PICTORIAL REVIEW

Review of congenital inner ear abnormalities on CT temporal bone

¹R S Z YIIN, BMBS, FRCR, ²P H TANG, MBBS, FRCR and ¹T Y TAN, MBBS, FRCR

¹Department of Diagnostic Radiology, Changi General Hospital, Singapore, and ²Department of Diagnostic and Interventional Imaging, KK Women's and Children's Hospital, Singapore

ABSTRACT. The aetiology of profound hearing loss in children is complex and multifactorial. Congenital inner ear abnormality is a major cause of hearing loss in children. CT temporal bone imaging is the modality of choice in the investigation of hearing loss. Recognising the congenital abnormalities of the inner ear guides the clinician's management of the condition. This pictorial essay illustrates the congenital abnormalities of the inner ear on high resolution CT temporal bone images and correlation with developmental arrest during embryology.

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Congenital inner ear abnormality is a major cause of sensorineural hearing loss in children [1]. The aetiology of profound congenital hearing impairment is divided into two main causes: environmental (50%) and genetic (50%). Environmental causes include viral infections (toxoplasma, rubella, cytomegalovirus and herpes simplex virus (TORCH)), bacterial meningitis, prematurity and foetal exposure to isoretinoin. Genetic causes can be divided into syndromic (30%) and non-syndromic (70%). Syndromic causes include Alport, Pendred, Waardenburg, CHARGE, branchio-oto-renal and X-linked progressive hearing loss with perilymphatic gusher. Non-syndromic causes include autosomal dominant, autosomal recessive, X-linked and mitochondria [1]. Recognising the congenital abnormalities of the inner ear guides the clinician's management of the condition.

Embryology

At approximately the third week of gestation, otic placodes arise from the surface ectoderm on each side of the rhombencephalon. The otic placodes subsequently invaginate and form otocysts, which are the otic and auditory vesicles. At around the fifth week, diverticulum buds from the otocysts form the endolymphatic sacs, followed by the cochleas and vestibules. The membranous cochlea achieves 1 to 1.5 turns at the end of 6 weeks, and 2.5 turns are formed at the end of the 7th week. The semicircular canals start to develop from the utricle segments of the otocysts at 7–8 gestational weeks. The superior canals form first, followed by the posterior and then the lateral canals. The inner ear structures have an

adult configuration by the end of 8 weeks [2]. The normal inner ear anatomy is depicted in Figure 1.

Classification

Jackler et al [2] proposed the most commonly used classification of inner ear abnormalities, which is based on a linear developmental model towards normal anatomy and the most likely time at which developmental arrest occurs during embryogenesis. Sennaroglu et al [3] further modified this classification. However, this classification has been challenged by other authors [4].

Michel aplasia

Michel aplasia, also known as complete labyrinthine aplasia, is a rare congenital inner ear abnormality, accounting for approximately 1% of cochlear bony malformations. This condition is defined as complete absence of inner ear structures (Figure 2) and is caused by developmental arrest of otic placode early during the third week of gestational age [2, 5]. A recent case series by Ozgen and Sennaroglu et al [5] described a spectrum of temporal bone and skull base anomalies that may be associated with this condition, such as petrous bone aplasia, narrowed or aplastic internal auditory canal, tegmental defects, mastoid and middle ear hypoplasia as well as jugular bulb variations.

Cochlear aplasia

Failure of cochlea development late in the third week of gestation results in this condition (Figure 3 and 4). The vestibule and semicircular canals are either normal, dilated or hypoplastic [4].

Address correspondence to: Dr Raphael Shih Zhu Yiin, Department of Diagnostic Radiology, Changi General Hospital, 2 Simei Street 3, Singapore 529889. E-mail: shihzhu.yiin@mohh.com.sg

Figure 1. CT images of normal right inner ear anatomy: (a–f) axial superior to inferior images. 1, internal auditory canal (IAC); 2, superior semicircular canal (SCC); 3, lateral SCC; 4, posterior SCC; 5, vestibule; 6, basal turn of cochlea; 7, apical turn of cochlea; 8, vestibular aqueduct; 9, facial nerve.

Common cavity

In common cavity malformation, developmental arrest occurs at the fourth week of gestation and is defined as a single cavity that represents the undifferentiated cochlea and vestibule (Figure 5).

Incomplete partition I

Incomplete partition I is also known as cystic cochleovestibular malformation, where the cochlea has no bony modiolus, resulting in an empty cystic cochlea (Figures 6 and 7). This is accompanied by a dilated cystic vestibule with developmental arrest at the fifth week of gestation [4].

Cochlea hypoplasia

The cochlea and vestibule can be differentiated from each other but the size of the cochlea is smaller than normal (Figure 8). Developmental arrest occurred at the sixth week of gestation.

Incomplete partition II

In this group, the cochlea consists of 1.5 turns; the apical and middle cochlea turns are undifferentiated and form a cystic apex (Figures 9 and 10). The vestibule is normal while the vestibular aqueduct is always enlarged. Developmental arrest occurs at the seventh week of gestation.

Figure 2. Michel deformity. (a) Axial image demonstrates total absence of inner ear structures in the right ear of an 8-year-old child, (b) coronal image of another patient with Michel deformity. Developmental arrest will have occurred at the third week of gestation. Note the narrow right internal auditory canal in the axial image (black arrow).

Figure 3. 1-year-old child with profound left sensorineural hearing loss. (a–c) Axial images show that the left cochlea is absent. Developmental arrest will have occurred late in the third week of gestation. There is also no separation of the lateral semicircular canal with the vestibule on the left (white arrow). The left posterior semicircular canal is also dilated (*). The left internal auditory canal is narrowed (black arrow).

Figure 4. 5-year-old child with bilateral cochlear aplasia. (a) Axial and (b) coronal images show absence of both cochleas. Note also the bilateral dilated vestibules (white arrows).

Figure 5. 1-year-old child with common cavity. Axial image of the right ear shows the cochlea and vestibule cannot be differentiated from each other. Developmental arrest will have occurred at the fourth week of gestation.

Figure 6. 1-year-old male with unilateral incomplete partition Type I (IP I). (a,b) Axial images show the cystic left cochlea (black arrow) and the mildly dilated cystic left vestibule (white arrow). The vestibular aqueduct is normal in size (arrowhead). (c,d) Coronal images show the cystic left cochlea (black arrow) and cystic left vestibule (black arrow). Note also a shortened lateral semicircular canal (white arrow). Developmental arrest will have occurred at the fifth week of gestation.

Semicircular canal abnormalities

The semicircular canals can either be absent, hypoplastic or enlarged (Figures 11–13). The most commonly recognised semicircular canal abnormality is a short lateral semicircular canal being confluent with the vestibule [6]. The semicircular canals develop at 6 to 8 weeks gestation and are completed between the 19th and 22nd weeks [6].

Enlarged vestibular aqueduct

Enlarged vestibular aqueduct (EVA) is commonly defined as having a width larger than 1.5 mm, measured at the midpoint of the common crus and external aperture (Figure 14). This condition was first described radiologically

Figure 7. 1-year-old male with bilateral incomplete partition Type I (IP I). (a,b) Axial images show that the cochleas (black arrows) and vestibules (*) are cystic.

Figure 8. 8-year-old female with bilateral cochlear hypoplasia. (a,b) Coronal images show small bilateral cochleas. Developmental arrest occurred at the sixth week of gestation.

Figure 9. 2-year-old male with bilateral incomplete partition II (Mondini deformity). (a) Coronal image shows both the cochlea only have 1.5 turns. (b) Axial image of the same patient shows the vestibules are normal (black arrows) and the vestibular aqueducts are enlarged (arrowheads). Developmental arrest occurred at the seventh week of gestation.

Figure 10. 10-year-old male with unilateral incomplete partition II (Mondini deformity). (a,b) Axial image show the cochlea only have 1.5 turns. The vestibules are normal (*) and the vestibular aqueduct is enlarged (black arrow). Developmental arrest occurred at the seventh week of gestation.

Figure 11. 12-year-old female with bilateral lateral semicircular canal aplasia. (a) Axial and (b) coronal images show both lateral semicircular canals are undifferentiated from the vestibules. White arrows indicate the location of where the lateral semicircular canals are expected to arise.

Figure 12. 4-year-old male with bilateral, lateral semicircular canal hypoplasia. (a) Axial shows both lateral semicircular canals are shortened (black arrowheads). (b) Normal right inner ear (black arrowheads) of a different patient to (a). (c) Coronal image shows the shortened left lateral semicircular canal, compared with a normal image of (b).

Figure 13. 2-year-old male with bilateral superior and posterior semicircular canals hypoplasia. (a) Coronal image shows the hypoplastic superior semicircular canals (black arrowheads). (b, c) Axial images of the same patient show bilateral posterior semicircular canal hypoplasia (black arrows). Note also the bilateral lateral semicircular canal aplasia.

Figure 14. 10-year-old male with profound sensorineural hearing loss. (a, b) Axial images show an isolated, enlarged left vestibular aqueduct (white arrows) without other inner ear abnormality. Image (a) is taken at the midpoint of the common crus and external aperture, which measures 3 mm. Image (b) is taken at the external aperture, which measures 7 mm.

by Valvassori and Clemis [7]. EVA has been reported to be the most common inner ear abnormality associated with sensorineural hearing loss and is also commonly associated with other inner ear abnormalities [6]. The aetiology of EVA is still under debate. One group of authors proposes that developmental arrest occurred at the fifth week of gestation. Another group suggests the abnormality occurred later in foetal and post-natal life. Indeed, Pyle et al [7] has demonstrated that non-linear growth of the vestibular aqueduct occurs throughout foetal life.

Conclusion

The aetiology of profound congenital hearing loss is multifactorial and complex. Congenital inner ear abnormality is a major cause of hearing loss in children. Hence, recognising the congenital abnormalities of the inner ear guides the clinician's management of the condition.

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