Central Vestibular Disorders
Dizziness

- Dysequilibrium
- **Vertigo** (a sense of motion of person or the visual surround)
- Presyncope (near-faint, light-headedness)
- Psychophysiologic dizziness (anxiety and panic)
- Overlapping
- Motion sickness
Acute vertigo

- A result of vestibular imbalance
  Asymmetry in tonic activity within the vestibular system
- Vegetative symptoms (nausea, vomiting, and diaphoresis)
- Get worse with head movement
- Ataxia

- Peripheral (labyrinth or vestibular nerve)
- Central (Brainstem or Cerebellum)
Acute vertigo

- Rotation reflects imbalance
  - Semicircular canal
  - Tendency to fall to the unaffected side (+ve)

- Upright tilting Imbalance
  - Otolith
  - Tendency to fall to the affected side.
Central vestibular disorders

- Identifying these is critical
  - *Common* 25% older patients presenting to ER with acute isolated vertigo have a cerebellar infarction
  - Life-threatening
  - The earlier the Dx the better the Px
  - Severe neurologic sequelae
  - 1st, 2nd & 3rd Prevention

Challenging

To differentiate central from peripheral

May have both peripheral and central components.

Urgency of the workup

Neurologic, medical, or psychiatric
<table>
<thead>
<tr>
<th></th>
<th><strong>Central</strong></th>
<th><strong>Peripheral</strong></th>
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<tr>
<td><strong>Imbalance</strong></td>
<td>Severe</td>
<td>Mild to moderate</td>
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<td>Frequent</td>
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<td><strong>Nausea</strong></td>
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<tr>
<td><strong>Recovery (central compensation)</strong></td>
<td>Rapid</td>
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Central nystagmus

- Multi-directional
- -ve Alexander's law
- Unaffected by removal of fixation
- Purely vertical or Purely torsional nystagmus
- Constant and does not wane with time
- Absence of a head thrust sign
- Multiple Gaze-evoked nystagmus (<30 degrees)
  Occasionally present in only one direction of gaze similar to peripheral nystagmus.
Gaze-paretic nystagmus RC

Weakness of the extraocular muscles

OR

their innervation

→ Gaze-evoked nystagmus

Gaze-evoked nystagmus is usually due to a defect in the central neural integrators controlling gaze-holding
Central Vestibular Disorders

- Vascular
- Inflammatory
- Neoplastic
- Craniocervical junction disorders
- Inherited ataxias
- Metabolic
- Others
Vascular

- Ischemic stroke/TIA
  - Brainstem
  - Cerebellar
  - Labyrinthine

- Hemorrhage
  - Brainstem
  - Cerebellar

- Migraine
  - Vertigo
  - Dysequilibrium
  - Benign paroxysmal vertigo
  - Paroxysmal torticollis
Inflammatory

- Cerebellitis
- Multiple sclerosis
- Susac syndrome
- Behçet's syndrome
- Systemic lupus erythematosus
- Sarcoidosis
- Infectious

Intracranial complications CSOM
Neoplastic

- Brainstem
- Cerebellar
- Fourth ventricle
- Paraneoplastic
  - Paraneoplastic cerebellar degeneration
  - Opsoclonus/myoclonus
Craniocervical junction disorders

- Chiari malformation
- Basilar impression
- Syringobulbia
Inherited ataxias

- Autosomal recessive
  - Friedreich ataxia
  - Ataxia-telangiectasia
  - Vitamin E deficiency
  - Refsum disease

- Autosomal dominant
  - Spinocerebellar ataxias
  - Episodic ataxias
Metabolic

- Wernicke's encephalopathy
- Diabetes Hypoglycemia
- Vitamin B12 deficiency
- Hypothyroidism
- Hyperventilation
Others

- Toxic
  - Medications
  - Alcohol
- Degenerative
  - Parkinson's disease
  - Progressive supranuclear palsy
  - Multiple systems atrophy
  - Normal pressure hydrocephalus
Others

- Epilepsy
- Trauma
  - Brain contusion
  - Post-concussion syndrome
- Physiologic
  - Mal de debarquement syndrome
  - Motion sickness
- Psychophysiologic
  - Chronic anxiety
  - Panic disorder
  - Phobic postural vertigo
- Psychogenic gait disorder
Others

- Global cerebral hypoperfusion
  - Vasovagal presyncope
  - Reduced cardiac output
  - Autonomic insufficiency
  - Hypovolemic
  - Multisensory disturbance

- Peripheral neuropathy
  - Cervical or thoracic myelopathy
  - Visual loss
  - Superior oblique myokymia
  - Voluntary nystagmus
Dissection of the vertebral artery

- 19 y male
- MVA
- Dizziness neck extension
- Visual disturbance
- Drop attacks
- Ataxia
Vascular

- Ischemic stroke/TIA
  - Brainstem
  - Cerebellar
  - Labyrinthine

- Hemorrhage
  - Brainstem
  - Cerebellar

- Migraine
  - Vertigo
  - Dysequilibrium
  - Benign paroxysmal vertigo
  - Paroxysmal torticollis
Ischemic Stroke

- Risk factors
  - Hypertension
  - Hyperlipidemia
  - Diabetes
  - Smoking.
  - Transient ischemic attacks (TIAs).
Vertebro-basilar arteries

Branches:
- Posterior inferior cerebellar artery (PICA)
- Anterior inferior cerebellar artery (AICA)
- Superior cerebellar artery (SCA)

Brainstem or Cerebellum
Figure 7–11. Principal arteries of the brain stem (ventral view).
Dissection of the vertebral artery

- 19 y male
- MVA
- Dizziness neck extension
- Visual disturbance
- Drop attacks
- Ataxia
Dissection of the vertebral artery

- Dissection → lumen stenosis or occlusion
- Minor neck trauma, neck manipulation, or spontaneously
- Signs of vertebrobasilar ischemia 4Ds
  Dizziness, Diplopia, dysphagia & drop attacks
- Young patient
- No vascular risk factors
- Neck or head pain
- (MRA) → recognized more commonly now
Vertebrobasilar Transient Ischemic Attacks

- common cause of spontaneous
- resolve within 24 hours TIA
- visual
  - Diplopia
  - visual illusions and hallucinations
  - visual field defects
  - Blindness
- Drop attacks
- Incoordination
- Weakness in the extremities
- Rx aspirin, Percutaneous stenting
Case

- 55 Y Female
- Sudden Vertigo
- Rt SSNHL
- Rt side weakness
- Rt Horner’s syndrome
- Hiccup
Right VA dissecting aneurysm
Wallenberg’s syndrome with ipsilateral hemiparesis
Wallenberg syndrome
(Lateral medullary syndrome)
PICA

- From the vertebral artery
- Supplies
  - Dorsolateral medulla
  - Inferior cerebellar peduncle
## Structures within LMS

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<th>Structure</th>
<th>Symptoms / sign</th>
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<tr>
<td>Spinothalamic and quintothalamic tracts</td>
<td>Loss of pain / thermal sensation in <strong>contralateral</strong> limbs and trunk +/- face. <strong>PICA</strong>*</td>
</tr>
<tr>
<td>Sympathetic fibers</td>
<td>Ipsiliteral Horner’s syndrome (constricted pupil, ptosis, decreased sweating).</td>
</tr>
<tr>
<td>Inferior cerebellar peduncle</td>
<td>Ataxia - falling to affected side.</td>
</tr>
<tr>
<td>Nucleus ambiguus</td>
<td>Vomiting.</td>
</tr>
<tr>
<td>Nucleus and tract of cranial nerve V</td>
<td>Impaired sensation of ipsilateral side of face.</td>
</tr>
<tr>
<td>Tractus solitarius</td>
<td>Loss of taste.</td>
</tr>
<tr>
<td>Reticular formation</td>
<td>Hiccup.</td>
</tr>
</tbody>
</table>

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*PICA* = Parasthesia (pain & tem) In Contra-laterally Abdomen
Lt Lateral Pontomedullary Syndrome

- Vertigo
- Lt SSNHL
- Ataxia
- Lt Horner's syndrome
- Lt facial anesthesia
- Died
AICAs

- Basilar artery
- Supply
  - Lateral pontomedullary region
  - Inner ear
  - Middle cerebellar peduncle
  - Anterior inferior cerebellum including the flocculus.
Lateral Pontomedullary Infarction

- Infarction of the lateral pontomedullary region
- Middle cerebellar peduncle
- Anterior inferior cerebellum
- Vestibular-masseter syndrome
- Occlusion of a small branch in the rostral medulla and caudal pons
- Acute vestibular imbalance
- Unilateral paresis of the muscles of mastication.
Lateral Pontomedullary Infarction

- AICA $\rightarrow$ Lab art
  - $\rightarrow$ infarction of the cochlea and labyrinth
  - $\rightarrow$ Vertigo
  - $\rightarrow$ Ipsilateral facial anesthesia
- Contralateral body anesthesia
- Ipsilateral Horner's syndrome
- NO dysphagia or dysphonia.
- Rich anastomotic network $\rightarrow$ spare 7&8 nerve
- Swelling and herniation of the cerebellar tonsils $\rightarrow$ quadriplegia, coma, and death.
- Surgical decompression and ventriculostomy to relieve hydrocephalus may be lifesaving
Cerebellar Infarction

Brainstem or Cerebellum

Mimic a peripheral vestibular disturbance,
Overlap with Brainstem PICA, AICA, and SCA.
Cerebellar Ataxia

- Lack of coordination without paresis
- Alteration in tone
- Loss of postural sense
- Involuntary movements.
Cerebellar Ataxia

- Lesions of the midline vermis → disturbances
  - Stance
  - Gait
  - Ocular movements
- 2 large hemispheres
  - Affect limb movements.
Cerebellar vermis hematoma

- 70 Y male
- HTN, CAD on LMWH
- Headache
- Vertigo
- Ataxia
<table>
<thead>
<tr>
<th>Symptoms and Signs</th>
<th>Lateral Medullary (PICA)</th>
<th>Lateral Pontomedullary (AICA)</th>
<th>Superior Lateral Pontine (SCA)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vertigo, nystagmus</td>
<td>Vestibular nuclei, posterior inferior cerebellum</td>
<td>Labyrinth, vestibular nerve, flocculus, paraflocculus</td>
<td>Superior cerebellum, vermis</td>
</tr>
<tr>
<td>Gait and ipsilateral limb ataxia</td>
<td>Inferior cerebellar peduncle, posterior inferior cerebellum</td>
<td>Middle cerebellar peduncle, anterior inferior cerebellum</td>
<td>Superior cerebellum, vermis, superior cerebellar peduncle</td>
</tr>
<tr>
<td>Tinnitus, hearing loss (ipsilateral)</td>
<td>None</td>
<td>Cochlea, auditory nerve, cochlear nucleus</td>
<td>None</td>
</tr>
<tr>
<td>Facial paralysis (ipsilateral)</td>
<td>Facial nerve (rare)</td>
<td>Facial nerve</td>
<td>None</td>
</tr>
<tr>
<td>Facial pain or numbness (ipsilateral)</td>
<td>Spinal nucleus and tract of trigeminal nerve</td>
<td>Trigeminal nerve or spinal nucleus/tract</td>
<td>Spinal nucleus and tract of trigeminal nerve</td>
</tr>
<tr>
<td>Body hemianesthesia (contralateral)</td>
<td>Spinothalamic tract</td>
<td>Spinothalamic tract</td>
<td>Spinothalamic tract</td>
</tr>
<tr>
<td>Horner's syndrome</td>
<td>Descending sympathetic fibers</td>
<td>Descending sympathetic fibers</td>
<td>Descending sympathetic fibers</td>
</tr>
<tr>
<td>Dysphagia, hoarseness, decreased gag, vocal cord weakness (ipsilateral)</td>
<td>Vagus nerve and nuclei</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>Impaired vibration and position sense (contralateral)</td>
<td>None</td>
<td>None</td>
<td>Medial lemniscus</td>
</tr>
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</table>
Imaging

- CT, MRI, MRA and angiography
- (CT) of the brain without contrast
  - Normal with cerebellar or brainstem infarction
  - The first imaging test
    - widely available
    - Fast
    - Shows intraparenchymal or subarachnoid blood
Investigations

- Blood tests for vascular risk factors
- Echo
- Transesophageal echocardiography (TEE)
Subclavian steal syndrome

Medicine consult

- 45 Y F
- Dizziness
- Exacerbated by exercising the arm.
- Difference in blood pressure may occur between the two arms.
Subclavian steal syndrome

- Exacerbated by exercising the arm.
- A bruit may be heard in the axilla or supraclavicular.
- Difference in blood pressure may occur between the two arms.
- Stenosis of the proximal subclavian artery prior to the origin of the vertebral artery → retrograde flow down the vertebral artery as blood is siphoned into the arm.
37 y F
+ve FHx of Migraine
Vertigo
  Once a week
  Headache
  N&V
Migraine
Migraine Classification

- In 1988 The International Headache Society
- Diagnostic criteria for all headache disorders.
- Gold standard classification
- Revised in 2002
Diagnostic criteria for migraine without aura

A. At least 5 attacks fulfilling B–D.
B. Headache attacks lasting 4 hours to 3 days (untreated).
C. Headache has at least 2 of the following characteristics: (+ plus)
   1. Unilateral location
   2. Pulsating quality
   3. Moderate or severe intensity (Limit daily activities)
   4. Aggravation by stairs or similar routine physical activity
D. During headache at least 1 of the following:
   1. Nausea or vomiting
   2. Photophobia and phonophobia

Ruled out other disorder by appropriate investigations (0)

Hagr 5, 4, 3, 2+, 1 & 0
Arterial Activation

Release of Neurotransmitter

Worsening of Pain
Incidence

- The prevalence of migraine is 16%
- higher in women (25%) than in men (8%)
- 75% Migraine without aura
- Begins in the first three decades of life
- Prevalence peaking in the fifth decade
- commonly undiagnosed
Migraine Triggers

- Stress – emotional or physical
- Insufficient food or long gaps between food
- Certain foods
- Environmental factors:
  - loud noise
  - bright, flickering or flashing lights/glare
  - strong smells
- Changes in routine – On-Call, shift work .....etc.
- Hormonal factors – menstruation, menopause, the pill, HRT
Vestibular migraine

- The nomenclature is inconsistent
- Migrainous vertigo
- Vestibular migraine
- Benign recurrent vertigo
- Migraine-related vestibulopathy
- Migraine-associated dizziness.
- Basilar migraine
Vestibular migraine

- 1861 by Ménière An association between Ménière's disease and migraine
- No universally accepted diagnostic criteria
- Overlap
  - 28% Ménière's have Migraine during attach
  - 25% of migraine
  - 68% vertigo with headache
  - 81% Phonophobia
  - Otolithic crises of Tumarkin
Self-help Measures

- Keep a diary
- Avoid triggers to which you know you are sensitive
- Eat regularly, avoid sugary snacks and include slow release carbohydrate foods in your diet
- Drink plenty of water
- Limit your intake of drinks containing caffeine and alcohol
- Take regular exercise
- Get plenty of fresh air and practise deep breathing
- Ensure that ventilation indoors is good and try to keep rooms at a constant temperature
- Avoid strong perfumes etc
- Avoid bright, flashing or flickering lights (e.g. fluorescent)
- Avoid large reflective surfaces (e.g. plain white walls)
- Wear sunglasses and/or a hat in bright sunlight
- Ensure that computer screens are properly adjusted and fitted with anti-glare filters
- Take regular breaks, especially if you are working at a VDU or if your work is repetitive
- Take care with your posture
- Ensure that your working environment is as ergonomically designed as possible
- Learn relaxation techniques
Flunarizine (SIBELIUM)

- 10 mg < 65 Y
- Drowsiness (at night)
- Extrapyramidal and depressive (5 mg > 65 Y)
- Increased appetite
Behçet's

- 55 Y male
- Mouth and genital ulcers
- Joints pain
- Vertigo 3 hours
- MRI report
  multifocal lesions resemble multiple sclerosis.
Case

- 35 Y M
- Vertigo
- Nystagmus
- Convergence is normal

Internuclear ophthalmoplegia
Inflammatory

- Cerebellitis
- Multiple sclerosis
- Susac syndrome
- Behçet's syndrome
- Systemic lupus erythematosus
- Sarcoidosis
- Infectious

Intracranial complications CSOM
Multiple Sclerosis

- Demyelinating disorder of the CNS
  (oligodendroglia not Schwann cells for peripheral nerve (unaffected))
- Unknown cause ?autoimmunity, infection and heredity
- 15 and 50 years of age (peak onset at age 24)
- women (2:1)
- Relapsing and remitting or progressive.
- Diagnosed
  - Disseminated S/S of CNS dysfunction in time and space
    UMNL, sensory, CN, optic neuritis, internuclear ophthalmoplegia …..
  - MRI demyelinating plaques
Normal Synapse  MS synapse
Vertigo & Multiple Sclerosis

- Initial symptom in about 5%
- 50% during the disease
- Sustained over days or weeks
- Paroxysmal or positional
- Oscillopsia
- Selective involvement of the vestibular nuclei
  - Indistinguishable from a peripheral deficit
  - +/- suppressed by visual fixation
- Oligoclonal bands or a raised IgG (CSF) support Dx
- Slow visual, auditory, or somatosensory Evoked potentials
- Vertigo ? May resemble vestibular neuritis

Demyelinating plaque of Oligodendroglia myelin at the root entry zone
Internuclear ophthalmoplegia

- Medial longitudinal fasciculus (MLF) lesion connects 6 nerve nuclei to the contralateral medial rectus muscle nucleus
- Conjugate gaze $\rightarrow$ loss of contralateral medial rectus adduction
- Abducting nystagmus of the ipsilateral eye
- The horizontal saccadic and pursuit
- Convergence is usually normal
Susac's syndrome
retinocochleocerebral vasculopathy

- 25 y woman
- Neurology ?MS
- Vertigo 3 hours+ HL
- Ophth:
  retinal artery occlusions
- PTA
  low-frequency hearing loss
Case

- 50 Y male
- Vertigo
- Rt SSNHL
- Brun's Nystagmus
  - Lt nystagmus of medium amplitude in the primary position
  - Lt fine horizontal rotatory nystagmus on gaze to the left
  - Slow, large-amplitude right beating nystagmus on gaze to the right
- ABR Rt I-III = 3 ms
- Hyperventilation Nystagmus
- ? Vertigo
Brun's nystagmus

- Combination of
  - Gaze-evoked central nystagmus
    (from compression of the pons)
  - Horizontal-rotatory peripheral nystagmus
    (from damage to the vestibular verve)
- Secondary to CPA lesions
Neoplastic

- Brainstem
- Cerebellar
- Fourth ventricle
- Paraneoplastic
  - Paraneoplastic cerebellar degeneration
  - Opsoclonus/myoclonus
Paraneoplastic syndromes

- ? autoimmune reaction
- Asymptomatic malignancies
- Abruptly and progresses rapidly
Case

- 7 y boy
- Vertigo
- Headache
- Dysphagia & dysartheria
- Normal hearing
Craniocervical junction disorders

- Chiari malformation
- Basilar impression
- Syringobulbia
Chiari malformation

- Group of craniocervical junction abnormalities.
- Chiari I malformation,
  - 5 mm caudal displacement of cerebellar tonsils through foramen magnum
  - 50% Syringomyelia
  - Suboccipital headaches
  - S/S of vertebrobasilar ischemia (4Ds )
    - Lower CN Dysphagia, dysphasia, dysarthria, drop attack
    - Dizziness, or dysequilibrium and gait instability
    - Worsen with neck extension
    - Downbeat nystagmus
    - Vertigo, tinnitus, and hearing loss are uncommon
1896 Hans Chiari

- German pathologist
- Relatively small posterior fossa

Types

- **Chiari type I**
  cerebellar tonsils that are displaced into the upper cervical canal through the foramen magnum.

- **Chiari type II**
  medulla, fourth ventricle, and cerebellum into the cervical spinal canal

- **Chiari type III**
  Displacement +cervical spina bifida

- **Chiari type IV**
  severe cerebellar hypoplasia without displacement of brain through the foramen magnum
Positional nystagmus

- No latent period
- Persist as long as the head position is maintained.
- Pure vertical positional nystagmus
- Down beating with respect to the head
- Large-amplitude nystagmus
- More than one head position
- Other neurologic signs or symptoms.
Case

- 50 y male
- HTN, DM
- Sudden Vertigo
- Rt Hearing loss
- Ataxia- falling to Rt
- Hoarseness. Dysphagia. Dysarthria
- Loss of pain / thermal sensation in Lt
Lateral medullary syndrome
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The End