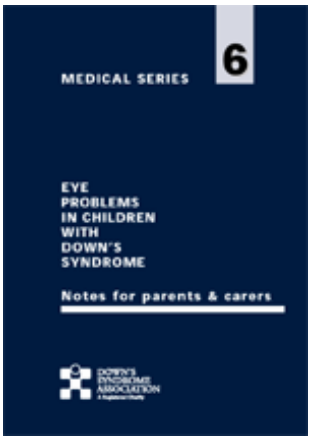

Down's Syndrome Association Medical Series

6. EYE PROBLEMS IN CHILDREN WITH DOWN'S SYNDROME

Notes for parents & carers

The eye problems which may affect children with Down's syndrome are commonly those which occur in any group of children – they just tend to occur more frequently and sometimes to a more marked degree. Health Visitors and General Practitioners will arrange for the children in their care to have their eyes checked and this is even more important if a child has Down's syndrome.

The treatment of children with Down's syndrome usually differs very little from that of other children. Because of their learning difficulties children with Down's syndrome sometimes need vision tests designed for younger children. They may find it easier to sign rather than name pictures verbally. Children with a heart condition do not usually have a problem if they need eye surgery, but advice from the cardiologist should be sought.

Common Eye Conditions

SQUINT

Around 20% of children with Down's syndrome have a squint. A squint is when the eyes are pointing in slightly different directions. Squints can be intermittent especially when they first appear; others are constant. In some cases the child alternates between squinting with the right eye and the left eye; in others, the child squints constantly with the same eye. When one eye moves out of alignment the brain receives two separate images so the one from the squinting eye is suppressed. The vision in a constantly squinting eye tends to be reduced.

Many children squint because they are long-sighted or short-sighted and consequently need glasses. Many children with a convergent squint, where one eye turns inwards towards the nose, are long-sighted (hypermetropic) and often, if such a child is given glasses to correct

this the squint can become less noticeable or even disappear completely while the child is wearing the glasses. Children who are short-sighted, or who are likely to become short-sighted when they grow older, may have a tendency for one eye to drift outwards which can be controlled.

If any child is suspected of having a squint or any visual problem, it is important to arrange referral to someone who can establish a diagnosis and arrange treatment. Usually, children see an orthoptist, who always works closely with an ophthalmologist and possibly an optometrist (see box).

PEOPLE WHO SPECIALISE IN EYE CONDITIONS

- An **ORTHOPTIST** is specially trained in the assessment of vision in people of all ages and all abilities, the recognition of squints and disorders of eye movements and the treatment of squints and related disorders.
- An **OPHTHALMOLOGIST** is a doctor specialising in eye conditions.
- An **OPTOMETRIST** is trained to test for glasses and other aspects of visual function and to recognise ocular abnormalities. They are also able to dispense glasses.
- A **DISPENSING OPTICIAN** is trained to fit spectacle frames and arrange to have these made up with the appropriate measurements and lenses for individual people.

Often it is more difficult to recognise a squint in children with Down's syndrome because of the distinctive appearance of the eyelids. For this reason, it is desirable for all children with Down's syndrome to have additional routine screening as recommended in the Down's Syndrome Medical Interest Group Health Check Guidelines (see page 7).

Treatment of Squints

Firstly, the child must be carefully examined to check the health of the eyes and to determine whether glasses are needed. This is usually done by putting drops into the eyes. These dilate and fix the pupil of the eye and make it possible to test accurately for glasses and examine the back of the eye with a light with only minimal co-operation from the child. Different drops are used in different clinics. Sometimes they may contain a substance called atropine. In the past some people were worried that atropine might have adverse effects for people with

Down's syndrome. In fact there is no evidence whatever of any harm. The only difference is that if atropine is used the pupil may stay dilated for longer than is usual.

The second aspect of treatment is to make sure that the child has equally good vision in both eyes. An experienced orthoptist will be able to establish this and treat younger children who have reduced vision in one eye. This is often done by covering the eye which can see well for a period of time each day to improve the vision in the squinting eye. The orthoptist aims to produce equal vision in both eyes so that the child is able to use either eye to fix on a test target picture.

Thirdly, if the child has a noticeable and unsightly squint, even when wearing glasses, then surgery can be arranged to correct this. At operation the position of the small muscles on the outside of the eye is adjusted so that they pull the eye into a straighter position.

LONG-SIGHTEDNESS (HYPERMETROPIA)

About 40% of pre-school children with Down's syndrome are long-sighted. This is often associated with a convergent squint. These children have to use extra effort to focus their eyes and this is more of a problem for close vision. If one eye is markedly more long-sighted than the other, the vision in this eye might be reduced (a lazy eye) and might require treatment from an orthoptist.

SHORT-SIGHTEDNESS (MYOPIA)

About 14% of pre-school children with Down's syndrome are short-sighted and the condition becomes more common up to adolescence. These children can often see near objects but have a problem with distance vision.

ASTIGMATISM

About 30% of pre-school children with Down's syndrome have astigmatism. This means that the image seen is distorted because the image is more out of focus in one direction than the other. The astigmatism can be either long-sighted or short-sighted or a mixture of the two.

FOCUSSING DIFFICULTY ('WEAK ACCOMMODATION')

Many children with Down's syndrome have difficulties focusing well on near tasks, and this applies whether they are long-sighted, short-sighted or even if they do not need glasses for general use. Furthermore, the problem with focussing persists even if the children wear their glasses.

Recent studies have suggested that children that have this problem with focusing benefit from wearing bifocals, at least in school. Some children with Down's syndrome choose to wear their bifocals all of the time, preferring them to the conventional 'single vision' glasses.*

NYSTAGMUS

About 10% of children with Down's syndrome have nystagmus. Nystagmus is a condition in which the eyes make small, involuntary, jerky movements. Often these movements are more noticeable when the child is looking sideways. Sometimes there is a position of gaze where the movements are considerably reduced. If this is the case, the child might adopt a compensatory head posture which allows the eyes to be in a position in which the nystagmus movements are minimised. If this happens the child should not be discouraged from adopting the head posture as this is likely to be the position where the vision is at its best. The vision is often better for near than distance. Children with nystagmus often prefer to hold books very close as this improves their vision and they should be allowed to do this. Any child with nystagmus should be referred to the Visual Impairment Support Service of the local Education Authority, which will provide advice at both the preschool stage and throughout school.

EYE INFECTIONS

Eye infections and watering eyes tend to be more common in people with Down's syndrome. Normally tears, which are formed continuously to keep the eyes moist and healthy, drain down the naso-lacrimal duct which connects the corner of the eye with the back of the nose. In people with Down's syndrome, this tube is often quite narrow and so it easily becomes blocked. This leads to watering of the eye and because clean tears are not rinsing through the system effectively, it is easier for infections to occur. Infections are usually treated with antibiotics given in the form of drops by day and ointment at night. If, however, the eyes are only slightly sticky and the discharge is not yellow or green, then bathing the eyes with cooled, boiled water in the morning and at night is usually sufficient treatment.

Children may grow out of this problem as the face grows bigger and the duct grows wider. If infections persist, it may be necessary to probe and syringe the tear ducts. Although this is a minor procedure, it is carried out under a general anaesthetic in young children.

As children with Down's syndrome often have rather dry skin they also tend to suffer from blepharitis. This is a condition affecting the eye lids where the skin around the eyelashes becomes flaky and inflamed. Usually the condition is mild and responds to simple measures such as bathing the lids with plain boiled water which has been cooled to a comfortable temperature. Sodium bicarbonate (a teaspoon to a pint of water) can be added. In more severe cases baby shampoo (normal shampoo would sting) can be used, in solution to clean the lid. Regular lid cleaning reduces irritation and lessens the likelihood of infections which would need to be treated with antibiotics.

Other Eye Conditions

CATARACTS

A cataract is when part or all of the lens of the eye has become cloudy. If the affected area is small it is possible to see round it, through the clear part of the lens. This kind of cataract does not cause a significant problem and is relatively common in people with Down's syndrome. A denser opacity of most of the centre of the lens is fortunately much less common as it causes a marked reduction in vision. Less than 1% of children with Down's syndrome have a dense cataract. These can be treated by removing the lens of the eye under general anaesthetic. As this leaves the eye unfocused, older people can have a lens implant at the time of the operation. Children often have an operation which makes it possible to have a lens implant inserted at a subsequent operation when the eye has reached adult size. If a lens implant is not inserted, the eye needs to be focused either by wearing thick glasses or contact lenses. Cataracts can be present at birth or develop later. They would be discovered at a routine check.

KERATOCONUS

This condition of the cornea (the clear structure covering the front of the eye) is more common in people with Down's syndrome but is still relatively rare. The cornea, instead of

being the normal curved shape, becomes conical. During the early stages this makes the person short-sighted, often with marked astigmatism making the vision distorted. Many cases do not progress any further than this stage. Other cases go on to develop scarring in the centre of the cornea. A small number of those affected develop sufficient thinning of the centre of the cornea to make them require a corneal graft. This is carried out under general anaesthetic. After a corneal graft the eye is vulnerable until it is completely healed. This can present some problems in people with Down's syndrome as the patient has to be discouraged from touching the eye.

The condition is extremely rare in childhood, may start to develop in adolescence and ultimately affects 10 – 15% of adults though for many the effects will not be serious. Although rare this is one reason why it is very important for people with Down's syndrome to have regular eye checks throughout the teenage years and beyond.

INFANTILE GLAUCOMA

This very rare condition is slightly more common in babies with Down's syndrome. Typically the eye looks larger than normal. The baby is distressed, particularly by bright lights, and the eyes may water more than usual. Urgent referral to an ophthalmologist is essential.

Recommended Schedule of Eye Checks

Fortunately, the more serious eye conditions are quite rare. However the more routine eye conditions are relatively common and can be much improved by treatment, especially if they are discovered early by vigilant screening. Therefore regular eye checks are essential for all people with Down's syndrome

As with all children cataract should be checked for at birth and at 6 weeks. From 6 weeks to 18 months the child is expected to be under the care of a paediatrician who will assess visual behaviour and the appearance of the eyes. It there are any concerns appropriate specialist referral will be made. All children with Down's syndrome should have formal ocular/visual assessment between 18 and 24 months and these formal eye checks should continue at least 2 yearly throughout life.

The Down's Syndrome Medical Interest Group (DSMIG) recommends the following.

	Birth and 6 weeks	6 weeks – 12 monthss	12 months	18-30 months	3-3½ years	4-4½ years
Eye check	Visual behaviour. Check for congenital cataract.	Visual behaviour. Check for squint.	Visual behaviour. Check for squint.	Orthoptic examination, refraction and ophthalmic examination.		Visual acuity, refraction and ophthalmic examination.

From 5 years onwards eye tests should be at least every 2 years.

Evidence based information for health care professionals about vision problems in Down's syndrome is available at

www.dsmig.org.uk

Go to 'Medical Library' and select 'Eye and Vision problems'. This section includes a useful article by Maggie Woodhouse and guidelines for professionals about dispensing bifocal spectacles.

Questions Often Asked By Parents

Q: When should a child with Down's syndrome first have an eye test and how often should this be repeated?

A: If a child starts to squint she should be referred for an eye test as soon as possible. For children with Down's syndrome without any obvious problem screening is recommended between 18 months and 2 years and again at the age of 4 years, prior to starting school. The children should have an assessment by an orthoptist, a test for glasses using drops and careful examination of the eyes.

During the school years the children should be checked regularly at least every 2 to 3 years. If any disorder is found, the children are usually seen more frequently, often once or twice a year. This assumes that the child will also have developmental checks by a paediatrician who would also refer her if a problem were suspected.

Q: How do you test vision at different ages?

A: Babies can fix and follow a light or small toy. They tend to look at more interesting things when given a choice. A series of tests have been devised which give a choice between either plain grey and a striped pattern or between plain grey and the outline of an object. The person carrying out the test has to watch where the baby chooses to look. The test cards are graded to show finer and finer stripes or fainter and fainter pictures which are shown until the baby shows no definite preference. This is noted and compared with standards expected for babies of different ages.

Toddlers are usually tested by showing them standardised black and white pictures held a measured distance away (Kay pictures) and asking them to name or match them. Some children with Down's syndrome prefer to sign to identify them rather than say what the pictures show.

Many children with Down's syndrome aged 3-4 years can be tested by showing that they can match letters. The child is given a card with five letters on it. The examiner shows a series of cards with letters of graduated sizes and the child points to a matching letter on the card he is holding. This test is carried out at an exact distance (3 or 6 metres). Each eye can be tested separately by covering one eye at a time. Slightly older children can use the matching card to identify letters on a line on the normal test type and this is even more accurate.

Q: How is it possible to determine how strong glasses need to be in a young child?

A: Small children are usually tested after drops have been put in the eyes. The drops relax the focusing of the eyes. The examiner is then able to shine a beam of light from a torch called a retinoscope across the child's eyes while holding a lens in front of the eye. The strength of lens needed to neutralise the movement of the light indicates the strength of lens required in the glasses. Most small children do not mind looking at the light for the short time although it takes an experienced person to test them.

Q. How is it possible to determine whether my child's near focusing is good?

A. Accommodation, or focusing at near, can be tested in the same way, with a retinoscope. This time the child needs to look at detailed pictures at near, and for this test drops are not used, as the eye must be in its natural state and able to change focus.

Q: If my child needs glasses will it be difficult to persuade him to wear them? How can I help him get used to them?

A: It can be difficult at the beginning. First of all it is vital that the glasses fit comfortably because a child will try to remove them if they are digging into the face or the back of the ear. It is a good idea to put on the glasses and immediately start an activity that the child enjoys such as looking at books or playing activity games. Television and videos are only helpful if an adult is sitting with the child to prevent him from removing the glasses. The child will associate the glasses with enjoyable activities and if he is sufficiently distracted will forget he has them on. If a parent has some glasses it sometimes helps if they wear them more of the time. Nursery school teachers and play-group leaders can be very useful allies. If the child takes the glasses of at the end of an activity, allow him to do so at first. Above all, don't turn it into a battle, because the child will quickly learn what fun it is to upset parents and teachers by throwing his glasses! Once the child has got used to the glasses they often become keen wearers when they realise they can see more clearly.

Q: Do I have to pay for my child's glasses or can I get them on the NHS?

A: All children (under 19 in full-time education) and older people who are either unemployed or have a low income are entitled to a voucher, which is meant to cover the price of a basic pair of glasses. The value of the voucher increases with stronger lenses. No additional help is given to single or unemployed parents.

It is sometimes necessary to pay extra money, in addition to the voucher price, to get glasses that fit well and are comfortable. You may, of course, wish to pay extra for a 'designer' frame, but remember that your child is very likely to lose glasses or break them and will certainly grow out of them fairly quickly.

Q: Can my child have two pairs of glasses - my child is always breaking or losing his glasses.

A. Ordinarily, the NHS only allows one pair of glasses to be supplied through the voucher scheme at each eye test (unless a child's eye condition means that two different prescriptions are required). However, a spare pair is permitted in individual cases if the prescription is significant (so that the child is handicapped when glasses are broken) and if the child's individual circumstances (e.g. their learning disability) mean that breakages are unavoidable. The optometrist or optician will need to apply to the Local Health Board for consent to issue a second voucher, and each case is considered on merit. You can, of course, pay privately for a spare pair. The cost of repairs can be covered by a repair voucher issued by your optician.

Q: Is it right to take my child to a high street optician or should she see a specialist?

A: It is desirable for all small children to attend a hospital clinic where they will be able to see a team of people - orthoptist, ophthalmologist and optometrist and possibly a specialised dispensing service. Once the child has been fully evaluated and any problems treated they are often only kept in the hospital service until they are mature enough to be discharged to outside optometrists/opticians. The service given by these opticians depends not so much on the name – often they are one of a chain – but on the care and expertise of the individuals working there. It is worth following recommendations given by parents of small children and of children with special needs.

Accessible resources for people with learning disabilities and visual problems can be obtained from

The RNIB Multiple Disability Team

RNIB

224 Great Portland Street

London W1N 6AA

Telephone: 020 7388 1266

Fax: 020 7388 2034

Barbara Crofts. Associate Specialist in Ophthalmology. Oxford

Revised March 2005 in conjunction with **Maggie Woodhouse** – Senior Lecturer Optometry and Vision Sciences, Cardiff and **Jennifer Dennis**, Paediatrician, DSMIG.