Best Practice Guidance for the management of hearing issues in Down syndrome

Background

Up to 50-70% of people with Down syndrome will experience impaired hearing at some point in their life [1 - 4]. Hearing losses may be conductive, sensorineural or mixed in nature, and may be temporary or permanent. The patterns of hearing loss presenting in Down syndrome change throughout life, with otitis media with effusion (glue ear) the most common cause in childhood, and sensorineural deafness becoming more prevalent with age.

Otitis media with effusion (OME or glue ear) affects up to 35% of children with Down syndrome at birth and 93% at 1 year old, reducing to 68% and 38% by the ages of 5 and 8 respectively [6.7,8]. There is a higher incidence of ossicular abnormalities in Down syndrome which may present with a conductive hearing loss [9]. The incidence of sensorineural hearing loss identified at newborn hearing screening in children with Down syndrome is higher than in the general population at 4-6% [3, 8]. Presbyacusis (age-related sensorineural hearing loss) is more common, and develops significantly earlier in adults with Down syndrome [1-4]

Early hearing loss has a significant impact on the speech and language development of children with Down syndrome, resulting in a disproportionately more severe speech delay than would be predicted by IQ alone [5]. It is important to consider hearing loss, either as a cause or an exacerbating factor, as a part of an assessment of behavioural problems, mental health or cognitive decline in children and adults with Down syndrome.

Health care professionals need to have a heightened awareness of the possibility of hearing loss, with the aim of identifying any impairment and offer appropriate intervention early, to maximise the educational and social development of children, and enabling independence and quality of life for adults.

It is important to take into consideration, at all ages, the person's communication needs, any additional medical needs and developmental and behavioural phenotype

when assessing and offering an intervention for the management of their hearing difficulties.

Recommendation:

A multidisciplinary team approach should be applied for the management of hearing issues in the child with Down syndrome. This should include a paediatric audiologist, otolaryngologist and paediatrician, with strong links to speech and language therapists and teachers for the deaf. Correspondence should be routinely shared with the parents/carers and, with parents' consent, with all members of the team [6].

Hearing Screening & Surveillance:

Studies have shown a poor correlation between caretakers' perception of hearing loss and objective hearing loss in children with Down syndrome, therefore regular objective hearing assessments should be performed as parents may underestimate hearing loss [10].

Hearing Assessment:

At all ages, people with Down syndrome have narrow ear canals which predispose to accumulation of wax. This may affect impedance testing and hearing [14].

Most people with Down syndrome are able to respond to standard test procedures e.g. distraction, speech discrimination, pure tone audiometry (play or standard) or visual reinforcement audiometrywhen carried out by testers with expertise in working with people with learning disability. However in the young infant it can be a challenge to undertake these tests. Where a dual diagnosis, e.g. autism, visual impairment etc. exists, the assessments can be particularly challenging.

Threshold measurement tests appropriate to developmental age must be used. Objective audiometry e.g. brain-stem evoked audiometry, may be required if a hearing level cannot be established reliably [11, 12, 13].

Because of an increased incidence of sensorineural as well as conductive loss, the frequency range tested should include 8000Hz whenever feasible, as this may be an early warning of impending sensorineural deafness [14], [15].

Recommendation:

All children with Down syndrome should have new-born hearing screening followed by targeted hearing assessments, initially at 6 to 10 months and then six monthly till the age of two and to continue at least annually throughout school-age years.

A hearing assessment is recommended at least two yearly throughout adult life or more frequently if there are concerns. This should include measurement of auditory thresholds, impedance testing (tympanometry) and otoscopy.

Guidance for management of hearing issues:

1. Children and adults with Down syndrome have relatively narrow ear canals predisposing them to accumulation of wax, which may affect hearing and impedance testing. Regular dewaxing should be part of their management. This is particularly important as progressive middle ear disease or cholesteatoma can be missed, if wax is not actively managed.

2. Otitis media with effusion (OME, glue ear) can be managed conservatively, with hearing aids (traditional hearing aids applied behind the ear or the BAHA Soft Band) or with ventilation tubes (grommets) [16 - 19]. Each child's case should be considered on its own merits by a multidisciplinary team with experience in dealing with glue ear in children with Down syndrome.

3. NICE Guidelines (2008) recommend hearing aids as the first-line treatment for hearing loss associated with glue ear in children with Down syndrome [16.]

4. Factors that should be considered when discussing the use of hearing aids versus grommets (ventilation tubes) for the management of glue ear include the severity of hearing loss, the presence of recurrent acute otitis media (where grommet insertion is beneficial), the age of the child, the practicality of grommet insertion

and the risks associated with grommets e.g. infection, perforation of tympanic membrane and the views of the family. [16, 20] .

5. Children with Down syndrome often have narrow ear canals and smaller grommets may need to be used, with the associated disadvantage of earlier extrusion.

6. Adenoidectomy should be considered in thesurgical management of recurrent OME, recurrent acute otitis media and also as part of the management of chronic nasal obstruction and obstructive sleep apnoea. The indications remain the same as those in a typically developing child.

7. Significant laryngopharyngeal reflux has also been shown to predispose to OME in children, which may be exaggerated in Down syndrome secondary to hypotonia [21].

8. At all ages, particular attention should be paid to the assessment and treatment of suppurative nasal and ear conditions, with the aim of reducing acute middle ear infections and exclusion of complications of chronic eustachian tube dysfunction e.g. cholesteatoma.

9. The BAHA Soft Band and bone-anchored hearing aids have an important role in the management of conductive hearing loss where grommets and conventional hearing aids cannot be tolerated or have failed [22].

10. All people with Down syndrome with sensorineural loss should be actively encouraged to use of hearing aids at all ages. Cochlear implantation has been shown to be successful in children with Down syndrome and referrals should be made based on the same audiological referral criteria as the typically developing child [17, 19, 20, 23].

11. People with Down syndrome should have life-long audiological follow-up with rapid access to services including speech and language therapy to enable them to meet their developmental potential and avoid social exclusion [5.]

12. Transition of care from paediatric audiological services to adult services should involve direct transfer of care to a named professional

Additional useful resources for parents is provided by the National Deaf Children's Society: http://downsyndromedevelopment.org.uk/wp-content/uploads/2013/03/Downs_Syndrome_And_Childhood_Deafness_2011.pdf

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