



## Hearing loss in Down Syndrome revisited – 15 years later



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### ABSTRACT

**Objective:** In 2001, the senior authors published a study investigating hearing loss in young children (ages 11 months to 3.8 years) with Down Syndrome (DS). We re-visit this same study population to review current audiologic status, the incidence of pressure equalization tube (PET) placement(s), and rate of tympanic membrane (TM) perforations. We aim to better understand the natural history of ear disease and hearing loss in DS and assess potential complications.

**Methods:** This retrospective chart review included 57 children with DS who previously completed in 2006, a 5 year, longitudinal study investigating otolaryngologic problems in DS. Updated audiologic data was available for 54. Audiograms, age of ear specific testing, PET placement(s), and tympanic membrane(TM) descriptions were reviewed.

**Results:** Ages ranged from 14 to 18 years (mean 16.34 years). PET placement occurred in 88.8%, with mean of 3.5 procedures. 30% of PET's were placed after age 6. Ear specific testing was obtained in 92.5% (mean age 4.54 years). Normal hearing was present in 44% (right ear) and 38% (left ear). "Functional" hearing levels, defined as normal or mild hearing loss and speech reception threshold  $\leq$  30 dB, occurred in 83.3%. Sensorineural/mixed hearing loss was present in 11% (right ear) and 9% (left ear). TM perforations rate was 17%. No cholesteatomas were found.

**Conclusion:** Chronic otitis media and indications for PET's persist as children with DS age. Although functional hearing occurred in 83.3%, there was an overall decrease in hearing levels as the children aged. Tympanic membrane perforations occurred in 17%. Continued surveillance of otologic and audiologic status in patients with Down syndrome is recommended.

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### 1. Introduction

In 1999, 65 children with Down Syndrome (DS) participated in a 5 year longitudinal study at Cincinnati Children's Medical Center, following common otolaryngologic problems seen in DS. Prior to this study, literature regarding otolaryngologic problems in children with DS was largely descriptive and rarely were clinical treatments suggested to improve the medical problems [1–4]. Instead, clinical findings were commonly referred to as being 'part of DS' and were not aggressively treated.

This 5 year longitudinal study provided 'state of the art' medical

and surgical care. This included evaluations and treatments similar to what typical children receive, but in some cases, more frequent surveillance was needed. For example, in the case of treating otologic disease, if a child with DS had stenotic external ear canals, examination under a microscope was done every 3 months so as not to miss a subclinical, asymptomatic chronic effusion. Pressure equalization tubes (PET) were placed after middle ear fluid persisted for greater than 3–4 months and/or after 3–4 consecutive episodes of acute otitis media.

We were not the first to be concerned about inadequate care for children with DS [5,6]. Roizen et al. had shown in their study in 1994 that it was common for children with DS to be underdiagnosed and under-treated for their chronic ear infections (5). In our treatment of otologic disease in DS, we endeavored to see what audiologic outcomes would occur with consistent diagnosis and treatment of the children's chronic ear infections. The motivation

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for this initial study came from our belief that the current literature at the time, which commonly quoted a 75% incidence of hearing loss in DS [2–4], was outdated, retrospective and likely represented consequences of untreated ear disease. With this study, we hoped to establish more accurate norms and expectations for both families and caregivers.

In addition to otologic examinations every 3–6 months, audiologic evaluations were done every 6 months. Our initial paper reported on hearing levels in the first 48 children in this study and was published in 2001 [7]. At the start of the study, 81% of the children had hearing loss ranging from borderline normal-mild loss to severe hearing loss. At the end of the first year in the study, most of the hearing loss seen in this cohort was found to be reversible with medical and surgical treatment. Ninety-eight percent of the children had normal to borderline normal hearing. At the end of the second year in the study, 93% had normal hearing [8].

We now revisit this same group of children, almost 15 years later. Specifically, we examine their current hearing levels, frequency of PET placement, and status of their tympanic membranes.

## 2. Materials and methods

This study was approved by the Institutional Review Board at Cincinnati Children's Hospital Medical Center (CHMC IRB ID 2014–4856) and included children with DS who were originally enrolled, between 1999 and 2001, in a 5 year, longitudinal study at Cincinnati Children's Medical Center, "Otolaryngology Manifestations of Down Syndrome". Sixty-five children were enrolled through the dissemination of information about the study through the Down Syndrome Association of Greater Cincinnati and the Thomas Down Syndrome Clinic at Cincinnati Children's Hospital. Fifty-seven completed the full 5 years of the study. All children were less than 2 years of age at their enrollment into the study and the only exclusion criterion in that study was the inability to speak and understand English.

Only children enrolled in the original study were deemed eligible for participation in this present study. As of June, 2015, there were medical records available for 57 of the 65 children. Of these 57, 3 were excluded due to lack of updated audiologic data from either moving away from Cincinnati or failure to keep follow up appointments and thus lacking up to date audiologic information. Therefore, 54 of the 57 subjects were included for data analysis in this present study.

All behavioral audiologic assessments were done in sound treated booths using audiometric equipment that meet ANSI, S3.6 standards (1996, 2004, 2010). Pure tone testing was done either in a soundfield environment or with supra-aural or insert transducers. Transducer use was determined by what was most appropriate to the test technique and developmental skills of each individual child being evaluated. Transducers were matched to the audiometer and were not interchanged without recalibration. For the majority of testing two audiologists conducted the assessments: one audiologist on the examination side with the child and an adult caregiver, and the other audiologist on the control side with the audiometric equipment.

The audiologic assessments were done in conjunction with the child's otolaryngology appointment and typically included tympanometry, otoacoustic emissions screening, pure tone audiometry and speech audiometry. Since children with Down Syndrome exhibit widely varying levels of developmental skills and sensory integration issues, which can affect their ability to participate in chronological age appropriate audiometric testing, an informal assessment of overall developmental levels was done prior to and during the test session. Using this information aided in determining the most appropriate test technique to assess the integrity of the

auditory system in each ear and measure hearing sensitivities across frequencies.

Using test procedures that are considered to be developmentally appropriate: behavioral observation audiometry (BOA), conditioned orienting response (COR), visual reinforcement audiometry (VRA), conditioned play audiometry or hand raising/response button; minimal response levels were obtained. As in our past study, a comparison of these minimal responses and the Auditory Behavioral Index [9] was used to describe the child's hearing level when the child being tested demonstrated significant developmental delays.

Each child's hearing was assigned a descriptive category by degree of hearing impairment by averaging hearing threshold/minimum responses at 500, 1000, and 2000 Hz: Normal/minimal hearing with hearing threshold/minimal responses between 0 and 25dBHL, mild hearing loss between 26 and 40 dBHL, moderate hearing loss between 41 and 55 dBHL, moderately severe hearing loss between 56 and 70 dBHL, severe hearing loss between 71 and 90 dBHL or profound hearing loss with hearing thresholds  $\geq$  91dBHL. Additionally, each hearing loss was further classified as conductive, mixed, or sensorineural.

In our discussion we also refer to a group of children who are considered to have "functional" hearing levels. This group was comprised of children with normal hearing and children with minimal hearing loss (ie. hearing threshold/minimal responses between 0dBHL and 30dBHL).

Although the entire audiology staff at CCHMC performed the hearing assessments, all test data were reviewed by a single audiologist (DH) to ensure consistency in labeling the minimal response levels and/or thresholds of all the children in this study.

Charts and operative reports were reviewed to determine the number of sets of PET's placed in each child and to determine the current status of the tympanic membranes, specifically for the presence or absence of tympanic membrane perforations.

## 3. Results

When we last reported hearing results on this study population in 2001, the age range was 11 months to 4 years of age. At the time of this current analysis, the ages of the subjects ranged from 14 to 18 years, with mean age of 16.34 years.

Of the 54 children, 48 children or 88.8% underwent PET placement, with "PET placement" representing a trip to the operating room for either unilateral or bilateral PET placement. The number of PET's per subjects ranged from 0 to 11, with 3.5 as the average number of PET placements. Table 1 reviews the incidence and frequency of PET placement in the study group. Seventy-five percent of the children required multiple PET placements, while 16% required just one set. A total of 190 PET sets were placed in the children throughout the 15 years of follow up. Of the 190 PET sets, 133 PET sets or 70% were placed in the children before the age of 6

**Table 1**  
Incidence and Frequency of PET placement in study population.

Number of PET	Frequency	Percent of children
0	4	7.41%
1	9	16.67%
2	10	18.52%
3	4	7.41%
4	12	22.22%
5	5	9.26%
6	5	9.26%
7	1	1.85%
8	1	1.85%
$\geq$ 9	3	5.56%

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