

Congenital Cataracts **Information Booklet**

It is always a difficult and frightening experience to discover that your baby has a rare and worrying condition. Be reassured that everyone in the Ophthalmic Department will be working to achieve the best possible future for your baby's vision.

We hope this leaflet will help you to further understand about cataracts, the treatment and the long term care.

At the end of this leaflet we have included a short account of Connagh, a child with cataracts, and how this affected him and his family. We hope it will provide you with some reassurance.

In a normal eye, light passes through the front of the eye (cornea) to the back of the eye (retina) through the lens. The lens and cornea focus the light so that an image is formed at the back of the eye (retina). This image is transferred via the optic nerve to the brain enabling us to see. All babies use their eyes from the day they are born, stimulating the visual area of the brain. If these areas are not stimulated permanent visual loss can occur. A normal lens is transparent and has a centre (nucleus) and a surround (cortex). The lens is able to change its shape and so focus on objects that are near and far.

What is a cataract?

A cataract is a lens that has become cloudy (opaque – rather like the white of an eye). When it is opaque, light is prevented from reaching the retina so the image is not clear and it is difficult to see properly. The degree of visual loss depends upon:

- Density of the cataract – the more dense the cataract the greater the visual loss
- Position of the cloudiness within the lens:-
Significant vision loss will occur if the cataract is at the back of the lens rather than the front of the lens (posterior nucleus cataract).

Vision will be affected if the cataract is on the outer edge of side of lens (cortical cataract) or in the front of the lens (anterior nucleus cataract).

What causes Cataracts?

All new born babies are examined prior to discharge from the maternity unit and part of that check is an examination of the eyes. This examination is repeated again at six weeks of age. If any cloudiness, or other eye abnormality, is suspected you and your baby will be referred to an Ophthalmologist (eye doctor). This examination will take place in the Outpatients Department and the doctor will be able to see whether your baby has a cataract or not. The eye doctor will put special drops into your baby's eyes to make the pupil large so the eyes can be examined more clearly. These drops will not harm your baby, but the pupils will remain enlarged for a few hours.

Your baby may have a cataract in one eye (unilateral) or both eyes (bilateral). Treatment of unilateral and bilateral cataracts is complex and different but early treatment, and therefore early stimulation to the brain can help to reduce the amount of visual loss.

Your baby's eye doctor will discuss in detail with you, whether an operation in the first few weeks of life is appropriate for your baby's cataract condition.

Unilateral cataract

Surgical Care:

If an operation is necessary within the first few weeks of life you and your baby will be admitted onto a baby ward for a day or two. Your baby's pupil will be dilated prior to removing the lens under general anaesthetic. Either an intra-ocular lens (artificial lens) or a contact lens will be used to improve your baby's vision. If your baby is to have an intra-ocular lens this will be inserted at the time of surgery after the cloudy lens has been removed. If a contact lens is to be used this will be fitted one or two weeks after surgery. Immediately after surgery a pad will be placed over your baby's eye until the following day, when you will both be seen by your ophthalmic doctor.

Your baby will go home on regular eye drops; these will need to be put in every 2 – 4 hours. You will be taught by nursing staff how to do this before you and your baby are discharged home. These drops help to reduce inflammation, scarring and minimise any problems that may occur after surgery.

Your baby may also need a course of occlusion therapy (patching) – this is discussed in the Long Term Management section.

Bilateral Cataracts

Surgical care:

If your baby has bilateral cataracts that are affecting his or her vision, surgery will be carried out as soon as possible. You and your baby will be admitted to a baby ward and may have to stay overnight. Generally both eyes will be operated on within a week of each other, but you and your baby will be able to go home and come back again for the second operation. Your baby's pupil will be dilated prior to removing the lens under general anaesthetic. Either an intra-ocular lens (artificial lens) or a contact lens will be used to improve your baby's vision. If your baby is to have an intra-ocular lens this will be inserted at the time of surgery after the cloudy lens has been removed. If a contact lens is to be used this will be fitted one or two weeks after surgery. Immediately after surgery a pad will be placed over your baby's eye until the following day, when you will both be seen by your ophthalmic doctor.

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Post-operative complications

Eyes are very delicate and complex organs and problems can occur after surgery has taken place. We have listed below a number of problems that can occur, but your baby's ophthalmic doctor will discuss them in greater detail if they do occur. In order to pick up any post operative problems your baby's eyes will be regularly checked.

Glaucoma (raised pressure within the eye).

Fluid in the eye is unable to drain away causing a build up of pressure. This increased pressure can eventually cause some degree of visual loss. It is seen in up to 25% of children undergoing congenital cataract surgery. Often it can be controlled with medicine and eye drops but may, in some infants, require further surgery.

Amblyopia (Lazy eye).

This can occur if the vision in one eye is stronger than in the other. The brain learns to ignore the vision it receives from the weak eye, causing varying degrees of visual loss in that eye. Occlusion therapy (patching) will be started – this involves your child wearing a 'patch' over the strong eye to try and improve the weak one.

Strabismus (Squint).

A squint may develop as a result of a 'lazy eye'. Treatment of the squint will depend upon its severity and further surgery may be required.

Pupil abnormalities.

Sometimes the pupil may become oval or off centre after surgery – this is quite common and will not affect your baby's vision.

Endophthalmitis.

This is an extremely rare bacterial infection which can have devastating consequences for the operated eye. Most cases are diagnosed and treated in the immediate post-operative period.

Long term management

All children who have had cataracts removed require long term follow-up. This follow-up takes place in the out-patients department where your child's vision will be tested on a regular basis by specially trained staff (Orthoptists) so that any change can be dealt with as soon as possible.

Vision, in the majority of children, can be further improved by the use of contact lenses or glasses. These will be fitted within a week or two of your child's operation. Contact lenses are used for babies as well as for older children. Inserting contact lenses into your child may seem daunting, but rest assured that you will be taught all about contact lens care and help will always be available.

Occlusion therapy (patching) will be necessary for the majority of children who have undergone cataract surgery. Patching is the covering of one eye each day with a specially fitted plaster, rather like a pirates patch, preventing that eye from seeing. As most children after surgery will have one eye stronger than the other, patching is done to encourage the use of the weak eye and so prevent further visual loss in that eye. Patching will not hurt your child although some children need a lot of encouragement to keep the patch on. The type of cataract your child, has and the degree of weakness in your child's eye, will determine when your child has to start patching and for how long. This will be discussed with you in detail by your child's orthoptist and ophthalmologist.

The treatment of cataracts is complex and varied and the outcome is difficult to predict. Some children will ultimately have poor vision but it is extremely rare for these children to have no vision. The majority of children attend mainstream school, with extra educational support, and many will have vision good enough to allow them to drive.

Connagh's Story

Joanne and Christopher were aware of congenital cataracts before their baby was born. Christopher had been born with one eye affected, as were a number of other members of his immediate family. However it was still a tremendous shock when Connagh was born with both eyes affected.

Joanne recalls how during the routine neonatal examination when Connagh was just two days old the paediatrician said, "*There's no red reflex*". All Joanne kept thinking was, "*Not Connagh, not our baby*". Confirmation from the Sunderland Eye Infirmary and an immediate referral to the RVI made the whole process more real. Joanne remembers with gratitude that it was Mr Clarke's secretary who rang them at home to make that initial appointment.

Christopher and Joanne found this first consultation the most traumatic – not only was Connagh's examination thorough and long, involving other professionals, but there was a detailed family history to be recorded. Mr Clarke explained the nature of Connagh's bilateral cataracts and the likely outcome. They could not take all the information in at once but Joanne felt immense relief that their new baby did not require surgery, instead there were regular check ups at the RVI, first every 3 months, then later every 6 months, always with the thought uppermost in their minds – would this visit bring the operation nearer.

At first it seemed that Connagh's development was not delayed by his restricted vision, this was partly due to the extra help given by his parents in adjusting the home environment to make it easier for Connagh to find his way safely around the home. The use of bright coloured mugs and dishes was helpful too. Most importantly Joanne and Christopher made sure that they allowed Connagh to explore things for himself, whilst keeping a watchful eye on him. However, as Connagh grew Joanne and Christopher noticed that he would always hold their hands when he was outside, even on the beach or at the park he would stay close to them and not run.

At 2 years Connagh was prescribed his first glasses. These helped and probably made Christopher and Joanne aware of how difficult life had been for him. When Connagh was 4 years old, as a result of the increasing discomfort and pain in his right eye, the decision was taken for surgery. Although his parents knew this would happen, and in many ways pleased that it would as they knew it could improve his sight, they were still very worried about the immediate after care. From personal experience with the extended family Christopher and Joanne knew that poor aftercare could lead to permanent sight loss. This was their fear. Connagh was accepting of the hospital procedures, the ward and even going to theatre. In fact he was a star patient. The operation to remove the cataract and replace it with a lens, led to a vast improvement of the vision of the right eye. An extremely careful four week aftercare followed, including the constant use of eye drops.

The positive side was gradually realising how much better Connagh's vision became. This realisation came from listening to how Connagh described the things he could now see. His grandma was astonished when he asked, "*What are the black things hanging from the lights?*" They were the wires holding the Christmas lights together – Connagh had never seen them before.

Connagh has been registered partially sighted and now has special needs input at his mainstream school following his educational statement. Connagh loves school and Joanne sees the educational input as very helpful.

Joanne and Christopher felt that being given as much information as possible has been right for them. Joanne's stressed the need for someone who will listen to your anxieties and fears. Joanne felt that the information they received should include the possible negative outcomes, as well as the hoped for positive improvements.

Soon Connagh will have an operation of his left eye, and although he will need regular check ups during his life, the future looks brighter for all the family.

The most helpful aspect of all has been Connagh himself. He has a sunny accepting disposition. Joanne acknowledges that without this she would have found the last few years much harder to cope with. Connagh has taken everything in his stride and still comes up smiling, incredibly chatty and friendly.

Useful contact number:

Cat's Eyes (parents caring from children with cataracts)

01730 815638