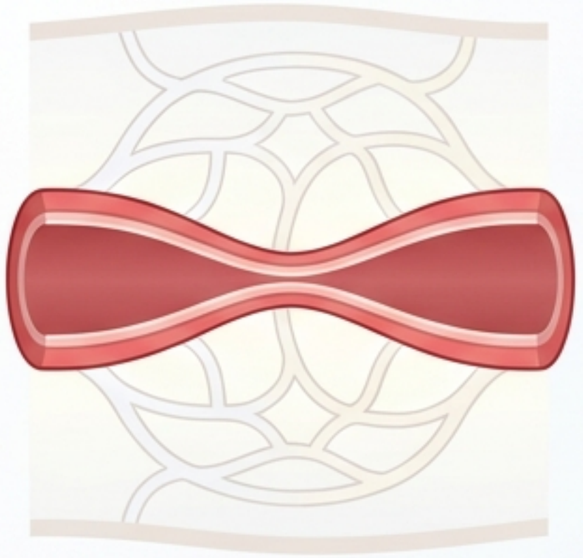
Raynaud Phenomenon

Clinical Guidelines & Management Workflow

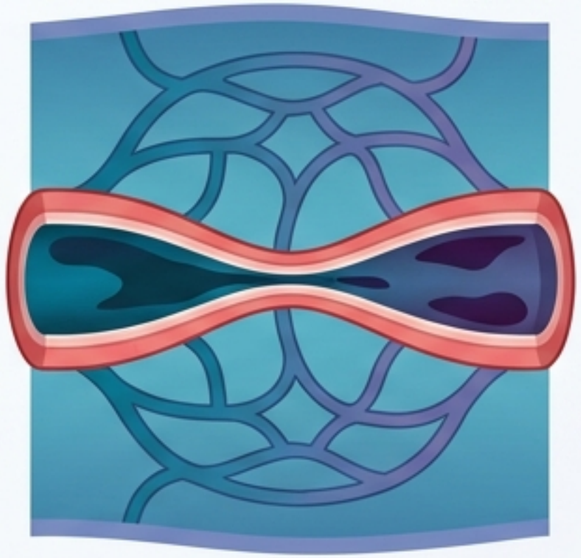
Synthesized from Med2Date Clinical Guidelines (Last updated 13 May 2026)

Stage 1:
White (Pallor)



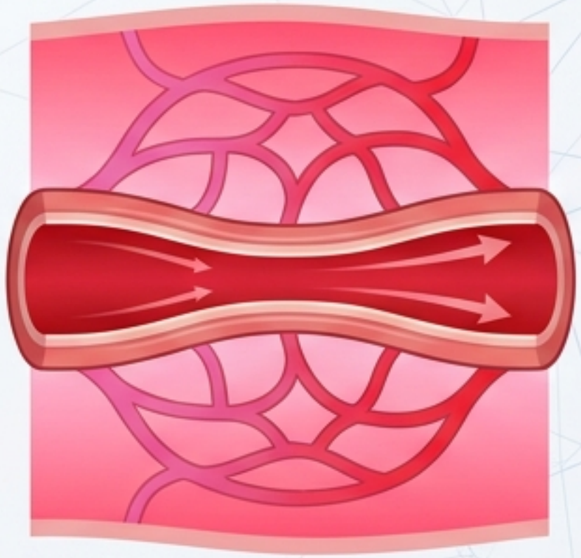
Artery tightly constricted (episodic vasospasm).
Trigger: Cold exposure or emotional stress.

Stage 2: Blue
(Cyanosis)



Artery remains constricted.
Blood pooling and deoxygenation.

Stage 3: Red
(Reactive Hyperaemia)

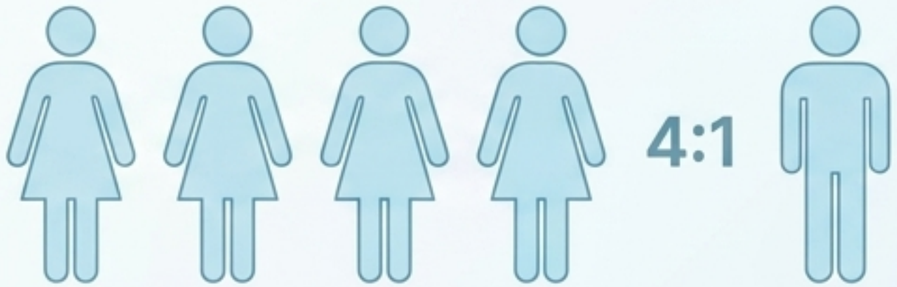


Artery wide open.
Rapid reperfusion of oxygenated blood.

Raynaud phenomenon (RP) is a reversible vasospastic disorder of the digital arteries and cutaneous microcirculation.

Australian Epidemiology & The Clinical Stakes

Primary RP: High Prevalence, Benign Course



- **3-5%** of the Australian general population.
- Young adults (15-30 years); Female-to-male ratio ~4:1.
- **Outcome**: Benign, self-limiting. Exacerbated by cold ambient temperatures in southern states.

Secondary RP: Rare, High Morbidity

- **0.5-1%** of the population.
- **Association**: >95% of systemic sclerosis cases present with RP.

Morbidity Alert

~30% of systemic sclerosis patients develop digital ulcers (Australian Scleroderma Cohort Study), risking severe infection and auto-amputation.

Primary vs. Secondary RP: The Diagnostic Matrix

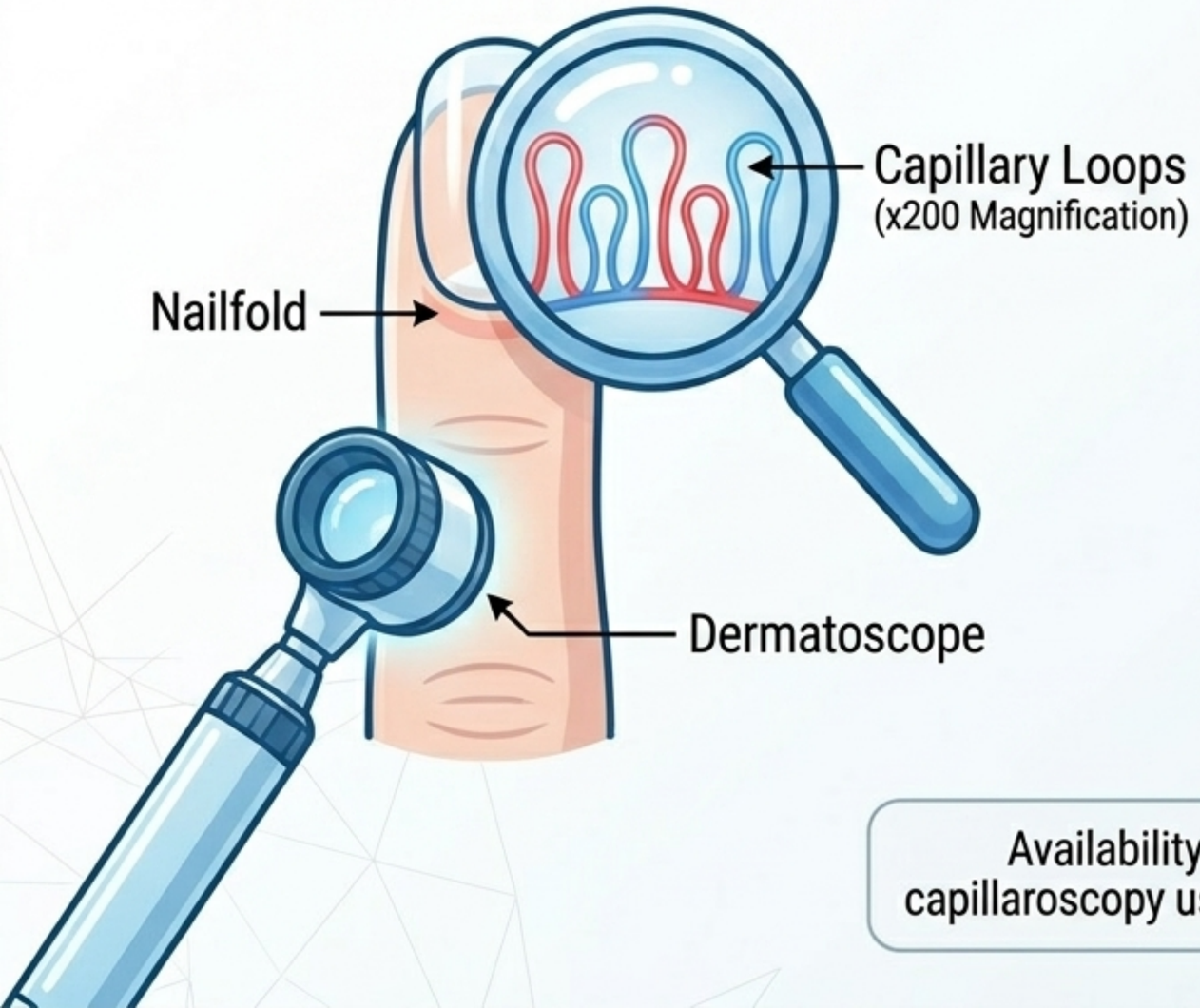
Primary RP (Raynaud Disease)		Secondary RP (Raynaud Syndrome)
Age of onset	15-30 years	>30 years (variable)
Symmetry	Symmetric, all digits	Often asymmetric, may spare thumbs
Tissue necrosis	Never	Digital pitting, ulcers, gangrene
Nailfold capillaries	Normal	Abnormal (giant, haemorrhages)
ANA	Negative or low-titre	Often positive (>1:160)
Associated features	None	Skin tightening, arthritis, sicca, dysphagia, ILD

Red Flags for Secondary RP

Refer to rheumatology if any are present: Age >30, asymmetric attacks, digital pitting/ulcers, abnormal capillaries, ANA >1:160, or signs of connective tissue disease. Note: 10-15% of primary diagnoses progress to secondary over 10 years.

The Microvascular Window: Nailfold Capillaroscopy

The single most important investigation to differentiate primary from secondary RP.



Examine 8 Fingers
Assess all digits. Exclude thumbs due to natural anatomical variation.

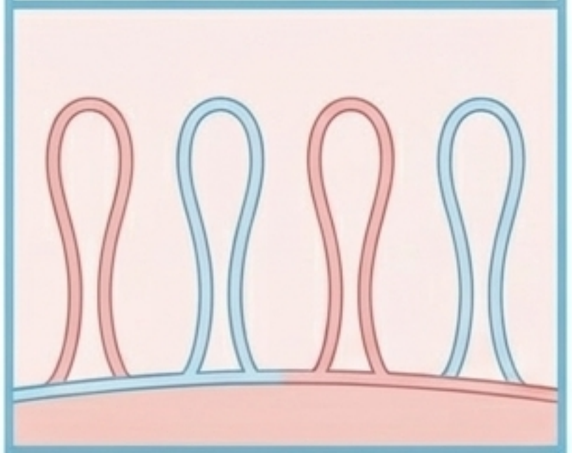
Preparation
Apply immersion oil/gel to the nailfold. Patient must acclimatise to room temperature for 15-20 minutes.

Equipment
Examine via stereomicroscope (x200 magnification) or use a dermatoscope / USB videocapillaroscope.

Availability: Standard at tertiary rheumatology departments. Telehealth capillaroscopy using patient-acquired images is emerging for remote access.

Capillaroscopy Progression Timeline

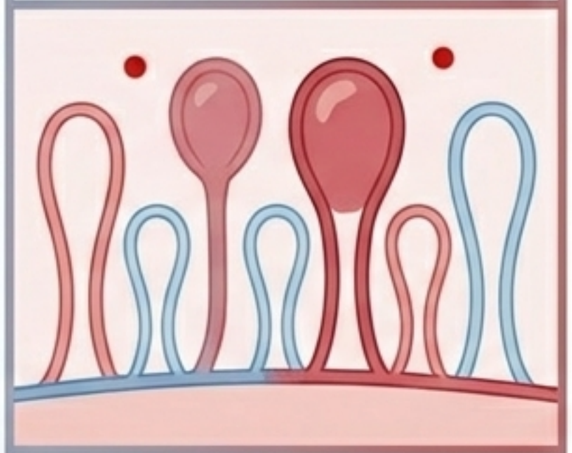
Normal
(Primary RP)



Normal Architecture

- 9-14 loops per mm.
- No enlargement or dropouts.

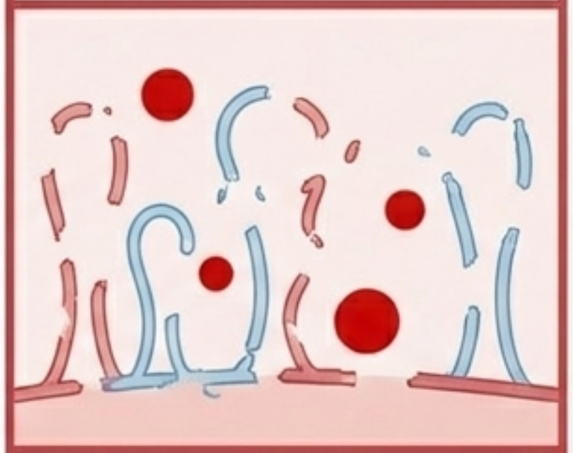
Early
(Scleroderma Pattern)



Early Phase

- Giant capillaries (>50 µm).
- Isolated microhaemorrhages.
- **Action:** Rheumatology monitor every 6-12 months.

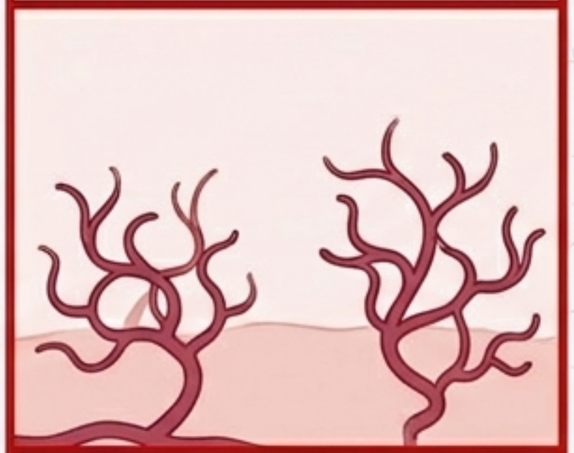
Active Phase



Active Phase

- Multiple giant capillaries.
- Frequent haemorrhages, reduced density.
- **Action:** Initiate/enhance immunosuppression.

Late Phase



Late Phase

- Extensive avascular zones.
- Bushy neoangiogenesis.
- **Action:** Aggressive management, ulcer prevention.

Autoantibody Risk Profiling & Biochemical Workup

The Essential Work-Up

Primary Test:

- ✓ ANA by indirect immunofluorescence (IIF).
- ✓ Negative/low-titre (<1:160) is reassuring.
- ✓ High-titre (≥1:160) warrants further ENA testing.

Supportive Labs:

- ✓ FBC, ESR, CRP, UEC, LFTs (baseline for therapies).
- ✓ TFTs (rule out hypothyroidism).
- ✓ SPEP, Cryoglobulins (transport warm at 37°C).

Antibody	Associated Risk/Significance
Anti-centromere (ACA)	Limited cutaneous SSc. Higher risk of pulmonary arterial hypertension (PAH).
Anti-Scl-70 (Topo I)	Diffuse cutaneous SSc. Higher risk of interstitial lung disease (ILD); worse prognosis.
Anti-RNA Pol III	Diffuse cutaneous SSc. Increased risk of scleroderma renal crisis.
Anti-Ro/La	Sjögren syndrome. Risk of neonatal lupus in pregnancy.

Baseline Management: Universal Lifestyle Interventions

1. Cold Avoidance

Insulated layered gloves, warm socks, hand-warmers. Pre-warm car interiors.

2. Smoking Cessation

The single most impactful modifiable factor. Offer NRT, varenicline, or bupropion (PBS-supported).

3. Meds to Avoid

β-blockers, ergotamine, sumatriptan, clonidine, amphetamines, pseudoephedrine. (Switch to cardioselective β-blockers if necessary).

4. Stress Management

CBT and biofeedback for anxiety-triggered RP attacks.

5. Exercise & Skin Care

Aerobic exercise for vasodilation. Protect digits from trauma (steel-capped boots). Keep hands moisturised.

6. Diet

Avoid excessive caffeine. Consider fish oil; ensure adequate Vitamin D.

First-Line Pharmacotherapy: Dihydropyridine CCBs

Trial for at least 4 weeks at optimal dose. Monitor for dose-dependent oedema, headache, flushing.

Nifedipine (Slow-release)

Dose:
20 mg SR daily, titrate to 30-60 mg SR daily (max 90 mg/day).

Adjustments:
Cautious titration in hepatic impairment.

Status:
PBS General Benefit.

Amlodipine

Dose:
5 mg daily, titrate to 10 mg daily.

Adjustments:
Start at 2.5 mg in hepatic impairment or elderly.

Status:
PBS General Benefit.

Second-Line & Emergency Therapies

Second-Line Therapies: PDE5 Inhibitors

Col 1: Sildenafil: 20 mg TDS.	**Col 2:** Tadalafil: 20 mg alternate days or daily.

Status: Off-label (Not PBS listed for RP)

⚠ **CONTRAINDICATED with nitrates (severe hypotension). Do not combine with CCBs without strict BP monitoring.**

Alternative Oral Options (Less Common)

Losartan (25-50mg) | Fluoxetine (20mg)

Emergency Salvage Therapy (Tertiary Centres Only)

- **IV Iloprost:** Prostacyclin analogue (0.5-2 ng/kg/min infusion).
- **Indication:** Critical digital ischaemia with threatened gangrene.

Requires continuous cardiac monitoring.

Managing Complications: Digital Ulcers

Classification of Digital Lesions

Digital Pitting

Small depressions from healed ulcers.

Management: Vasodilators.

Ischaemic Ulcer

Punched-out lesion with pale/necrotic base.

Management: Vasodilators, wound care (flucloxacillin/cephalexin; doxy/TMP-SMX for MRSA).

Calcific Ulcer

Overlying calcinosis deposits.

Management: Surgical debridement, topical sodium thiosulfate.

PBS Authority Pathway: Bosentan (Ulcer Prevention)

Mechanism: Endothelin receptor antagonist.
Dose: 62.5 mg BD for 4 weeks → 125 mg BD.
Status: PBS Authority Required (specifically for prevention of new digital ulcers in systemic sclerosis).
Black Box Warnings: Teratogenic (Category X – reliable contraception required).
Dose-dependent hepatotoxicity (monthly LFTs mandatory).

Special Populations: Tailoring Management

Pregnancy

- **Guidance:** Nifedipine is Category C but generally considered safe and is first-line. Avoid Amlodipine.

⚠ **Warning:** Bosentan is absolutely contraindicated (Category X).

Paediatrics

- **Guidance:** Primary RP is most common. Focus on lifestyle first. Use paediatric dosing for CCBs.

➡ **Action:** Secondary RP requires immediate paediatric rheumatology referral.

Elderly

- **Guidance:** Start CCBs at lower doses (amlodipine 2.5 mg). Monitor for hypotension and fall risk.

⚠ **Alert:** New-onset RP in elderly should prompt investigation for atherosclerotic or thromboembolic causes.

Immunocompromised

- **Guidance:** Patients on MTX, mycophenolate, or rituximab have higher infection risks with digital ulcers.

➡ **Action:** Maintain a lower threshold for antibiotics and surgical review.

Aboriginal and Torres Strait Islander Health Considerations

Access & Delivery Logistics

- Specialist services and capillaroscopy are limited remotely.
- Utilize telehealth, RFDS referral pathways, and emerging USB videocapillaroscopy.
- Medications available via Remote Area Aboriginal Health Services (RAAHS) Section 100 supply.

Culturally Safe Care

- Use accessible analogies (e.g, ‘your fingers are like pipes that squeeze shut in the cold’).
- Respect gender-specific preferences for hand examinations.
- Maintain a low threshold for ANA testing if skin changes present.

Comorbidities & Risk Factors

- Significantly higher smoking rates (~40% vs ~11%).
- Integrate proactively with Tackling Indigenous Smoking programmes.

Synthesis: The Raynaud's Clinical Pathway

Presentation: Patient presents with triphasic digital colour changes (White → Blue → Red).

Assessment & Investigation: Perform clinical exam & check Red Flags (Age >30, asymmetry, necrosis). Order ANA + ENA panel. Perform Nailfold Capillaroscopy.

Primary RP Profile
Normal capillaries, ANA negative, no ulcers.

Management: GP Follow-up. Lifestyle measures. If severe, prescribe CCBs (Nifedipine/Amlodipine).

Secondary RP Profile
Abnormal capillaries, ANA ≥1:160, ENA positive, or ulcers present.

Management: Rheumatology Referral. Add PDE5i if refractory. Bosentan for ulcer prevention. Monitor for complications (PAH/ILD).