

Retinopathy of Prematurity **Information Booklet**

You will have many worries and anxieties, as your baby has been born so prematurely. This is a very difficult time for you. Additionally you have now been told that your baby has Retinopathy of Prematurity, a potentially damaging condition. We hope that this short information booklet will help you to understand the condition and the doctor's explanation.

We have included a short account of one of our families who is further along the road. We hope that you will find this encouraging.

All the staff of the Ophthalmic Department will be striving to obtain the most positive outcome for your baby's vision.

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). The retina is a very complex and sensitive structure that converts the light image into nerve impulses that are 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.

To understand why infants develop retinopathy of prematurity (ROP) it is important to have some understanding of the normal development of the eyes in babies. Babies' eyes develop throughout the 40 week gestational period. Blood vessels begin to supply the retina with both oxygen and nutrients from 16 weeks gestation. They begin to grow from the optic disk and gradually fan out towards the edge of the retina. Between 28-30 weeks these blood vessels begin to grow much more rapidly fanning out over the back of the retina. The retina is not, however, fully supplied with blood vessels until the baby is full term. Once a baby is born and opens its eyes light passes through the lens and is focused onto the retina. The image is passed via nerve cells and fibres, through the optic nerve to the visual area in the brain. When babies are born more than 10 weeks early (30 weeks) problems may arise with the development of the retina.

Many babies born so soon require extra oxygen to help their developing brain but the oxygen can cause the very fine blood vessels in the developing eye to go into spasm. This can cause the blood vessels and surrounding tissue to become inflamed and swollen diminishing the blood supply to the developing tissue. Eventually scarring may occur on the retina. This damage is known as retinopathy and can lead to vision loss. The degree of vision loss is determined by the amount of damage the developing eye suffers.

The period of accelerated growth diminishes after the baby has reached 30 weeks and further damage to the eye recedes as the eye is beginning to mature.

There are five stages of retinopathy of prematurity.

Stage 1: The outer aspect of the retina is affected by the diminished blood supply and is shown by a clear demarcation line. About 80% of babies born more than 10 weeks early will suffer some degree of Stage 1 retinopathy. This usually resolves spontaneously without any significant effect on vision.

Stage 2: Any damage remains in the outer aspect of the retina but there is a demarcation line that becomes thickened and ridged.

Stage 3: New blood vessels developing on the ridge will be abnormal. These abnormal vessels can lose their elasticity and begin to contract leading to Stage 4.

Stage 4: The contracting blood vessels begin to pull the retina away from the back of the eye.

Stage 5: The retina is totally detached from the back of the eye.

Retinopathy of prematurity only occurs in premature infants. The survival of premature infants is improving all the time, but retinopathy remains a difficult problem to prevent. As more premature infants survive the more common retinopathy of prematurity becomes.

What are the symptoms of retinopathy of prematurity?

There are no symptoms of retinopathy therefore all small babies born less than 32 weeks gestation are screened by a specialist eye doctor (Ophthalmologist). Usually the screening begins when your baby is either six weeks old or is 33 weeks gestation and usually takes place fortnightly until the blood vessels in your baby's retina are mature.

What happens during the screening checks?

Drops will be put into your baby's eyes to enlarge (dilate) the pupils, this is necessary for the doctor to see your baby's retina. Once your baby's pupils are enlarged anaesthetic drops will be put into the eye so that your baby does not feel any pain during the examination. The doctor will hold your baby's eyelids open, either by hand or with a small instrument, and look closely at the back of your baby's eyes with a special light. The doctor may need to gently press around your baby's eyes with a special probe, in order to get a better look. The examination only takes a few minutes. Your baby will not like the examination but should only feel slight pressure, but no pain.

Your baby's nurse will keep you fully informed about the state of your baby's retina and let you know if treatment is needed.

How is retinopathy treated?

Stage 1 & 2: If your baby has either of these two stages no treatment is necessary, but your baby's eyes will still be closely monitored.

Stage 3: If your baby has stage 3 retinopathy, laser treatment will be given. The laser is a special piece of equipment that emits a very bright and fine beam of laser light. This laser light destroys the abnormal blood vessels that cause retinal scarring. Laser treatment involves your baby having a general anaesthetic and usually takes 2 – 3 hours. Normally only one laser treatment session is necessary, but the doctors will continue to check your baby's eyes on a regular basis. Laser treatment can cause your baby's eyes to become a little red and inflamed but this settle down in a few days.

The nurses will put special drops and ointment into your baby's eyes to help them to heal.

Stage 4 &5: Stage 4 and Stage 5 retinopathy are very rare and the treatment is complicated. Your baby's ophthalmologist will discuss this with you in great detail.

What is the long term outlook for babies with retinopathy?

Currently 80% of babies with Stage 3 retinopathy do well and have good vision, but will be regularly checked. Even with treatment a small number of babies with Stage 3 retinopathy will have significant visual loss.

Most babies, whose eyes do well initially, will continue to see well. However, all pre-term babies have an increased risk of developing eye problems as they grow older. These problems may include:

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.

Myopia (Short-sightedness):

This is when the rays of light passing through the cornea are unable to focus onto the retina, causing distant objects to become blurry. This is usually corrected by wearing the appropriate glasses or contact lenses.

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. Often it can be controlled with medicine and eye drops but may, in some infants, require further surgery.

All of these conditions, if they arise, will be discussed in much greater detail by your baby's eye doctor.

If your baby has had Stage 1 and Stage 2 retinopathy, regular visits to the Ophthalmic Outpatient's Department are not normally necessary. If any eye problems develop in the future your child will be referred back to the Ophthalmologist by your own GP.

However, if your baby has had either Stage 3, 4 or 5 retinopathy, your baby's eyes will need to be checked frequently. Initially you and your baby will attend the

Ophthalmic Outpatient's Department every two to three months, then every six months and finally on a yearly basis until your child is about 5 years old. Once discharged from the Ophthalmic Department your own GP will make the referral back if any further problems arise with your child's eyes.

For the vast majority of infants, referrals back to the Ophthalmic Department are not necessary.

Elliot's Story

As with other families whose baby is born prematurely Chris and Jonathan suffered many anxieties over the health of their twin boys, initially it was a question of their survival.

Chris had gone into premature labour at 26 weeks gestation and there was a frantic search for a Special Care Baby Unit that could undertake the delivery and care for the two infants. Jonathan remembers the fear and loneliness that gripped him after being left at the local hospital whilst his wife was urgently transferred to North Tees Hospital.

Elliot and Lewis were born by caesarean section to improve their chance of survival. Nevertheless they were extremely ill with the many problems which affect a premature birth. Chris and Jonathan were aware that particular medical interventions designed to assist breathing can contribute to retinopathy, but the most pressing need was just keeping the babies alive. The neonatologist spoke to Chris and Jonathan regularly in regard to the multitude of problems that were occurring or could develop, including retinopathy. The babies' eyes were checked fortnightly once the babies were about six weeks old, although Chris and Jonathan did not stay for the examination. As with so many of the invasive tests and procedures Chris and Jonathan felt unable to be a part of the, *"It didn't feel that the babies were ours."* However the expert intensive care was enabling the babies to survive and grow. Their confidence in the medical and nursing teams helped Chris and Jonathan to cope. For Chris and Jonathan developing a strong bond with the boys was put on hold for a few weeks.

As Elliot and Lewis grew older and stronger they were transferred back to Newcastle, first going to the RVI, and then Newcastle General. It was better to be closer to home, but difficult to leave the unit where the babies had spent the first 12 weeks of their lives. At Newcastle General, Mr Clarke the Ophthalmologist met the babies and once again went through the development and care of retinopathy. It was here that an examination showed that Elliot's retinopathy had developed to Stage 3. Chris and Jonathan grew anxious and worried that Elliot may become blind. The operation for laser treatment was agreed. Jonathan recalls watching protectively as Chris carried their tiny son through the corridors at the RVI, towards the ward. Elliot was in theatre for over two and a half hours, a time of intense anxiety. Thankfully as Chris and Jonathan recall, Elliot did not require to be ventilated again following the surgery. The operation had taken place just before Christmas. It seemed a long wait until the New Year, when they found out that the operation had been a success. Lewis's retinopathy was much less severe and he avoided this surgery but has developed a squint.

The boys have their vision tested every six months and it appears that it has not been affected. This is a tremendous bonus. The boys will continue to have their eyes checked regularly over the next few years. Although Elliot and Lewis had a very traumatic first two years of life, particularly so for Elliot, both parents now feel that they are sailing onto calmer waters.