Understanding Chromosome & Gene Disorders

Hearing Loss

rarechromo.org

Hearing loss

Growing up with a hearing loss myself, I am often approached by parents of childrenwith additional needs and hearing loss for information and support. Parents of children with additional special needs who receive a hearing loss diagnosis often experience the same emotional turmoil as other parents whose children have been diagnosed with a hearing loss. I've heard various stories about how these parents often feel even further isolated from the community than before. However, there is acommunity willing to help and offer support.

I was born with a mild case of Treacher Collins syndrome, a genetic condition affecting the development of the facial bones and tissues. As a result, I have a moderate conductive hearing loss in both ears. To help with this, I wear a bone conduction hearing aid that I had implanted at 5 years old. Many people who meet me for the first time are often surprised to learn of my hearing loss and that I wear a hearing aid, as it is covered with my long hair, even though I have no problem showing it off!

I am very fortunate to be able to share my experience of living with a genetic condition and growing up with a hearing loss. There were ups and downs, but I hope my experiences will give a bit of hope to others on their journey.

This guide has been compiled mainly as a signposting guide to help those with ahearing loss and their families to find the best available information.

Contents

Diagnosing a Hearing Loss

Diagnosing a hearing loss early is vital for every child, regardless of whether or not they have additional needs. However, it is often the case for children with additional needs that due to medical complications, the child's other needs take a higher priority. But it should be noted that any support regarding a child's hearing loss can help improve their quality of life and ensure the child can still be a part of their local environment.

If a child is born with a hearing loss, in the UK and many other countries, there is a national new-born hearing screening program which can detect a hearing loss just a few days after birth. If a baby fails this test, they are referred to an audiologist.

There can be various challenges associated when diagnosing a child with hearing loss who also has additional special needs. Various hearing assessments are used to test a child's hearing, depending on their developmental age. Audiologists may use a variety of different methods.

If the child is unable to complete the behavioural hearing assessments, it may be necessary to perform an Auditory Brainstem Response test when the child is asleep or sedated. This can give a more accurate result when children are unable to respond to sounds by moving, or when the child has sensory issues and may be uncomfortable with certain sounds or wearing headphones.

Hearing Assessments Used for Different Age Groups

Age group	Main hearing assessment used	Alternative assessments that can be used if results are inconsistent or if there is developmental delay
New-born – 3 months	OAE (Otto Acoustic Emissions), ABR (Auditory Brainstem Response)	
7 months – 3 years	VRA (Visual Reinforcement Audiometry)	Distraction, speech discrimination assessment, OAE, and ABR (if sedated)
3 years – 5 years	Performance test	VRA, distraction, OAE,ABR (if sedated) and speech discrimination assessment
5 years plus	PTA (Pure Tone Audiometry)	VRA, distraction, OAE,ABR (if sedated) and speech discrimination assessment

Different Types of Hearing Loss

In normal hearing, sound waves travel into the ear along the ear canal where they reach the middle ear (the ear drum and three tiny bones called the ossicles). The ear drum vibrates, and these vibrations travel to the ossicles and then to the cochlea. The cochlea is the inner part of the ear and generates signals which are passed along the auditory nerve before being processed by the brain as sound. Conventional hearing aids work with this principle.

Diagram of the Ear

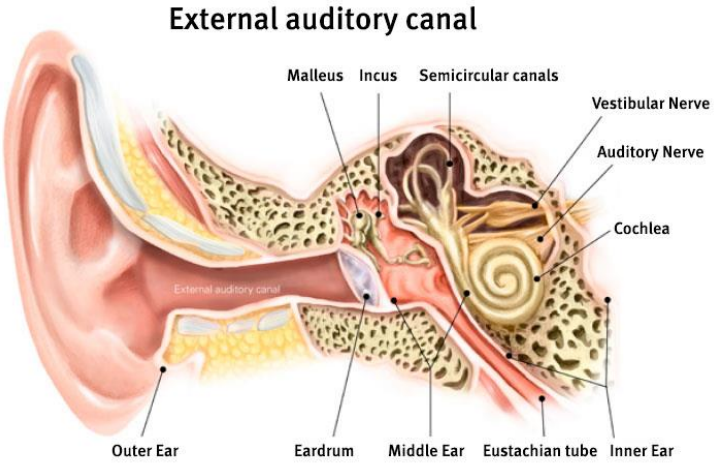

Image Courtesy of Oticon Medical

Glue ear

Many children experience 'glue ear' – where fluid builds up behind the ear drum. When this happens, it prevents sound travelling effectively to the inner ear. Having glue ear may cause temporary deafness. Most of the time, glue ear clears up spontaneously without any treatment and hearing usually returns to normal. Some children who have persistent glue ear may require 'grommets' to help drain the fluid from the ear. Grommets are tiny tubes which are inserted into the eardrum, to keep the middle ear ventilated and prevent further fluid build-up. This requires a short operation under a general anaesthetic. Some children may require more than one operation if the tubes fail to stay in place. Most of the time, grommets are successful. This type of hearing loss is called a 'conductive hearing loss'.

Conductive hearing loss

Conductive hearing loss is where sound is unable to travel through the ear to the auditory nerve. Conductive hearing loss also occurs with glue ear and when children have very narrow ear canals or are born with under-developed or absent parts of the ear.

Sensorineural hearing loss

A sensorineural hearing loss is where there are problems with the inner ear, sometimes with the cochlea or auditory nerve (the nerve that sends signals to the brain about sound).

Mixed hearing loss

Some children may have a mixed hearing loss – where they have both a conductive and sensorineural hearing loss.

Hearing Aids

Many types of hearing loss can be managed by using hearing aids. Most hearing aids are now digital. Digital hearing aids process sounds to allow the user to access sounds they are unable to hear. It is important to realise that hearing aids alone will not enable perfect hearing, and communication tactics will always be required.

There are different types of hearing aids for different types of hearing loss. The most common type of hearing aids are 'Behind-The-Ear' (BTE) hearing aids. These can be used for most children and adults of any age with a sensorineural, conductive, or mixed hearing loss. BTE's consist of an 'ear-mould' that fits in the outer ear and a receiver that sits behind the ear.

There are also 'In-The-Ear' or 'In-The-Canal" hearing aids that some children and adults prefer. However, for growing children, they may not be the best option as they will need to be replaced regularly as the child grows. They also may not have the same features and functionality of BTE hearing aids.

Cochlear Implants

A cochlear implant works by stimulating the auditory nerve cells with a signal produced by an implant. A transmitter worn on a hearing aid that sits behind the ear sends sounds to an internal implanted receiver. These sounds are then turned into electrical signals which are processed by electrodes in the cochlea which then carry sound from the auditory nerve to brain.

A Cochlear Implant Provided Cochlear

Image courtesy of Cochlear Europe Limited

More information and support for children with or considering a cochlear implant can be found at the Cochlear Implanted Children's Support Group website:

- https://www.cicsgroup.org.uk/

Bone Conduction Hearing Systems

Bone Conduction Hearing Systems (BCHS) also known as Bone Anchored Hearing Aids (BAHA) consist of an implant, abutment and processor. They work by conducting sound waves through an implant in the skull directly to the inner ear.

This type of hearing system fills an important gap for those people who are unable to wear conventional 'Behind-The-Ear' (BTE) hearing aids. This may be due to malformed ears, very narrow ear canals or frequent ear infections. Children awaiting the BAHS operation, or those undergoing assessment have the option to trial a BAHS on a soft-band, similar to a sports band or head band. This works with the same hearing principle except the BAHS is attached to a plastic disc on a soft-band.

Ponto series - Bone Conduction Hearing Aids by Oticon Medical

Image courtesy of Oticon Medical

Whichever hearing aid an audiologist decides is best for your child, there may or may not be some challenges. One common problem is trying to keep the hearing aids on. When I was little and wearing BTE hearing aids, my parents had to use double-sided sticky tape to keep my hearing aids in place due to the shape of my ears. Some babies will pull the aids out. This happens less as they grow and become used to the aids. Some children may not like wearing the hearing aids at first, and this is completely normal! It is advised to try to build up the minutes/hours of use. For example, try 30 minutes one day, then an hour the next day (if tolerated well enough). There can occasionally be feedback from the hearing aids which sounds like high pitched squealing or whistling, which can feel very unpleasant for the user. Try changing the batteries or checking that the aid is inserted correctly, if this fails, it's best to contact the audiologist.

Communication Methods

Learning some simple sign language can be very useful. But there are also things that may help to ease communication between parent and child, with or without hearing aids. The most important thing is to make certain the child or person with hearing loss can see your face, including your lips! I unconsciously lip-read and can get quite frustrated when I can't see the speakers' face. It also helps to speak 'normally', without exaggerating words. The National Deaf Children's Society (NDCS) have some useful tips for communicating with deaf children.

When in group situations, I like to be in a position where I can see everyone. It can be quite difficult to pinpoint where sounds are coming from, but when I can see most people, I can relax a little more and enjoy the atmosphere.

Alternative communication methods can be helpful if children or adults are unable or unsuitable for hearing aids, or even just during night-time or morning routines when the hearing aid is not in place. There are many different communication methods available but it may require a period of trial and error to find which methods the child or adult finds most comfortable.

Sign languages use hand shapes, facial expressions and body language to communicate. As with different spoken languages, many countries have their own sign language. In Britain there is British Sign Language (BSL) in USA there is American Sign Language (ASL).

In the UK, a health visitor will be able to sign post you to a speech and language therapist for help with learning Makaton sign language. Makaton uses signs and symbols. More information and support for Makaton can be found here:

- www.makaton.org

There are many different ways to learn sign languages, some charities may offer some sessions, whilst there are also some paid courses at schools and colleges. To find an approved centre in the UK, you can visit the Signature website:

- www.signature.org.uk

Some children with significant learning or development delay may not be able to learn sign or spoken language, and other communication methods may be more suitable, such as Picture Exchange Communication System (PECS). Many Unique families make their own pictures and some are available online:

- www.pecs-unitedkingdom.com

Unique publishes a separate Communication guide, freely available online in the Practical Guide series (https://rarechromo.org/practical-guides-for-families/).

Hearing Loss Jargon

When receiving a diagnosis of a hearing loss, there can be lots of different words used which can be confusing and overwhelming. You're not alone! A helpful glossary can be found on the NDCS website:

- www.ndcs.org.uk/information-and-support/glossary

Additional Support

It can be helpful to remember that audiologists are specialised health care providers but are not medical doctors. They need to know what conditions a child has and what impact they have on them as well as any planned surgery/treatment in order to plan future appointments so they don't clash and cause extra stress for families.

In an ideal world, all medical professionals would have some 'deaf awareness training' – when this is not the case, it may be useful to pass on your 'Unique' card that has a note about hearing loss too. Please send an email to info@rarechromo.org if you would like an awareness card.

A vital source of support in the UK for children with a hearing loss and their families is a Teacher of the Deaf (ToD) - a teacher who has an additional qualification to enable them to teach deaf children. They provide support to deaf children, their parents and families, and to other professionals who are involved with the child's education. Other specialists include specialist teaching assistants, communication support workers and educational audiologists.

There also numerous organisations who are able to offer support, such as the National Deaf Children's Society, The Royal National Institute for Deaf People and the American Speech Language Hearing Association, to name a few.

- www.ndcs.org.uk
- www.asha.org/public/hearing/Hearing-Loss-Organizations-and-Associations/
- www.rnid.org.uk

> **"It can be a challenge but it is important to stay consistent in having a child wear their hearing aids or device. Early diagnosis of hearing loss and intervention can significantly help a child's development."**

Audiograms

An audiogram is a visual representation used to record the results of hearing assessments. It is a visual representation of a person's hearing. In audiology, the grey shaded area on the audiogram is often referred to as a "speech banana". Speech information spreads throughout the "speech banana". People need to be able to hear speech sounds above and below the speech banana in order to hear all of the sounds in words.

Audiogram of Familiar Sounds

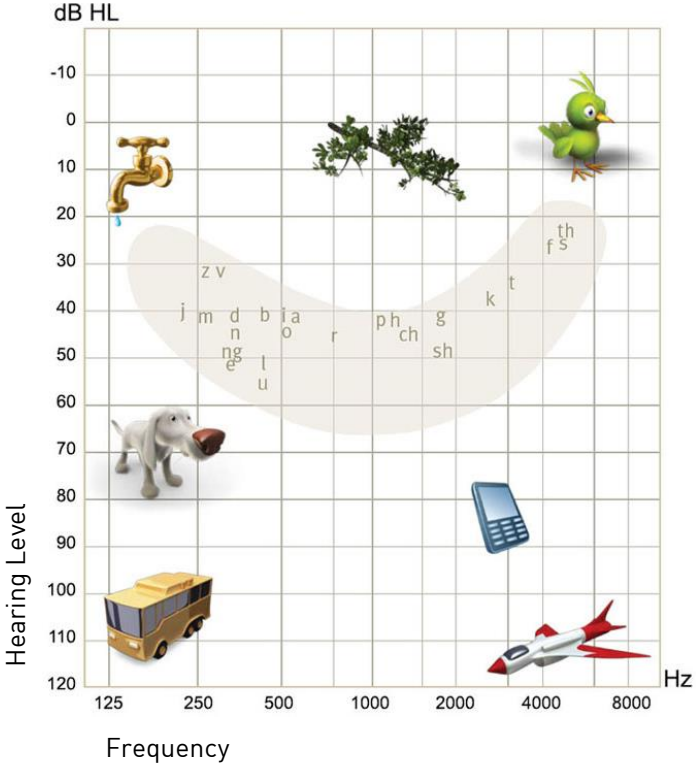

Image courtesy of Oticon Medical

Useful Organisations

National Deaf Children's Society (NDCS)

Website: https://www.ndcs.org.uk/
Email: helpline@ndcs.org.uk
Helpline: 0808 800 8880
Monday to Thursday 9am – 5pm and Friday 9am - 12:30pm
(Free from all UK landlines and major UK mobile providers)
SMS: 0786 00 22 888
Monday to Thursday 9am – 5pm and Friday 9am - 12:30pm
(Texts are charged at your standard network rate or taken from your monthly allowance).
Facebook page: https://www.facebook.com/NDCS.UK
Providing independent information, advice and guidance across the UK on a range of topics related to childhood deafness.

The Elizabeth Foundation

Website: https://elizabeth-foundation.org/
Email: info@elizabeth-foundation.org
Telephone: 023 9237 2735
Facebook: https://www.facebook.com/elizabethfoundation
UK charity providing services to help babies and preschool children with all degrees of deafness.

Royal Association for Deaf people (RAD)

Website: https://www.royaldeaf.org.uk/
Email: info@royaldeaf.org.uk
Telephone: 0300 688 2525
Text Phone: 0300 688 2527
SMS: 07851 423 866
Children and Families SMS: 07773 978 767
Email: cft@royaldeaf.org.uk
Offering support to children, young people and their families.

(RNID) Royal National Institute for Deaf People

Website: https://rnid.org.uk/
Telephone: 0808 808 0123
Address: Brightfield Business Hub, Bakewell Road,
Orton Southgate, Peterborough, PE2 6XU.
Supporting people who are deaf, have hearing loss or tinnitus.

DELTA (Deaf Education through Listening and Talking)

Website: https:// deafeducation.org.uk/
Email: enquiries@deafeducation.org.uk
Telephone: 0300 365 7200
Address: Care of Connevans Ltd Bridge House,
Nutfield Road, Merstham, Redhill, Surrey RH1 3EB
Providing information and resources for families and professionals.

Ewing Foundation

Website: https://ewing-foundation.org.uk/
Email: info@ewing-foundation.org.uk
Telephone: 07778 599939
Address: 15 Great College Street, London SW1P 3RX
A national charity, promoting inclusion and achievement for deaf children through listening and speaking.

I CAN

Website: https://www.icancharity.org.uk
Email: info@ican.org.uk
General enquiries: 020 7843 2510
Address: Angel Gate, Hall Street, London, EC1V 2PT
Offering practical help and information to children with communication difficulties.

The Listening Room

Website: https://thelisteningroom.com/
The Listening Room features free, fun activities and resources to support the development of speech, language, and listening skills in people of all ages with a hearing loss.

SENSE

Website: https://www.sense.org.uk/
Support for Adults: https://www.sense.org.uk/our-services/support-for-adults/
Support for Children: https://www.sense.org.uk/our-services/support-for-children/
Sense supports everyone living with complex disabilities. For everyone who is deafblind.

Sign Language

UK:

British Sign Language (BSL)

Website: https://www.british-sign.co.uk/
In the UK the most common form of Sign Language is called British Sign Language (BSL).

Makaton

Website: https://makaton.org/
Email: help@makaton.org
Telephone: 01276 606760
Address: The Makaton Charity, Suite 3, Unit 4.3
Frimley 4 Business Park, Frimley, Surrey GU16 7SG
Makaton uses speech with signs (gestures) and symbols (pictures) to help people communicate.

Signlive

Website: https://signlive.co.uk/
Email Us: hello@signlive.co.uk
Telephone: 0330 822 0288
Access to BSL interpreters

USA:

American sign language (ASL)

Website: https://www.nad.org/resources/american-sign-language/what-is-american-sign-language/
ASL is used predominantly in the United States but also in many parts of Canada.

Signing groups

There are also signing groups that are used for children with communication difficulties, who might not necessarily have hearing loss. These groups might be useful for children with or without hearing loss.

Sing and Sign

https://www.singandsign.co.uk/baby-signing/what-is-sing-and-sign

Schools for Deaf Children (UK)

There will be some mainstream school settings that have a separate unit for children with a hearing impairment. For those that aren't able to attend mainstream schools there are numerous specialist schools for deaf children throughout the UK. In Wales, there are no specialist schools for deaf children, most children attend a mainstream school but there are a few specialist schools for children with difficulties that are not specifically restricted to hearing.

The National Deaf Children's society has a list of UK schools for deaf children: https://www.ndcs.org.uk/information-and-support/education-and-learning/choosing-a-deaf-friendly-school/special-schools-for-deaf-children-in-the-uk/

Glue Ear App

The Hear Glue Ear app, developed by Cambridge Digital Health, in collaboration with Dr Tamsin Brown and the Cambridge Hearing Trust Charity is a valuable tool to help families manage their child's glue ear at home. The app includes audiobooks, songs and listening games for children. It also provides accurate, up-to-date glue ear information for parents and a hearing screen to monitor fluctuations in children's hearing levels over time.

- https://www.cambridgedigitalhealth.co.uk/hear-glue-ear-app

Notes:

Inform Network Support

Rare Chromosome Disorder Support Group
The Stables, Station Road West, Oxted, Surrey RH8 9EE,
United Kingdom
Tel: +44(0)1883 723356
info@rarechromo.org | www.rarechromo.org

Join Unique for family links, information and support.

Unique is a charity without government funding, existing entirely on donations and grants. If you can, please make a donation via our website at: www.rarechromo.org/donate

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This booklet was compiled by Unique in 2022 (AKP) and kindly reviewed by Reena Patel, Paediatric Audiology Clinical Lead at Whittington Health, London, and Marion Atkin, Chief Audiologist at Queen Elizabeth Hospital, Birmingham.

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