Policy and Procedure	
PHARMACY PRIOR AUTHORIZATION AND STEP THERAPY POLICY AND CRITERIA ORPTCONC058B.0222	**ANTINEOPLASTIC AGENTS** **RITUXIMAB** See Appendix A for medications covered by policy
Effective Date: 6/1/2022  **Robert Gluckman, M.D.** **Chief Medical Officer**	Review/Revised Date: 08/06, 04/07, 12/08, 02/09, 12/09, 04/10, 06/11, 02/13, 06/13, 02/14, 02/15, 06/15, 07/15, 01/16, 12/16, 01/18, 04/18, 08/18, 01/19, 03/19, 09/19, 12/19, 01/20, 12/20, 04/21, 07/21, 01/22. 04/22 MA) P&T Committee Meeting Date: 8/06, 4/07, 12/08, 2/09, 12/09, 04/10, 06/11, 02/13 , 06/13, 02/14, 02/15, 06/15, 07/15, 02/16, 02/17, 02/18, 06/18, 10/18, 02/19, 04/19, 10/19, 12/19, 02/20, 06/20, 02/21, 04/21, 08/21, 02/22, 04/22 Original Effective Date: 08/06 Approved by: Oregon Region Pharmacy and Therapeutics Committee Page 1 of 2

SCOPE:
Providence Health Plan, Providence Health Assurance, Providence Plan Partners, and Ayin Health Solutions as applicable (referred to individually as “Company” and collectively as “Companies”).

APPLIES TO:
Medicare Part B

POLICY CRITERIA:

COVERED USES:
All Food and Drug Administration (FDA) approved indications not otherwise excluded from the benefit, some medically- accepted indications (as outlined in the Required Medical Information section).

REQUIRED MEDICAL INFORMATION:

I. For initiation of therapy (new starts), both of the following criteria must be met:
   a. For **non-preferred rituximab** products: Documented trial and failure, intolerance, or contraindication to the use of both of the preferred biosimilar medications: Ruxience® (rituximab-pvvr) and Truxima® (rituximab-abbs). See Appendix A for preferred and non-preferred rituximab products
   b. Requests for rituximab may be approved for the following indications when the criteria below are met:
      i. For **Oncologic Diagnoses:** Use must be for a FDA approved indication or indication supported by National Comprehensive Cancer Network (NCCN) guidelines with recommendation 2A or higher
      ii. For **Rheumatoid Arthritis:**

1. Documentation of trial, failure, intolerance, or contraindication to at least one of the following targeted immune modulators: etanercept (Enbrel®), adalimumab (Humira®), or a preferred infliximab product **AND**
2. Documentation that rituximab will be used concurrently with methotrexate. If intolerance or contraindication to methotrexate, then in combination with another disease-modifying antirheumatic drug (DMARD) (for example, leflunomide, sulfasalazine, hydroxychloroquine), unless medical rationale is provided to support monotherapy.

iii. For **Vasculitis,** including antineutrophil cytoplasmic autoantibody (ANCA)-associated vasculitis [Granulomatosis with Polyangiitis (GPA) and Microscopic Polyangiitis (MPA)] and refractory polyarteritis nodosa (resistant to cyclophosphamide):
1. Documentation that rituximab will be given in combination with glucocorticoids, **AND**
2. Documentation of severe disease (for example, critical organ system involvement)

iv. For **Immune Thrombocytopenia (ITP):**
1. Documentation of trial, failure, intolerance, or contraindication to systemic corticosteroid therapy, **AND**
2. Documentation of active bleeding, or high-risk of bleeding, or a platelet count less than 30,000 cells per microliter

v. For **Relapsing and Remitting Multiple Sclerosis (RRMS):** One of the following:
1. Documentation of trial, failure, or intolerance, to at least two disease modifying therapies indicated for RRMS, OR
2. Documentation that patient has highly active or aggressive disease

vi. For **Refractory Myasthenia Gravis:**
1. Documentation that patient has severely impaired function due to myasthenia gravis, **AND**
2. Documented trial, failure, intolerance, or contraindication to at least two of the following conventional therapies:
    a. Acetylcholinesterase inhibitors (for example, pyridostigmine)
    b. Corticosteroids (for example, prednisone, methylprednisolone)
    c. Immunosuppressive agents (for example, azathioprine, cyclosporine, mycophenolate)
    d. Plasma exchange

vii. For **Autoimmune Hemolytic Anemia (AIHA):**
   1. Diagnosis of warm AIHA and documentation of trial, failure, intolerance, or contraindication to glucocorticoids, **OR**
   2. Diagnosis of cold AIHA or cold agglutinin disease

viii. Confirmed diagnosis of **Neuromyelitis Optica (NMO)**

ix. Confirmed diagnosis of **Moderate to Severe Pemphigus Vulgaris (PV)**

II. For **patients established on therapy** with the requested product (within the previous year): Documentation of adequate response to the medication must be provided.

EXCLUSION CRITERIA: N/A

AGE RESTRICTIONS: N/A

PRESCRIBER RESTRICTIONS:
Must be prescribed by, or in consultation with, a specialist for the respective indication such as: an oncologist, hematologist, rheumatologist, neurologist (in the case of RRMS, NMO), dermatologist (in the case of PV), or nephrologist (in the case of renal disease).

COVERAGE DURATION:
Initial authorization will be approved for six months and reauthorization will be approved until no longer eligible with the plan, subject to formulary and.or benefit changes

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For off-label use criteria please see the Chemotherapy Treatment Utilization Criteria; Coverage for Non-FDA Approved Indications ORPTCOPS105.

Requests for indications that were approved by the FDA within the previous six (6) months may not have been reviewed by the health plan for safety and effectiveness and inclusion on this policy document. These requests will be reviewed using the New Drug and or Indication Awaiting P&T Review; Prior Authorization Request ORPTCOPS047.

Requests for a non-FDA approved (off-label) indication requires the proposed indication be listed in either the American Hospital Formulary System (AHFS), Drugdex, or the National Comprehensive Cancer Network (NCCN) and is considered subject to evaluation of the prescriber's medical rationale, formulary alternatives, the available published evidence-based research and whether the proposed use is determined to be experimental/investigational.

Coverage decisions are made on the basis of individualized determinations of medical necessity and the experimental or investigational character of the treatment in the individual case.

INTRODUCTION:
Rituximab binds to the CD20 antigen on B-lymphocytes and the Fc portion recruits immune functions to mediate B-cell lysis. Recombinant human hyaluronidase is an endoglycosidase used to increase the dispersion and absorption of co-administered drugs when administered subcutaneously.

Rituximab has a boxed warning for severe mucocutaneous reactions, hepatitis B (HBV) reactivation, and progressive multifocal leukoencephalopathy (PML). Intravenous rituximab also has a boxed warning for fatal-infusion related reactions within 24 hours of administration. The majority of these reactions occur with the first infusion.

FDA APPROVED INDICATIONS:
Rituximab and biosimilars, injection for intravenous use

- Non-Hodgkin's Lymphoma (NHL)
    - Relapsed or refractory, low-grade or follicular, CD20-positive, B-cell NHL as a single agent
    - Previously untreated follicular, CD20-positive, B-cell NHL in combination with first line chemotherapy and, in patients achieving a complete or partial response to Rituxan in combination with chemotherapy, as single-agent maintenance therapy.
    - Non-progressing (including stable disease), low-grade, CD20-positive, B-cell NHL as a single agent after first-line CVP chemotherapy
    - Previously untreated diffuse large B-cell, CD20-positive NHL in combination with CHOP or other anthracycline-based chemotherapy regimens
- Chronic Lymphocytic Leukemia (CLL): in combination with fludarabine and cyclophosphamide (FC) for the treatment of patients with previously untreated and previously treated CD20-positive CLL
- Granulomatosis with Polyangiitis (GPA) (Wegener's Granulomatosis) and Microscopic Polyangiitis (MPA) in adult and pediatric patients two years of age and older in combination with glucocorticoids

Rituxan®, Ruxience®, Truxima®

- Rheumatoid Arthritis (RA): (Moderate to Severe), in combination with methotrexate, in patients who had an inadequate response to one or more tumor-necrosis-factor (TNF) antagonist therapies

Rituxan® only

- Moderate to severe Pemphigus Vulgaris (PV) in adult patients
- Mature B-cell NHL and mature B-cell acute leukemia (B-AL): previously untreated, advanced stage, CD20-positive, diffuse large B-cell lymphoma

(DLBCL), Burkitt-like lymphoma (BLL) or mature B-cell acute leukemia (B-AL) in combination with chemotherapy in pediatric patients age 6 months and older

Limitations of use: Rituxan® is not recommended for use in patients with severe, active infection.

Rituxan Hycela® (rituximab and hyaluronidase) injection

- Follicular Lymphoma
  - Relapsed or refractory, follicular lymphoma as a single agent
  - Previously untreated follicular lymphoma in combination with first line chemotherapy and, in patients achieving a complete or partial response to rituximab in combination with chemotherapy, as single-agent maintenance therapy
  - Non-progressing (including stable disease), follicular lymphoma as a single agent after first-line cyclophosphamide, vincristine, and prednisone (CVP) chemotherapy
- Diffuse Large B-cell Lymphoma
  - Previously untreated diffuse large B-cell lymphoma in combination with cyclophosphamide, doxorubicin, vincristine, prednisone (CHOP) or other anthracycline-based chemotherapy regimens
- Chronic Lymphocytic Leukemia
  - Previously untreated and previously treated CLL in combination with fludarabine and cyclophosphamide (FC)

Limitations of Use:

- Initiate treatment only after patients have received at least one full dose of a rituximab product by intravenous infusion.
- Not indicated for the treatment of non-malignant conditions.

POSITION STATEMENT:

Information to date suggests that patients with **rheumatoid arthritis** (RA) who receive rituximab have an increased risk of progressive multifocal leukoencephalopathy (PML). Physicians should consider the risk of PML in any patient treated with rituximab who presents with new onset neurologic manifestations. Consultation with a neurologist, brain magnetic resonance imaging (MRI) scan, and lumbar puncture should be considered as clinically indicated. The American College of Rheumatology guidelines, updated in 2015, recommend rituximab for use in certain populations including patients who were previously treated for lymphoproliferative disorders such as B-cell chronic lymphocytic leukemia, non-Hodgkin lymphoma, hairy cell leukemia.

Vasculitis is a term for a general condition that causes inflammation of blood vessels that can lead to occlusion or rupture of the vessels. This can have devastating effects to organ systems, including ischemia or hemorrhage. The cause is unknown in most cases, but certain infections [human immunodeficiency virus (HIV), Hepatitis B] and autoimmune conditions (e.g., rheumatoid arthritis) can be considered risk factors. Diagnosis is typically done through biopsy, angiography and other blood tests.

Granulomatosis with polyangiitis (GPA), also known as Wegener's granulomatosis, is an uncommon subset of vasculitis that is characterized by inflammation of blood vessels that primarily affect the upper airways, lungs, and kidneys. Typically, symptoms start in the sinuses and can progress rapidly to organ systems like the kidneys, ultimately causing organ failure or dysfunction (e.g., glomerulonephritis). Microscopic polyangiitis (MPA) is another uncommon form of vasculitis that primarily affects small to medium sized blood vessels in the kidneys, lung, nerves, skin, and joints. Symptoms are related to the affected organ system (e.g., muscle/joint pain, or dermatologic rash). Both GPA and MPA are commonly associated with antineutrophil cytoplasmic autoantibody (ANCA), approximately 80 to 90% of patients are found to have ANCA. Although GPA and MPA are distinct entities within ANCA-associated vasculitis, they have been classified together due to their overlapping manifestations and it can be extremely difficult to differentiate between the two diseases. Experts commonly recognize ANCA antigen types for myeloperoxidase (anti-MPO) or proteinase 3 (anti-PR3), rather than by disease type (GPA or MPA). [12, 13, 14, 28, 29]

The 2015 European League Against Rheumatism (EULAR) update for management of ANCA-associated vasculitis (AAV) was developed in collaboration with the European Renal Association-European Dialysis and Transplant Association (ERA-EDTA) and the European Vasculitis Association (EUVAS)[15]:

- For remission-induction and major relapse of new-onset organ-threatening or life-threatening AAV, it is recommended to treat with high-dose glucocorticoid therapy in combination with **rituximab** or cyclophosphamide for GPA and MPA (Grade A recommendation)
    - Two randomized controlled trials investigated the use of rituximab in AAV, the RAVE and RITUXVAS trials in patients with GPA and MPA.
    - In both studies, patients received high-dose glucocorticoids with rituximab 375 mg/m^2 weekly
    - Rituximab was non-inferior to cyclophosphamide in both trials

    - Rituximab appeared more effective for relapsing disease in RAVE trial, therefore it is preferred over cyclophosphamide for relapsing disease per EULAR/ERA-DTA recommendations
- Glucocorticoid plus rituximab or cyclophosphamide combination therapy is also recommended with a lower grade of evidence for eosinophilic granulomatosis with polyangiitis (EGPA) which is also known as Churg-Strauss syndrome.
- For remission-induction in non-organ-threatening disease in AAV, EULAR recommends combination of glucocorticoid and either methotrexate or mycophenolate mofetil. .
- Maintenance of remission is achieved by use of low-dose glucocorticoids plus either azathioprine, rituximab, methotrexate, or mycophenolate mofetil (listed in order of the strength of voting by the expert panel who developed the EULAR/ERA-DTA recommendations).
    - Leflunomide is no longer considered first-line therapy for remission maintenance due to more adverse effects compared to the immunosuppressants listed above
- For patients with recurrent infections, rituximab is associated with hypoimmunoglobulinemia, therefore it is recommended to test serum immunoglobulin levels prior to course of rituximab

The use of rituximab for polyarteritis nodosa (PAN) is supported by its efficacy in ANCA-associated vasculitis. PAN is a systemic vasculitis that is treated with glucocorticoids and cyclophosphamide in severe cases. Case reports has shown successful treatment by rituximab for life-threatening polyarteritis nodosa that did not respond to glucocorticoids and cyclophosphamide.

Immune thrombocytopenia (ITP) is also known as immune thrombocytopenic purpura or idiopathic thrombocytopenic purpura. This is an autoimmune disease characterized by immunologic destruction of otherwise normal platelets and is typically caused by an unknown trigger. First line treatment is typically with corticosteroids when the platelet count is < $30x10^9$/L. Immune globulin (IgG) can be considered for add-on therapy (one-time dose) when a rapid increase in platelet count in needed. Second-line treatments include splenectomy, TPO-receptor agonists (e.g., eltrombopag, romiplostim), and rituximab. Splenectomy is the only treatment that provides sustained remission off all treatments at one year and beyond in a high proportion of ITP patients.

Per the American Academy of Neurology, determining initial treatment **for relapsing remitting multiply sclerosis (RRMS)** should encompass consideration of safety, route of administration, lifestyle, cost, efficacy, common adverse effects (AEs), and tolerability. Rituximab has been used off-label to treat RRMS for many years. A 2021

systematic Cochrane review assessed the beneficial and adverse effects of rituximab as ‘first choice’ and as ‘switching’ therapy for adults with MS. In reviewing rituximab as a first choice agent for RRMS, one non-randomized study compared rituximab with interferon beta or glatiramer acetate, dimethyl fumarate, natalizumab, or fingolimod in active relapsing MS at 24 months' follow-up. Rituximab likely results in a large reduction in relapses compared with interferon beta or glatiramer acetate (hazard ratio (HR) 0.14, 95% confidence interval (CI) 0.05 to 0.39; 335 participants; moderate-certainty evidence). Rituximab may reduce relapses compared with dimethyl fumarate (HR 0.29, 95% CI 0.08 to 1.00; 206 participants; low-certainty evidence) and natalizumab (HR 0.24, 95% CI 0.06 to 1.00; 170 participants; low-certainty evidence). It may make little or no difference on relapse compared with fingolimod (HR 0.26, 95% CI 0.04 to 1.69; 137 participants; very low-certainty evidence). In those patients that were switching therapy, one RCT compared rituximab with placebo in relapsing MS at 12 months’ follow-up. Rituximab may decrease recurrence of relapses compared with placebo (OR 0.38, 95% CI 0.16 to 0.93; 104 participants; low-certainty evidence). The authors concluded, for preventing relapses in relapsing MS, rituximab as 'first choice' and as 'switching' may compare favorably with a wide range of approved DMTs. A comprehensive review on the treatment of multiple sclerosis by Gholamzad et al. 2019 suggested that rituximab for RRMS patients who did not respond to first- and second-line therapies and in cases where RRMS is stabilized after natalizumab treatment and is a candidate for a RRMS therapy with less PML risk.

Pemphigus vulgaris (PV) is an acquired autoimmune disease in which immunoglobulin (IgG) antibodies target desmosomal proteins to produce intraepithelial, mucocutaneous blistering. Rituxima in combination with short-term prednisone was compared to prednisone monotherapy (1:1) as first-line treatment in 90 newly diagnosed adult patients with moderate to severe pemphigus (74 PV). This was a randomized, open-label, controlled study. 66 of the patients with PV had severe disease according to disease severity defined by Harman’s criteria. Study treatment included an initial IV infusion of 1 gram of rituximab product in combination with a short-term regimen of 0.5mg/kg/day of oral prednisone tapered over 3 months for moderate disease and 1mg/kg/day for severe disease tapered over 6 months. All patients received a second IV infusion of 1g on day 15. Maintenance infusions of 500 mg were administered at months 12 and 18. In the prednisone arm, patients received 1 mg/kg/day of oral prednisone tapered off over 12 months for moderate disease and 1.5 mg/kg/day oral prednisone tapered over 18 months for severe disease. The primary endpoint was complete remission at month 24 without the use of prednisone therapy for two months or more. Rituximab plus prednisone had an 89% response rate and 90% response rate among the 38 PV patients compared to only a 34% response rate among prednisone monotherapy (28% of 36 PV patients).

Autoimmune hemolytic anemia (AIHA) is a group of disorders characterized by a malfunction of the immune system that produces autoantibodies, which attack red blood cells as if they were substances foreign to the body. There are no randomized, controlled prospective trials to compare relative effectiveness of the different treatment options.

- There are two main types of AIHA: Warm AIHA where the autoantibodies attach to and destroy red blood cells (RBC) at normal body temperature and cold AIHA (cold agglutinin disease) where the autoantibodies (IgM) become most active and attack RBC only at temperatures well below normal body temperature.
- Treatment strategy in warm AIHA includes reduction in autoantibody production (e.g., glucocorticoids, rituximab) and reduction in autoantibody effectiveness (e.g., splenectomy)
    - First-line agent is glucocorticoids at an initial dose of 1 to 1.5 mg/kg per day of prednisone or its equivalent in adults.
    - Second-line agents include splenectomy or rituximab, although splenectomy is more likely to achieve long-term cure.
    - Third-line agents include immunosuppressive or cytotoxic agents
- Treatment strategy in cold AIHA (cold agglutinin disease [CAD]) include minimizing cold-induced symptoms, maintaining an acceptable hemoglobin level, and addressing underlying disorders. Glucocorticoids and splenectomy are not effective therapy in CAD. Rituximab-containing regimens are usually recommended as first-line.

Neuromyelitis optica (NMO), previously known as Devic disease, is an autoimmune inflammatory disorder that typically affects the optic nerves and spinal cord.[32] Prophylactic treatment of NMO recurrence must be immediately performed when NMO is identified because the progression of NMO disability is related to the severity of attacks.

The pathogenesis of NMO is related to the presence of aquaporin-4 autoantibody, thus, rituximab has been often utilized as treatment given its activity against CD20. The depletion of CD20 provides a theoretical basis for treatment of autoimmune diseases, in which B cells and autoantibodies play a key role; for example, AQP4-Ab is associated with NMO. A meta-analysis of 26 studies with 577 participants was conducted to evaluate rituximab efficacy in terms of safety and tolerance and assessed the treatment efficacies based on relapse rates and disability. Antibodies against aquaporin-4 autoantibody were recorded in 435 of 577 (75.39%) patients with NMO. Rituximab therapy resulted in a mean − 1.56 (95% CI, − 1.82 to − 1.29)

reduction in the mean ARR ratio and a mean − 1.16 (95% CI, − 1.36 to − 0.96) reduction in the mean EDSS score. A total of 330 of 528 patients (62.9%) reached the relapse-free state. A total of 95 of 577 (16.46%) patients had adverse reactions.[33]

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APPENDIX A.

Brand Name	Generic Name	HCPCS Code
Preferred Products		
Ruxience®	rituximab-pvvr	Q5119
Truxima®	rituximab-abbs	Q5115
Non-preferred Products		
Riabni®	rituximab-arrx	J3590
Rituxan®	rituximab infusion	J9312
Rituxan Hycela®	rituximab & hyaluronidase infusion	J9311