Diagn Interv Radiol 2026; DOI: 10.4274/dir.2026.263733

HEAD AND NECK IMAGING

ORIGINAL ARTICLE



# Semicircular canal aplasia: radiologic spectrum and clinical correlations on computed tomography and magnetic resonance imaging

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**PURPOSE**

This study aimed to characterize the imaging patterns of semicircular canal (SCC) aplasia on high-resolution computed tomography (CT) and magnetic resonance imaging (MRI) to identify associated cochleovestibular and internal auditory canal (IAC) abnormalities and to explore potential syndromic correlations.

**METHODS**

We retrospectively evaluated 12 patients (24 ears) diagnosed with SCC aplasia between January 2021 and January 2025. Imaging findings were reviewed for canal type, cochlear and vestibular abnormalities, vestibular aqueduct morphology, and IAC configuration. Syndromic associations and clinical outcomes were also assessed.

**RESULTS**

Lateral SCC aplasia was most common (41.7%), followed by budding remnants (12.5%) and total aplasia (12.5%). Total aplasia consistently coexisted with vestibular hypoplasia ($P = 0.020$) and was frequently accompanied by narrowing of the IAC ($P = 0.04$). Cochlear hypoplasia was found in one patient with bilateral total aplasia. Sensorineural hearing loss was present in 75% of patients, and caloric testing revealed subclinical vestibular dysfunction in two clinically asymptomatic adults. Syndromic associations included two cases of DiGeorge syndrome and a newly identified coexistence with Gitelman syndrome.

**CONCLUSION**

SCC aplasia encompasses a spectrum of rare anomalies, most commonly affecting the lateral canal and closely related to vestibular dysplasia. Recognition of IAC narrowing is important, as it can complicate cochlear implantation. The identification of Gitelman syndrome as a new association expands the spectrum of systemic diseases associated with SCC abnormalities. Comprehensive imaging with both CT and MRI is essential not only for diagnosis but also for surgical planning and syndromic screening.

**CLINICAL SIGNIFICANCE**

Systematic assessment of the IAC and vestibular structures on CT and MRI is essential in patients with congenital hearing loss. Recognizing IAC narrowing and vestibular dysplasia aids in preoperative cochlear implant planning and supports targeted syndromic or genetic evaluation.

**KEYWORDS**

Aplasia, cochlear implantation, computed tomography, internal auditory canal, magnetic resonance imaging, semicircular canal

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Received 20 November 2025; revision requested 15 December 2025; accepted 25 January 2026.

Epub: 12.02.2026

DOI: 10.4274/dir.2026.263733

Semicircular canal (SCC) anomalies are rare congenital inner ear malformations that may present as part of syndromic entities such as CHARGE, DiGeorge, or Goldenhar syndromes, and they can occur in isolation. Although some patients may exhibit sensorineural hearing loss and vestibular dysfunction, others remain asymptomatic. Among SCC malformations, including aplasia and hypoplasia, lateral SCC (LSCC) aplasia is the most commonly reported type.[1-7]

You may cite this article as: Başpınar N, Atalar M, Aktı S, Bora A. Semicircular canal aplasia: radiologic spectrum and clinical correlations on computed tomography and magnetic resonance imaging. *Diagn Interv Radiol*. 12 February 2026 DOI: 10.4274/dir.2026.263733 [Epub Ahead of Print].

Inner ear development begins in the 6th week of gestation. The pars superior, comprising the utricle and SCCs, forms earlier than the cochlea and saccule of the pars inferior. Therefore, developmental disturbances affecting the pars superior may also influence the cochlea and vestibule. Embryologically, the superior SCC (SSCC) is the first to form, followed by the posterior SCC (PSCC), whereas the LSCC is the last to develop, making it more susceptible to isolated aplasia. Although SCC dysplasia is often accompanied by cochlear malformations, several reports have documented normal cochlear morphology in isolated LSCC aplasia.[7-9]

Imaging with computed tomography (CT) and magnetic resonance imaging (MRI) is critical for the evaluation of these anomalies. Although CT provides superior detail of osseous structures, MRI offers high-resolution visualization of the membranous labyrinth and associated neural structures.[6,8]

This study aimed to evaluate CT and MRI findings in patients with SCC aplasia, assess associated cochlear and vestibular abnormalities, and investigate possible syndromic relationships. Previous studies have often focused on isolated LSCC aplasia or syndromic malformations but have lacked combined CT–MRI correlation. This study addresses existing gaps in the literature by integrating high-resolution CT and MRI findings, systematically evaluating narrowing of the internal auditory canal (IAC) as a radiologic marker, and reporting a newly identified syndromic association with Gitelman syndrome.

**Main points**

- Lateral semicircular canal (SCC) aplasia is the most common anomaly, often presenting with vestibular dysplasia.
- Total SCC aplasia always coexists with vestibular hypoplasia, highlighting embryologic timing.
- Narrowing of the internal auditory canal is a key radiologic marker with surgical implications.
- The newly identified association with Gitelman syndrome expands the known syndromic spectrum.
- Computed tomography and magnetic resonance imaging offer complementary insights essential for diagnosis and preoperative mapping.

## Methods

### Study design and population

This retrospective study was conducted between January 2021 and January 2025 at a tertiary care university hospital. Patients who presented with hearing loss, dizziness, or balance disorders and were radiologically diagnosed with SCC aplasia by either CT or MRI were included. Of the 13 patients initially identified, 1 was excluded because motion artifacts significantly affected image quality, leaving 12 patients for final analysis. All patients with SCC aplasia diagnosed during the study period were included, regardless of referral indication, to minimize selection bias. However, some patients were assessed in the context of possible cochlear implantation, which may have influenced referral patterns. Vestibular testing was not performed uniformly in all patients because of differences in clinical presentation and patient cooperation, particularly among children. In adult patients who were able to cooperate, caloric testing was performed as part of the routine otoneurological evaluation. This limitation was mainly due to age- and cooperation-related constraints in pediatric patients. The patient flowchart of the study is shown in Figure 1.

### Imaging protocols

High-resolution CT scans were performed using a 128-slice multidetector scanner (GE Revolution EVO, Milwaukee, WI, USA). The imaging parameters were standardized as follows: 0.625-mm collimation, 0.375 pitch, 0.625-mm section thickness, 140 kVp (80 kVp for pediatric patients), 400 mAs (150 mAs for pediatric patients), and a 199-mm field of view.

MRI examinations were acquired using a 1.5-Tesla scanner (Magnetom Aera, Siemens Healthineers, Erlangen, Germany) with 20- and 64-channel head coils. The imaging protocol included axial T2-weighted fast spin–echo sequences [repetition time (TR): 5,136 ms, echo time (TE): 96 ms, slice thickness: 5 mm], T1-weighted volumetric interpolated breath-hold examination with fat saturation (TR: 18 ms, TE: 3.69 ms, slice thickness: 1 mm), three-dimensional T2-weighted sampling perfection with application-optimized contrasts using different flip-angle evolution sequences (TR: 1,200 ms, TE: 183 ms, slice thickness: 0.58 mm), and diffusion-weighted imaging (TR: 5,800 ms, TE: 98 ms, slice thickness: 5 mm, b values of 0 and 1,000 s/mm$^2$).

### Image evaluation

All imaging studies were retrospectively reviewed by two experienced neuroradiologists, one with 14 years and the other with 9 years of experience. In cases of discrepancy, consensus was reached through consultation with a senior neuroradiologist with more than 15 years of experience. High-resolution temporal bone CT scans were assessed in axial and coronal planes. The following structures were systematically evaluated in both ears of all patients: presence or absence of the LSCC, PSCC, and SSCC; cochlear morphology, categorized as normal or hypoplastic; vestibule morphology, defined as normal, hypoplastic, or enlarged/dysmorphic; integrity of the round and oval windows; presence or absence of the vestibular aqueduct; and IAC diameter, measured in the axial

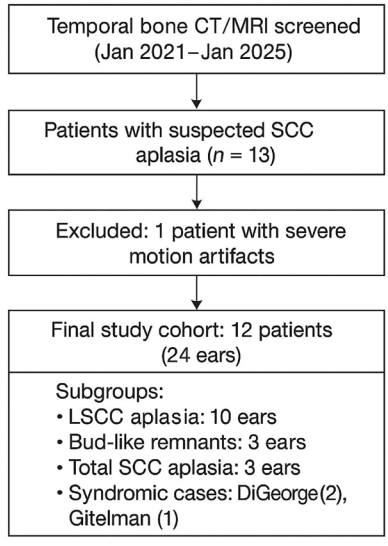


**Figure 1.** Study flow diagram. CT, computed tomography; MRI, magnetic resonance imaging; SCC, semicircular canal; LSCC, lateral semicircular canal.

plane and classified as narrow if ≤ 2.0 mm. The measurement was performed at the mid-porus level of the IAC on axial images, perpendicular to the canal axis, using standardized bone window settings to ensure reproducibility. SCC aplasia was defined as the complete absence of the canal, with or without a rudimentary bud. Bud-like remnants were also categorized as aplasia, consistent with the literature. This approach aligns with embryology-based classifications of inner ear malformations, in which rudimentary or bud-like IAC structures are considered aplastic variants rather than true hypoplasia.[1,5,8]

### Statistical analysis

All statistical analyses were performed using SPSS version 26.0 (IBM Corp., Armonk, NY, USA). Descriptive data are presented as mean ± standard deviation for continuous variables and as counts and percentages for categorical variables. Group comparisons were conducted using chi-square or Fisher's exact tests for categorical variables and independent-samples t-tests for continuous variables. Due to the limited sample size, nonparametric comparisons were preferred. A *P* value of <0.05 was considered statistically significant.

### Ethics

This study was conducted in accordance with the ethical standards of the Declaration of Helsinki. The Ethics Committee of Sivas Cumhuriyet University approved the study (approval number: 2025-04/127, date: April 24, 2025).

## Results

Twelve patients, seven men and five women, with a mean age of 23.3 ± 23.0 years (range: 2–66 years), were included in the analysis. A total of 24 ears were examined. Isolated aplasia of the LSCC was the most common anomaly, affecting 10 ears (41.7%) (Figure 2). Bilateral LSCC aplasia was identified in two patients, whereas bud-like LSCC remnants were present in three ears, two of which were in the same individual (Figure 3). Complete aplasia of all three IACs was observed in three ears, including one patient with bilateral involvement.

All cases of total SCC aplasia were associated with vestibular hypoplasia, and this association was statistically significant (*P* = 0.02). In most patients with LSCC aplasia, the vestibule was enlarged or dysmorphic, and the absence of the LSCC bony island was a consistent imaging feature (Figure 4). Multistructural involvement was more common in cases of total SCC aplasia, where vestibular and cochlear anomalies often coexisted (Figure 5). Representative CT and MRI images of different SCC aplasia patterns are illustrated in Figures 6 and 7.

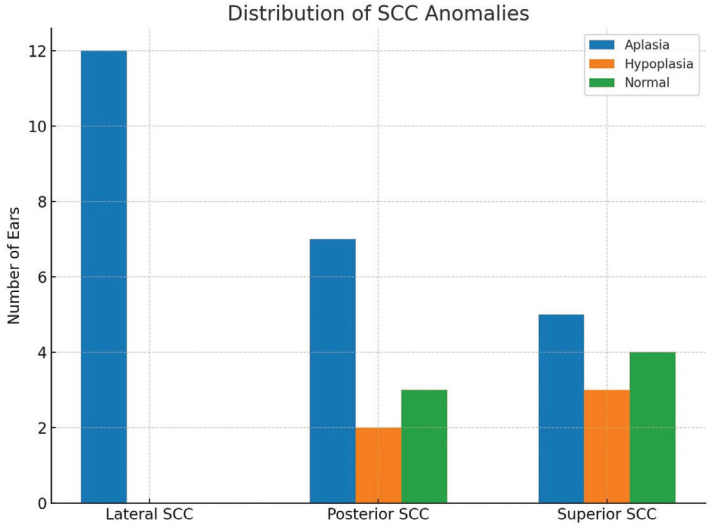


**Figure 2.** Distribution of semicircular canal anomalies. SCC, semicircular canal.

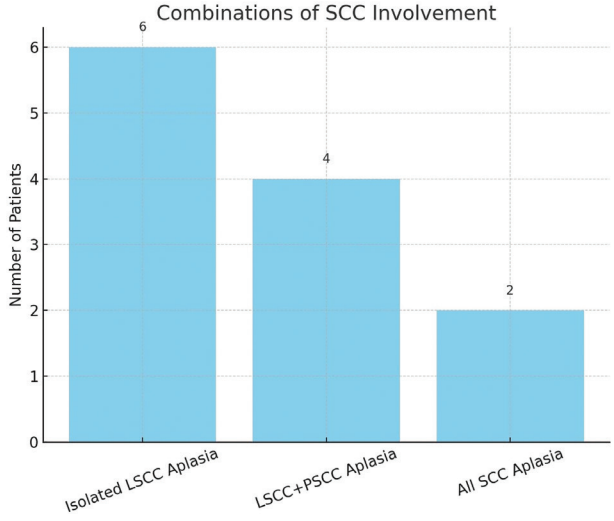


**Figure 3.** Combinations of semicircular canal anomalies. SCC, semicircular canal; LSCC, lateral semicircular canal; PSCC: posterior semicircular canal.

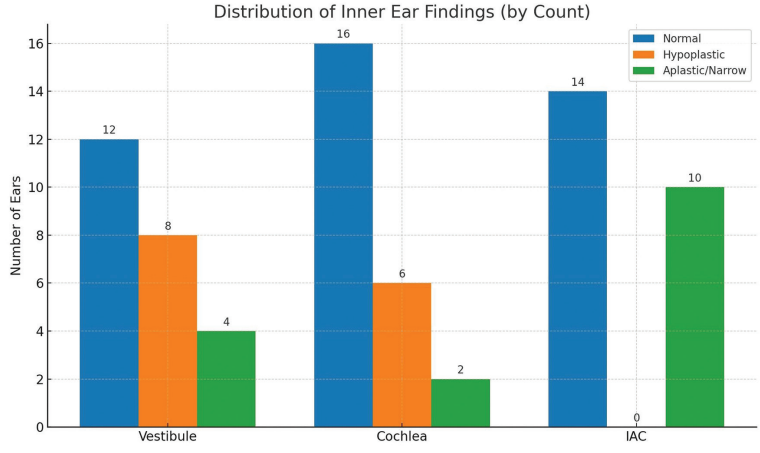


**Figure 4.** Distribution of inner ear findings. IAC, internal auditory canal.

Narrowing of the IAC, defined as a diameter of ≤ 2.0 mm, was detected in patients with bilateral total SCC aplasia and in some with bud-like remnants. Statistical analysis demonstrated a significant relationship between SCC aplasia and IAC narrowing ($P$ = 0.04). This finding underscores the relevance of IAC caliber assessment during preoperative evaluation. The vestibular aqueduct was identified in 6 of the 10 ears with LSCC aplasia, without evidence of enlargement. A summarized overview of radiologic patterns, associated inner ear anomalies, IAC findings, and clinical characteristics is provided in Table 1. The distribution of anomalies involving the SCCs, cochlea, vestibule, oval and round windows, and IAC measurements is summarized in Table 2. Overall, radiologic findings demonstrated a continuum from isolated LSCC aplasia to total labyrinthine dysplasia, reflecting embryologic severity. Given the limited sample size and small subgroup distributions, categorical variables were compared using Fisher's exact test.

Syndromic associations were documented in three patients. One patient with bilateral SCC aplasia had Gitelman syndrome, representing the first reported case of this coexistence in the literature. Two additional patients with LSCC aplasia were diagnosed with DiGeorge syndrome. Sensorineural hearing loss was observed in 9 of 12 patients (75%). Caloric testing, available in two adult patients, revealed unilateral canal paresis despite the absence of overt vestibular symptoms. Vertigo was less frequently reported in adults, although caloric testing revealed canal paresis in two cases, suggesting that vestibular dysfunction may remain clinically silent in some individuals.

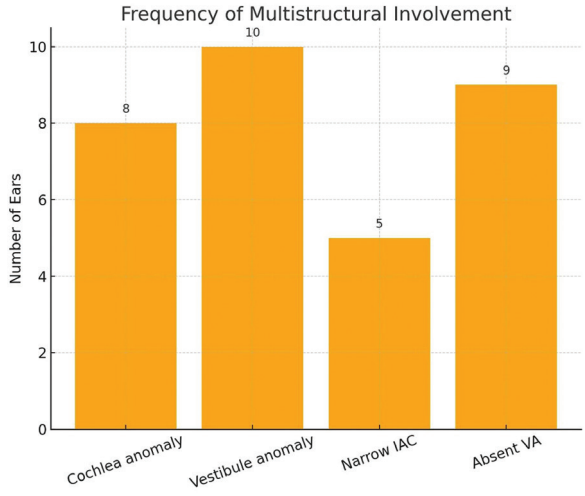


**Figure 5.** Frequency of multistructural involvement. IAC, internal auditory canal; VA, vestibular aqueduct.

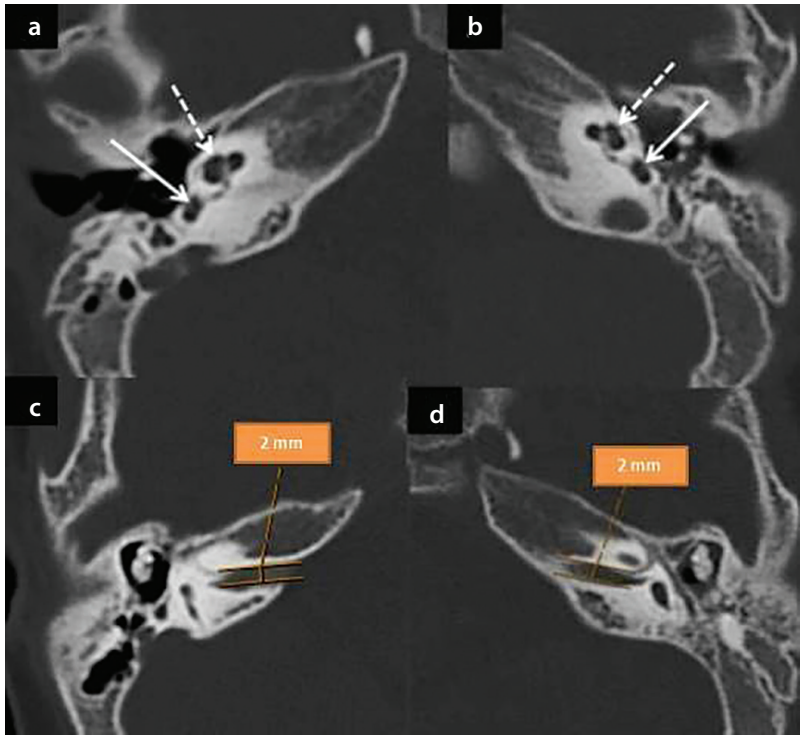


**Figure 6.** A 2-year-old girl with complete aplasia of the bilateral semicircular canals. **(a, b)** Non-contrast-enhanced axial temporal high-resolution computed tomography images show bilateral cochlear hypoplasia (dotted arrows) and hypoplastic vestibules (arrows). **(c, d)** The bilateral internal auditory canals are 2 mm below the lower threshold.

## Discussion

SCC aplasia and hypoplasia represent a broad spectrum of congenital inner ear malformations. In our series, LSCC aplasia was the most common anomaly, reflecting embryologic susceptibility due to its later development.[1,5] Interruption of development at earlier stages resulted in more extensive anomalies affecting the cochlea and vestibule, consistent with embryologic principles.[6,7]

In our cohort, LSCC aplasia was predominant, consistent with its later embryologic development and increased vulnerability to isolated developmental arrest. Earlier disruption of the pars superior resulted in more extensive malformations, explaining the consistent association between total SCC aplasia and vestibular hypoplasia. This developmental continuum was previously described by Sennaroğlu and Bajin[8] who emphasized that the timing of embryonic disruption determines the extent of associated anomalies. In our cohort, one patient with bilateral total SCC aplasia also demonstrated cochlear hypoplasia, providing further evidence that early developmental arrest produces more severe and combined anomalies.

Radiologically, the most reliable markers of SCC aplasia in our study were an enlarged or dysmorphic vestibule, absence of the LSCC bony island, and narrowing of the IAC. Previous reports have suggested that IAC narrowing may coexist with labyrinthine malformations and may even predict cochlear nerve deficiency.[9] In our series, IAC narrowing was significantly more common in patients with total SCC aplasia or bud-like remnants, indicating that evaluation of IAC caliber should be an integral part of imaging

assessment in suspected SCC malformations. From a surgical perspective, these structural variations have direct relevance to cochlear implantation outcomes. This finding has important implications for preoperative planning in candidates for cochlear implantation, where cochlear nerve integrity must be established before surgery.[10]

Previous studies have reported associations between SCC aplasia and Kallmann syndrome, CHARGE syndrome, and chromosomal abnormalities.[11] A notable feature of our cohort was the presence of syndromic associations in a quarter of the patients. Beyond classical syndromes such as CHARGE and DiGeorge, our study identified a previously unreported association. The coexistence of bilateral SCC aplasia with Gitelman syndrome is, to our knowledge, the first report of its kind in the literature. Although Gitelman syndrome is primarily a disorder of the renal tubules caused by mutations in the *SLC12A3* gene, experimental studies have demonstrated the expression of sodium–chloride cotransporters in the endolymphatic sac, suggesting a possible mechanism linking electrolyte imbalance to vestibular morphogenesis.[12] This observation broadens the spectrum of syndromes associated with SCC abnormalities. In addition, two patients with LSCC aplasia were diagnosed with DiGeorge syndrome (22q11.2 deletion), which is consistent with previous studies reporting frequent ear abnormalities in this syndrome due to abnormal development of the pharyngeal arch.[13,14] This finding warrants further investigation, as SLC12A3 mutations in Gitelman syndrome may disrupt inner ear fluid homeostasis.

Clinically, SCC aplasia presents with diverse manifestations. Most patients in our series had sensorineural hearing loss, but vestibular dysfunction was not always clinically evident. Notably, caloric testing revealed canal paresis in two adults who denied vertigo, suggesting the possibility of central compensation for chronic congenital vestibular dysfunction. Although vestibular dysfunction may remain clinically silent due to central compensation, vestibular tests such as caloric testing or video head impulse testing can reveal subclinical deficits and should be considered, particularly in adult patients and in cases of bilateral or total SCC aplasia. This highlights the importance of systematic vestibular testing, as subtle but clinically relevant deficits may otherwise be missed. Previous studies have also shown that congenital

**Table 1.** Summary of radiologic and clinical findings in patients with semicircular canal aplasia

| Parameter | Number (%) |
|---|---|
| Patients (n) | 12 |
| Ears evaluated (n) | 24 |
| Type of SCC aplasia (ears) | |
| • Isolated LSCC aplasia | 10 (41.7%) |
| • Bud-like LSCC remnants | 3 (12.5%) |
| • Total SCC aplasia | 3 (12.5%) |
| Laterality | |
| • Unilateral involvement | 9 patients |
| • Bilateral involvement | 3 patients |
| Associated inner ear findings | |
| • Vestibular hypoplasia (total SCC aplasia) | 3/3 (100%) |
| • Enlarged/dysmorphic vestibule (LSCC aplasia) | Majority |
| • Cochlear hypoplasia | 1 patient |
| Internal auditory canal | |
| • Narrow IAC (≤ 2.0 mm) | 3 patients |
| Clinical findings | |
| • Sensorineural hearing loss | 9/12 (75%) |
| • Vertigo symptoms | Rare |
| • Abnormal caloric test (tested patients) | 2 patients |
| Syndromic associations | |
| • DiGeorge syndrome | 2 patients |
| • Gitelman syndrome | 1 patient |

SCC, semicircular canal; LSCC, lateral semicircular canal; IAC, internal auditory canal.

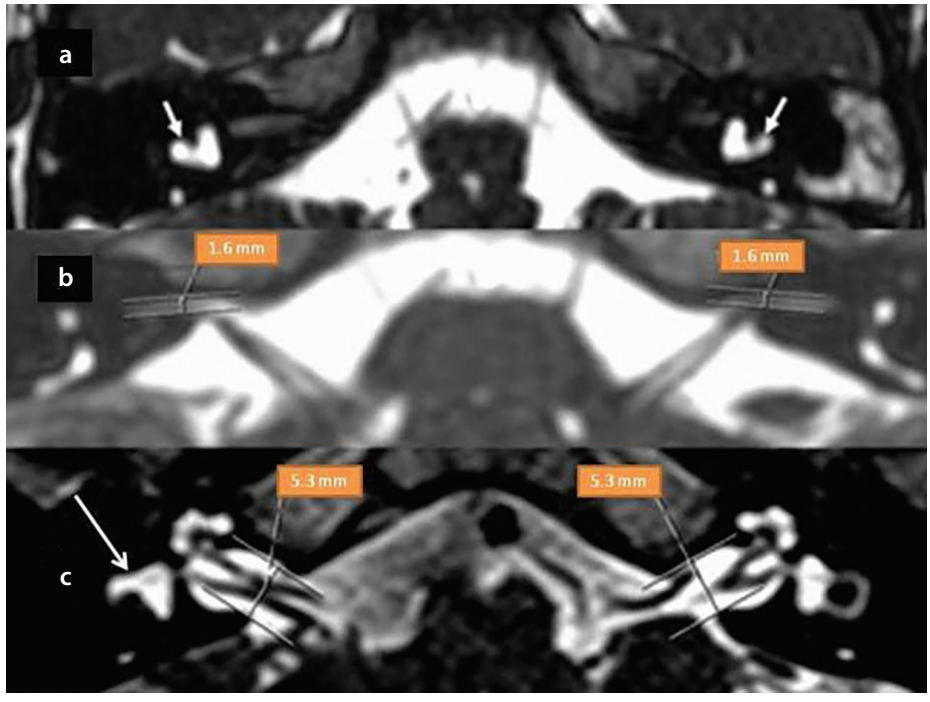


**Figure 7.** Three-dimensional T2-weighted SPACE magnetic resonance images in the axial plane. A 3-year-old boy with bilateral lateral semicircular canal (LSCC) aplasia shows bud-like structures (**a**, arrows) and bilateral narrow internal auditory canals (IACs) **(b)**. **(c)** A 2-year-old girl shows a bud-like formation of the right LSCC (arrow) with dysplastic and hypoplastic canal structures; the IAC diameters are normal.

**Table 2.** Distribution of findings of semicircular canal, cochlea, vestibular, oval and round window, and internal auditory canal

| Case number | Age (year) | Gender | Side | Lateral SCC | Posterior SCC | Superior SCC | Vestibular aquaduct | Oval window | Cochlea | Round window | Vestibule | IAC diameter (mm) |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| 1 | 23 | M | R | Absent | Absent | Absent | Absent | Absent | Normal | Normal | Hypoplastic | 4.1 |
| | | | L | Present | Present | Present | Present | Absent | Normal | Normal | Normal | 5.3 |
| 2 | 2 | M | R | Present | Present | Present | Absent | Absent | Normal | Normal | Normal | 3.2 |
| | | | L | Absent | Present | Present | Present | Absent | Normal | Normal | Enlarged/ dysmorphic | 2.6 |
| 3 | 22 | M | R | Absent | Present | Present | Present | Absent | Normal | Normal | Enlarged/ dysmorphic | 4.3 |
| | | | L | Absent | Present | Present | Present | Absent | Normal | Normal | Enlarged/ dysmorphic | 4.3 |
| 4 | 9 | M | R | Present | Present | Present | Present | Absent | Normal | Normal | Normal | 3.6 |
| | | | L | Absent | Present | Present | Present | Absent | Normal | Normal | Enlarged/ dysmorphic | 3.4 |
| 5 | 17 | F | R | Present | Present | Present | Absent | Absent | Normal | Normal | Normal | 3.8 |
| | | | L | Absent | Present | Present | Absent | Absent | Normal | Normal | Enlarged/ dysmorphic | 3.4 |
| 6 | 43 | M | R | Absent | Present | Present | Present | Absent | Normal | Normal | Enlarged/ dysmorphic | 4.7 |
| | | | L | Absent | Present | Present | Absent | Absent | Normal | Normal | Enlarged/ dysmorphic | 4.7 |
| 7 | 63 | F | R | Absent | Present | Present | Absent | Absent | Normal | Normal | Enlarged/ dysmorphic | 3.8 |
| | | | L | Present | Present | Present | Absent | Absent | Normal | Normal | Normal | 4.3 |
| 8 | 2 | F | R | Bud (rudimentary) | Present | Present | Not assessed | Not assessed | Not assessed | Not assessed | Enlarged/ dysmorphic | 5.3 |
| | | | L | Present | Present | Present | Not assessed | Not assessed | Normal | Not assessed | Normal | 5.3 |
| 9 | 3 | M | R | Bud (rudimentary) | Present | Present | Present | Absent | Aplasia | Aplasia | Enlarged/ dysmorphic | 1.6 |
| | | | L | Bud (rudimentary) | Present | Present | Absent | Absent | Normal | Normal | Enlarged/ dysmorphic | 1.6 |
| 10 | 2 | F | R | Absent | Absent | Absent | Absent | Hypoplastic | Normal | Normal | Hypoplastic | 2.0 |
| | | | L | Absent | Absent | Absent | Absent | Hypoplastic | Aplasia | Normal | Hypoplastic | 2.0 |
| 11 | 66 | M | R | Present | Present | Present | Present | Absent | Normal | Normal | Normal | 4.0 |
| | | | L | Absent | Present | Present | Present | Absent | Aplasia | Normal | Enlarged/ dysmorphic | 3.6 |
| 12 | 28 | F | R | Hypoplastic | Present | Present | Present | Absent | Normal | Normal | Normal | 5.0 |
| | | | L | Present | Present | Present | Present | Absent | Normal | Normal | Normal | 4.7 |

Note: Narrow IAC is defined as ≤ 2 mm. F, female; M, male; L, left; R, right; IAC, internal auditory canal; SCC, semicircular canal.

vestibular deficits, even when asymptomatic, may contribute to delayed motor milestones and impaired balance in children.[15]

Positioning our findings within the literature (Table 3), early tomography-era work by Phelps[13] first highlighted frequent LSCC dysmorphology but lacked the neural and membranous detail provided by modern cross-sectional imaging. Subsequent cochlear implant center series broadened the anatomic spectrum: Satar et al.[12] reported high rates of oval window anomalies and variable cochlear dysplasia with syndromic enrichment (predominantly CHARGE), whereas the CHARGE-focused cohort of Morimoto et al.[10] quantified cochlear aperture atresia and frequent posterior displacement of the facial nerve canal. Radiologic cohorts have also emphasized middle ear associations and aberrant courses of the facial nerve canal, which are crucial for surgical planning.[16] More recent vestibular function-oriented LSCC cohorts linked the absence of a central bony island with more severe caloric paresis and showed that bilateral LSCC dysplasia or aplasia often presents with bilateral vestibular loss.[14,15] Across these studies, LSCC anomalies consistently predominate, consistent with the embryologic vulnerability described by Jackler et al.[1] and by Sennaroglu and Saatci.[2] Our series aligns with this pattern but adds two practice-informing elements: first, a combined CT–MRI characterization that highlights IAC narrowing as an ancillary marker accompanying severe SCC malformations, and second, a novel syndromic association with Gitelman syndrome, expanding the syndromic spectrum beyond CHARGE and 22q11.2 deletion. Taken together, the comparative evidence supports routine scrutiny of the IAC and meticulous mapping of the facial and cochlear nerves before cochlear

**Table 3.** Comparison of reported cases of semicircular canal aplasia in the literature

| Author/year | Number of patients/ears | SCC type | Associated malformations | Syndromic associations | Key findings |
|---|---|---|---|---|---|
| Phelps,[13] 1974 | 34 | Mostly LSCC aplasia/dysplasia | Limited cochlear anomalies | Not specified | First systematic description using tomography |
| Satar et al.,[12] 2003 | 12 | LSCC aplasia, total SCC aplasia | Cochlear dysplasia, oval window anomalies | CHARGE | Emphasized surgical challenges and FN anomalies |
| Morimoto et al.,[10] 2006 | 15 (CHARGE) | Absent or hypoplastic SCC | Cochlear aperture atresia, aberrant FN canal | CHARGE | Highlighted absent SCC as radiologic marker in CHARGE |
| Shin et al.,[16] 2009 | 10 | LSCC aplasia, total SCC aplasia | Middle ear anomalies, FN canal anomalies | Mixed | CT/MR study; surgical mapping importance |
| Costa et al.,[11] 2020 | 8 | Bilateral SCC malformations | Variable cochlear and vestibular findings | Mixed/ chromosomal | Reported hearing loss profiles and rehabilitation approaches |
| Azzahiri et al.,[2] 2023 | 1 case | Bilateral LSCC aplasia | Vestibular enlargement/ none | Non-syndromic | Rare bilateral aplasia; in some reports without hearing loss |
| Michel et al.,[3] 2016 | 1 case | Isolated LSCC aplasia | Vestibular and cochlear dysfunction | Non-syndromic | VEMP and VNG showed vestibular impairment |
| Parnes and Chernoff,[4] 1990 | 2 | Bilateral SCC aplasia | Near-normal cochlea | Non-syndromic | Bilateral SCC aplasia with preserved cochlea |
| Kwak et al.,[14] 2020 | 22 | LSCC aplasia/ dysplasia | Cochlea usually normal | Non-syndromic | Strong correlation between LSCC aplasia and caloric paresis |
| Yun et al.,[15] 2024 | 18 | LSCC aplasia/ dysplasia | Absence of central bony island | Non-syndromic | Bilateral LSCC aplasia associated with bilateral vestibular loss |
| Present study | 12 | LSCC (41.7%), bud-like remnants, total SCC aplasia | Vestibular hypoplasia, IAC narrowing, cochlear hypoplasia (rare) | DiGeorge, Gitelman (novel) | First Gitelman association; IAC narrowing highlighted |

CT, computed tomography; LSCC, lateral semicircular canal; MRI, magnetic resonance imaging; SCC, semicircular canal; IAC, internal auditory canal; PSCC: posterior semicircular canal; FN, facial nerve.

implantation, as well as systematic syndromic screening, particularly in pediatric patients and those with bilateral or total SCC aplasia.

From a surgical perspective, accurate identification of SCC aplasia and its associated anomalies is essential for safe and effective cochlear implantation. Abnormal vestibular morphology, IAC narrowing, or cochlear nerve hypoplasia may complicate electrode insertion and affect postoperative outcomes. Therefore, both CT and MRI are indispensable in the preoperative evaluation of these patients, providing complementary information about osseous, membranous, and neural structures.

This study offers one of the few systematic evaluations of SCC aplasia using high-resolution CT and MRI, highlighting radiologic markers that have not been consistently emphasized in previous reports. Its main contribution is the identification of a novel syndromic association between bilateral SCC aplasia and Gitelman syndrome, expanding the spectrum of systemic diseases linked to these malformations. By correlating embryologic timing with imaging findings, the study clarifies the developmental mechanisms underlying cochleovestibular anomalies. The findings are clinically relevant, emphasizing IAC narrowing and vestibular hypoplasia as key imaging markers for surgical planning. The reproducibility of the results is supported by standardized imaging protocols and independent review by experienced neuroradiologists.

Nonetheless, certain limitations must be acknowledged. The study's retrospective, single-center design and limited sample size restrict generalizability and preclude multivariate or receiver operating characteristic analyses. Additionally, vestibular testing and genetic evaluations were not consistently performed, especially in pediatric patients, which may have resulted in underestimation of functional deficits and syndromic associations. Despite these constraints, the study offers valuable insights by integrating radiologic and embryologic perspectives and introducing a previously unreported syndromic association.

SCC aplasia is not a single entity but a spectrum of malformations with varying embryologic timing, radiologic features, and clinical effects. Recognition of characteristic imaging findings, such as vestibular dysplasia, absence of the bony LSCC island, and narrowing of the IAC, should alert radiologists to the possibility of SCC aplasia. Additionally, identifying syndromic associations highlights the importance of comprehensive systemic and genetic testing. Future multicenter studies with larger patient cohorts, systematic vestibular testing, and genetic profiling will be essential to refine classification, clarify pathogenesis, and improve clinical management strategies for these rare but clinically significant abnormalities.

In conclusion, SCC aplasia represents a heterogeneous spectrum of congenital inner ear malformations with clinically significant anatomic variability. More severe forms of SCC aplasia are consistently associated with vestibular hypoplasia and narrowing of the IAC, reflecting their shared embryologic origin and direct relevance to surgical planning. Systematic evaluation of the IAC and vestibular structures using both CT and MRI is therefore essential, particularly in candidates for cochlear implantation. Finally, the identification of a novel association with Gitelman syndrome underscores the importance of comprehensive syndromic assessment in patients with SCC anomalies.

### Footnotes

### Conflict of interest disclosure

The authors declared no conflicts of interest.

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