Otology seminar
Superior semicircular canal dehiscence syndrome

一、Introduction
- Definition: Absence of bone overlying superior semicircular canal resulting in a third window to the membranous labyrinth
- first described in 1998 by Minor et al: 8 patients of oscillopsia, vertigo induced by sound or pressure (Tullio phenomenon and Hennebert sign)
  - Sound: 250~4000Hz with intensity of 100~110 dB
  - Middle ear (pneumotoscopy, Valsalva with nose pinched)
  - Intracranial (Valsalva against closed glottis)
  - 1 mm slice thickness CT: superior semicircular canal dehiscence
- Smullen et al published 3 cases in 1999, two patients improved after surgical repair of the dehiscence via middle fossa approach
- Prevalence: Carey et. al. studied 1000 temporal bone (596 individuals)
  - 0.7% complete dehiscence
  - 1.3% had a very thin bony layer (< 0.1mm)
  - Symptomatic SCD difficult to assess in population
- Etiology: congenital or developmental defect -> trauma or increased intracranial pressure induced dehiscence

二、clinical manifestations
- Vestibular Manifestations
  - Chronic dysequilibrium
  - Oscillopsia: often in vertical-torsional plane
  - Tulio phenomenon
    - May be caused by syphilis, Perilymphatic fistula,
Meniere’s disease, Cholesteotoma

- Hennebert’s sign
  - May be caused by syphilis, Perilymphatic fistula, Cholesteotoma
- Vertigo induced by Valsalva maneuver

**Auditory Manifestations**

- Increased sensitivity to bone conducted sounds
  - Autophony: autophony of voice but not breathing, may relieve by head dependent position as Patulous Eustachian tube
  - Hear one’s footsteps, eye movement
- Fork Testing
  - Rinne: BC > AC
  - Weber: lateralize to dehiscence side
  - hear tuning fork when placed on distal bony skeleton(ankle)
- Audiometry
  - lowering BC thresholds < 0 dB HL at 250 and 500 Hz
  - --> ABG>10 dB
  - positive acoustic reflex
Lloyd B. et al, 2005 collect 65 SSCD patients based on CT and at least one sign

- Symptoms in Patients with Vestibular Manifestation
  - 90% with Tulio
  - 73% with pressure induced vestibular symptoms
  - 67% with both
  - 60% with autophony

- Signs in Patients with Vestibular Manifestation (90%)
  - 82% with sound evoked eye movements
  - 75% with Valsalva eye movements
  - 45% with Hennebert’s sign

### Pathophysiology

- **Third Window Hypothesis:**
  - Sound or pressure induced vestibular symptoms
  - Positive ear canal sound pressure pushes stapes into the vestibule-> pressure release point in the SSC-> endolymph flow-> excitation of the cupula
  - Increase vestibular pressure or an increase in CSF pressure-> inhibition of the cupula
  - Hirvonen et al. in 2001, vestibular nerve afferent in chinchilla
    - inhibited by negative pressure and excited by positive pressure on the EAC after fenestration of SSC
    - reversed after repair of fenestration
- decrease sensitivity of air conduction
  - some fraction of the perilymphatic volume is “shunted” through SSC from the cochlea, resulting in a decrease in the sound conducted to the cochlea

- increase sensitivity of bone conduction
  - Vibration of skull introduces oscillating compressions and rarefactions of the bone around the inner ear lymph—> set fluid into motion
  - SSCD allow larger than normal fluid motions—> increase in cochlea’s response to compressional wave
四、Diagnosis

- Oculography with sound and pressure

- Audiometry with acoustic reflex
  - Air bone gap at low frequency
  - Positive acoustic reflex differentiated from otosclerosis

- VEMP
  - Lower threshold in VEMP response (<80 dB)
  - Increased threshold or absent response in middle ear abnormality
  - Amplitude larger on the affected side than healthy side
  - Sensitive but some limitation:
    - Overlap between SSCD and normal groups
    - Not measurable in all patients
    - Enlarged vestibular aqueduct and posterior semicircular canal dehiscence
The roof of SSC can be composed of one to three layers. Otic capsule, trabecular bone (air cell), and cortical bone (petrous pyramid).

Reformatted Oblique coronal view

- Belden et al 2003, Temporal bone CT scans with 0.5-mm collimation and reformation improve specificity and positive predictive value

<table>
<thead>
<tr>
<th>TABLE 3</th>
<th>Diagnostic Discrimination of Temporal Bone CT for Detecting SSC Dehiscence When Scanning with 1.0-mm or 0.5-mm Collimation in Patients with Symptoms of Sound- or Pressure-induced Vertigo and Oscillopsia Combined with Data from the Control Populations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Performance Measure</td>
<td>1.0-mm-collimated CT</td>
</tr>
<tr>
<td>Sensitivity</td>
<td>27/27 (100)</td>
</tr>
<tr>
<td>Specificity</td>
<td>112/139 (81)</td>
</tr>
<tr>
<td>Positive predictive value</td>
<td>27/54 (50)</td>
</tr>
<tr>
<td>Negative predictive value</td>
<td>112/112 (100)</td>
</tr>
</tbody>
</table>

Note.—Data are number of ears. Numbers in parentheses are percentages. For subjects in the control population undergoing CT with 1.0-mm collimation, evaluations of “possible” or “definite” dehiscence were considered abnormal for the purposes of these statistical calculations.
Benjamin T. et al 2008

- 3D reconstruction CT
  - highly filtered to remove adjacent brain tissue and soft tissue
  - higher false positive and larger dehiscence
  - helpful for surgical planning
- Multiplanar reconstructions of HRCT are most sensitive
- diagnosis should be based on MPR of HRCT plus one additional objective finding (low threshold VEMP, conductive HL, tone evoked eye movement)

![Images of CT scans](image.png)

**Table 1. Statistical summary of various tests for SCD**

<table>
<thead>
<tr>
<th></th>
<th>TP</th>
<th>TN</th>
<th>FP</th>
<th>FN</th>
<th>PPV, %</th>
<th>NPV, %</th>
<th>Sensitivity, %</th>
<th>Specificity, %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Multiplanar CT</td>
<td>27</td>
<td>15</td>
<td>4*</td>
<td>0</td>
<td>87</td>
<td>100</td>
<td>100</td>
<td>79</td>
</tr>
<tr>
<td>3-D CT reconstruction</td>
<td>21</td>
<td>9</td>
<td>10</td>
<td>2</td>
<td>68</td>
<td>82</td>
<td>91</td>
<td>47</td>
</tr>
<tr>
<td>VEMP</td>
<td>16</td>
<td>12</td>
<td>3</td>
<td>4</td>
<td>84</td>
<td>75</td>
<td>80</td>
<td>80</td>
</tr>
<tr>
<td>Conductive hearing</td>
<td>19</td>
<td>18</td>
<td>1</td>
<td>4</td>
<td>95</td>
<td>82</td>
<td>83</td>
<td>95</td>
</tr>
<tr>
<td>Tone evoked nystagmus</td>
<td>14</td>
<td>18</td>
<td>0</td>
<td>7</td>
<td>100</td>
<td>72</td>
<td>67</td>
<td>100</td>
</tr>
</tbody>
</table>

*The 4 false-positive results on MPR were asymptomatic on that side and had no conductive hearing loss, normal VEMP, and no sound-induced nystagmus.
五、Treatment

- avoid stimulus
- debilitating symptoms need surgical procedure
  - middle fossa approach: plugging of SSC is better than resurfacing
    - 4 of 11 symptoms recurrence after resurfacing
  - transmastoid approach: Sumit K. Agrawal et al 2008
    - successful in 3 cases
    - limited in dura lower hanging or extensive cranial base dehiscence
  - plugging material: bone dust > muscle > bone wax
  - improve pressure- and noise-induced nystagmus, conductive HL, autophonia, and chronic imbalance
-并发症
  - SNHL
  - Vestibular hypofunction
Reference:


5. Minor LB, Labyrinthine fistulae: pathobiology and management., Curr Opin...


10. Murtaza Kharodawala, MD, The Third Window: Superior Semicircular Canal Dehiscence, UTMB grand round