Conductive hearing loss secondary to otosclerosis is a fairly common diagnosis in a busy otologic clinic. Stapedotomies are performed frequently with good outcomes in the majority of these patients. A rare but worrying occurrence of a perilymph gusher may occur during stapes surgery – though, thankfully at a reported incidence of only 0.03%.

Here we report a case of an unexpected intraoperative perilymph gusher, via video from and endoscopic approach.

This patient presented with progressive left sided conductive hearing loss, with normal clinical examination and imaging (fine cut CT temporal bones). The imaging revealed a lucent area at the region of the fissula ante fenestram. There was no evidence of enlarged vestibular aqueduct or internal auditory canal.

The patient was planned for an endoscopic left laser stapedotomy. Intraoperative, the footplate was noted to be fixed and laser stapedotomy followed by serial fenestration of the footplate was carried out. Unfortunately, a persistent high-volume gush of perilymph was encountered during the fenestration.

Multiple conservative attempts at stemming the flow was made – including the use of reverse Trendelenburg position, increasing respiratory rate and the administration of mannitol. When these efforts failed to produce any noticeable effects on the flow of perilymph, sealing of the fenestration with a muscle plug and vein graft was performed.

Postoperatively, the patient reported no vertigo and remained well throughout his admission. A repeat audiogram completed 2 months postoperatively showed preserved cochlear reserves.

In retrospect, this patient is suspected of having a “*X-Linked Stapes Gusher Syndrome”.* Mutations of POU3F4 are associated with X-linked (DFN3) deafness, resulting from both a conductive hearing loss caused by stapes fixation and progressive sensorineural pathology. Reports also indicate that those with this mutation seem to produce large volumes of perilymphatic flow during stapes surgery – which predisposes them to having a perilymphatic gusher and complicated surgery. Radiologic findings may include widening of the lateral internal auditory canal and dilation of the vestibule, although this may not be evident in all patients.

This case highlights the perils of stapes surgery should not be taken lightly, and one should always prepare the patient for unexpected outcomes. Those embarking on stapes surgery should also anticipate such complications and be well versed in tackling it. This is especially true when preoperative investigations suggest that the surgery will be routine and uncomplicated.