

PACES neurology

1. *Charcot Marie Tooth*
2. *Dystrophica myotonica*
3. *Peripheral neuropathy*
4. *Parkinsonism*
5. *Muscular dystrophy*
6. *Hemiplegia*
7. *Multiple sclerosis*
8. *INO*
9. *III*
10. *IV*
11. *VI*
12. *Horner's*
13. *Holmes Adie*
14. *Argyll Robertson*
15. *Spastic paraparesis*
16. *Flaccid paraparesis*
17. *Motor neurone disease*
18. *Myasthenia gravis*
19. *Cervical myelopathy*
20. *Pseudobulbar palsy*
21. *Syringomyelia*
22. *Proximal myopathy*
23. *Friedrich's ataxia*
24. *Cerebellar syndrome – unilateral or bilateral*
25. *T1 lesion*
26. *Lateral medullary syndrome*
27. *Ulnar palsy*
28. *Radial palsy*
29. *Bulbar palsy*
30. *Common peroneal nerve lesion*
31. *CPA lesion*
32. *Polio – old*

Charcot Marie Tooth Disease

Examination

- Distal wasting.
- Mixed motor and sensory polyneuropathy, mainly peripheral.
- Pes cavus
- High stepping gait.

What types are there?

5 types (axonal (CMT 2) vs demyelinating(CMT 1, 3, 4 X)) and many genotypes (39)
– hard to distinguish clinically.

Inheritance?

- AD.
- Commonest genetic neurological disease affecting 1/2500.
- Familial.

Natural History?

- Presents in teens or twenties.
- Can affect central muscles, and can cause severe neuropathic pain syndromes.

Investigations

Test via genetic testing and nerve conduction studies (prolonged conduction time in demyelination, decreased amplitude of compound APs in axonal).

Other conditions associated with Pes cavus?

- Charcot Marie Tooth (HMSN)
- Freidrich's Ataxia
- Idiopathic – 20%
- Fracture malunion
- Secondary to contracture
- Polio
- Syringomyelia
- CP

Myotonic dystrophy

Examination findings

Inspection:

- Myopathic facies
- Wasting of face neck and distal arms
- Frontal balding
- Cataracts
- Decreased power in face and bulbar muscles
- Slurred nasal speech
- Distal weakness
- Absent reflexes
- Hand grip myotonia
- Percussion myotonia

To conclude my examination:

- Cognitive examination
- CV and Resp system – they get cardiomyopathy
- Hypogonadism
- Urine dip for glucose – they get diabetes

To diagnose and investigate

- Serum CK
- EMG
- MD Type 1 – genetic

Lung function testing for neuromuscular

- ECG and Echo - Cardiomyopathy or conduction block
- FBG or OGTT - Diabetes
- Ophth RV - Cataracts
- Formal cognitive testing - Cognitive impairment

What is the inheritance?

- Trinucleotide repeat
- AD
- Anticipation

Natural history?

- Presents in early adulthood
- Progressive muscle wasting eventually leads the myotonia to disappear

Other myopathies?

- Duchenne's and Becker's – childhood
- Fascioscapulohumeral and Limb-girdle Muscular dystrophy – adulthood

Pathogenesis

Abnormalities of Na and K channels of the muscle membrane – prolonged discharge

Management:

- No direct treatment
- PT, OT, SW
- Geneticists
- GP – diabetes and cataracts
- Myotonia treated with phenytoin
- Cardio and Resp involvement

Peripheral neuropathy

- Feet first – longest fibres
- Achilles reflex often lost early (large myelinated fibres)
- Then glove distribution and then anterior midline of abdo wall
- Vibration lost early as large myelinated fibres. Then light touch, proprioception and then pain and temperature.

Dorsal columns – Proprioception and vibration (and 2 point discrimination)

Spino-thalamic tracts - Pain, temperature and light touch

- A fibres – Large and myelinated – Motor, vibration, light touch, proprioception
- B fibres – intermediate – autonomic
- C fibres – thin, unmyelinated – pain, temperature

Examination

- Light touch
- Proprioception
- Pain
- Vibration
- Peripheral neuropathy working your way up the limb.

Investigations

- Sugar and HbA1C
- B12
- TFTS
- IMMUNOGLOBULINS
- ESR
- (Consider Autoimmune screen, syphilis, HIV, lyme Dx in USA, heavy metals, porphyria)

Mixed motor and sensory polyneuropathy

Look for:

- Pes cavus – CMT Dx
- Diabetes – ulcers, injection sites, finger-prick marks
- Rheumatoid (Amyloid)

Causes:

Acute:

- Guillain-Barre - motor
- Diphtheria - mixed
- Porphyria - motor

Subacute:

Drugs - mainly mixed (predominantly sensory)

Isoniazid, Metronidazole, Nitrofurantoin

Dapsone - motor

Cisplatin, Vincristine

HIV medications

Alcohol

Toxins

Lead - motor

Solvents - mixed

Industrial compounds

Nutritional

B12 - painful sensory, with severe later motor symptoms

Chronic:

Paraneoplastic - sensory

Paraproteinaemia - mixed

CTD - mixed

Amyloid - mixed, with autonomic features and entrapments

Uraemia - mixed

Hypothyroidism - mixed

Diabetes - mixed

CIDP

Hereditary

CMT (HMSN)

Refsum's Dx

Causes by group:

- **Metabolic** – Hypothyroidism, **B12 and Diabetes**, Porphyria
- **Drugs** – **Alcohol**, chemotherapy, HIV medications, antibiotics
- **Paraneoplastic** (anti Hu)
- **Inflammatory** – SLE, RA, Guillan-Barre, Amyloid
- **Infectious** – HIV, Syphilis, Lyme, Leprosy
- **Hereditary** – CMT and Refsum's
- **Heavy metals and solvents**

Pure motor?

- Guillain-Barre
- Diphtheria
- Porphyria
- Lead
- Dapsone
-

Treatment for diabetic peripheral neuropathy

- Pain – Tricyclics and gabapentin
- Better glucose control
- Dietitian and diabetes nurse referral
- Footcare and podiatry

Facioscapulohumeral dystrophy

Examination:

- Permanent ptosis
- Difficulty closing eyes
- Facial weakness – cannot whistle, expressionless face
- Impaired articulation of speech
- Wasted SCMs
- Neck muscle weakness
- Winging of scapula
- Pectoralis and trapezius affected
- Biceps and triceps affected
- TJ BJ impaired

About it:

- AD, M and F
- Chromosome 4
- Normal IQ
- Normal CK
- No pseudohypertrophy
- Age of onset 10-40
- FHx

Becker MD

Examination:

- Young adult
- Prox weakness of lower extremities
- Pseudohypertrophy of calves
- Little facial weakness
- Kyphoscoliosis, decreased expansion
- No sensory loss

DD – Duchenne's

- Becker is less progressive – walk to 15-20
- Dystrophin gene
- X-linked also
- Look for respiratory and cardiac involvement.

Chorea

Hx:

- Sore throat
- Prev Rheumatic fever
- FHx – Huntington's (remember anticipation)
- OCP or pregnancy

Examination:

- 'Milk maid's grip'
- Tongue movements
- Pendular reflexes

Causes:

- Huntington's Chorea
- Chorea gravidorum
- St Vitus' dance (Sydenham's chorea)
- Drug related – more dystonic
- SLE
- Polycythaemia rubra vera
- Following stroke

Huntington's Dx:

- AD
- Full penetrance with anticipation
- Rx – Valproate or olanzepine

Parkinson's Disease/Parkinsonism

History:

HPC:

- Tremor
- Rigidity → falls, pain, stiffness
- Change in writing
- Change in speech
- Drooling saliva

- Poor sleep
- Seborrhoea
- Memory loss
- FALLS!!!

Autonomic symptoms including:

- Constipation
- Urinary problems
- Sexual dysfunction
- Sweating

Other:

- FHx
- Encephalitis
- Toxin exposure
- Neuroleptics

Examination

- Facies – paucity of blinking with fixed stare and ‘ironed’ wrinkles
- Drooling
- Resting tremor – coarse – BRING IT OUT WITH DISTRACTION – COUNT BACKWARDS WITH EYES CLOSED
- Often asymmetrical
- Bradykinesia – TEST WITH FINGER TAPPING AND TWISTING
- Increased tone with possible cog-wheeling
- Festinant gait with small steps and turning ‘en-bloc’ and reduced arm swing
- Speech problems – TEST SPEECH
- Writing – TEST WRITING

Also:

- BP problems (esp postural hypotension) – MSA-P – Striatonigral degeneration or Shy-Drager syndrome.
- Cerebellar syndrome – MSA-C (Olivopontocerebellar atrophy)
- Memory (DLB Dementia)
- Vertical ophthalmoplegia – progressive supranuclear palsy
- Dystonias – drug induced (and PSP also)

Other causes of tremor?

- PD – 4-6Hz – resting
- Cerebellar intention tremor
- Benign essential tremor
- Alcohol withdrawal tremor
- Thyrotoxicosis
- Flap of asterixis
- Neuroleptics

What is Parkinson's disease?

Idiopathic PD is a progressive degenerative disease of unknown cause most common in those in their 60s and 70s with a slight male predominance. Degeneration of dopaminergic neurons in the substantia nigra within the mid-brain leads to changes in basal ganglia functions of the control of movement, leading to the positive and negative motor symptoms, characteristically bradykinesia, tremor and rigidity. Other histological changes include the formation of intracellular Lewy bodies, as found in diffuse lewy body dementia.

What causes are there for Parkinsonism.

Idiopathic parkinson's disease does have some genetic component, although environmental factors such as toxin exposure are thought to play a large role.

Drug-induced parkinsonism is common, particularly with neuroleptic medication.

Post traumatic Parkinsonism is seen, so-called Boxer's dementia.

Vascular dementia with small vessel disease often affects the basal ganglia resulting in Parkinsonian features.

A number of infections can cause parkinsonism, including post-encephalitis, primary HIV infection, and prion disease.

Paraneoplastic phenomena have been described.

Parkinson's Plus?

A group of disorders characterised by Parkinsonian features, combined with additional neurological disorders. These include MSA (Parkinsonian and Cerebellar types – previously called striatonigral degeneration or olivopontocerebellar atrophy) and Progressive Supranuclear Palsy. Generally these conditions respond less well to anti-Parkinsonian treatments, and are more rapidly progressive degenerative diseases.

Treatments for PD?

1. Is treatment needed? Delay starting drug therapy for as long as possible, as trials have not shown any delay in the progression of the disease, although the recent NEJM article on Rasagiline is tantalising (2009). Often patients can manage with PT, OT and care input.
2. Levo-dopa combined with a peripheral decarboxylase inhibitor such as Co-Careldopa. However these formulations are only effective for a finite period of time until response to L-dopa is lost (when shorter acting formulations are helpful).

3. Dopamine agonists – ropinirole and pramipexole. May be neuroprotective (but little real life evidence) and less prone to freezing. Apomorphine is a useful drug which is given as infusion.
4. Anticholinergics – tremor
5. MAOIs – selegiline (stop dopamine breakdown)
6. COMT inhibitors – reduce metabolism of L-dopa

Ongoing trials such as PD med to evaluate how these strategies should be used and their relative effectiveness.

Other treatment strategies less often used (but with good data from some patients) include stereotactic thalamotomies and deep brain stimulator insertions.

Complications of PD:

- Falls
- Dysphagia
- Constipation
- Urinary problems
- Postural hypotension
- Cognitive decline
- Depression
- Positive symptoms including hallucinations, often provoked by medication

Multiple sclerosis

Hx

- Generally young (20-30) women > men, more in temperate climates
- General symptoms – low energy, headache, depression, aches
- Precise symptoms – sensory disturbance (incl. trigeminal neuralgia), visual loss, diplopia, limb weakness, vertigo, ataxia, sphincter problems

Ex

Anything upper motor neurone

What is MS?

MS is a common demyelinating disorder characterised by episodes of focal neurological disturbance with a classically relapsing and remitting course. These focal neurological deficits correlate with the anatomical position of plaques, areas of myelin destruction and inflammation in the white matter of the brain and spinal cord. Its cause is not fully understood, although genetic predisposition to have a disordered immune response to an environmental stimulus, possibly a viral infection, is likely.

How does MS present?

Transient sensory symptoms often precede the diagnosis as they are often minor and ignored by patients. Particular sensory symptoms typical of MS are trigeminal neuralgia in young patients, and limb sensory involvement with Lhermitte's sign (shocks in the limbs from neck flexion) or Uhthoff's phenomenon (where heat increases neurological symptoms). Optic neuritis, whether bulbar or retrobulbar, is a common presenting sign, as are gaze disorders, particularly INOs. Motor symptoms and any other cranial or central lesion may occur.

What are the different patterns of MS?

The classic pattern is one of relapsing/remitting disease, where acute episodes resolve totally for a period before returning. This pattern can last for many years and is the norm at presentation. However 20% of patients will develop secondary progressive disease, where after each event there is some improvement, however baseline function steadily deteriorates. Primary progressive disease is characterised by constant progression without significant improvement after episodes, with 15% of patients having this pattern. A few patients have what is termed benign disease, where relapses are very infrequent and there is little or no impact on patients' quality of life.

How would you investigate a patient with MS?

It would partly depend on how they presented, however common tests include:

Visual evoked potentials which will show increased latency in optic involvement. Brainstem and sensory involvement can be tested with somatosensory or brainstem auditory evoked potentials.

Neuroimaging has become a mainstay of investigation with the power of modern MRI scanners. Increased signal in the normally dark white matter on T2 images is characteristic of plaques. They may enhance during acute inflammation.

LP for oligoclonal bands are found in 95% of patients with established disease and 60% of patients at presentation. They persist between episodes. During an episode high Lymphocytes may be seen.

Hemiplegia

Ex

Unilateral:

- Fixed flexion deformity and posture
- Increased tone
- Weakness
- Brisk reflexes
- Upgoing plantar

Other signs may help localise site of lesion:

- UMN ipsilateral facial movements and visual field defect – contralat. hemisphere
- UMN ipsilateral facial movements – contralat. internal capsule
- LMN contralateral facial movements – pontine

To complete examination:

Causes:

- Ischaemic stroke
- Haemorrhage – SDH, EDH, in brain parenchyma
- SoL
- Less commonly demyelinating disease

Management:

Acute:

- Resuscitate as necessary incl. airway protection
- Hx and Ex
- Early brain imaging with CT
- Consideration of early thrombolysis or surgery at a HASU
- Aspirin, VTE prophylaxis
- Stroke ward care with specialist nurses, PT, OT

Chronic:

- Consideration of carotid endarterectomy and Rx or anticoag for AF
- Control of RFs incl BP, sugars, cholesterol
- Education of patient and family
- MDT with PT, OT, SW, Dietitian, Neuro, Nurses
- Prevention of complications like sores, contractures, chronic pain, depression

What are the Oxford Classification of Stroke?

Total Anterior circulation syndrome

All 3 of:

- Unilateral motor/sensory deficit of 2 of 3 of face, arm and leg

- Homonymous hemianopia
- Higher central dysfunction (dysphasia)

Partial Anterior Circulation Syndrome

2 of 3 of above or:

- Higher cerebral dysfunction alone
- Motor/sensory deficit limited to 1 limb.

Lacunar Circulation Syndrome:

- Pure motor (unilateral)
- Pure sensory (unilateral)
- Dysarthria/Dysphasia
- Hemiparesis with ipsilateral cerebellar ataxia

Posterior Circulation Syndrome:

- Ipsilateral cranial nerve palsy with contralateral motor/sensory signs
- Bilateral motor/sensory deficit
- EOM disorder
- Cerebellar signs
- Isolated homonymous visual field defect

ABCD2 score?

Risk of event post TIA

- A – Age >60
- B – BP >140/90
- C – Clinical features – weakness unilaterally 2 points, speech only 1 point
- D - >1 hour 2 points, 10-60 mins 1 point
- D – Diabetes

If score is 4 or above, or 2 episodes – admit for Ix (brain imaging, Echo, Dopplers, lipids, etc) . Start aspirin 300mg and control RFs.

Cerebellar syndrome

Examination:

- Arms
- Rebound with eyes closed
- Dysdiadochokinesis
- Intention tremor
- Dysmetria - Past pointing

Legs

- Heel-shin
- Gait – ataxic
- Hypotonic/ pendular reflexes

Face

- Nystagmus – towards lesion
- Speech – slurring, talting, staccato
- Titubation

What are the causes of bilateral cerebellar syndrome

- Developmental
 - Agenesis
 - Dandy-Walker
 - Arnold-Chiari
 - Von Hippel Lindau
- Neoplastic
 - Benign and malignant primary neoplasms of the CNS
 - Metastatic disease
 - Paraneoplastic – small cell lung Ca, Ovarian, Lymphoma
- Degenerative
 - Multi-system atrophy
 - Spino-cerebellar ataxias (more than 15)
 - Cerebellar ataxias – AD
 - Freidrich's - AR
 - Cerebellar degeneration
- Metabolic
 - Myxoedema
 - Thiamine deficiency
 - Hypoglycaemia
 - Inborn disorder
- Drugs
 - Alcohol
 - Phenytoin
 - Lead
 - Solvents

- Vascular
 - Stroke
 - Haemorrhage

- Infection/Inflammatory
 - Viral cerebellitis - children
 - Abscess
 - CJD

Unilateral cerebellar syndrome

Ipsilateral

Investigations for paraneoplastic cerebellar

- Anti-neuronal – anti Hu or anti Yo

Other signs of Friedreichs

- Kyphoscoliosis
- Optic atrophy
- Diabetes
- Sensory neuropathy
- Cardiomyopathy

Genetic abnormality – AR – Trinucleotide repeat - anticipation

INO – Internuclear ophthalmoplegia

- Conjugate lateral gaze disorder
- Failure of Adduction on lateral gaze often with nystagmus
- Due to a deficit in the coordination of vision in the medial longitudinal fasciculus in the pons (between the CN VI and CN III nuclei).

Causes:

Unilateral

- MS
- Stroke
- Trauma

Bilateral

- MS
- Toxicity – phenytoin
- Syringobulbia
- Trauma
- Brainstem infarction

What is 1 and a ½ syndrome?

Similar to INO but involving the PPRF and the MLF which causes failure of any lateral movement of the affected eye, and failure of adduction on the contralateral eye, leaving only abduction of the other eye still present.

III nerve palsy

Examination

- Ptosis (70% of L paplebrae by Parasympathetic III fibres) – may be complete
- Mydriasis with decreased direct and consensual constriction
- Inferio-lateral deviation of eye in primary position
- Diplopia on upwards and inwards gaze
- Loss of accomodation

Anatomy

- Nucleus in midbrain. Emerges ventrally by the cerebral peduncle
- Runs past the Post. Communicating artery
- Trough the cavernous sinus
- Through the superior orbital fissure
- Splits into superior and inferior

The nerve is structured with the parasympathetic fibres outermost, which means they are the most sensitive to compression, and are unaffected my infarction.

Syndromes and causes:

Bilateral – midbrain

Unilateral with tremor – Red nucleus – BENEDIKT'S SIGN

Unilateral with contralateral hemiparesis –cerebral peduncles – WEBER'S SYN

- Caused by demyelination, tumour, aneurysm, infarction

With Headache/retroorbital pain and pupils affected first

- post. Comm. Artery or basilar aneurysm

With Head injury/decreased consciousness

- transtentorial herniation

With meningism and other CNs affected

- basal meningitis (bacterial, viral, fungal, TB, cancer)

Sudden onset with papillary sparing – sudden infarction of the nerve 'MEDICAL'

- Diabetes
- Hypertension
- PAN
- SLE

In association with IV, VI and 1st division of V – Cavernous sinus pathology

- tumour
- Aneurysm
- Sinus thrombosis

With proptosis and IV, VI and 1st div of V – orbit/ fissure

- Tumour
- Granuloma
- Periosteitis

Investigations:

- BP
- BM
- ESR
- TFTs
- CT
- MRI
- Angiography

IV nerve palsy

Examination

- Often appear conjugate in primary position
- The eye is unable to look down when adducted (looking in).
- Head may tilt to compensate (to use abducens or oculomotor instead)

Anatomy

Nucleus in the mid-brain. Only cranial nerve to decussate and emerge dorsally by the superior aspect of the cerebellum. Then through the cavernous sinus and through the superior orbital fissure.

Syndromes and causes:

With contralateral hemiparesis/hemisensory loss – midbrain lesion

- infarction
- demyelination
- tumour

With cerebellar signs:

- Cerebellar tumour

In association with III, VI and 1st division of V – Cavernous sinus pathology

- tumour
- Aneurysm
- Sinus thrombosis

With proptosis and III, VI and 1st div of V – orbit/ fissure

- Tumour
- Granuloma
- Periosteitis

VI nerve palsy

Examination

Normal eye position in primary position. Diplopia and strabismus on lateral gaze towards the side of the lesion. True horizontal diplopia. Often the strabismus is not appreciable; check the diplopia is true (disappears when you test one eye) the direction of maximal diplopia, and the eye responsible for the outermost image.

What else would you check:

- Other cranial and peripheral nerves
- BP
- CBG
- Hearing

Anatomy:

Supplies lateral rectus which abducts the eye. The nucleus lies in the pons at the floor of the 4th ventricle. It is close to the VII nucleus. It then runs along the petrous portion of the temporal bone before entering the cavernous sinus and through the superior orbital fissure.

Syndromes and causes:

DIABETES

In association with III, VI and 1st division of V – Cavernous sinus pathology

- tumour
- Aneurysm
- Sinus thrombosis

With proptosis and III, VI and 1st div of V – orbit/ fissure

- Tumour
- Granuloma
- Periosteitis

False localising sign:

- **long intracranial course – susceptible to stretch damage from raised ICP of any cause**
- **Often bilateral**

In association with pain in V nerve (esp. 1st division)

- **Petrositis**
- (Gradenigo's syndrome)

Spastic paraparesis

History:

- Timings
- Back pain – general, localised, radicular?
- Sensory symptoms
- Bladder and bowels
- Functional status
- FHx and Birth Hx
- Complications – UTIs, retention, pressure sores, DVTs

Examination:

- Hypertonia
- Clonus
- Hyperreflexia
- Weakness
- Mild wasting without fasciculations

+/- clues to the cause:

- sensory involvement/ sensory level
- scars (on limbs – tendon release, on body/chest – malignancy)
- cerebellar signs (Friedrichs)
- Upper limb signs (lower MN – cervical myelopathy, UMN – cranial cause)
- Eye signs (MS)

Causes:

By type:

- Cord compression by – tumour, disc, EDH, abscess, haematoma
- Demyelinating – MS, transverse myelitis
- Hereditary spastic paraparesis
- Trauma
- MND
- Freidrich's ataxia
- Syringomyelia

By age:

In the young:

- **Freidrich's**
- **MS**
- **Trauma**
- **HIV**

In middle age:

- **MS**
- **Trauma**
- **MND**
- **Syringomyelia**
- Transverse myelitis
- Familial spastic paraplegia
- Tabes dorsalis
- SAC degeneration of the cord
- HIV
- Spinal cord tumour

In old age:

- **Spinal impingement from OA or tumour (likely metastatic)**
- **Vit B12**
- **Anterior spinal thrombosis**
- Atherosclerosis of spinal cord vasculature

Investigations:

- FBC,CRP, ESR, Cultures – infection
- Syphilis serology
- Vit B12/Folate
- PSA
- ALP
- Protein electrophoresis
- MRI/CT depending on indication
- CSF – oligoclonal bands
- ?HTLV-1

Causes of absent Ankle jerks and upgoing plantars?

- Vit B12 deficiency – SACDC
- Tabes dorsalis
- Friedrich's ataxia
- Dual pathology – Peripheral neuropathy (e.g. DM) and coexistent spastic paraparesis

Myasthenia gravis:

Examination:

- Fatigability – look up and count to 50 – ptosis and neutral gaze
- Smile – buccinator weakness with myasthenic snarl
- Nasal speech
- Distal weakness
- Normal sensation and reflexes

What is it?

Myasthenia gravis is a neuromuscular junction disorder characterised by weakness of some muscle groups with fatiguability. It is caused by autoimmune destruction of postsynaptic nicotinic receptors by AChR antibodies (complement mediated). Thymic dysfunction is often involved.

Ix:

- AChR Abs
- Tensilon test
- EMG
- CT Chest – Thymoma

Rx:

- Anticholinesterase drugs – pyridostigmine, etc
- Steroids
- Steroid sparing agents
- Consider thymectomy (if thymoma or v severe)
- IVIG and plasmaphoresis

Myasthenic crisis:

- Sit up
- Manage airway
- Oxygen and monitor CO₂
- IV neostigmine
- SC Atropine
- High dose Prednisolone
- Consider plasmaphoresis or IVIG
- NBM until swallow assessed

Complications:

- Cholinergic or myasthenic crisis
- Resp. failure
- Aspiration
- Neonatal MG from AB crossing placenta

Precipitants of a myasthenic crisis

- Infection
- Physical stressors – trauma, surgery
- Pregnancy
- Sleep deprivation
- Pain
- MEDICINES
 - Antibiotics – Gent, Ampicillin, Cipro, Erythro
 - Antiepileptics – phenytoin, gabapentin
 - Cardiac meds – beta blockers, Ca²⁺ channel blockers, antiarrhythmics
 - Some contrast
 - Prednisolone – short term

Motor neurone disease

On examination:

- Pseudobulbar palsy – weakness of V, VII, IX, X, XI, XII. Expressionless face. Speech monotonous. NOTE – jaw jerk brisk, tongue immobile, pointed and cannot protrude.
- Bulbar palsy – CN nuclei damage. Atrophy and fasciculations esp on tongue. Jaw jerk absent.
- Corticospinal tract involvement with increased tone, brisk reflexes and extensor plantars.
- Muscular atrophy – severe wasting and fasciculations from anterior horn cell death.

Key things to look/ask for:

- Bladder is NEVER involved
- EOMs are NEVER involved
- There are NEVER any sensory signs.

Differential diagnosis?

- Stroke can cause pseudobulbar palsy, but no LMN signs.
- Cervical myelopathy will cause mixed UMN LMN signs, but in a specific distribution and with sensory changes.

Others:

Frontal dementia

What is it:

Progressive, degenerative condition with damage to UMN and LMN. AKA Amyotrophic lateral sclerosis. Small amount is familial. Commoner in men. Onset generally after the age of 50. The cause is unclear, but may be triggered by a viral infection.

What can mimic it?

- HIV disease
- Lymphoproliferative disease
- Paraproteinaemia
- Lead poisoning

Ix:

- EMG – denervation and fibrillation
- NCS – neuropathy with conduction block
- MRI – exclude spinal cord Dx
- Check calcium, paraproteins, HIV

Treatment:

- Supportive - Emotional and physical
- MDT – SALT, PT, OT, DNs, Dietitians
- Considering feeding strategies
- Respiratory support is controversial
- 1 disease modifying Rx – Riluzole – anti-Glutamate properties.

Cervical myelopathy

LMN signs with wasting, anaesthesia and diminished reflexes at the level of compression.

UMN signs below in pyramidal distribution. Increased tone, clonus, extensor plantars. Less sensory loss.

Lesion may be due to:

Cervical spondylosis

Disc protrusion

SoL – neoplasm, abscess, haematoma

Syringomyelia

Development of a syrinx within the central spinal cord. Mainly in C spine but can extend down, or up into the brain stem. Often associated with obstruction of CSF flow from Chiari malformation or following trauma or infection.

Examination:

- Dissociated sensory loss (Loss of pain and temperature in cape distribution).
- INTACT VIBRATION, JPS AND LIGHT TOUCH
- Wasting of small muscles of the hand and winging of scapula.
- Then may develop long tract signs (spasticity and hyper-reflexia).
- Brain stem signs if affected. Incl HORNERS (Syringobulbia)

Treatments?

- Syringoperitoneal or SA shunting
- Foramen magnum release if Arnold-Chiari malformation

Friedreich's ataxia

Hx

- Age of onset
- High arched foot in family
- Scoliosis in childhood
- Cerebellar dysarthria/ataxia

Examination:

- Pes cavus
- Pyramidal weakness in legs
- Cerebellar signs
- Ataxia
- Impaired vibration and proprioception
- Romberg's positive
- Absence of reflexes with upgoing plantars
- Distal muscle wasting
- Optic atrophy

What is it?

- The commonest congenital inherited ataxia.
- AR disorder. Demyelination and gliosis of posterior columns, corticospinal tracts and spinocerebellar tracts. Peripheral nerves show demyelination.

Proximal myopathy

History:

- Weakness standing from a chair, walking up stairs, combing hair

Examination:

Proximal weakness and gait disturbance

Causes:

- Diabetic amyotrophy – KJ loss, sensory loss on thigh, v.v.painful
- Cushing's disease
- Thyrotoxicosis
- Polymyositis
- Drugs – alcohol, steroids
- Carcinomatous
- Osteomalacia
- Hereditary MD

Common peroneal nerve palsy

On examination:

- LOOK FOR ORTHOSES
- Weakness of dorsiflexion at ankle and toe
- Weakness of eversion
- PRESERVED INVERSION – differentiate an L5 lesion
- Intact reflexes
- Sensory loss to dorsum and outer aspect of the foot (via sural N.)
- High stepping gait with foot drop

ANATOMY:

Arises from the sciatic nerve in the popliteal fossa from where it goes laterally closely involving the fibular head. Inervated by the L4/5 nerve roots.

Caused by:

- Any cause of trauma, pressure or entrapment around the fibular head
 - o Rarely this may be a compartment syndrome following surgery
- Partial sciatic nerve lesion
- Sacral plexus damage
- Any cause of mononeuritis multiplex (DM, vasculitis, amyloid, renal failure, B12)

Ix:

- Hx of pressure
- XR Knee and Tib-Fib
- FBG, HbA1c
- CRP ESR
- Serum electrophoresis
- U&Es
- B12
- If cause still unclear consider nerve conduction studies

Ulnar palsy

Examination:

- Claw hand – 4th and 5th fingers
- Hypothenar eminence wasting
- Weakness of muscles (finger abduction, thumb adduction)
- Sensory loss of ulnar border of the hand

Caused by:

- Injury at elbow
- Entrapment at elbow
- Wrist and finger flexion

Radial palsy

Examination:

- Weakness of wrist extension, finger extension
- Sensory loss on dorsum of hand

Causes:

- Humerus fracture
- Prolonged pressure
- Lipoma, fibroma
- Systemic causes of a mononeuritis multiplex

Medial nerve

Examination:

- Sensory loss to medial border of the hand
- Loss of LOAF Muscles: LOAF – lateral 2 lumbricals, opponens pollicis, abductor pollicis brevis and flexor pollicis brevis
- Wasting of thenar eminence
- Look for CTS – Phalen's and Tinel's test

Causes of carpal tunnel syndrome

- Rheumatoid
- Acromegaly
- Hypothyroidism
- Amyloid
- Pregnancy
- Wt gain

Holmes Adie

A tonic (Adie's) pupil associated with absent reflexes and occasionally other autonomic features such as diarrhoea and anhidrosis. More common in young women.

The pupil is large and unresponsive to light, but will very slowly constrict to accommodation (most of the time!).

Likely viral cause which damages the parasympathetic neurons in the ciliary ganglion

Argyll Robertson Pupil

Bilateral small pupils

Irregular

Unreactive to light

Reactive to accommodation

Neurosyphilis (also any midbrain lesion from neoplastic, vascular or demyelinating processes). Likely caused by damage to Edinger-Westphal nuclei in midbrain (part of dorsal midbrain or parinaud's syndrome).

Causes of small pupils:

- Horner's
- Argyll Robertson
- Pilocarpine
- Pontine haemorrhage
- Y are you happy? Morphine

Subacute combined degeneration of the cord

History:

- FHx of pernicious anaemia or autoimmune disease
- Chronic diarrhoea
- Abdominal surgery
- Tingling or paraesthesia
- Diet

Examination

- Upgoing plantars and brisk knee reflexes
- ABSENT ANKLE JERKS
- Decreased light touch, vibration and JPS – Dorsal columns
- Romberg's positive

Also look for:

- Anaemia
- Abdo scars
- Argyll Robertson pupil
- Fundus (optic atrophy)
- MMSE (dementia)

Investigations:

- FBC
- Retics
- Haematinics
- Bone marrow
- Parietal cell and IF antibodies

Causes of B12 deficiency:

- Vegan
- Reduced IF secretion (gastric resection, pernicious anaemia)
- Ex consumption in the bowel (bacterial overgrowth, tapeworm)
- Lack of absorption (terminal ileal resection or inflammation)

Neurosyphilis

Tabes dorsalis

- Posterior spinal root and posterior column dysfunction
- Argyll Robertson pupil
- Optic atrophy
- Loss of pain sensation, JPS
- Romberg's negative
- Charcot's joints

May get urinary incontinence, visceral crises with abdo pain and diarrhoea and tenesmus

Jarisch Herxheimer reaction – tachy and fever post Penicillin – entotoxin release

Lyme Dx – *Borrelia burgdorferi*

- Bilateral Bell's palsy
- Peripheral neuropathy with radiculitis
- Arthritis
- Subacute encephalitis

Cerebellopontine angle tumour

VII and VIII (V, VI less often)

- Impaired hearing on affected side (sensorineural)
- Facial nerve weakness
- Trigeminal sensory and motor loss with absent corneal reflex and jaw jerk
- Cerebellar signs
- Papilloedema
- Is there neurofibromatosis?
- Papilloedema and symptoms of raised ICP

Causes:

- Acoustic neuroma
- Meningioma
- Pontine glioma
- Aneurysm
- Ca Nasopharynx
- TB/Syphilis

Investigations:

- CT or MRI head
- Syphilis serology
- Audiography
- Vertebral angiography
- Consider LP is above normal

Rx:

- Microsurgical resection
- Stereotactic radiosurgery
- Conservative

Pseudobulbar palsy

UMN lesion – common

Hx

- Dysphagia
- Nasal regurg
- Speech changes
- Emotional lability
- Hx stroke, MS, MND

Examination:

- Spastic, pointed tongue
- Donald Duck speech
- Poor palatal movements
- Brisk jaw jerk

Look for Upper limb signs

Causes:

- Stroke – internal capsule lesions
- MS
- MND

Rx

MDT- SALT – Ix with video fluoroscopy

Bulbar palsy

LMN – rarer (CN or medullary lesions)

Hx

- Dysphagia, nasal regurgitation
- Slurred speech
- Difficulty chewing
- Choking esp on liquids

Examination:

- Nasal speech – poor articulation
- Wasting of tongue with fasciculations
- Weakness of soft palate
- Accumulation of saliva
- Absent jaw jerk

ULs – wasting and fasciculations

Causes:

- MND
- GBS
- Syringobulbia
- Polio
- Neurosyphilis or neurosarcoid

Lateral medullary syndrome (Wallenberg's)

Hx:

Nausea, nystagmus

Limb ataxia

Dysphagia and hiccups

Examination:

- Ipsilateral:
 - Nystagmus
 - V, VI, VII, VIII
 - IX, X - Bulbar palsy
 - Horner's
- Contralateral:
 - SST signs – Pain and temperature loss

Caused by:

PICA or vertebral artery pathology involving medulla and cerebellum unilaterally

- Stroke
- Bleed
- Mass
- Dissection

Winged scapula:

Examination:

Unilateral or bilateral?

Are other UL muscles involved?

Nerve root – C5-7

Nerve – Long thoracic nerve

Muscle – Serratus anterior and Trapezius

Causes:

Facioscapulohumeral dystrophy

Brachial neuritis (triggered by virus or vaccination)

Diabetes

Proximal brachial plexus injury

C-spine pathology