

Quality Standards



Vision care for deaf children and young people

Guidelines for professionals

What parents have told us...

“We were impressed that they treated her as a child first, not just as a deaf child. They accommodated her needs and did various things to get her confidence, so they could get a good quality eye test done.”

“The initial diagnosis – although a shock to us as parents – was followed up with care and due information and explanation.”

“Families have the primary care and responsibility for their child. So it is vital to involve them from the outset as partners with health professionals in the care of their child – rather than as recipients of a service.”

Driving up vision care standards for deaf children

Several changes have taken place affecting deaf children since the publication of the first guidelines on vision care for all deaf children by Sense and NDCS in 2004. This new edition takes recent developments into account. Newborn Hearing Screening has been implemented throughout the UK. Babies born with permanent deafness may be identified in the first few weeks of life – which means that families can make earlier decisions on how their child can develop language skills. Developments in the cochlear implantation programme (and service, too) have seen many more parents choosing this option for their deaf child since 2004.

Most of the information a child will use to learn about their world comes through their ears and eyes. Deaf children are especially dependent on vision in order to acquire language and develop relationships. The role of vision and consequently the care of vision are crucial for deaf children if they are to fulfil their potential. These guidelines stress the importance of an ophthalmic examination following confirmation of deafness. A good vision assessment of a deaf child will require extra preparation and an understanding of the child's particular needs. It is vital that vision difficulties in a deaf child are picked up as soon as possible so that suitable treatment can be offered and, if appropriate, support provided to manage any dual sensory impairment.

This publication provides a valuable tool and reference point for all service providers and funders of services. It builds on what has been gained from good practice from professionals and in doing so aims to drive up standards in vision care for deaf children and young people throughout the UK.

We are proud that these guidelines have been developed through a strong partnership between NDCS and Sense. Both our organisations have long-standing expertise in issues relating to deafness and vision, especially in delivering services, and in the care of children, young people and their families.

We would like to thank everyone involved in the revision of *Vision care for deaf children and young people*. These guidelines were developed with help from ophthalmology, audiology and community professionals, parents and many others involved with deaf children.



Richard Brook
Chief Executive Sense



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Part one

Introduction

We hope that this document helps everyone towards ensuring that deaf children and young people fulfil their true potential.

Most of the information children use to learn about the world around them comes from what they see and hear. In the case of deaf children, vision plays a more significant role as the gateway for acquiring language and developing social cognition (Nikolopoulos et al. 2006). It is important to ensure there is an effective clinical pathway to enable recognition of any visual problem as early as possible in the deaf child's life and provision of the best vision care possible. Hall and Elliman (2006) state that "Forty per cent of children with sensorineural hearing impairments have eye problems, some very severe. All children with sensorineural hearing problems should undergo a specialist eye examination".

This document highlights the importance of an ophthalmic examination following confirmation of deafness, and that good vision assessment of a deaf child will require extra preparation and an understanding of the individual child's needs. The aim of this document is to enable professionals to review the ophthalmic care of deaf children and, where appropriate, develop services to meet the needs of deaf children, deaf young people, and their families.

The Royal College of Ophthalmologists (2005) states that "A child's health encompasses their physical, emotional and social development and visual loss in childhood can significantly impair progress in any of these dimensions." Children with a sensorineural deafness are considered a high-risk group, and clinical surveillance is recommended (Royal College of Ophthalmologists 2005). Vision care for deaf children is especially important, as deaf children use their vision to access and learn about the world around them.

These guidelines update the 2004 publication, *Vision care for deaf children and young people: guidelines for professionals working with deaf children*, and have again been produced by NDCS and Sense in close cooperation with professionals who work with and for deaf children, to provide guidance on how the vision of deaf children should be assessed and managed. The document identifies the optimal stages in a deaf child's development for assessments to be carried out, who should carry them out, and the type of assessments needed.

Aiming high for disabled children states that "The Government wants all children to have the best start in life and the ongoing support that they and their families need to fulfil their potential." (Department for Education and Schools (DfES) 2007). We hope that this document helps everyone towards ensuring that deaf children and young people fulfil their true potential.



The quality standards

Below is a list of the quality standards presented throughout this publication:

Quality standard 1

Information given to children, young people and parents must be accurate, valid, relevant, up to date, timely, understandable and developmentally, ethically and culturally appropriate. (Department of Health (DH) and Department for Education and Schools (DfES), 2004a).

Quality standard 2

All services must develop local written policies on working with families with a deaf child. It is essential to involve parents of deaf children in developing and reviewing these policies (DH and DfES, 2004b).

Quality standard 3

Every member of the multidisciplinary team must be aware of the importance of ophthalmic care for deaf children and the issues relating to dual sensory impairment.

Quality standard 4

Each child must have an identified lead clinician who coordinates the care and management of the child, and each child and family must be assigned a key worker as soon as the child's deafness has been identified.

Quality standard 5

Staff working with deaf children should receive training in deaf awareness, including issues relating to childhood deafness.

Quality standard 6

A deaf child or young person should be referred for ophthalmic assessment at any time if parents or the education service have concerns (Hall and Elliman, 2006).

Quality standard 7

The eye clinic should be informed of the family's communication preferences in advance so that suitable provision can be organised for appointments (DH and DfES, 2004a).

Quality standard 8

Written information about the visit to the clinic should be given to the family before the clinic appointment, and must be suitably worded for the family and in a format that the older child or young person can understand if appropriate.

Quality standard 9

"Staff working in the paediatric eye clinic should have a common core of skills, competencies and knowledge" as identified by the Royal College of Ophthalmologists, 2005.

"The initial diagnosis – although a shock to us as parents – was followed up with care and due information and explanation"

Parent

The quality standards

Quality standard 10

Following the diagnosis of permanent conductive or sensorineural deafness, or if a child needs hearing aids, the child should be referred for a full ophthalmic assessment, and this should be repeated where appropriate at key stages of their development.

Quality standard 11

All staff involved in the diagnosis of deafness must be aware of when and to whom to refer a deaf child. All staff should be aware of local protocol for referral to the ophthalmologist.

Quality standard 12

A report of the outcome of the visit should be sent to the parent, the referrer and the child's GP, and to all other key professionals involved. This includes copies of assessments and diagnostic results. Parental consent is needed before information is shared within the multidisciplinary team.

Quality standard 13

Ideally a follow-up appointment within two weeks should be offered for children who are priorities (for example, if they have been diagnosed with a serious eye condition) so that the parents and child can discuss further questions. The family may want to involve their key worker in appointments. The family should be provided with contact details for the vision team or the local authority visual impairment teaching service.

Quality standard 14

The clinician carrying out the assessment should be given specific information on the deaf child's additional support needs before the assessment.

Quality standard 15

If a child's vision impairment or dual sensory impairment is unexplained or suspected to have a genetic cause, the lead clinician is responsible for ensuring that a genetic referral is offered to the family and, if accepted, arranged.

Quality standard 16

To ensure continuity of care, all relevant information must be passed to the adult ophthalmology service (with copies sent to the GP) when the young person transfers from the paediatric service.

Quality standard 17

Before transfer to alternative services, the appropriate professional should arrange all relevant appointments with the new service and make sure that the young person and their family are notified.

Definitions and key words

Blindness: is defined as a visual acuity of less than 1.3 LogMAR or 3/60 (Snellen chart) in the better eye with refractive correction and/or where the person is so severely vision impaired that they cannot carry out any work for which eyesight is essential. As with partial sight, the extent of visual field contributes to certification.

Deaf: In this publication we use the term 'deaf' to mean all levels of permanent deafness, but not temporary deafness such as glue ear. The phrase 'all deaf children' includes children with complex needs.

Deafblind: sometimes known as dual sensory impairment or multi-sensory impairment. In addition to hearing and sight loss, deafblindness can cause difficulties with communication and mobility.

Partial vision or partial sight: is defined as a visual acuity of between 0.6 (LogMAR) or 6/24 (6/18 with large part of field missing) (Snellen chart) or 1.00 (LogMAR) or 6/60 (3/60 if full field) (Snellen chart), and where the person is substantially and permanently affected by defective vision caused by congenital defect, illness or injury. However, there is some flexibility and it is influenced by the extent of visual field.

Snellen: A clinical measurement of functional vision obtained by reading or matching letters on a chart. Vision is recorded as a fraction, eg 6/6 = normal vision, 6/60 = the person has to be 6 metres away in order to see what someone with normal vision could see at 60 metres (SERSEN, 2006).

Vision impairment or sight impairment: refers to any problem with vision from the very subtle to blindness. For certification purposes there are two categories of severe vision impairment, partial vision and blindness (see above).

Visual acuity: is a measure of how well a person is able to distinguish fine detail, such as the features of a letter, colour vision, and contrast sensitivity.

We use the words '**parent**' and '**family**' to include carers who have parental responsibility. If the child is a ward of court, or in the care of social services, appropriate permission must be sought before any medical or surgical procedure takes place.

Deaf children in the UK

More than two children a day are born in the UK with significant permanent deafness. A study by Fortnum et al. (2001) estimated that a further 0.6 to 0.9 per 1000 children will develop deafness by ten years of age (due to illness, progressive deafness or late onset deafness). Around 40% of deaf children will have additional needs (Davis et al., 1997) and/or ophthalmic problems (Guy et al., 2003).

The majority (92%) of deaf children will be born to hearing parents (Rawlings and Jensema, 1977; Schein and Delk, 1974). Fifty per cent of deafness in children has a genetic cause. The other half include causes such as prematurity, congenital infections, ototoxic medications and childhood infections (NDCS, 2008).

‘Vision provides the most effective stimulus for development in all areas and opens up the pathways to learning, especially during the first years of life. Vision and the development of communication are closely linked’ (Laffan, 1993).

Population studies over several decades have shown that there is a higher incidence of ophthalmic problems amongst deaf children. These higher rates of ocular pathology have been attributed to:

- important elements of the eye and ear (eg retina and cochlea) maturing during the same embryological stage, from the same embryonic layer, which may be susceptible to genetic or environmental factors, and
- environmental insults, such as hypoxia, toxic agents, viruses, meningitis, and other conditions which may affect both the eye and the ear.



For conditions that affect both hearing and vision see the ‘Conditions affecting both hearing and vision’ appendix 6 on page 47.

If a deaf child has a problem with their vision it is essential that it is identified and treated as soon as possible. Those identified with ocular abnormalities will require long-term ophthalmological review. Brinks et al. (2001) found that this is often overlooked. They reported that 63% of deaf students aged 10–22 had significant ocular pathology and had not seen an ophthalmologist in the previous two years.

For the majority of deaf children, when an ophthalmic problem is identified, it is unlikely to be of a serious nature. In Guy’s study (2003) of the 75 ocular abnormalities identified, 47 (60%) were refractive errors, for example astigmatism, myopia and hypermetropia which are usually correctable.

Where there are more serious eye conditions, this can create enormous challenges for the individual particularly in relation to communication, accessing information, mobility and social inclusion. However, with appropriate early assessment and support, children can be helped to fulfil their potential. Some local authorities employ specialist teachers to work with children and young people with multi-sensory impairment (MSI) as part of their sensory support team. You can find out more about MSI teachers in the ‘Key personnel’ appendix 2 on page 40.

Glue ear (otitis media with effusion)

Glue ear is one of the most common childhood conditions. In most cases glue ear will be short term and should not require additional assessment of vision. However, whilst glue ear is most common in children under five, and is usually temporary, it can persist into adolescence. Additionally, certain groups of children are likely to have persistent glue ear throughout childhood, for example those with Down’s syndrome or cleft palate. Persistent glue ear can have a significant effect on hearing, and children and young people who suffer from it should have their vision assessed. Children with glue ear may not have any amplification or support and therefore will place greater reliance on their vision. As these children are not at higher risk of an ophthalmic problem, referral to the local optometrist may be the most appropriate and effective route to prompt assessment of their visual acuity.

Glue ear is one of the most common childhood conditions.

Newborn hearing screening

All newborn babies in the UK should now be offered a hearing screen during the first few weeks of life. If a baby does not show a clear response to the screen they will be referred for a further screen, or for audiological assessment. Babies born with a permanent deafness may therefore be identified by eight weeks of age, although sometimes it may take longer to confirm the deafness.

All newborn babies in the UK should now be offered a hearing screen during the first few weeks of life.

Newborn hearing screening makes it possible for families to take very early action on behalf of their child and to intervene before language and communication deficit has become established. The NDCS website www.ndcs.org.uk has more information on newborn hearing screening, and links to the screening programmes in each part of the UK.

Types and degrees of deafness

Children may be deaf in one ear (unilateral deafness) or both ears (bilateral deafness). Types of deafness include:

- **Sensorineural (or nerve) deafness** is when there is a fault in the inner ear (most often because the hair cells in the cochlea are not working properly) or in the auditory (hearing) nerve. Sensorineural deafness is permanent.
- **Conductive deafness** is when sound cannot pass efficiently through the outer and middle ear to the cochlea and auditory nerve. This is most often temporary but can be permanent.
- **Mixed deafness** is when children who have a sensorineural deafness also have a conductive deafness, such as glue ear.

The following terms are used to define the degree of deafness. Decibel (dB) is a unit of measurement used to record the threshold of hearing (or the quietest sounds that a child can hear). This is a general guide only, and does not describe the impact of the deafness on the child.

- **Typical range (0–20dB)**
All speech sounds should be heard.
- **Mild deafness (21–40dB)**
A person with a mild level of deafness would hear a baby crying or music from a stereo but may be unable to hear whispered conversation.
- **Moderate deafness (41–70dB)**
A person with a moderate level of deafness may hear a dog barking or telephone ringing but may be unable to hear a baby crying.
- **Severe deafness (71–95dB)**
A person with a severe level of deafness may hear a chainsaw or drums being played but may be unable to hear a piano or a dog barking.
- **Profound deafness (>95 dB)**
A person with a profound level of deafness may hear a large lorry or an aeroplane, but may be unable to hear a telephone ringing.

Some children's deafness may fall into two categories; for example, moderate to severe. It is also important to remember that deaf children with the same level of deafness may experience sounds differently. The NDCS publication *Understanding Deafness* (2007, revised 2008) is useful for those who would like to find out more about childhood deafness. You can find details of this publication in the 'References' appendix 9 on page 56.



Key research findings

NDCS and Sense commissioned an evidence-based overview of ophthalmic disorders in deaf children. This was published in February 2006 as a supplement of *Otology and Neurotology* titled *Evidence-Based Overview of Ophthalmic Disorders in Deaf Children: A Literature Update* by Nikolopoulos et al. Over 1000 relevant research papers were identified, of which 191 were analysed in detail.

The notion that when a person is found to be deaf, vision impairment should be suspected is not new. Recently, a study by Bulmer et al. (unpublished, personal communication) on the aetiology of deafness carried out in Wolverhampton between 1995 and 2005 found refractive errors or ophthalmic abnormalities in 42% of deaf children sent for ophthalmic assessment. Studies as far back as 1933 on different populations of deaf children showed a high level of vision impairment. However, differences in definitions and classifications of vision impairment made comparison between these various studies difficult. Nevertheless, 40% to 60% of deaf children with ophthalmic problems has been a consistent finding. Here is a summary of some key studies identified:

- Guy et al. (2003) in Southern Derbyshire found that, out of 110 children who underwent ophthalmic assessment, 48 had ophthalmic abnormalities. Of the 82 children with uncomplicated deafness, 26 had refractive errors.
- A study by Brinks et al. (2001) found significant ocular pathology in 111 (48%) out of 231 young deaf people. They concluded that deaf students had a high frequency of eye disease, that early diagnosis and treatment could benefit their quality of life, and that the potential benefits of ophthalmologic screening in deaf children is enormous.
- Rogers et al. (1988) screened 360 deaf children and young people aged between 6 and 22 years for vision problems. Overall, 43% either had a significant refractive error or other eye abnormalities. They concluded that a deaf child is at greater risk of vision problems than a hearing child, and that a comprehensive examination should be performed at confirmation of deafness.
- Regenbogen and Godel (1985), in a comparative analysis with the study by Coleman (1970) on 3,623 hearing children, showed that more than two and a half times as many deaf children had eye abnormalities.
- Alexander (1973) found that out of 572 children, 35% had refractive errors and 22% had other ocular pathology.
- Stockwell (1952), in a study of 960 deaf children, reported that 46% needed glasses.
- Braly (1938) reported that 38% out of 422 deaf children had less than 20/20 vision on the Snellen chart.
- Burdge (1933) found that 40% of deaf children tested needed glasses.

Part two

Empowering parents, deaf children and deaf young people

Information for parents, deaf children and deaf young people

Quality standard 1

Information given to children, young people and parents must be accurate, valid, relevant, up to date, timely, understandable and developmentally, ethically and culturally appropriate. (Department of Health and DfES, 2004a).

Parents play a central role in the care of their deaf child. It is important that parents perceive vision care as a process that can have positive outcomes and not solely as a defect detecting exercise. Without causing them undue anxiety, parents need to be made aware of the importance of good vision care and ongoing monitoring as their child develops. They should be encouraged to report any concerns about their child's vision to the lead clinician. For more information on the role of a lead clinician, see the 'Key personnel' at appendix 2 page 40. Parents have a right to information so that they can make informed choices for their deaf child (NDCS and Sense, 2006, Vision 2020 UK, 2008):

“Informed Choice means that families can make knowledgeable decisions, which reflect their own culture, values and views. It is based on access to comprehensive, unbiased and evidence-based information, about the full range of options.” (Early Support, 2006)

Information for deaf children, young people and their families must be relevant, current, suitable for the intended audience, and accessible – for example, available in writing or verbally – (Early Support, 2006, Department of Health and DfES, 2004a, and NDCS, 2005a, Vision 2020 UK, 2008). *Aiming high for disabled children* states that “disabled children and their families [should be] able to access appropriate information at every stage of a child's life” (DfES 2007). Information should also be available in other languages and formats such as large print, audio or video, or Braille.

Professionals should continually question and evaluate their communication strategies to ensure that the information being shared is understood. In an NDCS and Sense survey of parents of deaf children conducted in 2001, 88% were satisfied with the information that eye care staff gave them. However, half the parents questioned did not know whether the audiology clinic had been informed of the results of the eye examination. Furthermore, one in three parents felt that some eye care staff were unaware of or insensitive to the communication needs of their deaf child.

It is important that parents perceive vision care as a process that can have positive outcomes

Young deaf people need to be fully informed so that they understand the importance of optimal vision and can take responsibility for their own vision care and know how to obtain an ophthalmic opinion, especially when they leave school. The lead clinician should ensure that young people are given the contact details of any key personnel, such as the social worker with deaf people, or youth worker, so that they can contact them for advice if their vision impairment causes practical problems in their social and educational development. You can read more about the transition from child to adult services in part six ‘Young people and transfer of care to another service’ on page 36.

Parents have a right to immediate access to information about their child’s condition as a means of informed choice and helping them accept the diagnosis. The names of relevant agencies and support groups should be provided to the child and family (Department for Education, Northern Ireland, 1996).

Multidisciplinary working

Quality standards

Quality standard 2

All services must develop local written policies on working with families with a deaf child. It is essential to involve parents of deaf children in developing and reviewing these policies (Department of Health and DfES, 2004b).

Quality standard 3

Every member of the multidisciplinary team must be aware of the importance of ophthalmic care for deaf children and the issues relating to dual sensory impairment.

Quality standard 4

Each child must have an identified lead clinician who coordinates the care and management of the child, and each child and family must be assigned a key worker as soon as the child’s deafness has been identified.

In order for parents to remain in control and to make effective and informed decisions about their child’s care, two-way communication with professionals is crucial. At the time of assessing a child’s hearing, any doubt about the diagnosis or condition should be conveyed clearly and sensitively to the parents or the young person and they should be informed when a more accurate diagnosis will be made. If a second opinion is requested, this should be respected and made available (General Medical Council, 2006).

Deaf children, young people, and their families should be consulted when developing or reviewing services and policies. This should be done in a meaningful way, ensuring that all communication requirements are catered for.

Many parents “value the opportunity to be part of the multi-agency team as an equal member” (Early Support, 2006). Including parents as part of the team helps the team to have a more holistic view of the child and family.

“Give children, young people and their parents increased information, power and choice over the support and treatment they receive, and involve them in planning their care and services.”

(Department of Health and DfES, 2004a).



“Professionals should share information about an individual child to allow other services to fulfil their duties”

Hall and Elliman, 2006

All of the professionals working with a deaf child should be aware of all the other professionals working with the child, and their roles and responsibilities. There should be close cooperation between the various professionals:

“Health professionals should share information about an individual child to allow other services to fulfil their duties and to facilitate smooth inter-agency working” (Hall and Elliman, 2006).

Professionals should also be in regular contact with the child, young person and their families, but in a managed way, so that the family is not overwhelmed with different messages and plans. Multidisciplinary working should include “pooling and sharing of data for planning

purposes and for the delivery of care and support to individuals” (Department of Health, 2008), and consent for this sharing of information must be gained from families. Clear communication between agencies should reduce the number of meetings and appointments that parents have to attend; for example joint visits could be arranged (Department of Health and DfES, 2004a):

“Where families have coordinated support, through joint planning, commissioning, assessment and provision, or through a key worker or lead professional, disabled children and their families are more likely to benefit from better coordinated support and accessible, knowledgeable professionals in both universal and specialist services.” (DfES, 2007)

With regard to vision care, each professional should be informed of the possible signs or symptoms and the significance of developing eye problems. They should be aware of the referral procedure and know who to refer to if there is concern about a deaf child’s vision, and be aware of issues relating to a dual or multiple sensory impairment (Vision 2020 UK, 2008). See the ‘Checklist for vision problems’ appendix 3 on page 43 for information on the signs that there may be problems with a child’s vision.

Exchange visits and interaction between ophthalmology and audiology staff would lead to a better understanding of each other’s working practices (for example, test procedures and technology used), enabling both disciplines to share knowledge and improve services to families.

“Working more closely together has allowed us to understand better how different members of the team can provide coordinated and focused care. We have a much better understanding of what each member of the team is doing and the sort of information that each requires. Communication has improved. Patients and their families can be better informed about what happens in the eye clinic before they arrive. Issues such as the appropriate timing of an ERG to investigate Usher syndrome can be discussed by several members of the team.”

Dr Nicky Bulmer, Associate Specialist, Community Paediatrician, Wolverhampton and Lucy Butler, Consultant Ophthalmologist, Birmingham.

The lead clinician is usually the doctor in charge of the child’s audiological care. This may be, for example, the community paediatrician, or the audiovestibular physician or ENT consultant, or another clinician, depending on local circumstances. It is the responsibility of education and health professionals to express concerns regarding a child’s vision to the lead clinician. The lead clinician is responsible for coordinating all aspects of the care and management of a deaf child, including their eye and vision care. They are also responsible for ensuring that all professionals who work with the child communicate effectively with the child and their family.

The lead clinician should ensure that a key worker is assigned to the child and family as soon as deafness has been identified. This can be any key member of the multidisciplinary team, for example the teacher of deaf children.

Aiming high for disabled children states that “Key working is a way of managing the package of support available and ensuring families access the services to which they are entitled, with workers being named individuals who act as a single point of contact for multiple services, empower families and help them navigate the system.” (DfES, 2007). The key worker plays a crucial role in supporting the family and providing an important link between parents and professionals.

They also have a major role in ensuring that professionals work together to provide an effective and seamless service. Ideally they should have counselling and communication skills. Families should be involved in deciding who their key worker will be and what the role involves.

“Studies of key workers consistently report positive effects on relationships with services, fewer unmet needs and greater family well-being.”

(Department of Health and DfES, 2004b)



Recommended additional training for vision care personnel

Quality standard 5

Staff working with deaf children should receive training in deaf awareness, including issues relating to childhood deafness.

To ensure a responsive child and family centred service, training in issues relating to childhood deafness, deaf awareness, communication approaches and technology is essential. Other areas in which staff can receive training include:

- developmental implications of a congenital deafness
- language development in deaf children
- using an interpreter and other communication services
- creating a deaf friendly environment, including information on acoustics, lighting, positioning of child, and glare control
- technology that deaf children may use
- multi-agency working practice (Department of Health, 2008).

Professionals' continued training and development plays an important part in providing informed choice for families:

“Training underpins many of the key concepts associated with Informed Choice, eg the provision of high quality, relevant information; increasing professional abilities to promote the empowerment of parents as active choosers; improving access to what is available; resource allocation decision-making and so forth. It is, therefore, a factor (like attitudes and values) that oils the wheels of Informed Choice as a successful process for both parents and professionals.” (Early Support, 2006).

For information about training, contact your local authority specialist services for deaf children. Further education colleges may offer sign language courses, which might include some deaf awareness training.

Part three

The local audiology service

The child's ongoing review by the local audiology service

Quality standard 6

A deaf child or young person should be referred for ophthalmic assessment at any time if parents or the education service have concerns (Hall and Elliman, 2006).

Audiologists should be aware of the importance of optimal vision and be responsive to any concerns about eyesight expressed by parents or school staff.

We have provided a checklist that can be photocopied and used with the child or parent by staff responsible for the deaf child, for example the audiovestibular physician, community paediatrician or teacher of deaf children, even if the child has been discharged from the eye clinic. The checklist contains useful questions that may help you to establish whether there should be concerns about a deaf child's eyesight. If there are concerns, the child should be referred to have their vision assessed by a paediatric ophthalmologist. Please see the 'Checklist for vision problems' appendix 3 on page 43.



The referral process

Quality standard 7

The eye clinic should be informed of the family's communication preferences in advance so that suitable provision can be organised for appointments (Department of Health and DfES, 2004a).

If the child or young person has not been referred for full ophthalmic assessment, if they have not attended appointments, or if there are any concerns or indicators, the child should be referred (or re-referred) to the ophthalmology service for assessment. Both the lead clinician and the GP should be informed of the referral.

The referral letter should state:

- the child or young person's name, date of birth, address, and NHS number
- the reason for referral
- information about the child's deafness, including age of onset, cause (if known), type and degree of deafness
- information about the language or communication methods that the child or family members use, and information on any interpreters or communication support required
- information on any technology that the child uses (for example, hearing aids or cochlear implant)
- clinical observations and results of any tests or examinations so far
- relevant information on family history of vision impairments or conditions related to vision impairments
- concerns of parents, other professionals, and the child or young person
- additional needs of the child or family that will need to be accommodated during appointments and when communicating with them
- the name of the child or young person's school or college
- information that has been given to the family.



This example of a referral letter may help you when writing your own letters:

Dear _____

Re: (name, address, DOB, NHS number)

Thank you for seeing this eight-year-old boy who has severe mid and high frequency deafness. He wears bilateral hearing aids and communicates well using spoken English. His parents have limited English and so a Punjabi speaking interpreter will be needed.

X's deafness was identified late and may be progressive. Investigations to date include imaging and serology have been negative. The result of CMV PCR on the dried blood spot is awaited. There is no family history of deafness. There have been no concerns about X's eyes or vision and there is no family history of visual problems.

On examination, eye movements appeared full and no squint was detected. Recent orthoptic screening in school was satisfactory.

I have explained the importance of vision assessment to X's parents using an interpreter and have also given them a copy of the *Vision Care* publication. X knows that you will need to use eye drops to examine his eyes fully, but please remind him and his parents during the appointment.

[Copies sent to GP, School Nurse, Paediatrician, ENT and Parents of X]

Copies of letters sent between professionals should be copied to the family. Alternatively, to ensure that parents are able to understand all letters that you send, you could write the letters directly to the parents, using appropriate language and formats, and then copy them to other professionals working with the child and family.



Part four

Assessment and care by eye specialists

Before the visit for eye assessment

Quality standard 8

Written information about the visit to the clinic should be given to the family before the clinic appointment, and must be suitably worded for the family and in a format that the older child or young person can understand if appropriate.

As soon as an appointment is made, the appropriate communication support should be booked if it is required.

Before the child and family visit the eye clinic, the key worker and the ophthalmology service should work together to ensure that the family's specific needs are met so that the maximum value of the assessment is obtained (see 'Good testing techniques for examining a deaf child's vision' section on page 25).

To make good use of the appointment and reduce any anxieties that the child or young person and their parents may have, the family should be told in advance what will happen during the appointment and how long it is likely to take. Information should include details about the tests that are to be carried out. If eye drops are likely to be used information regarding the effects and care requirements should be included in the appointment. You should also inform parents if there are any special arrangements they will need to make. Written information needs to be suitably worded for parents and age-appropriate for the child or young person if it is intended that it will be read by them. We have included an example of the information that could be sent to parents about their child's appointment in the appendix 4 on page 44.

There are a number of children's books on the subject of eye tests and eye care which can be helpful for families to use to familiarise children with the idea of going for eye tests. See the appendix 5 for details of some of these books on page 46. A preliminary visit to the assessment centre can also be beneficial to familiarise the child with the setting. The NDCS and Sense publication *Vision care for your deaf child* also has some useful information.

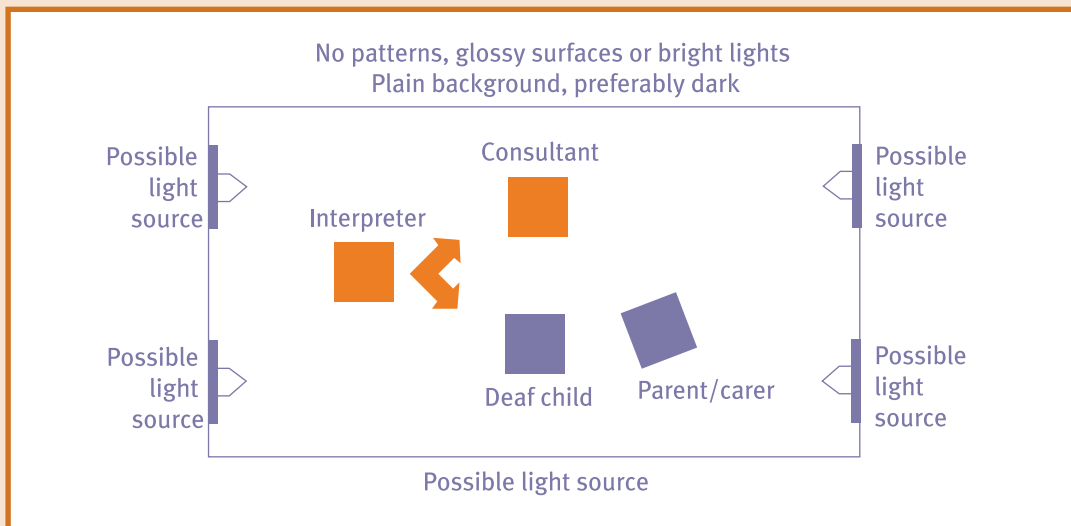
If standard procedures are inappropriate for children with multiple impairments, establish how the test can be carried out (see 'Deaf children with additional needs' section on page 32). It may be useful to consult with other professionals who work with the child to identify what can be done to ensure the tests are carried out appropriately. Other professionals may have more information on any needs the child and family may have.

The visit

On the day of the appointment at the eye clinic, the professional assessing the child should remind all team members of the child's visit and any specific needs that the child or family have. Before, and during the appointment, you should:

- check whether hearing aids or a cochlear implant are being used during the assessment
- check the level of spoken language that the child understands
- speak clearly using language at an appropriate level without covering the mouth, shouting, or exaggerating your lip patterns
- speak directly to the person being tested, but allow time for the parent or interpreter, if present, to explain what you are saying, if they are providing communication support
- create uncluttered backdrops so that the child can focus on the face in order to lipread more easily and/or to see sign language if it is being used
- ensure that the light falls on the face of the person talking, and that faces of people present are not in shadow (see Diagram 1)
- cut out glare from white/shiny surfaces or bright sunlight
- ensure any instructions are given and understood before turning out any lights.

Diagram Suggested room layout/positioning when assessing a deaf child



Notes

- 1 Use a chair with wheels for interpreter flexible positioning.
- 2 Parent's position may need to be in child's field of vision. Encourage them to move as needed.
- 3 Ideally interpreter, consultant, child and parent should be in a circular seating position.
- 4 If the child has visual field problems, interpreter may need to be very close or in a small sign frame.

Booking sign language interpreters and other communicators

Each hospital trust has a policy and guidance on booking sign and other language interpreters. Booking sign language interpreters and other communication support is usually done through agencies, local deaf organisations or directly with individuals offering services on a freelance basis. It is important to book any interpreters for a sufficient length of time in order to cover history, examination, medical tests, feedback of information, results, and next steps. Sign language interpreters are usually booked by the session with two hours as a minimum period. Some sessions may require two interpreters.



Before booking, find out what type of sign language is preferred by the child (or parents), for example British Sign Language (BSL), or sign supported English (SSE). Bookings must be made well in advance, whether booking through an agency or directly with interpreters, as interpreters are often booked weeks, sometimes months in advance. As soon as an appointment is made with a family, the appropriate communication support should be booked if it is required.

Types of communication support include: BSL/SSE/visual frame/tactile signing/deafblind manual interpreters, note takers, lip speakers, palantype and speech to text services. Check whether the services usually used by the hospital trust offers the communication support required. Agencies are listed in the Signature directory.

Booking freelance interpreters means having to make telephone calls to see who is available. However, there will not be a booking fee. There are three directories of sign language interpreters, all are regularly updated: Signature, ASLI, and SASLI. Some freelance interpreters will be listed in all three. You can find out more about these organisations in the 'Useful organisations' section. Some agencies specialise in short notice booking and other specialist services such as interpreters for deafblind people. The Signature directory lists registered deafblind interpreters.

Part five

The ophthalmic assessment

Ophthalmic assessments for deaf children and young people

Quality standard 9

“Staff working in the paediatric eye clinic should have a common core of skills, competencies and knowledge” as identified by the Royal College of Ophthalmologists, 2005.

Referral pathways for deaf children should differ from those recommended for the general child population. Orthoptists and optometrists involved in the routine screening of children should be aware of the increased risk of associated ophthalmic problems in deaf children and refer deaf children to an ophthalmologist if eye examinations highlight problems, or if the child has not previously seen an ophthalmologist.

The consultant ophthalmologist is responsible for the ophthalmic assessment of a deaf child. Ideally the consultant ophthalmologist will have experience of working with deaf children and knowledge of the different communication approaches and the different kinds of technology that deaf children may use.

At the ophthalmic assessment a full history should be taken and a thorough eye examination performed (see ‘Good testing techniques for examining a deaf child’s vision’ section on page 25). Certain findings – for example, the presence of a retinal dystrophy – may help to identify the cause of a child’s deafness. An orthoptic assessment forms part of the ophthalmic examination. It may include formal testing and recording of visual acuity, functional assessment of vision, diagnosis of strabismus and eye movement anomalies, and investigations for the presence, absence or potential for binocular vision. Some deaf children, including those with additional needs, might require “other specifically designed vision assessment tests.” (Nikolopoulos et al., 2006). It is important that the professional carrying out the assessment is able to be flexible to meet the needs of the individual child, and the tests being used must be suitable and likely to obtain accurate information about the child’s vision (Nikolopoulos et al., 2006).





The full clinical history should include specific questions about parental concerns about their child's vision, any concerns that the child or young person may have, the cause of their deafness, and other indicators such as problems with night vision, or delayed onset of walking. A family history of relevant eye or hearing problems should be noted. The comprehensive examination will also include visual acuity, refraction, visual field assessment, and ophthalmic examination.

For some deaf children, an electroretinogram (ERG) may be offered to find out how the retina responds to light. Before this is carried out it is important that the family understands why this test is being offered, what it entails, and that they can defer or refuse the test if they wish. It should be noted that the result from an ERG can help confirm the cause of the child's deafness. If the child's deafness is linked to a retinal dystrophy it is likely to be caused genetically. It is important that families have this information and are offered genetic counselling.

If the child's deafness is caused by one of the types of Usher syndrome it is important that the family knows that any subsequent deaf children they have are likely to have Usher syndrome. Children with Usher Type 1 can benefit from early cochlear implantation, which enables them to gain language through aided hearing at the optimum time for language and speech acquisition.

Current thinking is that an electroretinogram (ERG) may best be performed in children from eight years old. If there are any symptoms suggestive of Usher syndrome including night blindness, field loss or unexplained reduction in visual acuity and if the family history raises specific concerns an ERG can be carried out earlier.

An ERG is performed without anaesthesia. As much of the testing is performed in the dark, it is crucial that the child and their parents receive a clear explanation of the procedure in a well-lit room before starting the test. The family should also have received information about the test before the appointment. It is essential that departments performing the tests use a protocol which allows rod and cone and inner retinal responses to be identified. This may only be available at specialist centres, in which case the child should be referred to a centre with these facilities.

Good testing techniques for examining a deaf child's vision

The following can be adopted to help ensure the best outcome of the assessment:

- Avoid holding orthoptic fixation targets in front of your face. Position yourself so that the child can lipread you and can see your face to observe your facial expressions and gestures.
- Let the child know if and when you need to get close, and whether the contact will be by hand, occluder, torch, lens, etc.
- Rather than impose a key card for vision testing, accept signing, with interpretation, if required. Find out what is most appropriate for each child.
- Ensure that a deaf child knows what will happen during the appointment, and what they need to do before the lights are dimmed or turned off if this is required during a session. If you need to darken the room, you should only do so once you are sure the child understands what will happen. Allow the child to sit next to someone familiar to them during the test to reassure them if they would like this.
- Lipreading and sign reception may be restricted after eye drops are inserted. It is therefore important to explain the short-term effects to the child or young person and the parent before eye drops are used.
- Use toys, dolls or the parent for demonstration purposes as this can be a useful visual reinforcement. This is particularly helpful with occlusion, when each eye is covered in turn, or if the child seems apprehensive.
- During the test, visual reinforcement (such as smiles or thumbs up) will help to reassure the child. Encourage others in the room to do the same thing.

Ensure that a deaf child knows what will happen during the appointment.

The ophthalmic care pathway for a deaf child

The paediatric ophthalmologist who oversees the primary vision screening service for deaf children should liaise with the lead clinician and agree criteria to refer deaf children and for treating children diagnosed as deaf following the screening examination.

Routine screening at birth

All newborn babies will have their eyes examined as part of the routine screening programme (Department of Health and DCSF, 2008, Hall and Elliman, 2006). Babies with identified disabilities should be routinely examined by the paediatric team for congenital abnormalities that may have ophthalmological or audiological implications.

All newborn babies will be offered a hearing screen, either in hospital before discharge, or at home or in a community clinic at about 10 days of age. Babies who are born deaf will therefore be diagnosed in the first few months of life.

At diagnosis of deafness

Quality standard 10

Following the diagnosis of permanent conductive or sensorineural deafness, or if a child needs hearing aids, the child should be referred for a full ophthalmic assessment, and this should be repeated where appropriate at key stages of their development.

Quality standard 11

All staff involved in the diagnosis of deafness must be aware of when and to whom to refer a deaf child. All staff should be aware of local protocol for referral to the ophthalmologist.

“Specialist ophthalmic examination should be carried out as soon as the diagnosis of deafness is confirmed.”
Hall and Elliman, 2006

Nikolopoulos et al. recommend that “Specialist ophthalmic examination should be carried out as soon as the diagnosis of deafness is confirmed irrespective of age and may need to be repeated at intervals after diagnosis.” (Hall and Elliman, 2006, Nikolopoulos et al., 2006, NHSP, 2004).

At one to three years of age

Children with bilateral moderate or worse congenital sensorineural deafness should be referred for an assessment to a paediatric ophthalmologist. Ideally the child should be seen in the child development centre or specialist ophthalmic clinic where they can be accurately monitored and any relevant treatment or referral undertaken.

At four to five years of age

The Child Health Sub-group of the National Screening Committee recommends that all children have their vision screened by an orthoptist between the ages of four and five years of age, which will include an assessment of visual acuity (Department of Health and DCSF, 2008, Hall and Elliman, 2006). As part of this screening programme deaf children will have their vision assessed. However, this ‘routine general screen’ may not meet the additional needs of the deaf child, and may need to be adapted.

“It is important to ensure that vision screening is undertaken in schools for children with hearing impairment because (a) good vision is particularly vital for a person with impaired hearing, and (b) conditions such as Usher’s syndrome, which have important genetic and educational implications, may present with insidious loss of vision.” (Hall and Elliman, 2006).

At seven to nine years of age

Children with permanent sensorineural deafness where the cause of the deafness is unknown should be seen by an ophthalmologist to exclude Usher syndrome as the cause of their deafness (see ‘Usher syndrome’ appendix 7 on page 50). Presently, there is no guidance on the optimal age for testing for Usher syndrome, and eight years of age is considered the most appropriate. If an ERG is carried out, a normal result at this age may not exclude type 2 or type 3 Usher syndrome. A deaf baby who sits up and walks late, a deaf child who has poor night vision and poor balance in the dark, or whose deaf sibling has already been diagnosed with Usher syndrome should be referred for diagnostic testing earlier.

Transition to secondary school

Since myopia may increase during the adolescent years, all deaf children should have their visual acuity reassessed at this age by an optometrist or an orthoptist. If there are any concerns regarding night vision, field loss or visual loss that is not corrected by glasses, the child should be referred for a full ophthalmological assessment. Referral protocol may vary across different areas, but a child should still be referred if there are concerns such as those mentioned above.

At referral for cochlear implantation and other complex interventions

Some deaf children will need specific intervention methods to meet their individual needs, such as a cochlear implant or a bone anchored hearing aid. When a child is referred to a specialist service the receiving clinician should ensure that the child has had their vision checked as part of their specialist assessment. If there has been no assessment it should be arranged for the child to be seen by the ophthalmology service as soon as possible.



Local optometry services

The local optometrist will see some children with permanent deafness. All children should be:

- referred to the local ophthalmologist for further evaluation if there are concerns about their vision
- encouraged to show their prescription to their lead clinician, so that the information can be included in the child's case file, and the relevance discussed with the parents and other team members (with the family's consent). This is only relevant if the child knows about their prescription and is able to share this information with their lead clinician.

If a young deaf person has had no refractive problems during the school years, they should be encouraged to refer themselves for optometric testing every two years on transfer to adult services. (see 'Young people and transfer of care to another service' on page 36).

Following the visit

Parents need clearly written but sensitively expressed reports, correspondence and management plans relating to their child.

Quality standard 12

A report of the outcome of the visit should be sent to the parent, the referrer and the child's GP, and to all other key professionals involved. This includes copies of assessments and diagnostic results. Parental consent is needed before information is shared within the multidisciplinary team.

Parents need clearly written but sensitively expressed reports, correspondence and management plans relating to their child. This will promote a sense of partnership and trust between parents, the child or young person, and professionals. Age appropriate information will help young people to understand their vision care and any implications it may have (Department of Health, 2003).

A copy of any reports should be sent to the person who made the referral, the parent and/or child or young person, the GP and lead clinician, with the parents and child's agreement (Department of Health, 2003). Reports could include:

- identified pathology
- the management plan
- implications for the diagnosis and management of the deafness or other conditions
- information given to parents and (where relevant) age appropriate information to the child or young person
- whether an interpreter was present (where relevant)
- whether further verbal and/or written information about voluntary organisations, equipment, or other services were provided.

The key worker should be involved in any management planning. The child should already have a 'child and family plan or programme', and it is important that any visual difficulties are managed in conjunction with their deafness. Where there are implications arising from the vision assessment, a meeting should be arranged with key personnel to modify the plan. If there is new or potentially distressing information to be communicated to families, this should be discussed with the family before any written information is sent to them:

“Where the letter contains abnormal results or significant information that has not been discussed with the patient, it will be important for arrangements to be made to give the patient a copy of the letter after its contents have been discussed in a consultation with the receiving professional. As a general rule the contents of copied letters should reflect the discussion in the consultation with the sending healthcare professional, and there should be no new information in the letter that might surprise or distress the patient.”
(Department of Health, 2003)



Working with families

Families of deaf children may not be familiar with terminology relating to vision impairment and vision assessments. Check that parents understand any terminology that you use, and give them opportunities to ask questions (Scope, 2003).

Talk about the child or young person in a positive way, and be cautious in making long-term predictions (verbally or written), especially negative ones. The need for sensitive, honest and careful use of language, together with constructive statements about the future, cannot be overemphasised. Convey warmth, understanding and respect. Where a child has been diagnosed with a serious eye condition, be careful about using terms such as 'could lead to blindness' or 'on the blind register' as this may not help the family adjust to the diagnosis or in their planning for the future.

Every effort should be made for both parents (wherever possible) to receive their child's diagnosis or test results together, so that one parent is not left to give the news to the other parent. Single parent families may wish to bring a relative or friend with them, and professionals should accommodate this.

Parents need time to absorb any diagnosis or condition their child has, and should be given the opportunity to meet with professionals when they need to. Brief, written, easily understood information about the child's condition should be provided. Check that parents understand any terminology or explanations in the information. Written information does not reduce the need for professionals involved to discuss things with parents or young people. Provide a private, family friendly area for any meetings or discussions, and ensure that meetings are arranged at appropriate times (Department of Health and DfES, 2004a).

Encourage parents to contact specialist agencies, parents' support groups, and voluntary organisations for advice and help. Parents should be provided with relevant information about agencies and services (see the 'Useful organisations' appendix 11 on page 61).

Diagnosis of a serious eye condition

Quality standard 13

Ideally a follow-up appointment within two weeks should be offered to children who are priorities (for example, if they have been diagnosed with a serious eye condition) so that the parents and child can discuss further questions. The family may want to involve their key worker in appointments. The family should be provided with contact details for the vision team or the local authority visual impairment teaching service.

Where a child's deafness is linked to a serious and/or progressive eye condition, the child and family will need support in understanding the short- and long-term implications.

Where a child's deafness is linked to a serious and/or progressive eye condition, the child and family will need support in understanding the short- and long-term implications. For parents who have already had to assimilate the news that their child is deaf, the possibility of an additional eye problem or progressive visual deterioration may cause shock and anguish, especially if no treatment exists. Members of the eye care team will need to show particular care and sensitivity in giving the news (Scope, 2003). Some distress can be alleviated by the manner in which the diagnosis, and subsequent follow-up help and support is given. Deaf parents may not feel negative about their child's deafness. However, they could be unduly shocked by a diagnosis of a vision problem.

Information given to parents at the initial consultation will need further discussion at a later stage, when parents and the child have had the opportunity to assimilate the diagnosis. A follow-up appointment should be offered to discuss questions and concerns that occur as the parents and child consider the issues and impact of the diagnosis, especially if only one parent was present originally and/or the child has not been informed. Some parents may need even more time to absorb the diagnosis. Therefore flexibility for the return appointment will be necessary. This appointment could involve members of the ophthalmic team who have been trained to support families. The consultant need not necessarily be present.

Counselling should be available soon after diagnosis to enable parents and children to adapt to a serious dual sensory impairment. Some parents may feel unable to share the implications of the diagnosis of a serious eye condition with their child for some time and may themselves continue to be distressed, especially if their child's vision, hearing, or other conditions continue to deteriorate. It is important, therefore, that professional support is extended to families who need time to find their own way to adjustment and disclosure.

Hearing aids, cochlear implants, glasses and eye patches

For a child who is already wearing hearing aids or a cochlear implant, the prospect of another 'aid' may be difficult to accept and adjust to. Most deaf children will be fitted with hearing aids or a cochlear implant that sits behind the ear, and with advances in technology, aids have become smaller and slimmer. However, the question of whether there is enough room behind the child's ears to fit hearing aids and glasses may pose practical problems. The optometrist and the child's audiologist need to liaise closely to ensure that the child will be able to wear both as comfortably as possible, using additional soft supports if needed (NDCS and Sense, 2006). Wire-framed glasses' arms that wrap around the ear can be particularly useful for younger children and sit more easily next to hearing aids. For older children and young people there may be the option of wearing contact lenses or in the ear (ITE) hearing aids (depending on their level of deafness).

It is possible to get hearing aids that are incorporated into the arms of glasses. Although this type of aid may overcome the problem of space behind the ear they are not widely used, especially in children. This is mainly because when the glasses are removed the hearing aids also have to be taken off. Furthermore, if one of the aids needs to be modified or repaired the child will be without both their hearing aids and their glasses during this time. Cochlear implant processors have not, as yet, been incorporated into the arms of glasses but the same considerations would apply.

In some cases children with reduced visual acuity in one eye due to amblyopia may require patching treatment of the eye with the better visual acuity. Orthoptists and any other professionals working with the child should be aware of the possible effect of patching children who may have a visual field defect and use sign support and/or lipreading as an integral part of their communication.

Hearing aids: Information for families; Bone anchored hearing aids for children and young people: A guide for parents and families; and Cochlear implants: A guide for families by NDCS are all useful publications that can give you more information. You can find the details of these publications in the 'References' appendix 9 on page 56.



Deaf children with additional needs

Quality standard 14

The clinician carrying out the assessment should be given specific information on the deaf child's additional support needs before the assessment.

Up to half of all deaf children may have an additional disability (NDCS, 2007b, Cherow 1993, Fortnum et al. 1996, Stredler-Brown and Yoshinago-Itano, 1994), Carrying out detailed audiological and vision assessments on deaf children with complex needs may be time consuming and complex. A preliminary visit to the assessment centre can be beneficial to the child to get them used to the surroundings.

Children with multi-sensory impairment have a combination of visual and hearing difficulties. They are sometimes referred to as deafblind but may have some residual sight and/or hearing. Many also have additional disabilities but their complex needs mean that it may be difficult to ascertain their intellectual disabilities. Children with multi-sensory impairment have difficulties in perception, mobility and orientation, communication, and in the acquisition of information. Incidental learning is also more limited, and they may need alternative means of communication (SERSEN, 2006).

Specialist teachers of deafblind or multi-sensory impaired (MSI) children are employed in schools or as part of their sensory support team. It is advisable, where possible, for all children with dual sensory loss to have a specialist assessment of their needs from a team which includes a MSI teacher. For more information on MSI teachers, see the 'Key personnel' appendix 2 on page 40.

A standard approach to testing may not be successful for children who have problems recognizing objects, pictures, colours and shapes that are part of standard testing methods. Children with cortical damage may recognise objects through touch rather than by sight. Some children have difficulty seeing objects against complex backgrounds or if they are not presented in such a way as to maximise the child's ability to respond.

The following guidelines may be useful when assessing deaf children with additional needs (Brown, 1999):

- consult with those who know the child; for example the parents, health visitor and/or specialist teacher
- identify motivators (ie what interests the child), use all the senses and not just vision
- use a variety of sensory clues in order to alert the child to use their vision, eg a resonance board to show that a visual signal is coming
- develop strategies for relaxing or arousing a child who is overreacting to certain stimuli or unresponsive to what is happening around them
- position the child so that they are free to use their vision, eg allow the child to lie down rather than sit at a table if it is more appropriate for them.

The following provide more information on additional needs and multi-sensory impairment:

- *Vision for doing: Assessing functional vision of learners who are multiply disabled* (Aitken and Buultjens)
- *Essential elements in early intervention visual impairment and multiple disabilities* (Chen, D)
- *Eye to eye, measuring and describing vision in young children* (Woodhouse)
- *Visual Impairment: Access to education for children and young people* (Mason and McCall)
- *Teaching children who are deafblind* (Aitken and Buultjens)
- *Information for families: Multi-sensory impairment* (Early Support)
- *Deaf children with additional needs* (NDCS)

You can find the details of these publications in the 'References' appendix 9 on page 56.

You may also find it helpful to look at the resources on the Project Salute website www.projectsalute.net and the Lilliworks Active Learning website www.lilliworks.com

Deafness as part of a syndrome

Syndromic deafness (deafness associated with other features, such as vision impairment or kidney abnormalities for example.) accounts for 20%-30% of inherited or genetic childhood deafness (NDCS, 2008). Full diagnosis is important, as there may be associated medical problems or special educational needs or additional support needs for the child (Guest, 2000).

If the ophthalmologist diagnoses a deaf child with a genetic condition, the family, including the child or young person if appropriate, should be offered referral to a clinical genetics service.



The clinical genetics service

Quality standard 15

If a child's vision impairment or dual sensory impairment is unexplained or suspected to have a genetic cause, the lead clinician is responsible for ensuring that a genetic referral is offered to the family and, if accepted, arranged.

Every effort should be made to ensure that the family understands the diagnosis and care plan. Important information should be translated into a suitable format or language if it is needed.

Where a vision problem or multi-sensory impairment might be inherited or the cause is unknown, it is the responsibility of the lead clinician to offer a referral to the genetics service after discussion with the family. If a deaf child is diagnosed with a genetic condition, any sibling who is also deaf may have the same condition and they should be checked to ascertain whether this is the case. It is rare, though not impossible, for the sibling to be deaf for different reasons.

Young people who wish to talk about their condition, or siblings of an affected person, may be seen directly by the clinical genetics service that has previously seen their family and holds the details of their family history. They can be also be referred by their GP or Consultant.

For further information about genetics and deafness, and what families can expect from genetic counselling, you may like to read *Genetic counselling and deafness* by NDCS.

Genetic tests are only available for a few genetic conditions and such tests are individually arranged by the consultant in clinical genetics or the specialist ophthalmologist. Where no genetic test is available, information given to families on the risk for future generations is based on family history, experience with similar families in the past and medical literature.

Many genetics services hold outreach clinics in local hospitals and child development centres so that families do not always have to travel long distances.

Meeting the needs of ethnic minority families and hard-to-reach families

There is a higher incidence of deafness among some ethnic groups (Fortnum and Davis, 1997). Family centred services should take both linguistic and cultural diversity into account. Qualified language interpreters must be provided at hospital appointments and home visits for families who need them. Some languages do not contain words relative to the specific findings in deafness or other medical conditions. However, the family's cultural and linguistic needs must be met (Department of Health and DfES, 2004a). Every effort should be made to ensure that the family understands the diagnosis and care plan. Important information should be translated into a suitable format or language if it is needed.

Meetings, information and follow-up appointments should be handled sensitively and with respect for the needs of the individual family. Sometimes the venue for meetings, timing and day of appointments and the method of transport used to get there will dictate whether a family can attend (Vision 2020 UK, 2008). Networking with other parents and family members with similar needs and experiences should be offered (Vision 2020 UK, 2008).

Some families will be hard to reach and may need support to access services and transport. Non-attendance at appointments for children with hearing and vision problems should be followed up to understand the reasons why, and to offer support to ensure future attendance. The social worker for deaf children or another professional may be able to help. It should not be assumed that letters or reports sent home are understood.

Children from hard-to-reach families may have disjointed access to education and health services and they may miss the screening and health surveillance programmes (Department for Health and DfES, 2004b). Official letters or appointments may not be acted on because they may not be fully understood. Furthermore, families arriving in a new place may have limited means of finding out about education and health care. Some children may not be familiar with everyday objects such as pencils, pictures or bricks and may need longer to play with test material before assessment begins. The needs of these children and families must be assessed and there should be flexibility in protocols to accommodate their needs. The organisation Friends, Families and Travellers provides useful information on their website www.gypsy-traveller.org covering all aspects of supporting Gypsies and Travellers, regardless of ethnicity, culture or background. Most local authorities have useful information on hard-to-reach families, such as travellers, asylum seekers and ethnic minorities. You may find this information on your local authority's website, or contact them directly to find out more.



Part six

Young people and transfer of care to another service

Transfer from paediatric to adult services

Quality standard 16

To ensure continuity of care, all relevant information must be passed to the adult ophthalmology service (with copies sent to the GP) when the young person transfers from the paediatric service.

It is essential that policies and procedures are in place to ensure the smooth transition from the child to the adult ophthalmology service.

School leavers need to be informed about the services and resources available and the support they will require when they leave paediatric services and move into further education and employment.

“Disabled young people need high quality, multi-agency support to allow them to have choice and control over life decisions, and to be aware of what opportunities are open to them and the range of support they may need to access.” (Department of Health and DfES, 2004b)

Young people who wish to talk about a condition in their family that is known or suspected to be genetic, can be referred to the clinical genetic service, ideally to a specialist counsellor in deafness with sign language skills, if the deaf young person uses sign language. The clinical genetic service sees individuals of any age (see ‘The clinical genetics service’ section on page 34).

Young people with a significant ophthalmological condition will remain under the care of the hospital based ophthalmology service. It is essential that policies and procedures are in place to ensure the smooth transition from the child to the adult ophthalmology service.

The transition plan “should form part of the annual review in year nine, and any subsequent review. The purpose of the plan is to draw together information in order to plan coherently for the young person’s transition into adult life” (SERSEN, 2006). As a result of this, all relevant information about the young person must be transferred so that the adult service is fully aware of their medical history (Department of Health and DfES, 2004a).

Adult services should be made aware of any changing circumstances, for example, leaving home or moving into further or higher education, in order to initiate an assessment for assistive devices or other environmental adaptations. The key worker can play an important role in ensuring smooth transition to the adult service. If a refractive error is present, an optometrist should continue to check the young person’s vision, and the optometrist should advise on how often this must take place. If no refractive error has been present throughout their school life, optometry is recommended at two-yearly intervals.

Aiming high for disabled children states that “Disabled children and their families should be able to benefit from services which are easily accessible at key transition points in their life, designed around the child and family, and delivered in a coordinated and timely manner.” (DfES 2007). *The National Service Framework* suggests that “All transition processes are planned and focussed around preparation of the young person rather than the service organisation. Young People and their families are actively involved in transition planning” (Department of Health and DfES, 2004a).

Transfer to an alternative paediatric or adolescent service

Quality standard 17

Before transfer to alternative services, the appropriate professional should arrange all relevant appointments with the new service and make sure that the young person and their family are notified.

Where the child or young person moves out of the area, it is essential that the local ophthalmic service transfers all notes to the new service taking on the ophthalmic care of the child or young person (Hall and Elliman, 2006). The child or young person and family must be fully informed in writing of details relating to the new service. The GP in the new area may need to make referrals to the adult ophthalmic services in the new area.



Appendix 1: Communicating with deaf children

Deaf children have diverse and different needs and communicate in a variety of ways. This section outlines the range of communication approaches and methods that deaf children and young people may use:

British Sign Language (BSL)

A visual language using hand shapes, facial expressions, gestures and body language. BSL is an independent and complete language with a unique vocabulary; its structure and grammar differ from that of written and spoken English. Approximately 70,000 deaf people are estimated BSL users in the UK.

Cued Speech

Some words that sound different look very similar when they are lipread. Cued Speech uses hand shapes placed near the mouth to accompany spoken language to help make every sound and word clear to the deaf child. It can be used together with sign language or to complement an oral approach.

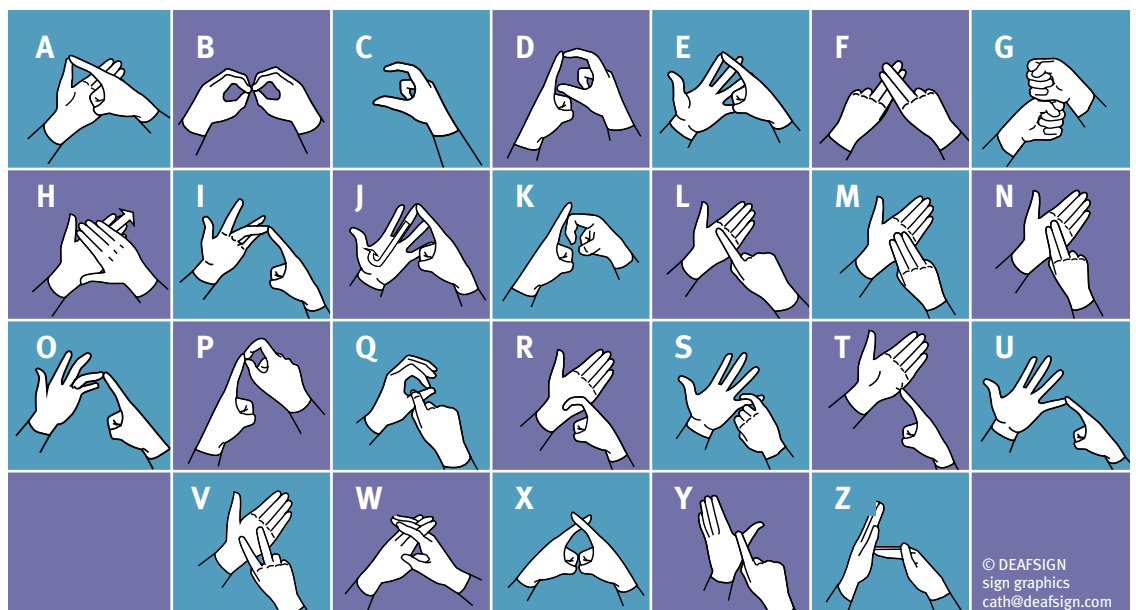
Deafblind manual

The alphabet is represented by placing the fingers of one hand on the hand of the receiver in a variety of ways to represent different letters.

Fingerspelling/manual alphabet

Each letter of the alphabet is given its own shape, and is used to spell names and places and other words that do not have their own sign.

Fingerspelling alphabet.



Lipreading/speechreading

This is the ability to read lip patterns. Although lipreading is difficult to learn, many deaf children naturally lipread when they are communicating. Lipreading is a poor communication tool when used on its own, as most deaf children can only understand a small percentage of what is being said. However, it is useful when used alongside other communication methods.

Makaton

A graded sign vocabulary system, based upon signs in BSL, used with children (deaf and hearing) who have severe communication and learning disabilities.

Signalong

A sign supported system for people who have language difficulties associated with learning disabilities and autism. The signs are mostly based on BSL and are used in English word order. Signalong is intended to support spoken language and is sometimes used with deaf children.

Signed English (SE)

An exact representation of the English language through signs, using a sign or fingerspelling for every spoken word. BSL signs are employed together with specially developed signs that give guidance on important points of grammar and fingerspelling. SE is not a communication method like BSL or SSE, but has been designed as a teaching tool to be used at the same time as spoken English.

Sign Supported English (SSE)

Uses signs taken from BSL. SSE is used in English word order but does not attempt to sign every word that is spoken.

Speech

Hearing is the primary sense through which language develops from birth. A significant proportion of deaf children use speech to communicate.

Tactile signing

“Signing approach based on a standard manual signing system (for example, BSL) in which the child’s hands are placed under the hands of an adult in order to receive signs and over the hands of the adult to give signs” (SERSEN, 2006).

Further information about communication methods can be obtained from NDCS and Sense.

Audiologist and audiological scientist

Non-medical professional who specialises in the identification, assessment and rehabilitation of deafness and balance disorders, including the fitting of hearing aids.

Audiovestibular physician or consultant (consultant in audiological medicine)

A doctor who investigates, diagnoses and manages children and/or adults with deafness, balance and communication disorders, including tinnitus and auditory processing disorders.

British Sign Language (BSL) interpreter

Interprets from spoken English to BSL, and BSL into spoken English.

Communication support personnel

People who are trained to provide appropriate communication support such as BSL interpreter, Relay interpreter, Note taker, Communication support worker, and Communicator guide (SERSEN, 2006).

Community paediatrician (audiology)

Children's doctor with specialist training in audiology and child development who is closely involved in the diagnosis, management and co-ordination of services for deaf children.

Educational audiologist

A qualified teacher of deaf children who has undertaken specialist training in audiology.

ENT surgeon (consultant otolaryngologist)

A doctor who specialises in diseases of the ears, nose, throat, head and neck, and who performs relevant surgery and may be responsible for the general medical assessment of a child (SERSEN, 2006).

Genetic counsellor

A member of the genetics team who usually has a background in nursing and/or genetics and is trained in clinical genetics and counselling.

Geneticist (consultant in clinical genetics)

A doctor who specialises in genetic disorders, identifying and providing advice about inheritance in diseases, disorders or syndromes which may involve deafness or visual problems.

Health visitor

Promotes public health as part of a primary care team. Works closely with families, monitors the child's development through the child health surveillance programme. Provides home visits and gives support, advice and information to parents.

Hearing therapist

Provides a rehabilitative service related to everyday living and functional use of hearing for deaf adults and young people. Also counsels on tinnitus and advises on assistive listening devices.

Key worker

A member of the multidisciplinary team assigned to work with the deaf child and family who will be familiar with local and national services. Also responsible for ensuring that professionals involved with the child and family communicate effectively.

Lead clinician

Is responsible for coordinating all aspects of the care and management of the deaf child, including their eye and vision care. Who the lead clinician is will depend on local circumstances; however, they may be the community paediatrician, the audiovestibular physician or consultant, or another clinician.

Link worker

Alternative title for Key worker (see above).

Ophthalmologist

A doctor who specialises in the medical and surgical management of eye disorders. Assesses visual function and has access to other investigations in order to assess the cause of visual dysfunction. A Paediatric Ophthalmologist specialises in children's eye diseases.

Optician

Is trained to adjust and dispense glasses and other optical aids. Ophthalmic opticians also carry out sight tests and prescribe glasses, although young children with MSI are more likely to be assessed at a specialist clinic (SERSEN, 2006).

Optometrist

Prescribes and dispenses glasses and contact lenses and may provide low vision rehabilitation. Examines ocular health, tests visual acuity, depth and colour perception, and the ability to focus and coordinate the eyes.

Orthoptist

Is trained to test vision and assess functional vision including binocular vision. Diagnoses and assesses strabismus, eye movement anomalies, and the reasons for failure of normal development of the eye. Many are involved in screening pre-school children for visual defects.

Paediatrician

A doctor who specialises in the care and development of children. They are usually based in hospitals and community clinics.

Peripatetic teacher

A teacher with specific expertise who visits homes and educational settings and is employed by the local authority to give specialist advice and support (SERSEN, 2006).

Social worker

Can assess children, young people and families for support, and advise on local and national services.

Social worker with deaf people

A qualified social worker with specialist training who has knowledge of deafness, deaf issues, deaf awareness, and often has sign language skills. Can also advise on benefits and equipment in the home. Social workers with deaf people may not be available in every area.

Specialist speech and language therapist

Assesses and advises on communication, language and speech perception and production and swallowing. They may be based in the hospital, the community or in the education environment. Some may offer treatment, depending on the context in which they work.

Teacher of the deaf (ToD)

Qualified teacher with postgraduate training. Promotes language development, listening skills and linguistic and cognitive access to the national curriculum. They play a key role with pre-school children supporting them and their family in their own home environment, and they work with deaf children in the classroom. They may also work as class teachers.

Teacher of MSI/deafblind

Qualified teacher with postgraduate specialist training, recognised by DCSF, who works with deafblind and multi-sensory impaired children, either in school or as part of the advisory service. They understand the unique impact of deafblindness on a child's ability to develop and learn. They assess children, advise on suitable environments, develop and deliver individual curriculum plans and work with families and other professionals to provide a specialist service to each child.

Teacher of the visually impaired (TVI)

Qualified teacher with postgraduate training in vision impairment. They will be involved in providing assessment, advice on pupil's vision function and a description of their educational needs. A TVI will provide training for staff in schools and advise on programmes of support and intervention. They may also work as class teachers.

Appendix 3: Checklist for vision problems

The eyes:

Child has quick movements in their eye or eyes	<input type="checkbox"/> Yes	<input type="checkbox"/> No
One eye turns in or out	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Wears or has glasses	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Child blinks or rubs eyes frequently	<input type="checkbox"/> Yes	<input type="checkbox"/> No

Near and distance vision:

Child holds things close to them to look at them	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Child has difficulty seeing written information on the black/white board in the classroom	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Child has difficulty finding small items which have fallen on the floor	<input type="checkbox"/> Yes	<input type="checkbox"/> No

Night vision and low contrast:

Child needs more light to read than expected	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Child has problems distinguishing the difference between brown and black	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Child has more problems than usual seeing in poor light, for example, the cinema	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Child becomes unusually disorientated in the dark	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Child reaches out for a handrail or someone's arm in low light or the dark	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Child has worse balance after dark	<input type="checkbox"/> Yes	<input type="checkbox"/> No

Visual fields:

Child has difficulty when going down steps	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Child bumps into things or trips over things more than usual	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Child doesn't read to the end of a sentence before going to the next	<input type="checkbox"/> Yes	<input type="checkbox"/> No

Adaptation to lighting changes:

Child pauses when coming indoors on sunny days	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Child has difficulty going from a light to a dark environment	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Child shows discomfort in bright light	<input type="checkbox"/> Yes	<input type="checkbox"/> No

Eye tracking:

Child moves head instead of eyes when reading or looking at pictures	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Child loses place when reading, and skips lines	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Child needs to use finger as a line marker	<input type="checkbox"/> Yes	<input type="checkbox"/> No

Eye teaming:

Child shows signs of fatigue during near visual tasks, and has poor levels of concentration	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Child has difficulty judging where things are in a space	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Child has difficulty seeing the scenery from the back of a car or difficulty seeing road signs	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Child makes maths errors due to misalignment of numbers	<input type="checkbox"/> Yes	<input type="checkbox"/> No
Child tires easily, is inattentive and has poor levels of concentration	<input type="checkbox"/> Yes	<input type="checkbox"/> No

This is a suggested template that you may want to use when writing information about your service for parents.

How to find us

The full address of the children's eye clinic is: _____

Reaching us by public transport:

Coming by bus, the buses _____ stop outside the clinic. The nearest train station is _____

Driving to the clinic:

Car parking at the hospital costs £_____ for a minimum of _____ hours.

How you can help us

Please tell us the best way of speaking to or communicating with your daughter or son. If you or your child uses sign language and a sign language interpreter is required for the appointment, we need to know well before the date of your appointment so that a sign language interpreter can be booked for you.

What to bring with you

If your child wears glasses, please bring them with you. If your child wears hearing aids, please make sure that the hearing aid is working and switched on. Please also bring your appointment letter. We have a play area for children. There is a café nearby for drinks and light food.

What may happen at the eye clinic

The eye doctor who sees your child may ask you some questions about their deafness, any illnesses in early childhood, and anything you have noticed about their vision; for example, does your child bring things close to her or his face to see them.

Eye drops may be used to help examine the eye carefully. Eye drops are used to examine the back of the eye, and to allow glasses to be checked. Sometimes eye drops can sting the eye a bit, but this wears off very quickly. Eye drops can make the pupil in the eye larger, and can make vision blurred, but this should wear off by the next day. If it is sunny, a pair of sunglasses to wear after the appointment may be useful. If eye drops are used during your child's appointment, you should tell school staff if your child is going back to school after the appointment.

The eye doctor may ask for different tests to be done. Some of these tests will be done by an orthoptist who checks how your child's eyes are working. The orthoptist uses tests appropriate to your child's age to check their level of vision. They will also look to see if there is a squint, and check how well your child uses both eyes together.

The appointment is likely to last for _____ hours.

After the appointment

We will send a letter to the person who referred your daughter or son to us, a copy to your GP and a copy to you. The letter will outline what was found during the appointment; for example, your child's level of vision, if the eyes look healthy, and if any further tests or appointments are needed. If we need to see your child again at the eye clinic we will send you another appointment.

How to contact us

The eye clinic is open from _____ to _____

The telephone number for appointments is _____

The textphone number for appointments is _____

We have included a map of the local area to help you find the clinic, and a map of the location of the clinic within the hospital.

If you have any questions about the appointment, please do not hesitate to contact us.

Arthur's Eyes, Marc Brown, Atlantic Monthly Press/Little Brown and Co, 2008

I Need Glasses: My Visit to the Optometrist, Virginia Dooley, Mondo Publishing, 2002

Luna and the Big Blur, Shirley Day. American Psychological Association, 2008

Magenta Gets Glasses, JP Reber, Simon & Schuster, 2002

Rugrats: Chuckie Visits the Eye Doctor, Luke David, Simon Spotlight/Nickelodeon, 1999

Sight, Kay Woodward, Hodder & Stoughton, 2005

The Good Luck Glasses, Sara London, Cartwheel Books, 2001

Topsy and Tim Have their Eyes Tested, Jean and Gareth Adamson, Blackie Children's Books, 1997

Forest Books specialises in books about deafness and deaf issues. They sell books for children, as well as families and professionals. There is a comprehensive online catalogue on their website www.forestbooks.com. You can find their contact details in the 'Useful organisations' section on page 61.

Appendix 6: Conditions affecting both hearing and vision

The following list is not exhaustive but includes some conditions associated with deafness and ophthalmic disorders. Terms that appear in the glossary have been referenced with the relevant glossary index number. The glossary starts on page 52.

Alport syndrome

The main characteristics are progressive renal disease and progressive sensorineural deafness (61). Ocular signs include cataract (10), conical shaped lens (35), an opaque line surrounding the margin of the cornea (4) and flecked pigment change on the retina with abnormal ERG (19) and EOG (18). It is usually inherited by X-linked recession (71), but may be autosomal dominant (7) or recessive (8) inheritance.

Alstrom syndrome

An autosomal recessive (8) disorder characterised by diabetes mellitus (17), severe sensorineural deafness (61), obesity and degeneration of the retina (59). Other features may include chronic renal disease, pigment changes of the skin, grey-black warty patches on the skin usually in the groin, elbows or knees (1), and incompetence of the testicles or ovaries (30). Poor vision and rapid involuntary movements of the eyes (44) are present in infancy.

Cerebral Palsy

A group of movement disorders resulting from damage to the developing brain. The extent of impairment varies widely. Sight, hearing, speech articulation and intellectual functioning may be affected (SERSEN, 2006).

CHARGE syndrome

CHARGE is an association of features of arrested development:

- Coloboma – a fissure in the lower part of the iris of the eye (12)
- Heart defects
- Atresia choanae – blockage of the nasal passages (6)
- Restricted growth and development
- Genital hypoplasia – impaired function of small testicles and ovaries (31)
- Ear abnormalities and/or deafness (conductive (13), sensorineural (61) or mixed)

A formerly under reported cause of deafblindness, first identified in 1979, it is believed that CHARGE may now be a leading cause of deafblindness among children, replacing congenital rubella syndrome. Recent research has located a mutation of the CHD (7) gene in a significant number of people with CHARGE.

Congenital cytomegalovirus

Ocular findings in infected neonates may include inflammation of the cornea (34), the choroid and retina (11) of the eye, small eyes (39), cataract (10), and degeneration of the optic nerve (46). Systemic involvement may include jaundice (33), enlargement of the liver and spleen (27) and the lymph nodes (36), delayed development, deposits of calcium in the brain, and a rash with reddish/purplish areas on the skin (51). Most affected infants do not present neonatally (42), but some 40% show features of congenital CMV by three years of age; deafness is mostly sensorineural (61). Retinal pigment changes may be seen.

Congenital rubella (rubella embryopathy (20))

The congenital rubella syndrome may include retinal pigmentary changes (60) (in 40% of the affected patients), cataract (10) (in 20%), small eyes (39), raised pressure within the eye (24), and inflammation of the cornea (34). Other systemic abnormalities may include congenital heart defects, abnormally small head (38), bone disease (47), enlargement of the liver and spleen (27) and the lymph nodes (36), deficiency of the blood platelets causing clotting problems (66) diabetes mellitus (17). Affects the child during pregnancy.

Congenital toxoplasmosis

The most common findings with sensorineural deafness (61) are inflammation of the choroid and retina of the eye (11) and deposits of calcium in the brain. Less common findings are seizures, abnormally large head due to increased fluid in the ventricles of the brain (28), enlargement of the liver and spleen (27), jaundice (33), a low red blood cell count (3) and fever. Some infected patients may be normal in the initial examination but some years later develop inflammation of the choroid and retina (11) of the eye, blindness, increased pressure in the skull caused by the large amount of fluid in the ventricles (28), learning difficulties and deafness.

Deafblind

This is a unique disability sometimes known as multi-sensory impairment or dual sensory impairment. It involves a combination of hearing and sight loss (SERSEN, 2006). (See 'Multi-sensory impairment' definition for more information).

DIDMOAD (Diabetes Insipidus (16), Diabetes Mellitus (17), Optic Atrophy (46) and Deafness)

This association of conditions is characterised by juvenile diabetes mellitus (17) and degeneration of the optic nerve (46), which has an onset between 2 and 24 years of age. Visual fields are constricted and the patient is colour-blind. Retinal pigment changes have also been reported. ERG (19) findings imply that a more widespread abnormality may be present than just degeneration of the nerve cells (46). It is inherited as an autosomal recessive disorder (8).

Duane syndrome

The patient is unable to move the eye laterally, medial movements being accompanied by upward rotation and retraction of the eyeball. This may be linked with permanent deafness, both conductive (13) and sensorineural (61), cleft palate and spinal anomalies.

Marshall syndrome (see Stickler syndrome)

This dominantly (7) inherited syndrome includes early onset of progressive sensorineural deafness (61), cataract (10), short-sightedness (41), saddle nose and short stature. It is not known whether it is a variant of Stickler syndrome.

Multi-Sensory impairment (MSI)

Children with multi-sensory impairment have a combination of visual and hearing difficulties. They are sometimes referred to as deafblind but may have some residual sight and/or hearing. Many also have additional disabilities but their complex needs mean that it may be difficult to ascertain their intellectual abilities. Children with MSI have difficulties in perception, communication, and in the

acquisition of information. Incidental learning is limited and they may need alternative means of communication. The children have highly individual needs arising from congenital or acquired loss and/or a degenerative condition.

Norrie syndrome

Abnormal development (58) or detachment of the retina lead to a congenital (14) or progressive loss of vision in early childhood. Progressive sensorineural deafness (61) usually occurs in late childhood. There may also be progressive loss of intellectual function. Inheritance is X-linked recessive (71).

Refsum syndrome

Progressive loss of the pigment of the retina (60) occurs in both child and adults types. Other symptoms include progressive hearing sensorineural deafness (61), loss of sense of smell, inflammation of the motor and sensory nerves leading to muscle weakness. Both types are due to a defect in the metabolism of phytanic acid found in grass and green vegetables. Treatment is dietary and requires supervision from a dietician and avoidance of foods which contain fat from animals which live on green plants, eg products from mutton and beef. Refsum syndrome is autosomal recessively inherited (8).

Stickler syndrome (see Marshall syndrome)

A disorder characterised by a flat facial profile, cleft palate, ocular changes and joint disease. Deafness is either conductive (13) or mixed (40) in nature. The ocular changes include progressive short-sightedness (41), detachment of the retina, and cataract (10), which may present early in life. It is autosomal dominant (7).

Usher syndrome (see Appendix 7)

Varicella embryopathy (chickenpox)

The key features include sensorineural deafness (61) (often moderate to severe), limb abnormalities, inflammation of the choroid and retina (11) and cataract (10). Other systems including the gut may be affected, and other eye problems may be found. Affects the child during pregnancy.

Usher syndrome is characterised by:

- congenital sensorineural deafness
- progressive retinal dystrophy
- in some types (see below) reduced or absent vestibular function.

Current understanding suggests that there are three main types: Usher 1, 2 and 3. The prevalence in the population of Usher syndrome varies. The frequency of Usher syndrome has been estimated to be 3.5/100,000 in Finland (Nuutila, 1970) and in Norway (Grondahl, 1986), 3.2/100,000 in Colombia (Tamayo, 1991), and 4.4/100,000 in the USA (Boughman, Vernon and Shaver, 1983). In the congenitally deaf population it is estimated to account for 3%-6% (Vernon 1969). More recent studies suggest 10% of congenitally deaf people have Type 1, and some data indicates that Usher 2 is almost twice as frequent as Usher 1. The overall frequency of Usher could be more than 1 in 10,000 for all Usher types. Kimberling (personal communication 2008) estimates frequency of detectable Usher is 8% among the deaf and hard of hearing child population.

Advances in molecular genetics in the last decade have, so far, located at least 10 genes implicated in Usher syndrome including 6 genes for Usher 1, in Usher type 2, three genes and in Usher type 3 one gene. With these advances and the successful outcome from clinical trials, the possibility of effective therapies being offered to people with Usher syndrome is now far more likely.

Retinitis pigmentosa (RP) is a progressive retinal dystrophy characterised by night blindness and a progressive peripheral field loss leading to extreme tunnel vision. As a result, in severe cases, the residual field may only be a few degrees wide. A number of secondary problems can arise. These include cataracts and macular oedema (water logging of the central retina), which may affect residual visual function. Both macular oedema and cataract may respond to treatment.

The retinopathy in Usher syndrome usually has the appearance of a typical RP. It is not possible at present to distinguish clinical types on the basis of the ophthalmic features, although some reports suggest that foveal lesions are more common in Type 1. Type 2 may be associated with a slightly milder course. Complete blindness is unusual and some people may retain vision of 6/60 or better into their fifties and sixties.

Type 1

Is characterised by congenital severe to profound sensorineural deafness affecting all frequencies. In most children, vestibular function is absent and leads to delayed motor development (poor head control, late sitting and rarely walking before eighteen months) and apparent clumsiness in later life. RP is often diagnosed earlier in Type 1, possibly because the combination of poor vision and balance problems leads to earlier functional difficulties.

Type 2

Is characterised by congenital mild to severe sensorineural deafness predominantly affecting the higher frequencies. Vestibular function is normal in childhood, and visual problems may not be apparent as early as in Type 1. Type 2 forms are more common in the UK than Types 1 or 3.

Type 3

This type has an initially mild, often late onset progressive deafness which is usually moderate to profound with a gradual decrease in vestibular function. However, the retinal appearance is indistinguishable from Types 1 and 2. This form is rare in the UK but more common in some populations; in Finnish and Ashkenazi Jewish, for example.

Changes in the retina can be detected by an ERG before the child has symptoms or is aware of problems and before changes can be seen in the retina. The time at which the ERG becomes abnormal depends upon the type of Usher syndrome. A normal ERG early on may not exclude a diagnosis of Usher syndrome.

Sense provides support and information to families and deaf people with Usher syndrome. You can find details of Sense Usher services in the 'Useful organisations' appendix 11 on page 61.

- 1 **Acanthosis nigricans:** skin disease characterised by grey-black warty patches, usually situated in the groin, elbows or knees.
- 2 **Amblyopia:** reduced visual acuity, associated with strabismus (squint) etc.
- 3 **Anaemia:** a reduction in the red blood cell pigment which may lead to pallor, weakness and breathlessness.
- 4 **Arcus:** An anatomical arch, ie an opaque line surrounding the margin of the cornea.
- 5 **Astigmatism:** occurs when the cornea/lens does not have the same degree of curvature in the horizontal and vertical planes, resulting in distorted images.
- 6 **Atresia choanae:** blockage of the nasal passages.
- 7 **Autosomal dominant (see also X-linked):** gene passed from one parent, who has the condition, to the child (1:2 chance at each pregnancy).
- 8 **Autosomal recessive (see also X-linked):** gene passed from both parents (known as carriers), who themselves do not have the condition, to the child (1:4 theoretical chance at each pregnancy).
- 9 **Binocular vision:** use of both eyes together.
- 10 **Cataract:** any opacity of the lens of the eye, resulting in blurred vision.
- 11 **Chorioretinitis:** inflammation of the retina and choroid of the eye.
- 12 **Coloboma:** a defect of eye development when a fissure that is normal but temporary fails to close up before birth.
- 13 **Conductive deafness:** where sound cannot pass efficiently through the outer and middle ear to the cochlea and auditory nerve. This can often be caused by fluid in the middle ear (glue ear). Some forms of conductive deafness are permanent; for example, meatal atresia and abnormalities of the middle ear ossicles.
- 14 **Congenital:** present at birth.
- 15 **Decibel (dB):** a unit used for measuring the intensity (loudness) of sound and plotted on a chart (audiogram).
- 16 **Diabetes insipidus:** excessive loss of water through urine and increased thirst and drinking as a result of deficiency in the hormone vasopressin.
- 17 **Diabetes mellitus:** Excessive drinking and urine output due to excess sugar in the blood and urine as a result of lack of/inability to use the hormone insulin produced in the pancreas gland.
- 18 **Electro-oculogram (EOG):** measures the potential difference between the cornea and the back of the eye. It reflects the activity of the retinal pigment epithelium (RPE).

- 19 **Electroretinogram (ERG):** records the electrical response of the retina to light. It reflects the activities of many parts of the retina including rods and cones. Using special techniques the responses of the different photoreceptors, including the rods and cones, can be separated.
- 20 **Embryopathy:** a disorder resulting from abnormal development of the embryo.
- 21 **Field of vision:** lateral and vertical extent of vision.
- 22 **Foveal/fovea:** this is the central area of the retina responsible for the best visual acuity.
- 23 **Fundoscopy, ophthalmoscopy:** examination of the retina and optic disc at the back of the eye by an instrument called an ophthalmoscope.
- 24 **Glaucoma (congenital):** raised pressure within the eye, causing sensitivity to light, excessive tearing, discomfort, enlargement of the eye, reduced vision, corneal clouding.
- 25 **Ganglion cells:** nerve cells.
- 26 **Glue ear:** see otitis media with effusion.
- 27 **Hepatospleno-megaly:** enlargement of the liver and spleen.
- 28 **Hydrocephalus:** abnormal increase in the amount of cerebral spinal fluid within the ventricles of the brain causing an enlarged head in childhood before the sutures have closed, and increased pressure within the skull in later years.
- 29 **Hypermetropia/hyperopia:** long-sightedness.
- 30 **Hypogonadism:** incompetence of the testicles or ovaries causing absence or impairment of secondary sex characteristics.
- 31 **Hypoplasia:** condition in which an organ or part is undeveloped.
- 32 **Intracranial:** in the skull.
- 33 **Jaundice:** yellow discolouration of the skin due to excess bile pigment. There are many different causes of jaundice.
- 34 **Keratitis:** inflammation of the cornea of the eye.
- 35 **Lenticonus (conical shaped lens):** rare and usually congenital condition of the lens of the eye in which the surface is conical, especially on the posterior side.
- 36 **Lymphadenopathy:** abnormal enlargement of the lymph nodes.
- 37 **Meatal atresia:** absent/extremely narrowed external ear canal.

- 38 Microcephaly:** abnormal smallness of the head.
- 39 Microphthalmos:** abnormal smallness of the eye.
- 40 Mixed deafness:** both a sensorineural and conductive deafness is present.
- 41 Myopia:** short-sightedness.
- 42 Neonatal:** occurring around birth or in the first week of life.
- 43 Neuropathy:** inflammation of the nerves.
- 44 Nystagmus:** rapid involuntary movements of the eyes.
- 45 Ophthalmoscopy:** see ‘fundoscopy’.
- 46 Optic atrophy:** degeneration of the optic nerve.
- 47 Osteopathy:** general term for bone disease or disorder.
- 48 Otitis media with effusion (glue ear):** an accumulation of fluid in the middle ear cavity, which commonly occurs in children with colds or after ear infections. With glue ear the deafness is temporary, usually mild to moderate, can affect one or both ears and fluctuate. It usually resolves spontaneously but may need specific treatment.
- 49 Ototoxic:** toxic to the ear, causing sensorineural deafness and/or balance disturbance. For example, certain medications.
- 50 Perinatal:** occurring around the time of childbirth.
- 51 Petechial rash:** minute reddish and purplish areas on the skin.
- 52 Photophobia:** abnormal sensitivity of the eyes to light.
- 53 Pigmentary retinopathy:** see retinitis pigmentosa.
- 54 Postnatal:** occurring after birth.
- 55 Prenatal:** occurring before birth.
- 56 Refraction:** the test which determines if someone needs glasses. Drops may be used when testing young people.
- 57 Refractive error (this includes short sight – myopia, and long sight – hypermetropia and astigmatism):** when the image received by the eye is blurred (unfocused) and can be corrected by lenses (for example, a magnifier), spectacles, contact lenses or other forms.
- 58 Retinal dysplasia:** abnormal development of the retina.

- 59 Retinal dystrophy:** degeneration of the retina.
- 60 Retinitis pigmentosa (RP)/pigmentary retinopathy:** group of hereditary disorders characterised by a progressive loss of photoreceptor and retinal pigment epithelium (RPE) function. Some types of RP are associated with non-ocular features (deafness in Usher syndrome).
- 61 Sensorineural deafness:** occurs in or beyond the inner ear, usually in the cochlea, more rarely in the auditory nerve. This type of deafness is permanent, ranges from mild to profound, and affects either one ear (unilateral) or both ears (bilateral) and may be progressive.
- 62 Snellen chart:** chart used for testing functional vision. A Snellen measurement is obtained by reading or matching letters on the chart. Vision is recorded as a fraction, for example, 6/6 is normal vision, and 6/60 means that the person has to be 6 metres away to see what someone with normal vision could see at 60 metres. The 3 metre LogMAR charts, a modified form of Snellen, are replacing Snellen as the 'gold-standard'. A LogMar reading of 0.0 shows normal vision. Both tests can be used with letter matching cards or plastic letters, so that the child does not have to name the letters.
- 63 Soundfield system:** an amplification system, which provides an even distribution of sound at low level around the room using a number of speakers. It improves quality of sound for all listeners and is especially useful for children with minor short term hearing variations.
- 64 Strabismus (squint):** condition in which the eyes are misaligned.
- 65 Syndrome:** a combination of signs and/or symptoms which form a clinical picture indicative of a particular disorder.
- 66 Thrombocytopenic purpura:** deficiency of blood platelets.
- 67 Usher syndrome:** see 'Usher syndrome' appendix 7 on page 50.
- 68 Vestibular:** used to refer to either the organ of balance in the ear or the function of balance.
- 69 Visual acuity:** the limit of spatial visual discrimination, commonly measured using letters or other geometrical forms.
- 70 Visual evoked response/potential:** visual evoked response of potential (VER or VEP) is a record of the electrical activity in the visual cortex of the brain in response to stimulation of the retina.
- 71 X-linked (see also autosomal dominant/recessive):** passed on the X-chromosome from the female: disease only apparent in male offspring (X-linked recessive) or in male or female (X-linked dominant).

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Appendix 11: Useful organisations

Association of Sign Language Interpreters (ASLI)

ASLI, Fortuna House, South Fifth Street, Milton Keynes, MK9 2EU

Phone: 0871 474 0522 • Textphone: 18001 0871 474 0522

Fax: 01908 32 52 59 • Email: office@asli.org.uk

Website: www.asli.org.uk

British Retinitis Pigmentosa Society

PO Box 350, Buckingham, MK18 1GZ

Phone: 01280 821 334 • Helpline: 01280 860 363

Fax: 01280 815 900 • Email: info@brps.org.uk

Website: www.brps.org.uk

Contact a Family

209-211 City Road, London, EC1V 1JN

Phone: 020 7608 8700 • Helpline (freephone): 0808 808 3555

Textphone: 0808 808 3556 • Fax: 020 7608 8701 • Email: helpline@cafamilly.org.uk

Website: www.cafamily.org.uk

Deafblind UK

National Centre for Deafblindness, John and Lucille van Geest Place, Cygnet Road, Hampton, Peterborough, Cambridgeshire, PE7 8FD

Phone: 01733 358 100 (voice and text) • Fax: 01733 358 356

Email: helpline@deafblinduk.org.uk

Website: www.deafblinduk.org.uk

Forest Books

The New Building, Ellwood Road, Milkwall, Coleford, Gloucestershire, GL16 7LE

Phone: 01594 833 858 • Textphone: 01594 833 858

Videophone: 01594 810 637 • Fax: 01594 833 446

Email: forest@forestbooks.com

Website: www.forestbooks.com

Look: UK Support for the Visually Impaired

c/o Queen Alexandra College, 49 Court Oak Road, Harborne, Birmingham, B17 9TG

Phone: 0121 428 5038 • Fax: 0121 427 9800

Email: info@look-uk.org

Website: www.look-uk.org

National Deaf Children's Society

15 Dufferin Street, London, EC1Y 8UR

Phone: 020 7490 8656 (voice and text)

Helpline (freephone): 0808 800 8880 (voice and text)

Fax: 020 7251 5020 • Email: helpline@ndcs.org.uk

Website: www.ndcs.org.uk

Royal College of Ophthalmologists

17 Cornwall Terrace, London, NW1 4QW

Phone: 020 7935 0702 • Fax: 020 7935 9838

Email: web@rcophth.ac.uk

Website: www.rcophth.ac.uk

Royal National Institute for Blind People (RNIB)

105 Judd Street, London, WC1H 9NE
Phone: 020 7388 1266 • Helpline: 0845 766 999
Fax: 020 7388 2034 • Email: helpline@rnib.org.uk
Website: www.rnib.org.uk

Royal National Institute for Deaf People (RNID)

19-23 Featherstone Street, London, EC1Y 8SL
Helpline (freephone): 0808 808 0123 • Textphone (freephone): 0808 808 9000
Fax: 020 7296 8199 • Email: informationline@rnid.org.uk
Website: www.rnid.org.uk

Scottish Association of Sign Language Interpreters (SASLI)

Baltic Chambers, Suite 317-319, 50 Wellington Street, Glasgow, G2 6HJ
Phone: 0141 202 0791 • Textphone: 0141 202 0790
Fax: 0141 202 0792 • SMS: 0779 559 7342 • Email: mail@sasli.org.uk
Website: www.sasli.org.uk

Sense

101 Pentonville Road, London, N1 9LG
Phone: 0845 127 0060 • Textphone: 0845 127 0062
Fax: 0845 127 0061 • Email: info@sense.org.uk
Website: www.sense.org.uk

Sense Usher Services

South East Regional Office, Newplan House, 41 East Street, Epsom, Surrey, KT17 1BL
Phone: 0845 127 0076 • Textphone: 0845 127 0078 • Fax: 0845 127 0077
Email: marylin.kilsby@sense.org.uk or seenquiries@sense.org.uk

Signature

Mersey House, Mandale Business Park, Belmont, Durham, DH1 1TH
Phone: 0191 383 1155 • Textphone: 0191 383 7915
Fax: 0191 383 7914 • Email: durham@signature.org.uk
Website: www.signature.org.uk

Vision 2020 UK

80 Elms Farm Road, Hornchurch, Essex, RM12 5RD
Phone: 01708 456832 • Fax: 01708 446310
Email: m.brace@vision2020uk.org.uk
Website: www.vision2020uk.org.uk

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Sense is the national charity that supports and campaigns for children and adults who have a combination of sight and hearing difficulties. We provide expert advice and information as well as specialist services to deafblind people, their families, carers and the professionals who work with them. In addition, we support people who have sensory impairments with additional disabilities.

Our services include:

Outreach services

Specialist workers who provide support to deafblind children and their families, adults and older people living in the community. They offer assessments, information, guidance, support and help people to access the services they are entitled to.

Branches

Sense branches bring together deafblind children, young people and adults, family members and carers to learn from and support each other, to campaign for local services, to raise money and to have fun.

Holidays

Sense holidays give deafblind people the chance to get away from it all, taste new experiences and make new friends. They also give families a much-needed break from the often demanding work of supporting a deafblind person.

Supported housing

Sense's supported housing ranges from houses where individuals have very high support needs, to accommodation where people require a lower level of support to live independently.

Day services

Sense runs a range of day services and resources centres around the UK – where deafblind individuals are supported to choose activities and programmes that help them to develop their skills and abilities.

Communicator-guides

Communicator guides offer communication support, guiding skills, and practical help – such as help with shopping or dealing with mail – to enable deafblind people to continue to live independently.

Membership

Sense Membership offers people a sense of belonging, the chance to be more actively involved in our work, plus the opportunity to receive our magazine, Talking Sense.

NDCS is the national charity dedicated to creating a world without barriers for deaf children and young people. We represent the interests and campaign for the rights of all deaf children and young people from birth until they reach independence.

NDCS provides the following services through our membership scheme. Registration is simple, fast and free to parents and carers of deaf children and professionals working with them. Contact the Freephone Helpline (see below) or register through www.ndcs.org.uk

- A Freephone Helpline 0808 800 8880 (voice and text) offering clear, balanced information on many issues relating to childhood deafness, including schooling and communication options.
- A range of publications for parents and professionals on areas such as audiology, parenting and financial support.
- A website at www.ndcs.org.uk with regularly updated information on all aspects of childhood deafness and access to all NDCS publications.
- A team of family officers who provide information and local support for families of deaf children across the UK.
- Advice on special educational needs (SEN) disability discrimination in education and welfare benefits.
- Representation at SEN and Social Security Appeals Tribunals from our Legal Casework Service.
- An audiologist and technology team to provide information about deafness and equipment that may help deaf children.
- A children's equipment grants scheme and the opportunity to borrow equipment to try out at home.
- Family weekends and special events for families of deaf children.
- Sports, arts and outdoor activities for deaf children and young people.
- A quarterly magazine and regular email updates.
- An online forum for parents and carers to share their experiences, at www.ndcs.org.uk/parentplace.

Vision care for deaf children

The National Deaf Children's Society (NDCS) and Sense, the National Deafblind and Rubella Association, have produced these quality standards and good practice guidelines.

Our goal is drive up standards in vision care for deaf children and young people, and to give busy professionals a framework for providing the best possible support and long-term care.

Deaf children are especially dependent on their sight in order to find their way in the world. They are also more likely to have visual difficulties than children without hearing impairments.

These guidelines stress the importance of an ophthalmic examination following confirmation of deafness – and provide helpful tips about how to offer effective vision assessments for children with hearing impairments.



Tel: **0845 127 0060** • Fax: **0845 127 0061**
Text: **0845 127 0062** • Email: **info@sense.org.uk**
www.sense.org.uk



NDCS Freephone Helpline: **0808 800 8880**
(voice and text) Open Monday to Friday, 9.30am to 5pm
Email: **helpline@ndcs.org.uk**
www.ndcs.org.uk

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NDCS is the national charity dedicated to creating a world without barriers for deaf children and young people.

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