Inner-Ear Disorders Presenting with Air–Bone Gaps: A Review

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Air–bone gaps (ABGs) are commonly found in patients with conductive or mixed hearing loss generally due to outer- and/or middle-ear diseases such as otitis externa, tympanic membrane perforation, interruption or fixation of the ossicular chain, and chronic suppurrative otitis media. ABGs can also be found in correlation with inner-ear disorders, such as endolymphatic hydrops, enlarged vestibular aqueduct syndrome, semicircular canal dehiscence, gusher syndrome, cochlear dehiscence, and Paget disease's as well cerebral vascular anomalies including dural arteriovenous fistula. The typical clinical presentation of inner-ear conditions or cerebral vascular anomalies causing ABGs includes audiological and vestibular symptoms like vertigo, oscillopsia, dizziness, imbalance, spinning sensation, pulsatile or continuous tinnitus, hyperacusis, autophony, auricular fullness, Tullio's phenomenon, and Hennebert's sign. Establishing a definitive diagnosis of the underlying condition in patients presenting with an ABG is often challenging to do and, in many patients, the condition may remain undefined. Results from an accurate clinical, audiological, and vestibular evaluation can be suggestive for the underlying condition; however, radiological assessment by computed tomography and/or magnetic resonance imaging is mandatory to confirm any diagnostic suspicion. In this review, we describe and discuss the most recent updates available regarding the clinical presentation and diagnostic workup of inner-ear conditions that may present together with ABGs.

KEYWORDS: Air–bone gap, inner-ear, Ménière's disease, mixed hearing loss

INTRODUCTION

An air–bone gap (ABG) is defined as the difference between air-conduction and bone-conduction audiometric thresholds. ABGs are found in patients with conductive (CHL) or mixed (MHL) hearing loss generally attributed to outer- and/or middle-ear diseases such as otitis externa, tympanic membrane perforation, interruption or fixation of the ossicular chain, and chronic suppurrative otitis media.

Air–bone gap can also follow the onset of inner-ear structural anomalies, such as enlarged vestibular aqueduct (EVA) syndrome, semicircular canal dehiscence, gusher syndrome, cochlear dehiscence, and Paget’s disease [1]. These conditions are mainly characterized by sensorineural hearing loss (SNHL) [2, 3]; however, an ABG can also be found due to the so-called “third-window” mechanism [4], where the depletion of acoustic energy from the inner-ear into the middle-ear and/or the cranial cavity [4, 5] decreases the amount of energy delivered to the round window through a third-window. This mechanism provokes a decrease in the pressure gradient across the basilar membrane, leading to a decreased perception of air-conducted sound that progresses to a low-frequency ABG [5]. ABGs have additionally been reported in patients with endolymphatic hydrops (EH); in this context, an ABG can be explained by increased perilymphatic pressure with a consequent decrease in stapes mobility or by saccular dilation leading to a reduc-
tion in stapedial mobility. Cerebral vascular anomalies including du-
ral arteriovenous fistula (DAVF) may also be responsible for an ABG
due to communication between the inner-ear and the cranial cavity
through the cochlear and vestibular aqueducts and multiple small
neurovascular foramina, facilitating contiguity between the perilym-
phatic and subarachnoid spaces [9].

The typical clinical presentation of inner-ear conditions or cerebral
vascular anomalies causing ABGs includes audiological and vestibu-
lar symptoms like vertigo, oscillopsia, dizziness, imbalance, spinning
sensation, pulsatile or continuous tinnitus, hyperacusis, autophony,
auricular fullness, Tullio's phenomenon, and Hennebert's sign [7].

Establishing a definitive diagnosis is often challenging and, in many
patients, the underlying condition remains unknown. This review
sought to describe and discuss the most recent updates available re-
garding the clinical presentation and diagnostic workup of inner-ear
conditions that may present together with an ABG.

CLINICAL AND RESEARCH CONSEQUENCES

Superior Semicircular Canal Dehiscence (SSCD)
Superior semicircular canal dehiscence (SSCD) is an idiopathic condi-
tion characterized by a loss of the bony wall of the superior semicir-
cular canal that promotes communication between the semicircular
canal and the middle cranial fossa. The incidence of SSCD in the gen-
eral population is difficult to estimate; for example, Minor [10] reported
65 cases in a nine-year period. Some risk factors have been reported
to correlate with SSCD including direct mechanical trauma, increased
cerebrospinal fluid pressure, increased pressure following Valsalva
maneuvers, underdevelopment of bone overlying the semicircular
canal, and progressive erosion due to vascular pulsations [30].

The clinical presentation of SSCD includes spontaneous or positional
dizziness, vertigo following pressure or sound, and vertical nystag-
us [10]. In some cases, SSCD can be asymptomatic, making the estab-
lishment of a clinical diagnosis harder [11].

The typical audiological feature of SSCD is an ABG in the low and
middle frequencies (≤ 2,000 Hz); bone-conduction thresholds can be
present at supranormal levels [12], which can facilitate a misdiagnosis
of CHL. At frequencies of 2000 Hz or higher, only a limited, or ab-
sent gap can be found because less acoustic energy passes through
the third window. The degree of ABG may be related to the location,
length, or cross-sectional area of the SSCD [13].

Many techniques have been proposed to diagnose SSCD, including
audiotometry, complete immittance testing with tympanometry and
acoustic reflexes, and air-conducted vestibular-evoked myogenic po-
tential (VEMP) testing. The diagnostic confirmation of a true case of
dehiscence should involve a high-resolution computed tomography
(CT) scan of the temporal bone using multiplanar reconstructions in
the planes of Stenver and Pöschl; Pöschl views parallel and Stenver
views perpendicular to the plane of the superior semicircular canal
have the highest diagnostic specificity [14].

The treatment of SSCD is exclusively surgical and includes capping
or plugging of the SSCD or reinforcement of the oval and/or round
windows [15].

Posterior Semicircular Canal Dehiscence (PSCD)
Posterior semicircular canal dehiscence (PSCD) is an uncommon ca-
nal dehiscence [16] that can be observed alone or in combination with
SSCD with frequencies of 0.3% and 4.5%, respectively [10]. The PSCD
can extend into the posterior fossa dura through communication
with a high-riding jugular bulb or via a bony defect. Often, PSCD re-
results from jugular bulb erosion or fibrous dysplasia [9].

Audiological symptoms associated with PSCD are similar to those in
SSCD; a clinical suspicion for PSCD should arise if bone-conduction re-
sponses are at 5 dB HL or better, a biphasic complex at VEMP test elicits
a lowered threshold at an increased amplitude, and vestibular signs
like nystagmus are present in the plane of the posterior semicircular
canal. Pure tone audiometry in the half-octave is useful to confirm the
characteristic low-frequency ABG correlated with this condition [17].

Lateral Semicircular Canal Dehiscence (LSCD)
Lateral semicircular canal dehiscence (LSCD) is often the result of
middle-ear diseases such as otitis media [17], cholesteatoma, and ca-
nal-wall-down mastoidectomy [9, 18]; isolated LSCD is extremely un-
usual [18] and has been reported only in patients with dysplastic bony
labyrinths to date. It is very difficult to ascertain whether ABG in LSCD
is related to abnormalities of the middle-ear or to a third-window
mechanism of the LSCD [19] (Figure 1).

Patients with LSCD can present with an ABG in the low-frequency
range. In cases of fistula secondary to cholesteatoma, vertigo and ipsi-
lateral nystagmus may be induced by pressure applied to the external
auditory canal [20]. In this scenario, it is controversial as to whether the
matrix of cholesteatoma should be completely removed or whether a
thin layer should be left in place to prevent possible deafness due

to labyrinthine exposure [9]. Previously Chen et al. [21] reported on the
complete surgical removal of cholesteatoma in 22 patients with LSCD;
two of them experienced postoperative hearing losses of 5 to 15 dB,
whereas the others presented with stable or improved hearing.

Enlarged Vestibular Aqueduct (EVA) Syndrome
The vestibular aqueduct have a diameter, on average, of 0.6 mm [22].
If the vestibular aqueduct is dilated to a diameter of greater than

MAIN POINTS

- An air-bone gap, typical of conditions that involve the exter-

nal and middle ear, can also be found in conditions affect-
ing the inner-ear due to a third-window mechanism or in
idiopathic or secondary endolymphatic hydrops
- Typical clinical presentation of inner-ear conditions with air-

bone gaps may include several audiologicalvestibular symptoms
such as vertigo, oscillopsia, dizziness, imbalance, tinnitus,
hyperacusis, autophony, auricular fullness, and Tullio's phe-

nomenon.
- Accurate clinical, audiological, and vestibular evaluations

can be suggestive of the underlying condition for hearing

loss presenting with an air-bone gap.
CT diagnostic criteria are based on the dimension of the vestibular aqueduct following the Valvassori [21] or the Cincinnati [28] criteria. MRI, especially involving T2-weighted images, is useful to visualize the membranous labyrinth and show the extraosseous portion of the endolymphatic sac [29].

To date, there is no evidence of a treatment able to limit the progression of EVA. Significant audiometric benefits have been observed in EVA patients receiving a cochlear implant [30, 31], while temporary hearing improvements were documented by Grimmer et al. [32] when treating EVA patients with corticosteroid therapy.

X-Linked Stapes Gusher
Stapes gusher syndrome is a congenital disorder linked to a mutation in the POU3F4 gene on the X chromosome affecting almost exclusively males. Female carriers can be asymptomatic or present a less severe degree of hearing loss, depending on the type of mutation [33]. Clinical features include congenital mixed hearing loss that worsens to severe deafness within eight to 10 years due to perilymphatic hydrops [34]. ABG is typically found in the low-frequency range, reflecting a third-window mechanism due to the absence of the lamina cribrosa and consequent communication between the perilymphatic and subarachnoid spaces; while the stapedial reflex may be preserved [4].

Computed tomography imaging can reveal abnormalities like cochlear hypoplasia, an enlarged internal auditory canal, absent modiolus and lamina cribrosa, and a labyrinthine facial nerve canal with a classic corkscrew appearance of the cochlea [34]. Cochlear implantation is indicated in patients with severe or profound SNHL, although careful insertion of the electrode is mandatory. Stapes surgery should be avoided due to the high risk of cerebrospinal fluid leak and meningitis; outcomes are not encouraging relative to those of other inner-ear abnormalities [35].

Bone Dyscrasias
Bone dyscrasias include a number of metabolic bone diseases such as Paget’s disease and osteogenesis imperfecta that show a pathologic bone turnover with involvement of the otic capsule. These conditions incite osteoclastic activation, inducing a third-window effect with a consequent low-frequency ABG at pure tone audiometry [36].

Osteogenesis imperfecta is caused by mutations in type I collagen synthesis, producing brittle bones and lax joints and ligaments as a result of existing fragile connective tissue [34]. Afflicted patients present with skeletal abnormalities and fractures due often to mild trauma; otic capsule mineralization can be facilitated with the use of bisphosphonates and dietary supplements [37]. Surgical options— which are, unfortunately, characterized by poor outcomes—include stapes surgery for fenestral disease [34] and cochlear implantation for retrofenestral disease.

Paget’s disease is characterized by a dystrophic remodeling of bone tissues and consists of an initial phase of bone resorption followed by a sclerotic phase and a final remodeling phase with, typically, lamellar bone; the bones show a typical mosaic-bone pattern [36]. Patients with Paget’s disease experience progressive bilateral hearing loss; Amilbabia Cabeza et al. [22] reported that subjects affected by Paget’s disease showed a more profound and higher incidence of CHL as compared...
Diagnostic strategy to differentiate middle-ear involvement, third-window lesions, and endolymphatic hydrops conditions

| Diagnostic measures | Middle ear involvement | Third-window lesion | Endolymphatic hydrops conditions |
|---------------------|------------------------|---------------------|---------------------------------|
| Type of hearing loss | Conductive hearing loss | Conductive or mixed hearing loss | Generally mixed or sensorineural hearing loss |
| Air-bone gap        | 0–60 dB, may involve all frequencies | 0–60 dB, greatest at frequencies <2000 Hz | 0–50 dB, greatest at frequencies <1000 Hz |
| Bone conduction thresholds | Rarely < 0 dB | Sometimes negative (−5 to −25 dB for low frequencies) | Rarely < 0 dB |
| Tympanometry        | Type A or B or C tympanogram | Type A tympanogram | Type A tympanogram |
| Acoustic reflex      | Absent                  | Present             | Generally present |
| Tullio phenomenon and/or Hennebert sign | Absent | May be present | Absent |
| Otoacoustic emissions | Absent                  | May be present | Generally absent |
| ECochG               | Normal                  | Generally elevated SP/AP ratio | SP/AP ratio generally > 0.37 |
| Cervical VEMP        | Absent                  | Low threshold and large amplitude | Generally absent or lower peak-to-peak amplitude at 500 Hz that 1000 Hz (tuning shift) |
| CT/MRI scan          | Middle ear abnormality  | Third-window lesion | Distension of the structures filled with endolymph at MRI |

ECochG: Electrocochleography; VEMP: vestibular evoked myogenic potentials; CT: computed tomography; MRI: magnetic resonance imaging; SP: Summating Potentials; AP: Action Potentials.

with among healthy controls. The pursuit of cochlear implantation is suggested in patients with severe and profound hearing loss.

Cochlear Dehiscence
Cochlear dehiscence refers to a thinning of the bone plate that covers the cochlea, which results in the establishment of direct communication between the inner-ear and the middle-ear cavity or neurovascular structure. When the scala vestibuli of the cochlea is compromised, a third-window mechanism occurs, decreasing cochlear input impedance and reducing the pressure within the scala vestibuli produced by sound. Kim and Wilson described the case of a dehiscence between the cochlea and carotid canal with an ABG that persisted after uneventful stapedectomy, while Fang reported a dehiscence that involved the geniculate ganglion, labyrinthine, or tympanic segments of the facial nerve.

Endolymphatic Hydrops (EH)
Endolymphatic hydrops (EH) is an idiopathic condition characterized by the enlargement of endolymphatic volumes in the inner ear with distension of Reissner’s membrane into the scala vestibuli of the cochlea and/or the saccule, utricle, and ampullae of the semicircular canals. EH is the pathogenic mechanism that underlies Ménière’s disease, a disorder of the inner-ear characterized by fluctuating hearing loss, vertigo attacks, tinnitus, and aural fullness.

EH typically results in SNHL; however, low-frequency ABGs can also be found in the absence of middle-ear pathology. An ABG in EH can be explained by the onset of increased perilymphatic pressure with a consequent decrease of stapes mobility or by saccular dilation, leading to a reduction in stapedial mobility as found by Okuno and Sando in the temporal bones of patients with Ménière’s disease.

Recently, 3-tesla MRI has been used to evaluate for the presence of EH, revealing a direct relationship between vertigo attacks and low-frequency ABGs due to a worsening of EH. Sugimoto et al. noted that the average bone-conduction thresholds in patients with EH were higher in those with EH adjacent to the stapes footplate, with more frequent ABGs at 250 Hz. Moreover, these authors found that patients with significant cochlear and vestibular EH adjacent to the stapes footplate had more frequent and longer Ménière’s disease crises relative to those with EH nonadjacent to the stapes footplate. Elsewhere, Maheu et al. found that the mean ABG frequency is clinically higher in patients with Ménière’s disease than in EH patients, without noticing a significant correlation between the mean ABG frequency and the summating potential (SP)/action potential (AP) area or amplitude ratio. However, Bess et al. reported three cases of unexplained CHL suggesting that the cause of secondary EH and subsequent ABGs in these cases could be an anomaly of the inner-ear fluids.

In 1965, Godlowski first described the causative relation between altered venous drainage of the vestibular and/or the cochlear veins into the venous cerebrospinal system and secondary EH. Cassandro et al. presented a case of dural arteriovenous fistulas in a 55-year-old female patient with unilateral left ear pulsatile tinnitus and CHL, suggesting that type II DAVF provokes subclinical intracranial hypertension with a prolonged parenchymal phase of the venous outflow. This mechanism could be responsible for a disturbance in the stria vascularis circulation, resulting in pulsatile tinnitus, and CHL. To our knowledge, only one case of hearing loss associated with DAVF has been reported in the literature; however, the hearing loss was sensorineural. An alteration in the venous cerebral circulation can indicate otologic symptoms like pulsatile tinnitus as a result of labyrinth hydromechanics in conditions such as bilateral transverse sinus stenosis or DAVF. In these conditions, there is a communication between intracranial fluids and the cochlear aqueduct.

Clinical Investigations
The typical conditions presenting with an ABG are pathologies af-
fecting the outer or middle ear. Therefore, the completion of accurate otoscopic, nasal, and rhinopharyngeal examinations is mandatory to assess the external auditory canal, tympanic membrane, and peritubal areas, followed by complete immittance testing. Details about the diagnostic strategy to differentiate middle-ear involvement, third-window lesions, and EH conditions are suggested in Table 1.

Findings of a type A tympanometry curve and normally evoked acoustic reflexes in patients presenting with an ABG generally exclude an alteration of the middle-ear and should suggest a third-window lesion [11]. Cervical VEMP testing and the measurement of otoacoustic emissions are also helpful in elucidating the ABG condition, as both cervical VEMP and otoacoustic emissions are present in third-window lesions and absent in patients with middle-ear disease [12,51]. Furthermore, cervical VEMP can be beneficial in evaluating EH, showing a biphasic complex greater in amplitude and lower in threshold for 1,000 Hz tone-burst stimuli relative to 500-Hz ones (tuning shift) [32]. An elevated SP/AP ratio as measured by electrocochleography has long been considered the electrophysiological correlate of EH-related clinical conditions. Similarly, an SP/AP ratio higher than 0.37 is considered indicative of EH [32].

Laser Doppler vibrometry can also help in differentiating middle-from the inner-ear diseases in patients with ABG when measuring umbo velocity [32]. Bedside vestibular examination can be useful to evaluate patients affected by third-window lesions given that sound stimulation (Tullio phenomenon), tragal compression (Hennebert sign), or pressure changes in the external ear canal (Valsalva maneuvers) may elicit dizziness and vertical-torsional nystagmus [53].

Imaging of third-window mechanism-based lesions relies on CT and MRI. High-resolution CT of the temporal bone is recommended to correctly evaluate third-window lesions [24]. CT has a central role in identifying semicircular canal dehiscence; MRI is equally sensitive in differentiating fluid from bone but should not be used as the preferred tool in the evaluation of third-window lesions [14]. MRI alongside magnetic resonance angiography may be useful for assessing central vascular abnormalities like DAVF. EH may be evaluated using MRI at 24 hours after the intratympanic administration of gadolinium-based contrast [53].

CONCLUSION
An ABG is an audiological finding typical of pathologies that involve the external or middle-ear; sometimes, an ABG can be found in conditions affecting the inner-ear due to a third-window mechanism or in idiopathic or secondary EH conditions such as central vascular abnormalities. Although making a definitive diagnosis is often challenging, accurate clinical, audiological, and vestibular evaluations can be suggestive of the underlying condition; however, radiological assessments by CT and/or MRI are mandatory to confirm any diagnostic suspicion.

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