Dermatomes

Investigations
Imaging
Skull and spinal x-rays
Brain CT
MRI
Doppler (carotids)
Digital cerebral and spinal angiography (little used, MRI and CT better)
PET (neoplasms outside CNS)
Dopamine transporter imaging (DAT)
fMRI
Isotope bone scanning (metastases)

Other
EEG (epilepsy, hepatic coma, hypoglycaemia, brain death)
Electromyography EMG (myopathic, myotonic and myasthenic changes)
Peripheral nerve conduction (neuropathies and nerve entrapments)
Cerebral evoked potentials (e.g. visual to confirm previous retrobulbar neuritis)
Biopsies inc. brain, meninges, muscle, peripheral nerve
Psychometric testing

Lumbar puncture
See ‘infectious disease’ notes

Special tests

<table>
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<th>Test</th>
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<tr>
<td>Stroke</td>
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<td>Anti-cardiolipin and lupus anticoagulant antibodies and</td>
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LMN
= From anterior horn cell or cranial nerve nucleus to muscle motor end-plate

Features that occur rapidly in voluntary muscles
• Weakness
• Wasting
• Hypotonia
• Reflex loss
• Fasciculation
• Fibrillation potentials (EMG)
• Muscle contractures
• Trophic changes in skin and nails

Possible causes
• Cranial nerve nuclei e.g. Bell’s palsy or anterior horn cell e.g. motor neuron disease
• Spinal root (radiculopathy) e.g. cervical and lumbar disc protrusion, neuralgic amyotrophy
• Peripheral or cranial nerve e.g. trauma, entrapment, polynuropathy

NMJ
Causes
• Myasthenia gravis
• Lambert-Eaton myasthenic-myoopathic syndrome (paraneoplastic manifestation of small-cell bronchial carcinoma)

Recognition
• Fatiguability

Muscle
Causes
• Inflammatory e.g. polymyositis, sarcoidosis
• Metabolic and endocrine e.g. Cushing’s, thyrotoxicosis, hypothyroidism
• Disorders calcium and vitamin D metabolism
• Hypokalaemia
• Alcohol (acute or chronic)
• Drug additions e.g. diamorphine, amphetamine
• Drugs e.g. steroids, lithium
• Muscular dystrophy

Identification
• Muscle pain may be present
• Serum creatine kinase
• EMG
• Muscle biopsy
• MRI of muscle
EEG: useful to categorise epilepsy and understand cause rather than confirm diagnosis
  • E.g. absence seizure attack: 3 Hz spike-and-wave activity

**Pharmacology**

**Indication for treatment**
  • Firm clinical diagnosis of epilepsy AND substantial risk of recurrent seizures

**General principles**
  • Titrate dose upwards
  • Aim for monotherapy
  • If first AED does not work, gradually add second agent then gradually withdraw first agent
  • Non-generic prescribing
  • Monitoring of AED levels only to assess compliance or toxicity
  • Consider drug interactions e.g. lamotrigine and sodium valproate

**General AED adverse effects**
  • General: headache, GI, rash
  • Toxicity: unsteadiness, ataxia, nystagmus, drowsiness
  • More common with multiple AEDs

**Carbamazepine**
  • General
    o Test FBC, U+E, LFT before starting
  • Indications
    o 1st line for *focal seizures* and *generalised tonic-clonic seizures*
  • Adverse reactions
    o May worsen myoclonic seizures
    o Rash (Stevens-Johnson, esp East and South Asians)
    o Blood dyscrasias: aplastic anaemia, agranulocytosis, neutropenia
    o Induction of hepatic enzymes (COCP, warfarin)
    o SIADH, hyponatraemia
    o (Teratogenic)

**Sodium valproate**
  • General
    o Test FBC, U+E, LFT before starting
  • Indications
    o 1st line for *generalised tonic-clonic seizures*, focal seizures and myoclonic seizures
  • Adverse reactions
    o Worry about *liver, haematopoiesis and pancreas*
    o Appetite increase (leading to weight gain)
    o Liver failure
    o Pancreatitis
    o Reversible hair loss
    o Oedema
Causes of diplopia other than cranial nerve lesions

• Local orbital lesions
• Wernicke’s encephalopathy
• NMJ lesions e.g. myasthenia gravis
• Muscle weakness e.g. muscular dystrophy

Facial nerve

Functions

• Muscles of facial expression
• Sensory taste from anterior 2/3 of tongue (chorda tympani)
• Motor fibres to stapedius muscle

Features of UMN lesion e.g. hemispheric stroke with hemiparesis

• Weakness of lower part of face on opposite side
• Frontalis spared giving normal brow furrowing
• Eye closure and blinking largely unaffected
• Earliest sign is slowing of one side of face e.g. baring teeth
• Sometimes relative preservation of spontaneous emotional movement e.g. smiling compared with voluntary movement

Features of complete unilateral LMN lesion

• Ipsilateral weakness of all facial expression muscles
• Angle of mouth falls causing unilateral dribbling
• Frontalis affected so weak frowning
• Weak eye closure giving risk of corneal exposure and ulceration
• Taste frequently impaired
• Hyperacusis

Causes of UMN lesions

• Demyelination
• Brainstem infarct
• Tumour

Causes of LMN lesions

• Cerebellopontine angle tumours e.g. acoustic neuroma, meningioma
  ○ Usually affects V first then VIII then VII (may also affect VI)
• Bell’s palsy
• Trauma
• Middle ear infection
• Ramsay Hunt syndrome (herpes zoster of geniculate ganglion)
• Skull base e.g. Paget’s disease
• Parotid gland e.g. parotid tumours, sarcoidosis (NB. will not affect taste or hearing)

Differential diagnosis of bilateral LMN lesions (often less easier to recognise)

• Infections e.g. Lyme disease (Bannwarth’s syndrome), HIV seroconversion
• Sarcoidosis
Diagnosis can be made EITHER if a second clinical event occurs indicative of a new lesion in a different anatomical location OR if a repeat brain MRI 1+ months later shows a new lesion or a gadolinium enhancing lesion even in the absence of new symptoms.

**Parkinson’s Disease**

**Disorders of the extrapyramidal system**

Disorders classified broadly as either akinetic-rigid (hypokinesias) or dyskinesias (hyperkinesias) though these may coexist.

These are caused by specific changes in neurotransmitters c.f. focal lesions.

**Definitions**

- Tremor = rhythmic sinusoidal oscillation of a body part
- Chorea = excessive, irregular movements flitting from one body part to another (dance-like)
- Myoclonus = brief electric shock-like jerks
- Tics = stereotyped movements or vocalisations
- Dystonia = sustained muscle spasms causing twisting movements and abnormal postures
- Hemiballismus = wild, flinging limb movements, often due to infarct in the contralateral subthalamic nucleus

**Causes**

Usually idiopathic but some rare familial forms exist.

Degeneration in substantia nigra causing loss of dopamine activity and formation of Lewy bodies (tangles of alpha-synuclein and ubiquitin).

**Clinical features**

Pre-motor, non-motor symptoms (in the approx 7 years before motor symptoms)

- Anosmia (90%, olfactory bulb affected early)
  - Depression or anxiety (50%)
  - Aches and pains
  - REM sleep behaviour disorder
  - Autonomic features e.g. urinary urgency, hypotension
  - Constipation
  - Restless legs syndrome

Classical triad of motor features (indious)

- Bradykinesia
  - Progressive fatiguing and decrement in amplitude of repetitive movements
  - Also difficulty initiating movement
  - When idiopathic, almost always initially more prominent on one side and upper limb usually affected first
  - E.g. micrographia, impassive face, reduced frequency of spontaneous blinking

- Tremor
  - Presenting symptom in 70%
Combined with dopa decarboxylase inhibitor e.g. benserazide (co-beneldopa) or carbidopa (co-careldopa) to reduce peripheral adverse effects

- May later add a COMT inhibitor e.g. entacapone to prolong duration of action

- **Indications**
  - Most effective treatment, all patients will eventually need it

- **Adverse effects**
  - Peripheral adverse effects e.g. nausea, hypotension
  - Wearing off
  - May induce unwanted dyskinesias

**Dopamine agonists**

- **Principles**
  - Non-ergot derived e.g. pramipexole, ropinirole, rotigotine via transdermal patch
  - (Ergot derived)
  - Domperidone used as an antiemetic when initiating (not other antiemetics as they block central dopamine receptors)

- **Indications**
  - In combination with levodopa
  - As initial monotherapy in younger patients (<65y) with mild to moderate impairment

- **Advantages**
  - Fewer motor complications than levodopa

- **Adverse effects**
  - Less effective and less well tolerated than levodopa
  - Ergot derived drugs associated with fibrotic reactions e.g. cardiac valvular fibrosis

**Anticholinergics e.g. trihexyphenidyl**

- **Indications**
  - Rarely used except in younger patients

- **Advantages**
  - May help tremor

- **Adverse effects**
  - Confusion in older patients

**Other**

- Selegilline or rasagilline (MAO-B inhibitor)
- Amantadine (mainly for dyskinesias in advanced disease)
- Apomorphine (potent, short-acting dopamine agonist injected s/c intermittently or by continuous infusion, advanced PD)
- Deep brain stimulation (motor side effects of treatment)
- L-dopa intestinal gel infusion (if the above two options are not possible)

**Prognosis**

Most respond well to treatment with several years of good control
• Fatigable = myasthenia

_Dysphonia_ = impairment in ability to produce voice sounds using the vocal organs, a phonation disorder

**Coma**

**Definitions**
Coma = state of unrousable unresponsiveness

**Consciousness**
Dependent on two separate things…

- Ascending reticular activating system (projects from brainstem to thalamus) determines arousal (level of consciousness)
- Cerebral cortex determines content of consciousness

Coma may be caused by impaired function of either or both systems

**Mechanisms and causes**

Diffuse brain dysfunction

- **Metabolic** (#1) e.g. hypoglycaemia, hyperglycaemia, metabolic acidosis
- **Drug overdose** (#2) either deliberate or accidental inc. alcohol
- **Infection** e.g. encephalitis, meningitis
- **Traumatic** brain injury
- Electrolytes e.g. hyponatraemia, hypernatraemia
- Organ failure e.g. severe uraemia, hepatic encephalopathy
- Hypoxic-ischaemic brain injury e.g. post-cardiac arrest
- CO₂ retention of type 2 respiratory failure, CO poisoning
- Hypothermia, hyperpyrexia
- Seizures inc. non-convulsive status epilepticus or post-ictal
- Sub-arachnoid haemorrhage
- Endocrine e.g. myxoedema coma

Direct effect within brainstem

- **Infarction** (stroke)
- Haemorrhage
- Demyelination
- Neoplasm e.g. glioma
- Wernicke-Korsakoff syndrome

Pressure effect on brainstem

- **SOL** (#3) e.g. tumour, haematoma, abscess
- Massive hemisphere infarction with oedema
- Cerebellar mass

**Coma ‘look-alikes’**

Brainstem death

Vegetative = intact brainstem function but no awareness or response to environmental stimuli except reflexes due to extensive cortical damage

Minimally conscious state

‘Locked-in’ syndrome = complete paralysis except vertical eye movements and blinking due to ventral pontine infarction
• Lateralising signs e.g. asymmetries of reflexes (suggest specific lesion but hypoglycaemia can also cause focal signs)

**Glasgow coma scale**
Possible scores: 3-15
Coma: 8 or lower

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<tr>
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<td>3</td>
<td>To speech</td>
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<td></td>
<td>2</td>
<td>To pain</td>
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<td>1</td>
<td>Absent</td>
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<tr>
<td>Verbal response</td>
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<td>Orientated</td>
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<td></td>
<td>4</td>
<td>Confused conversation</td>
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<tr>
<td></td>
<td>3</td>
<td>Inappropriate words</td>
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<td>Incomprehensible sounds</td>
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<tr>
<td>Motor response</td>
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<td>Obey</td>
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<td>Extension</td>
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**Possible investigations**
Other brainstem reflexes e.g. corneal or gag reflexes

**Blood**
- Drugs: alcohol, salicylates
- Biochemistry inc. U+Es, LFTs, glucose, Ca
- Metabolic and endocrine inc. TSH, cortisol
- ABG

**Urine**
- Toxicology: benzos, narcotics, amphetamines

**Brain imaging**
- CT
- MRI only if CT normal (harder to monitor patient)

**CSF** only post-CT and if it will affect management

**EEG** (metabolic coma, encephalitis, non-convulsive status)

**Delirium**
**Definition**
Acute confusional state in which reduced attention is a cardinal feature, usually also with altered behaviour, cognition, orientation and a fluctuating level of consciousness

**Key features**