Keratoconus Group
Newsletter Spring 2013

Diary Date – AGM and members' meeting in London

Saturday 23rd March 2013 at 10:30am at the Clinical Tutorial Complex, Moorfields Eye Hospital (2nd Floor) City Road, EC1

Our speaker will be Louise deBoard who has recently been appointed counsellor at Moorfields and will give an introduction to the new Patient Support Service at the hospital.

All welcome. Any queries call Anne on 020-8993 4759 or email anne@keratoconus-group.org.uk. We hope to see lots of people there.

Inside this issue

New Surgical Intervention in Keratoconus

This is a summary provided by Daniel Gore, Corneal Fellow at Moorfields at the October meeting of the KC Group in London.

Dan began his talk with some basic facts about KC.

In keratoconus, progressive steepening of the cornea distorts its shape, makes it difficult for light to focus on the retina and results in a deterioration in vision. KC is currently estimated to affect 1 in 1750 Caucasians and 1 in 440 Asians. Mild KC can be corrected by spectacles, but many people go on to contact lenses (usually RGPs) and up to 20% will need a corneal transplant.

Quality of life deteriorates even though people may be able to read a fair way down the eye chart due to issues of light sensitivity, variability of vision, limited wear time of contact lenses etc. An American study estimated the 40 year costs of having KC at 25,000 dollars (half of that sum being the costs associated with a corneal transplant). By contrast, the cost to the NHS of collagen crosslinking (CXL, covered in more detail later in the talk) is £800 - £900.

So we need to convince NICE (the body that approves new treatments for use in the NHS) to think in the long term.

Survival of corneal transplants

Dan quoted figures from a recent Australian study, which looked at survival rates of penetrating (full thickness) grafts. These

showed graft survival rates of 89% at 10 years, 49% at 20 years, and 17 % at 23 years post-op.

For those who therefore needed a re-graft, subsequent survival rates were shorter – 10 year survival rates for a 2nd graft were 53% and 33% for a 3rd graft.

Survival of corneal grafts is dependent on functioning endothelial cells (the back layer of the cornea). The cell count in this layer goes down with age in all eyes (approx 3% loss per year). In a grafted eye, this cell loss is faster, particularly in the first years post graft. Once too many cells are lost in this layer, the cornea starts to swell with fluid and vision becomes cloudy.

A French study has demonstrated that survival rates for partial (DALK) grafts, more commonly done these days, are considerably better. The endothelial cell loss in the first 5 years post graft was less than half of that seen in penetrating grafts (22% vs. 50%), and from this the French study estimated that 50% of DALK grafts would survive 49 years, compaired with less than 20 years for full-thickness grafts.

"Survival rates for partial grafts… are considerably better (than full thickness grafts)."

Collagen crosslinking (CXL) treatment may completely alter the proportion of KC eyes that need a corneal transplant. There is still grossly inadequate NHS provision of CXL, but Moorfields got the funding to carry out CXL in the summer of 2012.

Is CXL safe? Who to treat, who not to treat?

There are now different versions of CXL so it is important to know the evidence for each. CXL involves the use of riboflavin drops and UV light to bind the collagen fibrils in the cornea together.

In the original method of carrying out collagen crosslinking,

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the epithelium (the outer layer of the cornea) is peeled off to enable the drops to penetrate the cornea. Riboflavin drops are applied for 30mins followed by 30mins of UV light. The longest follow up data published is by the Siena group of researchers who followed up 44 eyes for 4 years. Their results showed an average reduction of corneal steepness of 2 dioptres, a reduction of myopia and astigmatism by 2 dioptres and a gain of 2 lines on the Snellen chart. So CXL stabilised keratoconus and in some cases reversed it.

This study, however, was not a randomised controlled trial so the results have to be treated with some caution. There is now some data from randomised control trials (each patient had one eye treated and the other untreated). Collectively, these showed KC getting worse in 32% of the untreated group and 3% of the treated group, so it is important to note that there is a risk, albeit small, of failure with CXL. 45% of the treated group improved, with significantly better vision. The effects of CXL can continue for up to 2 years before stability is achieved. The results also suggest that treating younger patients, and those with less advanced (steep) disease, is more effective.

The main risks of CXL are infection and scarring. The risk of infection is in the first few days after the epithelium is removed and before it heals. Scarring can occur either as a result of infection, or as part of an abnormal, exaggerated healing process. 3% of patients can be expected to lose up to 2 lines of vision, and may require a corneal graft for improved vision. This compares with a 20% risk of requiring a corneal graft in an 'average' KC eye without CXL. Some corneas will not be suitable for treatment because they are too thin, or because of severe ocular inflammation (e.g. allergies). There is a theoretical risk that UV light could accelerate long-term endothelial cell loss, but so far there is no evidence of this.

All the above results relate to CXL with removal or disruption of the epithelium (epi-off). But it is possible to alter the riboflavin drops to penetrate the epithelium so that CXL is done without removing the epithelium (epi on) which removes the need for a sterile operating theatre. So is 'epi on' CXL truly risk-free and is it as effective?

The published evidence comes from 2 small non-randomised trails in Italy involving around 60 patients.

These showed no complications at all after 18 months and similar effectiveness to '*epi-off*' CXL. However, these numbers are not yet big enough, and a larger randomised trial is required, ideally replicated at different trial sites. NICE (the body that approves treatments for use in the NHS) prefers to have data from more than one trial. The chemicals that were added to the riboflavin to enable the drops to penetrate the epithelium are quite toxic (preservatives, antibiotics) although there is a possibility of using novel molecular compounds that would enable it to penetrate the epithelium in a non-toxic, reversible, way.

Moorfields is currently rewriting the pathway for KC patients. Funding for CXL is only available where progression of KC can be demonstrated. So patients will likely be seen more frequently, with more scans being done in order to demonstrate progression. In future, at diagnosis a Pentacam scan will be done, the vision will be tested and 6 monthly checks will be done for 2 years. '*Epi-off*' CXL will be offered to those whose KC is progressing.

"Moorfields is currently rewriting the pathway for KC patients."

The large scale (>2,500 patients) Moorfields study by Stephen Tuft, quoted in previous newsletters, showed who is most at risk of progressing to the point of needing a corneal graft. The important variables were ethnicity (more patients of Asian or African origin showed faster progression) age (more patients diagnosed in the early teens), cornea already steep at diagnosis, and poorer vision (higher prescription) at diagnosis.

The criteria that will be used to demonstrate progression will be changes in one of the following between scans –an increase in corneal steepness of more than 1.5 dioptres, a reduction of corneal thickness of more than 13 microns, an increase in astigmatism of more than 1.5 dioptres or the loss of more than one line on the Snellen eye chart. CXL will be offered to those aged 16+ who show progression (it is possible to treat younger patients, but specific arrangements, including the choice of anaesthetic, may be required).

Visual rehabilitation

For a large number of people with KC, crosslinking has come too late to be of benefit. However, for those whose KC has now stabilised, there are options of shape correction to improve the vision, such as corneal ring implants, topography

guided PRK and myopic correction with lens implants.

Corneal ring implants (ICRS) are designed to flatten the cornea. The rings are made of PMMA and made in different diameters and different lengths.

Ring insertion can be combined with CXL or as a stand-alone procedure. If combined with CXL, should this be done pre or post CXL? A randomised trial showed that inserting rings first and performing CXL later produced better results with patients gaining more lines on the eye chart and greater reduction in astigmatism. ICRS is available at Moorfields Eye Hospital; the results of corneal rings, however, are unpredictable.

"Topography-guided PRK is likely in the future to be available at Moorfields Eye Hospital for patients who are intolerant of RGPs."

In topography-guided PRK, a laser is used to make the corneal surface more regular. The aim of this is be able to correct vision with glasses, rather than RGPs. While laser cannot normally be used in KC because it would further weaken and destabilise the cornea, it can be used after CXL or if the cornea has already stiffened with age and the KC has stabilised. If combined with CXL, there is again the question of the order in which the procedures should be done. Combining the two has the advantage of just one procedure (done epi off). A study in Athens compared the combined procedure with CXL followed by PRK 6 months later and showed that the combined procedure had better results (although this was not a randomised trial). Topography-guided PRK is likely in the future to be available at Moorfields Eye Hospital for patients who are intolerant of RGPs.

Future directions

Some of you will have read about a proposed clinical trial of '*epi on*' CXL at Moorfields. Funding for this has been turned down, but it is still a possibility for the future, and further applications for funding are in the pipeline.

In future years, it may be that CXL will be offered at the point of diagnosis.

Another possible development could be a genetic blood test for KC. But the fact that there seem to be a large number of genes implicated in KC makes this very complicated, so this is likely to take at least 10 years to develop.

Can you help raise the profile of corneal donation?

As you may remember from recent newsletters, the KC Group has been invited to several meetings with NHS British Transplant in which we have talked about the variations in waiting times for a corneal graft around the UK and more generally about corneal donation. NHSBT has been working hard to raise the profile of corneal donation to ensure there is enough corneal tissue to meet demand. At a recent meeting with their Communications department, we were asked if our members could provide some personal stories of how receiving a transplant transformed their life that could be used in future publicity. If you would be willing to help with this, please contact anne@keratoconus-group.org.uk or ring us on 020-8993 4759.

Fundraising for research into KC and for the KC Group

A big thank you to all the people who sponsored me (Anne) on the Fight for Sight Carrots Nightwalk last September to raise money for research into KC. I was joined by Rachel and her two daughters, Lizzie (who has KC) and Charlotte on our walk round the main sights of London on a dark, but mercifully mainly dry, evening. There were hundreds of other walkers raising money for all sorts of other eye conditions. Also in September the husband of one of our members, Mark, did the London to Paris bike ride, also raising money for KC research. Between the five of us, we raised just over £3000 for Fight for

Sight, the charity that funds research into various eye conditions, including the KC genetic research we reported on in a recent newsletter. Mark also donated another £400 of sponsorship money to KC Group funds, so an extra thank you to him!

"there is a choice of a 6 mile … or a 15 mile walk"

The 2012 Carrots Nightwalk was held only in London but in 2013, Fight for Sight will be organising Nightwalks in Birmingham, Cardiff and Glasgow as well as London. The date for all the walks is 20th September 2013 so I hope more of our members will sign up for this fundraiser this year.

In each city there is a choice of a 6 mile evening walk or a 15 mile night walk. When you register, you will be able to specify which eye condition you want to raise money for. So please members, friends and families, go to www.fightforsight.org.uk/carrots NOW and help raise money for KC research. Do contact us if you have any queries.

NICE consultation on revised guidelines for CXL

The National Institute for Health and Clinical Excellence (NICE) issued guidelines on the provision of corneal crosslinking on the NHS in 2009. Those guidelines said there there was then insufficient research evidence about the efficacy and safety of CXL, but allowed ophthalmologists to apply for funding for CXL from their Primary Care Trusts if treatments were then part of an audit which would provide more evidence. Over the last few years, this has meant that very few hospitals have been able to offer CXL on the NHS, as PCTs have been refusing requests on the grounds that it is still 'experimental'.

Register your interest to take part in consultation on the NICE web site

NICE is now revising its guidelines, and these will be put out for consultation shortly. Just 28 days are given for comments from the date the consultation starts, and we hope that our members (especially those of you who have had the treatment) will contribute to this. At time of writing, we have not yet been notified of the date but will announce it on the KC Group forum as soon as we have it. In the meantime, you can ensure that you'll be informed by registering your interest on the NICE web site: www.nice.org.uk/IP/724.

2013 conference

The KC Group 2013 conference will be held in Manchester on 15th June. It will be an interesting day, with speakers on surgical and contact lens options for KC, a speaker from NHS British Transplant and from the Manchester Eye Bank

Please put this date into your diary. We will be sending out more information and booking forms to our members nearer the time.

Support the KC Group with a standing order

Around 50 of our members have already signed a standing order form making a regular donation to KC Group funds. So we're including a form with this newsletter in the hope that many more of you could spare just £1 a month to keep this charity going. Being able to depend on a regular, predictable amount coming in each year would make a huge difference to our ability to plan for the future and to expand our activities to reach many more people with KC. Like all charities in these times of austerity, we've seen a drop in donations in recent years, although our membership continues to increase (currently over 2100).

So do support us if you can by filling in the form and returning it to us.

2013 Annual General Meeting announcement

The London meeting on 23rd March, announced on the front page, is also our AGM. So as well as hearing about the new patient support service at Moorfields, we will be giving a review of the KC Group's activities in the last year plus a financial report. It's also your chance to meet the trustees who will be standing for re-election. And, of course, if you'd like to become more active and join the committee that runs our charity, this is your chance to put yourself forward. We're always looking for fresh ideas and people who are committed to improving things for all of us with KC!

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