Medical Policy:

Rituximab Injectable (Riabni®, Rituxan®, Ruxience®, Truxima®) Intravenous

POLICY NUMBER	LAST REVIEW	ORIGIN DATE
MG.MM.PH.102	October 29, 2024	

The treating physician or primary care provider must submit to EmblemHealth, or ConnectiCare, as applicable (hereinafter jointly referred to as "EmblemHealth"), the clinical evidence that the member meets the criteria for the treatment or surgical procedure. Without this documentation and information, EmblemHealth will not be able to properly review the request preauthorization or post-payment review. The clinical review criteria expressed below reflects how EmblemHealth determines whether certain services or supplies are medically necessary. This clinical policy is not intended to pre-empt the judgment of the reviewing medical director or dictate to health care providers how to practice medicine. Health care providers are expected to exercise their medical judgment in rendering appropriate care. Health care providers are expected to exercise their medical judgment in rendering appropriate care.

EmblemHealth established the clinical review criteria based upon a review of currently available clinical information (including clinical outcome studies in the peer reviewed published medical literature, regulatory status of the technology, evidence-based guidelines of public health and health research agencies, evidence-based guidelines and positions of leading national health professional organizations, views of physicians practicing in relevant clinical areas, and other relevant factors). EmblemHealth expressly reserves the right to revise these conclusions as clinical information changes and welcomes further relevant information. Each benefit program defines which services are covered. The conclusion that a particular service or supply is medically necessary does not constitute a representation or warranty that this service or supply is covered and/or paid for by EmblemHealth, as some programs exclude coverage for services or supplies that EmblemHealth considers medically necessary.

If there is a discrepancy between this guideline and a member's benefits program, the benefits program will govern. Identification of selected brand names of devices, tests and procedures in a medical coverage policy is for reference only and is not an endorsement of any one device, test or procedure over another. In addition, coverage may be mandated by applicable legal requirements of a state, the Federal Government or the Centers for Medicare & Medicaid Services (CMS) for Medicare and Medicaid members. All coding and web site links are accurate at time of publication.

EmblemHealth may also use tools developed by third parties, such as the MCG™ Care Guidelines, to assist us in administering health benefits. The MCG™ Care Guidelines are intended to be used in connection with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice. EmblemHealth Services Company, LLC, has adopted this policy in providing management, administrative and other services to EmblemHealth Plan, Inc., EmblemHealth Insurance Company, EmblemHealth Services Company, LLC, and Health Insurance Plan of Greater New York (HIP) related to health benefit plans offered by these entities. ConnectiCare, an EmblemHealth company, has also adopted this policy. All of the aforementioned entities are affiliated companies under common control of EmblemHealth Inc.

Length of Authorization

Coverage will be provided for 6 months (12 months initially for pemphigus vulgaris) and may be renewed unless otherwise specified.

- Maintenance therapy for oncology indications (excluding ALL) may be renewed for up to a maximum of 2 years.
    - Adult Acute Lymphoblastic Leukemia (ALL) may be renewed for a maximum of 18 doses.
    - Mantle Cell Lymphoma may be renewed until disease progression or intolerable toxicity.
    - Hairy Cell Leukemia may be renewed for up to a maximum of 12 doses.
    - Induction/Consolidation of Pediatric B-Cell Acute Leukemia and Aggressive Mature B-Cell Lymphomas may NOT be renewed.
    - Pediatric Hodgkin Lymphoma may NOT be renewed.
- Management of Immunotherapy-Related Toxicities:
    - Myositis/Myasthenia Gravis/Encephalitis may NOT be renewed.
    - Bullous Dermatitis may be renewed for a maximum of 18 months (4 total doses).

- Chronic Graft-Versus-Host Disease (cGVHD) may NOT be renewed.
- Hematopoietic Cell Transplantation may NOT be renewed.
- Lupus Nephritis and Pediatric Idiopathic Nephrotic Syndrome may be renewed ONLY in patients experiencing a disease relapse.
- Complications of Transplanted Solid Organ may NOT be renewed.
- Relapse therapy for pemphigus vulgaris must be at least 16 weeks past a prior infusion

Dosing Limits [Medical Benefit]

A. **Max Units (per dose and over time):**

Oncology Indications
Chronic Lymphocytic Leukemia (CLL)/Small Lymphocytic Leukemia (SLL): • Initial therapy: ○ Loading dose: 100 billable units x 1 dose ○ Subsequent doses: 130 billable units every 28 days x 5 doses per 6 months • Renewal therapy: 130 billable units every 8 weeks
ALL • 100 billable units twice weekly x 18 doses
Hairy Cell Leukemia • 100 billable units weekly x 8 doses, 100 billable units every 14 days x 8 doses, then 100 billable units every 28 days x 4 doses
Histiocytic Neoplasms – Rosai-Dorfman Disease • 130 billable units weekly x 6 doses in a 6 month period
Pediatric Hodgkin Lymphoma • 100 billable units x 3 doses
Hematopoietic Cell Transplantation • Initial dose: 100 billable units x 1 dose before transplant • Subsequent doses: 250 billable units x 3 doses after transplant
cGVHD • 100 billable units weekly x 8 doses
Immunotherapy Toxicity Treatment: • 375mg/m^2 dose weekly x 4 doses in a 6-month period
All other oncology indications: • Initial therapy: 100 billable units weekly x 8 doses per 6 months • Renewal therapy: 100 billable units x 4 doses per 6 months
Non-Oncology Indications
Rheumatoid Arthritis (RA): • 100 billable units every 14 days x 2 doses in a 18 week period
Multiple Sclerosis (MS): • 100 billable units every 14 days x 2 doses every 6 months
Pemphigus Vulgaris: • Initiation: 100 billable units weekly x 4 doses in a 12 month period • Maintenance and Relapse: 50 billable units every 16 weeks
GPA(WG)/MPA: • Induction: 100 billable units weekly x 4 doses in a 20-week period • Initial Maintenance: 50 billable units x 2 doses in a 6 month period • Subsequent Maintenance: 50 billable units every 6 months
All other non-oncology indications: • 100 billable units weekly x 4 doses in a 6 month period

Guideline

I. Initial Approval Criteria

****For Medicare members – Rituximab-please refer to our separate LCD/NCD Medicare criteria**

For Commercial and Medicaid members:

1. Non-preferred agent: Rituxan and Riabni
2. Preferred agents: Ruxience and Truxima

Coverage is provided in the following conditions *(in addition to use supported by the National Comprehensive Cancer Network [NCCN] Clinical Practice Guidelines [NCCN Guidelines®] and/or NCCN Drugs & Biologics Compendium [NCCN Compendium®] with a recommendation of category level 1 or 2A*)*:

1. Patient must be screened for HBV infection (i.e., HBsAg and anti-HBc) prior to initiating therapy; **AND**
2. For newly started Rituxan or Riabni therapy, for Commercial, Medicaid, and Medicare members:

   Coverage may be considered medically necessary when:

   A. Patient has experienced a therapeutic failure or intolerance with the plan-preferred medications (Ruxience AND Truxima); **OR**

   B. Rituxan or Riabni is requested for an indication for which the plan-preferred biosimilar agents (Ruxience or Truxima) have not been FDA-approved OR are not supported by NCCN Guidelines® or NCCN Compendium® with a recommendation of category level 1 or 2A; **AND**

   **Please note: Coverage for an appropriate biosimilar substitution will be allowed where NCCN Guidelines or Compendium state that an FDA-approved biosimilar is an appropriate substitution for rituximab.*
3. Patient is at least 18 years of age, unless otherwise specified; **AND**
4. Patient has not received a live vaccine within 28 days prior to starting treatment and live vaccines will not be administered concurrently while on treatment; **AND**

Oncology Indications:

1. Patient is CD20-positive (*excluding use for cGVHD, Hematopoietic Cell Transplantation, and Management of Immunotherapy-Related Toxicity)*; **AND**

Pediatric Mature B-Cell Acute Leukemia †

1. Patient is at least 6 months of age; **AND**
2. Used in combination with chemotherapy for previously untreated disease

Adult* Acute Lymphoblastic Leukemia (ALL) ‡

1. Patient has Philadelphia chromosome-positive (Ph+) disease; **AND**

   A. Used in combination with a tyrosine kinase inhibitor (TKI)-based regimen; **AND**

      i. Patient is <65 years of age without significant comorbidities; **OR**

   B. Used in combination with MOpAD (methotrexate, vincristine, pegaspargase, dexamethasone) for TKI-refractory disease; **OR**
2. Patient has Philadelphia chromosome-negative (Ph-) disease; **AND**

A. Used as a component of a multiagent chemotherapy

**NCCN recommendations for Adult ALL may be applicable to adolescent and young adult (AYA) patients within the age range of 15-39 years.*

CNS Cancer ‡

1. Patient has leptomeningeal metastases from lymphomas; **OR**
2. Patient has primary CNS lymphoma; **AND**
    A. Used for induction therapy; **AND**
        i. Used as a single agent OR in combination with a methotrexate-containing regimen, temozolomide, or lenalidomide¥; **OR**
        ii. Patient has CSF positive or spinal MRI positive disease§; **OR**
    B. Used for consolidation (monthly maintenance) therapy; **AND**
        i. Used as continuation of induction regimen in patients with complete response or complete response unconfirmed (CRu) to induction therapy; **AND**
            a. Used as a single agent§; **OR**
            b. Used on combination with high-dose methotrexate¥; **OR**
    C. Used for relapsed or refractory disease; **AND**
        i. Used as a single agent OR in combination with systemic therapy in patients with prior whole brain radiation therapy§; **AND**
            a. Patient has CSF positive or spinal MRI positive disease; **OR**
        ii. Used as a single agent OR in combination with temozolomide, lenalidomide, or highdose methotrexate¥

§ For intrathecal administration ONLY; ¥ For intravenous administration ONLY

Adult Hodgkin's lymphoma ‡

1. Patient has nodular lymphocyte-predominant disease

Chronic lymphocytic leukemia/Small lymphocytic lymphoma (CLL/SLL) †

1. Used in combination with fludarabine and cyclophosphamide (FC) †; **OR**
2. Patient has disease without del(17p)/TP53 mutation; **AND**
    A. Used as first-line therapy in combination with bendamustine (*excluding use in frail patients*); **OR**
    B. Used as subsequent therapy in combination with one of the following:
        i. Bendamustine (patients <65 years of age without significant comorbidities; excluding use in frail patients)
        ii. Idelalisib
        iii. Lenalidomide

iv. Venetoclax; **OR**

3. Patient has disease <u>with</u> del(17p)/TP53 mutation; **AND**

A. Used as first-line therapy in combination with high-dose methylprednisolone; **OR**

B. Used as subsequent therapy in combination with one of the following:

i. Alemtuzumab

ii. High-dose methylprednisolone

iii. Idelalisib

iv. Lenalidomide

v. Venetoclax; **OR**

4. Used as initial therapy for histologic (Richter's) transformation to diffuse large B-cell lymphoma; **AND**

A. Used in combination with cyclophosphamide, doxorubicin, and vincristine-based regimens or as a component of OFAR (oxaliplatin, fludarabine, cytarabine, and rituximab)

<u>Waldenström's macroglobulinemia/Lymphoplasmacytic Lymphoma ‡</u>

<u>Non-Hodgkin's lymphomas (NHL) †</u> **<u>Adult B-Cell Lymphomas † ‡</u>** **Φ** including, but not limited to, the following:

1. **<u>AIDS-related B-Cell Lymphoma ‡ HIV-Related B-Cell Lymphomas ‡</u>**

A. Disease is related to Burkitt lymphoma, diffuse large B-cell lymphoma (DLBCL), HHV8-positive DLBCL (not otherwise specified), or primary effusion lymphoma (PEL)

2. **<u>Burkitt Lymphoma ‡</u>**

A. Used in combination with chemotherapy

3. **<u>Diffuse Large B-Cell Lymphoma †</u>**
4. **<u>Low-grade or Follicular Lymphoma †</u>**
5. **<u>Extranodal Marginal Zone Lymphoma (EMZL) of the Stomach & Nongastric Sites (Noncutaneous) ‡</u>**
6. **<u>High-Grade B-Cell Lymphomas ‡</u>**
7. **<u>Mantle Cell Lymphoma ‡</u>**
8. **<u>Nodal & Splenic Marginal Zone Lymphoma ‡</u>**
9. **<u>Post-transplant lymphoproliferative disorder (PTLD) (B-Cell Type) ‡</u>**
10. **<u>Histologic Transformation of Indolent Lymphomas to Diffuse Large B-Cell Lymphoma ‡</u>**

<u>Castleman's Disease ‡</u>

1.. Patient has multicentric disease; **OR**

2. Patient has unicentric disease; **AND**

A. Used as second-line therapy for relapsed or refractory disease; **OR**

B. Used for unresectable disease or symptomatic disease after incomplete resection

Primary Cutaneous B-Cell Lymphomas ‡

Pediatric Aggressive Mature B-Cell Lymphomas (Primary Mediastinal Large B-Cell Lymphoma, Diffuse Large B-Cell Lymphoma, Burkitt Lymphoma, & Burkitt-like Lymphoma) † ‡ Φ

1. Patient is at least 6 months of age*; **AND**
2. Used in combination with chemotherapy

**Pediatric Aggressive Mature B-Cell Lymphoma may be applicable to adolescent and young adult (AYA) patients older than 18 years of age and less than 39 years of age, who are treated in the pediatric oncology setting.*

Hairy Cell Leukemia ‡

1. Used as a single agent; **AND**

    A. Used for less than complete response or relapsed disease in patients unable to receive purine analogs (i.e., cladribine or pentostatin); **OR**

2. Used in combination with cladribine; **OR**
3. Used in combination with pentostatin; **AND**

    A. Used for less than complete response or relapsed disease; **OR**

4. Used in combination with vemurafenib; **AND**

    A. Used as initial therapy in patients unable to tolerate purine analogs (i.e., cladribine or pentostatin) including frail patients and those with active infection; **OR**

    B. Used for less than complete response or relapse within 2 years of complete response following initial treatment with cladribine or pentostatin; **OR**

    C. Used for progression after therapy for relapsed or refractory disease (if not previously given); **OR**

5. Used in combination with venetoclax; **AND**

    A. Used for progression after therapy for relapsed or refractory disease; **AND**

        i. Patient had disease resistance to BRAF inhibitor therapy

Histiocytic Neoplasms – Rosai-Dorfman Disease ‡

1. Used as a single agent for nodal, immune-cytopenia, or immunoglobulin G4 (IgG4) related diseases; **AND**
    A. Used for symptomatic unresectable unifocal disease; **OR**
    B. Used for symptomatic multifocal disease; **OR**
    C. Used for relapsed/refractory disease

Pediatric Hodgkin Lymphoma ‡

1. Patient is ≤ 18 years of age*; **AND**
2. Patient has nodular lymphocyte-predominant disease; **AND**

3. Used in combination with CVbP (cyclophosphamide, vinblastine, prednisolone); **AND**
4. Used as primary treatment for stage IA or IIA disease (incomplete resection and non-bulky disease)

**Pediatric Hodgkin Lymphoma may be applicable to adolescent and young adult (AYA) patients up to the age of 39 years*

Hematopoietic Cell Transplantation (HCT) ‡

1**.** Used as conditioning for allogeneic transplant as part of a non-myeloablative regimen in combination with cyclophosphamide and fludarabine

Chronic graft-versus-host disease (cGVHD) ‡

1. Patient is post-allogeneic stem cell transplant (generally 3 or more months); **AND**
2. Used as additional therapy in combination with corticosteroids; **AND**
3. Patient has no response (e.g., steroid-refractory disease) to first-line therapy options; **AND**
4. Patient must try and have an inadequate response, contraindication, or intolerance to at least a three (3) month trial of ibrutinib.

Management of Immunotherapy-Related Toxicities ‡

1. Patient has been receiving therapy with an immune checkpoint inhibitor (e.g., cemiplimab, nivolumab, pembrolizumab, atezolizumab, avelumab, durvalumab, ipilimumab, dostarlimab, nivolumab/relatlimab, tremelimumab, retifanlimab, toripalimab, etc.); **AND**
    A. Patient has encephalitis related to their immunotherapy; **AND**
        i. Patient is autoimmune-encephalopathy-antibody positive; **AND**
        ii. Patient has had limited to no improvement after 7 to 14 days on high-dose corticosteroids with or without intravenous immunoglobulin (IVIG); **OR**
    B. Patient has bullous dermatitis related to immunotherapy; **AND**
        i. Used as additional therapy for moderate (G2), severe (G3) or life-threatening (G4) disease; **OR**
    C. Patient has moderate, severe, or life-threatening steroid-refractory myositis (proximal muscle weakness, neck flexor weakness, with or without myalgias) related to immunotherapy; **AND**
        i. Used for significant dysphagia, life-threatening situations, or cases refractory to corticosteroids; **OR**
    D. Patient has myasthenia gravis related to immunotherapy; **AND**
        i. Used as additional therapy for severe (G3-4) disease that is refractory to plasmapheresis or IVIG

Non-Oncology Indications:

- Patient is not on concurrent treatment with another CD20-directed therapy, TNF-inhibitor, IL-inhibitor, biologic response modifier or other non-biologic agent (e.g., apremilast, abrocitinib, tofacitinib, baricitinib, upadacitinib, deucravacitnib, etc.); **AND**

Rheumatoid arthritis (RA) †

1. Medication must be requested by a Rheumatologist; **AND**
2. Adult patient (18 years or older); **AND**
3. Documented moderate to severe disease; **AND**
4. Must be used in combination with methotrexate unless the patient has a contraindication or intolerance; **AND**
5. Patient tried and failed at least a 3-month trial with ONE oral disease modifying anti-rheumatic drug (DMARD) (e.g., methotrexate, azathioprine, auranofin, hydroxychloroquine, penicillamine, sulfasalazine, leflunomide, etc.)*; **AND**
6. Previous failure with one or more preferred TNF antagonists at least one of which should be a self-injectable; **AND**
7. Physician has assessed baseline disease severity utilizing an objective measure/tool; **AND**
8. Patient has not had treatment with Rituxan, Riabni, Ruxience, or Truxima in the previous 4 months

** Note: For patients already established on biologic therapy, trial and failure of oral DMARDs is not required*

Pemphigus vulgaris †

1. Adult patient (18 years or older); **AND**
2. Patient has a diagnosis of pemphigus vulgaris as determined by one or more of the following:
   A. Patient has one or more of the following clinical features:
      i. Appearance of lesions, erosions and/or blisters
      ii. Nikolsky sign (induction of blistering via mechanical pressure at the edge of a blister or on normal skin)
      iii. Characteristic scarring and lesion distribution; **AND**
   B. Histopathologic confirmation by skin/mucous membrane biopsy; **AND**
   C. Positive direct immunofluorescence (DIF) microscopy result **OR** Presence of autoantibodies as detected by direct immunofluorescence or enzyme-linked immunosorbent assay (ELISA); **AND**
3. Patient has moderate to severe disease as assessed utilizing an objective measure/tool (i.e. PDAI, PSS, ABSIS); **AND**
4. Patient is on combination glucocorticoid therapy; **AND**
5. Other causes of blistering or erosive skin and mucous membrane diseases have been ruled out

Granulomatosis with Polyangiitis (GPA) (Wegener's granulomatosis) and Microscopic Polyangiitis (MPA) †

1. Adult and pediatric patients (2 years or older); **AND**

2. Used in combination with glucocorticoids

Thrombocytopenic purpura ‡

1. Patient diagnosis includes one of the following:
   A. Primary thrombocytopenia
   B. Idiopathic (Immune) thrombocytopenia purpura (ITP)
   C. Evan's syndrome; **AND**

2. Patient has previously failed or has a contraindication or intolerance to therapy with corticosteroids; **AND**

3. Patient is at increased risk for bleeding as indicated by platelet count (within the previous 28 days) less than 30×10^9/L (30,000/mm³)

Thrombotic Thrombocytopenic Purpura (TTP) ‡

1. Patient has immune-mediated or acquired disease with ADAMTS13-deficiency; **AND**
   A. Used in combination with corticosteroids and therapeutic plasma exchange (TPE); **OR**
   B. Used as a single agent as prophylactic therapy for patients in remission

Multiple Sclerosis (MS) ‡

1. Patient must have a confirmed diagnosis of multiple sclerosis (MS) as documented by laboratory report (i.e., MRI); **AND**
2. Patient has a diagnosis of a relapsing form of MS [i.e., relapsing-remitting MS (RRMS), active secondary progressive disease (SPMS), or clinically isolated syndrome (CIS)]

Autoimmune Hemolytic Anemia (AIHA) ‡

1. Patient has warm-reactive disease refractory to or dependent on glucocorticoids; **OR**
2. Patient has cold agglutinin disease with symptomatic anemia, transfusion-dependence, and/or disabling circulatory symptoms

Systemic Lupus Erythematosus (SLE) ‡

1. Patient has a confirmed diagnosis of SLE as evidenced by all of the following:
   A. Confirmed SLE classification criteria score > 10* (Note: must include clinical and immunologic domains criteria)
   B. Anti-nuclear antibody (ANA) titer of ≥ 1:80 measured via indirect immunofluorescence (IIF) on human epithelial (HEp-2) cells (or an equivalent ANA positive test) at least once; **AND**

2. Patient has failed to respond adequately to at least two (2) standard therapies such as anti-malarials (i.e. hydroxychloroquine, chloroquine), corticosteroids, non-steroidal anti-inflammatory drugs (NSAIDs), aspirin,

immunosuppressives (i.e. azathioprine, methotrexate, calcineurin inhibitors [cyclosporine, tacrolimus, voclosporin], oral cyclophosphamide, or mycophenolate); **AND**

3. Patient has moderate to severe active disease defined as a Physician's Global Assessment (PGA) score of > 1 **AND** one of the following:

A. Systemic Lupus Erythematosus Disease Activity Index 2000 (SLEDAI 2K) score of ≥ 6

B. Disease activity with ≥ 2 systems with British Isles Lupus Assessment Group-2004 (BILAG) B scores

C. ≥1 system(s) with British Isles Lupus Assessment Group-2004 (BILAG) A score(s)

* *A web-based scoring calculator as well as further definitions of each criterion are available at:*

https://rheumatology.org/criteria

Lupus Nephritis ‡

1. Patient has disease that is non-responsive or refractory to standard first-line therapy (i.e.,mycophenolate mofetil, mycophenolic acid, cyclophosphamide, or calcineurin inhibitors [e.g., tacrolimus, voclosporin, cyclosporine etc.]); **AND**

2. Used as a single agent OR as add-on therapy in combination with mycophenolate mofetil, mycophenolic acid, or cyclophosphamide

Myasthenia Gravis (unrelated to immunotherapy-related toxicity) ‡

1. Patient has muscle-specific tyrosine kinase (MuSK)-antibody positive disease; **AND**

2. Patient is refractory to standard first-line therapy (e.g., glucocorticoids, azathioprine, mycophenolate mofetil, etc.)

Complications of Transplanted Solid Organ (kidney, liver, lung, heart, pancreas) in Adult and Pediatric* Patients

1. Used for suppression of panel reactive anti-human leukocyte antigen (HLA) antibodies prior to transplantation; **OR**

2. Used for treatment of antibody-mediated rejection of solid organ transplantation

**Note: There is no minimum age requirement for this indication*

Neuromyelitis Optica Spectrum Disorder (NMOSD) ‡

1. Patient has a confirmed diagnosis based on the following:

A. Patient was found to be seropositive for aquaporin-4 (AQP4) IgG antibodies; **AND**

i. Patient has at least one core clinical characteristic § *(*Note: some core clinical characteristics require both clinical and typical MRI findings*); **AND**

ii. Alternative diagnoses have been excluded [e.g., myelin oligodendrocyte glycoprotein (MOG) antibody disease (MOGAD), multiple sclerosis, sarcoidosis, cancer, chronic

infection, etc.]; **OR**

B. Patient is seronegative for AQP4-IgG antibodies OR has unknown AQP4-IgG status; **AND**

i. Patient has at least two core clinical characteristics § occurring as a result of one or more clinical attacks; **AND**

ii. Patient has experienced ALL of the following:

a. At least 1 core clinical characteristic must be acute optic neuritis, acute myelitis, or area postrema syndrome

b. Fulfillment of typical MRI findings requirements for each area affected ψ; **AND**

iii. Alternative diagnoses have been excluded [e.g., myelin oligodendrocyte glycoprotein (MOG) antibody disease (MOGAD), multiple sclerosis, sarcoidosis, cancer, chronic infection, etc.]; **AND**

2. Used as a single agent or in combination with immunosuppressive therapy (e.g., azathioprine, methotrexate, mycophenolate, etc.)

§ Core Clinical Characteristics of NMOSD
•Acute optic neuritis •Acute myelitis •Acute area postrema syndrome (APS): episode of otherwise unexplained hiccups and/or nausea and vomiting (lasting for at least 48 hours or with MRI evidence of a dorsal brainstem lesion) •Acute brainstem syndrome other than APS •Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic lesion on MRI ¥ • Acute cerebral syndrome with NMOSD-typical brain lesion on MRI §
ψ Typical MRI findings in NMOSD related to clinical presentation (T2 unless noted otherwise)
• Optic neuritis: Normal cerebral MRI (or only nonspecific white matter lesions) OR longitudinally extensive optic nerve lesion (≥ half of the length of the optic nerve or involving optic chiasm; T2 or T1/Gd) •Myelitis: Intramedullary lesion ≥ 3 contiguous VS (LETM) OR focal atrophy ≥ 3 contiguous VS in patients with a history of acute myelitis •Area postrema syndrome (APS): Lesion in the dorsal medulla oblongata/area postrema •Other brainstem syndrome: Periependymal brainstem lesion (4th ventricle) •¥ Diencephalic syndrome: Periependymal lesion (3rd ventricle) OR hypothalamic/thalamic lesion • § Cerebral syndrome: Extensive periependymal lesion (lateral ventricle; often with Gd) OR long (> 1⁄2 length), diffuse, heterogeneous or edematous corpus callosum lesion OR long corticospinal tract lesion (unilateral or bilateral, contiguously involving internal capsule and cerebral peduncle) OR large, confluent (unilateral or bilateral) subcortical or deep white matter lesion
**LETM = longitudinally extensive transverse myelitis lesions*

<u>Antisynthetase Syndrome-Related Interstitial Lung Disease ‡</u>

1. Patient has antisynthetase antibody positive disease (e.g., anti-Jo-1, -PL-7, -PL-12, -OJ, -EJ, etc.); **AND**
2. Physician has assessed baseline disease severity utilizing an objective measure (i.e., baseline glucocorticoid use, pulmonary function testing [i.e., forced vital capacity (FVC%), total lung capacity (TLC%), diffusing capacity of the lungs for carbon monoxide (DLCO%)], or chest CT scan); **AND**

3. Patient has documented severe active disease; **AND**
4. Patient has recurrent or progressive disease despite treatment with glucocorticoids and/or other immunosuppressive agents (e.g., azathioprine, mycophenolate mofetil, cyclophosphamide, tacrolimus, etc.); **AND**
5. Will be used in combination with glucocorticoids or other immunosuppressive agents (e.g., azathioprine, mycophenolate mofetil, cyclophosphamide, tacrolimus, etc.), unless the patient has a contraindication or intolerance

Idiopathic Membranous Nephropathy ‡

1. Patient has a documented diagnosis of idiopathic (primary) membranous nephropathy; **AND**

2. Secondary causes of membranous nephropathy have been ruled out [e.g., infections, autoimmune diseases, malignancies, nutritional supplements (e.g., lipoic acid, etc.), nonsteroidal anti-inflammatory drugs (NSAIDs), etc.]; **AND**

A. Used as first-line therapy in patients with any of the following moderate to high risk factors for progressive disease:

i. Proteinuria > 3.5 g/day and no decrease > 50% after 6 months of therapy with an angiotensin converting enzyme inhibitor (ACEi) or angiotensin II receptor blocker (ARB); **OR**

ii. eGFR < 60 ml/min/1.73m2; **OR**

iii. Proteinuria > 8 g/d for > 6 months; **OR**

iv. Patient has experienced serious complications of nephrotic syndrome (e.g., acute kidney injury, infection, thromboembolic events, etc.); **OR**

B. Used for initial disease relapse following remission on first-line therapy with rituximab, a calcineurin inhibitor (e.g., tacrolimus, cyclosporine, etc.) or cyclophosphamide in combination with glucocorticoids; **OR**

C. Used for treatment-resistance to first-line therapy with rituximab, a calcineurin inhibitor (e.g., tacrolimus, cyclosporine, etc.) or cyclophosphamide in combination with glucocorticoids; **AND**

i. Patient has a stable eGFR; **AND**

ii. Will be used in combination with a calcineurin inhibitor if previously treated with rituximab alone in the first-line setting; **OR**

D. Used for disease recurrence following kidney transplant; **AND**

i. Patient has proteinuria > 1 g/d

Pediatric Idiopathic Nephrotic Syndrome ‡

1. Patient is 12 years of age or younger

2. Patient has symptomatic disease (i.e., nephrotic-range proteinuria and either hypoalbuminemia or edema when albumin level is not available)

3. Patient has been diagnosed with one of the following:

A. Frequently relapsing nephrotic syndrome (FRNS) with at least four relapses per year or at least two relapses within 6 months of initial presentation

B. Steroid dependent nephrotic syndrome (SDNS) with two consecutive relapses during steroid tapering or within 14 days of cessation of therapy

C. Steroid resistant nephrotic syndrome (SRNS) with failure to achieve complete remission within a 4-6 -week course of daily corticosteroids; **AND**

4. Patient has failed an adequate trial with at least one other steroid-sparing agent (e.g., cyclophosphamide, calcineurin inhibitor [e.g., tacrolimus, cyclosporine, etc.], mycophenolate mofetil, etc.)

IgG4-Related Disease ‡

1. Physician has assessed baseline disease severity utilizing an objective measure/tool (e.g., IgG4-RD Responder Index score, physician's global assessment [PGA], amount of glucocorticoid or other immunosuppressive use, incidence of disease flares, serum IgG4 level, etc.); **AND**

2. Other conditions that mimic IgG4-related disease have been ruled out (e.g., malignancy, infection, other autoimmune disorders, etc.); **AND**

3. Patient has documented active disease; **AND**

4. Documented failure or ineffective response to an adequate trial with glucocorticoids, unless there is a contraindication or intolerance to use

† FDA-labeled indication(s); **‡** Compendia recommended indication(s); Φ Orphan Drug

II. Renewal Criteria

Coverage can be renewed based upon the following criteria:

1. Continuation of documented current and/or successful therapy with a non-preferred agent (Rituxan or Riabni); **AND**
2. Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include the following: severe infusion reactions, tumor lysis syndrome (TLS), severe mucocutaneous reactions, progressive multifocal leukoencephalopathy (PML), viral hepatitis, serious bacterial, fungal, or viral infections, cardiac arrhythmias, renal toxicity, bowel obstruction or perforation; **AND**

Oncology Indications:

1. Tumor response with stabilization of disease or decrease in size of tumor or tumor spread; **AND**
2. Patient has not exceeded dosing or duration limits as defined above

Adult Acute Lymphoblastic Leukemia (ALL)

• Treatment response or stabilization of disease as indicated by CBC, bone marrow cytogenic analysis, QPCR, or FISH

Pediatric B-Cell Acute Leukemia and Aggressive Mature B-Cell Lymphomas (induction or consolidation therapy)

• Coverage may NOT be renewed

Pediatric Hodgkin Lymphoma

• Coverage may NOT be renewed

Chronic Graft-Versus-Host Disease (cGVHD)

• Coverage may NOT be renewed

Hematopoietic Cell Transplantation

• Coverage may NOT be renewed

Management of Immunotherapy-Related Toxicities

• Coverage for use in the treatment of myositis/myasthenia gravis/encephalitis may NOT be renewed

• Coverage for use in bullous dermatitis: Patient has not exceeded a maximum of 18 months of therapy (4 total doses)

All Other Oncology Indications

• Disease response with treatment as defined by stabilization of disease or decrease in size of tumor or tumor spread

Non-Oncology Indications:

Rheumatoid arthritis (RA)

1. Disease response as indicated by improvement in signs and symptoms compared to baseline such as the number of tender and swollen joint counts and/or an improvement on a disease activity scoring tool [e.g. an improvement on a composite scoring index such as Disease Activity Score-28 (DAS28) of 1.2 points or more or a ≥20% improvement on the American College of Rheumatology-20 (ACR20) criteria]; **AND**
2. Dose escalation (up to the maximum dose and frequency specified below) may occur upon clinical review on a case-by-case basis provided that the patient has:
    A. Shown an initial response to therapy; **AND**
    B. Received a minimum of one maintenance dose at the dose and interval specified below; **AND**
    C. Responded to therapy with subsequent loss of response

Thrombocytopenic purpura (ITP or Evans Syndrome)

1. Disease response as indicated by the achievement and maintenance of a platelet count of at least 50×10^9/L as necessary to reduce the risk for bleeding

Thrombotic Thrombocytopenic Purpura (TTP)

1. Disease response as indicated by an increase in ADAMTS13 activity with a reduction in thrombotic risk

Multiple Sclerosis (MS)

1.. Continuous monitoring of response to therapy indicates a beneficial response*

[manifestations of MS disease activity include, but are not limited to, an increase in annualized relapse rate (ARR), development of new/worsening T2 hyperintensities or enhancing lesions on brain/spinal MRI, and progression of sustained impairment as evidenced by expanded disability status scale (EDSS), timed 25-foot walk (T25-FW), 9-hole peg test (9-HPT)]

**Note: – Inadequate response, in those who have been adherent and receiving therapy for sufficient time to realize the full treatment effect, is defined as ≥ 1 relapse, ≥ 2 unequivocally new MRI-detected lesions, or increased disability on examination over a one-year period*

Granulomatosis with Polyangiitis (GPA) (Wegener's granulomatosis) and Microscopic polyangiitis (MPA)

1. Disease response as indicated by improvement in signs and symptoms of condition compared to baseline; **AND**
2. Decreased frequency in the occurrence of major relapses (defined by the reappearance of clinical and/or laboratory signs of vasculitis activity that could lead to organ failure or damage, or could be life threatening)

Pemphigus vulgaris

1. Patient is currently receiving tapering doses of corticosteroids or has discontinued use of corticosteroids; **AND**

   A. Disease response as indicated by complete epithelialization of lesions and improvement in signs and symptoms of condition compared to baseline; **OR**

   B. Patient has not experienced continued development of new lesions, continued extension of old lesions, or failure of established lesions to begin to heal despite therapy; **OR**

      i. For Relapses ONLY: Patient has had active disease control; **AND**

      ii. Patient has the appearance of 3 or more new lesions a month that do not heal spontaneously within 1 week, or by the extension of established lesions

Autoimmune hemolytic anemia (AIHA)

1. Disease response as indicated by improvement in anemia signs and symptoms (e.g., dyspnea, fatigue, etc.) ; **AND**
2. Patient has improvement in laboratory values (Hb/Hct), reduced transfusion needs, and/or reduced glucocorticoid use

Systemic Lupus Erythematosus (SLE)

1. Adequate documentation of disease stability and/or improvement as indicated by one or more of the following when compared to pre-treatment baseline:

   A. Improvement in the SELENA-SLEDAI-2K; **OR**

   B. Reduction of baseline BILAG-2004 from A to B or from B to C/D, and no BILAG-2004 worsening in other organ systems, as defined by ≥2 new BILAG-2004 B; **OR**

   C. No worsening (<0.30 points increase) in Physician's Global Assessment (PGA) score; **OR**

   D. Seroconverted (negative)

Lupus Nephritis

1. Coverage may only be renewed in patients experiencing a disease relapse (e.g., increased serum creatinine, increase in protein urine excretion, decrease in eGFR, etc.)

Myasthenia Gravis (unrelated to immunotherapy-related toxicity)

1. Disease response as indicated by a decrease in the daily dose of corticosteroids and/or an improvement in signs and symptoms compared to baseline.

Complications of Transplanted Solid Organ (kidney, liver, lung, heart, pancreas)

1. Coverage may NOT be renewed.

NMOSD

1. Disease response as indicated by stabilization/improvement in any of the following:

A. Decrease in acute relapses or improvement of stability
B. Reduced hospitalizations
C. Reduction/discontinuation in plasma exchange treatments
D. Reduction/discontinuation of corticosteroids without relapse

Antisynthetase Syndrome-Related Interstitial Lung Disease

1. Disease response as indicated by stabilization/improvement in any of the following:
    A. Reduction or stabilization of glucocorticoid use from baseline
    B. Improvement or stabilization of pulmonary function testing (i.e., improvement defined as ≥10% increase in FVC%, TLC%, or DLCO%; stabilization defined as < 10% decrease in FVC%, TLC%, or DLCO%)
    C. Improvement or stabilization of chest CT score (improvement defined as >10% decrease in CT score, stabilization defined as a ≤ 10% increase in CT score)

Idiopathic Membranous Nephropathy

1. Patient experienced beneficial disease response with improvement in symptoms and/or other objective measures compared to baseline (e.g., reduction in proteinuria, increase and/or normalization of serum albumin, improvement/stability of serum creatinine and/or eGFR, decrease in anti-PLA2R antibody levels, etc.); **OR**
2. Patient has resistant disease following first-line therapy with rituximab; **AND**
    A. Patient has stable eGFR; **AND**
    B. Will be used in combination with a calcineurin inhibitor if previously treated with rituximab alone in the first-line setting

Pediatric Idiopathic Nephrotic Syndrome ‡

1. Patient previously achieved beneficial disease response from the prior course of therapy; **AND**
2. Patient is experiencing signs and symptoms of recurrent active disease necessitating additional doses (e.g., recurrence of nephrotic-range proteinuria with a dipstick ≥ 3+ [≥300 mg/dL] for 3 consecutive days **OR** urinary protein creatinine ratio [UPCR] ≥200 mg/mmol [≥2 mg/mg] on a spot urine sample on 3 consecutive days, with or without reappearance of edema in a child who had previously achieved complete remission)

IgG4-Related Disease ‡

1. Patient experienced beneficial disease response with improvement in involved organ-related symptoms and/or other objective measures compared to baseline (e.g. improvement in the IgG4-RD Responder Index score of > 2 points, improvement in the physician's global assessment [PGA], reduction in glucocorticoid or other immunosuppressive use, reduction of disease flares, reduction in serum IgG4 level, etc.); **AND**
2. Patient meets one of the following:
    A. Ongoing maintenance therapy is required due to patient having a high-risk of relapse
    B. Patient is experiencing signs and symptoms of relapsed active disease necessitating an additional course of therapy

Dosage and Administration

Indication		Dose
CLL/SLL	Initial Therapy	375 mg/m² intravenously (IV) weekly for 8 doses; **OR** 375 mg/m² IV cycle 1, then 500 mg/m² every 28 days cycles 2-6 (6 total doses); **OR**

Indication		Dose
		375 mg/m² IV cycle 1, followed by 500 mg/m² every 2 weeks for 4 doses, then 500 mg/m² every 28 days for 3 doses (8 total doses)
	Renewal Therapy	375 mg/m² IV every 3 months; **OR** 500 mg/ m² IV every 8 weeks
Adult B-Cell Lymphomas, Castleman Disease, Primary Cutaneous B-Cell Lymphomas, Waldenström Macroglobulinemia, or Adult HL	Initial Therapy	375 mg/m2 IV once weekly for 4 – 8 doses in a 6 month period
	Renewal Therapy	375 mg/m² IV once weekly for 4 doses per 6 month period; OR 375 mg/ m² IV every 8 weeks
Pediatric Aggressive Mature B-Cell Lymphomas		Induction* [courses 1 and 2 (COPDAM1 and COPDAM2)] 375 mg/m2 IV, two doses during each of the induction courses (Day -2 and Day 1). During the 1st induction course, prednisone is given as part of the chemotherapy course, and should be administered prior to rituximab. Rituximab will be given 48 hours after the first infusion of rituximab. Consolidation* [courses 1 and 2 (CYM/CYVE)] 375 mg/m2 IV, one dose during each of the consolidation courses (Day 1) Relapsed/Refractory RCYVE – 375mg/m2 IV on day 1 of each 21-day cycle RICE – 375 mg/m2 IV on days 1 and 3 of courses 1 and 2, and on day 1 only of course 3 if needed. **Note: dosing and dosing schedules are highly variable and dependent on regimen used, please refer to NCCN and PI for additional protocols*
Pediatric Mature B-Cell Acute Leukemia		Induction* [courses 1 and 2 (COPDAM1 and COPDAM2)] 375 mg/m2 IV, two doses during each of the induction courses (Day -2 and Day 1). During the 1st induction course, prednisone is given as part of the chemotherapy course, and should be administered prior to rituximab. Rituximab will be given 48 hours after the first infusion of rituximab. Consolidation* [courses 1 and 2 (CYM/CYVE)] 375 mg/m2 IV, one dose during each of the consolidation courses (Day 1) **Note: dosing and dosing schedules are highly variable and dependent on regimen used, please refer to NCCN and PI for additional protocols*
CNS Lymphoma		Intravenous administration

Indication	Dose
	Initial Therapy: 375 mg/m2 IV once weekly for 4 – 8 doses in a 6 month period Renewal Therapy: 375 mg/m² IV once weekly for 4 doses per 6 month period; OR 375 mg/m² IV every 8 weeks Intrathecal/Intraventricular administration 10-40 mg weekly to every 3 weeks
ALL	375 mg/m2 IV up to twice weekly for a total of 16 to 18 infusions (e.g., induction [days 1 and 7], salvage reinduction when necessary [days 1 and 7], consolidation [4 infusions: blocks 1, 3, 4, and 6], late intensification [days 1 and 7], late consolidation [2 infusions: blocks 7 and 9], and maintenance [6 infusions])
Hairy Cell Leukemia	375 mg/m2 IV once weekly for 4 – 8 doses; OR 375mg/m2 IV on days 1 and 15 every 28 days for 4 cycles, then 375mg/m2 IV every 4 weeks for 4 cycles (up to 8 total cycles)
RA	1,000 mg on days 1 and 15, repeated every 24 weeks. May repeat up to every 16 weeks in patients requiring more frequent dosing based on clinical evaluation.
Pemphigus Vulgaris (PV)	*Initiation* *1,000 mg IV on days 1 and 15; OR* *375 mg/m² IV weekly for 4 doses* *Maintenance* *500 mg IV at month 12 and repeat every 6 months thereafter or based on clinical evaluation* *Relapse* *1,000 mg IV upon relapse, resumption of glucocorticoids may be considered* **Subsequent infusions (maintenance and relapse) should be no sooner than 16 weeks after the previous infusion.*
AIHA	Warm-reactive disease 375 mg/m² IV weekly for 4 doses in a 6 month period; OR 1,000 mg IV on days 1 and 15 Cold agglutinin disease 375 mg/m² IV weekly for 4 doses in a 6 month period
Thrombocytopenic Purpura or Thrombotic Thrombocytopenic Purpura (TTP)	375 mg/m² IV weekly for 4 doses; OR 1,000 mg IV on days 1 and 15
Management of ImmunotherapyRelated Toxicities	Bullous Dermatitis 1,000 mg IV every 2 weeks for 2 doses, then 500 mg IV at months 12 and 18 as needed Myositis

Indication	Dose
	375 mg/m² IV weekly for 4 doses Myasthenia Gravis 375 mg/m² IV weekly for 4 doses; OR 500 mg/m² IV every 2 weeks for 2 doses Encephalitis 1,000 mg IV every 2 weeks for 2 doses; OR 375 mg/m² IV weekly for 4 doses
Granulomatosis with Polyangiitis (GPA) (Wegener's Granulomatosis) and Microscopic Polyangiitis (MPA)	Induction (Pediatric and Adult) 375 mg/m² IV weekly for 4 doses; OR Adults: 1,000 mg IV on days 1 and 15; OR Pediatric (up to a maximum of 1,000 mg per dose): o 575 mg/m² IV on days 1 and 15 (BSA ≤1.5m²) o 750 mg/m² IV on days 1 and 15 (BSA >1.5m²) Maintenance Pediatric: o 250 mg/m² IV on days 1 and 15, then 250 mg/m² IV every 6 months thereafter based on clinical evaluation Adult: o 500 mg IV on days 1 and 15, then 500 mg IV every 6 months thereafter based on clinical evaluation *Initial MAINTENANCE infusions should be no sooner than 16 weeks and no later than 24 weeks after the previous infusion if rituximab was used for initial induction therapy. *Initial MAINTENANCE infusions should be initiated within 4 weeks following disease control when initial induction occurred with other standard of care immunosuppressants
cGVHD	375 mg/m² IV weekly for 4 doses, then 375 mg/m² IV monthly for 4 months -OR- 375 mg/m² IV weekly for 4 doses (Note: A second course of 4 weekly doses may be administered 8 weeks after initial therapy for patients with lack of or incomplete response.) -OR- 375 mg/m² IV weekly for 4 – 8 doses
Hematopoietic Cell Transplantation	Conditioning: 375 mg/m² IV for 1 day before transplant, then 1000 mg/m² IV on days 1,8, and 15 after transplant
Multiple Sclerosis	1,000 mg IV on days 1 and 15, repeat every 6 months
NMOSD	1,000 mg IV once on days 1 and 15, repeat every 6 months -OR 375 mg/m2 once weekly for 4 weeks, repeat every 6 months

Indication	Dose
Histiocytic Neoplasms – Rosai-Dorfman Disease	500 mg/m² IV every 1 – 2 weeks for 2 – 6 doses every 6 months
SLE	1,000 mg IV on days 1 and 15 -OR 375 mg/m2 IV once weekly for 4 doses
Lupus Nephritis	1,000 mg IV on days 1 and 15 -OR 375 mg/m2 IV once weekly for 4 doses
Myasthenia Gravis (unrelated to immunotherapy-related toxicity)	1,000 mg IV on days 1 and 15, may repeat a full or partial course every 6 months -OR 375 mg/m2 IV once weekly for 4 doses, may repeat a full or partial course every 6 months
Pediatric Hodgkin Lymphoma	375 mg/m² IV on day 1 of every 2-3 week cycle for a total of 3 cycles
Complications of transplanted solid organ (kidney, liver, lung, heart, pancreas)	Adults and pediatrics weighing ≥0.5 m2: 375 mg/m2 weekly for up to 4 doses – Pediatrics weighing <0.5 m2: 12.5 mg/kg weekly for up to 4 doses
Antisynthetase Syndrome-Related Interstitial Lung Disease	1,000 mg IV on days 1 and 15 repeated every 6 months -OR 375 mg/m2 IV once weekly for 4 doses repeated every 6 months
Pediatric Idiopathic Nephrotic Syndrome	375 mg/m2 IV once weekly for 1-4 doses
Idiopathic Membranous Nephropathy	375 mg/m2 IV once weekly for 1-4 doses every 6 months -OR 1,000 mg IV on days 1 and 15 every 6 months
IgG4-Related Disease	Induction: 375 mg/m2 IV once weekly for 1-4 doses -OR1,000 mg IV on days 1 and 15 *Subsequent infusions (maintenance and relapse) may be administered at either induction schedule above and should be repeated no sooner than every 6 months

Applicable Procedure Codes

Code	Description
J9312	Injection, rituximab, 10mg
Q5115	Injection, rituximab-abbs, biosimilar, (Truxima), 10 mg
Q5119	Injection, rituximab-pvvr, biosimilar, 10 mg (Ruxience). Effective Date: 07/01/2020
Q5123	Injection, rituximab-arrx, biosimilar, (Riabni), 10 mg

Applicable NDCs

Code	Description
50242-0051-xx	Rituxan 100 mg/10 mL single-use vial for injection
50242-0053-xx	Rituxan 500 mg/50 mL single-use vial for injection
63459-103-10	rituximab-abbs injection, carton containing one 100 mg/10 mL (10 mg/mL) single-dose vial
63459-104-50	rituximab-abbs injection, carton containing one 500 mg/50 mL (10 mg/mL) single-dose vial
55513-0326-01	rituximab-arrx injection, carton containing one 100mg/10ml (10mg/ml)-single dose vial
55513-0224-01	rituximab-arrx injection, carton containing one 500 mg/50 mL (10 mg/mL) single-dose vial
00069-0238-xx	Ruxience 100 mg/10 mL single-dose vial for injection
00069-0249-xx	Ruxience 500 mg/50 mL single-dose vial for injection

ICD-10 Diagnoses

Code	Description
B20	Human immunodeficiency virus [HIV] disease
C79.32	Secondary malignant neoplasm of cerebral meninges
C81.00	Nodular lymphocyte predominant Hodgkin lymphoma, unspecified site
C81.01	Nodular lymphocyte predominant Hodgkin lymphoma, lymph nodes of head, face, and neck
C81.02	Nodular lymphocyte predominant Hodgkin lymphoma, intrathoracic lymph nodes
C81.03	Nodular lymphocyte predominant Hodgkin lymphoma, intra-abdominal lymph nodes
C81.04	Nodular lymphocyte predominant Hodgkin lymphoma, lymph nodes of axilla and upper limb
C81.05	Nodular lymphocyte predominant Hodgkin lymphoma, lymph nodes of inguinal region and lower limb
C81.06	Nodular lymphocyte predominant Hodgkin lymphoma, intrapelvic lymph nodes
C81.07	Nodular lymphocyte predominant Hodgkin lymphoma, spleen
C81.08	Nodular lymphocyte predominant Hodgkin lymphoma, lymph nodes of multiple sites
C81.09	Nodular lymphocyte predominant Hodgkin lymphoma, extranodal and solid organ sites
C82.00	Follicular lymphoma grade I, unspecified site
C82.01	Follicular lymphoma grade I, lymph nodes of head, face and neck
C82.02	Follicular lymphoma, grade I, intrathoracic lymph nodes
C82.03	Follicular lymphoma grade I, intra-abdominal lymph nodes
C82.04	Follicular lymphoma grade I, lymph nodes of axilla and upper limb
C82.05	Follicular lymphoma grade I, lymph nodes of inguinal regional and lower limb
C82.06	Follicular lymphoma grade I, intrapelvic lymph nodes
C82.07	Follicular lymphoma grade I, spleen
C82.08	Follicular lymphoma grade I, lymph nodes of multiple sites
C82.09	Follicular lymphoma grade I, extranodal and solid organ sites
C82.10	Follicular lymphoma grade II, unspecified site
C82.11	Follicular lymphoma grade II, lymph nodes of head, face and neck
C82.12	Follicular lymphoma, grade II, intrathoracic lymph nodes
C82.13	Follicular lymphoma grade II, intra-abdominal lymph nodes
C82.14	Follicular lymphoma grade II, lymph nodes of axilla and upper limb
C82.15	Follicular lymphoma grade II, lymph nodes of inguinal region and lower limb
C82.16	Follicular lymphoma grade II, intrapelvic lymph nodes
C82.17	Follicular lymphoma grade II, spleen
C82.18	Follicular lymphoma grade II, lymph nodes of multiple sites
C82.19	Follicular lymphoma grade II, extranodal and solid organ sites
C82.20	Follicular lymphoma grade III, unspecified, unspecified site

C82.21	Follicular lymphoma grade III, unspecified, lymph nodes of head, face and neck
C82.22	Follicular lymphoma, grade III, unspecified, intrathoracic lymph nodes
C82.23	Follicular lymphoma grade III, unspecified, intra-abdominal lymph nodes
C82.24	Follicular lymphoma grade III, unspecified, lymph nodes of axilla and upper limb
C82.25	Follicular lymphoma grade III, unspecified, lymph nodes of inguinal region and lower limb
C82.26	Follicular lymphoma grade III, unspecified, intrapelvic lymph nodes
C82.27	Follicular lymphoma grade III, unspecified, spleen
C82.28	Follicular lymphoma grade III, unspecified, lymph nodes of multiple sites
C82.29	Follicular lymphoma grade III, unspecified, extranodal and solid organ sites
C82.30	Follicular lymphoma grade IIIa, unspecified site
C82.31	Follicular lymphoma grade IIIa, lymph nodes of head, face and neck
C82.32	Follicular lymphoma, grade IIIa, intrathoracic lymph nodes
C82.33	Follicular lymphoma grade IIIa, intra-abdominal lymph nodes
C82.34	Follicular lymphoma grade IIIa, lymph nodes of axilla and upper limb
C82.35	Follicular lymphoma grade IIIa, lymph nodes of inguinal region and lower limb
C82.36	Follicular lymphoma grade IIIa, intrapelvic lymph nodes
C82.37	Follicular lymphoma grade IIIa, spleen
C82.38	Follicular lymphoma grade IIIa, lymph nodes of multiple sites
C82.39	Follicular lymphoma grade IIIa, extranodal and solid organ sites
C82.40	Follicular lymphoma grade IIIb, unspecified site
C82.41	Follicular lymphoma grade IIIb, lymph nodes of head, face and neck
C82.42	Follicular lymphoma, grade IIIb, intrathoracic lymph nodes
C82.43	Follicular lymphoma grade IIIb, intra-abdominal lymph nodes
C82.44	Follicular lymphoma grade IIIb, lymph nodes of axilla and upper limb
C82.45	Follicular lymphoma grade IIIb, lymph nodes of inguinal region and lower limb
C82.46	Follicular lymphoma grade IIIb, intrapelvic lymph nodes
C82.47	Follicular lymphoma grade IIIb, spleen
C82.48	Follicular lymphoma grade IIIb, lymph nodes of multiple sites
C82.49	Follicular lymphoma grade IIIb, extranodal and solid organ sites
C82.50	Diffuse follicle center lymphoma, unspecified site
C82.51	Diffuse follicle center lymphoma, lymph nodes of head, face and neck
C82.52	Diffuse follicle center lymphoma, intrathoracic lymph nodes
C82.53	Diffuse follicle center lymphoma, intra-abdominal lymph nodes
C82.54	Diffuse follicle center lymphoma, lymph nodes of axilla and upper limb
C82.55	Diffuse follicle center lymphoma, lymph nodes of inguinal region and lower limb
C82.56	Diffuse follicle center lymphoma, intrapelvic lymph nodes
C82.57	Diffuse follicle center lymphoma, spleen
C82.58	Diffuse follicle center lymphoma, lymph nodes of multiple sites
C82.59	Diffuse follicle center lymphoma, extranodal and solid organ sites
C82.60	Cutaneous follicle center lymphoma, unspecified site
C82.61	Cutaneous follicle center lymphoma, lymph nodes of head, face and neck
C82.62	Cutaneous follicle center lymphoma, intrathoracic lymph nodes
C82.63	Cutaneous follicle center lymphoma, intra-abdominal lymph nodes
C82.64	Cutaneous follicle center lymphoma, lymph nodes of axilla and upper limb
C82.65	Cutaneous follicle center lymphoma, lymph nodes of inguinal region and lower limb
C82.66	Cutaneous follicle center lymphoma, intrapelvic lymph nodes

C82.67	Cutaneous follicle center lymphoma, spleen
C82.68	Cutaneous follicle center lymphoma, lymph nodes of multiple sites
C82.69	Cutaneous follicle center lymphoma, extranodal and solid organ sites
C82.80	Other types of follicular lymphoma, unspecified site
C82.81	Other types of follicular lymphoma, lymph nodes of head, face and neck
C82.82	Other types of follicular lymphoma, intrathoracic lymph nodes
C82.83	Other types of follicular lymphoma, intra-abdominal lymph nodes
C82.84	Other types of follicular lymphoma, lymph nodes of axilla and upper limb
C82.85	Other types of follicular lymphoma, lymph nodes of inguinal region and lower limb
C82.86	Other types of follicular lymphoma, intrapelvic lymph nodes
C82.87	Other types of follicular lymphoma, spleen
C82.88	Other types of follicular lymphoma, lymph nodes of multiple sites
C82.89	Other types of follicular lymphoma, extranodal and solid organ sites
C82.90	Follicular lymphoma, unspecified, unspecified site
C82.91	Follicular lymphoma, unspecified, lymph nodes of head, face and neck
C82.92	Follicular lymphoma, unspecified, intrathoracic lymph nodes
C82.93	Follicular lymphoma, unspecified, intra-abdominal lymph nodes
C82.94	Follicular lymphoma, unspecified, lymph nodes of axilla and upper limb
C82.95	Follicular lymphoma, unspecified lymph nodes of inguinal region and lower limb
C82.96	Follicular lymphoma, unspecified, intrapelvic lymph nodes
C82.97	Follicular lymphoma, unspecified, spleen
C82.98	Follicular lymphoma, unspecified, lymph nodes of multiple sites
C82.99	Follicular lymphoma, unspecified, extranodal and solid organ sites
C83.00	Small cell B-cell lymphoma, unspecified site
C83.01	Small cell B-cell lymphoma, lymph nodes of head, face and neck
C83.02	Small cell B-cell lymphoma, intrathoracic lymph nodes
C83.03	Small cell B-cell lymphoma, intra-abdominal lymph nodes
C83.04	Small cell B-cell lymphoma, lymph nodes of axilla and upper limb
C83.05	Small cell B-cell lymphoma, lymph nodes of inguinal region and lower limb
C83.06	Small cell B-cell lymphoma, intrapelvic lymph nodes
C83.07	Small cell B-cell lymphoma, spleen
C83.08	Small cell B-cell lymphoma, lymph nodes of multiple sites
C83.09	Small cell B-cell lymphoma, extranodal and solid organ sites
C83.10	Mantle cell lymphoma, unspecified site
C83.11	Mantle cell lymphoma, lymph nodes of head, face and neck
C83.12	Mantle cell lymphoma, intrathoracic lymph nodes
C83.13	Mantle cell lymphoma, intra-abdominal lymph nodes
C83.14	Mantle cell lymphoma, lymph nodes of axilla and upper limb
C83.15	Mantle cell lymphoma, lymph nodes of inguinal region and lower limb
C83.16	Mantle cell lymphoma, intrapelvic lymph nodes
C83.17	Mantle cell lymphoma, spleen
C83.18	Mantle cell lymphoma, lymph nodes of multiple sites
C83.19	Mantle cell lymphoma, extranodal and solid organ sites
C83.30	Diffuse large B-cell lymphoma unspecified site
C83.31	Diffuse large B-cell lymphoma, lymph nodes of head, face, and neck
C83.32	Diffuse large B-cell lymphoma intrathoracic lymph nodes

C83.33	Diffuse large B-cell lymphoma intra-abdominal lymph nodes
C83.34	Diffuse large B-cell lymphoma lymph nodes of axilla and upper limb
C83.35	Diffuse large B-cell lymphoma, lymph nodes of inguinal region and lower limb
C83.36	Diffuse large B-cell lymphoma intrapelvic lymph nodes
C83.37	Diffuse large B-cell lymphoma, spleen
C83.38	Diffuse large B-cell lymphoma lymph nodes of multiple sites
C83.39	Diffuse large B-cell lymphoma extranodal and solid organ sites
C83.70	Burkitt lymphoma, unspecified site
C83.71	Burkitt lymphoma, lymph nodes of head, face, and neck
C83.72	Burkitt lymphoma, intrathoracic lymph nodes
C83.73	Burkitt lymphoma, intra-abdominal lymph nodes
C83.74	Burkitt lymphoma, lymph nodes of axilla and upper limb
C83.75	Burkitt lymphoma, lymph nodes of inguinal region and lower limb
C83.76	Burkitt lymphoma, intrapelvic lymph nodes
C83.77	Burkitt lymphoma, spleen
C83.78	Burkitt lymphoma, lymph nodes of multiple sites
C83.79	Burkitt lymphoma, extranodal and solid organ sites
C83.80	Other non-follicular lymphoma, unspecified site
C83.81	Other non-follicular lymphoma, lymph nodes of head, face and neck
C83.82	Other non-follicular lymphoma, intrathoracic lymph nodes
C83.83	Other non-follicular lymphoma, intra-abdominal lymph nodes
C83.84	Other non-follicular lymphoma, lymph nodes of axilla and upper limb
C83.85	Other non-follicular lymphoma, lymph nodes of inguinal region and lower limb
C83.86	Other non-follicular lymphoma, intrapelvic lymph nodes
C83.87	Other non-follicular lymphoma, spleen
C83.88	Other non-follicular lymphoma, lymph nodes of multiple sites
C83.89	Other non-follicular lymphoma, extranodal and solid organ sites
C83.90	Non-follicular (diffuse) lymphoma, unspecified site
C83.91	Non-follicular (diffuse) lymphoma, unspecified lymph nodes of head, face, and neck
C83.92	Non-follicular (diffuse) lymphoma, unspecified intrathoracic lymph nodes
C83.93	Non-follicular (diffuse) lymphoma, unspecified intra-abdominal lymph nodes
C83.94	Non-follicular (diffuse) lymphoma, unspecified lymph nodes of axilla and upper limb
C83.95	Non-follicular (diffuse) lymphoma, unspecified lymph nodes of inguinal region and lower limb
C83.96	Non-follicular (diffuse) lymphoma, unspecified intrapelvic lymph nodes
C83.97	Non-follicular (diffuse) lymphoma, unspecified spleen
C83.98	Non-follicular (diffuse) lymphoma, unspecified lymph nodes of multiple sites
C83.99	Non-follicular (diffuse) lymphoma, unspecified extranodal and solid organ sites
C85.20	Mediastinal (thymic) large B-cell lymphoma, unspecified site
C85.21	Mediastinal (thymic) large B-cell lymphoma, lymph nodes of head, face and neck
C85.22	Mediastinal (thymic) large B-cell lymphoma, intrathoracic lymph nodes
C85.23	Mediastinal (thymic) large B-cell lymphoma, intra-abdominal lymph nodes
C85.24	Mediastinal (thymic) large B-cell lymphoma, lymph nodes of axilla and upper limb
C85.25	Mediastinal (thymic) large B-cell lymphoma, lymph nodes of inguinal region and lower limb
C85.26	Mediastinal (thymic) large B-cell lymphoma, intrapelvic lymph nodes
C85.27	Mediastinal (thymic) large B-cell lymphoma, spleen
C85.28	Mediastinal (thymic) large B-cell lymphoma, lymph nodes of multiple sites

C85.29	Mediastinal (thymic) large B-cell lymphoma, extranodal and solid organ sites
C85.80	Other specified types of non-Hodgkin lymphoma, unspecified site
C85.81	Other specified types of non-Hodgkin lymphoma, lymph nodes of head, face and neck
C85.82	Other specified types of non-Hodgkin lymphoma, intrathoracic lymph nodes
C85.83	Other specified types of non-Hodgkin lymphoma, intra-abdominal lymph nodes
C85.84	Other specified types of non-Hodgkin lymphoma, lymph nodes of axilla and upper limb
C85.85	Other specified types of non-Hodgkin lymphoma, lymph nodes of inguinal region of lower limb
C85.86	Other specified types of non-Hodgkin lymphoma, intrapelvic lymph nodes
C85.87	Other specified types of non-Hodgkin lymphoma, spleen
C85.88	Other specified types of non-Hodgkin lymphoma, lymph nodes of multiple sites
C85.89	Other specified types of non-Hodgkin lymphoma, extranodal and solid organ sites
C88.0	Waldenström macroglobulinemia
C88.4	Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT-lymphoma)
C91.00	Acute lymphoblastic leukemia not having achieved remission
C91.01	Acute lymphoblastic leukemia, in remission
C91.02	Acute lymphoblastic leukemia, in relapse
C91.1	Chronic lymphocytic leukemia of B-cell type
C91.10	Chronic lymphocytic leukemia of B-cell type not having achieved remission
C91.12	Chronic lymphocytic leukemia of B-cell type in relapse
C91.40	Hairy cell leukemia not having achieved remission
C91.42	Hairy cell leukemia, in relapse
D36.0	Benign neoplasm of lymph nodes
D47.Z1	Post-transplant lymphoproliferative disorder (PTLD)
D47.Z2	Other neoplasms of uncertain behavior of lymphoid, hematopoietic and related tissue-Castleman disease
D59.1	Other autoimmune hemolytic anemias
D69.3	Immune thrombocytopenic purpura
D69.41	Evans Syndrome
D69.42	Congenital and hereditary thrombocytopenia purpura
D69.49	Other primary thrombocytopenia
D89.811	Chronic graft-versus-host disease
G04.81	Other encephalitis and encephalomyelitis
L10.0	Pemphigus vulgaris
M05.10	Rheumatoid lung disease with rheumatoid arthritis of unspecified site
M05.111	Rheumatoid lung disease with rheumatoid arthritis of right shoulder
M05.112	Rheumatoid lung disease with rheumatoid arthritis of left shoulder
M05.119	Rheumatoid lung disease with rheumatoid arthritis of unspecified shoulder
M05.121	Rheumatoid lung disease with rheumatoid arthritis of right elbow
M05.122	Rheumatoid lung disease with rheumatoid arthritis of left elbow
M05.129	Rheumatoid lung disease with rheumatoid arthritis of unspecified elbow
M05.131	Rheumatoid lung disease with rheumatoid arthritis of right wrist
M05.132	Rheumatoid lung disease with rheumatoid arthritis of left wrist
M05.139	Rheumatoid lung disease with rheumatoid arthritis of unspecified wrist
M05.141	Rheumatoid lung disease with rheumatoid arthritis of right hand
M05.142	Rheumatoid lung disease with rheumatoid arthritis of left hand
M05.149	Rheumatoid lung disease with rheumatoid arthritis of unspecified hand
M05.151	Rheumatoid lung disease with rheumatoid arthritis of right hip

M05.152	Rheumatoid lung disease with rheumatoid arthritis of left hip
M05.159	Rheumatoid lung disease with rheumatoid arthritis of unspecified hip
M05.161	Rheumatoid lung disease with rheumatoid arthritis of right knee
M05.162	Rheumatoid lung disease with rheumatoid arthritis of left knee
M05.169	Rheumatoid lung disease with rheumatoid arthritis of unspecified knee
M05.171	Rheumatoid lung disease with rheumatoid arthritis of right ankle and foot
M05.172	Rheumatoid lung disease with rheumatoid arthritis of left ankle and foot
M05.179	Rheumatoid lung disease with rheumatoid arthritis of unspecified ankle and foot
M05.19	Rheumatoid lung disease with rheumatoid arthritis of multiple sites
M05.20	Rheumatoid vasculitis with rheumatoid arthritis of unspecified site
M05.211	Rheumatoid vasculitis with rheumatoid arthritis of right shoulder
M05.212	Rheumatoid vasculitis with rheumatoid arthritis of left shoulder
M05.219	Rheumatoid vasculitis with rheumatoid arthritis of unspecified shoulder
M05.221	Rheumatoid vasculitis with rheumatoid arthritis of right elbow
M05.222	Rheumatoid vasculitis with rheumatoid arthritis of left elbow
M05.229	Rheumatoid vasculitis with rheumatoid arthritis of unspecified elbow
M05.231	Rheumatoid vasculitis with rheumatoid arthritis of right wrist
M05.232	Rheumatoid vasculitis with rheumatoid arthritis of left wrist
M05.239	Rheumatoid vasculitis with rheumatoid arthritis of unspecified wrist
M05.241	Rheumatoid vasculitis with rheumatoid arthritis of right hand
M05.242	Rheumatoid vasculitis with rheumatoid arthritis of left hand
M05.249	Rheumatoid vasculitis with rheumatoid arthritis of unspecified hand
M05.251	Rheumatoid vasculitis with rheumatoid arthritis of right hip
M05.252	Rheumatoid vasculitis with rheumatoid arthritis of left hip
M05.259	Rheumatoid vasculitis with rheumatoid arthritis of unspecified hip
M05.261	Rheumatoid vasculitis with rheumatoid arthritis of right knee
M05.262	Rheumatoid vasculitis with rheumatoid arthritis of left knee
M05.269	Rheumatoid vasculitis with rheumatoid arthritis of unspecified knee
M05.271	Rheumatoid vasculitis with rheumatoid arthritis of right ankle and foot
M05.272	Rheumatoid vasculitis with rheumatoid arthritis of left ankle and foot
M05.279	Rheumatoid vasculitis with rheumatoid arthritis of unspecified ankle and foot
M05.29	Rheumatoid vasculitis with rheumatoid arthritis of multiple sites
M05.30	Rheumatoid heart disease with rheumatoid arthritis of unspecified site
M05.311	Rheumatoid heart disease with rheumatoid arthritis of right shoulder
M05.312	Rheumatoid heart disease with rheumatoid arthritis of left shoulder
M05.319	Rheumatoid heart disease with rheumatoid arthritis of unspecified shoulder
M05.321	Rheumatoid heart disease with rheumatoid arthritis of right elbow
M05.322	Rheumatoid heart disease with rheumatoid arthritis of left elbow
M05.329	Rheumatoid heart disease with rheumatoid arthritis of unspecified elbow
M05.331	Rheumatoid heart disease with rheumatoid arthritis of right wrist
M05.332	Rheumatoid heart disease with rheumatoid arthritis of left wrist
M05.339	Rheumatoid heart disease with rheumatoid arthritis of unspecified wrist
M05.341	Rheumatoid heart disease with rheumatoid arthritis of right hand
M05.342	Rheumatoid heart disease with rheumatoid arthritis of left hand
M05.349	Rheumatoid heart disease with rheumatoid arthritis of unspecified hand
M05.351	Rheumatoid heart disease with rheumatoid arthritis of right hip

M05.352	Rheumatoid heart disease with rheumatoid arthritis of left hip
M05.359	Rheumatoid heart disease with rheumatoid arthritis of unspecified hip
M05.361	Rheumatoid heart disease with rheumatoid arthritis of right knee
M05.362	Rheumatoid heart disease with rheumatoid arthritis of left knee
M05.369	Rheumatoid heart disease with rheumatoid arthritis of unspecified knee
M05.371	Rheumatoid heart disease with rheumatoid arthritis of right ankle and foot
M05.372	Rheumatoid heart disease with rheumatoid arthritis of left ankle and foot
M05.379	Rheumatoid heart disease with rheumatoid arthritis of unspecified ankle and foot
M05.39	Rheumatoid heart disease with rheumatoid arthritis of multiple sites
M05.40	Rheumatoid myopathy with rheumatoid arthritis of unspecified site
M05.411	Rheumatoid myopathy with rheumatoid arthritis of right shoulder
M05.412	Rheumatoid myopathy with rheumatoid arthritis of left shoulder
M05.419	Rheumatoid myopathy with rheumatoid arthritis of unspecified shoulder
M05.421	Rheumatoid myopathy with rheumatoid arthritis of right elbow
M05.422	Rheumatoid myopathy with rheumatoid arthritis of left elbow
M05.429	Rheumatoid myopathy with rheumatoid arthritis of unspecified elbow
M05.431	Rheumatoid myopathy with rheumatoid arthritis of right wrist
M05.432	Rheumatoid myopathy with rheumatoid arthritis of left wrist
M05.439	Rheumatoid myopathy with rheumatoid arthritis of unspecified wrist
M05.441	Rheumatoid myopathy with rheumatoid arthritis of right hand
M05.442	Rheumatoid myopathy with rheumatoid arthritis of left hand
M05.449	Rheumatoid myopathy with rheumatoid arthritis of unspecified hand
M05.451	Rheumatoid myopathy with rheumatoid arthritis of right hip
M05.452	Rheumatoid myopathy with rheumatoid arthritis of left hip
M05.459	Rheumatoid myopathy with rheumatoid arthritis of unspecified hip
M05.461	Rheumatoid myopathy with rheumatoid arthritis of right knee
M05.462	Rheumatoid myopathy with rheumatoid arthritis of left knee
M05.469	Rheumatoid myopathy with rheumatoid arthritis of unspecified knee
M05.471	Rheumatoid myopathy with rheumatoid arthritis of right ankle and foot
M05.472	Rheumatoid myopathy with rheumatoid arthritis of left ankle and foot
M05.479	Rheumatoid myopathy with rheumatoid arthritis of unspecified ankle and foot
M05.49	Rheumatoid myopathy with rheumatoid arthritis of multiple sites
M05.50	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified site
M05.511	Rheumatoid polyneuropathy with rheumatoid arthritis of right shoulder
M05.512	Rheumatoid polyneuropathy with rheumatoid arthritis of left shoulder
M05.519	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified shoulder
M05.521	Rheumatoid polyneuropathy with rheumatoid arthritis of right elbow
M05.522	Rheumatoid polyneuropathy with rheumatoid arthritis of left elbow
M05.529	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified elbow
M05.531	Rheumatoid polyneuropathy with rheumatoid arthritis of right wrist
M05.532	Rheumatoid polyneuropathy with rheumatoid arthritis of left wrist
M05.539	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified wrist
M05.541	Rheumatoid polyneuropathy with rheumatoid arthritis of right hand
M05.542	Rheumatoid polyneuropathy with rheumatoid arthritis of left hand
M05.549	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified hand
M05.551	Rheumatoid polyneuropathy with rheumatoid arthritis of right hip

M05.552	Rheumatoid polyneuropathy with rheumatoid arthritis of left hip
M05.559	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified hip
M05.561	Rheumatoid polyneuropathy with rheumatoid arthritis of right knee
M05.562	Rheumatoid polyneuropathy with rheumatoid arthritis of left knee
M05.569	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified knee
M05.571	Rheumatoid polyneuropathy with rheumatoid arthritis of right ankle and foot
M05.572	Rheumatoid polyneuropathy with rheumatoid arthritis of left ankle and foot
M05.579	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified ankle and foot
M05.59	Rheumatoid polyneuropathy with rheumatoid arthritis of multiple sites
M05.60	Rheumatoid arthritis of unspecified site with involvement of other organs and systems
M05.611	Rheumatoid arthritis of right shoulder with involvement of other organs and systems
M05.612	Rheumatoid arthritis of left shoulder with involvement of other organs and systems
M05.619	Rheumatoid arthritis of unspecified shoulder with involvement of other organs and systems
M05.621	Rheumatoid arthritis of right elbow with involvement of other organs and systems
M05.622	Rheumatoid arthritis of left elbow with involvement of other organs and systems
M05.629	Rheumatoid arthritis of unspecified elbow with involvement of other organs and systems
M05.631	Rheumatoid arthritis of right wrist with involvement of other organs and systems
M05.632	Rheumatoid arthritis of left wrist with involvement of other organs and systems
M05.639	Rheumatoid arthritis of unspecified wrist with involvement of other organs and systems
M05.641	Rheumatoid arthritis of right hand with involvement of other organs and systems
M05.642	Rheumatoid arthritis of left hand with involvement of other organs and systems
M05.649	Rheumatoid arthritis of unspecified hand with involvement of other organs and systems
M05.651	Rheumatoid arthritis of right hip with involvement of other organs and systems
M05.652	Rheumatoid arthritis of left hip with involvement of other organs and systems
M05.659	Rheumatoid arthritis of unspecified hip with involvement of other organs and systems
M05.661	Rheumatoid arthritis of right knee with involvement of other organs and systems
M05.662	Rheumatoid arthritis of left knee with involvement of other organs and systems
M05.669	Rheumatoid arthritis of unspecified knee with involvement of other organs and systems
M05.671	Rheumatoid arthritis of right ankle and foot with involvement of other organs and systems
M05.672	Rheumatoid arthritis of left ankle and foot with involvement of other organs and systems
M05.679	Rheumatoid arthritis of unspecified ankle and foot with involvement of other organs and systems
M05.69	Rheumatoid arthritis of multiple sites with involvement of other organs and systems
M05.70	Rheumatoid arthritis with rheumatoid factor of unspecified site without organ or systems involvement
M05.711	Rheumatoid arthritis with rheumatoid factor of right shoulder without organ or systems involvement
M05.712	Rheumatoid arthritis with rheumatoid factor of left shoulder without organ or systems involvement
M05.719	Rheumatoid arthritis with rheumatoid factor of unspecified shoulder without organ or systems involvement
M05.721	Rheumatoid arthritis with rheumatoid factor of right elbow without organ or systems involvement
M05.722	Rheumatoid arthritis with rheumatoid factor of left elbow without organ or systems involvement
M05.729	Rheumatoid arthritis with rheumatoid factor of unspecified elbow without organ or systems involvement
M05.731	Rheumatoid arthritis with rheumatoid factor of right wrist without organ or systems involvement
M05.732	Rheumatoid arthritis with rheumatoid factor of left wrist without organ or systems involvement
M05.739	Rheumatoid arthritis with rheumatoid factor of unspecified wrist without organ or systems involvement
M05.741	Rheumatoid arthritis with rheumatoid factor of right hand without organ or systems involvement
M05.742	Rheumatoid arthritis with rheumatoid factor of left hand without organ or systems involvement

M05.749	Rheumatoid arthritis with rheumatoid factor of unspecified hand without organ or systems involvement
M05.751	Rheumatoid arthritis with rheumatoid factor of right hip without organ or systems involvement
M05.752	Rheumatoid arthritis with rheumatoid factor of left hip without organ or systems involvement
M05.759	Rheumatoid arthritis with rheumatoid factor of unspecified hip without organ or systems involvement
M05.761	Rheumatoid arthritis with rheumatoid factor of right knee without organ or systems involvement
M05.762	Rheumatoid arthritis with rheumatoid factor of left knee without organ or systems involvement
M05.769	Rheumatoid arthritis with rheumatoid factor of unspecified knee without organ or systems involvement
M05.771	Rheumatoid arthritis with rheumatoid factor of right ankle and foot without organ or systems involvement
M05.772	Rheumatoid arthritis with rheumatoid factor of left ankle and foot without organ or systems involvement
M05.779	Rheumatoid arthritis with rheumatoid factor of unspecified ankle and foot without organ or systems involvement
M05.79	Rheumatoid arthritis with rheumatoid factor of multiple sites without organ or systems involvement
M05.80	Other rheumatoid arthritis with rheumatoid factor of unspecified site
M05.811	Other rheumatoid arthritis with rheumatoid factor of right shoulder
M05.812	Other rheumatoid arthritis with rheumatoid factor of left shoulder
M05.819	Other rheumatoid arthritis with rheumatoid factor of unspecified shoulder
M05.821	Other rheumatoid arthritis with rheumatoid factor of right elbow
M05.822	Other rheumatoid arthritis with rheumatoid factor of left elbow
M05.829	Other rheumatoid arthritis with rheumatoid factor of unspecified elbow
M05.831	Other rheumatoid arthritis with rheumatoid factor of right wrist
M05.832	Other rheumatoid arthritis with rheumatoid factor of left wrist
M05.839	Other rheumatoid arthritis with rheumatoid factor of unspecified wrist
M05.841	Other rheumatoid arthritis with rheumatoid factor of right hand
M05.842	Other rheumatoid arthritis with rheumatoid factor of left hand
M05.849	Other rheumatoid arthritis with rheumatoid factor of unspecified hand
M05.851	Other rheumatoid arthritis with rheumatoid factor of right hip
M05.852	Other rheumatoid arthritis with rheumatoid factor of left hip
M05.859	Other rheumatoid arthritis with rheumatoid factor of unspecified hip
M05.861	Other rheumatoid arthritis with rheumatoid factor of right knee
M05.862	Other rheumatoid arthritis with rheumatoid factor of left knee
M05.869	Other rheumatoid arthritis with rheumatoid factor of unspecified knee
M05.871	Other rheumatoid arthritis with rheumatoid factor of right ankle and foot
M05.872	Other rheumatoid arthritis with rheumatoid factor of left ankle and foot
M05.879	Other rheumatoid arthritis with rheumatoid factor of unspecified ankle and foot
M05.89	Other rheumatoid arthritis with rheumatoid factor of multiple sites
M05.9	Rheumatoid arthritis with rheumatoid factor, unspecified
M06.00	Rheumatoid arthritis without rheumatoid factor, unspecified site
M06.011	Rheumatoid arthritis without rheumatoid factor, right shoulder
M06.012	Rheumatoid arthritis without rheumatoid factor, left shoulder
M06.019	Rheumatoid arthritis without rheumatoid factor, unspecified shoulder
M06.021	Rheumatoid arthritis without rheumatoid factor, right elbow
M06.022	Rheumatoid arthritis without rheumatoid factor, left elbow
M06.029	Rheumatoid arthritis without rheumatoid factor, unspecified elbow
M06.031	Rheumatoid arthritis without rheumatoid factor, right wrist

M06.032	Rheumatoid arthritis without rheumatoid factor, left wrist
M06.039	Rheumatoid arthritis without rheumatoid factor, unspecified wrist
M06.041	Rheumatoid arthritis without rheumatoid factor, right hand
M06.042	Rheumatoid arthritis without rheumatoid factor, left hand
M06.049	Rheumatoid arthritis without rheumatoid factor, unspecified hand
M06.051	Rheumatoid arthritis without rheumatoid factor, right hip
M06.052	Rheumatoid arthritis without rheumatoid factor, left hip
M06.059	Rheumatoid arthritis without rheumatoid factor, unspecified hip
M06.061	Rheumatoid arthritis without rheumatoid factor, right knee
M06.062	Rheumatoid arthritis without rheumatoid factor, left knee
M06.069	Rheumatoid arthritis without rheumatoid factor, unspecified knee
M06.071	Rheumatoid arthritis without rheumatoid factor, right ankle and foot
M06.072	Rheumatoid arthritis without rheumatoid factor, left ankle and foot
M06.079	Rheumatoid arthritis without rheumatoid factor, unspecified ankle and foot
M06.08	Rheumatoid arthritis without rheumatoid factor, vertebrae
M06.09	Rheumatoid arthritis without rheumatoid factor, multiple sites
M06.80	Other specified rheumatoid arthritis, unspecified site
M06.811	Other specified rheumatoid arthritis, right shoulder
M06.812	Other specified rheumatoid arthritis, left shoulder
M06.819	Other specified rheumatoid arthritis, unspecified shoulder
M06.821	Other specified rheumatoid arthritis, right elbow
M06.822	Other specified rheumatoid arthritis, left elbow
M06.829	Other specified rheumatoid arthritis, unspecified elbow
M06.831	Other specified rheumatoid arthritis, right wrist
M06.832	Other specified rheumatoid arthritis, left wrist
M06.839	Other specified rheumatoid arthritis, unspecified wrist
M06.841	Other specified rheumatoid arthritis, right hand
M06.842	Other specified rheumatoid arthritis, left hand
M06.849	Other specified rheumatoid arthritis, unspecified hand
M06.851	Other specified rheumatoid arthritis, right hip
M06.852	Other specified rheumatoid arthritis, left hip
M06.859	Other specified rheumatoid arthritis, unspecified hip
M06.861	Other specified rheumatoid arthritis, right knee
M06.862	Other specified rheumatoid arthritis, left knee
M06.869	Other specified rheumatoid arthritis, unspecified knee
M06.871	Other specified rheumatoid arthritis, right ankle and foot
M06.872	Other specified rheumatoid arthritis, left ankle and foot
M06.879	Other specified rheumatoid arthritis, unspecified ankle and foot
M06.88	Other specified rheumatoid arthritis, vertebrae
M06.89	Other specified rheumatoid arthritis, multiple sites
M06.9	Rheumatoid arthritis, unspecified
M31.1	Thrombotic microangiopathy
M31.30	Wegener’s granulomatosis without renal involvement
M31.31	Wegener’s granulomatosis with renal involvement
M31.7	Microscopic polyangiitis
R59.0	Localized enlarged lymph nodes

R59.1	Generalized enlarged lymph nodes
R59.9	Enlarged lymph nodes, unspecified
Z85.71	Personal history of Hodgkin lymphoma
Z85.72	Personal history of non-Hodgkin lymphomas
Z85.79	Personal history of other malignant neoplasms of lymphoid, hematopoietic and related tissues

Revision History

Company(ies)	DATE	REVISION
EmblemHealth & ConnectiCare	10/29/2024	Annual Review: Updated length of authorization and dosage chart. Added Medicare NCD/LCD Statement. Initial Criteria: Added: 'Patient is at least 18 years of age, unless otherwise specified; AND Patient has not received a live vaccine within 28 days prior to starting treatment and live vaccines will not be administered concurrently while on treatment; AND" For Oncology Indications: Added portion in parenthesis Patient is CD20-positive "(*excluding use for cGVHD, Hematopoietic Cell Transplantation, and Management of Immunotherapy-Related Toxicity)*;" AND Adult* Acute Lymphoblastic Leukemia (ALL) ‡: added Adult to the title. Removed the following to reword portions: "Induction/Consolidation Treatment Patient's disease is Philadelphia chromosome-negative (Ph-); AND Patient is at least 15 years of age; AND Used in combination with an anthracycline, cyclophosphamide and vincristine-based regimen Relapsed/Refractory Treatment Used as a component of MOpAD regimen (methotrexate, vincristine, pegaspargase, dexamethasone); AND Patient's disease is Philadelphia chromosome-negative (Ph-); OR" Updated as: " Patient has Philadelphia chromosome-positive (Ph+) disease; AND Used in combination with a tyrosine kinase inhibitor (TKI)-based regimen; AND Patient is <65 years of age without significant comorbidities; OR Used in combination with MOpAD (methotrexate, vincristine, pegaspargase, dexamethasone) for TKI-refractory disease; OR Patient has Philadelphia chromosome-negative (Ph-) disease; AND Used as a component of a multiagent chemotherapy **NCCN recommendations for Adult ALL may be applicable to adolescent and young adult (AYA) patients within the age range of 15-39 years."* CNS Cancer ‡ Changed the AND after the following statement to "OR": Patient has leptomeningeal metastases from lymphomas; OR Adult Hodgkin's lymphoma ‡ Added Adult to the title. Chronic lymphocytic leukemia/Small lymphocytic lymphoma (CLL/SLL) † added the following criteria: Used in combination with fludarabine and cyclophosphamide (FC) †; OR Patient has disease without del(17p)/TP53 mutation; AND A. Used as first-line therapy in combination with bendamustine (*excluding use in frail patients*); OR B. Used as subsequent therapy in combination with one of the following: Bendamustine (patients <65 years of age without significant comorbidities; excluding use in frail patients) Idelalisib, Lenalidomide, Venetoclax; OR Patient has disease with del(17p)/TP53 mutation; AND Used as first-line therapy in combination with high-dose methylprednisolone; OR Used as subsequent therapy in combination with one of the following: Alemtuzumab, High-dose methylprednisolone, Idelalisib, Lenalidomide, Venetoclax; OR Used as initial therapy for histologic (Richter's) transformation to diffuse large B-cell lymphoma; AND Used in combination

		with cyclophosphamide, doxorubicin, and vincristine-based regimens or as a component of OFAR (oxaliplatin, fludarabine, cytarabine, and rituximab) Non-Hodgkin's lymphomas (NHL) † Adult B-Cell Lymphomas † ‡ Φ: Added Adult B-Cell Lymphomas to the title. AIDS-related B-Cell Lymphoma ‡ HIV-Related B-Cell Lymphomas ‡: added the following criteria:" Disease is related to Burkitt lymphoma, diffuse large B-cell lymphoma (DLBCL), HHV8-positive DLBCL (not otherwise specified), or primary effusion lymphoma (PEL)" Burkitt Lymphoma ‡: Added the following criteria:" Used in combination with chemotherapy" Post-transplant lymphoproliferative disorder (PTLD) (B-Cell Type) ‡: Added "B-cell type" Added: Histologic Transformation of Indolent Lymphomas to Diffuse Large B-Cell Lymphoma ‡ Castleman's Disease ‡: Added the following criteria: " Patient has multicentric disease; OR Patient has unicentric disease; AND Used as second-line therapy for relapsed or refractory disease; OR Used for unresectable disease or symptomatic disease after incomplete resection" Added: Primary Cutaneous B-Cell Lymphomas ‡ and Pediatric Aggressive Mature B-Cell Lymphomas (Primary Mediastinal Large B-Cell " Added: :Lymphoma, Diffuse Large B-Cell Lymphoma, Burkitt Lymphoma, & Burkitt-like Lymphoma) † ‡ Φ A. Patient is at least 6 months of age*; AND B. Used in combination with chemotherapy **Pediatric Aggressive Mature B-Cell Lymphoma may be applicable to adolescent and young adult (AYA) patients older than 18 years of age and less than 39 years of age, who are treated in the pediatric oncology setting."* Added: Hairy Cell Leukemia ‡ and criteria; Histiocytic Neoplasms – Rosai-Dorfman Disease ‡ and criteria; Pediatric Hodgkin Lymphoma ‡ and criteria; Hematopoietic Cell Transplantation (HCT) ‡ and criteria Management of Immunotherapy-Related Toxicities ‡ Patient has been receiving therapy with an immune checkpoint inhibitor (updated examples) Changed the word "pulse" to "high" in the following statement "Patient has had limited to no improvement after 7 to 14 days on high-dose" Non-Oncology Indications: Patient is not on concurrent treatment with another CD20-directed therapy, TNF-inhibitor, IL-inhibitor, biologic response modifier or other non-biologic agent (e.g., apremilast, abrocitinib, tofacitinib, baricitinib, upadacitinib, deucravacitnib, etc.); AND Rheumatoid arthritis: added: Note: For patients already established on biologic therapy, trial and failure of oral DMARDs is not required Added: Thrombotic Thrombocytopenic Purpura (TTP) ‡and criteria and Multiple Sclerosis and criteria Added: Systemic Lupus Erythematosus (SLE) ‡ and criteria; added Lupus Nephritis and criteria; added: Myasthenia Gravis (unrelated to immunotherapy-related toxicity) ‡ and criteria; Added: Complications of Transplanted Solid Organ (kidney, liver, lung, heart, pancreas) in Adult and Pediatric* Patients and criteria; Added Neuromyelitis Optica Spectrum Disorder (NMOSD) ‡ and criteria Added: Antisynthetase Syndrome-Related Interstitial Lung Disease ‡ and criteria; added: Idiopathic Membranous Nephropathy ‡ and criteria; added: Pediatric Idiopathic Nephrotic Syndrome ‡ and criteria; Added: IgG4-Related Disease ‡ and criteria

		Renewal Criteria: Oncology Indications: Added: Adult Acute Lymphoblastic Leukemia (ALL) • Treatment response or stabilization of disease as indicated by CBC, bone marrow cytogenic analysis, QPCR, or FISH Pediatric B-Cell Acute Leukemia and Aggressive Mature B-Cell Lymphomas (induction or consolidation therapy) • Coverage may NOT be renewed Pediatric Hodgkin Lymphoma • Coverage may NOT be renewed Hematopoietic Cell Transplantation • Coverage may NOT be renewed Management of Immunotherapy-Related Toxicities • Coverage for use in the treatment of myositis/myasthenia gravis/encephalitis may NOT be renewed • Coverage for use in bullous dermatitis: Patient has not exceeded a maximum of 18 months of therapy (4 total doses) All Other Oncology Indications • Disease response with treatment as defined by stabilization of disease or decrease in size of tumor or tumor spread Thrombocytopenic purpura (ITP or Evans Syndrome)- added ITP or Evans syndrome to the title Added: "Thrombotic Thrombocytopenic Purpura (TTP) 1. Disease response as indicated by an increase in ADAMTS13 activity with a reduction in thrombotic risk Multiple Sclerosis (MS) 1.. Continuous monitoring of response to therapy indicates a beneficial response* [manifestations of MS disease activity include, but are not limited to, an increase in annualized relapse rate (ARR), development of new/worsening T2 hyperintensities or enhancing lesions on brain/spinal MRI, and progression of sustained impairment as evidenced by expanded disability status scale (EDSS), timed 25-foot walk (T25-FW), 9-hole peg test (9-HPT)] **Note: – Inadequate response, in those who have been adherent and receiving therapy for sufficient time to realize the full treatment effect, is defined as ≥ 1 relapse, ≥ 2 unequivocally new MRI-detected lesions, or increased disability on examination over a one-year period*" Added: "Systemic Lupus Erythematosus (SLE) -Adequate documentation of disease stability and/or improvement as indicated by one or more of the following when compared to pre-treatment baseline: Improvement in the SELENA-SLEDAI-2K; OR Reduction of baseline BILAG-2004 from A to B or from B to C/D, and no BILAG-2004 worsening in other organ systems, as defined by ≥2 new BILAG-2004 B; OR No worsening (<0.30 points increase) in Physician's Global Assessment (PGA) score; OR Seroconverted (negative) Lupus Nephritis Coverage may only be renewed in patients experiencing a disease relapse (e.g., increased serum creatinine, increase in protein urine excretion, decrease in eGFR, etc.) Myasthenia Gravis (unrelated to immunotherapy-related toxicity) 1. Disease response as indicated by a decrease in the daily dose of corticosteroids and/or an improvement in signs and symptoms compared to baseline. Complications of Transplanted Solid Organ (kidney, liver, lung, heart, pancreas) 1. Coverage may NOT be renewed. NMOSD 1. Disease response as indicated by stabilization/improvement in any of the following: Decrease in acute relapses or improvement of stability, Reduced

		hospitalizations, Reduction/discontinuation in plasma exchange treatments, Reduction/discontinuation of corticosteroids without relapse" Antisynthetase Syndrome-Related Interstitial Lung Disease Disease response as indicated by stabilization/improvement in any of the following: Reduction or stabilization of glucocorticoid use from baseline, Improvement or stabilization of pulmonary function testing (i.e., improvement defined as >10% increase in FVC%, TLC%, or DLCO%; stabilization defined as < 10% decrease in FVC%, TLC%, or DLCO%), Improvement or stabilization of chest CT score (improvement defined as >10% decrease in CT score, stabilization defined as a < 10% increase in CT score) Idiopathic Membranous Nephropathy Patient experienced beneficial disease response with improvement in symptoms and/or other objective measures compared to baseline (e.g., reduction in proteinuria, increase and/or normalization of serum albumin, improvement/stability of serum creatinine and/or eGFR, decrease in anti-PLA2R antibody levels, etc.); OR Patient has resistant disease following first-line therapy with rituximab; AND Patient has stable eGFR; AND Will be used in combination with a calcineurin inhibitor if previously treated with rituximab alone in the first-line setting Pediatric Idiopathic Nephrotic Syndrome ‡ Patient previously achieved beneficial disease response from the prior course of therapy; AND Patient is experiencing signs and symptoms of recurrent active disease necessitating additional doses (e.g., recurrence of nephrotic-range proteinuria with a dipstick > 3+ [>300 mg/dL] for 3 consecutive days OR urinary protein creatinine ratio [UPCR] ≥200 mg/mmol [≥2 mg/mg] on a spot urine sample on 3 consecutive days, with or without reappearance of edema in a child who had previously achieved complete remission) IgG4-Related Disease ‡ Patient experienced beneficial disease response with improvement in involved organ-related symptoms and/or other objective measures compared to baseline (e.g. improvement in the IgG4-RD Responder Index score of > 2 points, improvement in the physician's global assessment [PGA], reduction in glucocorticoid or other immunosuppressive use, reduction of disease flares, reduction in serum IgG4 level, etc.); AND Patient meets one of the following: Ongoing maintenance therapy is required due to patient having a high-risk of relapse Patient is experiencing signs and symptoms of relapsed active disease necessitating an additional course of therapy Updated dosage and administration chart Removed limitations and exclusions
EmblemHealth & ConnectiCare	8/18/2023	Annual Review: <u>Length of Authorization:</u> Added "Adult Acute Lymphoblastic Leukemia (ALL) may be renewed for a maximum of 18 doses. o Mantle Cell Lymphoma may be renewed until disease progression or intolerable toxicity. o Hairy Cell Leukemia may NOT be renewed. o Induction/Consolidation of Pediatric B-Cell Acute Leukemia and Aggressive Mature B-Cell Lymphomas may NOT be renewed. o Pediatric Hodgkin Lymphoma may NOT be renewed. • Management of Immunotherapy-Related Toxicities: o Myositis/Myasthenia Gravis/Encephalitis may NOT be renewed. o Bullous Dermatitis may be renewed for a maximum of 18 months (4 total doses).

		• Chronic Graft-Versus-Host Disease (cGVHD) may NOT be renewed. • Hematopoietic Cell Transplantation may NOT be renewed. • Lupus Nephritis may be renewed ONLY in patients experiencing a disease relapse. • Complications of Transplanted Solid Organ may NOT be renewed. • Removed "Acute lymphoblastic leukemia (ALL) may not be renewed." <u>Updated Dosing Limits Chart</u> Added: <u>Pediatric Mature B-Cell Acute Leukemia † Indication and criteria</u> <u>Acute Lymphoblastic Leukemia (ALL) ‡ Initial Criteria:</u> Removed "Patient's disease is Philadelphia chromosome-positive (Ph+) and refractory to tyrosine kinase inhibitors (e.g. omacetaxine, imatinib, bosutinib, ponatinib, nilotinib, etc.)" <u>CNS Cancer ‡</u> Initial Criteria Removed: "Rituximab will be administered intrathecally; OR: Also Removed "Patient will receive in combination with a methotrexate-containing regimen as a component of induction therapy and/or consolidation therapy with a complete response to induction therapy; OR ii. Patient has relapsed or refractory disease and will receive rituximab as a single agent, or in combination with temozolomide, lenalidomide or high-dose methotrexate" Added "Used for induction therapy; AND A. Used as a single agent OR in combination with a methotrexate-containing regimen, temozolomide, or lenalidomide¥; OR B. Patient has CSF positive or spinal MRI positive disease§; OR iii. Used for consolidation (monthly maintenance) therapy; AND A. Used as continuation of induction regimen in patients with complete response or complete response unconfirmed (CRu) to induction therapy; AND 1.)Used as a single agent§; OR 2.)Used on combination with high-dose methotrexate¥; OR iv. Used for relapsed or refractory disease; AND A. Used as a single agent OR in combination with systemic therapy in patients with prior whole brain radiation therapy§; AND 1.)Patient has CSF positive or spinal MRI positive disease; OR B. Used as a single agent OR in combination with temozolomide, lenalidomide, or highdose methotrexate¥ *§ For intrathecal administration ONLY; ¥ For intravenous administration ONLY"* <u>Non-Hodgkin's lymphomas (NHL) † Initial Criteria</u>: Added "<u>Extranodal Marginal Zone Lymphoma (EMZL) of the Stomach & Nongastric Sites (Noncutaneous)</u> ‡ 11. High-Grade B-Cell Lymphomas ‡" 12. Removed "Gastric & Non-Gastric MALT Lymphoma ‡ 13. Hairy Cell Leukemia ‡ a. Used for relapsed or refractory disease" b. "Patient has had solid organ transplant or allogeneic hematopoietic stem cell transplantation 14. Primary Cutaneous B-Cell Lymphomas ‡

		a. Used for generalized (skin only), marginal zone or follicle center disease" <u>Management of Immunotherapy-Related Toxicities Initial Criteria:</u> Added "cemiplimab" "ipilimumab, dostarlimab, nivolumab/relatlimab-rmbw " to the Statement: Patient has been receiving therapy with an immune checkpoint inhibitor (e.g. cemiplimab ,nivolumab, pembrolizumab, atezolizumab, avelumab, durvalumab, ipilimumab, dostarlimab, nivolumab/relatlimab-rmbw etc.); AND Removed "non-viral" from the following statement: Patient has non-viral encephalitis related to their immunotherapy; AND Removed "Patient is refractory to methylprednisolone and/or IV immunoglobulin (IVIG)" Added "Patient has had limited to no improvement after 7 to 14 days on pulse-dose corticosteroids with or without intravenous immunoglobulin (IVIG); OR 2. Patient has bullous dermatitis related to immunotherapy; AND ii. Used as additional therapy for moderate (G2), severe (G3) or life-threatening (G4) disease; OR 3. Patient has moderate, severe, or life-threatening steroid-refractory myositis (proximal muscle weakness, neck flexor weakness, with or without myalgias) related to immunotherapy; AND ii. Used for significant dysphagia, life-threatening situations, or cases refractory to corticosteroids; OR 4. Patient has myasthenia gravis related to immunotherapy; AND ii. Used as additional therapy for severe (G3-4) disease that is refractory to plasmapheresis or IVIG" <u>Pemphigus vulgaris Initial Criteria:</u> Added "Positive direct immunofluorescence (DIF) microscopy result OR " Removed "or indirect" from the Statement: Presence of autoantibodies as detected by direct or indirect immunofluorescence" and added " or enzyme-linked immunosorbent assay (ELISA); AND" <u>Thrombocytopenic purpura ‡ Initial Criteria:</u> Removed "Congenital and hereditary thrombocytopenic purpura D. Thrombotic thrombocytopenic purpura in patients with ADAMTS13-deficiency" Added "Patient has previously failed or has a contraindication or intolerance to therapy with corticosteroids; AND 3. Patient is at increased risk for bleeding as indicated by platelet count (within the previous 28 days) less than 30 × 109/L (30,000/mm^3)" <u>Chronic graft-versus-host disease (cGVHD) ‡ Initial Criteria:</u> Removed "Patient has failed one or more previous lines of systemic therapy for the treatment of cGVHD (e.g., corticosteroids or immunosuppressants such as cyclosporine); AND" Added "Used as additional therapy in combination with corticosteroids; AND 5. Patient has no response (e.g., steroid-refractory disease) to first-line therapy options; AND" <u>Renewal Criteria:</u> Removed "Thrombotic thrombocytopenic purpura (TTP) Disease response as indicated by an increase in ADAMTS13 activity with a reduction in thrombosis risk" <u>Granulomatosis with Polyangiitis (GPA) (Wegener's granulomatosis) and Microscopic polyangiitis (MPA)</u>

		Added "Decreased frequency in the occurrence of major relapses (defined by the reappearance of clinical and/or laboratory signs of vasculitis activity that could lead to organ failure or damage, or could be life threatening)" Chronic graft-versus-host disease (cGVHD) Removed "Disease response as indicated by improvement in patient-reported symptoms or clinician assessments (e.g., manifestations of disease to the skin, oral cavity, musculoskeletal system, etc.)" Added "Coverage may NOT be renewed" Updated Dosing Chart Updated Truxima Indications: Granulomatosis with Polyangiitis (GPA) (Wegener's Granulomatosis) and Microscopic Polyangiitis (MPA) Removed "In combination with glucocorticoids, is indicated for the treatment of adult patients with Granulomatosis with Polyangiitis (GPA) (Wegener's Granulomatosis) and Microscopic Polyangiitis (MPA)." Added "In combination with glucocorticoids, is indicated for the treatment of adult and pediatric patients 2 years of age and older with Granulomatosis with Polyangiitis (GPA) (Wegener's Granulomatosis) and Microscopic Polyangiitis (MPA)" Removed "Pemphigus Vulgaris (PV) 1. Treatment of adult patients with moderate to severe pemphigus vulgaris." Added NDCs: 00069-0238-xx 00069-0249-xx
EmblemHealth & ConnectiCare	12/30/2022	Added FDA approved indication of Rheumatoid Arthritis to Ruxience and Riabni
EmblemHealth & ConnectiCare	09/16/2021	Confirmed non-preferred status of Riabni; Added Q-Code (Q5123) Injection, rituximab-arrx, biosimilar, (riabni), 10 mg
EmblemHealth & ConnectiCare	02/02/2021	Added Riabni biosimilar and its indications per FDA label
EmblemHealth & ConnectiCare	12/19/2020	Clarifications: • Step therapy will apply to NEW starts only • NCCN-supported use (with 1 or 2A recommendation) will be covered Renewal criteria updated: • Removed: "Patient continues to meet criteria identified above" Added coverage: "Continuation of documented current and/or successful therapy with a non-preferred agent (Rituxan)"
EmblemHealth & ConnectiCare	11/2/2020	Effective 01/01/2021 Member must fail trial of Ruxience AND Truxima, prior to using Rituxan. (Medicare members are subject to this step therapy).
EmblemHealth & ConnectiCare	06/11/2020	Added Q-Code (Q5119): Injection, rituximab-pvvr, biosimilar, 10 mg (Ruxience). Effective Date: 07/01/2020
EmblemHealth & ConnectiCare	05/07/2020	Added two newly approved indications and criteria for Truxima per FDA Label: • Rheumatoid Arthritis (RA) Granulomatosis with Polyangiitis (GPA) (Wegener's Granulomatosis) and Microscopic Polyangiitis (MPA)

EmblemHealth & ConnectiCare	03/31/2020	Added to the Initial Approval Criteria: Effective 07/01/2020, Ruxience and Truxima are the preferred agents for Commercial and Medicaid members. Failed trial of Ruxience AND Truxima for FDA approved indications prior to using Rituxan (Only Commercial and Medicaid members are subject to this step therapy).
EmblemHealth & ConnectiCare	03/31/2020	Ruxience and Truxima indications updated per FDA label.
EmblemHealth & ConnectiCare	01/01/2020	Added Ruxience biosimilar and its indications per FDA label
EmblemHealth & ConnectiCare	10/17/2019	Updated Truxima's indications for NHL and CLL, updated dosage and administration for CLL (initial therapy), updated Non-Hodgkin's Lymphoma dosing criteria, updated dosing criteria for Granulomatosis with Polyangiitis (GPA), Wegener's Granulomatosis and Microscopic Polyangiitis (MPA), updated dosing criteria for cGVHD
EmblemHealth & ConnectiCare	07/01/2019	Added Q5115 Truxima biosimilar
EmblemHealth & ConnectiCare	12/03/2018	Added J9312 and removed J9310 from Applicable Procedure Codes.

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