

## Isolated Aplasia of Lateral Semicircular Canal

Berat Demir<sup>1</sup>, Bülent Mamikoğlu<sup>2</sup>

<sup>1</sup>Department of Otorhinolaryngology-Head and Neck Surgery, Pendik Training and Research Hospital, Marmara University Medical Faculty, Mimar Sinan Caddesi No. 41, Fevzi Cakmak Mahallesi, Ust Kaynarca-Pendik, 34899 Istanbul, Turkey.

<sup>2</sup>University of Illinois College of Medicine Department of Neurosurgery 1 Illini Dr, Peoria, IL.  
*drberatdemir80@hotmail.com*

**Abstract:** We aim to review the recent literature about inner ear embryology and discuss the audio vestibular findings in a patient with isolated lateral semicircular canal (LSCC) aplasia. Patient presented with dizziness for the past two years along with lightheadedness and spinning sensations. Radiological and vestibular test results are reviewed. Isolated lateral semicircular canal aplasia is a problem of embryological origin, it may manifest itself with audio vestibular complaints in late ages.

**Keywords:** Semicircular canals; Sensorineural hearing loss; Dizziness; Vestibular labyrinth

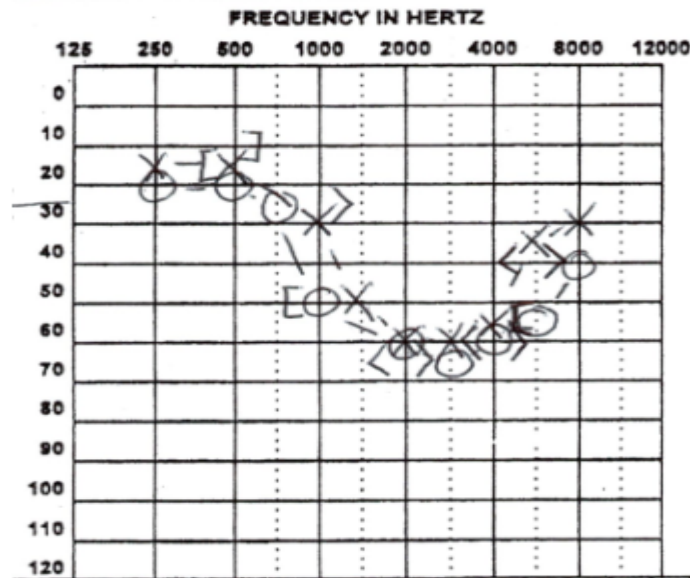
### INTRODUCTION

Aplasia of lateral semicircular canal (LSCC) is the most common congenital labyrinthine anomaly. LSCC aplasia is the result of incomplete partition from vestibular anlage<sup>[1]</sup> Usually aplasia of semicircular canal (SCC) cases are seen in association with syndromic or non-syndromic congenital anomalies of cochlea<sup>[2,3,4]</sup>. The LSCC is the last of the three SSCs which develops during the fetal period, and temporal bone histopathologist suspected that is why it is the most common labyrinthine anomaly<sup>[5]</sup>. Our report reviews the recent literature about inner ear embryology and discusses the audio vestibular findings in a patient with isolated LSCC aplasia.

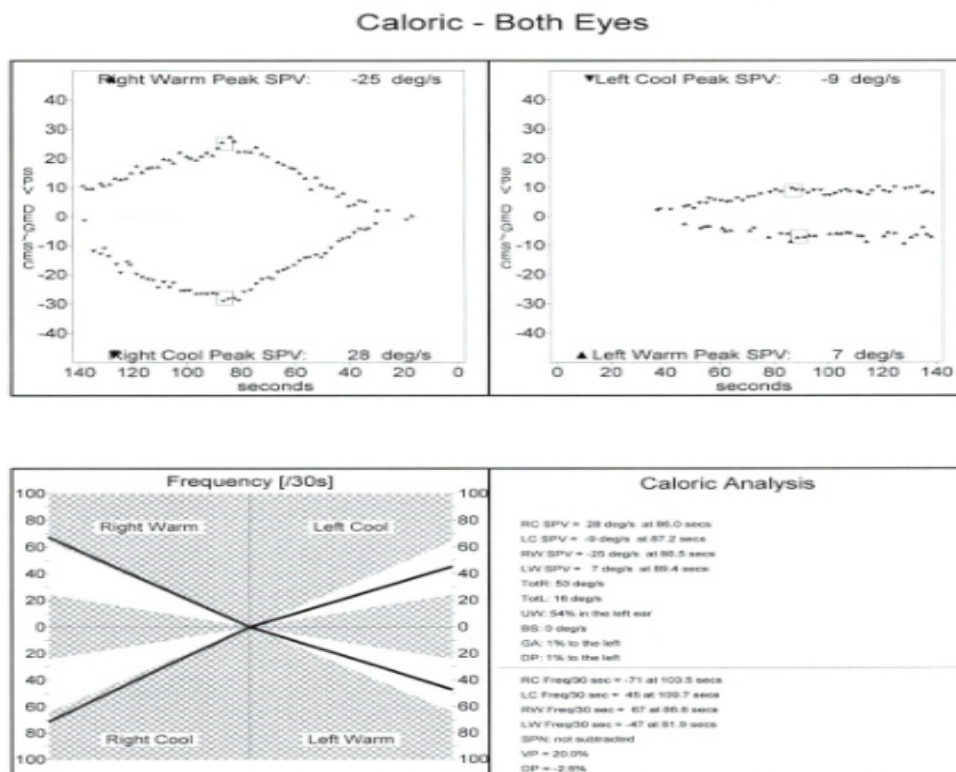
### CASE PRESENTATION

A 46-year-old woman presented with dizziness for the past two years with lightheadedness and spinning sensations. Her dizziness was characterized with spontaneous episodes which last seconds. Between episodes she felt normal. She could tell when she was going to have an attack as it starts with a pressure feeling in her head. She has history of severe migraine headaches and her mother had left sided Meniere's disease. Otoscopic examination was normal. Pure tone air and bone conduction audiometry revealed normal low frequency hearing, falling to a moderately-severe mid-frequency sensorineural hearing loss, improving to mild high frequency sensorineural hearing loss, slightly poorer for her right ear at 1000 and 6000-8000 Hz (Figure 1). There was a very weak spontaneous nystagmus to the right, which was barely noticeable at right gaze under examination with video Frenzel goggles. Vibration to mastoid triggered brisk right beating nystagmus. Caloric test revealed left unilateral labyrinthine weakness (Figure 2). VEMP (vestibular evoked myogenic potentials) showed normal response at the right but there was no response at left (Figure 3). Radiological assessment with Computerized Tomography (CT) and Magnetic Resonance imaging (MRI) showed us left LSCC aplasia (Figure 4A-B, 5A-B, 6A-B). No cochlear malformation was seen in either side. The internal auditory canal and the cerebellopontine angle were strictly normal.

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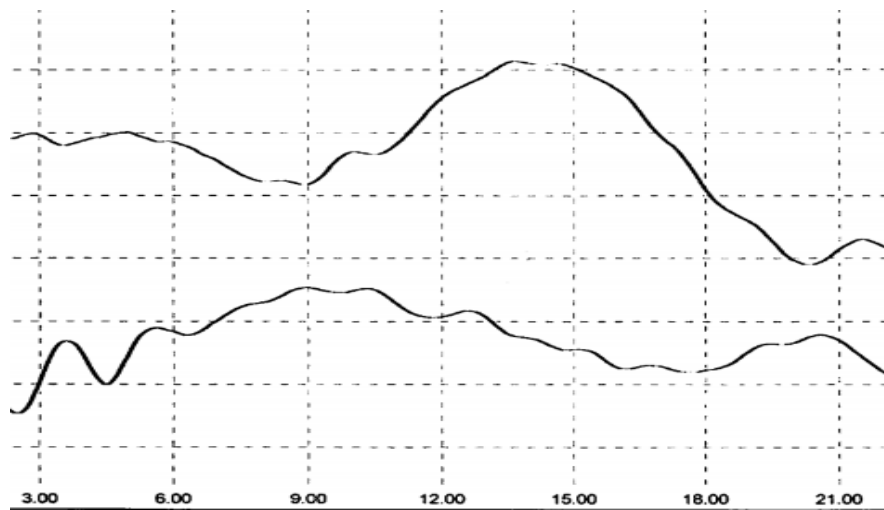
**Figure1.** Pure tone air and bone conduction audiometry shows normal low frequency hearing, falling to a moderately-severe mid-frequency sensorineural hearing loss, improving to mild high frequency sensorineural hearing loss, slightly poorer for her right ear at 1000 and 6000-8000 Hz. Speech reception thresholds were consistent with pure tone responses obtained, 25 dB at the right and 20 dB at the left ear. Word recognition scores were excellent for each ear, 92% at the right and 100 % at the left ear.



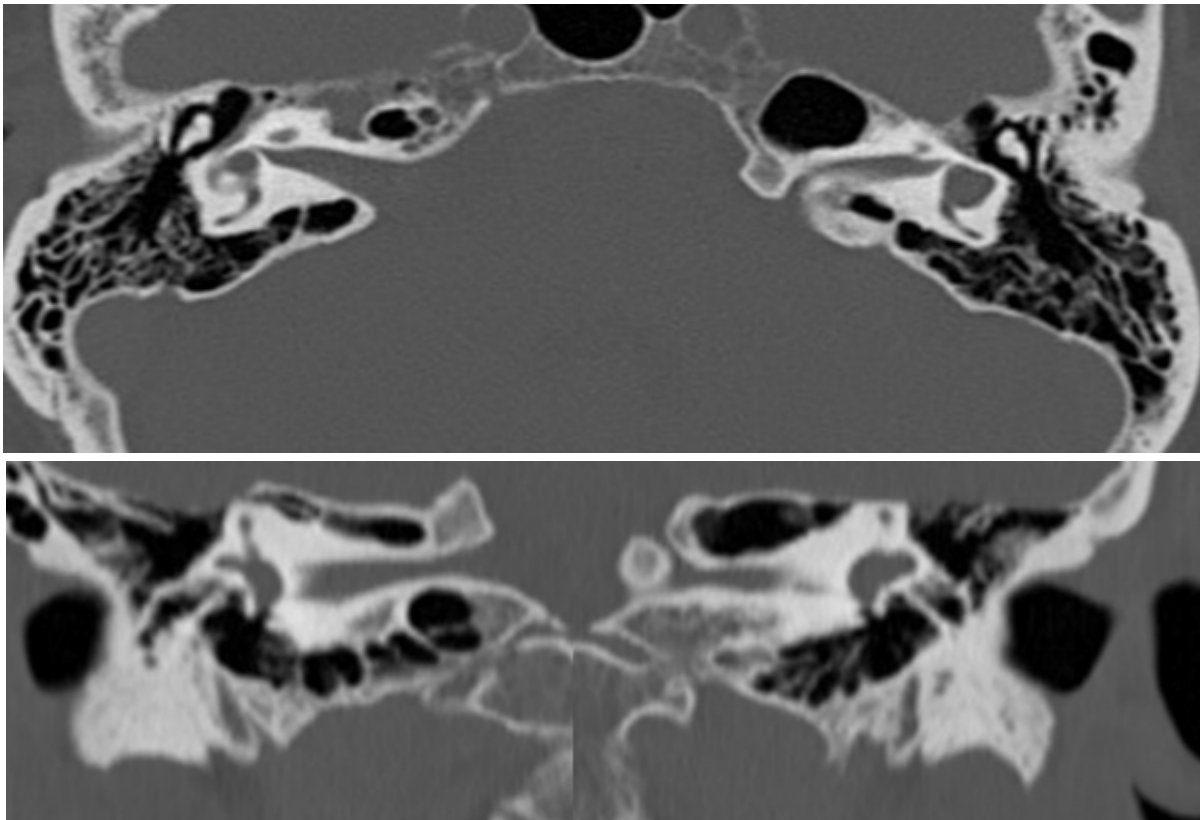
**Figure2.** Caloric test, total right ear is 53 deg/sn and total left ear is 16 deg/sn. 54% unilateral weakness at the left is seen.

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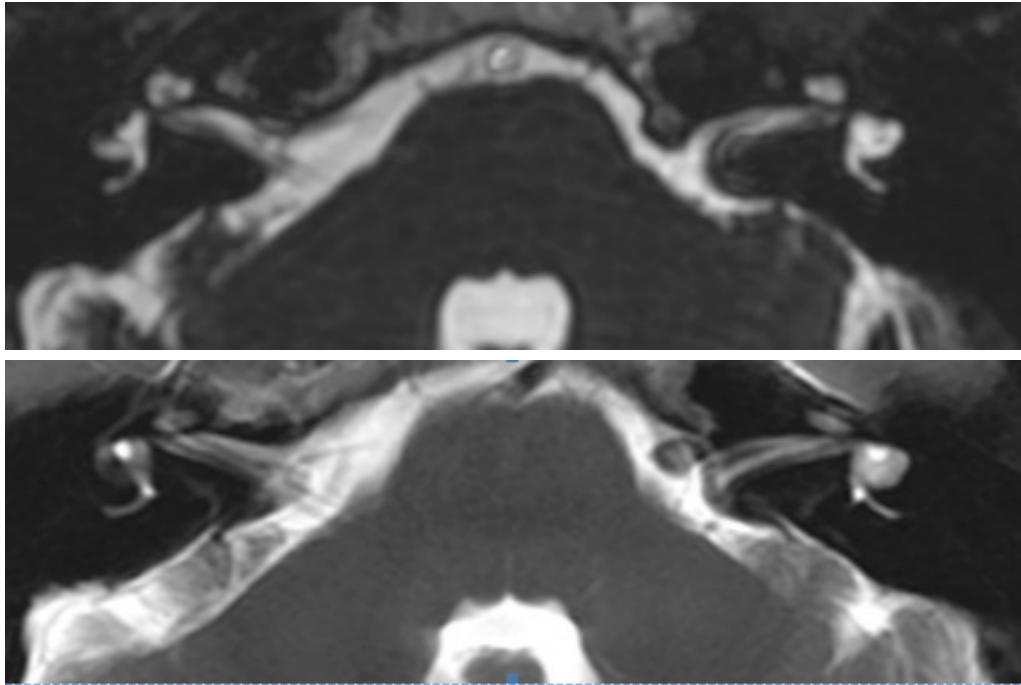
**Figure3.** VEMP testing was completed with air conduction clicks through inserts at intensity of 95dB. Tested subject kept 40 mm pressure through a cuff for each side to contract sternocleidomastoid muscle. There is good brisk response at the right side, P13 is detected at 13.1ms and N23 at 20.4 ms with amplitude difference of 12.8  $\mu$ V, no response at the left side.



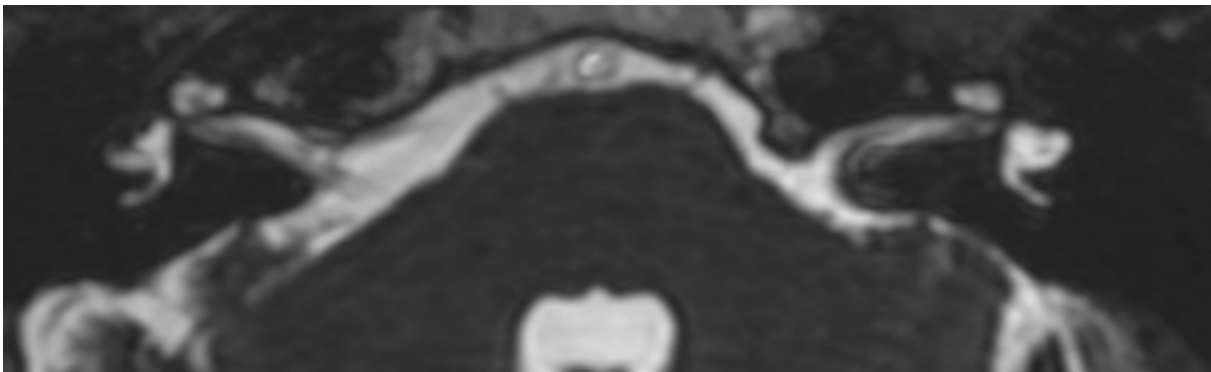
**Figure4 A, B.** Figure A is axial and B is coronal images. The right side is normal, the left vestibule is in common cavity with lateral canal, where the vestibule is seen as irregular cystic cavity, the other semicircular canal, cochlea, facial nerve and vestibular aqueduct is unremarkable. The internal auditory canal is normal and the "Bill's Bar" is clearly visible on axial images.

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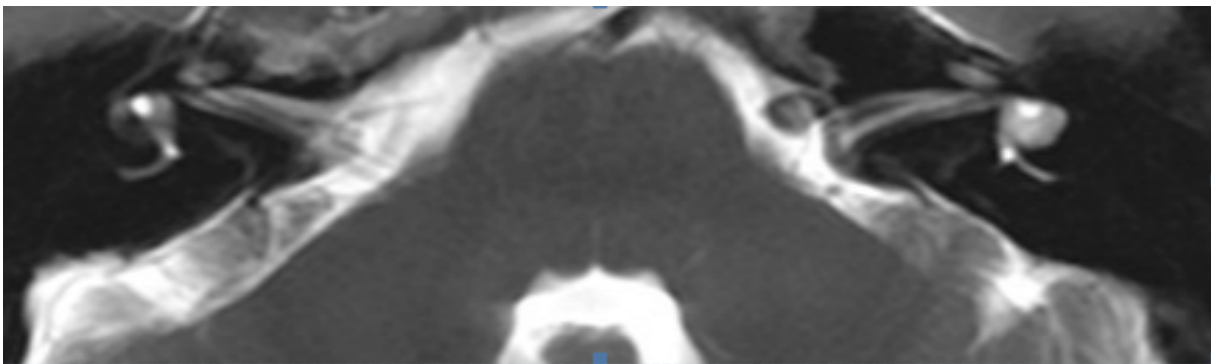
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**Figure 5 A & B.** These are axial MRI images, A is T2, and B is T2 with contrast. The right side is normal, on the left the vestibule is in common cavity with lateral semicircular canal. Other semicircular canals, cochlea and 7th/8th cranial nerve complex is normal.



**Figure 6a.** Axial MRI T2



**Figure 6b.** Axial MRI T1

### DISCUSSION

The development of the vestibular system is made up with Fgf (fibroblast growth factor) , RA (Retinoic acid) and Wnt signaling. RA and Fgf continue to pattern the ear at otic vesicle stages to set up sensory, neurogenic and non-neural domains in ventral otic epithelium. Fgf promotes, while RA restricts, the otic expression of *otx1b*, which has a conserved role in formation of the horizontal semicircular canal [6]. By week 5, dorsomedial part of the otic vesicle grows in order to form the utricle and semicircular canals. Towards the ventrocaudal area the pars sacculocochlearis develops, due to constitute the cochlear duct and saccule [2]. By week 6, the SCCs and the cochlea can be identified. Presumably, the SCC malformation is the result of an abnormal fusion during embryological development. Kiernan et al. showed that SCC development is encoded in the proximal portion of chromosome 4 near the centromere [7].

There is no consensus about caloric response findings in LSCC aplasia patients though clinicians predict to see a non-responsive ear [8]. Some will be seen, as in our case, to be responsive in the presence of significant weakness. Thus this suggests lateral canal may not be the sole area responsible for the caloric response [9].

A compensated peripheral vestibular nystagmus is often suppressed by visual fixation but could be seen during examination with Frenzel goggles. The response to vibration of mastoid can be helpful. Another use of vibration is in evaluating Meniere patients after gentamicin injection a brisk nystagmus towards the good ear confirms the gentamicin effect weeks after the injection. Without goggles no nystagmus would be seen in normal subjects until the frequency of oscillation approaches 1 Hz or the peak velocity is greater than 60°/s [10].

Patients with aplasia may also present with episodic spells as in our patient. There is an MRI study reported 76.7% endolymphatic hydrops in the aplasia group compared to 27.8% in the control group ( $P < 0.001$ ) [11]. Histopathologic reports do not support these MRI findings [2].

Sensorineural hearing loss is usually seen in association with LSCC malformations in the presence of cochlear abnormally but a number of LSCC patients presented with conductive hearing loss are also reported [12]. Our patient's imaging cochlear characteristics are normal, but she has significant bilateral high frequency hearing loss. Although in migraineurs, auditory symptoms are not as common as vestibular symptoms, this issue must be considered as hearing loss can be seen in some migraineurs [13,14]. Fluctuating or permanent sudden sensorineural hearing loss has been described in migraineurs, and these patients characteristically had concomitant with neurological phenomena (such as retinal migraine, hemiplegia, and/or visual aura) as a result of vasospasm. The other explanation is concurrent organ of Corti lesions (Alexander's dysphasia/aplasia) might be present, mostly at the basal turn of cochlea and leading to her hearing loss. Thus, in her hearing mostly high frequencies are affected. Given the severe labyrinthine abnormally this explanation is more likely possible.

### CONCLUSION

Aplasia of lateral semicircular canalis the most common congenital labyrinthine anomaly. Although aplasia of LSCC is a congenital anomaly, it may develop by giving symptoms in older ages. Therefore, aplasia of LSCC should not be forgotten in patients with complaints of dizziness in older ages.

### Informed Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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