

# **Rituximab:**

## **Rituxan<sup>®</sup>, Truxima<sup>®</sup>, Ruxience<sup>™</sup>, Riabni<sup>™</sup>**

### **(Intravenous)**

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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

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

- Maintenance therapy for oncology indications may be renewed for up to a maximum of 2 years, unless otherwise specified:
  - Adult Acute Lymphoblastic Leukemia (ALL) may be renewed for a maximum of 18 doses.
  - Mantle Cell Lymphoma may be renewed until disease progression or intolerable toxicity.
  - Hairy Cell Leukemia may NOT be renewed.
  - Induction/Consolidation of Pediatric B-Cell Acute Leukemia and Aggressive Mature B-Cell Lymphomas may NOT be renewed.
  - Pediatric Hodgkin Lymphoma may NOT be renewed.
- Management of Immunotherapy-Related Toxicities:
  - Myositis/Myasthenia Gravis/Encephalitis may NOT be renewed.
  - Bullous Dermatitis may be renewed for a maximum of 18 months (4 total doses).
- Relapse therapy for Pemphigus Vulgaris must be at least 16 weeks past a prior infusion.
- Chronic Graft-Versus-Host Disease (cGVHD) may NOT be renewed.
- Hematopoietic Cell Transplantation may NOT be renewed.
- Lupus Nephritis may be renewed ONLY in patients experiencing a disease relapse.
- Complications of Transplanted Solid Organ may NOT be renewed.

#### **II. Dosing Limits**

##### **A. Quantity Limit (max daily dose) [NDC Unit]:**

- Rituxan 100 mg/10 mL single-dose vial for injection: 12 vials per 28 day supply
- Rituxan 500 mg/50 mL single-dose vial for injection: 8 vials per 28 day supply

- Truxima 100 mg/10 mL single-dose vial for injection: 12 vials per 28 day supply
- Truxima 500 mg/50 mL single-dose vial for injection: 8 vials per 28 day supply
- Ruxience 100 mg/10 mL single-dose vial for injection: 12 vials per 28 day supply
- Ruxience 500 mg/50 mL single-dose vial for injection: 8 vials per 28 day supply
- Riabni 100 mg/10 mL single-dose vial for injection: 12 vials per 28 day supply
- Riabni 500 mg/50 mL single-dose vial for injection: 8 vials per 28 day supply

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

<b>Oncology Indications</b>
<b><u>Chronic Lymphocytic Leukemia (CLL)/Small Lymphocytic Leukemia (SLL):</u></b>
<ul style="list-style-type: none"> <li>• Initial therapy: <ul style="list-style-type: none"> <li>◦ Loading dose: 100 billable units x 1 dose</li> <li>◦ Subsequent doses: 130 billable units every 28 days x 5 doses per 6 months</li> </ul> </li> <li>• Renewal therapy: 130 billable units every 8 weeks</li> </ul>
<b><u>ALL</u></b>
<ul style="list-style-type: none"> <li>• 100 billable units twice weekly x 18 doses</li> </ul>
<b><u>Hairy Cell Leukemia</u></b>
<ul style="list-style-type: none"> <li>• 100 billable units weekly x 8 doses</li> </ul>
<b><u>Histiocytic Neoplasms – Rosai-Dorfman Disease</u></b>
<ul style="list-style-type: none"> <li>• 130 billable units weekly x 6 doses in a 6 month period</li> </ul>
<b><u>Pediatric Hodgkin Lymphoma</u></b>
<ul style="list-style-type: none"> <li>• 100 billable units x 3 doses</li> </ul>
<b><u>cGVHD</u></b>
<ul style="list-style-type: none"> <li>• 100 billable units weekly x 8 doses</li> </ul>
<b><u>Hematopoietic Cell Transplantation</u></b>
<ul style="list-style-type: none"> <li>• Initial dose: 100 billable units x 1 dose before transplant</li> <li>• Subsequent doses: 250 billable units x 3 doses after transplant</li> </ul>
<b><u>All other oncology indications:</u></b>
<ul style="list-style-type: none"> <li>• Initial therapy: 100 billable units weekly x 8 doses per 6 months</li> <li>• Renewal therapy: 100 billable units x 4 doses per 6 months</li> </ul>
<b>Non-Oncology Indications</b>
<b><u>Rheumatoid Arthritis (RA):</u></b>
<ul style="list-style-type: none"> <li>• 100 billable units every 14 days x 2 doses in a 16 week period</li> </ul>
<b><u>Multiple Sclerosis (MS):</u></b>
100 billable units every 14 days x 2 doses every 6 months
<b><u>Pemphigus Vulgaris:</u></b>
<ul style="list-style-type: none"> <li>• Initiation: 100 billable units weekly x 4 doses in a 12 month period</li> <li>• Maintenance: 50 billable units every 16 weeks</li> </ul>
<b><u>GPA(WG)/MPA:</u></b>
<ul style="list-style-type: none"> <li>• Induction: 100 billable units weekly x 4 doses in a 4 month period</li> <li>• Initial Maintenance: 50 billable units x 2 doses in a 6 month period</li> <li>• Subsequent Maintenance: 50 billable units every 6 months</li> </ul>
<b><u>All other non-oncology indications:</u></b>
<ul style="list-style-type: none"> <li>• 100 billable units weekly x 4 doses in a 6 month period</li> </ul>

**III. Initial Approval Criteria <sup>1-4</sup>**

Effective January 1, 2022, coverage for Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infections and Pediatric Acute-Onset

Neuropsychiatric Syndrome (PANDAS/PANS) [ICD-10 D89.89] is permitted under Massachusetts Division of Insurance guidance.

Coverage is provided in the following conditions:

- Patients must have failed, or have a contraindication, or intolerance to rituximab-abbs (Truxima™) OR rituximab-pvvr (Ruxience™) prior to consideration of rituximab (Rituxan®) or rituximab-arrx (Riabni™); **AND**

- Patient is at least 18 years of age (unless otherwise specified); **AND**

#### **Universal Criteria** <sup>1-4</sup>

- Patient does not have a severe, active infection; **AND**
- Patient has been screened for the presence of hepatitis B (HBV) infection (i.e., HBsAg and anti-HBc) prior to initiating therapy and patients with evidence of current or prior HBV infection will be monitored for HBV reactivation during treatment; **AND**
- Patient has not received a live vaccine within 28 days prior to starting treatment and live vaccines will not be administered concurrently while on treatment; **AND**

#### **Oncology Indications** <sup>1-5</sup>

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

#### **Pediatric Mature B-Cell Acute Leukemia † Φ** <sup>1</sup>

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

#### **Adult\* Acute Lymphoblastic Leukemia (ALL) ‡** <sup>5,93</sup>

- Patient has Philadelphia chromosome-negative (Ph-) disease; **AND**
  - Used for induction/consolidation therapy; **AND**
    - Used in combination with a regimen containing an anthracycline and vincristine; **OR**
  - Used for relapsed/refractory disease; **AND**
    - Used in combination with MOpAD regimen (methotrexate, vincristine, pegaspargase, dexamethasone)

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

#### **Central Nervous System (CNS) Cancers ‡** <sup>5</sup>

- Patient has leptomeningeal metastases from lymphomas§; **OR**

- Patient has primary CNS lymphoma; **AND**
  - Used for induction therapy; **AND**
    - Used as a single agent OR in combination with a methotrexate-containing regimen, temozolomide, or lenalidomide~~¥~~; **OR**
    - Patient has CSF positive or spinal MRI positive disease~~§~~; **OR**
  - Used for consolidation (monthly maintenance) therapy; **AND**
    - Used as continuation of induction regimen in patients with complete response or complete response unconfirmed (CRu) to induction therapy; **AND**
      - Used as a single agent~~§~~; **OR**
      - Used on combination with high-dose methotrexate~~¥~~; **OR**
  - Used for relapsed or refractory disease; **AND**
    - Used as a single agent OR in combination with systemic therapy in patients with prior whole brain radiation therapy~~§~~; **AND**
      - Patient has CSF positive or spinal MRI positive disease; **OR**
    - Used as a single agent OR in combination with temozolomide, lenalidomide, or high-dose methotrexate~~¥~~

*§ For intrathecal administration ONLY; ¥ For intravenous administration ONLY*

#### **Adult Hodgkin Lymphoma ‡<sup>5</sup>**

- Patient has nodular lymphocyte-predominant disease

#### **Chronic Lymphocytic Leukemia/Small Lymphocytic Lymphoma (CLL/SLL) † ‡ Φ<sup>1-5</sup>**

- Used in combination with fludarabine and cyclophosphamide (FC) †; **OR**
- Patient has disease without del(17p)/TP53 mutation; **AND**
  - Used as first-line therapy in combination with bendamustine (*excluding use in frail patients*); **OR**
  - Used as subsequent therapy in combination with one of the following:
    - Bendamustine (*patients <65 years of age without significant comorbidities; excluding use in frail patients*)
    - Idelalisib
    - Lenalidomide
    - Venetoclax; **OR**
- Patient has disease with del(17p)/TP53 mutation; **AND**
  - Used as first-line therapy in combination with one of the following:
    - Alemtuzumab
    - 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 first-line therapy for histologic (Richter's) transformation to diffuse large B-cell lymphoma; **AND**
  - Used in combination with cyclophosphamide, doxorubicin, and vincristine-based regimens or as a component of OFAR (oxaliplatin, fludarabine, cytarabine, and rituximab)

### **Waldenström Macroglobulinemia/Lymphoplasmacytic Lymphoma ‡<sup>5</sup>**

**Adult B-Cell Lymphomas † ‡ Φ<sup>1-5,44</sup>** 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), or primary effusion lymphoma (PEL)
- Burkitt Lymphoma ‡
  - Used in combination with chemotherapy
- Castleman Disease ‡
  - Patient has multicentric disease; **OR**
  - Patient has unicentric disease; **AND**
    - Used as second-line therapy for relapsed or refractory disease; **OR**
    - Used for unresectable disease or symptomatic disease after incomplete resection
- Diffuse Large B-Cell Lymphoma † Φ
- Low-Grade (grade 1-2) or Follicular Lymphoma † Φ
- Extranodal Marginal Zone Lymphoma (EMZL) of the Stomach & Nongastric Sites (Noncutaneous) ‡
- High-Grade B-Cell Lymphomas ‡
- Mantle Cell Lymphoma ‡
- Nodal & Splenic Marginal Zone Lymphoma ‡
- Histologic Transformation of Indolent Lymphomas to Diffuse Large B-Cell Lymphoma ‡
- Post-Transplant Lymphoproliferative Disorders (PTLD) (B-Cell type) ‡

### **Primary Cutaneous B-Cell Lymphomas ‡<sup>5</sup>**

**Pediatric Aggressive Mature B-Cell Lymphomas (Primary Mediastinal Large B-Cell Lymphoma, Diffuse Large B-Cell Lymphoma, Burkitt Lymphoma, & Burkitt-like Lymphoma) † ‡ Φ<sup>1,5,50,121</sup>**

- Patient is at least 6 months of age\*; **AND**

- Used in combination with chemotherapy

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

### **Hairy Cell Leukemia ‡<sup>5</sup>**

- Used as a single agent; **AND**
  - Used for less than complete response or relapsed disease in patients 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 less than complete response or relapsed disease; **OR**
- Used in combination with vemurafenib; **AND**
  - Used for less than complete response or relapsed disease; **OR**
  - Used for progression after therapy for relapsed or refractory disease

### **Histiocytic Neoplasms – Rosai-Dorfman Disease ‡<sup>5</sup>**

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

### **Pediatric Hodgkin Lymphoma ‡<sup>5,128</sup>**

- Patient is ≤ 18 years of age\*; **AND**
- Patient 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) patients up to the age of 39 years.*

### **Chronic Graft-Versus-Host Disease (cGVHD) ‡<sup>5,22-25,45</sup>**

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

### **Hematopoietic Cell Transplantation ‡<sup>5</sup>**

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

### **Management of Immunotherapy-Related Toxicities ‡ 5,62**

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

- Patient is not on concurrent treatment with another TNF-inhibitor, biologic response modifier or other non-biologic agent (i.e., apremilast, tofacitinib, baricitinib, upadacitinib, etc.); **AND**

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

- |  |
|--|
| <ul style="list-style-type: none"> <li>• Patient must have failed or experienced intolerable side effects to: <ul style="list-style-type: none"> <li>– TWO (2) or more of the following: certolizumab (Cimzia), adalimumab (Humira), or golimumab (Simponi) or golimumab (Simponi ARIA); <b>AND</b></li> <li>– BOTH sarilumab (Kevzara) and tofacitinib (Xeljanz)</li> </ul> </li> </ul> |
|--|
- Documented moderate to severe active disease; **AND**
  - Used in combination with methotrexate unless the patient has a contraindication or intolerance; **AND**



- Patient tried and failed at least a 3 month trial with ONE oral disease modifying anti-rheumatic drug (DMARD) (e.g., methotrexate, azathioprine, auranofin, hydroxychloroquine, penicillamine, sulfasalazine, leflunomide, etc.); **AND**
- Previous failure with one or more preferred TNF antagonists at least one of which should be a self-injectable; **AND**
- Physician has assessed baseline disease severity utilizing an objective measure/tool; **AND**
- Patient has not had treatment with rituximab in the previous 4 months

#### **Pemphigus Vulgaris † Φ 1,10,11,35,36,38,61,80,114,139**

- Patient has a diagnosis of pemphigus vulgaris as determined by the following:
  - Patient 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**
- Patient has moderate to severe disease as assessed utilizing an objective measure/tool (i.e., PDAI, PSS, 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**

- Patient is at least 2 years of age; **AND**
- Used in combination with 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**
- Patient has previously failed or has a contraindication or intolerance to therapy with corticosteroids; **AND**
- Patient is at increased risk for bleeding as indicated by platelet count (within the previous 28 days) less than  $30 \times 10^9/L$  (30,000/mm<sup>3</sup>)



**Thrombotic Thrombocytopenic Purpura (TTP) ‡ 16-18,20,21,126**

- Patient is at increased risk for bleeding as indicated by platelet count (within the previous 28 days) less than  $30 \times 10^9/L$  (30,000/mm<sup>3</sup>); **AND**
- Patient 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 patients in remission

**Multiple Sclerosis ‡ 144,148**

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

**\*Definitive diagnosis of MS with a relapsing-remitting course is based upon BOTH dissemination in time and space. Unless contraindicated, MRI should be obtained (even if criteria are met). 148**

<u>Dissemination in time</u> <i>(Development/appearance of new CNS lesions over time)</i>	<u>Dissemination in space</u> <i>(Development of lesions in distinct anatomical locations within the CNS; multifocal)</i>
<ul style="list-style-type: none"> <li>• <math>\geq 2</math> clinical attacks; <b>OR</b></li> <li>• 1 clinical attack <b>AND</b> one of the following:                             <ul style="list-style-type: none"> <li>○ MRI indicating simultaneous presence of gadolinium-enhancing and non-enhancing lesions at any time or by a new T2-hyperintense or gadolinium-enhancing lesion on follow-up MRI compared to baseline scan</li> <li>○ CSF-specific oligoclonal bands</li> </ul> </li> </ul>	<ul style="list-style-type: none"> <li>• <math>\geq 2</math> lesions; <b>OR</b></li> <li>• 1 lesion <b>AND</b> one of the following:                             <ul style="list-style-type: none"> <li>○ Clear-cut historical evidence of a previous attack involving a lesion in a distinct anatomical location</li> <li>○ MRI indicating <math>\geq 1</math> T2-hyperintense lesions characteristic of MS in <math>\geq 2</math> of 4 areas of the CNS (periventricular, cortical or juxtacortical, infratentorial, or spinal cord)</li> </ul> </li> </ul>

**\*\*Active secondary progressive MS (SPMS) is defined as the following: 145,148-150**

- Expanded Disability Status Scale (EDSS) score  $\geq 3.0$ ; **AND**
- Disease is progressive  $\geq 3$  months following an initial relapsing-remitting course (i.e., EDSS score increase by 1.0 in patients with EDSS  $\leq 5.5$  or increase by 0.5 in patients with EDSS  $\geq 6$ ); **AND**
  - $\geq 1$  relapse within the previous 2 years; **OR**
  - Patient has gadolinium-enhancing activity **OR** new or unequivocally enlarging T2 contrast-enhancing lesions as evidenced by MRI

**\*\*\*Definitive diagnosis of CIS is based upon ALL of the following: 148**

- A monophasic clinical episode with patient-reported symptoms and objective findings reflecting a focal or multifocal inflammatory demyelinating event in the CNS
- Neurologic symptom duration of at least 24 hours, with or without recovery
- Absence of fever or infection
- Resembles a typical MS relapse (attack and exacerbation) but occurs in a patient not known to have multiple sclerosis

### **Autoimmune Hemolytic Anemia (AIHA) ‡<sup>26-32</sup>**

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

### **Lupus Nephritis ‡<sup>115-117,132</sup>**

- Patient has disease that is non-responsive or refractory to standard first-line therapy (e.g., mycophenolate mofetil, mycophenolic acid, cyclophosphamide, calcineurin inhibitors [e.g., tacrolimus]); **AND**
- Used as a single agent **OR** as add-on therapy in combination with mycophenolate mofetil, mycophenolic acid, or cyclophosphamide

### **Myasthenia Gravis (unrelated to immunotherapy-related toxicity) ‡<sup>118-120</sup>**

- Patient has muscle-specific tyrosine kinase (MuSK)-antibody positive disease; **AND**
- Patient is refractory to standard first-line therapy (e.g., glucocorticoids, azathioprine, mycophenolate mofetil, etc.)

### **Complications of Transplanted Solid Organ (kidney, liver, lung, heart, pancreas) in Adult and Pediatric\* Patients<sup>133-138</sup>**

- 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) ‡<sup>90-92</sup>**

- Patient has a confirmed diagnosis based on the following:
  - Patient is seropositive for aquaporin-4 (AQP4) IgG antibodies; **AND**
    - Patient has at least one core clinical characteristic §; **AND**
    - Alternative diagnoses have been excluded (e.g., multiple sclerosis, sarcoidosis, cancer, chronic infection, etc.); **OR**
  - Patient is seronegative for AQP4-IgG antibodies **OR** has unknown AQP4-IgG status; **AND**
    - Patient has at least two core clinical characteristics § occurring as a result of one or more clinical attacks; **AND**
    - Patient has experienced **ALL** of the following:
      - At least 1 core clinical characteristic must be optic neuritis, acute myelitis with LETM\*, or area postrema syndrome
      - Dissemination in space (≥2 different core clinical characteristics §)
      - Fulfillment of additional MRI requirements, as applicable ¶; **AND**

- Alternative diagnoses have been excluded (e.g., 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 <sup>90</sup>
<ul style="list-style-type: none"> <li>▪ Optic neuritis</li> <li>▪ Acute myelitis</li> <li>▪ Area postrema syndrome: episode of otherwise unexplained hiccups or nausea and vomiting</li> <li>▪ Acute brainstem syndrome</li> <li>▪ Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic MRI lesions</li> <li>▪ Symptomatic cerebral syndrome with NMOSD-typical brain lesions</li> </ul>
ψ Additional MRI requirements for NMOSD without AQP4-IgG and NMOSD with unknown AQP4-IgG status <sup>90</sup>
<ul style="list-style-type: none"> <li>▪ Acute optic neuritis: requires brain MRI showing (a) normal findings or only nonspecific white matter lesions, OR (b) optic nerve MRI with T2-hyperintense lesion or T1-weighted gadolinium enhancing lesion extending over &gt;1/2 optic nerve length or involving optic chiasm</li> <li>▪ Acute myelitis: requires associated intramedullary MRI lesion extending over ≥3 contiguous segments (LETM) OR ≥3 contiguous segments of focal spinal cord atrophy in patients with history compatible with acute myelitis</li> <li>▪ Area postrema syndrome: requires associated dorsal medulla/area postrema lesions</li> <li>▪ Acute brainstem syndrome: requires associated periependymal brainstem lesions</li> </ul>
<i>*LETM = longitudinally extensive transverse myelitis lesions</i>

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

#### IV. Renewal Criteria <sup>1-4</sup>

Coverage may be renewed based upon the following criteria:

- Patient 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**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: severe infusion-related reactions, tumor lysis syndrome (TLS), severe mucocutaneous reactions, 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**

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- Patient 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 cytogenetic analysis, QPCR, or FISH

### **Hairy Cell Leukemia**

- Coverage may NOT be renewed

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

- Coverage may NOT be renewed

### **Pediatric Hodgkin Lymphoma**

- Coverage may NOT be renewed

### **Chronic Graft-Versus-Host Disease (cGVHD)**

- Coverage may NOT be renewed

### **Hematopoietic Cell Transplantation**

- Coverage may NOT be renewed

### **Management of Immunotherapy-Related Toxicities**

- Coverage for use in the treatment of myositis/myasthenia gravis/encephalitis may NOT be renewed.
- Coverage for use in bullous dermatitis: Patient has not exceeded a maximum of 18 months of therapy (4 total doses).

### **All Other Oncology Indications**

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

### **Non-Oncology Indications** <sup>1-4,7-12</sup>

#### **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]; **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 patient 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)**

- Disease response as indicated by the achievement and maintenance of a platelet count of at least  $50 \times 10^9/L$  as necessary to reduce the risk for bleeding

### **Thrombotic Thrombocytopenic Purpura (TTP)**

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

### **Multiple Sclerosis (MS) <sup>147,151</sup>**

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

**\*Note:**

- Inadequate response, in those who have been adherent and receiving therapy for sufficient time to realize the full treatment effect, is defined as  $\geq 1$  relapse,  $\geq 2$  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)**

- 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 <sup>1,10,11,35</sup>**

- Patient 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
    - Patient has not developed new lesions and established lesions begin to heal
    - For Relapses ONLY: Patient previously achieved disease control; **AND**
      - Patient has the appearance of 3 or more new lesions a month that do not heal spontaneously within 1 week, or by the extension of established lesions

### **Autoimmune Hemolytic Anemia (AIHA)**

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

#### **Lupus Nephritis** <sup>115-117</sup>

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

#### **Myasthenia Gravis (unrelated to immunotherapy-related toxicity)** <sup>118-120</sup>

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

#### **Complications of Transplanted Solid Organ (kidney, liver, lung, heart, pancreas)** <sup>133-138</sup>

- Coverage may NOT be renewed.

#### **NMOSD** <sup>90,91</sup>

- Disease response as indicated by stabilization/improvement in any of the following: neurologic symptoms as evidenced by a decrease in acute relapses, reduced hospitalizations, reduction/discontinuation in plasma exchange treatments, and/or reduction/discontinuation of corticosteroids without relapse

### **V. Dosage/Administration** <sup>1-5,9,23-26,32,34,40,42,44,50,62,80,83-89,91,94-98,102-111,115-118,122-125,128-133,135-137,140</sup>

<b>Indication</b>		<b>Dose</b>
CLL/SLL	Initial Therapy	375 mg/m <sup>2</sup> intravenously (IV) weekly for 8 doses; <b>OR</b> 375 mg/m <sup>2</sup> IV cycle 1, then 500 mg/m <sup>2</sup> every 28 days cycles 2-6 (6 total doses); <b>OR</b> 375 mg/m <sup>2</sup> IV cycle 1, followed by 500 mg/m <sup>2</sup> every 2 weeks for 4 doses, then 500 mg/m <sup>2</sup> every 28 days for 3 doses (8 total doses)
	<i>Renewal Therapy</i>	375 mg/m <sup>2</sup> IV every 3 months; <b>OR</b> 500 mg/ m <sup>2</sup> IV every 8 weeks
Adult B-Cell Lymphomas, Primary Cutaneous B-Cell Lymphomas, Waldenström Macroglobulinemia, or Adult HL	Initial Therapy	375 mg/m <sup>2</sup> IV once weekly for 4 – 8 doses in a 6 month period
	<i>Renewal Therapy</i>	375 mg/m <sup>2</sup> IV once weekly for 4 doses per 6 month period; <b>OR</b> 375 mg/ m <sup>2</sup> IV every 8 weeks
Pediatric Aggressive Mature B-Cell Lymphomas		<b><u>Induction* [courses 1 and 2 (COPDAM1 and COPDAM2)]</u></b> 375 mg/m <sup>2</sup> IV, two doses during each of the induction courses (Day -2 and Day 1).

Indication	Dose
	<p><i>During the 1<sup>st</sup> 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></p> <p><b><u>Consolidation* [courses 1 and 2 (CYM/CYVE)]</u></b></p> <p>375 mg/m<sup>2</sup> IV, one dose during each of the consolidation courses (Day 1)</p> <p><b><u>Relapsed/Refractory</u></b></p> <p>RCYVE – 375mg/m<sup>2</sup> IV on day 1 of each 21-day cycle</p> <p>RICE – 375 mg/m<sup>2</sup> IV on days 1 and 3 of courses 1 and 2, and on day 1 only of course 3 if needed.</p> <p><i>*Note: dosing and dosing schedules are highly variable and dependent on regimen used, please refer to NCCN and PI for different protocols.</i></p>
Pediatric Mature B-Cell Acute Leukemia	<p><b><u>Induction* [courses 1 and 2 (COPDAM1 and COPDAM2)]</u></b></p> <p>375 mg/m<sup>2</sup> IV, two doses during each of the induction courses (Day -2 and Day 1).</p> <p><i>During the 1<sup>st</sup> 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></p> <p><b><u>Consolidation* [courses 1 and 2 (CYM/CYVE)]</u></b></p> <p>375 mg/m<sup>2</sup> IV, one dose during each of the consolidation courses (Day 1)</p> <p><i>*Note: dosing and dosing schedules are highly variable and dependent on regimen used, please refer to NCCN and PI for different protocols.</i></p>
CNS Lymphoma	<p><b><u>Intravenous administration</u></b></p> <p><u>Initial Therapy:</u> 375 mg/m<sup>2</sup> IV once weekly for 4 – 8 doses in a 6 month period</p> <p><u>Renewal Therapy:</u> 375 mg/m<sup>2</sup> IV once weekly for 4 doses per 6 month period; <b>OR</b></p> <p>375 mg/m<sup>2</sup> IV every 8 weeks</p> <p><b><u>Intrathecal/Intraventricular administration</u></b></p> <p>10-40 mg weekly to every 3 weeks</p>
ALL	<p>375 mg/m<sup>2</sup> 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])</p>
Hairy Cell Leukemia	<p>375 mg/m<sup>2</sup> IV once weekly for 4 – 8 doses</p>
RA	<p>1,000 mg IV on days 1 and 15, repeated every 24 weeks. May repeat up to every 16 weeks in patients requiring more frequent dosing based on clinical evaluation.</p>



Indication	Dose
Pemphigus Vulgaris	<p><u>Initiation</u> 1,000 mg IV on days 1 and 15; <b>OR</b> 375 mg/m<sup>2</sup> IV weekly for 4 doses</p> <p><u>Maintenance</u> 500 mg IV at month 12 and repeat every 6 months thereafter or based on clinical evaluation</p> <p><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></p>
AIHA	375 mg/m <sup>2</sup> IV weekly for 4 doses in a 6 month period
Thrombocytopenic Purpura or Thrombotic Thrombocytopenic Purpura (TTP)	375 mg/m <sup>2</sup> IV weekly for 4 doses; <b>OR</b> 1,000 mg IV on days 1 and 15
Management of Immunotherapy-Related Toxicities	<p><u>Bullous Dermatitis</u> 1,000 mg IV every 2 weeks for 2 doses, then 500 mg IV at months 12 and 18 as needed</p> <p><u>Myositis</u> 375 mg/m<sup>2</sup> IV weekly for 4 doses</p> <p><u>Myasthenia Gravis</u> 375 mg/m<sup>2</sup> IV weekly for 4 doses; <b>OR</b> 500 mg/m<sup>2</sup> IV every 2 weeks for 2 doses</p> <p><u>Encephalitis</u> 1,000 mg IV every 2 weeks for 2 doses; <b>OR</b> 375 mg/m<sup>2</sup> IV weekly for 4 doses</p>
GPA (WG), MPA	<p><u>Induction (Pediatric and Adult)</u> 375 mg/m<sup>2</sup> IV weekly for 4 doses; <b>OR</b></p> <ul style="list-style-type: none"> <li>- Adults: 1,000 mg IV on days 1 and 15; <b>OR</b></li> <li>- Pediatric (up to a maximum of 1,000 mg per dose): <ul style="list-style-type: none"> <li>o 575 mg/m<sup>2</sup> IV on days 1 and 15 (BSA ≤1.5m<sup>2</sup>)</li> <li>o 750 mg/m<sup>2</sup> IV on days 1 and 15 (BSA &gt;1.5m<sup>2</sup>)</li> </ul> </li> </ul> <p><u>Maintenance</u></p> <ul style="list-style-type: none"> <li>- Pediatric: <ul style="list-style-type: none"> <li>o 250 mg/m<sup>2</sup> IV on days 1 and 15, then 250 mg/m<sup>2</sup> IV every 6 months thereafter based on clinical evaluation</li> </ul> </li> <li>- Adult: <ul style="list-style-type: none"> <li>o 500 mg IV on days 1 and 15, then 500 mg IV every 6 months thereafter based on clinical evaluation</li> </ul> </li> </ul>

Indication	Dose
	<p><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></p> <p><i>*Initial MAINTENANCE infusions should be initiated within 4 weeks following disease control when initial induction occurred with other standard of care immunosuppressants.</i></p>
cGVHD	<p>375 mg/m<sup>2</sup> IV weekly for 4 doses, then 375 mg/m<sup>2</sup> IV monthly for 4 months</p> <p><b>-OR-</b></p> <p>375 mg/m<sup>2</sup> IV weekly for 4 doses (<i>Note: A second course of 4 weekly doses may be administered 8 weeks after initial therapy for patients with lack of or incomplete response.</i>)</p> <p><b>-OR-</b></p> <p>375 mg/m<sup>2</sup> IV weekly for 4 – 8 doses</p>
Hematopoietic Cell Transplantation	375 mg/m <sup>2</sup> IV for 1 day before transplant, then 1000 mg/m <sup>2</sup> IV on days 1,8, and 15 after transplant
Multiple Sclerosis	1,000 mg IV on days 1 and 15, repeat every 6 months
NMOSD	<p>1,000 mg IV once on days 1 and 15, repeat every 6 months</p> <p><b>-OR-</b></p> <p>375 mg/m<sup>2</sup> once weekly for 4 weeks, repeat every 6 months</p>
Histiocytic Neoplasms – Rosai-Dorfman Disease	500 mg/m <sup>2</sup> IV every 1 – 2 weeks for 2 – 6 doses every 6 months
Lupus Nephritis	<p>1,000 mg IV on days 1 and 15</p> <p><b>-OR-</b></p> <p>375 mg/m<sup>2</sup> IV once weekly for 4 doses</p>
Myasthenia Gravis (unrelated to immunotherapy-related toxicity)	<p>1,000 mg IV on days 1 and 15, may repeat a full or partial course every 6 months</p> <p><b>-OR-</b></p> <p>375 mg/m<sup>2</sup> IV once weekly for 4 doses, may repeat a full or partial course every 6 months</p>
Pediatric Hodgkin Lymphoma	375 mg/m <sup>2</sup> 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)	<p>– Adults and pediatrics weighing ≥0.5 m<sup>2</sup>: 375 mg/m<sup>2</sup> weekly for up to 4 doses</p> <p>– Pediatrics weighing &lt;0.5 m<sup>2</sup>: 12.5 mg/kg weekly for up to 4 doses</p>
<p><i>Abbreviations: COP = Cyclophosphamide, Oncovin (vincristine), Prednisone; COPDAM = Cyclophosphamide, Oncovin (vincristine), Prednisolone, Adriamycin (doxorubicin), Methotrexate; CYM = Cytarabine (Aracytine, Ara-C), Methotrexate; CYVE = Cytarabine (Aracytine, Ara-C), Veposide (VP16)</i></p>	

## 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

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## Appendix 1 – Covered Diagnosis Codes

ICD-10	Description
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
C83.50	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
C88.0	Waldenström macroglobulinemia
C88.4	Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT-lymphoma)
C91.00	Acute lymphoblastic leukemia not having achieved remission
C91.01	Acute lymphoblastic leukemia, in remission
C91.02	Acute lymphoblastic leukemia, in relapse
C91.10	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
D59.10	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
D76.3	Other histiocytosis syndromes
D89.811	Chronic graft-versus-host disease
D89.812	Acute on chronic graft-versus-host disease
D89.813	Graft-versus-host disease unspecified
G04.81	Other encephalitis and encephalomyelitis
G04.89	Other myelitis
G04.90	Encephalitis and encephalomyelitis, unspecified
G35	Multiple sclerosis
G36.0	Neuromyelitis optica [Devic]
G70.0	Myasthenia gravis
G70.00	Myasthenia gravis without (acute) exacerbation
G70.01	Myasthenia gravis with (acute) exacerbation
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
M31.1	Thrombotic microangiopathy
M31.30	Wegener's granulomatosis without renal involvement
M31.31	Wegener's granulomatosis with renal involvement
M31.7	Microscopic polyangiitis
M32.14	Glomerular disease in systemic lupus erythematosus
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
T86.09	Other complications of bone marrow transplant
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
Z94.89	Other transplanted organ and tissue status
Z94.9	Transplanted organ and tissue status, unspecified

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

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

Determination (NCD), Local Coverage Articles (LCAs), and Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. They can be found at: <https://www.cms.gov/medicare-coverage-database/search.aspx>. Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD/LCA):

<b>Jurisdiction(s):</b> 6, K	<b>NCD/LCD/LCA Document (s):</b> A59101
<a href="https://www.cms.gov/medicare-coverage-database/new-search/search-results.aspx?keyword=a59101&amp;areaId=all&amp;docType=NCA%2CCAL%2CNCD%2CMEDCAC%2CTA%2CMCD%2C6%2C3%2C5%2C1%2CF%2CP">https://www.cms.gov/medicare-coverage-database/new-search/search-results.aspx?keyword=a59101&amp;areaId=all&amp;docType=NCA%2CCAL%2CNCD%2CMEDCAC%2CTA%2CMCD%2C6%2C3%2C5%2C1%2CF%2CP</a>	

<b>Jurisdiction(s):</b> 5,8	<b>NCD/LCD/LCA Document (s):</b> A55639
<a href="https://www.cms.gov/medicare-coverage-database/new-search/search-results.aspx?keyword=a55639&amp;areaId=all&amp;docType=NCA%2CCAL%2CNCD%2CMEDCAC%2CTA%2CMCD%2C6%2C3%2C5%2C1%2CF%2CP">https://www.cms.gov/medicare-coverage-database/new-search/search-results.aspx?keyword=a55639&amp;areaId=all&amp;docType=NCA%2CCAL%2CNCD%2CMEDCAC%2CTA%2CMCD%2C6%2C3%2C5%2C1%2CF%2CP</a>	

<b>Jurisdiction(s):</b> J, M	<b>NCD/LCD/LCA Document (s):</b> A56380
<a href="https://www.cms.gov/medicare-coverage-database/new-search/search-results.aspx?keyword=a56380&amp;areaId=all&amp;docType=NCA%2CCAL%2CNCD%2CMEDCAC%2CTA%2CMCD%2C6%2C3%2C5%2C1%2CF%2CP">https://www.cms.gov/medicare-coverage-database/new-search/search-results.aspx?keyword=a56380&amp;areaId=all&amp;docType=NCA%2CCAL%2CNCD%2CMEDCAC%2CTA%2CMCD%2C6%2C3%2C5%2C1%2CF%2CP</a>	

<b>Jurisdiction(s):</b> 15	<b>NCD/LCD/LCA Document (s):</b> A57160
<a href="https://www.cms.gov/medicare-coverage-database/new-search/search-results.aspx?keyword=a57160&amp;areaId=all&amp;docType=NCA%2CCAL%2CNCD%2CMEDCAC%2CTA%2CMCD%2C6%2C3%2C5%2C1%2CF%2CP">https://www.cms.gov/medicare-coverage-database/new-search/search-results.aspx?keyword=a57160&amp;areaId=all&amp;docType=NCA%2CCAL%2CNCD%2CMEDCAC%2CTA%2CMCD%2C6%2C3%2C5%2C1%2CF%2CP</a>	

<b>Jurisdiction(s):</b> 15	<b>NCD/LCD/LCA Document (s):</b> A58582
<a href="https://www.cms.gov/medicare-coverage-database/new-search/search-results.aspx?keyword=a58582&amp;areaId=all&amp;docType=NCA%2CCAL%2CNCD%2CMEDCAC%2CTA%2CMCD%2C6%2C3%2C5%2C1%2CF%2CP">https://www.cms.gov/medicare-coverage-database/new-search/search-results.aspx?keyword=a58582&amp;areaId=all&amp;docType=NCA%2CCAL%2CNCD%2CMEDCAC%2CTA%2CMCD%2C6%2C3%2C5%2C1%2CF%2CP</a>	

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)

**Medicare Part B Administrative Contractor (MAC) Jurisdictions**

<b>Jurisdiction</b>	<b>Applicable State/US Territory</b>	<b>Contractor</b>
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
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