Distortion products otoacoustic emissions in diagnosis of hearing loss in Down syndrome

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Abstract

This study was carried out to investigate the features of hearing impairment in subjects with Down syndrome. Forty-seven children and 14 adults with Down syndrome were included in the study. In all cases, a complete otorhinolaryngological examination was performed, followed by audiological assessment. Depending on age, intellectual level and middle ear status the following examinations were performed: pure-tone ‘play audiometry’, tympanometry, acoustic reflex, auditory brain response (ABR) and distortion products otoacoustic emissions (DPOAE). The results were compared with age matched control groups. Among the group of children with Down syndrome, we have frequently found impairment of the conductive function of the middle ear which was expressed by pathological tympanometry. Tympanometry of B and C type was detected in 56% of ears. The amplitude of DPOAE was lower in children with Down syndrome than in the control group. This difference was more expressed in adults with Down syndrome. DPOAE examination results in subjects with Down syndrome without conductive hearing loss indicate early age related inner ear impairment. © 1998 Elsevier Science Ireland Ltd. All rights reserved.

Keywords: Distortion products otoacoustic emissions; Down syndrome; Hearing loss

1. Introduction

Speech and language development in normal children can be affected by hearing loss, but in children with other intellectual disabilities such as Down syndrome, the development of speech is severely aggravated. Communication and perception skills are much more affected than other areas of the child’s development. This, in part, is due to hearing loss, as hearing is indispensable for normal speech development [1–3]. In Down syndrome, middle ear dysfunction is more frequently observed than in the healthy population [4,5,2,6,7]. This is the effect of anatomical malformations, such as Eustachian tube anomalies, persistent mesenchymal tissue in the tympanic cavity;
external auditory meatus stenosis and mastoid bone hypoplasia [4,8,9]. The Eustachian tube function is additionally compromised due to an anatomically constricted nasopharynx together with adenoid hypertrophy [10,11]. Conductive hearing loss due to the above causes decreases with age in a great number of cases. On the other hand, the number of cases with sensorineural hearing loss increases with age [12–15].

The diagnostic difficulties that always accompany a hearing examination in small children, especially those with Down syndrome, only add to the problem [6,16,17]. In many cases a hearing examination requires a lot of patience and experience as well as the use of different examination techniques.

Thus, in this study we have tried to assess the usefulness of distortion products otoacoustic emissions (DPOAE) techniques combined with tympanometry in auditory disorders diagnostics in subjects with Down syndrome. The pure-tone audiometry examination can be distorted by low intellectual abilities and thus does not allow the detection of small alterations. The auditory brain response (ABR) examination, which supplies reliable information, is troublesome because of the necessity of being carried out during pharmacological sleep. However, both the DPOAE and tympanometry do not require the cooperation of the patient and they seem to be adequate methods of examining patients with Down syndrome. Tympanometry provides data on the middle ear and the DPOAE examination is a very sensitive indicator of inner ear function. The efficient system conducting sound is an indispensable condition for DPOAE detection, which limits its use in middle ear disorders. Because of the different types of hearing loss in different age groups, the examination was carried out in children and adults with Down syndrome and compared to corresponding control groups.

2. Patients and methods

Forty-seven children and 14 adults with Down syndrome were studied. All examined subjects lived at home and all had cytogenetic confirmation of the diagnosis. Parental consent was obtained for all patients. In all cases, a complete otorhinolaryngological examination with otopscopy was performed, followed by audiological assessment. Audiological tests were carried out after removal of impacted cerumen. The children’s age varied from 5 months to 16 years with an average age of 5.8 years. The results of DPOAE were compared with the results in the group of otologically healthy children (20 ears) aged 3.5–10 years, mean 6.4. The adult patients age varied from 17 to 37 years, mean 25.3 years. The results of DPOAE were compared with those of the otologically healthy adults (20 ears) aged 16–37 years, average 25.8. Depending on age, intellectual level and middle ear status the following examinations were performed: pure tone ‘play audiometry’, tympanometry, acoustic reflex, ABR responses to click and DPOAE.

All tests were conducted in a sound-proof room. The tests performed in the groups of children and adults are presented in Tables 1 and 2, respectively.

Pure-tone air and bone conduction thresholds were obtained for each ear using play audiometric procedures (Vera 103 + Midimate 622 Madsen Electronics). The pure-tone average air conduction threshold level in dB HL for 500, 1000, 2000, 3000 Hz was used to represent hearing sensitivity. As suggested by Northern and Downs [18], this

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Audiological tests performed in children with Down syndrome</th>
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<tbody>
<tr>
<td></td>
<td>Number of patients</td>
</tr>
<tr>
<td>Pure-tone play audiometry</td>
<td>10</td>
</tr>
<tr>
<td>Impedance audiometry</td>
<td></td>
</tr>
<tr>
<td>Tympanometry</td>
<td>47</td>
</tr>
<tr>
<td>Stapedial reflex</td>
<td>23</td>
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<tr>
<td>ABR</td>
<td></td>
</tr>
<tr>
<td>Physiological sleep</td>
<td>4</td>
</tr>
<tr>
<td>Pharmacological sleep</td>
<td>3</td>
</tr>
<tr>
<td>DPOAE</td>
<td>44</td>
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</tbody>
</table>
study defined hearing loss as pure-tone average (PTA) level $>15\,\text{dB HL}$. A hearing loss by air conduction with air-bone gap $>10\,\text{dB}$ was classified as conductive hearing loss.

Impedance tests were carried out by means of the Zodiac 901 impedance meter (Madsen Electronics). The tympanograms were classified according to Jerger. Acoustic stapedial reflexes were also elicited by pure tones at 500, 1000, 2000 and 4000 Hz. ABR were recorded by means of EP-Test (Medical Electronics Workshop). The alternating click produced by rectangular wave pulses was used as auditory stimulus. Threshold of hearing was defined as the lowest intensity stimulus yielding a measurable response—wave V. DPOAE were recorded by means of Celesta 503 cochlear emissions analyzer (Madsen Electronics). Two simultaneous primary pure tone signals were presented at a level of 70 dB SPL, the ratio $f_1/f_2 = 1.22$. DP response amplitude above noise floor (NF) across a range of frequencies 500, 750, 1500, 2000, 3000, 4000, 6000 and 8000 Hz, were recorded. The responses above 1 S.D. of NF level were considered as valid. Results were analysed using nonparametric statistics (Mann–Whitney U-test).

### Table 3
Tympanometry results in children with Down syndrome

<table>
<thead>
<tr>
<th>Tympanometry</th>
<th>Number of ears</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>A type</td>
<td>10</td>
<td>10.7</td>
</tr>
<tr>
<td>A$_1$ type</td>
<td>31</td>
<td>33.3</td>
</tr>
<tr>
<td>B type</td>
<td>32</td>
<td>34.5</td>
</tr>
<tr>
<td>C type</td>
<td>20</td>
<td>21.5</td>
</tr>
<tr>
<td>Total</td>
<td>93</td>
<td>100</td>
</tr>
</tbody>
</table>

3. Results

3.1. Results in the group of children

The typical aural characteristics of Down syndrome were observed in 46 children. There was external auditory meatus stenosis of different degrees with epithelial ceruminous plug in all children. Normal tympanic membrane during otoscopy was noted only in eight out of 46 patients after cerumen was removed. In 38 patients in this group, the tympanic membranes were thickened and retracted. They were in more oblique position in many cases. Unilateral attic cholesteatoma was found in one case. Chronic rhinitis was not present in eight patients. Surgical indications for adenoid removal were reported in three patients. Play audiometry was performed in ten older children only, due to lack of cooperation. Hearing loss of the conductive type was found during this examination. The air-bone gap in audiograms was between 25 and 45 dB HL. The pure-tone average hearing loss (PTA) did not exceed 15 dB HL in two ears only. The mean hearing impairment (PTA) for the whole group was 27.4 dB HL.

Tympanometric examination was well tolerated and carried out in all the patients. The results are presented in Table 3. The stapedius reflex was detected in 23 children (46 ears) with tympanograms of A and C type, respectively.

ABR examination was performed in seven children using click as a stimulus. Because of the necessity of performing the ABR procedure while under anesthesia, it was carried out in suspected hearing disorders only. Sensorineural hearing loss was reported in one child, on the grounds of the ABR examination (auditory threshold 60 and 50 dB with Type A tympanograms). The child had...
Fig. 1. Pedigree of family with der (14,21) and sensorineural hearing loss.

Legend:
- ○ - sensorineural hearing loss
- ☐ - der(14;21) carrier
- ♦ - stillbirth
- ● - Down syndrome
- ⊙ - normal karyotype
- ◊ - death

Down syndrome genetically determined by trisomy 21 as the result of Robertson’s chromosomal translocation in its mother (14;21). An auditory examination carried out in this family revealed sensorineural hearing loss in the child’s father (III, 8), brother (IV, 5), sister (IV, 6) (Fig. 1). As shown in the genealogy, the inheritance of hearing impairment was independent of the chromosomal translocation appearance (14;21) and Down syndrome. It may be concluded that sensorineural hearing loss in the above mentioned child was an accidental combination of two genetically determined disorders—Down syndrome and sensorineural hearing impairment, which is inherent in this family.

DPOAE examination was performed in 44 children (80 ears). In 36 of all ears the tympanometry was of A and A₅ type. In those cases, the amplitude of DPOAE was within or above normal range. The mean values of the amplitude for a particular frequency were within the normal range and the DP-gram curve was similar to the one obtained in the control group. However, amplitudes in the group with Down syndrome were lower in comparison with those in the control group. The difference was statistically significant for frequencies of 1500, 2000, 3000 and 6000 Hz (Fig. 2).
3.2. Results in the adult group

As was presented in the previous group, the external auditory meatus was constricted in 12 out of 14 subjects and filled with cerumen in nine. Tympanic membranes were thickened in the majority of cases (8/14). A retraction pocket in the posterior–superior quadrant was found in one ear. Impaired nasal patency was noted in three out of 14 patients.

Pure-tone audiometry was carried out in all patients except one. The pure-tone average hearing loss was 32.3 dB HL in this group. Two patients had normal hearing, at PTA 15 dB HL, considered as within the normal range and four had normal hearing at PTA 25 dB HL, also considered as within the normal range. Severe hearing loss was reported in one case—PTA 56 and 82 dB HL.

The results of tympanometry are presented in Table 4. The stapedius reflex was detected in all but three ears with tympanometry of A type but was not detected in all ears with tympanometry of A, B and C type, respectively.

DPOAE procedure was performed in all adult patients with Down syndrome. In 21 of all ears there was tympanometry of A and A type. The amplitudes in the group with tympanometry of A and A type were significantly lower at all frequencies except at 500 Hz and 8 kHz in comparison with those in the control group (Fig. 3). They were also significantly lower at all frequencies than in the children with Down syndrome, despite the fact that a similar difference was not reported in both control groups (Fig. 4).

4. Discussion

Among the group of children with Down syndrome, we frequently found impairment of the conductive function of the middle ear which was expressed by pathological tympanometry. In our study, the tympanometry of B and C type was detected in 56% of ears. The proportion of abnormal tympanometry in children with Down syndrome as reported by other authors was 60–70% [4,12,19]. The proportion of tympanometry of As type in our study was 33.3%. Tympanometry of As type has also been reported by Keiser et al. [14] and Dahle et al. [18]. Schwartz et al. [20] found tympanometry of As type in 9% of patients. They suggest that this may be due to ossicular chain stiffness. Tympanometry of As, B and C type were present less often in the adult group. It seems that these changes are of a temporary character.

<table>
<thead>
<tr>
<th>Tympanometry results in adults with Down syndrome</th>
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<tbody>
<tr>
<td>Number of Down syndrome</td>
</tr>
<tr>
<td>-------------------------</td>
</tr>
<tr>
<td>A type tympanometry</td>
</tr>
<tr>
<td>A, type tympanometry</td>
</tr>
<tr>
<td>B type tympanometry</td>
</tr>
<tr>
<td>C type tympanometry</td>
</tr>
<tr>
<td>Total</td>
</tr>
</tbody>
</table>
ever Keiser et al. [14] reported abnormal tympanometry of B and C type in 39% of adult subjects with Down syndrome.

Taking into consideration the fact that 50% of children with otitis media with effusion have hearing better than 15 dB HL PTA, a hearing impairment \( > 15 \text{ dB HL PTA} \) in \( \approx 30–35\% \) of children with Down syndrome should be expected. In our study this proportion should be 28%. But in children with reliable results of pure-tone audiometry only in two ears out of 20 was the pure tone average hearing level \( > 15 \text{ dB HL} \). In all other ears, hearing loss was \( \approx 30 \text{ dB HL PTA} \). The same phenomenon has been reported in other papers [4,12,19]. It may be due to difficulties in obtaining reliable audiometric results in subjects with Down syndrome. It may also indicate an additional cause of hearing impairment. Dahle et al. [19] reported that children with Down syndrome, classified as having normal hearing, had a mean hearing level lower than children in the control group. This could explain the frequent lack of hearing improvement after ventilation tube insertion in children with Down syndrome [21]. DPOAE carried out in children with Down syndrome and with tympanometry of A and A\( _2 \) type had significantly lower amplitudes at many frequencies. It may be the result of alterations within the Organ of Corti. ABR examination performed in infants with Down syndrome shows a steeper latency-intensity slope which may also be associated with altered cochlear function [22].

A great number of children in this group had Type A\( _2 \) tympanograms. DPOAE procedure requires transmission of stimulus and evoked emission through the middle ear and is very susceptible to changing conditions in the middle ear.

To overcome this problem, audiometric examinations were carried out in the group of adults with Down syndrome and with normal functioning of the middle ear. Significantly lower DPOAE amplitudes at all frequencies, except at 500 Hz and 8 kHz, were detected in this group in comparison to those obtained in the controls. This would indicate that the onset of sensorineural hearing loss increases with age. This phenomenon has been reported by many authors. The first to describe it was Brooks et al. [12]. They found a sensori-neural hearing loss component in 55% of patients over 20 years of age with Down syndrome. This was confirmed by Keiser et al. [14]. In 1970, Krmpotic-Nemanic [15] described a phenomenon of age related bone apposition in the fundus of the internal auditory meatus. Similar findings were described by the same author in children with Down syndrome. She claims that this can be the cause of sensori-neural hearing loss in this group of children. Buchanan [13] found hearing impairment at high frequencies (6–8 kHz) in Down syndrome individuals in the second decade of life, which had a tendency to increase with age and would suggest the early onset of presbycusis.
In our study, decreasing DPOAE amplitude was not only associated with high frequencies. Had it been compared to presbycusis it would be a senile type of deafness caused by stria vascularis lesion of the cochlear duct, according to Schuknecht [23]. Developmental changes in the inner ear such as shortening of the cochlea and Mondini or Scheibe dysplasia were found during histopathological examination in subjects with Down syndrome [24]. This may also be the cause of sensori-neural hearing loss. Cell population of the spiral ganglion is also decreased [25]. However, severe and moderate hearing impairment of the sensorineural type was found in only two cases. In one case the cause was not related to Down syndrome.

DPOAE examination results in subjects with Down syndrome without conductive hearing loss indicate early, age related inner ear impairment. An explanation of the mechanisms responsible for these alterations can be found by in depth analysis of genes located in the additional aneuploid chromosome 21.

These investigations could become a source for a new therapeutic approach [26].

References


