INFLAMMATORY MYOPATHIES: THE CLINICAL PATHWAY CANVAS

A visual reference atlas for the diagnosis, risk stratification, and management of idiopathic inflammatory myopathies in Australian practice.

AUSTRALIAN EPIDEMIOLOGY & DISEASE BURDEN

10–15 per 100,000

Overall prevalence of **idiopathic inflammatory myopathies** (IIM) in Australia.

DERMATOMYOSITIS

Incidence 5–9 per million. Bimodal peaks at 5–15 years & 45–60 years.

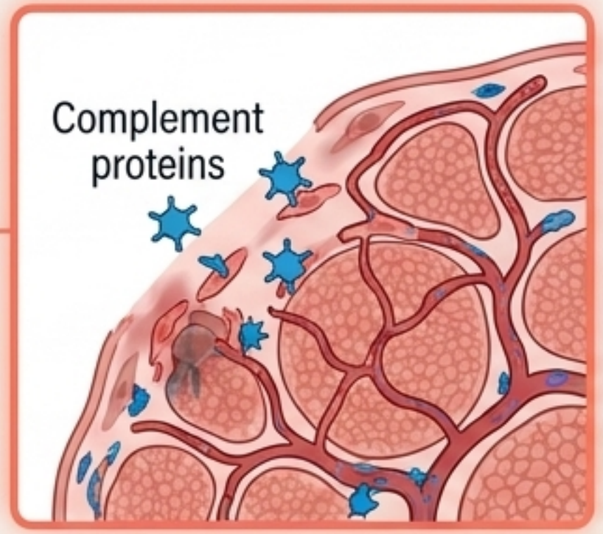
INCLUSION BODY MYOSITIS

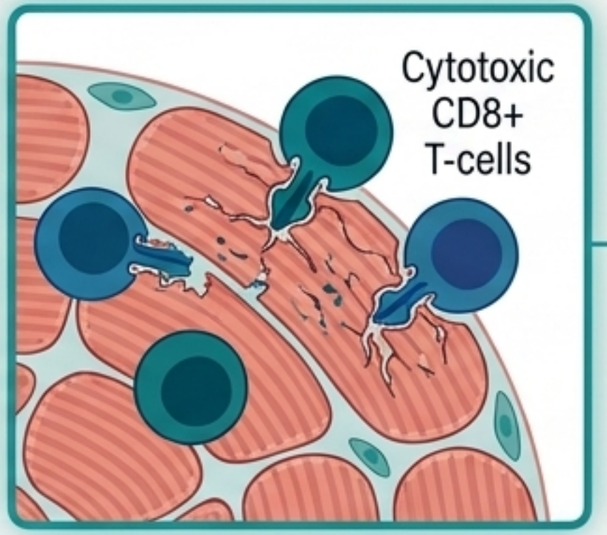
45–70 per million (in age ≥50). Male-to-female ratio 3:1. Most common acquired myopathy >50 yrs.

The Statin Factor: 2.5 Million+ PBS statin prescriptions annually highlight the critical diagnostic consideration of **statin-associated IMNM** in any patient with unexplained myalgia and marked CK elevation.

THE CELLULAR BATTLEFIELD: PATHOPHYSIOLOGY BY SUBTYPE

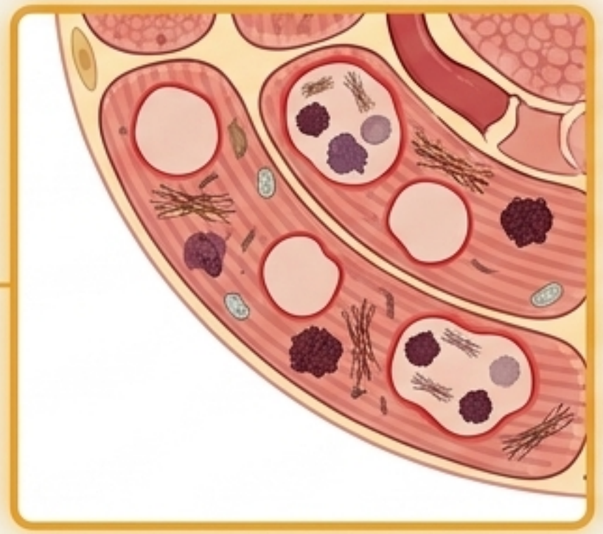
Dermatomyositis:
Complement-driven microangiopathy attacking intramuscular blood vessels and skin (Perifascicular atrophy).

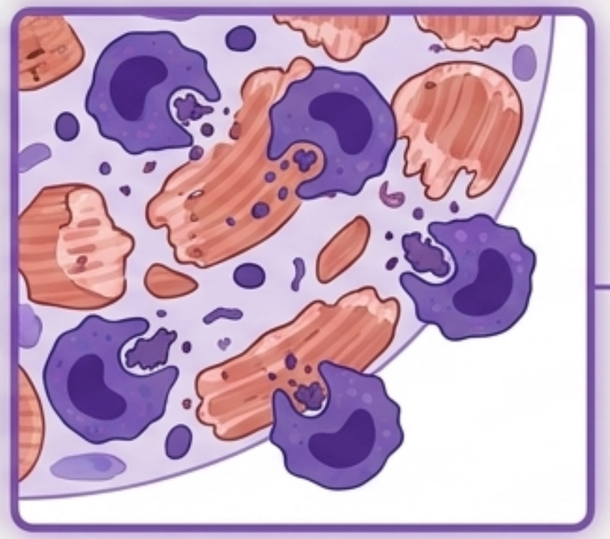




Polymyositis:
Direct CD8+ cytotoxic T-cell attack invading non-necrotic muscle fibres.

Inclusion Body Myositis:
Inflammatory AND degenerative pathology (rimmed vacuoles, amyloid deposits, TDP-43





IMNM:
Prominent myofibre necrosis and macrophage clearing, with minimal inflammatory infiltrate.

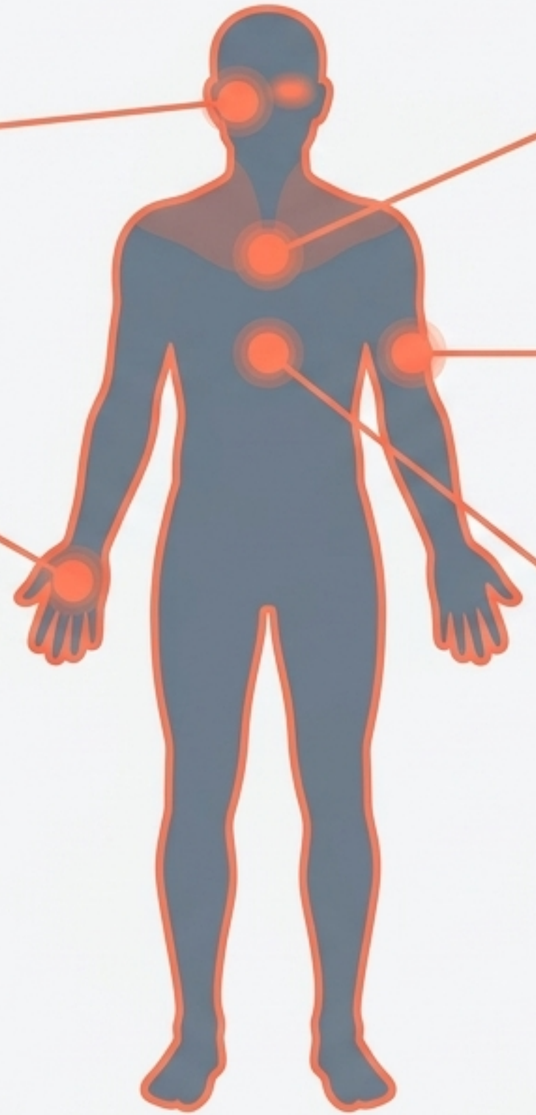
THE BIG FOUR: DIAGNOSTIC MATRIX

	DERMATOMYOSITIS (DM)	POLYMYOSITIS (PM)	INCLUSION BODY MYOSITIS (IBM)	IMNM
Pathophysiology Target	• Blood vessels (microangiopathy)	• Muscle fibers (direct attack)	• Degenerative + Inflammatory	• Severe myofibre necrosis
Clinical Presentation	• Proximal weakness + Distinctive skin rashes	• Subacute proximal weakness (No skin findings)	• Asymmetric, Distal focus (Finger flexors, Quadriceps)	• Severe proximal weakness
CK Levels	• Very High (10–50x ULN)	• Very High (10–50x ULN)	• Normal to Mildly Elevated (<15x ULN)	• Extremely High (Often >3000 U/L)
Biopsy Hallmark	• Perifascicular atrophy	• Endomysial CD8+ infiltrate	• Rimmed vacuoles	• Necrosis, minimal inflammation
Treatment Responsiveness	• High (Aggressive immunosuppression)	• High (Aggressive immunosuppression)	• Largely Refractory (Focus on supportive care)	• High (Combination immunosuppression often required)

DERMATOMYOSITIS: CLINICAL TOPOGRAPHY

Heliotrope rash (violaceous periorbital oedema).

Gottron papules (erythematous papules over MCP/PIP/DIP joints), **periungual erythema, mechanic's hands.**



Shawl sign, V-sign.

Symmetrical, progressive proximal weakness.

Dysphagia (20-30%) and **Interstitial Lung Disease** (ILD).

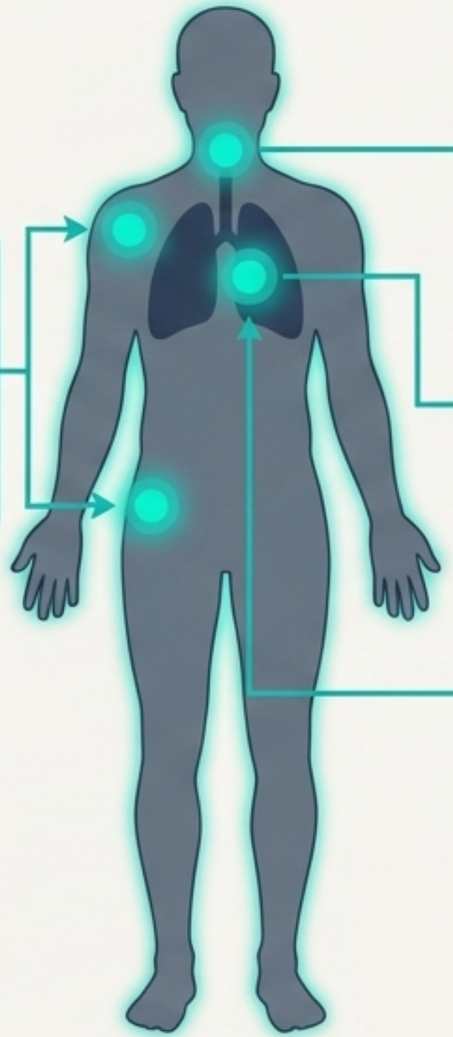
Cancer Risk: 4–7× increased malignancy risk in adult-onset DM, highest in first 3 years. Mandatory screening required.

TREATMENT SUMMARY

Systemic immunosuppression + strict sun protection (SPF 50+) + topical corticosteroids + HCQ for refractory skin.

POLYMYOSITIS: CLINICAL TOPOGRAPHY & DIAGNOSTIC CRITERIA

Symmetrical proximal weakn developing subacutely over weeks/months.



Symmetrical proximal weakness developing subacutely over weeks/months.

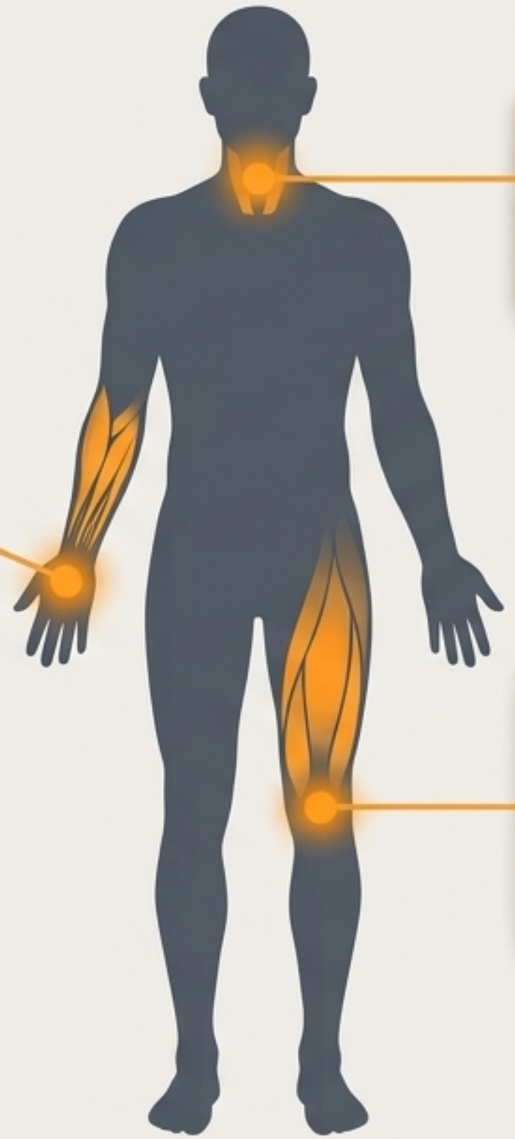
Dysphagia in 30–40% (often a presenting feature).

Cardiac involvement (10–30%) and potential overlap with ILD.

DIAGNOSTIC CRITERIA	
☒	Proximal weakness
☒	Elevated CK
☒	Myopathic EMG
☒	Biopsy: Endomysial CD8+ T-cells
☒	**Crucial:** Absence of skin findings.

Exclusion Mimics: Exclude IBM, muscular dystrophy, drug/toxin myopathy, and endocrine/metabolic myopathies before confirming PM.

INCLUSION BODY MYOSITIS: CLINICAL TOPOGRAPHY



Severe dysphagia (up to 60%, severe aspiration risk).

Finger flexor weakness (flexor digitorum profundus). Cannot grip objects, turn keys, open jars.

Early quadriceps weakness. Knee extensor failure leading to falls.

TREATMENT RESISTANCE

Largely unresponsive to corticosteroids and traditional immunosuppression.
Do NOT pursue prolonged high-dose therapy.

MANAGEMENT FOCUS

- Supervised progressive resistance training
- Fall prevention (home modifications)
- Speech pathology / modified diets
- IVIg (modest benefit for swallowing only)

THE IBM DIAGNOSTIC TRAP: REDIRECTING THE CLINICAL PATHWAY

Elderly male presents with proximal weakness + mildly elevated CK (<15x ULN).

Halt immunosuppression → Biopsy reveals rimmed vacuoles → **CORRECT DIAGNOSIS: INCLUSION BODY MYOSITIS** → Shift to supportive care, physiotherapy, and dysphagia management.

RECOGNIZE THE RED FLAGS:
Marked asymmetry, profound finger flexor weakness, knee buckling/ falls, prominent dysphagia.

Misdiagnosed as Polymyositis → Prescribed high-dose corticosteroids → **Result:** No clinical improvement, prolonged steroid toxicity.

IMMUNE-MEDIATED NECROTISING MYOPATHY (IMNM)

ANTI-HMGCR MYOPATHY

Trigger: Strongly associated with statin exposure (atorvastatin, rosuvastatin), though can be statin-naïve in youth.

Profile: Severe proximal weakness, CK often >3,000 U/L. Weakness persists after statin cessation.

ANTI-SRP MYOPATHY

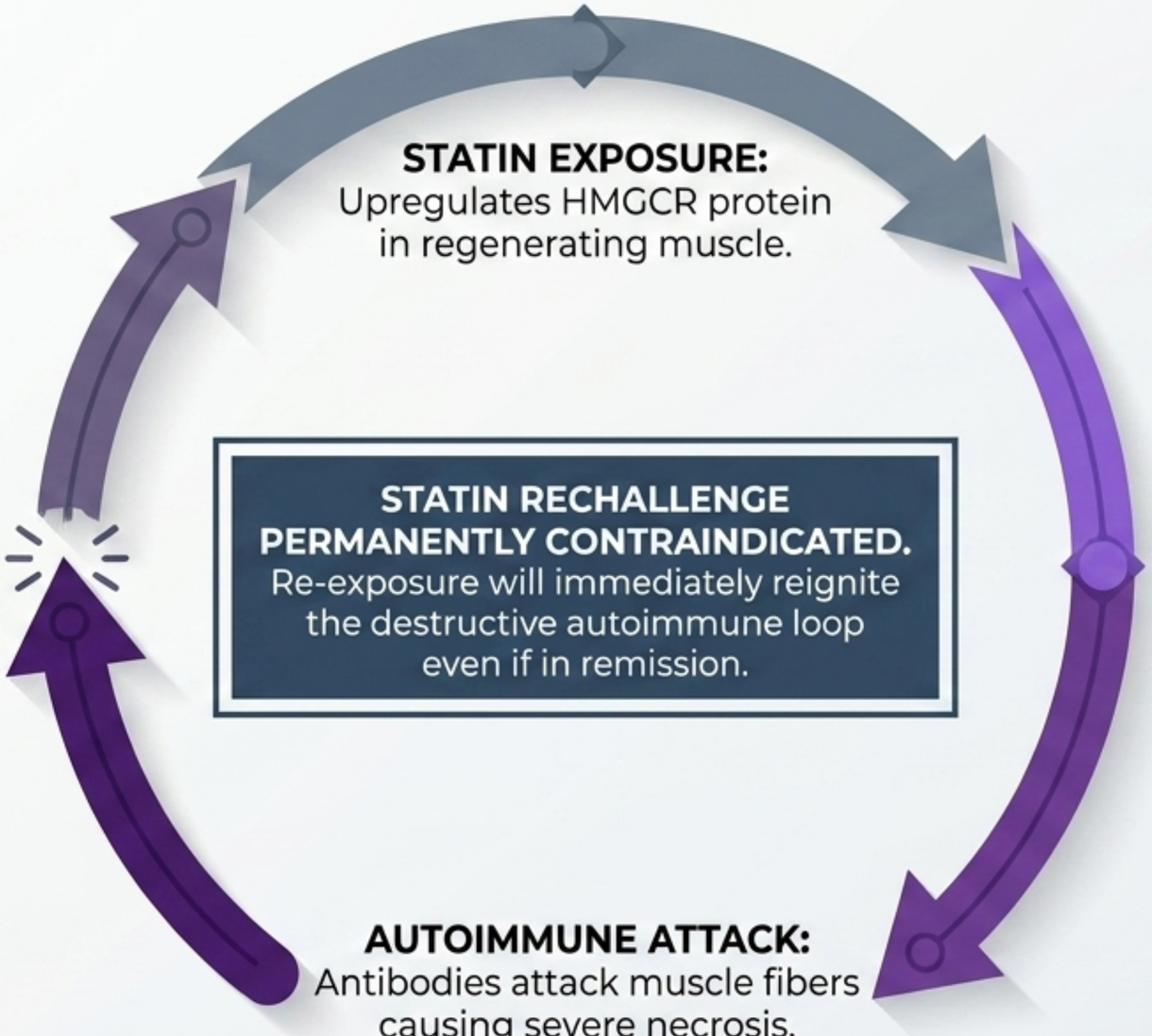
Trigger: Not associated with statins.

Profile: Rapidly progressive severe weakness, CK often >5,000 U/L. Higher rates of cardiac involvement (requires baseline ECG/Echo). Highly treatment-resistant.

Shared Baseline: Extreme CK Elevation

Requires aggressive immunosuppression (often combination: Prednisolone + Steroid-sparing + IVIg/Rituximab).

THE STATIN-IMNM FEEDBACK LOOP

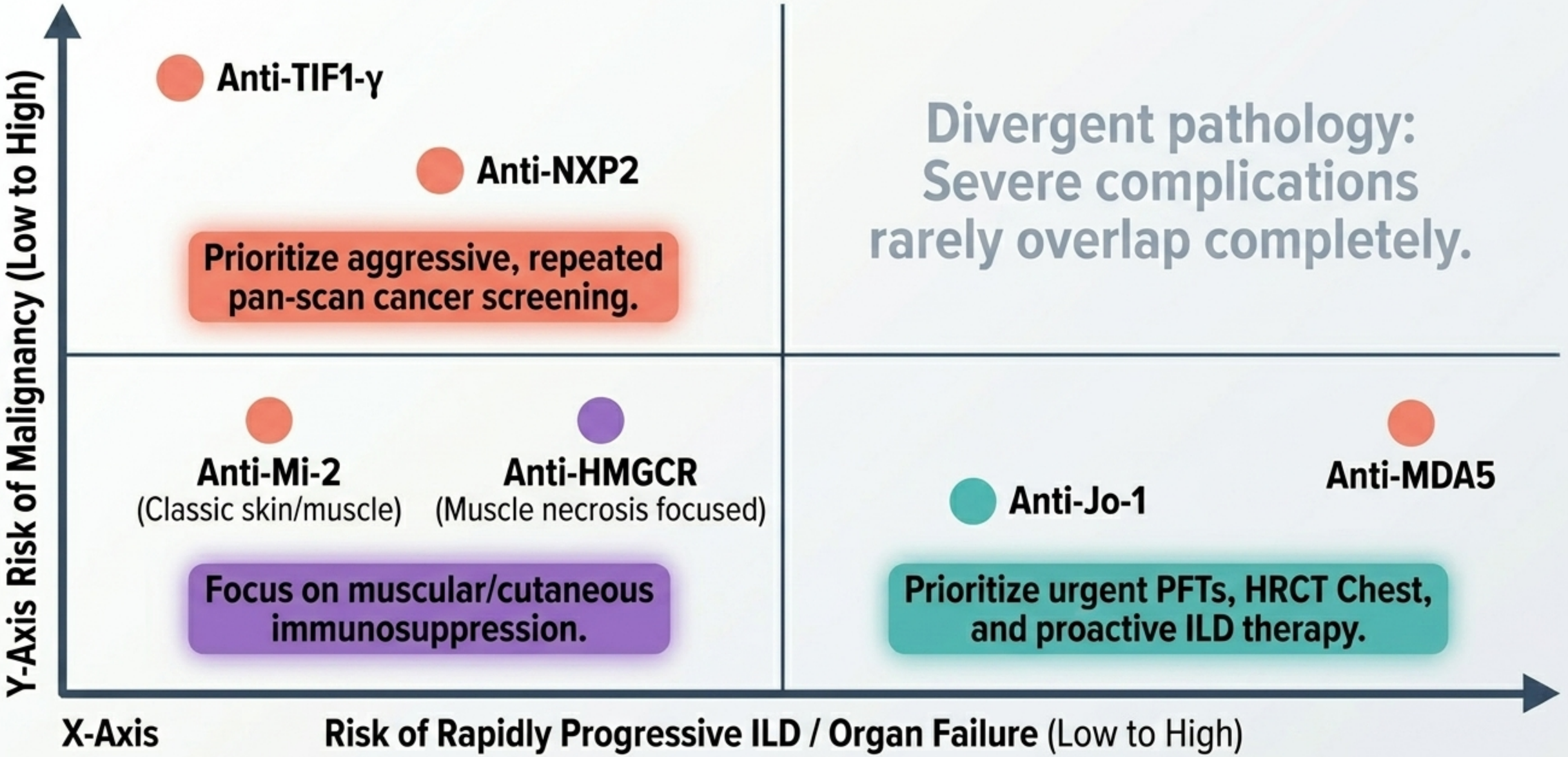


The MSA Toolkit: Myositis-Specific Antibodies

Antibody	Subtype Association	Key Clinical Phenotype	Cancer Risk
Anti-TIF1-γ	Adult DM	Prominent skin	High Cancer Risk (~80%)
Anti-NXP2	Adult/JDM	Calcinosis, muscle oedema	Moderate-High Cancer Risk
Anti-Mi-2	Classic DM	Good treatment response	Low Cancer Risk
Anti-SAE	Adult DM	Dysphagia, skin precedes myopathy	Moderate Cancer Risk
Anti-MDA5	Amyopathic DM	Rapid ILD, skin ulcers	Low Cancer (High Lung Risk)
Anti-Jo-1	Antisynthetase syndrome	ILD, arthritis, mechanic's hands	Low Cancer Risk
Anti-SRP	IMNM	Severe necrotising, cardiac risk	Low Cancer Risk
Anti-HMGCR	IMNM	Statin-associated necrotising	Low Cancer Risk

Testing Note: Tested via ALBIA/Euroline at major AU reference labs (MBS item via extended serology; specialist referral required).

PHENOTYPE RISK STRATIFICATION MAP



The Diagnostic Toolkit

BIOMARKERS

- **CK & Aldolase:** Serial monitoring guides treatment (CK MBS 66516). Aldolase elevated even if CK normal (amyopathic DM, IBM).
- **MSA Panel**: Guides phenotype (Specialist referral required).

IMAGING

- **Muscle MRI (MBS 63011):** T2-STIR for active oedema (inflammation), T1 for fatty replacement. Crucial for guiding biopsy site.
- **HRCT Chest & Echocardiogram**: Baseline organ screening.

FUNCTIONAL

- **EMG (MBS 11704):** Myopathic pattern (short-duration, polyphasic). Differentiates neuropathic causes.
- **PFTs:** FVC/DLCO for ILD screening.

PATHOLOGY & GENETICS

- **Muscle Biopsy**: Gold standard. Directs the primary subtype diagnosis.
- **Pharmacogenomics (MBS 73300):** TPMT/NUDT15 genotype testing prior to Azathioprine to prevent severe myelosuppression.

RISK STRATIFICATION & SEVERITY ASSESSMENT

SEVERE

CK >15× ULN. Bed-bound, severe aspiration/dysphagia, rapidly progressive ILD (anti-MDA5), cardiac involvement, Anti-SRP IMNM.

Setting:
Inpatient Tertiary/ICU.
Early IVIg/Rituximab.

MODERATE

CK 5–15× ULN. Difficulty with ADLs (stairs/dressing), mild dysphagia, mild ILD (FVC >70%), Antisynthetase syndrome.

Setting:
Outpatient MDT with close monitoring

MILD

CK <5× ULN. Normal ADLs, no dysphagia, no ILD. Skin-only (amyopathic DM).

Setting:
Outpatient rheumatology

VALIDATED TOOLS

MDAAT
(Disease activity)

MMT-8
(Muscle strength)

CMAS
(Paeds/Adults function)

HAQ-DI
(Patient-reported)

The Stepped Therapy Ladder (DM, PM, IMNM)

FIRST-LINE INDUCTION

Prednisolone

- 1 mg/kg/day PO (max 60mg). Taper aim: steroid-free remission in 12-24 mos.

FIRST-LINE STEROID SPARING

Methotrexate OR Azathioprine

- **MTX:** 7.5-25mg weekly + Folic acid.
- **AZA:** 2mg/kg/day.

SECOND-LINE / ILD FOCUS

Mycophenolate Mofetil OR Tacrolimus

- **MMF:** Authority required.
- **Tacrolimus:** Target trough 5-10 ng/mL, off-label PBS for myositis.

REFRACTORY / SEVERE ESCALATION

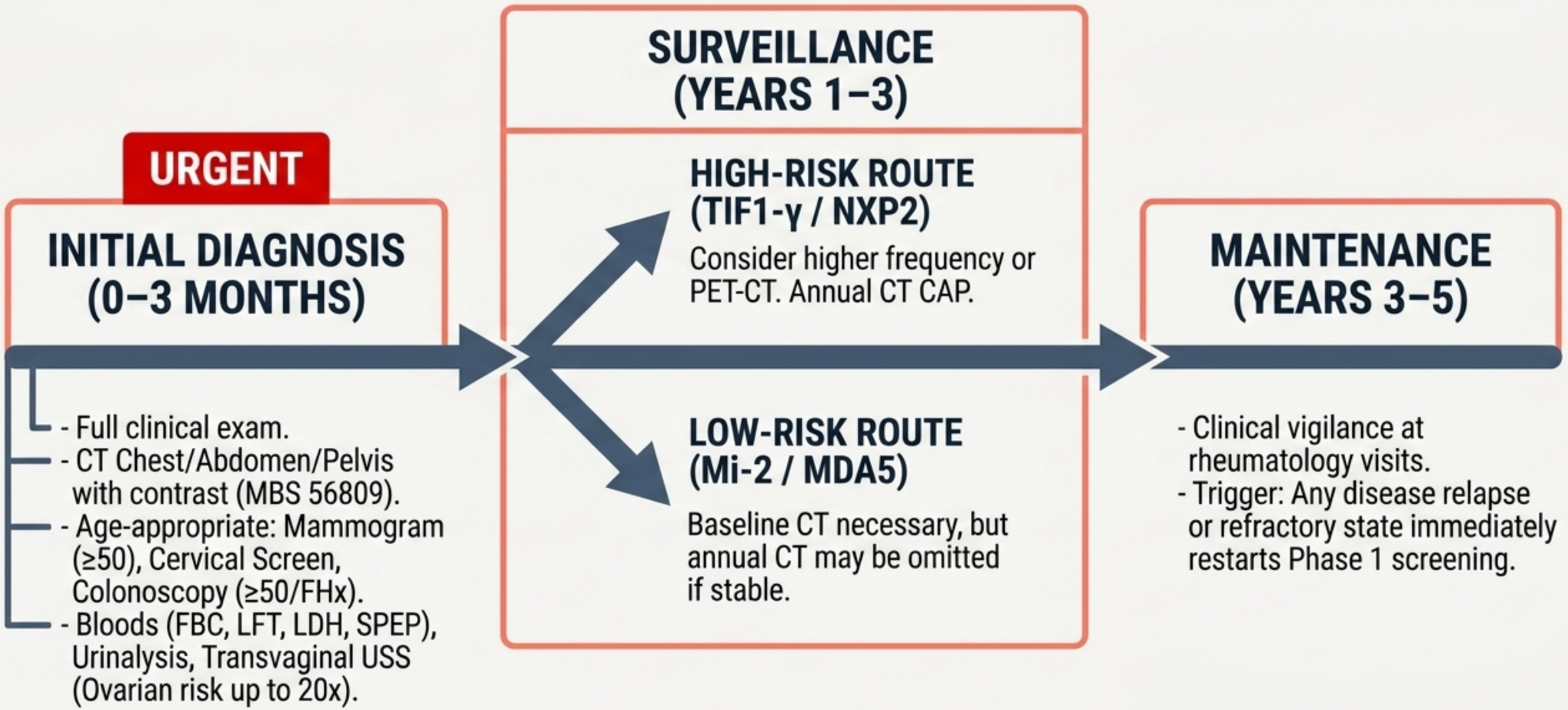
IVIg OR Rituximab

- **IVIg:** 2g/kg over 2-5 days (PBS Specialist Drug Program).
- **Rituximab:** Off-label, PBAC exceptional circumstances.

WARNING: MTX CI with Trimethoprim.

Check TPMT before AZA; reduce AZA by 75% with Allopurinol.

ADULT DM CANCER SCREENING TEMPORAL PROTOCOL



THE CLINICAL MONITORING PROTOCOL

Track A: Induction Phase (First 3 Months)

Cadence: Every 2–4 Weeks

Bloods:
CK, aldolase, CRP, ESR, FBC, LFTs, U&E.

Med Safety:
MTX bloods (fortnightly), AZA bloods (weekly FBC for 1st month).

Clinical:
Steroid side effects (BP, BGL, weight), MMT-8, MDAAT.

Track B: Maintenance Phase (Months 3+)

Cadence: Every 1–3 Months+

Routine:
1-3 monthly CK, anti-HMGCR titres (if IMNM).

Annual:
DEXA scan (corticosteroid bone protection + Calcium/Vit D).

Annual:
Ophthalmology review (if on HCQ for skin disease).

6–12 Monthly:
PFTs (FVC/DLCO) for any ILD involvement.

SPECIAL POPULATIONS TOOLKIT

PREGNANCY

Teratogenic/AVOID: Methotrexate, Mycophenolate (stop ≥3 mos prior).

Safe to continue: Azathioprine, Hydroxychloroquine, Prednisolone, IVIg.

PAEDIATRIC / JDM

Anti-NXP2 highly associated with hard-to-treat **calcinosis**.

First-line: Prednisolone + early MTX.

Note: Cancer screening not routinely indicated.

ELDERLY (≥65 YEARS)

IBM is top diagnostic differential for isolated quad/swallowing weakness.

High risk of **corticosteroid morbidity** (diabetes, osteoporosis) requires proactive DEXA and rapid steroid-sparing.

RENAL/HEPATIC & IMMUNE

MTX contraindicated if eGFR <30 or severe hepatic disease.

Sucrose-IVIg contraindicated in renal failure.

PJP prophylaxis required on heavy immunosuppression.

ABORIGINAL AND TORRES STRAIT ISLANDER HEALTH CONSIDERRATIONS

Barrier: Specialist Access	**Solution:** Proactive use of Telehealth (MBS 91822) and facilitation of Patient-Assisted Travel Schemes (PATS).
Barrier: Delayed Presentation / Diagnostic Masking	**Solution:** Education on recognizing cutaneous signs (heliotrope/Gottron) in darker skin tones; avoiding misattribution of proximal weakness to fatigue.
Barrier: High Comorbidity Burden	**Solution:** High baseline rates of diabetes/CKD make prolonged corticosteroids high-risk. Mandates early steroid-sparing agents and pre-MTX renal screening.
Barrier: Complex Regimens & Screening	**Solution:** Leverage Webster-paks, PBS Safety Net, and close liaison with Aboriginal Medical Services (AMS) to ensure culturally safe, community-supported screening.