Management of Raynaud’s Phenomenon

1. Scope

This guidance is applicable to primary care and secondary care clinicians involved in the management of Raynaud’s Phenomenon (RP). It has been adapted from a version written by the Highland Rheumatology Unit.

2. Diagnosis

Raynaud’s Phenomenon (RP) is the biphasic (blue/white) or triphasic colour change of the extremities when exposed to stimuli such as temperature change or emotional stimuli. It can occur in 3-20% of the population. Most RP does not need to be referred to rheumatology but it is important to make sure that you have considered secondary Raynaud’s in each case where referral to Rheumatology would be appropriate. Unilateral Raynaud’s may reflect underlying vascular pathology such as subclavian artery stenosis or aneurysm, and referral to the vascular service for investigation should be considered.

2.1 Primary Raynaud’s

Often occurs in late teens/ early twenties. It is not associated with any connective tissue disease (CTD) features and doesn’t lead on to digital ulceration.

2.2 Secondary Raynaud’s

Occurs in ~ 1% off all Raynaud’s where it is associated with underlying rheumatic disease.

Clinical features suggestive of an underlying Connective Tissue Disease (CTD) are:

- **Systemic sclerosis (Scleroderma)**: tightness/ thickening of skin of fingers, reflux/dysphagia, digital ulcers, calcinosis, telangectasiae, breathlessness, hypertension, active urinary sediment (blood/ protein on dipstick), occasionally small joint inflammatory arthritis

- **Lupus (SLE)**: arthralgia (occasionally small joint inflammatory arthritis), malar rash, discoid rash, alopecia, photosensitive rash, recurrent oral ulcers, cytopenia (low WCC or platelets), serositis (pleuritic/ pericarditic chest pain), hypertension, active urinary sediment (blood/protein on dipstick)
**Anti-phospholipid syndrome (APS)**: PE, DVT, recurrent unexplained early miscarriages (< 10 weeks), single unexplained late miscarriage (> 10 weeks) or unexplained death in utero, eclampsia/ pre-eclampsia, young stroke/MI, livedo reticularis

**Primary Sjogren’s syndrome**: persistent dry/ gritty eyes or need for frequent artificial tears, persistent dry mouth, unexplained parotitis/ salivary gland swelling, photosensitive rash, discoid rash, cytopenia (low WCC or platelets), occasionally small joint inflammatory arthritis

**Myositis**: Proximal limb girdle muscle weakness, elevated Creatinine Kinase (CK), photosensitive rash, typical dermatomyositis rash, occasionally small joint inflammatory arthritis

### 2.2.1 Investigations

Further investigations recommended as per clinical presentation:

- Blood pressure
- Urine dipstick
- CTD screen (C3/C4 if suspecting Lupus or Sjogren’s)
- CXR if breathless or serositis
- CTD screen (C3/C4 if suspecting Lupus or Sjogren’s)
- Lupus anticoagulant & anti-cardiolipin antibodies (if suspecting APS)
- Routine bloods
- CK (if suspecting Myositis)

If clinical features and/ or investigations suggestive of CTD refer to rheumatology.

### 3. Management

#### 3.1 Non-pharmacological

Essential to the management of RP.

- Smoking cessation
- Remove offending medication e.g. β blocker
- Refer patients to Scleroderma and Raynaud’s UK website for advice on self management - www.sruk.co.uk

#### 3.2 Pharmacological

All treatments are expected to give only a modest improvement

**First line**: Calcium channel blockers.
Nifedipine capsule 5mg is licensed for the treatment of Raynaud's phenomenon. However, this preparation is being discontinued. No other dihydropyridines are licensed for the treatment of Raynaud’s phenomenon. There is clinical experience suggesting that long-acting nifedipine is effective for the treatment of Raynaud's and has fewer adverse reactions than rapid-acting preparations. The long acting preparations of nifedipine are administered once daily and the slow release preparations twice daily.¹

Suggested dose:

Nifedipine slow release 10mg nocte increased by 10mg per week until on 30mg BD or maximum dose tolerated.

If ineffective or not tolerated Amlodipine 5mg – 10mg OD is an alternative.²

3.2.2. Secondary care rheumatology

Second line treatment options require referral to rheumatology.

**Second line: sildenafil³⁴** (Blanket off-label form for this cohort previously approved).

Commence sildenafil 25mg OD, titrating weekly up to 50mg TDS or maximum tolerated dose.

**Third line: Intravenous iloprost.** A licensed version is now available. Administered at Clinical Intervention Unit, QMH.

4. Management of Digital Ulceration

Requires urgent referral to rheumatology in cases of secondary Raynaud’s – exclusion of co-existent proximal vessel disease and infection.

Treatment options:

- Sildenafil
- IV iloprost
- Bosentan

5. References
