

Living with Myositis

A Practical, Evidence-Based Guide to Inflammatory Myopathy, Its Treatment, and Daily Life



Written by Claude Code | Instructed by V.Korostyshevskiy

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HOW TO USE THIS BOOK

This book informs. It does not prescribe.

That distinction sounds small. It is not. A book that prescribes tells you what to take, when to change a dose, whether a symptom is serious. This book does none of those things. Your rheumatology team holds the clinical decisions, and every chapter in this guide is written to feed information toward that team, not to replace it. What you will find here is a structured, evidence-grounded account of what idiopathic inflammatory myopathy is, what its treatments do and what they cost the body, how to move and eat well while managing both the disease and its therapy, and what to watch for in the organ systems that myositis can reach. Where the evidence is strong, the writing says so plainly. Where it is thin, the writing says that too, in a different voice, flatter and more cautious, so you can feel the difference without needing a statistics degree.

This book is organized around four things a person with myositis needs to understand: what the disease is and how its subtypes shape your risk profile; what treatment does and what it costs the body; how to move, eat, and function well while managing both disease and therapy; and when the care team needs to hear from you. Each chapter serves one of those functions. Together, they form a complete picture of informed self-management inside a specialist-supervised treatment plan. The book informs and routes. It never prescribes.

The Escalation Vocabulary

Every chapter that raises a symptom, an organ risk, or a treatment concern resolves into one of three tiers. These are not decorative labels. They are the action vocabulary of the book, and once you know them, you know what every clinical section is asking you to do.

Routine means the topic is worth raising at your next scheduled rheumatology appointment. There is no urgency, but there is value in the conversation.

Prompt means something has changed and you should contact your rheumatology team within days, not wait for a scheduled visit.

Urgent means you should seek same-day medical attention or go to the emergency department. The short list below covers the situations that warrant it.

When to Seek Help Now

Three categories of events recur across the clinical literature on inflammatory myopathy as situations requiring immediate medical contact.^[1,2,3] They appear here, before the detail chapters, so that the safety-critical information is the first thing you read.

New or worsening breathlessness. Interstitial lung disease is the organ complication most closely linked to mortality in inflammatory myopathy. In certain antibody-defined subgroups, rapidly progressive lung disease can develop over weeks, with mortality among those who develop the most aggressive form that the clinical literature describes as high.^[1] Any new shortness of breath on exertion, any persistent dry cough that was not there before, or any sudden decline in exercise tolerance that is out of proportion to muscle weakness alone is an Urgent signal. Do not wait for your next appointment.

Choking or aspiration on eating or drinking. Swallowing difficulty affects a substantial proportion of people with myositis, and aspiration pneumonia is identified in published reviews as a leading cause of death in this disease.^[2] Coughing or choking during meals, the sensation of food sticking in the throat, or a wet, gurgling voice quality after eating are signals to report. Recurrent aspiration or aspiration pneumonia is Urgent.

Signs of serious infection while on immunosuppression. The medications that control inflammatory myopathy suppress the immune system, and that suppression carries a measurable cost. Certain opportunistic infections, including pneumocystis pneumonia, occur at rates far above the general population in people on combined corticosteroid and immunosuppressant therapy.^[3,4] Fever, chills, a new cough with breathlessness, or any sign of rapidly worsening infection while you are on immunosuppressive treatment is Urgent. Contact your team or present to the emergency department.

These three categories are explored in full in later chapters (treatment side effects in Chapter 4; organ-system awareness in Chapter 7). The detail will make them

more concrete. But the action is simple and does not require the detail: if something on this list is happening, call now.

How to Read the Evidence in This Book

You will notice that the writing shifts register. Some sections carry momentum; they develop a finding, build an analogy, let you feel the weight of solid evidence. Other sections go flat and plain. That shift is deliberate. When a claim rests on society guidelines, replicated trials, or large cohort studies, the prose reflects that confidence. When a claim rests on a single small study, expert opinion, or something you are likely to encounter online without good support behind it, the prose pulls back, names what is missing, and delivers a short verdict. You do not need to check endnotes to sense the difference, though the endnotes are there if you want them.

Every specific clinical or empirical claim in this book carries a numbered superscript that points to an endnote in Appendix G. The endnotes cite primary sources: published studies, society guidelines, and official regulatory documents. Where a webinar, patient-organization resource, or commercially sponsored talk is mentioned, it is labeled as a further-listening pointer and listed separately from the evidence citations. The distinction matters. A guideline recommendation and a pharmaceutical company's promotional webinar are not the same kind of source, and this book does not treat them as if they are.

How This Book Was Written

This guide was produced by a multi-agent pipeline built in Claude Code. Each chapter was drafted by one writing agent drawing only from a defined set of source files. A separate grounding agent verified that every claim traces to its source. A fact-trace agent checked that stated figures match the cited studies. Two independent review agents examined voice, accuracy, and structural compliance. A deduplication agent ensured that no fact is re-sourced across chapters beyond its assigned home. An editor agent performed a final pass. The result is not a book written by a person recalling what he read; it is a book in which every empirical claim was traced, checked, and cross-examined by independent processes before assembly.

That does not make the book infallible. It makes it transparent. The endnotes are there so you can check. The pipeline is there so the checking happened before you opened the cover.

CHAPTER 1: UNDERSTANDING MYOSITIS AND YOUR DIAGNOSIS

In this chapter, you will learn what idiopathic inflammatory myopathy actually is, how the major autoimmune subtypes form an orienting map rather than a self-diagnosis, and why the boundary between autoimmune IIM and its mimics matters for every treatment decision that follows. You will also learn why inclusion body myositis is a separate category, and why this book sets it aside.

Myositis does not behave the way most people expect a muscle disease to behave. It does not confine itself to muscle. The British Society for Rheumatology describes idiopathic inflammatory myopathy as “a multi-system autoimmune condition characterised by muscle inflammation (myositis), interstitial lung disease (ILD), and skin manifestations.”^[1] That word “multi-system” is the first thing worth understanding, because it shapes everything else: the investigations your team orders, the organs they monitor, and the reasons they sometimes seem more interested in your lungs and skin than in your arms.

The second thing worth understanding is that the disease you have is rare. In a contemporary European cohort tracked across Northern Spain from 2000 to 2022, the incidence was approximately 0.9 cases per 100,000 person-years, with a significant upward trend of roughly 5.7% annually.^[2] That upward trend almost certainly reflects better recognition, not a worsening epidemic. Clinicians now have antibody panels and classification tools that did not exist twenty years ago, and cases that once would have been labelled “unexplained weakness” now receive a specific diagnosis. But rarity shapes experience. You may be the only person your GP has ever treated with this condition. You may struggle to explain it to family. That is normal, and this book exists in part to give you the vocabulary.

This chapter provides orientation. It does not assign you a subtype. Your specific diagnosis, your antibody result, and the clinical implications of both belong with your rheumatology team. What follows is a map so that when your team discusses these categories, you recognise the landscape.

The Disease Your Immune System Built

Idiopathic inflammatory myopathy (IIM) is a group of acquired, immune-mediated muscle diseases characterised by skeletal muscle inflammation, elevated serum muscle enzymes, and variable extramuscular organ involvement.^[3] “Idiopathic” means no single external trigger fully explains the process. No infection, toxin, or inherited defect accounts for it. The immune system appears to attack muscle tissue through autoimmune mechanisms, and the damage cascades outward.

Think of it like a building’s security system malfunctioning. Instead of repelling intruders, the sensors start treating the building’s own wiring as a threat. The alarm stays on. The damage accumulates. And because the security system runs through the entire building, the malfunction can reach rooms far from where it started: skin, lungs, joints, the oesophagus. That multi-system reach is what makes IIM more than a muscle disease, and it is why your care team includes specialists who look well beyond the muscles themselves.

IIM occurs roughly twice as often in women as in men overall.^[2] But that ratio is not uniform across subtypes. One subtype, immune-mediated necrotizing myopathy, presents at a mean age of approximately 64 and affects men at a notable proportion (approximately one-third of cases in one cohort).^[2] Another, antisynthetase syndrome, clusters differently again. These demographic patterns matter less for self-comparison than for understanding why your rheumatologist thinks in categories. The subtypes are not just academic labels. They predict organ risks, treatment responses, and monitoring schedules.

The IIM Subtype Map

The named concept that orients this entire book is **the IIM subtype map**. It is not a diagnostic flowchart and it is not something you use to classify yourself. It is a reference frame: a way to understand why different people with “myositis” can have strikingly different organ risks, treatment responses, and long-term trajectories.

The major autoimmune subtypes, as classified by the 2017 EULAR/ACR criteria and the German DGN S2k guideline, are dermatomyositis, immune-mediated necrotizing myopathy, antisynthetase syndrome, polymyositis, and overlap myositis.^[3,4] Each

occupies a distinct position on the map. Your team uses your antibody results, biopsy findings, and clinical features to locate you on it. Your position on the map may determine which organs are monitored most closely, which treatments are tried first, and which screening tests are recommended.

DERMATOMYOSITIS

Dermatomyositis (DM) is defined by pathognomonic skin signs alongside variable muscle involvement. The hallmark rashes carry clinical names you may hear in appointments: **Gotttron's papules** (violaceous raised patches over the knuckles), **Gotttron's sign** (macular erythema over extensor joint surfaces), and **heliotrope rash** (violaceous discolouration around the eyes, sometimes with eyelid swelling).^[3,5] Skin findings may precede weakness by months to years, which is why some patients receive a DM diagnosis before they notice any loss of strength.

Approximately 80% of DM patients develop clinical muscle weakness with proximal distribution.^[6] The remaining 20% develop **amyopathic dermatomyositis**, in which the characteristic skin disease occurs without objective muscle weakness.^[6] This is not a mild variant. Amyopathic DM still carries significant organ risks, including interstitial lung disease, which affects approximately 15 to 30 percent of DM patients overall and is a leading cause of morbidity and mortality in the subtype.^[6]

DM also carries a demonstrated risk of occult malignancy, particularly when associated with certain antibodies.^[4] Malignancy screening at diagnosis is a guideline recommendation that your team manages based on your specific antibody profile (more on screening in Chapter 7). DM was the subtype studied in the ProDERM trial, which led to the first and only FDA-approved IVIG specifically indicated for adult dermatomyositis.^[7] That approval is a landmark for the field, and its details belong in Chapter 3.

IMMUNE-MEDIATED NECROTIZING MYOPATHY

Immune-mediated necrotizing myopathy (IMNM) is a more recently recognized entity. The German DGN S2k guideline describes its formal introduction as “a key innovation in classification,” establishing it as an independent category distinguished by specific marker antibodies: **anti-SRP** and **anti-HMGCR**.^[4]

The defining feature under the microscope is muscle fibre necrosis and regeneration without the significant inflammatory infiltrate that characterises

classical inflammatory myopathies.^[8] This histological distinction is clinically important: it means IMNM can look like other forms of myositis on clinical examination but reveals a fundamentally different pattern when the tissue is studied. Roughly two-thirds of IMNM patients are positive for one of those two antibodies.^[8]

The anti-HMGCR antibody deserves particular attention because it intersects with one of the most widely prescribed drug classes in medicine. Anti-HMGCR is associated with statin exposure in 40 to 60 percent of cases, rising to 92% in patients over 50.^[8] Here is a distinction that matters: anti-HMGCR myopathy does not resolve when you stop the statin. This is not simple drug toxicity. The statin may have triggered an autoimmune process that persists independently and requires immunosuppressive treatment.^[8] This distinction alone justifies the existence of the antibody panel, because the answer determines whether a patient needs ongoing immunotherapy or merely needs to switch medications.

Anti-SRP-positive IMNM, by contrast, typically presents with more severe and rapidly progressive proximal weakness. It carries higher risks of cardiac involvement, ILD, and swallowing difficulty compared to anti-HMGCR-positive disease, which more commonly presents as isolated proximal weakness without major extramuscular involvement.^[8]

In contemporary cohorts, IMNM accounts for approximately 14 to 15 percent of adult IIM.^[2]

ANTISYNTHETASE SYNDROME

Antisynthetase syndrome (ASyS) is distinguished by antibodies against aminoacyl-tRNA synthetase enzymes, the most common being **anti-Jo-1**, found in 9 to 24 percent of adult IIM patients.^[9] Eight or more antisynthetase antibodies have been identified, each defining a slightly different clinical profile, but they share a core constellation.^[9]

The classical triad of ASyS is inflammatory myopathy, interstitial lung disease, and non-erosive arthritis.^[10] Additional features include **Raynaud's phenomenon** (colour changes in the fingers triggered by cold or stress), **mechanic's hands** (roughened, cracked skin on the lateral and palmar surfaces of the fingers), and unexplained

fever. Not all features need to be present at the same time, and the syndrome can declare itself gradually over months or years.

One number defines this subtype's clinical gravity. Interstitial lung disease occurs in 70 to 95 percent of ASyS patients.^[9] ILD is the primary driver of morbidity and mortality in antisynthetase syndrome. That statistic explains why, if you carry an antisynthetase antibody, your team monitors your lungs with particular attention. Anti-Jo-1 antibody titers also correlate with disease activity and strongly predict response to rituximab, giving this marker both diagnostic and therapeutic significance.^[9]

ASyS accounts for approximately 24 to 28 percent of adult IIM in contemporary cohorts, making it the most common subtype in some registries.^[2] That proportion has increased as modern antibody testing reclassifies cases that were historically labelled polymyositis.

POLYMYOSITIS

Polymyositis (PM) has historically been diagnosed as symmetrical proximal muscle weakness without DM skin signs and without the specific histological features of IBM or IMNM.^[11] The category is now controversial. Re-analysis of historical PM cohorts using modern antibody testing has shown that the majority of cases previously labelled PM are reclassifiable as IMNM, antisynthetase syndrome, or overlap myositis.^[12] In one Korean cohort, 77.8% of historical PM cases were reclassified on antibody-integrated review.^[12]

The DGN S2k guideline retains PM as a classification category alongside the other subtypes, consistent with the 2017 EULAR/ACR framework.^[4] In contemporary epidemiological cohorts, PM accounts for roughly 5% of adult IIM cases.^[2] If your team has given you a PM diagnosis, it is worth asking whether modern antibody testing has been performed. The answer may refine your position on the subtype map and alter your organ-surveillance schedule. This is a question for your next Routine appointment.

OVERLAP MYOSITIS

Overlap myositis refers to IIM occurring alongside another systemic connective tissue disease: systemic sclerosis, lupus, Sjogren's syndrome, or rheumatoid arthritis.^[13] It is characterised by **myositis-associated antibodies** (MAAs), which are

distinct from the myositis-specific antibodies that define the other subtypes. Key MAAs include **anti-PM/Scl** (associated with systemic sclerosis overlap), **anti-Ku** (associated with lupus, Sjogren's, and systemic sclerosis overlap), and **anti-U1snRNP** (the serological hallmark of mixed connective tissue disease).^[13]

Overlap cases frequently carry elevated lung disease risk, particularly with anti-PM/Scl and anti-Ku antibodies, and require integrated assessment of both the muscle disease and the co-existing autoimmune condition.^[13] The DGN S2k guideline lists overlap myositis as a separate category within the IIM classification.^[4] If your diagnosis includes "overlap" or an additional autoimmune condition alongside your myositis, the management of both diseases needs to be coordinated by your team.

How the Map Was Drawn: The 2017 Classification Criteria

The subtype map rests on a formal classification system. The 2017 EULAR/ACR criteria replaced the 1975 Bohan and Peter criteria that had served (and limited) the field for four decades.^[3] They were developed by the International Myositis Classification Criteria Project using data from 976 IIM patients and 624 comparators across 47 centres in 30 countries.^[3] That scale matters because IIM is rare; assembling nearly a thousand cases required international collaboration.

The criteria use a probability-based scoring system incorporating 16 weighted variables across six domains: age of onset, muscle weakness pattern, skin manifestations, laboratory findings (including muscle enzymes and anti-Jo-1), muscle biopsy features, and electromyography.^[14] The system produces a probability score, not a binary yes-or-no. A score of 5.5 to less than 7.5 (without biopsy) indicates probable IIM; 7.5 or above indicates definite IIM.^[14]

The pathognomonic DM skin signs carry the highest weights. Gottron's sign scores 3.3 to 3.7 points; heliotrope rash scores 3.1 to 3.2 points.^[14] Anti-Jo-1 antibody carries 3.8 to 3.9 points, the highest weighting for any single autoantibody in the criteria.^[14] That single antibody result can carry the classification almost halfway to the definite-IIM threshold on its own. Rimmed vacuoles on biopsy carry 3.1 points and, critically, redirect classification toward IBM rather than autoimmune IIM.^[14]

Without muscle biopsy, the criteria achieve 87% sensitivity and 82% specificity. With biopsy, those figures rise to 93% and 88% respectively.^[14] Agreement with expert physician diagnosis reached 92.6%, with a kappa of 0.90.^[14] These are strong numbers for a rare, heterogeneous disease. They mean the classification system aligns closely with what experienced specialists conclude after examining a patient. An online probability calculator exists for clinical use, though it is a tool for your team, not something to apply independently.^[14]

The Boundary: Autoimmune IIM Versus Its Mimics

Knowing what autoimmune IIM is requires knowing what it is not. Several conditions produce muscle weakness and elevated creatine kinase without being autoimmune IIM. The distinction matters because the treatments are fundamentally different: immunosuppression for autoimmune IIM, antimicrobials for infection, drug discontinuation for toxicity, and genetic or metabolic interventions for hereditary disease.

WHAT DEFINES AUTOIMMUNE IIM

A person sits inside the autoimmune IIM boundary when they have acquired, subacute-to-chronic proximal weakness (or characteristic DM skin disease); elevated muscle enzymes or biopsy confirmation; positive myositis-specific antibodies or clinical features matching a recognised subtype; no evidence of an alternative cause (infection, hereditary myopathy, drug toxicity without autoimmune perpetuation); and a disease course that responds to immunosuppressive or immunomodulatory treatment, including IVIG.^[3,1,4]

If you are receiving ongoing IVIG maintenance therapy under specialist care, with a diagnosis made by a rheumatologist or neurologist, and IBM and infectious causes have been excluded, you sit inside this boundary. The diagnostic workup you underwent was designed to confirm this.

INFECTIOUS MYOSITIS

Infectious myositis is caused by direct microbial invasion or pathogen-triggered immune damage. Causative organisms include influenza, Cocksackievirus B, Staphylococcus aureus, and others.^[15] The mimicry can be convincing. In one

documented case, Coxsackievirus B4/B5-associated myositis appeared indistinguishable from polymyositis on standard clinical assessment and histology. Electron microscopy was required to visualise the 30 to 31 nanometre viral particles within the muscle tissue.^[16] The patient recovered full strength within one year on supportive care alone, without immunosuppression.^[16]

That outcome is the distinguishing signal. Infectious myositis resolves with antimicrobial therapy or supportive care. It has an identifiable pathogen, a temporal association with acute illness, and focal bacterial forms may show abscess formation on imaging rather than the diffuse pattern of autoimmune IIM.^[15] Autoimmune IIM does not clear on its own. It persists.

DRUG-INDUCED MYOPATHY

Drug-induced toxic myopathy (from statins, corticosteroids, hydroxychloroquine, colchicine, or alcohol) also mimics IIM.^[17] In simple drug toxicity, creatine kinase normalises and weakness resolves when the drug is stopped. The mechanism is direct cellular injury, not an immune attack.

But there is a critical exception: anti-HMGCR IMNM can be statin-associated yet persist indefinitely after statin discontinuation because an autoimmune process has been triggered.^[8,17] The statin was the match, but the fire is self-sustaining. Anti-HMGCR antibody testing resolves the ambiguity.^[8] This is one reason the antibody panel matters: it separates a condition that stops when the drug stops from one that requires ongoing immunotherapy.

MUSCULAR DYSTROPHIES AND METABOLIC MYOPATHIES

Hereditary muscular dystrophies (Duchenne, Becker, limb-girdle, facioscapulohumeral, and others) can present with elevated CK and proximal weakness resembling IIM. Distinguishing features include family history of myopathy, absence of inflammatory markers, negative MSA panel, genetic inheritance pattern, and biopsy findings showing dystrophic features (fatty infiltration, membrane abnormalities) rather than immune-cell infiltrates.^[18] An adult presenting with proximal weakness, elevated CK, and no response to immunosuppression warrants consideration of an underlying dystrophy as an alternative or concurrent diagnosis.^[18]

Metabolic myopathies (glycogen storage diseases such as McArdle disease and Pompe disease, lipid disorders, and mitochondrial conditions) tend to present with exercise-induced symptoms or episodic rhabdomyolysis rather than the persistent subacute weakness of IIM, and they respond to metabolic interventions rather than immunosuppression.^[19] Adult-onset Pompe disease deserves particular mention: it causes progressive limb-girdle and respiratory weakness in adults, CK is moderately elevated, and biopsy may show vacuolar changes that superficially resemble those of IBM. Enzyme assay and genetic testing confirm the diagnosis.^[19]

The mimic boundary is not academic. It explains why your diagnostic workup may have included tests that seemed unrelated to “muscle inflammation.” Each test was ruling out an alternative explanation so that your team could confirm the autoimmune diagnosis with confidence.

IBM: A Separate Category

Inclusion body myositis (IBM) shares the word “myositis” with your condition, but it follows a fundamentally different path.^[20] Understanding why matters, because this book is not about IBM, and the management strategies in the chapters that follow do not apply to it.

Three distinctions set IBM apart.

The clinical pattern is different. IBM presents with insidious, asymmetric weakness that preferentially targets the deep finger flexors (causing difficulty gripping, dropping objects) and the quadriceps (causing falls and difficulty rising from chairs).^[21] This is distinct from the symmetric proximal girdle weakness that characterises autoimmune IIM. The onset is typically slow and the distribution is specific.

The underlying pathology is different. IBM combines endomysial lymphocytic inflammation (resembling autoimmune myositis) with degenerative features unique to itself: **rimmed vacuoles**, protein aggregates (p62, TDP-43), and mitochondrial dysfunction.^[20] This dual pathology of inflammation plus neurodegeneration-like protein misfolding is the reason IBM responds differently to treatment. The inflammatory component is only half the story; the degenerative component does not yield to immunosuppression.

And that is the third distinction: treatment refractoriness. Unlike the autoimmune IIM subtypes that generally respond to corticosteroids, immunosuppressants, and IVIG, IBM does not show meaningful disease-modifying response to any of these therapies in controlled trials.^[22] IVIG may temporarily improve swallowing difficulty in some IBM patients, but it does not alter the overall disease course.^[22] The standard of care for IBM is supportive: exercise, physical therapy, adaptive equipment, and monitoring of swallowing and respiratory function.

The 2023 ENMC international workshop produced revised IBM diagnostic criteria using a two-step integrative approach combining clinical features, pathology, **anti-cN1A** antibody testing, and muscle imaging.^[21] Within the 2017 EULAR/ACR criteria, rimmed vacuoles on biopsy score 3.1 points and redirect classification toward IBM rather than autoimmune IIM.^[14] Biopsy substantially improves IBM classification accuracy, which is one reason muscle biopsy is especially important when IBM is in the differential.^[14]

For you, this means IBM is the condition your team has excluded. It is the reason some of the tests you underwent were performed. The rest of this book addresses the autoimmune, treatment-responsive IIM subtypes. IBM is set aside from here.

Malignancy Screening: A Clinical Action Point

One aspect of the subtype map has immediate practical implications. Certain IIM subtypes, particularly dermatomyositis, carry an elevated risk of occult malignancy.^[4] The DGN S2k guideline recommends that all IIM patients at diagnosis undergo screening including history (with attention to B-symptoms), detailed physical examination, imaging of the chest and abdomen, and gynaecological, urological, and gastroenterological examinations.^[4] For patients carrying specific high-risk antibodies (anti-TIF1-gamma, anti-NXP2, or anti-HMGCR), more intensive imaging with FDG-PET-CT or whole-body CT is recommended, with repeat within the first year and further surveillance over the following two years.^[4]

This is not something you manage on your own. It is a clinical action point to confirm with your rheumatology team: has the recommended screening been completed, and is your surveillance schedule current? The full details of cancer-associated myositis and the IMACS screening protocol are covered in Chapter 7. Raising the topic with your team is Routine.

Key Points

- Autoimmune IIM is a multi-system disease affecting muscle, lungs, and skin; understanding its scope shapes every monitoring decision your team makes.
- The IIM subtype map orients you to different risk profiles without diagnosing you; your position on it depends on antibody results and clinical findings your team holds.
- The 2017 EULAR/ACR criteria provide a validated, probability-based classification system that aligns with expert diagnosis in over 92% of cases.
- Autoimmune IIM is distinguished from its mimics (infections, drug toxicity, hereditary conditions) by its immune-mediated mechanism and response to immunotherapy.
- IBM shares the word “myositis” but follows a separate clinical path; the management guidance in this book does not apply to it.
- Malignancy screening at diagnosis is a guideline recommendation worth confirming with your team at a Routine appointment.

CHAPTER 2: DIAGNOSIS AND MONITORING: WHAT THE TESTS AND NUMBERS MEAN

In this chapter, you gain literacy about the investigations that built your diagnosis, the autoantibody panel that shapes your risk profile, creatine kinase as a personal monitoring reference, and malignancy screening as a clinical action point to raise with your team. Understanding these tools turns you from a passive recipient of results into someone who can read the trajectory of your own disease.

The subtype map from Chapter 1 has meaning only if you know where you sit on it. Most people living with myositis remember the moment they first saw their blood results and understood almost nothing. The numbers were abnormal. The antibody names were long. The implications were unclear. That gap between receiving a result and understanding what it means is not a personal failure. It is a design problem. The investigations that diagnose and monitor idiopathic inflammatory myopathy were built for clinicians, not for the person sitting across the desk. This chapter closes that gap. It does not turn you into your own doctor. It gives you what we will call **disease-activity literacy**: the ability to read your own numbers, ask the right questions, and recognize when something has changed enough to matter.

The diagnostic investigation set

Your diagnosis rested on a defined set of investigations, each contributing a different kind of information. No single test confirms idiopathic inflammatory myopathy on its own. The 2017 EULAR/ACR classification criteria use sixteen weighted variables across six domains: age of onset, muscle weakness pattern, skin manifestations, laboratory findings, muscle biopsy features, and electromyography.^[1] Think of it as a composite score, like a credit rating assembled from multiple data points rather than a single pass-or-fail test.

The criteria achieve strong diagnostic performance, with agreement approaching expert physician diagnosis at the standard probability cutoff.^[1] The system works. But it works because no single piece carries the entire weight alone.

The investigations break into two broad groups: those that established what you have, and those that track how it behaves over time.

The autoantibody panel: your disease's fingerprint

The **myositis-specific antibody** (MSA) panel is, in practical terms, the single most informative laboratory result in your diagnostic workup. The British Society for Rheumatology 2022 guideline makes this explicit: patients should be tested for myositis autoantibodies, a recommendation carrying Grade 1B with 100% agreement among the guideline panel.^[2] The reason is straightforward. Each MSA defines a disease phenotype with characteristic organ-involvement patterns, treatment-response tendencies, and risk profile. MSAs are found in approximately 60 to 70% of adults with IIM.^[3]

Here is the detail that makes antibodies unusually useful: they essentially never co-occur in the same patient.^[3] If you carry anti-Jo-1, you are unlikely to also carry anti-MDA5. That mutual exclusivity means a positive MSA result points toward a single, defined phenotype rather than a blurred overlap of possibilities. One antibody, one phenotype. That clarity is rare in autoimmune diagnostics.

The antibody result does not assign you a fate. It assigns a probability landscape. Anti-TIF1-gamma, for example, carries an approximately 50% cancer risk in adults over 40, which is why it triggers enhanced malignancy screening at diagnosis.^[2,4] Anti-MDA5 signals the possibility of rapidly progressive interstitial lung disease, particularly in amyopathic dermatomyositis.^[4] Anti-SRP, one of the two marker antibodies for immune-mediated necrotizing myopathy, is associated with more severe weakness and carries higher risks of cardiac involvement and dysphagia.^[3] Anti-HMGCR, the other IMNM marker, is associated with statin exposure in a significant proportion of cases, though it also occurs in people who have never taken statins.^[3]

Anti-Jo-1, the most common antisynthetase antibody (found in 9 to 24% of adult IIM patients), points toward the classical triad of myopathy, ILD, and arthritis characteristic of antisynthetase syndrome, where ILD occurs in 70 to 95% of cases.^[3] Anti-Jo-1 titers also correlate with disease activity, making the antibody level itself a monitoring tool in that subtype.^[3]

These numbers are population-level risks, not personal predictions. Your antibody result tells your rheumatology team which organ systems need heightened surveillance and how aggressive the monitoring schedule should be. It does not tell either of you what will happen to you specifically.

If your MSA panel came back negative, that does not mean the diagnosis is wrong. Not all patients test positive for an MSA, meaning that a negative antibody panel does not rule out myositis. It means your team has less antibody-guided specificity for risk profiling and relies more heavily on clinical assessment, biopsy findings, and imaging to track your disease.

The distinction between MSAs and **myositis-associated antibodies** (MAAs) matters here. MAAs (such as anti-PM/Scl, anti-Ku, and anti-U1snRNP) are less specific: they appear in IIM patients with overlap features and also in other connective tissue diseases.^[3] An MAA result suggests the possibility that myositis coexists with another autoimmune condition. Your team interprets it in the broader clinical context.

Whether your result is a specific MSA, an MAA, or a seronegative panel, the next step is the same: ask your rheumatology team what your antibody result means for your monitoring schedule. If you do not know your result, this is worth requesting at your next visit.

Creatine kinase: one number, many meanings

Creatine kinase (CK) is the muscle enzyme most people with myositis learn to track first. It appears on nearly every blood panel. When muscle fibers are damaged, CK leaks into the bloodstream, and the level rises. In the 2017 EULAR/ACR criteria, elevated CK (along with LDH, AST, or ALT) contributes 1.3 to 1.4 scoring points toward classification.^[4]

The number is useful. It is also frequently misunderstood.

CK is not a diagnosis. It is a marker of muscle injury requiring contextual interpretation.^[5] Elevated CK appears in muscular dystrophies, metabolic myopathies, rhabdomyolysis, acute viral myositis, drug-induced myopathy, and after vigorous exercise. A high number on its own does not tell you whether the

immune system is attacking your muscles or whether you overdid it at physiotherapy yesterday. Context is everything.

Normal CK does not exclude active disease. In amyopathic dermatomyositis, CK may sit within the normal range despite significant skin disease and even organ involvement.^[5] In some cases of myositis where the inflammatory phase has partially burned out, CK may normalize while functional impairment persists. The number tracks muscle fiber damage in real time; it does not measure accumulated weakness or fibrosis.

The value of CK lies in its longitudinal trend, not in any single reading. In subtypes where CK correlates reliably with disease activity (IMNM, anti-Mi-2-positive DM, anti-Jo-1-associated antisynthetase syndrome), a rising CK from your established baseline may signal a flare before you feel it in your muscles.^[3] A falling CK on treatment may confirm that therapy is working at the tissue level. In anti-HMGCR-positive IMNM, the correlation is particularly strong: median CK in one cohort sat at approximately 4,977 IU/L, and antibody titers tracked with both CK levels and muscle strength.^[3]

The word “baseline” matters. Your reference point is not the laboratory’s printed normal range. It is your own resting CK on stable therapy, the number your blood draws consistently return when your disease is controlled and you have not recently exercised hard. Ask your rheumatology team what your resting CK baseline is. Write it down. Every future CK result gains meaning in relation to that personal number, not in relation to a generic range designed for the general population.

If your CK rises above your personal baseline, it is worth noting. If it rises significantly, that is a Routine conversation at your next visit. If it rises sharply and you also notice new weakness, that becomes a Prompt reason to contact your team within days rather than waiting for a scheduled appointment.

Muscle biopsy: what it determined and why it mattered

If you had a **muscle biopsy**, it likely answered questions that blood tests and imaging could not resolve on their own. The procedure involves removing a small

sample of muscle tissue (usually from the thigh or upper arm) and examining it under a microscope with specialized staining techniques.

The German DGN S2k guideline specifies that biopsy tissue should be processed in detail with immunohistology and, where appropriate, electron microscopy.^[4] That level of processing is not routine pathology. It is specialized work, typically performed at centers experienced with neuromuscular disease.

What biopsy reveals depends on the subtype. In dermatomyositis, the characteristic finding is **perifascicular atrophy**: muscle fibers at the edges of the fascicle (the bundles that make up a muscle) are preferentially damaged, with inflammation concentrated around blood vessels and in the perimysial connective tissue. In IMNM, the pattern is different: muscle fibers show necrosis and regeneration without the dense inflammatory cell infiltration seen in other subtypes. In IBM (the contrast condition excluded from this book's scope), the hallmark is endomysial inflammation alongside rimmed vacuoles and protein aggregates.^[1]

Biopsy substantially improves diagnostic accuracy. The 2017 criteria achieve 86% overall classification accuracy without biopsy and 91% with it. For distinguishing IBM from autoimmune subtypes, the improvement is dramatic: 58% accuracy without biopsy rises to 94% with it.^[1] If your team performed a biopsy, this is why.

A normal biopsy does not rule out IIM entirely. Inflammatory infiltrates can be patchy, and a sample taken from an unaffected area may miss the disease. This is one reason MRI sometimes guides the biopsy site: it identifies the actively inflamed region, reducing the chance of sampling normal-looking tissue next to the disease. Your team considered biopsy results alongside your clinical picture, antibody results, and imaging. No single investigation was definitive on its own. Together, they built the case.

MRI: seeing inflammation in the muscle

Magnetic resonance imaging (MRI) with T2 and STIR sequences identifies skeletal muscle edema, a marker of active inflammation.^[5] On these sequences, inflamed muscle appears bright against the surrounding tissue, mapping where the disease is currently active.

MRI serves two purposes in your diagnostic journey. Initially, it may have guided your biopsy by identifying an actively inflamed muscle to sample, improving the chance of a diagnostic yield. Over time, it can track disease activity and treatment response, showing whether areas of inflammation are resolving or persisting.^[5]

MRI also helps distinguish autoimmune IIM from conditions that mimic it. Autoimmune myositis typically shows diffuse, often symmetric T2 signal elevation in proximal muscles. Infectious myositis may show focal abscess formation. Muscular dystrophies show fatty replacement patterns rather than edema.^[6] These differences helped narrow your diagnosis.

Your team decides when MRI is warranted for monitoring. If you are told your disease appears controlled on clinical assessment and blood work, routine repeat MRI may not be necessary. If symptoms change or there is uncertainty about disease activity, MRI provides information that blood tests cannot.

Electromyography: confirming the pattern

Electromyography (EMG) involves inserting a thin needle electrode into a muscle to measure its electrical activity. In active IIM, EMG typically shows a myopathic pattern: short-duration, low-amplitude motor unit potentials combined with increased insertional activity (fibrillation potentials and positive sharp waves), reflecting muscle membrane irritability.^[5]

EMG's primary contribution is confirming that weakness comes from the muscle itself rather than from nerve damage. A neuropathic cause of weakness (which would require a different diagnostic pathway) shows a distinctly different electrical pattern. EMG does not distinguish among IIM subtypes with high specificity, but it confirms the myopathic origin of the problem.^[5]

One pattern worth understanding: if EMG shows myotonic discharges (a specific repetitive firing pattern), the differential shifts toward myotonic dystrophies or certain metabolic myopathies rather than classical autoimmune IIM.^[7] This is one of the ways EMG helps the team exclude conditions that look like myositis but are not.

Most people find EMG uncomfortable. It is not typically repeated for routine monitoring once the diagnosis is established, unless the clinical picture changes in a way that raises new diagnostic questions.

Pulmonary function tests: a baseline that matters

Pulmonary function tests (PFTs) measure how well your lungs move air and transfer oxygen. In IIM, PFTs serve as screening and surveillance for interstitial lung disease, which occurs across multiple subtypes and is a leading cause of morbidity and mortality.^[2]

The two values your team tracks most closely are **forced vital capacity** (FVC) and **diffusing capacity for carbon monoxide** (DLCO). FVC measures the total volume of air you can forcefully exhale after a full breath. DLCO measures how efficiently oxygen crosses from the air sacs into your blood. Both decline when ILD develops or worsens.

Your baseline PFT result is another personal reference number worth knowing. A meaningful decline in FVC or DLCO from your baseline is a trigger for clinical review, and your team will tell you what change matters for you. If you have not had PFTs, this is worth raising with your team. If you have, ask what your baseline values are. These are numbers that matter over years, not just at diagnosis.

If you notice new or worsening breathlessness, do not wait for a scheduled PFT. That is an Urgent reason to contact your team the same day.

Malignancy screening: a clinical action point

Certain MSAs carry an established association with cancer, and guideline-based screening at diagnosis is a defined part of IIM care. The German DGN S2k guideline recommends that all IIM patients at diagnosis receive a history review including B-symptoms, detailed physical examination, CT of the thorax and abdomen (or alternatively chest X-ray and abdominal ultrasound), and gynaecological, urological, and gastroenterological screening examinations.^[4]

For patients carrying anti-TIF1-gamma, anti-NXP2, or anti-HMGCR antibodies, the recommendation intensifies: FDG-PET-CT or whole-body CT at diagnosis, with repeat within the first year and further surveillance over the following two years based on individual risk.^[4]

This is not cause for alarm. It is cause for a conversation. If you do not know whether you have completed guideline-recommended malignancy screening, ask your team at your next Routine visit. If screening has been done, ask whether follow-up is recommended based on your antibody status. The specific protocol details (which screens apply, at what intervals, and for how long) are clinical decisions managed by your team with knowledge of your individual risk profile. The full epidemiological context for cancer-associated myositis is covered in Chapter 7.

Putting it together: disease-activity literacy in practice

Disease-activity literacy is a collaboration tool, not a self-diagnosis tool. It means knowing four things: what your antibody result is (or that you are seronegative), what your resting CK baseline is on stable therapy, what your PFT baseline is, and what screening has been completed. These four data points, combined with your team's clinical assessments, form the monitoring framework that follows you for years.

There is a practical asymmetry in chronic disease care that makes this literacy matter. Your rheumatology team sees you periodically. You live in your body every day. Between appointments, you are the one who notices that stairs have become harder, that a blood result is higher than last time, or that breathing feels different during exercise. Disease-activity literacy gives those observations a framework: you know which changes are Routine topics for the next visit, which are Prompt reasons to call within days, and which are Urgent signals to seek care now. Know your baseline numbers, report changes, and raise the clinical action points that may not come up unless you bring them.

Key Points

- Your antibody result (or seronegative status) shapes your monitoring schedule; ask your team what it means for you specifically.
- CK gains meaning from your personal resting baseline on stable therapy, not from the laboratory's printed normal range.

- A normal CK does not rule out active disease; amyopathic dermatomyositis and burned-out phases can produce misleadingly normal values.
- Muscle biopsy substantially improves diagnostic classification accuracy overall and is critical for distinguishing IBM from autoimmune subtypes.
- Your PFT baseline (FVC and DLCO) is a reference you should know, because a meaningful decline triggers clinical review.
- Malignancy screening at diagnosis is a guideline recommendation worth confirming has been completed with your team.

CHAPTER 3: TREATMENT, WITH IVIG AT THE CENTER

Treatment for autoimmune myositis is not a single medication; it is a layered architecture built to hold a disease that requires sustained, calibrated pressure over months and years. At the center of that architecture sits intravenous immunoglobulin, the only therapy in this disease backed by a large prospective randomized trial and an FDA approval. Understanding why IVIG occupies that central position, what the trial evidence established, and how corticosteroids, conventional immunosuppressants, rituximab, and emerging JAK inhibitors fit around it turns the treatment plan from a list of drugs into a system you can follow and question. That understanding belongs to you, not only to your team.

The treatment no one expected to work first

Most autoimmune diseases follow a predictable script. A patient receives a diagnosis, starts a drug that suppresses the immune system, and waits to see whether the disease quiets down. If it does, the drug stays. If it does not, another one replaces it. The logic is straightforward: overactive immunity causes the damage, so dampening immunity should reduce it.

Intravenous immunoglobulin does not follow that script. It is not an immunosuppressant. It does not dampen the immune system. It is pooled human antibody, collected from thousands of plasma donors, infused into your veins every month.^[1] And yet, in the largest prospective trial ever conducted for any myositis subtype, it outperformed placebo by a margin that changed the regulatory landscape of the disease.^[2]

That trial, and the FDA approval it produced, is where this chapter begins. But the story it tells is larger than one drug. It is a story about how a hierarchy of treatments fits together: which ones come first, which ones back them up, and where IVIG sits in a system designed to control a disease that requires sustained, calibrated pressure over months and years.

The hierarchy: how the pieces fit

Treatment for autoimmune inflammatory myopathy is not a single medication. It is a layered system, and the British Society for Rheumatology published the most methodologically rigorous guideline to date for that system in 2022, grading 213 publications through formal GRADE methodology.^[3]

The hierarchy has a clear structure.

CORTICOSTEROIDS: THE FIRST LINE

Glucocorticoids remain the established first-line treatment for all autoimmune IIM subtypes. No placebo-controlled trial of corticosteroids in myositis has ever been conducted, but decades of observational evidence showing mortality reduction and improvement in more than 60% of patients provide the foundation on which every subsequent treatment builds.^[4] The BSR recommends high-dose glucocorticoids to treat active muscle inflammation at the time of treatment induction, with strong confidence (Grade 1B) and unanimous panel agreement.^[3]

What corticosteroids do well, they do quickly. What they cost, they cost cumulatively. The side-effect burden (bone loss, glucose dysregulation, infection risk, weight gain, cataracts, and more) is common enough and serious enough that it is the primary driver for early co-prescription of steroid-sparing immunosuppressants.^[4] Your team does not wait until side effects appear. They begin building the next layer from the start.

The specifics of those side effects, and how to manage them, belong in Chapter 4. Here, the point is simpler: corticosteroids open the door, and the goal from the moment they start is to walk through it toward something that can hold the disease without the same cumulative cost. The standard approach reduces the dose gradually over months, guided by muscle strength assessments and laboratory markers, targeting a low maintenance dose that balances disease control against side-effect burden.^[4] That taper does not happen in isolation. It happens alongside the next layer.

STERIOD-SPARING IMMUNOSUPPRESSANTS: THE SUPPORTING CAST

Five conventional **disease-modifying antirheumatic drugs** (DMARDs) occupy the BSR's first-line steroid-sparing tier: methotrexate, azathioprine, mycophenolate mofetil, tacrolimus, and ciclosporin.^[3] Early combination therapy (corticosteroid plus immunosuppressant from the first phase of treatment) is associated with better functional outcomes than corticosteroids alone.^[4]

These are not interchangeable. Each has a distinct profile, and the choice among them depends on your subtype, your organ involvement, and considerations your rheumatology team weighs against your specific situation.

Methotrexate is often a first-choice conventional immunosuppressant for muscle involvement, with reported response rates of approximately 70 to 80% in retrospective and open series.^[4] It carries one important caveat for myositis patients: a risk of pulmonary toxicity that makes it less preferred when interstitial lung disease is present. A 2025 retrospective study of antisynthetase syndrome documented two cases of reversible methotrexate pneumonitis, reinforcing the pattern that azathioprine is preferred over methotrexate in patients with ILD.^[5]

Azathioprine has the longest evidence history as a steroid-sparing agent in myositis. A randomized placebo-controlled trial in polymyositis demonstrated steroid-sparing benefit and better functional outcome at three-year follow-up.^[4] It shows similar efficacy to methotrexate in head-to-head comparisons and avoids the pulmonary toxicity concern, making it the preferred choice when lung involvement is part of the clinical picture.^[5]

Mycophenolate mofetil is particularly reported as effective for IIM-related interstitial lung disease, including rapidly progressive forms.^[4] **Tacrolimus** and **ciclosporin**, the calcineurin inhibitors, block T-cell activation and have shown benefit on pulmonary function in patients with antisynthetase antibody positivity.^[4]

Which of these your team selects, and whether more than one is combined, is a clinical decision that depends on your antibody profile, your organ involvement, and your tolerance. The chapter that follows covers what living with these medications looks like day to day. Here, the point is their place in the architecture: they support

the corticosteroid taper, and they occupy the layer between first-line steroids and the more targeted therapies above.

HYDROXYCHLOROQUINE: THE SKIN-SPECIFIC OPTION

Hydroxychloroquine occupies a separate lane. It is used for skin manifestations in dermatomyositis (effective in 40 to 75% of patients in observational data) but appears ineffective for muscle involvement.^[4] If your disease includes significant skin disease, your team may prescribe it alongside the agents above. It does not replace them for muscle control.

IVIG: the mechanism and the evidence

This is the center of the chapter, because it is the center of your maintenance therapy.

WHAT IVIG IS

Intravenous immunoglobulin is pooled human IgG derived from thousands of plasma donors. It contains antibodies directed against a broad range of pathogens and autoantigens, and it is classified as immunomodulatory rather than immunosuppressive.^[1] The two are not the same, and confusing them carries consequences. Immunosuppressants turn the immune system down. IVIG recalibrates it.

HOW IT WORKS: ONE MECHANISM, LANDED

The mechanisms of IVIG in autoimmune disease are multiple, but one is particularly well characterized in dermatomyositis and illustrates the logic of the therapy.

Think of it this way. In dermatomyositis, the immune system's **complement** cascade, a chain reaction that normally helps destroy invaders, misfires. It deposits a structure called the **membrane attack complex** on the tiny blood vessels (capillaries) that feed muscle tissue, damaging them from the inside.^[6] The muscle damage you experience is downstream of that vascular injury.

IVIG interrupts the chain. Infused immunoglobulin has been shown to diminish complement activation and reduce the deposition of the membrane attack complex on muscle capillaries and fibers.^[6] It is not destroying the immune cells. It is intercepting the ammunition before it reaches the target.

A second mechanism works in parallel. Your body has a receptor called **FcRn** (the neonatal Fc receptor) that protects antibodies from being broken down, extending their lifespan in the bloodstream. Pathogenic autoantibodies, the ones causing damage, benefit from that protection too. When IVIG floods the system with a large dose of normal IgG, it saturates FcRn, effectively crowding out the harmful antibodies and accelerating their clearance.^[7,402] The harmful antibodies lose their bodyguard.

These two mechanisms (complement inhibition and accelerated autoantibody clearance) are the best-characterized pathways. Additional mechanisms include suppression of cytotoxic T-cell activation and migration into muscle tissue, reduction of pro-inflammatory cytokines, neutralization of disease-specific autoantibodies through anti-idiotypic binding, and inhibition of B-cell and dendritic-cell activation.^[7,402] The full picture is complex. The practical point is not.

IVIG works through modulation, not suppression. That is why it can be used safely during active infection, in patients with malignancy, and (with limited data) during pregnancy, settings where conventional immunosuppressants carry additional risks.^[4]

THE PRODERM TRIAL: THE EVIDENCE LANDMARK

The **ProDERM trial** is the pivotal study. It is the first large prospective randomized controlled trial of IVIG in any myositis subtype, and the largest study ever conducted to evaluate the therapy for this disease.^[2]

The numbers are worth sitting with. Ninety-five adults with active dermatomyositis were enrolled across 36 sites globally. They received either IVIG or placebo every four weeks for 16 weeks, followed by a 24-week open-label extension.^[2]

At Week 16, a large majority of patients receiving IVIG met the definition of clinical improvement. In the placebo group, 43.8% improved.^[2] That gap (nearly 35 percentage points) crossed the threshold that regulators require to approve a therapy. Skin disease, measured by the Cutaneous Dermatomyositis Disease Area and Severity Index, improved significantly. And efficacy was maintained through Week 40.^[8]

A 2023 post-hoc analysis confirmed that skin improvement occurred regardless of how severe the skin disease was at baseline.^[9,411] The trial did not just show that IVIG works. It showed that it works broadly, across a range of disease severity, and that it holds.

THE FDA APPROVAL

On July 15, 2021, the U.S. Food and Drug Administration approved Octagam 10% for the treatment of adult dermatomyositis. It is the first and only FDA-approved IVIG specifically indicated for this condition.^[10] The FDA's Office of Orphan Products Development subsequently granted seven years of marketing exclusivity.^[10]

That approval changed the regulatory landscape. Before ProDERM, IVIG for myositis was supported by smaller controlled trials, open-label studies, and clinical experience. The therapy was effective and widely used, but it lacked the regulatory standing that a Phase III trial provides.^[4] After ProDERM, dermatomyositis became one of the few autoimmune conditions with a specifically indicated IVIG product backed by large-scale RCT evidence.

IVIG-CENTERED MAINTENANCE

This is the concept at the heart of the chapter. **IVIG-centered maintenance** is the pattern in which IVIG, administered monthly, serves as the backbone of disease control while the team calibrates the drugs around it (tapering corticosteroids, selecting the right immunosuppressant, adjusting based on response).

The BSR recommends IVIG for severe and refractory muscle inflammation and for glucocorticoid- and DMARD-refractory skin disease, with strong confidence and unanimous agreement (Grade 1B, 100%).^[3] In practice, IVIG occupies the position of the sustained-control therapy: the one that keeps running while other elements of the regimen shift.

What does maintenance look like in practice? The standard approach used in the ProDERM trial, and consistent with prior literature, is a dose administered intravenously every four weeks.^[2] That dose is typically fractionated across two to five consecutive infusion days to reduce adverse-event risk and manage fluid load.^[1] Each infusion day involves sitting in a reclining chair at an infusion center or hospital day unit, often for several hours, while a nurse monitors for reactions throughout.

Premedication (typically acetaminophen and an antihistamine) is standard practice to mitigate infusion reactions.^[1]

The number of cycles varies by clinical response. Some patients receive IVIG for a defined course and then reassess. Others are maintained longer-term.

Subcutaneous immunoglobulin, self-administered via a programmable pump, has been reported as an alternative to hospital-based IVIG in some myositis patients, though the evidence base is still emerging.^[4]

Your specific IVIG product, your infusion schedule (how many days per cycle, at what rate), and the decision to continue, adjust, or eventually taper are all clinical decisions that belong to your team. If you do not know which product you receive, or whether your team has considered your IgA status in selecting it, those are worth asking about. The chapter's purpose is not to prescribe those details. It is to show you where the therapy sits in the evidence and why it occupies the position it does.

COMMON SIDE EFFECTS AND BOXED CAUTIONS

IVIG is well tolerated in most patients, but it carries defined risks that your infusion team monitors.

Common reactions occur in roughly 5 to 15% of infusions: headache, fever, chills, muscle aches, and fatigue, typically within 24 hours and typically self-resolving.^[4] Slowing the infusion rate usually manages acute reactions during the infusion itself.

Rare but serious effects include **aseptic meningitis** (disproportionately reported in patients with a history of migraines and with rapid infusion rates), hemolytic anemia, and hyperviscosity syndrome.^[11]

All IVIG products carry two FDA boxed warnings. The first concerns **thrombosis**: blood clots that may occur with any IVIG product, with risk factors including advanced age, prolonged immobilization, prior thrombosis, estrogen use, and cardiovascular risk factors.^[12] The second concerns **renal dysfunction**, with risk factors including pre-existing kidney impairment, diabetes, age over 65, and use of nephrotoxic drugs.^[12] The mitigation guidance is consistent: administer at the minimum effective infusion rate, ensure adequate hydration, and use the lowest dose practicable in patients with elevated risk.^[12]

The quantified safety data from ProDERM and the specific thromboembolism risk factors are covered in Chapter 4, where they sit alongside the broader side-effect landscape of all treatments. Here, the point is that these are known, monitored, and managed, not discovered after the fact.

One additional consideration: patients with selective **IgA deficiency** who have developed anti-IgA antibodies face an elevated risk of anaphylaxis when exposed to IgA-containing IVIG preparations.^[13] IgA deficiency screening before initiating IVIG is recommended practice and directly influences which product your team selects. If you have not discussed your IgA status with your team, it is worth raising at your next visit. This is a Routine question.

Rituximab: the refractory-disease option

When corticosteroids, conventional DMARDs, and IVIG are insufficient to control the disease, the treatment hierarchy moves to **rituximab**, a monoclonal antibody that depletes the B cells responsible for producing autoantibodies.^[3]

The landmark trial is the Rituximab in Myositis (RIM) study. It enrolled adults and juveniles with refractory dermatomyositis or polymyositis. The primary endpoint (time to improvement) was not met in the intention-to-treat analysis, but for a reason that tells its own story: both treatment arms responded at high rates, making it difficult to detect a statistical difference between them.^[14] Rituximab did achieve significant steroid-sparing, and a post-hoc analysis identified something clinically useful. Patients who carried anti-Jo-1 or anti-Mi-2 autoantibodies were strong predictors of a good response.^[14]

A 2022 systematic review and meta-analysis, pooling 26 studies, reported an overall response rate of 65%.^[15] Skin involvement showed the strongest response at 81%. Lung disease responded in 65% of cases. Muscle involvement responded in 59%.^[15]

The BSR recommends rituximab for refractory myositis (Grade 2A, conditional recommendation), noting particular effectiveness in juvenile-onset disease and in patients with a positive myositis autoantibody profile and lower accumulated damage.^[3]

Rituximab is not the same kind of therapy as IVIG. Where IVIG modulates, rituximab depletes. It eliminates the B cells that produce the autoantibodies driving the

disease. The response pattern reflects the different organ domains: skin responds most readily, followed by lung disease, with muscle involvement responding somewhat less consistently.^[15] This variation is one reason your team considers antibody profile and dominant organ involvement when deciding whether rituximab is the right choice.

The adverse-event profile includes infusion reactions (mainly at the first infusion), infections (bacterial and viral, including serious infections), and, rarely, progressive multifocal leukoencephalopathy.^[15] Vaccination against pneumococcus and influenza before starting rituximab is recommended, since B-cell depletion impairs the immune response to vaccines administered afterward.^[15]

Whether you are a candidate for rituximab depends on your autoantibody profile, your disease trajectory, and your team's assessment of where you sit in the treatment pathway. That assessment is theirs to make.

Refractory disease: the outer layers

Beyond rituximab, the hierarchy reaches treatments reserved for severe or refractory disease that has not responded to the options above.

Cyclophosphamide is used mainly for severe or progressive interstitial lung disease in myositis, particularly in patients with anti-MDA5 or anti-SRP antibodies. Its toxicity profile (myelosuppression, hemorrhagic cystitis, infections, infertility, secondary malignancy) limits it to situations where other options are inadequate.^[4] The BSR assigns it a strong recommendation for severe and refractory disease (Grade 1B).^[3]

Abatacept, a T-cell co-stimulation blocker, carries a conditional BSR recommendation (Grade 2B).^[3] A Phase 3 trial published in 2025 showed variable success, with the overall result not meeting its primary endpoint but a subgroup signal in non-dermatomyositis patients.^[16]

These are not treatments you are likely to encounter unless your disease has proven resistant to the layers closer to the center. They exist to ensure that the hierarchy has depth. There is always a next step. For completeness: anti-TNF agents are generally not recommended in adult myositis, with some patients reported to have

worsened on them, and plasma exchange showed no benefit in a randomized trial.^[4] These are closed doors, not open ones.

JAK inhibitors: the emerging layer

The most significant recent development in the treatment landscape sits at the boundary between established and emerging.

JAK inhibitors target the signalling pathways downstream of the interferons that drive dermatomyositis pathogenesis. The biological rationale is clear: type I and type II interferons are prominent in DM, the JAK-STAT pathway mediates their effects, and blocking that pathway should reduce the inflammatory cascade.^[17]

Case series and small meta-analyses of existing JAK inhibitors (tofacitinib, ruxolitinib, baricitinib) have shown promising skin improvement in refractory dermatomyositis.^[18] But the evidence remained at the case-series level until one trial changed the calculus.

The **VALOR trial** produced the first positive Phase III result for a targeted oral therapy in dermatomyositis, establishing JAK inhibition as a genuine treatment layer rather than a speculative one.^[19,20] The full trial data and the regulatory arc belong in Chapter 8. Whether it will be relevant to your treatment depends on your subtype and your team's assessment.

JAK inhibitors as a class carry defined safety considerations, including increased risk of infections (particularly herpes zoster), venous thromboembolism, cardiovascular events, and malignancy.^[18] These risks must be weighed against the potential benefits in any individual patient, which is another reason this decision belongs to your team.

The full VALOR trial data and the broader pipeline of investigational therapies are the subject of Chapter 8. Here, the point is that the hierarchy is not static. A treatment layer that was speculative five years ago now has Phase III evidence behind it. The architecture adapts.

The architecture, seen whole

Step back and look at the system.

Corticosteroids open the door. Conventional immunosuppressants support the taper. IVIG provides sustained control at the center, backed by the strongest RCT evidence in the field. Rituximab covers refractory disease. Cyclophosphamide and abatacept provide depth for severe cases. And JAK inhibitors are moving from the edge of the map toward the established center.

One thing this architecture reveals is how few of its medications were designed for myositis. Corticosteroids, methotrexate, azathioprine, rituximab: all came from other fields and were adapted. IVIG was pooled antibody used across multiple autoimmune conditions before the ProDERM trial gave it a myositis-specific indication. The treatment of rare disease often works this way, borrowing tools from larger fields and building evidence patiently, study by study. The 2022 BSR guideline itself was based on 213 publications for a disease group that most clinicians encounter only a few times in a career.^[3]

That context matters because it explains why so many decisions are routed to your team rather than resolved in a book. The evidence base, while growing, is smaller than for diseases that affect millions of people. The guideline grades reflect this: strong recommendations where the evidence is clear, conditional recommendations where it is still accumulating, and clinical judgment filling the gaps in between. Your team holds the clinical judgment. This chapter holds the map.

Your team navigates this hierarchy based on your subtype, your antibody profile, your organ involvement, and your response over time. The decisions about which drugs to combine, when to adjust, and when to add or remove a layer are clinical decisions that this book does not make. If your team proposes a change, the architecture in this chapter is the framework for understanding where the new plan fits. If you want to ask about alternatives, this chapter gives you the vocabulary. The decision remains theirs. The understanding is yours.

Key Points

- Corticosteroids remain the established first line for autoimmune myositis, but the goal from the start is to taper them through steroid-sparing agents.

- IVIG is immunomodulatory rather than immunosuppressive, recalibrating the immune response through complement inhibition and accelerated autoantibody clearance.
- The ProDERM trial established IVIG efficacy in dermatomyositis and produced the first and only FDA-approved IVIG indication for this disease.
- Rituximab is the primary option for refractory disease, with autoantibody profile predicting response; patients with a positive myositis antibody are more likely to benefit.
- JAK inhibitors are emerging, with the VALOR trial producing the first positive Phase III result for an oral targeted therapy in dermatomyositis.

CHAPTER 4: LIVING WITH TREATMENT: MANAGING SIDE EFFECTS

In this chapter: the medications that control myositis carry costs of their own, from bone thinning and blood sugar shifts to infection vulnerability and infusion-related risks. Understanding these treatment-driven complications, knowing what to watch for, and knowing when to call your team turns passive side-effect endurance into active, informed management.

The drugs that quiet an overactive immune system do not limit their effects to the immune system. Corticosteroids thin bone. They raise blood sugar. They can weaken the very muscles they were prescribed to protect. Immunosuppressants lower defenses against infections the body would ordinarily handle without difficulty. Intravenous immunoglobulin, for all its benefit, thickens the blood and stresses the kidneys. None of this is a reason to refuse treatment; untreated inflammatory myopathy is worse. But it is a reason to understand exactly what treatment costs, so you and your team can manage those costs together.

That cluster of downstream harms has a name worth keeping: **treatment-driven comorbidity**. It is the recognition that many of the complications a person with myositis faces are not caused by the disease itself but by the therapy required to control it. The distinction matters because the response to a treatment-driven problem is usually the opposite of the response to a disease-driven one. A flare needs more immunosuppression. A treatment complication needs less, or a different kind of protection layered on top. Confusing the two leads to harm in both directions.

When the Treatment Mimics the Disease: Steroid Myopathy

Here is a clinical paradox that every person on long-term corticosteroids should understand. The medication prescribed to reduce muscle inflammation can, over time, cause muscle weakness of its own. That weakness looks and feels similar to the disease it treats. Getting the distinction right is one of the most important conversations you will have with your rheumatology team.

The condition has a name worth knowing. **Steroid myopathy** presents as painless, proximal weakness, predominantly in the legs.^[1] Facial and swallowing muscles are spared. Deep tendon reflexes stay intact. The pattern can be subtle enough that the person (and sometimes the clinician) assumes the underlying disease is getting worse when the treatment is actually the problem.

The most reliable early indicator sits in a number you already know. In steroid myopathy, serum creatine kinase (CK) is typically normal or near-normal. A rising CK, by contrast, points toward inflammatory disease activity rather than steroid toxicity.^[1,2] This single lab value often provides the first clue.

Think of it the way you might think about a thermostat. CK is a signal. When inflammation is driving the weakness, the signal runs hot. When steroids are driving the weakness, the signal stays cool even though the muscles feel just as weak. The thermostat reading does not tell you why the house feels uncomfortable, but it narrows the possibilities substantially.

Further investigation separates the two patterns more precisely. Electromyography in steroid myopathy typically shows no spontaneous electrical activity, while inflammatory myopathy produces the characteristic fibrillations and sharp waves of active muscle-fiber irritation.^[1] Muscle biopsy, if performed, shows selective atrophy of fast-twitch (type 2) fibers without inflammatory infiltrate, a clean pattern that distinguishes treatment damage from disease damage.^[3]

Timing provides an additional clue. Weakness that appears early in a course of corticosteroids (the first weeks) is more likely disease activity breaking through. Weakness emerging after months of stable-dose therapy, particularly alongside other signs of steroid excess (weight gain concentrated around the face and trunk, thin skin, easy bruising), is more likely steroid myopathy.^[1]

Not all corticosteroids carry equal risk. Fluorinated glucocorticoids (dexamethasone, triamcinolone, betamethasone) are associated with higher myopathy risk than non-fluorinated agents such as prednisone and prednisolone.^[4] Your team's choice of agent already reflects this consideration. When steroid myopathy is suspected, reducing the corticosteroid dose is the primary intervention, and weakness typically improves over weeks to months after dose reduction. Alternate-day dosing may also reduce the risk.^[4] Moderate-intensity aerobic exercise and resistance training in

muscles that retain greater-than-antigravity strength play a preventive and rehabilitative role, though the evidence base for exercise as a specific countermeasure to steroid myopathy remains limited.^[4]

The management consequences are direct. If the weakness is inflammatory, immunosuppression needs to increase. If it is steroid myopathy, the corticosteroid dose needs to come down. Getting this wrong in either direction carries substantial harm: undertreating a flare lets the disease advance, while escalating steroids for steroid-induced weakness deepens the very problem it was meant to address.^[1,2] This distinction is not always resolvable without specialist review. If you notice new or worsening weakness on a stable steroid regimen, that is a conversation for your rheumatologist, not a problem to manage alone.

This is Prompt. New weakness on a stable corticosteroid dose warrants contact with your rheumatology team within days, not at the next scheduled visit.

Protecting Your Bones

Glucocorticoid-induced osteoporosis is the most common form of secondary osteoporosis, and it begins earlier than most people expect. Fracture risk starts rising within the first three to six months of corticosteroid use.^[5] It is detectable even at doses below 2.5 mg prednisolone equivalent per day. No safe dose threshold has been established.

The 2022 American College of Rheumatology guideline for glucocorticoid-induced osteoporosis (published in 2023) provides a clear framework.^[6] It strongly recommends a clinical fracture risk assessment within six months of starting or continuing corticosteroids at 2.5 mg per day or more for longer than three months. That assessment includes fracture history, a FRAX calculation (a validated fracture probability tool for people aged 40 and older), and bone mineral density measurement by DXA scan.

The guideline defines four fracture risk categories. Low risk corresponds to a FRAX ten-year major osteoporotic fracture probability below 10%. Moderate sits between 10% and 19%. High risk is 20% or above. Very high risk applies to anyone with a prior osteoporotic fracture, a bone density T-score at or below negative 3.5, a

glucocorticoid dose of 30 mg per day or more for over 30 days, or a cumulative dose exceeding 5 grams per year.^[6]

For people on doses above 7.5 mg per day, the guideline recommends applying a correction to the FRAX calculation: multiply the major osteoporotic fracture probability by 1.15 and the hip fracture probability by 1.2.^[6] Corticosteroids degrade bone quality in ways that standard density measurement alone does not fully capture, and the correction accounts for that hidden fragility.

High cumulative glucocorticoid exposure (greater than 1 gram prednisolone equivalent total) is an independent fracture risk factor beyond current daily dose.^[5] This is particularly relevant if your treatment history involved a high initial induction course. The bone damage from that early burst does not reset when the dose comes down.

Protection starts with the basics, regardless of risk category. Calcium intake of 1,000 to 1,200 mg per day (from food and supplements combined), vitamin D supplementation of 600 to 800 IU per day as a starting point, weight-bearing exercise, smoking cessation, and limiting excess alcohol are universally recommended.^[6] These measures are the floor, not the ceiling.

For people at high fracture risk, the guideline strongly recommends pharmacologic bone protection. Oral bisphosphonates (alendronate or risedronate) are the initial therapy for most high-risk patients. For those who cannot tolerate oral bisphosphonates, intravenous options and denosumab are alternatives.^[6] For very high fracture risk, anabolic agents (teriparatide or romosozumab) are conditionally recommended as initial therapy, though these carry their own considerations (cardiovascular cautions with romosozumab, for instance) and require specialist-level evaluation.^[6]

If you have been on corticosteroids for more than three months and have not had a DXA scan or a FRAX calculation, that is a concrete question for your next rheumatology or primary care visit: has bone protection been assessed, and does it need to start?

This is Routine. Bone density assessment and protection planning belong at your next scheduled appointment.

Blood Sugar on Steroids

Corticosteroids disrupt glucose regulation through a mechanism that catches many people off guard. The hyperglycemia they produce is predominantly postprandial, meaning it shows up after meals rather than in the morning fasting window.^[7] A fasting glucose test can come back normal while afternoon and evening glucose runs substantially higher than it should. This timing pattern is not a quirk; it is a predictable consequence of how corticosteroids interact with insulin sensitivity.

With a single morning dose of an intermediate-acting glucocorticoid (prednisolone, the most commonly prescribed in myositis), peak hyperglycemia typically arrives four to six hours after the dose, hitting late afternoon and early evening hardest.^[7] If you have ever wondered why you feel different in the afternoon on steroid days, this is one possible explanation.

Several factors raise the risk of clinically significant glucose elevation: a daily dose above 20 mg prednisolone, longer treatment duration, age over 65, higher body mass index, a baseline HbA1c at or above 6%, and a family history of diabetes.^[7] None of these factors means glucose problems are inevitable. They mean monitoring matters.

The metabolic effects of corticosteroids extend beyond glucose. Even low-dose use (prednisolone 2.5 to 7.5 mg per day) is associated with increased cardiovascular risk through a cluster of changes: central obesity, dyslipidemia, hypertension, and insulin resistance.^[8] This metabolic syndrome pattern is a recognized consequence of prolonged glucocorticoid use and contributes to cardiovascular risk independently of the underlying inflammatory disease.

One additional concern belongs here. Chronic glucocorticoid use above approximately 20 mg per day for more than two weeks, or any dose above physiologic replacement for more than three to four weeks, can suppress the hypothalamic-pituitary-adrenal (HPA) axis.^[8] In practical terms, this means the body's own cortisol production may shut down during extended steroid therapy. Recovery after stopping can take months and, in some cases, up to a year. If you are on long-term steroids and face surgery, significant illness, or another physiological stress, your team needs to know, because supplementary stress dosing may be required.

If you are on corticosteroids and your glucose monitoring has been limited to fasting blood draws, it is worth asking your team whether postprandial or random glucose checks would be more informative. Fasting glucose alone can miss the problem entirely.^[7]

This is Routine. Glucose monitoring and metabolic assessment are standing items for your next visit.

Infection Risk: What Immunosuppression Opens the Door To

Corticosteroids and immunosuppressants do their job by turning down the immune response. The disease needs that suppression. Infections exploit it. The highest absolute infection risk in inflammatory myopathy occurs in the first three months after starting immunosuppressive induction therapy, when cumulative immunosuppression is at its peak.^[9]

Two infections deserve specific attention because the data in myositis are concrete enough to quantify.

PNEUMOCYSTIS PNEUMONIA

Pneumocystis jirovecii pneumonia (PJP, formerly known as PCP) is a serious opportunistic lung infection. Across all forms of inflammatory myopathy, the estimated incidence is approximately 3.0 per 100 person-years, substantially higher than in the general population.^[10]

One subgroup carries dramatically higher risk. Patients with anti-MDA5-antibody-positive dermatomyositis face a PJP incidence of 7.5 per 100 person-years, and three-month mortality from PJP in this subgroup is approximately 83%.^[10] The independent predictors for PJP in inflammatory myopathy include presence of anti-MDA5 antibodies, a CD4+ T-cell count below 200 cells per microliter, and disease duration under three months post-treatment initiation.^[10]

PJP prophylaxis (typically trimethoprim-sulfamethoxazole) is inconsistently prescribed across rheumatic disease populations, and evidence for universal prophylaxis in all myositis patients is not yet definitive.^[11] Whether prophylaxis is appropriate for you depends on your antibody status, your immunosuppressant regimen, and your team's assessment of your individual risk. If you do not know

whether you are on PJP prophylaxis, or whether your antibody panel included anti-MDA5 testing, those are specific questions for your rheumatologist.

This is Prompt for high-risk subgroups. If you develop new fever, dry cough, or progressive breathlessness while on immunosuppression, contact your team promptly. Do not wait.

HERPES ZOSTER

The second infection with strong myositis-specific data is **herpes zoster** (shingles). A national cohort study found that people with dermatomyositis or polymyositis develop shingles at a rate of 35.8 per 1,000 person-years, compared with 7.01 per 1,000 person-years in matched controls without myositis.^[12] That is roughly a five-fold difference in the underlying disease alone.

Treatment amplifies the gap. Combined corticosteroid plus immunosuppressant therapy raises the hazard ratio to 7.43.^[12] Even myositis patients not on any immunosuppressant had nearly five times the zoster risk of the general population, suggesting that the disease itself, not only the treatment, contributes to vulnerability.^[12]

Shingles is painful, sometimes debilitating, and in immunosuppressed patients the episodes can be prolonged and more severe. But unlike PJP, shingles has a highly effective preventive measure. That brings us to vaccination.

Vaccination: What You Need and When

Vaccination is actively recommended for people with rheumatic diseases on immunosuppression, and the benefit of preventing infection substantially outweighs theoretical concerns about disease flare.^[13] Studies consistently fail to show increased flare rates after standard non-live vaccines in myositis and related conditions. The 2022 ACR vaccination guideline and the 2026 EULAR recommendations provide specific schedules for immunosuppressed adults.^[13,14]

Given the five-fold zoster risk described above, **Shingrix (recombinant zoster vaccine)** is the vaccine that most directly connects to the data already in this chapter. It is strongly recommended for all adults aged 18 and older on immunosuppressive therapy.^[13,14] This is a non-live, recombinant vaccine; it does not carry the live-virus contraindication that applies to the older zoster vaccine.

Trial data demonstrated high efficacy, and a post-hoc analysis in patients with autoimmune inflammatory rheumatic disease confirmed that efficacy without increased flares.^[15] If you have not received Shingrix, that gap is worth addressing at your next appointment.

Influenza vaccination is strongly recommended annually. High-dose or adjuvanted formulations are conditionally preferred over standard-dose for immunosuppressed patients, to improve the immune response.^[13]

Pneumococcal vaccination is strongly recommended for immunosuppressed adults of any age. Current guidance offers either PCV20 alone or PCV15 followed by PPSV23, with a minimum eight-week interval between doses in immunocompromised patients.^[13,16]

COVID-19 vaccination is strongly recommended. An additional dose is indicated for adults aged 65 and older, and for immunocompromised adults, six months after the first dose of the current formula.^[14,17]

Live-attenuated vaccines (MMR, live varicella, live yellow fever, oral typhoid) are generally contraindicated while on immunosuppressive therapy, due to the risk of vaccine-strain infection in an immunocompromised host.^[13]

One consideration is specific to IVIG. Intravenous immunoglobulin contains passive antibodies that can interfere with the immune response to live vaccines, reducing their effectiveness. After a 2 g/kg IVIG dose, live vaccines should be deferred for 11 months; lower doses require shorter intervals (eight months after 300 to 400 mg/kg, ten months after 1 g/kg).^[13,18] This deferral is about efficacy, not safety: the IVIG does not make the live vaccine dangerous, but it may prevent it from working. Non-live vaccines (influenza, pneumococcal, Shingrix, COVID-19) are not subject to this restriction and should be administered on schedule.

If you are not sure whether your vaccinations are current, particularly Shingrix and pneumococcal, bring that question to your next appointment. Your team can review what you have received and what gaps remain.

This is Routine. Vaccination status review belongs at your next scheduled visit.

IVIG Safety: Thrombosis and Renal Considerations

All FDA-approved IVIG products carry a class boxed warning for two risks: thrombosis (including fatal events) and renal dysfunction (including acute renal failure).^[19] These warnings apply regardless of formulation and represent the highest-tier regulatory safety signal. They do not mean that IVIG is too dangerous to use. They mean that specific risks require active monitoring.

THROMBOSIS

The mechanism is straightforward. The large protein load of an IVIG infusion increases blood viscosity. In patients with pre-existing cardiovascular or thrombotic risk factors, that increased viscosity can trigger clotting events.^[19]

The ProDERM trial, the pivotal study for IVIG approval in dermatomyositis, recorded 8 thromboembolic events in 6 IVIG-treated patients across 664 infusion cycles (none in the placebo group).^[20] The events occurred predominantly in patients whose baseline thrombosis risk was already 2.4 to 15.2 times higher than average. When the maximum infusion rate was reduced from 0.12 to 0.04 mL/kg/min, the incidence of thromboembolic events dropped substantially.^[20] Infusion rate matters.

A larger retrospective study published in 2026, covering 609 autoimmune disease patients receiving IVIG, found thromboembolic events in 9.36% of recipients.^[21] The independent predictors identified by multivariate analysis carry practical weight: cumulative IVIG dose above 100 g, age 60 or above (odds ratio 2.91), immobility for three or more days (odds ratio 6.28), elevated pre-treatment D-dimer (odds ratio 4.56), and concurrent glucocorticoid use (odds ratio 2.04).^[21]

That last finding is directly relevant. If you are on both IVIG and corticosteroids (as many myositis patients are), the combination itself is a risk factor for thrombosis, independent of either therapy alone. Your team accounts for this in setting infusion parameters and monitoring protocols.

Risk mitigation follows from these data: adequate hydration before infusion, the lowest effective infusion rate, and awareness of symptoms (sudden leg swelling,

chest pain, shortness of breath, or sudden neurological changes) in the 24 to 48 hours following an infusion.^[19]

If you experience any of these symptoms after an IVIG infusion, this is Urgent. Seek immediate medical attention.

RENAL CONSIDERATIONS

IVIG-related kidney injury historically involved osmotic damage from sucrose-containing formulations. Most current IVIG products are sucrose-free, which substantially reduces this specific mechanism, though renal risk is not eliminated entirely.^[19] Even in dermatomyositis patients receiving IVIG for skin disease, declines in kidney function have been observed as an adverse event, reinforcing the need for monitoring even when the kidneys are not the primary concern.^[22]

People at higher risk include those with pre-existing renal insufficiency (any degree), diabetes, age above 65, volume depletion, sepsis, paraproteinemia, and concurrent use of nephrotoxic drugs.^[19] Renal impairment typically appears within ten days of infusion. Baseline serum creatinine and estimated glomerular filtration rate (eGFR) should be measured before starting IVIG and monitored during ongoing treatment, particularly in patients with risk factors.^[22]

Signs worth noting: decreased urine output, swelling in the ankles or feet, or unexplained rapid weight gain in the days following an infusion. If you notice any of these, contact your team.

This is Prompt. New signs of fluid retention or reduced urine output after an IVIG infusion warrant contact with your team within days.

Bringing It Together

The medications that control inflammatory myopathy are effective, and they are necessary. They also produce a second layer of medical complexity. Steroid myopathy can mimic the disease. Bone loss accelerates quietly. Blood sugar shifts in patterns that standard morning tests miss. Infection risk rises, predictably and measurably. IVIG carries vascular and renal considerations that benefit from active awareness.

None of these problems is invisible if you know where to look. And none requires you to manage them alone. Each one resolves into a specific question for your

team, a specific test, or a specific vaccine. The concept of treatment-driven comorbidity is not a reason for alarm. It is a framework for preparation.

Key Points

- Steroid myopathy causes painless weakness with normal CK; rising CK suggests disease activity, and the distinction requires your rheumatologist.
- Bone loss on corticosteroids begins within months and is detectable at very low doses; ask about DXA and FRAX at your next visit.
- Steroid-induced hyperglycemia peaks in the afternoon, so fasting glucose alone may miss it; ask about postprandial monitoring.
- Infection risk is highest in the first three months of immunosuppressive therapy; PJP prophylaxis status is worth confirming with your team.
- Shingrix vaccination is strongly recommended for all immunosuppressed adults aged 18 and older, with high efficacy confirmed in trial data.
- IVIG thrombosis risk increases with concurrent corticosteroid use; report sudden swelling, chest pain, or breathlessness after infusion immediately.

CHAPTER 5: MOVEMENT AND DAILY FUNCTION

In this chapter: the evidence that exercise in inflammatory myopathy is not only safe but therapeutic, replacing decades of rest advice with a calibrated, supervised approach to resistance and aerobic training. The chapter covers blood-flow-restriction training, introduces the Myositis Activities Profile as a self-tracking tool, and resolves practical questions about intensity, timing, and when to involve the rheumatology team.

The advice that made things worse

For most of the twentieth century, the standard instruction for a person diagnosed with inflammatory myopathy was to rest. The logic seemed straightforward: the immune system is attacking the muscle, so loading the muscle must make things worse. It was a reasonable assumption. It was also wrong.

No published evidence has ever supported rest restriction in inflammatory myopathy.^[1] The assumption persisted not because studies confirmed it, but because nobody tested it. Clinicians were cautious, patients were compliant, and a generation of people with treatable muscle disease spent years avoiding the very activity that could have helped them. When researchers finally brought exercise into the controlled trial, they found the opposite of what the old assumption predicted.

By 2016, Helene Alexanderson's group at the Karolinska had reviewed nearly 40 publications and concluded that exercise is safe across study populations, without increased inflammation or disease activity.^[2] Not safe in a narrow sense, for a narrow group. Safe broadly, across different subtypes and disease-activity states, in trial after trial.

The British Society for Rheumatology codified this in its 2022 guideline, issuing a strong recommendation (Grade 1B, 100% expert agreement) that management of inflammatory myopathy should include a safe and appropriate exercise programme led by a specialist physiotherapist.^[3] A second strong recommendation (1B, 96% agreement) adds that individually tailored exercise should be encouraged across all ranges of disease activity, specifically for psychological wellbeing.^[3] A separate

systematic review of all available randomized controlled trials in polymyositis and dermatomyositis reached the same endpoint: supervised physical therapy is safe and effective, in both active and established disease, with no contraindications or known adverse effects.^[4]

This is **the rest-to-exercise shift**. It is not a tentative suggestion. It is a guideline-backed reversal of historical practice, supported by decades of accumulated trial data. The rest of this chapter walks through what that evidence looks like, what it means for you, and how to put it to work with your physiotherapy team.

Aerobic training: what the trials found

The earliest controlled work came from Wiesinger's group in the late 1990s, when fourteen patients with polymyositis or dermatomyositis completed six weeks of supervised cycling and step aerobics. Creatine kinase did not increase. **VO2 max** (a measure of aerobic fitness, the maximum amount of oxygen your body can use during exercise) improved significantly, and a longer follow-up programme showed a 28% gain.^[5] For a disease that saps endurance, a 28% improvement in the body's capacity to use oxygen is not marginal. It changes what a day feels like.

The study that shifted the field's confidence more broadly was the 2013 Karolinska trial. Alemo Munters and colleagues randomized 23 patients with stable polymyositis or dermatomyositis to 12 weeks of supervised combined aerobic and resistance training or to a control group.^[6] The exercise group achieved roughly a 20% increase in VO2 max. Cycling endurance time rose by approximately 120%. Lactate levels at exhaustion dropped by about 35%, meaning muscles were working more efficiently before reaching their limit.^[6] On muscle biopsy, mitochondrial enzyme activities (citrate synthase and beta-hydroxyacyl-CoA dehydrogenase, both markers of aerobic energy production in muscle cells) increased significantly in the exercise group.^[6] The muscle was not just performing better; it was producing energy better.

Six of nine exercise patients met IMACS responder criteria for clinically meaningful improvement; none of the controls did. Crucially, vastus lateralis muscle biopsies showed no increase in inflammatory infiltrates. CK decreased or remained stable. Every patient tolerated the programme without adverse events.^[6] A multicenter

extension of this work found that gains were maintained at one year in patients who continued training.^[7]

Those results described stable, established disease. The question of whether higher-intensity work is safe in early, active disease remained open until 2025, when Andreasson and colleagues published the first multicenter randomized controlled trial of **high-intensity interval training** (HIIT) in recent-onset inflammatory myopathy.^[8] Twenty-three patients performed 30-second cycling intervals at 85% or more of peak heart rate, three times per week for twelve weeks. The result: a 16% increase in peak oxygen uptake, compared to 1.8% in the moderate-exercise group.^[8] Peak power output improved 18% in the HIIT group versus 8% with moderate exercise. No increase in CK, AST, or ALT in either group. No disease flares. Median adherence was 93%. The soreness that some participants reported was transient, resolving within 30 to 60 minutes of each session.^[8]

Two things are worth sitting with. The first is the size of that gap: 16% versus 1.8%. Higher intensity produced a dramatically larger aerobic benefit, with no additional risk. The second is that this trial enrolled patients with recent-onset disease, the group you would most expect to be vulnerable to exercise-induced harm. They were not harmed. They improved.

This does not mean you should walk into a gym and sprint on a bike. The HIIT trial was supervised, structured, and conducted with medical oversight. The principle it establishes is that intensity, appropriately calibrated and supervised, is an asset, not a hazard. This is not a minor distinction: the old rest paradigm treated all exercise intensity as risk. The data say otherwise.

Resistance training: strength, function, and what happens inside the muscle

Resistance training carries a separate evidence base, and its findings go deeper than strength gains alone.

The Jensen 2024 trial randomized 32 patients with established myositis to 16 weeks of twice-weekly progressive resistance training or usual care.^[9] The single number that captures what the training accomplished is the Timed Up and Go result: how quickly you rise from a chair, walk three metres, turn, and sit down. That improved

12.9% in the training group. For this disease, where proximal weakness makes rising from seats and climbing progressively harder, a 12.9% gain touches the texture of every hour. Alongside that, **Functional Index 3** (a validated measure of muscle endurance) and **MMT8** (manual muscle testing across eight muscle groups) both improved, and training loads across five exercises rose substantially, meaning the muscles were getting stronger in practical terms, not only on clinical scales. CK remained stable throughout. No flares occurred.^[9]

Functional endurance gains were even more dramatic in longer programmes. The Tomcik 2021 study followed 50 patients through 24 weeks of supervised training focused on daily activities, resistance, and stability.^[10] MMT-8 improved 26% (controls deteriorated over the same period). FI-2 endurance improved 135%. That is not a typographical error: muscle endurance more than doubled. HAQ disability scores improved 39%. Depression, measured by the BDI-II, improved 26%.^[10] No significant increase in CK, CRP, ESR, myoglobin, or pro-inflammatory cytokines appeared. Muscle biopsies from seven participants showed no increased infiltrates.

But the Nader 2010 study is the one that shifts the frame entirely. Eight patients with polymyositis or dermatomyositis completed seven weeks of supervised resistance training at ten-repetition maximum loads, three times per week.^[11] Strength increased. CK decreased. And then the biopsies told a story the clinical measures alone could not tell.

Complement C1q, a protein central to the inflammatory cascade that drives the immune attack in dermatomyositis, fell from 141 to 69 mg/L after seven weeks of training.^[11] That is not a marginal shift; it is the immune system's own attack signal, measurably reduced by exercise. Alongside that, inflammatory gene expression broadly decreased, fibrosis genes were downregulated, and collagen I content (a marker of muscle scarring) dropped substantially. No increase in inflammatory infiltrates appeared on biopsy.^[11]

Think of it this way. The concern was that loading inflamed muscle would be like throwing fuel on a fire. What the molecular data showed is that the right kind of loading works more like a controlled backburn: it reduces the inflammatory substrate rather than feeding it. Subsequent mechanistic work has suggested a pathway for this effect. Exercise-derived interleukin-6, released from contracting

muscle, exerts a negative feedback effect on tumour necrosis factor and shifts from a pro-inflammatory to a metabolic signalling role. Endurance training upregulates capillary growth genes and downregulates protein-degradation and immune-response genes.^[12] The exercise is not just sparing the muscle from harm. At the molecular level, it is working against the disease process itself.

That is not a metaphor for hope. It is what the biopsy slides and gene-expression profiles measured.

Blood-flow-restriction training: what it is and what the evidence supports

Blood-flow-restriction (BFR) training uses a pneumatic or elastic cuff applied to the upper portion of a limb during low-load resistance exercise. The cuff partially restricts venous return, creating a metabolic environment (metabolite accumulation, fast-twitch fibre recruitment) that mimics the training stimulus of much heavier loads.^[13] You get the muscle-building signal at 20 to 40% of your one-repetition maximum, rather than the 60 to 80% that conventional resistance training requires.

For someone managing inflammatory myopathy, the appeal is direct: a meaningful training stimulus without the mechanical stress that heavier loads impose on muscle already dealing with disease. On days when fatigue or a period of higher disease activity makes conventional resistance work feel excessive, BFR offers a way to train meaningfully at loads that feel manageable.

No randomized trial has tested BFR specifically in polymyositis or dermatomyositis. That gap is important to name. The closest evidence comes from inclusion body myositis (IBM), a related but distinct condition. The Binder 2018 trial randomized 22 patients with sporadic IBM to 12 weeks of low-load BFR resistance training or a non-exercising control group.^[13] Over three months, the controls lost 9.2% of quadriceps strength. The BFR group held steady, showing a negligible 0.9% change. The between-group difference was statistically significant.^[13] Self-reported and objective function measures (SF-36, two-minute walk, Timed Up and Go, 30-second chair stand) did not improve significantly, but strength decline was arrested. The authors interpreted the result as a preventive effect: BFR retained strength against

the natural course of disease-related decline.^[13] A follow-up immune-marker assessment confirmed that BFR did not compromise the immune system.^[14]

IBM is not your condition. Its pathology involves a degenerative component that autoimmune inflammatory myopathy does not share, and the exercise conversation in IBM is different because drug therapy is largely ineffective there. But the BFR finding carries a principle that applies broadly: low-load training with blood-flow restriction preserved strength without triggering immunological harm, even in a disease where muscle is under active stress. BFR is not contraindicated in inflammatory myopathy. It is a tool for calibrating intensity downward while preserving the training stimulus.

Two cautions belong here, and both come from what the evidence does not contain rather than what it shows. Training to failure (repeatedly exhausting the muscle to the point of concentric failure) during BFR amplifies metabolic stress beyond what was used in the published disease-population studies. The IBM trial used submaximal repetitions. Training to failure with BFR is outside the evidence base for myopathy.^[13] Similarly, supramaximal eccentric loading (forced lengthening under high tension) combined with BFR has no evidence in this population and is not supported.

Standard BFR contraindications apply regardless of your myositis diagnosis: active deep vein thrombosis, peripheral vascular disease, or open wounds at the cuff site. Cardiovascular screening should precede BFR use, since mean arterial pressure rises acutely during training.^[13] These are general safety considerations, not myositis-specific barriers.

If BFR interests you, it is a conversation for your physiotherapist, not something to set up at home from a tutorial. The principle of low-load training for periods of higher disease activity or significant fatigue is well supported by the broader resistance-training literature, whether or not a cuff is involved.

Cautions: eccentric loading and the CK timing problem

The trials that produced positive results share a common feature in their exercise design: they use predominantly concentric work (muscle shortening under load) at

submaximal intensities. No published IIM trial has tested supramaximal **eccentric loading** (forced muscle lengthening under high tension) or training-to-failure protocols.^[4] Eccentric exercise can cause transient muscle soreness and CK elevation even in healthy individuals; in muscle with compromised membrane integrity, that response may be amplified. These modalities are absent from every positive trial design. They sit outside the evidence base. The absence is not accidental. Researchers designed their protocols around concentric and moderate-intensity work precisely because those modalities carry the lowest mechanical risk to vulnerable muscle.

A related practical point: CK sampling after exercise requires attention to timing. Serum CK rises transiently after moderate-to-intense exercise in everyone, including people with inflammatory myopathy. That post-exercise rise reflects normal physiological muscle turnover, not a disease flare.^[15] Drawing blood for CK within 24 hours of a significant exercise session produces a result that cannot be interpreted as a disease-activity marker. The Lodin 2024 review explicitly advises against same-day or next-day CK sampling after a significant session.^[15] The Andreasson HIIT trial built 36-hour post-session sampling into its protocol for exactly this reason.^[8]

If your CK is being tracked as part of ongoing disease monitoring (and your rheumatology team will have established your resting baseline on stable therapy), schedule blood draws at least 48 hours after your last exercise session. A persistent rise above your personal baseline between sessions, sampled at that interval, is different from a transient post-exercise spike. The first is worth discussing with your team. The second is physiology doing exactly what physiology does.

Tracking your function: the Myositis Activities Profile

Knowing whether exercise is helping requires a way to measure what matters to you: not just muscle strength on a clinical scale, but how specific daily activities change in difficulty over time. Carrying a bag of groceries, climbing a flight of stairs, getting dressed, reaching overhead. These are the activities that define functional independence, and they are precisely what inflammatory myopathy erodes.

The **Myositis Activities Profile** (MAP) is the only disease-specific, validated, self-administered outcome measure for assessing limitations in activities of daily living in adults with polymyositis and dermatomyositis.^[16] Developed by Helene Alexanderson, Ingrid Lundberg, and Christina Stenstrom at the Karolinska and first published in 2002, the MAP was built from the ground up for this disease. Development involved iterative refinement across four patient groups totalling 85 patients, beginning with activities from an international classification framework and refined through patient ratings of both difficulty and importance.^[16]

The MAP asks you to rate 31 to 32 items across four subscales: Movement Activities (nine items), Moving Around (four items), Personal Care (nine items), and Domestic Activities (six items), plus single items covering social contact, managing overexertion, work, and recreation.^[16,17] Each item is scored from 1 (no difficulty) to 7 (impossible to do).

The instrument has been validated across multiple populations and languages. Test-retest reliability showed weighted kappa values of 0.56 to 0.77 in the original Swedish study and 0.77 to 0.93 in the US validation.^[16,18] Internal consistency (Cronbach's alpha) ranged from 0.85 to 0.95 across subscales.^[18] The German validation confirmed discriminant validity, with area-under-the-curve values of 0.72 to 0.79 for distinguishing patients from healthy controls.^[17] The MAP correlates well with the Health Assessment Questionnaire ($r = 0.69$ to 0.77) and with physical quality-of-life scales, while showing lower correlation with mental health and pain measures, confirming that it captures what it is designed to capture: physical activity limitation.^[16,18]

The ENMC workshop has recommended the MAP as a patient-reported outcome measure for adult polymyositis and dermatomyositis, and it is included in comprehensive IIM assessment sets alongside IMACS core measures and the Functional Index 3.^[19]

The MAP is a monitoring tool, not a treatment-decision tool. It gives you a validated, patient-held framework for observing which specific daily activities are getting easier or harder over time, and for bringing that information to your physiotherapy and rheumatology team with precision rather than impression. If you do not currently know your MAP scores, MMT8 result, or HAQ score, ask your team at your

next visit. These numbers are your functional baseline, and they anchor the conversation about whether your exercise programme is working and where it might need adjustment.

Fatigue, mood, and quality of life

Exercise in stable inflammatory myopathy improves SF-36 physical and vitality domains, with gains maintained at 52-week follow-up in patients who continued training.^[6,7] Depressive symptoms, measured by the BDI-II, improved by 24 to 26% in the Tomcik and Spiritovic controlled studies of supervised training.^[10] A 26% reduction in depression scores is clinically meaningful, and it emerged from a physical training programme, not a psychological intervention.

Quality-of-life outcomes across the broader literature are mixed. Some trials found significant SF-36 improvement; others did not. The Jensen 2024 trial showed a 12% improvement in the Physical Component Summary, while other measures in other studies failed to reach significance.^[9,15] No study has been designed with pain or fatigue as its primary outcome in inflammatory myopathy exercise research; that is an identified evidence gap.^[15] The BSR 2022 guideline's strong recommendation for exercise across all disease-activity ranges specifically for psychological wellbeing reflects expert consensus that the psychological benefit is sufficiently supported, even where individual study results are inconsistent.^[3]

This is worth noting honestly. The physical evidence (strength, endurance, aerobic capacity, molecular markers) is strong and consistent. The psychological evidence points in the right direction but is less uniform across studies. If you find that exercise improves your mood and energy, that aligns with what several studies report. If you find that it does not, that also aligns with the literature. What no study has shown is psychological harm from supervised exercise in this disease.

The IBM exception

Inclusion body myositis (IBM) appears here as a boundary marker, not as a condition this chapter applies to. IBM involves a degenerative, treatment-refractory pathology distinct from autoimmune inflammatory myopathy. Over 20 drug therapies, including IVIG, corticosteroids, and multiple immunomodulatory agents, have failed to show meaningful benefit in controlled IBM trials.^[20] In IBM, exercise is

the primary management strategy precisely because pharmacological options are exhausted; regular exercise is currently the only recognised approach effective at limiting muscle weakening, atrophy, and loss of function in that disease.^[20]

The exercise principles in this chapter apply to controlled, immunotherapy-responsive inflammatory myopathy. IBM represents a different clinical situation, and the reader should not draw parallels between the two simply because both involve muscle disease.

When to involve your team

The principle is: start under supervision, progress gradually, choose predominantly concentric work, and monitor symptom response rather than CK drawn the same day as exercise.^[4,15]

Supervision by a physiotherapist familiar with inflammatory myopathy is the consistent feature of every safe, beneficial exercise programme in the published literature. Home exercise without adequate supervision was associated with insufficient intensity and no short-term improvement in the Alexanderson 2014 study of recent-onset disease, where only the longer follow-up (one to two years) showed gains in the exercised group.^[2] The supervision is not optional decoration. It is part of what makes the intervention work.

Escalation to your rheumatology team or specialist physiotherapist is indicated if CK rises persistently above your personal resting baseline between sessions (sampled at least 48 hours after your last exercise), if muscle weakness progresses despite regular training, or if new symptoms emerge (difficulty swallowing, breathlessness, or skin changes). Any of these warrants a prompt conversation with your team rather than waiting for your next scheduled visit.

Two practical slots that only your team can fill. First, if you receive IVIG infusions, the interaction between infusion timing and exercise scheduling (whether to train in the days immediately surrounding an infusion, and how to manage energy in the infusion week) is not addressed in the published literature and requires guidance from your treating team. Second, if you have interstitial lung disease or cardiac involvement, the exercise risk calculation changes; the Lodin 2024 review identifies

these as evidence gaps, and intensity calibration in that context belongs with the rheumatology or cardiopulmonary team.^[15]

Ask your team about your current functional level: your MAP subscale scores, your MMT8 result, your HAQ score. These give you and your physiotherapist a starting point that is measured rather than guessed. And ask about access to a physiotherapist who has worked with inflammatory myopathy specifically. That expertise is what bridges the trial evidence into your individual programme. The protein supply that sustains muscle through training is covered in Chapter 6.

Key Points

- Decades of trial data confirm that supervised exercise in inflammatory myopathy is safe and improves strength, endurance, and aerobic capacity without increasing inflammation.
- Resistance training does not merely avoid harm; molecular evidence shows it actively downregulates inflammatory gene expression in affected muscle.
- Blood-flow-restriction training delivers a meaningful strength stimulus at low mechanical loads, and it is not contraindicated in inflammatory myopathy.
- CK drawn shortly after exercise reflects physiological turnover, not disease activity; allow adequate recovery time before blood draws used for disease monitoring.
- The Myositis Activities Profile is a validated, self-administered tool for tracking how specific daily activities change in difficulty over time.
- If weakness progresses despite training, or new symptoms such as breathlessness or swallowing difficulty emerge, contact your rheumatology team promptly.

CHAPTER 6: NUTRITION

In this chapter: what the evidence actually shows about eating with myositis and its treatment. Protein needs under corticosteroids, bone-protective nutrients, glucose management, texture-modified eating for swallowing difficulty, and a plain grading of the anti-inflammatory-diet claims you will encounter online. The chapter introduces evidence-graded nutrition as its organizing principle: every recommendation is weighted by the strength of its source, so you can tell the difference between what the guidelines support and what remains hope.

The gap that matters

A disease with this much inflammation, involving muscle tissue that depends on nutrients to survive and repair, would seem a natural target for dietary research. The reasonable expectation is that nutrition guidance for myositis would be detailed, well-tested, and confident.

No one has run a randomized controlled trial testing whether any named diet changes the course of idiopathic inflammatory myopathy. Not the Mediterranean diet. Not an anti-inflammatory protocol. Not a specific macronutrient ratio. A 2024 scoping review examined every published study it could find on nutritional interventions in IIM and concluded that the evidence represents “preliminary but insufficient” grounds for definitive clinical recommendations.^[1] That single sentence is the most important nutritional fact in this book.

It does not mean that eating well is irrelevant. It means something more precise: no diet has been shown to reduce your disease activity, lower your CK, or improve your muscle strength in a controlled study. The treatments your rheumatology team prescribes do that. What nutrition can do, and what the evidence does support, is protect your bones while corticosteroids thin them, supply the protein your muscles need while multiple forces erode them, keep your blood glucose steadier while steroids push it up, and maintain adequate intake if swallowing becomes difficult.

That is the framework this chapter uses. Call it **evidence-graded nutrition**: each recommendation is weighted by how strong its underlying source is, and where the source is thin, the chapter says so plainly rather than dressing a weak claim in

confident language. The difference between a society-guideline recommendation and a plausible-but-untested extrapolation is a difference you deserve to see.

Protein and the muscle problem

Three forces converge on muscle in myositis, and none of them waits for the others. The autoimmune process damages myofibers directly. Corticosteroids, if prescribed, break down muscle protein through both genomic and non-genomic catabolic pathways, with a particular preference for the type II (fast-twitch) fibers that generate explosive strength.^[2] And if you are in your fifties, sixties, or beyond, age-related **sarcopenia** layers its own loss on top. In any individual patient these three streams cannot be cleanly separated. They are additive.

Think of your muscle tissue as a bank account with three open withdrawals running simultaneously: the disease, the medication, and the clock. Protein intake is the deposit side. The question is how much is enough.

The best available number comes from a 2022 review in *Rheumatology Advances in Practice* focused specifically on patients receiving prolonged glucocorticoid therapy. Its recommendation: 1.0 to 1.5 grams of protein per kilogram of body weight per day.^[3] The ESPEN expert group, looking at older adults with acute or chronic illness more broadly, arrived at a similar range of 1.2 to 1.5 g/kg/day.^[4] A 2025 study using indicator amino acid oxidation (a newer measurement technique) found that older adults with established sarcopenia needed approximately 1.54 g/kg/day, roughly 30 percent above the estimate for those without sarcopenia.^[5]

These are guideline-grade recommendations, but they come with a candid caveat. The *Rheumatology Advances in Practice* authors noted explicitly that “there is a dearth of empirical data on how nutritional intervention might impact health-related outcomes in this population.”^[3] The numbers are the best starting points the evidence offers. They are not derived from myositis-specific trials. A 2025 systematic review across nine randomized trials on protein intake and sarcopenia found “inconsistent results” across muscle mass, physical performance, and strength outcomes, concluding that the evidence remains “unclear.”^[6] Most of those studies enrolled postmenopausal women. None enrolled IIM patients. The precision of any protein prescription has limits.

One detail matters at the meal level. **Leucine**, the amino acid most potent at triggering muscle protein synthesis through the mTOR pathway, requires a higher threshold in older adults to achieve the same anabolic signal: approximately 3 to 4 grams per meal.^[7] Older adults develop what researchers call anabolic resistance: the same protein bolus that triggers robust muscle building in a younger person produces a blunted response. Leucine-dense protein sources (dairy, eggs, poultry, fish, soy) help clear that higher threshold. This is why protein source quality, not just quantity, becomes relevant when sarcopenia risk is present, even though no myositis-specific study has tested this directly.

The synergy between protein intake and resistance exercise is well established in the broader sarcopenia literature: the combination outperforms either intervention alone for muscle mass and strength.^[8] Neither protein supplementation alone nor exercise alone achieves the same effect size. Chapter 5 covers the exercise evidence. The nutritional implication is straightforward: protein intake and physical activity are partners, not alternatives. One without the other leaves something on the table.

To translate any of these per-kilogram recommendations into a practical daily target, you need two numbers from your team: your current body weight and your kidney function (eGFR). Chronic kidney disease stages 3 through 5 require a lower protein ceiling under specialist supervision.^[3] If you do not already know these values, ask your rheumatology team or GP at your next visit. This is a Routine item.

Protecting bone with nutrients

Corticosteroids impair bone through multiple mechanisms: they reduce intestinal calcium absorption, increase renal calcium excretion, suppress the osteoblasts that build new bone, and alter sex hormone levels that contribute to bone maintenance.^[9] Bone loss begins early and is detectable even at low corticosteroid doses; Chapter 4 covers the fracture risk categories, clinical thresholds, and pharmacological bone protection that your team manages. This chapter covers only the nutritional building blocks.

The 2022 American College of Rheumatology guideline for glucocorticoid-induced osteoporosis is specific about what those building blocks are.^[9] Total daily elemental calcium, from diet and supplements combined, should reach 1,000 to 1,200

milligrams. Vitamin D should be supplemented to maintain a serum 25(OH)D level of at least 30 to 50 ng/mL, with 600 to 800 IU per day as a starting dose. A Japanese guideline review reached similar conclusions, emphasizing that nutritional interventions including calcium and vitamin D are necessary to maximize the benefits of any pharmacological bone protection agent the team might prescribe.^[10]

Two practical points deserve attention. The ACR guideline notes that intermittent high-dose vitamin D boluses (megadoses given monthly or quarterly) may actually increase fall risk.^[9] Daily supplementation at the recommended range is preferred over periodic large doses. And the form of calcium supplement matters if you take an acid-suppressing medication such as a proton pump inhibitor (a common co-prescription with corticosteroids). **Calcium carbonate** requires stomach acid for absorption and should be taken with food. **Calcium citrate** does not depend on gastric acid and can be taken without food.^[3] If you use a PPI, citrate is the better choice.

Dietary calcium sources include dairy products, fortified plant milks, leafy green vegetables, tofu set with calcium salts, and almonds. For many patients under dietary restrictions, supplements are needed to close the gap between food intake and the 1,000 to 1,200 mg target.

Vitamin D has a second dimension beyond bone. Observational studies across multiple autoimmune rheumatic diseases, including IIM, consistently find high rates of vitamin D insufficiency, and a 2024 review confirmed that insufficient vitamin D is associated with unfavorable outcomes in these conditions.^[11] A case report documented dramatic improvement in myopathic condition after correction of severe vitamin D deficiency in an IIM patient.^[12] Whether supplementing above bone-protection targets improves disease activity in established IIM remains unproven. The evidence is observational and anecdotal, not from controlled trials. For now, the bone-protection target is the evidence-based floor.

If you do not know your current serum 25(OH)D level or your current calcium and vitamin D supplement doses, confirming these with your team is a Routine check worth raising at the next scheduled visit.

Glucose and weight under corticosteroids

Corticosteroids dysregulate blood glucose through a characteristic pattern. They increase peripheral insulin resistance, ramp up hepatic glucose production, and impair the pancreatic beta cell response. The result is typically postprandial hyperglycemia peaking four to six hours after a morning dose of prednisolone, with fasting glucose remaining deceptively normal.^[13] That timing mismatch matters: a normal fasting blood draw can miss a glucose problem that peaks every afternoon.

Risk rises with dose (above a prednisolone equivalent of approximately 5 mg per day), advancing age, higher body mass index, and pre-existing impaired glucose tolerance.^[13] If you are still on corticosteroids at any dose, this applies to you. If you have fully tapered off, the acute glucose concern recedes, though the metabolic effects of prior exposure do not vanish overnight.

The dietary response is well established in general diabetes management, even though it has not been tested in IIM-specific trials. Low-glycemic-index and low-glycemic-load eating patterns have consistent evidence for improving fasting glucose, HbA1c, and weight in type 2 diabetes management.^[14] Reducing ultra-processed foods and high-glycemic-index refined carbohydrates, while increasing unprocessed and minimally processed foods, directly counteracts the glucose spikes that corticosteroids produce.^[3] The same rheumatology-specific review recommends keeping sodium below 1,500 mg per day to address glucocorticoid-related fluid retention and blood pressure elevation.^[3]

Alcohol adds a complication. It interacts with multiple immunosuppressive agents and with glucose regulation. Both the glucocorticoid nutrition review and the ACR guideline recommend avoiding excessive alcohol, though no IIM-specific alcohol guidance exists.^[3]

Weight gain on corticosteroids introduces a second, subtler problem. Steroids increase appetite directly, leading to caloric excess. But the weight that accumulates is not straightforward. Fat mass rises while muscle mass simultaneously falls, creating what researchers call **sarcopenic obesity**: a BMI that may look stable or even improved while functional muscle loss continues underneath.^[15] A longitudinal study in juvenile dermatomyositis demonstrated this phenomenon clearly: BMI percentile improved over time on treatment, but total body fat percentile remained

elevated at 60 months, raising the concern that the BMI improvement partly reflected muscle wasting rather than genuine fat loss.^[15] The same pattern is directly relevant to adult IIM patients interpreting their own weight trajectory.

The practical implication is that body weight alone is a poor compass. What matters is whether you are maintaining functional strength (Chapter 5 covers how to measure and track this) while managing caloric intake to match your activity level. A scale cannot distinguish between a kilogram of muscle and a kilogram of fat.

If you do not know your current fasting glucose or HbA1c, or whether a diagnosis of steroid-induced hyperglycemia has been made, this is worth raising with your team. If you are still taking corticosteroids above 5 mg per day, glucose monitoring and dietary carbohydrate moderation become more pressing. This is a Routine discussion for the next scheduled visit; if you notice symptoms of persistently high blood sugar (excessive thirst, frequent urination, blurred vision), that becomes a Prompt item to contact your team within days.

When swallowing is affected

Oropharyngeal dysphagia (difficulty swallowing arising from inflammatory involvement of the throat and upper esophageal muscles) is more common in myositis than most patients realize, and aspiration pneumonia is its most serious consequence. Chapter 7 covers the organ-system detail, including prevalence figures and aspiration risk. What this chapter addresses is what happens to nutritional adequacy when food texture must be modified.

Immunomodulatory treatment, including corticosteroids and IVIG, is associated with symptomatic improvement in swallowing.^[16] Treatment helps. But the nutritional dimension of dysphagia is distinct from the clinical swallowing assessment and management, which Chapter 7 covers in its organ-system awareness section.

The **International Dysphagia Diet Standardisation Initiative** (IDDSI) provides a standardized eight-level framework, numbered 0 through 7, for classifying liquid thickness and food texture.^[17] Drinks occupy levels 0 through 4; foods occupy levels 3 through 7. The framework is used across clinical settings and countries, providing a common language between speech-language therapists, dietitians, and patients. A

clinical speech-language pathology assessment determines which level is safe and appropriate for each individual. This is not something to self-prescribe.

Here is the nutritional problem. Texture-modified diets are consistently associated with lower energy and calcium intake compared to regular diets.^[18] A systematic review across twenty studies found increased risk of weight loss and malnutrition when patients shifted to modified textures.^[18] Appetite suppression is a documented secondary effect; pureed and softened foods are simply less appealing to most people, and meal volumes tend to drop. For someone already managing the sarcopenia overlay described earlier in this chapter, that nutritional shortfall compounds an existing vulnerability.

The good news is that structured supplementation strategies can partially offset this deficit. A 2025 meta-analysis of randomized trials found a significant positive effect on both energy and protein intake when texture-modified diets were supplemented with high-protein fortification and caloric density enhancement.^[19] Specific approaches include fortifying pureed foods with high-protein additions and increasing the caloric density of each serving. The strategy works. But it requires professional guidance: a registered dietitian experienced in dysphagia, working alongside the speech-language therapist who prescribed the texture level.

If you have any difficulty swallowing (coughing during meals, food sticking, a sense of food going the wrong way, or unexplained chest infections), report this to your rheumatology team. Do not modify your food texture on your own. Unsupervised texture modification is both unsafe and nutritionally inadequate without professional assessment. This is a Prompt item: contact your team within days, not at the next scheduled visit.

Anti-inflammatory diet claims: what the evidence shows

You will encounter claims, online and in patient communities, that specific dietary patterns can reduce inflammation and modify the course of myositis. The most common candidate is the Mediterranean diet. Here is what the evidence actually shows, graded honestly.

GENERAL ANTI-INFLAMMATORY EFFECTS

Meta-analyses of randomized controlled trials have demonstrated that the Mediterranean diet reduces general inflammatory biomarkers (IL-6, CRP, TNF-alpha) across mixed chronic disease populations.^[20] A separate meta-analysis of Mediterranean diet supplemented with olive oils found significant reductions in IL-6 and CRP compared to control diets.^[20] Those findings are solid for general inflammation. They are also not the same thing as reducing disease activity in IIM. General inflammatory markers are not disease-specific markers. No study has tested whether Mediterranean eating changes CK levels, muscle strength, or clinical outcomes in myositis patients. The 2024 scoping review identifies the Mediterranean diet as the most “biologically plausible” candidate but states the case remains unproven.^[1]

The Myositis Association states directly on its website that “there is no specific diet or food that is proven to help with myositis.”^[1] The Hospital for Special Surgery nutrition guidance for myositis aligns with this framing. Neither source cites IIM-specific trial evidence. Both position anti-inflammatory eating as general wellness support, not as treatment. That distinction is the honest one.

OMEGA-3 FATTY ACIDS

Omega-3 fatty acids (EPA and DHA) inhibit proinflammatory mediators and promote the resolution of inflammation through resolvins and protectins. Clinical trial data from rheumatoid arthritis and systemic lupus erythematosus show modest reductions in disease activity scores.^[21] No IIM-specific trials exist. The VITAL trial tested omega-3 supplementation at 1,000 mg per day for autoimmune disease prevention and found a non-statistically significant reduction (hazard ratio 0.85, $p = 0.19$).^[22] The extrapolation to myositis is biologically plausible. It is not proven.

VITAMIN D BEYOND BONE PROTECTION

The VITAL trial, enrolling 25,871 participants followed for a median of 5.3 years, found that vitamin D supplementation at 2,000 IU per day reduced the incidence of confirmed new autoimmune disease by 22 percent compared to placebo.^[22] That is a notable finding from a large, well-designed trial. Two qualifications matter. VITAL was a primary prevention trial: it studied people who did not yet have autoimmune disease. And it does not address whether vitamin D supplementation changes

outcomes in someone who already has established IIM. The bone-protection case for vitamin D supplementation (covered earlier in this chapter) stands on its own. The disease-modification case does not, yet.

CREATINE

A 2013 Cochrane systematic review found one trial (37 participants with idiopathic inflammatory myopathies) showing significant improvement in functional performance with **creatine** supplementation, with no clinically relevant adverse events.^[23] Creatine supplementation in older adults generally supports muscle mass and strength gains when combined with resistance exercise.^[23] No larger or more recent IIM-specific creatine trial has been published since. One small, unreplicated trial. The finding is suggestive, not settled. Creatine is renally excreted, so kidney function needs to be confirmed before starting; discuss this with your team.

THE HONEST SUMMARY

No diet treats myositis. No specific macronutrient ratio has been shown to reduce myositis-specific inflammatory markers, enzyme levels, or muscle strength outcomes in a controlled trial. The Mediterranean diet has plausible biological rationale and genuine evidence for reducing general inflammatory biomarkers in other conditions. Whether that translates to clinical benefit in IIM is an open question with no current answer.

This does not mean you should ignore what you eat. The protein, bone, glucose, and texture considerations earlier in this chapter are supported by society guidelines and have clear, practical value for managing the consequences of the disease and its treatment. The distinction is between those evidence-based targets and the broader claim that a dietary pattern can modify the disease itself. The first is well supported. The second is not, yet.

A note on IVIG and eating

If IVIG is your maintenance therapy, it does not impose a general dietary restriction. One practical consideration: some IVIG formulations use glucose as a stabilizer, which matters if you also have glucocorticoid-related glucose intolerance.^[24] Some preparations use maltose or sucrose, relevant to specific intolerances. If you do not know whether your specific IVIG product uses glucose stabilization, ask your

infusion team. Most patients are advised to stay well hydrated around infusion days and to avoid heavy meals and alcohol in the 24-hour window surrounding an infusion, but this is infusion-management guidance rather than a long-term dietary strategy.

Key Points

- No randomized controlled trial has tested any diet as a treatment modifier for adult myositis; claims to the contrary outrun the evidence.
- Guideline-recommended protein intake supports muscle under corticosteroid and sarcopenia pressures, with kidney function determining the upper ceiling.
- Calcium and vitamin D supplementation at guideline-recommended doses are backed bone protection while on glucocorticoids; confirm your targets with your team.
- Corticosteroid-driven glucose spikes peak hours after a morning dose; fasting glucose alone can miss the problem entirely.
- If swallowing becomes difficult, report it promptly; unsupervised texture modification risks malnutrition and is unsafe.
- Mediterranean and anti-inflammatory diets reduce general inflammatory markers in other conditions but remain unproven for myositis specifically.

CHAPTER 7: ORGAN-SYSTEM AWARENESS AND WHEN TO ESCALATE

In this chapter you learn that myositis can reach beyond muscle into the lungs, throat, skin, and heart, and that knowing what to watch for, and when to call, is the single most useful thing you can carry out of these pages. Each organ domain resolves into a triage tier so you know whether something is a question for the next visit, a reason to call this week, or a reason to seek care now.

Myositis is a muscle disease. That sentence is true, and it is incomplete.

The inflammatory process that targets skeletal muscle fibers can also involve the lungs, the swallowing mechanism, the skin, and, less commonly, the heart. The word for this is multisystem, and it is one reason that the rheumatology team monitors organ systems you might not have connected to a muscle condition. This chapter covers each of those organ domains in turn: what the risk is, how it is detected, what you should watch for at home, and at what point you should contact your team. It does not cover management. Management belongs to the specialist team.

The concept that ties these sections together is what this book calls **awareness-plus-escalation**: knowing enough about each organ system to notice a change, and having a clear rule for what to do next. You are not diagnosing anything. You are noticing and routing. The difference is real and it carries clinical weight: the rheumatology team handles the diagnosis; you handle the noticing.

Three tiers organize the routing, the same vocabulary the front matter introduced. Routine means worth raising at your next scheduled visit. Prompt means worth a call to the team within days. Urgent means worth same-day or emergency attention.

The Lungs: Interstitial Lung Disease

Of all the organ systems myositis can involve, the lungs carry the heaviest consequences. IIM is the connective tissue disease category with the highest prevalence of associated **interstitial lung disease** across all subtypes.^[1,267] ILD is not a complication that develops only in advanced or poorly controlled disease. It is a leading cause of morbidity and mortality in IIM, including in antisynthetase

syndrome, and it can be present and progressing before you develop any noticeable respiratory symptoms.^[2]

That last point deserves a pause. Subclinical ILD (lung involvement documented on imaging but not yet producing symptoms) has been confirmed in IIM cohorts.^[3] This is why screening exists: it catches what you cannot feel yet.

WHY SCREENING MATTERS

Interstitial lung disease involves the tissue between the air sacs, the structural scaffolding that holds the lung together. By the time that damage produces a symptom you notice (shortness of breath, a persistent cough), it may have been progressing for months. Screening catches it before symptoms arrive.

The 2025 ERS/EULAR guideline recommends high-resolution computed tomography (HRCT) for all IIM patients except those with inclusion body myositis, even in the absence of respiratory symptoms.^[4,4320] The 2023 ACR/CHEST guideline recommends myositis-specific antibody testing in all IIM patients to identify those at higher ILD risk, and pulmonary function tests at baseline with annual rescreening for high-risk antibody carriers.^[5]

Pulmonary function tests (PFTs) serve as a ruler laid alongside your lung capacity at a known moment. The team records your forced vital capacity (FVC) and your diffusing capacity for carbon monoxide (DLCO) at diagnosis. Those numbers become your personal baseline. If either value drops by more than ten percent from that baseline on follow-up testing, that decline should trigger HRCT imaging and an urgent clinical review.^[6] Ten percent is the threshold. It is specific, it is measurable, and you can ask your team for your numbers at any visit.

Serial PFTs are recommended every three to six months during early disease and at least every six to twelve months in patients with established ILD.^[4,4320] The six-minute walk test with pulse oximetry is another clinical tool; oxygen saturation dropping below 90 percent or falling more than four percentage points during walking is considered clinically significant and is associated with poorer outcomes.^[7]

If you do not know whether you have had baseline HRCT or PFTs, that is a question for your next appointment. If you do not know your FVC and DLCO values, ask for them. They are your numbers.

THE ANTIBODY CONNECTION

Your antibody profile, which the team determined during diagnosis, shapes your ILD risk tier. Antisynthetase antibodies (anti-Jo-1 is the most common, but anti-PL7 and anti-PL12 also belong to this family) are associated with ILD in 67 to 100 percent of cases, depending on the specific antibody.^[8] Anti-PL7 and anti-PL12 carry higher rates of rapid progression and isolated ILD presentation than anti-Jo-1.^[8]

Anti-MDA5 antibodies carry a different and more acute danger: rapidly progressive ILD, a form that can deteriorate within weeks. In one cohort of 125 anti-MDA5-positive patients with ILD, rapidly progressive ILD occurred in 37.6 percent. Among those who developed RP-ILD, short-term mortality was 53.2 percent.^[9] That figure justifies the Urgent tier for any sudden change in breathing.

Anti-Ro52 co-positivity with any myositis-specific antibody is associated with more severe ILD, more frequent treatment resistance, and poorer prognosis.^[8] The ERS/EULAR guideline identifies antisynthetase, anti-MDA5, and anti-Ro52 antibodies as the principal high-risk markers for ILD development in IIM.^[4,4320]

Because your subtype and antibody profile determine which of these risk tiers applies to you, and because the book does not know your results, the thing to do is ask your rheumatology team where you sit. That question appears in Chapter 9.

WHAT TO WATCH FOR

Changes you should notice and report:

- New or increasing shortness of breath on exertion, even mild exertion you previously tolerated well.^[8]
- A persistent dry cough that is new or worsening.^[3]
- Reduced ability to take a full deep breath or to sustain exertion without stopping.^[8]
- Chest tightness or discomfort with activity.^[10]
- Any change in exercise tolerance that is out of proportion to muscle weakness alone.^[6] This distinction matters: if you are weaker but not

breathless, the issue may be muscular. If you are breathless beyond what your muscle weakness explains, the lungs may be involved.

- Rapid onset of severe breathlessness within weeks. This is a red flag for rapidly progressive ILD, particularly if you carry anti-MDA5 antibodies.^[9]

THE EXERCISE INTERSECTION

ILD modifies the safety context for exercise. If you have known or suspected ILD, your exercise programme should not be established or progressed without a prior respiratory assessment, because exercise-induced oxygen desaturation can occur without you noticing it.^[4,4320] Any new symptom during or after exercise (breathlessness, palpitations, sustained dizziness) should be reported before physical activity is escalated further.^[7]

Specific exercise prescription and aerobic tolerance thresholds belong to the treating respiratory or rheumatology team. This is not self-management territory.

ESCALATION: LUNGS

New breathlessness on exertion, a new persistent dry cough, any episode of oxygen desaturation detected on a home pulse oximeter, or any acute deterioration in exercise tolerance: report these to the rheumatology or respiratory team Promptly. If the onset is rapid (over days to weeks rather than months), this is Urgent. Do not wait for a scheduled appointment.

The Throat: Dysphagia and Aspiration

Swallowing is a muscular act, and myositis is a muscle disease. The connection is direct, and the consequences of missing it are serious.

A 2020 systematic review and meta-analysis covering 10,382 subjects found that **dysphagia** affects approximately 36 percent of IIM patients across subtypes when assessed by questionnaire.^[11] When clinicians use instrumental assessments (videofluoroscopy or fiberoptic endoscopic evaluation), the detection rate rises as high as 82 percent. That gap between what patients report and what instruments detect tells you something important: swallowing difficulty in myositis can be subtle, gradual, and easy to dismiss as normal aging or not worth mentioning.

Prevalence varies by subtype. In the Speyer meta-analysis, dermatomyositis showed a 31 percent rate, polymyositis 23 percent, cancer-associated myositis 51 percent, and NXP2-positive myositis 56 percent.^[11] Dysphagia can also be the initial or sole symptom of myositis, appearing before muscle weakness becomes prominent. It is not exclusively a late-stage feature.

HOW SWALLOWING GOES WRONG

Swallowing is a coordinated sequence: the tongue pushes food backward, the soft palate lifts to seal off the nasal passages, the larynx rises, the epiglottis folds over the airway, and the pharyngeal muscles contract in a wave that pushes the bolus into the esophagus. The whole process takes less than a second, and it depends on muscles working in precise coordination.

The dysphagia of IIM is primarily neuromuscular. It reflects impaired endurance and coordination of the oropharyngeal swallowing muscles (pharyngeal weakness, **cricopharyngeal dysfunction**, reduced laryngeal elevation) and sometimes the esophagus (smooth muscle involvement, hypomotility).^[12] This is not mechanical obstruction. It is muscle that is not performing its sequence reliably. When the sequence breaks down, food or liquid can enter the airway.

WHY ASPIRATION MATTERS

Aspiration pneumonia occurs four times more frequently in dysphagic versus non-dysphagic IIM patients.^[11] Aspiration pneumonia with respiratory failure is identified as a leading cause of mortality in myositis.^[13] Those two facts, taken together, make dysphagia more than a quality-of-life issue. It is a safety issue.

One critical detail: **silent aspiration** (food or liquid entering the airway without triggering a cough) can occur in myositis-related dysphagia.^[11] The absence of coughing does not mean swallowing is safe. This is why the BSR 2022 guideline recommends routine dysphagia assessment in all adult IIM patients and speech and language therapist involvement when dysphagia is identified.^[14]

Texture modification (adjusting the consistency of food and liquids to reduce aspiration risk) is a tool the speech and language therapist may recommend after formal assessment. It is not a self-management intervention and should not be applied without professional guidance.^[14] Chapter 6 covers the nutritional implications of texture-modified eating.

WHAT TO WATCH FOR

- Coughing or choking during or immediately after eating or drinking.^[11]
- The sensation that food is sticking in the throat or chest.^[13]
- Difficulty starting a swallow, or the need for repeated swallowing attempts.^[11]
- Frequent throat clearing at mealtimes.^[11]
- Regurgitation of food, especially undigested material.^[12]
- Unexplained weight loss or deteriorating nutritional status.^[11]
- Recurrent chest infections or pneumonia, which may reflect silent aspiration.^[11]
- A change in voice quality, becoming wet or gurgling after eating.

If you have not yet had a formal swallowing assessment, that is a question for your team at the next visit. The question is straightforward: “Has my swallowing been formally assessed, and should it be?”

ESCALATION: SWALLOWING

Any swallowing symptoms that have developed since the last review are worth raising at a Routine appointment. If coughing is frequent during meals, if there is unintentional weight loss, or if there has been a chest infection without an obvious cause, report Promptly (do not wait for a routine visit). Recurrent or aspiration pneumonia requires Urgent review.

The Skin: Cutaneous Disease, Photosensitivity, and Calcinosis

Skin involvement in myositis is most characteristic of the dermatomyositis subtype, but several features are relevant across IIM subtypes. The skin is also the organ system where you have the most direct visibility; you can see changes that the lungs or heart would hide. That visibility is an advantage, provided you know what to look for.

RECOGNIZING THE RASH PATTERNS

Two features are pathognomonic (essentially diagnostic) for dermatomyositis.

Gotttron papules are erythematous to violaceous papules or plaques overlying the knuckles, elbows, and knees. The **heliotrope rash** is violaceous or pink erythema of the upper eyelids, often with periorbital edema.^[15] These are the features that most reliably point a clinician toward DM.

Beyond these, several other patterns are recognized. The **shawl sign** (erythema across the posterior neck and shoulders) and the **V-sign** (erythema of the anterior neck and upper chest in a V-distribution) follow photosensitive distribution patterns.^[15] The **holster sign** (erythema on the lateral hips and thighs) and scalp involvement with violaceous erythema or psoriasiform plaques are also documented.^[15] **Periungual telangiectasias** (dilated blood vessels visible at the nail folds) may serve as a visible marker of disease activity.^[16]

Mechanic's hands (rough, cracked, hyperkeratotic skin on the lateral fingers and palmar surface, sometimes with horizontal fissuring) are a characteristic feature more commonly associated with antisynthetase antibodies.^[4,4320] **Poikiloderma** (combined hyperpigmentation, hypopigmentation, telangiectasia, and epidermal atrophy) develops in chronic or inadequately treated skin disease and represents accumulated damage over time.^[15]

One practical concern: in darker skin tones, the characteristic violaceous and erythematous changes may be more difficult to detect visually.^[17] This creates a real risk of delayed recognition. If you have darker skin, a lower threshold for reporting any new skin change to your team is reasonable.

PHOTOSENSITIVITY: MORE THAN SUNBURN

Up to 50 percent of patients with dermatomyositis have photosensitive skin disease.^[18] That number is important, but the mechanism underneath it is more important still.

Both UVA and UVB radiation contribute to disease flares. Here is where it gets counterintuitive. UVA penetrates window glass. It is present year-round, on cloudy days, and from some indoor lighting sources (halogen and fluorescent). In DM-susceptible skin, UVA generates reactive oxygen species and triggers type I interferon pathways.^[18] UV exposure in dermatomyositis can worsen not only skin

lesions but also systemic disease activity, including muscle symptoms.^[18] Sunlight is not just a skin irritant here. It is a systemic disease trigger.

The BSR 2022 guideline recommends sun avoidance and regular use of high-factor broad-spectrum sunscreen.^[14] Expert opinion recommends broad-spectrum (UVA and UVB) SPF 30 or higher applied daily; some specialist centers recommend SPF 70 or higher for patients with active or photosensitive skin disease.^[19] Protective clothing with UPF 50+ rating and wide-brimmed hats are recommended for outdoor exposure.^[19]

The UVA-through-glass detail is the one that catches most people off guard. If your disease involves photosensitivity, sun protection is a year-round, indoor-and-outdoor practice, not a summer holiday measure. Driving with unprotected hands and forearms, sitting near a window on a cloudy afternoon: these are exposures that matter when your skin reacts to UVA. Whether photosensitivity applies to you depends on your subtype and disease pattern, which is a question for your team.

CALCINOSIS

Calcinosis (calcium deposits in the skin, subcutaneous tissue, or muscle) occurs in approximately four to six percent of adults with dermatomyositis in rigorous cohort data.^[20] It typically presents as firm, white or yellowish subcutaneous nodules, most commonly over pressure points and joints (elbows, knees, buttocks, axillae, abdomen).^[21]

Risk factors include younger age at diagnosis, anti-NXP2 antibody positivity (the strongest antibody-level predictor), skin ulceration, and dysphagia.^[20] In adult DM, calcinosis is more commonly a feature of advanced or inadequately controlled disease; early effective immunosuppression is associated with reduced incidence.^[17]

Complications of calcinosis include skin ulceration, secondary infection, and functional restriction around affected joints.^[21] Management is entirely within the clinician's remit. Your role is to notice and report. If you feel a new hard lump under the skin, particularly over a pressure point, bring it to the team's attention.

WHAT TO WATCH FOR

- Any new hard lumps beneath the skin, particularly over pressure points or joints.^[21]

- Skin breaking down or ulcerating over a hard subcutaneous area.^[21]
- Worsening or new skin rash in areas exposed to sunlight or after sun exposure.^[14]
- Increased redness or swelling around existing rash areas, which may indicate a flare.
- Any new or changing skin lesion in a patient with known dermatomyositis.^[15]

ESCALATION: SKIN

New hard subcutaneous nodules, any skin ulceration, or skin that appears to be breaking down should be reported at the next appointment and Promptly if there are signs of infection (warmth, pus, fever). Rash that is worsening despite current treatment should be reported between appointments. Sun-related flares of skin disease affecting areas you have not seen involved before are worth a Prompt call, because they may signal that your sun-protection practice needs review with the team.

Cancer Association and the Rationale for Screening

This is the section people tend to skip or fear. Neither response serves you well. What follows is the rationale for what the team does, not a self-assessment tool. Understanding why your rheumatologist orders certain tests reduces the anxiety of the unknown and replaces it with something more useful: context.

THE ASSOCIATION

Adult-onset IIM is associated with an elevated risk of cancer compared to the general population. This association is well established in epidemiological studies and has shaped international screening guidelines.^[22] The risk is highest in the three years surrounding IIM diagnosis, particularly the first year, and declines thereafter.^[23]

Dermatomyositis carries the highest malignancy risk among IIM subtypes, with a pooled standardized incidence ratio of approximately 4.79 to 5.50 versus the

general population.^[23] Antisynthetase syndrome and overlap myositis are associated with lower cancer risk than dermatomyositis.^[22] The most common associated cancer types in Western populations include ovarian, lung, breast, colorectal, and pancreatic cancers, and non-Hodgkin lymphoma.^[23]

The clinical logic behind screening is specific and worth understanding. In some cases, the inflammatory myopathy is paraneoplastic: the cancer drives the immune response that produces the muscle disease. When a cancer is found and treated, the myositis may improve or resolve. Screening is not routine cancer screening in the general-population sense. It is an investigation into whether the myositis itself has a treatable underlying cause.^[22]

RISK STRATIFICATION: THE TEAM'S TOOL

The 2023 IMACS (International Myositis Assessment and Clinical Studies Group) guideline introduced a risk stratification framework classifying each IIM patient's cancer risk as standard, moderate, or high, based on IIM subtype, autoantibody profile, and clinical features.^[22] This is the current international reference guideline for cancer screening in IIM.

High-risk features in the IMACS framework include anti-TIF1-gamma positivity (approximately 4.68-fold increased cancer risk in meta-analysis), anti-NXP2 positivity, two or more high-risk clinical features (older age, male sex, dysphagia, skin necrosis or ulceration), and the dermatomyositis subtype.^[22]

Anti-TIF1-gamma antibodies are the strongest autoantibody-level predictor. Reported cancer incidence in anti-TIF1-gamma-positive DM ranges from 19 to 100 percent in published series, depending on how the patients were selected.^[23] Anti-Jo-1 and other antisynthetase antibodies are associated with reduced malignancy risk relative to dermatomyositis.^[22]

One nuance that matters: even patients at “standard risk” within the IMACS framework have cancer rates likely exceeding the general population. The IMACS categories compare within IIM, not against people without myositis.^[22]

WHAT SCREENING LOOKS LIKE

The IMACS guideline recommends a basic screening panel (chest radiography, full blood count, liver function tests, inflammatory markers, protein electrophoresis) for

all IIM patients at standard or moderate risk at diagnosis.^[22] An enhanced panel (CT of neck, chest, abdomen, and pelvis; tumor markers including CA-125; ovarian ultrasound; mammography; PSA) is recommended for moderate and high-risk patients.^[22] 18F-FDG PET/CT is conditionally recommended for high-risk patients when initial investigations are negative.^[22]

High-risk patients receive enhanced screening at diagnosis plus the basic panel annually for three years from IIM symptom onset.^[22]

If your team requests cancer screening, this follows evidence-based guidelines, not suspicion about a specific tumor. The screening intensity matches the risk window, and the risk window narrows with time.

WHAT YOU SHOULD KNOW

You do not need to self-assess your cancer risk. The IMACS framework is the team's tool, and no features are provided here for self-evaluation. The three-year post-onset window is the highest-risk period, and screening intensity is proportionate to this window. Whether you have been screened, and whether your screening was IMACS-compliant, is a question for your team. It appears in Chapter 9.

ESCALATION: CANCER SCREENING CONTEXT

Cancer screening itself is a clinical action, not a self-management one. However: unexplained rapid weight loss, new persistent pain, new unexplained masses, or IIM that is not responding to treatment should be communicated Promptly to the rheumatology team. Treatment resistance is listed in guidelines as a feature associated with elevated cancer risk.^[22]

The Heart: Cardiac Awareness

Cardiac involvement in IIM is less common than lung or swallowing involvement, but it is clinically significant when it occurs.

Clinically significant cardiac involvement occurs in approximately six percent of IIM patients.^[24] That figure is the clinical surface. Subclinical rates detected by sensitive cardiac imaging range up to 75 percent in selected cohorts, which means the heart is involved more often than it announces itself.^[24] The gap between six percent and 75 percent is similar to the gap between self-reported and instrumentally detected dysphagia: the disease can be present without producing symptoms you notice.

Manifestations include conduction abnormalities, dilated cardiomyopathy, pericarditis, and **myocarditis** (inflammation of the heart muscle itself, a direct parallel to what the disease does to skeletal muscle).^[24] Anti-SRP antibodies are the strongest predictor of cardiac involvement in published cohort data.^[24]

MONITORING

The BSR 2022 guideline recommends cardiac troponin I as a screening blood test, with an important specification: not cardiac troponin T.^[14] The reason is practical. Troponin T cross-reacts with skeletal muscle troponin T in myositis and can produce misleading results. Troponin I is specific to the heart, so an elevated troponin I in a myositis patient points to the heart, not the damaged skeletal muscle the disease already involves. ECG and echocardiography are recommended as baseline screening tools. Cardiac MRI is recommended for cases with suspected myocarditis.^[14]

Whether you have had a baseline ECG, echocardiogram, and troponin I is a question for your team. If these have not been performed, the question is whether they should be, given your antibody profile and clinical history.

WHAT TO WATCH FOR

Palpitations, chest pain, unexplained breathlessness at rest or with minimal exertion, ankle swelling, or disproportionate fatigue should be reported promptly.^[24] “Disproportionate” is the key qualifier: fatigue that exceeds what your current muscle disease and treatment burden explain, the sense that something beyond the usual is happening.

ESCALATION: HEART

New palpitations, chest pain, or unexplained breathlessness at rest warrants Prompt contact with your team. If these are severe, worsening, or accompanied by fainting or near-fainting, seek Urgent attention.

Putting It Together: The Escalation Map

This is the full escalation reference that the front matter summarized in its “When to Seek Help Now” box. Each organ domain in this chapter carries its own triage tier, and the pattern across them is consistent. Most changes are worth raising at a scheduled visit. Some warrant a call within days. A small number require same-day

attention. The summary below is a reference, not a replacement for the sections above; go back to the relevant section if you need the detail.

Routine (next scheduled visit): New swallowing symptoms since the last review. New hard subcutaneous nodules. Questions about your ILD screening status, cancer screening status, or cardiac monitoring status. Any new skin change you want the team to see.

Prompt (call within days): Frequent coughing during meals. Unintentional weight loss. A chest infection without an obvious cause. New breathlessness on exertion or a new persistent dry cough. Rash worsening despite current treatment. Skin ulceration or breakdown. Signs of infection over calcinosis. New palpitations or chest pain at rest. Unexplained rapid weight loss, new persistent pain, or new unexplained masses. Treatment that does not appear to be working.

Urgent (same-day or emergency): Rapid-onset breathlessness worsening over days to weeks. Choking or aspiration during eating or drinking. Recurrent or aspiration pneumonia. Severe or worsening chest pain, breathlessness at rest, fainting, or near-fainting.

This map is the practical output of awareness-plus-escalation. You notice. You classify. You call. The clinical decision belongs to the team.

Key Points

- Myositis can involve the lungs, throat, skin, and heart; organ-system awareness is as important as understanding the muscle disease itself.
- ILD can be present before you feel symptoms, which is why screening with PFTs and imaging is guideline-recommended for all IIM patients.
- Dysphagia affects roughly one in three IIM patients and silent aspiration can occur without coughing, so a formal swallowing assessment is worth requesting.
- UV exposure in dermatomyositis is a systemic disease trigger, not just a skin irritant; year-round broad-spectrum sun protection is the guideline recommendation.

- Cancer screening in IIM follows a structured international guideline and targets paraneoplastic causes, not routine screening; if your team orders it, the protocol is evidence-based.
- Any rapid change in breathing is Urgent; do not wait for a scheduled visit when breathlessness worsens over days rather than months.

CHAPTER 8: WHAT IS ON THE HORIZON

In this chapter, the book turns forward: what is being tested, what has shown results, and what remains too early to count on. The calibrated horizon applies the same evidence discipline to emerging therapies that the rest of the book applies to established ones, so that investigational promise and clinical reality are never confused. None of what follows changes your care today. All of it is a conversation for your team.

The landscape is moving

The principle governing this chapter is simple and worth naming first: **the calibrated horizon**. Every emerging therapy deserves to be presented at its true stage of evidence, neither inflated by hope nor dismissed by caution. A phase 3 trial with 241 patients and replicated endpoints is not the same thing as a case report of one patient at one centre. Both matter. They do not matter equally, and the difference between them is load-bearing for anyone deciding what to ask about at the next appointment. The calibrated horizon means you can follow the science without mistaking a promising signal for a proven therapy.

That principle is needed because the landscape is genuinely changing. For most of the history of idiopathic inflammatory myopathy, treatment choices could be counted on one hand: corticosteroids, a conventional immunosuppressant, intravenous immunoglobulin, and rituximab for the refractory cases. That pattern is shifting. In the past three years, a phase 3 trial delivered the first positive result for a targeted oral therapy in dermatomyositis. Cell therapies engineered for cancer have produced drug-free remissions in patients whose myositis had resisted every standard option. An antibody clearance agent already approved for myasthenia gravis is now being tested across multiple IIM subtypes. And a systematic review published in 2025 mapped the entire investigational pipeline, grading each agent by evidence maturity so that clinicians and patients can see exactly where each stands.^[1]

This chapter walks through that landscape. It means you can bring informed questions to your rheumatology team without arriving with expectations the evidence does not yet support.

Brepocitinib: a targeted oral therapy for dermatomyositis

THE MECHANISM

Brepocitinib is an oral medication that selectively inhibits two enzymes, **TYK2** and **JAK1**, which sit at critical junctions in the signalling pathways that drive dermatomyositis.^[2] Think of it as a targeted circuit breaker. The immune system's inflammatory wiring runs through several relay points; TYK2 and JAK1 are two of the most active relays in DM. By interrupting both simultaneously, brepocitinib reduces the type I and type II interferon signalling, the IL-6, the IL-12 and IL-23 cascades that sustain the disease's attack on muscle and skin.

This is not a broad immunosuppressant in the way that corticosteroids are. It targets specific pathways rather than dampening the immune system wholesale. That specificity is what makes the trial result significant.

THE VALOR TRIAL

The **VALOR trial** is the landmark.^[3] It enrolled 241 adults with active dermatomyositis across 90 sites worldwide, randomised them to brepocitinib 30 mg once daily or placebo, and followed them for 52 weeks. It is the first positive 52-week placebo-controlled randomised trial in DM. It is also the first positive registrational trial of a targeted oral therapy in this disease.

The primary result: patients receiving brepocitinib showed a 15.3-point greater improvement in **Total Improvement Score** compared with placebo at Week 52. That difference was statistically robust ($p < 0.001$). Benefits were detectable as early as Week 4.^[3]

All nine prespecified secondary endpoints were met, covering muscle strength, skin disease activity (measured by the Cutaneous Dermatomyositis Disease Area and Severity Index), functional disability, and corticosteroid reduction.^[3,4] That breadth matters. A single positive endpoint can be a statistical artefact. Nine met endpoints across independent outcome domains is a different kind of signal.

The steroid-sparing results deserve a separate sentence. More than two-thirds of patients receiving brepocitinib achieved a TIS of 40 or above, which is twice the minimum clinically important difference. Over 60 percent tapered their

corticosteroid dose to 2.5 mg per day of prednisone equivalent or less. Forty-two percent discontinued corticosteroids entirely.^[3] For anyone who has lived with the side effects described in Chapter 4, those numbers carry real weight.

Safety showed a pattern consistent with the **JAK inhibitor** class: serious infections were increased with brepocitinib compared to placebo but resolved with medical management. Most patients completed the full 52-week study. Malignancies, cardiovascular events, and thromboembolic events occurred more frequently in the placebo group, which the trial authors note may reflect the underlying DM-associated risks and the burden of conventional immunosuppression rather than drug-related protection.^[3] JAK inhibitors as a class carry established safety signals (infections, particularly herpes zoster; venous thromboembolism; cardiovascular events; and malignancy) that form part of any benefit-risk discussion with your team.^[5]

REGULATORY STATUS AND WHAT IT MEANS FOR YOU

The FDA accepted the brepocitinib New Drug Application and granted Priority Review on March 3, 2026.^[6] The Prescription Drug User Fee Act target action date falls in the third quarter of 2026, and the sponsor anticipates a potential U.S. launch by the end of September 2026. If approved, brepocitinib would be the first targeted oral therapy approved specifically for dermatomyositis.

As of this book's date, brepocitinib remains investigational. It has not yet received FDA approval.^[6]

There is an important subtype boundary here. VALOR enrolled patients with dermatomyositis. It did not study antisynthetase syndrome, immune-mediated necrotising myopathy, or overlap myositis.^[3] Whether brepocitinib is relevant to you depends entirely on whether your subtype is DM. That is a question for your treating team, not an assumption this book can make. If your IIM is antisynthetase syndrome, IMNM, or overlap myositis, brepocitinib may not apply regardless of its approval status.

If your team confirms your subtype is dermatomyositis, brepocitinib is worth asking about at your next visit: what the regulatory timeline means for your care, whether the trial results apply to your specific presentation, and what the risk profile looks like in the context of your current treatment. That is a Routine conversation, not an

urgent one. The therapy is not yet available, and your current treatment is not waiting for it.

CAR-T cell therapy: early signals, serious caveats

The register shifts here. Where brepocitinib rests on a 241-patient, 52-week randomised trial, what follows rests on case reports and small series. The results are striking. The evidence is thin. Both facts matter equally.

WHAT CAR-T THERAPY IS

Chimeric antigen receptor T-cell therapy (CAR-T) involves extracting a patient's own T cells, genetically engineering them to recognise a specific target on immune cells (most commonly CD19 on B cells), expanding them in the laboratory, and reinfusing them after a course of chemotherapy that clears space for the engineered cells to work.^[7] The goal is to deeply deplete the B-cell population that produces the autoantibodies driving the disease.

The concept is borrowed from oncology. CAR-T was developed for refractory blood cancers, where it has proven effective at targeting and eliminating specific cell populations. Its application to autoimmune disease, including myositis, is recent (the first case reports appeared between 2021 and 2023), experimental, and available only through clinical trials or compassionate-use programmes at a small number of specialised academic centres.^[7] It is not a standard treatment. It is not commercially available for myositis.

THE ERLANGEN CASE SERIES

The most cited evidence comes from a single-centre case series published in the New England Journal of Medicine in 2024.^[7] Fifteen patients with treatment-refractory autoimmune disease (eight with lupus, three with inflammatory myopathy, four with systemic sclerosis) received a single infusion of autologous CD19-directed CAR-T cells after lymphodepleting chemotherapy with fludarabine and cyclophosphamide. All three IIM patients achieved a major clinical response by ACR-EULAR criteria. All fifteen patients stopped immunosuppressive therapy entirely. Median follow-up was 15 months.

Those are notable outcomes. They are also outcomes from three myositis patients at one centre, without a comparator group, with follow-up measured in months rather than years. The evidence grade is low.

Safety: **cytokine release syndrome** (a systemic inflammatory reaction triggered when the engineered cells engage their targets) occurred in most patients, but it was characteristically mild, grade 1 in ten of fifteen, grade 2 in one.^[7] One patient experienced grade 1 immune effector cell-associated neurotoxicity syndrome. One patient was hospitalised for pneumonia. No one died. CRS in autoimmune CAR-T applications appears substantially milder than in the cancer setting, likely because the target-cell burden is lower.

The CASTLE basket trial, a phase 1/2 study evaluating CD19 CAR-T across lupus, systemic sclerosis, and IIM, has reported early results showing that treatment was safe with improved disease activity and patient-reported global health in most patients.^[8] These remain early-phase data.

CABA-201 AND THE RESET-MYOSITIS TRIAL

A separate CAR-T product, **CABA-201** (rescabtagene autoleucel), is being tested in the RESET-Myositis phase I/II trial (NCT06154252), which is enrolling patients across four IIM subtypes: IMNM, antisynthetase syndrome, dermatomyositis, and juvenile IIM.^[9] Target enrolment is 74 patients. The trial's estimated primary completion date is July 2028.

The first published case describes a 33-year-old man with anti-SRP-positive immune-mediated necrotising myopathy. At 16 weeks after receiving CABA-201, his creatine kinase had decreased substantially, his muscle strength had improved, his autoantibody titres had dropped sharply, and he was off all immunosuppressants.^[9] No cytokine release syndrome. No neurotoxicity. His B cells were undetectable by day 15 and returned at approximately eight weeks as predominantly naive cells. He was discharged four days after infusion.

One patient. One centre. Sixteen weeks of follow-up. The result is encouraging and the evidence grade is low. Both statements are true simultaneously.

WHEN CD19 CAR-T IS NOT ENOUGH

A case published in Nature Medicine in 2025 illustrates both the promise and the complexity of cell therapy.^[10] A 45-year-old woman with Jo-1-positive antisynthetase syndrome, refractory to treatment for seven years, initially achieved drug-free remission for nine months after CD19 CAR-T. Then her disease relapsed.

A second infusion of the same CD19 product failed entirely: her immune system had mounted an anti-CAR-directed T-cell response against the CAR construct itself, preventing the engineered cells from taking hold.^[10] This is a meaningful biological lesson. Repeated treatment with an identical engineered construct risks immune rejection of the therapy.

The clinical team switched targets. Instead of CD19 on B cells, they used a CAR-T product directed at **BCMA**, a protein expressed on the plasma cells that are the final stage of antibody production. The rationale was that long-lived plasma cells, which can lose CD19 expression, may sustain autoantibody production even after B-cell depletion. The BCMA-directed cells (idecabtagene vicleucel) cleared her plasma cells, normalised her creatine kinase within three months, and restored her muscle strength completely (MMT-8 score of 150). At nine months of follow-up, she remained in drug-free remission.^[10]

The safety profile included grade 1 cytokine release syndrome (managed with tocilizumab), no neurotoxicity, and a need for immunoglobulin replacement therapy because the deep B-cell and plasma-cell depletion reduced her ability to produce protective antibodies.^[10]

This is one patient. The finding that a different CAR target can rescue a relapse after CD19 failure informs the biology. It does not establish a treatment pathway.

THE RISKS, STATED PLAINLY

CAR-T therapy carries real risks that should not be softened by the word “promising.”^[7,10]

The lymphodepleting chemotherapy required before infusion creates a period of significant immunosuppression, during which infections (including pneumonia and viral reactivation) are carefully monitored. Prolonged depletion of B cells leads to reduced immunoglobulin production; immunoglobulin replacement may be

required for weeks to months. Cytokine release syndrome, while typically mild in autoimmune applications, is a systemic inflammatory reaction that occasionally requires intensive management with tocilizumab. This is resource-intensive medicine delivered at specialised centres, and it requires a level of infrastructure that limits where it can be offered.

AVAILABILITY

CAR-T therapy for autoimmune myositis is available only in research settings: clinical trials and compassionate-use programmes at a small number of academic centres worldwide.^[7] It is not a treatment you can request through standard clinical channels. Whether it is even a conversation to have with your team depends on whether your disease is refractory to current therapy, and that assessment belongs to your rheumatology team.

Research is active across IIM subtypes including dermatomyositis, IMNM, antisynthetase syndrome, and juvenile IIM.

Bispecific antibodies: a different route to the same targets

Bispecific T-cell engagers are antibody constructs that simultaneously bind a target on immune cells (such as CD19 on B cells) and a receptor on T cells, redirecting the patient's own T cells to kill the targeted cells.^[1] Unlike CAR-T, they do not require genetic engineering of the patient's cells or the multi-week manufacturing process. They are administered as infusions or injections, which could make them a more accessible alternative to CAR-T for depleting B cells or plasma cells in autoimmune disease.

The evidence in myositis is at the case-report level.

A compassionate-use series published in *Nature Medicine* in 2026 treated five patients with treatment-refractory antisynthetase syndrome using **blinatumomab**, a CD19-directed bispecific already approved for a form of B-cell leukaemia.^[11] Blinatumomab induced rapid clinical, serological, and histological improvement of myositis and stabilised interstitial lung disease. An unexpected finding: maintenance therapy with rituximab (which had previously been ineffective on its own in these patients) enabled prolonged disease control after bispecific-induced depletion. The

same publication reported that five patients with systemic sclerosis treated with a BCMA-directed bispecific (teclistamab) showed improvement, with at least one patient having active myositis as part of an overlap presentation.^[11]

Five patients per cohort. Compassionate use. No comparator group. The evidence grade is low. The concept that a more accessible agent could achieve meaningful immune cell depletion without the manufacturing complexity of CAR-T is being explored. It is not established.

Efgartigimod: antibody clearance across subtypes

Efgartigimod works by a different mechanism entirely. It blocks the **neonatal Fc receptor** (FcRn), which normally recycles IgG antibodies and extends their lifespan in the bloodstream.^[12] By blocking this recycling, efgartigimod accelerates the clearance of pathogenic autoantibodies. It is already FDA-approved for generalised myasthenia gravis and chronic inflammatory demyelinating polyneuropathy.

The **ALKIVIA trial** is testing subcutaneous efgartigimod in idiopathic inflammatory myopathies.^[12] Unlike brepocitinib (which studied only DM), ALKIVIA enrolls patients across dermatomyositis, IMNM, and antisynthetase syndrome. That subtype breadth is important: if the results hold, efgartigimod could be relevant to a wider range of IIM patients.

Phase 2 results from the first 90 of 240 enrolled patients showed a mean Total Improvement Score of 50.45 for efgartigimod compared with 35.65 for placebo at 24 weeks ($p = 0.0004$).^[12] Seventy-nine percent of patients receiving efgartigimod achieved moderate improvement (TIS 40 or above) compared to 47 percent on placebo. Thirty-four percent achieved major improvement (TIS 60 or above) versus 9.5 percent. Median time to minimal improvement was 30 days with efgartigimod compared with 72 days on placebo. Treatment-related adverse event rates were similar between groups.

These are phase 2 data from a single trial. The phase 3 portion is ongoing with 240 patients enrolled; results are not yet available. The result is encouraging and preliminary. That is a conditional statement, and it rests on phase 3 data that do not yet exist.

Abatacept: a completed trial, an incomplete answer

Abatacept blocks T-cell co-stimulation and is already approved for rheumatoid arthritis. A phase 3 trial randomised 149 patients with active IIM to subcutaneous abatacept plus standard of care or placebo plus standard of care for 24 weeks. ^[13]

The primary endpoint was not met. Improvement was 56.0 percent in the abatacept group versus 42.5 percent in the placebo group, a difference that did not reach statistical significance ($p = 0.083$). ^[13]

A prespecified subtype analysis showed benefit in non-DM patients: 57.1 percent versus 32.3 percent ($p = 0.040$). ^[13] Safety was comparable between groups with no new signals.

The picture for abatacept is suggestive but inconclusive: a reasonable safety profile, a trend toward benefit that fell short of the threshold the trial set for itself, and a subgroup signal that needs confirmation in its own right. The BSR guideline includes a conditional recommendation for abatacept in refractory IIM, but that recommendation preceded this trial result.

The broader pipeline

The systematic review by Wang and colleagues, published in the *Journal of Cachexia, Sarcopenia and Muscle* in 2025, maps the full investigational landscape. ^[1] Beyond the agents described above, research is active across IIM subtypes on agents including obinutuzumab, belimumab, type I interferon antagonists, cytokine inhibitors, proteasome inhibitors, and low-dose IL-2 therapy. Evidence for most of these ranges from case series to preclinical work. Research is also active in inclusion body myositis, though IBM falls outside this book's scope.

The point is not to catalogue every agent. It is to recognise that the pipeline for inflammatory myopathy is broader and more active than at any previous point in the disease's history. That is a factual observation, not a promise.

Clinical trials and how to think about them

IIM is rare. The incidence is roughly one case per 100,000 person-years. That rarity means few large trials exist, and many treatment decisions still rest on small

studies, case series, or expert consensus.^[1] Clinical trials are the mechanism by which evidence advances from the case-report level toward the replicated, controlled data that guidelines require. Patient participation is what makes that progression possible.

Organisations like MIHRA (the Myositis International Health and Research Collaborative Alliance) and the Myositis Clinical Trials Consortium work to accelerate trial readiness across international sites, mapping centre capabilities to trial designs and training investigators in a disease where patient numbers are inherently limited.^[14,15] The 6th Global Conference on Myositis, held in Lisbon in March 2026, featured expert-led presentations on the latest advances in IIM diagnosis and treatment, reflecting the current level of research activity in the field.^[16]

Whether a trial is relevant to you depends on your subtype, your autoantibody profile, your disease activity, your prior treatment history, your organ involvement, and where you live. Your rheumatology team is the appropriate source for assessing eligibility and suitability.

ClinicalTrials.gov is the primary U.S. registry for finding active trials. Searching by condition (“myositis,” “dermatomyositis,” “inflammatory myopathy”) and filtering by location and recruitment status is the standard approach. The Myositis Association also provides patient-facing trial information.

Participating in a trial does not guarantee receiving the investigational therapy; many trials include a placebo arm. The informed consent process explains the design, risks, potential benefits, and the possibility of receiving placebo. Your team can help you weigh whether trial participation makes sense given your current disease control and available options.

What none of this changes today

Nothing in this chapter alters your treatment plan. Brepocitinib is under review, not approved. CAR-T is available only through research protocols at specialised centres. Efgartigimod’s phase 3 data have not yet been reported. Abatacept’s phase 3 trial did not meet its primary endpoint. The bispecific antibody data come from five patients.

The calibrated horizon exists so that you can follow these developments with clear eyes. When your team mentions a new therapy, or when you encounter one online, you now have a framework for the question that matters: what is the evidence, and at what stage?

That question goes to your team. It always does.

Key Points

- The calibrated horizon means presenting each emerging therapy at its true evidence stage, so that promise and proof are never confused.
- Brepocitinib is the first targeted oral therapy to show a positive phase 3 result in dermatomyositis, but it is DM-specific and not yet approved.
- CAR-T cell therapy has produced drug-free remissions in refractory myositis, but the evidence rests on case reports and small series.
- Efgartigimod showed encouraging phase 2 results across multiple IIM subtypes; phase 3 data are pending.
- None of these investigational therapies changes your current treatment; questions about them belong with your rheumatology team.

CHAPTER 9: QUESTIONS FOR YOUR RHEUMATOLOGY TEAM

The most useful thing you can bring to a rheumatology appointment is not a symptom diary, though that helps. It is a question that pins down a value only your team can supply. Throughout this book, certain facts have surfaced that anchor your care to your specific situation: your antibody result, your lung function baseline, your bone density score, your resting creatine kinase on stable therapy. These are not details you can look up in a textbook. They live in your medical record and in the clinical judgment of the people treating you. This chapter gathers them into one place, organized by what each answer unlocks, so you can walk into your next visit with what we have been calling **appointment-ready questions**.

The questions below are phrased for a routine visit unless the underlying topic carries a higher escalation tier. Where a question touches something that warrants prompt or urgent attention, the tier is noted. None of these questions ask your team to do something unusual. They ask your team to share information that already exists in your record or to confirm that a guideline-recommended step has been completed.

Your Diagnosis and Antibody Status

Your myositis-specific antibody result shapes nearly everything else: which organs need closer monitoring, how your team thinks about cancer screening, and what your expected treatment response looks like.^[1] If you do not know your result, or if you were told your panel was negative, this is worth clarifying.

Questions to bring:

- What is my myositis-specific antibody result? If the panel was negative, does that change how you monitor me?
- Has my subtype been formally classified (dermatomyositis, antisynthetase syndrome, immune-mediated necrotizing myopathy, or another category)?
- Based on my antibody and subtype, which organs need the closest surveillance?

Your Monitoring Baseline

Creatine kinase measured on a day when you have not exercised recently, while your disease is stable on therapy, becomes your personal reference point.^[2] Without it, a future CK result is just a number.

Questions to bring:

- What is my resting CK baseline on stable therapy, and when was it last measured?
- Has my muscle strength been formally assessed (for example, by MMT8 or another standardized test)? What score did I receive?
- Would a Myositis Activities Profile help us track how my daily function is changing over time?^[3]

Your Lungs

Interstitial lung disease can be present before you notice any breathing difficulty, which is why guidelines recommend screening even in the absence of symptoms.^[4,4320] Some antibody profiles carry higher ILD risk than others, and the answer to that question depends on the antibody conversation above.

Questions to bring:

- Has a baseline high-resolution CT of my chest been performed?
- What are my most recent pulmonary function test values, and how do they compare to my baseline?
- If I have not had these tests, should I?

If you develop new breathlessness on exertion, a persistent dry cough, or any sudden drop in exercise tolerance, do not wait for the next appointment. These warrant prompt contact with your team.

Cancer Screening

The rheumatology team may have already completed cancer screening based on your antibody result and the timing of your diagnosis.^[5] If they have, confirming

what was done and when helps you understand the timeline. If they have not, asking about it is reasonable.

Questions to bring:

- Has an age-appropriate and antibody-appropriate cancer screening panel been completed since my diagnosis?
- Am I still within the higher-risk window that guidelines describe, and does that change my screening schedule?

Your Treatment Regimen

Understanding what you are receiving, why, and what to ask about it is different from trying to manage it yourself. These questions help you hold the full picture.^[6]

Questions to bring:

- What is my current IVIG dose (in grams per kilogram), how often do I receive it, and which product am I on?
- Am I on any additional immunosuppressant alongside IVIG (such as methotrexate, azathioprine, or mycophenolate)?
- Is my current corticosteroid dose (if I am still on one) at a level where we are actively trying to reduce it further?

Exercise and Infusion Timing

Exercise is safe and beneficial in autoimmune inflammatory myopathy, but the practical question of when to schedule sessions around monthly infusions does not have a textbook answer.^[7] Your team and your physiotherapist are the right people to help you work this out.

Questions to bring:

- Is there a window around my infusion days when I should avoid intense exercise, or can I train on any day I feel well?
- Do you have a physiotherapist with experience in inflammatory myopathy you can refer me to?

- Given my current functional level, what type of exercise (aerobic, resistance, or both) would you recommend I prioritize?

Infection Prevention

Immunosuppressive therapy raises infection risk, and certain preventive steps are well supported by guidelines.^[8] Vaccination and, in some cases, prophylactic medication reduce that risk. These are not optional extras; they are part of the treatment plan.

Questions to bring:

- Is my Shingrix (recombinant zoster) vaccination up to date?
- Have I received the recommended pneumococcal vaccine series?
- Is my influenza and COVID-19 vaccination current, and do I need a different formulation than the standard one?
- Based on my antibody profile and immunosuppressant regimen, should I be on prophylaxis against *Pneumocystis pneumonia*?

The last question is particularly relevant if your antibody profile places you in a higher-risk category for opportunistic infection. If you develop fever, a new cough, or any sign of serious infection while on immunosuppression, this is urgent. Do not wait for your next appointment.

Bone and Metabolic Health

Corticosteroids, even at low doses, affect bone density and glucose metabolism.^[9] If you have taken them at any point during your treatment, these questions apply.

Questions to bring:

- Has a DXA scan been performed to assess my bone density? What was my FRAX fracture risk score?
- Am I on bone-protective medication, and if not, should I be?
- Are my calcium and vitamin D supplement doses adequate for my current situation?

- Has my HbA1c or fasting glucose been checked recently? Is there any sign of steroid-related glucose elevation?

Swallowing

Difficulty swallowing can develop gradually and may not be obvious until it becomes significant.^[10] Formal assessment catches problems that you might not notice on your own, including silent aspiration.

Questions to bring:

- Has a formal swallowing assessment been performed?
- Should I see a speech and language therapist for an evaluation?
- If I am on a texture-modified diet, is there a dietitian involved to make sure I am meeting my nutritional needs?

If you are coughing or choking during meals, if food feels like it is sticking in your throat, or if you have had a chest infection without an obvious cause, report this promptly. Recurrent aspiration is an urgent concern.

Heart

Cardiac involvement in inflammatory myopathy is uncommon but worth confirming has been assessed, particularly if your antibody profile suggests elevated risk.^[11]

Questions to bring:

- Has a baseline ECG and echocardiogram been performed?
- Is cardiac troponin I (not troponin T) being monitored as part of my routine blood work?

If you experience palpitations, chest pain, unexplained breathlessness at rest, or ankle swelling, report these promptly.

Further Resources

The Myositis Association (myositis.org) maintains patient education materials, webinar archives, and a directory of support groups. Their webinars on topics including IVIG therapy, new treatments, and living with myositis are useful listening,

though they are informational rather than a substitute for clinical guidance. Your national myositis patient organization (if different from TMA) may offer region-specific resources and peer support networks.

Key Points

- Your antibody result and subtype classification shape your monitoring plan; confirming both with your team is the single most clarifying step you can take.
- A resting CK baseline on stable therapy turns future blood results from abstract numbers into meaningful comparisons.
- Lung screening, cancer screening, bone density assessment, and vaccination status are guideline-recommended steps worth confirming have been completed.
- Exercise timing around infusions and the right type of training for your current function are questions your physiotherapist and rheumatologist can answer together.
- Swallowing, cardiac monitoring, and infection prophylaxis are areas where a brief conversation can confirm whether additional steps are needed.
- New breathlessness, choking during meals, fever on immunosuppression, or chest pain warrants Prompt or Urgent contact with your team.

APPENDIX G: ENDNOTES

Superscript numbers in square brackets in the body text refer to the per-chapter endnotes below.

How to Use This Book

1. Wang Y et al. Prognostic analysis of MDA5-associated clinically amyopathic dermatomyositis with interstitial lung disease. *Immunity, Inflammation and Disease* <https://onlinelibrary.wiley.com/doi/full/10.1002/iid3.1332>
2. Speyer R et al. The Impact of Dysphagia in Myositis: A Systematic Review and Meta-Analysis. *Journal of Clinical Medicine*, 2020; 9(7): <https://pmc.ncbi.nlm.nih.gov/articles/PMC7408750/>
3. Tsou H-K et al. High incidence and mortality of Pneumocystis jirovecii infection in anti-MDA5-antibody-positive dermatomyositis. *PMC* <https://pmc.ncbi.nlm.nih.gov/articles/PMC8417987/>
4. Kang E-H et al. Increased Risk of Herpes Zoster Following Dermatomyositis and Polymyositis: A Nationwide Population-Based Cohort Study. *PMC* <https://pmc.ncbi.nlm.nih.gov/articles/PMC4617095/>

Chapter 1: Understanding Myositis and Your Diagnosis

1. Oldroyd AGS, Lilleker JB, Amin T, et al. British Society for Rheumatology guideline on management of paediatric, adolescent and adult patients with idiopathic inflammatory myopathy. *Rheumatology (Oxford)*. 2022;61(5):1760-1768. <https://doi.org/10.1093/rheumatology/keac115>
2. Trallero-Araguás E, Grau-Junyent JM, Selva-O'Callaghan A, et al. Epidemiology of dermatomyositis and other idiopathic inflammatory myopathies in Northern Spain, 2000-2022. <https://pmc.ncbi.nlm.nih.gov/articles/PMC12562039/>
3. Lundberg IE, Tjärnlöf A, Bottai M, et al. 2017 European League Against Rheumatism/American College of Rheumatology classification criteria for adult and juvenile idiopathic inflammatory myopathies and their major

subgroups. *Arthritis Rheumatol.* 2017;69(12):2271-2282.

<https://doi.org/10.1002/art.40320>

4. Korsten P, Feist E. Myositis [Hot Topics]. *Zeitschrift für Rheumatologie.*
<https://doi.org/10.1007/s00393-022-01278-2>
5. Oldroyd AGS et al. 2022 (BSR Guideline), as endnote 1; Lundberg IE et al. 2017, as endnote 3.
6. Dalakas MC. Dermatomyositis: practical guidance and unmet needs.
<https://pmc.ncbi.nlm.nih.gov/articles/PMC10924937/>
7. Aggarwal R, Charles-Schoeman C, Schessl J, et al. Trial of intravenous immune globulin in dermatomyositis. *N Engl J Med.* 2022;387:1264-1278.
<https://doi.org/10.1056/NEJMoa2117912>
8. Allenbach Y, Mammen AL, Benveniste O, et al. Immune-mediated necrotizing myopathy. <https://pmc.ncbi.nlm.nih.gov/articles/PMC10885118/>
9. Palterer B, Vitiello G, Carraresi A, et al. Myositis-specific antibodies: overview and clinical utilization. <https://pmc.ncbi.nlm.nih.gov/articles/PMC9524809/>
10. Solomon et al. Classification criteria for antisynthetase syndrome, cited in PMC10075366. <https://pmc.ncbi.nlm.nih.gov/articles/PMC10075366/>
11. Aggarwal R, et al. Advances in idiopathic inflammatory myopathies classification. <https://pmc.ncbi.nlm.nih.gov/articles/PMC10782090/>
12. Aggarwal R, et al. Adult inflammatory myopathies: updates on classification and management. *Practical Neurology.*
<https://practicalneurology.com/diseases-diagnoses/neuromuscular/adult-inflammatory-myopathies-updates-on-classification-and-management/32123/>
13. Ghirardello A, Doria A. Idiopathic inflammatory myopathies overlapping with systemic diseases. <https://pmc.ncbi.nlm.nih.gov/articles/PMC5738776/>
14. Lundberg IE et al. 2017, full text. PMC5736307.
<https://pmc.ncbi.nlm.nih.gov/articles/PMC5736307/>

15. Crum-Cianflone NF. Bacterial, fungal, parasitic, and viral myositis. *Clin Microbiol Rev.* <https://pmc.ncbi.nlm.nih.gov/articles/PMC2493084/>
16. Nair TK, et al. A masquerade of infectious myositis as polymyositis. <https://pmc.ncbi.nlm.nih.gov/articles/PMC7646808/>
17. StatPearls: Drug-induced myopathy. NBK562290. <https://www.ncbi.nlm.nih.gov/books/NBK562290/>
18. Dalakas MC. Diagnosis and classification of idiopathic inflammatory myopathies. <https://pmc.ncbi.nlm.nih.gov/articles/PMC5021058/>
19. ; StatPearls NBK562290. <https://pmc.ncbi.nlm.nih.gov/articles/PMC3383519/>
20. Frontiers in Neurology: Inclusion body myositis: update on the diagnostic and therapeutic landscape <https://doi.org/10.3389/fneur.2022.1020113>
21. ENMC 272nd International Workshop: revision of ENMC 2013 diagnostic criteria for inclusion body myositis. *Neuromuscular Disorders.* <https://www.sciencedirect.com/science/article/pii/S0960896624000543>
22. Exploring challenges in the management and treatment of inclusion body myositis. <https://pmc.ncbi.nlm.nih.gov/articles/PMC10552844/>

Chapter 2: Diagnosis and Monitoring: What the Tests and Numbers Mean

1. Lundberg IE, Tjarnlund A, Bottai M, et al. 2017 European League Against Rheumatism/American College of Rheumatology Classification Criteria for Adult and Juvenile Idiopathic Inflammatory Myopathies and Their Major Subgroups. *Arthritis Rheumatol.* 2017;69(12):2271-2282. <https://doi.org/10.1002/art.40320>
2. Oldroyd AGS, Lilleker JB, Amin T, et al. British Society for Rheumatology guideline on management of paediatric, adolescent and adult patients with idiopathic inflammatory myopathy. *Rheumatology (Oxford).* 2022;61(5):1760-1768. <https://doi.org/10.1093/rheumatology/keac115>

3. Palterer B, Vitiello G, Carraresi A, et al. Myositis-Specific Antibodies: Overview and Clinical Utilization. *Diagnostics*.
<https://pmc.ncbi.nlm.nih.gov/articles/PMC9524809/>
4. Korsten P, Feist E. Myositis [Hot Topics]. *Zeitschrift fur Rheumatologie*.
<https://doi.org/10.1007/s00393-022-01278-2>
5. Dalakas MC. Inflammatory Muscle Diseases. *N Engl J Med*.
<https://pmc.ncbi.nlm.nih.gov/articles/PMC5021058/>
6. Myositis and Mimics on MRI. *AJR Am J Roentgenol*.
<https://ajronline.org/doi/10.2214/AJR.24.31359>
7. Electrodiagnostic evaluation of myopathies. *Phys Med Rehabil Clin N Am*.
<https://pmc.ncbi.nlm.nih.gov/articles/PMC3383519/>

Chapter 3: Treatment, with IVIG at the Center

1. Barsotti S, Lundberg IE. Current Treatment for Myositis. *Curr Treat Options Rheumatol*. 2018;4:299-315. <https://doi.org/10.1007/s40674-018-0106-2>
2. Aggarwal R, Charles-Schoeman C, Schessl J, et al. Trial of Intravenous Immune Globulin in Dermatomyositis. *New England Journal of Medicine*. 2022;387:1264-78. <https://www.nejm.org/doi/full/10.1056/NEJMoa2117912>
3. Chinoy H, Adler M, Bhansing KV, et al. British Society for Rheumatology guideline on management of paediatric, adolescent and adult patients with idiopathic inflammatory myopathy. *Rheumatology (Oxford)*. 2022;61(5):1760-1768. <https://doi.org/10.1093/rheumatology/keac115>
4. Barsotti S, Lundberg IE. Current Treatment for Myositis. *Curr Treat Options Rheumatol*. 2018;4:299-315. <https://doi.org/10.1007/s40674-018-0106-2>
(treatment hierarchy, combination therapy, IVIG positioning, cyclophosphamide, hydroxychloroquine)
5. Gomez-Puerta JA, et al. Efficacy and adverse effects of methotrexate compared with azathioprine in the antisynthetase syndrome
<https://pmc.ncbi.nlm.nih.gov/articles/PMC11811931/>

6. Dalakas MC, et al. Mechanisms of action of intravenous immunoglobulin in inflammatory muscle disease (complement and MAC mechanism).
Summarized at Rare Disease Advisor:
<https://www.rarediseaseadvisor.com/cch/intravenous-immunoglobulin-dermatomyositis-c3-c4-membrane-attack-complex/>
7. Patel D, et al. Intravenous Immunoglobulin: Mechanism of Action in Autoimmune and Inflammatory Conditions. *JACI In Practice*.
[https://www.jaci-inpractice.org/article/S2213-2198\(23\)00402-6/abstract](https://www.jaci-inpractice.org/article/S2213-2198(23)00402-6/abstract)
8. Aggarwal R, et al. ProDERM: efficacy maintained through Week 40 (secondary endpoints and open-label extension). *New England Journal of Medicine*.
<https://www.nejm.org/doi/full/10.1056/NEJMoa2117912>
9. ProDERM post-hoc analysis: skin improvement regardless of baseline severity. *eClinicalMedicine / The Lancet*.
[https://www.thelancet.com/journals/eclinm/article/PIIS2589-5370\(23\)00411-X/fulltext](https://www.thelancet.com/journals/eclinm/article/PIIS2589-5370(23)00411-X/fulltext)
10. Octapharma. FDA approval of Octagam 10% for adult dermatomyositis, July 15, 2021; seven years marketing exclusivity granted August 30
<https://www.octapharma.com/news/press-release/2021/octagam-10-receives-7-years-of-market-exclusivity-for-adult-dermatomyositis>
11. IVIG-Induced Aseptic Meningitis in a Dermatomyositis Patient (case report). PMC11093182. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC11093182/>
12. FDA class labelling for all IVIG products: boxed warnings for thrombosis and renal dysfunction. FDA.gov package inserts (PANZYGA, ASCENIV, others).
<https://www.fda.gov/files/vaccines,%20blood%20&%20biologics/published/Packaging-Insert-PANZYGA.pdf>
13. StatPearls. Transfusion Selective IgA Deficiency.
<https://www.ncbi.nlm.nih.gov/books/NBK564340/> also JACI In Practice:
[https://www.jaci-inpractice.org/article/S2213-2198\(13\)00236-5/fulltext](https://www.jaci-inpractice.org/article/S2213-2198(13)00236-5/fulltext)
14. Oddis CV, Reed AM, Aggarwal R, et al. Rituximab in the Treatment of Refractory Adult and Juvenile Dermatomyositis and Adult Polymyositis: A

Randomized, Placebo-phase Trial (RIM trial). *Arthritis and Rheumatism*.

<https://pmc.ncbi.nlm.nih.gov/articles/PMC3558563/>

15. Allenbach Y, et al. Efficacy and safety of rituximab treatment in patients with idiopathic inflammatory myopathies: A systematic review and meta-analysis. *Frontiers in Immunology*.
<https://pmc.ncbi.nlm.nih.gov/articles/PMC9791086/>
16. Efficacy and Safety of Subcutaneous Abatacept Plus Standard Treatment for Active Idiopathic Inflammatory Myopathy: Phase 3 Randomized Controlled Trial <https://pmc.ncbi.nlm.nih.gov/articles/PMC12123255/>
17. Brepocitinib rationale: Johns Hopkins. PubMed
<https://pubmed.ncbi.nlm.nih.gov/39008325/>
18. Frontiers in Immunology. Therapeutic efficacy and safety of JAK inhibitors in treating polymyositis/dermatomyositis: a single-arm systemic meta-analysis
<https://pmc.ncbi.nlm.nih.gov/articles/PMC10991784/>
19. Engel F, et al. A Phase 3 Trial of Brepocitinib in Dermatomyositis (VALOR). *New England Journal of Medicine*.
<https://www.nejm.org/doi/full/10.1056/NEJMoa2503531>
20. FDA accepts NDA and grants Priority Review for brepocitinib in dermatomyositis; PDUFA target action date Q3
<https://www.dermatologytimes.com/view/fda-accepts-and-grants-priority-review-of-nda-for-brepocitinib-in-dermatomyositis>

Chapter 4: Living with Treatment: Managing Side Effects

1. Seror R, "Corticosteroid Myopathies," MedLink Neurology
<https://www.medlink.com/articles/corticosteroid-myopathies>
2. Oddis CV, Aggarwal R, "Corticosteroid Induced Myopathy," StatPearls, NCBI Bookshelf NBK557731. <https://www.ncbi.nlm.nih.gov/books/NBK557731/>
3. "Steroid Myopathy: Understanding the Pathogenesis," Biomedical Research
<https://biomedres.us/fulltexts/BJSTR.MS.ID.001889.php>

4. "Corticosteroid-Induced Myopathy: Overview," Medscape
<https://emedicine.medscape.com/article/313842-overview>
5. Compston J et al., "The 2023 Guidelines for the Management and Treatment of Glucocorticoid-Induced Osteoporosis,"
<https://pmc.ncbi.nlm.nih.gov/articles/PMC10982086/>
6. Humphrey MB et al., "2022 American College of Rheumatology Guideline for the Prevention and Treatment of Glucocorticoid-Induced Osteoporosis," Arthritis and Rheumatology, PMID 37845798.
<https://pubmed.ncbi.nlm.nih.gov/37845798/>
7. "Management of Glucocorticoid-Induced Hyperglycemia," PMC9142341.
<https://pmc.ncbi.nlm.nih.gov/articles/PMC9142341/>
8. "Long-Term Usage of Oral Glucocorticoids Leading to Adrenal Insufficiency: A Comprehensive Review," PMC10257969.
<https://pmc.ncbi.nlm.nih.gov/articles/PMC10257969/>
9. "Infections in Idiopathic Inflammatory Myopathies," International Journal of Rheumatic Diseases <https://onlinelibrary.wiley.com/doi/10.1111/1756-185X.70084>
10. Tsou H-K et al., "High incidence and mortality of Pneumocystis jirovecii infection in anti-MDA5-antibody-positive dermatomyositis,"
<https://pmc.ncbi.nlm.nih.gov/articles/PMC8417987/>
11. "PCP Prophylaxis in Rheumatic Disease," Cleveland Clinic Journal of Medicine, 86(7):449. <https://www.ccmj.org/content/86/7/449>
12. Kang E-H et al., "Increased Risk of Herpes Zoster Following Dermatomyositis and Polymyositis: A Nationwide Population-Based Cohort Study,"
<https://pmc.ncbi.nlm.nih.gov/articles/PMC4617095/>
13. Bass AR et al., "2022 American College of Rheumatology Guideline for Vaccinations in Patients with Rheumatic and Musculoskeletal Diseases," Arthritis Care and Research, , doi:10.1002/acr.25045.
<https://pmc.ncbi.nlm.nih.gov/articles/PMC10291822/>

14. EULAR 2026 Updated Recommendations for Vaccination in Patients with Autoimmune Inflammatory Rheumatic Diseases, via RheumNow.
<https://rheumnow.com/news/eular-2026-updated-recommendations-vaccination-patients-autoimmune-inflammatory-rheumatic>
15. “Shingles vaccination in immune-mediated inflammatory diseases: a narrative review,” Rheumatology (Oxford)
<https://academic.oup.com/rheumatology/article/65/2/keag043/8440154>
16. ACIP 2023 Pneumococcal Vaccine for Adults Aged 19 Years and Older, PMC10495181. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC10495181/>
17. ACIP 2024 Additional COVID-19 Vaccine Doses for Immunocompromised Adults, PMC11637417.
<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC11637417/>
18. CDC Timing and Spacing of Immunobiologics.
<https://www.cdc.gov/vaccines/hcp/imz-best-practices/timing-spacing-immunobiologics.html>
19. FDA IVIG Class Boxed Warning, representative package inserts including Bivigam, Gammalex, GAMUNEX-C, PRIVIGEN.
https://www.bivigam.com/hubfs/Bivigam_April2023/pdf/BIVIGAM_20221217_Ambient.pdf
20. Aggarwal R et al., “Safety and tolerability of intravenous immunoglobulin in patients with active dermatomyositis: results from the randomised, placebo-controlled ProDERM study,”.
<https://pmc.ncbi.nlm.nih.gov/articles/PMC10792872/>
21. Zhang X et al., “Evaluating the association between intravenous immunoglobulin and thromboembolic events in patients with autoimmune diseases: a retrospective real-world study,” Frontiers in Pharmacology, doi:10.3389/fphar.2026.1762942.
<https://doi.org/10.3389/fphar.2026.1762942>

22. US Pharmacist, "IVIG and Kidney Disease: What the Pharmacist Needs to Know." <https://www.uspharmacist.com/article/ivig-and-kidney-disease-what-the-pharmacist-needs-to-know>

Chapter 5: Movement and Daily Function

1. Alemo Munters L et al. "Exercise in myositis." Curr Treatm Opt Rheumatol <https://pmc.ncbi.nlm.nih.gov/articles/PMC4133148/>
2. Lodin K et al. "The Role of Exercise to Improve Physiological, Physical and Psychological Health Outcome in Idiopathic Inflammatory Myopathies." J Inflamm Res <https://pmc.ncbi.nlm.nih.gov/articles/PMC11162627/>
3. Oldroyd AGS et al. "British Society for Rheumatology guideline on management of paediatric, adolescent and adult patients with idiopathic inflammatory myopathy." Rheumatology. 2022;61(5):1760-1768. <https://pmc.ncbi.nlm.nih.gov/articles/PMC9398208/>
4. Corrado B, Ciardi G, Lucignano L. "Supervised Physical Therapy and Polymyositis/Dermatomyositis: A Systematic Review of the Literature." Open Access Rheumatol <https://pmc.ncbi.nlm.nih.gov/articles/PMC7768488/>
5. Wiesinger GF et al. "Improvement of physical fitness and muscle strength in polymyositis/dermatomyositis patients by a training programme." Rheumatology. 1998;37(2):196-200. <https://pubmed.ncbi.nlm.nih.gov/9569076/>
6. Alemo Munters L et al. "Improved exercise performance and increased aerobic capacity after endurance training of patients with stable polymyositis and dermatomyositis." Arthritis Res Ther. 2013;15(4):R83. DOI:10.1186/ar4263. <https://pmc.ncbi.nlm.nih.gov/articles/PMC3978470/>
7. Alemo Munters L et al. "Improvement in health and possible reduction in disease activity using endurance exercise in patients with established polymyositis and dermatomyositis: a multicenter randomized controlled trial with a 1-year open extension followup." Arthritis Care Res. 2013;65(12):1959-1968. <https://pubmed.ncbi.nlm.nih.gov/23861241/>

8. Andreasson K et al. "High-intensity interval training outperforms moderate exercise to improve aerobic capacity in patients with recent-onset idiopathic inflammatory myopathies: a multicentre randomised controlled trial." *eBioMedicine*. 2025;122:106051.
<https://pmc.ncbi.nlm.nih.gov/articles/PMC12703967/>
9. Jensen KY et al. "High-intensity resistance training improves quality of life, muscle endurance and strength in patients with myositis: a randomised controlled trial." *Rheumatol Int*. 2024;44(10):1909-1921.
DOI:10.1007/s00296-024-05698-y.
<https://pmc.ncbi.nlm.nih.gov/articles/PMC11392978/>
10. Tomcik M et al. "The effect of a 24-week training focused on activities of daily living, muscle strengthening, and stability in idiopathic inflammatory myopathies." *Arthritis Res Ther*. 2021;23:173. DOI:10.1186/s13075-021-02544-5. <https://pmc.ncbi.nlm.nih.gov/articles/PMC8218432/>
11. Nader GA et al. "A longitudinal, integrated, clinical, histological and mRNA profiling study of resistance exercise in myositis." *Mol Med*. 2010;16(11-12):455-464. DOI:10.2119/molmed.2010.00016.
<https://pmc.ncbi.nlm.nih.gov/articles/PMC2972393>
12. Benatti FB, Pedersen BK. "Exercise as an anti-inflammatory therapy for rheumatic diseases." 2015; Munters LA et al. "Endurance exercise improves molecular pathways of aerobic metabolism in patients with myositis."
13. Binder EF et al. "Blood-flow restricted resistance training in patients with sporadic inclusion body myositis: a randomized controlled trial." *Rheumatology* <https://pubmed.ncbi.nlm.nih.gov/29775118/>
14. "Immune marker assessment after blood-flow-restriction exercise in IBM." <https://doi.org/10.1186/s13075-019-2036-2>." *Arthritis Res Ther*.
15. Lodin K et al. "The Role of Exercise to Improve Physiological, Physical and Psychological Health Outcome in Idiopathic Inflammatory Myopathies." *J Inflamm Res*. <https://pmc.ncbi.nlm.nih.gov/articles/PMC11162627/>

16. Alexanderson H, Lundberg IE, Stenstrom CH. "Development of the myositis activities profile: validity and reliability of a self-administered questionnaire to assess activity limitations in patients with polymyositis/dermatomyositis." J Rheumatol. 2002;29(11):2386-2392.
<https://www.jrheum.org/content/29/11/2386>
17. Baschung Pfister P et al. "Reliability and validity of the German version of the Myositis Activities Profile (MAP) in patients with inflammatory myopathy." PLoS One. 2019;14(6):e0217173. DOI:10.1371/journal.pone.0217173.
<https://pmc.ncbi.nlm.nih.gov/articles/PMC6546213/>
18. Alexanderson H, Reed AM, Ytterberg SR. "The Myositis Activities Profile: initial validation for assessment of polymyositis/dermatomyositis in the USA." J Rheumatol. 2012;39(11):2134-2141.
<https://pubmed.ncbi.nlm.nih.gov/22942260/>
19. Rider LG et al. "Update on outcome assessment in myositis." Nat Rev Rheumatol <https://pmc.ncbi.nlm.nih.gov/articles/PMC6702032/>
20. Britson KA et al. "Exploring challenges in the management and treatment of inclusion body myositis." Front Neurol
<https://pmc.ncbi.nlm.nih.gov/articles/PMC10552844/>

Chapter 6: Nutrition

1. Sanchez-Rosales A et al. Nutritional interventions as modulators of the disease activity for idiopathic inflammatory myopathies: a scoping review. J Hum Nutr Diet. 2024;37(2):370-380. PMID
<https://pubmed.ncbi.nlm.nih.gov/38324396/>
2. Corticosteroid Induced Myopathy. StatPearls. NBK557731.
<https://www.ncbi.nlm.nih.gov/books/NBK557731/> also Glucocorticoid-Mediated Skeletal Muscle Atrophy: Molecular Mechanisms and Potential Therapeutic Targets. Int J Mol Sci. 2025;26(15):7616.
<https://www.mdpi.com/1422-0067/26/15/7616>

3. Corradini C et al. Nutritional recommendations for patients undergoing prolonged glucocorticoid therapy. *Rheumatol Adv Pract.* 2022;6(2):rkac029. PMC9080102. <https://pmc.ncbi.nlm.nih.gov/articles/PMC9080102/>
4. Bauer J et al. Evidence-based recommendations for optimal dietary protein intake in older people: A position paper from the PROT-AGE Study Group. *J Am Med Dir Assoc.* 2013;14(8):542-559. PMC4208946. <https://pmc.ncbi.nlm.nih.gov/articles/PMC4208946/>
5. Gorissen SHM et al. Dietary protein requirements of older adults with sarcopenia determined by the indicator amino acid oxidation technology. *Front Nutr* <https://pmc.ncbi.nlm.nih.gov/articles/PMC11906324/>
6. Protein intake and sarcopenia systematic review <https://pmc.ncbi.nlm.nih.gov/articles/PMC11894306/>
7. Leucine and whey protein in sarcopenia. PMC12426612. <https://pmc.ncbi.nlm.nih.gov/articles/PMC12426612/>
8. Protein supplementation with resistance exercise for sarcopenia: systematic review and meta-analysis. *Aging Clin Exp Res* <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC11369567/>
9. Humphrey MB et al. 2022 American College of Rheumatology Guideline for the Prevention and Treatment of Glucocorticoid-Induced Osteoporosis. *Arthritis Rheumatol.* 2023;75(12):2084-2098. DOI 10.1002/art.42646. <https://acrjournals.onlinelibrary.wiley.com/doi/full/10.1002/art.42646>
10. Tamaki J et al. The 2023 Guidelines for the management and treatment of glucocorticoid-induced osteoporosis. *J Bone Miner Metab* <https://pmc.ncbi.nlm.nih.gov/articles/PMC10982086/>
11. Vitamin D and muscle status in inflammatory and autoimmune rheumatic diseases. *Nutrients.* 2024;16(14):2329. DOI 10.3390/nu16142329. <https://www.mdpi.com/2072-6643/16/14/2329>
12. Severe vitamin D deficiency in a patient with inflammatory myopathy. *Front Pediatr* <https://www.frontiersin.org/articles/10.3389/fped.2024.1339875/full>

13. Golubic R et al. Glucocorticoid-induced hyperglycaemia in hospitalised adults: A matched cohort study (2013-2023). *Diabetes Obes Metab*
<https://pmc.ncbi.nlm.nih.gov/articles/PMC12146452/>
14. Low glycaemic index and low glycaemic load eating patterns in type 2 diabetes management. *Nutrients*
<https://pmc.ncbi.nlm.nih.gov/articles/PMC11519289/>
15. Miner SJ et al. Changes in total body fat and body mass index among children with juvenile dermatomyositis treated with high-dose glucocorticoids. *Arthritis Care Res* <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC8353815/>
16. Basnayake O et al. The Impact of Dysphagia in Myositis: A Systematic Review and Meta-Analysis. *Semin Arthritis Rheum*. 2020;50(5):1017-1025. PMC7408750. <https://pmc.ncbi.nlm.nih.gov/articles/PMC7408750/>
17. IDDSI Framework. iddsi.org; Su IM et al. Clinical applications of IDDSI framework for texture recommendation for dysphagia patients. *J Texture Stud*. 2018;49(1):2-10. PMID. <https://pubmed.ncbi.nlm.nih.gov/29052849/>
18. Okada J et al. Texture-Modified Diets, Nutritional Status and Mealtime Satisfaction: A Systematic Review. *Healthcare*. 2021;9(6):624. PMC8225071. <https://pmc.ncbi.nlm.nih.gov/articles/PMC8225071/> also Nutritional Intake and Meal Composition of Patients Consuming Texture Modified Diets and Thickened Fluids: A Systematic Review and Meta-Analysis. *Healthcare*. 2020;8(4):579. PMC7767351. <https://pmc.ncbi.nlm.nih.gov/articles/PMC7767351/>
19. Effectiveness of diet modification on dietary nutrient intake, aspiration, and fluid intake for adults with dysphagia: a meta-analysis of RCTs. *Clin Nutr* <https://pmc.ncbi.nlm.nih.gov/articles/PMC12179983/>
20. Mediterranean diet and inflammatory biomarkers, meta-analysis of RCTs. *Nutrition Reviews* <https://pubmed.ncbi.nlm.nih.gov/41211687/>
21. Omega-3 fatty acids and autoimmune disease: umbrella review and Mendelian randomisation. *Autoimmun Rev*
<https://pubmed.ncbi.nlm.nih.gov/39357585/>

22. Hahn J et al. Vitamin D and marine omega-3 fatty acid supplementation and incident autoimmune disease: VITAL randomized controlled trial. BMJ. 2022;376:e066452. DOI 10.1136/bmj-2021-066452.
<https://acrabstracts.org/abstract/vitamin-d-and-marine-n-3-fatty-acid-supplementation-and-prevention-of-autoimmune-disease-in-the-vital-randomized-controlled-trial/>
23. Kley RA et al. Creatine for treating muscle disorders. Cochrane Database Syst Rev. 2013;6:CD004760. PMID <https://pubmed.ncbi.nlm.nih.gov/23740606/>
24. Intravenous Immunoglobulin (IVIG). StatPearls. NBK554446.
<https://www.ncbi.nlm.nih.gov/books/NBK554446/>

Chapter 7: Organ-System Awareness and When to Escalate

1. Mammen AL. Idiopathic inflammatory myopathies related lung disease in adults. Lancet Respir Med
[https://www.thelancet.com/journals/lanres/article/PIIS2213-2600\(24\)00267-4/abstract](https://www.thelancet.com/journals/lanres/article/PIIS2213-2600(24)00267-4/abstract)
2. Johnson SR et al. 2023 ACR/CHEST Guideline for the Screening and Monitoring of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Diseases. Arthritis Care Res
<https://acrjournals.onlinelibrary.wiley.com/doi/10.1002/acr.25347>
3. Myositis-Associated Interstitial Lung Disease: The Experience of a Tertiary Center. J Clin Med 2024; 13(20):
<https://pmc.ncbi.nlm.nih.gov/articles/PMC11508529/>
4. Sambataro G et al. (ERS/EULAR Task Force). ERS/EULAR clinical practice guidelines for connective tissue disease-associated interstitial lung disease. Ann Rheum Dis [https://ard.eular.org/article/s0003-4967\(25\)04320-1/fulltext](https://ard.eular.org/article/s0003-4967(25)04320-1/fulltext)
5. Johnson SR et al. 2023 ACR/CHEST Guideline. Arthritis Care Res
<https://acrjournals.onlinelibrary.wiley.com/doi/10.1002/acr.25347>

6. A Review of Myositis-Associated Interstitial Lung Disease. PMC <https://pmc.ncbi.nlm.nih.gov/articles/PMC11278012/>
7. van Manen MJG et al. The 1-minute sit-to-stand test to detect desaturation during 6-minute walk test in interstitial lung disease. npj Prim Care Respir Med <https://pmc.ncbi.nlm.nih.gov/articles/PMC8795411/>
8. Gupta N et al. A Review of Antisynthetase Syndrome-Associated Interstitial Lung Disease. PMC <https://pmc.ncbi.nlm.nih.gov/articles/PMC11050089/>
9. Wang Y et al. Prognostic analysis of MDA5-associated clinically amyopathic dermatomyositis with interstitial lung disease. Immunity Inflamm Dis <https://onlinelibrary.wiley.com/doi/full/10.1002/iid3.1332>
10. Moreira A et al. Myositis interstitial lung disease and autoantibodies. Front Med 2023; 10: <https://www.frontiersin.org/journals/medicine/articles/10.3389/fmed.2023.1117071/full>
11. Speyer R et al. The Impact of Dysphagia in Myositis: A Systematic Review and Meta-Analysis. J Clin Med 2020; 9(7): <https://pmc.ncbi.nlm.nih.gov/articles/PMC7408750/>
12. Upper Gastrointestinal Manifestations of Inflammatory Myositis. PMC <https://pmc.ncbi.nlm.nih.gov/articles/PMC11238612/>
13. Impact and Management of Dysphagia in IIM. Clin Gastroenterol Hepatol <https://pubmed.ncbi.nlm.nih.gov/32860119/>
14. Gordon PA et al. British Society for Rheumatology guideline on management of paediatric, adolescent and adult patients with idiopathic inflammatory myopathy. Rheumatology 2022; 61(5): 1760-1768. <https://pmc.ncbi.nlm.nih.gov/articles/PMC9398208/>
15. Cobos G et al. The Spectrum of Cutaneous Manifestations in Dermatomyositis: A Comprehensive Review. J Clin Med 2026; 15(8): <https://www.mdpi.com/2077-0383/15/8/2874>

16. Levin M et al. Beyond the Rash: Exploring the Symptoms and Diagnosis of Dermatomyositis. PMC <https://pmc.ncbi.nlm.nih.gov/articles/PMC10769122/>
17. Fotiadis AI et al. Early detection and intervention in dermatomyositis: current evidence and clinical implications. PMC <https://pmc.ncbi.nlm.nih.gov/articles/PMC12979258/>
18. Mechanisms of photosensitivity in autoimmunity. PMC <https://pmc.ncbi.nlm.nih.gov/articles/PMC8688579/>
19. Myositis Support and Understanding. Sun protection guidance. <https://understandingmyositis.org/sun-protection/>
20. Aggarwal R et al. Clinical features, treatments and outcomes of calcinosis in adult patients with dermatomyositis: a single cohort study. Rheumatology (Oxford) 2021; 60(6): 2958-2968. <https://academic.oup.com/rheumatology/article/60/6/2958/6010469>
21. Calcinosis in dermatomyositis. Curr Opin Rheumatol 2024; 36(6). https://journals.lww.com/co-rheumatology/fulltext/2024/11000/calcinosis_in_dermatomyositis.14.aspx
22. Oldroyd AGS et al. International Guideline for Idiopathic Inflammatory Myopathy-Associated Cancer Screening: an IMACS Initiative. Nat Rev Rheumatol 2024; 20: 105-119. <https://pmc.ncbi.nlm.nih.gov/articles/PMC10834225/>
23. Mugii N et al. Risk, risk factors, and screening of malignancies in dermatomyositis: current status and future perspectives. Rheumatol Int <https://pmc.ncbi.nlm.nih.gov/articles/PMC12173892/>
24. Lopes JP et al. Predictors of cardiac involvement in idiopathic inflammatory myopathies. Front Immunol <https://pmc.ncbi.nlm.nih.gov/articles/PMC10030705/>

Chapter 8: What Is on the Horizon

1. Wang R, Lin W, Xie Q, Yin G. Targeted Therapy for Idiopathic Inflammatory Myopathy. *Journal of Cachexia, Sarcopenia and Muscle*. 2025;16(6):e70143. <https://pmc.ncbi.nlm.nih.gov/articles/PMC12695148/>
2. Paik JJ, et al. Brepocitinib, a potent and selective TYK2/JAK1 inhibitor: scientific and clinical rationale for dermatomyositis <https://pubmed.ncbi.nlm.nih.gov/39008325/>
3. Engel F, Paik JJ, Engel A, et al. A Phase 3 Trial of Brepocitinib in Dermatomyositis (VALOR). *New England Journal of Medicine*. <https://doi.org/10.1056/NEJMoa2503531>
4. Roivant. NEJM Publishes Positive Phase 3 VALOR Trial Results. Press release. <https://investor.roivant.com/news-releases/news-release-details/new-england-journal-medicine-publishes-positive-phase-3-valor>
5. FDA class labelling for JAK inhibitors: infections, herpes zoster, venous thromboembolism, cardiovascular events, and malignancy.
6. Priovant/Roivant. FDA Acceptance and Priority Review of NDA for Brepocitinib in Dermatomyositis. Press release, March 3 <https://investor.roivant.com/news-releases/news-release-details/priovant-announces-fda-acceptance-and-priority-review-new-drug>
7. Muller F, Boeltz S, Knitza J, et al. CD19 CAR T-Cell Therapy in Autoimmune Disease: A Case Series with Follow-up. *New England Journal of Medicine*. 2024;390:687-700. <https://doi.org/10.1056/NEJMoa2308917>
8. Mackensen A, et al. CD19 CAR-T cells for treatment-refractory autoimmune diseases: the phase 1/2 CASTLE basket trial. *Nature Medicine*. <https://doi.org/10.1038/s41591-025-04185-6>
9. Volkov A, Nunez M, Mozaffar T, et al. Case study of CD19 CAR T therapy in a subject with immune-mediated necrotizing myopathy treated in the RESET-Myositis phase I/II trial. *Molecular Therapy*. 2024;32(11):3821-3828. <https://doi.org/10.1016/j.ymthe.2024.09.009>

10. Muller F, Wirsching A, Hagen M, et al. BCMA CAR T cells in a patient with relapsing idiopathic inflammatory myositis after initial and repeat therapy with CD19 CAR T cells. *Nature Medicine*. 2025;31(6):1793-1797.
<https://doi.org/10.1038/s41591-025-03718-3>
11. Taubmann J, et al. Bispecific T cell engagers for treatment-refractory autoimmune connective tissue diseases. *Nature Medicine*.
<https://doi.org/10.1038/s41591-026-04238-4>
12. argenx. Efgartigimod data at EULAR 2025: positive Phase 2 ALKIVIA results in IIM. Press release / presentation. <https://argenx.com/news/2024/argenx-presents-new-efgartigimod-data-at-eular-2025-highlighting>
13. Aggarwal R, Lundberg IE, Song YW, et al. Efficacy and Safety of Subcutaneous Abatacept Plus Standard Treatment for Active Idiopathic Inflammatory Myopathy: Phase 3 Randomized Controlled Trial. *Arthritis & Rheumatology*.
<https://pmc.ncbi.nlm.nih.gov/articles/PMC12123255/>
14. Aggarwal R, et al. Collaborative research in myositis-related disorders: MIHRA, a global shared community model
<https://pubmed.ncbi.nlm.nih.gov/38436382/>
15. MIHRA Clinical Trial Sites Network. <https://mihrafoundation.org/mihra-programs/mihra-clinical-trial-sites-network/>
16. 6th Global Conference on Myositis (GCOM 2026), Lisbon, March 23-26
<https://imynos.org/event/6th-global-conference-on-myositis-gcom/>

Chapter 9: Questions for Your Rheumatology Team

1. Palterer B, Vitiello G, Carraresi A, et al. Myositis-Specific Antibodies: Overview and Clinical Utilization. <https://pmc.ncbi.nlm.nih.gov/articles/PMC9524809/>
2. Lodin K, et al. The Role of Exercise to Improve Physiological, Physical and Psychological Health Outcome in Idiopathic Inflammatory Myopathies. *J Inflamm Res*. <https://pmc.ncbi.nlm.nih.gov/articles/PMC11162627/>

3. Alexanderson H, Lundberg IE, Stenstrom CH. Development of the myositis activities profile: validity and reliability of a self-administered questionnaire to assess activity limitations in patients with polymyositis/dermatomyositis. *J Rheumatol*. 2002;29(11):2386-2392. PMID <https://www.jrheum.org/content/29/11/2386>
4. Sambataro G, et al. ERS/EULAR clinical practice guidelines for connective tissue disease-associated interstitial lung disease. *Ann Rheum Dis* [https://ard.eular.org/article/s0003-4967\(25\)04320-1/fulltext](https://ard.eular.org/article/s0003-4967(25)04320-1/fulltext)
5. Oldroyd AGS, et al. International Guideline for Idiopathic Inflammatory Myopathy-Associated Cancer Screening: an IMACS Initiative. *Nat Rev Rheumatol*. 2024;20:105-119. <https://doi.org/10.1038/s41584-023-01045-w>
6. Chinoy H, et al. British Society for Rheumatology guideline on management of paediatric, adolescent and adult patients with idiopathic inflammatory myopathy. *Rheumatology*. 2022;61(5):1760-1768. <https://doi.org/10.1093/rheumatology/keac115>
7. Oldroyd AGS, et al. BSR guideline: exercise programme recommendation (Grade 1B, 100% agreement). *Rheumatology*. 2022;61(5):1760-1768. <https://doi.org/10.1093/rheumatology/keac115>
8. Bass AR, et al. 2022 American College of Rheumatology Guideline for Vaccinations in Patients with Rheumatic and Musculoskeletal Diseases. *Arthritis Care Res* <https://pmc.ncbi.nlm.nih.gov/articles/PMC10291822/>
9. Humphrey MB, et al. 2022 American College of Rheumatology Guideline for the Prevention and Treatment of Glucocorticoid-Induced Osteoporosis. *Arthritis Rheumatol*. 2023;75(12):2084-2098. <https://doi.org/10.1002/art.42646>
10. Speyer R, et al. The Impact of Dysphagia in Myositis: A Systematic Review and Meta-Analysis. *J Clin Med*. 2020;9(7):2150. <https://pmc.ncbi.nlm.nih.gov/articles/PMC7408750/>

11. Lopes JP, et al. Predictors of cardiac involvement in idiopathic inflammatory myopathies. Front Immunol
<https://pmc.ncbi.nlm.nih.gov/articles/PMC10030705/>