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By Disease

Down's Syndrome

Preoperative

- = genetic disorder (Trisomy 21)
- commonest genetic abnormality
- 1.6:1000

CLINICALLY

- characteristic dysmorphic features: brachycephaly (short flattened cranium), flat nasal bridge, upward slopping palpebral fissures (space between upper & lower eyelids), small mouth and ears, single palmer crease, small stature
- AIRWAY: large tongue, crowding of midface structures, high arched palate, micrognathia, short broad neck, hypersalivation, subglottic and tracheal stenosis, TMJ laxity
- CNS: impaired global development, epilepsy, hypotonia, squint, cataracts, deafness
- CVS: VSD, Eisenmenger's syndrome (chronic L \Rightarrow R shunt \Rightarrow pHTN), ASD, PDA, TOF and PHT, rhythm abnormalities -> pacemaker, AR, MVP
- RESP: OSA, recurrent chest infections, tonsillar/adenoidal hypertrophy
- MUSCU: atlanto-axial instability 30% (frequently asymptomatic), cervical spondylosis
- GI: GORD, gastrointestinal atresia's, gall stones and Coeliac's disease
- NEURO: epilepsy 10%
- HAEM: immunodeficient, leukaemia's and lymphoma's,
- ENDO: obesity, hypothyroid 40%, DM
- SKIN: difficult IV access
- level of functioning @ home and community
- previous operations and anaesthetics
- medications
- allergies

INVESTIGATIONS

- ECG:
- ECHO: (if indicated)
- c-spine xrays - if neck pain, \downarrow AROM, neurology on neck positioning
- TFT's

MANAGEMENT

- may require a pre-medication (midazolam 0.5mg/kg PO, ketamine 5mg/kg PO)
- optimise if able

Intraoperative

- intubate (use a size smaller tube and insure has a leak)
- IPPV
- GORD cares
- avoid excessive movement of neck
- often need lower MAC
- consider RA + GA where possible

Postoperative

- get parents in room when waking (can help manage agitation)
- may have hypotonia (up to 75%) -> airway compromise
- HDU

- continuous SpO₂, supplemental O₂
- sedation score & RR
- minimise opioid consumption
- physiotherapy
- get mobile
- at risk of post-operative chest infections

Ehlers-Danlos Syndrome

Preoperative

- = group of defective collagen cross-linking disorders
- 6-10 major types depending on classification
 - ↳ each target diff locations:
 - type 1&2 = mostly skin - but also hypermobility
 - type 3 = hypermobility
 - type 4 = vascular (severe form)
 - ie vessels & organs prone to rupturing
 - (80% life threatening probs by age 40)
 - others v rare

HISTORY

- recurrent dislocations
- prolonged spontaneous bleeding
- rupture of cerebral or other vessels
- bowel perforation
- spontaneous pneumothorax

EXAMINATION

- extensible, fragile skin
- joint laxity
- hypermobile joints
- ocular abnormalities
- kyphoscoliosis

INVESTIGATIONS

- CXR: pneumothorax

MANAGEMENT

- supportive

Intraoperative

- careful positioning and padding
- lung protective ventilation
- gentle intubation (can cause severe tracheal bruising)

Postoperative

- low threshold for CXR

Marfan's Syndrome

Preoperative

- = connective tissue disorder
- autosomal dominant

CLINICALLY

GENERAL: tall, long thin fingers

AIRWAY: cervical spine/ligamentous abnormality, high arch palate, crowded teeth

RESP: emphysema, spontaneous pneumothorax, pectus excavatum, tracheomalacia, OSA, kyphoscoliosis

CVS: dilated ascending aorta, dissecting aneurysms, AR, MVP, MR, coronary thrombosis

CNS: cataracts, retinal detachment, lens dislocation, dural ectasia (60%) - causing back pain, abdo pain

MUSCULO: easy joint dislocation

MANAGEMENT

- supportive
- fix stuff as it breaks (valves, aorta's, scoliosis)
- antibiotic prophylaxis as indicated

Intraoperative

- may be a difficult intubation
- pressure cares
- lung protective ventilation
- neuraxial blocks no problems - risk of inadequate block if dural ectasia (consider MRI spine if symptoms and time)

Postoperative

- chest infection cares

Pierre Robin

Preoperative

- = syndrome characterised by mandibular hypoplasia, glosptosis, macroglossia, cleft palate and cardiac defects
- receding mandible fails to hold tongue forward -> obstruction
- 1/8500 live births

Intraoperative

- difficult laryngoscopy (tends to get better with age)
- gas inductions usually used
- can move into prone position
- LMA's have been used successfully
- can topicalise with finger - suckling then do either:
 - place LMA
 - awake laryngoscopy
- other adjuncts may be used: intubating stylets, BURP, external manipulation of larynx

Post-operative

- extubate awake and on side