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Original Article

# Bilateral Hearing Loss in Primary Ciliary Dyskinesia: A Study of Conductive and Sensorineural Mechanisms from Pediatric and Adult Cases

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**BACKGROUND:** Primary ciliary dyskinesia (PCD) is a rare genetic disorder that affects the respiratory and auditory systems. This study aims to assess the prevalence, type, and severity of bilateral hearing loss (HL) in PCD and Kartagener syndrome (KS) patients, examining age-related differences and chronic impacts of otologic pathologies.

**METHODS:** A total of 19 patients (38 ears), including 6 children and 13 adults, were evaluated from June to September 2021. Comprehensive clinical examinations included otoscopy, tympanometry, and pure tone audiometry (PTA) for air and bone conduction. Tympanometry findings were compared with otoscopic results. Statistical analyses were conducted using SPSS v16.0 (SPSS Inc.; Chicago, IL, USA), with a significance threshold of $P \leq .05$.

**RESULTS:** Hearing loss was identified in 42.1% of patients, with conductive HL predominant in children (3 out of 4), while mixed HL was more common in adults (3 out of 4). Tympanometry results showed 57.9% type A and 42.1% type B findings, correlating with otoscopic observations. Chronic otitis media with effusion (OME) and tympanosclerosis (TS) were the primary pathologies contributing to middle ear damage. Age was significantly correlated with HL severity ($P = .005$). Mild HL was most common (62.5%), followed by moderate HL (25%) and moderately severe HL (12.5%).

**CONCLUSION:** This study identifies distinct age-related patterns in the type and severity of HL among PCD patients, with sensorineural components observed in adults due to progressive middle ear damage. Audiological evaluations are essential for identifying these complications. Further research is needed to optimize treatment approaches and understand the progression of HL in PCD/KS patients.

**KEYWORDS:** Primary ciliary dyskinesia, hearing loss, Kartagener syndrome, otitis media with effusion, bone conduction

## INTRODUCTION

Primary ciliary dyskinesia (PCD) is a congenital and genetically heterogeneous disorder that primarily impacts the upper and lower respiratory systems, leading to symptoms such as chronic rhinitis and recurrent respiratory infections. When PCD is accompanied by situs inversus totalis, the condition is referred to as Kartagener syndrome (KS).[1,2]

In addition to respiratory and nasal symptoms, individuals with PCD frequently experience otologic manifestations, with a prevalence ranging from 25% to 75%. These manifestations include hearing loss (HL), otitis media with effusion (OME), tympanic membrane perforation, cholesteatoma, and tympanosclerosis (TS).[2-6] The broad spectrum of ear-related complications can be attributed to the presence of cilia in both the middle and inner ear, particularly along the anterior wall proximal to the eustachian tube.

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The evolutionary significance of cilia highlights their initial sensory functions, which later evolved to support motility—an essential role within auditory structures such as the middle and inner ear.[7]

Although previous studies have primarily focused on pediatric populations, emphasizing OME and related conductive HL, our study broadens this perspective by prioritizing bone conduction assessments as part of comprehensive audiological evaluations.

## MATERIAL AND METHODS

This study was conducted between June and September 2021 in collaboration with the Second University Otolaryngology Department and the Third University Pediatric Department of Attikon University Hospital (IRB approval: 285, date: May 29, 2022). Informed consent was obtained by all participants' parents.

Patients diagnosed with PCD or KS underwent a comprehensive clinical examination that included otoscopy, tympanometry, and pure tone audiometry (PTA), assessing both air and bone conduction. Testing frequencies ranged from 250 Hz to 8000 Hz for air conduction and 250 Hz to 4000 Hz for bone conduction, excluding 8000 Hz for the latter.

All adult patients presenting with OME or TS underwent nasopharyngeal endoscopy to rule out the presence of a nasopharyngeal tumor. Informed consent was obtained from all individual participants included in the study.

Statistical analysis were conducted using SPSS version 16.0 (SPSS Inc.; Chicago, IL, USA), with the significance threshold set at $P \leq .05$. Patients under the age of 3 were excluded from the study due to equipment limitations.

## RESULTS

The study sample comprised 19 patients (38 ears), consisting of 6 children and 13 adults. Fourteen participants suffered from PCD, while the other 5 were diagnosed with KS. Male participants outnumbered females nearly 2 to 1 (12 males vs. 7 females). The mean age was 22.84 ± 11.72 years, ranging from 3 to 45 years.

All patients exhibited bilateral audiological and otological manifestations. Eleven out of 19 patients (2 children and 9 adults) exhibited normal findings in otomicroscopy, tympanometry, and PTA. In the remaining 8 cases (4 children, 4 adults), OME was the predominant pathological finding in children (3 out of 4), with one case demonstrating grommets without associated otorrhea upon otoscopic examination. Among adults, TS was present in 3 out of 4 cases, while the remaining case exhibited both TS and OME. No instances of otorrhea or cholesteatoma were observed. The results of tympanometry were consistent with the otoscopic findings, thus 11 out of 19 patients had a type A tympanogram, and the remaining 8tympanograms were type B (7 with reduced and 1 with increased ear canal volume, ECV).

About 62.5% of our patients had mild HL, followed by moderate HL (25%), while one case had moderately severe HL (12.5%). No patient had severe HL—deafness. Mild conductive HL predominated among pediatric patients (3 out of 4), whereas moderate mixed HL was more common among adults (3 out of 4). Comparing the age of the subgroups according to the grade of HL was statistically significant in our study ($P = .005$) but not in comparison to the types of HL ($P = .077$) (Table 1).

### MAIN POINTS

- Conductive HL predominantly affects pediatric PS/KS patients.
- Mixed and sensorineural HL are more common in PS/KS adult patients due to progressive middle ear damage linked to conditions like OME and TS.
- Tympanometry results closely correlate with otoscopic findings, revealing a significant association between age and hearing loss severity but not with the type of hearing loss.
- Regular audiological evaluations are needed, and conservative management is recommended over grommet insertion due to associated risks and potential complications.

## DISCUSSION

Hearing loss was identified in 42.1% of our sample, with the distribution as follows: 15.8% had mixed HL, 10.5% had sensorineural HL, and 15.8% had conductive HL. A statistically significant correlation was observed between age and the grade of HL, though not between age and the type of HL.

These findings contradict the study by Andersen et al, which reported a trend of improving HL with age, accompanied by a reduction in the air-bone gap and the predominance of sensorineural HL.[8] Similarly, Rubbo et al[9] noted that hearing impairment in PCD patients tends to improve with age.

While conductive HL is well-established as the most common type in PCD, our data revealed that 26.3% of participants exhibited a sensorineural component. In contrast, Piatti et al[10] found sensorineural HL in only 1 out of 23 participants, attributing it to causes unrelated to the primary disease. Our findings align more closely with Bequignon et al,[4] who observed sensorineural HL in nearly half of their pediatric patients and OME in one-quarter of the cohort. Similarly, Goutaki et al,[11] in their mixed-age study, reported that adults experienced hearing issues more frequently than children, with age over 30 identified as a risk factor for HL.

It is our opinion that this age-related discrepancy may result from the chronic effects of middle ear inflammation and pressure due to persistent OME, leading to fibrosis and calcification of the middle ear. This hypothesis is supported by the PTA findings of moderate to moderately severe mixed HL in this subgroup. The possible pathophysiological mechanism is that while OME and tympanosclerosis primarily cause conductive HL, prolonged inflammation and recurrent infections can extend to the cochlea via the round window membrane. This can introduce inflammatory mediators or toxins into the inner ear, damaging the hair cells and auditory nerve, ultimately leading to sensorineural or mixed HL. Thus, the cascade of ciliary dysfunction, middle ear effusion, and tympanosclerosis highlights a complex pathway linking middle ear pathology to irreversible cochlear damage.[12-15]

Kreicher et al raised the possibility of concurrent sensorineural pathology in PCD patients, reporting mixed HL in 30% of their cohort.[3] The

**Table 1.** Demographics, Clinical, Otological, and Audiological Findings

| Patient | Sex | Age | Grade of HL | Type of HL | Lost Frequencies | Tympanometry | Otoscopic Findings |
|---|---|---|---|---|---|---|---|
| PCD | F | 22 | Moderate | Mixed, BL | All | B | TS |
| PCD | F | 19 | | | | A | |
| PCD | M | 18 | | | | A | |
| PCD | F | 33 | | | | A | |
| KS | M | 12 | Mild | Conductive, BL | All | B | OME |
| PCD | M | 11 | Mild | Conductive, BL | All | B | Grommets without otorrhea |
| PCD | M | 20 | | | | A | |
| PCD | M | 16 | | | | A | |
| KS | M | 29 | | | | A | |
| PCD | M | 3 | Mild | Conductive, BL | All | B | OME |
| PCD | F | 29 | | | | A | |
| PCD | F | 19 | | | | A | |
| PCD | M | 44 | | | | A | |
| PCD | M | 43 | | | | A | |
| PCD | M | 45 | Moderately severe | Mixed, BL | All | B | TS, OME |
| KS | M | 20 | Mild | Sensorineural, BL | High | B | TS |
| KS | M | 24 | Moderate | Mixed, BL | | B | TS |
| KS | F | 12 | Mild | Sensorineural, BL | High | B | OME |
| PCD | F | 14 | | | | A | |

BL, bilateral; KS, Kartagener syndrome; OME, Otitis Media with Effusion; PCD, primary ciliary dyskinesia; TS, tympanosclerosis.

primary distinction between their study and our results lies in participant demographics, as their research exclusively involved children. This may explain the absence of sensorineural HL in their sample, as OME tends to subside after the age of 12 when mucociliary clearance improves.

The majority of cases in our study demonstrated mild HL, with moderate HL being the next most common. Mixed HL was associated with more severe impairment, as anticipated, given the involvement of both air and bone conduction. All cases exhibited symmetrical HL, consistent with the findings of Campbell et al,[16] though contrasting with studies in the general pediatric population with OME, where HL is often asymmetrical, short-term, and fluctuating.

The tympanograms were type A in 57.9% of cases, correlating with normal otoscopic findings, while 42.1% exhibited type B tympanograms. Among those with type B tympanograms, 37.5% had OME, another 37.5% had tympanosclerosis (TS), and 12.5% presented with a combination of TS and OME. Notably, most type B cases exhibited reduced ear canal volume (ECV <1.5 mL), except for one patient with grommets who had an increased ECV (2.1 mL). These findings align with literature emphasizing the persistence of OME in PCD patients even beyond the age of 18.[17-19] The 11-year-old child with grommets had properly functioning devices with no observed discharge, although mild conductive HL was present. The decision to insert grommets in PCD patients remains controversial.

Conservative management is often preferred due to the high risk of persistent otorrhea and the potential for secondary cholesteatoma. While only 2 studies support grommet insertion, Lam et al argue for comprehensive upper respiratory disease management as an interdependent approach for PCD patients.[20-22]

Campbell et al[16] reported persistent otorrhea in 33% of PCD patients with grommets, compared to 3.8%-13.2% in the general population. Similarly, Zawawi et al[23] observed chronic otorrhea in 23% of children with ventilation tubes. Günaydın et al[6] highlighted increased tympanic membrane injuries, including sclerosis, perforation, and retraction, in patients with OME, potentially exacerbated by ventilation tube insertion. These complications may stem from poor mucociliary clearance in the nasopharynx, bacterial contamination, and foreign body reactions to the grommets. Although no cases of otorrhea or cholesteatoma were observed in our study, Wolter et al[24] reported bilateral cholesteatoma in 3 out of 31 participants. In our opinion, grommet insertion does not significantly improve hearing, a conclusion supported by previous studies.[3,8]

**Limitations**

Limitations include a small sample size due to the rarity of the disease, exacerbated by the COVID-19 pandemic and patient reluctance to visit hospitals. Additionally, focusing solely on the middle ear and anterior labyrinth overlooks potential impacts on the posterior labyrinth, where cilia are present. In the Rimmer et al study,[25] the impact of the disease on the anterior labyrinth was considered minimal, on the contrary to the posterior labyrinth, as the VOR tests were pathological.

## CONCLUSION

Primary ciliary dyskinesia/KS is a rare genetic disorder that affects the auditory system. This study highlights the prevalence of bilateral

hearing loss in PCD/KS patients, with conductive HL predominantly observed in children and mixed or sensorineural HL more common in adults. The condition involves impaired mucociliary function and sensorineural mechanisms, with the contribution of each varying by age. Chronic otitis media with effusion significantly contributes to progressive middle ear damage over time. Tympanometry findings, consistent with otoscopic results, underscore the persistence of middle ear pathologies in PCD/KS patients. Audiological evaluations and conservative management are crucial due to the risks associated with surgical interventions such as grommet insertion. Further research is essential to refine treatment strategies and better understand the progression of HL in PCD.

**Availability of Data and Materials:** The data that support the findings of this study are available on request from the corresponding author.

**Ethics Committee Approval:** This study was approved by the Ethics Committee of Attikon University Hospital (approval no: 285, date: May 29, 2022).

**Informed Consent:** Written informed consent was obtained from the participants' parents who agreed to take part in the study.

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