

A general reference manual, offering practical advice on how to prepare for a consultation with a person who is deaf, deafblind or Deaf (i.e. uses sign language as their first language), it describes the issues which need to be considered with respect to language, communication and culture. Every health professional, medical student and nursing student should have a copy of this book.

ANNA MIDDLETON is a Consultant Research Genetic Counsellor and Research Fellow at the Institute of Medical Genetics, Cardiff University, UK.

'This is a brilliant book that every health professional should read as part of their ongoing training.' **Steve Powell, CEO SignHealth, UK**

'This book presents some excellent material in a wide and complex field, written with such clarity that it will be useful to almost any reader whatever their background.' **Dr Lorraine Gailey, CEO Hearing Concern LINK, UK**

'This is a must-read book for all health professionals. Well worth keeping for those times when you may be confronted by a patient who requires different communication tactics that you have never used before.'

Paul Redfern, Consultant in Disability and Diversity, Middlesex, UK

'This book may be of particular interest to clinical geneticists and genetic counsellors.' **Rachel Belk, Genetic Counsellor, St Mary's Hospital, Manchester, UK**

'For those who have little experience of meeting deaf people, the case studies emphasize common pitfalls in communication, and I will be recommending the book to trainees who are new to the field.' **Dr Maria Bitner-Glindzicz, Consultant Geneticist, UCL Institute of Child Health, London, UK**

'I have worked with patients with deafblindness and NF2 for over 23 years, and I realize now that this is the first book to have addressed the basic knowledge of how to interact with people who are Deaf or Deafblind. I sincerely recommend this book.' **Professor Claes Möller, Head of Audiology and Medical Disability Research, University Hospital Örebro, Sweden**

'Most impressive. The more I look at it, the better it seems to be.'

Professor Dafydd Stephens, Honorary Professor of Audiological Medicine, Cardiff University, UK

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Working with Deaf People

A Handbook for
Healthcare Professionals

EDITED BY
Anna Middleton

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Medicine

She has conducted public investigations of services for people with deafblindness on behalf of the Swedish Ministry of Social Affairs and the Swedish National Agency for Education. She has done consultancy for the Nordic Staff Training Centre for Deafblind Services, the Swedish Resource Centre for Matters regarding Deafblindness and the Swedish Association of Rare Disorders. She teaches students, professionals and people with deafblindness and their next of kin in deafblindness know-how. She works as a supervisor at Research & Development in Sörmland and is associated to the Swedish Institute of Disability Research including HEAD and Audiological Research Centre at Örebro University Hospital.

Facts and figures about deafness, NF2 and deafblindness

Anna Middleton, Wanda Neary
and Kerstin Möller

Overview of deafness and hearing loss

The clinical impact of deafness is variable. It may occur at any stage of life, it may impact on the individual's ability to function on a day-to-day basis and it may or may not be disabling.

Conversational speech can be measured as having a loudness of approximately 60 decibels (dB). Hearing is considered significantly restricted when the ear cannot interpret or process sounds of 25 dB or more.

The following is adapted from work by Prosser and Martini (2007).

- An individual with a **'mild'** hearing loss can only begin to hear sound if it is between 20 and 40 dB. They may have difficulty understanding conversations in a noisy room, or if the speaker is far away, but should be able to hear one-to-one conversations if the speaker's voice is not too soft. They may have problems hearing a person in front of them who is whispering.

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- An individual with a '**moderate**' hearing loss can begin to hear sound between 41 and 70 dB; people with such a loss have difficulty understanding normal conversational levels of speech, but may be able to hear loud noises, for example the sound of a lawnmower (about 90 dB).
- An individual with a '**severe**' hearing loss begins to hear sound between 71 and 95 dB. Such people will only be able to hear an individual speaker if they are at close range and the speaker is shouting; they may be able to hear a car horn (110 dB). A person with a mild, moderate or severe hearing loss may receive benefit from the use of a hearing aid.
- An individual with a '**profound**' hearing loss can only hear sound equivalent to or over 95 dB, for example a gunshot (140 dB). Profoundly deaf people will not be able to hear loud speech or background noise and may prefer to use sign language rather than speech as their form of communication. Some profoundly deaf people may also not receive much benefit from a hearing aid and as such may choose not to wear one.

(Prosser and Martini 2007)

Terminology

There is often confusion about the terms used to describe deafness. This is largely because health professionals, academics and deaf and hard of hearing individuals themselves may use different terminology to describe related concepts (Grundfast and Rosen 1992).

- Generally speaking, clinicians and molecular deafness academics often use the term 'hearing impaired' rather than 'deaf' as this latter term is often considered too non-specific. 'Hearing impaired' has a precise medical definition, as per the International Classification of Functioning, Disability and Health (Stephens and Danermark 2005).
- However, the term 'hearing impaired' is often not viewed as politically correct these days. It tends to be avoided by deaf and hard of hearing people themselves, as they do not like to be perceived as being defective or 'impaired'.
- Use of the phrase '*the deaf*' is also not perceived as politically correct either as it has a slightly condescending air to it. This has been replaced by the use of 'deaf people' or 'people with deafness' instead. For example, in the UK the charity 'Hearing Dogs for *the Deaf*' is now termed 'Hearing Dogs for Deaf People'.
- Many people with disabilities have suggested it is most politically correct to refer to the *person* first and the 'disability' second, for example, 'people with Down's syndrome' rather than 'the Down's client'. This phraseology may only partly apply to deafness – it is still acceptable to say 'deaf people' because deafness can be tied up with identity. But for the hard of hearing group it may be considered more sensitive to use the term 'people with hearing loss'.

- Those with a mild or moderate level of deafness will often refer to themselves as being hard of hearing or having a hearing loss. Such people may also find great benefit from wearing a hearing aid and tend to use spoken language rather than signed language. Within interactions with hearing people they will often be able to use their residual hearing and amplified hearing as well as lip-reading to help their communication. People who have elderly-onset hearing loss often call themselves hard of hearing.
- People who are 'deafened' tend to have a profound level of deafness and usually will have started life as a hearing person. Their deafness may be progressive or may have a sudden onset. Deafened people usually feel they belong to the Hearing World rather than the Deaf community as they often do not use sign language. They may also receive little benefit from hearing aids, although cochlear implants may work well for this group. Within interactions with hearing people, deafened people may rely very heavily on lip-reading, writing and reading to communicate.

deaf and Deaf

The following text provides a very general guide to the use of the terms deaf, hard of hearing, deafened and Deaf in the UK. These concepts are fluid and changeable and there are no universally accepted definitions, used by affected

people themselves, that translate across the world. However, we have attempted to capture the way the terms are broadly used.

- People who refer to themselves as 'deaf' usually have a profound level of deafness, which may also be static rather than progressive.

People who refer to themselves as 'Deaf' (written with an uppercase D) are indicating that they have a culture and identity that is linked in with their deafness and that they use sign language as their preferred communication (Padden and Humphries 2005).

- Those who refer to themselves as 'deaf' may use some spoken language and some sign language in different contexts. Alternatively, they may prefer to only use sign language. They may also feel most comfortable within the Deaf World/community/culture (a minority group within mainstream society where those who belong mainly communicate in sign language).
- Some who refer to themselves as 'hard of hearing' may do so to indicate that they *are not* profoundly deaf and yet they may still mix within the Deaf community, and thus may use both sign language and spoken communication in different situations.
- Conversely some people call themselves 'hard of hearing' or 'deafened' to indicate that they are not part of the Deaf community.

People who refer to themselves as 'hard of hearing', 'hearing impaired' or 'deafened' often feel they belong to the Hearing World and/or Hard of Hearing World (consisting of other people with a similar perception of hearing loss). This tends to be a group that is not fluent in sign language and relies on using spoken language.

Case study: developing a Deaf identity

Jo from Northern Ireland had been profoundly deaf since birth, as were his mother and maternal grandfather. Jo had a severe level of deafness, together with a white forelock and different-coloured eyes. He was diagnosed by his paediatrician as having Waardenburg syndrome.

At age 5, Jo found it difficult to fit into the mainstream hearing school recommended by his Local Education Authority and so his parents decided to send him to a specialist school for deaf children, similar to the one his mother had been to. Here he was offered a Total Communication approach – and so had speech therapy to help him learn speech, but also he learnt sign language.

By the age of 10 he found mixing with other deaf children gave him a sense of normality that he didn't feel amongst hearing children and gradually he found he felt more comfortable using sign language rather than speech. Both his parents were members of the local Deaf club and also used sign language at home.

Over time Jo found that his experiences in school and at the local Deaf club helped him to develop a Deaf identity – he felt more comfortable mixing with other Deaf people and communicating in sign language. After leaving school he trained to become a social worker for deaf people.

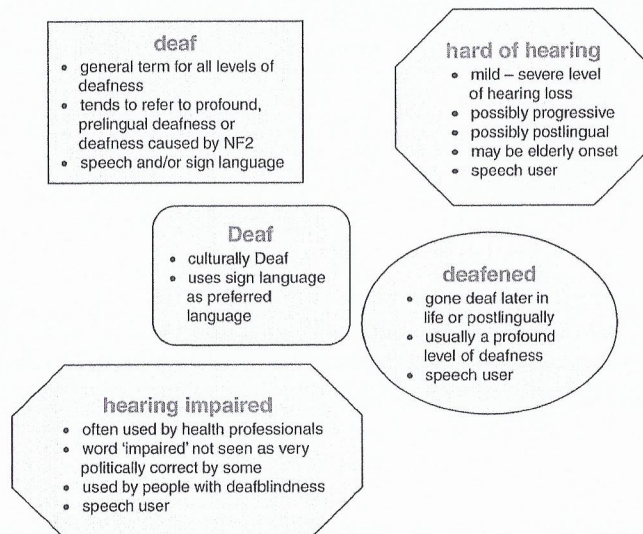
Terms used by people with NF2

- People with NF2 are born into the Hearing World; they usually use spoken language when communicating.
- When they develop hearing loss, which is most commonly in their 20s or 30s, people with NF2 usually describe themselves as deaf.
- Most people with NF2 learn to lip-read; they use speech and written communication.
- Some receive a cochlear implant, or an auditory brainstem implant, which aids their lip-reading.
- Only a small minority of people with NF2 learn to communicate using an NSL and consider themselves part of the Deaf community.

Terms used by people with deafblindness

- People with deafblindness are a heterogeneous group. Those who are born with a hearing loss will often call themselves hearing impaired or deaf.
- As their sight deteriorates they may call themselves 'deaf with visual impairment'.

- If they have learnt sign language as their first language they may call themselves Deaf.
- Some people call themselves deafblind; however, many are not aware that the combination of their hearing and visual loss is significant and so may just mention one sensory loss.
- Using the term 'impairment' in the label 'visual impairment' is politically correct; there is not the same stigma attached to this term as there is for 'hearing impairment' for those who have deafness in isolation.
- For ease of language and reading the term 'deafness' and 'deaf people' is used collectively in this book to refer to people with any level and perception of hearing loss.



Deafness can be perceived in different ways

- A person who considers themselves hard of hearing or deafened may find their hearing loss is an irritation for them and their significant others. They may also have to make large adjustments in their life both practically and emotionally in order to adapt to this.
- A person who considers themselves culturally Deaf may feel a sense of pride in their deafness and may not feel it is a problem at all (Ladd 1988, 2003) – in fact within their family and social position, they may feel that it is preferable to be deaf rather than hearing.

Deaf sign language users and hard of hearing speech users may have completely opposing views and perspectives towards deafness.

Having a clear family history of deafness can be very positive for individuals who are deaf. Several research studies have shown that, irrespective of the level of deafness, age of onset or impact of deafness, if there are other relatives who can be role models for the individual who is deaf, then this can be of enormous psychological benefit (Stephens 2007).

Frequency of deafness, NF2 and deafblindness

- In the developed world deafness is the most common congenital disorder (Hilgert, Smith et al. 2008).

- Approximately 1 in 500 babies are born with a hearing loss greater than 40 dB (Morton and Nance 2006).
- Hearing loss increases with age; 16% of adults have a bilateral hearing loss greater than 25 dB (Davis 1989, Morton 1991) and by the age of 80 nearly half the population will have a hearing loss greater than 25 dB (Morton 1991).
- The diagnostic prevalence of NF2 is 1 in 210,000 and the birth incidence has been estimated to be approximately 1 in 40,000 (Evans, Huson et al. 1992a); many individuals do not develop features of the condition until the third decade or later, although individuals with aggressive disease die before the third decade.
- The vast majority of people with NF2 develop deafness either due to the presence of vestibular Schwannomas (tumours of the eighth nerve) or due to the surgical removal of vestibular Schwannomas.
- Deafblindness is very rare, especially in young people. Prelingual deafblindness affects 1 in 10,000 children (Möller 2007).
- Usher syndrome affects about 3 per 100,000 (Sadeghi 2005).
- Developing a combination of visual and hearing impairment increases with age and particularly after the 7th decade.
- Visual impairment is the most common impairment in people with hearing loss. Thirty per cent of children with hearing loss or deafness have been found to have visual impairment (Nikolopoulos, Lioumi et al. 2006).

Describing deafness and hearing loss

A variety of different categories are used by health professionals to describe deafness. Classification can be made in different ways dependent on the cause, the form and the stage of life at which it occurred:

Type of deafness:	sensorineural vs. conductive
Timing of deafness:	congenital vs. acquired prelingual vs. postlingual late vs. early onset
Progression of deafness:	progressive vs. non-progressive
Cause of deafness:	genetic vs. environmental
Form of deafness:	syndromal vs. non-syndromal

A brief discussion of each of these follows.

Sensorineural vs. conductive deafness

When the structures within the inner ear or auditory nerve are changed or missing, for example, involving the cochlear hair cells, the result is *sensorineural* deafness, whereas structural changes in the outer and middle ear result in *conductive* deafness. A mixture of sensorineural and conductive factors leads to *mixed* deafness.

Congenital vs. acquired

Congenital means that a person has been born deaf whereas *acquired* means that a person has become deaf as

a result of environmental factors, for instance noise or industrial damage or infection. Congenital deafness is mostly sensorineural in origin. Deafness occurring in older age is sometimes referred to as 'an acquired sensorineural' form of deafness.

Prelingual vs. postlingual

Prelingual means that deafness existed before the development of language skills. This term is sometimes used interchangeably with the term *congenital*. If deafness occurs after the development of language skills this is described as *postlingual*.

Early vs. late onset

Early onset and *late onset* refer to the time when the deafness occurred, i.e. in childhood or adulthood.

Progressive vs. non-progressive

Progressive refers to the increasing severity of the condition over time, whereas *non-progressive* means that the severity has not changed over time and remains fairly consistent.

Genetic vs. environmental

Babies born with severe-profound, congenital or early-onset deafness have their deafness due to genetic causes

in >50% of cases, whereas environmental causes are believed to account for <50% of cases, with the remainder being of unknown cause (Parving 1983, 1984, Newton 1985).

Syndromal vs. non-syndromal

One-third of all cases of genetic deafness are *syndromal*; *non-syndromal* deafness accounts for the other two-thirds (Reardon and Pembrey 1990). Syndromal deafness includes other clinical features, for example blindness, craniofacial defects and pigmentation problems. The most common of these are Waardenburg syndrome, Usher syndrome, Pendred syndrome, CHARGE, neurofibromatosis Type II and branchio-oto-renal syndrome. Deafness is involved in over 400 genetic syndromes (Toriello, Reardon et al. 2004).

At least 20 different genetic syndromes are associated with prelingual deafblindness (Möller 2007). More than 50 hereditary syndromes are known to cause acquired deafblindness (Möller 2007).

When genetic deafness occurs in isolation with no other clinical features, it is known as *non-syndromal* deafness. There are over 50 non-syndromal deafness genes where alterations (mutations) have been identified. It is therefore technically possible to offer genetic testing for these genes, but testing may not be available clinically (Smith and Van Camp 2008).

Causes of deafness

There are many different causes of deafness; these include environmental and genetic factors.

- More than half of congenital or prelingual deafness has a genetic cause (Smith and Van Camp 2008).
- Several hundred genes are known to be involved with deafness (Smith and Van Camp 2008).
- Approximately 50% of non-syndromal prelingual deafness, with a genetic basis, is caused by alterations in the GJB2 and GJB6 genes. Approximately 1 in 50 people are carriers of the alterations in the GJB2 gene (Estivill, Fortina et al. 1998, Kelley, Harris et al. 1998, Smith and Van Camp 2008).
- Late-onset or 'elderly' hearing loss has always been thought to be due to environmental causes, but more recently it has been discovered that this has a probable genetic basis. This basis is still being investigated (Smith and Van Camp 2008).
- The most frequent environmental cause of congenital deafness is infection with cytomegalovirus (CMV) (Smith and Van Camp 2008).
- People who call themselves 'deafened' may be deaf due to a number of different reasons; for example, meningitis, head injury, otosclerosis, viral infections and ototoxic medications.

Case study: moderate hearing loss, use of speech

Jane was born to a hearing family in Wales – her parents, grandparents and siblings were all hearing. So, when she was diagnosed as having a moderate level of deafness by the Newborn Hearing Screening project, this was a great shock to the family.

Jane had congenital, bilateral, sensorineural deafness and was fitted with hearing aids. Jane's parents decided that they wanted to raise her within the wider hearing society and so with the help of a teacher for deaf children and additional speech therapy support at home Jane learnt to lip-read and use speech.

Jane's parents had genetic counselling in order to determine whether there was a genetic basis to Jane's deafness. Genetic testing revealed that Jane had inherited two alterations in the GJB2 gene, one from each of her parents, who carried these. Parents who are both carriers for these gene alterations have a 1 in 4 or 25% chance of having deaf children.

Comment

The GJB2 or Connexin 26 gene is the most common gene causing deafness. It mainly results in a child being born with deafness for the first time in a hearing family, where both parents of the child with deafness are hearing but carry an alteration in the gene.

Neurofibromatosis Type 2 (NF2)

- NF2 is a syndrome which usually results in total deafness. It is characterised by the presence of bilateral vestibular Schwannomas of the eighth nerve (benign tumours of the hearing nerve on both sides).
- Other tumours of the central and peripheral nervous system, such as meningiomas, gliomas, ependymomas and peripheral Schwannomas, are associated with NF2.
- NF2 is a dominantly inherited genetic condition caused by a defect on chromosome 22.
- There is a family history of NF2 in 50% of cases, but in the remaining 50% the condition arises as a result of a new genetic change.
- There is an aggressive form of the condition with an early onset, the presence of multiple nervous system tumours and premature death.
- In some individuals who are affected only with bilateral eighth-nerve tumours, the condition may progress slowly and they may retain their hearing into their seventh decade.
- NF2 is a clinically and genetically different condition from neurofibromatosis Type 1, which is a much more common genetic condition.

Presenting symptoms of NF2

- The majority of adult individuals with NF2 present with unilateral hearing loss, tinnitus, imbalance and vertigo,

related to the presence of bilateral vestibular

Schwannomas. Bilateral vestibular Schwannomas are usually present at diagnosis though only one may initially be causing hearing loss.

- The mean age at onset of symptoms is in the second and third decades.
- Two types of NF2 can be recognised – Wishart and Gardner.
- The Wishart type presents at an early age, the disease progression is rapid, and the individual has multiple other tumours of the central and peripheral nervous system in addition to bilateral vestibular Schwannomas.
- The onset of the Gardner type is at a later age and it has a more benign course with bilateral vestibular Schwannomas but no other tumours of the nervous system.
- Children aged 10 years or younger who are diagnosed with NF2 (a relatively rare occurrence) frequently present with symptoms of an isolated tumour of the nervous system rather than with symptoms due to bilateral vestibular Schwannomas (Evans, Birch et al. 1999).
- The diagnostic criteria for NF2 are given in the Appendix.

Diagnosis

- An MRI scan of the head and spine is the gold standard in diagnostic terms, with the possibility of revealing tumours as small as 2 to 3 mm in size.

- Presymptomatic DNA diagnosis of NF2 can be undertaken using molecular genetic techniques.

Case study: a new diagnosis of NF2

Tariq is a young man of 17 years. He has noted increasing difficulty with his left-sided hearing over a period of 12 months, and is now unable to hear a telephone conversation with his left ear. During the last 3 months he has also been troubled with continuous high-pitched left-sided tinnitus. There is no family history of deafness. His doctor refers him to the local ENT clinic, where the presence of a left-sided sensorineural hearing loss is confirmed. An MRI scan indicates bilateral vestibular Schwannomas, left greater than right. Tariq is referred to a specialist multidisciplinary NF2 clinic for further management.

Tariq is shocked at the diagnosis, and has many questions to put to the specialist multidisciplinary team regarding the impact of NF2 on his future career, his personal relationships and his general health. Further management of his eighth-nerve tumours is in the hands of the neuro-otologist and neurosurgeon. The geneticist explains the implications for Tariq's future children. Tariq is introduced to an NF2 support worker.

Comment

Members of the specialist multidisciplinary team must be sensitive to the psychological impact of NF2 as

a potentially life-changing disease for Tariq and his family. The situation is particularly difficult as there is no family history of NF2, and Tariq's illness has presented for the first time. The prospect of total eventual hearing loss and change in Tariq's plans for future employment may be devastating. Parental anxieties and possible feelings of guilt may be considerable. Members of the multidisciplinary team must ensure, as much as possible, that Tariq is prepared for the psychological blow of eventual total hearing loss and initiate appropriate training in non-auditory communication skills.

Management and treatment of NF2

NF2 presents marked challenges to clinicians with regard to the diagnosis as well as medical and surgical treatment (Neary, Stephens et al. 2006). Members of the affected person's family require screening for the condition, together with genetic counselling. The specialist role of regional and national centres with multidisciplinary teams involved in the management of all aspects of NF2 has been emphasised (Evans, Baser et al. 2005).

Clinicians need to be sensitive to the potentially life-changing psychological impact of the diagnosis of NF2, with the knowledge of eventual total hearing loss in the affected person (Neary, Stephens et al. 2006).

In families where the condition has appeared for the first time, the impact is even more marked. The prospect of future total deafness faces a person who on the surface appears healthy, and may have presented with only a mild hearing loss. Feelings of guilt and anxiety are common at this stage. A treatment strategy needs to be formulated which conserves useful hearing and aims for a good quality of life without causing complications that involve the facial nerve or neurological status. Training in lip-reading as well as other communication methods needs to be implemented as soon as possible.

The eighth nerve is very close to the facial nerves and so when vestibular Schwannomas are surgically removed, even by very skilled surgeons, it is possible to damage the facial nerve in this process. Minimising surgical damage to the facial nerve is paramount as bilateral facial weakness may result in physical difficulties relating to eating and smiling etc. and is often cosmetically distressing to the client. In NF2, the eyes may become affected, for example through damage to tear production as well as the blink reflex.

The primary goal of management is to foresee and take steps to avoid life-threatening events. The affected individual should be reviewed by the specialist team on a regular basis, when MRI of the neural axis should be undertaken. In this way impending complications should be identified and the need for surgery indicated in a timely manner.

When both auditory nerves are affected and the client with NF2 is totally deaf, an auditory brainstem implant

(ABI) is usually inserted. The ABI is reported to enhance lip-reading skills as well as providing an awareness of environmental sound, although the ABI usually does not result in good levels of hearing.

Deafblindness

In everyday terms, deaf and blind denote individual impairments and deafblind denotes a combination of the two impairments. People who have complete loss of both vision and hearing are very rare. Six per cent of those regarded as having deafblindness were found to have total visual and hearing impairment (Wolf-Schein 1989). Thus denoting deafblindness solely by total visual impairment (blindness) and total hearing impairment (deafness) is, in general, used neither by professionals in the field nor by organisations for people with combined visual and hearing impairment. There is no consensus definition of deafblindness.

In the USA, at the Helen Keller National Center for Youths and Adults who are Deaf-Blind (HKNC), deafblindness is defined as the degree of visual function and degree of hearing function. Historically, the primary education system in Sweden has referred to children with deafblindness as 'three-sensed' (Liljedahl 1993), which emphasises the senses left. In 2007, the Nordic Staff Training Centre adopted a new Nordic definition for the Deafblind Service which highlights special requirements rather than the degree of the two impairments.

'Deafblindness is a distinct disability. Deafblindness is a combined vision and hearing disability. It limits activities of a person and restricts full participation in society to such a degree that society is required to facilitate specific services, environmental alterations and/or technology' (www.nud.dk).

Finally, there is the classification based on International Classification of Functioning, Disability and Health (ICF) that combines the degree of sensory impairment, activity limitations and participation restrictions with environmental factors. The fact that visible and audible signals are not processed normally is an impairment of body function. Thus seeing/ watching and hearing/listening activities become limited and the visible and audible information cannot be interpreted in an intelligible way (Möller 2008).

Based on self-reports within deaf studies, deafblindness is regarded to be an alternative way of perceiving the world, though often as an 'isolated island' (Barnett 2002c). Reports from people with deafblindness show strategies and habits used for 'negotiating a place in a hostile world' (Schneider 2006).

Heterogeneity of the group

The population is distributed across all ages, with onset of one or both impairments at different ages. Some people first develop hearing impairment, others vice versa, while a few develop severe to profound visual and hearing impairment at the same time.

The group is often subdivided according to the age of the observed onset. In *prelingual onset*, the impact of both impairments comes before or during the development of language skills. Professionals usually call this subgroup congenital deafblindness. *Postlingual onset* is often called acquired deafblindness. The third group is *elderly* people who develop combined visual and hearing impairment at an old age.

Some conditions which cause deafblindness

The rarity of these conditions and difficulties in assessment increase the risk of an incorrect diagnosis, which also may be 'hidden' due to other dysfunctions and thus attributed to other conditions (McInnes and Treffry 1982, Möller 2007).

Usher syndrome

- Usher syndrome (USH) is a genetic disorder with autosomal recessive inheritance.
- USH is the most common cause of deafblindness before old age (Kimberling and Möller 1995, Sadeghi 2005). The

syndrome is divided into three distinct clinical types (types I–III). Different gene mutations and clinical features distinguish these types.

- USH affects the structure of the cochlea, the vestibular organ (types I and III) and the retina bilaterally (Kimberling and Möller 1995).
- In the inner ear (cochlea and the labyrinth) the hair cells are affected.
- The disorder in the eye, retinitis pigmentosa (RP), is one of several genetic disorders affecting the retina (Hartong, Berson et al. 2006). The visual deterioration is slower in USH compared to some other forms of RP (Sadeghi 2005).

USH type I is associated with profound deafness, while types II and III are associated with moderate to severe hearing impairment. In type III this is usually progressive (Kimberling and Möller 1995, Sadeghi 2005).

- Vestibular function is absent bilaterally in USH type I. This will result in a delayed walking age (>18 months) and clumsiness, especially in dimly lit situations or in darkness (Möller 2007).
- Type II has normal vestibular function, while type III has progressive loss of vestibular function.
- All three types of USH cause progressive visual impairment (Kimberling and Möller 1995).

- The RP in the three types of USH does not differ as much as the auditory or vestibular functions. In many cases the progression of visual impairment can be the same in types I, II and III.

Rehabilitation of children with USH type 1 has changed dramatically with the introduction of cochlear implants (CIs). Children with USH type 1 in Sweden are fitted with two implants at the age of 6–12 months. The aim is for them to hear spoken language and to develop speech. From around 1997, nearly 95% of all children with congenital profound deafness (including USH type 1) have received cochlear implants (Möller 2007). ‘Treating’ deafness when it occurs in conjunction with blindness tends to be approached from a medical model; however, this is not always the case. There are still some people with Usher syndrome who prefer to view their deafness from the cultural model and would prefer not to have a treatment such as a cochlear implant.

Alström syndrome

- Alström syndrome is a hereditary autosomal recessive disorder.
- All individuals with the disease have visual impairment, hearing loss, obesity problems, cardiovascular problems, early-onset diabetes type 2 and susceptibility to infection. Other organs such as kidneys, pancreas and liver can also be affected.

- An early symptom in infancy is light sensitivity.
- Vision problems due to RP will decline rapidly, although the degree of visual impairment varies. The vast majority of young adults with Alström will be blind by the age of 14 years.
- At the age of 4–6 years hearing loss is evident in most children with Alström syndrome.
- Over 500 cases of Alström syndrome are known worldwide.
- This is a condition causing gradual degeneration of the retina. As a result, the eye gradually loses its ability to capture and process light, night vision decreases and the child experiences loss of peripheral vision.
- An early sign of loss of vision may be that the child trips often and has trouble finding things. Vision will decline rapidly, although the degree of visual impairment varies.

Charge

- This is a syndrome that is autosomal dominant.
- The name is derived from the first letter of the organs that are most often affected. **C** stands for coloboma (defect in the development of the eye), **H** for heart defect, **A** for atresia choane (narrowing of the opening between the nasal cavity and the pharynx), **R** for retardation of growth and/or development, **G** for genital anomalies and **E** represents ear anomalies with or without hearing loss.

- The incidence is around 1 in 12,000 (Firth and Hurst 2005).
- Malformation of the outer and middle ear is common; malformation of the bones of the middle ear results in a conductive hearing loss.
- A malformed cochlea occurs in 90%. Small or absent semicircular canals cause sensorineural hearing loss and balance impairment.
- Many children with CHARGE have severe problems with breathing, swallowing or speech, and some have severe learning difficulties.

Congenital rubella syndrome (CRS)

- CRS is also known as German measles.
- Pregnant women who become infected with rubella in the first 12 weeks of pregnancy can infect their baby with the virus.
- The classic three features of congenital rubella syndrome are deafness, blindness and heart disease, although there are other possible symptoms that can occur such as learning difficulties and developmental delay.

Older people

- Degeneration of the macula, which is associated with older age, causes central visual field loss.

- Age-related hearing loss in combination with different diseases affecting the eye is very common over the age of 65 and prevalence increases with age.

Older people with combined visual and hearing loss comprise the largest group of people who have deafblindness.

General themes to consider when working with deaf and hard of hearing clients

Anna Middleton

This chapter considers general communication issues that are relevant to deafness and hearing loss. Specific issues pertinent to people who are deaf due to NF2 and people who have deafblindness are considered in Chapters 4 and 5 respectively.

Seeing a hearing aid

- When meeting a deaf or hard of hearing person for the first time, seeing a hearing aid may offer some clues as to the form of communication the person uses.
- It is possible that they identify more with the Hearing World and if so will use lip-reading and speech to communicate.
- Alternatively they may still be culturally Deaf and prefer to use sign language, but for interactions with hearing people prefer to utilise the residual hearing they have as

Working with Deaf People – a Handbook for Health Professionals, ed. Anna Middleton. Published by Cambridge University Press. © Cambridge University Press 2010.