

Department of Ophthalmology

Paediatric Ophthalmology

Congenital cataract

Parent Information Leaflet

The aim of this leaflet is to ensure that you are informed about your child's diagnosis of congenital cataract.

The eye is made of three parts:

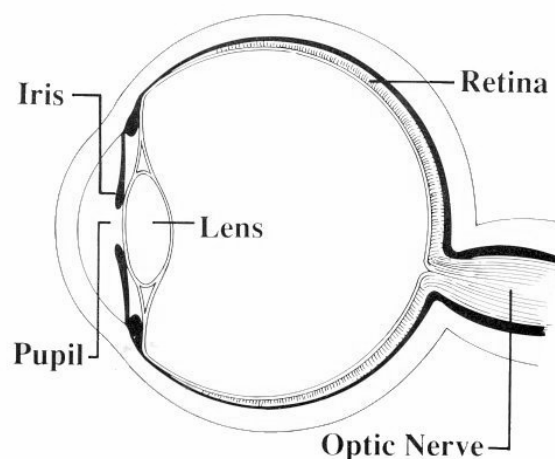
1. A light focussing bit at the front (cornea and lens)
2. A light sensitive film at the back of the eye (retina)
3. A large collection of communication wires to the brain (optic nerve)

A curved window called the cornea first focuses the light. The light then passes through a hole called the pupil. A circle of muscle called the iris surrounds the pupil. The iris is the coloured part of the eye. The light is then focused onto the back of the eye by a lens. Tiny light sensitive patches (photoreceptors) cover the back of the eye. The covering of photoreceptors at the back of the eye forms a thin film known as the retina. Photoreceptors collect information about the images you see. Each photoreceptor sends its signals down very fine wires to the brain. The wires joining each eye to the brain are called the optic nerves. The information then travels to many different special 'vision' parts of the brain. All parts of the brain and eye need to be present and working for us to see normally.

What is a cataract?

A cataract is cloudiness in the normally clear lens of the eye. If the lens is not clear then not all the light can get into the eye or it may be scattered by the cataract, blurring the vision and causing visual problems with dazzle and glare.

A cross section of the eye:



Congenital cataract

A congenital cataract is cloudiness in the lens of the eye that is present at, or develops shortly after birth. About 1 baby in 10,000 is affected by congenital cataracts. Sometimes the cataract only affects one eye (unilateral), but often both eyes are affected (bilateral).

What are the causes of cataract?

Unilateral cataracts:

Most children with a cataract in only one eye have good vision in the other. Often there is no history of childhood cataracts in the family, the child is healthy in every other way and no cause for the cataract can be found. Sometimes there are other structural problems in the eye besides the cataract, such as it being smaller than the other, which suggest that a problem occurred during the development of the eye before birth.

Bilateral cataracts:

There are four main groups of conditions that cause congenital cataracts:

1. Inherited, genetic cataract conditions
2. Infection of the unborn baby in the womb
3. Conditions that affect the normal metabolism of the child
4. Some specific eye conditions that cause cataract

What are inherited or genetic cataracts?

About one fifth of children with congenital cataracts will have close relative with the same condition. This is caused by a misprint in the genes responsible for the development of the lens. There are many different kinds of inherited cataract conditions. Additionally, congenital cataracts are common in children with chromosomal abnormalities, such as Down syndrome. If your baby has bilateral cataracts but no family tendency for this, some blood and urine tests will be performed to exclude other causes. Often no cause for bilateral cataracts can be found and these are called idiopathic cataracts.

What infections might cause cataract in the unborn baby?

If the mother catches certain infections during pregnancy, the unborn baby is more likely to have cataracts. These include:

1. German measles (rubella)
2. Toxoplasmosis
3. Chicken pox (varicella)
4. Herpes and cytomegalovirus infections

These germs can infect the baby in the womb or during birth and can damage the lens causing cataract.

What metabolic problems can cause cataracts?

Metabolism is a word used to describe the way our bodies make energy from food. Metabolic conditions where cataract is more common include:

1. Galactosaemia
2. Lowe syndrome
3. Diabetes

What other eye conditions cause cataracts?

There are some eye conditions that occur with cataracts. These include:

1. Leber congenital amaurosis and retinitis pigmentosa
2. Aniridia
3. Retinopathy of prematurity
4. Microphthalmia and anterior segment dysgenesis
5. Uveitis (eye inflammation)

How is a cataract diagnosed?

Soon after a baby is born (and at their six week check) a doctor will check your baby's eyes for signs of cataract. If a cataract is noted at either of these examinations an urgent consultation with a specialist (paediatric ophthalmologist) will be made. Sometimes cataracts develop after these screening tests and you might notice:

1. The baby has a white spot or a totally white pupil, in one or both eye
2. A red eye reflection is not visible in one or both eyes with flash photography
3. The baby does not respond to faces, pay attention to and follow toys
4. The baby's eyes wander and wobble (nystagmus)
5. The baby's eyes are not aligned, one turns in or out (squint)

How does cataract affect the way a child sees?

Cataracts can affect the vision in different ways depending on the child's age and the severity of the cataract.

If the cataract is mild your baby's vision may not be affected and the ophthalmologist may decide that early surgery is not required. Your child will be prescribed glasses if necessary and will be reviewed quite frequently to ensure that his/ her vision is developing normally. If the mild cataract is in one eye only, you may be asked to patch your baby's other eye in order to prevent the eye with the cataract becoming lazy (amblyopic).

If a severe cataract has been present from birth, the child may develop lazy eyes (bilateral amblyopia) if surgery is not carried out within the first 3 months of life. The specialized vision part of the brain develops very rapidly over the first few months of life. If a clear, sharp picture is prevented because severe cataracts are present during this critical period, the brain will never develop the ability to see clearly.

What happens during the operation?

Your baby will be admitted to the children's ward for their surgery. He/ she will have to fast before the anaesthetic is given. Before the operation, drops will be instilled into your baby's eye(s) to dilate the pupils. You will be able to go with your baby to the anaesthetic room. Usually anaesthetic gas is used to put babies and young children to sleep for the operation.

If you would like to know more please ask a member of staff for a copy of the parent information leaflet "Advice for parents whose child is having an operation".

The operation usually takes 1 1/2 to 2 hours. During the operation the surgeon will remove the cataract and some of the support structure of the lens and vitreous (the clear gel that fills the space between the lens and the retina of the eyeball). An acrylic intra-ocular lens (a lens that is implanted within the eye) will be used in suitable cases; a contact lens may be used in other cases. The small incision sites

are carefully sewn up with dissolvable stitches. An antibiotic and anti-inflammatory medicine is injected under the conjunctiva (the thin, transparent tissue that covers the outer surface of the eye) – this often results in a raised white patch and some blood staining on the white of the eye.

Following the operation

After the operation your baby will have a small see-through shield on his/ her eye to stop him/ her from rubbing it. You will be asked to start on very frequent drops, these are very important and reduce the inflammation in the eye following surgery and keep the pupil dilated. The surgeon will need to examine your baby's eye the day after the operation before you go home.

At home

Your baby will need to continue on frequent drops and will be reviewed within a week in eye clinic. If you have difficulty putting in the drops, lie your baby down on his/ her back and drop a pool of 3-4 drops in the inside corner of the eye, these will run into the eye when he/ she blinks. It is better to be sure and get more drops in the eye than to miss!

Try to stop the baby rubbing his/ her eye and avoid swimming for 1 month.

The eye will look red and may be sticky for a few days; if it is difficult to open the eye in the mornings then bathe gently with sterile water or boiled **cooled** water and a make-up pad, not cotton wool.

Benefits

To support your child to develop the best vision they can.

Risks

If left untreated your child will be visually impaired.

What are the risks of surgery?

1. Capsular thickening (the capsule is the outer bag of the natural lens that is left behind during surgery to hold the artificial lens or implant) and membrane formation
2. Retinal detachment
3. Infection
4. Glaucoma

Babies and young children develop much more scar tissue in their eyes after surgery than adults. Almost all babies undergoing cataract surgery will develop scar tissue over the lens implant, and will need further surgery. This usually happens within 4-12 months after the initial cataract surgery.

Rarely, the retina (the light sensitive film lining the inside of the eye) becomes detached. If detected quickly the retina can be re-attached surgically but usually the visual outcome is poor.

Infection in the eye after surgery only occurs in 1 in 1000 cases. Unfortunately this results in very poor vision. Surgeons avoid operating on both eyes under the same anaesthetic to reduce the risk of an infection occurring in both eyes.

Glaucoma is a high pressure in the eye, which damages the optic nerve and tends to occur years after the initial surgery. Glaucoma can affect up to 30% of children who have had early cataract surgery. If detected early, glaucoma can be treated with eye drops although some children do need an operation to keep the pressure controlled. Your child will need regular measurements of his/her eye pressures taken and this often means repeated examination under anaesthetic in the toddler years.

Other problems to consider:

1. Glasses and contact lens correction
2. Patching therapy for lazy eye
3. Wobble (nystagmus)
4. Eye misalignment (squint)

Glasses and contact lens correction

Since the eyes grow quite rapidly in the first years, your baby's glasses prescription will need to change to keep pace. If a lens implant has been used, the baby will need to grow into the lens strength and will require quite thick glasses or a contact lens correction for the first few years after cataract surgery. If needed the contact lenses are inserted by a contact lens practitioner in the clinic and are changed every few months. The artificial lens implant does not change focus like a natural lens and so your child will always need glasses.

Patching therapy

If the cataract affects only one eye and the other eye has good vision. The brain will continue to prefer to use the vision in the normal eye to the one that has cataract surgery. The eye will be lazy (amblyopic). In order to improve the vision in the lazy eye, your baby will need to have the normal eye patched for the majority of the time he/ she are awake and patching will need to continue until the child is 8 years old. This is a considerable burden on the child and family. A child only needs good vision in one eye to be able to drive and undertake most occupations in later years. Some parents decide not to go ahead with surgery for a unilateral cataract for this reason.

Wobble

Nystagmus (a to and fro movement in the eyes) is commonly seen in children with bilateral cataracts. It is sometimes also seen in children who have unilateral cataracts, especially if the vision in the eye is very poor. Because the eyes are constantly moving, fine detail in the vision is lost. Often the child will tilt or turn his or her head to place the eyes in a position where they wobble less to improve the vision. Glasses do not correct nystagmus, although they may help a little.

Squint

Most children who have had congenital cataracts develop a squint or misalignment of the eyes. Usually one eye will tend to turn in. This is another factor that may make the squinting eye lazy and patching of the other eye may be advised. If the squint becomes a social issue, squint surgery can be performed to improve the alignment. This is usually delayed until the child starts school.

How can parents, family and teachers make a difference?

There are lots of things that you can do to help your baby make the most of his/ her vision. If your child has been prescribed glasses, contact lenses or a Low Visual Aid it is important that they are encouraged to wear and use them.

This will help your child see more clearly and ensure the vision parts of the brain grow and develop. If your child dislikes bright lights ask your ophthalmologist for prescription sunglasses.

Your child may have difficulty learning to read and keeping up with schoolwork. Your ophthalmologist will arrange for the visual impairment teachers to visit you at home to help you stimulate your baby's vision and will liaise with nursery and school teachers to ensure that your child has the help they need to have a successful mainstream education.

Alternatives

Please be sure to speak to ask about any alternatives to your child's plan of care during their consultation.

Contacts/Further information

If you have any concerns about your child's eye, the eye becomes more red and your child stops feeding, starts vomiting, or seems in pain, ring the emergency eye clinic number on (01223) 216778 or contact your General Practitioner (GP) for advice.

Hospital Contacts:

Consultant Paediatric Ophthalmologist,
Department of Ophthalmology,
Clinic 3, Box 41,
Addenbrooke's Hospital,
Cambridge University Hospitals NHS Foundation Trust,
Hills Road, Cambridge, CB2 0QQ
Secretary: 01223 216700

Clinical Nurse Specialist Paediatric Ophthalmology,
Department of Ophthalmology,
Clinic 3, Box 41,
Addenbrooke's Hospital,
Cambridge University Hospitals NHS Foundation Trust,
Hills Road, Cambridge, CB2 0QQ
Bleep: 156 - 2251 (via Addenbrooke's Switchboard = 01223 245151)

Privacy & Dignity

We are committed to treating all patients with privacy and dignity in a safe, clean and comfortable environment. This means, with a few exceptions, we will care for you in same sex bays in wards with separate sanitary facilities for men and women.

In some areas, due to the nature of the equipment or specialist care involved, we may not be able to care for you in same sex bays. In these cases staff will always do their best to respect your privacy and dignity, eg with the use of curtains or, where possible, moving you next to a patient of the same sex. If you have any concerns, please speak to the ward sister or charge nurse.



We are currently working towards a smoke free site. Smoking is only permitted in the designated smoking areas.

For advice and support in quitting, contact your GP or the free NHS stop smoking helpline on 0800 169 0 169

Help with this leaflet:



If you would like this information in another language, large print or audio format, please ask the department to contact Patient Information: 01223 216032 or patient.information@addenbrookes.nhs.uk

Polish

Jeżeli chciałbyś uzyskać te informacje w innym języku, w dużej czcionce lub w formacie audio, poproś pracownika oddziału o kontakt z biurem Informacji Pacjenta (Patient Information) pod numerem telefonu: 01223 216032 lub pod adresem patient.information@addenbrookes.nhs.uk

Portuguese

Se precisar desta informação noutra língua, em impressão de letras grandes ou formato áudio, por favor peça ao departamento que contacte a secção de Informação aos Doentes (Patient Information) pelo telefone 01223 216032 ou através do e-mail patient.information@addenbrookes.nhs.uk

Arabic

إذا كنت تود الحصول على هذه المعلومات بلغة أخرى، بالأحرف الكبيرة أو بشكل شريط صوتي، يمكنك أن تطلب من القسم الاتصال بمعلومات المريض على الرقم: 01223216032 أو عبر البريد الإلكتروني: patient.information@addenbrookes.nhs.uk

Cantonese

如您需以另一語言版本、特大字體或錄音形式索取本資料，請要求部門聯絡病人諮詢服務：電話 01223 216032，電郵地址 patient.information@addenbrookes.nhs.uk

Turkish

Eğer bu bilgileri başka bir dilde veya büyük baskılı veya sesli olarak isterseniz, lütfen bulunduğunuz bölümdeki görevlilere söyleyin Hasta Bilgilendirme servisini arasinlar: 01223 216032 veya patient.information@addenbrookes.nhs.uk

Urdu

اگر آپ کو یہ معلومات کسی دیگر زبان میں، بڑے الفاظ میں یا آڈیو طریقہ سے درکار ہوں تو برائے مہربانی اس شعبہ سے پیشینٹ انفارمیشن سے ذیل کے ذریعہ رابطہ کریں: 01223 216032 یا patient.information@addenbrookes.nhs.uk کی درخواست کریں:

Bengali

আপনি যদি এই তথ্য অন্য কোন ভাষায়, বড় অক্ষরে বা অডিও রেকর্ডিং পোতে চান তাহলে 'প্যাশেন্ট ইনফরমেশান' এর সঙ্গে 01223 216032 নম্বরে ফোন করে বা patient.information@addenbrookes.nhs.uk ঠিকানায় ই-মেইল করে যোগাযোগ করার জন্য ডিপার্টমেন্টটিতে অনুরোধ জানান।

Document history

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