Session Learning Outcomes

At the end of the session, you should be able to:

- Describe the common presentation of CNS disorders
- Outline the different aetiologies, symptoms and management of hydrocephalus, and raised intracranial pressure.
- Discuss the aetiology, pathophysiology, symptoms and management of stroke.
- Describe the aetiology and clinical presentation of the common vascular disorders of the CNS
Session Plan

- Overview of the central nervous system
- Presenting problems in patients with nervous system diseases:
  - Headache
  - Facial pain
  - Loss of consciousness
  - Seizures and Epilepsy
  - Restless leg syndrome
  - Dementia
  - Personality change
  - Abnormal perception
Session Plan

- Visual disturbance
- Papilloedema
- Bladder, rectal and sexual dysfunction

  - Disorders of the CSF:
    - Raised ICP
    - Hydrocephalus

  - Cerebrovascular Diseases:
    - Stroke and TIA
    - Epidural Haematoma
    - Subdural Haematoma
    - Subarachnoid haemorrhage
    - Intracerebral Haemorrhage
Overview of the central nervous system
Presenting Problems in CNS Diseases
Central Nervous System

- Consists of Brain and Spinal cord
- Main cell types:
  - Neurones: found in the grey matter
  - Astrocytes: specialised support cells
  - Oligodendrocytes: form myelin
  - Microglia: resident cells of the monocyte/macrophage type

Tortora, GJ & Derrickson, B 2014, Principles of anatomy and physiology, 14th edn, John Wiley & Sons, Hoboken, NJ.
<table>
<thead>
<tr>
<th>Presenting Problems in CNS Diseases</th>
</tr>
</thead>
<tbody>
<tr>
<td>o Headache and facial pain</td>
</tr>
<tr>
<td>o Dizziness and blackouts</td>
</tr>
<tr>
<td>o Seizures</td>
</tr>
<tr>
<td>o Epilepsy</td>
</tr>
<tr>
<td>o Restless leg syndrome</td>
</tr>
<tr>
<td>o Sleep disorders</td>
</tr>
<tr>
<td>o Sensory Disturbance</td>
</tr>
<tr>
<td>o Disturbance of memory</td>
</tr>
<tr>
<td>o Change in personality</td>
</tr>
<tr>
<td>o Disorders of Perception</td>
</tr>
<tr>
<td>o Visual disturbance</td>
</tr>
<tr>
<td>o Papilloedema</td>
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<td>o Sphincter disturbance</td>
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# Headache

## Aetiology:

<table>
<thead>
<tr>
<th>Primary headache syndromes</th>
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<tbody>
<tr>
<td>• Migraine (with or without aura)</td>
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<tr>
<td>• Tension-type headache</td>
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<tr>
<td>• Trigeminal autonomic cephalalgia (including cluster headache)</td>
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<tr>
<td>• Primary stabbing/coughing/exertional/sex-related headache</td>
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<tr>
<td>• Thunderclap headache</td>
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<td>• New daily persistent headache syndrome</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>Secondary causes of headache</th>
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</thead>
<tbody>
<tr>
<td>• Medication overuse headache (chronic daily headache)</td>
</tr>
<tr>
<td>• Intracerebral bleeding (subdural haematoma, subarachnoid or intracerebral haemorrhage)</td>
</tr>
<tr>
<td>• Raised intracranial pressure (brain tumour, idiopathic intracranial hypertension)</td>
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<tr>
<td>• Infection (meningitis, encephalitis, brain abscess)</td>
</tr>
<tr>
<td>• Inflammatory disease (temporal arteritis, other vasculitis, arthritis)</td>
</tr>
<tr>
<td>• Referred pain from other structures (orbit, temporomandibular joint, neck)</td>
</tr>
</tbody>
</table>

Headache

- Diagnosis:
  - Based upon headache history:
    - The overall pattern (intermittent or continuous)
    - The tempo of onset
    - The time of day of onset of maximal pain
    - The effect of posture, coughing and straining
    - The location of the pain
    - Any associated symptoms
Facial Pain

- Aetiology:
  - Idiopathic
  - Neurological
    - Trigeminal neuralgia: bouts of brief (seconds), lancinating pain (‘electric shocks’), most frequently felt in the second and third divisions of the nerve and often triggered by talking or chewing.
    - Facial shingles: commonly affects the first (ophthalmic) division of the trigeminal nerve, and pain usually precedes the rash.
    - Post-herpetic neuralgia: may follow, typically a continuous burning pain throughout the affected territory, with marked sensitivity to light touch and resistance to treatment.
Loss of consciousness

○ Definition: It is defined as loss of awareness of the environment and ability to respond to it other than in sleep and suggests a global dysfunction of the brain.

○ Aetiology:
  • Neurological
    – Infection
    – Vestibular neuronitis
    – Benign positional vertigo
    – Meniere's disease
  – Physiological (visual–vestibular mismatch)
    – Demyelination
    – Migraine
    – Epileptic seizure
Seizures and Epilepsy

- Definition: A Seizure is defined as the occurrence of signs and/or symptoms due to abnormal, excessive or synchronous neuronal activity in the brain.
- ‘Epilepsy’ is the tendency to have unprovoked seizures.
- Status epilepticus: Seizures lasting longer than 30 minutes

Classification:
- Generalised seizures
- Focal seizures

Seizures and Epilepsy

26.35 Trigger factors for seizures
- Sleep deprivation
- Missed doses of anti-epileptic drugs in treated patients
- Alcohol (particularly withdrawal)
- Recreational drug misuse
- Physical and mental exhaustion
- Flickering lights, including TV and computer screens (generalised epilepsy syndromes only)
- Intercurrent infections and metabolic disturbances
- Uncommon: loud noises, music, reading, hot baths

Diagnosis:
- EEG
- Brain imaging

Management:
- First aid management of seizures
- Restrictions/Lifestyle modifications
- Anticonvulsant drug therapy

Restless Leg Syndrome

- **Definition:** It is a sleep disorder characterized by an urge to move the limbs with or without sensations, that is worsening in the evening or night, at rest or inactivity and improving with activity or movement.

- **Aetiology:**
  - Genetic factors
  - Iron deficiency
  - Spinal cord and peripheral nerve lesions
  - Pregnancy
  - Uremia
  - Medications

- **Management:**
  - Dopaminergic agents
  - Anti-seizure agents
  - Manipulation of the muscles
  - Iron supplement
  - Good sleep habits
Dementia

Definition: A global impairment of cognitive function, and is typically progressive and non-reversible.

Aetiology:
- Vascular
- Degenerative
- Neoplastic
- Inflammatory
- Traumatic
- Hydrocephalus
- Toxic/ nutritional
- Prion disease

Diagnosis:
- Imaging of head
- Blood tests
- Chest X-ray
- EEG

Management:
- Treatment of underlying condition
- Psychotropic drugs
Personality change

○ Aetiology:
  • Structural damage to Frontal lobe
    – Stroke, trauma, tumour or hydrocephalus

○ Clinical presentation:
  • Mesial frontal lesions: Increasingly withdrawn, unresponsive and mute, urinary incontinence, gait apraxia, and an increase in muscle tone
  • Dorsolateral pre-frontal lesions: Difficulties with speech, motor planning and organisation
  • Orbitofrontal lesions: Disinhibited, irresponsible behaviour
Abnormal Perception

- Damage to sensory areas in cortex (Trauma or Lesions) results in reduction or loss of the ability to perceive the sensation.
  - Apraxia: Inability to perform complex, organised activity
  - Agnosia: Inability to recognise familiar objects

- Abnormal excitation of these areas (Epilepsy, Migraine) results in apparent perception which is not based in physical reality.
  - Illusions: a sensory experience of something different than its reality
  - Hallucinations: a sensory experience of something that does not exist.
Visual disturbance

- Visual loss: can occur as the result of lesions in any areas between the retina and the visual cortex
- Positive visual phenomena: occurs in migraine as silvery zigzag lines or flashing coloured lights preceding headache
- Double vision occurs: from disturbance of the ocular motor nerves.

Visual pathways and visual field defects

Papilloedema

- Definition: swelling of optic disc secondary to raised intracranial pressure.

- Pathophysiology:
  - Raised ICP $\rightarrow$ reduced CSF circulation $\rightarrow$ Swollen nerve fibers $\rightarrow$ capillary and venous congestion $\rightarrow$ Papilloedema
Bladder Dysfunction

- **Atonic:** Damage to the lower motor neuron pathways that produces a flaccid bladder and sphincter with overflow incontinence.

- **Hypertonic:** Damage to upper motor neuron leading to uncontrolled over-activity of the parasympathetic supply resulting in frequency, urgency and urge incontinence.

- **Detrusor–sphincter dyssynergia:** Loss of coordinating control of the pontine micturition centre leading to uncoordinated detrusor contraction and sphincter relaxation resulting in both urgency and an inability to pass urine.
Rectal Dysfunction

- Constipation: Occurs due to damage to the autonomic components; usually a common early symptom in Parkinson’s disease.

- Diarrhoea: usually associated with Diabetic neuropathy

- Faecal incontinence: occurs due to lesions affecting the conus medullaris, the somatic S2–4 roots and the pudendal nerves
Erectile and Ejaculatory Failure

- Erectile failure: Erection is largely parasympathetic, and may be impaired by a number of drugs, including anticholinergic, antihypertensive and antidepressant agents. Neuropathic causes include diabetes mellitus, alcohol excess, and multiple sclerosis.

- Ejaculatory failure: Sympathetic activity is important for ejaculation, and may be inhibited by α-adrenoceptor antagonists (α-blockers).
Disorders of the CSF
Cerebrospinal Fluid (CSF)

- The cerebrospinal fluid (CSF) supports and protects the delicate nervous tissue.

- CSF examination: Lumbar puncture is used to sample CSF

- The fluid is then examined by:
  - Naked eye inspection
  - Microscopy
  - Microbiology
  - Biochemistry
### Cerebrospinal Fluid (CSF)

<table>
<thead>
<tr>
<th>How to interpret CSF results</th>
<th>Normal</th>
<th>Subarachnoid haemorrhage</th>
<th>Acute bacterial meningitis</th>
<th>Viral meningitis</th>
<th>Tuberculous meningitis</th>
<th>Multiple sclerosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pressure</td>
<td>50–250 mm of water</td>
<td>Increased</td>
<td>Normal/ increased</td>
<td>Normal</td>
<td>Normal/increased</td>
<td>Normal</td>
</tr>
<tr>
<td>Colour</td>
<td>Clear</td>
<td>Blood-stained Xanthochromic</td>
<td>Cloudy</td>
<td>Clear</td>
<td>Clear/cloudy</td>
<td>Clear</td>
</tr>
<tr>
<td>Red cell count (x 10⁶/L)</td>
<td>0–4</td>
<td>Raised</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>White cell count (x 10⁶/L)</td>
<td>0–4</td>
<td>Normal/ slightly raised</td>
<td>1000–5000 polymorphs</td>
<td>10–2000 lymphocytes</td>
<td>50–5000 lymphocytes</td>
<td>0–50 lymphocytes</td>
</tr>
<tr>
<td>Glucose</td>
<td>&gt; 50–60% of blood level</td>
<td>Normal</td>
<td>Decreased</td>
<td>Normal</td>
<td>Decreased</td>
<td>Normal</td>
</tr>
<tr>
<td>Protein</td>
<td>&lt; 0.45 g/L</td>
<td>Increased</td>
<td>Increased</td>
<td>Normal/ increased</td>
<td>Increased</td>
<td>Normal/ increased</td>
</tr>
<tr>
<td>Microbiology</td>
<td>Sterile</td>
<td>Sterile</td>
<td>Organisms on Gram stain and/or culture</td>
<td>Sterile/virus detected</td>
<td>Ziehl–Nielson/auramine stain or tuberculosis culture positive</td>
<td>Sterile</td>
</tr>
</tbody>
</table>
Raised Intracranial Pressure

- As the brain is encased in a rigid skull, if the brain swells it results in mounting intracranial pressure.

**Aetiology:**

- Intracranial haemorrhage
- Cerebral tumour
- Cerebral abscess
- Cerebral venous thrombosis
- Hydrocephalus
- Trauma
- Subarachnoid haemorrhage
Raised Intracranial Pressure

- Clinical features:
  - Headache worse on lying/straining
  - Vomiting
  - Diplopia (6th nerve involvement)
  - Papilloedema
  - Bradycardia, raised blood pressure
  - Impaired conscious level

- Management:
  - Relieving the underlying cause
  - Supportive treatment
  - Intensive care support
Hydrocephalus

- Definition: It is the excessive accumulation of CSF within the brain, and may be caused either by increased CSF production, by reduced CSF absorption, or by obstruction of the circulation.

Hydrocephalus

- Aetiology:
  - Congenital causes:
    - Aqueduct stenosis
    - Chiari malformations
    - Benign intracranial cysts
    - Congenital CNS infections
  - Acquired causes:
    - Tumour
    - Colloid cyst of third ventricle
    - Abscess
    - Haematoma
    - Meningitis, sarcoidosis
    - Intracranial haemorrhage
Hydrocephalus

- Clinical features:
  - Congenital:
    - Expansion of head, bulging of the fontanels
    - Signs of increased ICP may be absent
    - Seizures, Weakness and uncoordinated movement
    - Optic nerve atrophy leads to blindness.
  - Acquired:
    - Symptoms of increased ICP in case of acute
    - progressive dementia and gait changes in slowly progressive cases.
Hydrocephalus

- **Diagnosis:**
  - CSF examination
  - Brain imaging: MRI
  - EEG

- **Management:**
  - Congenital: Surgical placement of a shunt
  - Acquired: Surgical decompression and shunting
Cerebrovascular Diseases
Stroke

- Definition: Stroke is the most common clinical manifestation of cerebrovascular disease, and results in episodes of brain dysfunction due to focal ischemia or haemorrhage.

- Aetiology:
  - Cerebral infarction
    - Thromboembolic disease
    - Vasculitis
    - Endocarditis
  - Intracerebral haemorrhage
  - Subarachnoid haemorrhage
Stroke

- Pathophysiology:
  - Thromboembolic disease → obstruction to vascular supply → ischemia → further decrease in blood flow → irreversible cell death → release of inflammatory mediators by microglia and astrocytes → further death of all cell types in the area of maximum ischaemia
  
  - Intracerebral haemorrhage → explosive entry of blood into the brain parenchyma → immediate cessation of function in that area → Infarct may acts like a mass lesion to cause progression of the neurological deficit → if big death occurs.
Stroke

- Clinical features:
  - Facial droop, arm weakness, and slurred speech
  - Unilateral numbness, vision loss in one eye
  - Language disturbance and sudden, unexplained imbalance or ataxia

- Diagnosis:
  - CT/MRI scans
  - EEG
  - Cerebral angiography

- Management:
  - Neurosurgery
  - Vasodilators
  - Corticosteroids
  - Anticoagulants
  - Aspirin
Transient Ischaemic Attack

○ Definition: TIA is a short lived anoxic episode with minimal damage to CNS

○ Clinical features:
  • Symptoms do NOT persist >24 hrs
  • Amaurosis fugax – transient vision loss in one eye
  • Hemiparesis - Partial paralysis
  • Paraesthesia - Sensory disturbances
  • Vertigo, diplopia (double vision), rarely loss of consciousness
Epidural Haematoma

○ Definition: A hematoma that develops between the inner table of the bones of the skull and the dura mater.

○ Aetiology:
  • Fracture of the temporal area of the skull leading to tear in the middle meningeal artery.

○ Clinical features:
  • History of head injury and a brief period of unconsciousness followed by a lucid period in which consciousness is regained, followed by rapid progression to unconsciousness.
  • Death if untreated.
Epidural Haematoma

Subdural Haematoma

- **Definition:** A hematoma that develops in the area between the dura and the arachnoid (subdural space).

- **Aetiology:**
  - Tear in the small bridging veins that connect veins on the surface of the cortex to dural sinuses.

- **Clinical features:**
  - Clinical picture is similar to that of epidural hematoma, except that there usually is no lucid interval.
  - Brain herniation & death occur in most cases.
Subarachnoid Haemorrhage

- **Definition:** It refers to bleeding into the subarachnoid space between the pia and arachnoid membranes.

- **Clinical features:**
  - Sudden, severe, ‘thunderclap’ headache (often occipital), which lasts for hours or even days,
  - Often accompanied by vomiting, raised blood pressure and neck stiffness or pain.
  - Commonly occurs on physical exertion, straining and sexual excitement
  - Loss of consciousness, comatose
Intracerebral Haemorrhage

- **Definition:** Rupture of a blood vessel within the brain parenchyma. Lenticulostriate branch of the middle cerebral artery (“artery of cerebral haemorrhage”) is mainly ruptured.

- **Aetiology:**
  - Arterial disease and hypertensive crises
  - Rupture of intracerebral vessel aneurysms
  - Arteriovenous malformations
  - Neoplasms and leukaemia
  - Haemorrhagic diathesis
  - Trauma
Intracerebral Haemorrhage

- Clinical features:
  - Following physical or emotional exertion, intense headache and vomiting, rapid loss of consciousness
  - Patient’s head is thrown back with the face congested, and heavy, laboured breathing being apparent
  - One month survival is only approximately 17%
Reading and Resources

- Crowley LV, 2012, *An Introduction to Human Diseases – Pathology and Pathophysiology Correlations*, 9th edn, Jones and Bartlett Learning
Reading and Resources

- Mosby’s dictionary of medicine, nursing and health professions 2013, 9th edn, Elsevier, St. Louis, MO.
- VanMeter, KC & Hubert, RJ 2014, *Gould's pathophysiology for the health professions*, 5th edn, Elsevier, St Louis, MO.
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