Superior Semicircular Canal Dehiscence
Deborah Farragher, Judith White
Section of Vestibular and Balance Disorders

Superior Semicircular Canal Anatomy and Physiology

Superior semicircular canal dehiscence (SSCD) syndrome

- First described by Minor et al. 1998
- Sound or pressure induced eye movements
- Defect in the bone overlying the SCC forms an abnormal connection between the vestibular system and intracranial compartment.

SSCD Nystagmus

Positive pressure in the external auditory canal and Valsalva against pinched nostril causes upwards slow phase nystagmus directed away from the affected ear.

Negative pressure in the external canal and Valsalva against a closed glottis cause slow phase nystagmus downwards and towards the affected ear.

Fixation will suppress the nystagmus, and videonystagmography is helpful.

Eye movements align with the abnormal canal

- Tonic upwards and ipsilateral intorsion is seen with ampullofugal endolymph flow - activation (loud noise, positive middle ear pressure)
- Tonic downwards and ipsilateral extorsion is seen with ampullopetal endolymph flow - inhibition (Valsalva against a closed glottis, jugular compression, raised intracranial pressure)

SSCD

- Positive middle ear pressure or loud sound stimulates ampullofugal endolymph flow - canal activation
- Eye movements are tonic upwards with ipsilateral intorsion.
SSCD

- Valsalva against a closed glottis or jugular compression causes increased intracranial pressure and ampullopetal endolymph flow - canal inhibition
- Eye movements are tonic downwards with ipsilateral extorsion

SSCD

- Hypothesis: vibration acts on the bony dehiscence when applied in a cranio-caudal direction to set up bidirectional superior semicircular canal endolymph flow and cause the characteristic torsional nystagmus.
- Are the otolith organs (saccule and utricle) involved in the response, particularly in bilateral cases where vertical nystagmus is seen?

Physical Exam

- Loud tones in left ear showed nystagmus with vertical and torsional components

Proposed Mechanism

Tullio Phenomenon

- Initially described by Tullio based on experiments in animals where the bone was fenestrated overlying the semicircular canal and eye and head movement were evoked in the plane of the canal with loud noise (Alexander's law). Later used to describe findings seen in patients with syphilitic erosion of the labyrinth or advanced Meniere's with labyrinthine adhesions.
- Test at 2000 Hz at 110 dB

Symptoms Associated with SSCD
Typical presenting symptoms

• Chronic disequilibrium (76 %)
• Sound evoked disequilibrium or visual instability
• Pressure evoked disequilibrium or visual instability
• Conductive hyperacusis (unusually sensitive to vibration) 39%

Common Presenting complaints

• Sound induced disequilibrium
  – Loud noises cause imbalance or frank vertigo
  – Organ music
  – Fire alarm
  – School bells (teachers)
  – Loud telephone or music
  – Audiologist (demonstrating audiologic screening on herself)

Common Presenting Symptoms

• Hearing Loss
  – Muffled, blocked or echo in hearing
  – Usually low frequency
  – Typically reduced hearing for sounds presented thru the air, but unusually sensitive hearing for sounds presented via vibration (bone conduction)
  – Patient may hear a vibrating tuning fork applied to their ankle in their ear (conducts vibration thru skeleton)

Common Presenting Symptoms

• Pressure Induced Disequilibrium
  – Elevators
  – Scuba Diver
  – Sneezing
  – Flying
  – Straining
  – Lifting Weights

Less Common Associated Symptoms

• Hearing Eyes Move
• Hearing Heartbeat
• Hearing other usually imperceptible physiologic sounds (breathing, blood flow, joint movement)
• Visual instability with running, walking
• Atypical Positional Vertigo (minimal prolonged torsion in ear down positions)
  – Dura may pull on canal

Case Descriptions

47 yo with a 10 yr. hx. of gradually worsening vertigo and oscillopsia induced by loud noises, lifting or straining.

Audiogram showed a low frequency 20-30 dB conductive hearing loss with bone thresholds at –5 to –15 dB.
### More Cases

- 36 yo with oscillopsia with loud noises or tragal pressure/Valsalva. Constant disequilibrium. s/p stapedectomy for presumed otosclerosis (20-25 dB CHL, no benefit from surgery).
- CT confirmed SSCD

### More Cases

- 62 yo male intolerant to supine positions and pressure changes for 1 year. Prior outside audiogram did not test bone conduction. MRI negative.
- Vestibular physical therapy with repositioning unsuccessful.
- CT obtained, which showed large left superior semicircular canal dehiscence.

### More Cases

- 50 yo male seen in consultation from neurology. Complains of pulsatile tinnitus right ear and lightheadedness, disequilibrium with loud noise, clearing ears or pressure changes. Very difficult to work as a machinist. Having difficulty driving Harley Davidson.
- Audiogram showed characteristic low frequency conductive hearing loss right ear, with normal acoustic reflexes.
- CT confirmed large right SSCD

### More Cases

- 76 yo female with gait problems and frequent falls. Prior canal wall down mastoid surgery right, age 17. Right ear always sensitive to cold wind. MRI normal.
- Audiogram showed conductive hearing loss right ear (likely secondary to prior mastoid surgery).
- CT showed right SSCD

### More Cases

- 74 yo male with positional vertigo rolling to right and on awakening, frequent falls, poor balance.
- Vestibular physical therapy and repositioning maneuvers not successful
- Right torsional nystagmus in right ear down positioning, with persistent atypical positional nystagmus following 11 trials of repositioning
- No tinnitus or hearing change. Audiogram normal for age. MRI normal.
- Bilateral SSCD on CT

### More Cases

- 42 yo male professional scuba diver complaining of severe vertigo with pressure equalization
- Audiogram suggested low frequency conductive hearing loss bilaterally
- Vestibular testing normal
- CT showed bilateral SSCD, larger on the left
- Patient elected to continue diving against medical advice
**Diagnostic Tools**

**THE HISTORY**
- History is the most sensitive, useful and valuable test in diagnosing this disorder.
- ASK about noise sensitivity, pressure sensitivity and hearing loss.

**NEXT TEST: AUDIOGRAM**
- Look for bone conduction hearing above average, with low frequency reduced hearing for other sounds — low frequency conductive hearing loss. Affects 39% of patients with SSCD.
- Normal acoustic reflexes (these will be absent in other forms of conductive hearing loss, including otosclerosis and middle ear disease)

**Audiologic Analysis**
- Left ear low frequency conductive hearing loss

**Audiogram**
**Vestibular Evoked Myogenic Potential - VEMP**

- Audiology Test
- Applies loud sound to the ear
- Triggers relaxation of a contracting sternocleidomastoid muscle thru a reflex (saccule-brainstem-motor nerve)
- Unusually active in SSCD
- A useful screening test
- May be false negative if patient cannot tense neck, but rarely false positive

**Recommendations for Audiologists**

- For audiograms suggesting conductive hearing loss with normal tympanograms, consider VEMP testing. VEMP will be absent in conductive hearing loss due to otosclerosis or ossicular problems, but present in SSCD. Also consider testing for bone thresholds above 0 dB in this group. SSCD is associated with supra-normal bone thresholds (conductive hyperacusis).

**Imaging**

- High resolution temporal bone CT-scan showed dehiscence overlying left superior semicircular canal.

**Radiological Correlates**

- High resolution CT in plane of the canal
- Radiographically present in 20% of imaging studies
- Histologically present in 10% of temporal bones
- Greater than 5 mm in size, dural compression causes loss of superior canal function.

**Vibration-induced nystagmus**

Useful office procedure to screen for SSCD, uses vibration applied to base of skull while observing nystagmus (Frenzel lenses helpful)

**Vibration protocol**
**Signs and Findings in SSCD**
- Low frequency conductive hearing loss with supra-threshold bone conduction
- Low threshold of vestibular evoked myogenic potential (VEMP)
- CT temporal bones with fine cuts reconstructed in the plane of the superior semicircular canal demonstrate bony dehiscence between the top of the canal and the middle cranial fossa

**Treatment Options In SSCD**
- Diagnosis itself is very reassuring to patients
- Watchful waiting is a good management strategy. Symptoms often improve as dehiscence expands.

**Disabling Symptoms**
- Pressure Equalizing tubes may help
- Treatment of co-morbid exacerbating conditions such as migraine is very helpful

**Disabling Symptoms**
- Benzodiazepines are not indicated for the long term management of vestibular disorders
- Superior Canal Plugging
  - Middle cranial fossa or trans mastoid procedure with low risk of hearing loss or vestibular ablation in experienced hands

**Pearls**
- Dehiscence is often bilateral, since skull shape is usually symmetric
- Other canals may be dehiscent, with similar presenting symptoms
  - Mastoid surgery may thin lateral canal
  - Posterior canal dehiscence is possible, but less frequent
References


