Neurology

Brain death

Tests

EEG in Monitoring 3.B.2

Neuromuscular disorders and anaesthesia
Brain death
Coma GCS < 8, potential for recovery
Persistent coma
Persistent vegetative state: brainstem function intact
Brain death: brainstem function lost (no spontaneous ventilation)
Death: “Irreversible cessation of circulatory and respiratory functions or irreversible cessation of all functions of the entire brain, including the brainstem.”

Diagnostic criteria of brain death
Deep coma
confirm clearance of depressant drugs
normal body temperature
no gross electrolyte or metabolic disturbance
Apnoea
no relaxants, opioids or other depressants
raised PaCO₂, normal PaO₂
Irreversible structure brain damage

Procedure
Pupillary response to light absent
Corneal touch reflex absent
Vestibulo-ocular reflexes absent
20 ml ice cold water in ear canals, no eye movement
Facial motor response to trigeminal distribution pain absent
Gag and cough reflexes absent
Apnoea in the presence of PaCO₂ > 60 mmHg, pH < 7.30
Two examinations at least 2 h apart by separate qualified doctors
Angiography required if no diagnosis or tests incomplete (e.g. eye injury)
Modified criteria < 1 year of age due to greater recovery potential
Tests

Flicker fusion test
- Increasing frequency of flashing light
- Frequency at which light appears steady is recorded
- Used to assess degree of hypotension producing significant cerebral ischaemia
Neuromuscular disorders and anaesthesia

Literature
- Little high-quality evidence
- Many case reports, few series
- Small numbers for all conditions except myasthenia gravis (thymectomy)

General concerns
- Pre-op assessment
  - Weakness
  - Respiratory failure
  - Upper airway maintenance
  - Assessment of need for back-up ventilated bed
  - Decreased fitness and exercise tolerance
  - Possible myocardial involvement
  - Documentation of functional state before anaesthesia
- Deformity
  - Airway assessment
  - Likely ease of intubation
  - Positioning
- Investigations
  - Respiratory function testing
  - Blood gas analysis
  - ECG, echocardiography if indicated

Intra-op management
- Risk of aspiration
- Decision regarding need for muscle relaxants
- Altered sensitivity to relaxants, need for NMJ monitoring
- Variable distribution of weakness makes relaxant monitoring unreliable
- Appropriate management of steroids or other drugs

Post-op
- Careful monitoring of recovery to adequate tidal volume
- Monitoring for respiratory failure post-extubation
- Possible ICU ventilation

Motor neurone disorders

Multiple Sclerosis
- Aetiology
  - Unknown, environmental and genetic associations
  - Patchy demyelination in the CNS
- Features
  - Signs
    - Typical onset in females 20-40 years of age
    - Variation in severity over minutes to years
    - Common involvement of optic nerve and oculomotor pathways
    - Exacerbated by
      - Parturition, elevated temperature, other stresses
    - Progression to motor weakness of limbs
- Investigations
  - MRI shows “plaques”
  - Abnormal sensory evoked potentials
  - CSF IgG and myelin basic protein elevated
- Treatment
  - Steroids, ACTH, immunosuppressants used with some effect

Anaesthetic considerations
Hyperthermia may cause exacerbation of weakness
Induction agents, volatiles, relaxants (including suxamethonium) all known to be safe
Relaxants may exacerbate weakness directly
Regional
  Possible increased permeability of blood-brain barrier
  Increased risk of CNS toxicity from local anaesthetics
  Possible increased risk of histotoxicity from spinal local anaesthetic
  Not supported by clinical data

Guillain-Barré syndrome
Aetiology
  Cell-mediated autoimmune response
  Causes demyelination of peripheral nerves
  Commonly post-viral (herpes viruses, influenza, para-'flu, HIV)
  Possibly post-vaccination (TB, tetanus, typhoid)
Features
  Signs
    Progressive motor weakness or more than one limb
    Areflexia or hyporeflexia
    Symmetrical and progressive
    Results in flaccid paralysis
    Mild sensory involvement, mostly vibration and proprioception
    Cranial nerve involvement in 45%
    Autonomic dysfunction
      Circulatory instability: hypo- and hyper-tension
      Bradycardia, tachyarrhythmias
      Ileus, urinary retention
Investigations
  CSF: low WCC, high protein
  Abnormal nerve conduction studies
Timecourse
  Recovery 2-4 weeks after onset
Treatment
  Plasma exchange proven effective
  Immunoglobulin therapy is as effective, but more relapses
  Steroids commonly used but unproven
  Supportive management: ventilation etc.
Anaesthetic considerations
  Increased aspiration risk with bulbar palsy
  Respiratory muscle weakness
  Circulatory instability on induction
  Exaggerated response to pressors and vasodilators
  Suxamethonium contraindicated

Motor neurone disease
Aetiology
  Some inherited, most sporadic (amyotrophic lateral sclerosis)
  Progressive degeneration of upper and lower motor neurones
Features
  Signs
    Sensory and autonomic pathways spared
    Cerebral function largely spared
    Several patterns of progression
      Cranial vs somatic
      Upper vs lower
Similar to post-polio syndrome

Investigations
None specific
EMG shows denervation

Anaesthetic considerations
Upper airway and respiratory muscle weakness
Lack of specific treatment raises ethical problems
Suxamethonium contraindicated due to potassium release

Neuromuscular junction disorders

Myasthenia gravis

Aetiology
Autoantibodies (IgG) to α-subunit of ACh receptors on skeletal muscle
Thymic abnormalities in 75% of patients
Associated with other autoimmune diseases
Hypothyroidism, RA, SLE, pernicious anaemia

Neonatal variants
Children of myasthenic mothers
Transient weakness from maternal IgG
Hereditary myasthenia
No autoantibodies, structurally abnormal receptors

Paraneoplastic variant (Eaton-Lambert syndrome)
Autoantibodies against voltage-gated Ca^{2+} channels
Decreased ACh release from nerve terminals
Association with autonomic dysfunction, reduced gastric motility
Predominantly limb involvement
Little bulbar involvement

Features

Signs
Ocular involvement first: ptosis and diplopia
Commonly bulbar weakness
Asymmetrical trunk and limb involvement

Investigations
Edrophonium (Tensilon™) test
1 mg-6 mg edrophonium, 0.6mg atropine
Autoantibody assay
False positives in RA and some family of affected patients
False negative immediately after anaesthesia

Treatment
Anticholinesterase drugs
Pyridostigmine 60 mg qid (up to 750 mg/day)
Plasma exchange
Immunoglobulin (unknown mechanism)
High dose corticosteroids
Azathioprine, cyclophosphamide, cyclosporin
Thymectomy after medical optimization gives the best results

Anaesthetic considerations
Decision to continue anticholinesterase drugs depends on severity
Anticholinergic agents may be required to cover bowel anastomoses
Myasthenic crisis (acute exacerbation)
Described worsening with local anaesthetics, muscle relaxants, narcotics, ether, aminoglycosides
Weakness can also be due to anticholinesterase overdose
Regional anaesthesia is well tolerated
General anaesthesia
Propofol plus opioid, or
Volatile only
Produces 50% twitch fade at 1.0 MAC
Suxamethonium is safe
Possibly decreased sensitivity
Decreased metabolism if on high anticholinesterase dose
High incidence of phase II block at normal dose
Non-depolarizing agents are usually avoided
Increased sensitivity, difficulty reversing

**Muscle disorders**

**Myotonia dystrophia**

**Aetiology**
- Disorder of relaxation of skeletal muscle (AD 19q)
- Slow reuptake of $Ca^{2+}$ into sarcoplasmic reticulum
- Multiple tissues affected
- Myotonia congenita variant present from birth
- Paramyotonia variant manifest only with cold

**Features**

**Signs**
- Weakness with myotonia
  - Involving pharyngeal muscles as well as limbs and face
- Cataracts
- Frontal balding
- Variable intellectual disability, somnolence
- Cardiomyopathy, conduction abnormalities
- Testicular failure
- Reduced gastric motility

**Investigations**
- ECG increased PR interval, atrial flutter, other arrhythmias
- RFT marked reduction in maximal expiratory pressure, small reduction in VC

**Treatment**
- Myotonia can be treated with phenytoin, but is not usually a problem
- Atrophy is not treatable

**Anaesthetic considerations**
- Prolonged contraction in response to depolarizing relaxants
  - Suxamethonium absolutely contraindicated
  - Also triggered by propranolol, clofibrate, $K^+$
- Prolonged contraction with shivering
  - Aim to maintain normothermia
  - Relative contraindication to volatiles
- Myotonia provoked by mechanical stimulus and diathermy
- Increased risk of apnoea with sedative drugs
- Myotonia antagonized only by intramuscular local anaesthetic
- Some relief with quinine, procainamide or phenytoin
- Non-depolarizing relaxants are effective at normal doses
- Reversal appears to be safe despite theoretical risk of myotonia
- Intravenous regional anaesthesia should be effective

**Muscular dystrophy**

**Aetiology**

**Familial**
- Duchenne (X-linked), Limb girdle (AR), Facioscapulohumeral (AD)
- Atrophy of skeletal muscle with fatty infiltration and fibrosis
Features

Signs
- Progressive limb weakness
- Diaphragm function relatively preserved
- Late cardiomyopathy, arrhythmias, mitral valve prolapse
- Kyphoscoliosis with respiratory compromise common

Investigations
- CK typically elevated
- ECG abnormalities (RSR' in V1, deep Q in lateral V leads, arrhythmias)
- RFT VC<30% predicted indicated high risk with GA

Anaesthetic considerations
- Progressive disease so earlier operation is preferable
- Increased incidence of malignant hyperthermia
- Suxamethonium contraindicated due to potassium release
- Decreased margin of safety with non-depolarizing relaxants
- Gastroparesis reported pre- and post-operatively
- Increased risk of aspiration with bulbar weakness
- Avoid tachycardia as increased risk of arrhythmia
- Rhabdomyolysis and renal failure

Malignant hyperthermia in Complications 3.A.4

Neuroleptic malignant syndrome