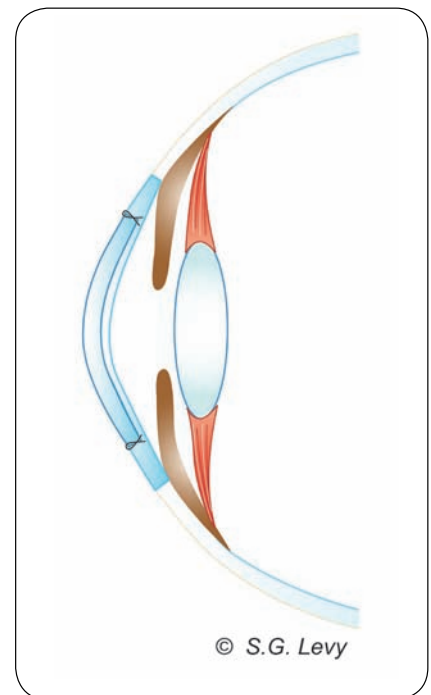
Air is injected into the cornea to separate the two layers



The keratoconic layer is completely removed



A healthy cornea is transplanted to replace the abnormal layer



The cornea transplant is secured with ultra-fine sutures which are removed after healing

Full thickness cornea transplantation

A long-established treatment for the most severe cases

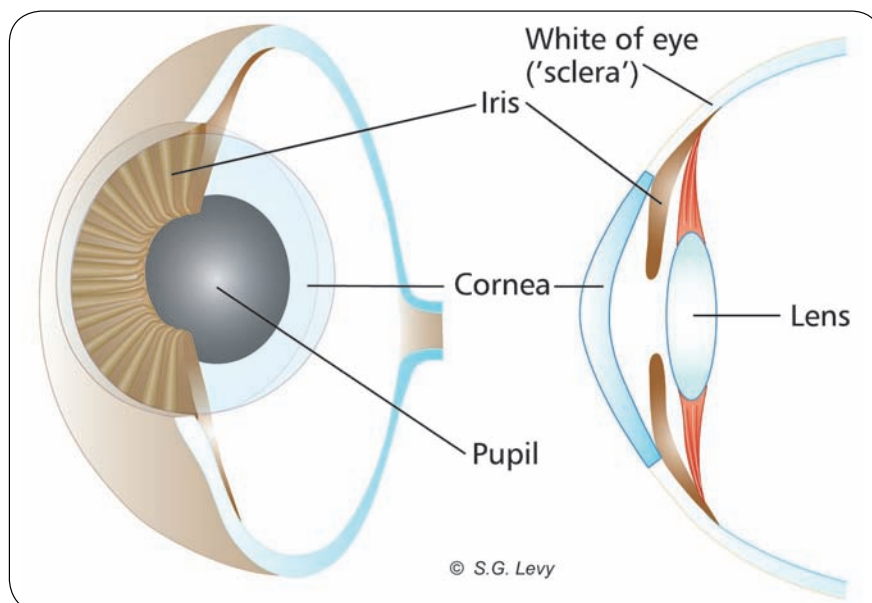
The centre of the patient's own cornea is completely removed and replaced by a transplant. This is the longest-established treatment for severe keratoconus. It is appropriate when all the cornea is abnormal, not only the outer layer.

The technical name for this procedure is 'penetrating keratoplasty'.

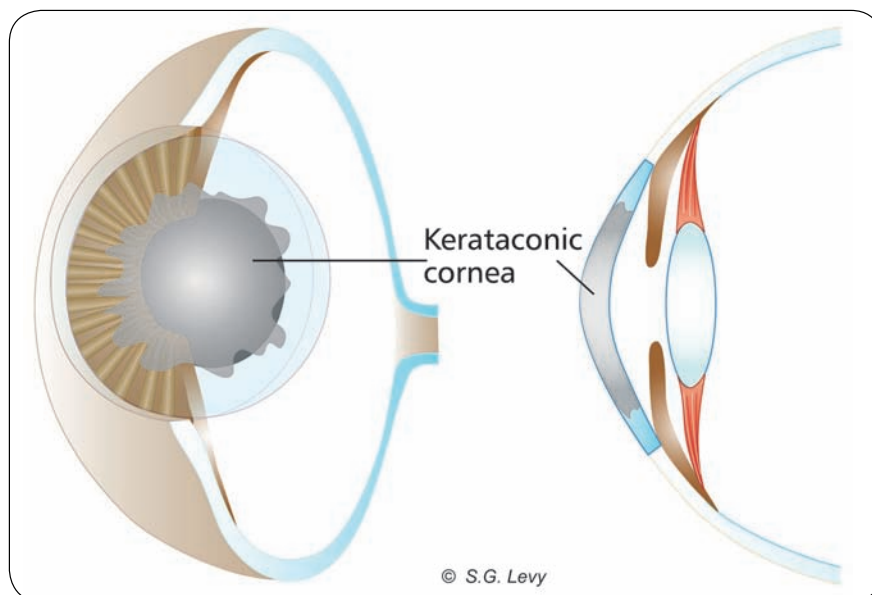
Risks include rejection of the foreign tissue by the patient's immune system (greater than after partial transplantation but still infrequent), wearing out of the transplant after many years and astigmatism (an uneven corneal shape) post-operatively requiring gas permeable contact lens wear.

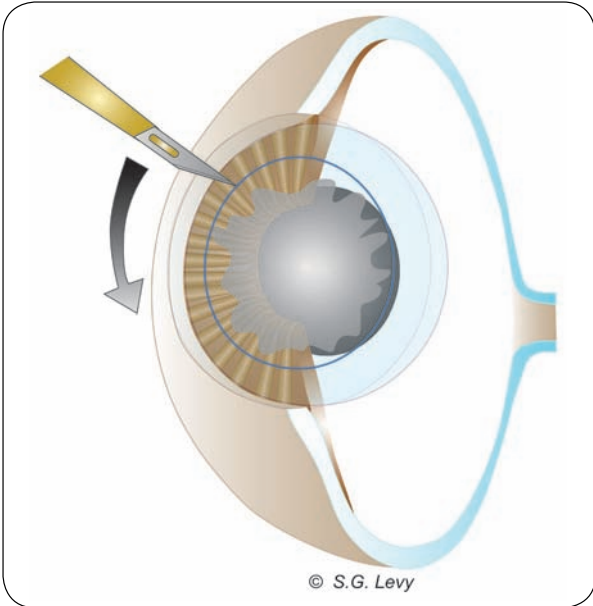
These illustrations of full thickness cornea transplantation ('penetrating keratoplasty') are a general guide, not an exact description.

In health, the cornea is perfectly clear

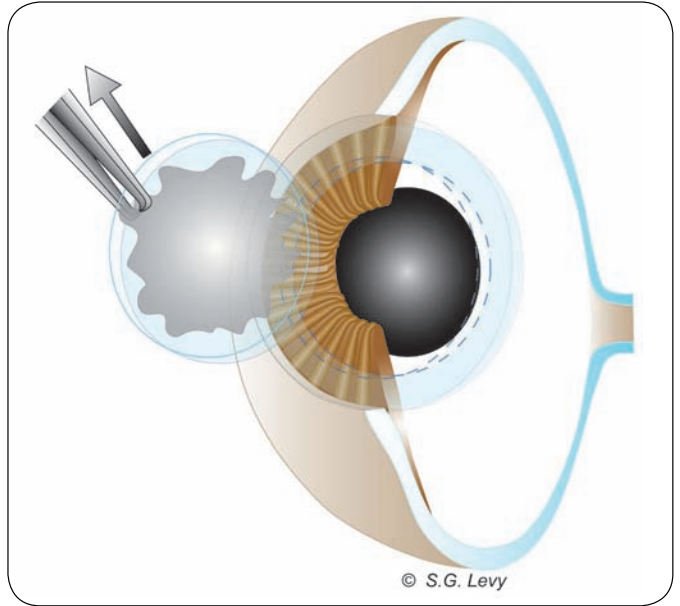


The technique is suitable when keratoconus involves the full thickness of the cornea

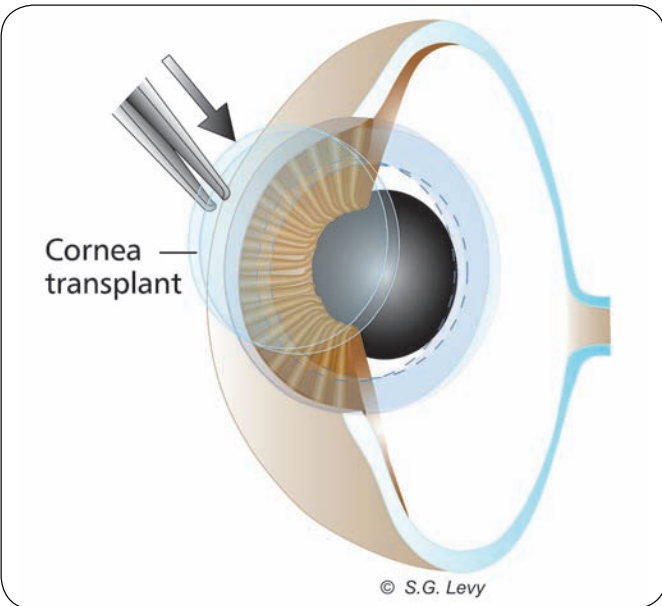




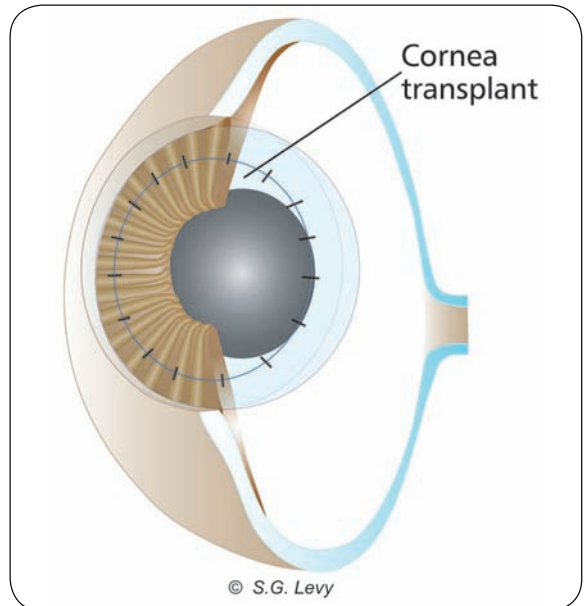
A circular cut is made in the keratoconic cornea



The keratoconic cornea is removed



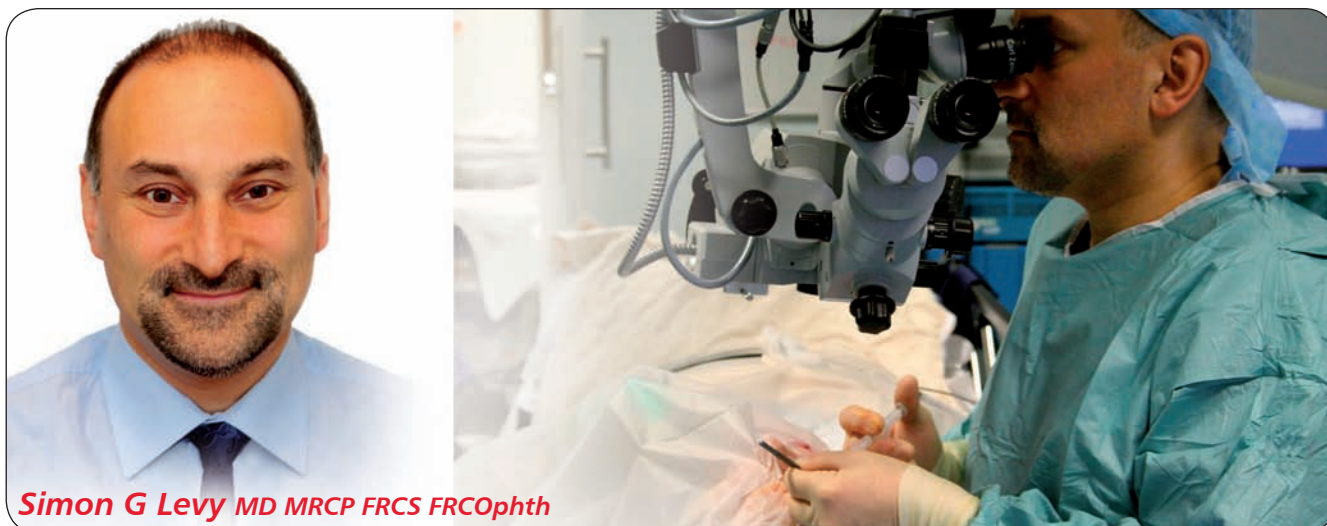
A healthy cornea is transplanted



The cornea transplant is held in place with ultra-fine sutures which are removed after healing



I performed this full thickness cornea transplant for a patient with keratoconus



I am a Consultant Ophthalmologist in private and NHS practice, whose expertise is in the 'front' of the eye:

- Cataract surgery
- Refractive surgery
- Cornea surgery
- Eye surface disorders

My specialist Fellowships were at Moorfields Eye Hospital, London and the prestigious Gimble Eye Centre in Canada where I was trained by Dr Howard V Gimble, a founding father of modern eye surgery. I hold the FRCS and FRCOphth Diplomas in clinical ophthalmology and a research MD on cornea pathology. Before becoming an ophthalmologist I trained in internal medicine and was awarded the MRCP (UK) Diploma.

My clinical emphasis is on – maximising quality of vision and minimising spectacle dependence after cataract and refractive surgery; optimising my patients' physical and mental comfort during surgery; innovative surgical procedures.

I was amongst the first surgeons in the UK to perform Lasik. Other career highlights include performing the first no-suture cornea transplant in the UK, advising the National Institute of Clinical Excellence (NICE) on Intacs cornea implants for keratoconus and implanting the first lens implant in the UK made by Rayner (the company that in 1949 manufactured the world's first lens implant) capable of correcting presbyopia and astigmatism simultaneously.

My private surgical practice is at selected centres with superb equipment – Spire Bushey Hospital, the Wellington Hospital London and Optegra Eye Care Guildford. In the NHS I am at North West London Hospitals NHS trust.

Outside work I have a young family, enjoy the visual arts, travelling and hill walking and have gourmet tendencies.

Appointments

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