Tympanic floor reconstruction for conductive hearing loss due to a dehiscent high jugular bulb in the only hearing ear

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\section*{Abstract}
A 10-year-old girl with left-sided congenital deafness, who was treated for recurrent otitis media effusion, presented with conductive hearing loss in her only hearing ear. Otoscopy showed a blue mass in the tympanic cavity, and a dehiscent high jugular bulb (DHJB) was diagnosed. Computed tomography showed that the jugular bulb (JB) was located above the inferior wall of the tympanic cavity and was in contact with the tympanic membrane and obstructing the round window niche. The patient underwent surgical fixation of the DHJB and reconstruction of the tympanic floor with a tragus cartilage autograft. Two years after the operation, the JB was still situated in an appropriate location, and the patient's hearing had improved. Observation is one of the management options for DHJB. However, surgical treatment should be considered for DHJB, even if the patient only has one hearing ear.

\section*{Introduction}
The jugular bulb (JB) is a dilated region of the jugular vein, which is normally located below the inferior tympanic cavity. A high jugular bulb (HJB) is a vascular anomaly within the temporal bone, in which the jugular fossa is located higher than usual and protrudes into the tympanic cavity or pyramidal region. A dehiscent HJB (DHJB) refers to a defect in the bony wall covering the JB, resulting in a portion of the JB protruding into the tympanic cavity. The frequency of DHJB increases with age into adulthood and then stabilizes \cite{1–3}. DHJB is often asymptomatic and can be detected incidentally by imaging examinations. Still, in some cases, they are known to cause various symptoms, such as tinnitus, conductive hearing loss, and sensorineural hearing loss. Tinnitus and sensorineural hearing loss are relatively rare symptoms of HJB. Conductive hearing loss occurs through contact with the tympanic membrane, interference with the ossicles, or the blocking of the round window niche \cite{4}.

Various surgical treatments for DHJB, such as compression of the JB with autologous tissue or hydroxyapatite cement or endovascular treatment, have been reported \cite{5–9}. We report the case of a patient who presented with conductive hearing loss due to a DHJB in her only hearing ear. In this case, rigid reconstruction of the tympanic floor with autologous tissue improved the patient’s hearing.

\section*{Case presentation}
a 10-year-old girl with left-sided congenital deafness had been treated for recurrent otitis media effusion (OME) for several years at a local otolaryngology clinic. At that time, hearing in the right ear was normal. She was referred to the Department of Otolaryngology at a general hospital because she continued to experience hearing loss on the right side and pulsatile tinnitus even after she underwent treatment for the OME.

An otoscopic examination revealed a pulsatile dark-red mass on the tympanic membrane in the right tympanic cavity (Figure 1(A)). Computed tomography (CT) showed a JB that protruded into the tympanic cavity and was in contact with the tympanic membrane (Figure 2(A,B)). Although the JB protruded into the tympanic cavity, the bone of the tympanic floor had disappeared. Pure-tone audiometry...
revealed an A-B gap hearing loss of 30 dB in the low-frequency range on the right side (Figure 3(A)). Tympanometry showed a type A tympanogram. These findings were consistent with conductive hearing loss caused by a DHJB.

The sudden appearance of the DHJB, which had not previously been detected, was considered to indicate that the DHJB was progressing. Based on a fear that bilateral hearing loss would develop if the DHJB-induced conductive hearing loss on the better hearing ear worsened, we decided to perform surgical treatment to prevent the advancement of the DHJB and improve the patient’s hearing. To obtain an adequate view of the hypotympanum, we chose a postauricular incision.

After general anesthesia was induced, a small postauricular incision was made, and then a tympanomeatal flap was raised under microscopy. We opted for microsurgery because it was necessary to use both hands to fix the cartilage. The DHJB protruded into the tympanic cavity through the bony defect, was in contact with the tympanic membrane, and had blocked the round window niche (Figure 4(A)). It was not in contact with the ossicular chain. We compressed the DHJB with tragus cartilage, pushed it out of the tympanic cavity, and closed the bone defect. The cartilage was fixed with fibrin glue while holding it down using a suction tube. After confirming that the cartilage was no longer being pushed back, we filled hypotympanum with gelfoam. As a result, the round window could be seen (Figure 4(B)).

After the operation, there were no symptoms of venous reflux disorder, such as edema or visual field disturbance, and the wound was stable. One year after the surgery, an audiological examination showed that the air-bone conduction difference had almost unchanged (Figure 3(B)). The tympanic membrane showed that the DHJB had not recurred, although OME was still noted. Since there was no risk of intratympanic hemorrhaging due to pressure changes, we recommended self-ventilation (Otovent®) at home. Two years after the surgery, a hearing test showed that the air-bone gap had disappeared, and the improvements in the patient’s hearing had been maintained (Figures 1(B) and 3(C)).

**Discussion**

HJB is defined as when the top of a JB is located beyond the lower edge of the tympanic ring. A DHJB involves the protrusion of a JB into the tympanic sinus due to a defect in the bone of the tympanic floor covering the JB [1,2,10–13]. Since HJB are not present at birth, they are considered to be caused by hemodynamic changes during growth and development.

In this case, we found a DHJB in the patient’s only hearing ear, resulting in conductive hearing loss. Rigid reconstruction of the tympanic floor with a cartilage graft successfully repaired the DHJB and prevented its progression. Growth and negative pressure due to OME may have contributed to the development of the DHJB in this case.

Regarding the management strategy for DHJB, observation is often chosen, except when tinnitus or conductive hearing loss needs to be ameliorated. The mechanisms responsible for tinnitus and conductive hearing loss involve contact with the eardrum, interference with the ossicles, and/or blocking of the round window [4]. The goal of surgical or interventional treatment is to eliminate these factors.
As for the treatments reported for DHJB, pulsatile tinnitus was ameliorated in 5/7 cases using autologous tissue [5]. There have also been case reports about the amelioration of tinnitus using hydroxyapatite as a reconstructive material (in 3/3 patients) and the improvement of conductive hearing loss using Gelfoam® [6,7]. In addition, compression of the sigmoid sinus to change the patient’s hemodynamic status ameliorated hearing loss in 1/2 of patients [14,15]. In other case reports, the endovascular treatment of tinnitus due to DHJB using stents or coils has been reported, both of which resulted in success [8,9].

In the current case, the contralateral ear was deaf, and we decided to perform surgery to protect the hearing ability of the other ear. Since the risk of coils falling out due to growth and body movement was unacceptable, we did not perform a radiological intervention. Instead, we reconstructed the ear with autologous cartilage graft. In this procedure, there is a risk of intra-operative bleeding from JB because the dura overlying the JB is often attenuated. Although such profusion could be managed to some extent by compression, the risk of impaired hearing, especially when performed in the only hearing ear, should also be fully considered. However, even after taking such risks into consideration, treating the DHJB helped to stabilize the patient’s hearing. In addition, it allowed adequate treatment for the OME because performing self-ventilation in the presence of a DHJB can cause bleeding in the tympanic cavity due to pressure changes.

Conclusion

We reported a rare case of a child with a DHJB in her only hearing ear, which may have progressed with age and been caused by OME. In such cases, surgical treatment should be considered if the hearing loss significantly reduces the patient’s quality of life. Rigid reconstruction of the tympanic floor with tragus cartilage is a robust method, and the tympanic floor did not collapse during tympanic self-ventilation therapy.

Informed consent statement

This manuscript was written in accordance with the Code of the Ethics of the World Medical Association, Declaration of Helsinki. Written informed consent was obtained from the patient’s parents for the publication of this case report and any accompanying images.

Disclosure statement

No potential conflict of interest was reported by the author(s).
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