

Rituximab:

Riabni®, Rituxan®, Ruxience®, Truxima® (Intravenous/Intrathecal/Intraventricular)

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I. Length of Authorization 1-5,23-25,34,44,62,80,94-98,102-104,108,115-118,128-130,133-138,152,154,169-173,191-198

Oncology Indications

- Initial: Prior authorization validity for oncology indications will be provided initially for 6 months (180 days).
- Renewal: Prior authorization validity for oncology indications may be renewed every 6 months (180 days) for up to a maximum of 2 years, unless otherwise specified.
 - Prior authorization validity may NOT be renewed for the following:
 - ❖ Pediatric B-Cell Acute Leukemia (induction/consolidation)
 - ❖ Pediatric Aggressive Mature B-Cell Lymphomas (induction/consolidation)
 - ❖ Pediatric Hodgkin Lymphoma
 - ❖ Pediatric Aggressive Mature B-Cell Lymphomas (Post-Transplant Lymphoproliferative Disorders - treatment and prevention)
 - ❖ KSHV-Associated Inflammatory Cytokine Syndrome
 - ❖ Management of Immune Checkpoint Inhibitor-Related Toxicities (excluding Bullous Dermatitis/Bullous Pemphigoid)
 - ❖ Hematopoietic Cell Transplantation (HCT)
 - ❖ Transplant Associated-Thrombotic Microangiopathy (TA-TMA)
 - ❖ Adult B-Cell Lymphomas - Post-Transplant Lymphoproliferative Disorder (prevention of Epstein-Barr virus (EBV)-related PTLTD)
 - Adult B-Cell Lymphomas (regimen containing brentuximab and lenalidomide or treatment of Mantle Cell Lymphoma): Prior authorization validity may be renewed until disease progression or intolerable toxicity.

- Adult Acute Lymphoblastic Leukemia (ALL): Prior authorization validity may be renewed for a maximum of 18 doses.
- Hairy Cell Leukemia and Chronic Graft-Versus-Host Disease (cGVHD): Prior authorization validity may be renewed for up to a maximum of 12 doses.
- Management of Immune Checkpoint Inhibitor-Related Toxicities (Bullous Dermatitis/Bullous Pemphigoid): Prior authorization validity may be renewed for a maximum of 18 months (4 total doses).

Non-Oncology Indications

- Initial: Prior authorization validity for non-oncology indications will be provided initially for 6 months (180 days), unless otherwise specified.
 - Prior authorization validity will be provided initially for 12 months (365 days) for the following:
 - ❖ Pemphigus Vulgaris
 - ❖ Systemic Lupus Erythematosus
 - ❖ Lupus Nephritis
- Renewal: Prior authorization validity for non-oncology indications may be renewed every 6 months (180 days) thereafter, unless otherwise specified.
 - Prior authorization validity may NOT be renewed for the following:
 - ❖ Pediatric Idiopathic Nephrotic Syndrome (*May be renewed ONLY in members experiencing a disease relapse*)
 - ❖ Complications of Transplanted Solid Organ
 - ❖ Wiskott-Aldrich Syndrome
 - Prior authorization validity may be renewed every 12 months (365 days) for the following:
 - ❖ Systemic Lupus Erythematosus
 - ❖ Lupus Nephritis (*May be renewed ONLY in members experiencing a disease relapse*)

II. Dosing Limits

Max Units (per dose and over time) [HCPCS Unit]:

Oncology Indications
<u>Chronic Lymphocytic Leukemia (CLL)/Small Lymphocytic Leukemia (SLL):</u> • Initial therapy: 100 billable units x 1 dose, then 130 billable units every 7 days x 11 doses • Renewal therapy: 130 billable units every 8 weeks
<u>ALL</u> • 100 billable units twice weekly
<u>Waldenström Macroglobulinemia/Lymphoplasmacytic Lymphoma</u> • Initial therapy: 100 billable units every 7 days x 12 doses • Renewal therapy: 400 billable units every 6 months
<u>B-Cell Lymphoma</u> • Initial therapy: 100 billable units every 7 days x 8 doses in a 6 month period • Renewal therapy: 800 billable units every 168 days
<u>CNS Cancers</u>

- Initial therapy: 190 billable units every 7 days x 8 doses - Renewal therapy: 100 billable units every 28 days
<u>Hairy Cell Leukemia</u> 100 billable units every 7 days x 8 doses, 100 billable units every 14 days x 8 doses, then 100 billable units every 28 days x 4 doses
<u>KSHV-Associated Inflammatory Cytokine Syndrome</u> 100 billable units every 21 days for 6 doses
<u>Histiocytic Neoplasms – Rosai-Dorfman Disease</u> 780 billable units every 6 months
<u>Chronic Graft-Versus-Host Disease (cGVHD)</u> 100 billable units every 7 days
<u>Hematopoietic Cell Transplantation</u> - Initial dose: 100 billable units x 1 dose before transplant - Subsequent doses: 250 billable units every 7 days x 3 doses after transplant
<u>Immunotherapy-Related Toxicities</u> <i>Bullous Dermatitis/Bullous Pemphigoid</i>: 100 billable units every 14 days x 2 doses, then 50 billable units at months 12 & 18 <i>Myositis, Encephalitis, Hemolytic Anemia, Thrombocytopenia, Acute Kidney Injury</i>: 100 billable units every 7 days x 4 doses <i>Myasthenia Gravis</i>: 130 billable units every 7 days x 4 doses
<u>Transplant Associated-Thrombotic Microangiopathy (TA-TMA)</u> 100 billable units every 7 days x 4 doses
<u>All other oncology indications (Castleman Disease, Primary Cutaneous B-Cell Lymphomas, or HL):</u> - Initial therapy: 100 billable units every 7 days x 8 doses in a 6 month period - Renewal therapy: 400 billable units every 6 months
Non-Oncology Indications
<u>Rheumatoid Arthritis (RA):</u> 200 billable units every 24 weeks
<u>Multiple Sclerosis (MS):</u> 200 billable units every 6 months
<u>Wiskott-Aldrich Syndrome:</u> 100 billable units x 1 dose
<u>Pemphigus Vulgaris (PV):</u> - Initiation: 100 billable units every 7 days x 4 doses in a 12 month period - Maintenance: 50 billable units every 6 months
<u>GPA(WG)/MPA:</u> - Induction: 100 billable units every 7 days x 4 doses - Initial Maintenance: 100 billable units every 14 days x 2 doses - Subsequent Maintenance: 100 billable units every 6 months
<u>Thrombocytopenic Purpura or Thrombotic Thrombocytopenic Purpura, Complications of Transplanted Solid Organ, IgG4-Related Disease:</u> 100 billable units every 7 days x 4 doses
<u>All other non-oncology indications (AIHA, SLE or LN, Myasthenia Gravis, NMOSD, Antisynthetase Syndrome-Related Interstitial Lung Disease, Idiopathic Membranous Nephropathy, Pediatric Idiopathic Nephrotic Syndrome):</u> 400 billable units every 6 months

III. Initial Approval Criteria ¹⁻⁴

Prior authorization validity is provided in the following conditions:

For Commercial Members ONLY

- Patient must have a contraindication, intolerance, or failure to Ruxience, Truxima, AND Riabni prior to the consideration of another rituximab product; **AND**

For Medicaid Members ONLY

- Patient must have a contraindication, intolerance, or failure of Ruxience prior to the consideration of another rituximab product; **AND**
- Member is at least 18 years of age, unless otherwise specified; **AND**

Universal Criteria ¹⁻⁴

- Member does not have a severe, active infection; **AND**
- Member has been screened for the presence of hepatitis B (HBV) infection (i.e., HBsAg and anti-HBc) prior to initiating therapy and members with evidence of current or prior HBV infection will be monitored for HBV reactivation during treatment; **AND**
- Provider will confirm that member 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 ¹⁻⁵

- Member is CD20 antigen expression positive (*excluding use for cGVHD, Hematopoietic Cell Transplantation, Transplant Associated-Thrombotic Microangiopathy, KSHV-Associated Inflammatory Cytokine Syndrome, and Management of Immune Checkpoint Inhibitor-Related Toxicity*); **AND**

Pediatric Mature B-Cell Acute Leukemia (B-AL) † ¹

- Member is at least 6 months of age; **AND**
- Used in combination with chemotherapy for previously untreated disease

Adult* Acute Lymphoblastic Leukemia (ALL) ‡ ^{5,93}

- Member has Philadelphia chromosome-positive (Ph+) disease; **AND**
 - Used in combination with MOPAD (methotrexate, vincristine, pegaspargase, dexamethasone) for TKI-refractory disease; **OR**
- Member 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) members within the age range of 15-39 years.*

Central Nervous System (CNS) Cancers ‡ ⁵

- Member has leptomeningeal metastases from lymphomas; **AND**
 - Used as intra-cerebrospinal fluid (CSF) therapy; **OR**
- Member has primary CNS lymphoma (including primary vitreoretinal lymphoma/PCNSL ocular variant without other CNS involvement); **AND**

- Used as induction OR for relapsed or refractory disease; **AND**
 - Used as intravenous systemic therapy as a single agent or in combination with one of the following:
 - Temozolomide
 - Lenalidomide
 - Methotrexate-containing regimen; **OR**
 - Used as intra-CSF therapy if CSF positive or spinal MRI positive disease; **OR**
- Used as consolidation (monthly maintenance) therapy as continuation of induction regimen in members with complete response or complete response unconfirmed (CRu) to induction therapy; **AND**
 - Used as a single agent or in combination with high-dose methotrexate

Adult Hodgkin Lymphoma ‡ ⁵

- Member has nodular lymphocyte-predominant disease

Chronic Lymphocytic Leukemia/Small Lymphocytic Lymphoma (CLL/SLL) † ‡ Φ ¹⁻⁵

- Used in combination with fludarabine and cyclophosphamide (FC) †; **OR**
- Member has disease without del(17p)/TP53 mutation; **AND**
 - Used as first-line therapy in combination with bendamustine (*excluding use in frail members*); **AND**
 - Covalent Bruton Tyrosine Kinase inhibitor and B-cell Lymphoma-2 inhibitor are not available or not feasible; **OR**
 - Used as subsequent therapy in combination with one of the following:
 - Bendamustine (*members <65 years of age without significant comorbidities; excluding use in frail members*)
 - Idelalisib
 - Lenalidomide
 - Venetoclax; **OR**
- Member 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) transformation; **AND**

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

Waldenström Macroglobulinemia/Lymphoplasmacytic Lymphoma ‡⁵

Adult B-Cell Lymphomas † ‡ Φ^{1-5,44,208,209} including, but not limited to, the following:

- HIV-Related B-Cell Lymphomas ‡
 - Disease is related to Burkitt lymphoma, diffuse large B-cell lymphoma (DLBCL), HHV8-positive DLBCL (not otherwise specified), primary effusion lymphoma (PEL), or plasmablastic lymphoma
- Burkitt Lymphoma ‡
- Diffuse Large B-Cell Lymphoma † ‡ Φ
- Low-Grade or Follicular Lymphoma † ‡ Φ
- Extranodal Marginal Zone Lymphoma (EMZL) of the Stomach & Nongastric Sites (Noncutaneous) ‡
- Nodal & Splenic Marginal Zone Lymphoma ‡
- High-Grade B-Cell Lymphomas ‡
- Mantle Cell Lymphoma ‡
- Histologic Transformation of Indolent Lymphomas to Diffuse Large B-Cell Lymphoma ‡
- Post-Transplant Lymphoproliferative Disorders (PTLD) ‡
 - Used for B-Cell type PTLD; **OR**
 - Used for prevention of Epstein-Barr virus (EBV)-related PTLD; **AND**
 - Member has EBV reactivation

Castleman Disease ‡⁵

- Member has multicentric disease; **OR**
- Member has unresectable or incomplete resection of unicentric disease; **AND**
 - Used as first-line therapy; **OR**
 - Used as alternate first-line therapy; **OR**
 - Used as subsequent therapy for relapsed, refractory, or progressive disease

Primary Cutaneous B-Cell Lymphomas ‡⁵

Pediatric Aggressive Mature B-Cell Lymphomas † ‡ Φ^{1,5,50,121,208,209}

- Member is at least 6 months of age*; **AND**
 - Used in combination with chemotherapy for one of the following:
 - Primary Mediastinal Large B-Cell Lymphoma

- Diffuse Large B-Cell Lymphoma
- Burkitt Lymphoma
- Burkitt-like Lymphoma; **OR**
- Used for treatment of Post-Transplant Lymphoproliferative Disorders (PTLD); **OR**
- Used for prevention of Epstein-Barr virus (EBV)-related PTLD; **AND**
 - Member has EBV reactivation

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

Hairy Cell Leukemia ‡⁵

- Used as a single agent; **AND**
 - Used for incomplete hematologic recovery or relapsed disease in members unable to receive purine analogs (i.e., cladribine or pentostatin); **OR**
- Used in combination with cladribine; **OR**
- Used in combination with pentostatin; **AND**
 - Used for incomplete hematologic recovery or relapsed disease; **OR**
- Used in combination with vemurafenib; **AND**
 - Used as initial therapy OR for relapse ≥ 2 years after initial therapy in members who are not candidates for purine analogs including members who are frail and those with active infection; **OR**
 - Used for incomplete hematologic recovery or relapsed disease; **OR**
 - Used for progression after therapy for relapsed or refractory disease; **OR**
- Used in combination with venetoclax; **AND**
 - Used for progression after therapy for relapsed or refractory disease; **AND**
 - Member had disease resistance to BRAF inhibitor therapy

Histiocytic Neoplasms – Rosai-Dorfman Disease ‡⁵

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

Pediatric Hodgkin Lymphoma ‡^{5,128}

- Member is ≤ 18 years of age*; **AND**
- Member has nodular lymphocyte-predominant disease; **AND**
- Used in combination with CVbP (cyclophosphamide, vinblastine, prednisolone); **AND**

- 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) members up to the age of 39 years.*

Kaposi Sarcoma - Kaposi-sarcoma associated herpesvirus (KSHV)-Associated Inflammatory Cytokine Syndrome (KICS) ‡⁵

- Used as systemic therapy in combination with liposomal doxorubicin; **OR**
- Used as add-on therapy to alternative Kaposi Sarcoma (KS)-directed systemic therapies

Chronic Graft-Versus-Host Disease (cGVHD) ‡^{5,22-25}

- Member is post-allogeneic hematopoietic cell transplant (generally 3 or more months); **AND**
- Used as additional therapy in combination with systemic corticosteroids; **AND**
- Member has no response (e.g., steroid-refractory disease) to first-line therapy options

Hematopoietic Cell Transplantation (HCT) ‡⁵

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

Transplant Associated-Thrombotic Microangiopathy (TA-TMA) in Adult and Pediatric Members* ‡^{198,199}

- Member is post-hematopoietic stem cell transplant (HSCT); **AND**
- Alternative diagnoses have been excluded [e.g., thrombotic thrombocytopenia purpura, Coombs-positive hemolytic anemia, medication toxicities, infectious complications, disseminated intravascular coagulation, graft-versus-host disease (GVHD), etc.]; **AND**
- Member has high-risk disease defined as any of the following:
 - Random urine protein to creatinine ratio (rUPCR) ≥ 1
 - Elevated serum sC5b-9 > upper limit of normal (ULN)
 - LDH $\geq 2 \times$ ULN
 - Concurrent grade II-IV acute GVHD
 - Concurrent infections (bacterial or viral)
 - Clinical evidence of TA-TMA related organ dysfunction (*excluding KDIGO stage I acute kidney injury, defined as serum creatinine 1.5-1.9-time baseline*)

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

Management of Immune Checkpoint Inhibitor-Related Toxicities ‡^{5,62}

- Member has been receiving therapy with an immune checkpoint inhibitor; **AND**
 - Member has encephalitis related to immunotherapy; **AND**
 - Member is autoimmune-encephalopathy-antibody positive; **OR**

- Member has had limited to no improvement after 7 to 14 days on high-dose corticosteroids with or without intravenous immunoglobulin (IVIG); **OR**
- Member has bullous dermatitis related to immunotherapy; **AND**
 - Used as additional therapy for severe (G3) or life-threatening (G4) disease; **OR**
- Member has bullous pemphigoid related to immunotherapy, confirmed by biopsy or serology; **AND**
 - Used as additional therapy for moderate (G2) disease; **OR**
- Member has hemolytic anemia with hemolysis related to immunotherapy; **AND**
 - Used as additional therapy for G3 disease if no response to corticosteroids after 5-7 days; **OR**
 - Used as additional therapy for G4 disease if no response to corticosteroids after 3-5 days; **OR**
- Member has thrombocytopenia related to immunotherapy; **AND**
 - Used as additional therapy for G3 or G4 disease if no response to corticosteroids after 1-2 weeks; **OR**
- Member has stage 3 acute kidney injury/elevated serum creatinine related to immunotherapy; **AND**
 - Toxicity remains >stage 2 after 4-6 weeks of corticosteroids; **OR**
 - Creatinine increases during corticosteroid taper (or once off corticosteroids); **OR**
- Member has mild, moderate, severe, or life-threatening steroid-refractory myositis (proximal muscle weakness, neck flexor weakness, dysphagia, with or without myalgias) related to immunotherapy; **AND**
 - Used for significant dysphagia, life-threatening situations, or cases refractory to corticosteroids; **OR**
- Member has myasthenia gravis related to immunotherapy; **AND**
 - Used as additional therapy for severe (G3-4) disease that is refractory to plasmapheresis or IVIG

Non-Oncology Indications

- Member is not on concurrent treatment with another CD20-directed therapy, biologic agent, or targeted synthetic therapies; **AND**

Rheumatoid Arthritis (RA) † 1-4,46-49,112,113,191

- Physician has assessed baseline disease severity utilizing an objective measure/tool; **AND**
- Documented moderate to severe active disease; **AND**
- Used in combination with methotrexate unless the member has a contraindication or intolerance; **AND**

- Member tried and failed at least a 3-month trial with ONE conventional synthetic disease modifying anti-rheumatic drug (csDMARD) (e.g., methotrexate, azathioprine, auranofin, hydroxychloroquine, penicillamine, sulfasalazine, leflunomide, etc.); **OR**
- Member is already established on biologic or targeted synthetic therapy for the treatment of RA; **AND**
- Previous failure with one or more TNF antagonists; **AND**
- Member has not had treatment with rituximab in the previous 4 months

Pemphigus Vulgaris † Φ^{1,10,11,35,36,38,61,80,114,139}

- Member has a diagnosis of pemphigus vulgaris as determined by the following:
 - Member has one or more of the following clinical features:
 - Appearance of lesions, erosions and/or blisters
 - Nikolsky sign (induction of blistering via mechanical pressure at the edge of a blister or on normal skin)
 - Characteristic scarring and lesion distribution; **AND**
 - Histopathologic confirmation by skin/mucous membrane biopsy; **AND**
 - Positive direct immunofluorescence (DIF) microscopy result OR the presence of autoantibodies as detected by indirect immunofluorescence (IIF) or enzyme-linked immunosorbent assay (ELISA); **AND**
- Member has moderate to severe disease as assessed utilizing an objective measure/tool (e.g., Pemphigus Disease Activity Index [PDAI], Pemphigus Severity Score [PSS], Autoimmune Bullous Skin Disorder Intensity Score [ABSIS], etc.); **AND**
- Used in combination with glucocorticoids (e.g., prednisone, prednisolone, etc.); **AND**
- 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) † ‡ Φ^{1-4,125}

- Member is at least 2 years of age; **AND**
 - Used as induction therapy; **AND**
 - Used in combination with glucocorticoids (e.g., prednisone, methylprednisolone, etc.); **OR**
 - Used as maintenance therapy; **AND**
 - Used with or without glucocorticoids (e.g., prednisone, methylprednisolone, etc.)

Thrombocytopenic Purpura ‡^{6-9,63,127}

- Diagnosis includes one of the following:
 - Primary thrombocytopenia or idiopathic (immune) thrombocytopenia purpura (ITP)
 - Evans syndrome; **AND**

- Member has previously failed or has a contraindication or intolerance to therapy with corticosteroids; **AND**
- Member is at increased risk for bleeding as indicated by platelet count (within the previous 28 days) of less than $30 \times 10^9/L$ (30,000/mm³)

Thrombotic Thrombocytopenic Purpura (TTP) ‡ ^{16-18,20,21,196}

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

Multiple Sclerosis (MS) ‡ ^{144,148,203}

- Member must have a confirmed diagnosis of multiple sclerosis (MS) as documented by laboratory report (i.e., MRI); **AND**
- Member 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) ‡ ²⁶⁻³²

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

Systemic Lupus Erythematosus (SLE) ‡ ^{153-155,158-163,169}

- Member has a diagnosis of active systemic lupus erythematosus (SLE) WITHOUT active lupus nephritis (LN); **AND**
 - BOTH of the following:
 - ONE of the following:
 - The member has ONE of the following:
 - Has tried and had an inadequate response to hydroxychloroquine; **OR**
 - Has intolerance, or hypersensitivity to hydroxychloroquine; **OR**
 - The member has an FDA labeled contraindication to hydroxychloroquine; **AND**
 - ONE of the following:
 - The member has ONE of the following:
 - Has tried and had an inadequate response to ONE corticosteroid OR immunosuppressive agent (i.e., azathioprine, methotrexate, mycophenolate, cyclophosphamide); **OR**
 - Has an intolerance or hypersensitivity to ONE corticosteroid OR immunosuppressive agent (i.e., azathioprine, methotrexate, mycophenolate, cyclophosphamide); **OR**

- Member has an FDA labeled contraindication to ALL corticosteroids AND immunosuppressive agents (i.e., azathioprine, methotrexate, mycophenolate, cyclophosphamide); **AND**
- Member is currently treated with and will continue standard SLE therapy (i.e., corticosteroids, hydroxychloroquine, azathioprine, methotrexate, mycophenolate, cyclophosphamide); **AND**
- Member does not have severe active central nervous system (CNS) lupus; **AND**
- Member will NOT be using in combination Lupkynis; **AND**
- ONE of the following:
 - The member will NOT be using the requested agent in combination with another immunomodulatory agent (e.g., TNF inhibitors, JAK inhibitors, IL-4 inhibitors, etc.); **OR**
 - The member will be using the requested agent in combination with another immunomodulatory agent AND BOTH of the following:
 - The prescribing information for the requested agent does NOT limit the use with another immunomodulatory agent; **AND**
 - There is support for the use of combination therapy (submitted copies of clinical trials, phase III studies, or guidelines required)

Lupus Nephritis (LN) ‡ ^{115-117,132,153,155,159,166,169}

- Member has a diagnosis of active LN; **AND**
- Member has class III, IV, V disease confirmed via kidney biopsy; **AND**
- Member will be using background immunosuppressive LN therapy (e.g., corticosteroids plus mycophenolate, azathioprine, or cyclophosphamide) in combination; **AND**
- Member does not have severe active central nervous system (CNS) lupus; **AND**
- ONE of the following:
 - The member will NOT be using the requested agent in combination with another immunomodulatory agent (e.g., TNF inhibitors, JAK inhibitors, IL-4 inhibitors, etc.); **OR**
 - The member will be using the requested agent in combination with another immunomodulatory agent AND BOTH of the following:
 - The prescribing information for the requested agent does NOT limit the use with another immunomodulatory agent; **AND**
 - There is support for the use of combination therapy (submitted copies of clinical trials, phase III studies, or guidelines required)

Myasthenia Gravis (unrelated to immunotherapy-related toxicity) ‡ ^{118-120,156}

- Member has muscle-specific tyrosine kinase (MuSK)-antibody positive disease; **AND**
- Member 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* Members ¹³³⁻¹³⁸

- Used for suppression of panel reactive anti-human leukocyte antigen (HLA) antibodies prior to transplantation; **OR**
- 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) ‡ ^{90-92,157,165}

- Member has a confirmed diagnosis based on the following:
 - Member was found to be seropositive for aquaporin-4 (AQP4) IgG antibodies; **AND**
 - Member has at least one core clinical characteristic § (**Note: some core clinical characteristics require both clinical and typical MRI findings*); **AND**
 - Alternative diagnoses have been excluded [e.g., myelin oligodendrocyte glycoprotein (MOG) antibody disease (MOGAD), multiple sclerosis, sarcoidosis, cancer, chronic infection, etc.]; **OR**
 - Member is seronegative for AQP4-IgG antibodies OR has unknown AQP4-IgG status; **AND**
 - Member has at least two core clinical characteristics § occurring as a result of one or more clinical attacks; **AND**
 - Member has experienced ALL of the following:
 - At least 1 core clinical characteristic must be acute optic neuritis, acute myelitis, or area postrema syndrome
 - Fulfillment of typical MRI findings requirements for each area affected ψ; **AND**
 - Alternative diagnoses have been excluded [e.g., myelin oligodendrocyte glycoprotein (MOG) antibody disease (MOGAD), multiple sclerosis, sarcoidosis, cancer, chronic infection, etc.]; **AND**
- Used as a single agent or in combination with immunosuppressive therapy (e.g., azathioprine, methotrexate, mycophenolate, etc.)

§ Core Clinical Characteristics of NMOSD ^{90,157}

- 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) ¹⁵⁷

- Optic neuritis: Normal cerebral MRI (or only nonspecific white matter lesions) OR longitudinally extensive optic nerve lesion ($\geq$ half of the length of the optic nerve or involving optic chiasm; T2 or T1/Gd)
- Myelitis: Intramedullary lesion $\geq$ 3 contiguous VS (LETM) OR focal atrophy $\geq$ 3 contiguous VS in members 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)
- $\nexists$ 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; VS = vertebral segments

Antisynthetase Syndrome-Related Interstitial Lung Disease $\pm$ ^{167,168,174,186}

- Member has antisynthetase antibody positive disease (e.g., anti-Jo-1, -PL-7, -PL-12, -OJ, -EJ, etc.); **AND**
- 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**
- Member has documented severe active disease; **AND**
- Member has recurrent or progressive disease despite treatment with glucocorticoids and/or other immunosuppressive agents (e.g., azathioprine, mycophenolate mofetil, cyclophosphamide, tacrolimus, etc.); **AND**
- Will be used in combination with glucocorticoids or other immunosuppressive agents (e.g., azathioprine, mycophenolate mofetil, cyclophosphamide, tacrolimus, etc.), unless the member has a contraindication or intolerance

Idiopathic (Primary) Membranous Nephropathy $\pm$ ^{172,175-177}

- Member has a documented diagnosis of idiopathic (primary) membranous nephropathy; **AND**
- 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**
 - Used as first-line therapy in members with any of the following moderate to high risk factors for progressive disease:
 - 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**
 - eGFR < 60 ml/min/1.73m²; **OR**
 - Proteinuria > 8 g/d for > 6 months; **OR**

- Member has experienced serious complications of nephrotic syndrome (e.g., acute kidney injury, infection, thromboembolic events, etc.); **OR**
- 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**
- 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**
 - Member has a stable eGFR; **AND**
 - Will be used in combination with a calcineurin inhibitor if previously treated with rituximab alone in the first-line setting; **OR**
- Used for disease recurrence following kidney transplant; **AND**
 - Member has proteinuria > 1 g/d

Pediatric Idiopathic Nephrotic Syndrome ‡ ^{170,171,173,197}

- Member is 12 years of age or younger; **AND**
- Member has symptomatic disease (i.e., nephrotic-range proteinuria and either hypoalbuminemia or edema when albumin level is not available); **AND**
- Member has been diagnosed with one of the following:
 - Frequently relapsing nephrotic syndrome (FRNS) with at least three relapses per year or at least two relapses within 6 months following remission of the initial episode
 - Steroid dependent nephrotic syndrome (SDNS) with two consecutive relapses during steroid therapy (either at full-dose or during tapering) or within 14 days of cessation of steroid therapy
 - Steroid resistant nephrotic syndrome (SRNS) with failure to achieve complete remission within a 4-week course of daily corticosteroids; **AND**
- Member 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 ‡ ^{178-182,195}

- Member has a confirmed diagnosis of IgG4-RD (e.g., physical exam findings, imaging results, laboratory tests, pathological findings in involved organ/sites, etc.); **AND**
- Other conditions that mimic IgG4-related disease have been ruled out (e.g., malignancy, infection, other autoimmune disorders, etc.); **AND**
- Member is experiencing (or recently experienced) an IgG4-RD flare that required corticosteroid treatment; **AND**
 - Member has disease that is refractory to corticosteroids; **OR**
 - Member has a contraindication or intolerance to corticosteroid treatment; **AND**

- Member is at high risk of recurrent disease flares based on a history of disease in ≥ 2 organs/sites; **AND**
- At least one of the following organs are affected:
 - Pancreas, bile ducts/biliary tree, orbits, lungs, kidneys, lacrimal glands, major salivary glands, retroperitoneum, aorta, pachymeninges, sinonasal tract, and/or thyroid gland

Wiskott-Aldrich Syndrome ‡²⁰⁵

- Used as pretreatment prior to infusion of etuvetidigene autotemcel; **AND**
 - Used to deplete autoreactive B-cells; **OR**
 - Used as pre-emptive treatment for potential lymphoproliferative disorder due to Epstein Barr Virus infection

† FDA Approved Indication(s); ‡ Compendia Recommended Indication(s); Φ Orphan Drug

IV. Renewal Criteria¹⁻⁴

Prior authorization validity may be renewed based upon the following criteria:

- Member continues to meet the universal and other indication-specific relevant criteria such as concomitant therapy requirements (not including prerequisite therapy), performance status, etc. identified in section III; **AND**
- Duration of authorization has not been exceeded (*refer to Section I*); **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: severe infusion-related reactions, tumor lysis syndrome (TLS), severe mucocutaneous reactions (e.g., paraneoplastic pemphigus, Stevens-Johnson syndrome, lichenoid dermatitis, etc.), progressive multifocal leukoencephalopathy (PML), hepatitis B virus reactivation, serious infections (bacterial, fungal or viral), cardiovascular adverse reactions (e.g., ventricular fibrillation, myocardial infarction, cardiogenic shock, cardiac arrhythmias), renal toxicity, bowel obstruction and perforation, etc.; **AND**

Oncology Indications^{1-5,23-25,34,44,50,62,94-98,102-104,128-130}

- Member has not exceeded dosing or duration limits as defined in Sections I, II, and V; **AND**

Adult Acute Lymphoblastic Leukemia (ALL)

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

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 Indications¹⁻⁴

Rheumatoid Arthritis (RA)

- Disease response as indicated by improvement in signs and symptoms compared to baseline such as the number of tender and swollen joint counts, reduction of C-reactive protein, improvement of patient global assessment, 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 $\geq 20\%$ improvement on the American College of Rheumatology-20 (ACR20) criteria, or improvement of disease severity on RAPID3 assessment]; **AND**
- 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 member has:
 - Shown an initial response to therapy; **AND**
 - Received a minimum of one maintenance dose at the dose and interval specified below; **AND**
 - Responded to therapy with subsequent loss of response

Thrombocytopenic Purpura (ITP or Evans Syndrome) ^{7-9,63}

- Disease response as indicated by the achievement and maintenance of a platelet count of at least $30 \times 10^9/L$ and at least doubling the baseline platelet count

Thrombotic Thrombocytopenic Purpura (TTP)¹⁹⁶

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

Multiple Sclerosis (MS) ^{147,151,203}

- 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 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) ^{1-4, 125}

- Disease response as indicated by disease control and improvement in signs and symptoms of condition compared to baseline; **AND**
- 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 ^{10,11,35,61}

- Member is currently receiving tapering doses of corticosteroids or has discontinued use of corticosteroids; **AND**
 - Disease response as indicated by one of the following:
 - Complete epithelialization of lesions and improvement in signs and symptoms of condition compared to baseline; **OR**
 - Member has not developed new lesions and established lesions begin to heal; **OR**
 - For Relapses ONLY:
 - Member previously achieved disease control; **AND**
 - Member 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) ^{31,152}

- Disease response as indicated by improvement in signs and symptoms of anemia (e.g., dyspnea, fatigue, etc.); **AND**
- Member has had an improvement in laboratory values (e.g., hemoglobin, hematocrit, etc.), reduced transfusion needs, and/or reduced glucocorticoid use

Systemic Lupus Erythematosus (SLE) ^{153,155,158,161-163}

- Member has experienced clinical benefit with the requested agent; **AND**
- The member is currently treated with and will continue standard SLE therapy (i.e., corticosteroids, hydroxychloroquine, azathioprine, methotrexate, mycophenolate, cyclophosphamide)

Lupus Nephritis ¹¹⁵⁻¹¹⁷

- Member has experienced clinical benefit with the requested agent (*Note: Prior authorization validity may only be renewed in members 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) ¹¹⁸⁻¹²⁰

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

NMOSD ^{90,91}

- 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 ^{167,168,174}

- 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 $\geq 10\%$ increase in FVC%, TLC%, or DLCO%; stabilization defined as $< 10\%$ decrease in FVC%, TLC%, or DLCO%)
 - Improvement or stabilization of chest CT score (i.e., improvement defined as $\geq 10\%$ decrease in CT score; stabilization defined as a $\leq 10\%$ increase in CT score)

Idiopathic Membranous Nephropathy ^{172,175,177}

- Member 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**
- Member has resistant disease following first-line therapy with rituximab; **AND**
 - Member 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 ‡ ^{170,171,173,197}

- Member previously achieved beneficial disease response from the prior course of therapy; **AND**
- Member is experiencing signs and symptoms of recurrent active disease necessitating additional doses (e.g., recurrence of nephrotic-range proteinuria with a dipstick $\geq 3+$ [≥ 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 ‡ ^{178-182,195}

- Disease response as indicated by one or more of the following:
 - Reduction in corticosteroid requirement for IgG4-RD flare treatment from baseline
 - Reduction in IgG4-RD flares from baseline
 - Stabilization/improvement in symptoms, physical exam findings, imaging results, laboratory tests, and/or pathological findings in IgG4-RD involved organ/sites compared to baseline

V. Dosage/Administration 1-5,9,19,23-26,32,34,40,42,44,50,62,80,83-89,91,94-98,102-111,115-118,122-125,128-133,135-

137,140,151,163,164,166,167,169-172,174,177-184,186-189,191-193,195,198-199,206-209

Indication		Dose
CLL/SLL	Initial Therapy	375 mg/m ² intravenously (IV) weekly for 12 doses; OR 375 mg/m ² IV cycle 1, then 500 mg/m ² every 28 days cycles 2-6 (6 total doses); OR 375 mg/m ² IV cycle 1, followed by 500 mg/m ² every 2 weeks for 4 doses, then 500 mg/m ² every 28 days for 3 doses (8 total doses)
	Renewal Therapy	375 mg/m ² IV every 3 months; OR 500 mg/ m ² IV every 8 weeks
Waldenström Macroglobulinemia/ Lymphoplasmacytic Lymphoma	Initial Therapy	375 mg/m ² IV weekly for 12 doses
	Renewal Therapy	375 mg/m ² IV once weekly for 4 doses per 6 month period; OR 375 mg/ m ² IV every 8 weeks
Castleman Disease, Primary Cutaneous B-Cell Lymphomas, or Adult Hodgkin Lymphoma	Initial Therapy	375 mg/m ² IV once weekly for 4 – 8 doses in a 6 month period
	Renewal Therapy	375 mg/m ² IV once weekly for 4 doses per 6 month period; OR 375 mg/ m ² IV every 8 weeks
Adult B-Cell Lymphomas		<u>In combination with brentuximab vedotin and lenalidomide</u> 375 mg/m ² IV every 21 days until progression or unacceptable toxicity <u>All other regimens</u> Initial Therapy: 375 mg/m ² IV once weekly for 4 – 8 doses in a 6 month period Renewal Therapy: 375 mg/m ² IV once weekly for 4 doses per 6 month period; OR 375 mg/m ² IV every 8 weeks
Pediatric Aggressive Mature B-Cell Lymphomas (Primary Mediastinal Large B-cell Lymphoma, DLBCL, Burkitt Lymphoma, Burkitt-like Lymphoma)		<u>Induction* [courses 1 and 2 (COPDAM1 and COPDAM2)]</u> 375 mg/m ² IV, two doses during each of the induction courses (Day -2 and Day 1). <i>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.</i> <u>Consolidation* [courses 1 and 2 (CYM/CYVE)]</u> 375 mg/m ² IV, one dose during each of the consolidation courses (Day 1) <u>Relapsed/Refractory</u> RCYVE – 375mg/m ² IV on day 1 of each 21-day cycle RICE – 375 mg/m ² IV on days 1 and 3 of courses 1 and 2, and on day 1 only of course 3 if needed.

Indication	Dose
	<i>*Note: dosing and dosing schedules are highly variable and dependent on regimen used, please refer to NCCN and PI for additional protocols.</i>
Pediatric Aggressive Mature B-Cell Lymphomas (Post-Transplant Lymphoproliferative Disorders)	375 mg/m ² IV once weekly for 4 – 8 doses in a 6 month period
Pediatric Mature B-Cell Acute Leukemia	<u>Induction* [courses 1 and 2 (COPDAM1 and COPDAM2)]</u> 375 mg/m² IV, two doses during each of the induction courses (Day -2 and Day 1). <i>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.</i> <u>Consolidation* [courses 1 and 2 (CYM/CYVE)]</u> 375 mg/m² IV, one dose during each of the consolidation courses (Day 1) <i>*Note: dosing and dosing schedules are highly variable and dependent on regimen used, please refer to NCCN and PI for additional protocols.</i>
CNS Cancers	<u>Systemic (intravenous) administration*</u> <u>Initial Therapy:</u> Up to 750 mg/m² weekly for 4 – 8 doses <u>Renewal Therapy:</u> 375 mg/m² IV once weekly for 4 doses every 6 months; OR 375 mg/m² IV every 4-8 weeks <u>Intra-CSF (intrathecal/intraventricular) administration*</u> 25 mg weekly to twice weekly <i>*Note: dosing and dosing schedules are highly variable and dependent on regimen used, please refer to NCCN</i>
ALL	375 mg/m ² 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/m ² IV once weekly for 4 – 8 doses; OR 375mg/m ² IV on days 1 and 15 every 28 days for 4 cycles, then 375mg/m ² IV every 4 weeks for 4 cycles (up to 8 <u>total</u> cycles)
KSHV-Associated Inflammatory Cytokine Syndrome	375 mg/m ² IV on day 1 of each 21-day cycle for a total of 6 cycles
Rheumatoid Arthritis	1,000 mg IV on days 1 and 15, repeated every 24 weeks. May repeat up to every 16 weeks** following the previous infusion in members requiring more frequent dosing based on clinical evaluation. <i>**Dose escalation criteria detailed in section IV must be met prior to increasing dosing frequency.</i>

Indication	Dose
Pemphigus Vulgaris	<u>Initiation</u> 1,000 mg IV on days 1 and 15; OR 375 mg/m² IV weekly for 4 doses <u>Maintenance</u> 500 mg IV at month 12 and repeat every 6 months thereafter or based on clinical evaluation <u>Relapse</u> 1,000 mg IV upon relapse, resumption of glucocorticoids may be considered <i>*Subsequent infusions (maintenance and relapse) should be no sooner than 16 weeks after the previous infusion.</i>
AIHA	<u>Warm-reactive disease</u> 375 mg/m² IV weekly for 4 doses every 6 months; OR 1,000 mg IV on days 1 and 15 every 6 months <u>Cold agglutinin disease</u> 375 mg/m² IV weekly for 4 doses every 6 months
Thrombocytopenic Purpura or Thrombotic Thrombocytopenic Purpura (TTP)	375 mg/m² IV weekly for 4 doses every 6 months; OR 1,000 mg IV on days 1 and 15 every 6 months
Management of Immune Checkpoint Inhibitor-Related Toxicities	<u>Bullous Dermatitis/Bullous Pemphigoid</u> 1,000 mg IV every 2 weeks for 2 doses, then 500 mg IV at months 12 and 18 as needed <u>Thrombocytopenia, Hemolytic Anemia, Encephalitis, Acute Kidney Injury/Elevated Serum Creatinine</u> 375 mg/m² IV weekly for 4 doses; OR 1,000 mg IV on days 1 and 15 <u>Myositis</u> 375 mg/m² IV weekly for 4 doses <u>Myasthenia Gravis</u> 375 mg/m² IV weekly for 4 doses; OR 500 mg/m² IV every 2 weeks for 2 doses
GPA (WG), MPA	<u>Induction (Pediatric and Adult)</u> 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): ○ 575 mg/m² IV on days 1 and 15 (BSA ≤1.5m²)

Indication	Dose
	750 mg/m² IV on days 1 and 15 (BSA >1.5m²) <u>Maintenance</u> - Pediatric: 250 mg/m² IV on days 1 and 15, then 250 mg/m² IV every 6 months thereafter based on clinical evaluation - Adult: 500 mg to 1,000 mg IV on days 1 and 15, then 500 mg to 1,000 mg IV every 6 months thereafter based on clinical evaluation <i>*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.</i> <i>*Initial MAINTENANCE infusions should be initiated within 4 weeks following disease control when initial induction occurred with other standard of care immunosuppressants.</i>
cGVHD	375 mg/m ² IV weekly for 4 doses, then 375 mg/m ² IV monthly for 4 months -OR- 375 mg/m ² IV weekly for 4 doses (Note: If no response or an incomplete response, a second course of 4 weekly doses may be administered 8 weeks after initial therapy. If relapse to one or two 4-week courses, a second or third course of 4 weekly doses may be administered) -OR- 375 mg/m ² IV weekly for 4 – 8 doses
Hematopoietic Cell Transplantation	<u>Conditioning:</u> 375 mg/m ² IV for 1 day before transplant, then 1000 mg/m ² IV on days 1,8, and 15 after transplant
Transplant Associated-Thrombotic Microangiopathy (TA-TMA)	375 mg/m ² IV weekly for 4 doses
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/m ² once weekly for 4 weeks, repeat every 6 months
Histiocytic Neoplasms – Rosai-Dorfman Disease	500 mg/m ² IV every 1 – 2 weeks for 2 – 6 doses every 6 months
SLE or Lupus Nephritis	1,000 mg IV on days 1 and 15 every 6 months -OR- 375 mg/m ² IV once weekly for 4 doses every 6 months
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

Indication	Dose
	-OR- 375 mg/m ² IV once weekly for 4 doses, may repeat a full or partial course every 6 months
Pediatric Hodgkin Lymphoma	375 mg/m ² 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 m ² : 375 mg/m ² IV weekly for up to 4 doses – Pediatrics weighing <0.5 m ² : 12.5 mg/kg IV 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/m ² IV once weekly for 4 doses repeated every 6 months
Pediatric Idiopathic Nephrotic Syndrome	375 mg/m ² IV once weekly for 1-4 doses
Idiopathic Membranous Nephropathy	375 mg/m ² 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	<u>Induction:</u> 375 mg/m ² IV once weekly for 1-4 doses -OR- 1,000 mg IV on days 1 and 15 <i>*Subsequent infusions (maintenance and relapse) may be administered at either induction schedule above and should be repeated no sooner than every 6 months.</i>
Wiskott-Aldrich Syndrome	375 mg/m ² IV as a single dose approximately 22 days prior to etuvetidigene autotemcel administration
<i>Abbreviations: COP = Cyclophosphamide, Oncovin (vincristine), Prednisone; COPDAM = Cyclophosphamide, Oncovin (vincristine), Prednisolone, Adriamycin (doxorubicin), Methotrexate; CYM = Cytarabine (Ara-C), Methotrexate; CYVE = Cytarabine (Ara-C), Vepesid (Etoposide, VP-16); RICE = Rituximab, Ifosfamide, Carboplatin, Etoposide (VP-16)</i> For adults only: upon renewal, following at least 12 months of sustained remission and stability at current dose and frequency, consider incremental dose reduction and/or interval extension to the following maintenance dosing: Rheumatoid Arthritis: • Goal maintenance dose: 500 mg IV every 6 months ○ Dose decrease by no more than 500 mg Multiple Sclerosis: • Goal maintenance dose: 500 mg IV every 12 months ○ Dose decrease by no more than 500 mg; OR ○ Interval increase by no more than 6 months <i>Note: This information is not meant to replace clinical decision making when initiating or modifying medication therapy and should only be used as a guide. Member-specific variables should be taken into account.</i>	

VI. Billing Code/Availability Information

HCPCS Code(s):

- J9312 – Injection, rituximab, 10 mg; 1 billable unit = 10 mg (*Rituxan IV only*)
- Q5115 – Injection, rituximab-abbs, biosimilar, (truxima), 10 mg; 1 billable unit = 10 mg
- Q5119 – Injection, rituximab-pvvr, biosimilar, (ruxience), 10 mg; 1 billable unit = 10 mg
- Q5123 – Injection, rituximab-arrx, biosimilar, (riabni), 10 mg; 1 billable unit = 10 mg

NDC(s):

- Rituxan 100 mg/10 mL single-dose vial for injection: 50242-0051-xx
- Rituxan 500 mg/50 mL single-dose vial for injection: 50242-0053-xx
- Truxima 100 mg/10 mL single-dose vial for injection: 63459-0103-xx
- Truxima 500 mg/50 mL single-dose vial for injection: 63459-0104-xx
- Ruxience 100 mg/10 mL single-dose vial for injection: 00069-0238-xx
- Ruxience 500 mg/50 mL single-dose vial for injection: 00069-0249-xx
- Riabni 100 mg/10 mL single-dose vial for injection: 55513-0224-xx
- Riabni 500 mg/50 mL single-dose vial for injection: 55513-0326-xx

VII. References

1. Rituxan [package insert]. South San Francisco, CA; Genentech, Inc.; December 2021. Accessed March 2026.
2. Truxima [package insert]. Incheon, Republic of Korea; Celltrion, Inc.; June 2025. Accessed March 2026
3. Ruxience [package insert]. New York, NY; Pfizer, Inc; June 2025. Accessed March 2026.
4. Riabni [package insert]. Thousand Oaks, CA; Amgen, Inc.; July 2025. Accessed March 2026.
5. Referenced with permission from the NCCN Drugs & Biologics Compendium (NCCN Compendium®) rituximab. National Comprehensive Cancer Network, 2026. The NCCN Compendium® is a derivative work of the NCCN Guidelines®. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc. To view the most recent and complete version of the Compendium, go online to NCCN.org. Accessed March 2026.
6. Arnold DM, Dentali F, Crowther MA, et al. Systematic review: efficacy and safety of rituximab for adults with idiopathic thrombocytopenic purpura. *Ann Intern Med* 2007; 146:25-33.
7. Zaja F, Baccarani M, Mazza P, et al: Dexamethasone plus rituximab yields higher sustained response rates than dexamethasone monotherapy in adults with primary immune thrombocytopenia. *Blood* 2010; 115(14):2755-2762.
8. Stasi R, Pagano A, Stipa E, et al: Rituximab chimeric anti-CD10 monoclonal antibody treatment for adults with chronic idiopathic thrombocytopenic purpura. *Blood* 2001; 98(4):952-957.
9. Neunert C, Lim W, Crowther M, Cohen A, Solberg L Jr, Crowther MA. The American Society of Hematology 2011 evidence-based practice guideline for immune thrombocytopenia. *Blood*. 117(16):4190-4207.

10. Joly P, Mouquet H, Roujeau JC, et al. A single cycle of rituximab for the treatment of severe pemphigus. *N Engl J Med* 2007; 357:545-52.
11. Ahmed AR, Spigelman Z, Cavacini LA et al. Treatment of pemphigus vulgaris with rituximab and intravenous immune globulin. *N Engl J Med* 2006; 355:1772-9.
12. Singh JA, Saag KG, Bridges SL Jr, et al. 2015 American College of Rheumatology Guideline for the Treatment of Rheumatoid Arthritis. *Arthritis Care Res (Hoboken)*. 2015 Nov 6. Doi: 10.1002/acr.22783.
13. Smolen JS, Landewé R, Bijlsma J, et al. EULAR recommendations for the management of rheumatoid arthritis with synthetic and biological disease-modifying antirheumatic drugs: 2016 update. *Ann Rheum Dis*. 2017 Mar 6. Pii: annrheumdis-2016-210715.
14. González-Barca E, Domingo-Domenech E, Capote FJ, et al. Prospective phase II trial of extended treatment with rituximab in patients with B-cell post-transplant lymphoproliferative disease. *Haematologica*. 2007 Nov; 92(11):1489-94.
15. Chamberlain MC, Johnston SK, Van Horn A, et al. Recurrent lymphomatous meningitis treated with intra-CSF rituximab and liposomal ara-C. *J Neurooncol*. 2009 Feb;91(3):271-7.
16. Scully M, Cohen H, Cavenagh J, et al. Remission in acute refractory and relapsing thrombotic thrombocytopenic purpura following rituximab is associated with a reduction in IgG antibodies to ADAMTS-13. *Br J Haematol* 2007;136:451-461.
17. Fakhouri F, Vernant JP, Veyradier A, et al. Efficiency of curative and prophylactic treatment with rituximab in ADAMTS13-deficient thrombotic thrombocytopenic purpura: a study of 11 cases. *Blood*. 2005;106:1932-37.
18. Elliott MA, Heit JA, Rajiv K, et al. Rituximab for refractory and or relapsing thrombotic thrombocytopenic purpura related to immune-mediated severe ADAMTS13-deficiency: a report of four cases and a systematic review of the literature. *Eur J Haematol* 2009. Epub ahead of print, doi:10.1111/j.1600-0609.2009.01292.
19. Scully M, McDonald V, Cavenagh J, et al. A phase 2 study of the safety and efficacy of rituximab with plasma exchange in acute acquired thrombotic thrombocytopenic purpura. *Blood*. 2011;118(7):1746-1753.
20. Tun NM, Villani GM. Efficacy of rituximab in acute refractory or chronic relapsing non-familial idiopathic thrombotic thrombocytopenic purpura: a systematic review with pooled data analysis. *J Thromb Thrombolysis*. 2012;34(3):347-359.
21. Froissart A, Buffet M, Veyradier A, et al. Efficacy and safety of first-line rituximab in severe, acquired thrombotic thrombocytopenic purpura with a suboptimal response to plasma exchange. Experience of the French Thrombotic Microangiopathies Reference Center. *Crit Care Med* 2012; 40(1):104-111.0
22. van Dorp S, Resemann H, te Boome L, et al. The immunological phenotype of rituximab-sensitive chronic graft-versus-host disease: a phase II study. *Haematologica* 2011;96(9):1380-1384.

23. Kim SJ, Lee JW, Jung CW, et al. Weekly rituximab followed by monthly rituximab treatment for steroid-refractory chronic graft-versus-host disease: results from a prospective, multicenter, phase II study. *Haematologica* 2010;95(11):1935-1942.
24. Cutler C, Miklos D, Kim HT, et al, Rituximab for Steroid-Refractory Chronic Graft-Versus-Host Disease. *Blood*. 2006, 108(2):756-62.
25. Wolff D, Schleuning M, von Harsdorf S, et al. Consensus Conference on Clinical Practice in Chronic GVHD: Second-Line Treatment of Chronic Graft-versus-Host Disease. *Biol Blood Marrow Transplant*. 2011 Jan;17(1):1-17. Doi: 10.1016/j.bbmt.2010.05.011.
26. Frame JN, Fichtner R, McDevitt PW. Rituximab for the treatment of autoimmune hemolytic anemia (AIHA) in adults: an analysis of literature reports in 92 patients. *Blood* 2004;104:Abstract 3721.
27. Birgens H, Frederiksen H, Hasselbalch HC, et al: A phase III randomized trial comparing glucocorticoid monotherapy versus glucocorticoid and rituximab in patients with autoimmune haemolytic anaemia. *Br J Haematol* 2013; 163(3):393-399.
28. Schollkopf C, Kjeldsen L, Bjerrum OW, et al. Rituximab in chronic cold agglutinin disease: a prospective study of 20 patients. *Leuk Lymphoma* 2006; 47(N2):253-260.
29. Berentsen S, Ulvestad E, Gjertsen BT, et al. Rituximab in chronic cold agglutinin disease: a prospective study of 20 patients. *Blood* 2004; 103(8):2925-2928.
30. Reynaud Q, Durieu I, Dutertre M, et al. Efficacy and safety of rituximab in auto-immune hemolytic anemia: A meta-analysis of 21 studies. *Autoimmun Rev*. 2015;14(4):304-313.
31. Barcellini W, Zaja F, Zaninoni A, et al, "Low-dose Rituximab in Adult Patients With Idiopathic Autoimmune Hemolytic Anemia: Clinical Efficacy and Biologic Studies," *Blood*, 2012, 119(16):3691-7.
32. Roumier M, Loustau V, Guillaud C, et al. Characteristics and outcome of warm autoimmune hemolytic anemia in adults: New insights based on a single-center experience with 60 patients. *Am J Hematol*. 2014;89(9):E150-E155.
33. Gobert D, Bussel JB, Cunningham-Rundles C, et al. Efficacy and safety of rituximab in common variable immunodeficiency-associated immune cytopenias: a retrospective multicentre study on 33 patients. *Br J Haematol*. 2011;155(4):498-508.
34. Shin YW, Lee ST, Park KI, et al. Treatment strategies for autoimmune encephalitis. *Ther Adv Neurol Disord*. 2017 Aug 16;11:1756285617722347. Doi: 10.1177/1756285617722347. eCollection 2018. Review.
35. Murrell DF, Dick S, Ahmed AR, et al. Consensus statement on definitions of disease, end points, and therapeutic response for pemphigus. *J Am Acad Dermatol*. 2008 June; 58(6): 1043–1046. Doi:10.1016/j.jaad.2008.01.012. Avail at: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2829665/pdf/nihms82304.pdf>
36. Grover, S. Scoring Systems in Pemphigus. *Indian J Dermatol*. 2011 Mar-Apr; 56(2): 145–149. doi: 10.4103/0019-5154.80403
37. Daniel BS, Hertl M, Weth VP, et al. Severity score indexes for blistering diseases. *Clin Dermatol*. 2012 Jan-Feb; 30(1): 108–113. Doi: 10.1016/j.clindermatol.2011.03.017

38. Joly P, Litrowski N. Pemphigus group (vulgaris, vegetans, foliaceus, herpetiformis, brasiliensis). Clin Dermatol 2011; 29:432.
39. Lambert MP, Gernsheimer TB. Clinical updates in adult immune thrombocytopenia. Blood. 2017. 129:2829-2835. Doi:10.1182/blood-2017-03-754119
40. Schulz H, Pels H, Schmidt-Wolf I, et al. Intraventricular treatment of relapsed central nervous system lymphoma with the anti-CD20 antibody rituximab. Haematologica January 2004 89: 753-754.
41. Fahrenbruch R, Kintzel P, Bott AM, et al. Dose Rounding of Biologic and Cytotoxic Anticancer Agents: A Position Statement of the Hematology/Oncology Pharmacy Association. J Oncol Pract. 2018 Mar;14(3):e130-e136.
42. Hematology/Oncology Pharmacy Association. *Intravenous Cancer Drug Waste Issue Brief*. Updated February 2024. Retrieved from https://www.hoparx.org/documents/287/HOPA_Drug_Waste_Issue_Brief_-_Updated_02.22.24.pdf
43. Bach PB, Conti RM, Muller RJ, et al. Overspending driven by oversized single dose vials of cancer drugs. BMJ. 2016 Feb 29;352:i788.
44. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for B-Cell Lymphomas, Version 2.2026. National Comprehensive Cancer Network, 2026. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc. To view the most recent and complete version of the Guidelines, go online to NCCN.org. Accessed March 2026.
45. Imbruvica [package insert]. Horsham, PA; Janssen Biotech, Inc. October 2025.
46. Keystone E, Burmester GR, Furie R, et al. Improvement in patient-reported outcomes in a rituximab trial in patients with severe rheumatoid arthritis refractory to anti-tumor necrosis factor therapy. Arthritis Rheum. 2008 Jun 15;59(6):785-93. Doi: 10.1002/art.23715.
47. Mease PJ, Cohen S, Gaylis NB, et al. Efficacy and Safety of Retreatment in Patients with Rheumatoid Arthritis with Previous Inadequate Response to Tumor Necrosis Factor Inhibitors: Results from the SUNRISE Trial. The Journal of Rheumatology May 2010, 37 (5) 917-927; DOI: <https://doi.org/10.3899/jrheum.090442>
48. Tak PP, Rigby W, Rubbert-Roth A, et al. Sustained inhibition of progressive joint damage with rituximab plus methotrexate in early active rheumatoid arthritis: 2-year results from the randomised controlled trial IMAGE. Ann Rheum Dis. 2012 Mar;71(3):351-7. Doi: 10.1136/annrheumdis-2011-200170. Epub 2011 Oct 19.
49. Emery P, Deodhar A, Rigby WF, et al. Efficacy and safety of different doses and retreatment of rituximab: a randomised, placebo-controlled trial in patients who are biological I with active rheumatoid arthritis and an inadequate response to methotrexate (Study Evaluating Rituximab's Efficacy in MTX iNadequate rEsponders (SERENE)). Ann Rheum Dis. 2010 Sep;69(9):1629-35. Doi: 10.1136/ard.2009.119933. Epub 2010 May 20. Erratum in: Ann Rheum Dis. 2011 Aug;70(8):1519.

50. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Pediatric Aggressive Mature B-Cell Lymphomas, Version 2.2025. National Comprehensive Cancer Network, 2026. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc. To view the most recent and complete version of the Guidelines, go online to NCCN.org. Accessed March 2026.
51. Lee KH, Lee J, Bae JS, et al. Analytical similarity assessment of rituximab biosimilar CT-P10 to reference medicinal product. *Mabs*. 2018;10(3):380-396
52. Ogura M, Sancho JM, Cho S-G, et al. Efficacy, pharmacokinetics, and safety of the biosimilar CT-P10 in comparison with rituximab in patients with previously untreated low-tumour-burden follicular lymphoma: a randomised, double-blind, parallel-group phase 3 trial. *Lancet Haematol*. 2018;5:e543-e553.
53. Gulácsi L, Brodszky V, Baji P, et al. The rituximab biosimilar CT-P10 in rheumatology and cancer: a budget impact analysis in 28 European countries. *Adv Ther*. 2017; 34: 1128-1144.
54. Yoo DH, Suh CH, Shim SC, et al. A multicentre randomised controlled trial to compare the pharmacokinetics, efficacy and safety of CT-P10 and innovator rituximab in patients with rheumatoid arthritis. *Ann Rheum Dis*. 2017; 76: 566-570.
55. Suh C, Berrocal Kasay A, Chalouhi El-Khoury E, et al. Pharmacokinetics and safety of three formulations of rituximab (CT-P10, US-sourced innovator rituximab and EU-sourced innovator rituximab) in patients with rheumatoid arthritis: results from phase 3 randomized controlled trial over 24 weeks. *Arthritis Rheumatol*. 2016; 68: 1634.
56. Kim WS, Buske C, Ogura M, et al. Efficacy, pharmacokinetics, and safety of the biosimilar CT-P10 compared with rituximab in patients with previously untreated advanced-stage follicular lymphoma: a randomised, double-blind, parallel-group, non-inferiority phase 3 trial. *Lancet Haematol*. 2017; 4: e362-e373.
57. Cohen S, Emery P, Greenwald M, et al. A phase I pharmacokinetics trial comparing PF-05280586 (a potential biosimilar) and rituximab in patients with active rheumatoid arthritis. *Br J Clin Pharmacol*. 2016 Jul;82(1):129-38.
58. Williams JH, Hutmacher MM, Zierhut ML, et al. Comparative assessment of clinical response in patients with rheumatoid arthritis between PF-05280586, a proposed rituximab biosimilar, and rituximab. *Br J Clin Pharmacol*. 2016 Dec;82(6):1568-1579.
59. Sharman JP, Liberati AM, Ishizawa K, et al. A Randomized, Double-Blind, Efficacy and Safety Study of PF-05280586 (a Rituximab Biosimilar) Compared with Rituximab Reference Product (MabThera®) in Subjects with Previously Untreated CD20-Positive, Low-Tumor-Burden Follicular Lymphoma (LTB-FL). *BioDrugs*. 2019 Dec 9. Doi: 10.1007/s40259-019-00398-7.
60. Cohen SB, Burgos-Vargas R, Emery P, et al. Comparative assessment of clinical response in patients with rheumatoid arthritis between PF-05280586, a proposed rituximab biosimilar, and rituximab. *Br J Clin Pharmacol*. 2016 Dec;82(6):1568-1579.
61. Murrel DF, Peña S, Joly P, et al. Diagnosis and management of pemphigus: Recommendations of an international panel of experts. *JAAD*: Mar2020;82;3;575-585. DOI:<https://doi.org/10.1016/j.jaad.2018.02.021>.

62. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Management of Immune Checkpoint Inhibitor-Related Toxicities, Version 1.2026. National Comprehensive Cancer Network, 2026. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc. To view the most recent and complete version of the Guidelines, go online to NCCN.org. Accessed March 2026.
63. Neunert C, Terrell DR, Arnold DM, et al. American Society of Hematology 2019 guidelines for immune thrombocytopenia [published correction appears in Blood Adv. 2020 Jan 28;4(2):252]. Blood Adv. 2019;3(23):3829-3866. Doi:10.1182/bloodadvances.2019000966.
64. McLaughlin P, Grillo-López AJ, Link BK, et al. Rituximab chimeric anti-CD20 monoclonal antibody therapy for relapsed indolent lymphoma: half of patients respond to a four-dose treatment program. J Clin Oncol. 1998 Aug;16(8):2825-33.
65. Piro LD, White CA, Grillo-López AJ, et al. Extended Rituximab (anti-CD20 monoclonal antibody) therapy for relapsed or refractory low-grade or follicular non-Hodgkin's lymphoma. Ann Oncol. 1999;10(6):655-661. Doi:10.1023/a:1008389119525.
66. Davis TA, Grillo-López AJ, White CA, et al. Rituximab anti-CD20 monoclonal antibody therapy in non-Hodgkin's lymphoma: safety and efficacy of re-treatment. J Clin Oncol. 2000;18(17):3135-3143. Doi:10.1200/JCO.2000.18.17.3135.
67. Marcus R, Imrie K, Belch A, et al. CVP chemotherapy plus rituximab compared with CVP as first-line treatment for advanced follicular lymphoma. Blood. 2005;105(4):1417-1423. Doi:10.1182/blood-2004-08-3175.
68. Salles G, Seymour JF, Offner F, et al. Rituximab maintenance for 2 years in patients with high tumour burden follicular lymphoma responding to rituximab plus chemotherapy (PRIMA): a phase 3, randomised controlled trial [published correction appears in Lancet. 2011 Apr 2;377.
69. Hochster H, Weller E, Gascoyne RD, et al. Maintenance rituximab after cyclophosphamide, vincristine, and prednisone prolongs progression-free survival in advanced indolent lymphoma: results of the randomized phase III ECOG1496 Study. J Clin Oncol. 2009;27(10):1607-1614. Doi:10.1200/JCO.2008.17.1561.
70. Habermann TM, Weller EA, Morrison VA, et al. Rituximab-CHOP versus CHOP alone or with maintenance rituximab in older patients with diffuse large B-cell lymphoma. J Clin Oncol. 2006;24(19):3121-3127. Doi:10.1200/JCO.2005.05.1003,
71. Coiffier B, Thieblemont C, Van Den Neste E, et al. Long-term outcome of patients in the LNH-98.5 trial, the first randomized study comparing rituximab-CHOP to standard CHOP chemotherapy in DLBCL patients: a study by the Groupe d'Etudes des Lymphomes de l'Adulte. Blood. 2010;116(12):2040-2045. Doi:10.1182/blood-2010-03-276246.
72. Pfreundschuh M, Kuhnt E, Trümper L, et al. CHOP-like chemotherapy with or without rituximab in young patients with good-prognosis diffuse large-B-cell lymphoma: 6-year results of an open-label randomised study of the MabThera International Trial (MinT) Group. Lancet Oncol. 2011;12(11):1013-1022. Doi:10.1016/S1470-2045(11)70235-2.

73. Dakhil S, Hermann R, Schreeder MT, et al. Phase III safety study of rituximab administered as a 90-minute infusion in patients with previously untreated diffuse large B-cell and follicular lymphoma. *Leuk Lymphoma*. 2014;55(10):2335-2340. Doi:10.3109/10428194.2013.877135.
74. Fischer K, Bahlo J, Fink AM, et al. Long-term remissions after FCR chemoimmunotherapy in previously untreated patients with CLL: updated results of the CLL8 trial. *Blood*. 2016;127(2):208-215. Doi:10.1182/blood-2015-06-651125.
75. Robak T, Dmoszynska A, Solal-Céligny P, et al. Rituximab plus fludarabine and cyclophosphamide prolongs progression-free survival compared with fludarabine and cyclophosphamide alone in previously treated chronic lymphocytic leukemia. *J Clin Oncol*. 2010 Apr 1;28(10):1756-65.
76. Stone JH, Merkel PA, Spiera R, et al. Rituximab versus cyclophosphamide for ANCA-associated vasculitis. *N Engl J Med*. 2010;363(3):221-232. Doi:10.1056/NEJMoa0909905.
77. Guillevin L, Pagnoux C, Karras A, et al. Rituximab versus azathioprine for maintenance in ANCA-associated vasculitis. *N Engl J Med*. 2014;371(19):1771-1780. Doi:10.1056/NEJMoa1404231.
78. Niles JL, Merkel PA, Mertz L, et al. Long-Term Safety of Rituximab in Granulomatosis with Polyangiitis or Microscopic Polyangiitis: Results of the Four-Year Study of Rituximab in ANCA-Associated Vasculitis Registry [abstract]. *Arthritis Rheumatol*. 2018; 70 (suppl 10).
79. Brogan P, Cleary G, Hersh AO, et al. Pediatric Open-Label Clinical Study of Rituximab for the Treatment of Granulomatosis with Polyangiitis (GPA) and Microscopic Polyangiitis (MPA) [abstract]. *Arthritis Rheumatol*. 2018; 70 (suppl 10).
80. Joly P, Maho-Vaillant M, Prost-Squarcioni C, et al. First-line rituximab combined with short-term prednisone versus prednisone alone for the treatment of pemphigus (Ritux 3): a prospective, multicentre, parallel-group, open-label randomised trial. *Lancet*. 2017;389(10083):2031-2040. Doi:10.1016/S0140-6736(17)30070-3.
81. Thomas DA, O'Brien S, Faderl S, et al. Chemoimmunotherapy with a modified hyper-CVAD and rituximab regimen improves outcome in de novo Philadelphia chromosome-negative precursor B-lineage acute lymphoblastic leukemia. *J Clin Oncol*. 2010;28(24):3880-3889. Doi:10.1200/JCO.2009.26.9456.
82. Kadia TM, Kantarjian HM, Thomas DA, et al. Phase II study of methotrexate, vincristine, pegylated-asparaginase, and dexamethasone (MopAD) in patients with relapsed/refractory acute lymphoblastic leukemia. *Am J Hematol*. 2015;90(2):120-124. Doi:10.1002/ajh.23886.
83. Goldman S, Smith L, Anderson JR, et al. Rituximab and FAB/LMB 96 chemotherapy in children with Stage III/IV B-cell non-Hodgkin lymphoma: a Children's Oncology Group report. *Leukemia*. 2013;27(5):1174-1177. Doi:10.1038/leu.2012.255.
84. Griffin TC, Weitzman S, Weinstein H, et al. A study of rituximab and ifosfamide, carboplatin, and etoposide chemotherapy in children with recurrent/refractory B-cell (CD20+) non-Hodgkin lymphoma and mature B-cell acute lymphoblastic leukemia: a report from the Children's Oncology Group. *Pediatr Blood Cancer*. 2009;52(2):177-181. Doi:10.1002/pbc.21753.

85. Choquet S, Leblond V, Herbrecht R, et al. Efficacy and safety of rituximab in B-cell post-transplantation lymphoproliferative disorders: results of a prospective multicenter phase 2 study. *Blood*. 2006;107(8):3053-3057. Doi:10.1182/blood-2005-01-0377.
86. Trappe R, Oertel S, Leblond V, et al. Sequential treatment with rituximab followed by CHOP chemotherapy in adult B-cell post-transplant lymphoproliferative disorder (PTLD): the prospective international multicentre phase 2 PTLD-1 trial. *Lancet Oncol*. 2012;13(2):196-206.
87. Ghobrial IM, Hong F, Padmanabhan S, et al. Phase II trial of weekly bortezomib in combination with rituximab in relapsed or relapsed and refractory Waldenstrom macroglobulinemia. *J Clin Oncol*. 2010;28(8):1422-1428. Doi:10.1200/JCO.2009.25.3237.
88. Advani RH, Horning SJ, Hoppe RT, et al. Mature results of a phase II study of rituximab therapy for nodular lymphocyte-predominant Hodgkin lymphoma. *J Clin Oncol*. 2014;32(9):912-918. Doi:10.1200/JCO.2013.53.2069.
89. Godeau B, Porcher R, Fain O, et al. Rituximab efficacy and safety in adult splenectomy candidates with chronic immune thrombocytopenic purpura: results of a prospective multicenter phase 2 study. *Blood*. 2008;112(4):999-1004. Doi:10.1182/blood-2008-01-131029.
90. Wingerchuk DM, Banwell B, Bennett JL, et al. International consensus diagnostic criteria for neuromyelitis optica spectrum disorders. *Neurology*. 2015 Jul;85(2):177-89. Epub 2015 Jun 19.
91. Trebst C, Jarius S, Berthele A, et al. Update on the diagnosis and treatment of neuromyelitis optica: recommendations of the Neuromyelitis Optica Study Group (NEMOS). *J Neurol* 2014; 261:1.
92. Nikoo Z, Badihian S, Shaygannejad V, et al. Comparison of the efficacy of azathioprine and rituximab in neuromyelitis optica spectrum disorder: a randomized clinical trial. *J Neurol*. 2017;264(9):2003. Epub 2017 Aug 22.
93. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Acute Lymphoblastic Leukemia, Version 2.2025. National Comprehensive Cancer Network, 2026. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc. To view the most recent and complete version of the Guidelines, go online to NCCN.org. Accessed March 2026.
94. Thomas DA, O'Brien S, Bueso-Ramos C, et al. Rituximab in relapsed or refractory hairy cell leukemia. *Blood*. 2003 Dec 1;102(12):3906-11. Doi: 10.1182/blood-2003-02-0630.
95. Nieva J, Bethel K, Saven A. Phase 2 study of rituximab in the treatment of cladribine-failed patients with hairy cell leukemia. *Blood*. 2003 Aug 1;102(3):810-3.
96. Chihara D, Kantarjian H, O'Brien S, et al. Long-term durable remission by cladribine followed by rituximab in patients with hairy cell leukaemia: update of a phase II trial. *Br J Haematol*. 2016 Sep;174(5):760-6.
97. Else M, Dearden CE, Matutes E, et al. Rituximab with pentostatin or cladribine: an effective combination treatment for hairy cell leukemia after disease recurrence. *Leuk Lymphoma*. 2011 Jun;52 Suppl 2:75-8. Doi: 10.3109/10428194.2011.568650.

98. Zenhäusern R, Simcock M, Gratwohl A, et al; Swiss Group for Clinical Cancer Research (SAKK). Rituximab in patients with hairy cell leukemia relapsing after treatment with 2-chlorodeoxyadenosine (SAKK 31/98). *Haematologica*. 2008 Sep;93(9):1426-8.
99. Birgens H, Frederiksen H, Hasselbalch HC, et al. A phase III randomized trial comparing glucocorticoid monotherapy versus glucocorticoid and rituximab in patients with autoimmune haemolytic anaemia. *Br J Haematol*. 2013 Nov;163(3):393-9. Doi: 10.1111/bjh.12541.
100. Niederwieser, D., Hamm, C., Cobb, P. et al. Efficacy and Safety of ABP 798: Results from the JASMINE Trial in Patients with Follicular Lymphoma in Comparison with Rituximab Reference Product. *Targ Oncol* 15, 599–611 (2020). <https://doi.org/10.1007/s11523-020-00748-4>.
101. Burmester, G., Drescher, E., Hrycaj, P. et al. Efficacy and safety results from a randomized double-blind study comparing proposed biosimilar ABP 798 with rituximab reference product in subjects with moderate-to-severe rheumatoid arthritis. *Clin Rheumatol* 39, 3341–3352 (2020). <https://doi.org/10.1007/s10067-020-05305-y>.
102. Solimando AG, Crudele L, Leone P, et al. Immune Checkpoint Inhibitor-Related Myositis: From Biology to Bedside. *Int J Mol Sci*. 2020;21(9):3054. Published 2020 Apr 26. Doi:10.3390/ijms21093054.
103. Kong SS, Chen YJ, Su IC, et al; CHEESE Study Group. Immunotherapy for anti-NMDA receptor encephalitis: Experience from a single center in Taiwan. *Pediatr Neonatol*. 2019 Aug;60(4):417-422. Doi: 10.1016/j.pedneo.2018.10.006.
104. Feng S, Coward J, McCaffrey E, et al. Pembrolizumab-Induced Encephalopathy: A Review of Neurological Toxicities with Immune Checkpoint Inhibitors. *J Thorac Oncol*. 2017 Nov;12(11):1626-1635. Doi: 10.1016/j.jtho.2017.08.007.
105. Chamberlain MC, Johnston SK, Van Horn A, et al. Recurrent lymphomatous meningitis treated with intra-CSF rituximab and liposomal ara-C. *J Neurooncol*. 2009 Feb;91(3):271-7. Doi: 10.1007/s11060-008-9707-1. Epub 2008 Sep 27.
106. Rituximab in the treatment of Rosai-Dorfman syndrome with IgG4 disease. *Journal of the American Academy of Dermatology* 2019; 81: AB269.
107. Abla O, Jacobsen E, Picarsic J, et al. Consensus recommendations for the diagnosis and clinical management of Rosai-Dorfman-DeStombes disease. *Blood* (2018) 131 (26): 2877–2890.
108. Maury S, Chevret S, Thomas X, et al; for GRAALL. Rituximab in B-Lineage Adult Acute Lymphoblastic Leukemia. *N Engl J Med*. 2016 Sep 15;375(11):1044-53. Doi: 10.1056/NEJMoa1605085.
109. Wieduwilt MJ, Jonas BA, Schiller GJ, et al; A Phase II Study of Pegylated Asparaginase, Cyclophosphamide, Rituximab, and Dasatinib Added to the UCSF 8707 (Linker 4-drug) Regimen with Liposomal Cytarabine CNS Prophylaxis for Adults with Newly Diagnosed Acute Lymphoblastic Leukemia (ALL) or Lymphoblastic Lymphoma (LBL): University of California Hematologic Malignancies Consortium Study (UCHMC) 1401. *Blood* 2018; 132 (Supplement 1): 4018. Doi: <https://doi.org/10.1182/blood-2018-99-117469>.

110. Thomas DA, Faderl S, O'Brien S, et al. Chemoimmunotherapy with hyper-CVAD plus rituximab for the treatment of adult Burkitt and Burkitt-type lymphoma or acute lymphoblastic leukemia. *Cancer*, 106: 1569-1580. <https://doi.org/10.1002/cncr.21776>.
111. Kreitman RJ, Wilson W, Calvo KR, et al. Cladribine with immediate rituximab for the treatment of patients with variant hairy cell leukemia. *Clin Cancer Res*. 2013 Dec 15;19(24):6873-81. Doi: 10.1158/1078-0432.CCR-13-1752.
112. Fraenkel L, Bathon JM, England BR, et al. 2021 American College of Rheumatology Guideline for the Treatment of Rheumatoid Arthritis. *Arthritis Rheumatol*. 2021 Jul;73(7):1108-1123. Doi: 10.1002/art.41752.
113. Smolen JS, Landewé RBM, Bijlsma JWW, et al. EULAR recommendations for the management of rheumatoid arthritis with synthetic and biological disease-modifying antirheumatic drugs: 2019 update. *Annals of the Rheumatic Diseases* 2020;79:685-699.
114. Venugopal SS, Murrell DF. Diagnosis and clinical features of pemphigus vulgaris. *Dermatol Clin*. 2011 Jul;29(3):373-80, vii. Doi: 10.1016/j.det.2011.03.004. PMID: 21605802.
115. Fanouriakis A, Kostopoulou M, Cheema K, et al: 2019 update of the Joint European League Against Rheumatism and European Renal Association-European Dialysis and Transplant Association (EULAR/ERA-EDTA) recommendations for the management of lupus nephritis. *Ann Rheum Dis* 2020; 79(6):713-723.
116. Vigna-Perez M, Hernández-Castro B, Paredes-Saharopulos O, et al. Clinical and immunological effects of Rituximab in patients with lupus nephritis refractory to conventional therapy: a pilot study. *Arthritis Res Ther*. 2006;8(3):R83. Doi: 10.1186/ar1954. Epub 2006 May 5.
117. Melander C, Sallée M, Trolliet P, et al. Rituximab in severe lupus nephritis: early B-cell depletion affects long-term renal outcome. *Clin J Am Soc Nephrol*. 2009 Mar;4(3):579-87. Doi: 10.2215/CJN.04030808. Epub 2009 Mar 4.
118. Bird SJ. (2026). Chronic immunotherapy for myasthenia gravis. In Shefner JM, Goddeau RP (Eds.), *UptoDate*. Last updated: January 06, 2026. Accessed: March 2026. Available from <https://www.uptodate.com/contents/chronic-immunotherapy-for-myasthenia-gravis>.
119. Topakian R, Zimprich F, Iglseder S, et al. High efficacy of rituximab for myasthenia gravis: a comprehensive nationwide study in Austria. *J Neurol*. 2019;266(3):699-706. Doi:10.1007/s00415-019-09191-6.
120. Li T, Zhang GQ, Li Y, et al. Efficacy and safety of different dosages of rituximab for refractory generalized AchR myasthenia gravis: a meta-analysis. *J Clin Neurosci*. 2021;85:6-12. Doi:10.1016/j.jocn.2020.11.043.
121. Colin V, Auperin A, Pilon M, et al. Rituximab for High-Risk, Mature B-Cell Non-Hodgkin's Lymphoma in Children. *Clinical Trial*. *N Engl J Med*. 2020 Jun 4;382(23):2207-2219. Doi: 10.1056/NEJMoa1915315.
122. Furman RR, Sharman JP, Coutre SE, et al. Idelalisib and rituximab in relapsed chronic lymphocytic leukemia. *N Engl J Med*. 2014;370(11):997-1007. Doi:10.1056/NEJMoa1315226.
123. Greil R, Obřtlíková P, Smolej L, et al. Rituximab maintenance versus observation alone in patients with chronic lymphocytic leukaemia who respond to first-line or second-line rituximab-

- containing chemoimmunotherapy: final results of the AGMT CLL-8a Maintenance randomised trial. *Lancet Haematol*. 2016 Jul;3(7):e317-29. Doi: 10.1016/S2352-3026(16)30045-X.
124. Dartigeas C, Van Den Neste E, Léger J, et al. Rituximab maintenance versus observation following abbreviated induction with chemoimmunotherapy in elderly patients with previously untreated chronic lymphocytic leukaemia (CLL 2007 SA): an open-label, randomised phase 3 study. *Lancet Haematol*. 2018 Feb;5(2):e82-e94. Doi: 10.1016/S2352-3026(17)30235-1.
 125. Chung SA, Langford CA, Maz M, et al. 2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Antineutrophil Cytoplasmic Antibody-Associated Vasculitis. *Arthritis Care Res (Hoboken)*. 2021;73(8):1088. Epub 2021 Jul 8.
 126. Zheng XL, Vesely SK, Cataland SR, et al. ISTH guidelines for treatment of thrombotic thrombocytopenic purpura. *J Thromb Haemost*. 2020;18(10):2496-2502. Doi:10.1111/jth.15010.
 127. Michel M, Chanut V, Dechartres A, et al. The spectrum of Evans syndrome in adults: new insight into the disease based on the analysis of 68 cases. *Blood*. 2009 Oct 8;114(15):3167-72. Doi: 10.1182/blood-2009-04-215368.
 128. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Pediatric Hodgkin Lymphoma, Version 2.2025. National Comprehensive Cancer Network, 2026. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc. To view the most recent and complete version of the Guidelines, go online to NCCN.org. Accessed March 2026.
 129. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Hematopoietic Cell Transplantation, Version 3.2025. National Comprehensive Cancer Network, 2026. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc. To view the most recent and complete version of the Guidelines, go online to NCCN.org. Accessed March 2026.
 130. Khouri IF, Saliba RM, Giralt SA, et al. Nonablative allogeneic hematopoietic transplantation as adoptive immunotherapy for indolent lymphoma: low incidence of toxicity, acute graft-versus-host disease, and treatment-related mortality. *Blood*. 2001 Dec 15;98(13):3595-9. Doi: 10.1182/blood.v98.13.3595.
 131. Kharfan-Dabaja MA, Mhaskar AR, Djulbegovic B, et al. Efficacy of rituximab in the setting of steroid-refractory chronic graft-versus-host disease: a systematic review and meta-analysis. *Biol Blood Marrow Transplant*. 2009 Sep;15(9):1005-13. Doi: 10.1016/j.bbmt.2009.04.003.
 132. Rovin BH, Furie R, Latinis K, et al; LUNAR Investigator Group. Efficacy and safety of rituximab in patients with active proliferative lupus nephritis: the Lupus Nephritis Assessment with Rituximab study. *Arthritis Rheum*. 2012 Apr;64(4):1215-26. Doi: 10.1002/art.34359.
 133. McDonald RA. (2025). Kidney transplantation in children: Immunosuppression. In Niaudet P, Kremen J (Eds.), *UptoDate*. Last updated: December 10, 2025 Accessed: March 2026. Available from <https://www.uptodate.com/contents/kidney-transplantation-in-children-immunosuppression>.
 134. Parajuli S, Brennan DC. (2025). Kidney transplantation in adults: Prevention and treatment of antibody-mediated rejection. In Legendre C, Vella J, Lam AQ (Eds.), *UptoDate*. Last updated:

September 24, 2025. Accessed: March 2026. Available from <https://www.uptodate.com/contents/kidney-transplantation-in-adults-prevention-and-treatment-of-antibody-mediated-rejection>.

135. Colvin MM, Cook JL, Chang P, et al. Antibody-mediated rejection in cardiac transplantation: emerging knowledge in diagnosis and management: a scientific statement from the American Heart Association. *Circulation*. 2015 May 5;131(18):1608-39. Doi: 10.1161/CIR.0000000000000093.
136. Hachem RR. (2024). Evaluation and treatment of antibody-mediated lung transplant rejection. In Kotloff RM, Dieffenbach P (Eds.), *UptoDate*. Last updated: Jun 27, 2024. Accessed: March 2026. Available from <https://www.uptodate.com/contents/evaluation-and-treatment-of-antibody-mediated-lung-transplant-rejection>.
137. Alhamad T, Kukla A, Stratta RJ. (2024). Pancreas allograft rejection. In Brennan DC, Nathan DM, Lam AQ (Eds.), *UptoDate*. Last updated: October 31, 2024. Accessed: March 2026. Available from <https://www.uptodate.com/contents/pancreas-allograft-rejection>.
138. Sakamoto S, Akamatsu N, Hasegawa K, et al. The efficacy of rituximab treatment for antibody-mediated rejection in liver transplantation: A retrospective Japanese nationwide study. *Hepatol Res*. 2021 Sep;51(9):990-999. Doi: 10.1111/hepr.13643.
139. Joly P, Horvath B, Patsatsi A, et al. Updated S2K guidelines on the management of pemphigus vulgaris and foliaceus initiated by the European academy of dermatology and venereology (EADV). *Journal of the European Academy of Dermatology & Venereology*. 2020 Sept; 34(9):1900-1913.
140. Senff NJ, Noordijk EM, Kim YH, et al.; European Organization for Research and Treatment of Cancer; International Society for Cutaneous Lymphoma. European Organization for Research and Treatment of Cancer and International Society for Cutaneous Lymphoma consensus recommendations for the management of cutaneous B-cell lymphomas. *Blood*. 2008 Sep 1;112(5):1600-9. Doi: 10.1182/blood-2008-04-152850. Epub 2008 Jun 20. PMID: 18567836.
141. Gawronski KM, Rainka MM, Patel MJ, Gengo FM. Treatment Options for Multiple Sclerosis: Current and Emerging Therapies. *Pharmacotherapy*. 2010; 30(9):916-927.
142. Goodin DS, Frohman EM, Garmany GP Jr, et al. Disease modifying therapies in multiple sclerosis: report of the Therapeutics and Technology Assessment Subcommittee of the American Academy of Neurology and the MS Council for Clinical Practice Guidelines. *Neurology*. 2002 Jan 22; 58(2):169-78.
143. Freedman MS, Selchen D, Arnold DL, et al. Treatment optimization in MS: Canadian MS Working Group updated recommendations. *Can J Neurol Sci*. 2013 May;40(3):307-23.
144. Polman CH, Reingold SC, Banwell B, et al. Diagnostic criteria for multiple sclerosis: 2010 Revisions to the McDonald criteria. *Ann Neurol*. 2011 Feb; 69(2): 292–302. doi: 10.1002/ana.22366.
145. Lublin FD, Reingold SC, Cohen JA, et al. Defining the clinical course of multiple sclerosis: the 2013 revisions. *Neurology*. 2014 Jul 15;83(3):278-86.

146. Multiple Sclerosis Coalition. The use of disease-modifying therapies in multiple sclerosis: principles and current evidence. 2017 March.
http://www.nationalmssociety.org/getmedia/5ca284d3-fc7c-4ba5-b005-ab537d495c3c/DMT_Consensus_MS_Coalition_color. Accessed 4/2018.
147. Rae-Grant, A, Day GS, Marrie RA, et al. Practice guideline recommendations summary: Disease-modifying therapies for adults with multiple sclerosis. Report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology. *Neurology*® 2018;90:777-788.
148. Thompson AJ, Banwell BL, Barkhof F, et al. Diagnosis of multiple sclerosis: 2017 revisions of the McDonald criteria. *Lancet Neurol*. 2018 Feb;17(2):162-173. doi: 10.1016/S1474-4422(17)30470-2.
149. Kappos L, Bar-Or A, Cree BAC, et al. Siponimod versus placebo in secondary progressive multiple sclerosis (EXPAND): a double-blind, randomised, phase 3 study. *Lancet*. 2018;391(10127):1263. Epub 2018 Mar 23.
150. Lorscheider J, Buzzard K, Jokubaitis V, et al, on behalf of the MSBase Study Group. Defining secondary progressive multiple sclerosis. *Brain*, Volume 139, Issue 9, September 2016, Pages 2395–2405, <https://doi.org/10.1093/brain/aww173>.
151. Freedman MS, Devonshire V, Duquette P, et al; Canadian MS Working Group. Treatment Optimization in Multiple Sclerosis: Canadian MS Working Group Recommendations. *Can J Neurol Sci*. 2020 Jul;47(4):437-455. doi: 10.1017/cjn.2020.66.
152. Miche M, Terriou L, Roudot-Thoraval F, et al. A randomized and double-blind controlled trial evaluating the safety and efficacy of rituximab for warm auto-immune hemolytic anemia in adults (the RAIHA study). *Am J Hematol*. 2017;92:23-27. <https://doi.org/10.1002/ajh.24570>
153. Fanouriakis A, Kostopoulou M, Alunno A, et al: 2019 update of the EULAR recommendations for the management of systemic lupus erythematosus. *Ann Rheum Dis* 2019; 78(6):736-745.
154. American College of Rheumatology Ad Hoc Committee on Systemic Lupus Erythematosus Guidelines. Guidelines for referral and management of systemic lupus erythematosus in adults. *Arthritis Rheum*. 1999;42(9):1785–1796.
155. Gordon C, Amissah-Arthur MB, Gayed M, et al. The British Society for Rheumatology guideline for the management of systemic lupus erythematosus in adults. *Rheumatology (Oxford)*. 2018 Jan 1;57(1):e1-e45. doi: 10.1093/rheumatology/kex286.
156. Narayanaswami P, Sanders DB, Wolfe G, et al. International Consensus Guidance for Management of Myasthenia Gravis: 2020 Update. *Neurology*. 2021 Jan 19;96(3):114-122. doi: 10.1212/WNL.0000000000011124.
157. Jarius, S., Aktas, O., Ayzenberg, I. et al. Update on the diagnosis and treatment of neuromyelitis optica spectrum disorders (NMOSD) – revised recommendations of the Neuromyelitis Optica Study Group (NEMOS). Part I: Diagnosis and differential diagnosis. *J Neurol* 270, 3341–3368 (2023). <https://doi.org/10.1007/s00415-023-11634-0>

158. Fanouriakis A, Tziolos N, Bertsias G, et al. Update on the diagnosis and management of systemic lupus erythematosus. *Ann Rheum Dis* 2021; 80:14-25. doi:10.1136/annrheumdis-2020-218272
159. Aringer M, Costenbader K, Daikh D, et al. 2019 European League against Rheumatism/American College of rheumatology classification criteria for systemic lupus erythematosus. *Arthritis Rheumatol* 2019;71:1400–12.
160. Lam NC, Brown JA, Sharma R. Systemic Lupus Erythematosus: Diagnosis and Treatment. *Am Fam Physician*. 2023 Apr 107(4):383-395.
161. Yee CS, Cresswell L, Farewell V, Rahman A, Teh LS, Griffiths B et al. Numerical scoring for the BILAG-2004 index. *Rheumatology (Oxford)* 2010; 49(9):1665-9.
162. Gladman DD, Ibanez D, Urowitz MB. Systemic lupus erythematosus disease activity index 2000. *J Rheumatol* 2002; 29(2):288-91.
163. Chessa E, Piga M, Floris A, Devilliers H, Cauli A, Arnaud L. Use of Physician Global Assessment in systemic lupus erythematosus: a systematic review of its psychometric properties. *Rheumatology (Oxford)*. 2020 Dec 1;59(12):3622-3632. doi: 10.1093/rheumatology/keaa383.
164. Tiacchi E, De Carolis L, Santi A, Falini B. Venetoclax in relapsed or refractory hairy-cell leukemia. *N Engl J Med* 2023;388:952-954.
165. Kümpfel T, Giglhuber K, Aktas O, et al. Neuromyelitis Optica Study Group (NEMOS). Update on the diagnosis and treatment of neuromyelitis optica spectrum disorders (NMOSD) - revised recommendations of the Neuromyelitis Optica Study Group (NEMOS). Part II: Attack therapy and long-term management. *J Neurol*. 2023 Sep 7. doi: 10.1007/s00415-023-11910-z. Epub ahead of print.
166. Kidney Disease: Improving Global Outcomes (KDIGO) Lupus Nephritis Work Group. KDIGO 2024 Clinical Practice Guideline for the Management of Lupus Nephritis. *Kidney Int*. 2024;105(1S):S1–S69.)
167. Doyle TJ, Dhillion N, Madan R, et al. Rituximab in the Treatment of Interstitial Lung Disease Associated with Antisynthetase Syndrome: A Multicenter Retrospective Case Review. *J Rheumatol*. 2018 Jun;45(6):841-850. doi: 10.3899/jrheum.170541.
168. Anderson H, Sem M, Lund M, et al. Long-term experience with rituximab in anti-synthetase syndrome-related interstitial lung disease, *Rheumatology*, Volume 54, Issue 8, August 2015, Pages 1420–1428. doi.org/10.1093/rheumatology/kev004
169. Fanouriakis A, Kostopoulou M, Andersen J, et al. EULAR recommendations for the management of systemic lupus erythematosus: 2023 update. *Ann Rheum Dis*. 2024 Jan 2;83(1):15-29. doi: 10.1136/ard-2023-224762.
170. Trautmann A, Boyer O, Hodson E, et al. International Pediatric Nephrology Association. IPNA clinical practice recommendations for the diagnosis and management of children with steroid-sensitive nephrotic syndrome. *Pediatr Nephrol*. 2023 Mar;38(3):877-919. doi: 10.1007/s00467-022-05739-3.

171. Trautmann A, Vivarelli M, Samuel S, et al. International Pediatric Nephrology Association. IPNA clinical practice recommendations for the diagnosis and management of children with steroid-resistant nephrotic syndrome. *Pediatr Nephrol*. 2020 Aug;35(8):1529-1561. doi: 10.1007/s00467-020-04519-1.
172. Kidney Disease: Improving Global Outcomes (KDIGO) Glomerular Diseases Work Group. KDIGO 2021 Clinical Practice Guideline for the Management of Glomerular Diseases. *Kidney Int*. 2021 Oct;100(4S):S1-S276. doi: 10.1016/j.kint.2021.05.021.
173. Kallash M, Smoyer WE, Mahan JD. Rituximab Use in the Management of Childhood Nephrotic Syndrome. *Front Pediatr*. 2019 May 10;7:178. doi: 10.3389/fped.2019.00178.
174. Sawal N, Mukhopadhyay S, Rayancha S, et al. A narrative review of interstitial lung disease in anti-synthetase syndrome: a clinical approach. *J Thorac Dis*. 2021 Sep;13(9):5556-5571. doi: 10.21037/jtd-20-3328. PMID: 34659821; PMCID: PMC8482343.
175. Fervenza FC, Appel GB, Barbour SJ, et al. Rituximab or Cyclosporine in the Treatment of Membranous Nephropathy. *N Engl J Med*. 2019 Jul 4;381(1):36-46. doi: 10.1056/NEJMoa1814427.
176. Beck LH, Salant DJ. (2025). Membranous nephropathy: Pathogenesis and etiology. In Glassock RJ, Fervenza FC, Lam AQ (Eds.), *UptoDate*. Last updated: February 04, 2026. Accessed: March 2026. Available from <https://www.uptodate.com/contents/membranous-nephropathy-pathogenesis-and-etiology>
177. De Vriese AS, Wetzels JFM, Cattran DC. (2025). Membranous nephropathy: Treatment and prognosis. In Glassock RJ, Fervenza FC, Lam AQ (Eds.), *UptoDate*. Last updated: September 19, 2025. Accessed: March 2026. Available from <https://www.uptodate.com/contents/membranous-nephropathy-treatment-and-prognosis>
178. Khosroshahi A, Wallace ZS, Crowe JL, et al. International Consensus Guidance Statement on the Management and Treatment of IgG4-Related Disease. *Arthritis Rheumatol*. 2015;67(7):1688-1699. doi:10.1002/art.39132
179. Ebbo M, Grados A, Samson M, et al. Long-term efficacy and safety of rituximab in IgG4-related disease: Data from a French nationwide study of thirty-three patients. *PLoS One*. 2017;12(9):e0183844. Published 2017 Sep 15. doi:10.1371/journal.pone.0183844
180. Carruthers MN, Topazian MD, Khosroshahi A, et al. Rituximab for IgG4-related disease: a prospective, open-label trial. *Ann Rheum Dis*. 2015;74(6):1171-1177.
181. Khosroshahi A, Bloch DB, Deshpande V, Stone JH. Rituximab therapy leads to rapid decline of serum IgG4 levels and prompt clinical improvement in IgG4-related systemic disease. *Arthritis Rheum*. 2010;62(6):1755-1762.
182. Khosroshahi A, Carruthers MN, Deshpande V, Unizony S, Bloch DB, Stone JH. Rituximab for the treatment of IgG4-related disease: lessons from 10 consecutive patients. *Medicine (Baltimore)*. 2012;91(1):57-66.
183. Ly KI, Crew LL, Graham CA, Mrugala MM. Primary central nervous system lymphoma treated with high-dose methotrexate and rituximab: A single-institution experience. *Oncol Lett*. 2016;11:3471-3476.

184. Smith R, Jones R, Guerry M, et al. Rituximab for remission maintenance in relapsing antineutrophil cytoplasmic antibody-associated vasculitis. *Arthritis Rheum.* 2012 Nov;64(11):3760-9. doi: 10.1002/art.34583.
185. Reiss SN, Yerram P, Modelevsky L, Grommes C. Rituximab, methotrexate, carmustine, etoposide, and prednisone (RMBVP) for the treatment of relapsed/ refractory primary central nervous system lymphoma: a retrospective single-center study. *Leuk Lymphoma* 2022;63:627-632
186. Narváez J, Cañadillas E, Castellví I, et al. (2024). Rituximab in the treatment of progressive interstitial lung disease associated with the antisynthetase syndrome. *Arthritis Research & Therapy*, 26(1), 122.
187. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Temozolomide + Rituximab: Central Nervous System Cancers Chemotherapy Order Template, CNS69. National Comprehensive Cancer Network, 2026. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc. To view the most recent and complete version of the Guidelines, go online to NCCN.org. Accessed March 2026.
188. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for High-Dose Methotrexate/Leucovorin/Ibrutinib + Rituximab: Central Nervous System Cancers Chemotherapy Order Template, CNS82. National Comprehensive Cancer Network, 2026. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc. To view the most recent and complete version of the Guidelines, go online to NCCN.org. Accessed March 2026.
189. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for HDMP (High-Dose Methylprednisolone) + Rituximab: Chronic Lymphocytic Leukemia/Small Lymphocytic Lymphoma Chemotherapy Order Template, CLL25. National Comprehensive Cancer Network, 2026. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc. To view the most recent and complete version of the Guidelines, go online to NCCN.org. Accessed March 2026.
190. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Carfilzomib/Dexamethasone + Rituximab: Waldenström Macroglobulinemia/Lymphoplasmacytic Lymphoma Chemotherapy Order Template, WAL24. National Comprehensive Cancer Network, 2026. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc. To view the most recent and complete version of the Guidelines, go online to NCCN.org. Accessed March 2026.
191. Smolen JS, Landewé RBM, Bergstra SA, et al. EULAR recommendations for the management of rheumatoid arthritis with synthetic and biological disease-modifying antirheumatic drugs: 2022 update. *Ann Rheum Dis.* 2023 Jan;82(1):3-18. doi: 10.1136/ard-2022-223356. Epub 2022 Nov

10. Erratum in: *Ann Rheum Dis*. 2023 Mar;82(3):e76. doi: 10.1136/ard-2022-223356corr1. PMID: 36357155.
192. Webber S, Harmon W, Faro A, et al. Anti-CD20 Monoclonal Antibody (rituximab) for Refractory PTLT after Pediatric Solid Organ Transplantation: Multicenter Experience from a Registry and from a Prospective Clinical Trial [abstract]. *Blood* 2004;104:Abstract 746.
193. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Lenalidomide + Brentuximab vedotin + Rituximab: Diffuse Large B-Cell Lymphoma Chemotherapy Order Template, DBL89. National Comprehensive Cancer Network, 2026. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc. To view the most recent and complete version of the Guidelines, go online to NCCN.org. Accessed March 2026.
194. Kremer, N., Snast, I., Cohen, E.S. et al. Rituximab and Omalizumab for the Treatment of Bullous Pemphigoid: A Systematic Review of the Literature. *Am J Clin Dermatol* **20**, 209–216 (2019). <https://doi.org/10.1007/s40257-018-0401-6>
195. Moutsopoulos HM, Fragoulis GE, Stone JH (2025). Treatment and prognosis of IgG4-related disease. In Helfgott SM, Seo P (Eds.), *UptoDate*. Last updated: April 15, 2025. Accessed: March 2026. Available from: <https://www.uptodate.com/contents/treatment-and-prognosis-of-igg4-related-disease>
196. Zheng XL, Al-Housni Z, Cataland SR, et al. 2025 focused update of the 2020 ISTH guidelines for management of thrombotic thrombocytopenic purpura. *J Thromb Haemost*. Published online June 17, 2025. doi:10.1016/j.jtha.2025.06.002
197. Floege J, Gibson KL, et al. KDIGO 2025 Clinical Practice Guideline for the Management of Nephrotic Syndrome in Children. *Kidney Int*. 2025;107(5S):S241-S289. doi:10.1016/j.kint.2024.11.007
198. Carreras E, Diaz-Ricart M, Jodele S, et al. Early Complications of Endothelial Origin. 2024 Apr 11. In: Sureda A, Corbacioglu S, Greco R, et al., editors. *The EBMT Handbook: Hematopoietic Cell Transplantation and Cellular Therapies* [Internet]. 8th edition. Cham (CH): Springer; 2024. Chapter 42. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK608292/> doi: 10.1007/978-3-031-44080-9_42
199. Stavrou E, Lazarus HM. Thrombotic microangiopathy in haematopoietic cell transplantation: an update. *Mediterr J Hematol Infect Dis*. 2010;2(3):e2010033. doi: 10.4084/MJHID.2010.033. Epub 2010 Nov 3. PMID: 21776339; PMCID: PMC3134219.
200. Holdhoff M, Ambady P, Abdelaziz A, et al. High-dose methotrexate with or without rituximab in newly diagnosed primary CNS lymphoma. *Neurology*. 2014 Jul 15;83(3):235-9. doi: 10.1212/WNL.0000000000000593. Epub 2014 Jun 13. PMID: 24928128; PMCID: PMC4117362.
201. Gonçalves PH, Uldrick TS, Yarchoan R. HIV-associated Kaposi sarcoma and related diseases. *AIDS*. 2017 Sep 10;31(14):1903-1916. doi: 10.1097/QAD.0000000000001567. PMID: 28609402; PMCID: PMC6310482.
202. Biddle K, Jade J, Wilson-Morkeh H, et al. British Society for Rheumatology Guideline Steering Group, The 2025 British Society for Rheumatology management recommendations for ANCA-

- associated vasculitis, *Rheumatology*, Volume 64, Issue 8, August 2025, Pages 4470–4494, <https://doi.org/10.1093/rheumatology/keaf240>.
203. Montalban X, Lebrun-Fréney C, Oh J, et al. Diagnosis of multiple sclerosis: 2024 revisions of the McDonald criteria. *Lancet Neurol*. 2025 Oct;24(10):850-865. doi: 10.1016/S1474-4422(25)00270-4.
 204. Rovin BH, Barratt J, Cook HT, et al. KDIGO 2025 Clinical Practice Guideline for the Management of Immunoglobulin A Nephropathy (IgAN) and Immunoglobulin A Vasculitis (IgAV). *Kidney Int*. 2025 Oct;108(4S):S1-S71. doi: 10.1016/j.kint.2025.04.004. PMID: 40975564.
 205. Waskyra [package insert]. Rome, Italy; Fondazione Telethon ETs; December 2025. Accessed March 2026.
 206. Chandramohan P, Jain A, Antony G, Krishnan N, Shenoy P. Low-dose rituximab protocol in rheumatoid arthritis-outcome and economic impact. *Rheumatol Adv Pract*. 2021 Jan 7;5(1):rkaa077. doi: 10.1093/rap/rkaa077. PMID: 33605940; PMCID: PMC7878847.
 207. Boremalm M, Sundström P, Salzer J. Discontinuation and dose reduction of rituximab in relapsing-remitting multiple sclerosis. *J Neurol*. 2021 Jun;268(6):2161-2168. doi: 10.1007/s00415-021-10399-8. Epub 2021 Jan 21. PMID: 33475825; PMCID: PMC7818716.
 208. Styczynski J, Gil L, Tridello G, et al. Infectious Diseases Working Party of the European Group for Blood and Marrow Transplantation, Response to Rituximab-Based Therapy and Risk Factor Analysis in Epstein Barr Virus–Related Lymphoproliferative Disorder After Hematopoietic Stem Cell Transplant in Children and Adults: A Study From the Infectious Diseases Working Party of the European Group for Blood and Marrow Transplantation, *Clinical Infectious Diseases*, Volume 57, Issue 6, 15 September 2013, Pages 794–802, <https://doi.org/10.1093/cid/cit391>
 209. Styczynski J, Giebel S. Posttransplant Lymphoproliferative Syndromes. 2024 Apr 11. In: Sureda A, Corbacioglu S, Greco R, et al., editors. *The EBMT Handbook: Hematopoietic Cell Transplantation and Cellular Therapies* [Internet]. 8th edition. Cham (CH): Springer; 2024. Chapter 45. Available from: https://link.springer.com/chapter/10.1007/978-3-031-44080-9_45
 210. National Government Services, Inc. Local Coverage Article: Billing and Coding: Off-label Use of Rituximab and Rituximab Biosimilars (A59101). Centers for Medicare & Medicaid Services, Inc. Updated on 09/17/2025 with effective date 10/01/2025. Accessed March 2026.
 211. Wisconsin Physicians Service Insurance Corp. Local Coverage Article: Billing and Coding: Chemotherapy Agents for Non-Oncologic Conditions (A55639). Centers for Medicare & Medicaid Services, Inc. Updated on 11/18/2025 with effective date 11/27/2025. Accessed March 2026.
 212. Palmetto GBA. Local Coverage Article: Billing and Coding: Rituximab (A56380). Centers for Medicare & Medicaid Services, Inc. Updated on 08/26/2025 with effective date 10/01/2025. Accessed March 2026.
 213. CGS Administrators, LLC. Local Coverage Article: Billing and Coding: Immune Thrombocytopenia (ITP) Therapy (A57160). Centers for Medicare & Medicaid Services, Inc. Updated on 03/09/2026 with effective date 03/05/2026. Accessed March 2026.
 214. CGS Administrators, LLC. Local Coverage Article: Billing and Coding: Off-label Use of Rituximab and Rituximab Biosimilars (A58582). Centers for Medicare & Medicaid Services, Inc. Updated on 09/18/2025 with effective date 10/01/2025. Accessed March 2026.

Appendix A – Non-Quantitative Treatment Limitations (NQTL) Factor Checklist

Non-quantitative treatment limitations (NQTLs) refer to the methods, guidelines, standards of evidence, or other conditions that can restrict how long or to what extent benefits are provided under a health plan. These may include things like utilization review or prior authorization. The utilization management NQTL applies comparably, and not more stringently, to mental health/substance use disorder (MH/SUD) Medical Benefit Prescription Drugs and medical/surgical (M/S) Medical Benefit Prescription Drugs. The table below lists the factors that were considered in designing and applying prior authorization to this drug/drug group, and a summary of the conclusions that Prime's assessment led to for each.

Factor	Conclusion
Indication	Yes: Consider for PA
Safety and efficacy	Yes: Consider for PA
Potential for misuse/abuse	No: PA not a priority
Cost of drug	Yes: Consider for PA

Appendix 1 – Covered Diagnosis Codes

ICD-10	Description
C46.0	Kaposi's sarcoma of skin
C46.1	Kaposi's sarcoma of soft tissue
C46.2	Kaposi's sarcoma of palate
C46.3	Kaposi's sarcoma of lymph nodes
C46.4	Kaposi's sarcoma of gastrointestinal sites
C46.50	Kaposi's sarcoma of unspecified lung
C46.51	Kaposi's sarcoma of right lung
C46.52	Kaposi's sarcoma of left lung
C46.7	Kaposi's sarcoma of other sites
C46.9	Kaposi's sarcoma, unspecified
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

C81.19	Nodular sclerosis Hodgkin lymphoma, extranodal and solid organ sites
C81.29	Mixed cellularity Hodgkin lymphoma, extranodal and solid organ sites
C81.39	Lymphocyte depleted Hodgkin lymphoma, extranodal and solid organ sites
C81.49	Lymphocyte-rich Hodgkin lymphoma, extranodal and solid organ sites
C81.79	Other Hodgkin lymphoma, extranodal and solid organ sites
C81.99	Hodgkin lymphoma, unspecified, 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
C83.390	Primary central nervous system lymphoma
C83.398	Diffuse large B-cell lymphoma of other extranodal and solid organ sites
C83.50	Lymphoblastic (diffuse) lymphoma, unspecified site
C83.51	Lymphoblastic (diffuse) lymphoma, lymph nodes of head, face, and neck
C83.52	Lymphoblastic (diffuse) lymphoma, intrathoracic lymph nodes
C83.53	Lymphoblastic (diffuse) lymphoma, intra-abdominal lymph nodes
C83.54	Lymphoblastic (diffuse) lymphoma, lymph nodes of axilla and upper limb
C83.55	Lymphoblastic (diffuse) lymphoma, lymph nodes of inguinal region and lower limb
C83.56	Lymphoblastic (diffuse) lymphoma, intrapelvic lymph nodes
C83.57	Lymphoblastic (diffuse) lymphoma, spleen
C83.58	Lymphoblastic (diffuse) lymphoma, lymph nodes of multiple sites
C83.59	Lymphoblastic (diffuse) 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
C84.09	Mycosis fungoides, extranodal and solid organ sites
C84.19	Sézary disease, extranodal and solid organ sites
C84.49	Peripheral T-cell lymphoma, not classified, extranodal and solid organ sites
C84.69	Anaplastic large cell lymphoma, ALK-positive, extranodal and solid organ sites
C84.79	Anaplastic large cell lymphoma, ALK-negative, extranodal and solid organ sites
C84.99	Mature T/NK-cell lymphomas, unspecified, extranodal and solid organ sites
C84.A9	Cutaneous T-cell lymphoma, unspecified, extranodal and solid organ sites
C84.Z9	Other mature T/NK-cell lymphomas, extranodal and solid organ sites
C85.10	Unspecified B-cell lymphoma, unspecified site
C85.11	Unspecified B-cell lymphoma, lymph nodes of head, face, and neck
C85.12	Unspecified B-cell lymphoma, intrathoracic lymph nodes
C85.13	Unspecified B-cell lymphoma, intra-abdominal lymph nodes
C85.14	Unspecified B-cell lymphoma, lymph nodes of axilla and upper limb
C85.15	Unspecified B-cell lymphoma, lymph nodes of inguinal region and lower limb
C85.16	Unspecified B-cell lymphoma, intrapelvic lymph nodes
C85.17	Unspecified B-cell lymphoma, spleen
C85.18	Unspecified B-cell lymphoma, lymph nodes of multiple sites

C85.19	Unspecified B-cell lymphoma, 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
C85.99	Non-Hodgkin lymphoma, unspecified, extranodal and solid organ sites
C88.00	Waldenström macroglobulinemia not having achieved remission
C88.40	Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT-lymphoma) not having achieved remission
C91.00	Acute lymphoblastic leukemia not having achieved remission
C91.01	Acute lymphoblastic leukemia, in remission
C91.02	Acute lymphoblastic leukemia, in relapse
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
D47.Z1	Post-transplant lymphoproliferative disorder (PTLD)
D47.Z2	Other neoplasms of uncertain behavior of lymphoid, hematopoietic and related tissue-Castleman disease
D59.0	Drug-induced autoimmune hemolytic anemia
D59.2	Drug-induced nonautoimmune hemolytic anemia
D59.10	Autoimmune hemolytic anemia, unspecified

D59.11	Warm autoimmune hemolytic anemia
D59.12	Cold autoimmune hemolytic anemia
D59.13	Mixed type autoimmune hemolytic anemia
D59.19	Other autoimmune hemolytic anemia
D69.3	Immune thrombocytopenic purpura
D69.41	Evans Syndrome
D69.42	Congenital and hereditary thrombocytopenia purpura
D69.49	Other primary thrombocytopenia
D69.59	Other secondary thrombocytopenia
D76.3	Other histiocytosis syndromes
D82.0	Wiskott-Aldrich syndrome
D89.811	Chronic graft-versus-host disease
D89.812	Acute on chronic graft-versus-host disease
D89.813	Graft-versus-host disease unspecified
D89.84	IgG4-related disease
D89.89	Other specified disorders involving the immune mechanism, not elsewhere classified
D89.9	Disorder involving the immune mechanism, unspecified
G04.81	Other encephalitis and encephalomyelitis
G04.89	Other myelitis
G04.90	Encephalitis and encephalomyelitis, unspecified
G35.A	Relapsing-remitting multiple sclerosis
G35.B0	Primary progressive multiple sclerosis, unspecified
G35.B1	Active primary progressive multiple sclerosis
G35.B2	Non-active primary progressive multiple sclerosis
G35.C0	Secondary progressive multiple sclerosis, unspecified
G35.C1	Active secondary progressive multiple sclerosis
G35.C2	Non-active secondary progressive multiple sclerosis
G35.D	Multiple sclerosis, unspecified
G36.0	Neuromyelitis optica [Devic]
G70.00	Myasthenia gravis without (acute) exacerbation
G70.01	Myasthenia gravis with (acute) exacerbation
J84.9	Interstitial pulmonary disease, unspecified
L10.0	Pemphigus vulgaris
L13.8	Other specified bullous disorders
L13.9	Bullous disorder, unspecified
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
M05.7A	Rheumatoid arthritis with rheumatoid factor of other specified 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
M05.8A	Other rheumatoid arthritis with rheumatoid factor of other specified 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
M06.0A	Rheumatoid arthritis without rheumatoid factor, other specified 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
M06.8A	Other specified rheumatoid arthritis, other specified 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
M31.10	Thrombotic microangiopathy, unspecified
M31.11	Hematopoietic stem cell transplantation-associated thrombotic microangiopathy [HSCT-TMA]
M31.30	Wegener's granulomatosis without renal involvement
M31.31	Wegener's granulomatosis with renal involvement
M31.7	Microscopic polyangiitis
M32.10	Systemic lupus erythematosus organ or system involvement unspecified
M32.11	Endocarditis in systemic lupus erythematosus
M32.12	Pericarditis in systemic lupus erythematosus
M32.13	Lung involvement in systemic lupus erythematosus
M32.14	Glomerular disease in systemic lupus erythematosus
M32.15	Tubulo-interstitial nephropathy in systemic lupus erythematosus
M32.19	Other organ or system involvement in systemic lupus erythematosus
M32.8	Other forms of systemic lupus erythematosus
M32.9	Systemic lupus erythematosus, unspecified
M60.80	Other myositis, unspecified site
M60.811	Other myositis, right shoulder
M60.812	Other myositis, left shoulder
M60.819	Other myositis, unspecified shoulder
M60.821	Other myositis, right upper arm
M60.822	Other myositis, left upper arm
M60.829	Other myositis, unspecified upper arm
M60.831	Other myositis, right forearm
M60.832	Other myositis, left forearm
M60.839	Other myositis, unspecified forearm
M60.841	Other myositis, right hand
M60.842	Other myositis, left hand
M60.849	Other myositis, unspecified hand

M60.851	Other myositis, right thigh
M60.852	Other myositis, left thigh
M60.859	Other myositis, unspecified thigh
M60.861	Other myositis, right lower leg
M60.862	Other myositis, left lower leg
M60.869	Other myositis, unspecified lower leg
M60.871	Other myositis, right ankle and foot
M60.872	Other myositis, left ankle and foot
M60.879	Other myositis, unspecified ankle and foot
M60.88	Other myositis, other site
M60.89	Other myositis, multiple sites
M79.10	Myalgia, unspecified site
M79.11	Myalgia of mastication muscle
M79.12	Myalgia of auxiliary muscles, head and neck
M79.18	Myalgia, other site
N04.0	Nephrotic syndrome with minor glomerular abnormality
N04.1	Nephrotic syndrome with focal and segmental glomerular lesions
N04.2	Nephrotic syndrome with diffuse membranous glomerulonephritis
N04.21	Primary membranous nephropathy with nephrotic syndrome
N04.3	Nephrotic syndrome with diffuse mesangial proliferative glomerulonephritis
N04.4	Nephrotic syndrome with diffuse endocapillary proliferative glomerulonephritis
N04.5	Nephrotic syndrome with diffuse mesangiocapillary glomerulonephritis
N04.6	Nephrotic syndrome with dense deposit disease
N04.621	Primary membranous nephropathy with isolated proteinuria
N04.7	Nephrotic syndrome with diffuse crescentic glomerulonephritis
N04.8	Nephrotic syndrome with other morphologic changes
N04.9	Nephrotic syndrome with unspecified morphologic changes
N17.9	Acute kidney failure, unspecified
T86.09	Other complications of bone marrow transplant
Z85.3	Personal history of malignant neoplasm of breast
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
Z94.81	Bone marrow transplant status
Z94.84	Stem cells transplant status
Z94.89	Other transplanted organ and tissue status
Z94.9	Transplanted organ and tissue status, unspecified

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

The preceding information is intended for non-Medicare coverage determinations. Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determinations (NCDs) and/or Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. Local Coverage Articles (LCAs) may also exist for claims payment purposes or to clarify benefit eligibility under Part B for drugs which may be self-administered. The following link may be used to search for NCD, LCD, or LCA documents: <https://www.cms.gov/medicare-coverage-database/search.aspx>. Additional indications, including any preceding information, may be applied at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes		
Jurisdiction	NCD/LCA/LCD Document (s)	Contractor
5,8	A55639	Wisconsin Physicians Service Insurance Corp (WPS)
15	A57160, A58582	CGS Administrators, LLC
6,K	A59101	National Government Services, Inc
J,M	A56380	Palmetto GBA

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	Applicable State/US Territory	Contractor
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC