Rituximab:
Rituxan®, Truxima®, Ruxience®, Riabni™
(Intravenous)

Document Number: IC-0109

Last Review Date: 08/01/2024
Date of Origin: 7/20/2010
Dates Reviewed: 09/2010, 12/2010, 02/2011, 03/2011, 05/2011, 06/2011, 09/2011, 12/2011, 03/2012, 06/2012, 09/2012, 12/2012, 03/2013, 06/2013, 09/2013, 12/2013, 03/2014, 06/2014, 09/2014, 12/2014, 03/2015, 05/2015, 08/2015, 11/2015, 02/2016, 05/2016, 08/2016, 10/2016, 02/2017, 05/2017, 08/2017, 10/2017, 02/2018, 05/2018, 07/2018, 09/2018, 12/2018, 03/2019, 06/2019, 09/2019, 10/2019, 12/2019, 03/2020, 06/2020, 09/2020, 12/2020, 01/2021, 03/2021, 06/2021, 09/2021, 12/2021, 01/2022, 03/2022, 06/2022, 09/2022, 12/2022, 03/2023, 06/2023, 09/2023, 12/2023, 03/2024, 05/2024, 08/2024

I. Length of Authorization [1-5,23-25,34,44,62,80,94-98,102-104,108,115-118,128-130,133-138, 153,155,170-174]

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 be renewed for up to a maximum of 12 doses.
  - Induction/Consolidation of Pediatric B-Cell Acute Leukemia and Aggressive Mature B-Cell Lymphomas may NOT be renewed.
  - Pediatric Hodgkin Lymphoma may NOT be renewed.
- Management of Immunotherapy-Related Toxicities:
  - Myositis/Myasthenia Gravis/Encephalitis may NOT be renewed.
  - Bullous Dermatitis may be renewed for a maximum of 18 months (4 total doses).
- 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 and Pediatric Idiopathic Nephrotic Syndrome 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]:

Oncology Indications

Chronic Lymphocytic Leukemia (CLL)/Small Lymphocytic Leukemia (SLL):
• Initial therapy:
  ○ Loading dose: 100 billable units x 1 dose
  ○ Subsequent doses: 130 billable units every 28 days x 5 doses per 6 months
• Renewal therapy: 130 billable units every 8 weeks

ALL • 100 billable units twice weekly x 18 doses
Waldenström Macroglobulinemia/Lymphoplasmacytic Lymphoma • Initial therapy: 100 billable units weekly x 12 doses • Renewal therapy: 400 billable units every 6 months
CNS Cancers • Initial therapy: 190 billable units weekly x 8 doses • Renewal therapy: 400 billable units every 6 months
Hairy Cell Leukemia • 100 billable units weekly x 8 doses, 100 billable units every 14 days x 8 doses, then 100 billable units every 28 days x 4 doses
Histiocytic Neoplasms – Rosai-Dorfman Disease • 130 billable units weekly x 6 doses in a 6 month period
Pediatric Hodgkin Lymphoma • 100 billable units x 3 doses
Chronic Graft-Versus-Host Disease (cGVHD) • 100 billable units weekly x 8 doses
Hematopoietic Cell Transplantation • Initial dose: 100 billable units x 1 dose before transplant • Subsequent doses: 250 billable units x 3 doses after transplant
All other oncology indications: • Initial therapy: 100 billable units weekly x 8 doses per 6 months • Renewal therapy: 400 billable units every 6 months
Non-Oncology Indications
Rheumatoid Arthritis (RA): • 100 billable units every 14 days x 2 doses in an 18-week period
Multiple Sclerosis (MS): • 100 billable units every 14 days x 2 doses every 6 months

Pemphigus Vulgaris (PV): • Initiation: 100 billable units weekly x 4 doses in a 12 month period • Maintenance and Relapse: 50 billable units every 16 weeks
GPA(WG)/MPA: • Induction: 100 billable units weekly x 4 doses in a 20-week period • Initial Maintenance: 100 billable units x 2 doses in a 6 month period • Subsequent Maintenance: 100 billable units every 6 months
All other non-oncology indications: • 400 billable units every 6 months

III. Initial Approval Criteria [1-4]

Coverage is provided in the following conditions:

- Patient must try and have an inadequate response, contraindication, or intolerance to Ruxience AND Truxima; **OR**
- Patient is continuing treatment with a different rituximab product

Step therapy does not apply to MN residents with metastatic cancer per statute 62Q.1841. https://www.revisor.mn.gov/statutes/cite/62Q.1841

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

Universal Criteria [1-4]

- 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 [1-5]

- 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 (B-AL) † [1]

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

Adult* Acute Lymphoblastic Leukemia (ALL) ‡ [5,93]

- Patient has Philadelphia chromosome-positive (Ph+) disease; **AND**
  - Used in combination with a tyrosine kinase inhibitor (TKI)-based regimen; **AND**
    - Patient is <65 years of age without significant comorbidities; **OR**
  - Used in combination with MOpAD (methotrexate, vincristine, pegaspargase, dexamethasone) for TKI-refractory disease; **OR**

- Patient has Philadelphia chromosome-negative (Ph-) disease; **AND**
  - Used as a component of a multiagent chemotherapy

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

Central Nervous System (CNS) Cancers ‡ [5]

- 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 or intraventricular administration. ¥ For intravenous administration

Adult Hodgkin Lymphoma ‡ [5]

- Patient 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**
- Patient has disease <u>without</u> 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 high-dose methylprednisolone; **OR**
    - Used as subsequent therapy in combination with one of the following:
        - Alemtuzumab
        - High-dose methylprednisolone
        - Idelalisib
        - Lenalidomide
        - Venetoclax; **OR**
- Used as initial therapy for histologic (Richter's) transformation to diffuse large B-cell lymphoma; **AND**
    - Used in combination with cyclophosphamide, doxorubicin, and vincristine-based regimens or as a component of OFAR (oxaliplatin, fludarabine, cytarabine, and rituximab)

Waldenström Macroglobulinemia/Lymphoplasmacytic Lymphoma ‡ [5]

Adult B-Cell Lymphomas † ‡ Φ [1-5,44] 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
- 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) ‡
- 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) (B-Cell type) ‡

Castleman Disease ‡ [5]

- 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

Primary Cutaneous B-Cell Lymphomas ‡ [5]

Pediatric Aggressive Mature B-Cell Lymphomas (Primary Mediastinal Large B-Cell Lymphoma, Diffuse Large B-Cell Lymphoma, Burkitt Lymphoma, & Burkitt-like Lymphoma) † ‡ Φ [1,5,50,121]

- 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 ‡ [5]

- 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 as initial therapy in patients unable to tolerate purine analogs (i.e., cladribine or pentostatin) including frail patients and those with active infection; **OR**
  - Used for less than complete response or relapse within 2 years of complete response following initial treatment with cladribine or pentostatin; **OR**
  - Used for progression after therapy for relapsed or refractory disease (if not previously given); **OR**
- Used in combination with venetoclax; **AND**
  - Used for progression after therapy for relapsed or refractory disease; **AND**
    - Patient 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]

- 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) ‡ [5,22-25]

- Patient is post-allogeneic hematopoietic cell transplant (generally 3 or more months); **AND**
- Used as additional therapy in combination with systemic corticosteroids; **AND**
- Patient 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

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, tremelimumab, retifanlimab, toripalimab, tislelizumab, 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 high-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 CD20-directed therapy, biologic agents (e.g., TNF-inhibitor, IL-inhibitor, integrin receptor antagonist, T cell costimulation modulator, etc.) or targeted synthetic therapies (e.g., apremilast, abrocitinib, tofacitinib, baricitinib, upadacitinib, deucravacitnib, ritlecitinib, ruxolitinib, etrasimod, ozanimod, etc.); **AND**

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

- 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 patient has a contraindication or intolerance; **AND**
  - Patient 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**
  - Patient is already established on biologic or targeted synthetic therapy for the treatment of RA; **AND**
- Previous failure with one or more preferred TNF antagonists at least one of which should be a self-injectable; **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 (e.g., 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) of less than 30×10^9/L (30,000/mm^3)

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

- 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 (MS) ‡ [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]

Dissemination in time (*Development/appearance of new CNS lesions over time*)	**Dissemination in space** (*Development of lesions in distinct anatomical locations within the CNS; multifocal*)

• ≥ 2 clinical attacks; **OR**
• 1 clinical attack AND one of the following:
  ○ 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
  ○ CSF-specific oligoclonal bands
	• ≥ 2 lesions; **OR**
	• 1 lesion AND one of the following:
	  ○ Clear-cut historical evidence of a previous attack involving a lesion in a distinct anatomical location
	  ○ MRI indicating ≥ 1 T2-hyperintense lesions characteristic of MS in ≥ 2 of 4 areas of the CNS (periventricular, cortical or juxtacortical, infratentorial, or spinal cord)

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

- Expanded Disability Status Scale (EDSS) score ≥ 3.0; **AND**
- Disease is progressive ≥ 3 months following an initial relapsing-remitting course (i.e., EDSS score increase by 1.0 in patients with EDSS ≤5.5 or increase by 0.5 in patients with EDSS ≥6); **AND**
  - ≥ 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
- Patient is not known to have multiple sclerosis

Autoimmune Hemolytic Anemia (AIHA) ‡ [26-32]

- 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

Systemic Lupus Erythematosus (SLE) ‡ [153-155,158-163,169]

- Patient has a confirmed diagnosis of SLE as evidenced by all of the following:
  - Confirmed SLE classification criteria score ≥ 10***** (*Note: must include clinical and immunologic domains criteria)*
  - Anti-nuclear antibody (ANA) titer of ≥ 1:80 measured via indirect immunofluorescence (IIF) on human epithelial (HEp-2) cells (or an equivalent ANA positive test) at least once; **AND**
- Physician has assessed baseline disease severity utilizing an objective measure/tool (i.e., Systemic Lupus Erythematosus Disease Activity Index 2000 (SLEDAI 2K), British Isles Lupus Assessment Group-2004 (BILAG 2004), and/or Physician's Global Assessment (PGA) score); **AND**
- Patient has failed to respond adequately to at least two (2) standard therapies****** such as anti-malarials (i.e. hydroxychloroquine, chloroquine), corticosteroids, non-steroidal anti-inflammatory drugs (NSAIDs), aspirin, immunosuppressives (i.e. azathioprine, methotrexate, calcineurin inhibitors [cyclosporine, tacrolimus, voclosporin], oral cyclophosphamide, or mycophenolate); **AND**
- Patient has moderate to severe active disease as defined by ONE of the following:
  - Systemic Lupus Erythematosus Disease Activity Index 2000 (SLEDAI 2K) score of ≥ 6
  - Disease activity with ≥ 2 systems with British Isles Lupus Assessment Group-2004 (BILAG) B scores
  - ≥ 1 system(s) with British Isles Lupus Assessment Group-2004 (BILAG) A score(s)

***Classification Criteria for Systemic Lupus Erythematosus (SLE)** [159]	
Clinical Score Δ (range: 0-39)	**Clinical Domains and Criteria**
2	**Constitutional:** Unexplained fever > 101°F
3 4	**Hematologic:** White blood cell count < 4,000/mm^3 Platelet count < 100,000/mm^3 or Autoimmune hemolysis
2	**Neuropsychiatric:** Delirium

3 5	Psychosis Primary generalized seizure or partial/focal seizure
 2 4 6	**Mucocutaneous +:** Non-scarring alopecia or oral ulcers Subacute cutaneous or discoid lupus Acute cutaneous lupus
 5 6	**Serosal:** Pleural or pericardial effusion Acute pericarditis
6	**Musculoskeletal:** Joint involvement with either synovitis involving 2 or more joints with swelling or effusion OR tenderness in 2 or more joints with at least 30 minutes of morning stiffness
 4 8 10	**Renal:** Proteinuria > 0.5g/24 hr by a 24-hour urine or equivalent spot urine protein-to-creatinine ratio Renal biopsy class II or V lupus nephritis Renal biopsy Class III or IV lupus nephritis
Immunologic Score ᐞ (range: 0-12)	**Immunologic Domains and Criteria**
2	**Presence of antiphospholipid antibodies** (i.e., positive lupus anticoagulant, positive anti-ß2GP1 antibodies, and/or anti-cardiolipin antibodies at medium or high titer)
 3 4	**Presence of low complement proteins** (below lower limit of normal): Low C3 OR low C4 Low C3 AND C4
6	**Presence of anti-Sm and/or anti-dsDNA antibodies**
***** A web-based scoring calculator as well as further definitions of each criterion are available at: https://rheumatology.org/criteria ᐞ Occurrence on at least one occasion is sufficient to count toward score when all other causes have been ruled out. Count only the highest weighted score within each of the 10 domains (7 clinical and 3 immunologic) and any additional criteria within the same domain will not count. **+** Observed by a physician via clinical exam or photograph review	

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

Lupus Nephritis (LN) ‡ [115-117,132,153,155,159,166,169]

- Patient has disease that is non-responsive or refractory to standard first-line therapy (i.e., mycophenolate mofetil, mycophenolic acid, cyclophosphamide, or calcineurin inhibitors [e.g., tacrolimus, voclosporin, cyclosporine etc.]); **AND**
- 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) ‡ [118-120,156]

- 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 [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]

- Patient has a confirmed diagnosis based on the following:
  - Patient was found to be seropositive for aquaporin-4 (AQP4) IgG antibodies; **AND**
    - Patient 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**
  - 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 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 (≥ half of the length of the optic nerve or involving optic chiasm; T2 or T1/Gd) ▪ Myelitis: Intramedullary lesion ≥ 3 contiguous VS (LETM) OR focal atrophy ≥ 3 contiguous VS in patients with a history of acute myelitis ▪ Area postrema syndrome (APS): Lesion in the dorsal medulla oblongata/area postrema ▪ Other brainstem syndrome: Periependymal brainstem lesion (4th ventricle) ▪ ¥ Diencephalic syndrome: Periependymal lesion (3rd ventricle) OR hypothalamic/thalamic lesion ▪ ** Cerebral syndrome: Extensive periependymal lesion (lateral ventricle; often with Gd) OR long (> 1⁄2 length), diffuse, heterogeneous or edematous corpus callosum lesion OR long corticospinal tract lesion (unilateral or bilateral, contiguously involving internal capsule and cerebral peduncle) OR large, confluent (unilateral or bilateral) subcortical or deep white matter lesion
LETM = longitudinally extensive transverse myelitis lesions; VS = vertebral segments

Antisynthetase Syndrome-Related Interstitial Lung Disease ‡ [167,168,174, 186]

- Patient 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**
- Patient has documented severe active disease; **AND**
- Patient 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 patient has a contraindication or intolerance

Idiopathic Membranous Nephropathy ‡ [172, 175-177]

- Patient 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 patients 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/1.73m^2; **OR**

- Proteinuria > 8 g/d for > 6 months; **OR**
- Patient 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**
  - Patient 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**
  - Patient has proteinuria > 1 g/d

Pediatric Idiopathic Nephrotic Syndrome ‡ [170-173]

- Patient is 12 years of age or younger; **AND**
- Patient has symptomatic disease (i.e., nephrotic-range proteinuria and either hypoalbuminemia or edema when albumin level is not available); **AND**
- Patient has been diagnosed with one of the following:
  - Frequently relapsing nephrotic syndrome (FRNS) with at least four relapses per year or at least two relapses within 6 months of initial presentation
  - Steroid dependent nephrotic syndrome (SDNS) with two consecutive relapses during steroid tapering or within 14 days of cessation of therapy
  - Steroid resistant nephrotic syndrome (SRNS) with failure to achieve complete remission within a 4-6 -week course of daily corticosteroids; **AND**
- Patient has failed an adequate trial with at least one other steroid-sparing agent (e.g., cyclophosphamide, calcineurin inhibitor [e.g., tacrolimus, cyclosporine, etc.], mycophenolate mofetil, etc.)

IgG4-Related Disease ‡ [178-182]

- Physician has assessed baseline disease severity utilizing an objective measure/tool (e.g., IgG4-RD Responder Index score, physician's global assessment [PGA], amount of glucocorticoid or other immunosuppressive use, incidence of disease flares, serum IgG4 level, etc.); **AND**
- Other conditions that mimic IgG4-related disease have been ruled out (e.g., malignancy, infection, other autoimmune disorders, etc.); **AND**
- Patient has documented active disease; **AND**

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

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

IV. Renewal Criteria [1-4]

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 (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 [1-5,23-25,34,44,50,62,94-98,102-104,129,130,128]

- 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 cytogenic analysis, QPCR, or FISH

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

- Coverage may NOT be renewed

Pediatric Hodgkin Lymphoma

- Coverage may NOT be renewed

Chronic Graft-Versus-Host Disease (cGVHD)

- Coverage may NOT be renewed

Hematopoietic Cell Transplantation

- Coverage may NOT be renewed

Management of Immunotherapy-Related Toxicities

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

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

All Other Oncology Indications

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

Non-Oncology Indications [1-4]

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 ≥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 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) [7-9, 63]

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

Thrombotic Thrombocytopenic Purpura (TTP)

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

Multiple Sclerosis (MS) [147,151]

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

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

unequivocally new MRI-detected lesions, or increased disability on examination over a one-year period.

Granulomatosis with Polyangiitis (GPA) (Wegener's granulomatosis) and Microscopic Polyangiitis (MPA) [1-4, 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]

- 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) [31,152]

- 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

Systemic Lupus Erythematosus (SLE) [153,155,158,161-163]

- Adequate documentation of disease stability and/or improvement as indicated by one or more of the following when compared to pre-treatment baseline:
  - Improvement in the SELENA-SLEDAI-2K; **OR**
  - Reduction of baseline BILAG-2004 from A to B or from B to C/D, and no BILAG-2004 worsening in other organ systems, as defined by ≥2 new BILAG-2004 B; **OR**
  - No worsening (<0.30 points increase) in Physician's Global Assessment (PGA) score; **OR**
  - Seroconverted (negative)

Lupus Nephritis [115-117]

- 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) [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.

Complications of Transplanted Solid Organ (kidney, liver, lung, heart, pancreas) [133-138]

- Coverage may NOT be renewed.

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 ≥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 ≥10% decrease in CT score; stabilization defined as a ≤ 10% increase in CT score)

Idiopathic Membranous Nephropathy [172,175,177]

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

Pediatric Idiopathic Nephrotic Syndrome ‡ [170-173]

- Patient previously achieved beneficial disease response from the prior course of therapy; **AND**
- Patient is experiencing signs and symptoms of recurrent active disease necessitating additional doses (e.g., recurrence of nephrotic-range proteinuria with a dipstick ≥ 3+ [≥300 mg/dL] for 3 consecutive days OR urinary protein creatinine ratio [UPCR] ≥200 mg/mmol

[≥2 mg/mg] on a spot urine sample on 3 consecutive days, with or without reappearance of edema in a child who had previously achieved complete remission)

IgG4-Related Disease ‡ [178-182]

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

V. Dosage/Administration [1-5,9,19,23-26,32,34,40,42,44,50,62,80,83-89,91,94-98,102-111,115-118,122-125,128-133,135-137,140,152,164,165, 167,168, 170-173,175,178-184-185, 187-190]

Indication		Dose
CLL/SLL	Initial Therapy	375 mg/m² intravenously (IV) weekly for 12 doses; **OR** 375 mg/m² IV cycle 1, then 500 mg/m² every 28 days cycles 2-6 (6 total doses); **OR** 375 mg/m² IV cycle 1, followed by 500 mg/m² every 2 weeks for 4 doses, then 500 mg/m² every 28 days for 3 doses (8 total doses)
	Renewal Therapy	375 mg/m² IV every 3 months; **OR** 500 mg/ m² IV every 8 weeks
Waldenström Macroglobulinemia	Initial Therapy	375 mg/m² IV weekly for 12 doses
	Renewal Therapy	375 mg/m² IV once weekly for 4 doses per 6 month period; **OR** 375 mg/ m² IV every 8 weeks
Adult B-Cell Lymphomas, Castleman Disease, Primary Cutaneous B-Cell Lymphomas, or Adult HL	Initial Therapy	375 mg/m² IV once weekly for 4 – 8 doses in a 6 month period
	Renewal Therapy	375 mg/m² IV once weekly for 4 doses per 6 month period; **OR** 375 mg/ m² IV every 8 weeks
Pediatric Aggressive Mature B-Cell Lymphomas		<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). *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.* <u>**Consolidation* [courses 1 and 2 (CYM/CYVE)]**</u>

Indication	Dose
	375 mg/m² IV, one dose during each of the consolidation courses (Day 1) **Relapsed/Refractory** RCYVE – 375mg/m² IV on day 1 of each 21-day cycle RICE – 375 mg/m² IV on days 1 and 3 of courses 1 and 2, and on day 1 only of course 3 if needed. **Note: dosing and dosing schedules are highly variable and dependent on regimen used, please refer to NCCN and PI for additional protocols.*
Pediatric Mature B-Cell Acute Leukemia	**Induction* [courses 1 and 2 (COPDAM1 and COPDAM2)]** 375 mg/m² IV, two doses during each of the induction courses (Day -2 and Day 1). *During the 1st induction course, prednisone is given as part of the chemotherapy course, and should be administered prior to rituximab. Rituximab will be given 48 hours after the first infusion of rituximab.* **Consolidation* [courses 1 and 2 (CYM/CYVE)]** 375 mg/m² IV, one dose during each of the consolidation courses (Day 1) **Note: dosing and dosing schedules are highly variable and dependent on regimen used, please refer to NCCN and PI for additional protocols.*
CNS Lymphoma	**Intravenous administration** Initial Therapy: Up to 750 mg/m² weekly for 4 – 8 doses Renewal Therapy: 375 mg/m² IV once weekly for 4 doses per 6 month period; **OR** 375 mg/m² IV every 8 weeks **Intrathecal/Intraventricular administration** 25 mg weekly to twice weekly
ALL	375 mg/m² IV up to twice weekly for a total of 16 to 18 infusions (e.g., induction [days 1 and 7], salvage reinduction when necessary [days 1 and 7], consolidation [4 infusions: blocks 1, 3, 4, and 6], late intensification [days 1 and 7], late consolidation [2 infusions: blocks 7 and 9], and maintenance [6 infusions])
Hairy Cell Leukemia	375 mg/m² IV once weekly for 4 – 8 doses; **OR** 375mg/m² IV on days 1 and 15 every 28 days for 4 cycles, then 375mg/m² IV every 4 weeks for 4 cycles (up to 8 total cycles)
RA	1,000 mg IV on days 1 and 15, repeated every 24 weeks. May repeat up to every 16 weeks** following the previous infusion in patients requiring more frequent dosing based on clinical evaluation. ***Dose escalation criteria detailed in section IV must be met prior to increasing dosing frequency.*

Indication	Dose
Pemphigus Vulgaris	Initiation 1,000 mg IV on days 1 and 15; **OR** 375 mg/m² IV weekly for 4 doses Maintenance 500 mg IV at month 12 and repeat every 6 months thereafter or based on clinical evaluation Relapse 1,000 mg IV upon relapse, resumption of glucocorticoids may be considered **Subsequent infusions (maintenance and relapse) should be no sooner than 16 weeks after the previous infusion.*
AIHA	Warm-reactive disease 375 mg/m² IV weekly for 4 doses in a 6 month period; **OR** 1,000 mg IV on days 1 and 15 Cold agglutinin disease 375 mg/m² IV weekly for 4 doses in a 6 month period
Thrombocytopenic Purpura or Thrombotic Thrombocytopenic Purpura (TTP)	375 mg/m² IV weekly for 4 doses; **OR** 1,000 mg IV on days 1 and 15
Management of Immunotherapy-Related Toxicities	Bullous Dermatitis 1,000 mg IV every 2 weeks for 2 doses, then 500 mg IV at months 12 and 18 as needed Myositis 375 mg/m² IV weekly for 4 doses Myasthenia Gravis 375 mg/m² IV weekly for 4 doses; **OR** 500 mg/m² IV every 2 weeks for 2 doses Encephalitis 1,000 mg IV every 2 weeks for 2 doses; **OR** 375 mg/m² IV weekly for 4 doses
GPA (WG), MPA	Induction (Pediatric and Adult) 375 mg/m² IV weekly for 4 doses; **OR** – Adults: 1,000 mg IV on days 1 and 15; **OR** – Pediatric (up to a maximum of 1,000 mg per dose): o 575 mg/m² IV on days 1 and 15 (BSA ≤1.5m²) o 750 mg/m² IV on days 1 and 15 (BSA >1.5m²) Maintenance – Pediatric:

Indication	Dose
	○ 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 **Initial MAINTENANCE infusions should be no sooner than 16 weeks and no later than 24 weeks after the previous infusion if rituximab was used for initial induction therapy.* **Initial MAINTENANCE infusions should be initiated within 4 weeks following disease control when initial induction occurred with other standard of care immunosuppressants.*
cGVHD	375 mg/m² IV weekly for 4 doses, then 375 mg/m² IV monthly for 4 months **-OR-** 375 mg/m² IV weekly for 4 doses *(Note: A second course of 4 weekly doses may be administered 8 weeks after initial therapy for patients with lack of or incomplete response.)* **-OR-** 375 mg/m² IV weekly for 4 – 8 doses
Hematopoietic Cell Transplantation	Conditioning: 375 mg/m² IV for 1 day before transplant, then 1000 mg/m² IV on days 1,8, and 15 after transplant
Multiple Sclerosis	1,000 mg IV on days 1 and 15, repeat every 6 months
NMOSD	1,000 mg IV once on days 1 and 15, repeat every 6 months **-OR-** 375 mg/m² once weekly for 4 weeks, repeat every 6 months
Histiocytic Neoplasms – Rosai-Dorfman Disease	500 mg/m² 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 **-OR-** 375 mg/m² IV once weekly for 4 doses
Myasthenia Gravis (unrelated to immunotherapy-related toxicity)	1,000 mg IV on days 1 and 15, may repeat a full or partial course every 6 months **-OR-** 375 mg/m² IV once weekly for 4 doses, may repeat a full or partial course every 6 months
Pediatric Hodgkin Lymphoma	375 mg/m² IV on day 1 of every 2-3 week cycle for a total of 3 cycles

Indication	Dose
Complications of transplanted solid organ (kidney, liver, lung, heart, pancreas)	– Adults and pediatrics weighing ≥0.5 m^2: 375 mg/m^2 weekly for up to 4 doses – Pediatrics weighing <0.5 m^2: 12.5 mg/kg weekly for up to 4 doses
Antisynthetase Syndrome-Related Interstitial Lung Disease	1,000 mg IV on days 1 and 15 repeated every 6 months **-OR-** 375 mg/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	Induction: 375 mg/m^2 IV once weekly for 1-4 doses **-OR-** 1,000 mg IV on days 1 and 15 **Subsequent infusions (maintenance and relapse) may be administered at either induction schedule above and should be repeated no sooner than every 6 months.*
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)	

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 June 2024.
2. Truxima [package insert]. Incheon, Republic of Korea; Celltrion, Inc.; February 2024. Accessed June 2024.
3. Ruxience [package insert]. New York, NY; Pfizer, Inc; October 2023. Accessed June 2024.
4. Riabni [package insert]. Thousand Oaks, CA; Amgen, Inc.; February 2023. Accessed June 2024.
5. Referenced with permission from the NCCN Drugs & Biologics Compendium (NCCN Compendium®) rituximab. National Comprehensive Cancer Network, 2024. 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 June 2024.
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.
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 (2019). Intravenous Cancer Drug Waste Issue Brief. Retrieved from http://www.hoparx.org/images/hopa/advocacy/Issue-Briefs/Drug_Waste_2019.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 2.2024. National Comprehensive Cancer Network, 2024. 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 June 2024.
45. Imbruvica [package insert]. Horsham, PA; Janssen Biotech, Inc. May 2024. Accessed June 2024.
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 1.2024. National Comprehensive Cancer Network, 2024. 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 July 2024.
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-Khouri 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 Immunotherapy-Related Toxicities, Version 1.2024. National Comprehensive Cancer Network, 2024. 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 July 2024.

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 1.2024. National Comprehensive Cancer Network, 2024. 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 June 2024.
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 JWJ, 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. (2024). Chronic immunosuppressive therapy for myasthenia gravis. In Shefner JM, Goddeau RP (Eds.), *UptoDate*. Last updated: February 27, 2024. Accessed: July 2024. 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, Pillon 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, Obrtlíková 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, Chanet 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 1.2024. National Comprehensive Cancer Network, 2024. 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 July 2024.
129. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Hematopoietic Cell Transplantation, Version 1.2024. National Comprehensive Cancer Network, 2024. 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 July 2024.
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. (2023). Kidney transplantation in children: Immunosuppression. In Niaudet P, Hoppin AG (Eds.), *UptoDate*. Last updated: Dec 14,2023. Accessed: July 2024. Available from https://www.uptodate.com/contents/kidney-transplantation-in-children-immunosuppression?search=pediatric%20antibody%20mediated%20rejection&source=search_result&selectedTitle=1~150&usage_type=default&display_rank=1#H181637276.

134. Parajuli S, Brennan DC. (2022). Kidney transplantation in adults: Prevention and treatment of antibody-mediated rejection. In Legendre C, Vella J, Lam AQ (Eds.), *UptoDate*. Last updated: September 16, 2022. Accessed: July 2024. Available from https://www.uptodate.com/contents/kidney-transplantation-in-adults-prevention-and-treatment-of-antibody-mediated-rejection?search=Antibody%20mediated%20rejection&source=search_result&selectedTitle=1~150&usage_type=default&display_rank=1.

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: July 2024. Available from https://www.uptodate.com/contents/evaluation-and-treatment-of-antibody-mediated-lung-transplant-rejection.

137. Alhamad T, Kukla A, Stratta RJ. (2022). Pancreas allograft rejection. In Brennan DC, Nathan DM, Lam AQ (Eds.), *UptoDate*. Last updated: December 7, 2022. Accessed: July 2024. 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 neuromyelits 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. Tiacci 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, Dhillon 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. (2024). Membranous nephropathy: Pathogenesis and etiology. In Glasscock RJ, Fervenza FC, Lam AQ (Eds.), *UptoDate*. Last updated: March 4, 2024. Accessed: July 2024. Available from https://www.uptodate.com/contents/membranous-nephropathy-pathogenesis-and-etiology

177. De Vriese AS, Wetzels JFM, Cattran DC. (2024). Membranous nephropathy: Treatment and prognosis. In Glasscock RJ, Fervenza FC, Lam AQ (Eds.), *UptoDate*. Last updated: March 8, 2024. Accessed: July 2024. 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, at 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 singlecenter 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, 2024. 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 July 2024.
188. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Temozolomide + Rituximab: Central Nervous System Cancers Chemotherapy Order Template, CNS82. National Comprehensive Cancer Network, 2024. 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 July 2024.
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, 2024. 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 July 2024.

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, 2024. 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 July 2024.
191. 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 01/26/2024 with effective date 02/01/2024. Accessed June 2024.
192. 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 05/21/2024 with effective date 05/30/2024. Accessed June 2024.
193. Palmetto GBA. Local Coverage Article: Billing and Coding: Rituximab (A56380). Centers for Medicare & Medicaid Services, Inc. Updated on 08/11/2023 with effective date 10/01/2023. Accessed June 2024.
194. CGS Administrators, LLC. Local Coverage Article: Billing and Coding: Immune Thrombocytopenia (ITP) Therapy (A57160). Centers for Medicare & Medicaid Services, Inc. Updated on 02/29/2024 with effective date 03/07/2024. Accessed June 2024.
195. 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 11/15/2023 with effective date 11/22/2023. Accessed June 2024.

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
D89.84	IgG4-related disease
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
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
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
M31.30	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
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)

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

Nondiscrimination & Language Access Policy

Discrimination is Against the Law. Aspirus Health Plan, Inc. complies with applicable Federal civil rights laws and does not discriminate on the basis of race, color, national origin, age, disability, or sex, (including sex characteristics, including intersex traits; pregnancy or related conditions; sexual orientation, gender identity and sex stereotypes), consistent with the scope of sex discrimination described at 45 CFR § 92.101(a)(2). Aspirus Health Plan, Inc. does not exclude people or treat them less favorably because of race, color, national origin, age, disability, or sex.

Aspirus Health Plan, Inc.:
Provides people with disabilities reasonable modifications and free appropriate auxiliary aids and services to communicate effectively with us, such as:

- Qualified sign language interpreters.
- Written information in other formats (large print, audio, accessible electronic formats, other formats).

Provides free language assistance services to people whose primary language is not English, which may include:

- Qualified interpreters.
- Information written in other languages.

If *you* need reasonable modifications, appropriate auxiliary aids and services, or language assistance services, contact the Nondiscrimination Grievance Coordinator at the address, phone number, fax number, or email address below.

If *you* believe that Aspirus Health Plan, Inc. has failed to provide these services or discriminated in another way on the basis of race, color, national origin, age, disability, or sex, *you* can file a *grievance* with:

Nondiscrimination Grievance Coordinator
Aspirus Health Plan, Inc.
PO Box 1890
Southampton, PA 18966-9998
Phone: 1-866-631-5404 (TTY: 711)
Fax: 763-847-4010
Email: customerservice@aspirushealthplan.com

You can file a *grievance* in person or by mail, fax, or email. If *you* need help filing a *grievance*, the Nondiscrimination Grievance Coordinator is available to help *you*.

You can also file a civil rights complaint with the U.S. Department of Health and Human Services, Office for Civil Rights, electronically through the Office for Civil Rights Complaint Portal, available at https://ocrportal.hhs.gov/ocr/portal/lobby.jsf, or by mail or phone at:

U.S. Department of Health and Human Services
200 Independence Avenue, SW
Room 509F, HHH Building
Washington, D.C. 20201
1.800.368.1019, 800.537.7697 (TDD)

Complaint forms are available at http://www.hhs.gov/ocr/office/file/index.html. This notice is available at Aspirus Health Plan, Inc.'s website: https://aspirushealthplan.com/webdocs/70021-AHP-NonDiscrim_Lang-Assist-Notice.pdf.

Language Assistance Services

Albanian: KUJDES: Nëse flitni shqip, për ju ka në dispozicion shërbime të asistencës gjuhësore, pa pagesë.Telefononi në 1-800-332-6501 (TTY: 711).

Arabic تنبيه: إذا كنت تتحدث اللغة العربية، فإن خدمات المساعدة اللغوية متاحة لك مجاناً. اتصل بن اعلى رقم الهاتف1-800-332-6501(رقم هاتف الصم والبك : 711)

French: ATTENTION: Si vous parlez français, des services d'aide linguistique vous sont proposés gratuitement. Appelez le 1-800-332-6501 (ATS: 711).

German: ACHTUNG: Wenn Sie Deutsch sprechen, stehen Ihnen kostenlos sprachliche Hilfsdienstleistungen zurVerfügung. Rufnummer: 1-800-332-6501 (TTY: 711).

Hindi: _यान द_: य_द आप ि हंदी बोलते ह_ तो आपके ि लए मु_त म_ भाषा सहायता सेवाएं उपल_ध ह_।1-800-332-6501 (TTY: 711) पर कॉल कर_।

Hmong: LUS CEEV: Yog tias koj hais lus Hmoob, cov kev pab txog lus, muaj kev pab dawb rau koj. Hu rau 1-800-332-6501 (TTY: 711).

Korean: 주의: 한국어를 사용하시는 경우, 언어 지원 서비스를 무료로 이용하실 수 있습니다.1-800-332-6501 (TTY: 711)번으로 전화해 주십시오.

Polish: UWAGA: Jeżeli mówisz po polsku, możesz skorzystać z bezpłatnej pomocy językowej. Zadzwoń pod numer1-800-332-6501 (TTY: 711).

Russian: ВНИМАНИЕ: Если вы говорите на русском языке, то вам доступны бесплатные услуги перевода.Звоните 1-800-332-6501 (телетайп: 711).

Spanish: ATENCIÓN: si habla español, tiene a su disposición servicios gratuitos de asistencia lingüística. Llame al1-800-332-6501 (TTY: 711).

Tagalog: PAUNAWA: Kung nagsasalita ka ng Tagalog, maaari kang gumamit ng mga serbisyo ng tulong sa wika nangwalang bayad. Tumawag sa 1-800-332-6501 (TTY: 711).

Traditional Chinese: 注意：如果您使用繁體中文， 您可以免費獲得語言援助服務。請 致電 1-800-332-6501 (TTY: 711)

Vietnamese: CHÚ Ý: Nếu bạn nói Tiếng Việt, có các dịch vụ hỗ trợ ngôn ngữ miễn phí dành cho bạn. Gọi số 1-800-332-6501 (TTY: 711).

Pennsylvania Dutch: Wann du Deitsch (Pennsylvania German / Dutch) schwetzscht, kannscht du mitaus Koschte ebbergricke, ass dihr helft mit die englisch Schprooch. Ruf selli Nummer uff: Call 1-800-332-6501 (TTY: 711).

Lao: ໂປດຊາບ: ຖ້າວ່າ ທ່ານເວົ້າພາສາ ລາວ, ການບໍລິການຊ່ວຍເຫຼືອດ້ານພາສາ,ໂດຍບໍ່ເສັງຄ່າ, ແມ່ນມີພ້ອມໃຫ້ທ່ານ. ໂທຣ 1-800-332-6501 (TTY: 711).