Facial nerve stimulation following cochlear implantation for X-linked stapes gusher syndrome

Kevin A. Peng MD1, Edward C. Kuan MD2, Frederick Yoo MD2, Rebecca Lewis AuD2, Ali R. Sepahdari MD2, Gail Ishiyama MD2, Akira Ishiyama MD2

Departments of 1Head and Neck Surgery, 2Auudiology and Speech Pathology, 3Radiological Sciences, and 4Neurology, David Geffen School of Medicine, University of California, Los Angeles, Los Angeles, CA

ABSTRACT
At the conclusion of this presentation, the participants should be able to recognize the presentation of X-linked stapes gusher syndrome and describe the anatomical characteristics that predispose these patients to facial nerve stimulation and/or facial nerve injury following cochlear implantation.

Objectives To present a case of facial nerve stimulation following cochlear implantation in a patient with X-linked deafness with stapes gusher syndrome.

Study design Retrospective case report.

Methods The medical records of a patient with X-linked stapes gusher syndrome status post cochlear implantation were reviewed.

Results A nine-year-old male presented to the otology clinic for continued care after cochlear implantation with a 21-electrode device in another country five years prior. Four years following implantation, and one year prior to presentation at our clinic, the patient began to experience intermittent ipsilateral facial nerve stimulation. The cochlear implant coursed normally through the basal turn of the cochlea, but at the level of the geniculate ganglion, the electrode protruded into the facial nerve canal, likely secondary to the observed absence of bony separation between the facial nerve and the cochlea. Taken together, radiological and clinical findings were diagnostic of X-linked stapes gusher syndrome. Inactivation of electrodes 1 and 19-21 successfully abated facial nerve stimulation.

Conclusions X-linked stapes gusher syndrome presents with a congenital hearing loss that usually bilateral, mixed, and progressive. Cochlear implantation is almost always accompanied by a CSF fistula at the time of cochleostomy. Patients typically derive useful hearing from the cochlear implant, but owing to the anatomical anomalies associated with this condition, the facial nerve is at increased risk for intraoperative injury as well as excessive stimulation following implant activation.

INTRODUCTION
The association between X-linked deafness and stapes gusher has been well described for nearly a half-century, and characteristically involves a dilation of the lateral internal auditory canal (IAC) as well as a deficiency of bone between the lateral IAC and cochlea, both of which are evident on computed tomography imaging. This deficiency likely permits the transmission of harmful intracranial pressure peaks to the cochlea, leading to a progressive hearing loss. When sensorineural loss becomes profound, cochlear implantation may be necessary. Although the inheritance pattern may suggest that only males are affected, heterozygous females also display varying degrees of hearing loss. An important differential diagnostic consideration is far-advanced otosclerosis, although the patient’s age and radiological findings are usually able to segregate these diagnoses.

Figure 1. Axial CT image through the level of the cochlear basal turn shows normal course of the cochlear implant electrode through the basal turn (black arrow).

Figure 2. Axial CT image through the level of the geniculate ganglion shows protrusion of the electrode into the facial nerve canal, near the geniculate ganglion (long white arrow). The tympanic segment of the facial nerve is also seen on this image (short white arrow).

Figure 3. Coronal CT image through the level of the geniculate ganglion, showing the electrode coursing through the cochlear basal turn (black arrow) and protruding into the geniculate ganglion (white arrow). Note the absence of bony separation between the facial nerve canal and the cochlea.

Patients with profound hearing loss secondary to inner ear anomalies generally derive benefit from cochlear implantation, but the rate of perioperative complications is, not surprisingly, higher than that seen with implantation of normal cochleae. In particular, performing a cochleostomy in X-linked stapes gusher patients virtually guarantees a cerebrospinal fluid (CSF) fistula. Furthermore, given the deficiency of bone at the basal turn of the cochlea, there is an additional, albeit small, risk of inadvertent passage of the implant beyond the confines of the cochlea. Postoperatively, facial nerve stimulation may be encountered, as demonstrated in the case here.

CASE PRESENTATION
A nine-year-old male presented to the otology clinic for continued care after cochlear implantation. He was diagnosed with progressive congenital hearing loss in infancy, with hearing in the left ear affected more than the right. There was no family history of deafness. By age 4, he demonstrated a profound loss in his left ear, and underwent cochlear implantation with a 21-electrode device.

Four years following implantation, and one year prior to presentation at our clinic, the patient began to experience intermittent facial nerve stimulation, deemed iatrogenic secondary to electrical stimulation by the implant. Electrodes 1 as well as 19-21 were switched off, which successfully abated the facial nerve stimulation. On examination in our clinic, facial nerve function was full and symmetric. Computed tomography (CT) was performed, revealing a dilated internal auditory meatus. The cochlear implant coursed normally through the basal turn of the cochlea and the lateral IAC. Phelps et al. describe a third phenomenon — CSF fistula at the time of cochleostomy, dehiscence of the bony partition between the basal turn of the cochlea and the IAC, leading to iatrogenic facial nerve stimulation. Fortunately, the patient’s volitional facial nerve function was unaffected.

CONCLUSION
Special considerations must be taken into account when performing cochlear implantation on patients with inner ear malformations. In particular, the X-linked stapes gusher is associated with three specific phenomena — CSF fistula at the time of cochleostomy, dehiscence of the bony partition between the basal turn of the cochlea and the IAC, and dehiscence of the bony partition between the cochlea and the facial nerve canal — the last of which may predispose to facial nerve stimulation upon implant activation.

REFERENCES