

Hearing-impaired young people – a physician’s guide

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ABSTRACT

Physicians reading this will have a broad range of in-depth knowledge about their own subspecialty. However, in daily medical practice there are topics of which all physicians should have some knowledge. Those who deal with young people should have some knowledge of the needs of the hearing-impaired population within this group of patients. This article is intended to provide an overview of young people with hearing impairment (HIYP), the challenges they face and what we can do to help them. In this paper, we assume that data published regarding hearing-impaired children apply to HIYP from 13 years (the age at which the transition process begins) to 25 years of age (the age at which ‘youth’ according to the World Health Organization and the Education Health Care Plan ends).

KEYWORDS: Deaf awareness, hearing impaired, young people

Introduction

In 2015, there were at least 48,932 children with hearing impairment across the UK. This had increased 2% from the preceding year. In school-aged children in the UK, 87% communicate using spoken English or Welsh and only 10% use sign language in some form.¹ The proportion of deaf children with cochlear implants stood at 7% in 2011 and 8% in 2013.¹

At least 40% of young people with hearing impairment (HIYP) have complex health needs involving several other conditions requiring medical care, such as cerebral palsy, epilepsy, renal impairment and Down’s syndrome; therefore, they may be seen in a wide range of inpatient and outpatient settings.

It’s important to understand that every HIYP is different – with a different level of deafness, hearing equipment and communication preferences. Some of these young people will be from deaf families and therefore may be part of the deaf community, which will offer them help and support.

The National Deaf Children’s Society (NDCS) has given 11 tips for communicating with HIYP (Box 1). These should be used in clinical settings. Most are common sense, but how many of us can say we are truly deaf aware? Some trusts run deaf awareness courses, which can be very beneficial for all physicians and patients. Given that hearing loss affects 1 in

Box 1. 11 tips for communicating with deaf young people

1. Find out how they communicate – not all deaf children use British Sign Language (BSL). Every deaf child will have a preferred way of communicating, so find out if they use speech, BSL or a mixture of both. If they do use BSL, ask their parents if they will need an interpreter
2. Get their attention
3. Face them when you are talking
4. Speak clearly and naturally
5. Don’t cover your mouth
6. Use visual cues where possible
7. Make it clear what the topic of conversation is
8. Stand with your face to the light
9. Reduce background noise
10. Never give up or say ‘I’ll tell you later’

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6 adults overall and that the proportion of older patients in the population is increasing, one could argue for deaf awareness being included in mandatory training.

Flexible communication with patients

HIYP may communicate in a range of ways. While most use spoken language as their first language, some will use British Sign Language (BSL) or sign-supported English. Many also use lip-reading, to variable degrees. Some use picture systems and those with dual sensory impairment may use hands-on signing. Others use a combination of these. They may also change communication type depending on the context in which they are communicating and gradually over time, if their hearing loss or vision worsens. Some patients carry a personal communication passport describing their most effective means of communication.² The most important thing to do is to ask them how they want to communicate and then help to facilitate this. From 1 August 2016 onwards, all organisations that provide NHS care are legally required to follow the Accessible Information Standard. This standard aims to make sure that people who have a disability, impairment or sensory loss are provided with information that they can easily read or understand, and support, so that they can communicate effectively with health and social care services.³ This may

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require booking a BSL interpreter for a patient's outpatient appointment or to come to the ward to explain their care. If BSL is their first language and you need to send a letter, this may have to be converted from written English into BSL recorded onto a DVD that can then be posted to the patient. Importantly, this process shouldn't take any longer than it does for a written report to be sent out.

HIYP also need to be able to contact services, so you should be contactable by email, text, video message or online booking service. Patients given only a telephone contact number have to rely on others to help them, and thus cannot be independent.

HIYP may use a variety of equipment to help them to hear, such as hearing aids or cochlear implants, and to connect with mainstream technologies, such as radio aids and streaming devices that send sound from TV, mobile phones and other portable electronic equipment to their hearing aids. If they come into hospital, it will be essential to make sure they have their equipment with them and that it is working. Lack of access to appropriate communication support poses significant clinical risk.

Educational attainment in HIYP

Hearing impaired young people face many challenges. Just 41.1% of deaf children achieved five GCSEs (including English and maths) at grades A* to C in 2014, compared with 64.2% of children with no identified special educational need. Expressed in a different way, nearly two thirds of hearing impaired young people (58.9%) are failing to achieve five good GCSEs.⁴ A child with mild hearing loss can miss up to 50% of what is being said in the classroom. However, with the right support of professionals they can achieve as well as other children.⁵

Unlike for GCSEs, no breakdown is provided for A-level results in England, Wales and Northern Ireland by type of special educational need or disability. However, success rates for deaf students at Scottish Highers (Scottish equivalent of A-levels) and destination data are published, showing a considerably lower attainment in deaf children compared with hearing students at Highers level. Significantly, the proportion of deaf young people leaving school for higher education is less than half that of hearing students.⁶

Support in education

78% of school-aged deaf children attend mainstream schools where there is no specialist provision; 7% attend mainstream schools with resource provisions, 3% attend special schools for deaf children and 12% attend special schools not specifically for deaf children.¹

HIYP can get support in school from two main sources:

- from their school special educational needs co-ordinator
- there are at least 1,461 teachers employed as 'teachers of the deaf' in the UK who work directly with children in a school setting.¹ They provide practical support to the children and their teachers on how to use their hearing equipment and how to optimise a hearing-impaired child's learning. On average, each 'teacher of the deaf' has a theoretical caseload of 47 deaf children each, but there is considerable local variation in resource provision.

Outside school, young people and their families have access to the NDCS, which has a wealth of information online.

The NDCS has its own website for young people, 'The Buzz', and also runs local groups for deaf young people and their families. The society also provides information for professionals.

HIYP may be able to get disabled students allowance for extra equipment and support if they attend college or university. Each provider of further education should provide advice and support for sensory impairment.⁷ HIYP may be able to apply for a supported internship or an access to work grant to help them to start work or to stay in work.⁸ HIYP also have access to their local social services' hearing impaired provisions.

Adolescence and transition into adult services

Adolescence is the time when we notice a significant dropout rate from audiology services in our HIYP. In secondary school, HIYP with mild and moderate losses don't appreciate how much information they are missing without amplification.

In a recent local survey of professionals working with adolescent hearing aid users, we asked what reasons the adolescents had given for not wanting to wear their aids. The most common reason was feeling self-conscious and not wanting to be singled out from their peers. When we asked the professionals what reasons they felt were important in the dropout rate, they stated self-image and lack of family support as the most important factors, along with fear of being bullied.

'If transfer to adult services is handled badly, there is a risk that the young person will "drop out" from medical services altogether.'⁹ Government guidance strongly recommends that transition planning should begin at the latest by the age of 13–14 (Year 9 in schools in the UK).¹⁰ Involvement of children in the gradual development of independence can start from the age of about 6–7 year.¹¹

In 2011, the NDCS produced a report entitled *Over to you*. It was based on a 1-year project, meeting with 100 HIYP from 12–18 years of age to examine their experiences of audiology services.¹² One of its key messages was that HIYP want greater involvement in making the transition from children's to adult services. This includes receiving information and guidance on the process, and on where their new service will be.

Bridging the gap

The withdrawal of HIYP from audiological services has been recognised and steps are being taken to try to prevent this.^{13,14} Young people are encouraged to understand their diagnosis and to self-manage their hearing aids and appointments. They are given information on tinnitus, balance, vision care and relevant services, such as hearing therapy, social care, careers and local support groups. Dedicated young person or transition clinics have been introduced in many areas. It is important to provide an adult learning disability clinic in audiology as well. Guidelines recommend the allocated keyworker follows up with young people who do not attend the adult service.¹³ Audiology departments should have transition protocols and pathways that are agreed by all key stakeholders and commissioners.¹³ Medical and audiological staff in paediatric and adult audiology need access to relevant training to support successful transition of HIYP. Physicians who recognise that an HIYP has dropped

out of follow-up, or is having audiovestibular difficulties, should offer to re-refer them.

Cause of the hearing impairment

As audiovestibular physicians, it is our role to look into the cause of childhood hearing impairment in line with national guidelines.^{15–17} In some areas, this service is provided by community paediatricians in association with ear, nose and throat surgeons. This knowledge can help families to deal with the diagnosis of hearing impairment and help to provide vital information to the patient and their families about whether their hearing loss is likely to progress, is inherited, or is part of a syndrome with associated medical problems requiring further management, such as Alport or Pendred Syndromes. In some diagnoses, steps to be taken to prevent further hearing loss, such as avoiding head injury in widened vestibular aqueduct patients or aminoglycosides in the mitochondrial mutation A1555G, should be outlined. The common aetiological investigations are listed in Table 2. These aetiological investigations need to be revisited during transition to identify certain medical conditions: for example, the retinal pigmentary changes that may occur in the second decade in Usher syndrome, and renal dysfunction in early adult life in Alport and Branchio-Oto-Renal syndrome. It is important that HIYP with such diagnoses attend their renal, genetic or ophthalmological follow-up.

Approximately half of hearing impairment in young people is genetic in origin. Many HIYP ask what has caused their hearing loss, and whether it will affect their own children. We work with clinical geneticists to provide them with genetic testing and counselling. It should be noted that some parts of the deaf community are strongly opposed to prenatal genetic testing as they feel their hearing loss isn't a disability. They have a real fear that genetic technology could reduce the number of deaf children being born thereby threatening the viability of the deaf community.¹⁸

Help for tinnitus

Tinnitus in young people is under recognised, yet can be effectively managed in the right hands, especially with early intervention. The 2015 British Society of Audiology guidelines help relevant professionals to understand and refer these patients to the right places for help.¹⁹ Hearing therapists and some audiologists have training in this field. In some cases, tinnitus can be a marker of a mental health problem (such as anxiety and/or depression), which may need specialist psychological management. Tinnitus may also coexist with hyperacusis and/or misophonia.

Help for balance

Published reports have identified vestibular dysfunction in 30–70% of hearing-impaired children.^{20–25} This is under recognised. These children have delayed motor skills or may be described as 'clumsy'. Referral to audiovestibular medicine or a dedicated ear, nose and throat balance clinic for a balance assessment can help in several ways: vestibular physiotherapy helps children with vestibular deficits and some may require safety advice and the presence or absence

Table 2. Common aetiological investigations into bilateral hearing loss

Investigation	Possible finding
MRI IAMs and brain	<ul style="list-style-type: none"> > Absent auditory nerve > Mondini dysplasia > Widened vestibular aqueduct
Genetic testing	<ul style="list-style-type: none"> > Connexin 26 and 30 – commonest genetic mutation causing non-syndromic hearing loss > A1555G, which predisposes to aminoglycoside ototoxicity
Blood tests	<p>CMV IgG: a negative result excludes congenital CMV, a positive result requires retrieval of the Guthrie card (newborn bloodspot) to test for CMV DNA to confirm congenital CMV</p> <p>Positive test commonest non-genetic cause of childhood hearing loss in UK</p> <p>Autoimmune screen in cases of sudden/progressive/fluctuating hearing loss</p>
Ophthalmology review	<p>40% of children with permanent congenital hearing loss have ophthalmic conditions, such as refractive errors²⁶</p> <p>Particularly important to identify retinitis pigmentosa to detect Usher syndrome</p>
Urine dipstick	Haematuria and proteinuria are found in Alport syndrome
Renal ultrasound	Especially if signs of Branchio-Oto-Renal syndrome
Family audiograms	Approximately half of hearing impairment is genetic
ECG	If hearing loss is severe to profound, to identify Jervell Lange Nielsen syndrome (a cause of sudden death)

Causes of hearing loss that can be progressive are shown in bold
 CMV = cytomegalovirus; ECG = electrocardiogram; IAM = internal auditory meati; IgG = immunoglobulin G; MRI = magnetic resonance imaging

of vestibular dysfunction helps to determine the aetiology of their hearing loss.

Vertigo in HIYP may be related to underlying vestibular dysfunction or be due to migraine, but cardiac, neurological problems, drugs and psychological causes are also included in the differential diagnosis.

Conclusions

- > Hearing loss is common in medical practice.
- > Physicians need to be both deaf aware, flexible and holistic in their approach to enable effective and safe communication with HIYP.
- > It is important for physicians to be aware of the medical conditions associated with hearing loss and to refer them appropriately.

- > HIYP no longer attending audiology should be encouraged to attend or be re-referred.
- > We can all work harder at being deaf aware, not just in young people but in those of all ages. ■

Conflicts of interest

The authors have no conflicts of interest to declare.

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