Original Article

Prevalence and Parental Awareness of Hearing Loss in Children with Down Syndrome

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Abstract

Background: To establish the prevalence of hearing deficit in children with Down syndrome (DS) in Hong Kong as measured by brainstem auditory evoked potentials (BAEP). The secondary objective is to examine the agreement between BAEP and clinical questioning in detecting hearing deficit in DS.

Methods: Consecutive DS patients attending the Down's Clinic in a regional pediatric referral center were recruited into this cross-sectional study. BAEP data performed within 12 months were retrieved. The care-taker was interviewed with a structured questionnaire to detect any symptom of hearing impairment. BAEP findings and clinical questionings were compared in an agreement analysis using quadratic weighted kappa statistics.

Results: Fifty DS patients (35 male, 15 female, mean age 11.70 years ± 5.74 standard deviation) were recruited. Eighteen patients (36.0%) were identified having hearing deficit by BAEP. Among patients with hearing impairment, 13 patients (72.2%) had a conductive deficit, and most have mild to moderate hearing loss. Five patients (27.8%) had sensorineural deficit and most have moderate to severe degree. Eight (44.4%) had bilateral hearing deficit. Care-takers of 13 patients (26.0%) reported symptoms of hearing impairment, with 9 (69.2%) having mild symptoms, 3 (23.1%) had moderate symptoms and 1 (7.7%) had severe symptoms. The weighted kappa was 0.045 (95.0% confidence interval − 0.138–0.229), indicating very poor strength of agreement between BAEP and clinical questioning. For patients with conductive hearing impairment, only 1 patients (7.7%) recalled history of otitis media.

Conclusions: The estimated point prevalence of hearing impairment in Chinese DS children in Hong Kong is 36%. Our finding of poor strength of agreement between objective testing and symptom questioning reflects significant underestimation of hearing impairment by history taking alone. In view of the high prevalence and low parental awareness, continuous surveillance of hearing is mandatory for DS patients throughout childhood and adolescence.

Key words: Brainstem Auditory Evoked Potential; Down Syndrome; Hearing; Prevalence

Introduction

Down syndrome (DS, trisomy 21) affects 1 in 700 live births in Hong Kong. It is one of the most frequent genetic causes of mild to moderate intellectual disability. DS is associated with a number of congenital disorders such as cardiac defects, gastrointestinal abnormalities, eye problems, hearing loss and thyroid diseases.

Hearing loss is found in 38–78% of patients with DS.[1] Conductive hearing loss occurs in most cases because of higher incidence of middle ear effusion, anatomical anomalies of Eustachian tube and stenosis of ear canals.[2] Prevalence of sensorineural hearing loss is also higher than the general population, and the incidence tends to increase with age. A population-based cross-sectional study in Norway revealed a prevalence of 35% of hearing loss in children with DS at age 8.[3] Another study from Poland involved 70 DS children from 2 months to 17 years (3.42 years on average) undergoing brainstem auditory evoked potentials (BAEP) study. Among 140 ears examined, only 43 (31%) were found to have hearing threshold below 30 dB.[4] However, it was unclear of the degree of parental awareness in those with abnormal BAEP results.

Data on hearing impairment of DS in Chinese population are scarce. Kwong and Wong[1] reviewed the retrospective data on 109 DS children with a mean age of 32.7 months attending a single child assessment center from 1985 to 1993. Among the 72 patients who failed in free field distraction test, 49 were identified to have hearing impairment on subsequent audiological assessment, yielding a period prevalence of 45% in the preschool population. As over 90% of children were below 5 years of age, the degree of hearing deficit in relation to age could not be demonstrated. The point

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prevalence of school age children and the degree of parental awareness were unknown.

McPherson *et al*.[2] performed otoacoustic emission (OAE) in 78 Chinese school DS children with a mean age of 12.1 years, and 90% failed the screening test. Only 11 out of 86 referred children attended diagnostic pure tone audiometry (PTA). Eight were subsequently confirmed to have hearing deficits. Only 15.2% of parents in this cohort reported a positive history of hearing loss. As only a small proportion of screened subjects underwent the diagnostic test, the point prevalence of 72.7% (8/11) was probably an overestimation. This study was also inconclusive in differentiating different types of hearing loss in DS.

The primary objective of this study was to identify the point prevalence and types of hearing loss in a cohort of ethnic Chinese DS children actively followed up in a specialized clinic for DS patients at a regional pediatric center. The secondary objective was to ascertain the parental awareness of their children's hearing deficits. The severity of the deficit on BAEP was correlated to standardized symptom-reporting from parents/care-takers to determine the degree of agreement between BAEP findings and parental awareness of hearing loss.

Methods

This was a cross-sectional study to determine the hearing status of children with DS. Consecutive DS children who were followed up in the DS Clinic at Caritas Medical Centre were included in this study. The hospital is a regional referral center in Hong Kong, China, and the DS Clinic receives referrals from a pediatric population of 149,000, equivalent to 17% of the Hong Kong child population. The annual delivery rate in the catchment was 5500 in 2012. The clinic provides comprehensive medical care and health surveillance for DS children from 0 to 18 years of age. The diagnosis of DS had been confirmed by karyotype studies. At the time of this study, around 100 patients were actively followed up in the clinic.

Hearing screening has been part of our health supervision protocol. BAEP is utilized to determine the hearing threshold and types of hearing loss. As compared to OAE and PTA, BAEP has the advantage of high sensitivity and specificity (>90%) and is suitable for younger (<4 years) or noncooperative children.[5] Patients with acute otitis media or externa were given treatment till recovery before proceeding to BAEP study.

Brainstem auditory evoked potentials studies were performed in the electrodiagnostic unit under a standard protocol. BAEP was obtained with the Viking Select machine manufactured by VIASYS Healthcare. The lower filter was 150 Hz and upper one 3 KHz. BAEP was recorded after administration of a transient stimulus of click-tip sound (75 dB at a rate of 11.4 Hz) to each ear at first 10 ms. When the BAEP was obtained, the threshold intensity was established using descending methods (every 10 dB). Air conduction studies were performed in all subjects. In patients with raised hearing thresholds, bone conduction was performed, and the air-bone gap was estimated. Hearing threshold of 20 dBnHL was regarded as normal. Hearing loss was defined as mild (20–40 dBnHL), moderate (40–60 dBnHL), and severe to profound (more than 60 dBnHL). The types and severity of hearing loss were determined. Conductive hearing deficit was defined as absent of wave I and normal wave III–V interpeak latency. An air-bone gap of at least 20 dBnHL in hearing threshold should also be present for defining the conductive deficit. Sensorineural deficit was defined as prolonged wave III–V interpeak latency with absence of air-bone gap in hearing threshold.

A structured questionnaire was used to obtain participant's case history. Consent was obtained with the care-taker prior to the phone interview. Concomitant BAEP study performed within 12 months was retrieved for analysis. BAEP would be arranged if the participant did not have one done within 12 months. The questionnaire was conducted by one of the investigators (WLL) who was blinded to the BAEP findings at the time of interview. Demographic data (including age, gender, degree of intellectual disability), history of otitis media, use of hearing aids and previous ear, nose and throat (ENT) surgery including cochlear implantation were documented. Symptoms of hearing impairment were also obtained. Symptoms of hearing impairment were classified into four groups (normal, mild, moderate and severe). Mild symptom was defined as the inability to identify soft sound such as whispering; moderate as failure to hear daily conversation or telephone ring. Severe symptom was defined as only able to hear very loud sound such as shouting or vacuum cleaner noise or difficultly to perceive any sound.[6] Data obtained from questionnaires were counter checked with computerized data from Clinical Management System of the Hong Kong Hospital Authority, and any missing information was also retrieved.

Data analysis was carried out using SPSS version 12.0 (IBM Corporation, USA). MedCalc Statistical Software version 12.7.2 (MedCalc Software, Acacialaan 22, B-8400 Ostend, Belgium) was used for calculating weighted kappa, which was a measure of agreement between BAEP and clinical questionings in identifying different levels of hearing loss in each individual. Quadratic weights were used instead of linear weights because we believed a difference between mild to moderate degree of hearing impairment was less important than a difference between moderate and severe impairment. Kappa value (strength of agreement) was interpreted as follows: <0.20 (poor), 0.20–0.40 (fair), 0.41–0.60 (moderate), 0.61–0.80 (good), 0.81–1.00 (excellent).[7]

This study was approved by the Hong Kong Hospital Authority Kowloon West Cluster Ethical Committee on November 1, 2012.

Results

Participants

A total of 50 subjects were recruited in our study, and their characteristics are shown in Table 1. There were total

102 patients following up in our DS Clinic. Twenty-two patients aged more than 18 years old were excluded from the current study. Thirty patients were further excluded because of failure to obtain consent for the phone interview or BAEP study.

There were 35 male and 15 female patients with mean age 11.70 ± 5.74 years. Twenty-three patients (46%) had mild grade intellectual disability, 22 patients (44%) had moderate grade intellectual disability and the remaining 5 patients were too young for classification of intellectual disability. Only 6 patients (12%) could recall history of otitis media. Thirteen cases (26%) had been actively followed up by ENT specialists for various reasons, including history of otitis media, cerumen impaction and obstructive sleep apnea.

Estimated prevalence of hearing loss

As shown in Table 2, hearing threshold in BAEP was normal bilaterally in 32 cases (64%). Eighteen patients (36%) were identified having hearing deficit by BAEP. Hearing threshold ranged from 30 to 70 dBnHL. Eight cases (44.4%) had bilateral hearing deficit, right ear hearing loss was found in 2 cases (11.2%) and 8 cases (44.4%) over the left side. Among patients with hearing impairment, 13 patients (72.2%) were found to have conductive hearing deficit, and 5 patients (27.8%) had sensorineural hearing deficit. For cases with conductive hearing problem, 6 patients (46.1%) had mild hearing loss, 2 patients (15.4%) had moderate loss and 5 patients (38.5%) had severe loss. For patients with conductive hearing impairment, only 1 patients (7.7%) recalled history of otitis media. For those who had sensorineural hearing deficit, 40% were identified as moderate hearing loss and 40% as severe loss.

Correlation between clinical symptoms and brainstem auditory evoked response results

Among all subjects, care-takers of 13 patients (26.0%) perceived their children had symptoms of hearing impairment, with nine (69.2%) having mild symptoms, three (23.1%) had moderate symptoms and one (7.7%) had severe symptoms.

Table 3 showed the inter-rater agreement between BAEP findings and symptoms of hearing impairment obtained by questionnaire. The quadratic weighted kappa was 0.045 (95% confidence interval [*CI*] −0.138–0.229), indicating very poor strength of agreement between BAEP and clinical questioning in identifying the degree of hearing loss. When unilateral hearing loss on BAEP was reclassified as normal hearing (according to WHO criteria in defining hearing threshold as the deficit in the best hearing ear),[3] quadratic weighted kappa was increased to 0.097 (95% *CI* −0.109–0.303), but the adjusted strength of agreement remained poor [Table 4].

Confirmation of brainstem auditory evoked response results

Subjects with abnormal brainstem auditory evoked response (BAER) results were referred to ENT specialists for assessment. By the end of the study, eight subjects had undergone ENT assessment. Three subjects had defaulted referral appointments, one subject had refused further

Table 1: Characteristics of 50 subjects in this study

Gender, *n* (%)	
Male	35 (70)
Female	15 (30)
Age (years)	
Mean	11.7 (SD, ± 5.7)
Median	13.9
Intellectual disability, *n* (%)	
Mild	23 (46)
Moderate	22 (44)
Unavailable	5 (10)
History of otitis media, *n* (%)	
Yes	6 (12)
No	44 (88)
Use of hearing aids, *n* (%)	
Yes	1 (2)
No	49 (98)
Active follow-up in ENT, *n* (%)	
Yes	13 (26)
No	37 (74)

SD: Standard deviation; ENT: Ear, nose and throat.

Table 2: BAEP results in this study

BAER results	Number of subjects, *n* (%)
Normal	32 (64.0)
Conductive deficit	13 (26.0)
Mild (20–40 dBnHL)	6 (46.1)
Moderate (>40–60 dBnHL)	2 (15.4)
Severe (>60 dBnHL)	5 (38.5)
Sensorineural deficit	5 (10.0)
Mild (20–40 dBnHL)	1 (20.0)
Moderate (>40–60 dBnHL)	2 (40.0)
Severe (>60 dBnHL)	2 (40.0)
Total subjects with hearing deficit	18 (36.0)
Bilateral	8 (44.4)
Unilateral	
Left	8 (44.4)
Right	2 (11.2)

BAEP: Brainstem auditory evoked potentials; BAER: Brainstem auditory evoked response.

Table 3: Correlation between BAEP results and clinical symptoms, *n*

Degree of hearing problem by questionnaire	Severity of hearing deficit estimated by BAEP				Total
	Normal	Mild	Moderate	Severe	
Normal	25	5	3	4	37
Mild	4	1	1	3	9
Moderate	3	0	0	0	3
Severe	0	1	0	0	1
Total	32	7	4	7	50

Quadratic weighted κ = 0.045 (95% *CI*: 0.138–0.229). *CI*: Confidence interval; BAEP: Brainstem auditory evoked potentials.

referral, and six were still awaiting for ENT assessments. Among the 8 subjects who were assessed by ENT specialists, all were found to have hearing problems that correlated with BEAR results, either with the use of pure tone audiogram or clinical assessments.

Discussion

This is the first study on the hearing status of Chinese DS children across the pediatric age range of 0–18 years. The point prevalence of hearing loss is estimated to be 36%, which is similar to a recent population-based study in Norway (35%).[3] Our current study was not population-based; the subjects consisted of a convenient sample attending a regional specialty clinic. The number of subjects in each age group was also small. Nonetheless, our sample had a heterogeneous age distribution, and the proportion of subjects in each age group was similar to that of a concomitant local population survey [Table 5].[8] All the subjects underwent the diagnostic BAEP study without prior screening by the symptom enquiry, distraction test or OAE, thereby reflecting a relatively unbiased snapshot of the hearing status in this population. Most patients had conductive hearing impairment, which concurs with findings in previous studies.[1-3]

Patients with DS have various structural anomalies that predispose to conductive hearing impairment. These include midface hypoplasia, small size of the pinna, stenotic external auditory canal predisposing to cerumen impaction (40–50%) and small eustachian tubes openings.[9,10] Generalized hypotonia may lead to dysfunction of tensor veli palatini muscle of palate, increasing the risk of acute otitis media and chronic effusion from Eustachian tube collapse.[10] The presence of residual mesenchymal tissue in the middle ear (75%), malformed ossicular chain (25%), recurrent upper respiratory tract infections due to impaired immune function also contribute to conductive hearing impairment.[9,10]

There is also higher prevalence of sensorineural hearing deficit among children with DS. This may result from anomalies such as shortened organ of Corti, reduced spiral ganglion cells in temporal bones and inner ear abnormalities such as mondini deplasia.[2,9] Progressive compression of the auditory nerve in internal auditory meatus may lead to nerve degeneration.[9] In our study, all patients with sensorineural hearing impairment were older than 10 years of age [Table 5]. Sensorineural deficit was observed to occur later in life in DS but much earlier compared with normal population, suggestive of early presbycusis.[2,9] Although there was a high prevalence of hearing impairment among our subjects, only a small percentage of parents reported a positive history of hearing deficit and associated risk factors in their children. In this study, the agreement between BAEP findings and parental reporting of hearing problems was poor and remained weak even after adjustment for the best hearing ears. Our finding provides strong evidence to reflect poor parental awareness of hearing problems among DS patients. Notably, seven children with severe deficits on BAEP were reported to have normal hearing or only mild loss by their care-takers. On the other hand, three children with normal BAEP studies were mistaken to have moderate hearing loss. It is unreliable to ascertain hearing impairment by clinical questionings alone; the deficit is often masked by coexisting intellectual disability and behavioral problems. Moreover, our study also showed a high correlation between BEAR results and ENT findings, reflecting that BAER can be a reliable screening tool in this setting.

Hearing evaluation is particularly difficult in this population. DS children may have inconsistent voluntary response and poor attention during behavioral testing. The latter cannot differentiate the type of hearing impairment and side of diseased ears. BAEP, which can evaluate auditory performance regardless of patient's age and mental status, is particularly useful in this setting.

Early detection of hearing problem is crucial to language development. Shott *et al*.[11] demonstrated that with aggressive medical and surgical interventions of chronic otitis media in young DS children, 98% attained normal hearing after treatment. According to *the American Academy of Pediatrics guideline*, in 2011, after universal newborn hearing screening at birth, further evaluation should be performed at 6 months of age for confirmation. Referral

Table 4: Correlation between BAEP results and clinical symptoms when unilateral hearing deficit was reclassified as normal hearing, *n*

Degree of hearing problem by questionnaire	Severity of hearing deficit estimated by BAEP				Total
	Normal	Mild	Moderate	Severe	
Normal	31	0	1	3	35
Mild	8	0	0	3	11
Moderate	3	0	0	0	3
Severe	0	1	0	0	1
Total	42	1	1	6	50

Quadratic weighted κ = 0.097 (95% *CI*: 0.109–0.303). *CI*: Confidence interval; BAEP: Brainstem auditory evoked potentials.

Table 5: Distribution of types of hearing loss across age group

Age group (years)	Conductive deficit, *n*	Sensorineural deficit, *n*	Normal, *n*	Total, *n*	Mid-2013 Hong Kong population (percentage of total population)[8]
0–4	5	0	4	9	257,500 (3.6)
5–9	3	0	8	11	245,700 (3.4)
10–14	0	3	9	12	287,400 (4.0)
15–19	5	2	11	18	398,100 (5.5)

to an otolaryngologist is necessary if the screening test is failed. Diagnostic BAEP should also be performed to establish hearing status.[12] By 1-year of age, behavioral audiogram can be performed but additional screening by BAEP is needed if the child is unable to complete the test.[13] Thereafter, behavioral audiogram should be carried out every 6 months up to 4 years of age, and then annually.[12-14] Additional BAEP study should be performed if normal hearing is not established by behavioral testing. Referral to otolaryngologists should be arranged in any stage in case of suspected hearing impairment.[12]

In conclusion, the estimated point prevalence of hearing impairment in Chinese DS children in Hong Kong is 36%. Our finding of poor strength of agreement between objective testing and symptom questioning reflects significant underestimation of hearing impairment by history taking alone. In view of the high prevalence and low parental awareness, continuous surveillance of hearing is mandatory for DS patients throughout childhood and adolescence. BAEP is a useful objective test, particularly in young and uncooperative children who are not suitable for behavioral audiogram or PTA.

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