

## Reclassification of inner ear malformations according to the updated Sennaroğlu classification: which categories change?

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### ABSTRACT

**Aims:** To determine how many ears change category, and which categories are affected, when a cochlear implant (CI) candidate cohort classified in 2009 is reassessed according to the up-to-date Sennaroğlu classification.

**Methods:** Preoperative temporal bone computed tomography studies of 100 consecutive CI candidates (200 ears) examined between 2006 and 2008 were re-evaluated, and the measurements repeated, by the radiologist who had made the original assessment, with the earlier category concealed. Ears were re-categorized according to Sennaroğlu's current 2017 classification.

**Results:** A congenital cochlear malformation was present in 38 ears (19.0%) of 20 patients. Of the 200 ears, 176 (88.0%) kept a category with a direct equivalent in the 2017 scheme, 12 were subtyped within cochlear hypoplasia (CH) and 2 remained unclassifiable because of labyrinthine ossification; no ear was reassigned to a different entity. Ten ears with an isolated abnormality of the vestibule or the semicircular canals could not be categorized under the 2017 scheme. A subdivision of CH applied locally in 2009 mapped onto the current subtypes without crossover. At a threshold of 3.0 mm, all 8 ears with a narrow internal auditory canal in the coronal plane had a malformed cochlea; the axial plane performed less consistently at every threshold tested.

**Conclusion:** Following reclassification based on current criteria, redistribution was limited to cochlear hypoplasia, but ten ears were left without a category. Because the boundaries between some cochlear malformations are defined descriptively rather than numerically, their applicability may be limited.

**Keywords:** Cochlea, temporal bone, cochlear implantation, computed tomography, sensorineural hearing loss

### INTRODUCTION

Congenital sensorineural hearing loss (SNHL) is most often caused by membranous malformations that leave the bony labyrinth intact and cannot be demonstrated radiologically. In the remaining fifth of cases the bony labyrinth is involved, and these malformations can be shown by computed tomography (CT) and magnetic resonance imaging.<sup>1</sup> Because this group presents both surgical difficulties and problems in decision making, its preoperative recognition is a routine part of the assessment of cochlear implant (CI) candidates.<sup>1</sup>

The category is not merely descriptive: it guides electrode choice, signals the risk of cerebrospinal fluid gusher and facial nerve injury, and bears on the choice between cochlear and auditory brainstem implantation and on the expected outcome.<sup>1,2</sup> A change in category is therefore a change in clinical meaning.

Classification schemes have evolved over time: Jackler et al.<sup>3</sup> first grouped these malformations by the stage of arrested embryogenesis, Sennaroğlu and Saatci<sup>4</sup> later divided incomplete partition (IP) into types I and II (IP-I and IP-II),<sup>4</sup> and the scheme was revised in 2010 and again in 2017.<sup>1,5</sup> In the earlier version, cochlear malformations were classified into six groups, while the vestibule, the semicircular canals (SCCs), the internal auditory canal (IAC) and the aqueducts were assessed on separate axes.<sup>4</sup> The current version brings all of these under a single heading of inner ear malformations, in eight categories, and adds subgroups such as IP type III (IP-III) and four subtypes of cochlear hypoplasia (CH), designated CH-I to CH-IV. Cochlear aperture (CA) abnormalities are included as a category of their own.<sup>1</sup> Other schemes have been proposed, but the Sennaroğlu classification remains the most widely accepted.<sup>6-8</sup>

These revisions have a consequence that is rarely addressed directly. A large body of work published before 2017, including prevalence figures, surgical series, and audiological correlations, was categorized under earlier schemes. If the boundaries between categories have moved, the comparability of that work with contemporary reports cannot simply be assumed. To our knowledge, the extent to which a cohort classified under the earlier scheme is redistributed when the current criteria are applied has not been quantified.

We therefore reexamined the preoperative temporal bone CT studies of a CI candidate cohort that had been classified in 2009, applying the 2017 criteria, in order to determine how many ears change category and which categories are affected.

## METHODS

### Ethics

This study was approved by the Gaziantep University Medical Ethics Committee of the Faculty of Medicine, (Date: 19.02.2009, Decision No: 02-2009/20). All procedures were carried out in accordance with the ethical rules and the principles of the Declaration of Helsinki.

### Study Design and Patients

This retrospective study was carried out at a tertiary referral center. Consecutive patients assessed by the department of otorhinolaryngology, referred as CI candidates and examined by temporal bone CT in our department between January 2006 and December 2008 were included. All had bilateral severe to profound SNHL. Patients were excluded if their imaging could not be retrieved from the archive or if the CT had been performed at another institution. Examinations degraded by motion were repeated at the time, so no study was excluded on grounds of image quality. Ears with acquired pathology were retained, and the acquired finding was recorded separately from the congenital category. The final cohort comprised 100 patients (200 ears), 52 male and 48 female, aged 1 to 36 years (mean 4.9; median 3.0). Ninety-six patients were younger than 18 years. Adults were retained because the unit of analysis is the ear rather than the patient.

The cohort and its original radiological assessment were reported in an academic thesis in 2009.<sup>9</sup> The present study reanalyzes the same imaging material under a different classification system, using the original findings only as the comparator; the reclassification reported here has not been published previously.

### Imaging Technique

CT was performed on a six-detector-row scanner (Brilliance CT; Philips Medical Systems, Best, The Netherlands), with sedation where required. The temporal bones were scanned continuously in the axial plane at 120 kV and 250 mAs, and images were reconstructed with a bone algorithm at 0.8 mm section thickness.

To allow consistent comparison between sides, axial reformats were generated with both inner ears at the same level and with the anterior and posterior limbs of the lateral SCC visible in a single section. Coronal reformats were then obtained perpendicular to this plane, at 0.8 mm section

thickness. Images were reviewed at a window width of 4095 and a window level of 600.

### Image Analysis

For the present study all CT examinations were re-evaluated and all measurements were repeated by a single radiologist with 17 years of experience in temporal bone imaging, who had also performed the original assessment in 2009. The earlier category was concealed until the new category had been recorded.

### Classification

Ears were classified according to the eight categories of Sennaroğlu and Bajin.<sup>1</sup> Acquired pathology was recorded separately from the congenital category, so that an ear could carry both.

In ears with labyrinthine ossification, a congenital category was assigned when cochlear morphology, that is the number of turns and the presence of the modiolus and the interscalar septa, remained assessable; when ossification obscured these structures the ear was recorded as unclassifiable.

We drew the boundary between CH-II and IP-I at a cochlear height of 4.4 mm, the lower limit of the normal range reported in a normative series;<sup>10</sup> this threshold was adopted for the present study.

### Measurements

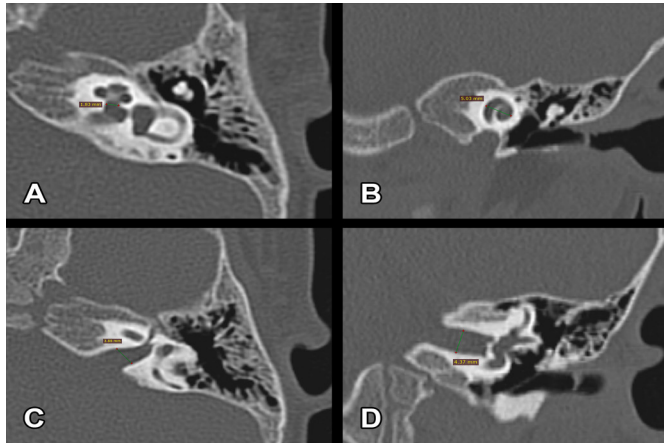
The CA,<sup>11,12</sup> the cochlear height<sup>10,13</sup> and the IAC<sup>1,14</sup> were measured as previously described and as illustrated in **Figure 1**. Reported thresholds differ. For the CA we used 1.5 mm, as in recent reports from the group that proposed the classification,<sup>11,12</sup> rather than the 1.4 mm stated in the classification itself;<sup>1</sup> a CA was recorded as aplastic when no bony cochlear nerve canal could be identified. For cochlear height we used the operational threshold of 4.4 mm described above,<sup>10</sup> a value that does not require adjustment for age.<sup>13</sup> For the IAC, criteria differ in both the value and the site of measurement, so four values were applied in both planes as a sensitivity analysis rather than as validated criteria for either plane: 2.0 mm,<sup>15</sup> 2.5 mm,<sup>1</sup> 3.0 mm<sup>9</sup> and 3.2 mm.<sup>14</sup> Because the CA measurement alone determines a category, it was obtained twice in separate sessions and the mean of the two values was used.

The vestibular aqueduct (VA) was considered enlarged when it appeared wider than the neighboring posterior SCC on an axial section displaying both.<sup>14</sup> Ears in which the aqueduct was not visualized were recorded separately and analyzed as not enlarged.

The vestibule, the SCCs and the ossicular chain were assessed qualitatively.

### Comparison Between Classifications

The scheme used in the original assessment comprised six categories of cochlear malformation: Michel deformity, cochlear aplasia, common cavity (CC), CH, IP-I and IP-II.<sup>4,9</sup> Categories were treated as concordant when the earlier label had a direct equivalent in the current system; ears originally



**Figure 1.** Measurement techniques

(A) Cochlear aperture, measured in the axial plane at the mid-modiolar level from the inner margins of its bony walls. (B) Cochlear height, measured in the coronal plane on the section of greatest height showing both the basal and the apical turn, between the centers of these two turns. (C) Internal auditory canal, measured at its midpoint in the axial plane. (D) Internal auditory canal, measured at its midpoint in the coronal plane.

assigned to CH, a single group at that time, were recorded as subtyped rather than changed.

The original assessment had also used a local subdivision of CH according to the degree of development of the internal architecture, and separate local categories for an isolated hypoplastic vestibule and for isolated dysplastic SCCs.<sup>9</sup> None of these is part of a published classification, and they are reported here as secondary observations.

### Statistical Analysis

Categorical data are reported as counts and percentages, and continuous data as mean with standard deviation and range. Agreement between the two CA measurements was assessed with the intraclass correlation coefficient (ICC; two-way random effects, absolute agreement, single measures) and

the Bland-Altman method, excluding ears in which no canal could be identified. Because both ears of every patient were included, ear-level proportions are reported descriptively. Analyses were performed with SPSS Statistics 20.0 (IBM Corp., Armonk, NY, USA).

## RESULTS

### Cohort

A congenital cochlear malformation was present in 38 ears of 20 patients (19.0% of ears, 20.0% of patients). Acquired pathology was recorded in 9 ears: labyrinthitis ossificans in 7 and otosclerosis in 2.

### Redistribution Between the Two Classifications

Of the 200 ears, 176 (88.0%) kept a category with a direct equivalent in the 2017 scheme, and no ear was reassigned to a different entity (Table 1).<sup>9</sup> The 12 ears grouped as CH in the earlier scheme were distributed across all four current subtypes: CH-I in 1 ear, CH-II in 1, CH-III in 8 and CH-IV in 2 (Figure 2). Two ears remained unclassifiable because labyrinthine ossification obscured cochlear morphology.

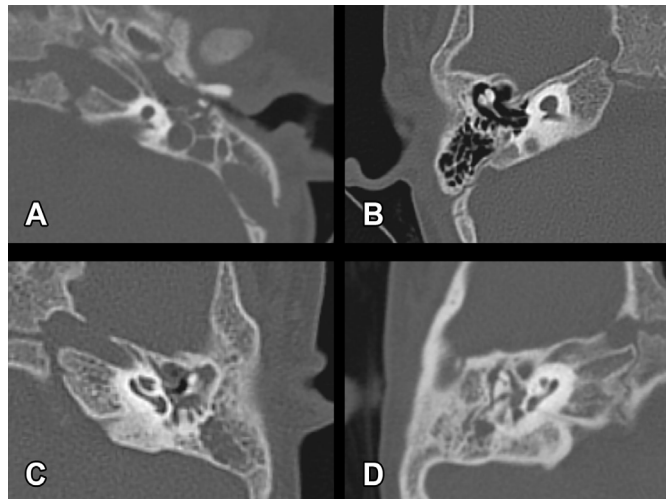
Ten further ears had been recorded as normal under the earlier scheme, which classified cochlear malformations, but have no category in the current one, which covers inner ear malformations: each had an isolated abnormality of the vestibule or the SCCs.

None of the entities added in the 2017 revision was identified in this cohort. There were no ears with rudimentary otocyst, IP-III, isolated enlarged VA or isolated CA abnormality. Every ear with an abnormal CA also had a cochlear malformation that had already been recognized in the original assessment.<sup>9</sup>

**Table 1.** The 200 ears distributed under the 2002 and the 2017 classifications, with the local subdivision used in 2009 and the modifications suggested here

| 2002 scheme    |     | 2009 local subdivision            |     | 2017 scheme    |             |     | Suggested modification |             |     |
|----------------|-----|-----------------------------------|-----|----------------|-------------|-----|------------------------|-------------|-----|
| Category       | n   | Category                          | n   | Category       | Subgroup    | n   | Category               | Subgroup    | n   |
| MD             |     | MD                                |     | MD             | 3 subgroups |     | MD                     | 3 subgroups |     |
|                |     | Not in scheme                     |     | RO             | -           |     | RO                     | -           |     |
| CAP            |     | CAP                               |     | CAP            | 2 subgroups |     | CAP                    | 2 subgroups |     |
| CC             | 1   | CC                                | 1   | CC             | -           | 1   | CC                     | -           | 1   |
|                |     | Type 1                            | 2   |                | CH-I        | 1   |                        | CH-I        | 1   |
| CH             | 12  |                                   |     | CH             | CH-II       | 1   | CH                     | CH-II       | 1   |
|                |     | Type 2                            | 10  |                | CH-III      | 8   |                        | CH-III      | 8   |
|                |     |                                   |     |                | CH-IV       | 2   |                        | CH-IV       | 2   |
| IP-I           | 11  | IP-I                              | 11  | IP-I           | IP-I        | 11  |                        | IP-I        | 11  |
| IP-II          | 12  | IP-II                             | 12  | IP             | IP-II       | 12  | IP                     | IP-II       | 12  |
|                |     | Not in scheme                     |     |                | IP-III      |     |                        | IP-III      |     |
|                |     | Categorized under another heading |     | EVA            | -           |     | IEVA*                  | -           |     |
|                |     | Not in scheme                     |     | CAA            | -           |     | ICAA*                  | -           |     |
|                |     | Categorized under another heading |     | No category    |             | 10  | IVSA**                 | -           | 10  |
| Normal         | 162 | Normal                            | 162 | Normal         |             | 152 | Normal                 |             | 152 |
| Unclassifiable | 2   | Unclassifiable                    | 2   | Unclassifiable |             | 2   | Unclassifiable         |             | 2   |
| Total          | 200 | Total                             | 200 | Total          |             | 200 | Total                  |             | 200 |

Category gives the name of the category in each scheme, and Subgroup the subgroup where a scheme subdivides it; n is the number of ears assigned. Blank cells indicate that no ear was assigned, and a dash that the scheme defines no subgroup. "Not in scheme" indicates a category that did not exist at that time, "categorized under another heading" a structure assessed in 2002 outside the list of cochlear categories, and "no category" ears for which the 2017 scheme provides none. Italics indicate entries that are not part of the corresponding published scheme; \* denotes a suggested change of name and \*\* a suggested new category. n: Number of ears, MD: Michel deformity, RO: Rudimentary otocyst, CAP: Cochlear aplasia, CAA: Cochlear aperture abnormalities, CC: Common cavity, CH: Cochlear hypoplasia, CH-I to CH-IV: Cochlear hypoplasia types I to IV, EVA: Enlarged vestibular aqueduct, ICAA: Isolated cochlear aperture abnormalities, IEVA: Isolated enlarged vestibular aqueduct, IP: Incomplete partition, IP-I to IP-III: Incomplete partition types I to III, IVSA: Isolated vestibular and/or semicircular canal abnormality



**Figure 2.** Cochlear hypoplasia subtypes on axial or axial-oblique computed tomography, one ear of each subtype

(A) CH-I, a bud-like cochlea in which no internal architecture can be recognized. (B) CH-II, a small cochlea with defective internal architecture; the ossicles within the field of view are also dysplastic. (C) CH-III, a proportionally small cochlea. (D) CH-IV, a normal basal turn with hypoplastic middle and apical turns. CH-I to CH-IV: Cochlear hypoplasia types 1 to IV

### Local Subdivision of Cochlear Hypoplasia

In the 2009 assessment the hypoplastic ears had been divided locally into those in which the internal architecture was absent (type 1) and those in which it was partially developed (type 2).<sup>9</sup> This subdivision mapped onto the current subtypes without overlap (Table 1). Both type 1 ears became CH-I or CH-II, and all 10 type 2 ears became CH-III or CH-IV. No ear crossed between the two pairs.

### Cochlear Aperture

The CA could be measured in 198 ears. In the remaining 2 ears, both belonging to the same patient, no bony cochlear nerve canal could be identified and the CA was recorded as aplastic. The mean of the two measurements was 1.94 mm (SD 0.22, range 0.83 to 2.60). Agreement between them was good, with an ICC of 0.87, a mean difference of -0.01 mm and 95% limits of agreement of -0.24 to 0.22 mm.

In 5 ears (2.5%) the CA measured below 1.5 mm and was classified as hypoplastic. Together with the 2 aplastic apertures, 7 ears met the current criteria for a CA abnormality. These were the same 7 ears in which a narrow CA had been reported in the original assessment.<sup>9</sup> All 7 belonged to patients in whom a cochlear malformation had already been recognized.

### Cochlear Height

Cochlear height could be measured in 197 ears. It was not measurable in 2 ossified ears and in 1 ear with a CC. The mean was 4.96 mm (SD 0.45, range 2.90 to 6.00). Twelve ears fell below 4.4 mm, and all 12 belonged to the CH group identified in the original assessment.<sup>9</sup>

### Internal Auditory Canal

The mean mid-portion width was 4.48 mm (SD 0.92, range 2.10 to 8.80) in the axial plane and 4.69 mm (SD 1.04, range 1.50 to 8.80) in the coronal plane.

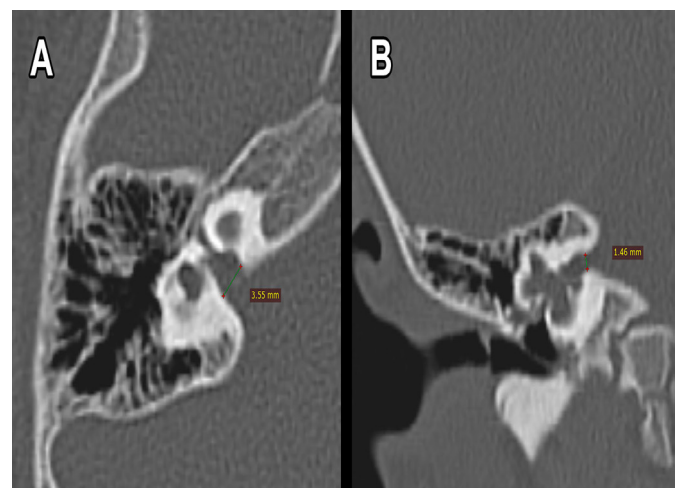
The two planes did not identify the same ears (Table 2). In the coronal plane, every ear that measured below 3.0 mm had a malformed cochlea, and only at 3.2 mm did 2 of the

**Table 2.** Ears identified as having a narrow internal auditory canal at four thresholds, in each plane

| Threshold | Coronal plane narrow IAC (mal; norm) | Axial plane narrow IAC (mal; norm) | Both planes narrow IAC (mal; norm) |
|-----------|--------------------------------------|------------------------------------|------------------------------------|
| 2.0 mm    | 1 (1; 0)                             | 0 (0; 0)                           | 0 (0; 0)                           |
| 2.5 mm    | 6 (6; 0)                             | 2 (2; 0)                           | 1 (1; 0)                           |
| 3.0 mm    | 8 (8; 0)                             | 7 (4; 3)                           | 4 (4; 0)                           |
| 3.2 mm    | 11 (9; 2)                            | 11 (4; 7)                          | 4 (4; 0)                           |

Values in parentheses give the number of ears with and without a cochlear malformation. Measurements were made at the mid-portion of the canal in the axial and coronal plane. The thresholds are the most widely used criterion (2.0 mm), the value stated in the 2017 classification (2.5 mm), the criterion of the original assessment (3.0 mm), and two standard deviations below the normative coronal mid-portion mean (3.2 mm). None of the ears listed had acquired pathology, and every ear counted as having a normal cochlea also had a normal vestibule and normal semicircular canals. IAC: Internal auditory canal, mal; ears with a cochlear malformation, norm, ears with a normal cochlea

11 ears identified have a normal one. In the axial plane, fewer malformed ears were identified at every threshold, and ears with a normal cochlea appeared earlier and in greater number. At 3.0 mm, 8 ears were narrow in the coronal plane and 7 in the axial plane, but only 4 were narrow in both. In one patient with bilateral IP-I the canal exceeded 3.0 mm in the axial plane on both sides but was narrow in the coronal plane (Figure 3).



**Figure 3.** Internal auditory canal in the right ear of a patient with bilateral incomplete partition type I

In the axial plane (A) the canal appears unremarkable, whereas in the coronal plane (B) the same canal is narrow. Measurement confined to the axial plane would not have identified this canal as narrow

### Accompanying Findings

The vestibule was normal in 166 ears, dilated in 15, hypoplastic in 14 and absent in 2, and formed part of a CC in 1 ear. In 1 ear it was considered probably normal despite partial ossification, and in 1 ear ossification prevented assessment. The SCCs were normal in 165 ears, dysplastic in 26 and aplastic in 2; 3 ears were considered probably normal despite partial ossification, and in 4 ears ossification prevented assessment. The VA was enlarged in 15 ears of 9 patients, all of which had a malformed cochlea (IP-II in 12 and IP-I in 3), and was not visualized in 5 ears.

Ten ears in 5 patients had an isolated abnormality of the vestibule or of the SCCs with a normal cochlea: A hypoplastic vestibule in 4 ears and dysplastic SCCs in 6.

Ossicular chain malformation was present in 9 ears of 5 patients, 7 of them in ears with a cochlear malformation.

## DISCUSSION

Applying the 2017 criteria to our cohort left most ears where they had been. Eighty-eight percent retained a category with a direct equivalent and no ear was reassigned to a different entity; the only redistribution occurred within CH, which is a single group in the earlier scheme and four subtypes in the current one. Ten ears, however, were left without any category at all.

The redistribution itself could have practical implications for reading the older literature. Prevalence figures and surgical series published before 2017 appear to remain broadly comparable with contemporary reports, at least for the categories represented in this cohort. The exception is CH. In the earlier scheme it was one category; it is now four, and these four have been reported to differ in hearing level.<sup>16</sup> A prevalence figure for CH published before 2017 therefore covers a mixed group and cannot be read as equivalent to a figure for any one of the current subtypes.

The way the hypoplastic ears redistributed is worth noting. The 2009 assessment had divided them locally according to whether the internal architecture could be recognized,<sup>9</sup> and this division mapped onto the current subtypes without a single crossover. Both schemes separated the same ears on the same feature, namely whether the modiolus and the interscalar septa can be identified. The finding is limited by the small number of ears and by the fact that the same observer made both assessments, but it does suggest that this feature is a natural dividing line and can be applied consistently.

None of the categories introduced in the 2017 revision was found in our cohort. Rudimentary otocyst and IP-III are uncommon. In a recent series of 155 malformed ears, rudimentary otocyst accounted for 3.2%, an isolated enlarged VA for 3.9%, and IP-III was likewise not encountered;<sup>8</sup> a cohort containing 38 malformed ears would not be expected to include them. Although no isolated CA abnormality or isolated enlarged VA was newly identified in this cohort, that absence cannot be generalized, since the 2009 protocol had already assessed both structures independently of the state of the cochlea.<sup>9</sup> How far an older report is comparable with a current one may therefore depend as much on how thoroughly the temporal bone was assessed at the time as on which categories were then available.

The 2017 revision also changed the scope of the scheme. The earlier version classified cochlear malformations and assessed the vestibule, the SCCs, the IAC and the aqueducts on separate axes,<sup>4</sup> whereas the current version brings all inner ear structures into a single list of eight categories.<sup>1</sup> Ears with an isolated abnormality of the vestibule or the SCCs, which had a place in the earlier scheme, therefore have none in the current one: by the heading of the classification they are inner ear malformations, yet no category accommodates them. Ten ears in this cohort were of this kind (Table 1).

Two of the eight categories are also defined differently from one another. The enlarged VA category requires a normal cochlea, vestibule and SCCs and is thus restricted to the isolated form,

although its title does not say so. No such restriction is placed on CA abnormalities, which are explicitly said to occur either with other malformations or with a normal cochlea;<sup>1</sup> an ear with both a cochlear malformation and an abnormal CA therefore has no stated rule for assignment, and all seven such ears in this cohort were of that kind. A recent series classified under the current scheme meets the same difficulty: enlarged VA had to be reported separately as isolated and as associated with other anomalies, and CA abnormality was reported as a finding across the whole cohort rather than as one of the eight categories.<sup>8</sup> A category for isolated vestibular and/or SCC abnormalities, and titles that state whether the isolated form is meant, would remove both difficulties (Table 1).

Two boundaries in the current scheme rest on size without a numerical definition. CH-I, CH-II and IP-I all lack the modiolus and the interscalar septa, and are separated by how large the cochlea is; yet the 2017 classification states no measurement for this. We had to adopt a threshold from an independent normative study,<sup>10</sup> derived from normal cochleae rather than malformed ones. A classification whose categories are separated by size would be easier to apply if the measurement were specified within the classification itself. The distinction between CH-III and CH-IV is also made without a numerical criterion. The published definitions, fewer than two turns versus hypoplastic middle and apical turns with a normal basal turn, are not mutually exclusive. In practice the distinction rests on whether the cochlea is proportionally or disproportionately small, with the middle and apical turns affected more than the basal turn. In one morphometric series the basal turn length of the two subtypes fell in a similar range,<sup>17</sup> and although one series found CH-IV to differ clearly from the other three in hearing level,<sup>16</sup> that comparison has not been repeated.

Two further difficulties affect measurement in malformed ears. The axial plane is conventionally aligned with the lateral SCC, but that canal is itself dysplastic in some malformed ears, so neither the axial plane nor the coronal plane derived from it can be standardized. Cochlear height is defined between the centers of the basal and the apical turn, which presupposes that both can be identified; in a cystic cochlea there are no turns and the endpoints become a matter of judgment. The ears in which size determines the category may thus be those in which size is hardest to measure consistently.

An incidental observation concerns the IAC. The coronal and the axial plane did not identify the same ears. At every threshold the coronal plane identified more of the ears with a cochlear malformation than the axial plane did, and it included no ear with a normal cochlea until 3.2 mm. The axial plane identified fewer malformed ears at each threshold and, from 3.0 mm upward, an increasing proportion of the ears it identified had a normal cochlea. In one patient with bilateral IP-I the canal appeared unremarkable in the axial plane and markedly narrow in the coronal plane.

Part of the explanation may lie in where the measurements that underlie the published criteria were taken. Normative data exist for the coronal mid-portion, the site used here: a mean midpoint width of 5.06 mm<sup>14</sup> and a mean maximum height of 4.62 mm<sup>18</sup> have been reported in controls. To our

knowledge, no comparable normative data exist for the axial mid-portion. The axial measurements in the literature were taken at the porus or a short distance inside it,<sup>14,15,18</sup> where the canal is wider, or at its narrowest point.<sup>19</sup> The thresholds themselves are equally varied: below 2.0 mm is the most widely used criterion,<sup>15</sup> the 2017 classification states 2.5 mm at the midpoint without giving a source,<sup>1</sup> and 3.0 mm at the porus has also been used.<sup>20</sup> The width of a normal canal ranges from about 2 to 12 mm, with a mean near 5 mm,<sup>21</sup> and in one series the variation between normal subjects was large enough in the axial plane that no threshold could be derived, whereas one could be derived for coronal measurements.<sup>19</sup> A single number applied without reference to the site of measurement is therefore unlikely to travel well between series.

The reliability of the measurement is a further difficulty. The canal has been described as too variable in shape and course for measurement to be consistent,<sup>14,22</sup> and volumetric assessment is more reliable between raters than diameter, with ICCs of 0.92 and 0.77 respectively.<sup>15</sup> A study that measured the height, width and cross-sectional area of the canal found that none of them distinguished children with cochlear nerve deficiency from controls, whereas the CA did.<sup>23</sup> A single axial diameter may therefore be a weak basis for calling a canal narrow, and the coronal plane should not be omitted.

### Limitations

This study has several limitations. The imaging dates from 2006 to 2008 and was acquired on a six-detector-row scanner; although the 0.8 mm reconstructions allowed the modiolus and the interscalar septa to be assessed, the thinner sections available today may allow better assessment of the internal architecture. The present analysis was based on CT alone. Magnetic resonance imaging had been performed in part of the cohort at the time of the original assessment, but cochlear nerve status was not reassessed here, so the categories assigned in this study rest on bony anatomy alone. All readings were made by one radiologist who had also performed the original assessment; although the earlier category was concealed during review, recall could not be excluded, and neither intraobserver nor interobserver reliability could be established for the categorical assessment. The IAC was measured only once, so the reliability of that measurement in this series is unknown, and the comparison between planes should be read with that in mind. Finally, several categories were represented by few ears or by none at all, so the study says nothing about how ears in the unrepresented categories would be redistributed, and the secondary analysis rests on only 12 hypoplastic ears.

### CONCLUSION

Reclassification of this cohort under the 2017 criteria changed little. No ear moved to a different entity, and redistribution was confined to CH, where a single group became four subtypes; ten ears, however, were left without a category. Since the boundaries between the cystic cochlear malformations, that is IP-I, CH-I and CH-II, and between CH-III and CH-IV, are defined descriptively rather than numerically, their application rests on observer judgment. The scheme leaves ears with an isolated vestibular and/or SCC

abnormality without a category, and defines its enlarged VA and CA categories on different terms; both could be clarified. In this cohort coronal measurement of the IAC identified the affected ears more consistently than axial measurement did, although the number of ears was small.

## ETHICAL DECLARATIONS

### Ethics Committee Approval

This study was approved by the Gaziantep University Medical Ethics Committee of the Faculty of Medicine, (Date: 19.02.2009, Decision No: 02-2009/20).

### Informed Consent

As this was a retrospective study, formal written informed consent was not required and was therefore not obtained. This retrospective study used pre-existing anonymized patient data. No additional intervention was performed, and there was no direct patient contact. The study was approved by the Ethics Committee, and the requirement for written informed consent was waived by the ethics committee.

### Peer Review Process

This manuscript was subject to external peer review.

### Conflict of Interest

The authors declare no conflicts of interest related to this study.

### Financial Disclosure

The authors received no financial support for the conduct or publication of this research.

### Author Contributions

Concept: MA, ASK; Design: MA, ASK; Control: ASK; Data Collection and/or Processing: MA; Analysis and/or Interpretation: MA, ASK; Literature Review: MA; Article Writing: MA; Critical Review: MA, ASK.

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