been successful in head and neck oncology. The study by Lee et al 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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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. In 1993 we described an 18-year-old woman with AN and normal hearing thresholds, normal otoacoustic emission test results, and absent ABRs.

We retested her at age 24 years. Audiological results were identical (Figure 1). 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 previously); 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 al 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. Dially, 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. 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.

This patient’s speech-in-noise perception improved when she listened with 2 ears, likely owing to interaural level
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
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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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 artenoids, with most arising from the superior laryngeal nerve.2-4 In 1 series 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 described4 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