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RESEARCH ARTICLE

Middle Ear Pressure Changes Over Time in Children with Down Syndrome

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Abstract

Objective: There is debate regarding the management of otitis media with effusion (OME) in children with Down syndrome. Information about the progression of middle ear function in this group is limited.

Methods: A retrospective study reviewing case notes and audiometric data of children with Down syndrome, recorded changes in middle ear pressure over time using tympanograms as a surrogate marker.

Results: The first and last tympanograms of 24 children were compared along with clinical findings. 50% (12/24) with Type B tympanograms at their initial review persisted to have the same trace at follow up (42% having had at least one intervention). Overall, 25% (6/24) showed a drop in pressure group, with 4/6 having had interventions.

Conclusion: Middle ear pressures in children with Down syndrome is a complex issue. Most OME will persist, which contrasts to children without Down syndrome, where up to 80% improve over a period of 3 months.

Keywords

Otitis media, Otitis media with effusion, Tympanogram, Down syndrome, Middle ear, Otolaryngology

Introduction

Down syndrome is one of the most common genetic conditions affecting approximately 1 in every 1000 live births [1]. There are a multitude of potential clinical manifestations associated with this condition including dysmorphic features, organic disorders such as congenital cardiac defects, gastrointestinal defects, ocular abnormalities, celiac disease and endocrine disorders along with haemato-oncological, immunological and disorders affecting the ears, nose and throat [2].

Despite this, studies in developed countries have documented vast improvements in child survival, from 25 years in the early 1980s [2] to an estimated life expectancy of around 50-60 years in recent years [3]. In particular, this increased life expectancy has been thought largely due to advancements in the ability to repair congenital heart defects, along with a multidisciplinary approach to care.

A survey of parents attending a Down syndrome association conference showed that 50% of Down syndrome children saw an ENT surgeon regularly [4] and 55-75% of children suffered from conductive hearing loss. It should be noted that there is a high prevalence of otitis media with effusion (OME) in children with Down syndrome [5,6] although this is not the only cause of hearing difficulty. Other causes of a conductive hearing loss in patients with Down syndrome include structural abnormalities of the mastoid and ossicular chain themselves [7] as demonstrated in both neuroradiological and post mortem studies of the temporal bones [8]. The incidence of sensorineural and mixed hearing loss is higher in children with Down syndrome and is estimated to effect approximately 4-9% [5,9].

Currently, there is little evidence guiding the treatment of OME in children with Down syndrome [10]. This study aims to assess the changes in tympanometry over time in children with down syndrome as a marker of middle ear pressure change and to see if any intervention had an overall impact on the end tympanometry suggesting improvement.

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Methods

Following local institutional approval, a retrospective search was undertaken, identifying all patients with the diagnosis of Down syndrome that had attended for consultation in ENT outpatients over the previous 10 years.

Adults at the time of initial review were excluded from the study population along with individuals with no audiological data available.

Medical notes were reviewed and details of patient demographics, duration of follow up, ear nose and throat interventions performed along with first and last audiometric tests were collected and analysed using simple statistics.

Comparison between tympanometry, using a 226 Hz probe, of the first and last attendances occurred using Jergers classification (See Table 1) to estimate change [11].

During analysis, patient improvement was based on a positive change in one or both ears in the child i.e a change in tympanometry curve from type B trace to either type C1, C2 or Type A trace.

A change in tympanometry curve, in one or both ears, with an increase in negative pressure i.e from type A to type C or B trace was recorded as deterioration in middle ear pressure.

At both initial presentation and at follow up, clinical otoscopic examination findings were noted to assess if there were clinical findings of otitis media with effusion and to confirm the presence or absence of any ventilation tubes or evidence of any residual perforation or concurrent disease such as cholesteatoma.

Results

Overall 39 patients were identified having attended the department in the specified period of ten years. Of these, 9 were adults and as such excluded. Of the remaining 30 patients, 6 were excluded as no audiological testing results were available. The remaining 24 patients were included.

14 males and 10 females were present within our patients. Male to female ratio was 0.7:1. The average age of patients on their last audiological testing was 83.83 months (range 13-203, Standard deviation = 51.38 months). The length of follow up (time between tympanograms) was, on average, 46.5 months (Range 5-129 months).

Of the children identified, a quarter of patients (6/24) demonstrated a drop in tympanometry curve over the follow up period, with 4/6 having had at least one intervention. Half of all patients who had a Type B tympanogram at their first clinic appointment (12/24) continued to have the same curve at their final follow up. 14 children had undergone surgical intervention whereas 10 patients had not undergone any interventional procedures.

Table 1: Jergers 1970 classification of tympanometry [11].

Classification	Compliance (ml)	Pressure (dPa)
A	> 0.2	> -99
C1	> 0.2	-100 to -199
C2	> 0.2	-200 to -399
B	< 0.2	> -400

Table 2: Pressure changes over time in children who had no interventions.

No Intervention Group		
	Number (%)	Pressure Group Change
Better	1 (10%)	Type B to C1
Same	7 (70%)	All Maintained Type B
Worse	2 (20%)	Both Type A to C2
	10	

Table 3: Pressure changes in children who had interventions.

No Intervention Group		
	Number (%)	Pressure Group Change
Better	5 (35.7%)	2 × Type B to C2/Type B to C1/Type C2 to A/Type B to A
Same	5 (35.7%)	All Remained Type B
Worse	4 (28.6%)	Type C1 to C2/Type C1 to B/Type A to C1/Type A to C2
	14	

The change in tympanometry curve over time in those children who had no intervention can be seen in Table 2. Within this group it can be seen that 90% who had no operative intervention remained in the same pressure group or had a worsening in middle ear pressure defined as a drop in pressure group. No child had evidence of a perforation present on otoscopic examination or on measurement of canal volumes to suggest this.

Of the remaining 14 children who had operative intervention, the changes in pressure group can be seen in Table 3. No child at follow up had remaining ventilation tubes present on otoscopic examination and no evidence of perforation.

It can be seen that within this group, despite intervention, most children (64.3%) remained within the same pressure group or dropped down into a worse pressure group. 4 of the 6 children who showed a drop in pressure group had received an intervention.

57% of children had only one intervention between tympanograms while the remaining 6 children had two or more procedures. The commonest procedure performed was bilateral grommet insertion with adenoidectomy as the second commonest procedure performed. The details of procedures performed within the intervention group is shown in Table 4.

Discussion

Hearing loss has been well documented in children with Down syndrome. The principal cause of this is mid-

Table 4: Interventions performed in between Tympanograms.

No.	First Procedure	Second Procedure	Third Procedure
2	Unilateral Grommet Insertion		
2	Bilateral Grommet Insertion		
2	Bilateral Grommet Insertion + Adenoidectomy		
1	EUA Ears + Adenoidectomy + Tonsillectomy		
1	Bilateral Myringotomy + Tonsillectomy		
1	Bilateral Grommet Insertion	Bilateral Grommet Insertion + Adenoidectomy	
1	Bilateral Grommet Insertion	Bilateral Grommet Insertion	
1	Bilateral Grommet Insertion + Adenoidectomy	Bilateral Myringotomy + Right Grommet Insertion	
1	Bilateral Grommet Insertion + Adenoidectomy	Bilateral Grommet Insertion + EUA PNS + Tonsillectomy	
1	Bilateral Grommet Insertion + Adenoidectomy	Bilateral Myringotomy + Left Grommet Insertion + EUA PNS	EUA Ears + Left Grommet Insertion
1	Bilateral Grommet Insertion	Adenotonsillectomy + Bilateral Grommet Insertion	Bilateral Grommet Insertion

*EUA: Examination Under Anaesthesia; PNS: Post Na.

dle ear effusion which has been reported to be the commonest cause of moderate hearing loss in up to 90% of cases [12,13].

It was found in our patients, that children who had no interventions, 70% (7/10) had no change in middle ear pressure over time. These children all had type B tympanograms during the follow up period with confirmation of OME on otoscopy. There were a small number of children who were difficult to examine, either due to patient cooperation or structural variation. The persistence of the middle ear effusions in these patients is in keeping with other research following the course of OME which demonstrated long term persistence of middle ear effusions [14].

Of those patients who had interventions, most had at least one set of grommets inserted. When comparing the intervention group to the no intervention group it can be seen that a higher proportion, 35.7% versus 10%, of children had some improvement suggesting that those who have intervention are more likely to have an improvement in tympanometry although this may not be back to "normal". Unfortunately, when comparing this to children who do not have Down syndrome, where it can be expected that 66% of OME will resolve within 3 months and a further 20% will resolve within 6 months [15], the differences are clear.

At follow up, no ventilation tubes were present but the majority of patients had recurrent middle ear effusion or worsening of middle ear pressures. The relatively poor outcomes in our study are in keeping with reported low success rates for hearing improvement following ventilation tube insertion, in the short term, for patients with Down syndrome [16].

There have been a myriad of reasons that explain the poor prognosis of OME in Down syndrome. Anatomical differences, such as underdevelopment and collapse of the Eustachian tubes [17], along with mid-facial hypoplasia causing relatively small post nasal space meaning that normal sized adenoidal tissue may cause Eustachian tube dysfunction [18]. Generalised hypotonia has been implicated to cause decreased tensor veli palatini function, thereby making opening of the Eustachian tube ineffectual [19]. Recurrent acute upper respiratory tract infections have been noted in children with Down syndrome which may be due to reduced T and B cell function [20] or defective neutrophil chemotaxis [21] and explains why some patients suffer from persistent OME. This also explains persistent OME and the mechanism may lead to chronic inflammatory changes and impairment of mucociliary transport systems in the nasopharynx and tubal mucosa.

During analysis of our cases, there was no documentation of chronic perforations of the tympanic membrane on review and this was confirmed on tympanography. This is in contrast to previous work that has highlighted the incidence of perforation may be as high as 17-18% [22,23] when compared to normal rates which are estimated from 0.5% to 3.8% [24].

Limitations

Due to the retrospective nature of this study there are some limitations. Firstly, it was found that tympanography was not always performed at every follow up appointment thereby making trend analysis difficult. Furthermore, there was a wide range of ages and time between clinic appointments; hence, the first and last traces were compared with the aim of providing a "snap shot" of any gross changes. There are additional challenges in relation to carrying out tympanometry in children with Down syndrome due to the narrow ear canal. The authors advice caution regarding the ability to apply these results to the whole population with Down syndrome as, clearly, the results may be prone to incidental changes in tympanometry dependant on multiple factors such as: Season, current allergy status and recent upper respiratory tract infections or middle ear infections. Also the results can clearly be affected by any longstanding previously mentioned causes of middle ear dysfunction. If a prospective study were performed then other, more sensitive, methods of measuring middle ear pressure could have been employed including using both 226 hz and 1000 hz probes, as standard, in very young children which have been shown to be better at detecting OME. Furthermore, there is evidence to suggest that 1000 hz probes may be more reliable in identifying middle ear effusions in children with Down syndrome compared to 226 hz probes [25]. The overall numbers within the study make definitive conclusions difficult however there is a lack of data within the literature documenting long term middle ear pressure changes within this group of patients.

Conclusion

This work adds evidence to the effectiveness of ventilation tubes in children with Down syndrome in the long term, although it certainly is not conclusive. In those children who have had no intervention, any middle ear effusion seemed to persist. The current trend to identify and aggressively treat hearing impairment in this patient group, with the aim to help overall development, is important but the treatment modality recommended still requires significant clinical judgement and collective agreement with parents and patients if possible.

The use of ventilation tubes may be effective in certain circumstances; however, this work suggests that even despite intervention, most middle ear effusions recur once the ventilation tube have extruded. There is undoubtedly scope for further prospective research in this field.

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Disclosures

None.

Conflicts of Interest

None.

Mr Mitchell and Mr Turner were involved in data collection, analysis and writing the paper. Dr Holmes was involved in writing the paper.

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