Intravenous Immunoglobulin for Dermatomyositis: What the Approval Evidence Shows

Written by The Life Science Feed Editorial Team · Edited by Mara Voss

Dermatomyositis (DM) is a rare, chronic inflammatory myopathy affecting muscle, skin, and other organs, often leading to significant morbidity. Clinicians have long sought effective, evidence-based treatments beyond corticosteroids and immunosuppressants, which carry their own burden of side effects and often fail to control disease activity adequately. The recent approval of intravenous immunoglobulin (IVIg) offers a new therapeutic option for these challenging patients, but understanding the underlying evidence is important for its judicious application.

KEY TAKEAWAYS

- The Pivot: IVIg is now an approved treatment for dermatomyositis, providing a new option for a rare and challenging condition.
- The Data: A Brazilian registry of idiopathic inflammatory myopathies (REMAS) found 18.3% of patients received IVIg, highlighting its existing use in clinical practice.
- The Action: Clinicians should consider IVIg as a treatment option for dermatomyositis, particularly in patients with refractory disease or contraindications to other therapies, while acknowledging the need for more robust trial data.

Idiopathic inflammatory myopathies (IIMs) represent a complex group of rare autoimmune diseases, with dermatomyositis being the most prevalent subtype. These conditions are characterized by muscle weakness, skin rashes, and systemic involvement, posing diagnostic and therapeutic challenges for clinicians. The rarity of IIMs has historically hampered large-scale clinical trials, leading to a severe lack of high-quality evidence for many existing treatment protocols. This scarcity of data often forces clinicians to rely on observational studies, expert consensus, or off-label use of immunosuppressive agents.1

To address this evidence gap, a Brazilian network of nine research centers established the Registry of Idiopathic Inflammatory Myopathies (REMAS). This multicenter collaboration aims to improve knowledge, facilitate research, and ultimately enhance outcomes for patients with IIMs. Since December 2022, REMAS has prospectively recruited consecutive patients diagnosed with various IIMs, including dermatomyositis (DM), clinically amyopathic DM (CADM), inclusion body myositis (IBM), polymyositis (PM), immune-mediated necrotizing myopathy (IMNM), and anti-synthetase syndrome (ASyS).1

Understanding the Patient Population in REMAS

The REMAS registry currently includes 284 patients, providing a valuable snapshot of the IIM population in Brazil. The majority of these patients, 53.1%, received a diagnosis of dermatomyositis. This predominance of DM aligns with global epidemiological trends, where DM often represents the most common form of IIM in many regions. The patient cohort was predominantly female, comprising 77.1% of the total, and Caucasian, accounting for 44%. These demographic characteristics are consistent with the known epidemiology of autoimmune diseases, which frequently show a female predilection.1

The median age of patients at inclusion in the registry was 51 years (interquartile range, IQR: 41, 62 years), with a median age at disease onset of 43 years (IQR: 33, 55 years). The median duration of disease was 6 years (IQR: 2, 11 years), indicating that the registry captures patients across various stages of their disease journey, from relatively recent diagnoses to those with long-standing conditions. This broad representation is important for understanding the natural history and treatment patterns of IIMs.1

Clinical and Laboratory Manifestations

Clinical assessments within the REMAS registry provided insights into disease severity and impact. The median Manual Muscle Test 8 (MMT-8) score was 77 (IQR: 74, 80), reflecting a moderate degree of muscle weakness across the cohort. Patient-reported outcomes included a median visual analogue scale (VAS) score of 3 (IQR: 0, 5) for overall well-being, while physician VAS was 2 (IQR: 0, 5). The Health Assessment Questionnaire (HAQ) yielded a median score of 0.33 (IQR: 0.00, 1.00), indicating varying levels of functional impairment. These scores collectively paint a picture of patients experiencing significant, but often manageable, disease burden.1

Laboratory markers further characterized the patient population. Median creatine kinase (CK) levels at inclusion were 145 U/L (IQR: 74, 327 U/L). While elevated, these levels suggest that many patients were not in an acute flare with extremely high CK, possibly reflecting ongoing treatment or chronic disease activity. Myositis-specific autoantibodies (MSAs) were also assessed. Among those positive for MSAs, anti-Jo-1 was the most frequent, detected in 10.5% of patients, followed closely by anti-Ro-52, present in 10.1%. These autoantibodies are essential for subclassifying IIMs and can predict specific clinical phenotypes and prognoses, guiding treatment decisions.1

Treatment Patterns and IVIg Use

The REMAS registry documented the treatment market for IIMs in Brazil, highlighting the common use of corticosteroids. A substantial 87% of patients received prednisone, with a median daily dose of 5 mg (IQR: 0, 20 mg). This indicates that prednisone remains a cornerstone of therapy, often used at maintenance doses. Pulse therapy with methylprednisolone was administered to 32.7% of patients, typically reserved for acute exacerbations or severe disease presentations.1

Intravenous immunoglobulin (IVIg) was part of the treatment regimen for 18.3% of patients in the registry. This figure is particularly relevant for understanding the real-world application of IVIg in dermatomyositis and other IIMs. The use of IVIg in nearly one-fifth of the cohort suggests that it is already considered a viable option by clinicians, likely for patients with refractory disease, those intolerant to other immunosuppressants, or individuals with severe extramuscular manifestations. This aligns with the increasing recognition of IVIg's immunomodulatory properties in various autoimmune conditions.1

The mechanism of action for IVIg in autoimmune diseases like dermatomyositis is complex. IVIg contains pooled

IgG antibodies from thousands of healthy donors, which can modulate the immune system through several pathways. These include blocking Fc receptors, neutralizing pathogenic autoantibodies, suppressing B and T cell activation, inhibiting complement activation, and modulating cytokine production. For a deeper dive into the broader mechanisms of immune modulation, clinicians might find our coverage on peptide therapies relevant for understanding similar complex biological interactions.

Limitations and Future Directions

While the REMAS registry provides valuable real-world data on IIMs and the use of IVIg, it is important to acknowledge its limitations. As an observational registry, it cannot establish causality or definitive efficacy for any specific treatment. The data reflect current clinical practice rather than controlled trial outcomes. The registry also noted limited representation from some regions of Brazil, which could introduce selection bias and limit the generalizability of certain findings to the broader Brazilian population or other global cohorts.1

The registry's primary objective was to establish a multicenter collaboration and advance knowledge on IIMs, which it successfully achieved. The data on IVIg use, while not from a randomized controlled trial, highlights its integration into clinical practice. This observational evidence supports the need for more rigorous, prospective studies, including randomized controlled trials, to precisely define the efficacy, optimal dosing, and long-term safety profile of IVIg in dermatomyositis. Such trials would provide the definitive evidence required for guideline development and broader adoption. Clinicians looking for comprehensive guidance on managing complex autoimmune conditions might consult the Oxford Handbook of Rheumatology (5th ed) for practical, evidence-based recommendations.

The absence of detailed outcome data specifically linked to IVIg treatment within this abstract means clinicians must interpret its reported use with caution. The registry did not provide information on the duration of IVIg therapy, the specific indications for its use in individual patients, or the response rates observed. These granular details are essential for a complete understanding of IVIg's role and effectiveness in this patient population. Future analyses from the REMAS registry, or dedicated clinical trials, will need to address these gaps to provide a clearer picture of IVIg's clinical utility.

The establishment of the REMAS registry itself is a significant step forward for the IIM community. By facilitating multicenter collaboration, it creates a platform for future research, including the potential for interventional studies. This collaborative infrastructure is vital for studying rare diseases, where individual centers often lack sufficient patient numbers to conduct adequately powered trials. The registry's ongoing recruitment and data collection will continue to enrich our understanding of these complex conditions and their management.

The approval of IVIg for dermatomyositis marks a significant development for patients and clinicians alike. But the data from registries like REMAS show the ongoing need for robust clinical trial evidence to optimize its use. The current evidence base, while growing, still leaves many questions unanswered regarding patient selection, treatment duration, and comparative effectiveness against other therapies. The field awaits further insights from dedicated studies to fully integrate IVIg into the therapeutic algorithm for dermatomyositis.

> **CLINICAL IMPLICATIONS**
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> The approval of intravenous immunoglobulin for dermatomyositis provides a much-needed therapeutic option in a disease area historically plagued by limited evidence and challenging management. Clinicians now have a

formally recognized treatment, which should streamline access and potentially reduce the reliance on off-label prescribing. This is particularly relevant for patients who are refractory to conventional immunosuppressants or who experience significant side effects from corticosteroids.

But the evidence, as highlighted by the REMAS registry, largely reflects real-world use rather than definitive trial outcomes. The 18.3% of patients receiving IVIg in the Brazilian cohort suggests it is already part of the therapeutic armamentarium, likely for more severe or complex cases. This observational data, while valuable for understanding practice patterns, does not replace the need for rigorous, placebo-controlled trials to establish precise efficacy and optimal patient selection criteria.

For patients, this approval offers hope for improved disease control and quality of life, especially for those who have exhausted other options. However, the high cost and logistical challenges associated with IVIg administration mean that its use will likely be reserved for specific patient subsets. Payers and healthcare systems will need to balance the clinical benefit with the economic burden, emphasizing the importance of clear guidelines for appropriate use.

The industry, having secured this approval, now faces the imperative to support further research. This includes head-to-head trials against other immunosuppressants and studies evaluating long-term outcomes and cost-effectiveness. The current approval is a starting point, not an endpoint, for optimizing care in dermatomyositis.

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REFERENCES

1. 1. de Souza FHC, Miossi R, de Araujo DB. Registry of idiopathic inflammatory myopathies (REMAS) from nine Brazilian research centers linked to tertiary care and teaching hospitals. Adv Rheumatol. 2026.

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