Best practice guidance for the management of hearing issues in children with Down syndrome

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On behalf of Down Syndrome Medical Interest Group (DSMiG)

Down Syndrome Medical Interest Group

In Down syndrome, most associated medical conditions are not unique but are over-represented within the condition with a preventable secondary handicap. DSMiG are a network of health professionals who aim to provide equitable treatment for all with Down syndrome (DS), and develop evidence-based guidelines for a minimum safe standard of essential care for the management of relevant medical issues. The guidelines for the management of hearing issues in children with Down syndrome in the United Kingdom and Ireland were updated in 2017.

Hearing issues in Down syndrome

A number of hearing issues are more common in Down syndrome, compared to the typically developing child. 4-6% of children with DS have a sensorineural hearing loss (SNHL) identified at newborn screening (c.f. 0.4%). This increases to 20% by early adulthood (60% unilateral). Presbyacusis occurs 20-30 years earlier than in typically ageing adults. Otitis media with effusion (OME) is significantly more common, with 93% & 68% incidence at 1 and 5 years old respectively (c.f. 45% & 12%). Children with DS are vulnerable to chronic middle ear disease and cholesteatoma, due to chronic eustachian tube dysfunction. Conductive hearing loss can result from ossicular malformations, and from excess wax build up in a congenitally small external auditory canal.

Standard of care

A multidisciplinary approach should be taken by a team of professionals with an interest in the management of Down syndrome

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<th>Paediatric audiologist</th>
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<td>Speech and Language therapist</td>
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<td>Teacher for Hearing Impaired</td>
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Guidelines

Protocol for Audiological Assessment
- Newborn hearing screening
- 8-10 months old
- 6 monthly until 2 years old
- At least yearly until adulthood, then 2-yearly

Audiological assessment
- Developmental age, rather than chronological. ERA can be used if necessary
- Regular dewaxing is an essential part of follow-up

Management of OME
- Hearing aids are first line treatment (NICE 2008)
- Grommet insertion is decided on an individual patient basis
- Softband/BAHA are well tolerated for recurrent OME and middle ear disease
- Indications for adenoid surgery are as for typically developing children
- Clinicians must be aware of the higher risk of cholesteatoma

Sensorineural loss
- Hearing aid use should be actively encouraged
- Audiological indications for a cochlear implant are the same as for a typically developing child
- Cognitive impairment is not a contraindication for a cochlear implant

Transition from Paediatric to Adult care
- There should be direct transfer of care to a named practitioner for ongoing follow-up

Figure 1. Multidisciplinary team approach to the management of hearing loss in the child with Down syndrome

Conclusion

Children with Down syndrome should have their hearing issues managed via a multidisciplinary approach, by clinicians with a specific interest and areas of expertise. Follow-up should be cohesive, to maximise educational and social development of children, and enable independence and quality of life for adults.

References
2. NICE guidance – Otitis media with effusion in the under 12s surgery. At - https://www.nice.org.uk/guidance/cg80