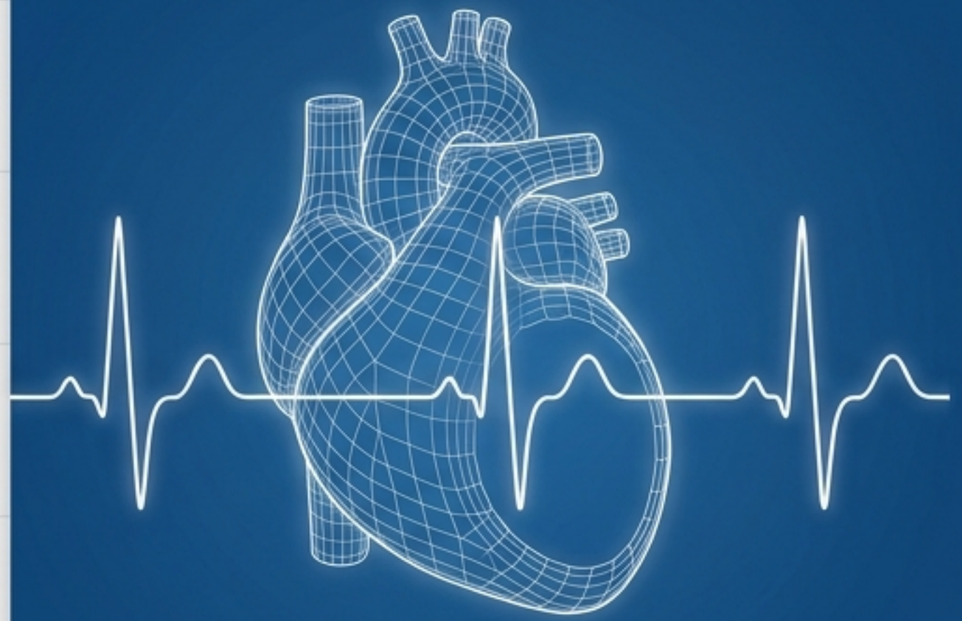


Dilated Cardiomyopathy Clinical Care Continuum

Navigating Diagnosis, Guideline-Directed Therapy, and Genetic Substrates in Australian Practice

[Clinical Guidelines] [Target Audience: HCPs] [Australian Healthcare Context]

The Australian DCM Snapshot

Prevalence & Impact

1:250 to 1:500 Prevalence

Drives 30–40% of all non-ischaemic HFrEF presentations. Accounts for 2.5% of all >65yo hospitalisations.

The Ultimate Endpoint

Leading Transplant Indication

Represents ~45% of adult primary diagnoses for heart transplantation in the ANZOD registry.

Economic Burden

$2.7B Annual Cost

Financial impact driven by PBS pharmacotherapy, device implantation, readmissions, and advanced therapies.

Hallmark Definition: Left ventricular dilatation and systolic dysfunction (LVEF ≤40%) **without abnormal loading conditions or CAD** sufficient to explain the impairment.

The Dichotomy of Dilatation: Ischaemic vs. Non-Ischaemic

Ischaemic DCM		Non-Ischaemic DCM
Significant epicardial CAD (≥70%) or prior MI.	Coronary Anatomy	Normal or non-obstructive.
Regional hypokinesis (corresponds to coronary territory).	Wall Motion	Global hypokinesis.
Subendocardial/transmural in coronary territory.	CMR LGE Pattern	Mid-wall fibrosis (non-territorial) or diffuse oedema.
Worse survival. Potential benefit from revascularisation.	Prognosis	Generally better 5-year survival if idiopathic.

MANDATORY CORONARY ASSESSMENT: All newly diagnosed DCM patients aged >40 years (or younger with ≥2 CV risk factors) MUST undergo CT or invasive coronary angiography. Unrecognised ischaemia accounts for 30% of 'idiopathic' cases in older cohorts.

Systematic Aetiological Evaluation

Toxic / Drug-Related

Causes: Alcohol (≥80g/day for >5yrs), Anthracyclines, Trastuzumab, Methamphetamine.

Action: Substance history, Urine drug screen.

Infectious / Inflammatory

Causes: Viral (Coxsackie, Parvovirus, SARS-CoV-2), Giant Cell, Sarcoidosis.

Action: CMR, Serology, FDG-PET.

Endocrine / Metabolic

Causes: Thyroid dysfunction, Phaeochromocytoma, Haemochromatosis.

Action: TFTs, Catecholamines, Iron studies.

Autoimmune / Systemic

Causes: SLE, Rheumatoid Arthritis, Scleroderma, EGPA.

Action: ANA, anti-dsDNA, Eosinophil count.

Infiltrative / Storage

Causes: Amyloidosis (AL/ATTR), Fabry disease.

Action: Serum free light chains, Tc-99m PYP scan.

Arrhythmia-Mediated

Causes: Persistent AF, incessant SVT.

Action: ECG, Holter monitoring (Rate control/ablation may reverse DCM).

The Diagnostic Imaging Arsenal

Transthoracic Echo (TTE)

First-Line

Utility: LVEDD >58mm, LVEF ≤40%, global/regional wall motion, GLS.

MBS Item 55118

Cardiac MRI (CMR)

Gold Standard Tissue

Utility: Late gadolinium enhancement (LGE) distinguishes ischaemic vs non-ischaemic; maps fibrosis/oedema.

MBS Item 63484

Coronary Angiography

Mandatory if >40yo

Utility: CT (low/int risk) vs Invasive (revascularisation likely).

MBS Item 57360 / 38218

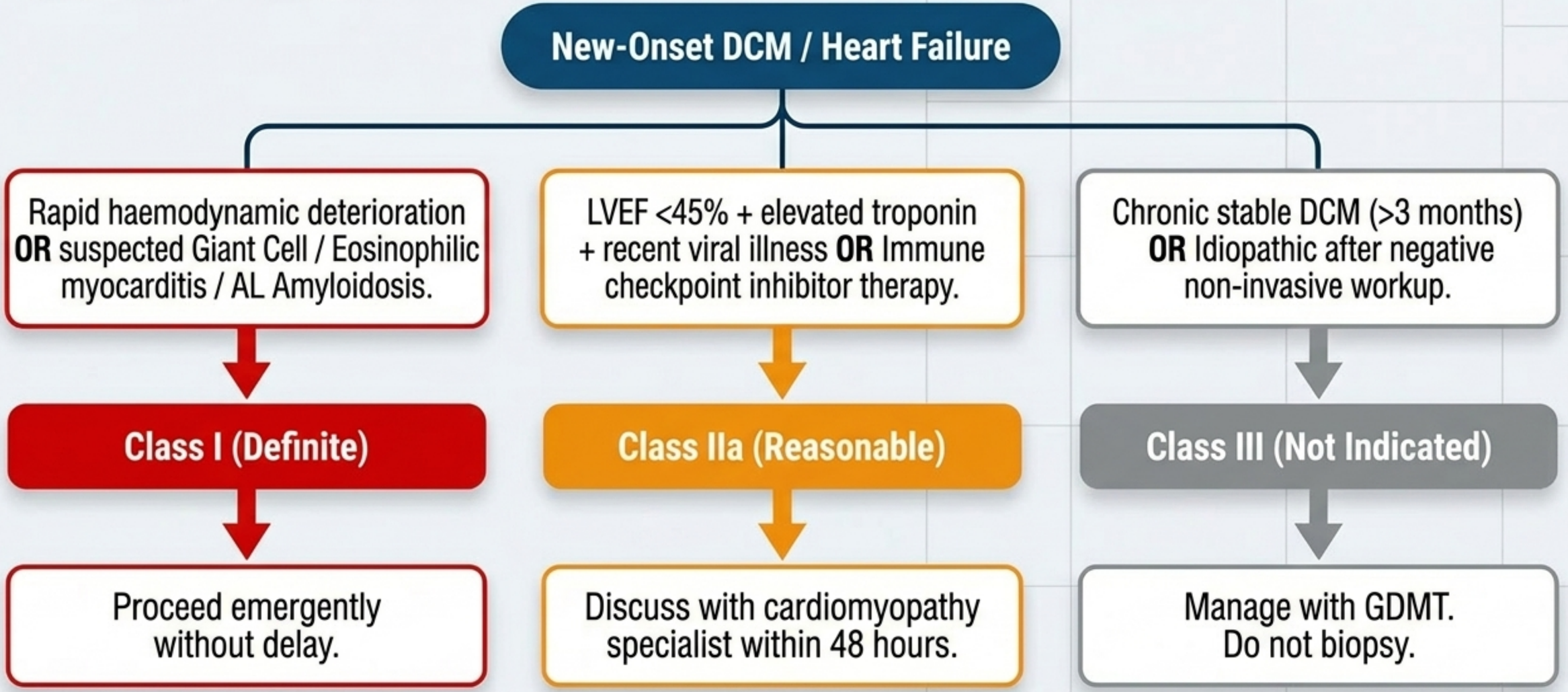
FDG-PET / CT

Targeted

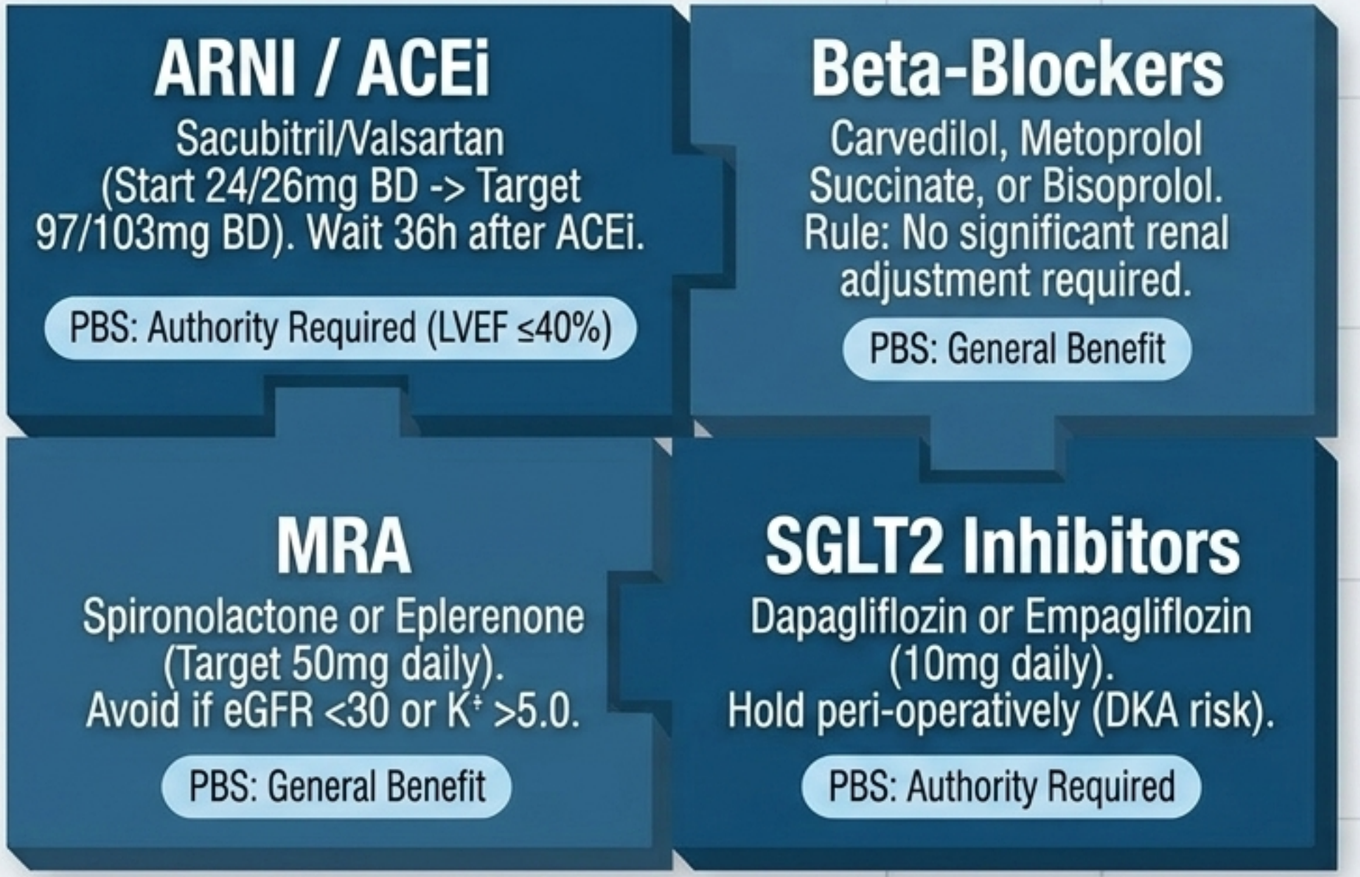
Utility: Preferred for diagnosing active cardiac sarcoidosis inflammation.

MBS Item 61406

Endomyocardial Biopsy (EMB) Decision Tree

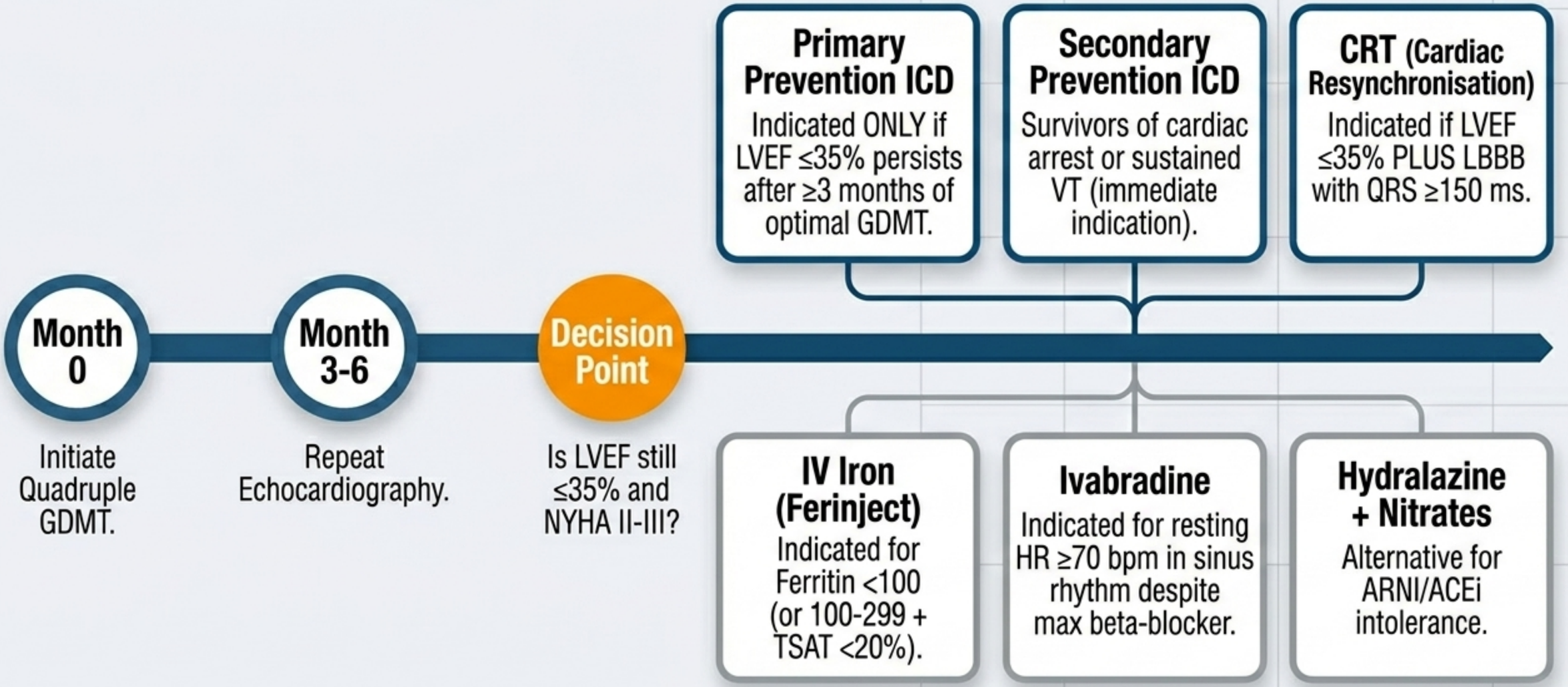


The Quadruple Therapy Foundation (GDMT)



MANDATORY RULE: Early Sequential Initiation. Do not wait for full uptitration of one agent. Initiate all four pillars at low doses within the first 4 weeks.

Device Therapy and Adjunct Pharmacotherapy

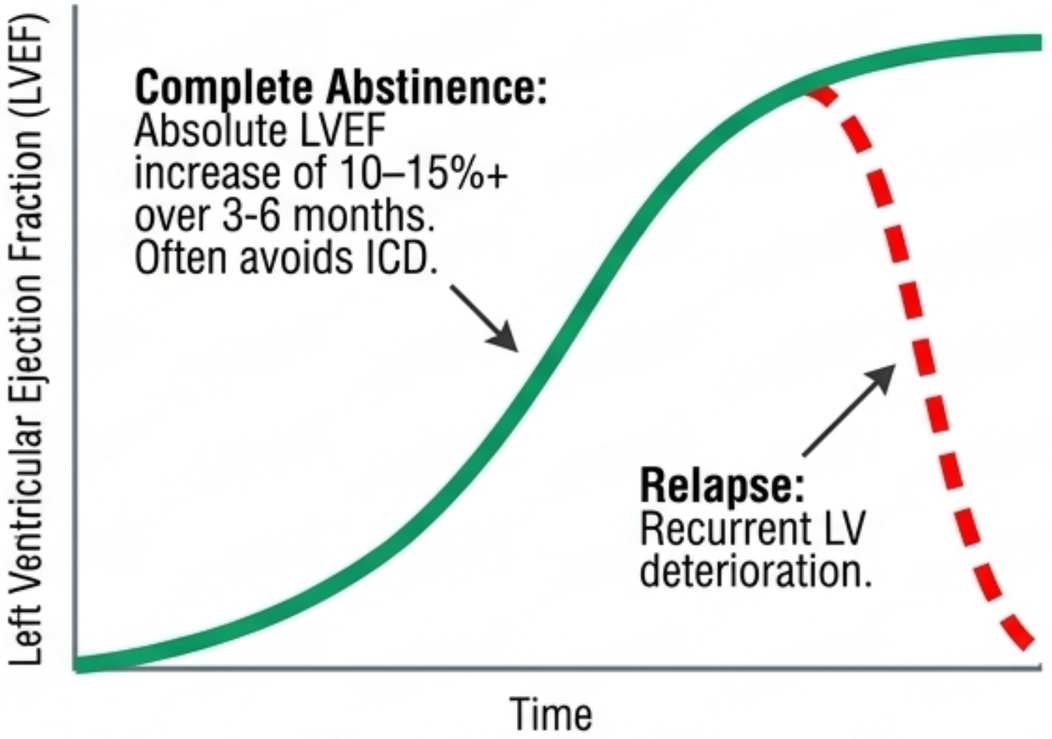


Targeted Aetiological Management

Inflammatory & Autoimmune DCM	• **Giant Cell Myocarditis:** Mandatory immunosuppression (Methylprednisolone IV -> > Prednisolone + Cyclosporine/Azathioprine). Untreated 1-year mortality >90%. • **Viral Lymphocytic:** Immunosuppression NOT recommended unless viral PCR negative on biopsy.
Immune Checkpoint Inhibitor (ICI) Myocarditis	• **Action:** Hold ICI immediately. High mortality requires urgent oncology-cardiology MDT. • **Therapy:** High-dose IV Methylprednisolone. Re-challenge is generally contraindicated.
Anthracycline Cardiotoxicity	• **Prevention:** Serial echo during chemo. GLS >15% relative reduction flags subclinical toxicity. • **Intervention:** Initiate GDMT immediately upon LVEF decline. (Trastuzumab-relatoed is usually reversible; Anthracycline is often irreversible).

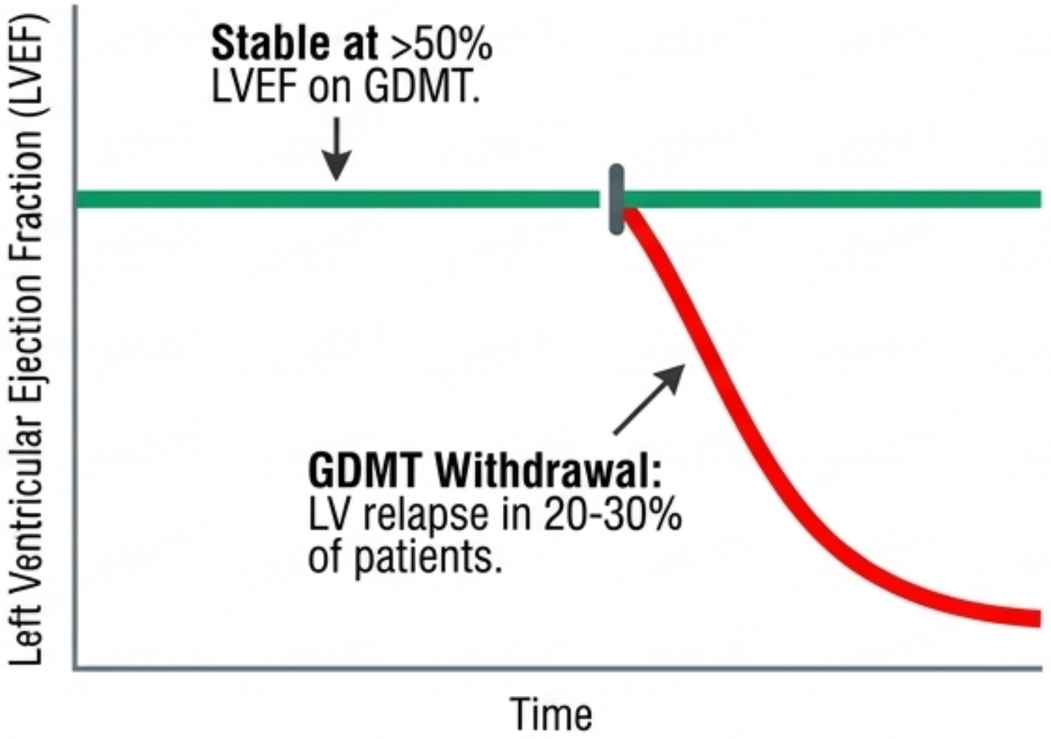
The Reversibility Curves: Recovery vs. Relapse

Alcoholic Cardiomyopathy



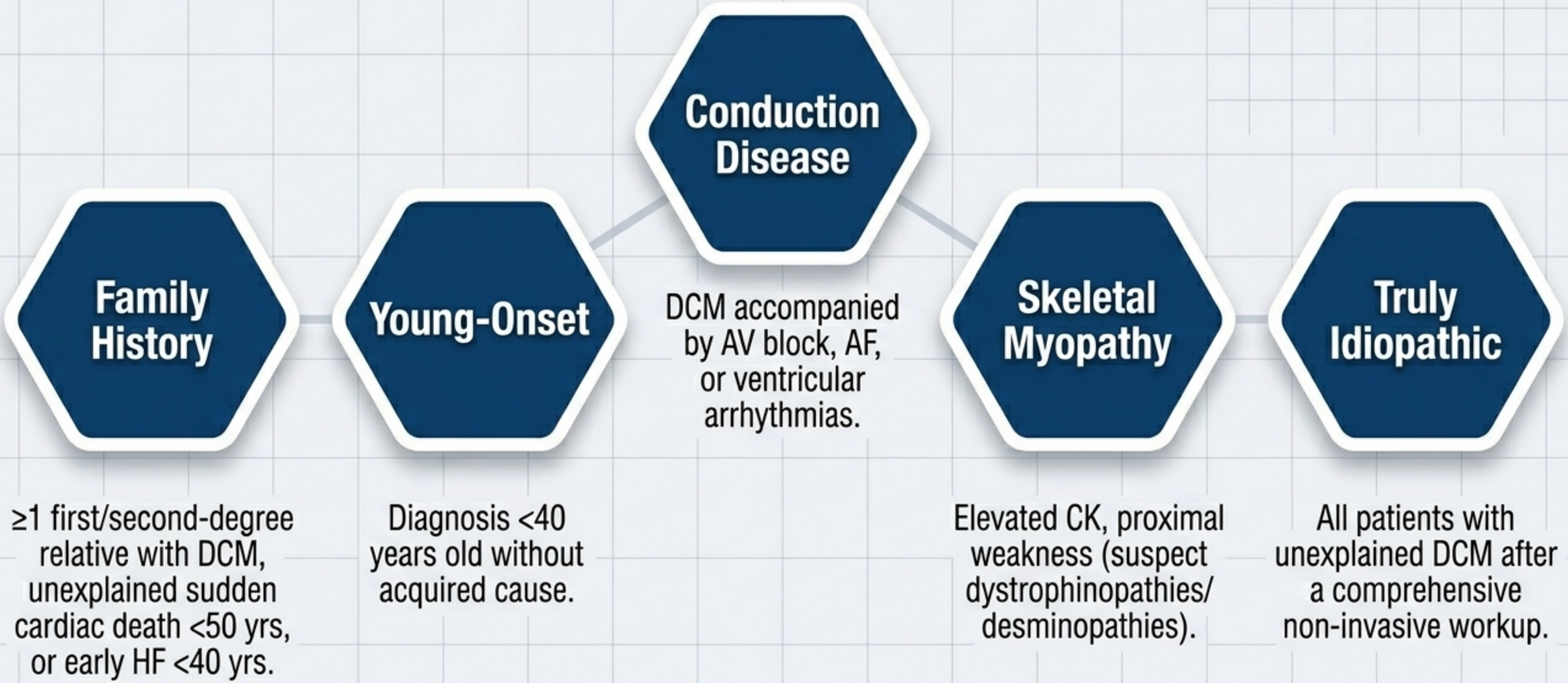
Counsel all patients on threshold (≥80g/day for >5yrs). Co-prescribe Naltrexone/Acamprosate and Thiamine.

The TRED-HF Trap (Recovered Idiopathic DCM)



WARNING: Recovered DCM is NOT Cured DCM. Lifelong GDMT maintenance is mandatory even if LVEF normalises.

Identifying the Genetic Phenotype: When to Refer



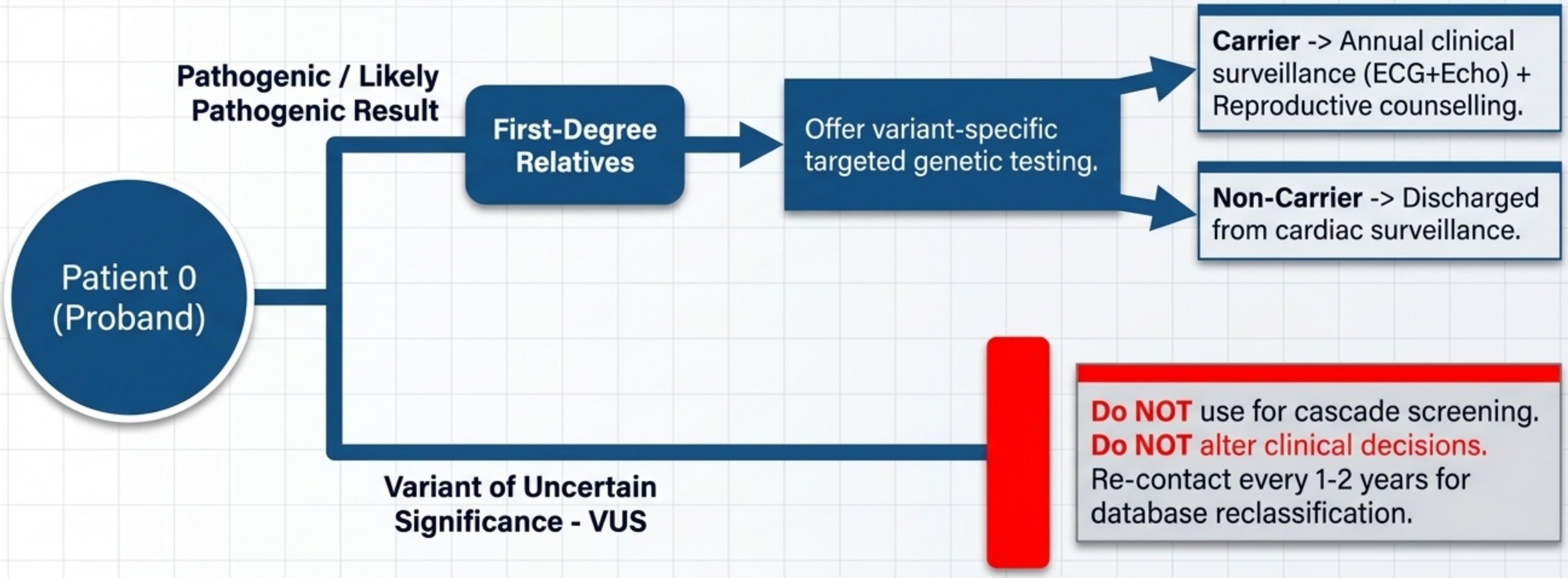
The Genetic Substrate Matrix

Favorable Response		
Gene	**Frequency**	**Clinical Profile**
TTN (Titin truncating variants)	~25% of familial DCM (Most common)	Reduced penetrance. Highly favorable LVEF recovery on standard GDMT.

High-Risk/Aggressive		
LMNA (Lamin A/C)		High-risk for AV block, AF, and aggressive arrhythmias. Lower threshold for ICD implantation.
PLN (Phospholamban)		Aggressive end-stage HF risk and sudden death (p.Arg14del variant).
RBM20		Very early-onset, high arrhythmia burden, LV thrombus risk. Early transplant consideration.

Insight: Current panels solve 30-50% of familial DCM; negative tests do not rule out genetic aetiology.

Executing the Family Cascade Screen



Context: Australian genetic testing available via State Clinical Genetics Services or private panels (~$400-800). Refer relatives to the Australian Genetic Heart Disease Registry.

Prognostic Horizons and Risk Stratification

Low Risk

Idiopathic or TTNtv-related.

LVEF >30%.

NT-proBNP <1000 pg/mL.

Normal renal function.

Setting: GP-led monitoring + 6-12 month cardiology review.

Moderate Risk

Persistent AF.

Renal impairment (eGFR 30-60).

NT-proBNP 1000-5000 pg/mL.

CMR shows Mid-Wall LGE (OR 4.6 for adverse events).

Setting: Multidisciplinary HF Clinic.

High Risk

LVEF <25%.

LMNA, PLN, or RBM20 variant.

Hyponatraemia (<135 mmol/L).

eGFR <30.

Peak VO_2 <14 mL/kg/min.

Setting: Urgent Advanced Therapy Evaluation.

Global Longitudinal Strain (GLS) decline >15% relative change is more **sensitive** than LVEF for detecting early deterioration.

Special Population Nuances

Pregnancy

Contraindicated: ACEi, ARB, ARNI, SGLT2i, MRA are teratogenic.

Safe: Labetalol/Metoprolol, Hydralazine + Nitrates.

Insight: Bromocriptine may inhibit prolactin-driven pathophysiology in Peripartum Cardiomyopathy (PPCM).

Paediatrics

Aetiologies: Infants (<1yr) = Myocarditis/Metabolic; Older children = Genetic/Idiopathic.

Therapy: Enalapril and Carvedilol are standard. SGLT2i NOT approved.

Renal Impairment

Action: Accept up to 30% rise in creatinine on RAASi. Do not stop unless K^+ >6.0 or AKI.

SGLT2i Rule: Can initiate if eGFR ≥20 mL/min (reduces loop diuretic requirements).

Hepatic Impairment

Action: Carvedilol is extensively hepatically metabolised (reduce dose).

Eplerenone contraindicated in Child-Pugh C.

Warfarin requires strict INR monitoring due to congestion.

Aboriginal and Torres Strait Islander Health Considerations

The RHD Endgame

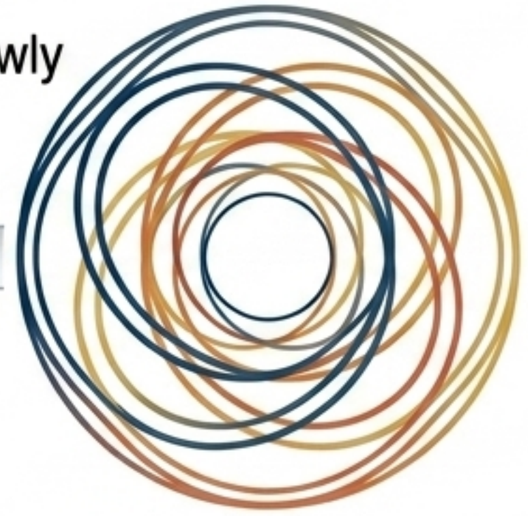
Secondary DCM from Rheumatic Heart Disease remains endemic in remote NT, WA, and QLD.

RHD must be excluded with echo in all newly diagnosed ATSI patients from these regions.

Burden & Demographics

2–3-fold excess heart failure burden.

Mean age at diagnosis is ~15 years earlier (~55 vs ~70 years).



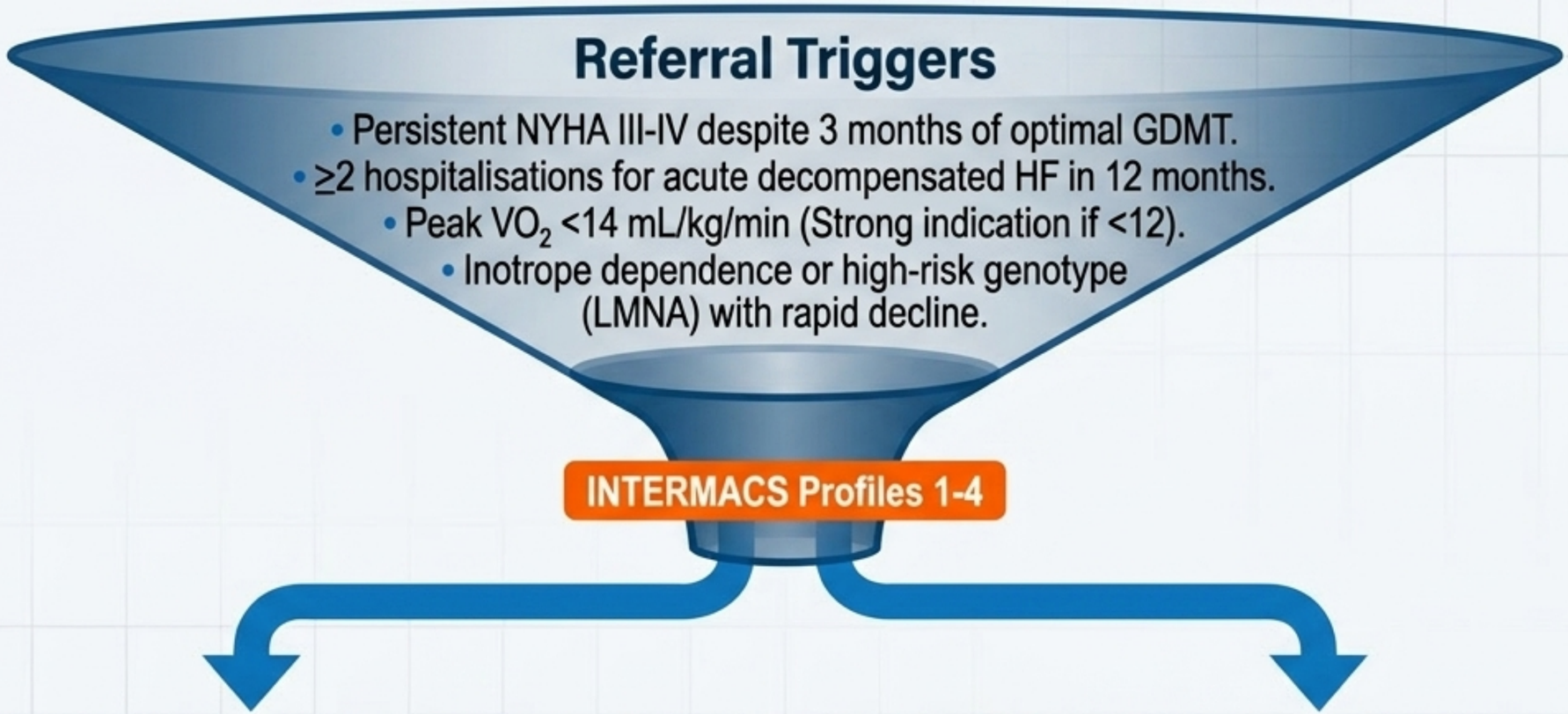
Culturally Safe Pathways

- Utilize PBS Closing the Gap (CTG) co-payments to ensure complex GDMT adherence.
- Partner with Aboriginal Community Controlled Health Organisations (ACCHOs) for integrated management of comorbid diabetes and CKD.

Culturally Safe Pathways

Utilize NT Cardiac telehealth and portable handheld echocardiography to overcome severe access barriers in remote communities.

The Advanced Therapy Threshold



LVAD

Bridge to transplant or destination therapy (e.g., HeartMate 3). 2-year survival ~75-80%.

Heart Transplant

Median wait time in Australia is 6-18 months.
Critical Rule: Refer before the patient becomes critically unwell.
1-year post-transplant survival exceeds 90% at Australian centres.

The Patient Care Continuum Map

Month 0 (Diagnosis & Initiation)

- Echo + CMR + Mandatory Ischaemia Exclusion.
- Initiate all 4 pillars of GDMT simultaneously at low doses.
- Refer for Genetic Counselling based on 5 trigger criteria.

Months 1-2 (Optimization)

- Uptitrate GDMT every 2-4 weeks. Monitor UEC, K^+, eGFR.

Month 3 (The Checkpoint)

- Repeat Echocardiography.
  - ⚠️ If LVEF ≤35% -> Primary Prevention ICD.
  - ✅ If LVEF >50% -> Maintain lifelong GDMT (Beware TRED-HF trap).

Month 6+ (The Prognostic Horizon)

- Assess functional capacity (CPET/6MWT).
- If NYHA III/IV persists -> Refer for Transplant/LVAD evaluation.
- Commence annual surveillance. Family cascade screening depled based on genetic results.

Closing Footnote: Best practice demands integrated multidisciplinary heart failure management programs to reduce readmissions and mortality.