Otoneurology

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Vestibular Anatomy

Ear Structures of importance

Inner ear is within the temporal bone – hardest in body.

Miniaturization: Everything is in the bony labyrinth (size of dime)

Check an MRI of IAC if you are dubious.

Membranous Labyrinth

The Labyrinth is filled with Endolymph and Perilymph

MRI of inner ear
Clinical Correlations

- Meniere’s disease
  - Meningitis – cochlear aqueduct
    - Labyrinth may later ossify – no fluid on MRI
- Perilymphatic fistula

Vestibular Hair cells – measure force

- Relative movement of hair cells to head causes change in electrical potential

Clinical Correlation – Hair Cells

- Aminoglycosides kill hair cells
  - Some just vestibular hair cells (gentamicin)
- Loop diuretics and NSAIDS are hair cell toxins (cochlear only)

STARTING AND STOPPING = ACCELERATION (F=MA)

Membranous Labyrinth

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

Circulation to inner ear

- AICA
  - Labyrinthine aa
  - Vestibulocochlear aa
    - PC, Saccule
  - Anterior vestibular aa
    - AC, LC, Utricle
Clinical correlates

AICA strokes generally take out everything (hearing and balance) on one side (labyrinthine artery)
PICA strokes rarely affect hearing
Neither type of stroke can affect both ears at the same time
Very improbable to get a branch occlusion

Vestibular Nerve

- Superior vestibular nerve: AC, LC, Utricle
- Inferior vestibular nerve: PC, Saccule
- Scarpa’s ganglion

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

Clinical Correlations

- Vestibular neuronitis – Mainly affects superior division. Spares inferior.
- Acoustic Neurinoma
- Bad design – IAC/brain junction vulnerable to shear trauma.

Vertebral/PICA

AICA

VN is a BIG nucleus
Vestibular Physiology

The ear is an inertial navigation device
- Semicircular Canals are rate sensors.
- Otoliths (utricle and saccule) are linear accelerometers
- Bilateral symmetry means redundant design.

Vestibular Reflexes
- VOR: Vestibulo-ocular reflex
- VSR: Vestibulospinal reflex

Inertial navigation

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.

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

Utricle and Saccule orientation

How the system deals with imperfections in Vestibular Sensors

- Imbalance
- Timing
- Gain
- Noise
Imbalance between ears

- 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 canal – mixed vertical/rotatory
5. Vertical or torsional nystagmus – 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

Velocity Storage for VOR

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


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Ewald’s 2nd Law

Wilson/Melville Jones

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Ewald’s Compensation need for both eyes and neck

Saturation  \rightarrow  Anti-Saturation  \rightarrow  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.


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Imperfections in Vestibular Sensors

- Gain
  - Ewald’s 2nd law – built in problem
  - Growth and development – adapt to bigger head, loss of 50% inner ear by age 80
  - Disease – bilateral vestibular loss

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Noise in Vestibular Sensors

- Noise – a common problem
  - Fluctuations in vestibular function
  - Menière’s, Fistula
- Noise makes vestibular input unreliable
  - Logical consequence is to decrease weighting of vestibular input
Clinical correlations

- Grocery Store Syndrome (AKA visual dependence)
  - Unable to tolerate busy visual environments
  - Normally people switch between most salient sensory modes –
    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

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.

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
- Formal method of forming "optimal estimate."
- Integrates efference with afference
- Accounts for noise, sensor differences.

Wolpert, 1997
Vestibular Disorders

Positional Vertigo
The most common syndrome

- Benign Paroxysmal Positional Vertigo (BPPV) -- bed spins
  - Orthostatic hypotension (dizzy upright)
  - Central positional nystagmus (dizzy everywhere)
  - Low CSF pressure syndrome (dizzy upright)

Benign Paroxysmal Positional Vertigo (BPPV)

61 Y/O man slipped on wet floor.
LOC for 20 minutes.
In ER, unable to sit up because of dizziness
Hallpike Maneuver: Positive

Positional Vertigo Dix-Hallpike Maneuver

Benign Paroxysmal Positional Vertigo (BPPV)

- 20% of all vertigo
- Brief and strong
- Provoked by change of head position
- Definitively diagnosed by Hallpike test
BPPV Mechanism: Utricular debris migrates to posterior canal

BPPV treatment
- Medication (e.g. antivert) – minor benefit
  - May avoid vomiting by pretreating
- Excellent response to PT
- Surgery – canal plugging if rehab fails (need more rehab after plug). Rarely done.


Unilateral Vestibular lesions
- Vestibular Neuritis/Labyrinthitis (common)
- Meniere’s disease (unusual, 1/2000 prevalence)
- Acoustic Neuroma (very rare)
- Vestibular paroxysmia (not sure how common)

Vestibular Neuritis: Case
56 y/o woman began to become dizzy after lunch. Dizziness increased over hours, and consisted of a spinning “merri-go-round” sensation, combined with unsteadiness.
Vomiting ensued 2 hours later, and she was brought by family members to the ER.

Vestibular Spontaneous Nystagmus
Vestibular Spontaneous Nystagmus seen with video Frenzel Goggles

Aside : how to examine for SN
- Frenzel Goggles (best)
- Ophthalmoscope (good –but backwards)
- Gaze-evoked nystagmus (Alexander’s Law)
- Sheet of white paper (neat, Ganzfeld)
Vestibular Neuritis -- rx

- Disturbance of unknown cause (Viral ? Vascular) involving vestibular nerve or ganglion
- Disability typically lasts 2 weeks.
- Symptomatic Rx (meclizine, phenergan)
- Rehab if still symptomatic after 2 months.
- These patients can still get BPPV!

Meniere’s Disease

- Prosper Meniere
  - Hearing loss (fluctuating)
  - Episodic Vertigo
  - Fluctuating (roaring) Tinnitus
  - Aural Fullness
- About 1/2000 people in population
- Chronic condition – lasts lifetime

Etiology of Meniere’s (Dogma)

- Dilation and episodic rupture of inner ear membranes (Endolymphatic Hydrops)
- As endolymph volume and pressure increases, the utricular/saccular and Reissner’s membranes rupture, releasing potassium-rich endolymph into the perilymph causing cochlear/vestibular paralysis

Meniere’s disease – symptoms

- Progressive hearing loss -- sometimes go deaf
- Episodic vertigo – dizzy for several days
- Ataxia – gradually increases over years
- Visual sensitivity →

Visual Sensitivity is common

- Sensory integration disorder – upweight vision, downweight everything else
- Grocery store, Omnimax, Target, etc
- Typical of disorders with intermittent vestibular problems

Otolithic Crises of Tumarkin

- Drop attacks
- Go from upright to on floor in fraction of second
- No LOC
- Very dangerous
- Destructive treatment is best
Treatments of Meniere’s

- Medical management –
  - Usually ineffective
- Bad rehab candidate while fluctuating
- Surgery
  - Low dose gentamicin treatment works nicely
  - High dose gentamicin treatment (overkill)


Acoustic Neuroma

- Rare cause of unilateral loss
- Generally also deaf
- Slowly progressive – little or no vertigo
- 1 mm/year growth

Treatment of Acoustic Neuroma

- Watchful waiting (about 25%)
- Operative removal (about 50%) – losing ground
- Gamma Knife (about 25%) – gaining ground because effective and noninvasive
- Good rehab candidate

Vestibular Paroxysmia (AKA microvascular compression)

- Irritation of vestibular nerve
- Quick spins, tilts, dips
- May follow 8th nerve surgery, Gamma knife treatment, acoustic neuroma, vestibular neuritis, vascular loop
- Wastebasket syndrome in some cases?

Clinical Diagnosis of MVC

- Quick spins
- May have nystagmus on hyperventilation
- Response to anticonvulsant
- No rehab potential
Bilateral Vestibular Loss

A stewardess developed a toe-nail infection. She underwent 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 unsuccessfully sued her doctor for malpractice.

SYMPTOMS OF BILATERAL VESTIBULAR LOSS

- Oscillopsia
- Ataxia
- Oscillopsia

Gentamicin is a problem

DIAGNOSIS IS EASY

- History of recent IV antibiotic medication
- Eyes closed tandem Romberg is positive
- Dynamic illegible ‘E’ test (DIE) failed
- Ophthalmoscope test failed

Dynamic Illegible ‘E’ test (DIE test)

- Distance vision with head still
- Distance vision with head moving
- Normal: 0-2 lines change.
- Abnormal: 4-7 lines change

**Rapid Dolls failed**

- VOR: Vestibulo-ocular reflex

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**Ophthalmoscope Test**

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**LABORATORY DIAGNOSIS**

Everything should be “dead”

- ENG
- Rotatory chair
- VEMP

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**DIAGNOSIS Continued**

- Rotatory chair confirms diagnosis

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**DIAGNOSIS Continued**

- ENG shows little or no response

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**Treatment Bilateral**

- No medical management (other than avoiding more damage)
- Outstanding rehab candidate
- Be prepared for a deposition
Case: WS
Retired plastic surgeon, with impaired hearing related to war injuries, found that when he went to church, when organ was playing, certain notes made him stagger. His otolaryngologist noted that during audiometry (with hearing aid in), certain tones reliably induced dizziness and a mixed vertical/torsional nystagmus. This “Tullio’s phenomenon” could be easily reproduced experimentally. MRI scan was normal.

Diagnosis of SCD
- History of sound and pressure sensitivity
- Valsalva test is easiest bedside test
- Temporal Bone CT scan (0.6 mm high resolution)
- VEMP: Vestibular evoked myogenic potentials, bigger on bad side
Superior Canal Dehiscence

- Etiology:
  - Congenital bone defect (2% ?)
  - Trauma/Age may exacerbate

- Treatment:
  - Surgical
    - Plug
    - Resurface

- Most patients chose to “live with it”

Summary of Otologic Vertigo

More details


More movies

www.dizziness-and-hearing.com