

Bone conduction hearing device use for the management of hearing loss in children and young adults with Down syndrome

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Background

Hearing loss is common among individuals with Down Syndrome (DS), with an estimated prevalence of 40-80% throughout childhood and early adulthood.¹ Hearing loss may have significant impacts on speech, language and cognitive development.

- **Conductive hearing loss (CHL)** is the most commonly diagnosed type of hearing loss, comprising ~80% cases
 - CHL involves impaired transmission of sound passed the outer and middle ear
 - 40-60% of cases of CHL are attributed to **chronic otitis media with effusion (COME)**²
 - Middle ear disease and CHL may persist or fluctuate, despite treatment with pressure equalization tubes (PET)³

- **Bone conduction devices (BCDs)**, worn externally on a head band or implanted into bone behind the ear, transmit sound through bone directly to the inner ear (Fig. 1)
 - Bypasses outer and middle ear obstruction in CHL
 - Beneficial in **single-sided deafness**, defined as profound unilateral hearing loss with normal hearing in the contralateral ear, as sound is transmitted via bone to functioning ear (Fig. 2)
 - Conventional hearing aids utilize air conduction, which requires functional outer and middle ear spaces

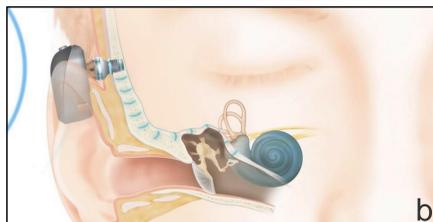


Figure 1. Sound conduction in a normal outer and middle ear (a) and with a bone conduction device, bypassing the outer and middle ear (b).

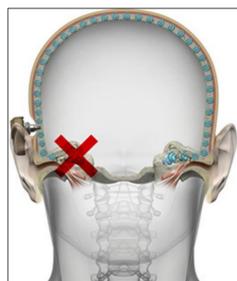


Figure 2. Sound conduction in single-sided deafness: sound from the side of the hearing loss is transmitted via BCD through bone to the contralateral, normally functioning ear.

Objectives / Aims

This case series aims to:

- Describe the clinical characteristics and indications associated with BCD use, and
- Evaluate outcomes and complications related to BCD use among individuals with DS and hearing loss

Design / Methods

- Case series of 15 patients (1.5 – 27 years) with DS and hearing loss managed with a BCD
- Patients identified from an audiological database at Cincinnati Children's Hospital Medical Center
- Retrospective chart review with descriptive analysis, utilizing clinical encounters in pediatric otolaryngology, audiology, and developmental pediatrics

Results

Indications for BCD Placement:

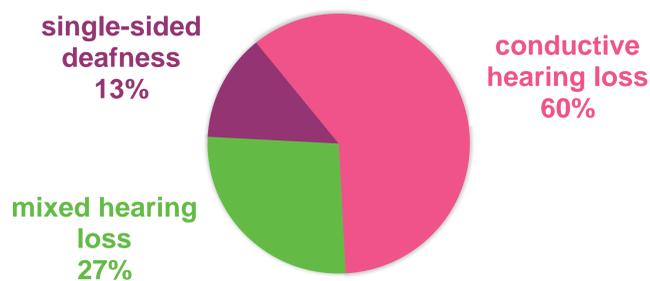


Figure 3. Indications for intervention with BCD in the studied patient population.

Outcomes:

- 66.7% successful BCD use, defined as long-term use, use of BCD until resolution of underlying hearing loss, and intermittent use of BCD when symptomatic
- 20% were lost to follow-up
- Complications were documented in 53% of patients. However, most were minor or transient complications, and 62.5% ultimately used BCD successfully.
- Of the 40% of patients who underwent aided audiogram testing, all demonstrated improved hearing thresholds when aided (compared to unaided thresholds)

Results

Table 1: Patient Characteristics

	Number of patients (% of total study)	% successful BCD use [†]
Gender		
Male	6 (40.0%)	50.0%
Female	9 (60.0%)	77.8%
Ethnicity		
White, non-Hispanic	13 (86.7%)	69.2%
Black or African-American	1 (6.7%)	0.0%
Hispanic	1 (6.7%)	100.00%
Type and Severity of Hearing Loss[‡]		
Conductive	9 (60.0%)	55.6%
Mild	1 (6.7%)	0.0%
Moderate	3 (20.0%)	100.0%
Severe	5 (33.3%)	40.0%
Mixed	4 (26.7%)	75.0%
Mild	-	-
Moderate	2 (13.3%)	100.0%
Severe	2 (13.3%)	50.0%
Single-Sided Deafness	2 (13.3%)	100.0%
Age at Initiation of BCD		
0-3 years	7 (46.7%)	57.1%
3-10 years	5 (33.3%)	60.0%
10-18 years	-	-
18+ years	3 (20.0%)	100.0%
Type of BCD		
Soft band	11 (73.3%)	72.7%
Adhesive adapter	1 (6.7%)	100.0%
Surgical implant	3 (20.0%)	33.3%
Prior Conventional Hearing Aid Use		
Yes	4 (26.7%)	100.0%
No	11 (73.3%)	45.4%

Associated ENT Diagnoses		
COME	14 (93.3%)	71.4%
EAC stenosis	2 (13.3%)	50.0%
Aural atresia	1 (6.7%)	0.0%
CSOM or cholesteatoma	1 (6.7%)	100.0%
TM perforation	3 (20.0%)	100.0%
EVA	1 (6.7%)	100.0%
Cochlear nerve hypoplasia	1 (6.7%)	100.0%
Prior ENT Interventions		
Myringotomy	3 (20.0%)	66.7%
PET placement	10 (66.7%)	60.0%
Tympanoplasty +/- mastoidectomy	3 (20.0%)	100.0%

[†] Successful user defined as patients with long-term use of BCD, use of BCD until underlying hearing loss resolved, and intermittent use of BCD when symptomatic

[‡] Severity of hearing loss determined by degree of hearing loss in worse hearing ear, when applicable

COME – chronic otitis media with effusion
EAC – external auditory canal
CSOM – chronic suppurative otitis media
TM – tympanic membrane
PET – pressure equalization tube
EVA – enlarged vestibular aqueducts

Results

Table 2: Patient Outcomes

Duration of Use ^{‡‡}	Median (Range)	
	340 days (36-1173 days)	
Device Use		
Number of patients (% of total study)		
Successful use	10 (66.7%)	
Long-term	7 (46.7%)	
Until resolution of HL	2 (13.3%)	
As needed	1 (6.7%)	
Declined long-term use	1 (6.7%)	
Complex behavioral issues prevented use	1 (6.7%)	
Device Complications		
Number of patients (% of total study)		
% successful BCD use		
Any complication	8 (53.3%)	62.5%
Frequent removal	4 (46.7%)	57.1%
Damaged device	2 (13.3%)	100.0%
Skin or soft tissue complication	1 (6.7%)	100.0%
Sound distortion	1 (6.7%)	100.0%

^{‡‡} Duration of use calculated as date of initiation of BCD to date of most recent follow-up or date BCD discontinued. Excludes three patients lost to followup.

Conclusions

- BCDs are an acceptable and effective intervention for HL in selected patients with DS
- BCDs provide consistent access to sound for those with long-standing chronic or fluctuating middle ear disease
- Further research aims to evaluate quality of life and developmental outcomes associated with BCD use

References

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- 3 Sidell D, Hunter L, Lin L, Arjmand E. Risk factors for preoperative and postoperative hearing loss in children undergoing pressure equalization tube placement. *Otolaryngol Head Neck Surg.* 2014; 150(6):1048-1055.

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