Congenital hearing loss and anomalies of the internal auditory canal: imaging findings and implications for surgical and clinical management

Jessica Somerville, MD1; C. Bruce Macdonald, MD1
1University of Tennessee, Department of Otolaryngology Head and Neck Surgery

Abstract

Introduction: Approximately 20% of patients with congenital sensorineural hearing loss are found to have a radiologically proven inner ear anomaly. Of the inner ear abnormalities, duplication of the internal auditory canal is rare. The nomenclature of this finding has been described as duplicated internal auditory canal and as anomalous facial nerve course associated with internal auditory canal atresia. We sought to describe clinical characteristics and treatment results of patients found to have duplicated internal auditory canal on imaging.

Methods: This case series identified patients from over a 2 year period obtained in a random manner from teaching files and daily case material. Imaging findings, clinical characteristics, and audiologic and treatment were reviewed.

Results: A total of 7 patients were identified with a total of 12 ears (2 patients had unilateral presentation). 3 patients in this series had a history of branchio-oto-renal (BOR) syndrome. All patients had high resolution computed tomography (CT) performed. 4 patients had Magnetic resonance imaging (MRI) performed. All patients had audiogram evaluation. 6 patients had sensorineural hearing loss, and one patient had conductive hearing loss. As hearing loss became more severe as documented by decibels of hearing loss on audiogram, increasing number of middle and inner ear anomalies were found on imaging.

Conclusions: This series is one of the largest series to evaluate duplicated internal auditory canal in conjunction with cochlear abnormalities. MRI is a useful adjunct to evaluate the cochlear nerve and should be considered in patients undergoing cochlear implantation with findings of duplicated internal auditory canal on CT scan. Identification of the facial nerve course is important in patients with IAC stenosis, and anomalies of the facial nerve canal may not manifest on physical exam. Hearing loss became more severe, there number of structural abnormalities in the inner and middle ear increased.

Discussion

The incidence of severe congenital hearing loss has been estimated at one in every 1000 live births. Of patients with congenital hearing loss, a smaller percentage will have anomalies noted on radiographic imaging even without physical exam abnormalities. For example, congenital hearing loss may be associated with facial nerve abnormalities and the patient may not have evidence of facial nerve dysfunction on physical exam. Patients with abnormalities of the external ear have a higher percentage of facial nerve abnormalities on imaging than patients with inner ear abnormalities. The more common finding in patients with abnormal facial nerve anatomy and congenital hearing loss is a conductive loss related to the tympanic segment of the facial nerve. The labyrinthine segment of the facial nerve may be related to a malformed cochlea in a similar fashion that the tympanic segment is anatomic aid to malformed middle ear structures. The course of the facial nerve is an important surgical landmark, and thus its course is crucial in determining the approach to patients with hearing loss undergoing surgical procedures for hearing amplification.

The facial nerve travels in the internal auditory canal in the anterior superior division. Although rare, both unilateral and bilateral duplication of the internal auditory canal has been described. On imaging, this is seen when the internal auditory canal is of smaller caliber and the facial nerve travels in a “duplicated” canal instead. Most patients with atresia of the internal auditory canal have normal facial nerve function. In addition, duplication of the internal auditory canal has been classically associated with the lack of a vestibulocochlear nerve. The defect is most commonly defined as a stenotic/atretic posteriorinferior canal with the facial nerve running inside a more antero-superior canal. The normal caliber of the IAC ranges from 2 to 8 millimeters with an average diameter of 4 millimeters. A canal of less than 2 millimeters is considered stenotic. Often inner ear anomalies are frequently present in addition to IAC anomalies. A 2011 study by Corlin et al. of 11 patients with anterior displacement of the labyrinthine segment of the facial nerve also had associated cochlear abnormalities. External ear anomalies have also been described with IAC stenosis.

The facial nerve canal develops from the primordial otic capsule and Reicher’s cartilage from the second branchial arch. The canalicular segment of the facial canal develops in the primordial otic capsule, and the fusion of the otic capsule with Reicher’s cartilage provides the remaining cartilaginous base for the labyrinthine and tympanic segments. Previous studies have explained anteromedial migration of the facial nerve canal as a natural course in development with a hypoplastic cochlea. Anteromedial position of the facial nerve has not previously been reported with Mondini malformation but has been associated with a hypoplastic cochlea. Some authors have postulated that the large cochlear size in Mondini’s deformity would preclude anterior migration of the labyrinthine segment of the facial nerve. Other theories regarding anterior migration of the facial nerve at various points in its course through the temporal bone relate to structures derived from Reicher’s cartilage. For example, the absence of the stapes crura may allow for anterior migration of the tympanic or mastoid segment of the facial nerve in an attempt to take a more direct route to its end organ, the muscle of facial expression. This case series is the first, to our knowledge, to identify anteromedial displacement of the labyrinthine facial nerve in association with Mondini’s dysplasia of the cochlea as seen in 2 of our patients.

BOR syndrome patients frequently have inner ear abnormalities. Kemptner et al. described the common presence of inner ear anomalies in patients with BOR, however, they were unable to identify a pathogenic change in their series of 35 patients. Almost half of the patients in our study had BOR syndrome. Our patients presented on combined MRIs or PET in 4 patients that suggest that internal auditory canal diameter does not correlate with the presence or absence of cranial nerve 8 and thus may not present with significant hearing loss. Our patient series revealed that patients with normal hearing had deep and middle ear structures that did not contribute to the hearing loss. Patients with more profound hearing loss required hearing amplification for speech and language development.

Conclusions

This case series is one of the largest series to evaluate bilateral and unilateral duplicated internal auditory canal in conjunction with cochlear abnormalities. Contrary to previous reports, a stenotic IAC is not a reliable indicator of presence or absence of the vestibulocochlear nerve. Our patient series concurs with other more recent suggestions in the literature that canal diameter does not correlate with nerve presence or absence. MRI is a useful adjunct to evaluate presence of the cochlear nerve and should be considered in patients undergoing cochlear implantation with findings of duplicated internal auditory canal on CT scan. Identification of the facial nerve course is important in patients with IAC stenosis, and anomalies of the facial nerve canal may not manifest on physical exam. As hearing loss became more severe, there number of structural abnormalities in the inner and middle ear increased.

Individual cases were accrued in a random manner from teaching files and daily case material over a 2 year time period. It is routine practice at our institution to obtain temporal bone CT scans on patients presenting with congenital idiopathic sensorineural hearing loss. Imaging findings, clinical characteristics, audiologic testing and treatment were reviewed. 2 neuroradiologists reviewed available imaging, and all audiologic testing was performed at the same institution.

References

5. Girensaman AV, Medawar L, Leifer L, Goetz J. Abnormal course of the facial sensory fibers in cases of atresia of the internal auditory canal.