RAYNAUD'S PHENOMENON

Mean age at onset: primary (CTD not apparent)= 14; secondary (established CTD)= 36
10% of all women

CONNECTIVE TISSUE DISEASE
SSc (90%), SLE (40%), PM/DM (40%), Sjogren's (33%), MCTD (85%), RA (10%)
Usually referred due to severe 1^{o} RP; significant portion transition to CTD
ARA CTD criteria low sens/high spec so isolated RP suspicious for CTD (30% of children; old pt)
Predictors: abnormal nailfold vessels (80x risk), immuno tests, ulcers (not in RD), asymmetrical colour changes, fewer digits affected, younger or older age of onset
OBSTRUCTIVE DISEASE: athscl esp men>60/Buerger's/ATOCS/microembolism
DRUGS: B-blockers, ciclosporin, ergotamine, sulfasalazine. cytotoxics
OCCUPATIONAL: hand-arm vibration syndrome(WFVS)/cold/ammo workers/vinyl chloride disease
MISCELLANEOUS: reflex sympathetic dystrophy, hypothyroidism, cryoglobulinaemia, malignancy

Pathophysiology (not precisely known; 4 aetiologies considered)
1. Neurogenic (a-ceptor density increased, B-ceptor presynaptic vesicle increase in peripheral vessels)
2. Blood/VEC interactions
3.Inflamm/immune mechanisms (most severe form occurs with CTD; abnormal WCC behaviour in HAVS)
4. Genetics (higher concordance in monozygotic twins)

Clinical features
COLOUR CHANGE: *PALLOR:* vasospasm *CYANOSIS:* deoxygenation of static blood *RUBOR:* reactive hyperaemia
PAIN/PARAESTHESIA: reactive hyperaemia
TISSUE LOSS: ulcers/gangrene only in RP, not RD
*Triggers: cold(drop in core temperature, not just extremities)/vibration/emotion
*Distribution: fingers and toes (can affect any extremity)
*Differentiating from RD: asymmetrical, fewer digits, ulcer/gangrene, abnormal nail fold/younger;older, immunology

Investigations (confirm diagnosis, differentiate RP from RD)
1. Measure digital SBP before and after cooling at 15degC (drop >30mmHg significant)
2. Strain gauge plethysmography
3. Computerised thermography (skin temperature as indicator of finger flow)
4. Nailfold capillaroscopy (opthalmoscope on high power visualises enlarged vessels; dilatation and tortuosity with patches of "drop-out" ie vessel obliteration)
5. Associated diseased (autoantibody screen, ESR, FBC, renal function, cryoglobulins, TFTs CXR, urinalysis)
Nailfold changes + abnormal immune tests detects 90% of CTDs

Management
1. General: (a)avoid triggers (b)stop smoking (c)ulcer care (beware absence of erythema when infected)
2. Vasodilators:
Nifedipine 20mg TDS (CaChB, gold standard): VDer+antiplatelet||SEs: flush, headache, ankle swelling (abate)
Praxilene (naftidofuryl oxalate): VD+SRS antagonist/addition allows lower dose CaChB
Prostaglandins (PGI2/PGE2 analogues, second line): VD+antiplatelet eg Iloprost||SEs:flush, headache, hypotension
3. Sympathectomy: poor response so c-symp not indicated for upper limb RP

NICE CKS: MANAGEMENT OF RAYNAUD'S (2014)

- Lifestyle: keep whole body including hands and feet warm; devices
  stop smoking
  exercise
  Minimize stress
- Nifedipine prophylaxis immediate release/moderate to reduce SEs (happen in 75%); can use intermittently eg when participating in outdoor activities; periodically stop as can be in remission
- Remission: 2 successive cold seasons or 12 months symptom free
- Refer: rheumatologist or occupational medicine as appropriate;
- Admission: severe digital ischaemia
  -Iloprost reduces frequency and severity of attacks vs placebo/used for threatened digital loss
  -Bosentan (dual ET-1RA) reduces new ulcer formation vs placebo
  -Surgery: digital sympathectomy, stellate ganglion block, lumbar sympathetic blocks, local or regional sympathetcomy if threatened digital loss, debdribement of infected/necrotic tissue