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# Summary

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

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*By A Hollingworth & J Fernando*
Practical Points

Immunosuppressed Patients

Risks

Short term
- Staph mediastinitis
- 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 ⇒ ↑↑ ciclosporin levels . 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), crico-arytenoid 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:
  ‣ TNFx 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, palpate 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
- ABD0: splenomegally

INVESTIGATIONS

- routine bloods: organ dysfunction, drug levels, anaemia, thrombocytopenia
- 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 \( \Rightarrow \) C1 forward on C2
    • significant if \( >3 \text{mm} \) between odontoid & arch of atlas
    • worsened by neck flexion
  - Post AAS (rare)
    • destruction of PEG \( \Rightarrow \) C1 backward on C2
    • worsened by neck ext ie laryngoscopy
  - Vertical AAS:
    • destruction of lat masses of C1 \( \Rightarrow \) odontoid upwards through foramen magnum \( \Rightarrow \) compression brainstem
  - Lateral AAS (uncommon):
    • involvement of C1/C2 facet joints \( \Rightarrow \) compression spinal nerves & vertebral artery
    • \( >2 \text{mm} \) 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 \( \Rightarrow \) 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)
By A Hollingworth & J Fernando

- regular paracetamol
- NSAIDS if not contraindicated
- physio (incentive spirometry and percussion therapy)
- monitoring
- DVT prophylaxis
- re-institute DMARD ASAP (gold, penicillamine, 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 haematoma - paramedian approach
  › external cardiac massage impossible
  › difficult positioning- tenderness and stiffness of back
- Non articular:
  › fibrosing alveolitis
  › AR (1%). MV & arrhythmias = rare
  › renal involvement - amyloid
  › 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/hyoxia 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 β blockade if not already treated
- minimise SNS response peri-op
- invasive monitoring
- RA is acceptable