Anatomy and Pathophysiology of the Vestibular System

Lecture Plan
1. Vestibular Physiology
2. Anatomy and clinical syndromes

Vestibular Physiology
- Vestibular sensors and reflexes
  - VSR
  - VOR
- Sensor imperfections and local brainstem compensation
- Central problems and higher level processing

Vestibular Overview

Two main reflexes
- VOR - vestibulo-ocular reflex
- VSR - vestibulo-spinal reflex

Vestibulo-ocular reflex V.O.R.
- Stabilizes eye in space
- Necessary to see while head is in motion
Vestibulo-spinal reflex
V.S.R.
- Stabilizes body
- Helps maintain desired orientation to environment

Vestibular system is needed to walk reasonably safely in the dark.

The vestibular system incorporates considerable redundancy.

6 degrees of freedom problem
- Three axes of rotation
  - Roll, pitch and yaw
- Three axes of translation
  - AP, Lateral, Vertical

The Navigation Problem.
- Motion sensing is a "mission critical" task -- for example, vestibular system is needed to walk reasonably safely in the dark.
- The vestibular system incorporates considerable redundancy.

The vestibular inner ear is an inertial navigation device
- Semicircular Canals are angular rate sensors.
- Otoliths (utricle and saccule) are linear accelerometers
- Bilateral symmetry means redundant design.
**5 sensors, 2 tests**
- Clinical Correlate: can only measure 2/5 -- lateral canal and saccule with available vestibular tests.

**Starting and Stopping = Acceleration**

**Otolithic Membrane**

**Utricle and Saccule orientation**

**Imperfections in Vestibular Sensors**
- Imbalance
- Timing
- Gain
- Noise
Imbalance

- Push-pull arrangement
- Common mode rejection
- Illusion of motion when one side goes bad

Vestibular Nystagmus

1. Both sides – no nystagmus
2. One side – lateral/rotatory
3. One horizontal canal – lateral nystagmus.
4. One vertical – mixed vertical/rotatory
5. Vertical or horizontal – usually central

Imperfections in Vestibular Sensors

- Timing of canals isn’t good for eyes or body
  - Need to extend timing for eyes
  - Need phasic emphasis for neck

Built in Timing Problems

Velocity Storage for VOR
Adjust to agile eye

Different timing needed for sluggish neck
In vestibular lesions

- Velocity storage goes away for eyes (VOR). Time constant drops from 21 to 7 sec.
- Not clear what happens to timing in the neck/body – may be unchanged.


Ewald’s 3 Laws (1892)

Observations made upon the exposed membranous labyrinth of Pigeons (Ewald's pneumatic hammer)

- Eye and head movements occur in the plane of the canal being stimulated and in the direction of endolymph flow
- In the lateral canal, ampullopetal flow causes a greater response than ampullofugal flow
- In the vertical canal the reverse is true

Ewald’s 2nd Law


Imperfections in Vestibular Sensors

- Gain
  - Ewald’s 2nd law – built in problem
  - Growth and development
  - Disease – bilateral vestibular loss

Ewald’s Compensation

need for both eyes and neck

Saturation → Anti-Saturation → Linear behavior

In unilateral vestibular loss, Ewald’s 2nd law probably causes head-shaking nystagmus, positive rapid-dolls head reflex. We are not sure what happens to VCR/VSR.

Imperfections in Vestibular Sensors

- Noise - a common problem
  - Positional vertigo (BPPV mainly)
  - Fluctuations in vestibular function
    - Ménière's, Fistula
- Noise makes vestibular input unreliable
  - Logically consequence is to decrease weighting

Clinical correlations

- Grocery Store Syndrome (AKA visual dependence)
  - Unable to tolerate busy visual environments
  - Normally people switch between most salient sensory mode
    - Visual-vestibular/somatosensory
  - Can't switch -> bothered by target

Higher Level Vestibular Problems

- Coordinate rotation is needed to communicate with VCR and VSR
- Integration is needed of vision and somatosensation with vestibular input
- Estimation is needed to process multiple unreliable sensors

Sensory Integration

- Visual, vestibular, somatosensory senses must be integrated to form best estimate.
- If incorrect estimate
  - Motion sickness
  - Visual dependence
    - Grocery store syndrome
    - Simulator sickness
  - Can't switch -> bothered by target

Internal Model Theory (how the brain works?)

- Outgrowth of Space program
- Space Shuttle – 100's of inputs and outputs
  - Some intermittent
  - Some more reliable than others
  - Some sluggish, some rapid
  - Some are noisy
- Needed a method of formally computing best estimate of Space Shuttle State

Kalman Filter (internal model)

- Grew out of work by Kalman at MIT
  - Formal method of forming "optimal estimate"
  - Integrates efference with afference
  - Accounts for noise, sensor differences.

Wolpert, 1997
Vestibular Anatomy and Disorders

Membranous Labyrinth

MRI of inner ear

Clinical Correlations

- Meniere’s disease (?)
- Meningitis in children
- Perilymphatic fistula

Vestibular Hair cells – measure force

- Relative movement of hair cells to head causes change in electrical potential
- Same general design for hearing
STARTING AND STOPPING = ACCELERATION

Clinical Correlation – Hair Cells
- Aminoglycosides kill hair cells
- Loop diuretics and NSAIDS are hair cell toxins

Membranous Labyrinth
Narrow lumen increases effect of viscosity
Allows mechanical integration to take place

Clinical correlates
Vestibular Atelectasis
Collapse of membranous labyrinth
May correlated with dysequilibrium in elderly population.

Peripheral circulation to inner ear
- AICA
  - Labyrinthine
  - Vestibulocochlear
  - Anterior vestibular
    - AC, IC, Utricle

Cupula to Brain
Cupula
Scarpa’s ganglion
Vestibular Nerve
Vestibular Nucleus
Cortex
**Vestibular Nerve**

- Superior vestibular nerve: AC, IC, Ovalis
- Inferior vestibular nerve: PC, Sacule
- Scarpa's ganglion

**Clinical Correlations**

- Vestibular neuronitis – infection of Scarpa’s ganglion?
- Acoustic Neurinoma
- Microvascular compression syndrome

**Vestibular Nucleus**

Major Nuclei (4)

1. Superior, ‘S’, Bechterew, vertical canals, VOR
2. Lateral (‘L’, Deiters), VSR
3. Medial (‘M”, Schwalbe), lateral canals, VOR
4. Descending (‘D’), cerebellar connections

**Vascular supply** = almost everything affects the vestibular nucleus

- Big nucleus
- Vertebral/PICA
- AICA
- Basilar branches

**Nystagmus**

- Involuntary movement of the eye
- Obscures vision
  - “Jerk” nystagmus - usually vestibular
  - Congenital nystagmus
- Horizontal direction usually vestibular
- Vertical or torsional often central

**Diving injuries**
Perilymph Fistula
Both middle and inner ear
- Blowout due to pressure (Scuba), explosion, trauma
- Dizziness and hearing loss
- Fix with a patch

Membranous Labyrinth
MRI of inner ear

Superior Canal Dehiscence
- Opening between top of superior canal and dura
- Tullio’s phenomenon
- Pressure sensitivity
- Valsalva produces nystagnus

Disorders of the canal mechanics
- Cogan’s syndrome
  - Autoimmune inner ear disease
  - High sed rate, interstitial keratitis, deafness
  - Canal is plugged by fibrous tissue
- Canal Fistula
  - Opening in the canal – SCD
- BPPV

BPPV Mechanisms
canalithiasis (animation)
Vestibular Hair Cells

Ototoxicity – aminoglycosides selectively damage vestibular hair cells

Disorders of Vestibular Hair Cells

Gentamicin Toxicity (Inner ear problem)

48 YO Airline stewardess developed an ingrown toe-nail infection. She underwent a course of gentamicin and vancomycin. 12 days after starting therapy she developed imbalance. 21 days after starting, she was “staggering like a drunk person”. Meclizine was prescribed. Gentamicin was stopped on day 29. One year later, the patient had persistent imbalance, visual symptoms, and had not returned to work. Hearing is normal. She presently does volunteer work.

Bedside diagnosis of Gentamicin toxicity

- Romberg – unable ECTR
- DIE test
  - Distance vision with head still
  - Distance vision with head moving (horizontal or vertical, 1-2 Hz)
  - Normal: 0-2 lines change.
  - Abnormal: 4-7 lines drop with movement

Gentamicin toxicity

- Causes permanent imbalance and oscillopsia
- Hair cells do not regenerate
- No medical treatment is available
- Physical therapy is helpful to promote compensation

Oscillopsia
Otolith disorders

- Not much is known
- Tilt disorders?
- Rocking?

Cochlear hair cells

- Arranged in a tonotopic spiral
- High frequencies at base
- Spiral Ganglion wound around cochlea

Endolymph and Perilymph

- 45 year old woman, speech pathologist
- Troubled by spells of dizziness, nausea, vomiting, gradually increasing hearing loss
- Many years of symptoms

Meniere's Disease

- Low frequency sensorineural hearing loss is typical of early Meniere's disease
Meniere's disease

- Eventually people with Meniere's disease go deaf on their "bad" ear.
- Meniere's disease is a chronic disease.

Treatment of Meniere's

- Medical – salt restriction, diuretic, vestibular suppressants
- Surgical – various destructive options
  - Gentamicin injection is currently favored
  - Labyrinthectomy if deaf
  - Vestibular neurectomy if gentamicin fails
- Strange devices and placebos
  - Meniett

Eighth Cranial Nerve

Things that can go wrong with the ganglia or the nerve

- Vestibular neuritis
- Sudden hearing loss (the viral type)
- Acoustic neuroma
- Microvascular compression

Vestibular neuritis

- Sudden severe vertigo
- Loss for several weeks
- Usually vomiting, imbalance
- Attributed to herpes virus (herpes-1)
- Symptomatic treatment (initially)
- Recently shown that steroids are helpful
- May use PT after 2 months to encourage compensation

Case

- ML – 21 year old man
- Gradually increasing hearing loss on right side
- Developed a headache and had a CT scan done in ER
- Huge tumor was seen
Acoustic Neuroma

Treatment of Acoustic Neuroma
- Watchful waiting (about 25%)
- Operative removal (about 50%) – losing ground
- Gamma Knife (about 25%) – gaining ground because effective and noninvasive

Microvascular compression and vestibular neuralgia
- Irritation of vestibular nerve
- Quick spins
- Motion sensitivity
- May follow 8th nerve surgery
- Wastebasket syndrome in some cases?

Diagnosis of MVC
- Normal ENG/Audiogram
- May have nystagmus on hyperventilation
- Response to oxcarbazepine (Trileptal)
- 3D CISS MRI may show blood vessel

Summary
- Vestibular system – angular and linear motion sensors
- Integration with eyes/feet/internal model
- Patterns of vestibular disturbance
  - Unilateral loss – nystagmus
  - Bilateral loss – oscillopsia
  - Position sensitivity – positional vertigo
  - Pressure sensitivity – exercise intolerance
  - Irritability – paroxysms