HAP33: Weakness not due to stroke

Myasthenia Gravis

Prevalance ~5/10000 of UK population.
Peak incidence in 3rd and 6th decades of life.
Autoimmune disease – targeted at post-synaptic ACh receptors (contrast Lambert-Eaton Myasthenic Syndrome – target is pre-synaptic voltage-gated calcium channels)

Presents with weakness and fatigability – diagnosis is generally based on this. The Edrophonium test is now rarely used, and generally only when diagnosis is uncertain. Making the primary diagnosis is rare in ED, but may need to manage crisis.

First line treatment is with acetylcholinesterase inhisbitors (eg Pyridostigmine) – symptom control. Disease progression can be improved with steroids, immunosuppressants (Azathioprine), IVIG, Plasmapheresis, Thymectomy.

Mortality from a crisis is around 4-8%. Any shortness of breath or dysphagia in a known MG patient should be taken seriously. Assess need for BiPAP or intubation. For RSI need to increase dose of sux and reduce dose of non-depolarising neuromuscular blocking drugs.

Infection, trauma, and drugs (including many antibiotics) can trigger a myasthenic crisis. Aminoglycosides, quinolones, macrolides are contra-indicated, as are multiple other drug classes.

Multiple sclerosis

Multicentric, multiphasic, idiopathic, inflammatory demyelinating condition of brain and spinal cord.

Prevalence ~80 per 100000, twice as common in females. Onset typically age 20-50, rare in those over 60. It is a leading cause of non-traumatic disability in young adults.

Three patterns:

- Relapsing remitting (70% of cases)
- Primary progressive
- Acute fulminant (Death or severe disability within days)

New episodes rarely occur more often that 1.5 times/year

Characteristically see episodes of multiple neurological dysfunction, usually referable to CNS, typically with recovery – may be complete in early stages, but cumulative defects occur with time.

MRI will usually make the diagnosis. In known patients, investigations are to rule out alternative diagnoses – CT for intracranial bleed, MRI for transverse myelitis, BM for hypoglycaemia, etc.

Treatment is usually with steroids for acute episodes. Control of pyrexia is important – elevated temperature can exacerbate weakness. Treatment is otherwise targeted at complications – treatment of infection, catheterisation for retention, etc.

2/3 of patients die as a result of the disease and its complications (including suicide); life expectancy is 5-10 years less than for the general population.

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Guillain-Barré syndrome

~1200 cases per year in the UK.
2/3 report infection in preceding weeks.

Group of conditions:

- Acute Inflammatory Demyelinating Polyradiculoneuropathy (most common in UK)
- Acute Motor Axonal Neuropathy)Axon directly affected
- Acute Motor and Sensory Axonal Neuropathy)
- Miller-Fisher (Ophthalmoplegia, ataxia, areflexia – most common form in Far East)

Onset may be with pain or paraesthesia, prior to weakness. Pain may be severe and difficult to control.

Rapidly progressive bilateral weakness, usually spreads proximally, with reduced or absent deep tendon reflexes. Sensory symptoms are typically mild.

The condition usually progresses over four weeks, with a gradual and variable recovery – better in children. There is usually no fever.

Respiratory failure and autonomic failure can occur – regular lung function testing is required.

MRI and nerve conduction tests may confirm the diagnosis, but neither is 100% sensitive. Elevated CSF protein with normal WCC is also suggestive.

Severity is arbitrarily defined as mild or severe based on ability to walk.

IVIG or plasma exchange show some benefit. Treatment is otherwise supportive, with particular attention to pressure areas, DVT risk, pain control, and monitoring for respiratory failure.

Tetanus

Clostridium tetani – obligate anaerobe
UK: ~10 cases annually, 29% mortality
Cases can be generalised, localised or cephalic (both can generalise), or neonatal (early onset of generalised)

Immunisation status	Clean wound	Tetanus-prone wound	
	Immunisation	Immunisation	HTIG
Full primary course + two boosters	No	No	If high risk
Full primary course, last booster <10 years	No	No	If high risk
Incomplete primary or complete primary but <2 boosters, last >10 years	Yes (then complete)	Yes (then complete)	Yes
Not immunised/not known-	Yes (then complete)	Yes (then complete)	Yes

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Tetanus prone:

- wounds or burns that require surgical intervention that is delayed for more than six hours
- wounds or burns that show a significant degree of devitalised tissue or a puncture-type injury, particularly where there has been contact with soil or manure
- wounds containing foreign bodies
- compound fractures
- wounds or burns in patients who have systemic sepsis.

High risk:

- Heavy contamination with spores
- Extensive devitalised tissue

HTIG: 250iu; 500iu if delayed >24 hours/heavy contamination/burns

Usual presentation – trismus and facial muscle ('risus sardonicus') initially, with spasm and rigidity spreading distally, then involving pharyngeal muscles and causing laryngospasm. As it progresses, respiratory compromise, dislocations, fractures, and rhabdomyolysis can occur.

Autonomic features typically commence several days after clinical infection – unstable BP, arrhythmias, hyperthermia, sweating, urine retention, high output renal failure, ileus, diarrhoea.

There is no alteration of consciousness.

Tetanus toxin in a serum sample (prior to HTIG) confirms tetanus; negative does not exclude. Negative antibodies make diagnosis more likely. Positive wound culture/swab rare.

Death usually due to respiratory failure if untreated or cardiac arrhythmia if ventilated.

Treatment:

- Debride wounds
- Antibiotics (metronidazole)
- HTIG
- Benzodiazepines for spasms
- Treat complications; avoid beta-blockers

Botulism

Clostrium botulinum – obligate anaerobe
UK: ~11 cases/year, most from wounds (IVDU most at risk)

Presents with cranial nerve palsy, bulbar palsy, descending paralysis, with developing autonomic dysfunction. Typically no fever and no altered conscious level.

Treatment:

- Human botulinum immunoglobulin
- Supportive measures