

CASE REPORT

Bone Anchored Hearing Aids in Children with Cleft Palate

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Ear pathology, conductive hearing loss and associated communicative disorders are often present in cleft palate children.

These children usually require multiple grommets insertion as well as other middle ear surgeries in early years of life because of increased prevalence of chronic otitis media, retraction pockets and cholesteatoma. These sequelae arise from the long standing of the otitis media with effusion itself, as from the complication of multiple ventilation tube insertions.

The BAHA system should be viewed as an important and possible way of bypassing these problems and have a definitive indication in some of these children.

In the ENT Department of our hospital (tertiary care children's hospital) we have six cleft palate children fitted with unilateral BAHA.

It is important to review all the therapeutic options within the members of the team, involving the parents or caregivers of these children. The audiologist and the otorhinolaryngologist involved in the cleft palate team must always bear in mind that bone-anchored hearing aids are a useful tool in management of children with cleft palate.

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The incidence of cleft palate is 1:500-1,000 living births, making it one of the most common congenital defects. The presence of middle ear effusion in children with cleft palate is almost universal^[1, 2].

The pathogenesis of otitis media with effusion (OME) is due to poor tensor veli palatini function and increased Eustachian tube compliance with its inability to open actively on swallowing. Middle ear pathology leads to conductive hearing loss (CHL) in 50 to 93% of patients. This high incidence of OME and hearing loss is also found after surgical repair of the cleft leading to persistent otologic pathology. Long-term hearing loss with cleft palate patients occurs in up to 24%^[1-3].

Case Presentations

Case 1

A 6-year-old caucasian boy was first seen in our ENT Department with a history of hearing loss.

The otoscopy revealed an atelectatic right ear and chronic otitis media in the left ear.

Pure-tone audiometry disclosed a 35 dB CHL on the right ear and a 40 dB CHL on the left.

Under general anesthesia lateral adenoidectomy was performed with VT insertion on right ear (grommet). Lateral adenoidectomy was performed because adenoids obstructed the Eustachian tube bilaterally and the central part of the adenoids was left in place to prevent velopharyngeal insufficiency. VT remained in place for only 7 months, with recurrence of pathology after extrusion.

At the age of 8 a large cholesteatoma developed from the central perforation in the left ear and he was operated on a canal wall down procedure because the extension of the disease.

At the age of 9 we achieved a reaction free cavity in the left ear and had a right atelectatic ear. The parents refused reconstructive surgery on right ear (Figure 1a). The audiogram at that time revealed 35 dB CHL in the right ear and 55 dB CHL on the left (Figure 1b). The

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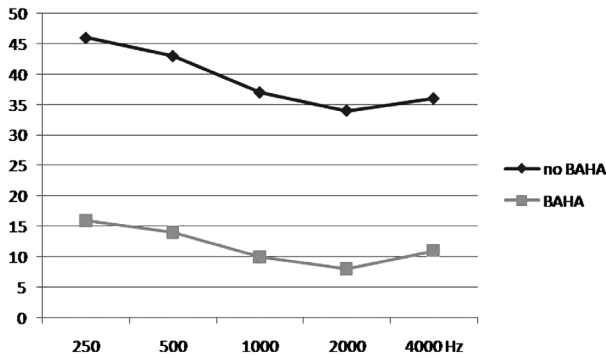


Figure 1a. Right atelectatic ear at the age 9.

large meatoplasty in the left ear precluded the utilization of an air conduction hearing aid and it was not well tolerated in the right ear in a trial of six weeks. He underwent a surgery for BAHA application in the left mastoid at 10 years, on a two-stage procedure with very good audiologic results (hearing thresholds of 15 dB on free-field audiometry with BAHA Divino) (Figure 1b) .

We have a follow-up period of 23 months without complications.

Case 2

A 2-year-old caucasian girl was first observed in our ENT Department with a history of hearing loss. Diagnosis of bilateral otitis media with effusion and episodes of suppurative otitis media was established.

As the pathology persists, at the age of 3.5 she had the first surgery - bilateral VT insertion. The VT extruded in less than 6 months and at the age of 5 she presented

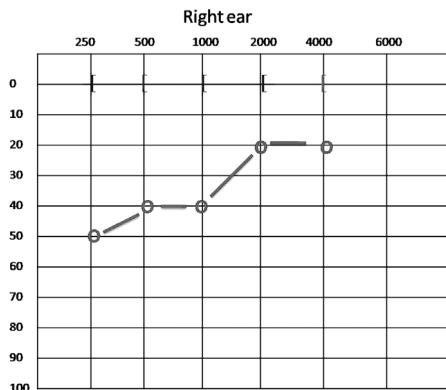


Figure 2a. Chronic suppurative otitis media in left ear.

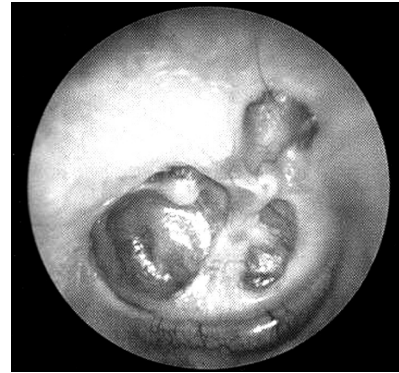


Figure 1b. Pure tone audiometry with bilateral conductive hearing loss and superimposed free field audiometry with BAHA.

with a large central perforation in left year and a very retracted drum in the right.

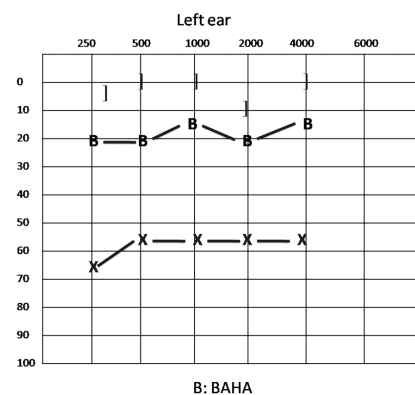
Conductive hearing loss subsided and at the age of 7 she underwent right ear VT insertion (grommets) and conventional hearing aid for the left ear. Caregivers refused reconstructive surgery on the right ear.

At the age of 9 a cholesteatoma developed in the right ear leading to canal wall down surgery and the repeated episodes of otorrhea in left ear precluded the use of the hearing aid.

A complication free cavity was achieved in the right ear and right ear surgery was further refused.

Figure 2a shows left central perforation and Figure 2b reveals pure tone audiometry at age of 9, after mastoid surgery .

She underwent a surgery for BAHA application at the age of 11, in the left side, on a two-stage procedure with good audiologic results (hearing thresholds of 20



dB on free-field audiometry with BAHA Divino) (Figure 2b). Right ear infections decreased in number and severity afterwards. The explanation for tympanoplasty refusal by the caregivers was that they did not want any more surgery on the ears, and BAHA despite the need for close care was a procedure that bypasses the ear itself.

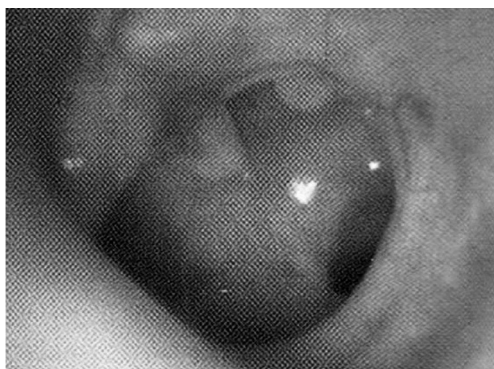


Figure 2b. Pure tone audiometry showing a severe bilateral conductive/mixed hearing loss and superimposed free field audiometry with BAHA.

Cases 3, 4, 5 and 6

Case 3 is now a 12-year-old Caucasian boy, with a long history of otologic procedures: grommets insertion at the age 2 and 4 who subsequently developed bilateral otorrhoea at six years old; at the age 10, he underwent a right tympanomastoidectomy with ipsilateral BAHA surgery and left side tympanoplasty at the age of 12.

Case 4 is a 13-year-old Caucasian boy who underwent grommets insertion at the age of 2 and 5; later on he developed a bilateral cholesteatoma and underwent a right modified radical mastoidectomy at the age 7 and subsequently combined approach tympanomastoidectomy and BAHA insertion on the left side.

Case 5 is a 15-year-old Caucasian girl with a history much similar to case 4, and BAHA surgery combined with ipsilateral tympanomastoidectomy at the age 13.

Case 6 is a 10-year-old Caucasian girl with three sets of ventilation tubes insertion from the age 3 to 7 who subsequently developed bilateral draining ears; her caregivers refused reconstructive surgery and she was fitted with the BAHA system.

Figure 3 reveals pure tone audiometry of cases 3, 4, 5 and 6.

In Table 1 we present free-field scores of the six patients with and without BAHA.

Discussion

With the advent of ventilation tube insertion, there were significant improvements in hearing. Early placement of ventilation tubes (VT) to correct the hearing impairment improve speech and language development and prevent long term complications which is advocated by many authors, some of them suggesting that VT should be routinely placed at the time of palatal repair^[4,5].

It is important to bear in mind the impact of hearing loss, even if transient, on the development of speech and language in children. Children who receive VT early have been shown to have better hearing thresholds, but its use fullness is questioned by several authors who point out the fact that patients receiving repeated sets of tubes were at great risk of persistent CHL and other sequelae. Also, the long term ventilation tubes have an incidence up to 30% of residual perforation^[1,2,6].

Lifelong otologic evaluation is recommended due to the 5.9% risk of cholesteatoma.

It is unclear that this higher incidence of long term sequelae such as CHL, chronic suppurative otitis media, atelectasia and cholesteatoma are related to the consequences of VT insertion per se or to the underlying inflammatory process itself.

Nevertheless, it would seem reasonable to try to manage OME in cleft palate children in a way as low risk as possible. Ventilation tubes should only be inserted when there is evidence of hearing loss and persistent effusion for more than 6 months^[7]. Non-surgical alternatives should be counselled with the child's parents; nowadays however, VT insertion in children with cleft palate is still a controversial issue^[8]. These alternatives are conventional hearing aids and in selected cases, bone anchored hearing aids, as in our cases^[9-11], and detailed information must be given to

the caregivers during the discussion of the therapeutic options. This conservative approach to the management of CHL associated with OME in cleft palate children seems to have no deleterious long-term consequences.

Conclusion

Middle ear disease is common in children with cleft palate, and, unlike the case for children without clefts, has a prolonged recovery, and a substantial incidence of late sequelae.

In cleft palate children we must put bone anchored hearing aids option on schedule, independent of other surgical interventions on the ears.

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