



**Down's Syndrome
Association**

A Registered Charity No. 1061474

Health Series



Eye conditions in children



The
Down Syndrome
Medical Interest
Group

A Down's Syndrome Association publication

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The Down's Syndrome Medical Interest Group (DSMIG)

This resource has been produced in collaboration with the Down's Syndrome Medical Interest Group (DSMIG).

DSMIG was launched in 1996 and is a registered charity. It is a network of healthcare professionals – mainly doctors – from the UK and Republic of Ireland whose aim is to share and disseminate information about the medical aspects of Down syndrome and to promote interest in the specialist management of the syndrome.

The eye conditions 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, General Practitioners or a Paediatrician 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 disability 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.

We have produced a separate resource about children, vision, eye tests and glasses which can be downloaded from the health section of our website (under 'vision').

Common eye conditions

Squint

Around 20% of children with Down's syndrome have a squint. A squint means that 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, in which one eye turns inwards towards the nose, are long-sighted (hypermetropic) and often, if given glasses to correct this, the squint can become less noticeable or even disappear completely while the child is wearing the glasses.

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.

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).

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 eye 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.

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 whatsoever of any harm caused. 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 try to ensure that the child has equally good vision in both eyes. An experienced orthoptist will be able to measure the vision in each eye 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 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 may be 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.

Focusing 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 focusing persists even when the children are wearing their glasses.

Recent studies have suggested that children who 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 in which the nystagmus movements are considerably reduced. If this is the case, the child may adopt a compensatory head posture, which allows the eyes to be in this 'null' position. 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. Seating in school should be arranged so that the child can easily use their null position. Vision is often better for near rather 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 Authority, which will provide advice at both the pre-school stage and throughout school.

Eye infections

Blocked tear ducts

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.

Blepharitis

As children with Down's syndrome often have rather dry skin they also tend to suffer from blepharitis. This is a condition affecting the eyelids in which 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 that 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 occurs when part or all of the lens of the eye has become cloudy. Cataracts can be present at birth or develop later. They would be discovered at a routine baby check either in hospital or by your GP. 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 covering most of the centre of the lens is much less common and causes a marked reduction in vision. Fewer than 1% of children with Down's syndrome have a dense cataract. It 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 that 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. Contact lenses can be successfully worn by people with Down's syndrome.

Keratoconus

This condition of the cornea (the clear structure covering the front of the eye) is more common in people with Down's syndrome. The cornea, instead of being the normal curved shape, becomes conical. 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. During the early stages keratoconus makes vision distorted, but as many young people with Down's syndrome are unable to report this, it can go unnoticed.

Many cases do not progress any further than this stage, but other cases progress and if not treated, vision can be badly affected. There is a new treatment called 'cross-linkage' becoming available, although patients may currently have to travel to specialist centres for treatment through the NHS. Cross-linkage is available privately throughout the UK. The treatment stops progression of keratoconus and maintains good vision. However, it is only feasible in the early stages of the condition, so early diagnosis is crucial. This is why it is very important for people with Down's syndrome to have regular eye checks throughout the teenage years and beyond. Further, research has shown that retinoscopy (used to test for glasses) is the most reliable method of detecting early keratoconus, but some optical practices have moved away from retinoscopy in favour of automatic instruments, and these are unlikely to pick up keratoconus. If a child has been discharged from the hospital service, it is essential for parents to find an optometrist who is very experienced with retinoscopy, as well as, of course, with working with people with learning disabilities. If keratoconus is diagnosed, then the first step is to monitor, and if the condition progresses, a referral to a corneal specialist should be made without further delay.

If keratoconus is not progressing, or treatment is not feasible, then keratoconus can be managed very successfully with contact lenses.

Untreated cases may go on to develop scarring in the centre of the cornea and a small number of those affected develop sufficient thinning of the centre of the cornea to require a corneal graft.

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 health 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, cataracts 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. If there are any concerns appropriate specialist referral will be made.

All children with Down's syndrome should have formal ocular/visual assessment (at a hospital eye clinic or specialist optometrist practice) between 18 and 24 months and these formal eye checks should continue at least every 2 years throughout life.

Age Check

- Birth - 6 weeks Visual behaviour. Check for congenital cataract.
- 6 weeks - 12 months Visual behaviour. Check for squint.
- 12 months Visual behaviour. Check for squint.
- 18 - 30 months Orthoptic examination, refraction and ophthalmic examination.
- 4 - 4 1/2 years 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 <https://www.dsmig.org.uk/information-resources/by-topic/vision/>

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 they 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 years. If any disorder is found, the children are usually seen more frequently, often once or twice a year. This recommendation assumes that the child will also have developmental checks by a paediatrician who would also refer them 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 watches where the baby chooses to look. The test cards are graded to show finer and finer stripes or pictures that 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 examiner shows a series of cards with letters of graduated sizes at a measured distance and the child points to a matching letter or symbol on the card they are holding. Whichever test is used; each eye can be tested separately by covering one eye at a time.

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

A: Small children are often 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 and watch the light reflected back from the eye. The strength of lens needed to focus the reflected 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. It is possible to test children without drops, but greater experience is required on the part of the optometrist. As children get older, the need for drops reduces.

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 them to wear them? How can I help my child 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 them 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 playgroup leaders can be very useful allies. If the child takes the glasses off at the end of an activity, allow them 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 their glasses!

If a child has a squint, especially one that is straightened by glasses, the aim will be for the child to wear their glasses all of the time. If there is no squint, then there is no need for full-time wear, unless the child chooses that.

Once the child has got used to the glasses they often become keen wearers when they realise they can see more clearly, and if you encourage wear for important activities like learning, you can allow the child to decide whether they want to wear them more often.

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 their 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 unable to see 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 their local Primary Care Organisation 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 and replacements can be covered by a repair voucher issued by your optician.

Q: Is it alright to take my child to a high street optician or should they 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 specialized dispensing service (although there are some specialised optometrists working in the community who have the skills to see small children). If children attend the hospital, once they have 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.

Annual health checks for people with Down's syndrome (aged 14 years plus)

In the past people with learning disabilities have not had equal access to healthcare compared to the general population. This, amongst other reasons, has given rise to poorer mental and physical health and a lower life expectancy for people with learning disabilities. Free annual health checks for adults with learning disabilities, with their GP, were introduced in 2008 as a way to improve people's quality of life.

The annual health check for people with learning disabilities is a Directed Enhanced Service (DES). This is a special service or activity provided by GP practices that has been negotiated nationally. Practices can choose whether or not to provide this service. The Learning Disability DES was introduced to improve healthcare and provide annual health checks for adults on the local authority learning disability register. To participate in this DES, staff from the GP practice need to attend a multi-professional education session run by their local Trust. The GP practice is then paid a sum of money for every annual health check undertaken.

Who can have one?

Annual health checks have been extended to include anyone with learning disabilities aged 14 years or above. So anyone with Down's syndrome aged 14 years or over can have an annual health check.

The benefits of annual health checks

- additional support to get the right healthcare
- increased chance of detecting unmet, unrecognised and potentially treatable health conditions
- action can be taken to address these health needs.

How to get an annual health check

- The GP may get in touch with the person with Down's syndrome to offer an annual health check but this doesn't always happen.
- A person with Down's syndrome and/or a supporter can ask their GP for an annual health check. You do not need to be known to social services to ask for an annual health check.

Not all GPs do annual health checks for people with learning disabilities but they should be able to provide details of other GPs in your area who offer this service.

What happens next?

- The GP practice may send out a pre-check questionnaire to be filled out before the annual health check takes place.
- The GP may arrange for the person with Down's syndrome to have a routine blood test a week or so before the annual health check.

Who attends the annual health check?

If the person with Down's syndrome (age 16 years or over) has capacity and gives their consent, a parent or supporter can attend the health check as well.

How long should an annual health check be?

Guidance from the Royal College of GPs suggests half an hour with your GP and half an hour with the Practice nurse.

What areas of health should be looked at as part of the annual health check?

We have produced a check list for GPs which contains information about what should be included as part of a comprehensive and thorough annual health check. This includes a list of checks that everyone with a learning disability should undergo as part of an annual health check and a list of checks specific to people with Down's syndrome. You can find the health check list at the 'annual health checks' section of our website under 'families and carers and 'health and wellbeing'.

What happens after the annual health check?

Your GP should tell you what they and the nurse have found during the annual health check. You should have a chance to ask any questions you have. Your GP may refer you to specialist services for further tests as appropriate. Your GP should use what they have found during your annual health check to produce a health action plan. This should set out the key actions agreed with you and (where applicable) your parent or carer during the annual health check. Your GP has to do this as part of the annual health check service.

Information about health issues for GPs

There is information at our website for GPs about some of the more common health conditions seen in people with Down's syndrome. You will find this information at the 'annual health checks' section of our website under 'families and carers and 'health and wellbeing'.

GPs learning disability register

People with learning disabilities experience poorer health compared to the rest of the population, but some ill health is preventable. Over one million people in the UK have a learning disability but only 200,000 are on their GPs learning disability register.

We know that people with a learning disability often have difficulties accessing health services and face inequalities in the service they receive. The Government is asking parents and supporters to speak to their GP and ensure their sons/daughters or the people whom they support are registered. It is hoped that this drive will ensure better and more person centered health care for people with learning disabilities.

The Learning Disability Register is a record of people with a learning disability who are registered with each GP practice. The Register is sometimes referred to as the Quality Outcomes Framework (QOF) Register.

If you are not sure you are on the Register, you can ask the receptionist at your GP Practice to check for you.

The doctor may have made a note on the record that a person has Down's syndrome but this does not automatically mean they have been put on the Register. When you speak to the GP about being registered, the needs and support of the person in health settings can be discussed. This information can be entered on the person's Summary Care Record (SCR) so that all health professionals at the practice know about their needs and how best to support them.

If the person is over 16 years of age or older, they must give their consent (see section in this resource about the Mental Capacity Act 2005):

- for information about their support needs to be added to their SCR
- to which information can be shared and with whom

It's never too early (or late) to join your GP's Learning Disability Register; you can join at any age. It's a good idea for children with a learning disability to join the learning disability register at an early age. This means adjustments and support can be put in place before they reach adult services.

Reasonable adjustments in health care

You may have heard of the term 'reasonable adjustments' and wondered what it meant. Since the Disability Discrimination Act (1995) and the Equality Act (2010) (this does not apply to Northern Ireland) public services are required by law to make reasonable adjustments to help remove barriers faced by people with disabilities when trying to use a service. The duty under the Equality Act to make reasonable adjustments applies if you are placed at a substantial disadvantage because of your disability compared to people without a disability or who don't have the same disability as you.

So for people with physical disabilities reasonable adjustments may include changes to the environment like ramps for the ease of wheelchair users. For people with learning disabilities 'reasonable adjustments' may include easy read information, longer appointments, clearer signs at the practice, help to make decisions, changes to policies, procedures and staff training.

If a patient with Down's syndrome is NOT on their GP's Learning Disability Register then reasonable adjustments to care for that person cannot be anticipated and made.

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The Down's Syndrome Association provides information and support on all aspects of living with Down's syndrome.

We also work to champion the rights of people with Down's syndrome, by campaigning for change and challenging discrimination.

A wide range of Down's Syndrome Association publications can be downloaded free of charge from our website.

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