# LISRAYA® (brepocitinib) Authorization Checklist for Dermatomyositis

When submitting an authorization request for LISRAYA, it is important to document:
- The basis for the patient's DM diagnosis
- The patient's disease manifestations
- Other prior and current therapies tried for DM, and any reasons why alternative DM therapies would be inappropriate for this patient

The tables below provide potential information to include, if applicable, in each of these areas. Most patients will not have all of the diagnostic features, disease manifestations, and prior/current therapies listed, but it is valuable to document any that are present.

| Patient Name: | Patient Date of Birth: / / |
|---|---|

Diagnosis ☐ ICD-10 M33.1: Other dermatomyositis ☐ ICD-10 M33.9: Dermatopolymyositis

## DOCUMENTING DIAGNOSIS

| Diagnostic Features | Example Criteria |
|---|---|
| Characteristic Skin Findings | Heliotrope rash, Gottron's papules, Gottron's sign, shawl sign, V sign, holster sign, inverse palmar papules, calcinosis, Mechanic's hands, flagellate erythema, poikiloderma, scalp erythema. |
| Proximal Muscle Weakness | Symmetric proximal weakness. |
| Elevated Muscle Enzymes | CK, Aldolase, AST/ALT, or LDH. |
| Electromyography (EMG) | Myopathic pattern present. |
| MRI Evidence | Symmetric muscle edema on T2/STIR imaging with or without fascial/subcutaneous edema or fatty atrophy. |
| Myositis-Specific Autoantibody (MSA) | Positive (e.g., Mi-2, TIF1-γ, MDA5, NXP2, SAE, Jo-1, PL-7, PL-12, EJ, OJ). |
| Muscle Biopsy | Perifascicular myofiber atrophy, perimysial inflammatory infiltrate, perivascular inflammation and vasculopathy. |
| Skin Biopsy | Interface dermatitis, cutaneous vasculopathy. |
| Nailfold Capillaroscopy | Periungual erythema, cuticular hypertrophy, capillary loop dilation/dropout, microhemorrhages, tufting. |
| Other Associated Systemic Manifestations | Raynaud's phenomenon, interstitial lung disease, polyarthritis, myocarditis, esophageal dysfunction/dysphagia, vasculopathy. |
| Exclude Other Causes | Thyroid-induced, statin-induced, and/or genetic muscular dystrophies ruled out. |

**Reference tool: UpToDate®: Diagnostic approach to dermatomyositis**
https://uptodate.uphs.upenn.edu/contents/image/print?imageKey=RHEU...771&search=dermatomyositis&source=outline_link&selectedTitle=1~150

**Reach out to mcs@priovant.com or 1-888-736-9788 if you have trouble accessing the above link.**

## EXAMPLES OF DM-SPECIFIC DISEASE MANIFESTATIONS

| Disease Manifestation | Examples of Patient-Specific Evidence |
|---|---|
| Cutaneous disease | Characteristic DM lesions (Gottron's papules/sign, heliotrope rash, shawl/V-sign, periungual changes); persistent erythema, ulceration, or other inflammatory lesions; pruritus, burning, pain, photosensitivity, sleep disturbance, functional limitations, or psychosocial burden. |
| Inflammatory muscle disease | Proximal muscle weakness; impaired mobility or activities of daily living; reduced muscle endurance; elevated CK, aldolase, or other muscle enzymes; muscle MRI abnormalities (e.g., edema); EMG findings consistent with inflammatory myopathy; muscle biopsy findings; dysphagia attributable to pharyngeal muscle involvement. |
| Interstitial lung disease | HRCT abnormalities (e.g., ground-glass opacities, organizing pneumonia, reticulation) or radiographic progression; reduced or declining FVC/DLCO; dyspnea, cough, exercise intolerance, oxygen requirements, or other pulmonary functional impairment. |
| Cardiac involvement | Myocarditis, myocardial inflammation, or cardiomyopathy; cardiac MRI abnormalities; elevated cardiac troponin I; echocardiographic abnormalities; arrhythmias or conduction disturbances; reduced exercise tolerance or heart failure symptoms attributable to DM. |
| Inflammatory arthritis / arthralgia | Documented synovitis; joint swelling, tenderness, or morning stiffness; ultrasound or MRI evidence of inflammatory arthritis; pain or functional limitations attributable to joint inflammation. |
| Gastrointestinal vasculopathy | Persistent or severe abdominal pain; gastrointestinal bleeding; imaging or endoscopic findings consistent with vasculopathy or ischemia; bowel ulceration, perforation, or other gastrointestinal complications attributable to DM. |
| Esophageal / oropharyngeal involvement | Dysphagia, odynophagia, choking, aspiration, or impaired swallowing; abnormal swallow study, esophageal manometry, or other diagnostic testing; nutritional compromise or aspiration-related complication. |
| Vascular manifestations / vasculopathy | Digital ulceration, ischemic lesions, nailfold capillary abnormalities, Raynaud's phenomenon, livedoid changes, or other evidence of vascular inflammation or injury; pain, tissue loss, or functional impairment associated with vascular involvement. |

## EXAMPLES OF DM-SPECIFIC DISEASE MANIFESTATIONS *(CONTINUED)*

| Disease Manifestation | Examples of Patient-Specific Evidence |
|---|---|
| Calcinosis cutis | Clinically or radiographically documented calcinosis; painful or inflamed deposits; ulceration, recurrent infection, restricted joint mobility, or functional impairment; progression or new lesion formation. |
| Subcutaneous involvement / panniculitis | Tender subcutaneous nodules or plaques; biopsy findings consistent with panniculitis; associated pain, inflammation, or functional limitations. |
| Fascial involvement | Fascial pain, tenderness, stiffness, or restricted range of motion; MRI evidence of fascial inflammation; functional impairment attributable to inflammatory fascial disease. |
| Constitutional manifestations | Persistent fatigue, fever, malaise, unintended weight loss, or reduced exercise tolerance attributable to active DM; associated impairment in daily activities, work, or quality of life. |

## PRIOR AND ONGOING TREATMENT (Common treatments for DM included below)

| Medication name | Date Prescribed | Dosage and Frequency | Duration of Use | Response to the Medication *(eg, no or incomplete response with persistent symptoms and disease activity; poor tolerability due to side effects)* |
|---|---|---|---|---|
| Systemic Corticosteroids | | | | |
| Hydroxychloroquine | | | | |
| Methotrexate | | | | |
| Mycophenolate mofetil / mycophenolic acid | | | | |
| Azathioprine | | | | |
| Cyclophosphamide | | | | |
| Cyclosporine | | | | |
| Tacrolimus | | | | |
| Leflunomide | | | | |
| IVIg | | | | |
| Rituximab / B-cell depleting agents | | | | |
| Abatacept | | | | |
| Repository corticotropin injection | | | | |

## CONTRAINDICATIONS

| Medication Name | Reason for Contraindication *(such as relevant patient-specific clinical history, including comorbidities and treatment-related risk factors)* |
|---|---|
| | |
| | |
| | |