# Contents

# By Disease

# Epilepsy

## Preoperative

= recurrent, chaotic brain dysfunction leading to behavioural disorder -> convulsions

### HISTORY

- make provision if oral anticonvulsant medications cannot be given
- quantify severity of seizure disorder
- full drug history (including specific timing)
- effect on lifestyle (drivers licence)

- associated co-morbid diseases/ syndromes

### EXAMINATION

- neurological
- as appropriate

### INVESTIGATIONS

- electrolytes
- glucose
- anti-convulsant plasma drug

### MANAGEMENT

- maintain GI function so anti-convulsent medications can be continued
- discuss with neurologist if concerned
- avoid prolonged fasting
- benzodiazepines for premedication
- can convert oral into other routes:
    - carbamazepine (PR),
    - phentobarbital (IM or IV),
    - phenytoin (IV),
    - fosphenytoin (IM),
    - sodium valproate (IV) - IV dose = PO dose.
    - clonazepam (IV)
- day case is fine if well controlled epilepsy ie seizure free for 1 yr or nocturnal only
- warn of risk of peri-op seizure $\Longrightarrow$ affected ability to drive

## Intraoperative

- can consider thiopentone for induction (powerfully anti-convulsants)
- use a NDNMBD without a steroid nucleus (atracurium):
    ↳ many anti-convulsant drugs lead to rapid metabolism of vecuronium and rocuronium
- avoid hypocarbia -> as it decreases seizure threshold
- use RA
- use anti-emetics that decrease risk of dystonic reactions (dex, cyclizine, ondansetron)
- avoid:
    - ketamine - cerebral excitatory effects
    - etomidate - assoc with myoclonus although not strictly pro-epileptic
    - antiemetics: high incidence of dystonia prochlorperazine, metoclopramide, droperidol
    - enflurane - abnormal EEG esp with ↓$CO_2$
- propofol:
    - assoc with abnormal movements during induction & emergence
    - unlikely to represent true seizure - normal EEGs seen during
    - TIVA:

- profound suppression of abnormal EEG during infusion
- reported effective in status
- epileptic pts may be prone to seizures during rapid emergence from propofol anaesthesia

▸ ∴ caution advised in administration of propofol to epileptics with drivers licenses
↳ co-induce with midaz - also helps emergence

## Postoperative

- careful documentation of epileptiform activity peri-op:
  - ▸ post op shivering
  - ▸ post op dystonic movements

  ↳ do not mis label as epilepsy as sig legal ramifications
- monitor for post operative seizures
  - ▸ any seizure means cannot drive for 6 months
  - ▸ should be warned pre-op
- keep anti-convulsants going

# Cerebrovascular Disease

## Preoperative

= cerebral damage from a vasculopathy (infarction or haemorrhage)
- classified by area:
  - ▸ global cerebral dysfunction = Multi-infarct dementia
  - ▸ focal disorder = TIA or stroke

### TIA

= focal neurological deficit that must fully resolves within 24 hours
- embolism of platelet and fibrin aggregates released from areas of atherosclerotic plaque
- high risk of stroke ~5%/yr with mortality of 30%
- indication for CUSS +/- angio:
  - ▸ recurrent TIA
  - ▸ TIA despite aspirin
- All should be risk stratified ABCD2 criteria (to give 2 day stroke risk):
  - ▸ Age >60
  - ▸ Bp >140/90
  - ▸ Clinical -
    - speech = 1 point
    - limb mm weakness = 2 point
  - ▸ Duration -
    - 10-60 min = 1 point
    - >60min = 2 point
  - ▸ Diabetes

  ↳ max 7 points :
    - 1-3 = 1% risk
    - 4-5 = 4% risk
    - 6-7 = 7% risk

### HISTORY

- sudden onset: dysphasia, weakness, numbness, collapse, in-coordination
- risk factors: IHD, HT, CVA, DM, smoker, cholesterol, age, obesity

### EXAMINATION

- neurological to find lesion
- cardiovascular

### INVESTIGATION
- U+E
- FBC
- cholesterol
- BSL
- ESR
- CXR (>60 or smoker)
- ECG
- CT Head
- carotid dopplers

### MANAGEMENT
- aspirin 300mg daily
- Dipyridamole
- omeprazole 20mg daily
- simvastain 40mg nocte
- anti-hypertensive if BP >140/90
- stop smoking
- CEA if > 70% stenosis or symptomatic TIA or CVA
- may need to manage peri-operative anti-coagulation (may be on warfarin)
- ask about vertebrobasillar insufficiency precipitated by postural changes and neck position
- continue anti-hypertensive medications

### ANAESTHESIA
- delay elective surgery if stoke/TIA:
  - for at least 6 weeks -> 20 fold increase in post-operative stroke
  - better = delay for 3-6 months
- careful documentation of prop neuro deficit

## Intraoperative
- no sux if has the onset of a hemiplegia within 9 months
- thromboprophylaxis - stop aspirin only in high surgical risk bleeding ie tonsils/neurosurg
- maintain normotension - pronounced CVS instability common
- large bore access
- invasive monitoring
- neutral neck position
- gentle induction
- cover for hypertensive response to intubation (remi, alfentanil and beta-blocker)
- avoid hyperventilation

## Post-operative
- examine early for new neurology

# Parkinsonism

## Preoperative
- Parkinsonism = imbalance of mutually antagonistic dopaminergic & cholinergic systems in basal ganglia
- pigmented cells in substantia nigra are lost -> reduced dopaminergic activity
- Causes of Parkinsonism:
  - Parkinson's disease - unknown cause
  - Drugs - esp neuroleptic agents
  - post traumatic
  - post encephalitic

## HISTORY
- Tremor
- Bradykinesia
- Rigidity
- Postural instability
- Micrographia
- Unsteadiness on feet
- Difficulty turning once walking
- dysphagia

## EXAMINATION
- postural hypotension
- neurological examination - extra pyramidal signs

## INVESTIGATIONS
- clinical diagnosis
- ECG - PVCs common and not significant
- lying & standing bps/tilt table study: autonomic dysfunction or effect of drugs (beware of position changes)
- PFTs - may be compromised because of bradykinesia and muscle rigidity.

## MANAGEMENT
- manage with help of PD physician
- excessive salivation + dysphagia -> risk of aspiration
- GORD is common
- urinary retention common

## DRUGS
- Aim = increase dopaminergic tone & decrease cholinergic tone within CNS
- Treatment limited by SEs: nausea & confusion esp in elderly
- 20% unresponsive to therapy

**Dopaminergic Drugs**

L-dopa
- inactive form of dopamine -> decarboxylated to dopamine in the brain.
- best for bradykinesia & rigidity (limited action on tremor)
- usually administered with decarboxylase inhibitors (benserazide, carbidopa) that do not cross the BBB -> reduces peripheral conversion into dopamine

MAO-I's
- eg selegiline = MAO B inhibitor
- less interactions than non-specific MAO-Is
- reduce central breakdown of dopamine
- must avoid:
    - pethidine ⟹ HTN++
    - SSRI's & TCA's ⟹ CNS excitability

Ergot derivatives
- bromocriptine, cabergoline, lisuride and pergolide -> are all dopamine receptor agonists
- used as adjuntcs to L DOPA if intolerant of side effects

Entacapone
-> adjuvant to those on L-dopa
- results in
    - ↓dose L-dopa
    - ↑duration of action)

- other agents = ropinirole, pramipexole, amantadine, apomorphine, tolcapone.

**Anti-cholinergic Drugs**
- indicated when symptoms are mild

- good for:
  - tremor
  - +/- rigidity
  - +/- sialorrhoea

  (bradykinesia not affected)
- useful in drug induced parkinsonism
- agents = benztropine, procyclidine, benzhexol & orphenadine
- parenteral formulations for procyclidine & benzatropine (useful for acute drug induced dystonias)

**Surgical procedures**
- usually performed in awake patient with stereotatic probes
- techniques include:
  - thalamotomy - tremor & rigidity
  - pallidotomy - rigidity & bradykinesia
  - deep brain implantable devices (avoid diathermy - use bipolar. check device post op)

**Drugs Interactions**
Pethidine -> hypertensive crisis & rigidity (similar to MH)
Synthetic opioids -> rigidity in high doses
Inhalational agents -> can produce arrhythmias
Anti-emetics (anti-dopaminergics) -> exacerbate extra-pyramidal symptoms (∴ use ondansetron)
Anti-psychotics (anti-dopinergics) -> exacerbate symptoms (∴ use atypicals ie risperidone, clozapine)
TCA's -> induce arrhythmias
SSRI's -> hypertensive crises & cerebral excitation
Anti-hypertensives -> postural hypotension (esp clonidine & reserpine)

### ANAESTHESIA
- continuation of normal anti-parkinson drug regime is of upmost importance - symptoms develop from 3hr post missed dose
- give meds at anytime preop & resume asap post op
- if missed:
  - NG tube intra-op ⟹ give crushed meds
  - apomorphine only parenteral formulation:
    - dosing conversion from oral meds unclear without previous trial
    - if predicted NBM post op then should have neurology plan for periop anti-parkinsons meds
    - if problems getting oral dose in then use escalating doses subcut:
      - 1.5mg wait 30mins
      - 3mg then wait in 40min periods
      - rpt 3mg doses until get response
      - usual dose range 3-30mg
  - rotigotine - available as patch

# Intra-operative
- continue medications up to time of operation
- can use an anti-sialogogue if really needed- glyco
- RSI - may be needed (Gi dysfunction, excessive saliva)
- normothermia to avoid shivering
- no one anaesthesia technique superior
- IV morphine for pain
- PCA may be difficult for them to operate

# Post-operative
- may need post-operative care in HDU
- physio
- N/G tube placement may be needed to allow oral feeding and meds
- anti-emetics ->
  - 1st choice = domperidone 10-20mg Q4-6hrly PO should be first choice

↳ it doesn't cross BBB and ∴ doesn't produce extra-pyramidal side effects
- 2nd line:
  - ondansetron
  - cyclizine

# Spinal Cord Lesions

## Preoperative

3 phases:

1. INITIAL – (minutes)
   - extreme hypertension and arrhythmias -> MI, LVF, APO
2. SPINAL SHOCK – (immediately to 3 days - 8 weeks):
   - features = hypotension, bradycardia and loss of sympathetic tone
   - parasympathetic tone remains ∴ risk of extreme brady/aystole esp on intubation/tracheal suction
   - common after lesions above T7
   - loss of muscle tone and reflexes below lesion
   - paralytic ileus common
3. REFLEX PHASE – up to 9months
   - neuronal re-wiring with re-establishment of sympathetic tone + muscle tone and reflexes.

### HISTORY

- injury: level, how, when
- ongoing symptoms?
- complications?
- autonomic dysreflexia?
- stabilization surgery? - neuraxial diff. intubation difficult
- complete or incomplete lesion - complete ⟹ autonomic dysreflexia greater

- functional ability
- help @ home
- medications
- allergies (latex)
- chart review for previous anaesthetics

### Complications

- CVS: reduction in blood volume (20%), postural hypotension
- RESP:
  - initial level may get worse with oedema before improvement later
  - based on level:
    - >C3 = apnoea,
    - C3-C5 = possible diaphragmatic sparing,
    - below C5 = phrenic sparing, intercostals paralysis, recruitment of accessory muscles over 6 months, decreased ability to cough and FEV1,
  - FVC better in horizontal position c/o diaphragmatic excursion,
  - bronchial hypersecretion
  - recurrent pneumonia's
- METABOLIC: poor thermoregulation, can't shiver, decreased control of peripheral vasculature
- MUSCULOSKELETAL:
  - muscle spasms, spasticity - baclofen & diazepam
  - reduced bone density -> #'s, heterotopic calcification
- SKIN: pressure sores, difficult IV access
- HAEMATOLOGICAL: anaemia, increased risk of VTE (some warfarinised after d5)

- GI: delayed gastric emptying
- GU: recurrent UTI's
- PAIN: chronic pain often a problem

### EXAMINATION
- tracheostomy gives you a clue of the level of lesion (ie. high)
- formal neurological assessment
- LMN signs @ level of lesion
- UMN signs below lesion
- sensory loss below lesion
- airway and precordium assessment

### INVESTIGATIONS
- BLDs
    - FBC: anaemia
    - U+E: renal impairment
    - LFT's
- PFTs
- CXR:
- spine xrays, CT and MRI
- ECG: for high thoracic lesions
- urine (microscopy and culture)

### MANAGEMENT
- avoid suxamethonium from 2 days -> 9months or 2 years
    ↳ denervated motor end plate ⟹ spread of MEP across entire mm ⟹ if depolarised ⟹ ↑↑K eflux ⟹ asystole
- supportive care until haemodynamics settle
- MDT input
- gabapentin for neuropathic pain
- screen for depression
- if requires minor operations assess whether anaesthesia required

### ANAESTHESIA REQUIRED?
- if planned op would require GA in normal pt ⟹ then needed in cord patient (even if complete)
- minor periph surgery with LA likely fine below complete surgery
    ↳ risk of mm spasm remains
- urology procedure with lesion above T5 ⟹ high risk of autonomic dysreflexia

# Intraoperative
### GENERAL ANAESTHESIA
- Spinal shock phase:
    - only life threatening surgeries
    - prior to intubation: atropine 300 or glyco 200
    - Cx spine care
    - preload with IVF +/- CVL
- Reflex phase:
    - as history above - impt
    - standard monitoring
    - fluid load
    - high C spine lesions -> IPPV
    - no apparent ↑risk of aspiration despite GORD
    - vasopressors as required
    - NMB after intubation not required - unless mm spasm
    - hypothermia cares
    - position cares

### REGIONAL ANAESTHESIA (central neuraxial)

- advantages: prevents autonomic dysreflexia, unlikely to cause cardiovascular instability c/o sympathetic tone already decreased, avoids GA, no benefit from intrathecal opioid
- disadvantages: may be difficult, difficulty in testing success and level of block (loss of hypertonicity, disappearance of lower limb reflexes, loss of sweating)

# Postoperative

- nurse supine + slight head up -> improves ventilation
- monitor temperature
- monitor for dysreflexia

# Important Related Issues

## Autonomic Dysreflexia

= massive disordered autonomic response to stimulation below level of the lesion
- loss of descending inhibitory control on regenerating presynaptic fibres
- rare with lesions below T7
- ↑common the higher the lesion
- occurs within 3 weeks of lesion, less likely after 9 months

### TRIGGERS

GU: bladder distension, UTI
OB: labour, cervical dilation
GI: constipation, bowel obstruction, acute abdomen
MUSCU:#
SKIN: minor trauma, cutaneous infection

### CLINICAL

- hypertension (commonest)
- headache
- flushing
- pallor
- nausea
- anxiety
- sweating
- bradycardia
- penile erection
- seizures
- APO
- coma

### MANAGEMENT

- treat cause! :
    - unrevealed trauma or infection
    - catheterise
    - check for faecal impaction
- drug Rx:
    - phentolamine 2-10mg IV
    - GTN: transdermal -> SL -> IV
    - clonidine 150-300mcg IV (if HTN & spasticity)
    - esmolol 10mg IV (only if also tachycardic)

# Obstetric Anaesthesia

## Effect of Pregnancy on Spinal Cord Injury

- CVS: exaggerated postural hypotension and response to caval compression
- RESP: reduced respiratory reserve, increased risk of respiratory failure, increased O2 demand
- HAEM: increased anaemia and VTE

- SEPSIS: increased risk of infection (pressure areas and urinary)
- LABOUR:
  - increased risk of premature labour,
  - increased risk of autonomic dysreflexia with lesions >T5 (may be the first sign of labour)

### MANAGEMENT
- review early
- MDT input
- plan
- discuss use of neuraxial technique
  - epidural:
    - generally possible
    - do early = most effective way of preventing autonomic dysreflexia
    - load with fluid, pressors ready
    - leave catheter in for 48 hours post delivery ⟹ prevent risk of autonomic dysreflexia which remains for 48hr post partum
    - failure to get epidural ⟹ prophylactic drug Rx to prevent autonomic dysreflexia
  - spinal - generally possible even if spinal instrumentation
- GA Caesar risks as GA above

# Myasthenia Gravis

## Preoperative
- autoimmune disruption of post-synaptic acetylcholine receptors @ NMJ
- up to 80% of functional receptors loss
- typically young woman or older men
- may have mediastinal mass/thymus hyperplasia

### HISTORY
- mild ptosis -> bulbar palsy and respiratory failure
- severity of MG (duration, functional capacity, doses of medications)
  - ↳ if isolated chronic ocular symptoms unlikely to have progressive disease
  - ↳ missed dose - ? significant effect
- Rx normally with:
  - mild: oral anticholinesterase meds +/- steroids
  - severe: immunosuppressive agents, plasmapheresis or immunoglobulin infusion
- bulbar symptoms (requirement for post-operative ventilation)
- views on epidural pain relief vs PCA vs rectus sheath catheters +/- PCA
- significant other cardio/respiratory disease –
  - heart failure,
  - COPD,
  - restrictive lung disease,
  - recurrent aspiration pneumonia

### EXAMINATION
- swallow
- functional capacity
- effectiveness of cough
- habitus
- airway assessment
- focused RESP and CVS examination
- evidence of proximal myopathy and strength

### INVESTIGATION
- spirometry
- PEFR
- others as indicated

- previous anaesthetics (easy of intubation and ventilation)
- CXR

### MANAGEMENT
- discussion with neurology about patient degree of optimisation required for surgery
- plan for post-operative ventilation if required (ICU)
- plan for analgesic technique as indicated
- introduction to physiotherapy
- GORD/aspiration prophylaxis: H2 antagonists, Na+ citrate, metoclopramide, appropriate starvation
- keep anti-cholinesterase and other drugs going if practical

# Intraoperative
- continue all drugs right up to induction…theoretical inhibition of NMB never shown to be a problem
- avoid premeds
- consider regional ⟹ ↓post op opioids, & risk of resp depression
- avoid muscle relaxation if possible (may not be given major abdominal surgery):
    - atracurium - predictable offset
    - roc & suggamadex ideal
- if RSI needed: sux 1.5mg/kg
- avoid ester LAs eg prilocaine. Bupiv & ropiv are safe
- keep warm
- use PNS
- intubation
- controlled ventilation
- volatile maintenance vs TIVA depending on NMJ control
- good analgesia
- intraoperative hydrocortisone/dexamethasone if indicated
- avoid reversal if possible
    - (increased risk of cholinergic crisis) -> if need to reverse use standard doses
- extubate once wide awake and obey commands (able to lift head off pillow for 5 seconds)
- N/G tube may be required so can have regular medication

### PeriOp Anit-Cholinesterase Management
- oral 30mg pyridostigmine = 1mg parenteral neostigmine
- neo reversal:
    - if at least 1 twitch TOF (no twitches = no reversal)
    - 2.5mg bolus
    - give 1mg bolus every 2-3min to max equivalent oral pydridostigmine dose (1:30)
        ↳ eg pyridostigmine dose = 120mg 4hrly then give max 4mg neostigmine
    ↳ better to avoid all this and use roc + sugammadex

### Drugs

| Drug | Interaction | Notes |
|---|---|---|
| NMBs | - ↑sensitivity | - Use short acting & 10% norm dose eg atracurium,<br>- monitor PNS |
| Sux | - Resistance to depolarisation<br>- Delayed onset of action | - No reported ill effects at 1.5mg/kg<br>- Delayed recovery if on plasmapheresis & anticholinesterases<br>- Follow with NDMBs only when full recovery achieved |
| Volatiles | - All ↓NMJ transmission by up to 50% | - May allow avoidance of need for NMBs |
| TIVA | - No effect on NMJ transmission | - Useful if NMJ function precarious |
| LAs | - Prolonged action<br>- ↑toxicity in ester linked agents (if on plasmapheresis & anticholinesterase)<br>- Exacerbation of MG reported | - Use min doses<br>- monitor resp function |
| Esterases eliminated drugs | - Prolonged effect<br>- ↑toxicity (plasmapheresis/ anticholinesterases) | eg Sux, remi, miv, ester linked LAs, esmolol |
| ABx | - NMB effects may become imp | - Avoid aminoglycosides (gent), erythrmoycin, cipro |
| Misc | - Other drugs effect NMJ transmission:procainmide, ßblocker (esp propanolol), phenytoin, Mg | |
| Pryridostigmine | - Adult: 30-120mg 4 to 6hrly (max daily dose 720mg) | - Useful duration of action. Less potent & slower onset than neostigmine |
| Neostigmine (IV) | - Adult: 1-2.5mg 2 to 4hrly (max daily 5-20mg | - IV ⟹ ↑side effects with ↓duration of action<br>- if used IV then coadminster anticholinergic agents (glyco) |
| Neostigmine (oral) | - Adult: 15-30mg (up to 2hrly) (max daily 75-300mg) | - ↑Gi effects than pyridostigmine<br>- useful if parenteral therapy required |
| Edrophonium | Adult: 2mg IV injection then 30s later 8mg if no adverse reaction | - Used in diagnosis of myasthenia & differentiation of myasthenic & cholinergic crises |

# Postoperative

- ICU
- physio
- good analgesia
- restart oral medications as soon as possible (may need IV neostigmine (30mg pyridostigmine : 1mg neostigmine) or hydrocortisone if not able to tolerate PO medications)
- incentive spirometry
- have a low threshold for starting antibiotics if develops clinical indication for pneumonia or LRTI

### Predictors of Post operative Ventilation:

- major body cavity surgery
- duration of disease (> 6 years)
- history of chronic respiratory disease
- dose requirements of >750mg/day
- preoperative VC of <3L

# Thymectomy

- consensus now suports thymectomy in all adults with generalised MG
- remission rates high with 96% gain benefit in symptoms
- best done if normal or hyperplastic thymus
- Approachs =
    - trans-sternal (commonest)

    - thoracoscopic - unproven but ↑ing use
    - (transcerivcal less satisfactory)
- Anaesthetic management as for norm MG patient
- <10% require ventilaiton for >3hr post op
- Most will require some mm relxation for op
- epidural or PCA best analgesia

# Eaton-Lambert Syndrome

- = myasthenic syndrome
- = proximal mm weakness assoc with cancer - most common = lung small cell ca
- thought due to ↓release of Ach fro pre-synaptic membrane
- is not reversed by anticholinesterase meds
- mm weakness improves with exercise
- assoc symptoms (dysautonomia):
    - dry mouth
    - impaired accomodation
    - urinary hesitance
    - constipation
- Pts sensitive to all NMBs (ie ↑ed potency of NMBs incl sux) but use ↓doses
- high index of suspicion for all pts with suspected lung Ca

# Multiple Sclerosis

## Preoperative

= acquired demyelinating plaques in CNS
- early adulthood
- 30% benign course
- 5% rapid deterioration
- Europe, NZ & North America

### CLINICAL

- deterioration & remitting course

- visual disturbances
- nystagmus
- weakness
- paralysis
- bulbar palsy
- respiratory muscle failure
- sensitivity to heat - 0.5deg C may cause marked deterioration symptoms

- document neurological lesions (so you can compare post operatively)

### MANAGEMENT

- steroids & interferon decrease symptoms
- baclofen & dantrolene used for painful mm spasms
- bulbar symptoms ≈ peri-op airway problems

## Intraoperative

- Regional anaesthesia:
    - does not affect symptoms
    - but medico-legally wise to avoid
    - document neurology & warn of symptom exacerbation
- Neuraxial :
    - associated with recurrence of symptoms (reduced by using weaker concentrations of LA + opioid)

    - obstetrics:
        - epidural for labour not contraindicated (use low dose LA)
        - spinal acceptable & widely used. low dose LA
- GA does not affect course of illness
    - avoid sux -> marked increase in K+
- normal response to NDNMBD (reduce dose)
- measure haemodynamics carefully -> marked autonomic instability
- aggressive normothermia:
    - avoid pyrexia -> worsens symptoms - use antipyrexics ++
    - avoid ↓temp ⟹ delay recovery

## Postoperative

- bulbar palsy & respiratory function may affect -> increased risk of aspiration

# Guillain-Barre Syndrome

## Preoperative

= immune mediated progressive demyelinated disorder characterised by acute or subacute proximal skeletal muscle paralysis
- distal to proximal weakness
- 85% achieve full recovery although perhaps after months
- rapidity of symptoms onset ≈ more likely progression to resp failure
- bulbar palsy (diff swallow or phonation, or unable to cough) impending sign of need for intubation

? viral aetiology

### HISTORY

- limb paresthesia
- back pain
- SOB
- dysphagia

### EXAMINATION

- loss of reflexes
- distal -> proximal loss of power
- bulbar dysfunction
- autonomic dysfunction

### MANAGEMENT

- ⅓ need ventilatory support if develops respiratory failure
- steroid use controversial
- plasmapheresis
- immunoglobulin

## Intraoperative

- IV access
- fluid load
- pressors ready
- autonomic dysfunction ⟹
    - hypotension on induction
    - may have dramatic tachycardia to surgical stimuli
    - may have paradoxical brady with atropine
- no sux
    - c/o extreme hyperkalaemia
    - can persist for months post recovery
- cautious use of NDNMBD - may not be required

- epidural analgesia useful - is used to help with distressing paresthesia in some pts

## Postoperative

- epidural analgesia can be effective for distressing Paresthesia
- monitor

# Motor Neuron Disease (ALS)

(amyotrophic lateral sclerosis)
- pts mentally normal until terminal resp failure ⟹ ethical problems of long term ventilation

## Preoperative

= degeneration of upper and lower motor neurons in the spinal cord

### HISTORY

- weakness
- starts in proximal muscles of hand -> progressive to axial and bulbar weakness

### EXAMINATION

- muscular atrophy
- fasciculation
- weakness
- respiratory failure
- autonomic dysfunction

### INVESTIGATION

- EMG

### MANAGEMENT

- symptomatic

## Intraoperative

- bulbar dysfunction -> ETT (many with have trachy)
- IPPV
- autonomic dysfunction ⟹ hypotension on
    - induction
    - start of IPPV
    - position changes
    - ↳ careful CVS monitoring
- wide bore IV access
- fluids
- pressors ready
- avoid sux
- use NDNMBD @ lower dose

## Postoperative

- may need respiratory support

# Dystrophia Myotonica

= myotonic dystrophy, myotonia atrophica

## Preoperative

- = persistent contraction of skeletal muscles following stimulation

- progressive deterioration/atrophy of skeletal, cardiac and smooth muscle ⟹ decreased cardiorespiratory function

### HISTORY
- autosomal dominant
- 20-30yrs
- prefrontal balding
- cataracts
- weakness
- bulbar palsy
- respiratory reserve
- mental deterioration after 2nd decade
- death in 5th - 6th decade

- PMHX of endocrine dysfunction: DM, hypothyroidism, adrenal insufficiency and gonadal atrophy

### EXAMINATION
- atrophy of facial, sternomastoid and peripheral muscles
- progressive atrophy of skeletal, cardiac and smooth muscles
- respiratory failure from weakness & bulbar signs
- cardiomyopathy signs
- MVP (20%)

### INVESTIGATIONS
- U+E: and glucose – endocrine issues
- ECG: degeneration of cardiac conduction system -> dysrhythmias and AV block
- CXR:
- ABG:
- spirometry:
- ECHO: structural defects: MV prolapse in ~20%

### MANAGEMENT
- supportive
- anti-myotonic medications (procainamide, phenytoin, quinine and mexileline)
- GORD and delayed gastric emptying cares - antacids
- pregnancy may aggravate disease -> C/S may be required c/o inadequate uterine contraction

## Intraoperative
- sux -> prolonged muscle contraction and K+ release (avoid)
- use of NDNMBD:
  - always with PNS (PNS may cause mm contraction/tetany)
  - use short acting as need to avoid neostigmine may ⟹ prolonged contraction
  - best to avoid or use roc/sugammadex
- invasive monitoring: profound risk of CVS depression
- balanced anaesthetic
- ETT
- RA doesn't prevent muscular contractions
- muscular spasms: LA infiltration, quinine, phenytoin
- hypothermia cares (avoid shivering as can provoke myotonia)

## Postoperative
- HDU
- respiratory monitoring
- physio
- analgesia (ideally RA)

# Muscular Dystrophy

= range of congenital disorders characterised by progressive weakness of affected muscle groups

1. X linked (Duchenne's, Becker's)
2. Autosomal recessive (limb-girdle, childhood, congenital)
3. Autosomal dominant (faciocapulohumeral, oculophargeal)

Duchenne = commonest & most severe form - see Paeds

# Duchenne Muscular Dystrophy

## Preoperative

- most common and most severe form of muscular dystrophy

### CLINICAL

- sex-linked (males)
- 2-5 years
- muscular weakness
- wheelchair bound by 12 years
- death by 25 years (cardiac failure or pneumonia)

CVS: myocardial degeneration -> heart failure, MV prolapse
RESP: respiratory muscle weakness, restrictive ventilation pattern, inadequate cough, eventual respiratory failure and infection
HAEM: vascular muscle dysfunction -> increased bleeding
MUSCULOSK: progressive, severe kyphoscoliosis

### INVESTIGATION

- CK - tracts disease level: elevated early on then ↓ to below normal late
- CXR
- spirometry
- ECHO - mandatory if pt wheelchair bound

### MANAGEMENT

- liase with paediatrician
- GORD prophylaxis

## Intraoperative

- antisialogogue & antacids/prokinetic
- balanced induction
- TIVA - avoids risk of anaesthetic induce rhabdomyolysis with volatiles
- avoid sux
- NDNMBD safe (reduce dose + PNS)
- keep ETT if concerned about bulbar dysfunction
- caudal great

## Postoperative

- standard care

# Malignant Hyperthermia

= pharmacogenetic disease of skeletal muscle induced by exposure to certain anaesthetic agents

-> excess Ca2+ release during muscle contraction -> increased muscle metabolism + heat production.
- prolonged and intensified interaction between actin and myosin.
    ↳ error anywhere in pathway but most likely site is DHPR & RyR receptor
- 200 mutations identified in RYR1 (19q) ⟹ 29 proven causality
- enhanced anaerobic metabolism -> lactic acidosis -> accumulation of intramitochondrial calcium -> deconjugation of oxidative phosphorylation -> cytolysis.

- incidence 1:5,000 -> 1:65,000 anaesthetics (suspected)
- mutation in the gene coding for the **ryanodine receptor**.
- autosomal dominant
- gene on chromosome 19
- thymidine instead of cytosine
- produces a cysteine for arginine substitution at position 615 of the receptor.

- pig model = soft exudative pig disorder
- mortality rates fallen markedly 80% to 2-3%
- previously uneventful anaesthetic is not indicative of safety

## TRIGGERS
- stress (in pigs & humans)
- all volatile agents (except N2O)
- sux

-> these all either enhance Ca2+ influx or slowing its efflux.
- some may have tolerated the same agents previously
- rare in barbiturate-N2O-opiate-tranquiliser-non-depolarising muscle relaxant anaesthesia.

- sux potent trigger (first exposure)
- volatiles (median exposure till fulminant -> 3)

## CLINICAL
- in lower north island trigger names = Harvey, Harwere & Cook
- history of Central Core Disease

(1) increased ETCO2
(2) tachycardia
(3) tachypnoea
(4) masseter spasm:
    - = spasm after sux impeding intubation for ~2min
    - 30% with MMS alone go on to be MH susceptible
    - if poss abandon surgery, if not convert to MH free anaesthetic:
        - TIVA, charcoal filters, high flow O2
        - allow ~15min to ensure pt stabilised
(5) muscle rigidity
(6) temp increase (late) - 1 C\15min

- varied presentation:
    - intra-op & or 4\24 post op
    - or post op 2-3days
    - acute florid or indulent chronic presentation
- tachyarrhythmias
- difficulty ventilation
- hypertension
- sweating
- DIC
- hyperkalaemia
- cardiac arrest
(fever developing post anaesthetic is not indicative of MH)

## INVESTIGATIONS
- PaCO2 >60mmHg
- PvCO2 >90mmHg and increasing
- BE -5 and falling
- mixed met/resp acidosis
- intial & 24hr CK >50,000 IU/L
- K+ increases
- Na+ increases
- 1st void urine for myoglobinuria

## ACUTE MANAGEMENT
- call for help
- convert to TIVA, stop volatile
- maintain anaesthesia with hypnotics and opioids.
- muscle relaxation with NDNMBD - only if required
- terminate surgery
- hyperventilate
- 100% O2
- cool (N/S stomach lavage)
- maintain urine output
- inotropes as needed
- HCO3 2-4mEq/kg
- Dantrolene 2.5mg/kg every 5min (total dose 10mg/kg/day - continuous infusion **or** treat deterioration (25%))
- cardiac arrhythmias -> beta blockers & lignocaine.
- high K+ -> glucose-insulin & frusemide
- watch for DIC
- CK Q6hrly

## PROGNOSIS
- mortality without dantrolene = 70%
- mortality with dantrolene = 5%

## LONG-TERM MANAGEMENT
- referral to an MH centre
- warn patient and family of impending consequences
- if uncertain about diagnosis -> must screen
- IVCT = in vitro contracture test = gold standard:
    - open invasive procedure either under regional or trigger free anaesthetic
    - vastus medialis taken at Mh centre - exposed to halothane & caffeine
    - if positive search for genetic mutations
        - if found then can screen family for that mutation.
        - if none found: then rest of family must have IVCT
- medic alert and appropriate documentation

## PROPHYLACTIC MANAGEMENT
- take history
- decrease anxiety with midazolam
- machine:
    - use vapour free machine if able
    - otherwise:
        - remove vapourisers, flush with O2 @ 10-15 L\min for 20min
        - new circuit and airway devices
- TIVA anaesthetic with NDNMBs only
- all LAs safe
- ETCO2 monitoring
- nasal temp probe - establish pre-exposure baseline temp
- dantrolene available

### SUSPECTED PREV ANAESTHETIC HISTORY
- unexplained/expected cardiac arrest/death ⟹ 50% risk of MH
- Hx of post-op myoglobinuria (red/black urine)
- renal failure in otherwise healthy pt
- post op fever - poor association but cannot be excluded

### OBSTETRIC PATIENTS
- baby = 50% chance of having MH
- mother MH susceptible:
    - planned delivery with early anaesthetic advice
    - anticipate airway problems -> AFOI
    - roc & TIVA ideal
    - RA safe and preferred
    - MH safe drugs
    - uterine drugs fine
- father MH:
    - sux doesn't cross the placenta so baby is fine
    - volatiles only once baby delivered

## Associated Diseases
- central core disease =
    - non progressive inherited condition
    - periph mm weakness & cardiac problems
    - = only condition known to be assoc with MH
    - treat as MH susceptible
- Heatstroke & King-Denborough syndrome - controversial
- Neuroleptic malignant syndrome - controversial but v. unlikely not related
- SIDS - not associated