



## Pharmacy Medical Policy

### Nononcologic Uses of Rituximab

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 NCD/LCD: N/A

#### Related Policies

- Quality Care Cancer Program Oncology Policy [099](#)

#### Policy

#### Commercial Members: Managed Care (HMO and POS), PPO, and Indemnity

**Note:** All requests for outpatient retail pharmacy for indications listed and not listed on the medical policy guidelines may be submitted to BCBSMA Clinical Pharmacy Operations by completing the Prior Authorization Form on the last page of this document. Physicians may also call BCBSMA Pharmacy Operations department at (800)366-7778 to request a prior authorization/formulary exception verbally. Patients must have pharmacy benefits under their subscriber certificates.

Please refer to the chart below for the formulary and Prior Authorization of the medications affected by this policy.

Standard Formulary	
Drug	Coverage Status
Rituxan <sup>®</sup> (rituximab) *^	Non-preferred with Prior Authorization
Riabni <sup>™</sup> (rituximab -arrx) *^	Non-preferred with Prior Authorization
Ruxience <sup>™</sup> (rituximab - pvvr)	Preferred with Prior Authorization
Truxima <sup>®</sup> (rituximab- abbs)	Preferred with Prior Authorization

**NOTE:** This policy is only for nononcologic uses of Rituximab. For any oncology diagnosis please refer to [policy 099](#) and submit request to AIM.

Rituximab<sup>\*^</sup> may be considered **MEDICALLY NECESSARY** for the following indications:

- Rheumatoid Arthritis<sup>%%</sup>.
- Granulomatosis with Polyangiitis (GPA) (Wegener's Granulomatosis) and Microscopic Polyangiitis (MPA).
- Pemphigus Vulgaris (PV) <sup>%\*</sup>.

<sup>\*^</sup> - Rituxan or Riabni approvals require the use of two preferred biosimilars except where noted.

<sup>%%</sup> - Only requires one biosimilar which is FDA approved before allowing the originator product Rituxan.

<sup>%\*</sup> - Does not require any biosimilar as they are not FDA approved before allowing the originator product Rituxan.

Rituximab may be considered **MEDICALLY NECESSARY** for the following off-label indications:

- the following autoimmune hemolytic anemias (AIHA):
  - warm AIHA in glucocorticoid-refractory or glucocorticoid-dependent patients;
  - cold agglutination syndrome;
- thrombotic thrombocytopenic purpura in patients with refractory disease or relapse (ie, lack of response to plasma exchange therapy and glucocorticoids);
- Churg-Strauss syndrome (eosinophilic granulomatosis with polyangiitis):
  - first-line treatment in combination with glucocorticoids for patients with severe (organ-threatening) disease;
  - add-on therapy for treatment-refractory disease;
- factor inhibitors in patients with hemophilia who are refractory to conventional first-line treatments (eg, immune tolerance induction, glucocorticoids with or without cyclophosphamide), preferably as add-on therapy
- add-on therapy for patients with hepatitis C virus-associated cryoglobulinemic vasculitis who have:
  - active disease resistant to antiviral drugs; or
  - severe or life-threatening cryoglobulinemic vasculitis;
- multicentric Castleman disease (first- or second-line therapy);
- primary Sjögren syndrome that is refractory to glucocorticoids and other immunosuppressive agents;
- add-on therapy for systemic lupus erythematosus refractory to standard first-line treatment;
- add-on therapy for lupus nephritis refractory to standard first-line treatment regimens;
- systemic sclerosis (scleroderma) in patients refractory to first-line treatment;
- neuromyelitis optica for relapse prevention;
- idiopathic membranous nephropathy;
- glucocorticoid-refractory chronic graft-versus-host disease; and
- desensitization of human leukocyte antigen-sensitized renal transplant candidates before transplantation.

Rituximab is **INVESTIGATIONAL** for all other nononcologic uses, including but not limited to:

- idiopathic thrombocytopenic purpura in patients who do not respond to first-line treatments;
- paroxysmal cold hemoglobinuria;
- mixed connective tissue disease;
- multiple sclerosis;
- treatment of myasthenia gravis;
- treatment of minimal change disease;

- prophylaxis for graft-versus-host disease;
- induction immunosuppressive therapy for kidney transplantation;
- induction immunosuppressive therapy for heart transplantation
- treatment of antibody-mediated rejection in solid organ transplant recipients; and
- treatment of antibody-mediated rejection after pancreatic islet transplantation.

## Prior Authorization Information

### Outpatient

- For services described in this policy, see below for products where prior authorization **IS REQUIRED** if the procedure is performed **outpatient**.

	Outpatient
Commercial Managed Care (HMO and POS)	Prior authorization is <b>required</b> .
Commercial PPO	Prior authorization is <b>required</b> .

## CPT Codes / HCPCS Codes / ICD Codes

*Inclusion or exclusion of a code does not constitute or imply member coverage or provider reimbursement. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage as it applies to an individual member.*

*Providers should report all services using the most up-to-date industry-standard procedure, revenue, and diagnosis codes, including modifiers where applicable.*

*The following codes are included below for informational purposes only; this is not an all-inclusive list.*

**The above medical necessity criteria MUST be met for the following codes to be covered for Commercial Members: Managed Care (HMO and POS), PPO, Indemnity, Medicare HMO Blue and Medicare PPO Blue:**

### No specific CPT Codes

## Description

### Rituximab

Rituximab (Rituxan) is a chimeric murine-human monoclonal antibody directed against the CD20 surface antigen, which is expressed on pre-B and mature B lymphocytes. Rituximab induces lysis of normal and malignant CD20-expressing B cells; possible mechanisms of cell lysis include complement-dependent cytotoxicity and antibody-dependent cell-mediated cytotoxicity.<sup>1</sup>

B cells are thought to play a role in the pathogenesis of rheumatoid arthritis and other autoimmune diseases by producing auto-antibodies and proinflammatory cytokines, and by activating T lymphocytes.<sup>1</sup> Rituximab reduces the number of B cells in the peripheral blood and in lymphoid tissues, thereby interrupting pathogenic processes of autoimmune diseases.

Rituximab is infused intravenously.

### Adverse Events

Rituximab carries the following black box warnings<sup>2</sup>:

- Fatal infusion reactions within 24 hours of rituximab infusion; approximately 80% of fatal reactions occurred with the first infusion.
- Severe mucocutaneous reactions, some with fatal outcomes.

- Hepatitis B virus reactivation, in some cases resulting in fulminant hepatitis, hepatic failure, and death.
- Progressive multifocal leukoencephalopathy resulting in death.

Labeled warnings and precautions include:

- Tumor lysis syndrome (for patients with hematologic malignancies)
- Infections
- Cardiac arrhythmias and angina
- Bowel obstruction and perforation
- Not administering live virus vaccines before or during rituximab therapy
- Cytopenias.

## Summary

### **Hematologic Disorders and Vasculitides**

For individuals who have AIHA-warm AIHA and cold agglutinin syndrome-refractory to first-line therapy who receive rituximab, the evidence includes RCTs and observational studies. Relevant outcomes are symptoms, change in disease status, functional outcomes, quality of life, and treatment-related morbidity. Two RCTs have found that overall response rates were better with rituximab than a control condition at 1 year in patients with newly diagnosed warm AIHA. Serious adverse events were higher with rituximab than corticosteroids (1 RCT) but lower than placebo (the other RCT). Response rates from observations studies have supported these findings and found lesser yet substantive response rates in patients with cold agglutinin syndrome. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals who have relapsed or refractory ITP who receive rituximab, the evidence includes an RCT of second-line therapy and observational studies. Relevant outcomes are symptoms, change in disease status, functional outcomes, quality of life, and treatment-related morbidity. Rituximab as second-line treatment for adult thrombocytopenia trial failed to demonstrate improved outcomes with rituximab as second-line therapy in adults with ITP. The evidence is insufficient to determine the effects of the technology on health outcomes.

For individuals who have relapsed or refractory TTP who receive rituximab, the evidence includes a nonrandomized trial (phase 2), a cohort study, and case series. Relevant outcomes are symptoms, change in disease status, functional outcomes, quality of life, and treatment-related morbidity. These studies have provided consistent evidence of improved health outcomes. For example, a phase 2 trial reported substantially lower relapse rates than historical controls. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals who have Churg-Strauss syndrome (eosinophilic granulomatosis with polyangiitis) who receive rituximab, the evidence includes a single-center retrospective observational study and 3 case series. Relevant outcomes are symptoms, change in disease status, functional outcomes, quality of life, and treatment-related morbidity. Response and remission rates have generally been high, but treatment-related adverse events-some severe-have been reported. The evidence is insufficient to determine the effects of the technology on health outcomes.

For individuals who have congenital or acquired hemophilia A with inhibitory antibodies, refractory to first-line therapy, who receive rituximab, the evidence includes a phase 2 trial, a cohort study, and case series. Relevant outcomes are symptoms, change in disease status, functional outcomes, quality of life, and treatment-related morbidity. Response rates have varied among reports (25% to 50%), depending on whether rituximab was administered as mono- or combination therapy; remission rates have generally been high. Treatment-related adverse events-some severe-have been reported. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals who have HCV-associated cryoglobulinemic vasculitis who receive rituximab, the evidence includes 2 RCTs, a phase 2 nonrandomized trial, and observational studies. Relevant outcomes are symptoms, change in disease status, functional outcomes, quality of life, and treatment-related morbidity. The reported response rates in these studies are consistent with improved health outcomes. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

### **Autoimmune-Related Connective Tissue Disorders**

For individuals who have MCTD who receive rituximab, the evidence includes 2 case series. Relevant outcomes are symptoms, change in disease status, functional outcomes, quality of life, and treatment-related morbidity. In one of the series, 3 of 5 patients with MCTD achieved partial remission with rituximab and, in the other, which focused on MCTD related to interstitial lung disease, there was no significant change in forced vital capacity at 1 or 2 years after initiating rituximab. The evidence is insufficient to determine the effects of the technology on health outcomes.

For individuals who have multicentric Castleman disease (angiofollicular lymph node hyperplasia) who receive rituximab, the evidence includes 2 prospective and 3 retrospective cohort studies. Relevant outcomes are symptoms, change in disease status, functional outcomes, quality of life, and treatment-related morbidity. Although the evidence base consists of nonrandomized studies, rituximab has significantly improved overall survival and markedly reduced the incidence of non-Hodgkin lymphoma. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals who have primary Sjögren syndrome, refractory to first-line therapy, who receive rituximab, the evidence includes a large RCT (disease onset <10 years prior) and smaller observational studies. Relevant outcomes are symptoms, change in disease status, functional outcomes, quality of life, and treatment-related morbidity. The efficacy of rituximab has not been consistently demonstrated in this population. For example, a large (N=120) randomized trial showed no difference in response rates compared with placebo, and a small (N=41) nonrandomized trial showed statistically significant differences in response rates compared with disease-modifying antirheumatic drugs in previously treated patients. The incidence of adverse events did not appear to increase above that observed in other patient populations. The evidence is insufficient to determine the effects of the technology on health outcomes.

For individuals who have SLE, refractory to first-line therapy, who receive rituximab, the evidence includes a large RCT and systematic reviews that also included observational studies. Relevant outcomes are symptoms, change in disease status, functional outcomes, quality of life, and treatment-related morbidity. The single RCT failed to show improved response rates at 1 year with rituximab add-on therapy. Cohort studies and case series of refractory patients have generally reported higher response rates than controlled studies. The evidence is insufficient to determine the effects of the technology on health outcomes.

For individuals who have lupus nephritis, refractory to first-line therapy, who receive rituximab, the evidence includes an RCT and noncomparative studies. Relevant outcomes are symptoms, change in disease status, functional outcomes, quality of life, and treatment-related morbidity. The single RCT did not show improved response rates at 1 year with rituximab add-on therapy. Noncomparative studies have reported complete and partial response rates of 30% to 40% and approximately 35%, respectively, in patients with mostly refractory disease. Adverse events occurred in approximately 20% of patients. The evidence is insufficient to determine the effects of the technology on health outcomes.

For individuals who have systemic sclerosis, refractory to first-line therapy, who receive rituximab, the evidence includes observational studies and a small, unblinded trial. Relevant outcomes are symptoms, change in disease status, functional outcomes, quality of life, and treatment-related morbidity. Add-on rituximab therapy has generally improved skin symptoms and pulmonary function tests; adverse events, including sepsis deaths, occurred in 21% to 47% of patients. Long-term follow-up for efficacy and safety is limited. The evidence is insufficient to determine the effects of the technology on health outcomes.

### **Other Autoimmune-Related Conditions and Disorders**

For individuals who have MS who receive rituximab, the evidence includes 2 RCTs, a registry study, and case series. Relevant outcomes are symptoms, functional outcomes, quality of life, and treatment-related morbidity. One RCT in patients with relapsing-remitting MS showed reductions in the number of lesions detected by gadolinium-enhanced magnetic resonance imaging and at 24 and 48 weeks, and in clinical outcomes at 24-week follow-up. However, methodologic limitations restrict the conclusions drawn from these data. One well-designed RCT in patients with primary-progressive MS demonstrated no effect of rituximab on disease progression. A large registry study found that rituximab was associated with a relatively low rate of adverse events and relapses and little change in disability scores; this study lacked a comparison group. The evidence is insufficient to determine the effects of the technology on health outcomes.

For individuals who have NMO (prevention relapse), refractory to first-line therapy, who receive rituximab, the evidence includes uncontrolled observational studies and systematic reviews. Relevant outcomes are symptoms, change in disease status, functional outcomes, quality of life, and treatment-related morbidity. A 2016 systematic review of 46 uncontrolled studies found significant reductions in the relapse rate and Expanded Disability Status Scale scores after beginning treatment with rituximab. Based on adverse events reported, the safety of rituximab in NMO appeared comparable to the safety in other patient populations. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals who have refractory and nonrefractory myasthenia gravis who receive rituximab, the evidence includes observational studies and a systematic review. Relevant outcomes are change in disease status, quality of life, and treatment-related morbidity. A systematic review found a significant reduction in a myasthenia gravis symptom score after beginning rituximab treatment and a relatively low rate of adverse events. A limitation of the studies was that adverse event reports were not available for all patients. An uncontrolled observational study found significantly better clinical outcomes in patients with anti-MuSK myasthenia who were treated with rituximab compared with those who did not receive rituximab. However, few controlled studies and no RCTs are available. The evidence is insufficient to determine the effects of the technology on health outcomes.

For individuals who have idiopathic membranous nephropathy who receive rituximab, the evidence includes an RCT and observational studies. Relevant outcomes are change in disease status, quality of life, and treatment-related morbidity. Rituximab may have moderate benefit in patients with idiopathic membranous nephropathy who have failed previous treatment with other immunosuppressive regimens or those with a moderate risk of progression who have not previously received immunosuppressive therapy. However, an RCT with longer follow-up is needed to confirm the benefits of rituximab and to determine the optimal schedule, dose, and long-term safety and efficacy. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals who have minimal change in disease (adults and children) who receive rituximab, the evidence includes observational studies in adults and 2 RCTs and observational studies in children. Relevant outcomes are change in disease status, quality of life, and treatment-related morbidity. Rituximab benefit children with nephrotic syndrome associated with minimal change disease. However, because of the risk of severe and potentially life-threatening complications, rituximab use should be restricted to children with frequent relapses and serious adverse events from their medications (because the long-term efficacy and safety of rituximab in this group of patients remain unclear). The evidence is insufficient to determine the effects of the technology on health outcomes.

### **Transplant-Related Conditions and Disorders**

For individuals who have corticosteroid-refractory chronic GVHD who receive rituximab, the evidence includes multiple cohort studies. Relevant outcomes are change in disease status, quality of life, and treatment-related morbidity. Treatment with rituximab has demonstrated response rates in most patients, with sustained response and steroid reduction or discontinuation in some. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals who have sensitization to HLA and are renal transplant candidates who receive rituximab, the evidence includes an RCT and cohort studies. Relevant outcomes are change in disease status, quality of life, and treatment-related morbidity. An RCT comparing desensitization regimens with and without rituximab was terminated due to excess serious adverse events in the control arm. There may be a higher risk of polyomavirus BK virus infection. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals who are kidney transplant candidates who are receiving induction immunosuppressive therapy, the evidence includes cohort studies with historical controls and case series RCTs and systematic reviews. Although observed improvements in outcomes have suggested potential benefit with rituximab, data are retrospective or from small prospective studies. Dose-response studies and larger RCTs with longer follow-up are needed to demonstrate improved health outcomes. For individuals who are heart transplant candidates who are receiving induction immunosuppressive therapy, the recommendation for the use of rituximab as part of a combination regimen is based on consensus reporting of case reports and expert opinion.

For individuals who have ABMR of a solid organ transplant who receive rituximab, the evidence includes cohort studies with historical controls and case series. Relevant outcomes are change in disease status, quality of life, and treatment-related morbidity. Although observed improvements in outcomes have suggested potential benefit with rituximab, data are retrospective or from small prospective studies. Dose-response studies and larger RCTs with longer follow-up are needed to demonstrate improved health outcomes. The evidence is insufficient to determine the effects of the technology on health outcomes.

For individuals who have ABMR after pancreatic islet transplantation who receive rituximab, the evidence includes a case report. Relevant outcomes are change in disease status, quality of life, and treatment-related morbidity. The evidence is insufficient to determine the effects of the technology on health outcomes.

## Policy History

Date	Action
12/2021	BCBSA National medical policy review. No changes to policy statements. New references added.
7/2021	Updated to add drug table to the policy along with July changes.
12/2020	BCBSA National medical policy review. No changes to policy statements. New references added.
11/2020	New medical policy describing medically necessary and investigational indications. Effective 11/2020

## Information Pertaining to All Blue Cross Blue Shield Medical Policies

Click on any of the following terms to access the relevant information:

[Medical Policy Terms of Use](#)

[Managed Care Guidelines](#)

[Indemnity/PPO Guidelines](#)

[Clinical Exception Process](#)

[Medical Technology Assessment Guidelines](#)

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