



# Evaluation of co-morbid medical conditions in adults with Down syndrome: Treatment of hearing loss - grading the evidence and identifying research gaps

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## BACKGROUND

- Individuals with Down syndrome (DS) are at increased risk of hearing loss compared to the general population (38-78% vs 1-15%)<sup>1-2</sup>
- Narrowed ear canals, hypotonic eustachian tubes, and higher incidence of otitis media are factors that predispose those with DS to persistent hearing loss requiring repeated treatments<sup>2-3</sup>.
- While studies have evaluated the treatment options for hearing loss in children with DS<sup>4</sup>, there is a scarcity of evidence for the treatment of hearing loss in adults with DS.

## RESULTS/SUMMARY

Search Term: DS + (CONDITION) No limits applied	NLM/PubMed Limits applied (Human, English, >18yr)
Unfiltered Hits = 224 articles (hearing loss) 24 articles (hearing aids)	Filtered Hits = 67 articles (hearing loss) 8 articles (hearing aids)
Reviewed Title/Abstract	Excluded articles=XX <ul style="list-style-type: none"> <li>Not adults: 63 (hearing loss) 2 (hearing aids)</li> <li>Nonquantitative: 1 (hearing aids)</li> <li>Case report: 1 (hearing loss) 2 (hearing aids)</li> </ul>
Reviewed Article	Included articles = 3 total (found in both search terms)

### DATA SUMMARY

- All three reviewed studies focused exclusively on bone anchoring hearing aids (BAHA) as a treatment modality and demonstrated a subjective benefit to patients who used BAHA.
- Two studies (Sheehan 2006, Kunst 2006) described complications of BAHA treatment, which were relatively common and included skin and soft tissue complications and osseointegration failure.
- In the majority of studies excluded, children with DS were the studied population.
- The included studies had a wide range of ages including children and included subjects with varying degrees of conduction or mixed hearing loss (mild to severe).
- Two of these studies (Sheehan 2006, Kunst 2006) had inadequate control groups.
- All three studies were limited to the European setting and had small sample sizes.

## CONCLUSIONS

### Major gaps identified in the literature:

- Overall, there are very few studies evaluating the treatment of hearing loss in adults with DS, and no studies that focus exclusively on this patient population.
- No comparison of air-conducting hearing aids to bone-conducting hearing aids.
- A lack of prospective large sample size studies with proper control groups.
- Results were not stratified by age group in studies that included both children and adults.
- A lack of studies evaluating the financial costs and accessibility of treatment options.
- A lack of data examining whether medical comorbidities influence the availability and success of treatment options.
- Limited longitudinal follow up on complications and quality of life after BAHA implantation.

### For further consideration towards practical guidelines:

- Adults with DS who are treated with BAHA have high rates of daily BAHA use and report high satisfaction with the system despite some difficulty cleaning the abutment and the relatively frequent occurrence of post-operative skin and soft tissue complications.
- For adults with DS with chronic otitis media or those who have failed conventional hearing aids, BAHA should be considered.
- There is insufficient data to adequately compare the efficacy of BAHA in adults with DS to other treatment options such as air conducting hearing aids.

## OBJECTIVES/AIMS

- Identify original research articles in peer-reviewed medical journals that focus on key questions about the treatment of hearing loss in adults with DS
- Evaluate the quality of the existing evidence, identify deficiencies in current clinical knowledge and suggest directions for future research.
- Begin to formulate practical guidelines to support best medical practices for the treatment of hearing loss in adults with DS.

## DESIGN/METHODS

Using the National Library of Medicine biomedical literature database PubMed (NCBI 1960-2021), we performed a literature search on the treatment of hearing loss in adults with DS. Critical key questions were formulated *a priori* to inform the search strategy. The MeSH terms [Down syndrome] and "Hearing Loss" or "Hearing Aids" were combined in both the title and abstract search fields.

By consensus the following key questions were formulated:

- What are the treatment options for adults with DS?
- What other comorbidities affect treatment for these patients?
- For adults with DS what is the availability and comparison of treatment options for hearing loss?
- What is the impact of treatment on morbidity or mortality?
- What are the financial costs or harms of treatment?
- What are the treatment differences for those with DS versus the general population?

PubMedID (PBID)	16310862	16788427	17721366
Included Articles	Sheehan, Hans	Kunst, Hol	Kunst, Hol
Publication Year	2006	2006	2007
Subjects studied (N)	43 (14 above age of 18) with mild to moderate hearing loss	22 (12 with Down syndrome; 20 above age of 18) with moderate to severe hearing loss	22 (12 with Down syndrome; 20 above age of 18) with moderate to severe hearing loss
Age range	3 to 64	10 to 74	10 to 74
Source of subjects	UK Centers performing bone anchoring hearing aids	Netherlands; patients selected for bone anchored hearing aids	Netherlands; patients selected for bone anchored hearing aids
Methods	Survey	Retrospective	Retrospective
Study design	Descriptive study	case series	case control study
Comorbidities considered	Chronic suppurative otitis media, otitis media with effusion, canal stenosis, prior use of air-conducting hearing aids	Prior use of air-conducting hearing aids	Prior use of air-conducting hearing aids

Key Questions			
Treatment options	bone anchoring hearing aids	bone anchoring hearing aids	bone anchoring hearing aids, middle ear surgery, cochlear implantation
Medical comorbidities considered	chronic suppurative otitis media, otitis media with effusion, canal stenosis, otitis externa	chronic otitis externa, chronic otitis media, severe pain/skin irritation/headaches from pressure of BAHA against skin	Chronic otitis media, mastoiditis
Availability and comparison of treatment options	na	audiometry for BAHA vs prior hearing aids in patients was compared and found to be nonsignificant (n=9)	by Glasgow Benefit Inventory (GBI), BAHA more beneficial than middle ear surgery but slightly less beneficial than cochlear implant
Morbidity or mortality affected	96% patient/caregiver satisfaction with BAHA	Subjective remarkable improvements in daily activity in at least 23% of patients; all patients continued to use BAHA daily at ≥5 months post-implantation	subjective overall patient benefit demonstrated by GBI scores
Costs and harms of treatment	post-op soft tissue complications; most commonly excessive healing of graft site with hypertrophy of soft tissue onto abutment of BAHA	Implant loss, post-op soft tissue complications	na
Treatment differences to general population	na	similar implant loss, rate of skin complications compared to general population with BAHAs	na
USPSTF Ratings*			
Research design	III	II-3	II-2
Internal validity	Poor	Fair	Fair
External validity	Poor	Fair	Fair

## REFERENCES

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## \*USPSTF Research Design Reference

- I: Properly conducted randomized controlled trial (RCT)
- II-1: Well-designed controlled trial without randomization
- II-2: Well-designed cohort or case-control analytic study
- II-3: Multiple time series with or without the intervention; dramatic results from uncontrolled experiments
- III: Opinions of respected authorities, based on clinical experience; descriptive studies or case reports; reports of expert committees

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