Otoneurology

Lecture Plan (two lectures)
1. Ear and associated brain function
2. Ear and associated brain anatomy
3. Clinical Disorders

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
Video Frenzel Goggles

Vestibulo-spinal reflex
V.S.R.
- Stabilizes body
- Helps maintain desired orientation to environment

V.S.R

Inertial navigation

6 degrees of freedom problem
- Three axes of rotation
  - Roll, pitch and yaw

- Three axes of translation
  - AP, Lateral, Vertical

Sailors Sea Chanty

Roll, roll roll you son of a bitch!
The more you roll
The less you pitch

(anon)
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

The otoliths sense tilt and linear acceleration

OTOLITHIC MEMBRANE
OTOCONIA - CALCITE CRYSTALS

UTRICLE AND SACULE 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
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
Imperfections in Vestibular Sensors

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

Ewald's 2nd Law

- Gain is a problem built into the vestibular system.
- It affects growth, development, and diseases like 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.


Clinical correlations

- **Grocery Store Syndrome**
  - AKA visual dependence
  - Unable to tolerate busy visual environments
  - Normally people switch between most salient sensory modes – visual/vestibular/somatosensor y
  - 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
Coordinate Rotation is needed between head and body
- Ears are in head which can turn on body
- Must rotate vestibular signals into body coordinates (Nashner, 1974)
- This is probably computationally intensive and slow.
- Is there a clinical correlation? Must be, but we haven’t figured it out yet.

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

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

Kalman Filter diagram
- Formal method of forming “optimal estimate”.
- Integrates efference with afference
- Accounts for noise, sensor differences.

Miniaturation: Everything is in the bony labyrinth (size of dime)
Membranous Labyrinth

MRI of inner ear

The Labyrinth is filled with Endolymph and Perilymph

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 correlate with dysequilibrium in elderly population.

Peripheral circulation to inner ear

- AICA
  - Labyrinthine
  - Vestibulocochlear
    - PC, Saccule
  - Anterior vestibular
    - AC, IC, Uttricle

Cupula to Brain

- Cupula
- Scarpa’s ganglion
- Vestibular Nerve
- Vestibular Nucleus
- Cortex

Vestibular Nerve

- Superior vestibular nerve: AC, IC, Uttricle
- Inferior vestibular nerve: PC, Saccule
- Scarpa’s ganglion

Clinical Correlations

- Vestibular neuritis — 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

**Hearing Disorders: Function**

- Sound
- Middle ear – conductive hearing loss
- Cochlea – sensory hearing loss
- 8th nerve and beyond
  - Sensorineural hearing loss
  - Aphasia
  - Central auditory processing disorders

**Hearing disorders are common**

- 9.75% of population 45-54
- 38.55% of population 75 and older

- Source: NCHS 1982b (1977 data)

**Sound basics**

- Human ear – works over 20-20K Hz
- Loudness quantified in dB, 0-120 dB. Audiograms use HL units – normalized to normal young adults

**Spectrograms**

- We process the spectrum, not just a single frequency at a time.
- Massively parallel processing
Hearing overview

- External ear: focus sound
- Middle ear: impedance matching
- Inner ear: transform sound into array of neural firing
- Spiral Ganglion
- Cochlear nerve
- Brainstem

Sound transmission into inner ear

- External ear, TM
- Middle ear – ossicular chain
- Round window – pressure relief

Clinical Correlations

Conductive hearing loss
Nearly always remediable

Cochlear Slice

Wake up question

What and where is this building?
Organ of Corti

- Reissners membrane – separates scala media from scala vestibuli
- Stria vascularis – lateral wall of scala media, produces endolymph.

Inner and outer hair cells – inner respond to sound, outer amplify/filter

Tonotopy

- Incoming spectrum of sound gets processed by cochlea/spiral ganglion into an array of frequency specific channels. A “spectrum analyzer”
- Tonotopic channels ascend to auditory cortex
- Cortex parses out
  - Speech
  - Frequency information of components!
  - Loudness of components!

Innies and Outies

- Inner hair cells – conventional hearing, one row.
- Outer hair cells – motile.
  - They boost hearing at centrally selected frequencies.
  - Three rows: "W" pattern
  - Most sensitive to ototoxins

Clinical correlation

- Otoacoustic emissions (OAE testing)
- Listen for outer hair cells
- Newborn screening
- Malingering

Hearing neural wiring

- Spiral ganglion (wrapped around the cochlea)
- Cochlear Nucleus
- Trapezoid body (crossover)
- Lateral Lemniscus
- Inferior Colliculus (major integrating center)
- Medial Geniculate (attention?)
- Auditory Cortex (Brodman’s 41 and 42)

Important central auditory structures in Superior Olive

Efferents: olivocochlear body (bundle of Rasmussen). Goes to outer hair cells

- Sound localization based on interaural time differences and intensity.
Hearing localization rules

- Cochlear and above: sensorineural hearing loss
- The further up the lesion, the less likely unilateral loss
- The further up the lesion, the greater resemblance to aphasia rather than deafness.

Nystagmus

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