

Rituxan® (rituximab) (Intravenous)

Document Number: IC-0109

Last Review Date: 12/04/2018

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

I. Length of Authorization

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

- Maintenance therapy for oncology indications (excluding ALL) may be renewed for up to a maximum of 2 years.
- Acute lymphoblastic leukemia (ALL) may not be renewed.
- Relapse therapy for pemphigus vulgaris must be at least 16 weeks past a prior infusion

II. Dosing Limits

A. Quantity Limit (max daily dose) [Pharmacy Benefit]:

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

B. Max Units (per dose and over time) [Medical Benefit]:

Oncology Indications
<p><u>Chronic Lymphocytic Leukemia (CLL)/Small Lymphocytic Leukemia (SLL):</u></p> <ul style="list-style-type: none"> • Initial therapy: <ul style="list-style-type: none"> ○ Loading dose: 100 units x 1 dose ○ Subsequent doses: 130 units every 28 days x 5 doses per 6 months • Renewal therapy: 100 units per dose every 8 weeks x 4 doses per 6 months
<p><u>Immunotherapy Toxicity Treatment:</u></p> <ul style="list-style-type: none"> • 100 units per dose weekly x 4 doses in a 6 month period
<p><u>All other oncology indications:</u></p> <ul style="list-style-type: none"> • Initial therapy: 100 units per dose weekly x 8 doses per 6 months • Renewal therapy: 100 units per dose every 8 weeks x 4 doses per 6 months
Non-Oncology Indications

<ul style="list-style-type: none"> • Initiation: 100 units every 14 days x 2 doses in a 12 month period • Maintenance: 50 units every 16 weeks
<u>GPA(WG)/MPA:</u> <ul style="list-style-type: none"> • Induction: 100 units per dose weekly x 4 doses in a 4 month period • Initial Maintenance: 100 units x 2 doses in a 6 month period • Subsequent Maintenance: 50 units every 6 months
<u>cGVHD</u> <ul style="list-style-type: none"> • 100 units per dose weekly x 4 doses, then 100 units monthly x 4 months
<u>All other non-oncology indications:</u> <ul style="list-style-type: none"> • 100 units per dose weekly x 4 doses in a 6 month period

III. Initial Approval Criteria

Coverage is provided in the following conditions:

- Patient must be screened for HBV infection (i.e., HBsAg and anti-HBc) prior to initiating therapy; **AND**

Oncology Indications:

- Patient is CD20-positive; **AND**

Acute Lymphoblastic Leukemia (ALL) ‡

- Induction/Consolidation Treatment
 - Patient's disease is Philadelphia chromosome-negative (Ph⁻); **AND**
 - Patient is at least 15 years of age; **AND**
 - Used in combination with an anthracycline, cyclophosphamide and vincristine based regimen
- Relapsed/Refractory Treatment
 - Used as a component of MOpAD regimen (methotrexate, vincristine, pegaspargase, dexamethasone); **AND**
 - Patient's disease is Philadelphia chromosome-negative (Ph⁻); **OR**
 - Patient's disease is Philadelphia chromosome-positive (Ph⁺) and refractory to tyrosine kinase inhibitors (e.g., imatinib, bosutinib, ponatinib, nilotinib, etc.)

CNS Cancer ‡

- Patient has leptomeningeal metastases from lymphomas; **AND**
 - Rituximab will be administered intrathecally; **OR**
- Patient has primary CNS lymphoma; **AND**
 - Used intrathecally if patient has positive disease on lumbar puncture or spinal MRI; **OR**
 - Patient will receive in combination with a methotrexate-containing regimen as a component of induction therapy and/or consolidation therapy with a complete response (CR) or a complete response unconfirmed (CRu) to induction therapy; **OR**
 - Patient has relapsed or refractory disease and will receive rituximab as a single agent, or in combination with temozolomide, lenalidomide or high-dose methotrexate

Hodgkin's lymphoma ‡

- Patient has nodular lymphocyte-predominant disease

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

- Used in combination with fludarabine and cyclophosphamide (FC) †; **OR**
- Used as first-line therapy in combination with chlorambucil or bendamustine, or alemtuzumab in patients without del(17p)/TP53 mutation; **OR**
- Used as first line therapy in combination with fludarabine in patients without del(17p)/TP53 and del(11q) mutations; **OR**
- Used as first line therapy in combination with high-dose methylprednisolone and alemtuzumab with del(17p)/TP53 mutations; **OR**
- Used for relapsed or refractory disease; **AND**
 - Used in combination with venetoclax or idelalisib or lenalidomide or high-dose methylprednisolone; **OR**
 - Used in combination with chlorambucil without del(17p) mutations

Waldenström's macroglobulinemia/Lymphoplasmacytic Lymphoma ‡

Non-Hodgkin's lymphomas (NHL) † including, but not limited to, the following:

- AIDS-related B-Cell Lymphoma ‡
 - Used in combination with other chemotherapy; **AND**
 - Disease is related to Burkitt Lymphoma or diffuse large B-cell lymphoma
- Burkitt Lymphoma ‡
 - Used in combination with other chemotherapy
- Castleman's Disease ‡
 - Patient has multicentric disease; **OR**
 - Patient has unicentric disease; **AND**
 - Used as second-line therapy for relapsed or refractory disease; **OR**
 - Used for patients with symptoms after resection or unresectable disease
- Diffuse Large B-Cell Lymphoma †
- Low-grade or Follicular Lymphoma †
- Gastric & Non-Gastric MALT Lymphoma ‡
- Hairy Cell Leukemia ‡
 - Used for relapsed or refractory disease; **OR**
 - Used in patients with a less than complete response (CR) to initial therapy
- Mantle Cell Lymphoma ‡
- Nodal & Splenic Marginal Zone Lymphoma ‡
- Post-transplant lymphoproliferative disorder (PTLD) ‡
 - Patient has had solid organ transplant or allogeneic hematopoietic stem cell transplantation; **AND**

- Used as first-line or subsequent therapy for monomorphic or polymorphic disease;
OR
- Used as maintenance therapy for polymorphic disease after achieving a complete response on first-line therapy
- Primary Cutaneous B-Cell Lymphomas ‡
 - Used for generalized (skin only), marginal zone or follicle center disease

Management of Immunotherapy-Related Toxicities ‡

- Patient has been receiving therapy with an immune checkpoint inhibitor (e.g., cemiplimab, nivolumab, pembrolizumab, atezolizumab, avelumab, durvalumab, ipilimumab, etc.); **AND**
- Patient has non-viral encephalitis related to their immunotherapy; **AND**
- Patient is autoimmune-encephalopathy-antibody positive; **AND**
- Patient is refractory to methylprednisolone and/or IV immunoglobulin (IVIG).

Non-Oncology Indications:

Rheumatoid arthritis (RA) †

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

Pemphigus vulgaris †

- Adult patient (18 years or older); **AND**
- Patient has a diagnosis of pemphigus vulgaris as determined by 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**
 - Presence of autoantibodies as detected by direct or indirect immunofluorescence; **AND**

- Patient has moderate to severe disease as assessed utilizing an objective measure/tool (i.e. PDAI, PSS, ABSIS); **AND**
- Patient is on combination glucocorticoid therapy; **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) †

- Adult patient (18 years or older); **AND**
- Used in combination with glucocorticoids (e.g., prednisone, methylprednisolone, etc.)

Thrombocytopenic purpura ‡

- Patient has previously failed or has a contraindication or intolerance to therapy with corticosteroids; **AND**
- The patient is at increased risk for bleeding as indicated by platelet count (within the previous 28 days) less than $30 \times 10^9/L$ (30,000/mm³); **AND**
- Patient diagnosis includes one of the following:
 - Primary thrombocytopenia
 - Idiopathic (Immune) thrombocytopenia purpura (ITP)
 - Evan’s syndrome
 - Congenital and hereditary thrombocytopenic purpura
 - Thrombotic thrombocytopenic purpura in patients with ADAMTS13-deficiency

Chronic graft-versus-host disease (cGVHD) ‡

- Patient is post-allogeneic stem cell transplant (generally 3 or more months); **AND**
- Patient has failed one or more previous lines of systemic therapy for the treatment of cGVHD (e.g., corticosteroids or immunosuppressants such as cyclosporine); **AND**
- Patient must try and have an inadequate response, contraindication, or intolerance to at least a three (3) month trial of ibrutinib.

Autoimmune Hemolytic Anemia (AIHA) ‡

- 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

† FDA-labeled indication(s); ‡ Compendia recommended indication(s)

IV. Renewal Criteria

Coverage can be renewed based upon the following criteria:

- Patient continues to meet criteria identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include the following: severe infusion reactions, tumor lysis syndrome (TLS), severe mucocutaneous reactions, progressive multifocal leukoencephalopathy (PML), viral

hepatitis, serious bacterial, fungal, or viral infections, cardiac arrhythmias, renal toxicity, bowel obstruction or perforation; **AND**

Oncology Indications:

- Tumor response with stabilization of disease or decrease in size of tumor or tumor spread; **AND**
- Patient has not exceeded dosing or duration limits as defined in Sections I, II, and V.

Non-Oncology Indications:

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 and/or an improvement on a disease activity scoring tool [e.g. an improvement on a composite scoring index such as Disease Activity Score-28 (DAS28) of 1.2 points or more or a $\geq 20\%$ improvement on the American College of Rheumatology-20 (ACR20) criteria]; **AND**
- Dose escalation (up to the maximum dose and frequency specified below) may occur upon clinical review on a case by case basis provided that the patient has:
 - Shown an initial response to therapy; **AND**
 - Received a minimum of one maintenance dose at the dose and interval specified below; **AND**
 - Responded to therapy with subsequent loss of response

Thrombocytopenic purpura

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

Thrombotic thrombocytopenic purpura (TTP)

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

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

- Disease response as indicated by disease control and improvement in signs and symptoms of condition compared to baseline; **AND**
- A decrease 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

- Patient is currently receiving tapering doses of corticosteroids or has discontinued use of corticosteroids; **AND**
 - Disease response as indicated by complete epithelialization of lesions and improvement in signs and symptoms of condition compared to baseline; **OR**
 - Patient has not experienced continued development of new lesions, continued extension of old lesions, or failure of established lesions to begin to heal despite therapy; **OR**

- For Relapses ONLY: Patient has had active 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

Chronic graft-versus-host disease (cGVHD)

- Disease response as indicated by improvement in patient-reported symptoms or clinician assessments (e.g., manifestations of disease to the skin, oral cavity, musculoskeletal system, etc.)

Autoimmune hemolytic anemia (AIHA)

- Disease response as indicated by improvement in anemia signs and symptoms (e.g., dyspnea, fatigue, etc.) as well as: improvement in laboratory values (Hb/Hct), reduced transfusion needs, and/or reduced glucocorticoid use

V. Dosage/Administration

Indication		Dose
CLL/SLL	Initial Therapy	375 mg/m ² weekly x 8 doses; OR 375 mg/m ² cycle 1, then 500 mg/m ² every 28 days cycles 2-6 (6 total doses)
	<i>Renewal Therapy</i>	375 mg/m ² once weekly for 4 doses per 6 month period; OR 375 mg/ m ² every 8 weeks
NHL, PTLD, Waldenström's, Castleman's, or HL	Initial Therapy	375 mg/m ² once weekly for 4 - 8 doses in a 6 month period
	<i>Renewal Therapy</i>	375 mg/m ² once weekly for 4 doses per 6 month period; OR 375 mg/ m ² every 8 weeks
CNS Lymphoma		<u>Intravenous</u> <u>Initial:</u> 375 mg/m ² once weekly for 4 - 8 doses in a 6 month period <u>Renewal Therapy:</u> 375 mg/m ² once weekly for 4 doses per 6 month period; OR 375 mg/ m ² every 8 weeks <u>Intrathecal/Intraventricular</u> 10-40 mg weekly to every 3 weeks
ALL		375 mg/m ² once weekly for 4 - 8 doses in a 6 month period
RA		1,000 mg on days 1 and 15, repeated every 24 weeks. May repeat up to every 16 weeks in patients requiring more frequent dosing based on clinical evaluation.
Pemphigus Vulgaris		<u>Initiation</u> – Administer 1,000 mg on days 1 and 15 in combination with tapering doses of glucocorticoids – Used in combination with prednisone (or equivalent): ▪ Moderate disease: 0.5 mg/kg/day tapered over 3 months ▪ Severe disease: 1 mg/kg/day tapered over 6 months

	<p><u>Maintenance</u></p> <ul style="list-style-type: none"> Administer 500 mg at month 12 and repeat every 6 months thereafter or based on clinical evaluation. <p><u>Relapse</u></p> <ul style="list-style-type: none"> Administer 1000 mg upon relapse, resumption of glucocorticoids may be considered. <p><i>*Subsequent infusions (maintenance and relapse) should be no sooner than 16 weeks after the previous infusion.</i></p>
Thrombocytopenia, AIHA, or Immunotherapy Toxicity Treatment	375 mg/m ² weekly x 4 doses in a 6 month period
GPA (WG), MPA	<p><u>Induction</u></p> <ul style="list-style-type: none"> 375 mg/m² weekly x 4 doses, initially. <p><u>Maintenance</u></p> <ul style="list-style-type: none"> Administer 1,000 mg (as 2 x 500mg doses) for two doses separated by two weeks, then followed by 500mg every 6 months thereafter based on clinical evaluation. <p><i>*Initial MAINTENANCE infusions should be no sooner than 16 weeks and no later than 24 weeks after the previous infusion if Rituxan was used for initial induction therapy.</i></p> <p><i>*Initial MAINTENANCE infusions should be initiated within 4 weeks following disease control when initial induction occurred with other standard of care immunosuppressants.</i></p>
cGVHD	375 mg/m ² weekly x 4 doses, then 375 mg/m ² monthly x 4 months

VI. Billing Code/Availability Information

JCode:

- J9310 – Injection, rituximab, 100 mg; 1 billable unit = 100 mg (*Discontinued after 12/31/2018*)
- J9312 – Injection, rituximab, 10 mg; 1 billable unit = 10 mg (*Effective 1/1/2019*)

NDC:

- Rituxan 100 mg/10 mL single-use vial for injection: 50242-0051-xx
- Rituxan 500 mg/50 mL single-use vial for injection: 50242-0053-xx

VII. References

- Rituxan [package insert]. South San Francisco, CA; Genentech, Inc; October 2018. Accessed November 2018.
- Referenced with permission from the NCCN Drugs & Biologics Compendium (NCCN Compendium®) rituximab. National Comprehensive Cancer Network, 2018. 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 November 2018.

3. 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.
4. 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.
5. 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.
6. 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.
7. 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.
8. 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.
9. 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.
10. 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.
11. 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.
12. 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.
13. 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.
14. 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.
15. 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.

16. 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.
17. 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.
18. 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.
19. 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.
20. 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.
21. Cutler C, Miklos D, Kim HT, et al, "Rituximab for Steroid-Refractory Chronic Graft-Versus-Host Disease," *Blood*, 2006, 108(2):756-62.
22. 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.
23. 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.
24. 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.
25. 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.
26. Berentsen S, Ulvestad E, Gjertsen BT, et al: Rituximab for primary chronic cold agglutinin disease: a prospective study of 37 courses of therapy in 27 patients. *Blood* 2004; 103(8):2925-2928.
27. 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.
28. 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.
29. 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.

30. 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.
31. YW Shin, ST Lee, KI Park, et al. Treatment strategies for autoimmune encephalitis. *Ther Adv Neurol Disord.* 2017 Aug 16;11:1756285617722347. doi: 10.1177/1756285617722347. eCollection 2018. Review.
32. 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>
33. Grover, S. Scoring Systems in Pemphigus. *Indian J Dermatol.* 2011 Mar-Apr; 56(2): 145–149. doi: 10.4103/0019-5154.80403
34. 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
35. Lambert MP, Gernsheimer TB. Clinical updates in adult immune thrombocytopenia. *Blood.* 2017. 129:2829-2835. doi:10.1182/blood-2017-03-754119
36. H Schulz, H Pels, I Schmidt-Wolf, et al. Intraventricular treatment of relapsed central nervous system lymphoma with the anti-CD20 antibody rituximab. *Haematologica* January 2004 89: 753-754
37. First Coast Service Options, Inc. Local Coverage Determination (LCD): Rituximab (Rituxan®) (L33746). Centers for Medicare & Medicaid Services, Inc. Updated on 9/22/2017 with effective date 10/1/2017. Accessed November 2018.
38. National Government Services, Inc. Local Coverage Article: Rituximab (Rituxan®) (effective 2010) - Related to LCD L33394 (A52452). Centers for Medicare & Medicaid Services, Inc. Updated on 7/6/2018 with effective date of 6/7/2018. Accessed November 2018.
39. Palmetto GBA. Local Coverage Determination: Rituximab (Rituxan®) (L35026). Centers for Medicare & Medicaid Services, Inc. Updated on 05/11/2018 with effective date of 06/13/2018. Accessed November 2018.
40. Wisconsin Physicians Service Insurance Corp. Local Coverage Article: Chemotherapy Agents for Non-Oncologic Conditions (A55639). Centers for Medicare & Medicaid Services, Inc. Updated on 09/18/2018 with effective date 10/01/2018. Accessed November 2018.

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
C82.00	Follicular lymphoma grade I, unspecified site
C82.01	Follicular lymphoma grade I, lymph nodes of head, face and neck
C82.02	Follicular lymphoma, grade I, intrathoracic lymph nodes
C82.03	Follicular lymphoma grade I, intra-abdominal lymph nodes
C82.04	Follicular lymphoma grade I, lymph nodes of axilla and upper limb
C82.05	Follicular lymphoma grade I, lymph nodes of inguinal regional and lower limb
C82.06	Follicular lymphoma grade I, intrapelvic lymph nodes
C82.07	Follicular lymphoma grade I, spleen
C82.08	Follicular lymphoma grade I, lymph nodes of multiple sites
C82.09	Follicular lymphoma grade I, extranodal and solid organ sites
C82.10	Follicular lymphoma grade II, unspecified site
C82.11	Follicular lymphoma grade II, lymph nodes of head, face and neck
C82.12	Follicular lymphoma, grade II, intrathoracic lymph nodes
C82.13	Follicular lymphoma grade II, intra-abdominal lymph nodes
C82.14	Follicular lymphoma grade II, lymph nodes of axilla and upper limb
C82.15	Follicular lymphoma grade II, lymph nodes of inguinal region and lower limb
C82.16	Follicular lymphoma grade II, intrapelvic lymph nodes
C82.17	Follicular lymphoma grade II, spleen
C82.18	Follicular lymphoma grade II, lymph nodes of multiple sites
C82.19	Follicular lymphoma grade II, extranodal and solid organ sites
C82.20	Follicular lymphoma grade III, unspecified, unspecified site
C82.21	Follicular lymphoma grade III, unspecified, lymph nodes of head, face and neck
C82.22	Follicular lymphoma, grade III, unspecified, intrathoracic lymph nodes
C82.23	Follicular lymphoma grade III, unspecified, intra-abdominal lymph nodes
C82.24	Follicular lymphoma grade III, unspecified, lymph nodes of axilla and upper limb
C82.25	Follicular lymphoma grade III, unspecified, lymph nodes of inguinal region and lower limb
C82.26	Follicular lymphoma grade III, unspecified, intrapelvic lymph nodes
C82.27	Follicular lymphoma grade III, unspecified, spleen
C82.28	Follicular lymphoma grade III, unspecified, lymph nodes of multiple sites
C82.29	Follicular lymphoma grade III, unspecified, extranodal and solid organ sites
C82.30	Follicular lymphoma grade IIIa, unspecified site
C82.31	Follicular lymphoma grade IIIa, lymph nodes of head, face and neck
C82.32	Follicular lymphoma, grade IIIa, intrathoracic lymph nodes

C82.33	Follicular lymphoma grade IIIa, intra-abdominal lymph nodes
C82.34	Follicular lymphoma grade IIIa, lymph nodes of axilla and upper limb
C82.35	Follicular lymphoma grade IIIa, lymph nodes of inguinal region and lower limb
C82.36	Follicular lymphoma grade IIIa, intrapelvic lymph nodes
C82.37	Follicular lymphoma grade IIIa, spleen
C82.38	Follicular lymphoma grade IIIa, lymph nodes of multiple sites
C82.39	Follicular lymphoma grade IIIa, extranodal and solid organ sites
C82.40	Follicular lymphoma grade IIIb, unspecified site
C82.41	Follicular lymphoma grade IIIb, lymph nodes of head, face and neck
C82.42	Follicular lymphoma, grade IIIb, intrathoracic lymph nodes
C82.43	Follicular lymphoma grade IIIb, intra-abdominal lymph nodes
C82.44	Follicular lymphoma grade IIIb, lymph nodes of axilla and upper limb
C82.45	Follicular lymphoma grade IIIb, lymph nodes of inguinal region and lower limb
C82.46	Follicular lymphoma grade IIIb, intrapelvic lymph nodes
C82.47	Follicular lymphoma grade IIIb, spleen
C82.48	Follicular lymphoma grade IIIb, lymph nodes of multiple sites
C82.49	Follicular lymphoma grade IIIb, extranodal and solid organ sites
C82.50	Diffuse follicle center lymphoma, unspecified site
C82.51	Diffuse follicle center lymphoma, lymph nodes of head, face and neck
C82.52	Diffuse follicle center lymphoma, intrathoracic lymph nodes
C82.53	Diffuse follicle center lymphoma, intra-abdominal lymph nodes
C82.54	Diffuse follicle center lymphoma, lymph nodes of axilla and upper limb
C82.55	Diffuse follicle center lymphoma, lymph nodes of inguinal region and lower limb
C82.56	Diffuse follicle center lymphoma, intrapelvic lymph nodes
C82.57	Diffuse follicle center lymphoma, spleen
C82.58	Diffuse follicle center lymphoma, lymph nodes of multiple sites
C82.59	Diffuse follicle center lymphoma, extranodal and solid organ sites
C82.60	Cutaneous follicle center lymphoma, unspecified site
C82.61	Cutaneous follicle center lymphoma, lymph nodes of head, face and neck
C82.62	Cutaneous follicle center lymphoma, intrathoracic lymph nodes
C82.63	Cutaneous follicle center lymphoma, intra-abdominal lymph nodes
C82.64	Cutaneous follicle center lymphoma, lymph nodes of axilla and upper limb
C82.65	Cutaneous follicle center lymphoma, lymph nodes of inguinal region and lower limb
C82.66	Cutaneous follicle center lymphoma, intrapelvic lymph nodes
C82.67	Cutaneous follicle center lymphoma, spleen
C82.68	Cutaneous follicle center lymphoma, lymph nodes of multiple sites
C82.69	Cutaneous follicle center lymphoma, extranodal and solid organ sites
C82.80	Other types of follicular lymphoma, unspecified site
C82.81	Other types of follicular lymphoma, lymph nodes of head, face and neck

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

C83.31	Diffuse large B-cell lymphoma, lymph nodes of head, face, and neck
C83.32	Diffuse large B-cell lymphoma intrathoracic lymph nodes
C83.33	Diffuse large B-cell lymphoma intra-abdominal lymph nodes
C83.34	Diffuse large B-cell lymphoma lymph nodes of axilla and upper limb
C83.35	Diffuse large B-cell lymphoma, lymph nodes of inguinal region and lower limb
C83.36	Diffuse large B-cell lymphoma intrapelvic lymph nodes
C83.37	Diffuse large B-cell lymphoma, spleen
C83.38	Diffuse large B-cell lymphoma lymph nodes of multiple sites
C83.39	Diffuse large B-cell lymphoma extranodal and solid organ sites
C83.70	Burkitt lymphoma, unspecified site
C83.71	Burkitt lymphoma, lymph nodes of head, face, and neck
C83.72	Burkitt lymphoma, intrathoracic lymph nodes
C83.73	Burkitt lymphoma, intra-abdominal lymph nodes
C83.74	Burkitt lymphoma, lymph nodes of axilla and upper limb
C83.75	Burkitt lymphoma, lymph nodes of inguinal region and lower limb
C83.76	Burkitt lymphoma, intrapelvic lymph nodes
C83.77	Burkitt lymphoma, spleen
C83.78	Burkitt lymphoma, lymph nodes of multiple sites
C83.79	Burkitt lymphoma, extranodal and solid organ sites
C83.80	Other non-follicular lymphoma, unspecified site
C83.81	Other non-follicular lymphoma, lymph nodes of head, face and neck
C83.82	Other non-follicular lymphoma, intrathoracic lymph nodes
C83.83	Other non-follicular lymphoma, intra-abdominal lymph nodes
C83.84	Other non-follicular lymphoma, lymph nodes of axilla and upper limb
C83.85	Other non-follicular lymphoma, lymph nodes of inguinal region and lower limb
C83.86	Other non-follicular lymphoma, intrapelvic lymph nodes
C83.87	Other non-follicular lymphoma, spleen
C83.88	Other non-follicular lymphoma, lymph nodes of multiple sites
C83.89	Other non-follicular lymphoma, extranodal and solid organ sites
C83.90	Non-follicular (diffuse) lymphoma, unspecified site
C83.91	Non-follicular (diffuse) lymphoma, unspecified lymph nodes of head, face, and neck
C83.92	Non-follicular (diffuse) lymphoma, unspecified intrathoracic lymph nodes
C83.93	Non-follicular (diffuse) lymphoma, unspecified intra-abdominal lymph nodes
C83.94	Non-follicular (diffuse) lymphoma, unspecified lymph nodes of axilla and upper limb
C83.95	Non-follicular (diffuse) lymphoma, unspecified lymph nodes of inguinal region and lower limb
C83.96	Non-follicular (diffuse) lymphoma, unspecified intrapelvic lymph nodes
C83.97	Non-follicular (diffuse) lymphoma, unspecified spleen
C83.98	Non-follicular (diffuse) lymphoma, unspecified lymph nodes of multiple sites
C83.99	Non-follicular (diffuse) lymphoma, unspecified extranodal and solid organ sites

C85.20	Mediastinal (thymic) large B-cell lymphoma, unspecified site
C85.21	Mediastinal (thymic) large B-cell lymphoma, lymph nodes of head, face and neck
C85.22	Mediastinal (thymic) large B-cell lymphoma, intrathoracic lymph nodes
C85.23	Mediastinal (thymic) large B-cell lymphoma, intra-abdominal lymph nodes
C85.24	Mediastinal (thymic) large B-cell lymphoma, lymph nodes of axilla and upper limb
C85.25	Mediastinal (thymic) large B-cell lymphoma, lymph nodes of inguinal region and lower limb
C85.26	Mediastinal (thymic) large B-cell lymphoma, intrapelvic lymph nodes
C85.27	Mediastinal (thymic) large B-cell lymphoma, spleen
C85.28	Mediastinal (thymic) large B-cell lymphoma, lymph nodes of multiple sites
C85.29	Mediastinal (thymic) large B-cell lymphoma, extranodal and solid organ sites
C85.80	Other specified types of non-Hodgkin lymphoma, unspecified site
C85.81	Other specified types of non-Hodgkin lymphoma, lymph nodes of head, face and neck
C85.82	Other specified types of non-Hodgkin lymphoma, intrathoracic lymph nodes
C85.83	Other specified types of non-Hodgkin lymphoma, intra-abdominal lymph nodes
C85.84	Other specified types of non-Hodgkin lymphoma, lymph nodes of axilla and upper limb
C85.85	Other specified types of non-Hodgkin lymphoma, lymph nodes of inguinal region of lower limb
C85.86	Other specified types of non-Hodgkin lymphoma, intrapelvic lymph nodes
C85.87	Other specified types of non-Hodgkin lymphoma, spleen
C85.88	Other specified types of non-Hodgkin lymphoma, lymph nodes of multiple sites
C85.89	Other specified types of non-Hodgkin lymphoma, extranodal and solid organ sites
C88.0	Waldenström macroglobulinemia
C88.4	Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT-lymphoma)
C91.00	Acute lymphoblastic leukemia not having achieved remission
C91.01	Acute lymphoblastic leukemia, in remission
C91.02	Acute lymphoblastic leukemia, in relapse
C91.1	Chronic lymphocytic leukemia of B-cell type
C91.10	Chronic lymphocytic leukemia of B-cell type not having achieved remission
C91.12	Chronic lymphocytic leukemia of B-cell type in relapse
C91.40	Hairy cell leukemia not having achieved remission
C91.42	Hairy cell leukemia, in relapse
D36.0	Benign neoplasm of lymph nodes
D47.Z1	Post-transplant lymphoproliferative disorder (PTLD)
D47.Z2	Other neoplasms of uncertain behavior of lymphoid, hematopoietic and related tissue-Castleman
D59.1	Other autoimmune hemolytic anemias
D69.3	Immune thrombocytopenic purpura
D69.41	Evans Syndrome
D69.42	Congenital and hereditary thrombocytopenia purpura
D69.49	Other primary thrombocytopenia
D89.811	Chronic graft-versus-host disease

G04.81	Other encephalitis and encephalomyelitis
L10.0	Pemphigus vulgaris
M05.10	Rheumatoid lung disease with rheumatoid arthritis of unspecified site
M05.111	Rheumatoid lung disease with rheumatoid arthritis of right shoulder
M05.112	Rheumatoid lung disease with rheumatoid arthritis of left shoulder
M05.119	Rheumatoid lung disease with rheumatoid arthritis of unspecified shoulder
M05.121	Rheumatoid lung disease with rheumatoid arthritis of right elbow
M05.122	Rheumatoid lung disease with rheumatoid arthritis of left elbow
M05.129	Rheumatoid lung disease with rheumatoid arthritis of unspecified elbow
M05.131	Rheumatoid lung disease with rheumatoid arthritis of right wrist
M05.132	Rheumatoid lung disease with rheumatoid arthritis of left wrist
M05.139	Rheumatoid lung disease with rheumatoid arthritis of unspecified wrist
M05.141	Rheumatoid lung disease with rheumatoid arthritis of right hand
M05.142	Rheumatoid lung disease with rheumatoid arthritis of left hand
M05.149	Rheumatoid lung disease with rheumatoid arthritis of unspecified hand
M05.151	Rheumatoid lung disease with rheumatoid arthritis of right hip
M05.152	Rheumatoid lung disease with rheumatoid arthritis of left hip
M05.159	Rheumatoid lung disease with rheumatoid arthritis of unspecified hip
M05.161	Rheumatoid lung disease with rheumatoid arthritis of right knee
M05.162	Rheumatoid lung disease with rheumatoid arthritis of left knee
M05.169	Rheumatoid lung disease with rheumatoid arthritis of unspecified knee
M05.171	Rheumatoid lung disease with rheumatoid arthritis of right ankle and foot
M05.172	Rheumatoid lung disease with rheumatoid arthritis of left ankle and foot
M05.179	Rheumatoid lung disease with rheumatoid arthritis of unspecified ankle and foot
M05.19	Rheumatoid lung disease with rheumatoid arthritis of multiple sites
M05.20	Rheumatoid vasculitis with rheumatoid arthritis of unspecified site
M05.211	Rheumatoid vasculitis with rheumatoid arthritis of right shoulder
M05.212	Rheumatoid vasculitis with rheumatoid arthritis of left shoulder
M05.219	Rheumatoid vasculitis with rheumatoid arthritis of unspecified shoulder
M05.221	Rheumatoid vasculitis with rheumatoid arthritis of right elbow
M05.222	Rheumatoid vasculitis with rheumatoid arthritis of left elbow
M05.229	Rheumatoid vasculitis with rheumatoid arthritis of unspecified elbow
M05.231	Rheumatoid vasculitis with rheumatoid arthritis of right wrist
M05.232	Rheumatoid vasculitis with rheumatoid arthritis of left wrist
M05.239	Rheumatoid vasculitis with rheumatoid arthritis of unspecified wrist
M05.241	Rheumatoid vasculitis with rheumatoid arthritis of right hand
M05.242	Rheumatoid vasculitis with rheumatoid arthritis of left hand
M05.249	Rheumatoid vasculitis with rheumatoid arthritis of unspecified hand
M05.251	Rheumatoid vasculitis with rheumatoid arthritis of right hip

M05.252	Rheumatoid vasculitis with rheumatoid arthritis of left hip
M05.259	Rheumatoid vasculitis with rheumatoid arthritis of unspecified hip
M05.261	Rheumatoid vasculitis with rheumatoid arthritis of right knee
M05.262	Rheumatoid vasculitis with rheumatoid arthritis of left knee
M05.269	Rheumatoid vasculitis with rheumatoid arthritis of unspecified knee
M05.271	Rheumatoid vasculitis with rheumatoid arthritis of right ankle and foot
M05.272	Rheumatoid vasculitis with rheumatoid arthritis of left ankle and foot
M05.279	Rheumatoid vasculitis with rheumatoid arthritis of unspecified ankle and foot
M05.29	Rheumatoid vasculitis with rheumatoid arthritis of multiple sites
M05.30	Rheumatoid heart disease with rheumatoid arthritis of unspecified site
M05.311	Rheumatoid heart disease with rheumatoid arthritis of right shoulder
M05.312	Rheumatoid heart disease with rheumatoid arthritis of left shoulder
M05.319	Rheumatoid heart disease with rheumatoid arthritis of unspecified shoulder
M05.321	Rheumatoid heart disease with rheumatoid arthritis of right elbow
M05.322	Rheumatoid heart disease with rheumatoid arthritis of left elbow
M05.329	Rheumatoid heart disease with rheumatoid arthritis of unspecified elbow
M05.331	Rheumatoid heart disease with rheumatoid arthritis of right wrist
M05.332	Rheumatoid heart disease with rheumatoid arthritis of left wrist
M05.339	Rheumatoid heart disease with rheumatoid arthritis of unspecified wrist
M05.341	Rheumatoid heart disease with rheumatoid arthritis of right hand
M05.342	Rheumatoid heart disease with rheumatoid arthritis of left hand
M05.349	Rheumatoid heart disease with rheumatoid arthritis of unspecified hand
M05.351	Rheumatoid heart disease with rheumatoid arthritis of right hip
M05.352	Rheumatoid heart disease with rheumatoid arthritis of left hip
M05.359	Rheumatoid heart disease with rheumatoid arthritis of unspecified hip
M05.361	Rheumatoid heart disease with rheumatoid arthritis of right knee
M05.362	Rheumatoid heart disease with rheumatoid arthritis of left knee
M05.369	Rheumatoid heart disease with rheumatoid arthritis of unspecified knee
M05.371	Rheumatoid heart disease with rheumatoid arthritis of right ankle and foot
M05.372	Rheumatoid heart disease with rheumatoid arthritis of left ankle and foot
M05.379	Rheumatoid heart disease with rheumatoid arthritis of unspecified ankle and foot
M05.39	Rheumatoid heart disease with rheumatoid arthritis of multiple sites
M05.40	Rheumatoid myopathy with rheumatoid arthritis of unspecified site
M05.411	Rheumatoid myopathy with rheumatoid arthritis of right shoulder
M05.412	Rheumatoid myopathy with rheumatoid arthritis of left shoulder
M05.419	Rheumatoid myopathy with rheumatoid arthritis of unspecified shoulder
M05.421	Rheumatoid myopathy with rheumatoid arthritis of right elbow
M05.422	Rheumatoid myopathy with rheumatoid arthritis of left elbow
M05.429	Rheumatoid myopathy with rheumatoid arthritis of unspecified elbow

M05.431	Rheumatoid myopathy with rheumatoid arthritis of right wrist
M05.432	Rheumatoid myopathy with rheumatoid arthritis of left wrist
M05.439	Rheumatoid myopathy with rheumatoid arthritis of unspecified wrist
M05.441	Rheumatoid myopathy with rheumatoid arthritis of right hand
M05.442	Rheumatoid myopathy with rheumatoid arthritis of left hand
M05.449	Rheumatoid myopathy with rheumatoid arthritis of unspecified hand
M05.451	Rheumatoid myopathy with rheumatoid arthritis of right hip
M05.452	Rheumatoid myopathy with rheumatoid arthritis of left hip
M05.459	Rheumatoid myopathy with rheumatoid arthritis of unspecified hip
M05.461	Rheumatoid myopathy with rheumatoid arthritis of right knee
M05.462	Rheumatoid myopathy with rheumatoid arthritis of left knee
M05.469	Rheumatoid myopathy with rheumatoid arthritis of unspecified knee
M05.471	Rheumatoid myopathy with rheumatoid arthritis of right ankle and foot
M05.472	Rheumatoid myopathy with rheumatoid arthritis of left ankle and foot
M05.479	Rheumatoid myopathy with rheumatoid arthritis of unspecified ankle and foot
M05.49	Rheumatoid myopathy with rheumatoid arthritis of multiple sites
M05.50	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified site
M05.511	Rheumatoid polyneuropathy with rheumatoid arthritis of right shoulder
M05.512	Rheumatoid polyneuropathy with rheumatoid arthritis of left shoulder
M05.519	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified shoulder
M05.521	Rheumatoid polyneuropathy with rheumatoid arthritis of right elbow
M05.522	Rheumatoid polyneuropathy with rheumatoid arthritis of left elbow
M05.529	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified elbow
M05.531	Rheumatoid polyneuropathy with rheumatoid arthritis of right wrist
M05.532	Rheumatoid polyneuropathy with rheumatoid arthritis of left wrist
M05.539	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified wrist
M05.541	Rheumatoid polyneuropathy with rheumatoid arthritis of right hand
M05.542	Rheumatoid polyneuropathy with rheumatoid arthritis of left hand
M05.549	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified hand
M05.551	Rheumatoid polyneuropathy with rheumatoid arthritis of right hip
M05.552	Rheumatoid polyneuropathy with rheumatoid arthritis of left hip
M05.559	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified hip
M05.561	Rheumatoid polyneuropathy with rheumatoid arthritis of right knee
M05.562	Rheumatoid polyneuropathy with rheumatoid arthritis of left knee
M05.569	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified knee
M05.571	Rheumatoid polyneuropathy with rheumatoid arthritis of right ankle and foot
M05.572	Rheumatoid polyneuropathy with rheumatoid arthritis of left ankle and foot
M05.579	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified ankle and foot
M05.59	Rheumatoid polyneuropathy with rheumatoid arthritis of multiple sites

M05.60	Rheumatoid arthritis of unspecified site with involvement of other organs and systems
M05.611	Rheumatoid arthritis of right shoulder with involvement of other organs and systems
M05.612	Rheumatoid arthritis of left shoulder with involvement of other organs and systems
M05.619	Rheumatoid arthritis of unspecified shoulder with involvement of other organs and systems
M05.621	Rheumatoid arthritis of right elbow with involvement of other organs and systems
M05.622	Rheumatoid arthritis of left elbow with involvement of other organs and systems
M05.629	Rheumatoid arthritis of unspecified elbow with involvement of other organs and systems
M05.631	Rheumatoid arthritis of right wrist with involvement of other organs and systems
M05.632	Rheumatoid arthritis of left wrist with involvement of other organs and systems
M05.639	Rheumatoid arthritis of unspecified wrist with involvement of other organs and systems
M05.641	Rheumatoid arthritis of right hand with involvement of other organs and systems
M05.642	Rheumatoid arthritis of left hand with involvement of other organs and systems
M05.649	Rheumatoid arthritis of unspecified hand with involvement of other organs and systems
M05.651	Rheumatoid arthritis of right hip with involvement of other organs and systems
M05.652	Rheumatoid arthritis of left hip with involvement of other organs and systems
M05.659	Rheumatoid arthritis of unspecified hip with involvement of other organs and systems
M05.661	Rheumatoid arthritis of right knee with involvement of other organs and systems
M05.662	Rheumatoid arthritis of left knee with involvement of other organs and systems
M05.669	Rheumatoid arthritis of unspecified knee with involvement of other organs and systems
M05.671	Rheumatoid arthritis of right ankle and foot with involvement of other organs and systems
M05.672	Rheumatoid arthritis of left ankle and foot with involvement of other organs and systems
M05.679	Rheumatoid arthritis of unspecified ankle and foot with involvement of other organs and systems
M05.69	Rheumatoid arthritis of multiple sites with involvement of other organs and systems
M05.70	Rheumatoid arthritis with rheumatoid factor of unspecified site without organ or systems involvement
M05.711	Rheumatoid arthritis with rheumatoid factor of right shoulder without organ or systems involvement
M05.712	Rheumatoid arthritis with rheumatoid factor of left shoulder without organ or systems involvement
M05.719	Rheumatoid arthritis with rheumatoid factor of unspecified shoulder without organ or systems involvement
M05.721	Rheumatoid arthritis with rheumatoid factor of right elbow without organ or systems involvement
M05.722	Rheumatoid arthritis with rheumatoid factor of left elbow without organ or systems involvement
M05.729	Rheumatoid arthritis with rheumatoid factor of unspecified elbow without organ or systems involvement
M05.731	Rheumatoid arthritis with rheumatoid factor of right wrist without organ or systems involvement
M05.732	Rheumatoid arthritis with rheumatoid factor of left wrist without organ or systems involvement
M05.739	Rheumatoid arthritis with rheumatoid factor of unspecified wrist without organ or systems involvement
M05.741	Rheumatoid arthritis with rheumatoid factor of right hand without organ or systems involvement
M05.742	Rheumatoid arthritis with rheumatoid factor of left hand without organ or systems involvement
M05.749	Rheumatoid arthritis with rheumatoid factor of unspecified hand without organ or systems involvement
M05.751	Rheumatoid arthritis with rheumatoid factor of right hip without organ or systems involvement
M05.752	Rheumatoid arthritis with rheumatoid factor of left hip without organ or systems involvement
M05.759	Rheumatoid arthritis with rheumatoid factor of unspecified hip without organ or systems involvement

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

M06.032	Rheumatoid arthritis without rheumatoid factor, left wrist
M06.039	Rheumatoid arthritis without rheumatoid factor, unspecified wrist
M06.041	Rheumatoid arthritis without rheumatoid factor, right hand
M06.042	Rheumatoid arthritis without rheumatoid factor, left hand
M06.049	Rheumatoid arthritis without rheumatoid factor, unspecified hand
M06.051	Rheumatoid arthritis without rheumatoid factor, right hip
M06.052	Rheumatoid arthritis without rheumatoid factor, left hip
M06.059	Rheumatoid arthritis without rheumatoid factor, unspecified hip
M06.061	Rheumatoid arthritis without rheumatoid factor, right knee
M06.062	Rheumatoid arthritis without rheumatoid factor, left knee
M06.069	Rheumatoid arthritis without rheumatoid factor, unspecified knee
M06.071	Rheumatoid arthritis without rheumatoid factor, right ankle and foot
M06.072	Rheumatoid arthritis without rheumatoid factor, left ankle and foot
M06.079	Rheumatoid arthritis without rheumatoid factor, unspecified ankle and foot
M06.08	Rheumatoid arthritis without rheumatoid factor, vertebrae
M06.09	Rheumatoid arthritis without rheumatoid factor, multiple sites
M06.80	Other specified rheumatoid arthritis, unspecified site
M06.811	Other specified rheumatoid arthritis, right shoulder
M06.812	Other specified rheumatoid arthritis, left shoulder
M06.819	Other specified rheumatoid arthritis, unspecified shoulder
M06.821	Other specified rheumatoid arthritis, right elbow
M06.822	Other specified rheumatoid arthritis, left elbow
M06.829	Other specified rheumatoid arthritis, unspecified elbow
M06.831	Other specified rheumatoid arthritis, right wrist
M06.832	Other specified rheumatoid arthritis, left wrist
M06.839	Other specified rheumatoid arthritis, unspecified wrist
M06.841	Other specified rheumatoid arthritis, right hand
M06.842	Other specified rheumatoid arthritis, left hand
M06.849	Other specified rheumatoid arthritis, unspecified hand
M06.851	Other specified rheumatoid arthritis, right hip
M06.852	Other specified rheumatoid arthritis, left hip
M06.859	Other specified rheumatoid arthritis, unspecified hip
M06.861	Other specified rheumatoid arthritis, right knee
M06.862	Other specified rheumatoid arthritis, left knee
M06.869	Other specified rheumatoid arthritis, unspecified knee
M06.871	Other specified rheumatoid arthritis, right ankle and foot
M06.872	Other specified rheumatoid arthritis, left ankle and foot
M06.879	Other specified rheumatoid arthritis, unspecified ankle and foot
M06.88	Other specified rheumatoid arthritis, vertebrae

M06.89	Other specified rheumatoid arthritis, multiple sites
M06.9	Rheumatoid arthritis, unspecified
M31.1	Thrombotic microangiopathy
M31.30	Wegener's granulomatosis without renal involvement
M31.31	Wegener's granulomatosis with renal involvement
M31.7	Microscopic polyangiitis
R59.0	Localized enlarged lymph nodes
R59.1	Generalized enlarged lymph nodes
R59.9	Enlarged lymph nodes, unspecified
Z85.71	Personal history of Hodgkin lymphoma
Z85.72	Personal history of non-Hodgkin lymphomas
Z85.79	Personal history of other malignant neoplasms of lymphoid, hematopoietic and related tissues

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

Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. They can be found at: <http://www.cms.gov/medicare-coverage-database/search/advanced-search.aspx>. Additional indications may be covered at the discretion of the health plan.

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

Jurisdiction(s): 6, K	NCD/LCD Document (s): A52452
https://www.cms.gov/medicare-coverage-database/search/article-date-search.aspx?DocID=A52452&bc=gAAAAAAAAAAAAAA ==	

Jurisdiction(s): J, M	NCD/LCD Document (s): L35026
https://www.cms.gov/medicare-coverage-database/search/lcd-date-search.aspx?DocID=L35026&bc=gAAAAAAAAAAAAAA ==	

Jurisdiction(s): N	NCD/LCD Document (s): L33746
https://www.cms.gov/medicare-coverage-database/search/lcd-date-search.aspx?DocID=L33746&bc=gAAAAAAAAAAAAAA ==	

Jurisdiction(s): 5,8	NCD/LCD Document (s): A55639
https://www.cms.gov/medicare-coverage-database/search/article-date-search.aspx?DocID=A55639&bc=gAAAAAAAAAAAAAA	

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, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC