Utility of vestibular testing and new technologies in a complex cholesteatoma

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ABSTRACT
This paper reports a patient with a large recidivist cholesteatoma who underwent audio-vestibular tests and used customized 3D technologies (3D printing, augmented reality, virtual reality) to understand risks of the surgery. The patient was extremely concerned with her clinical findings and it found difficult to understand them. Customized 3D models helped the patient to understand the spatial relations and possible complications of surgery. The benefits of using new technologies in preoperative surgical planning for the surgeon and patient are also explained in a setting when radiological findings indicate high risks for surgery. Computed tomography scan showed a posterior semicircular canal fistula, which would add a significant challenge to the surgery. The fistula was not found in the result of the physiological test (cervical vestibular evoked myogenic potentials) and not found intra-operatively. Application of modern audio-vestibular investigations and use of customized 3D technologies may prove useful aids.

Abbreviations: AR: Augmented Reality; CT: Computed Tomography; DICOM: Digital Imaging and Communication in Medicine; ENT: Ear, Nose and Throat; JPEG: Joint Photographic Experts Group; STL: Stereolithography; VEMPs: Vestibular Evoked Myogenic Potentials; vHIT: video-Head Impulse Test; VR: Virtual Reality; WBB: Wii Balance Board

1. Introduction
Pediatric cholesteatoma is known to be more aggressive than cholesteatoma acquired later in age [1]. The incidence of recidivist disease in pediatric cholesteatoma is much higher than in adult patients [2]. Moreover, the incidence of cholesteatoma in children with cleft palate is reported higher (1.8–9.2%) than those without (0.3–0.6%) [3,4]. This is attributed to poor Eustachian tube function due to poor development and insertion of the tensor veli palatini muscle [5,6]. Kopcsányi et al. [1] reported recidivist disease in 40% of the pediatric cleft palate patients but no significant difference compared to recidivist rate of non-cleft palate children undergoing a second look surgery.

Though follow-up is routine for many years for pediatric patients and includes a variety of clinical, audiological and radiological techniques (such as diffusion weighted magnetic resonance imaging), it is extremely rare to remain asymptomatic for many years and then present with advanced disease [7]. When this occurs, it could be anticipated that the patient may experience a state of shock or disbelief and then sadness and frustration due to the recurrence of a chronic problem, especially if surgery risks further damage to hearing, balance, facial and vestibular nerve function. When faced with such challenges, application of modern audio-vestibular investigations and use of customized 3D technologies (such as 3D printing, augmented reality [AR] and virtual reality [VR]) may prove useful aids in communicating to the patient the likelihood of certain risks that radiological evaluation may allude to. We wish to explore the targeted use of such tools in a complex cholesteatoma surgery.

2. Case study
This paper reports a case of a 40 years old lady with a large recidivist cholesteatoma who was tested with a battery of vestibular tests and with new customized 3D technologies in order to understand her pathology and risks of the surgery. The benefits of the technology in preoperative surgical planning for the surgeon and patient are explained in a setting when radiological findings indicate high risks for surgery.
2.1. Past history

The patient’s background included a history of cleft palate surgery aged eight months, recurrent middle ear infections with grommets aged 3 and intact canal wall surgery and atticotomy surgery in left and right ear, respectively for cholesteatoma during her teenage years, elsewhere. She presented with moderate hearing loss requiring bilateral aiding since the age of 4. Her left ear has always been her better hearing ear.

2.2. Presenting symptoms

The patient presented with a history of feeling off-balance for a few seconds after a roller coaster ride and worsening pressure symptoms in both ears during flights a few months prior to presentation. Symptoms of imbalance were reportedly worse with pressure on her mastoid, though there was no history of sound or pressure induced vertigo, pain, discharge, deteriorating hearing, facial weakness or abnormalities of swallowing. Other symptoms included a metallic taste in her mouth and triggering nausea on pressure on her conchal bowl.

On examination, there were signs of previous surgery, with atticotomy defects and a thickened though intact tympanic membrane bilaterally. The bony part of the posterior ear canal wall in the left ear seemed eroded laterally near the bony-cartilaginous junction giving the appearance of a dry white pearl under the skin without impairing the contours of the canal. This appeared suspicious of a left cholesteatoma. Facial nerve and lower cranial nerves function was intact. Examination showed a negative fistula sign but she did experience discomfort and subjective imbalance on left mastoid palpation without nystagmus. Examination of her nose and throat revealed signs consistent with previous cleft palate surgery.

Computed tomography (CT) scan revealed a large soft tissue density (25 × 24 × 29 mm) in the left mastoid with extensive erosion of bone over the posterior ear canal wall, the posterior fossa dura, tegmen, the genu and the vertical facial nerve and suspicions of a fistula over the posterior semicircular canal (Figure 1). MRI scan showed no intracranial extension, normal fluid uptake in the otic capsule and significant compression of the sigmoid sinus by this mass with its epicenter in the mastoid cavity. Diffusion weight imaging (DWI) sequences suggested this mass was a cholesteatoma (Figure 1).

2.3. Audio-vestibular battery testing

Audio-vestibular testing included: pure tone audiogram (Figure 2(A)), speech audiometry (Figure 2(B)), cervical vestibular evoked myogenic potentials (cervical VEMPs, Figure 2(C)), video head impulse test (vHIT) of all six semicircular canals (Figure 2(D)),

![Figure 1. Pictures of the MRI (A–C) and of the CT scan (D and E) showing the extensive cholesteatoma (A, red circle) and the posterior semicircular canal fistula (B,C and D,E, red arrow). Noticed that C and E are enlargement from B and D, respectively.](image-url)
Figure 2. Results of the audio-vestibular battery testing including: pure tone audiometry (A), speech audiometry (B), cervical vestibular evoked myogenic potentials (cervical VEMPs, C), video-head impulse test (vHIT, D), and balance test (E). The hearing test (A) showed a hearing loss of 68.7 dB (PTA 0.5, 1, 2 and 4 kHz) on the right ear and of 55 dB on the left ear. The cervical VEMPs (C, using air-conducted sound stimulation short tone burst 500 Hz at 100 dB) showed normal responses from both ears. The vHIT (D) showed normal canal function on all six canals (gain values close to 1) with some overt catch up saccades for head impulses to the left horizontal side. The balance test (E) was abnormal: the patient fell on the foam with visual perturbation at amplitude 0.2.
and a balance test using VR (Figure 2(E)) on a Wii balance board (WBB) and on the WBB+foam. Caloric testing was not performed due to concern over causing complications with irrigation into an ear canal with cholesteatoma protruding through the posterior canal wall. VEMPs in response to air-conducted sounds did not provide a result due to an air bone gap and thus Fz tap evoked VEMPs were used.

Audiogram of air-conducted thresholds revealed a pure tone average (PTA 0.5, 1, 2 and 4kHz) of 68.7 dB and 55 dB in her right and left ears, respectively (Figure 2(A)). Thus, the affected ear was her better hearing ear and had been aided bilaterally for many years. Cervical VEMPs showed normal responses: peak to peak amplitude of 248 μV and 136.5 μV from the left and right ears, respectively (Figure 2(C)). This result indicated no physiological evidence of posterior canal fistula (as had been suggested by the CT scan and MRI). The vHIT showed normal canal function of all six canals with some catch up saccades for horizontal head impulses to the left side. This was not thought to be significant (Figure 2(D)). The patient fell on foam with a visual perturbation induced using VR at low amplitude of stimulation (VR 0.2, Figure 2(E)) which is abnormal for her age [8].

The diagnosis of large cholesteatoma was extremely upsetting given the fact that the ear was relatively trouble free compared to what the patient recalled from her childhood and that the treatment included surgery with significant risks to the damage hearing, balance and facial nerve function in addition to other serious risk of bleeding, CSF leak, infection, meningitis, and lower cranial nerve weakness. This was worsened as she found difficult to visualize the spatial relations and the related findings to the possible complications of surgery especially given this was her better hearing ear. To aid both the surgeon and the patient, three-dimensional (3D) surface models such as the cholesteatoma and its affected temporal bone and relevant structures were created from patient’s coronal high-resolution multi-slice CT scan using volume rendering module, and threshold range growing and model making technique within 3D Slicer version 4.6.0 and polygonal model editing module within Autodesk 3dsMax 2016 Student version (Autodesk Inc., Mill Valley, CA, USA) on a MSI GT80S 6QE TITAN SLI laptop. Supplementary labels and titles were also created and rendered in 3dsMax 2016 using polygonal modeling technique and mental ray rendering engine. A patient’s CT slice was chosen, and edited to create a 512 × 512 pixel square image at 72 dpi resolution, and was saved in the joint photographic experts group (JPEG) format in Adobe Creative Cloud Photoshop CC (2017 Adobe Systems Software Ireland Ltd., Dublin, Ireland). This image was also printed on paper for patient counseling. Both 3D and 2D assets were uploaded, linked in an AR development platform called Hyperspaces which was then published to the hyperspaces cloud server to build an AR application for free access using a predefined keyword on an iOS mobile device. Patient’s temporal bone in stereolithography (STL) format was 3D fabricated (Figure 3) using PolyLactic Acid (PLA) filament on a fused deposition modeling (FDM) 3D printer called Up Box (Beijing Tiertime Technology Co., Ltd., Beijing, China) connected to the MSI laptop via a USB AB cable. The advantage of the AR content is that the patient was able to view her disease through a free app (Hyperspaces Inglobe technologies Ltd., Latina, Italy) using a 2D picture or marker [9]. The value of the VR was that the surgeon was able to walk through the complex pathology and view the cholesteatoma and its relations from inside the ear, such as the middle ear.

**2.4. Surgery decisions and post op**

Left untreated, complications of the cholesteatoma included facial nerve palsy, thrombosis of her venous sinuses and its sequelae, compression of lower cranial nerves and the structures of the posterior and middle cranial fossa, meningitis, encephalitis, stroke and death. Unfortunately, with the left ear being her better hearing ear, the risks of surgery especially with a suspected fistula on radiological imaging also included partial or total hearing and vestibular loss. In addition, if CSF leak was encountered, blind sac closure would be required which would create a risk of recurrence and ongoing surgical treatment. Moreover, since blind sac closure would preclude air conduction aids, rehabilitation of hearing would be complicated with her current bone thresholds (PTA 41.3 dB in the left ear and 47.5 dB in the right ear) which would give suboptimal long-term hearing outcome with bone conduction aids. In the setting of a cholesteatoma, with ongoing MRI required for monitoring and infection risk, the use of a Cochlear Implant was felt to be contraindicated. Alternatively, an open exteriorized cavity came with its own challenges with the amount of exposed dura, facial nerve and if a fistula was indeed encountered, with planning for a meatoplasty needing to take into account hearing aid use. Thus, the surgical discussion was complex both from a surgical point of view as especially so from the point of view as especially so from the patient’s perspective and her hearing for her age [8].
view of the patient and the family. To assist in the discussion information from the VEMPS and vHITs and new technologies including a 3D print of her own temporal bone showing her the extent of the cholesteatoma (Figure 3), and AR showing the anatomy of the cholesteatoma and relevant relations to the facial nerve and middle ear and inner ear structures was found to be helpful (Figure 4).

Intraoperative findings were consistent with the imaging except no fistula was found on the posterior semicircular canal, though the bone was thinned. It was noted that the intra-operative findings of the posterior semicircular canal were consistent with the physiological tests of its function (cervical VEMP) rather than the radiological findings. As a result, a large modified radical mastoidectomy cavity was performed and the cholesteatoma was exteriorized. The cavity was kept open so the patient was able to continue to use her hearing aid in this ear. The histopathological report post-operated confirmed the cholesteatoma nature of the tissue. Postoperatively, the patient experienced minimal vestibular symptoms and intact facial nerve function and was discharged two days post operatively. Following recovery, she was also shown an interactive VR video of her removed cholesteatoma, which correlate her post operative management and ongoing management with relevance to the defect present. 3D VR visualization is available online: without the temporal bone (Figure 5(A), https://skfb.ly/VuLF) or with the temporal bone (Figure 5(B), https://skfb.ly/UAx6).

At three months post-surgery, the audio-vestibular test battery was repeated (hearing test, VEMPs, vHIT, and balance test). The results were unchanged: no deterioration of the hearing and vestibular function. Her balance was objectively improved (she felt on the foam with a VR visual perturbation at 0.3) as well as subjectively (with a dizziness handicap inventory’s score at 0).

3. Discussion

Technologies such as 3D printing, AR, and VR are well established and are readily available at economical costs for clinical practice. However, much of the use of these technologies has not been utilized or translated in the health industry which means its benefits to patients and the medical community are largely unexplored. The main limitation in the application of this technology is that surgeons (who are at the interface of solving the clinical problem) do not have easy access to
such technology due to lack of availability, high commercial cost and time delay caused in addressing the first two factors. Nevertheless, use of 3D printed temporal bones in surgical training and simulation has been reported [10].

The use of these tools in pre-surgical planning has also been reported by the authors [11]. Customized surgical instruments can be designed, developed and 3D printed for a particular patient using 3D software and also 3D printed. Given that the patient’s pathology was encased in an ear with post-operative changes and with bone eroded between major important structures, 3D printing alone (Figure 3) could not demonstrate the complex relational anatomy of the cholesteatoma to the major structures. For instance, since 3D printing did not print the soft tissue structures, the relationship between the cholesteatoma and the facial nerve could not be visualized. AR allowed the ability to place the relevant soft tissue structures with different color coding and the viewer was able to slice through the content of the temporal bone so spatial relationships to the cholesteatoma of the ossicles, otic capsule, facial nerve and sigmoid sinus could be visualized at different depths and unique to the patient’s disease. Thus, AR allowed a patient friendly tool which could be accessed anywhere at no cost on the patient’s own smart device. Together with the 3D printed bone, it allowed explanation of not only surgical access but also risks and relations of the structures.

The presence of these tools in every day practice enables better communication and planning between surgeon and patient with complex cases. Most importantly, in an age where medicine allows a diverse range of tools, it allows surgical care to be customized to the needs of the patient and the patient’s pathology. In this case, the patient and her family was an engaged and intelligent individual who had reasonable concerns and fears regarding her surgery. Being able to better inform the patient and her family with technology that is freely accessible with technologies that people carry on their person everyday (such as an iOS device), enables patients to be empowered and able to follow the clinician through the complexity of the decision making and then post-operative care. In addition, communication between colleagues and trainees is also aided. Instead of requiring to transport digital radiological files between different geographical locations, an image of the marker can be transferred online, or via message, enabling anyone with access to the 2D marker (whether it is in color, black and white, in print or in a digital format), to view the 3D AR content using Hyperspaces application on their smart device. Thus, information transfer and communication between medical personnel is immediate which is very useful in procedures where multiple teams may be involved. Expressing the digital content in VR enables the possibility of using such content in surgical training, as anatomical structures can be drawn into the file such as the relation of the facial nerve in relation to the cholesteatoma.

One major concern was that the cholesteatoma had caused a fistula of the posterior canal wall. Indeed, a semicircular canal fistula could be induced by extensive cholesteatoma [12,13]. However, the distinction between a fistula and thinning of the bone over the otic capsule could be difficult to detect on CT scan due to resolution even of modern imaging [14,15]. It does alert the surgeon to significant challenges for surgery. Moreover, the prevalence of a posterior semicircular canal fistula is low and the clinical symptoms associated are very wide [16,17]. The cervical vestibular evoked myogenic potentials (cervical VEMPs) record EMG on tensed neck muscles of the brief short

![Figure 5. Picture of the online interactive virtual reality (VR) tool of the cholesteatoma without the temporal bone (A) and with the temporal bone (B). These interactive virtual reality reconstructions of the lesion could be visualized online on a computer’s screen and in a virtual reality headset (cardboard) on a mobile phone.](https://skfb.ly/VuLF)
latency inhibitory potential to air conducted sound. In cases of posterior semicircular canal fistula, these potentials are enhanced due to hypersensitivity to sounds \[12,18,19\]. Zhou et al. \[20\] found that VEMPs resulted in 91.4% sensitivity and 95.8% specificity, assuming CT scan as a gold standard. But the best gold standard is the anatomical observation intraoperatively. Here, cervical VEMPs were normal giving good cues for the fact that the bony wall of the posterior canal was intact which was confirmed during the surgery. Preoperatively, cervical VEMPs provided reassurance in surgical decision making that although care should be taken to manage a fistula as alerted by the radiology, that it was possible that no fistula may be present. To our knowledge there are no reports of studies of cholesteatoma’s presenting with inner ear fistula correlating VEMPs, radiological findings and intraoperative confirmation. Thus, this case raises important insights into the value of functional vestibular testing, though larger sample is required for definitive conclusions.

### 4. Conclusions

This case illustrates the importance of not only of preoperative audiological but also functional vestibular testing in relation to complex cholesteatoma as an adjunct to radiological tests. Although radiological tests provide crucial information about diagnosis and anatomy in skull base cases, vestibular function tests provide physiological tests of function. Both in combination provide additional data that assists clinical decision-making. In addition, the evolution and ease of access to 3D technologies aid both patients and surgeons in the decision-making process. Advantages of these tools include rapid transfer of 3D information, free access which requires no additional infrastructure for the surgeon or the patient and ability to view it in a few different formats such as through a smart device using AR, or through VR goggles. Each tool has its strengths and limitations and its use can be tailored to the case and the particular pathology and challenges presented. Nevertheless, these tools are useful and cost effective, which is highly valuable in otological practice given the complex 3D relations that routinely challenge surgeons and patients in clinical practice.

### Disclosure statement

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### References

1. Kopcsányi G, Vincze O, Bagdán V, et al. Retrospective analysis of tympanoplasty in children with cleft palate: a 24-year experience. II. Cholesteatomatous cases. Int J Pediatr Otorhinolaryngol. 2015;79:698–706.
2. Lynrah ZA, Bakshi J, Panda NK, et al. Aggressiveness of pediatric cholesteatoma. Do we have an evidence?. Indian J Otolaryngol Head Neck Surg. 2013;65:264–268.
3. Goldman JL, Martinez SA, GANZEL TM. Eustachian tube dysfunction and its sequelae in patients with cleft palate. South Med J. 1993;86:1236–1237.
4. Vincenti V, Marra F, Bertoldi B, et al. Acquired middle ear cholesteatoma in children with cleft palate: experience from 18 surgical cases. Int J Pediatr Otorhinolaryngol. 2014;78:918–922.
5. Hartzell LD, Dornhoffer JL. Timing of tympanoplasty in children with chronic otitis media with effusion. Curr Opin Otolaryngol Head Neck Surg. 2010;18:550–553.
6. Meyerhoff WL, Shea DA, Foster CA. Otitis media, cleft palate, and middle ear ventilation. Otolaryngol Head Neck Surg. 1981;89:288–293.
7. Mukherjee P, Saunders N, Liu R, et al. Long-term outcome of modified radical mastoidectomy. J Laryngol Otol. 2004;118:612–616.
8. Chiarovano E, Wang W, Rogers SJ, et al. Balance in virtual reality: effect of age and bilateral vestibular loss. Front Neurol. 2017;8:5.
9. Cheng K, Mukherjee P, Curthoys I. Development and Use of Augmented Reality and 3D Printing in Consulting Patient with Complex Skull Base Cholesteatoma. Virtual and Physical Prototyping. 2017;12:241–248.
10. Rose AS, Kimbell JS, Webster CE, et al. Multi-material 3D models for temporal bone surgical simulation. Ann Otol Rhinol Laryngol. 2015;124:528–536.
11. Mukherjee P, Cheng K, Flanagan S, Greenberg S, et al. Utility of 3D printed temporal bones in pre-surgical planning for complex Bonebridge cases. Eur Arch Otorhinolaryngol. 2017;274:3021.
12. Brantberg K, Bagger-Sjöbäck D, Mathiesen T, et al. Posterior canal dehiscence syndrome caused by an apex cholesteatoma. Otol Neurotol. 2006;27:531–534.
13. Vashishth A, Singh Nagar TR, Mandal S, et al. Extensive intratemporal cholesteatomas: presentation, complications and surgical outcomes. Eur Arch Otorhinolaryngol. 2015;272:289–295.
14. Belden C, Weg N, Minor LB, et al. CT evaluation of bone dehiscence of the superior semicircular canal as a cause of sound- and/or pressure-induced vertigo. Radiology 2003;226:337–343.
15. Chien WW, Carey JP, Minor LB. Canal dehiscence. Curr Opin Neurol. 2011;24:25–31.
[16] Russo JE, Crowson MG, DeAngelo EJ, et al. Posterior semicircular canal dehiscence: CT prevalence and clinical symptoms. Otol Neurotol. 2014;35:310–314.

[17] Krombach GA, DiMartino E, Schmitz-Rode T, et al. Posterior semicircular canal dehiscence: a morphologic cause of vertigo similar to superior semicircular canal dehiscence. Eur Radiol 2003;13:1444–1450.

[18] Friedmann DR, Le BT, Pramanik BK, et al. Clinical spectrum of patients with erosion of the inner ear by jugular bulb abnormalities. Laryngoscope. 2010;120:365–372.

[19] Gopen Q, Zhou G, Poe D, et al. Posterior semicircular canal dehiscence: first reported case series. Otol Neurotol. 2010;31:339–344.

[20] Zhou G, Gopen Q, Poe DS. Clinical and diagnostic characterization of canal dehiscence syndrome: a great otologic mimicker. Otol Neurotol. 2007;28:920–926.