Neurodegenerative Disorders

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Multiple Sclerosis

- Relasping remitting disorder with plaques of demyelination throughout CNS
- Commoner in temperate areas
- Diagnosis
  - Clincal demonstration of lesion
  - MRI scan via T2
  - CSF
    - Oligoclonal bands of IgG
    - Upto 50 lymphocytes
  - Evoked potentials - delayed
- Clinical Features
  - Sensory: pins and needles, decreased vibration sense, trigeminal neuralgia
  - Motor: spastic weakness, hyper-reflexia
  - Cord: urgenecy / retention, erectile dysfunction
  - Visual defects: diplopia, heminopia, optic neuritis, nystagmus
  - Cerebeallar: truncal / limb ataxia, intention tremor, scanning speech
  - Fatigue, depression, decreased cognition
- Treatments
  - Disease Modifying Agents
    - Interferon
      - Decrease relapses; expensive
      - SE: flu like symptoms, depression, abortion
    - Glatiramer Acetate
      - Mimics myelin protein
    - Anti TNF
      - Decreases relapses significantly
      - SE: opportunisitic infections
  - Relapse
    - Methylprednisolone
      - 1g / 24hr IV / PO for 3 days
      - Use sparingly - less than 3 times a year

Parkinsonism

- Syndrome
  - Tremor
    - Resting, pill rolling tremor
    - Coarser than a cerebelair tremor
  - Rigidity
    - Throughout ROM (lead pipe)
    - Present equally in flexors and extensors
  - Bradykinesis
    - Slowness of movement initiation
    - Progressive reduction in speed / amplitude of repetitive actions
  - Other features
    - Monotonous speech, expressionless face, dribbling
    - Decreased peristalsis, micrographia
- Parkinson's Disease
  - Due to degeneration of substantia nigra dopaminergic neurones (presence of Lewy bodies in this area)
  - Usually starts after 50, becomes more common with age
  - Management
    - Alliance between doctors, physios, nurses, GP, patients and carers
    - Respite care
    - Rehabilitation
  - Medical Management
    - L Dopa
      - Start when PD interferes with life
      - Effect wears off with time
      - Balance symptom relief with bad side effects
      - SE: nausea, unwanted movements, orthostatic hypotension, arrhythmias, psychosis, compulsions
    - Dopa Decarboxylase Inhibitor
      - Prevents peripheral breakdown of L Dope
    - On-off
      - Switching between times of exaggerated involuntary movment and of immobility
    - Early use of dopamine agonists may reduce on-off and allow lower doses of L Dopa
    - MAO-B inhibitors
      - Inhibitor breakdown of available dopamine
      - For example selegiline
  - Neuropsychiatric Complications
    - Seen in at least 50% of cases
    - Often due to interplay of illness and drug SE
    - Depression is very common
- Parkinson's Plus / Other causes
  - Drug induced
    - Drugs for schizophrenia - these reduce available dopamine
  - VODKA signs
    - Vascular causes
      - Orthostatic hypotension + atonic bladder = multi system atrophy
        - Parkinsonism + autonomic dysfunction + cerebeallar ataxia
        - Extra pyramidal symptoms may precede autonomic failure
        - MSA may be unmasked by sudden worsening of postural hypotension after treatment with L Dopa
        - Survival: less than 10 yrs following diagnosis
    - Dementia + vertical gaze paralysis = supranuclear palsy
      - No effective treatment
      - Prognosis is less than 10 yrs following diagnosis
      - Vertical gaze palsy is most significant clinical feature
    - Kayser-Fleischer Rings
      - Wilson's disease
      - Hereditary copper accumulation
    - Apaxic gait = communicating hydrocephalus

Motor Neurone Disease

- Degeneration of neurones in motor cortex, CN nuclei and anterior horn cells
  - UMN and LMN may be affected but there is no sensory loss or sphincter disturbance
  - Never affects external eye movement
- Signs, Symptoms and Diagnosis
  - Over 40 with stumbling spastic gait and weak grip
  - UMN signs (weakness, spasticity, brisk reflexes, upgoing plantars) and LMN signs (weakness, wasting, fasiculations)
  - Progressive UMN and LMN signs
- Clinical Pattern
  - Bulbar Palsy
    - Speech and swallowing difficulties
    - Signs of LMN lesion with wasting / fasiculation of the tongue
    - Contrast with pseudo bulbar palsy
  - Amyotrophic Lateral Sclerosis
    - Combined LMN wasting and UMN signs with weakness
    - Greater incidence in Gulf war veterans
  - Progressive Musuclar Atrophy
    - Anterior horn cell lesion affecting distal muscles before proximal
    - Better prognosis than ALS
- Treatment
  - MDT approach
  - Anti glutamate drugs (prolongs life by 3 months)
  - Symptomatic relief
    - Drooling - TCAs
    - Dysphagia - blend food
    - Joint pain - analgesic ladder
- Prognosis
  - Incurable; often fatal within 5 years
  - Prognosis is worse with bulbar onset disease
  - Patients die of respiratory failure

Dementia

- Progressive deficits in several cognitive domains
  - Assume confusion is due to acute illness until proved otherwise
- Epidemiology
  - Rare < 55 yrs; 10% prevalence > 65 yrs
  - 20% prevalence > 80 yrs
- Causes
  - Alzheimer's
    - Leading cause of dementia
      - Mean survival 7 yrs from onset
    - Acquired deficits of visual spatial skill, memory and cognition
    - Causes
      - Accumulation of beta amyloid peptide resulting in progressive neuronal damage, increased number of senile plaques and loss of acetycholine
      - Selective neuronal loss, particuarly hippocampus, amygdala, temporal cortex and subcortical nuclei
  - Vascular
    - 25% if dementias, cumulative effects of many small strokes
    - Sudden onset and stepwise deterioration is characteristic
    - Look for other evidence of vascular pathology
  - Lewy Body
    - Characterised by Lewy bodies in brainstem and neocortex, fluctuating cognitive loss, alertness, Parkinsonismand visual hallucinations
  - Fronto-temporal dementia
    - Frontal / temporal atrophy without Alzheimer histology
    - Behavioural / personality change, preservation of episodic memory and spatial orientation, disinhibition, emotional unconcern
  - Ameliorable causes
    - Low T4, low B12, syphilis, tumour, sub dural haematoma
    - Parkinson's, HIV, normal pressure hydrocephalus, Whipple's disease
- Tests
  - FBC, ESR, U&E, Ca2+, LFT
  - TSH, autoantibodies, folate B12
  - CT / MRI to indentify any structural pathologies
- Management
  - Neuropsychiatric referral
  - Key worker / care management plan
  - Carer stress is inevitable
  - Give relevant specific treatments and treat any concurrent illness (these may add to confusion)