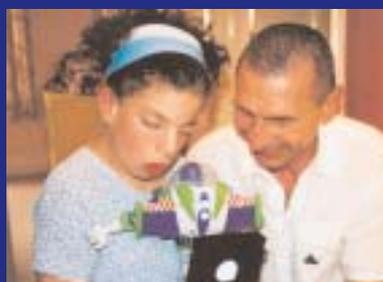


# Vision care for deaf children and young people

Guidelines for professionals working with all deaf children



Comments from  
parents

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

*“We always tell them that our son cannot hear, but there is no attempt to help him or to stand so that he can see them talking. They talk to us instead of him”*

*“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”*

*“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 all deaf children

The National Deaf Children's Society (NDCS) and Sense, The National Deafblind and Rubella Association have jointly produced 'Quality standards in vision care for deaf children and young people'. The document provides a valuable tool and reference point for all service providers and funders of services to help them to drive up standards in vision care for deaf children and young people, and aims to give busy professionals a framework in which to provide the best possible support and long-term care.

The document identifies 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.

Most of the information a child will use to learn about the world around them comes through their ears and their eyes. This means that for the deaf child eye care plays a vital role, and they will be especially dependent on vision in order to acquire language and develop relationships with others.

It is of utmost importance that vision difficulties are picked up as early as possible, so that appropriate treatment and support can be offered. Yet there is little clinical guidance on good vision care, and the crucial role that multi-disciplinary professionals will play in quality eye care for the deaf child and young deaf person. These guidelines stress the need for joined up working between services and service professionals to ensure the best delivery of care.

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

We would like to thank everyone involved in the development of this document, which we are confident will help both NDCS and Sense fulfil their ambitions for all deaf children and young people to develop their full potential. The guidelines were developed over 5 years, with crucial input from ophthalmology, audiology, and community professionals, parents and lay people involved with deaf children.

**Susan Daniels**, NDCS Chief Executive

**Tony Best**, Sense Chief Executive



# Contents

<b>Part one</b>	<b>Introduction</b>	
	Key messages	4
	Figure 1: Definitions and key words	5
	Deaf children in the UK	6
	Glue ear (otitis media with effusion)	7
	Newborn hearing screening	7
	Figure 2: Degrees of hearing loss	8
	Figure 3: Key research findings	9
<b>Part two</b>	<b>Empowering parents and young deaf people</b>	10
	The need for multi-disciplinary working	11
	The lead clinician	13
	The key worker	13
	Recommended additional training: vision care personnel	13
<b>Part three</b>	<b>The local audiology service</b>	
	The child's ongoing review by the local audiology service	15
	The referral process	15
<b>Part four</b>	<b>Assessment and care by eye specialists</b>	
	Before the visit for eye assessment	17
	The visit	17
	Figure 4: Booking sign language interpreters and other communicators	18
	Diagram 1: Suggested room layout/positioning when assessing a deaf child	19

<b>Part five</b>	<b>The ophthalmic assessment</b>	20
	Figure 5: Good testing techniques for the deaf child	21
	The ophthalmic care pathway for the deaf child	22
	At birth – routine screening programme	22
	At diagnosis of deafness	22
	At 2-4 years	23
	At 4-5 years - routine vision screening in the 'general child population'	23
	At 7-9 years	23
	At 10 years and above	23
	At referral for cochlear implantation and other complex interventions	23
	Local optometry services	24
	Figure 6: Useful questions when assessing night vision, field loss, etc.	24
	Following the visit	25
	Figure 7: Practical points when working with families	26
	Diagnosis of a serious eye condition	26
	Hearing aids (including cochlear implants), spectacles and eye patches	27
	Deaf children with additional needs	28
	Deafness as part of a syndrome	29
	The clinical genetic service	29
	Meeting the needs of ethnic minority families	30
	Difficult-to-reach families	30
<b>Part six</b>	<b>Young people and transfer of care to another service</b>	31
	Transfer to an alternative paediatric/adolescent service	31
	Acknowledgements	32
<b>Part seven</b>	<b>Appendices</b>	
	1 Communicating with deaf children	33
	2 Key personnel	35
	3 Conditions affecting both hearing and vision	37
	4 Usher syndrome	39
	5 Useful organisations	40
	6 Useful publications	42
	7 Glossary	43
	8 References	47

# Part one

## Introduction



The majority of information a child will use to develop understanding and to learn comes via vision and hearing – with vision as the co-ordinator. In the case of the deaf child, vision plays a more significant and compensatory role as the gateway for acquiring language and the skills to communicate effectively. It is important to recognise any impairment of vision as early as possible in the child's life to ensure there is an effective clinical pathway.

Currently there is little clinical guidance on good vision care for deaf children. The aim of this document is to enable professionals to review the ophthalmic care of the deaf child and, where appropriate, develop services to meet the individual needs of the child, young person, and their family.

**The Royal College of Ophthalmologists (2002) recommends that the visual behaviour of all children should be observed. This may be carried out by health or educational professionals (which can include the health visitor, the child's GP or other doctor).**

These guidelines have been produced, in close co-operation with families and with professionals working in health and education, to provide guidance on how the vision of deaf children should be assessed. The document identifies the optimal stages in a deaf child's development when assessments should be carried out, who should carry them out, and the type of assessments needed.

**Health for All Children states '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' (Hall & Elliman, 2002).**

Quality standards are identified to help evaluate and monitor services. This document will enable those responsible for planning and setting up services to bid for funds to develop good practice in working with deaf children and young deaf people.

### Key messages

- All deaf children should have an ophthalmic examination following confirmation of their deafness and they should be reviewed at key stages of their development.
- All deaf children need an ophthalmic assessment not only if there are concerns but also to detect conditions which have no obvious signs or symptoms.

‘Forty per cent of children with sensorineural hearing impairments have eye problems’

- A vision assessment of a deaf child will require extra preparation and an understanding of that child's particular needs. A carefully planned assessment will contribute to positive experiences for the child and their family.
- Quality services have a culture of learning continuously from families, service users and colleagues.
- Parents, with their intimate knowledge of their child, should play a key role in the multi-disciplinary team and work in partnership with the professionals involved in their child's care.
- Effective multi-disciplinary collaboration and information sharing will ensure better standards in vision care for deaf children.

These quality standards and good practice guidelines will be monitored and revised by 2008, therefore we welcome comment and feedback from all users of these guidelines.

### **Figure 1: Definitions and key words**

**Deaf:** refers to sensorineural and permanent conductive hearing loss of any degree, including unilateral hearing loss. This includes born deaf children and those with later onset of deafness.

The phrase '**all deaf children**' includes children with complex needs.

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

**Partial sight:** is defined as a visual acuity of between 6/24 and 6/60 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.

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

**Visual acuity or eyesight:** is a measure of how well a person is able to distinguish fine detail with adjacent visual stimuli, such as the features of a letter.

**Deafblind:** is used where the 'combined sight and hearing impairment cause difficulties with communication, access to information and mobility' (Department of Health, 1995).

The words '**parent**' and '**family**' include the child's carer who has 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 hearing loss. A recent study by Fortnum et al (2001) estimated that a further 0.6-0.9 per 1000 children will develop hearing loss by 10 years of age (due to illness, progressive hearing loss 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 & Jensema, 1977; Schein & Delk, 1974). In half of these cases the causes of permanent hearing loss include prematurity, congenital infections, prenatal, perinatal and postnatal illness and craniofacial abnormalities.

**'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).**

Research has shown that there is a higher incidence of ophthalmic problems amongst deaf children (Guy et al, 2003; Armitage, Burke & Buffin, 1995; Leguire et al, 1992). For conditions that affect both hearing and vision see Appendix 3. These higher rates of ocular pathology have been attributed to:

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

If a deaf child has a problem with their vision it is essential that this is identified and treated as soon as possible. Those identified with ocular abnormalities will require long-term ophthalmological review. Brinks et al (2001) found this is often overlooked. He reported that 63% of deaf students aged 10-22 had significant ocular pathology and had not seen an ophthalmologist in the previous 2 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.

*'If a deaf child has a problem with their vision, it is essential that this is identified and treated as soon as possible'*

Where there are more serious eye conditions (deafblindness), 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 educational support children can be helped to fulfil their potential. Some local education authorities employ specialist teachers to work with children and young people with multi-sensory impairment (MSI) as part of their sensory support team.

### **Glue ear (otitis media with effusion)**

Glue ear in children is one of the most common audiological childhood conditions. In most cases glue ear will be short term and should not require assessment of vision. However, a child with persistent glue ear, which has a significant effect on their hearing, should have their vision assessed. Most of these children are unlikely to 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.

### **Newborn hearing screening (see [www.nhsp.info](http://www.nhsp.info))**

Newborn hearing screening is being introduced, and it is anticipated that it will be implemented throughout the UK by 2005.

Newborn babies will have their hearing screened during the first few weeks after birth. If a baby does not show a clear response to the screen they will be referred for audiological assessment. It is anticipated that babies born with a permanent hearing loss will be identified by eight weeks of age, although sometimes the hearing loss may take longer to confirm. Newborn hearing screening will make 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 (DfES, 2003).



## Figure 2 Degrees of hearing loss

The following terms are used to define the degree of hearing loss. This is a general guide only, and does not describe the impact of the disability on the child\*.

- **Normal range (< 20dB Hearing Level (HL))**

All speech sounds should be heard.

- **Mild hearing loss (21–40dB HL)**

The child is unlikely to have difficulty hearing a one-to-one conversation, particularly if listening at close range. They may find it difficult to understand softly spoken speech especially from a distance or if there is any background noise. Preferential seating is recommended in the classroom. Children with a mild hearing loss may benefit from hearing aids. In the classroom a soundfield system may also be useful.

- **Moderate hearing loss (41–70dB HL)**

Conversational levels of speech are usually difficult to hear, even in a quiet environment. Listening in noise will be extremely hard. Spoken language will be heard with the use of hearing aids. Children with this degree of loss usually hear their own vocalisations and loud environmental sounds.

- **Severe hearing loss (71–95dB HL)**

Normal conversational speech cannot be heard without amplification. Speech may not develop spontaneously, although it can do so with appropriate intervention. Some children may use sign language, with or without hearing aids.

- **Profound hearing loss (> 95+ dB HL)**

The child is unable to hear speech and possibly only very loud environmental sounds without amplification. Speech development may be absent or very delayed unless hearing loss is recognised early. With appropriate amplification they may be able to access some speech and environmental sounds. Some children may use sign language to communicate, with or without amplification.

\* 'Understanding Deafness' (2003), produced by the NDCS, is a useful booklet for those who would like to know about childhood deafness.

### Figure 3: Key research findings

In 2002 NDCS and Sense commissioned an evidence-based overview of ophthalmic disorders in deaf children. Over 1000 relevant research papers were identified, of which 190 were analysed in detail. A summary of the research is being prepared (NDCS/Sense, in preparation).

The notion that when a person is found to be deaf, a vision impairment should be suspected, is not new. Over 70 years, studies on different populations of deaf children have shown consistently a high level of vision impairment. However, differences in definitions and classifications of vision impairment make comparison between the various studies difficult.

Nevertheless, 30% to 60% of deaf children with significant vision impairment is a consistent finding, for example:

- Burdge (1933) found that 40% of deaf children tested needed spectacles, and Braly (1938) reported that 38% out of 422 deaf children had less than 20/20 vision on the Snellen chart.
- Stockwell (1952), in a study of 960 deaf children, reported that 46% needed spectacles.
- Alexander (1973) found that out of 572 children, 35% had refractive errors and 22% had other ocular pathology.
- Regenbogen and Godel (1985) showed, in a comparative analysis with the study by Coleman (1970) on 3623 hearing children, that more than two and a half times as many deaf children had eye abnormalities.
- Rogers et al (1988) screened 360 deaf children and young people aged between 6-22 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 child who is not deaf and that a comprehensive examination should be performed at identification of deafness.
- More recently 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.
- 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.

# Part two

## Empowering parents and young deaf people



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 'lead clinician' definition see page 13).

Parents have a right to information so that they can make informed choices for their deaf child. Information should be clear, accurate and accessible, and provided in spoken and written form, for example leaflets that can be taken away (Department of Health, 2003). Information should also be available in other languages and formats such as large print, audio or video tape or Braille.

### *Quality standard*

**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, 2003).

Professionals should continually question and evaluate their communication strategies to ensure that the information being shared is understood. In an NDCS/Sense survey of parents of deaf children (2001) 88% were satisfied with the information 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 eye care staff were unaware of or insensitive to the communication needs of their deaf child.

**'Children and young people should receive care that is integrated and co-ordinated around their particular needs, and the needs of their family. They, and their parents, should be treated with respect, and should be given support and information to enable them to understand and cope with the illness or injury, and the treatment needed. They should be encouraged to be active partners in decisions about their health and care, and, where possible, be able to exercise choice.'**  
(Department of Health, 2003).

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

In order for parents to remain in control and to make effective decisions about their child's care, two-way communication with professionals working with their child is crucial. 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 made available.

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 (Sense, 1999). The lead clinician should ensure that young people are given the telephone/text numbers and address of the social worker with deaf people and other key personnel so that they can make contact for advice if their vision impairment causes practical problems in their social and educational development.

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; HMSO, 1996; SOEID, 1996).

### *Quality standard*

**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.**

## **The need for multi-disciplinary working**

(see appendix 2 for key personnel)

**'The evidence strongly favours a holistic health-promoting approach, which crosses agencies and disciplines, rather than a narrow defect-detecting programme.'** (Hall & Elliman, 2002).

Parents are part of the multi-disciplinary team and will be involved with a number of professionals from different disciplines. Good communication between personnel within the team will help to detect any signs of deterioration in the child's vision and allow their needs to be addressed holistically.

Multi-disciplinary working can also reduce disruption to family life. Clear communication between agencies should reduce the number of meetings and appointments that parents have to attend, for example joint visits (Department of Health, 2003).

Each professional should:

- be informed of the possible signs/symptoms and significance of developing eye problems
- be aware of the referral procedure and know who to refer to if there is concern about the child's vision
- be aware of issues relating to a dual/multiple sensory impairment (see pages 26 and 28).

‘Clear communication between agencies should reduce the number of meetings and appointments that parents have to attend.’

#### Quality standard

**Every member of the multi-disciplinary team must be aware of the importance of ophthalmic care in deaf children and issues relating to dual sensory impairment.**

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 co-ordinated 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*

*Miss Lucy Butler, Consultant Ophthalmologist, Birmingham*

*\*Usher syndrome, see Appendix 4*

## The lead clinician

### *Quality standard*

**Each child must be appointed an identified lead clinician and the parents informed.**

The lead clinician is responsible for co-ordinating all aspects of the care and management of the deaf child, including their eye and vision care. They are also responsible for ensuring that all involved professionals communicate effectively with the child and family. The lead clinician may be, for example, the community paediatrician, or the audiological physician, or another clinician, depending on local circumstances (Department of Health, 2003).

The lead clinician has a duty to ensure that the child is known to the child health team, the education service, and others working with the child. It is the responsibility of education and health professionals to express concerns regarding a child's vision to the lead clinician.

## The key worker

### *Quality standard*

**Each child and family must be assigned a key worker as soon as the child's deafness has been identified.**

The lead clinician should ensure that a key worker (sometimes called a link worker) is assigned to the child and family (Hall & Elliman, 2002) as soon as deafness has been identified. This can be any key member of the multi-disciplinary team, for example the teacher of the deaf.

The key worker plays a crucial role in supporting the family, 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.

## Recommended additional training: vision care personnel

**All staff treating or caring for children and young people should have appropriate training, and should undergo regular updating and refreshment of skills. This training should cover both the technical clinical skills and the personal and communication skills necessary to treat children and their parents.**

*(Department of Health, 2003).*

To ensure a responsive child and family-centred service, training in issues relating to childhood deafness, deaf awareness, sign language and finger spelling is essential.

Details of approved sign language and finger spelling courses are available from organisations such as the Council for the Advancement of Communication with Deaf People (CACDP)

Other areas in which staff can receive training include:

- developmental implications of a congenital hearing loss
- language development in deaf and hearing impaired children
- using an interpreter
- creating an uncluttered environment, including lighting and glare control
- using a text phone/Typetalk (Sense, 2002).

For information about courses, contact the local education authority specialist services for deaf and hearing impaired children. Further education colleges may offer sign language courses, which will include deaf awareness issues.

### *Quality standard*

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



# Part three

## The local audiology service

### The child's ongoing review by the local audiology service

Audiologists should be aware of the importance of optimal vision and be responsive to any concerns about eyesight expressed by parents or school staff. The following check list can be used with the child or parent by staff responsible for the deaf child, for example the audiology physician, community paediatrician or school doctor, even if the child has been discharged from the eye clinic.

Does the child:

- have difficulty seeing the black/white board in the classroom?
- have more than usual problems seeing in poor light?
- become unusually disorientated in the dark?
- have difficulties reading a book or magazine?
- have problems seeing the scenery from the back of a car?
- have problems spotting a person in the crowd?
- have any discomfort in bright light?
- trip over things more frequently than other members of the family?
- have difficulty going from a light to a dark environment?
- blink frequently?
- rub her/his eyes frequently?
- have difficulty finding small items which have fallen onto the floor?



### The referral process

#### *Quality standard*

**The child should be referred for ophthalmic assessment at any time if parents or the education service have concerns (see pages 20-23).**

If the child has not been referred for full ophthalmic assessment, 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 reason for the referral (e.g. to address worrying symptoms or to look for signs of an identified disorder or as part of routine screening of all deaf children)
- the nature, onset, cause (if known) and degree of deafness
- any general developmental and health concerns
- the concerns of parents, nursery or school staff, teacher of the deaf, health visitor and other healthcare professionals involved with the child and/or the concerns of the child
- parents' and/or child's special needs so that the clinic can plan/make the necessary arrangements in advance, for example planning the assessment and the language/phrasing that will be used or booking a sign and/or language interpreter.

### *Quality standard*

**The eye clinic should be informed of the family's communication preferences so that suitable provision can be organised (Department of Health, 2003).**

For communicating with deaf children see Appendix 1.



# Part four

## Assessment and care by eye specialists

### Before the visit for eye assessment

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 Figure 5, page 21).



#### *Quality standard*

**Written information about what the visit to the clinic will involve, how long it is likely to take and the effect of the eye drops should be given to the family before the clinic appointment and be suitably worded for the family and in a format the older child/young person can understand.**

To make good use of the appointment and reduce any anxiety in the child and parents 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.

Written information needs to be suitably worded for parents and age-appropriate for the child. There are a number of children's books on the subject of eye tests which can be helpful (see Appendix 6).

For children with multiple impairments, if standard procedures are inappropriate, establish how the test can be carried out (see 'Deaf children with additional needs' page 28).

A preliminary visit to the assessment centre can be beneficial to familiarise the child.

### 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.

Other issues to be addressed are:

- check whether hearing aids or a cochlear implant are being used
- check the level of spoken language that the child understands

*‘Speak directly to the person being tested, but allow time for the parent or interpreter, if present, to explain’*

- if appropriate, ask the parent/child whether they wish to use their radio aid or the loop system, if this would assist communication
- speak clearly using language at an appropriate level without covering the mouth, shouting, or over-enunciating
- speak directly to the person being tested, but allow time for the parent or interpreter, if present, to explain
- the interpreter will relay everything spoken in the room. Talk through the interpreter, not to them
- create uncluttered backdrops so that the child can focus on the face in order to lip-read more easily and/or to see sign language
- ensure that the light falls on the face of the person talking (see Diagram 1)
- cut out glare from white/shiny surfaces or bright sunlight.

#### **Figure 4 Booking sign language interpreters and other communicators**

Booking sign language interpreters and other communicators can be done through agencies or directly with individuals offering services on a freelance basis. It is important to book any interpreters for sufficient time 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. Check what type of sign language is preferred, for example British Sign Language (BSL), sign supported English (SSE), etc.

- Booking through agencies usually involves one phone call to give details of the request. The agency will make the booking and charge a fee for their services. A disadvantage is that the request may not receive the urgent attention the booking might require. Some agencies offer a complete range of BSL/SSE/visual frame/tactile signing/deafblind manual interpreters, notetakers, lipspeakers, palantype and speech to text services, while others may only focus on one or two skills. Agencies are listed in the Council for the Advancement of Communication with Deaf People (CACDP) 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: \*CACDP, \*ASLI (the Association of Sign Language Interpreters), and \*SASLI (the Scottish Association of Sign Language Interpreters). Some freelance interpreters will be listed in all three.
- Specialist services. Some agencies specialise in short notice booking and interpreters for deafblind people: the CACDP directory lists registered deafblind interpreters.

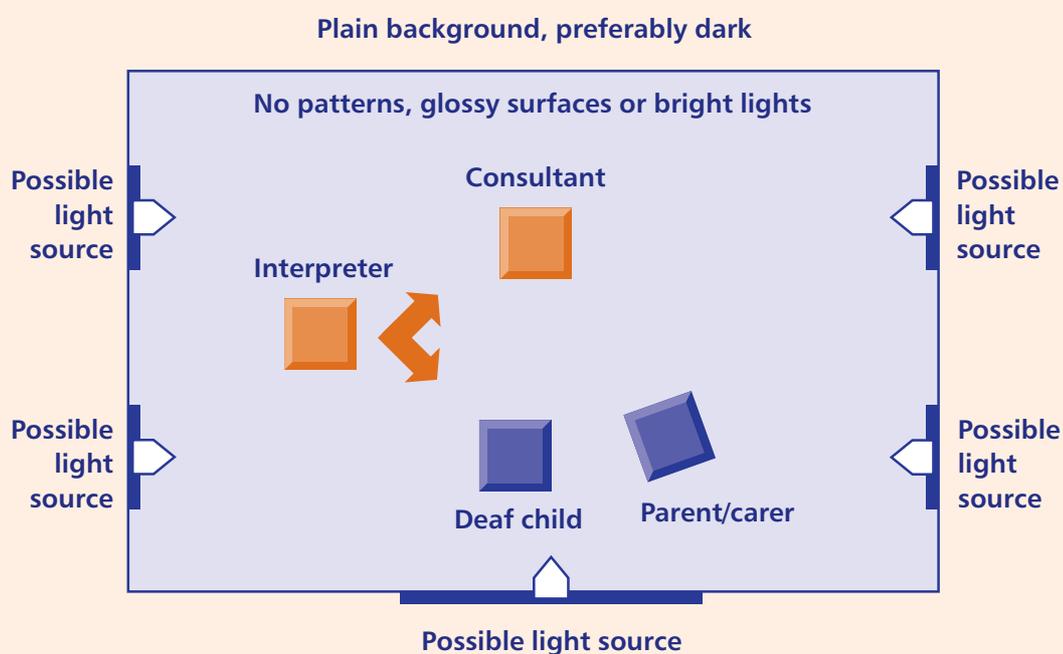
\*Directories are available from:

CACDP: tel: 0191 383 1155, text: 0191 383 7915, fax: 0191 388 7914,  
web: www.cacdp.org.uk. Or from the Forest Bookshop: tel/text: 01594  
833858, fax: 01594 833446.

ASLI: PO Box 32152, London N4 2YZ.

SASLI, Donaldson's College, West Coates, Edinburgh EH12 5JJ.

### Diagram 1 Suggested room layout/positioning when assessing a deaf child



#### Notes

- 1 Use chair with wheels for interpreter for 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.

# Part five

## The ophthalmic assessment



### Quality standard

**'All members of the ophthalmic team should be confident that they have the appropriate skills and expertise before managing any child. Any member of the ophthalmic team who does not spend a significant proportion of their time in management of children is discouraged from participation in children's eye care'** (Royal College of Ophthalmologists, 2002).

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 the deaf child to an ophthalmologist if the eye examination is abnormal or if the deaf child has not previously seen an ophthalmologist.

The consultant ophthalmologist is responsible for the ophthalmic assessment of the deaf child. Ideally the consultant ophthalmologist will have experience of working with children.

At the ophthalmic assessment a full history should be taken and a thorough eye examination performed (see figure 5, page 21). Certain eye findings, for example the presence of a retinal dystrophy, may help to identify the cause of the 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.

The full clinical history should include specific questions about parental (or child) concerns about the child's vision, the cause of their hearing loss, and other indicators such as problems with night vision, delayed onset of walking, etc. A family history of relevant eye or hearing problems should be sought. The comprehensive examination will also include:

- visual acuity
- refraction
- visual field assessment
- ophthalmic examination.

An electroretinogram (ERG) should be performed in:

*‘Referral pathways for deaf children should differ from those recommended for the general child population’*

- children who have any symptoms suggestive of Usher syndrome including night blindness or who have field loss or unexplained reduction in visual acuity
- children with unexplained profound congenital sensorineural hearing loss and delayed motor milestones (walking beyond the age of 18 months or possibly earlier in Afro-Caribbean or Asian children)
- children with retinal dystrophy.

All children with unexplained congenital bilateral profound or sloping moderate to severe sensorineural hearing loss should have ERG testing undertaken between the ages of 7 to 9 years to assess for Usher syndrome (see page 23).

An ERG is performed without anaesthesia. As much of the testing is performed in the dark, it is crucial that the child and parents receive a clear explanation of the procedure in a well lit room before starting the test. 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. Therefore, the child should be referred to a centre with these facilities.

An ERG should be considered in those children with late onset progressive sensorineural hearing loss in case of Usher type 3, Refsum disease, etc.

### **Figure 5 Good testing techniques for the deaf child**

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

- avoid holding orthoptic fixation targets in front of your face. Adopt a different position so that the child can lip-read and have a full view of your face to observe expression and gesture
- 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
- before testing in the dark, and whilst the surroundings are well lit, ensure that the child understands what to do. Only when you are sure that the child understands should you darken the room. Allow the child to sit next to someone familiar to them during the test to reassure them
- lip-reading and sign reception may be restricted after eye drops are inserted. It is, therefore, important to explain the short-term effects to the child and the parent before this happens
- 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, thumbs-up etc.) will help to reassure the child. Encourage others in the room to do the same thing.

## The ophthalmic care pathway for the deaf child

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

**'At any age, when parents, health visitors, GPs, paediatricians or clinical medical officers are concerned about visual disability or ophthalmic disease, including manifest squint, urgent referral should be made to an ophthalmologist'**  
*(Royal College of Ophthalmologists, 2002).*

### At birth – routine screening programme

All newborn babies will have their eyes examined as part of the routine screening programme. Babies with identified disabilities should be routinely examined by the paediatric team for congenital abnormalities that may have ophthalmological or audiological implications.

By 2005, all babies should have their hearing screened within the first few weeks after birth.

### At diagnosis of deafness

#### *Quality standard*

**Following the identification of a permanent hearing loss the child should be referred for a full ophthalmic assessment, and where appropriate at key stages of their development.**

The majority of children with a congenital sensorineural hearing loss will be identified soon after birth. However, it is important that parents and professionals are aware that some children will acquire a hearing loss or develop late onset deafness in later childhood.

#### *Quality standard*

**It is the responsibility of the ophthalmologist to ensure that all staff involved in the screening programme are aware of when and to whom to refer a deaf child.**

## At 2-4 years

Children with bilateral profound congenital sensorineural hearing loss of unknown aetiology should be re-referred to the paediatric ophthalmologist for assessment. 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 4 to 5 years – routine screening in the general child population

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. 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.

## At 7 to 9 years

Children with a sensorineural hearing loss, where the cause of the deafness is unknown should be re-referred to ophthalmology where they can be tested for Usher syndrome (see Appendix 4) using electroretinography (ERG). An ERG can be performed at any age and there are a number of conditions for which this test is useful.

Presently, there is no guidance on the optimal age for testing for Usher syndrome, and seven years of age is considered the most appropriate. However, a normal ERG at this age may not exclude type II Usher syndrome.

## At 10 years and above

Since myopia may increase during the adolescent years, all deaf children should have their visual acuity re-assessed at this age by an optometrist. If there are any concerns regarding night vision, field loss or visual loss, that is not corrected by spectacles, the child should be referred for a full ophthalmological assessment.

## 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 they should ensure that the child has had their vision checked as part of their assessment. If there has not been an ophthalmic assessment they should arrange for the child to be seen by the ophthalmology service as soon as possible.

## Local optometry services

Some children with permanent childhood hearing impairment will be seen by their local optometrist who may prescribe corrective lenses. All children should be:

- referred to the local ophthalmologist for further evaluation
- 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).

Where no refractive error is present during the school years, young deaf people should be encouraged to refer themselves for optometric testing every two years (see section 6).

### *Figure 6 Useful questions when assessing night vision, field loss etc.*

Observations by health visitors, teachers, care staff, as well as parents can help build up a picture of how the vision of the child is functioning. Children themselves can be excellent informants if they are asked language appropriate questions in an approachable manner.

- Does the child reach for someone's arm or the handrail after dark or in low light?
- Does the child have worse balance after dark?
- Is the child unduly sensitive to bright light, glare and shiny objects?
- Does the child explore the floor with her/his feet before going upstairs?
- Does the child have frequent mishaps, such as bumping into lamp posts, falling over low-lying objects, misjudging the depth of steps?
- Does the child have difficulty tracking moving objects, such as a ball?
- Does the child withdraw from ball games?
- Can the child distinguish between black and brown?
- Is the child startled by people or objects close to her/him?
- Does the child blink or rub her/his eyes frequently?

## Following the visit

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 and professionals. Age-appropriate information will help the older child or young person to understand their vision care and any implications it may have (Department of Health, 2003).



### *Quality standard*

**Ideally, copies of assessments, diagnostic results and what they mean should be sent to parents within ten working days of the clinic visit. Some results may take longer and parents should be made aware when they can expect to receive them.**

A copy of the report should be sent to the referrer, the parent and/or child, the GP and lead clinician, with the parents and child's agreement. This could include:

- identified pathology
- the management plan
- implications for the diagnosis and management of the hearing loss or other conditions
- information given to parents and (where relevant) age appropriate information to the child
- 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 hearing problems. Where there are implications arising from the vision assessment a meeting should be arranged with key personnel to modify the plan.

### *Quality standard*

**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. Parental consent is needed to share information within the multi-disciplinary team.**

*‘Some distress can be alleviated by the manner in which the diagnosis, subsequent follow-up help and support is given’*

### **Figure 7 Practical points when working with families**

- Talk about the baby or child 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.
- Families of deaf children may not be familiar with terminology relating to vision impairment. In order to check understanding, it may be helpful to ask parents to explain in their own words what they understand the diagnosis or disability to be. Any misunderstandings are likely to become apparent, which can then be clarified (Thameside Community Healthcare, 2000).
- 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.
- Single parent families may wish to bring a relative or friend with them.
- Parents need time to absorb any diagnosis or condition their child has and given the opportunity to meet with professionals when they need to.
- Brief, written, easily understood information about the child’s condition should be provided. Written information does not reduce the need of the professional involved to discuss things with parents.
- Provide a private, family-friendly area (Department of Health, 2003).
- Encourage parents to contact specialist agencies for advice and help. Parents should be provided with relevant information about agencies and services (see Appendix 5, ‘Useful organisations’).

### **Diagnosis of a serious eye condition**

Where the 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 extreme 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 (Thameside Community Healthcare NHS Trust, 2000; RNIB, 1997). Some distress can be alleviated by the manner in which the diagnosis, subsequent follow-up help and support is given. Deaf parents may not feel negatively about their child’s deafness. However, they could be unduly shocked by a diagnosis of a visual problem.

Information given to parents at the initial consultation will need further discussion at a later stage, when parents and 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.

### **Quality standard**

**Ideally a follow-up appointment within two weeks should be offered 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.**

Counselling should be available soon after diagnosis to enable parents and the child 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 a long time and may themselves continue to be distressed, especially if their child's vision/hearing/other condition continues to deteriorate. Recognising this should allow for professional support to be extended to families who need time to find their own way to adjustment and disclosure.

## **Hearing aids (including cochlear implants), spectacles and eye patches**

(for further information about hearing aids and their function see the NDCS booklet on 'Hearing aids: a guide', 2003).

For a child who is already wearing hearing aids or a cochlear implant processor 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 processor that sit behind-the-ear, and with advances in technology aids have become smaller and slimmer. However, whether there is enough room behind the child's ears to fit hearing aids and spectacles 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. For older children and young people there may be the option of wearing contact lenses or in-the-ear (ITE) hearing aids (if their hearing loss is less severe).

It is possible to get hearing aids that are combined into the arm(s) of spectacles. 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 spectacles 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 their aids during this time. Cochlear implant processors have not, as yet, been combined into the arm of spectacles but the same considerations apply.

Patching the good eye in an amblyopic deaf child may present particular challenges especially if they use lip-reading or sign support as an integral part of their communication.

### Deaf children with additional needs

Up to half of all deaf children may have an additional disability (Cherow 1993, Fortnum et al 1996, Stredler-Brown & Yoshinago-Itano, 1994). While all children should have detailed audiological and vision assessments, carrying out these assessments may be complex and time consuming. A preliminary visit to the assessment centre can be beneficial to familiarize the child.

Some LEAs employ specialist teachers of deafblind or multi-sensory impaired (MSI) children 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.

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 recognize objects through touch rather than by sight. Some children have difficulty seeing objects against complex backgrounds.

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

- consult those who know the child, for example the health visitor and/or specialist teacher
- identify motivators (i.e. 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, e.g. a resonance board to show that a visual signal is coming
- develop strategies for relaxing or arousing a child who is over-reacting to certain stimuli or unresponsive to what is happening around them
- position the child so that they are free to use their vision, e.g. allow the child to lie down rather than sit at a table.

Vision for doing (Aitken & Buultjens, 1992) is a useful guide to vision assessment in children with MSI. ‘Eye to eye’, measuring and describing vision in young children (Woodhouse, 2002) is also helpful.

‘Up to half of all deaf children may have an additional disability’

### *Quality standard*

**The clinician carrying out the assessment should be accurately informed of the child's special needs before the assessment.**

## **Deafness as part of a syndrome**

Syndromic deafness (i.e. deafness associated with other features, such as vision impairment, kidney abnormalities etc.) accounts for 20-30% of childhood deafness (Steel, 1999). Full diagnosis is important as there may be associated medical problems or special educational needs for the child (Guest, 2000).

If the ophthalmologist diagnoses a deaf child with a genetic condition, the family (and where relevant the child) should be offered referral to a clinical genetic service.

## **The clinical genetic service**

Where vision or dual sensory impairment is thought to be inherited or the cause is unknown, it is the responsibility of the lead clinician, with parental consent, to offer a referral to the genetic service. If this is declined, the family's wishes should be respected.

If a deaf child is diagnosed with a genetic condition, any sibling who is also deaf may have the same syndrome 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, especially in families where there is consanguinity.

Young people who wish to talk about their inherited condition, or the implications for themselves of a sibling's condition, can be referred to the genetic service. If the family is already known to the service, they can make the referral themselves.

One of the roles of the clinical genetic service is to identify and give information about conditions that may be inherited. DNA testing is only available for a few genetic conditions and most of these are not routine but have to be individually arranged by the consultant in clinical genetics. However, at present, for the majority of genetic conditions there may be no test because the gene(s) has/have not been identified. 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 the medical literature.

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

### *Quality standard*

**If the vision impairment or dual sensory impairment is unexplained or suspected to have a genetic cause, the lead clinician is responsible for ensuring that genetic testing and counselling are offered to the family and, if accepted, arranged.**

## Meeting the needs of ethnic minority families

There is a higher incidence of deafness among some ethnic minority families. Family-centred services should take both linguistic and cultural diversity into account. It is acknowledged that it can be difficult to access sign language interpreters from diverse ethnic groups. 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, 2003). Meetings, information and follow-up should be handled sensitively and respect the needs of the individual family; sometimes the venue for meetings and the method of transport used to get there will dictate whether a mother can attend. Networking with other mothers and other family members with similar needs and experiences should be offered.

## Difficult-to-reach families

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 reason why and to offer support to ensure future attendance. The social worker with deaf people may be able to help. These children may have disjointed access to education and health services and they may miss the screening and health surveillance programmes. 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.

‘There is a higher incidence of deafness among some ethnic minority families’

## Young people and transfer of care to another 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 study and employment.

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, where relevant, sign language skills. The clinical genetic service sees individuals of any age (see page 29).

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 be in place to ensure the smooth transition from the child to the adult ophthalmology service. 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, 2003).

Adult services should be made aware of any changing circumstances (e.g. leaving home, 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 and the optometrist should advise how often this must take place. If no refractive error has been present throughout their school life, optometry is recommended at two-yearly intervals.



### *Quality standard*

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

### **Transfer to an alternative paediatric/adolescent service**

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. The child/young person and family must be fully informed in writing of details relating to the new service.

### *Quality standard*

**All relevant appointments with the new service must be arranged by the existing service provider prior to transfer and the family (or young person) notified.**

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## 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 options.

#### Language

- **Oral language (speech):** hearing is the primary sense through which language develops from birth. A significant proportion of deaf children use speech to communicate.
- **Sign language in the UK (known as British Sign Language BSL):** a visual language using handshapes, 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.
- **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.
- **Signed English (SE):** an exact representation of the English language through signs, using a sign or finger spelling for every spoken word. BSL signs are employed together with specially developed signs that give guidance on important points of grammar and finger spelling. 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.

#### Modes of communication

- **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
- **Deafblind manual:** uses letter shapes (similar to the manual alphabet) spelt into the hand of the deafblind person
- **Lipreading/speechreading:** 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, 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

- **Cued Speech:** some words that sound different look very similar when they are lipread. Cued Speech uses handshapes 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.
- **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 relatively new sign supported system for children 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.

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

## Appendix 2

### Key personnel

**Audiological physician (consultant in audiological medicine):** a doctor who investigates, diagnoses and manages children and/or adults with hearing loss, balance and communication disorders, including tinnitus and auditory processing disorders.

**Audiologist and audiological scientist:** non-medical professionals who specialise in the identification, assessment and rehabilitation of hearing loss and balance disorders, including the fitting of hearing aids.

**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 the deaf child.

**Educational audiologist:** a qualified teacher of the deaf 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 the child.

**Genetic counsellor:** often a nurse, who is specifically trained in genetics and works with families.

**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.

**Hearing therapist:** provides a rehabilitative service related to everyday living and functional use of hearing for deaf adults and young people. They may also counsel on tinnitus, fit hearing aids and advise on assistive listening devices.

**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.

**Key worker:** a member of the multi-disciplinary team assigned to work with the deaf child and family who will be familiar with local and national services.

**Lead clinician:** is responsible for co-ordinating all aspects of the care and management of the deaf child, including their eye and vision care. They are also responsible for ensuring that professionals involved with the child and family communicate effectively. Who the lead clinician is will depend on local circumstances, however, they may be, for example the community paediatrician, the audiological physician, 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.

**Orthoptist:** tests vision and assesses functional vision including binocular vision. Diagnoses and assesses strabismus, eye movement anomalies, and the reasons for failure of normal development. Many are involved in screening pre-school children for visual defects.

**Optometrist:** prescribes and dispenses spectacles 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 co-ordinate the eyes.

**Social worker with deaf people:** specialist social worker with sign language skills, and with knowledge of deaf issues and deaf awareness. Advises on benefits and equipment in the home.

**Specialist speech and language therapist:** some SLTs are additionally trained and specialise in working with deaf babies, children and adults and therefore have sign language skills. They assess and advise on communication, language and speech perception and production. They may be based in the hospital, the community and in the education environment. Some may offer treatment, depending on the context in which they work.

**Teacher of the deaf (ToD):** qualified teacher with specialist 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.

**Teacher of MSI/deafblind:** qualified teacher with post-graduate training, who works in an advisory role, often alongside the ToD and TVI, or in the classroom. 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 who will have an additional qualification and specific 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 classroom teachers.

# Appendix 3

## Conditions affecting both hearing and vision

The following list is not exhaustive but includes some conditions associated with deafness and ophthalmic disorders.

### **Alport syndrome**

The main characteristics are progressive renal dysfunction and progressive sensorineural deafness. Ocular signs include cataract, lenticonus, corneal arcus and flecked retinopathy with abnormal ERG and EOG. It is usually transmitted by X-linked recession, but may be dominant or autosomal recessive.

### **Alstrom syndrome**

An autosomal recessive disorder characterised by diabetes mellitus, severe sensorineural deafness, obesity and retinal dystrophy. Other features include chronic renal disease, pigmentary skin lesions, acanthosis nigricans and male hypogonadism. Poor vision and nystagmus are present in infancy.

### **CHARGE syndrome or association**

CHARGE is an association of features of arrested development of unknown cause: Coloboma, Heart defects, Atresia choanae, Restricted growth and development, Genital hypoplasia and Ear abnormalities and/or hearing loss (conductive, sensorineural or mixed). A formerly under reported cause of deafblindness, it is believed that CHARGE may now be a leading cause of deafblindness among children, replacing congenital rubella syndrome.

### **Congenital cytomegalovirus (CMV embryopathy)**

Ocular findings in infected neonates include chorioretinitis, microphthalmos, cataracts, keratitis and optic atrophy. Systemic involvement includes jaundice, hepatosplenomegaly, microcephaly, psychomotor retardation, cerebral calcifications and a petechial rash. Most affected infants do not present neonatally, but some 40% show features of congenital CMV by 3 years of age, mostly sensorineural hearing loss. Retinal pigment changes may be seen.

### **Congenital rubella (rubella embryopathy)**

The congenital rubella syndrome includes retinal pigmentary retinopathy (in 40% of the affected patients), cataract (in 20% of the children), microphthalmos, glaucoma and keratitis. Other systemic abnormalities include; congenital heart defects, sensorineural deafness, growth and developmental delay, microcephaly, osteopathy, lymphadenopathy, hepatospleno-megaly, thrombocytopenic purpura and diabetes.

### **Congenital toxoplasmosis**

The most common findings with sensorineural deafness are chorioretinitis and intracranial calcifications. Less common findings are seizures, hydrocephalus, hepatosplenomegaly, jaundice, anaemia and fever. Some infected patients may be normal in the initial examination and develop chorioretinitis, blindness, hydrocephalus, mental retardation and deafness, some years later.

### **DIDMOAD (Diabetes Insipidus, Diabetes Mellitus, Optic Atrophy, and Deafness)**

Characterised by juvenile diabetes mellitus and optic atrophy, which has an onset between 2 and 24 years of age. Visual fields are constricted and the patient is colour blind. Pigmentary changes of the retina have also been reported. ERG findings imply that a more widespread abnormality may be present than just ganglion cell degeneration. It is inherited as an autosomal recessive disorder.

### **Duane syndrome**

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

### **Marshall syndrome (see Stickler syndrome)**

This dominantly inherited syndrome includes early onset of progressive sensorineural hearing loss, cataracts, myopia, saddle nose and short stature. It is not known whether it is a variant of Stickler.

### **Norrie syndrome**

Retinal dysplasia or detachment lead to a congenital or progressive loss of vision in early childhood. Progressive sensorineural hearing loss usually occurs in late childhood. There may also be progressive loss of intellectual function. Inheritance is X-linked recessive.

### **Refsum syndrome**

Retinitis pigmentosa occurs in both the infantile and adult-onset types. Other features include progressive sensorineural deafness and motor and sensory neuropathy. Both are due to a defect in phytanic acid metabolism and are autosomal recessively inherited.

### **Stickler syndrome (see Marshall syndrome)**

A craniofacial disorder characterised by a flat facial profile, cleft palate, ocular changes and joint disease. Hearing loss is either conductive or mixed in nature. Ophthalmic features include progressive myopia and retinal detachment, which may present in early life. It is autosomal dominant.

### **Usher syndrome (see Appendix 4)**

### **Varicella embryopathy (chickenpox)**

The key features include sensorineural deafness (often moderate/severe), limb abnormalities and chorioretinitis and cataracts. Other systems including the gut may be affected, and other eye problems may be found.

# Appendix 4

## Usher syndrome

Usher syndrome is characterised by:

- congenital sensorineural hearing loss
- 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 et al., 1991), and 4.4/100,000 in the USA (Boughman, Vernon & Shaver, 1983). In the congenitally deaf population it is estimated to account for 3%-6% (Vernon 1969).

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 but may be amenable 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 commoner in Type 1. Type 2 may be associated with a slightly milder course. Complete blindness is unusual and some people will retain vision of 6/60 or better into their 60s.

**Type 1:** is characterised by congenital severe to profound sensorineural hearing loss affecting all frequencies. In most children, vestibular function is absent and leads to delayed motor development (poor head control, late sitting and rarely walks 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 hearing loss predominantly affecting the higher frequencies. Vestibular function is normal, and visual problems may not be apparent as early as in Type 1.

**Type 3:** this rarer type has an initially mild, often late onset progressive hearing loss which is usually moderate to profound with a gradual decrease in vestibular function. However, the retinal appearance is indistinguishable from Types 1 and 2.

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 Ushers.

# Appendix 5

## Useful organisations

### **Association of Sign Language Interpreters (ASLI)**

PO Box 32152  
London N4 2YZ  
Tel: 020 8809 4353 (voice & text)  
Fax: 020 8800 3489  
Email: [info@asli.org.uk](mailto:info@asli.org.uk) Web: [www.asli.org.uk](http://www.asli.org.uk)

### **British Retinitis Pigmentosa Society**

PO Box 350  
Buckingham MK18 1GZ  
Tel: 01280 821334  
Helpline: 01280 860 363  
Fax: 01280 815900  
Email: [info@brps.org.uk](mailto:info@brps.org.uk) Web: [www.brps.org.uk](http://www.brps.org.uk)

### **Council for the Advancement of Communication with Deaf People (CACDP)**

Durham University Science Park, Block 4  
Stockton Road  
Durham DH1 3UZ  
Tel: 0191 383 1155 (voice & text)  
Fax: 0191 383 7914  
Email: [durham@cacdp.org.uk](mailto:durham@cacdp.org.uk) Web: [www.cacdp.org.uk](http://www.cacdp.org.uk)

### **Contact a Family**

209-211 City Road  
London EC1V 1JN  
Tel: 020 7608 8700; helpline (freephone): 0808 808 3555  
Text: 0808 808 3556  
Fax: 020 7608 8701  
Email: [info@cafamily.org.uk](mailto:info@cafamily.org.uk) Web: [www.cafamily.org.uk](http://www.cafamily.org.uk)

### **Deafblind UK**

National Centre for Deafblindness  
John and Lucille van Geest Place  
Cygnet Road  
Hampton  
Peterborough  
Cambridgeshire, PE7 8FD  
Tel: 01733 358100  
Text: 01733 358858  
Fax: 01733 358356  
Email: [info@deafblinduk.org.uk](mailto:info@deafblinduk.org.uk) Web: [www.deafblinduk.org.uk](http://www.deafblinduk.org.uk)

**Look: National Federation of Families with Visually Impaired Children**

C/O Queen Alexandra College, 49 Court Oak Road  
Harborne  
Birmingham B17 9TG  
Tel: 0121 428 5038  
Fax: 0121 427 9800  
Email: [info@look-uk.org](mailto:info@look-uk.org) Web: [www.look-uk.org](http://www.look-uk.org)

**The National Deaf Children's Society**

15 Dufferin Street  
London EC1Y 8UR  
Tel: 020 7490 8656 (voice & text);  
helpline (freephone): 0808 800 8880 (voice & text)  
Fax: 020 7251 5020  
Helpline email: [helpline@ndcs.org.uk](mailto:helpline@ndcs.org.uk)  
Email: [ndcs@ndcs.org.uk](mailto:ndcs@ndcs.org.uk) Web: [www.ndcs.org.uk](http://www.ndcs.org.uk)

**Royal National Institute of the Blind (RNIB)**

105 Judd Street  
London WC1H 9NE  
Tel: 020 7388 1266; helpline: 0845 766 999  
Fax: 020 7388 2034  
Email: [helpline@rnib.org.uk](mailto:helpline@rnib.org.uk) Web: [www.rnib.org.uk](http://www.rnib.org.uk)

**Royal National Institute for Deaf People (RNID)**

19-23 Featherstone Street  
London EC1Y 8SL  
Tel: 0808 808 0123 (freephone)  
Text: 0808 808 9000 (freephone)  
Fax: 020 7296 8199  
Email: [informationline@rnid.org.uk](mailto:informationline@rnid.org.uk) Web: [www.rnid.org.uk](http://www.rnid.org.uk)

**Sense, The National Deafblind and Rubella Association**

11-13 Clifton Terrace  
London N4 3SR  
Tel: 020 7272 7774; text: 020 7272 9648  
Fax: 020 7272 6012  
Email: [enquiries@sense.org.uk](mailto:enquiries@sense.org.uk) Web: [www.sense.org.uk](http://www.sense.org.uk)

**SPECS (Specific Eye Conditions)**

49 Southfield Avenue  
Paignton  
South Devon  
TQ3 1LH  
Tel: 01803 524238  
Email: [info@eyeconditions.org.uk](mailto:info@eyeconditions.org.uk) Web: [www.eyeconditions.org.uk](http://www.eyeconditions.org.uk)

## Appendix 6

### Useful publications

**Ben's Brand New Glasses**, Caroline Dina, Faber, 1987, ISBN 0 571 14567-1

**Going to the Optician**, 1999, booklet published by Sense, The National Deafblind and Rubella Association

**The Mole Who Needed Glasses**, Adam Storer, Tesco Stores/Brilliant Books, 2001  
ISBN 1 84221 146 3

**Topsy and Tim Have their Eyes Tested**, Jean and Gareth Adamson, Blackie Children's Books, 1995, ISBN 0 216 94169 5 paperback, ISBN 0 216 94168 7 hardback

### Other useful sources of publications

**RNIB Library** (see appendix 5)

**RNID Library** (see appendix 5)

**Forest Bookshop** (for books, videos and CD-ROMs about deafness and deaf issues)

The Forest Bookshop Warehouse, Unit 2, The New Building

Ellwood Road, Milkwall, Coleford

Gloucester GL16 7LE

Tel: 01594 833858 (24hr)

Text: 01594 833858, Text answerphone: 01594 833507

Videophone: 01594 810637

Fax: 01594 833446

Email: [forest@forestbooks.com](mailto:forest@forestbooks.com) Web: [www.ForestBooks.com](http://www.ForestBooks.com)

# Appendix 7

## Glossary

**Acanthosis nigricans:** skin disease characterised by grey-black warty patches, usually situated in the groin, elbows or knees.

**Amblyopia:** reduced visual acuity, associated with strabismus, obstruction to vision such as by cataract, or refractive error, even after correction by glasses of a refractive error, without any apparent disease of the eye. It is usually only present in one eye.

**Anaemia:** a reduction in the red blood cell pigment which may lead to pallor, weakness and breathlessness.

**Arcus:** An anatomical arch, ie an opaque line surrounding the margin of the cornea.

**Astigmatism:** occurs when the cornea/lens does not have the same degree of curvature in the horizontal and vertical planes, resulting in distorted images.

**Atresia choanae:** blockage of the nasal passages.

**Autosomal dominant (see also X-linked):** passed from one parent, who has the condition, to the child (1:2 chance at each pregnancy).

**Autosomal recessive (see also X-linked):** passed from both parents (carriers), who themselves have the condition, to the child (1:4 theoretical chance at each pregnancy).

**Binocular vision:** use of both eyes together.

**Cataract:** any opacity of the lens of the eye, resulting in blurred vision.

**Chorioretinitis:** inflammation of the retina and choroid of the eye.

**Coloboma:** a defect of eye development when a fissure that is normal but temporary fails to close up before birth.

**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.

**Congenital:** present at birth.

**Decibel hearing level (dBHL):** a unit used for measuring the intensity (loudness) of sound and plotted on a chart (audiogram).

**Diabetes insipidus:** excessive loss of water through urine and increased thirst and drinking as a result of deficiency in the hormone vasopressin.

**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.

**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).

**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.

**Field of vision:** lateral and vertical extent of vision.

**Foveal/fovea:** this is the central area of the retina responsible for the best visual acuity.

**Fundoscopy, ophthalmoscopy:** examination of the retina and optic disc at the back of the eye by an instrument called an ophthalmoscope.

**Glaucoma (congenital):** raised pressure within the eye, causing sensitivity to light, excessive tearing, discomfort, enlargement of the eye, reduced vision, corneal clouding.

**Glue ear:** see otitis media with effusion.

**Hepatospleno-megaly:** enlargement of the liver and spleen.

**Hydrocephalus:** abnormal increase in the amount of cerebral spinal fluid within the ventricles of the brain.

**Hypermetropia/hyperopia:** long-sightedness.

**Hypogonadism:** functional incompetence of the gonads.

**Hypoplasia:** condition in which an organ or part is undeveloped.

**Intracranial:** in the skull.

**Jaundice:** yellow discolouration of the skin due to excess bile pigment. There are many different causes of jaundice.

**Keratitis:** inflammation of the cornea of the eye.

**Lenticonus:** rare and usually congenital condition of the lens of the eye in which the surface is conical, especially on the posterior side.

**Lymphadenopathy:** abnormal enlargement of the lymph nodes.

**Meatal atresia:** absent/extremely narrowed external ear canal.

**Microcephaly:** abnormal smallness of the head.

**Microphthalmos:** abnormal smallness of the eye.

**Mixed hearing loss:** both a sensorineural and conductive hearing loss is present.

**Myopia:** short-sightedness.

**Ophthalmoscopy:** see 'fundoscopy'.

**Optic atrophy:** degeneration of the optic nerve.

**Osteopathy:** general term for bone disease or disorder.

**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 hearing loss is temporary, usually mild to moderate, can affect one or both ears and fluctuate. It usually resolves spontaneously but may need specific treatment.

**Oto-toxic:** toxic to the ear, causing sensorineural hearing loss and/or balance disturbance. Examples include noise and some drugs.

**Perinatal:** occurring around the time of childbirth.

**Petechial rash:** minute reddish and purplish areas on the skin.

**Photophobia:** abnormal sensitivity of the eyes to light.

**Pigmentary retinopathy:** see retinitis pigmentosa.

**Postnatal:** occurring after birth.

**Prenatal:** occurring before birth.

**Refraction:** the test which determines if someone needs glasses. Drops may be used when testing young people.

**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 (eg, a magnifier), spectacles, contact lenses or other forms.

**Retinal dystrophy:** degeneration of the retina.

**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).

**Sensorineural hearing loss:** 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.

**Snellen chart:** commonest chart used for testing sharpness of distant vision made up of lines of letters of decreasing size. The 3 metre logMAR charts, a modified form of Snellen, are replacing Snellen as the 'gold-standard'. Both tests can be used with letter matching cards or plastic letters, so that the child does not have to name the letters.

**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.

**Strabismus (squint):** condition in which the eyes are misaligned.

**Thrombocytopenic purpura:** deficiency of blood platelets.

**Usher syndrome:** see Appendix 4.

**Vestibular:** used to refer to either the organ of balance in the ear or the function of balance.

**Visual acuity:** the limit of spatial visual discrimination, commonly measured using letters or other geometrical forms.

**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.

**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).

# Appendix 8

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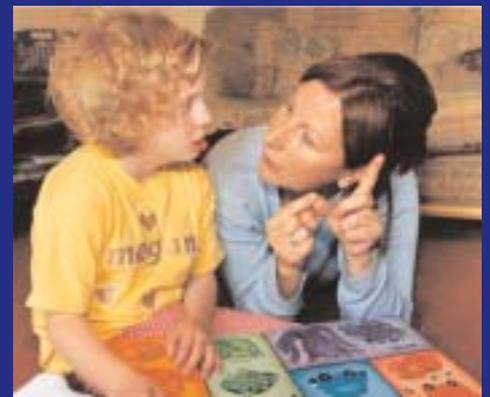
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## NDCS and Sense – working together

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

The National Deaf Children's Society is an organisation of families, parents and carers which exists to support parents in enabling their child to maximise their skills and abilities, and works to facilitate this process by every possible means. NDCS is working for all deaf children and young people. NDCS provides information on all aspects of childhood deafness and can also offer practical support, advice and advocacy.

Sense is the pioneering national charity that supports and campaigns for children and adults with sight and hearing difficulties. We provide specialist information, advice and services to deafblind people, their families, carers and the professionals who work with them. This includes: offering in-depth assessments of children with complex needs; supporting families through a national network and local branches; running a holiday programme for deafblind children and adults; and providing advice and support to people with Usher syndrome.



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London N4 3SR  
☎ tel 020 7272 7774  
☎ fax 020 7272 6012  
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Patron: Her Royal Highness The Princess Royal

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Website: [www.ndcs.org.uk](http://www.ndcs.org.uk)

Registered Charity No: 1016532