Positional nystagmus in superior semicircular canal dehiscence syndrome

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SNOF årsmöte Marstrand 2014
Third window: fysiopathological implications

- SCDS is one of the third window syndromes (LVAS, inner ear malf.)
- The presence of a third window implies:

A. Reduced inner ear impedance (modified thresholds for cochlear and vestibular stimulation)
B. Alternative pathway to the inner ear for inappropriate stimuli (sound, vibration and pressure changes)
### Vestibular

**Symptoms**
- Vertigo/oscillopsi induced by sound (Tullio phenomenon)
- Vertigo induced by pressure changes (Hennebert phenomenon)
- Chronic dizziness
- Vertical oscillopsi
- Drop attacks

**Signs**
- Vertical torsional nystagmus induced by loud sound
- Vertical torsional nystagmus induced by pressure changings in the ear/Valsalva
- Pulse-synchronous torsional vertical nystagmus
- Positional vertical torsional nystagmus

### Auditory

**Symptoms**
- Autophoni
- Pulse-synchronous tinnitus
- Intolerance for loud sounds
- Hearing loss

**Signs**
- Conductive hearing loss with normal tympanometri/stapedial reflexes
- Conductive hyperacusi
- Weber test lateralized
- Weber at malleolus lateralized
SCDS: diagnostic criteria (Audiology and Neurotology Department Karolinska)

- conductive hyperacusis/”false” middle ear air-bone gap
- sound/pressure induced nystagmus/vertigo
- enhanced response at VEMP
- canal dehiscence evident at parasagittal TC scans
1/8 patients with SCDS showed positional nystagmus

Vertigo and paroximal vertical-torsional nystagmus when sitting up from supine or prone position
M, 59 years old

– Head trauma
– Vertigo attack after some weeks
– Chronic dizziness and intolerance for fast head movements
– One side hearing loss after head trauma
– No positional vertigo referred
Videooculography (vertical recording)

sitting

prone

sitting right dix hallpike sitting
Tone burst 500 Hz 130 dB spl

Click bil 90 dB HL
CT
M, 49 years old

- After a dental drilling debut of positional vertigo, left hearing loss, autophonia (footstep sound)
- No sound/pressure induced symptoms
Videooculography (vertical recording)
Tone burst 500 Hz 130 dB spl

Click bil 90 dB HL
CT left
CT right
J. 40 years old

- Unprovoked debut of dizziness with increasing intensity under 2 years
- Increasing intolerance for sound/vibration and pressure changes
- Impossibility to take a plane for enhanced dizziness with pressure changes and intolerance for accelerations.
VEMP

Tone burst 500 Hz 130 dB spl

Click bil 90 dB HL

Right: 3.90  
Left: 3.0

Right: 1.7  
Left: 1.3
CT left
CT right
Videooculography (vertical recording)
Clinical features of positional nystagmus in SCDs

– Vertical/torsional in sagittal/parasagittal head positioning
– Downbeating, torsional towards the affected side in the up sitting from supine or prone positioning
– Upbeating, torsional towards the unaffected side in the supine or prone positioning
– Paroxismal dynamic, sometimes persistent
– Not responding to repeated repositioning maneuvers
Pathophysiology – sound/pressure induced nystagmus
Pathophysiology– sound/pressure induced nystagmus
Pathophysiology – positional nystagmus

Movement of Cerebrospinal Fluid within the Craniospinal Space when Sitting Up and Lying Down

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Positional nystagmus in BPV and SCDS: similarities and differences

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<td>Up beating in Dix Hallpike and down beating in up sitting</td>
<td>Up beating in Dix Hallpike and down beating in up sitting</td>
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<td>Torsional component towards the affected side in Dix-Hallpike</td>
<td>Torsional component away from the affected side in Dix-Hallpike</td>
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<td>Torsional component away from the affected side in up sitting</td>
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<td>Nystagmus more consistent in Dix-Hallpike</td>
<td>Nystagmus more consistent in up sitting</td>
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<td>Paroxysmal</td>
<td>Paroxysmal/persitent</td>
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<td>Responding to repositioning maneuvers</td>
<td>Not responding to repositioning maneuvers</td>
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<td>Latence</td>
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Conclusion

– SCDS can show a positional/positioning nystagmus
– this nystagmus is vertical/torsional and points out the stimulation of the superior semicircular canal
– it is probably due to the intracranial pressure changing during positioning
– the SCDS nystagmus can resemble the typical nystagmus in posterior canal BPV of the other side
– in case of atypical BPV consider the SCDS of the other side.