

been successful in head and neck oncology. The study by Lee et al¹ demonstrates that the other subspecialties are also ready to learn the patient perspective. One important lesson I have learned from my work with patients is that it is impossible to predict where engaging patients will lead, but it is imperative to have an open mind and be ready to listen. In head and neck surgery, engagement has led to patient-reported outcome-based survivorship research. In other subspecialties, the outcomes and methods will be different. As discussed, there are many methods to engage patients, including surveys and focus groups. Furthermore, there is considerable funding available to support this type of work. In addition, patient engagement does not need be limited to academic practices. Many private practice groups leverage patient engagement to improve the patient experience at their practice.

Treating patients as partners and understanding the patient perspective is a remarkable resource for quality improvement and research. The field of patient-centered research is an important complement to other traditional approaches to improving patient care and new discoveries.

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1. Lee K, Vijayendra VK, Bedri E-h, Honnurappa V, Redleaf M. Concerns of patients with Ménière disease in Ethiopia, India, and the United States. *JAMA Otolaryngology*. Published online April 9, 2020. doi:10.1001/jamaoto.2019.2663
2. Kitahara T, Okamoto H, Fukushima M, et al. A two-year randomized trial of interventions to decrease stress hormone vasopressin production in patients with Meniere's disease—a pilot study. *PLoS One*. 2016;11(6):e0158309. doi:10.1371/journal.pone.0158309
3. Movsas B, Hu C, Sloan J, et al. Quality of life analysis of a radiation dose-escalation study of patients with non-small-cell lung cancer: a secondary analysis of the Radiation Therapy Oncology Group O617 randomized clinical trial. *JAMA Oncol*. 2016;2(3):359-367. doi:10.1001/jamaoncol.2015.3969
4. Movsas B, Moughan J, Sarna L, et al. Quality of life supersedes the classic prognosticators for long-term survival in locally advanced non-small-cell lung cancer: an analysis of RTOG 9801. *J Clin Oncol*. 2009;27(34):5816-5822. doi:10.1200/JCO.2009.23.7420
5. Iwata AJ, Olden HA, Kippen KE, Swegal WC, Johnson CC, Chang SS. Flexible model for patient engagement: achieving quality outcomes and building a research agenda for head and neck cancer. *Head Neck*. 2019;41(4):1087-1093. doi:10.1002/hed.25584
6. Pcobelli G, Ziebell R, Fujii M, et al. Symptom burden in long-term survivors of head and neck cancer: patient-reported versus clinical data. *EGEMS (Wash DC)*. 2019;7(1):25. doi:10.5334/egems.271

OBSERVATION

Long-term Follow-up of a Patient With Auditory Neuropathy and Normal Hearing Thresholds

Few data are available regarding long-term outcomes in patients with auditory neuropathy (AN), absent auditory brainstem responses (ABRs) despite normal cochlear function.^{1,2} In

1993 we described an 18-year-old woman with AN and normal hearing thresholds, normal otoacoustic emission test results, and absent ABRs.³

We retested her at age 24 years. Audiological results were identical (**Figure 1**).⁴ Background noise severely impaired word recognition.

Herein we report results when the patient was aged 41 years. The patient provided written consent, and the Northwestern University institutional review board approved study procedures. The patient has pursued a successful career and raised a family despite continued difficulties hearing in noise. She struggles to understand unfamiliar accents; yet, she is English-Hebrew bilingual and understands Israeli accents well. She described inconsistent sound awareness, particularly for alarms, such as phones and doorbells.

The patient's air-conduction thresholds had increased in the interim (approximately +10 dB HL from 0.5-4 kHz) and were consistent with a mild high-frequency hearing loss, although still relatively normal (**Figure 1**). Distortion product otoacoustic emissions (DPOAEs) were robust bilaterally, and were approximately 12 dB signal-to-noise ratio (SNR) on average (compared with approximately 20 dB previously⁴); ABRs remained absent bilaterally.

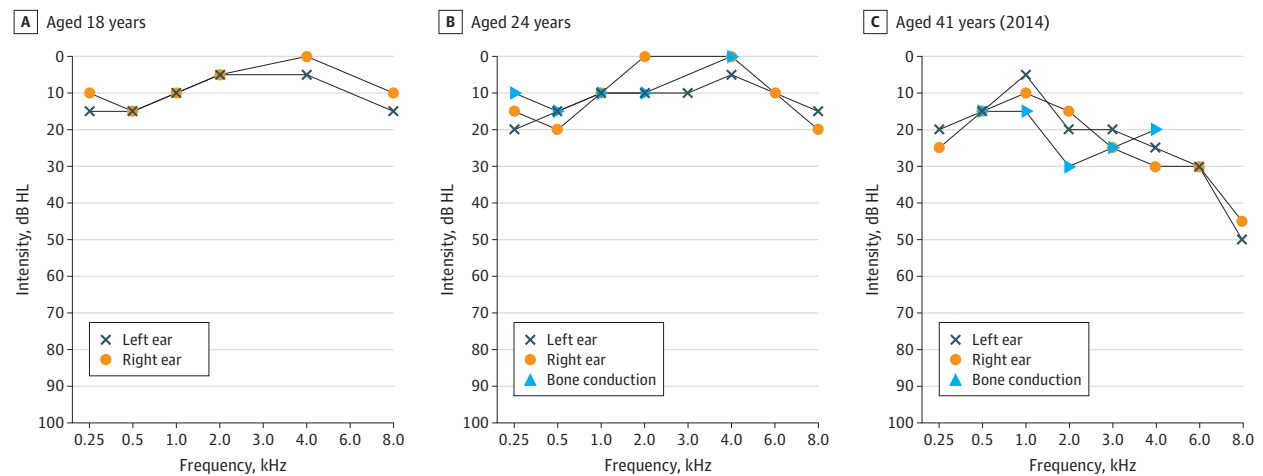
Sentence perception in sound field was measured with the Hearing in Noise Test (HINT). In quiet, the patient did not notice sentences until 39.1 dB SPL (<1st percentile) but, once she heard the sentences, she understood 100% of the words. The patient's speech reception threshold (SRT) in noise was +2 dB signal-to-noise ratio (SNR, <1st percentile). When the speech and noise sources were separated by 180°, her SRT improved to -2.9 dB SNR (noise right/speech left) and -1.2 dB SNR (noise left/speech right), indicating a spatial release from masking of 3.2 to 4.9 dB. Rance et al⁵ reported a spatial release of 7.6 dB (range, 0-13 dB) in patients with AN.

The HINT was conducted under headphones with speech and noise colocated (SRT, +4.5 dB SNR; <1st percentile). Performance was worsened by applying amplification algorithms to the speech signal to improve audibility (National Acoustics Lab-Revised +7.2 dB SNR; House Ear Amplification Routine: +6.2 dB SNR; both <1st percentile). The QuickSIN measured sentence recognition under headphones. The patient's average SNR loss was 10 dB and 13.5 dB in the right and left ears, respectively. Diotically, the patient scored 1.5 dB on the first run (within normal range) and 7.5 dB on the second (**Figure 2**).

Discussion | We know of 1 other long-term follow-up in a patient with AN, who as a young adult who had normal thresholds but 22 years later developed moderate-to-severe hearing loss.⁶ Although the case presented herein is subtler, elevated hearing thresholds are consistent. Paradoxically, the patient's DPOAEs were present in this frequency range. She noted a buzzing sound when certain audiogram tones were presented, which may have caused masking. Poor awareness of quiet sounds may affect her performance. Still, DPOAEs were lower than in our previous report.⁴

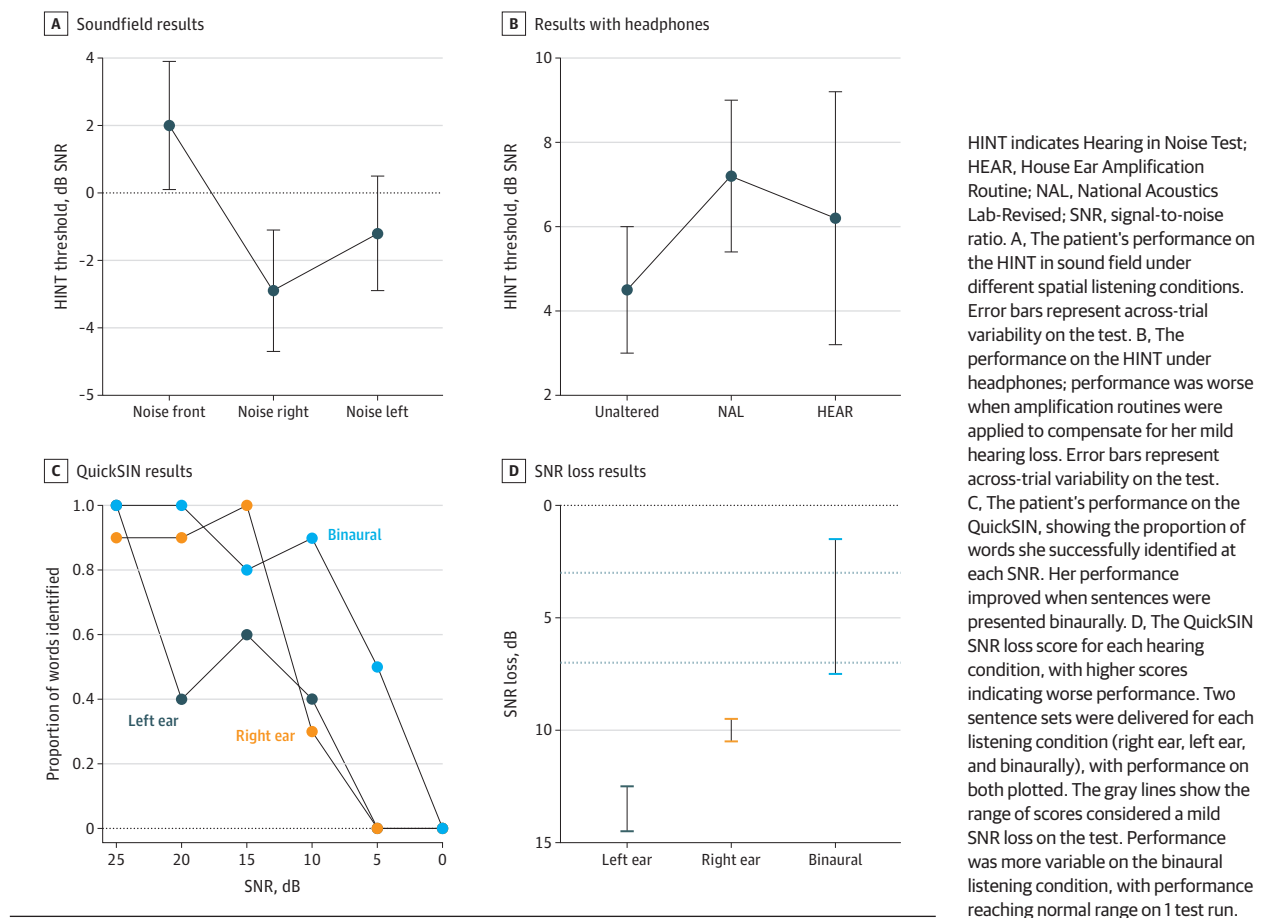
This patient's speech-in-noise perception improved when she listened with 2 ears, likely owing to interaural level

Figure 1. Changes in Patient Hearing Over a 23-Year Period



Patient audiograms at ages 18, 24, and 41 years. A mild high-frequency hearing loss emerged at age 41 years. Air-conduction thresholds are shown (right ear in orange, left ear in gray) and bone conduction (blue triangles).

Figure 2. Patient Speech Perception at Age 41 Years



differences (better ear effect). Importantly, her performance worsened under the HINT amplification condition.

This follow-up motivates new hypotheses about AN and its treatment:

1. A lack of synchrony prevents triggering protective mechanisms in noise (middle ear and medial olivocochlear reflexes), accounting for elevated hearing thresholds and decreased DPOAEs, indicating accelerated onset of

HINT indicates Hearing in Noise Test; HEAR, House Ear Amplification Routine; NAL, National Acoustics Lab-Revised; SNR, signal-to-noise ratio. A, The patient's performance on the HINT in sound field under different spatial listening conditions. Error bars represent across-trial variability on the test. B, The performance on the HINT under headphones; performance was worse when amplification routines were applied to compensate for her mild hearing loss. Error bars represent across-trial variability on the test. C, The patient's performance on the QuickSIN, showing the proportion of words she successfully identified at each SNR. Her performance improved when sentences were presented binaurally. D, The QuickSIN SNR loss score for each hearing condition, with higher scores indicating worse performance. Two sentence sets were delivered for each listening condition (right ear, left ear, and binaurally), with performance on both plotted. The gray lines show the range of scores considered a mild SNR loss on the test. Performance was more variable on the binaural listening condition, with performance reaching normal range on 1 test run.

age-related hearing loss. Alternatively, quiet sounds are easily masked and difficult to detect, accounting for the discrepancy between audiometry and DPOAEs.

2. Subtle, albeit diminished, binaural cues improve perception in noise. Screening for residual binaural sensitivity might be important when evaluating a patient with AN's candidacy for hearing aids and/or cochlear implants.
3. The patient's ability to learn a new accent suggests auditory processing is amenable to training in patients with AN.

To the extent this patient's case generalizes, her follow-up illuminates the possibilities and persistent challenges endemic to AN.

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1. Kraus N, Ozdamar O, Stein L, Reed N. Absent auditory brain stem response: peripheral hearing loss or brain stem dysfunction? *Laryngoscope*. 1984;94(3):400-406. doi:10.1288/00005537-198403000-00019
2. Starr A, Picton TW, Sininger Y, Hood LJ, Berlin CI. Auditory neuropathy. *Brain*. 1996;119(Pt 3):741-753. doi:10.1093/brain/119.3.741
3. Kraus N, McGee T, Ferre J, et al. Mismatch negativity in the neurophysiologic/behavioral evaluation of auditory processing deficits: a case study. *Ear Hear*. 1993;14(4):223-234. doi:10.1097/00003446-199308000-00001
4. Kraus N, Bradlow AR, Cheatham MA, et al. Consequences of neural asynchrony: a case of auditory neuropathy. *J Assoc Res Otolaryngol*. 2000;1(1):33-45. doi:10.1007/s101620010004
5. Rance G, Ryan MM, Carew P, et al. Binaural speech processing in individuals with auditory neuropathy. *Neuroscience*. 2012;226:227-235. doi:10.1016/j.neuroscience.2012.08.054
6. Berlin CI, Hood LJ, Morlet T, et al. Multi-site diagnosis and management of 260 patients with auditory neuropathy/dys-synchrony (auditory neuropathy spectrum disorder). *Int J Audiol*. 2010;49(1):30-43. doi:10.3109/14992020903160892

An Atypical Cause of Difficulty Swallowing

Schwannomas are benign nerve sheath cell tumors. Whereas 25% to 45% of schwannomas occur in the head and neck region, laryngeal schwannomas are rare.¹ Representing 0.1% of all benign laryngeal tumors, schwannomas are most commonly found in the aryepiglottic folds and arytenoids, with most arising from the superior laryngeal nerve.²⁻⁴ In 1 series³

of 55 laryngeal schwannomas, only 2 were reported in the pyriform sinus. We present here the third reported case of pyriform schwannoma, to our knowledge.

Report of a Case | A woman of Indian descent in her 70s was referred with a 10-month history of globus pharyngeus, intermittent dysphagia, odynophagia, and unintentional weight loss. The remaining medical history was unremarkable.

Flexible laryngoscopy and flexible endoscopic evaluation of swallow were unremarkable preoperatively. Computed tomographic findings demonstrated a 2×2-cm hypodense mass in the cervical esophagus, with its superior aspect approximating the inferior border of the cricoid cartilage (Figure 1A). Additional radiographic characteristics included soft tissue fullness at the left pyriform sinus, anterior displacement of the membranous trachea, and scalloped contour of the anterior C7 vertebral body.

Operative direct microlaryngoscopy and rigid esophagoscopy was performed. A Dedo laryngoscope was suspended in the postcricoid space. Only a subtle stalk was visualized extending into the esophageal inlet (Figure 2A). A right-angle laryngeal probe was passed distal to the mass and used to deliver a pendulous mass into the hypopharynx. The mass was firm, 3 cm in cranio-caudal dimension, and was encapsulated with mucosal tissue. It was transected at its pedicle on the medial surface of the left pyriform sinus with an AccuBlade CO₂ laser (Figure 2B). Histopathologic examination revealed a schwannoma positive for S-100, negative for desmin, with Ki67 proliferation index of 1% and negative margins (Figure 2C and D). One week postoperatively, she was tolerating a mechanical soft diet and had mild odynophagia. Examination findings showed a healing left pyriform eschar and normal vocal fold movement. At 3 months, she had complete resolution of symptoms and a normal examination.

Discussion | Patients with laryngeal schwannoma most commonly present with dysphonia.³ The true vocal fold ipsilateral to the lesion is often immobile or hypomobile, often secondary to nerve compression.^{2,3} Other mass effect-related symptoms of laryngeal schwannoma include dysphagia, globus pharyngeus, and stridor.^{1,2} One case of asphyxial death has been reported.⁴

The rare nature of laryngeal schwannoma makes diagnosis challenging. These nerve sheath tumors can arise from the pharyngeal plexus, the internal branch of the superior laryngeal nerve, or branches of the recurrent laryngeal nerve, all of which innervate the mucosal and submucosal layers of the pyriform sinus.⁵ On flexible laryngoscopy, these tumors appear as round submucosal tissue fullness.³ Cystic change has been described³ and was observed in this case. Imaging may be used to establish mass extent and differentiate benign from malignant tumors, but these modalities are not always able to differentiate schwannomas from other benign laryngeal tumors.³ Definitive histopathologic diagnosis is based on the presence of a clear capsule, Antoni A and Antoni B regions, and S-100 positivity.^{2,3}