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The sooner you complete it, the more chances you have to win!

Session Learning Outcomes

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

- Review and revise the basics of motor nervous system.
- Define and describe various types of abnormal gait and involuntary movements and describe their causes.
- Describe various conditions affecting the spine and spinal cord and discuss their clinical features and management.
- Describe the clinical features, investigations and management of peripheral neuropathies.
- Define Facial Nerve Palsy (Bell’s palsy) and discuss its clinical features and management.
- Describe the pathophysiology, clinical features, differential diagnosis, investigations, and management of Myasthenia Gravis and muscular dystrophies.
Session Plan

- Overview of the motor nervous system
- Neuromuscular disorders
  - Gait disorders
  - Involuntary movement
  - Disorders of the spine and spinal cord
    - Compression of spinal cord
    - Cervical Spondylosis
    - Lumbar disc herniation
    - Lumbar canal stenosis
    - Syringomyelia
Session Plan

• Diseases of nerves
  – Entrapment neuropathies
  – Trigeminal neuralgia
  – Bell’s palsy (Facial nerve palsy)
  – Hemifacial spasm

• Diseases of the neuromuscular junction
  – Myasthenia gravis

• Muscular dystrophies
Overview of the Motor Nervous System
Motor Nervous System

- The neuromuscular system consists of
  - Motor unit (motor neuron and the muscle fibers it innervates)
  - The spinal cord
  - Brain stem
  - The cerebellum, and basal ganglia,
  - The motor cortex
Organization of Movement

- The motor control system: It is the final common pathway that transmits all central nervous system commands to the skeletal muscles.
- This path is influenced by sensory input from the muscle spindles and tendon organs and descending signals from the cerebral cortex and brain stem.
- The cerebellum and basal ganglia influence the motor function indirectly, using brain stem and cortical pathways.

Grossman, S, Porth, CM 2013, Porth's pathophysiology, Concepts of Altered Health States, 9th edn, Lippincott Williams & Wilkins
Spinal Reflexes

- Spinal Reflexes: Coordinated, involuntary motor responses that are initiated by a stimulus applied to peripheral receptors
  - The flexor-withdrawal reflex: Initiate movements to avoid hazardous situations
  - *The Stretch and Deep Tendon Reflexes: Serve to integrate motor movements so they function in a coordinated manner*
Somatic Motor Pathways

- Somatic Motor Pathways: Neural circuits in the brain and spinal cord orchestrate all voluntary and involuntary movements.
  - Consist of:
    - Lower motor neurons
    - Local circuit neurons
    - Upper motor neurons
    - Basal nuclei neurons
    - Cerebellar neurons

Somatic motor pathways for coordination and control of movement.
Somatic Motor Pathways

- Organization of Upper Motor Neuron Pathways:
  - Direct motor pathways (pyramidal pathways)
  - Indirect motor pathways (extra-pyramidal pathways)

- Characteristic features of motor neurone lesions
  - Upper motor neuron lesion
  - Lower motor neuron lesion

<table>
<thead>
<tr>
<th>26.19 Distinguishing signs in upper versus lower motor neuron syndromes</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Upper motor neuron lesion</strong></td>
</tr>
<tr>
<td>Inspection</td>
</tr>
<tr>
<td>Tone</td>
</tr>
<tr>
<td>Pattern of weakness</td>
</tr>
<tr>
<td>Deep tendon reflexes</td>
</tr>
<tr>
<td>Plantar response</td>
</tr>
</tbody>
</table>

Assessment of Motor Function

- Assessing the motor system should include assessment of:
  - Body position
  - Involuntary movements
  - Muscle characteristics (strength, bulk, and tone)
  - Spinal reflexes
  - Coordination
Gait Disorders &
Involuntary Movement
Gait Disorders

- **Pyramidal gait:** Arm immobile against body, flexion of the shoulder, elbow, wrist and fingers, adduction of shoulder. Leg stiff and extended. Circumducts hip with each step (drags toes in a semicircle)
  - **Cause:** UMNL of the corticospinal tract e.g. CVA, cerebral trauma

- **Extra-pyramidal:** Posture stooped; trunk pitched forward; elbows, hips and knees flexed. Steps are short and shuffling. Hesitation to begin walking, difficult to stop suddenly. Person holds their body rigid, walking and turning as one fixed unit. Difficulty with any change in direction.
  - **Cause:** Parkinson’s disease, certain drugs
Gait Disorders

- Cerebellar ataxia: Staggering, wide based gait; difficulty with turns, uncoordinated movement. Positive Romberg sign.
  - Cause: Alcohol or barbiturate effect on cerebellum, cerebellar tumor, MS

- Steppage or Footdrop: Slapping quality – looks as if walking up stairs and finds no stair there. Lifts knee and foot high and slaps it down hard and flat to compensate for footdrop
  - Causes: Weakness of peroneal and anterior tibial muscles; due to LMN lesion at spinal cord e.g. poliomyelitis, herniated IV disc
Gait Disorders

- **Waddling**: Weak hip muscles – when the person takes a step, the opposite hip drops, which allows compensatory, lateral movement of the pelvis. The person often has marked lumbar lordosis and a protruding abdomen.
  - **Cause**: Hip girdle muscle weakness due to muscular dystrophy, dislocation of the hips

- **Scissor**: Rigidity and flexion of the knee, hyper-adduction at the hip joint that causes the patient to walk with the knees close together. Contractures in the adductor muscles pull the thigh region medially giving the scissor-like appearance.
  - **Cause**: Spastic cerebral palsy
# Involuntary Movement

<table>
<thead>
<tr>
<th>Description</th>
<th>Features</th>
<th>Causes</th>
</tr>
</thead>
</table>
| Tremor      | Rhythmical oscillation of body part | Essential tremor  
Parkinson's disease  
Drug-induced |
| Chorea      | Jerky, brief, involuntary movements | Huntington's disease  
Drug-induced |
| Athetosis   | Slower, writhing movement of the limbs,  
often combined with chorea | Same as chorea |
| Ballism     | A more dramatic form of chorea, causing  
often-violent flinging movements of one limb  
or one side of the body. | Stroke |
| Tics        | Stereotyped, repetitive movements,  
briefly suppressible | Tourette's syndrome |
| Myoclonus   | Shock-like muscle jerks | Epilepsy |
| Dystonia    | Sustained muscle contraction causing  
abnormal postures +/- tremors | Genetic  
Generalised dystonic syndromes |
Disorders of the Spine and Spinal Cord
Spinal Cord Compression

- **Definition:** It is caused by a space occupying lesion within the spinal canal that may damage nerve tissue either directly by pressure or indirectly by interfering with blood supply causing oedema or ischemia.

- **Aetiology:**
  - **Vertebral (80%)**
    - Tumours, trauma, disc prolapse, TB
  - **Meninges (intradural extra medullary) (15%)**
    - Tumours around the meninges, epidural abscess
  - **Spinal cord (intradural, intramedullary) (5%)**
    - Tumours within the spinal cord
Spinal Cord Compression

- **Clinical features:**
  - Gradual onset, can be acute in case of trauma

### Symptoms of Spinal Cord Compression

<table>
<thead>
<tr>
<th>Pain</th>
</tr>
</thead>
<tbody>
<tr>
<td>Localised over the spine or in a root distribution, which may be aggravated by coughing, sneezing or straining</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Sensory</th>
</tr>
</thead>
<tbody>
<tr>
<td>Paraesthesia, numbness or cold sensations, especially in the lower limbs, which spread proximally, often to a level on the trunk</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Motor</th>
</tr>
</thead>
<tbody>
<tr>
<td>Weakness, heaviness or stiffness of the limbs, most commonly the legs</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Sphincters</th>
</tr>
</thead>
<tbody>
<tr>
<td>Urgency or hesitancy of micturition, leading eventually to urinary retention</td>
</tr>
</tbody>
</table>

### Signs of Spinal Cord Compression

**Cervical, above C5**
- Upper motor neuron signs and sensory loss in all four limbs
- Diaphragm weakness (phrenic nerve)

**Cervical, C5–T1**
- Lower motor neuron signs and segmental sensory loss in the arms; upper motor neuron signs in the legs
- Respiratory (intercostal) muscle weakness

**Thoracic cord**
- Spastic paraplegia with a sensory level on the trunk
- Weakness of legs, sacral loss of sensation and extensor plantar responses

**Cauda equina**
- Spinal cord ends approximately at the T12/L1 spinal level and spinal lesions below this level can only cause lower motor neuron signs by affecting the cauda equina

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Spinal Cord Compression

- **Diagnosis:**
  - MRI of spine
  - Myelography
  - Plain X-rays of spine
  - Chest X-ray
  - CSF analysis
  - Serum B12

- **Management:**
  - depend on nature of underlying lesion.
    - Surgical treatments
    - Radiotherapy
    - Antituberculous chemotherapy
Cervical Spondylosis

- **Definition:** It is osteoarthritis in the cervical spine characterised by degeneration of the intervertebral discs and osteophyte formation.

- **Classification:**
  - **Cervical spondylotic radiculopathy:** Compression of a nerve root when disc prolapses laterally.
    - C4, C5, C6 and C7 are mostly affected
  - **Cervical spondylotic myelopathy:** Compression and pressure on anterior spinal artery and spinal cord due to dorsomedial herniation of the disc
## Cervical Spondylosis

<table>
<thead>
<tr>
<th>Cervical spondylotic radiculopathy</th>
<th>Cervical spondylotic myelopathy</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Pain in neck, radiating in the distribution of affected nerve root</td>
<td>• Insidious onset</td>
</tr>
<tr>
<td>• Movements exacerbate pain</td>
<td>• Painless</td>
</tr>
<tr>
<td>• Paraesthesia and sensory loss in affected segment</td>
<td>• Acute deterioration after trauma</td>
</tr>
<tr>
<td>• LMNL signs in limbs</td>
<td>• UMNL signs in limbs</td>
</tr>
<tr>
<td>− Weakness, wasting and impaired reflexes</td>
<td>− Spasticity of legs and arms</td>
</tr>
<tr>
<td></td>
<td>− Sensory loss in upper limbs</td>
</tr>
</tbody>
</table>
Cervical Spondylosis

- **Diagnosis:**
  - Plain X-rays of spine
  - MRI of spine
  - Myelography

- **Management:**
  - Analgesics
  - Physiotherapy
  - Surgical procedures

MRI showing cervical cord compression (arrow) in cervical spondylosis.

Lumbar Disc Herniation

- **Definition:** The nucleus pulposus may bulge or rupture through the annulus fibrosus, giving rise to pressure on nerve endings in the spinal ligaments, changes in the vertebral joints or pressure on nerve roots.

<table>
<thead>
<tr>
<th>Disc level</th>
<th>Root</th>
<th>Sensory loss</th>
<th>Weakness</th>
<th>Reflex loss</th>
</tr>
</thead>
<tbody>
<tr>
<td>L3/L4</td>
<td>L4</td>
<td>Inner calf</td>
<td>Inversion of foot</td>
<td>Knee</td>
</tr>
<tr>
<td>L4/L5</td>
<td>L5</td>
<td>Outer calf and dorsum of foot</td>
<td>Dorsiflexion of hallux/toes</td>
<td></td>
</tr>
<tr>
<td>L5/S1</td>
<td>S1</td>
<td>Sole and lateral foot</td>
<td>Plantar flexion</td>
<td>Ankle</td>
</tr>
</tbody>
</table>
Lumbar Disc Herniation

- Clinical features:
  - Sudden or gradual onset,
  - Sometimes repeated episodes of low back pain may precede sciatica
  - Constant aching pain in lumbar region, may radiate to the buttock, thigh, calf and foot
  - Aggravated on coughing or straining, better by lying flat
  - Loss of lumbar lordosis and spasm of paraspinal muscles
  - Sensory loss in surrounding dermatome
Lumbar Disc Herniation

- **Diagnosis:**
  - X-ray
  - CT scan
  - MRI

- **Management:**
  - Analgesics
  - Early mobilisation
  - Back strengthening exercises
  - Avoid physical movements likely to strain the lumbar spine

Lumbar Canal Stenosis

- Definition: Congenital narrowing of the lumbar spinal canal, exacerbated by the degenerative changes that commonly occur with age and develop exercise induced weakness and paraesthesia in the legs in elderly.

Syringomyelia

- Definition: Syringomyelia is the formation of a fluid-filled cyst within the spinal cord. The most common site is the cervical spine in the neck region. As the cyst grows, it presses on the spinal cord and interferes with the transmission of nerve impulses.
Diseases of Nerves
Entrapment Neuropathies

- Definition: Focal compression or entrapment of a nerve leading to myelin sheath damage and slow conduction of nerve impulses over the relevant site.

Common entrapment neuropathies:

<table>
<thead>
<tr>
<th>Nerve</th>
<th>Signs and symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median nerve</td>
<td><strong>Carpal tunnel syndrome</strong>&lt;br&gt;Pain and paraesthesia on palmar aspects of hands and fingers, waking the patient from sleep.</td>
</tr>
<tr>
<td>Ulnar nerve</td>
<td><strong>Paraesthesia on medial border of hand, wasting and weakness of hand muscles</strong></td>
</tr>
<tr>
<td>Common peroneal nerve</td>
<td><strong>Foot drop, trauma to head of fibula</strong></td>
</tr>
<tr>
<td>Lateral cutaneous nerve</td>
<td><strong>Tingling and dysaesthesia on lateral border of the thigh</strong></td>
</tr>
</tbody>
</table>
Carpal Tunnel Syndrome

- Definition: is a relatively common compression-type mononeuropathy caused by compression of the median nerve as it travels with the flexor tendons through a canal made by the carpal bones and transverse carpal ligament.

- Clinical features:
  - Pain, paresthesia, and numbness of the thumb and first two and one half digits of the hand;
  - Pain in the wrist and hand, which worsens at night; atrophy of the abductor pollicis muscle;
  - Weakness in precision grip.
  - Clumsiness of fine motor activity
Carpal Tunnel Syndrome

- **Diagnosis:**
  - Tinel sign
  - Phalen test
  - Electromyography
  - Nerve conduction studies

- **Management:**
  - Decrease the causative repetitive movements
  - Splints
  - Corticosteroids injections into the carpal tunnel
  - Surgery
Trigeminal Neuralgia

- **Definition:** This is characterised by unilateral lancinating facial pain, most commonly involving the second and/or third divisions of the trigeminal nerve territory, usually in patients over the age of 50 years.

- **Clinical features:**
  - Repetitive, severe and very brief pain
  - Triggered by touch, a cold wind or eating
  - Spasms may make the patient wince and sit silently
  - May remit and relapse over many years

- **Management:**
  - Carbamazepine
  - Surgery for decompression
  - Localised injection of alcohol or phenol
Facial Nerve Palsy (Bell’s Palsy)

- **Definition:** It is a partial or complete paralysis of facial nerve and is mostly due to infection of herpes simplex virus.

- **Clinical features:**
  - Develop subacutely over a few hours
  - Pain around the ear preceding the unilateral facial weakness
  - Hyperacusis and diminished salivation and tear secretion
  - Vesicles in the ear or on the palate

- **Management:**
  - Steroids
  - Taping the eye shut overnight
  - Plastic surgery
Hemifacial Spasm

- Definition: It is unilateral spasms of facial muscles most likely due an aberrant arterial loop irritating the facial nerve just outside the pons.

- Clinical features:
  - Usually presents after middle age
  - Intermittent twitching around one eye, spreading ipsilaterally to other facial muscles.
  - Spasms are exacerbated by talking, eating and stress

- Management:
  - Injections of botulinum
  - Microvascular decompression
Diseases of the Neuromuscular Junction
Myasthenia Gravis

- **Definition:** It is an autoimmune disorder of transmission at the neuromuscular junction that affects communication between the motor neuron and the innervated muscle.

- **Aetiology:**
  - Autoimmune attack by antibodies to acetylcholine receptors in the post-junctional membrane of the neuromuscular junction or autoantibodies to muscle-specific kinase (MuSK)
Myasthenia Gravis

○ Pathophysiology:
  → Sensitized helper T cells and an antibody-directed attack on the acetylcholine receptor in the neuromuscular junction
  → Shedding of the acetylcholine receptor–rich terminal portions of the folds in the end plate of the muscle fibre
  → A decreased number of receptors, and a widened synaptic space that impairs signal transmission
Myasthenia Gravis

Lambert-Eaton syndrome
Antibodies to pre-synaptic calcium channels

Motor neuron

Acetylcholinesterase removes acetylcholine from neuromuscular junction

Acetylcholine packets released by calcium influx

Voltage-gated calcium channel

Ca++

Acetylcholine

Acetylcholine receptor

Myasthenia gravis
Antibodies to acetylcholine receptors

In myasthenia, end plate is subject to cell-mediated immune assault (end plate simplified)

Depolarisation of muscle membrane

Sodium channels in clefts amplify potential change
Myasthenia Gravis

- Clinical features:
  - Fatigable muscle weakness
  - Movement rapidly weakens as muscle use continues.
  - Worsening of symptoms towards the end of the day or following exercise.
  - Weakness of the oculomotor muscles:
    - Intermittent ptosis or diplopia,
  - Weakness of chewing, swallowing, speaking or limb movement
  - Shoulder girdle muscles most commonly affected:
    - the patient is unable to undertake tasks above shoulder level, such as combing the hair, without frequent rests
  - Respiratory muscles may be involved and respiratory failure is an avoidable cause of death.
    - Aspiration may occur if the cough is ineffectual.
Myasthenia Gravis

- Differential diagnosis
  - Lambert-Eaton myasthenic syndrome (LEMS)

- Diagnosis:
  - Tensilon® test
  - EMG with repetitive stimulation
  - Acetylcholine receptor and Anti-MuSK antibodies in blood
  - CT scan

- Management:
  - Anticholinesterase drugs
  - Thymectomy
  - Plasma exchange
  - Intravenous Igs
  - Corticosteroids
Muscular Dystrophies

- **Definition:** inherited disorders with progressive muscle destruction, and may be associated with cardiac and/or respiratory involvement and sometimes non-myopathic features.

- **Clinical features:**
  - Onset often in the childhood
  - Myotonic dystrophy may present as adults
  - Wasting and weakness of the muscles, usually symmetrical
  - Weakness is usually proximal, except in myotonic dystrophy, when it is distal
Muscular Dystrophies

- **Diagnosis:**
  - Molecular genetic testing
  - EMG
  - Muscle biopsy
  - Creatine kinase levels
  - Screening for an associated cardiac abnormality

- **Management:**
  - No specific therapy
  - Physiotherapy and occupational therapy
  - Steroids
  - Treatment of associated cardiac and respiratory complications
  - Genetic counselling
Reading and Resources

- Crowley LV, 2012, An Introduction to Human Diseases – Pathology and Pathophysiology Correlations, 9th edn, Jones and Bartlett Learning
- Kumar, P & Clark, M 2012, Kumar and Clark’s clinical medicine, 8th edn, Saunders Elsevier, Edinburgh.
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.