Semicircular canal malformations: clinical and instrumental findings

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Anomalies involving only the semicircular canal (SCC) are classified as a mild inner ear malformation. Among these the lateral SCC dysplasia (shortening and widening) is the most frequent. The results of several studies have not shown any consistent relationship between SCC malformations and hearing loss. As for cochlear function, few reports have examined the vestibular symptoms and any correlation between the severity of the canal and the vestibular malformations and vestibular impairing exists.

The purpose of our study was to describe the cochleovestibular assessment in patients with sporadic semicircular canal malformations and to suggest possible explanations for the variability of symptoms and signs induced by SCC anomalies.

The study was performed in 22 adults patients affected by lateral semicircular canal dysplasia identified in the ENT and Audiology Unit of the University of Bologna from January 2010 to December 2014. All patients were examined by means of appropriate temporal bone high resolution computed tomography (HRCT) study with multiplanar reconstruction and were screened to exclude central nervous system and eighth cranial nerve enhancing lesions using contrast-enhanced brain magnetic resonance imaging (MRI). A careful anamnestic evaluation and a full audiovestibular test battery were attempted for each patient (pure tone audiometry, infra-red videonystagmoscopy (head pitch test, Hallpike maneuver, bilateral mastoid 100 Hz-vibration, head shaking test and Valsalva maneuver with pinched nostrils), bithermal caloric test, cervical and oculovestibular evoked myogenic potentials study (air and bone conducted stimuli), head impulse test using video-HIT in the planes of all 6 semicircular canals).

All the patients bilaterally had the SCC malformation: 15 of 22 patients had bilateral lateral SCC dysplasia; 3 patients had left LSCC dysplasia and right LSCC aplasia; 1 patient had aplasia LSCC bilaterally; 1 patient had SCC dysplasia of all 6 SCC; 1 patient had posterior SCC aplasia and contralateral posterior SCC dysplasia; 1 patient had LSCC dysplasia and anterior arm superior SCC aplasia. None of the patients had cochlear malformations or enlarged vestibular aqueduct. No external ear, middle ear or osseous abnormalities were noted on imaging. 6 of 22 patients had Meniere-like clinical history.

We will describe in detail the cochleovestibular instrumental findings.

n. 18 Adults- LSCC DYSPLASIA
F/M 10/8
Bilateral 100%

Associated inner ear malformations (EVA o Cochlea o SCD): 0

36 ears
- Isolated LSCC Dysplasia 30
- 6 SCC 2 (asynchronous bilateral recurrent hydrops)
- PSCC Dysplasia + PSCC Aplasia 1
- LSCC Dysplasia CSL 2 + anterior arm SSCC Aplasia 1 (no vertigo)

VESTIBULAR AQUEDUCT
- Normal 18
- Hypoplastic 7
- Borderline 2
- Long 1
- deisc GG 6
- deisc PSCC 2

Thinning SSCC 5

Hearing loss absent 17
36 au
HF SNHL 7
LF SNHL 4
Panotal SNHL 6 (MD)
U-shaped SNHL 2

Vestibular symptoms absent 7
- Atypical BPPV 3
- MENIERE-like 6
- Dizziness 1
- Barogenic vertigo 1

Vestibular function LSCC dysplasia
8 pt (TC + v-HIT) - monolateral hyporeflexia + v-HIT NN - 6
- Bilateral hyporeflexia + v-HIT NN - 1
- Bilateral normal reflexia + v-HIT NN - 1

VEMPs – NN 100%