High-resolution computed tomography of isolated congenital anomalies of the stapes: A pictural review using oblique multiplanar reformation in the ‘‘axial stapes’’ plane

Tomodensitométrie haute résolution des anomalies congénitales du stapes : revue iconographique des différents aspects dans le plan « axial stapes »

Jean-Christophe Gentric\textsuperscript{a,}\textrm{*}, Jean Rousset\textsuperscript{b}, Marc Garetier\textsuperscript{b}, Douraied Ben Salem\textsuperscript{a}, Philippe Mériot\textsuperscript{a}

\textsuperscript{a} Department of Neuroradiology, Cavale Blanche Brest University Hospital, boulevard Tanguy-Prigent, 29200 Brest, France
\textsuperscript{b} Department of Radiology, Clermont-Tonnnerre Hospital, rue Colonel-Fonferrier, 29019 Brest, France

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Computed tomography; Ears ossicles; Stapes; Congenital abnormalities; Middle ear; Auditory ossicles

**Summary**

**Purpose:** Isolated congenital anomalies of the stapes are infrequent but highly variable. The goal of this study is to present the numerous observed anomalies based on a large number of cases, and to describe anatomical variations and malformations of the stapes using high-resolution computed tomography (CT), after proper reorientation in the ‘‘axial stapes’’ plane.

**Materials and methods:** The 1805 CT of temporal bones performed during the past 5 years have been retrospectively studied. After reconstructing the images in the stapes axial plane, the ears presenting a congenital anomaly of the stapes were included in this study. All the ears with acquired lesions were excluded. The anomalies have been sorted according to the affected part of the stapes: the superstructure, the footplate or the obturator foramen. Two neuroradiologists classified the anomalies as either anatomical variation, malformation, or undetermined.

**Results:** Sixty-one stapes in 47 patients were found to have one or more congenitally abnormal shapes (bilateral anomalies were found in 14 of these patients). The abnormal part of the stapes was the superstructure in 17 cases, the footplate in 13 cases, the obturator foramen in 19 cases (with a high frequency of ‘‘double stapes’’ shape) while in 12 cases multiple parts were affected.

\* Corresponding author. Tel.: +33 2 98 34 74 87; fax: +33 2 98 34 78 24. E-mail address: jean-christophe.gentric@chu-brest.fr (J.-C. Gentric).
Introduction

The stapes is the smallest bone of the human body. As a part of the ossicle chain, it transmits vibrations from the tympanic membrane to the cochlea in the inner ear. The stapes grows at the cranial end of the second branchial arch. Its footplate originates from the superior part of the stapedial anlage whereas the inferior part of this anlage will constitute the limbs and the head of the stapes. The development of the otic capsule remains independent. The stapes ossification is initiated by a solitary ossification center on the footplate that secondly spreads toward the crura and the head [1–3]. The ossification occurs on the obturator surface and later to the external surface. Then, vacuolation progressively erases the obturator surface and remodels the stapes to its final shape.

Dissection works showed that the stapes present considerable morphological variations [4–7], but high-resolution CT scanners have more recently made the non-invasive study of the stapes available in patients. Swarz et al. focused on CT imaging of normal anatomy and anomalies of the temporal bone [8–16]. More recently, Hagr et al. studied human cadaver stapes footplate using very high-resolution CT to explore minor details of shape and orientation of the footplate [17]. Other authors as Trojanowska et al. compared 2D and 3D reconstructions and virtual endoscopy in assessment of the middle ear [18]. Blanco et al. highlighted the advantages of oblique multiplanar reformation in multislice temporal bone CT for the analysis of various structures such as the facial nerve, the cochlear basal turn and the ossicles. These useful reformations provided biometric landmarks in pathological conditions [19,20].

The acquired lesions of the stapes are well known and have been widely described [21]. Even if isolated congenital affections of the stapes are infrequent, the ossicle is the most commonly congenitally affected middle ear structure [1,22–24]. In this work, we retrospectively analyzed congenital anomalies as malformations and variations of the stapes using "axial stapes reformation".

Materials and methods

Data acquisition

CT with isolated congenital stapes anomaly were searched for retrospectively. These CT were part of the 1805 temporal bones CT performed in our institution for various indications between January 2005 and April 2011. Standardized acquisition protocol was used on two Siemens multislices CT scans: the VZ "Volume Zoom" for the first three years (four rows CT system) and the "Somatom AS+" (64×2 rows CT system) for the two remaining years (Siemens healthcare, Forchheim, Germany). All acquisitions were made in the axial plane. The patients were positioned in supine decubitus and scans were oriented parallel to the orbitomeatal line. The acquisition parameters were 140 kV, 350 mAs for adults and 120 kV, 180 mAs for children. The other parameters were a 0° gantry tilt, slice width 0.6 mm and a reconstruction algorithm H90s. The mean dose length product was 350 mGy.cm.

Data processing

Images were displayed using window width of 4000 and window level of 700. A Leonardo Workstation (Siemens healthcare, Forchheim, Germany) was used for reformation. The reformatted images consisted of overlapping sections of 0.6 mm each 0.4 mm in the stapes axial plane. Nine or ten slices were placed to analyze the stapes. To obtain the stapes axial plane, a triple orientation was required. First, the axial plane was placed in the plane of the semi-circular canal. Secondly, the coronal oblique plane was obtained after placing the slices perpendicular to the footplate on the axial plane. In this plane, the V-shape formed by the incudo stapedial articulation and the stapes was visible. Thirdly, the "axial stapes" plane was obtained in the coronal oblique plane, the slices were placed parallel to the stapes crura (Fig. 1). This plane contained the superstructure and the major axis of the footplate (Fig. 2).

Inclusion and exclusion criteria

The only inclusion criterion was the presence of an unusual aspect of the stapes including morphologic and morphometric anomalies. The exclusion criterion was any acquired affection of the ear, or the presence of another malformation of the ear. An anomaly of the long process of the incus was not an exclusion criterion because the stapes and the long process of the incus both originated from the second branchial arc. For each ear audiometric data were kept for analysis and were divided in "normal hearing" (NH) or "hearing loss" (HL).

Data analysis

To classify the shapes of the stapes, the ossicle has been divided into three parts: the superstructure, the obturator foramen and the footplate. The head and the crura of the stapes make the superstructure, the space between the footplate and the crura demarcates the obturator foramen. When the anomaly affected more than one of those parts, the anomaly was classified as "multiple". After the screening of congenital stapes anomalies, two neuroradiologists (PM, DBS) classified anomalies based on consensus. The used criteria to class the abnormal stapes as malformation
or anatomical variation was morphologic and morphometric. The various criteria were the absence of part(s) of the superstructure, one supplementary abnormal structure or a thickening of one constituent as the footplate. When to classify the anomaly either as a malformation or an anatomical variation was uncertain, it was ranked as “undetermined”.

Results

Of the 1805 CT of temporal bones with reconstructions in the axial stapes plane, 61 ears in 47 patients presented a pattern of isolated congenital anomaly of the stapes. In 16 cases, the anomaly was on the left side, in 17 cases on the right side and bilateral in 14 cases. The younger patient was 5 years old and the older one 67 years old.

As shown in Fig. 3, the superstructure was affected alone in 17 cases (27.8%), the footplate in 13 cases (21.3%) and the obturator foramen in 19 cases (31.2%). In 12 cases, the anomalies affected at least two of the three parts of the stapes (19.7%).

Regarding the congenital anomalies of the superstructure, the head was affected in one case, the anterior crus in three cases, the posterior crus in seven cases (Fig. 4). Both crura were affected in six cases. Among those 17 anomalies of the superstructure, nine were classed as malformations, three as anatomical variations and five as “undetermined”. The observed variations were crus fenestration (n = 2) and length variation of crus (n = 1). The encountered malformations were crus aplasia (n = 2), head aplasia (n = 1), crura hypoplasia (n = 3) and three anomalies of the junction between the crus and the footplate. The undetermined anomalies were crus dehiscence (n = 2) and shape variation of the superstructure (n = 3). Four of 17 patients presented normal hearing whereas others presented audiometric anomalies (Table 1).

Concerning the footplate, seven malformations, four variations and two undetermined patterns were observed. The variations were the thickening of the union between the crus and the footplate, the concavity or the convexity of the footplate area (Fig. 5). A hypoplasia of the footplate was demonstrated in seven cases. The footplate thickenings

**Figure 1** Coronal oblique plane shows the “V”-shape outlined by the ossicles (A). Nine or ten slices are obtained parallel to the stapes crura to obtain the stapes in the “axial stapes” plane. Reformatted images consisted of overlapping sections of 0.6 mm each 0.4 mm in the stapes axial plane (B).

**Figure 2** Normal stapes in the axial stapes plane (A, B). Stapes head (white arrow), anterior crus (white arrowhead), posterior crus (double white arrowhead), footplate (black arrow).

**Figure 3** Number and fractions of congenital anomalies observed according to their location.
Figure 4  Congenital stapes superstructures anomalies (A–D); absence of stapes head (A). Monopodal stapes with aplasia of anterior crus and large posterior crus (B). Hypoplasia of both crura without contact with the footplate (C). Anterior crus anomaly without contact with the footplate (D).

Table 1  Classification of stapes anomalies and consequences on the hearing.

<table>
<thead>
<tr>
<th>Superstructure</th>
<th>Footplate</th>
<th>Obturator foramen</th>
<th>Multiple</th>
<th>Total</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>NH</td>
<td>HL</td>
<td>NH</td>
<td>HL</td>
</tr>
<tr>
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<td>1</td>
<td>4</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Sub Total</td>
<td>4</td>
<td>13</td>
<td>1</td>
<td>12</td>
</tr>
</tbody>
</table>

NH: normal hearing; HL: hearing loss.

were classified as “undetermined” in two cases. Twelve of 13 patients presented audiometric anomalies (Table 1).

For the obturator foramen, one shape variation was encountered and the other 18 others were classified as “undetermined”. These anomalies all consisted of a “double stapes” pattern due to the presence of a linear hyperdensity within the obturator foramen (Fig. 6). This “third branch” was located between the anterior crus and the footplate (n=2), between the posterior crus and the footplate (n=12) or between the neck and the footplate (n=3). In one case, a third branch appeared in front of the anterior crus. Ten of 19 patients presented audiometric anomalies for the concerned ears (Table 1).

Figure 5  Congenital footplate anomalies (A, B). Footplate thickening on posterior part of the footplate (A). Footplate hypoplasia with bony band (B).
Multiple anomalies were observed in 12 cases (Fig. 7), ten cases were classified as malformations and two as "undetermined". Eight of 12 patients presented audiometric anomalies (Table 1).

Discussion

The analysis of the stapes is difficult due to the thinness of its parts [7,25], and specific acquisition and reconstruction protocols with appropriate parameters are required. It can be the only affected ossicle. Anomalies of the other ossicles or the outer and inner ear structures can be associated in complex malformations. In numerous studies, the average length of the entire bone was about 3.2 mm, while the average breadth was about 2.2 mm [6,7]. The shape of the obturator foramen is most often triangular. The axial stapes plane is perpendicular to the footplate and allows better appreciation of the thickness of the footplate by limiting the zoom effects related to CT detectors. In this plane, the footplate mean thickness is about 0.3 mm whereas it is 0.4 mm in the axial standard plane.

Congenital isolated anomalies of the stapes are infrequent [26]. We observed 61 congenital anomalies on the 3610 imaged ears (1.69%). The diagnostic can be clinically suspected on non-progressive transmission deafness. These anomalies are often bilateral and 14 of the 47 patients of this study were affected bilaterally. According to the literature, the congenital fixation of the footplate is the most frequent malformation [1,22–24,26]. This malformation is not visible in CT. The other isolated malformations of the stapes are infrequent but conspicuous on CT images because they affect the superstructure and/or the obturator foramen. In this retrospective study, we focused on these ones.

Seventeen congenital anomalies of the superstructure were encountered: nine malformations, three variations and five undetermined anomalies. Various shapes of the stapes head have been described; the shape can be oval, irregular, circular or it can take more rare patterns. The most described stapes head malformation is its absence [23,24,26]. However, this anomaly is rare and only one patient presented it in this study. Concerning the crura, their

length has been studied and the average value is about 2 mm for both. Posterior crus tends to be more curved and a little wider than the anterior one. The most described stapes malformations are crus aplasia or hypoplasia [23, 24, 26], five patients presented such an anomaly. Five other patients presented an anomaly of the junction between crus and footplate. These kinds of anomalies are more difficult to analyze than aplasia or hypoplasia of the crura. It can be helpful to reconstruct in a plane perpendicular to the stapes crura to analyze the medial part of the abnormal branch. Two patients presented patterns of discontinuity of one branch: in these cases, it is necessary to be aware of artifacts and traumatic history.

Thirteen congenital anomalies of the footplate were seen: seven malformations, four variations and two undetermined anomalies. Two variations have been found, the first observed variation was the thickening of the footplate near the junction of the crura in one case. For this kind of variation, the mean thickness in front of the anterior crus is about 0.37 mm [25], whereas the mean thickness in front of the posterior crus is about 0.4 mm [25]. It is sometimes difficult to separate a normal variant thickening of the footplate especially in front of the crus of a malformed thickening of the footplate. Pathological thickening of the footplate can be suspected when the middle part of the footplate is larger than 0.5 mm in this plane. The second variation is various patterns of concavity or convexity of the footplate in three cases. In the literature, the medial surface is most often concave and rarely flat [6]. The most frequently observed malformation is the footplate hypoplasia. Five patients presented this kind of anomaly in the study and they all presented hearing loss. Anatomic studies demonstrated that normal length of the footplate is about 2.8 ± 0.2 mm. Theses values are consistent with a footplate hypoplasia [6]. The use of reformation in the “axial stapes” plane allows appreciating the length of the footplate to search for footplate hypoplasia.

Twelve of the congenital anomalies observed affected the superstructure and the footplate. Theses anomalies are often bilateral (n = 3) and most of them (n = 10) were classified as malformations. Most of them presented hearing loss (8/12).

In this study, 19 abnormal congenital patterns of the obturator foramen were observed. Ten of 19 patients presented audiometric anomalies. Numerous variations of the obturator foramen shape are known without pathologic meaning: equilateral or isosceles triangles, triangle with unequal sides, oval, circular, irregular, kidney shaped, absent and double. All these variations have certainly not been underlined in this study because the observer considers them as normal. Most of the abnormal patterns noticed are the presence of a linear hyperdensity within the obturator foramen lying between the superstructure and the footplate. The radiological, and surgical literatures have scarcely addressed this anomaly of the obturator foramen. Park et al. highlighted the possibility of a malformation consisting in the closure of the obturator foramen [23, 24]. Dass et al. underlined the possibility of the presence of a foramen at the base of the posterior crus [7]. Concerning the pathological significance of this anomaly, the literature is poor and these radiological findings raise the question of whether this abnormality is a normal variation or a malformation.

The limits of this study are, the monocentric approach, the subjectivity of criteria used to classify the anomaly. The clinical significance of various patterns and especially the “double stapes” pattern is not evident and need to be complete by audiometric studies.

Conclusion

Oblique multiplanar reformations of temporal bones CT enhance diagnostic performances especially for minor anomalies of ossicles as congenital affections of the stapes. The “axial stapes” plane is appropriate for a morphologic and morphometric global analysis of the superstructure, the obturator foramen and the footplate.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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References


