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Case Report

# Endoscopic Repair of Traumatic Perilymphatic Fistula in Children: A Case Series

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Traumatic perilymphatic fistula (PLF) is an uncommon cause of acute vestibular symptoms and hearing loss following head injury in children. We describe the management of 3 pediatric patients with traumatic PLF using an endoscopic ear surgery (EES) approach. Three pediatric patients with traumatic PLF underwent repair via an EES approach between August and October 2018. Patients included a 14-year-old female (oval window), a 13-year-old male (round window), and a 10-month-old male (oval and round window). Ossicular chain injury was identified and repaired in 2 patients. The 10-month-old patient required a second-stage surgery that included lumbar drain placement and a post-auricular, endoscopic-assisted approach due to an especially brisk leak. All patients had complete resolution of vestibular symptoms post-operatively with no recurrence at a mean follow-up of 8.3 months. Traumatic PLF can be safely and effectively diagnosed and managed via an EES approach in children, though an endoscopic-assisted approach may be necessary in select cases due to factors such as patient age and leak severity.

**KEYWORDS:** Perilymphatic fistula, endoscopic ear surgery, vestibular, pediatric

## INTRODUCTION

Perilymphatic fistula (PLF) is the egress of perilymph from a defect in the otic capsule presenting with hearing loss and/or vestibular symptoms that are induced by blunt/penetrating trauma, barotrauma, chronic inflammation, cholesteatoma, or iatrogenic injury.[1,2] Diagnostic testing is often normal, and definitive diagnosis requires surgical middle ear exploration.[3] Conservative management is appropriate in some cases, as patients may have spontaneous resolution of symptoms.[1] However, the vestibular symptoms can often be debilitating and there is a risk of permanent sensorineural hearing loss (SNHL) with a persistent, unrepaired fistula, so a minimally invasive approach to early surgical confirmation and repair of PLF is appealing.[2]

The use of endoscopes to diagnose and treat PLF has been described in a handful of case reports, both as an adjunct and as the sole technique.[3-6] This approach offers the potential for improved visualization, shorter hospital stays, reduced post-operative analgesia, and avoidance of an external incision.[5-7] We describe our experience with the management of 3 pediatric patients with traumatic PLF using an endoscopic ear surgery (EES) approach. Key features of the 3 cases are summarized in Figure 1.

## CASE 1

A 14-year-old female had sustained a head injury 15 months prior to presentation to our pediatric vestibular program. She had persistent imbalance, episodic vertigo, and a right-sided hearing loss. She also had daily episodes of tinnitus, aural fullness, autophony, and unilateral hyperacusis that were provoked by motion or loud noises.

Her neurotologic examination was normal. Her audiogram showed a unilateral mild-moderate mixed hearing loss, tympanic membrane hypermobility, and absent acoustic reflexes. A vestibular test battery was normal. A computed tomography (CT) of the temporal bone was interpreted as normal.

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| Patient | Age (y) | Gender | Side | Time to repair (m) | Location of PLF | Ossicular damage | TM Perf | Surgical approach | Graft material | Need for LD | FU (m) | Post-op Audio | Post-op Vestibular Sx | Recurrent PLF |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| ES | 14 | F | R | 15 | OW | Long process incus | N | TEES | TP | N | 3 | Worse mixed | Resolved | N |
| JT | 13 | M | L | 0 | RW | N | Y | TEES | TP | N | 4 | Severe-profound mixed in high f | Resolved | N |
| IS | 1.2 | M | R | 0 | OW, RW, prom | SF fx, AC avulsed | Y | TEES, then endoscopic-assisted | TF | Y | 4 | Mod-severe mixed | NA | N |

y = years, m = months, Sx = symptoms, CT = computed tomography, PLF = perilymphatic fistula, Perf = perforation, LD = lumbar drain, TM = tympanic membrane, FU = follow-up, fx = fracture, TP = tragal perichondrium, f = frequency perichondrium, TF = temporalis fascia, mod = moderate, OW = oval window, RW = round window, prom = promontory, SF = stapes footplate, AC = anterior crus, F = female, M = male, R = right, L = left, Y = yes, N = no, NA = not applicable (secondary to age)

**Figure 1.** Summary of key features of cases.

The patient underwent middle ear exploration via a total EES approach. Intraoperative findings included an incus long process fracture and displaced anterior footplate into the proximal vestibule with a perilymphatic leak from the anterior oval window (Figure 2). The stapes were rotated back into anatomical position, the leak was sealed with tragal perichondrium, and the incudostapedial joint was repaired with hydroxyapatite bone cement.

Post-operatively, the patient's dizziness fully resolved within a few days without recurrence. A repeat audiogram showed stable bone thresholds with improved air conduction thresholds in the low frequencies.

## CASE 2

A 13-year old male presented to the emergency room with vertigo, nausea, vomiting, and blood from his left ear after a helmeted bicycle crash. Examination showed a small external auditory canal laceration and right-upbeating torsional nystagmus. Pneumolabyrinth was noted in the left vestibule on the CT (Figure 3). An audiogram showed moderate sloping to profound mixed hearing loss in the left ear.

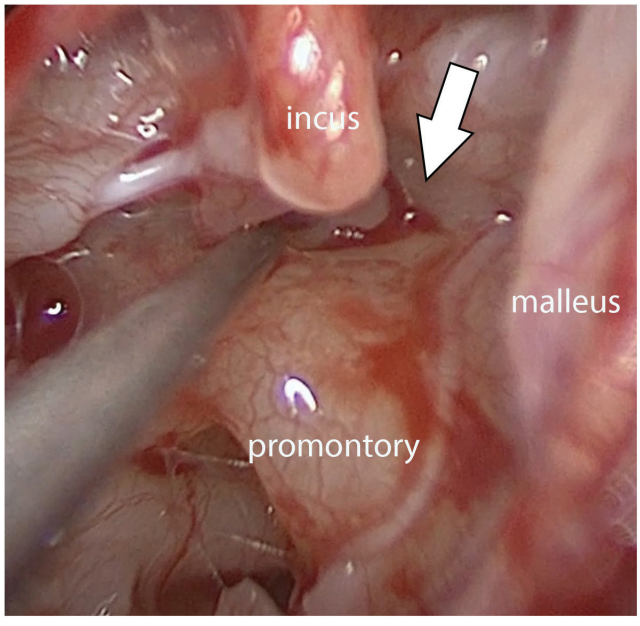


**Figure 2.** Case 1: Perilymphatic leak from the anterior oval window (arrow) adjacent to the fractured stapes footplate. Image is of the right ear (right is anterior; top is superior).

A middle ear exploration was conducted via a total EES approach. A perilymphatic leak from the round window was identified and repaired with tragal perichondrium.

Post-operatively, the patient's vertigo resolved. Four-month post-operative audiogram showed significant improvement in hearing in the low frequencies into the normal range up through 1500 Hz with a stable, mixed, severe to profound downsloping hearing loss from 2000 Hz through 8000 Hz. He had a stable hearing and no vertigo recurrence at the 12-month follow-up.

## CASE 3

A 10-month-old male presented to the emergency room with sudden-onset, brisk, clear, right otorrhea, and nystagmus following an unwitnessed fall. He was distressed at the presentation and his parents expressed that he seemed off-balance relative to his baseline. Examination revealed left-beating nystagmus and brisk, clear right otorrhea. Neurological examination was otherwise normal and there were no signs of other injuries. CT showed a fracture through the round window promontory. The fracture extended deep to the internal auditory canal, near the round window, and up through the jugular bulb. The patient underwent a middle ear exploration via an EES

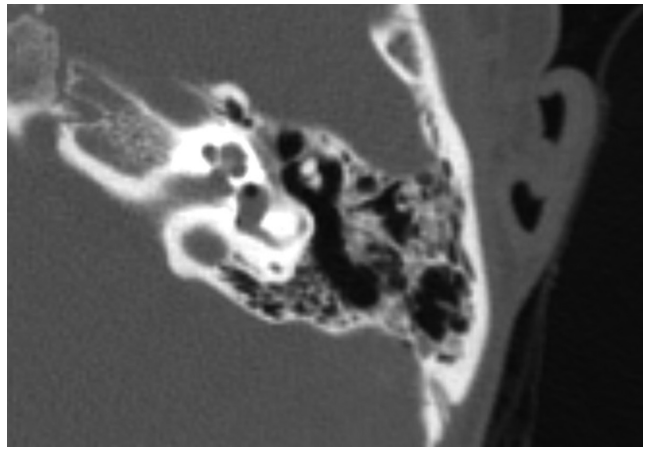

**Figure 3.** Case 2: Pneumolabyrinth in the vestibule. Image is axial CT of the left ear.

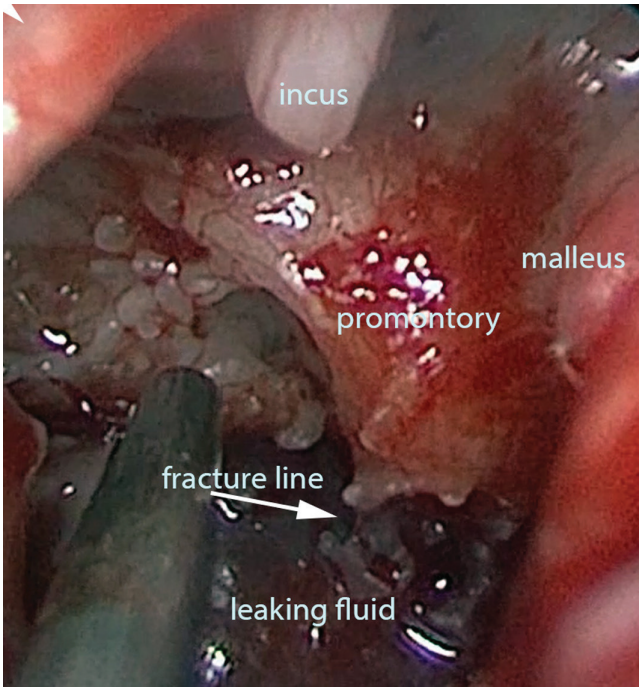


**Figure 4.** Case 3: Perilymphatic and/or cerebrospinal fluid leak from oval window defect and round window promontory fracture. Image is of the right ear (right is anterior; top is superior).

approach, which showed a brisk leak from both the oval window and the round window (Figure 4).

The flow volume was too high to seal the leak with a single-handed endoscopic approach, while reduced visualization with a 2-handed microscopic approach prohibited identification of the source of the oval window leak. Neurosurgery was unable to successfully place a lumbar drain. The middle ear was packed, and a lumbar drain was placed under fluoroscopic guidance the next day. An auditory brainstem response test showed a severe to profound mixed hearing loss in the affected ear. A second-stage repair was conducted 2 days later via a postauricular, microscopic approach with endoscopic assistance with the aid of the lumbar drain. The oval window leak was found to be emanating from the anterior oval window where the anterior third of the stapes footplate and the anterior stapedial crus were found to have been fractured and rotated forward off of the remaining stapes footplate and superstructure. The anterior stapes footplate and crus were removed, and the leak was controlled with a temporalis fascia graft. Additional leakage from the round window promontory fracture was controlled with bone wax and fascia. The lumbar drain was removed 2 days later without further leak.

Post-operative audiogram showed a moderate-to-severe mixed hearing loss on the right and normal hearing on the left. Post-operative vestibular testing showed an absent unilateral vestibulo-ocular reflex response on rotary chair testing and an absent right cervical vestibular evoked myogenic potential test. He had a stable hearing and no signs of vestibular symptoms over 10 months of follow-up.

## DISCUSSION

The differential diagnosis of PLF is broad, and a single test or imaging study is not pathognomonic. The history, physical examination, vestibular testing, audiometry, biomarkers, and imaging may be helpful in making the correct diagnosis.[1] The gold standard for diagnosing PLF remains middle ear exploration, and the minimally invasive EES approach lends itself well to this disease entity, particularly in pediatric patients.

Although single cases have been reported in adults, we have presented here a series of endoscopic PLF repairs in children.[5,8] Patient selection, operative set-up, correct instrumentation, and surgeon comfort are paramount.[3,4,7] The surgeon must be prepared to convert to a microscopic and/or postauricular approach, if needed, and a lumbar drain should be considered for brisk leaks.

The EES approach offers several advantages over the microscope that was evident in our cases, including greater magnification, increased field of view, increased angulation, and the absence of an external scar.[5] However, the microscope provides the advantages of binocular vision and 2 operating hands, which may be valuable in some PLF cases. The decision for a second-stage, post-auricular approach in the third patient was made to allow a lumbar drain to be placed under fluoroscopic guidance and to allow for greater manipulation and access.

Active surveillance for spontaneous resolution without surgical intervention is a viable option in traumatic PLF, but carries the risks of persistent vestibular symptoms and permanent SNHL.[9] These considerations must be addressed with the patient and family when determining the optimal treatment approach.

This study is limited by a retrospective approach without a control arm, so comparisons cannot be made to PLF patients who underwent active surveillance or microscopic repair.

## CONCLUSION

Traumatic PLF can be safely and effectively diagnosed and managed via an EES approach in children. The decision to pursue a middle ear exploration via an EES approach should be made on a case-by-case basis, and conversion to an endoscopic-assisted, traditional post-auricular, and/or staged approach must be considered in select cases. Additional study is warranted to evaluate the feasibility of this approach in a larger pediatric population.

**Informed Consent:** Written informed consent was obtained from the patients' parents who participated in this study.

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