# Hearing rehabilitation with bone conduction implant in patient with primary ciliary dyskinesia – a particular indication

Clinical Case


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## Abstract

Primary ciliary dyskinesia (PCD) is a rare genetic disease characterized by an abnormality in the function or structure of the cilia, coursing with pulmonary, otological and nasosinusal manifestations. Hypoacusis affects 60% of patients with PCD and results mainly from otitis media with effusion (OME) and otological complications such as simple or cholesteatomatous chronic otitis media (COM).
Clinical case: Female patient, 52 years old, with PCD, followed in the Otorhinolaryngology department for bilateral COM and chronic rhinosinusitis. Her surgical history includes nasosinusal endoscopic surgery and repeated myringotomies with placement of transtympanic ventilation tubes (TVTT), bilaterally. Despite the placement of TVTT, the simple tone audiogram revealed bilateral severe chronic mixed deafness, with mean bone thresholds of 36.25 dB on the right and 30 dB on the left. Ear tomography excluded erosion or discontinuity of ossicular chains, presence of cholesteatoma and pathology of the inner ear. The patient underwent surgery to place a bone conduction implant in her right ear. After 4 weeks, the processor was activated. The free field audiogram revealed a mean threshold of 26.25dB.
Conclusion: In PCD patients, insertion of TVTT is still a controversial treatment for OME. Persistent postoperative otorrhea is a frequent side effect and may complicate the use and tolerance of hearing aids. Bone conduction implants should be considered in auditory rehabilitation, with excellent results.


Keywords: primary ciliary dyskinesia; bone conduction implant; mixed deafness; transmission deafness

## Introduction

Primary ciliary dyskinesia (PCD) is a rare genetic disease characterized by impaired mucociliary clearance due to functional or structural ciliary abnormalities with a prevalence of 1/20,000 births and autosomal recessive or X-linked transmission.[1] PCD patients usually present with pulmonary changes (chronic bronchiectasis or recurrent infections) and sinonasal and otological manifestations (chronic rhinosinusitis and otitis media). Other conditions may include, less frequently, fertility disorders and, rarely, hydrocephalus, cardiac malformations, esophageal pathology or biliary atresia.[2] By contrast, 50% of patients with PCD show *situs inversus*, which leads to Kartagener's syndrome when associated with bronchiectasis and chronic rhinosinusitis.[3]

The main otological manifestation of PCD is otitis media with effusion (OME).[4] The middle ear mucosa consists of respiratory-type (ciliated columnar) epithelium.[5] Thus, in patients with PCD, mucociliary function is compromised, as is effusion viscosity, leading to OME. In addition, otological complications, such as tympanic perforation, retraction pockets and cholesteatoma are observed in more advanced stages.[6,7] Consequently, hearing loss affects nearly 50% of adults with PCD.[8,9]

In the general population, myringotomy with transtympanic ventilation tube insertion is indicated for persistent OME with 3 months of progression, unresponsive to medical treatment, resulting in conductive hearing loss (average air conduction thresholds higher than 20 dB) or recurrent acute otitis media (RAOM), with 3 episodes in 6 months or 4 episodes in 12 months.[10]

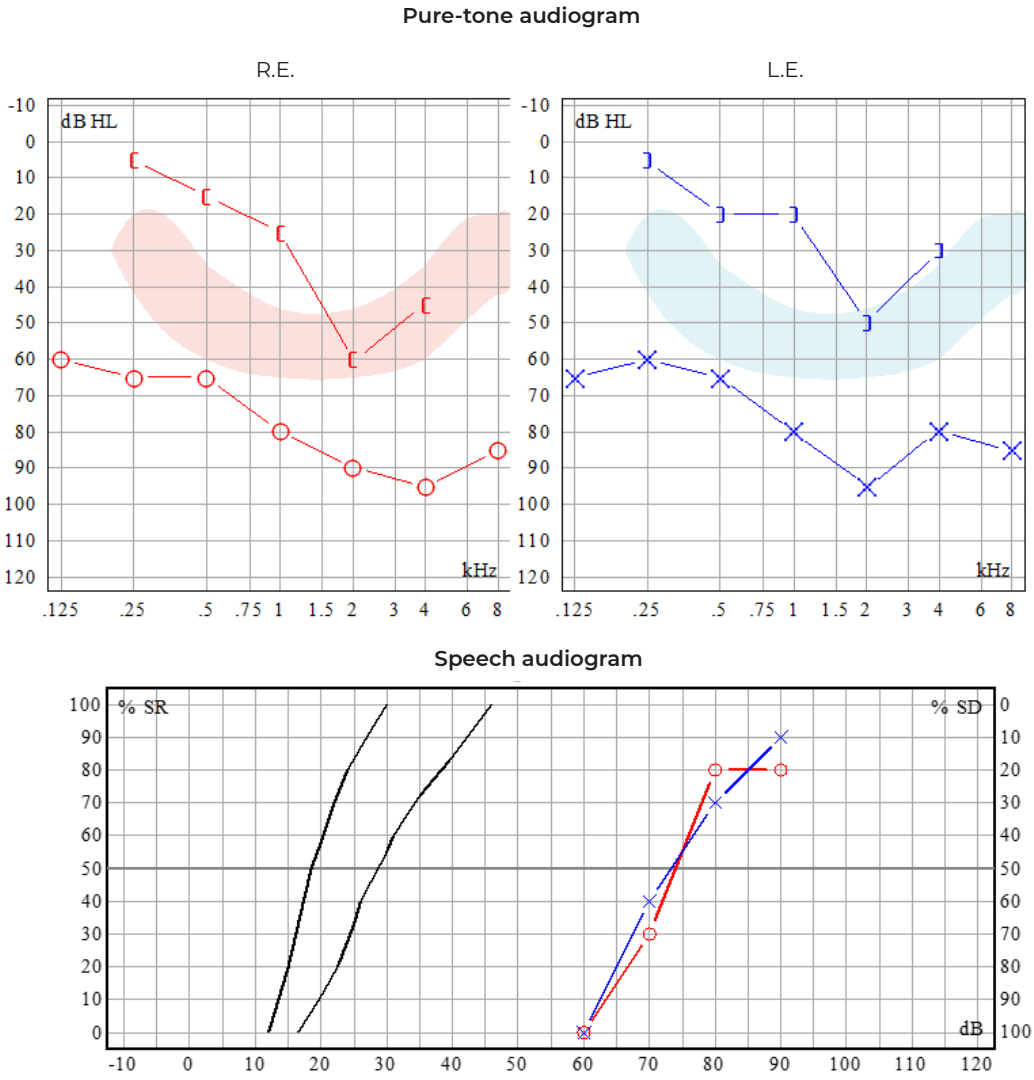
However, in PCD, TTVT insertion remains a controversial treatment for OME and RAOM.[11,12] In fact, no evidence has shown that TTVT insertion outperforms medical treatment in improving hearing.[13,14] Furthermore, persistent postoperative otorrhea is a frequent and problematic side effect of myringotomy with TTVT insertion, particularly in patients with PCD, affecting approximately 50% of cases, which may complicate hearing aid use and tolerance.[15]

An aural rehabilitation option for patients with PCD is bone conduction device (BCD) implantation. BCDs use bone vibration to directly stimulate the cochlea, bypassing outer and middle ear structures. They are divided into percutaneous (penetrate the skin) and transcutaneous (do not penetrate the skin) BCDs. The latter are further divided into passive and active transcutaneous BCDs. They are indicated in cases of conductive/mixed hearing loss or unilateral sensorineural hearing loss when conventional hearing aids are not indicated or tolerated.[16]

## Clinical case

SMFR, a 52-year-old Caucasian woman, with a medical history of Kartagener's syndrome and major depression, has been followed up in the Otorhinolaryngology (ORL) outpatient clinic at the Coimbra University Hospital Centre (*Centro Hospitalar e Universitário de Coimbra* – CHUC) for chronic rhinosinusitis and bilateral chronic otitis media since 2000. She is usually medicated with formoterol combined with budesonide nebulizer suspension, fluticasone propionate nasal spray, montelukast, mirtazapine and alprazolam and is unaware of drug allergies. Her surgical history includes long-term myringotomy with TTVT insertion 3 times (2001, 2008 and 2017) and functional nasal surgery with antrostomy and complete bilateral ethmoidectomy, in 2013. Despite complaints, her nasal condition was stable under frequent nasal hygiene and topical medical therapy. Her main complaints were hearing loss and decreased discrimination capacity, with no improvement after the last targeted surgery. General ORL examination revealed bilateral otoscopy with normally positioned long-term TTVTs, with mucus in the left ear, but not in the right ear. On anterior rhinoscopy, only bilateral inferior turbinate hypertrophy was noted, without bilateral rhinorrhea. Rhinoscopy showed no nasal polyposis or mucopurulence. No significant changes were detected by oral cavity and

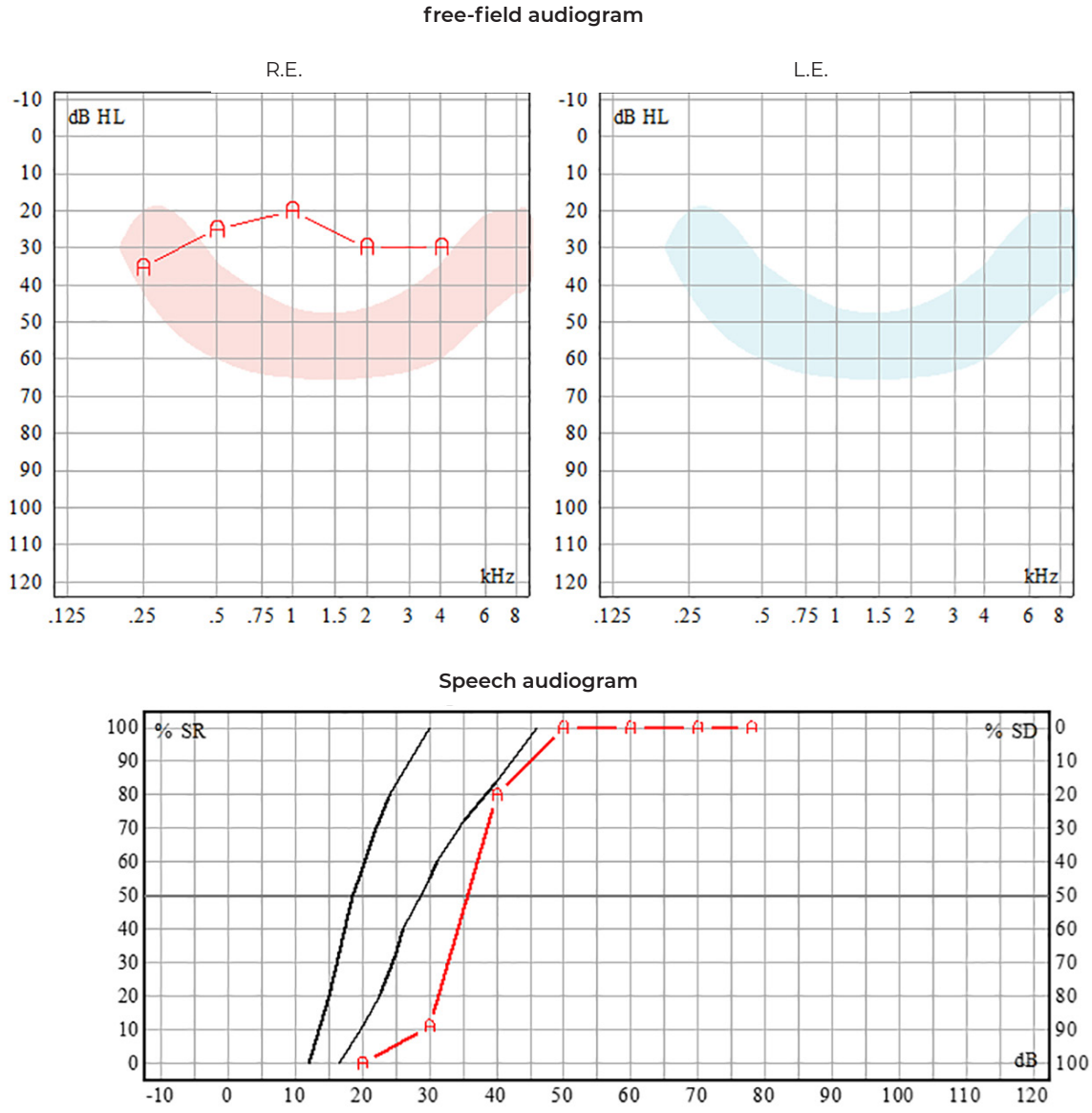
**Figure 1**
Pure-tone and speech audiograms prior to bone conduction device implantation




oropharynx examination or cervical palpation. A pure-tone audiogram showed bilateral severe mixed hearing loss, with a Pure-Tone Average (PTA) of 82.5 dB on the right ear and 80 dB on the left ear. The average bone conduction thresholds were 36.25 dB on the right ear and 30 dB on the left ear. A speech audiogram revealed a Speech Reception Threshold (SRT) of 75 dB, bilaterally.

Computed tomography (CT) imaging revealed a sclerotic pattern in the mastoid process of the temporal bone in both ears, with tissue filling the remaining mastoid cells and mastoid antrum and even the tympanic cavities, more significantly in the left ear. In this ear, this process extended to the anteromedial aspect of the external auditory canal (EAC), bilaterally highlighting TTVTs and window recess opacification. No erosive changes were identified in ossicular chains, scuta or tegmen tympani. Therefore, these images suggested chronic otomastoiditis, albeit without cholesteatoma or marked changes in the morphology or permeability of the cochleae, vestibules, semicircular canals or vestibular aqueducts. The internal auditory canals (IACs) had normal dimensions and morphology, without anomalous filling of the

**Figure 2**
Free-field and speech audiograms after bone conduction device implantation




cerebellopontine angle cisternae detectable on tomographic imaging. The EACs were permeable. The CT sections showed signs of chronic pansinusitis in the sphenoidal and maxillary regions.

The patient was followed up for several years in an ORL outpatient clinic, repeating serial audiometry. After the last TTVT insertion, she maintained auditory thresholds, objective examination findings and complaints of disabling hearing loss and communication difficulties. The option of aural rehabilitation with hearing aids was considered but ultimately disregarded due to persistent otorrhea. Accordingly, the patient performed a bone conduction test (BCT), which showed favorable results in the right ear, undergoing surgery for bone anchored hearing aid (BAHA) implantation in this ear, without complications. The right ear was chosen based on the BCT results and patient's preference and for spatial organization reasons. Although the bone threshold and, initially, device performance were better in the left ear, the BCD was

implanted in the right ear considering the BCT results. In addition, the patient was a secretary and, for spatial organization reasons, used the right ear more. Thus, the right ear was whose to implant the device. In all these cases, we prioritized the BCT over the speech audiogram.

The sound processor (BAHA T Power) was placed four weeks after the surgery. The patient adapted well to the device, which was used, on average, 10 hours per day. No complications were recorded in the postoperative period. The average free-field hearing threshold with the BCD was 26.25 dB, and the SRT in the speech audiogram was 35 dB, reaching maximum intelligibility (100%) at 50 dB.

The patient is continuing her outpatient ORL follow-up and is quite satisfied with the audiometric outcome.

## Discussion

The otological manifestations of patients with PCD derive from dysfunctional cilia of the epithelium of the middle ear and auditory tube. This functional deficit impairs mucociliary clearance and, thus, predisposes patients with PCD to OME and RAOM, which occur almost invariably in this population.[17] In the general population, hearing loss associated with OME is an indication for TTVT insertion. However, there is no consensus on this aural rehabilitation approach for the population with PCD.[6] TTVT insertion in patients with PCD shows limited efficacy due to systemic and persistent otorrhea, which is more common in these patients, and its obstruction by viscous glue in the middle ear. Moreover, repeated TTVT insertion is associated with a high risk of persistent tympanic perforation.[18,19]

Unlike OME in the general population, OME in patients with PCD is not caused by poor ventilation of the middle ear, which explains why TTVT insertion fails in these patients. Therefore, for patients with PCD and with a hearing loss above 25 dB, the decision to undergo treatment with myringotomy with TTVT insertion should be made considering its risks and benefits, the almost inevitable occurrence of persistent otorrhea. In this context, hearing aids emerge as an aural rehabilitation option.[20] TTVT insertion may avoid OMC complications resulting from persistent OME in patients with PCD, but research has shown that the number of cases of retraction pockets and cholesteatomas remains low without TTVT insertion. For this reason, this approach remains controversial, requiring further studies.[6]

In our clinical case, the patient repeatedly underwent myringotomy with TTVT insertion. These surgical procedures initially helped to improve her hearing, but the patient subsequently showed no significant hearing gain. In turn, due to persistent otorrhea, the patient was unable to tolerate hearing aids for improving hearing loss and decreased discrimination capacity, which impaired her communication on a daily basis. Therefore, we selected BAHA percutaneous BCDs, which were surgically implanted, using the processor T Power.

Currently, whenever possible, transcutaneous BCDs tend to be use because they are more aesthetically appealing and entail fewer skin complications. However, percutaneous implants are still indicated in cases of mixed hearing loss where transcutaneous implants are not effective enough, as in the case presented in this study.

In comparison with percutaneous devices, passive transcutaneous devices, including the BAHA Attract system (Cochlear) system, suffer from attenuation resulting from signal loss during transmission through the skin and soft tissues. This attenuation is more evident at high frequencies and can reach up to 25 dB at frequencies ranging from 6000 to 8000 Hz.[21]

Active transcutaneous devices include the Bonebridge bone conduction implant (MED-EL) and the Osia® system (Cochlear Americas), which was recently introduced in the market. Bonebridge is indicated for patients with conductive and mixed hearing loss with bone conduction hearing thresholds equal to or better than 45 dB HL or unilateral hearing loss.[22] The Osia® system is indicated

for patients with mixed or conductive hearing loss with bone conduction thresholds equal to or better than 55 dB HL, and unilateral hearing loss.[23] Among percutaneous BCDs, the Ponto (Oticon Medical) and Baha® Connect (Cochlear) systems stand out. They are indicated for cases of conductive or mixed hearing loss with bone conduction thresholds lower than 45 dB. Nevertheless, thanks to processors developed for the Baha® Connect system, such as the 5 SuperPower processor, these devices can be applied for hearing losses of up to 65 dB bone conduction.[24] Therefore, the Baha® Connect system and the 5 SuperPower processor were selected based on 60 dB bone thresholds at 2,000 Hz in the right ear.

BCD implantation is primarily indicated for otological diseases such as EAC atresia, chronic otitis media (COM), EAC stenosis, otosclerosis, unilateral hearing loss and post-mastoidectomy. Among specific indications, enlarged vestibular aqueduct, Mondini dysplasia and primary ciliary dyskinesia stand out. The last condition is the disease analyzed in this clinical case.[25] The patient presented with severe mixed hearing loss. Once the implant was placed, the average free-field hearing threshold increased to 26.25 dB, and on the speech audiogram, SRT was 35 dB.

## Conclusion

Patients with PCD must be closely followed up, and physicians should adopt a specific therapeutic approach. In these patients, TTVT insertion remains a controversial treatment for OME and da RAOM. Persistent postoperative otorrhea is a common side effect, which may complicate hearing aid use and tolerance. Moreover, BCDs should also be considered in aural rehabilitation in patients with PCD because they show excellent audiometric outcomes.

### Conflicts of Interest

The authors declare that there is no conflict of interests regarding the publication of this paper.

### Data Confidentiality

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

### Protection of humans and animals

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the 2013 Helsinki Declaration of the World Medical Association.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### Availability of scientific data

There are no datasets available, publicly related to this work.

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