ABSTRACT

Objective: The objective of this case report is to describe a newborn patient presenting after abnormal newborn hearing screening who was found to have cochleae with three turns bilaterally. A discussion of the limited relevant literature is presented, along with possible implications of this rare abnormality.

Methods: Case report and review of the limited relevant literature.

Case Report: The case of a newborn presenting after a failed newborn auditory screening is given, along with diagnostic evaluation, imaging findings, treatment, and follow-up.

Conclusion: Cochleae with three turns are a rare malformation that are not included in current classifications schemes. They may represent a distinct type of anomaly not caused by developmental arrest, and further research is needed to understand the effect of increased cochlear turns on central auditory processing and mechanisms of hearing loss.

INTRODUCTION

The human cochlea has been widely reported to have 2 ½ turns. However, the actual number of turns in the normal human cochlea has shown to vary, and the majority of normal cochleae contain between 2 ½ and 2 ¼ turns.1 We report a patient with incomplete sensorineural hearing loss (SNHL) who was found to have cochleae with 3 turns. Cochleae with 3 turns have been previously reported, however this represents the first report of this rare finding during evaluation of a patient with hearing loss.2

CASE REPORT

A 4 month old male with no known congenital abnormalities or medical problems was referred after a failed right newborn hearing screening. The child had no family history of hearing loss, though his father had complete heart block since childhood.

The child had been discharged from the newborn nursery without issue and otoacoustic emission (OAE) testing, at that time, was normal. The child developed hyperbilirubinemia, requiring readmission, and was treated with gentamicin for 72 hours for a fever. Distortion product otoacoustic emissions (DPOAE) testing, shortly after this event, was normal bilaterally. Unsedated Auditory brainstem response (ABR) testing indicated a marked wave V at 15 dB in the left ear and at 40 dB in the right. Unsedated ABR was repeated 1 month later, in an attempt to gain an improved response in the right ear. This demonstrated a progressing 50 dB hearing loss in the left ear and a stable 40 dB loss in the right.

The child was referred to our institution at 4 months of age for a sedated ABR, as the child had inconsistent results on previous testing. The child appeared normal and otoscopy was unremarkable. Sedated ABR revealed that the hearing had again worsened, with a 70 dB loss in the right ear and 65 dB loss in the left. However, bone levels were at a 35 dB hearing level, demonstrating a mixed loss. Interestingly, tympanometry revealed type A tympanograms. Imaging was performed and T2 weighted MRI revealed bilateral mild bilateral cochlear dysplasia with an increased number of cochlear turns. (Figure 1-4).

CASE REPORT

Tymanostomy tubes were placed bilaterally for recurrent infection at 6 months of age, no fluid was seen at time of placement. Sedated ABR testing was repeated at the same time, revealing results that were unchanged from the previous sedated ABR. Thus, it was postulated that both the sensorineural and conductive components of the hearing loss may have been related to the cochlear malformation.

DISCUSSION

The cochlear duct begins to form from the saccule of the otic vesicle in week 8 of development. The cochlear duct penetrates the surrounding mesenchyme, in a spiral fashion, and completes its turns by week 8.3 As development proceeds, the mesenchyme surrounding the cochlear duct forms cartilage, involutes to form the scala tympani and scala vestibuli, and finally ossifies as the otic capsule by birth.

Cochleae with three turns are a rare malformation that cause SNHL can be divided into membranous (80%) and osseous malformations (20%).3 Imaging evaluation reveals normal inner ear anatomy in the membranous type. In the osseous type, high-resolution computed tomography or high-resolution T2-weighted MRI provide accurate visualization of the inner ear and allow identification of osseous cochlear deformities.

The currently accepted categorization, proposed by Jackler et al, classifies cochlear deformities based on arrest during progressive stages of embryological development. The Jackler classification lists four types of osseous cochlear malformations: 1) cochlear aplasia, 2) cochlear hypoplasia, 3) incomplete partition, and 4) common cavity malformation.4 This classification was further refined by Sennaroglu and colleagues, who suggest that incomplete partition be divided further into cystic cochleovestibular malformation (IP-I) and the Mondini deformity (IP-II).5 As stated by Tian et al, human cochleae with three turns are a malformation not reflected in the current classification scheme.

FIGURE LEGEND

Figures 1-4 demonstrate axial (Figure 1,2) and coronal (Figure 3,4) T2 weighted MRI views of the 3 turn cochlea.

REFERENCES