Appreciating the Spectrum of Audiological Issues in Children with Down Syndrome

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Introduction

Down’s syndrome (DS) affects 1 in every 732 live births and is the most common cause of intellectual impairment affecting children. This increases to 1 in every 84 births when maternal age exceeds 40. It is a genetic disorder, caused by the presence of trisomy of chromosome 21st. Children with Down’s syndrome are at risk of a multitude of health related problems such as visual difficulties, congenital heart disease, thyroid disease, obstructive sleep apnoea and immunodeficiency. Crainofacial features such as cleft or high-arched palate, hypoplastic maxilla and narrow external auditory canal are often found in Down’s syndrome, as well as hypodontia or anodontia.

The most common health problem experienced in Down’s syndrome is hearing loss. At least 75% of children are affected along a mild to profound spectrum of hearing loss. Although there is considerable variation most individuals with DS have speech and language deficits particularly in language production, syntax and speech intelligibility. Optimising hearing is essential to minimise speech and cognitive delay.

The main cause of conductive hearing loss is persistent otitis media with effusion (OME); Park et al reported the incidence of conductive hearing loss at 37.9% of DS patents. In the general population up to 80% of children are reported as having at least one episode of OME by age four. The most effective management of OME is still contentious. In non syndromic children 70-80% will resolve without intervention within 3-4 months. Down’s syndrome children with almost universally get OME, the etiology of which is secondary to both anatomical and physiological dysfunction. Anatomical factors including mid face hypoplasia, contracted nasopharynx, abnormal shape in the Eustachian tube and poor function of the tensor veli muscle responsible for opening and closing the Eustachian tube. Poor physiological function of the immune system also predisposes to recurrent OME. Otitis media with effusion occurs at a younger age and persists to an older age than in other children. Sensorineural hearing loss is also relatively high in DS with up to 5% being affected.

Other otological disorders occurring at higher frequency in DS include wax impaction, tympanic membrane retraction, auditory canal stenosis and poor mastoid pneumatization. Inner ear abnormalities are also common in Downs Syndrome. A study of fifty nine patients by Blaser et al found universally hypoplastic inner ear structures with vestibular malformations particularly common. Downs syndrome children require special consideration and early intervention to avert the secondary handicap caused by hearing loss.
Impact

Hearing loss in any child can have a detrimental effect on their emotional, educational, linguistic and social development.\textsuperscript{16,17} Identifying a hearing loss is the fundamental tenet towards helping these children. International best practice now aims for early identification of permanent hearing loss through newborn hearing screening programs. It has been shown that children with hearing deficits who are identified and managed early have accelerated speech development.\textsuperscript{18,19} Hearing difficulties in DS compound existing auditory-verbal processing difficulties. A failure to address their hearing difficulties results in DS patients requiring more time to identify spoken words.\textsuperscript{20} Continued vigilance and treatment remains important as the prevalence of permanent hearing loss in children doubles by the age of nine.\textsuperscript{21,10} Up to 25\% of children with Downs syndrome will enter adulthood with a significant hearing loss \textsuperscript{22}. Jensen recently reported significant failures in primary care screening for adults with DS, including a failure to screen for hearing loss.\textsuperscript{23} Adults with Down’s syndrome may have an increased risk for development of depression and there is evidence of under treatment of depression in DS.\textsuperscript{24} Hearing assessment is essential in the differential diagnosis of depression and dementia.\textsuperscript{25}

Assessment

Children with Down’s syndrome are at a greater risk of developing a hearing impairment. Raut \textit{et al} reported the incidence of hearing loss in the first year of life at 34.1\%. Frequent hearing assessments should be undertaken to identify and treat this often reversible hearing disability.\textsuperscript{26} International guidelines recommend testing with oto-acoustic emissions (OAE), followed by automated auditory brainstem response (AABR) within six weeks.\textsuperscript{27} Testing should be carried out by suitably trained audiologists using modern calibrated technology.\textsuperscript{28} Most young children less than 3.5 years with DS are unable to cooperate with conventional behavioural audiological testing.\textsuperscript{29} Assessment with brain auditory evoked potentials (BAEP) is recommended in DS children.\textsuperscript{29,30} It should be noted that the average latency values of BAEP established for healthy children should not be used as a reference for children with DS.\textsuperscript{30} DS children have shortened latency values in BAEP which may be secondary to accelerated maturation of the nervous system or anatomical/functional disturbances of the central nervous system.\textsuperscript{30} The presence of narrow canals also predisposes to wax accumulation which may affect hearing and impedance testing.\textsuperscript{25} Recommended monitoring protocol includes screening at birth followed by full assessment by 10 months of age to include auditory thresholds, impedance testing and otoscopy. At 18 months of age a further review is carried out with testing annually thereafter until 5 years of age and then 2 yearly for life.\textsuperscript{31} More frequent assessments and intervention may be necessary should a deficit be found.\textsuperscript{31}

Management options

Virtually all children with Down’s syndrome develop OME and with it a reversible conductive hearing loss \textsuperscript{12}. Shott \textit{et al} found that treatment of OME either medically or surgically resulted in 98\% of children regaining normal hearing levels.\textsuperscript{32} DS individuals present with particular problems of assessment
and management. This difficulty is multifactorial including: earlier age of onset, prolonged course, greater risk of complications and potential diagnostic difficulties. Where a child with Down’s syndrome has OME for 3 or more months, surgical management such as insertion of pressure equalization tubes is often considered. This is to alleviate otitis media with effusion and prevents recurrence, however the benefits may be short lived compared to other children. Insertion of ventilation tubes is made more difficult or occasionally impossible by the narrow external auditory canals. Irrespective of these challenges, aggressive medical and surgical treatment of chronic ear disease is indicated as it provides significantly improved hearing levels. 32 It also lowers the incidence of tympanic membrane perforations and cholesteatoma.32, 33 Chronic otorrhoea can occur and may hamper the wearing of hearing aids.12

The use of hearing aids is indicated if there is a patient/parent preference, contraindication to anaesthetic, previous multiple pressure equalization tube insertion, chronic otorrhoea or canal stenosis. In these circumstances we should consider early provision of hearing aids.12 Small hearing aids that fit neatly behind the ear are widely available and when fitted appropriately can help mitigate the handicap caused by the hearing loss. Disadvantages to hearing aids include acceptance by the child and if there is a change in hearing levels, the hearing aids may be rejected as a result of being too loud or being too quiet. However with gradual introduction and persistence most children with accept the hearing aids.

Another tool in our armoury is the use of bone conduction amplification. This is where sound reaches the cochlea through bone vibration on the skull. These hearing aids when appropriate are highly successful. Options include aids that attach onto the leg of spectacles, aids that attach onto a head band or cap and aids that are attached to surgically implanted devices known as bone anchored/implanted hearing aids (BAHA). BAHA should be considered after conventional hearing aids and/or ventilation tubes have already failed.34 High levels of patient and carer satisfaction with the use of BAHA have been found.35 One of the disadvantages include the cost of the devices which are markedly more expensive than standard hearing aids, however a value cannot be placed on the sense of hearing.

Conclusions

It is important to be cognisant of hearing loss in any child and more so in those with Downs syndrome. Optimising hearing is essential to achieve best possible speech and language development. Down syndrome patients require specialist multidisciplinary assessment. An appreciation of the importance of life long surveillance for DS patients, with a need to coordinate assessment between tertiary centres and community audiological services is vital. Parents should be clearly informed of the increased risk of hearing loss as well as being prepared for ongoing assessment/treatment. In order to achieve best care for Down’s syndrome patients the medical profession must firstly ensure adequate knowledge of the specific patient needs as outlined above. Secondly it is implicit that adequate infrastructure and personnel are available to implement the recommended frequency of audiological assessment.
References