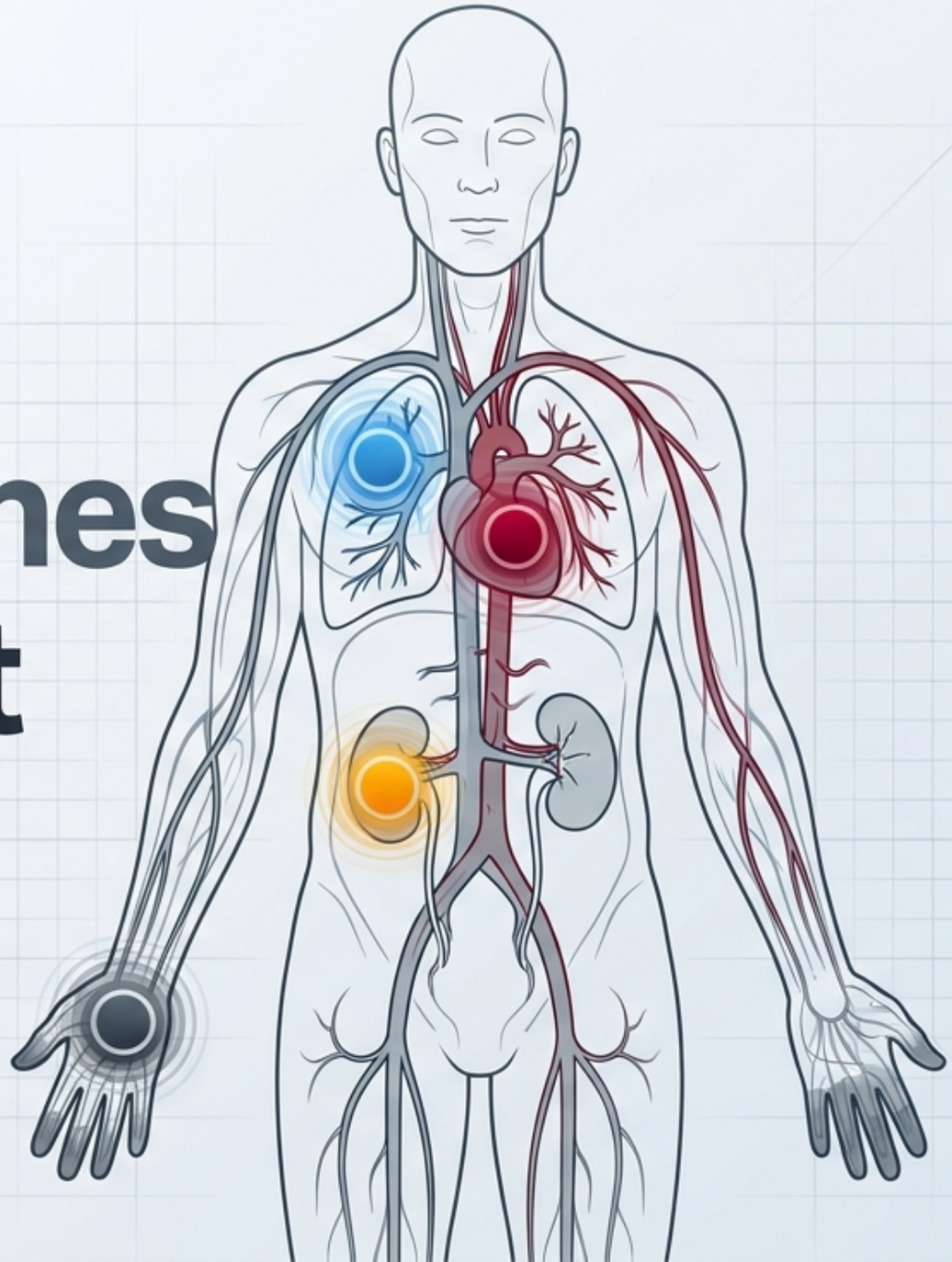
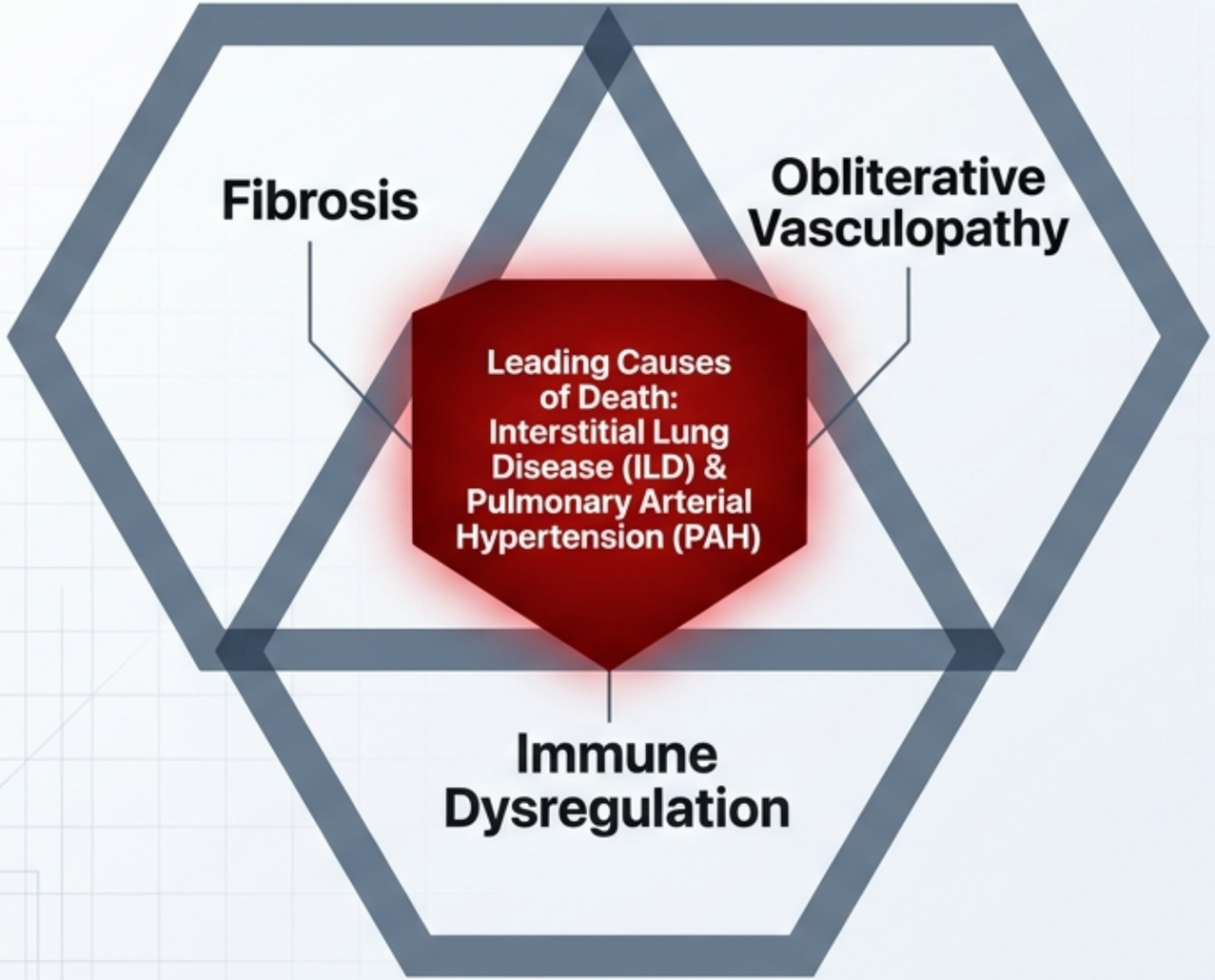
Systemic Sclerosis: Clinical Guidelines & Management Playbook

Med2Date Australian Reference Edition



The Pathological Triad Driving SSc Mortality



Standardised mortality ratio:

3–5×

the general population.

Australian Epidemiology and Patient Demographics

Prevalence

~20 per 100,000 adults

(Incidence: ~2 per 100k)

Gender Gap

4 : 1

(Female to Male ratio)

Peak Onset

30–50 years

of age

Phenotype Split

Diffuse SSc accounts for ~40% of the Australian Scleroderma Registry cohort, carrying significantly higher mortality.

40% Diffuse SSc	60% Limited SSc

Diagnostic Risk Stratification: Limited vs. Diffuse SSc

Limited Cutaneous SSc (lcSSc)

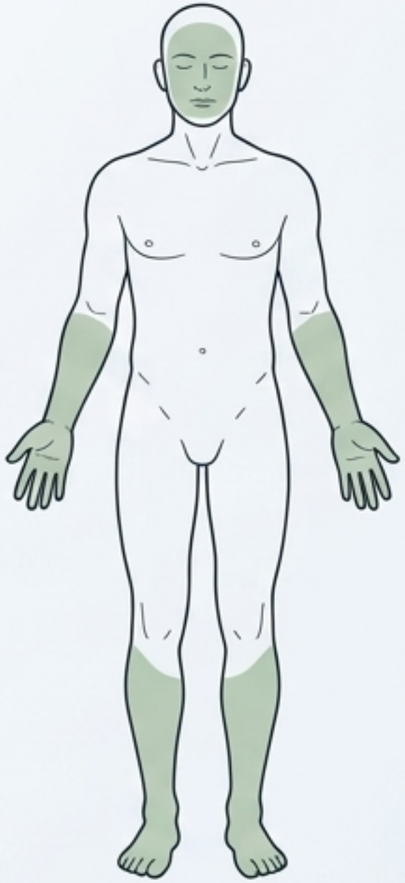
Antibodies: ACA (70-80%)

ILD Risk: Moderate

PAH Risk: Higher

SRC Risk: Low (~2%)

10-Year Survival: ~75%



Diffuse Cutaneous SSc (dcSSc)

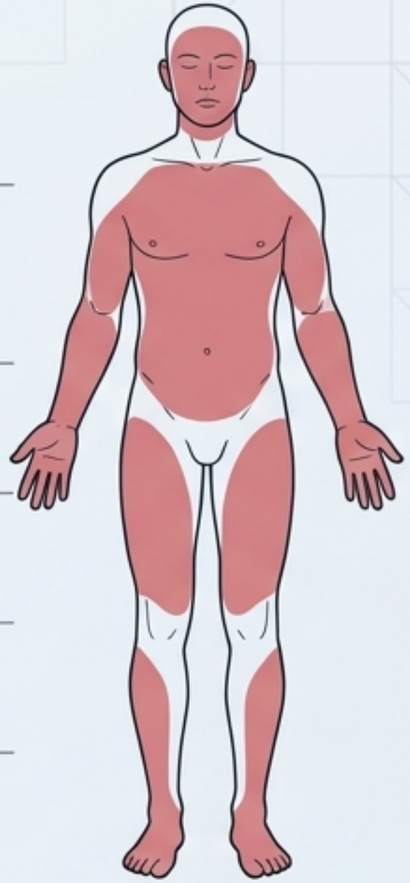
Antibodies: Scl-70 (30%), RNA Pol III (20%)

ILD Risk: High (~80%)

PAH Risk: Lower

SRC Risk: Higher (~15%)

10-Year Survival: ~55-65%

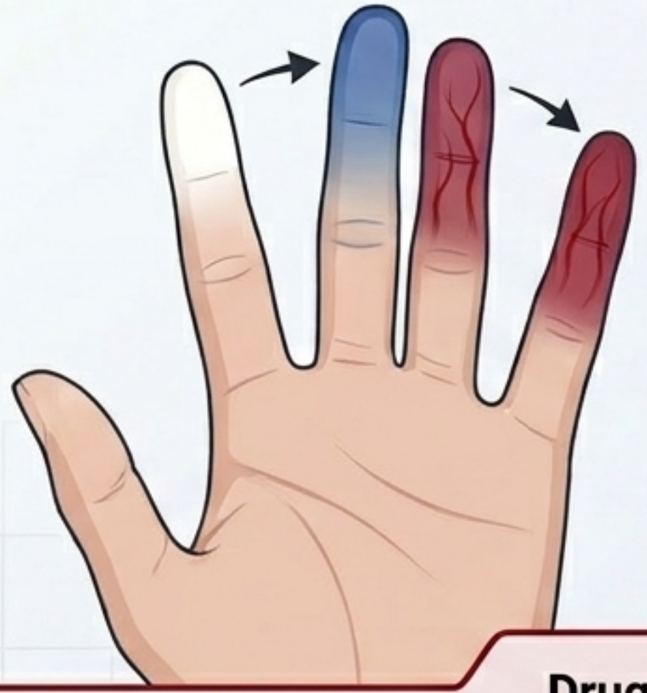


⚠ **ALERT:** RNA Polymerase III indicates high risk of scleroderma renal crisis and concurrent cancer (especially within 3 years of onset). Cancer screening mandatory.

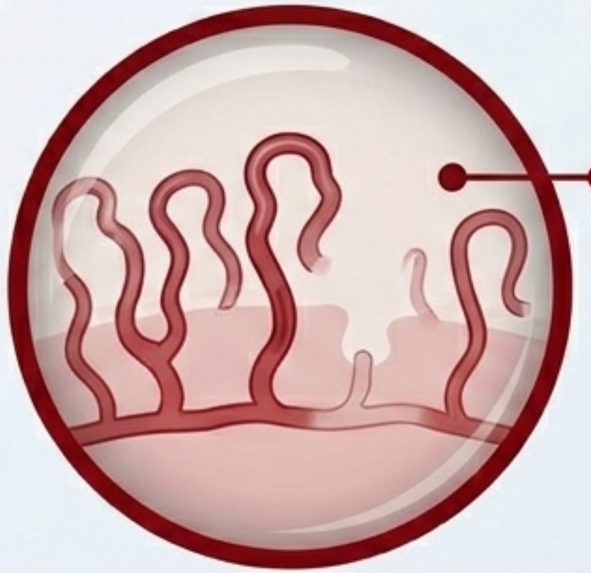
Identifying and Managing SSc-Associated Raynaud Phenomenon

Present in >95% of SSc patients; often the earliest manifestation.

Clinical Vasospasm



Nailfold Capillaroscopy



Abnormal capillaries: Key predictor of internal organ involvement in secondary RP.

Pharmacology Callout Widget

✔ PBS General Benefit

Drug: Nifedipine XR (Adalat®)
Role: First-line Calcium Channel Blocker vasodilator.
Dose: 30–60 mg PO daily.

Step-Up Treatment Escalation for Digital Ulcers

1. Conservative

Wound care, cold avoidance, smoking cessation, CCBs.

2. Add Vasodilator

PDE-5 inhibitor (Sildenafil 20 mg PO TDS) or Iloprost IV infusion.

3. Refractory Prevention

Bosentan (62.5 mg BD → 125 mg BD) for recurrent ulcers.
Botulinum toxin injection.

⚠ PBS Status: Authority Required (Specialist script)

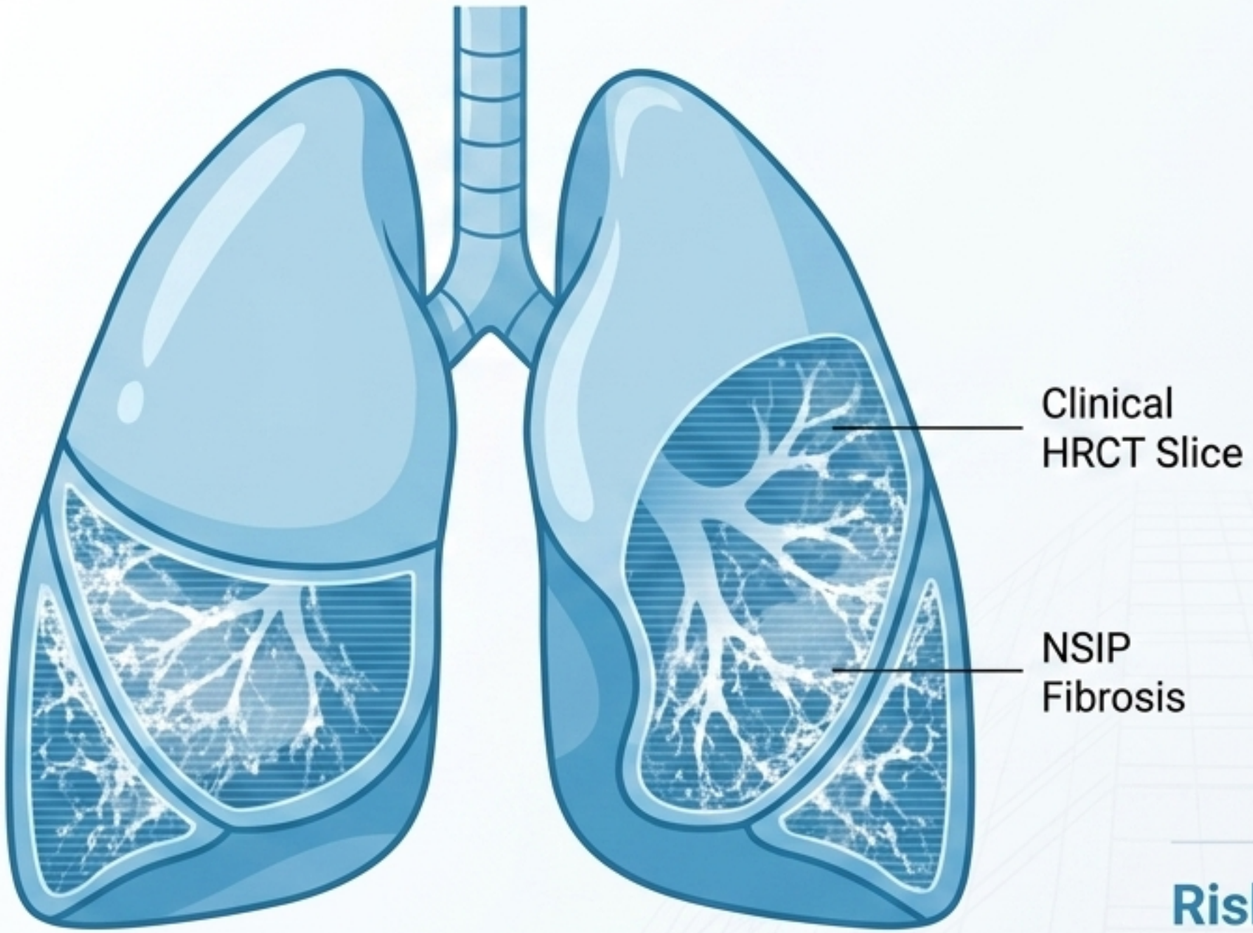
ACUTE DIGITAL ISCHAEMIA

Protocol: IV Iloprost (2 ng/kg/min escalating to max 6 over 6hrs x 3-5 days).

Add heparin if thrombosis suspected.

Surgical consult for gangrene.

Interstitial Lung Disease (SSc-ILD): The Leading Killer



Screening Protocol Flow

Baseline

HRCT + PFTs (FVC, DLCO) at diagnosis.

Annual Cadence

PFTs annually (more frequent if dcSSc or Scl-70 positive).

Trigger for Repeat HRCT

FVC decline >10% or new respiratory symptoms.

Risk Factors

- dcSSc phenotype
- Anti-Scl-70 positive
- Anti-U3 RNP positive

SSc-ILD Treatment Stratification

Mild → Moderate → Severe

Mild

- **Criteria:**
  <10% ILD extent on HRCT,
  FVC >70%.

Action:
Monitor closely (PFTs 6-12 mo).

Setting:
Outpatient Rheumatology.

Moderate

- **Criteria:**
  10-20% extent or declining FVC.

Action:
Mycophenolate mofetil (MMF)
1.5–2 g/day PO preferred 1st-line.
(Alt: IV Cyclophosphamide).

Setting:
Rheum + Respiratory.

Severe

- **Criteria:**
  Progressive fibrosing phenotype.

Action:
MMF/Cyclophosphamide induction
+ Nintedanib 150mg BD add-on.
Lung transplant assessment.

Setting:
Multidisciplinary ILD Clinic.

Pharmacology Callout

MMF (CellCept®)

✓ PBS General Benefit. Target 2g/day. Monitor FBC/LFTs.

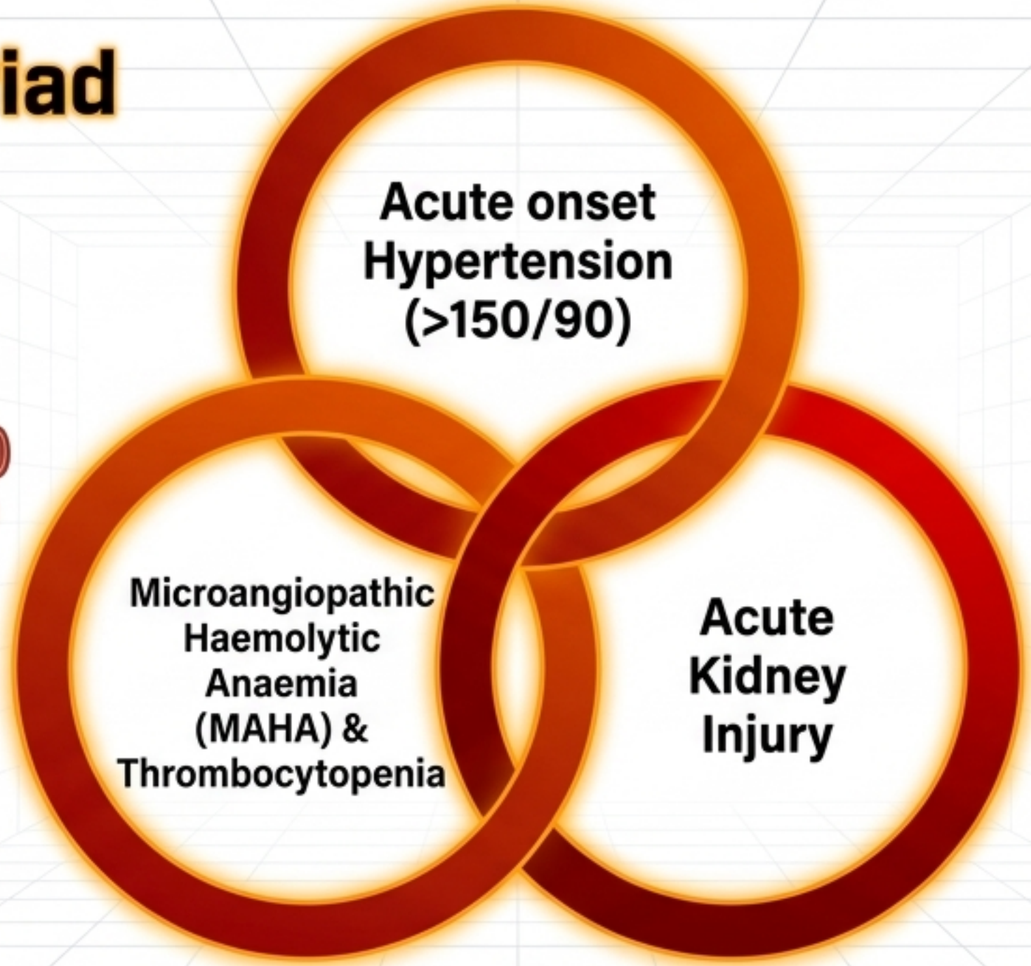
Pharmacology Callout

Nintedanib (Ofev®)

⚠ PBS Authority. Antifibrotic (TKI). Side effect: Diarrhoea.

Medical Emergency: Scleroderma Renal Crisis (SRC)

The Acute Triad



Risk Factors

- dcSSc (within first 4 years)
- RNA Pol III positive
- New anaemia / pericardial effusion

MAJOR IATROGENIC TRIGGER: Corticosteroids >15 mg/day prednisolone. Cyclosporine/Tacrolimus use also implicated.

SRC Immediate Response Protocol

1. Immediate ACE Inhibition

Captopril (Capoten®) 6.25–25 mg PO TDS.

Titrate aggressively every 24-48 hrs to BP <130/80.

Note: Short-acting preferred for rapid titration.
PBS Gen Benefit.

❌ 2. Avoid Harmful Agents

NO Corticosteroids >15mg.
NO NSAIDs.
NO Cyclosporine.

Note: ARBs do NOT substitute for ACE inhibitors in SRC.

3. Renal Replacement Trajectory

Haemodialysis if required. Continue ACEi regardless.

⚠ **Critical note:** Do not perform early nephrectomy; renal recovery is possible over 12–24 months on ACEi.

Pulmonary Arterial Hypertension (SSc-PAH) Screening Screening

Step 1: Annual Non-Invasive Screening

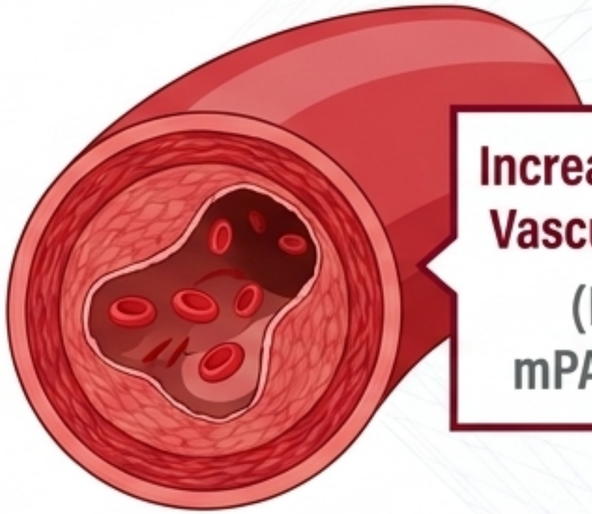
Echocardiography (TR velocity, RV function) + DLCO testing.

Step 2: Trigger for Escalation

DLCO <60% predicted OR dropping >15%.
Worsening dyspnoea (WHO functional class).

Step 3: Definitive Diagnosis

Right Heart Catheterisation (RHC).



Increased Pulmonary Vascular Resistance

(PVR >3 WU, mPAP >20 mmHg)

Risk Profile

Affects 10–15% of patients.

Higher risk in lcSSc, ACA positive patients, and those with a history of **digital ulcers.**

PAH Pharmacotherapy and Specialist Pathways

Class	Therapy
ERA	Ambrisentan 5-10 mg daily PO.
PDE-5i	Sildenafil 20 mg TDS PO.
Prostacyclin	Treprostinil SC / Epoprostenol IV.
sGC stimulator	Riociguat 1-2.5 mg TDS PO.

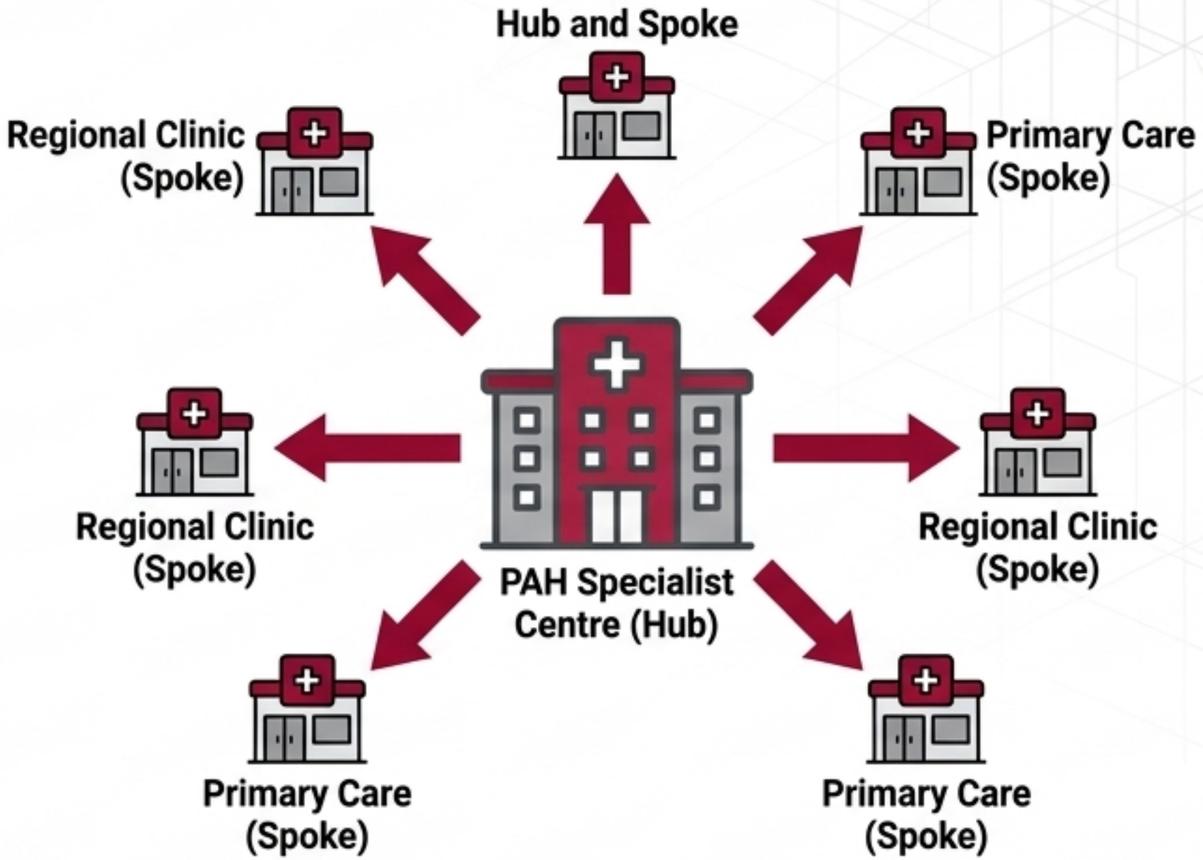
Standard of Care Highlight Block

✓ **Combination Therapy:** ERA + PDE-5i is first-line dual therapy.

Clinical Warnings

⚠ Riociguat is strictly contraindicated with PDE-5 inhibitors.

⚠ Anticoagulation is **NOT** routinely recommended in **SSc-PAH** (unlike idiopathic PAH).



Referral Callout

Mandatory referral to designated PAH centres (e.g., Royal Adelaide, St Vincent's Sydney, Alfred Melbourne) for RHC, initiation, and PBS Authority access.

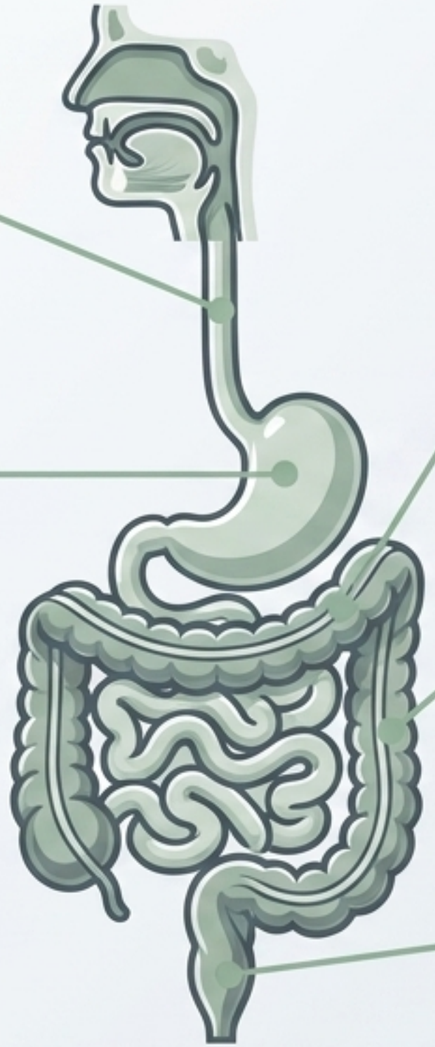
Gastrointestinal Dysmotility: An Anatomical Approach

Oesophagus: GORD / Dysphagia

Tx: Omeprazole 20-40mg, elevate head of bed

Stomach: Gastroparesis / Early Satiety

Tx: Metoclopramide 10mg TDS or Domperidone pre-meals



Small Bowel: Bacterial Overgrowth

Tx: Rotating antibiotics (Ciprofloxacin, Doxycycline, Amox-Clav 1 week/month)

Colon: Constipation / Pseudo-obstruction

Tx: Prucalopride (Resotrans®) 2mg PO daily (✓ PBS Gen), osmotic laxatives

Anorectal: Faecal Incontinence

Tx: Biofeedback, sacral nerve stimulation

GI involvement affects >90%. Severe weight loss/dysphagia requires immediate dietitian referral.

Special Populations: Modifying Care Pathways

Pregnancy

Teratogen washout required: Stop
- Stop Mycophenolate (≥6 wks), Methotrexate (≥3 mos).
- Switch Bosentan to Sildenafil.

Contraindicated: ACE inhibitors (use labetalol/nifedipine).

High SRC risk persists postpartum.

Elderly

Late-onset SSc (>65) is often dcSSc with a worse prognosis.

Adjust doses for renal function.

High falls risk with vasodilators.

Renal Impairment

Mycophenolate: Reduce dose if eGFR <25; avoid if severe.

Cyclophosphamide: Reduce by 25-50% if eGFR <30.

STRICT AVOIDANCE: NSAIDs (trigger SRC risk).

Immunocompromised

Administer Influenza, Pneumococcal (13+23), COVID, and Shingles (>50yr) vaccines prior to starting immunosuppression.

Monitor for PCP and TB.

Aboriginal & Torres Strait Islander Health Delivery

Telehealth & Diagnostics

Utilize MBS items 91822/91823 for specialist follow-up. Train local health workers to facilitate capillaroscopy imaging and strict BP monitoring remotely.

Medication Access

Route PBS Authority applications (Bosentan, Nintedanib) through Aboriginal Medical Services (AMS) pharmacies to leverage Closing the Gap co-payments.

Cultural Safety

Integrate Aboriginal Health Workers in care planning. **Respect kinship** obligations, especially regarding complex discussions around SRC dialysis, end-of-life care, and travel for RHC. Note high baseline rates of renal/cardiovascular comorbidities.

The Comprehensive SSc Annual Review Dashboard

Organ System	Baseline / Diagnostic	Annual Cadence	Ongoing / PRN
Lungs	HRCT + PFTs	FVC and DLCO testing	Repeat HRCT if symptoms
Heart/PAH	Echocardiography	Echo (TR velocity) + Clinical dyspnoea assessment	RHC if deteriorating
Kidneys	Baseline Creatinine	Monitor FBC for MAHA/thrombocytopenia	Strict BP monitoring (target <130/80)
Skin/Vascular	LeRoy typing, Autoantibody panel (ACA, Scl-70, RNA Pol III), Nailfold Capillaroscopy	Skin score review	Monitor for digital ulceration

Holistic SSc management requires hyper-vigilance across all organ systems.
Early detection of ILD, PAH, and SRC is the primary determinant of survival.