Contents

Summary

	RA	Anky Spond	SLE	Systemic Sclerosis
Prevelance (%)	1	0.15	0.03	0.001
Airway & intubation	C spine unstable Cricoarytenoid arthritis TMJ arthritis	Cx kyphosis TMJ arthritis Occult #s		↓mouth opening
Resp	Fibrosing alveolitis pleural effusions nodules on CXR	Fixed chest wall apical fibrosis	LRTI PE pleuritis	Fibrosing alveolitis
CVS	IHD	AR (1%)	Raynauds IHD pericarditis endocarditis	Raynauds >90% HTN & pHTN myocardial fibrosis arrhythmias pericardial effusions
Neuro	Periph neuropathy Radiculopathy Myelopathy	Cauda equina (rare)	Periph neuropathy Psychosis Seizures	
Renal	mild CRF (common)		glomerulonephritis	CRF HTN renal crisis
GI	Drug gastritis	Drug induced reflux	NSAP Nausea mesenteric vasculitis	Reflux ++
Haem	↓Hb - drug & disease Felty's syndrome (splenomegaly & ↓WCC)	↓Hb - drug & disease	Antiphospholipid syndrome ↓Hb ↓platelets	
Neuraxial Blocks	Often difficult infection risk	Difficult- lat approach required ↑risk epidural haematoma	Check coags Infection risk	

Practical Points

Immunosupressed Patients

Risks

Short term

- Staph mediastinits
- CMV reactivation
- Toxoplasma gondii

Long term

- infection

    -> susceptible to intracellular bacteria (Listeria, non-thyphoid salmonella, mycobacteria)
    -> Legionella and Pneumocystis
    -> invasive aspergillosus
    -> Candida

- SCC of skin
- lymphoma (EBV associated)
- musculoskeletal problems (osteoporosis)
- skin fragility
- nephrotoxicity (cyclosporin)
- IHD from steroid
- hypertension (cyclosporin)
- epilepsy
- pancreatitis
- cholelithiasis
- immunosuppression-induced gingival hyperplasia
- higher incidence of PET

Anaesthetic Care

- strict asepsis on all invasive procedures incl IV cannulation
- avoid dexamethasone
- consider HPA axis suppression & need for stress dose steroids if been on prev
- consider drug interactions:
    - ciclosporin -
        - renal dysfunction & HTN
        - prolongs action of NDNMBs
        - CCBs $\Longrightarrow$ $\uparrow\uparrow$ciclosporin levels $\therefore$ avoid giving or changing dose if able peri-op
    - tacrolimus - renal dysfunction

By Disease

Rheumatoid Arthritis

Preoperative

- multisystem disease which can range from mild -> severe
- autoimmune disorder characterised by a bilateral, peripheral symmetrical inflammatory arthritis with a number of other systemic associations

ASSOCIATED PROBLEMS

- AIRWAY: decreased TMJ mobility -> poor mouth opening, limited head and neck extension, potential atlanto-axial subluxation (anterior, posterior, lateral and vertical), cricoi-aretynoid arthritis (neck pain, arm symptoms, neurological symptoms)
- RESPIRATORY: restrictive lung disease, kyphoscoliosis, pulmonary fibrosis, pneumonitis, pleural effusions, bronchiolitis obliterans
- CARDIOVASCULAR: amyloid infiltration of myocardium, restrictive pericarditis, conduction abnormalities, valve pathology, IHD, Raynauds
- RENAL: CRF
- MUSCULOSKELETAL: joint changes and decreased ROM and pain, manual dexterity
- ANALGESIA: chronic pain management (adjuncts like ketamine and gabapentin may be required)
- HAEMATOLOGICAL: anaemia may be multi-factorial (chronic disease, NSAID induced GI blood loss, drug induced marrow suppression)
- CNS: peripheral neuropathy
- EYES: dry and inflamed
- co-morbid conditions: IHD, PMR, obstructive lung disease, smoking, systemic vasculitis

MEDICATIONS

- DMARDS (gold, pencillamine, methotrexate, azathioprin) - should cont periop. No evidence ↑infection
- steroids - supplementation as required for HPA axis
- Others:
    - TNFα blockers (biologics) eg infliximab
    - Monoclonal antibodies & IL 6 eg rituximab
    - ↳ non consensus but suggestion of risk of severe injury. Some suggest stop for 2/52 perioperatively
- SYMPTOM CONTROL: analgesics, antacids

HISTORY

- number of years
- disease control
- functional assessment
- past and present treatment
- activities of daily living

EXAMINATION

- AIRWAY: neck movement, mouth opening,
- MUSCULOSKELETAL: swan neck and boutonniere deformities, z deformity of thumb, ulnar deviation and palmar subluxation @ wrist, muscle wasting in palmar aspect of hands, rheumatoid nodules, inspect, palapate and move joints, get them to undo their buttons and test power briefly
- CVS: vasculitis, AR, pericardial rub
- CNS: dry eyes, nodular scleritis, hoarseness
- RESP: creps or effusions
- ABDO: splenomegally

INVESTIGATIONS

- routine bloods: organ dysfunction, drug levels, anaemia, thrombocytopaenia
- c-spine xrays:
    - (major destruction, duration > 5yrs, symptoms or signs)

- role is controversial
- suggested mandatory if neuro signs/symptoms or persistent neck pain
  ↳ stabilisation surgery may be required pre surgery

1. **atlanto-axial subluxation**
   - (25% RA pts but only 25% of these have symptoms)
   - Ask about tingling hands or feet/neck pain & Ax AROM
   - Diff types:
     - Anterior AAS (80%):
       - destruction of transverse lig ⟹ C1 forward on C2
       - significant if >3mm between odontoid & arch of atlas
       - worsened by neck flexion
     - Post AAS (rare)
       - destruction of PEG ⟹ C1 backward on C2
       - worsened by neck ext ie laryngoscopy
     - Vertical AAS:
       - destruction of lat masses of C1 ⟹ odontoid upwards through foramen magnum ⟹ compression brainstem
     - Lateral AAS (uncommon):
       - involvement of C1/C2 facet joints ⟹ compression spinal nerves & vertebral artery
       - >2mm lat alignment significant
       - need open mouth peg view to Ax
2. **subaxial subluxation**
   - >2mm loss of alignment is significant ie C2-C7 step ladder deformity
   - more common if fusion at higher level

- CXR: restrictive changes, effusions, nodules
- spirometry: restrictive pattern, decreased volumes
- ENT nasendoscopy if hoarseness/signs of resp obstruction
- CT or MRI
- ECHO: rule out structural heart disease (restrictive pericarditis\cardiomyopathy), pHTN -> RVF
- xrays of joints: erosion, osteopenia, loss of joint space, swelling
- ECG: pericarditis

MANAGEMENT

- DMARDS: steroids, methotrexate, anti-malarials, gold, sulfasalzine, TNF alpha inhibitors (etanercept, leflunomide)
- symptomatic medications: paracetamol, NSAIDS, intra-articular injections, opioids
- rheumatologist referral
- orthopaedic spinal/neurosurgical referral (C1-C2 fusion)
- cervical collars
- physio
- discussion about RA vs GA
- spinal precautions

Intraoperative

- GA (may require AFOI):
  - unless certain assume all pts have Cx spine instability ie AFOI or inline stabilisation/asleep FOI, VL
  - extreme caution if Post AAS (rare though)
- RA (may be difficult and could fail)
- careful positioning and pressure area care
- steroid supplementation
- good analgesia
- IVF fluid
- temperature cares
- aseptic techniques for invasive procedures (IVC insertion)

Postoperative

- PCA for pain relief (may need to be nurse controlled c/o of difficulty using)

- regular paracetamol
- NSAIDS if not contraindicated
- physio (incentive spiromertry and percussion therapy)
- monitoring
- DVT prophylaxis
- re-institute DMARD ASAP (gold, pencillamine, methotrexate, azathioprin) -> doesn't seem to change rates of post-operative wound infection
- caution with TNF-alpha blockers -> severe infections reported

Ankylosing Spondylitis

Preoperative

= inflammatory arthropathy of the SI joints and spine -> ankylosis
- male:female (4:1). peak onset in 30's

HISTORY

- back pain (worse in mornings and eased by movement)
- fatigue
- sweats
- weight loss

ANAESTHETIC CONSIDERATIONS

- Articular:
  - progressive kyphosis -> limited chest expansion
  - limited mouth opening and TMJ dysfunction
  - cricoarytenoid arthritis - cords susceptible to trauma
  - difficult neuraxial block & ↑risk of epidural haemoatoma - paramedian approach
  - external cardiac massage impossible
  - difficult positioning- tenderness and stiffness of back
- Non articular:
  - fibrosing alveolitis
  - AR (1%). MV & arrhythmias = rare
  - renal involvement - amyoid
  - Cauda equina - in long standing cases
  - assoc NSAIDs & DMARDS

INVESTIGATIONS

- spine xrays
- HLA B27
- increased ESR and CRP
- increased WCC
- renal function (amyloid)

MANAGEMENT

- paracetamol
- NSAIDS
- DMARDS

Intraoperative

- may required AFOI or ILMA
- spinal -> paramedian approach
- positioning and pressure cares

Postoperative

- standard care

Systemic Lupus Erythematous

Preoperative

= chronic, multi-system disease commonest in young females
- wide spread antibodies -> produce tissue damage
- no specific bony erosion or airway implications
- main risks like in assoc problems with major organs:

CLINICAL ASSESSMENT

- SKIN: skin and joint involvement common, oral and pharyngeal ulceration
- CARDIOVASCULAR:
  - pericarditis 15%, myocarditis, endocarditis (less common),
  - Raynauds phenomenon 30%,
  - IHD, arteritis -> ischaemia
- RESPIRATORY: infections and PE's, pleuritis, pleural effusion, pulmonary fibrosis
- NEUROLOGICAL:
  - cranial and peripheral nerve lesions,
  - transverse myelitis -> weakness and paraplegia,
  - depression, psychosis,
  - seizures,
  - stroke - if antiphospholipid antibodies
- RENAL: glomerulonephritis -> renal failure
- HAEMATOLOGICAL:
  - clotting disorders, hypercoagulable state:
    - immune related ↓platelets or antibodies to factor VIII,
    - anti-phospholipid syndrome 33%:
      - hypercoagulable state with paradoxical lupus anticoagulant ⟹ prolonged APTT
      - should seek haem advice if abnormal coags

INVESTIGATIONS

- thrombocytopaenia
- coagulopathy
- U+E
- CXR
- ECHO
- CT/MRI

MANAGEMENT

- medications: steroids, immunosuppressants

Intraoperative

- RA:
  - careful anti-coagulation testing
  - periph nerve lesions may suggest other technique better
- normothermia (decreases Raynauds)
- avoid trauma to airway - laryngeal oedema common
- invasive monitoring
- steroid supplementation
- strict asepsis

Systemic Sclerosis (Scleroderma)

Preoperative

= autoimmune mediated widespread collagenous deposition:

    - deranged cellular & immune function
    - microvascular insult ⟹ T cell accumulation in skin ⟹ fibroblast over proliferation ⟹ too much collagen
- genetic vs environment pathogenesis
- very varied severity

- 2 major types:
    - limited cutaneous form =
        - commoner 60%, milder
        - skin manifestation only present face & distal to elbow
        - can progress to CREST: Calcinosis, Raynauds, Esophageal dysfunction, Sclerodacytly and Telangiectasia
    - diffuse cutaneous form:
        - more aggressive
        - widespread skin hardening & internal organ involvement often lung (fibrosis ⟹ pHTN)
        - high mortality

HISTORY
- above
- fatigue
- decreased ROM @ joints
- tough skin

EXAMINATION
Airway: mouth narrowing and tightening of skin around neck
CVS: Raynauds, pericarditis, myocardial fibrosis, conduction defects, pulmonary hypertension
RESP: fibrosing alveolitis (40% in diffuse form)
RENAL failure
GI: GORD

INVESTIGATIONS
- FBC
- U+E
- PFTs
- ECG

MANAGEMENT
- immunosuppression (methotrexate)
- steroids

ANAESTHETIC IMPLICATIONS
- General - ↑risk of infection
- Skin - difficult cannulation - use US
- MSK:
    - Raynauds ⟹ warm OT & warm fluids
    - TMJ dysfunction & ↓C spine movement ⟹ AFOI, VL, asleep FOI
- GI - antacids
- Resp - ABG, high FiO2, avoid hypercarbia/hypoxia ie IPPV
- CVS - invasive monitoring. consider ECHO if concerns
- avoid renal toxic drugs

Intraoperative
- AFOI
- no consensus GA vs RA
- difficult IV access
- protective lung ventilation
- strict asepsis

Sarcoidosis

See Resp disease notes

Scoliosis

Preoperative

= progressive lateral curvature of the spine with added rotation
- can lead to restrictive lung disease -> hypoxia, hypercarbia, pulmonary hypertension
- causes:

1. idiopathic (75%)
2. secondary to muscular dystrophies, poliomyelitis, CP, Friedreich's ataxia

CLINICAL

- co-morbid conditions
- respiratory reserve
- signs of PHT and RHF

INVESTIGATIONS

- CXR
- PFTs - prior to correction
- ECHO - if concern about muscular dystrophies

Intraoperative

- regional + GA
- intraop spinal cord function monitoring
- prone or lateral issues

Postoperative

- HDU
- monitor Hb, U/O and Na+

Achondropasia

Preoperative

= premature ossification of bones combined with periosteal bone formation -> short limbs, normal cranium
- commonest form of dwarfism

CLINICAL

- AIRWAY:
  - small larynx -> smaller tube,
  - foramen magnum stenosis -> avoid hyperextension of neck
- CVS:
  - difficult IV access
  - use paeds bp cuff
- RESP:
  - restrictive lung disease,
  - OSA common
  - restrictive vent defects can ⟹ pHTN
- BACK:
  - epidural space often narrow, small frequent doses
  - spinal dosing very variable. some suggest spinal should not be used
- CNS: normal intelligence

Ehlers Danlos Syndrome

Preoperative

= group of defective collagen cross-linking disorders

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)

Marfan's Syndrome

- = autosomal dominant disorder of connective tissue metabolism

Clinical Features

- tall with long/thin fingers
- CVS:
  - dilation ascending aorta
  - dissecting aortic aneurysms
  - AR/MR
  - coronary thrombosis
- Resp:
  - emphysema
  - spont PTX
  - pectus excavatum
- Airway:
  - beware tracheomalacia
  - high arch palate
  - OSA
- MSK:
  - easy joint dislocation
  - Cx spine/ligament abnormal - routine XRs not required
  - kyphoscoliosis

Anaesthetic Implications

- control BP - periop ß blockade if not already treated
- minimise SNS response peri-op

- invasive monitoring
- RA is acceptable