

Imaging of Congenital Middle Ear Deafness

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Abstract

Introduction: Conductive hearing loss, in the presence of a normal external ear, is usually due to an interference with the normal transmission of sound from the tympanic membrane across the ossicular chain to the oval window. If conductive hearing loss occurs in a young child, congenital middle ear pathology is the most likely cause. **Materials and Methods:** High resolution computed tomography of the middle ear is the diagnostic modality of choice. Details of the imaging technique are outlined in this article. Eight cases of congenital middle ear deafness imaged in the radiological departments of Changi General Hospital and Tan Tock Seng Hospital in Singapore are illustrated. **Results:** Congenital middle ear deafness may be classified into ossicular chain abnormalities, congenital ossicular fixation, congenital oval window atresia and congenital round window atresia. The imaging features for each category are described. **Conclusion:** With proper imaging technique and a systemic analysis of the images, the abnormalities that result in congenital middle ear deafness can usually be demonstrated on most occasions.

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Introduction

When a child or young adult with morphologically normal external ears and no previous history of ear discharge or trauma presents with conductive hearing deficit, a congenital middle ear anomaly should be sought. Although fenestral otosclerosis is a differential diagnosis, the early onset of symptoms and a lack of progression would tend to favour congenital middle ear deafness. Fenestral otosclerosis usually presents in the second and third decades, and rarely in the first decade, of life.^{1,2}

The introduction of high resolution computed tomography (HRCT) of the middle ear has revolutionised the assessment of congenital middle ear deafness.³ In the past, the cause of deafness could only be revealed after exploratory tympanotomy; many congenital abnormalities that cause hearing loss could now be demonstrated by HRCT. However, exploratory surgical evaluation is still more accurate than radiological assessment at the present level of technological advancement, even though the introduction of multi-slice computed tomography (CT) scanner, with its submillimetre scanning, has further narrowed the gap between radiological and surgical assessments of congenital middle ear deafness.

Imaging Technique

The assessment of middle ear anomalies requires meticulous attention to the acquisition of images. HRCT of

the middle ear is the modality of choice. Images are acquired in the axial and coronal planes. A contiguous slice thickness of 1 mm is obtained. The stapes is best evaluated in the axial plane by overlapping the slices at 0.5 mm intervals over the oval window region. Positioning of the head is important for the axial scan and a frontal scout is employed. The head is adjusted such that the petrous ridge is projected over the lower one-third of the orbit. No gantry tilt is required. Over-flexion of the head, with the petrous ridge projecting over more than half of the orbit, would result in the facial nerve obscuring the stapes superstructure and excessive radiation to the lens. Over-extension of the head, with the petrous ridge being projected below the orbit, would result in distortion of the architecture of the middle ear.³ In the axial plane, images are acquired from the head of the malleus superiorly to the round window inferiorly. This can be achieved by imaging from a point about 5 mm below the top of the petrous ridge and scanning about 12 sections below this point. For coronal images, a lateral scout is used. The scan plane is parallel to the ramus of the mandible. Images are acquired from the head of the malleus anteriorly to the round window posteriorly. This is achieved by scanning over the round lucency of the external auditory canal in the lateral scout image, from the anterior to the posterior margins.

With multi-slice CT scanners, images are acquired at 0.5-mm intervals in the axial plane and reconstructed at

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0.3-mm intervals over the oval window region for better delineation of the stapes superstructure. Coronal images may be obtained by reformatting the raw data from the axial images. As such, the area of coverage in the axial plane should extend from the top of the petrous ridge to the tip of the mastoid bone. A bone algorithm is used in the acquisition of the images. Filming is done using intermediate window settings, so that both the bone and soft tissue are well demonstrated. The authors use a window width of 4000 and a window level of 800 for filming. However, different CT machines require slightly different window settings to achieve optimal images on film. It is better to customise these settings for individual machines rather than to use the prescribed window settings when filming.

Embryology of Middle Ear Structures

Knowledge of the embryology of the middle ear structures is the key to understanding the various anomalies that occur in the middle ear. The development of the ossicles and their attachments occurs between the 4th and 11th weeks of gestation. These structures are derived from the first and second branchial arches. The head of the malleus and the body and short process of the incus are derived from the first branchial arch (Meckel's cartilage). These structures are essentially within the epitympanum, forming the upper ossicular chain. The structures of the lower ossicular chain (within the mesotympanum) are the malleus manubrium, the long and lenticular processes of the incus and the stapes. These are derived from the second branchial arch (Reichert's cartilage), although the stapes has a dual origin. The head (capitulum), neck, anterior crus and posterior crus and tympanic surface of the stapedial footplate are derived from the second branchial arch. The vestibular surface of the footplate and the annular ligament are derived from the otic capsule.⁴

At 4.5 weeks of gestation, the stapes precursor, or blastema, appears. This structure encloses the stapedial artery in the 6th week of gestation, forming a dense mesenchymal ring with an obturator foramen. The stapedial artery atrophies in the 10th week of gestation, leaving the 2 crura with an empty obturator foramen. The facial nerve grooves the exterior aspect of the blastema, resulting in the development of the interhyale (future stapedius muscle and tendon) and laterohyale (which would eventually form the lateral margin of the bony facial nerve canal). The malleus and incus develop later than the stapes, starting in the 6th week of gestation. Incudostapedial articulation develops when the long process of the incus advances toward the stapedial ring to fuse with the head of the stapes. Between the 6th and 8th weeks of gestation, malleo-incudal articulation develops after these ossicles become separated by a zone of undifferentiated mesenchyme.

From the 11th to the 26th weeks of gestation, the ossicles undergo cartilage reabsorption, marrow obliteration, ossification and increase in size. Ossification of the incus and malleus occur in the 16th week of gestation, whereas that of the stapes occurs in the 18th week. The malleus and incus achieve adult size in the 16th week of gestation.⁵ Complete formation of the stapes is only achieved by the 38th week.⁶

Classification of Congenital Middle Ear Deafness

Anomalies of the middle ear that result in conductive hearing deficit can be associated with external auditory dysplasia, a known syndrome such as Klippel-Feil syndrome, or may occur in isolation.² Those that occur in isolation are known as congenital middle ear deafness. Many classifications have been proposed to categorise congenital middle ear deafness. One of these divides the causes into 4 classes.⁷ Another subdivides the lesions into malleo-incudal fixations, stapes fixations and incudostapedial disconnections.⁴ For our purpose, congenital middle ear deafness is divided into 4 categories: ossicular chain deformities, ossicular fixation, oval window atresia and round window atresia.

Congenital Ossicular Chain Deformities

Most ossicular chain deformities detected by imaging occur around the incudostapedial joint.² The structures of the incudostapedial joint are all derived from the second branchial arch. The fault may lie with the improper development of the long process of the incus, leading to failure of union between this structure and the stapes capitulum. Examples of this include an elongated and posteriorly oriented long process of incus, absent long process or a shortened long process.⁸ A shortened long process gives the appearance of a "blunted beak" on the axial scan (Figs. 1a & 1b). Stapes abnormalities have a common association with abnormalities of the long process of the incus. One or both stapedial crura may be deformed or even absent (Fig. 1a).² When only 1 crus is present, it is known as a unicrural stapes (Fig. 2). If both crura are absent and a bony beam is seen protruding from the footplate and articulating with the lenticular process of the incus, it is known as a monopod stapes (Fig. 3).⁹ Since the tympanic portion of the facial nerve canal also develops from the second branchial arch, deformity of the incudostapedial joint could be associated with abnormalities of the canal. This may take the form of canal dehiscence with facial nerve protrusion or an anomalous course with the facial nerve lying on the cochlear promontory, or even on the floor of the middle ear cavity.

The least common form of ossicular chain deformity detectable by imaging is malleo-incudal fusion (Fig. 4).

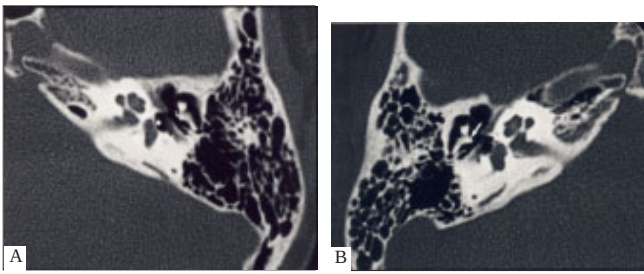


Fig. 1. (A) Axial HRCT scan of the left ear of an 8-year-old Chinese boy with conductive hearing loss. The small white arrow points to the congenitally shortened long process of the incus, giving a "blunted beak" appearance. Note that the stapedia crura (white asterisk) are also absent. (B) Axial HRCT scan of the normal right ear shows the "beak" (white arrow) of a normal long process of the incus. The small arrow points to the anterior crus of the normal stapes, which is just visible in the oval window.

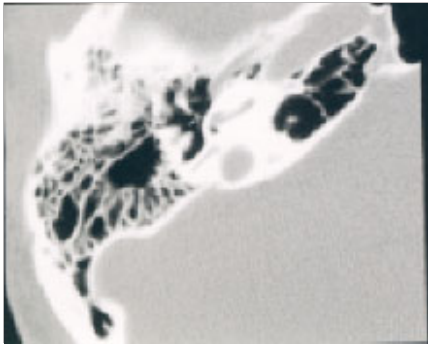


Fig. 4. Axial HCRT scan of the right ear of a 22-year-old Chinese lady with a long history of right-sided hearing loss shows malleo-incudal fusion (small black arrow), with the fused complex ossicles articulating with the stapes (small white arrow). The external auditory canal is normal (not shown).

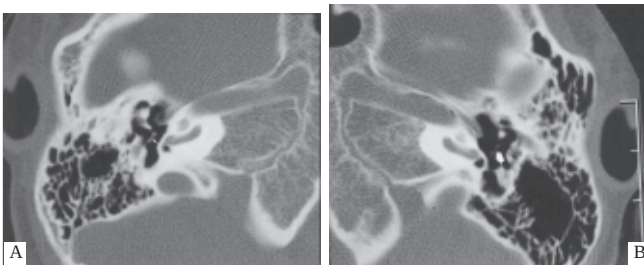


Fig. 6. (A) Axial HRCT scan of the right ear of a 10-year-old Chinese girl with right-sided conductive hearing loss for 5 years shows an ossified stapedius tendon (small white arrow). (B) Axial HRCT scan of the normal left ear shows the corresponding section. The broad white arrow indicates to the location of the stapedius tendon, which is only faintly visible on HRCT in normal cases.

This occurs when there is failure of interposition of a zone of undifferentiated mesenchyme between the malleus and incus during their pre-cartilaginous stage.² Though rare as an isolated anomaly, malleo-incudal fusion is a common ossicular deformity associated with external auditory canal dysplasia. Malleo-incudal fusion is best appreciated on the axial CT projection.

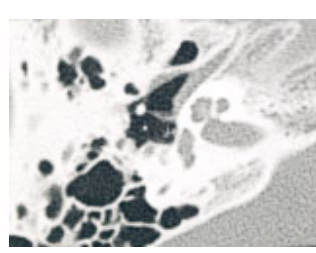


Fig. 2.

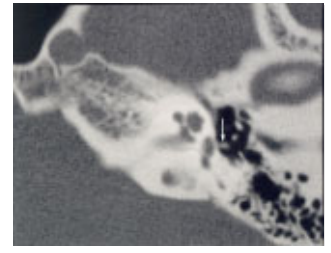


Fig. 3.

Fig. 2. Axial HRCT scan of the right ear of an 18-year-old Chinese boy with a history of right-sided hearing loss since childhood shows that the stapes has only the anterior crus (small white arrow); the posterior crus is absent. There is an associated absence of the distal incus which, in normal cases, would be found in the location indicated by the white asterisk.

Fig. 3. Axial HCRT scan of the left ear of a 20-year-old Chinese girl with bilateral conductive hearing loss since childhood shows a monopod stapes (white arrow). The stapes in the right ear appears similar (not shown).

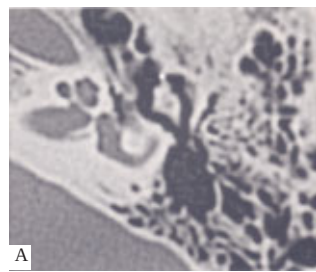


Fig. 5. (A) Tympanogram of a 32-year-old Chinese lady with a mild left conductive hearing loss shows a Type B pattern, indicating an impedance to the ossicular vibration. Axial HRCT scan of the left ear shows that the head of the malleus is fixed to the wall of the middle ear cavity by an ossified anterior malleolar ligament (white asterisk). (B) Axial HRCT scan of the normal right ear shows the corresponding section. The small white arrow indicates the location of the anterior malleolar ligament, which is usually not visible on HRCT.

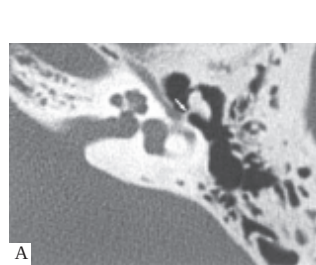


Fig. 7. (A) Axial HRCT scan of the left ear of a 43-year-old Chinese man with a history of left conductive hearing loss since childhood shows an anomalous bony bar (white arrow) fixing the body of incus to the medial wall of the middle ear cavity. (B) Axial HRCT scan of the normal right ear at the corresponding section. Note the absence of an anomalous bony bar.

Congenital Ossicular Fixation

Normal-appearing ossicles can be fixed to the wall of the middle ear cavity by calcified/ossified supporting ligaments or tendons, or by anomalous bony bars. When this occurs, the mobility of the affected ossicle is reduced, leading to conductive hearing loss. The malleus may be fixed to the



Fig. 8. Coronal HRCT scan of the left ear of a 28-year-old Malay man with bilateral conductive hearing loss since childhood shows a tapered configuration of the air in the middle ear (white asterisk) in the expected position of the oval window. The small arrow indicates the bony plate sandwiched between the vestibule and the middle ear. The small white arrow points to the anomalous tympanic portion of the facial nerve. The right ear also demonstrates all these features (not shown).

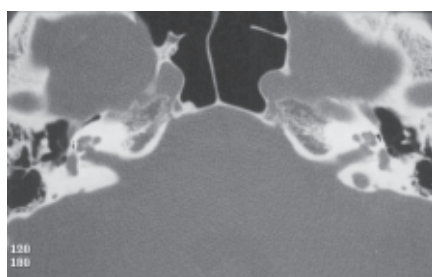


Fig. 9. Axial HRCT scan in a 32-year-old Chinese lady with a progressive right conductive hearing loss shows a lucent otosclerotic plaque in the anterior oval window region on the right side (small arrow). The corresponding region on the normal left side is of pearly white density (arrowhead).

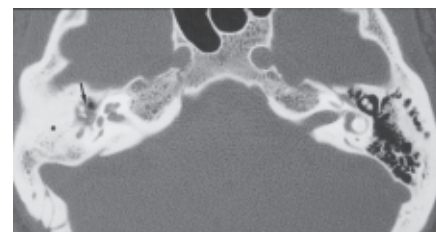


Fig. 10. Axial HRCT scan in a 33-year-old Malay lady with chronic right ear discharge and conductive hearing loss shows a sclerotic right mastoid bone (asterisk) with inflammatory debris within the middle ear cavity (arrow). The normal left middle ear cavity is well aerated and the mastoid bone is well pneumatized.

wall of the middle ear cavity by a calcified/ossified anterior or lateral malleal ligament (Figs. 5a & 5b). An ossified or calcified stapedial tendon can cause fixation of the stapes (Figs. 6a & 6b).⁷ Calcification or ossification of ligaments and tendons can also result from chronic middle ear inflammation, that is, tympanosclerosis. However, the age of the patient, the lack of a history of ear discharge, the well pneumatized mastoid air cells and the lack of any inflammatory debris within the middle ear cavity or mastoid air cells would tend to point to a congenital cause. In some cases, anomalous bony bars fixing the incus or the malleus to the wall of the middle ear cavity are responsible for the conductive hearing deficit (Figs. 7a & 7b).

If the CT scan shows a normal ossicular chain and no evidence of fixation to the wall of the middle ear cavity, congenital stapes fixation should be considered as the cause of the conductive hearing deficit. This is due to the failure of differentiation of the lamina stapedialis into the annular ligament, leading to footplate ankylosis. Since the footplate cannot be discerned from adjacent annular ligament even on HRCT, it is often not possible to make a definite diagnosis of footplate ankylosis.² Only when the footplate is physically thickened can such a diagnosis be made on HRCT. Fenestral otosclerosis would then be a differential diagnosis. However, the lack of a lucent otosclerotic plaque in the anterior oval window region and the young age of the patient would make a congenital cause more likely.

Oval Window Atresia

This is an uncommon embryologic defect, defined by the absence of structure, with the presence of a bony plate sandwiched between the vestibule and the middle ear.¹⁰ Imaging in the coronal plane best demonstrates this entity. The bony prominence over the cochlear promontory and

under the lateral semi-circular canal results in a tapered configuration and a dimple-like depression on the medial wall of the middle ear, centred at the oval window region.¹¹ Under normal circumstance, the air within the middle ear cavity is separated from the vestibule by a very thin bony plate, representing the stapes footplate. In oval window atresia, the air in the middle ear cavity is separated from the vestibule by a thick bony plate (Fig. 8). A normal-appearing stapes is not seen in the oval window region. The stapes is either absent entirely, dysplastic or displaced, usually inferiorly and posteriorly.¹⁰ There is an associated anomalous course of the tympanic portion of the facial nerve. In oval window atresia, this portion of the facial nerve either courses through the expected position of the oval window or lies on top of the cochlear promontory (Fig. 8).^{10,11} There is also a high incidence of associated malformation of the incus. The lenticular process of the incus could be deformed or absent.^{10,11}

In normal embryological development, the oval window is formed when the rudimentary stapes grows medially and makes contact with the otic capsule in the 7th week of gestation. If the primitive stapes fails to fuse with the primitive vestibule, the oval window cannot develop, thereby resulting in an atresia. One widely accepted theory to explain the failure of fusion between the primitive stapes and primitive vestibule is the abnormal anterior and inferior displacement of the tympanic portion of the facial nerve during the 5th to 6th weeks of gestation. This results in the facial nerve lying between the primitive stapes and the primitive vestibule, thus preventing their fusion and leading to oval window atresia.¹⁰

Round Window Atresia

An extremely rare congenital abnormality, it is associated with congenital stapes fixation.¹² The hearing deficit is

usually about 30 dB to 40 dB. It is not certain how much round window atresia contributes to the hearing deficit since there is still a relative lack of understanding in auditory contribution by a normal round window. However, stapedectomy in patients with round window atresia associated with stapes fixation usually yields poor results, suggesting that round window atresia probably plays a significant part in the hearing loss. On HRCT, a normal air-filled round window niche is not seen in round window atresia. Instead, an occlusion or cleft-like defect is demonstrated in the expected location of the round window.¹²

Embryologically, the round window starts to form in the 11th week of gestation, where a condensation of embryonic connective tissue appears. When the otic capsule ossifies, a cartilage is formed in the location of the future round window niche to prevent ossification of its opening. Subsequently, this cartilage ring differentiates into the round window. If the cartilage ring does not develop, there is osseous obliteration of the future round window niche, leading to an atresia.¹²

Conclusion

Congenital middle ear deafness must be considered if a child or an adolescent with normal-appearing external ears presents with a conductive hearing deficit, especially if there is no prior history of ear discharge or trauma. A proper imaging technique using HRCT can demonstrate the cause of the hearing deficit in most cases. Analysis of the CT images starts with the exclusion of two non-congenital causes of middle ear deafness: fenestral otosclerosis and chronic otomastoiditis. The former is rare in this age group and the absence of a lucent otosclerotic plaque in the anterior oval window region (Fig. 9) would exclude the diagnosis. The latter is easily excluded by the absence of sclerotic mastoid bone, the presence of well pneumatized air cells and the absence of inflammatory debris or fluid in the middle ear cavity and mastoid air cells (Fig. 10).

Following the exclusion of these 2 conditions, a systematic analysis of the middle ear structures is essential. The configuration of the ossicles and status of the incudostapedial and malleo-incudal joints are analysed. Calcified/ossified supporting ligaments and tendons must be excluded. Any

abnormal bony bars fixing the ossicles to the wall of the middle ear cavity should also be noted. The configuration of the oval window, round window and round window niche should be evaluated. The course of the tympanic portion of the facial nerve must be analysed, as an anomalous course could be a clue of oval window atresia.

Finally, if a systematic and careful analysis of the above structures yields a normal scan, the cause of the conductive hearing deficit would most likely be congenital ankylosis of the stapes footplate.

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