

Congenital Glaucoma **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 that this leaflet will go some way in helping you to understand about cataracts, its treatment and the long term care.

At the end of this leaflet we have included a short account of how Congenital Glaucoma affected one of our families. We hope it will provide some reassurance to you.

For an eye to be healthy, aqueous fluid (aqueous means 'watery' in Latin) circulates in the eye, and then flows out through the eye's drainage angle. The drainage angle consists of sponge like tissue (Trabecular meshwork) through which the fluid passes via a pipe (the canal of Schlemm) into blood vessels around the eye.

What is Congenital Glaucoma?

Baby's eyes develop in the womb. In babies with Congenital Glaucoma the drainage angle does not develop properly and the aqueous fluid is unable to drain away. This increases the pressure in the eye/s.

In a small number of babies Glaucoma is inherited but for the vast majority of babies the cause is unknown.

How will the increased pressure affect my baby's sight?

The increased pressure may cause:

- The cornea/s to become cloudy. In some babies this cloudiness may be obvious at birth. A cloudy cornea makes it difficult for the brain to receive a clear picture, so the brain begins to stop using this eye/s making it lazy (Amblyopic).
- The eye/s slightly change shape or become enlarged. This too alters the way your baby sees, making your baby short sighted with 'blurry' vision.
- The eventual damage of the optic nerve, causing varying degrees of vision loss.

Why would you suspect Congenital Glaucoma?

Congenital Glaucoma is rare and therefore not always easy to detect. But it is suspected if:

- The colour of your baby's eye/s seem to change gradually and become cloudy, or when they are cloudy at birth.
- The eyeball/s may become enlarged or gradually change shape.
- You baby may cry a lot and be generally miserable.
- Feeding your baby maybe difficult as s/he may be reluctant to feed or may vomit more than you would expect.
- S/he may have red and watery eye/s and be upset by bright lights, turning his/her head away from the light or try and bury his/her head into a pillow.
- Sometime there is a family history of Glaucoma.

How is Congenital Glaucoma treated?

All babies born with Congenital Glaucoma will require surgical intervention on the affected eye/s. Quite often the surgical operation is carried out at the same time as the examination, under an anaesthetic (EUA). You and your baby may well have to stay in overnight.

There are a number of different types of operation that can be performed on your baby. Two of the more common ones are:

Goniotomy – a small opening is made into the Trabecular meshwork so that the fluid can drain away. Sometimes there is some bleeding after the operation and blood can be seen in front of the eye, but this is slowly absorbed by the eye. If the eye is very cloudy, it may be necessary for the surface ‘skin’ of the cornea to be stripped away so that the doctor can see the eye better. It will be necessary to put eye drops into your baby’s eye after the operation as this helps the healing. You will be taught how to do this by the nurses before you and your baby go home.

Trabeculectomy – a small trapdoor is made in the white of the eye (Sclera) and a piece of the angle tissue (Trabecular meshwork) is removed before the trapdoor is replaced. Fluid in the eye filters out through the trapdoor forming a small blister in the conjunctiva. Drops will need to be put in the baby’s eye afterwards to help healing. This procedure may have to be repeated if the trapdoor heals over.

One of the main problems after any operation is the formation of scar tissue. This tissue can block the flow of fluid out of the eye and cause a rise in pressure. To try and minimise this problem a small amount of an anti-scarring drug, either Mytomycin or 5FU, is applied to the operation area for a few minutes during surgery.

As your baby’s eyes will be examined under an anaesthetic (EUA) on a regular basis to monitor the pressures, this anti-scarring treatment maybe repeated if your baby’s consultant feels it is necessary.

Your baby will need to wear an eye patch after the operation, usually until the following day when you will both be seen again by the doctor in the out-patients department. You will be shown how to put eye drops into your baby’s eye/s as they may need to be put in every two hours during the day. These drops will help heal your baby’s eye/s and prevent infection. Your baby’s eye/s will be examined again, in the Out Patient Clinic, three to four days after surgery.

At this time your baby’s doctor will also make, if appropriate, a referral to the Advisory Service for children with a Visual Impairment. This service works specifically with babies and children who have a visual impairment as the condition will have an impact on their learning and development. They will work with you and your baby, in your own home, and will advise and introduce you to different activities that will help you baby develop.

Longer term management

Babies with Congenital Glaucoma sometimes have more day to day problems than other babies, although the problems will not be there all the time. It is unclear why this should be but it is thought that it may be related to the raised pressures in their

eye/s, and/or the medication they have to take, or the eye drops used to help control the eye pressures. These problems are real and worrying to you as a parent.

Your baby may be fractious and irritable; nothing seems to pacify him/her. He or she will have difficulty in sleeping because of his or her fractiousness. At times it may be difficult to establish a regular feeding pattern because your baby feels unable to feed and may vomit feeds back. There may be periods when your baby does not open his or her eyes because the eye/s are sore and sensitive to light. These problems come and go but gradually improve as your baby grows older.

Your baby's eyes will be checked regularly to make sure the pressures are not rising. The most accurate way to measure your baby's eye pressure is with a EUA (examination under anaesthetic) as a day case. Eye drops will need to be put in before your baby is examined to enable the doctor to examine the back of the eye.

Your baby's eyes will need to be checked regularly throughout his/her life. How often these examinations take place will depend on your baby's progress. Your baby may require a number of surgical procedures to control the build up of pressure. As your baby grows eye patches or glasses may need to be worn to help improve and maintain the eyesight.

On a positive note, many babies treated for Congenital Glaucoma have good vision and lead a normal active life.

Verity's Story

What a joy there is in the birth of a child! The anticipation, excitement and wonder at this new member of the family.

Verity's birth was no exception. Jayne and Steven, her parents, were overcome by the birth of their beautiful baby daughter. Everyone who saw her commented, "What a beautiful baby, with huge blue eyes!" Although Jayne wondered about Verity's seeming inability to focus as she got older, she thought it was normal for her new baby. And the periods of intense crying was put down to colic.

It was at Verity's six week check that the excitement surrounding her birth was shattered by the realization that the 'normalities' of large blue eyes and the 'colic' could be due to some problem with Verity's eyes. The General Practitioner arranged an immediate referral to the Ophthalmic Department at the RVI. Just a few days later Jayne and Steven's life was turned upside down. The intense crying was due to increased pressure in Verity's eyes which also explained their size and her inability to focus. Verity had Congenital Glaucoma. Without warning or preparation Jayne and Steven had to try and accept that their daughter's future sight was endangered and the treatment to save Verity's vision was going to be long, and at times, distressing and painful.

What helped at that moment was the time given by the Ophthalmic Consultant in patiently explaining the problem and answering their questions. This enabled Jayne and Steven to consent to an emergency surgical procedure, the first of many. What has continued to help them is the support given by the staff at the hospital.

"It was hard, sometimes you just felt you had been to hell and back, what kept you going was holding on to the long term aim. Yes the surgery, visits to the hospital, the regular long term use of eye drops, are all constantly wearing, and often you feel it is you causing distress to your baby, but without all this Verity's vision would have deteriorated. She would have suffered more pain with the pressure in her eyes and maybe, eventually, she would have lost her sight.

Now Verity is 21 months old and we can see the light at the end of the tunnel. Watching Verity look around and see flowers in the garden, and point out the birds and aeroplanes in the sky, even point to crumbs on the carpet!, makes it all worthwhile."

Verity is accepting of the daily routine of eye drops and regular hospital visits. These are now part of her life.

"You do need help and support. Steven and I have grown closer together, the help and support from family is invaluable. We know that Verity's vision is as good as it can be and we have been part of the team which achieved this."