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PHARMACY POLICY STATEMENT
Kentucky Medicaid

DRUG NAME	Immune globulin: Intravenous: Bivigam, Carimune NF, Flebogamma DIF, Gammagard Liquid, Gammagard S/D, Gammaked, Gammaplex, Gamunex-C, Octagam, Privigen, and Thymoglobulin Subcutaneous: Cuvitru, Hizentra, and HyQvia
BILLING CODE	J1556-Bivigam; J1566-Carimune NF; J1572-Flebogamma DIF; J1569-Gammagard Liquid; J1566-Gammagard S/D; J1561-Gammaked; J1557-Gammaplex; J1561-Gamunex-C; J1568-Octagam; J1459-Privigen; J7511-Thymoglobulin; J1555-Cuvitru; J1559-Hizentra; J1575-HyQvia
BENEFIT TYPE	Medical
SITE OF SERVICE ALLOWED	Outpatient/Office/Home
COVERAGE REQUIREMENTS	Prior Authorization Required QUANTITY LIMIT— N/A
LIST OF DIAGNOSES CONSIDERED **NOT** MEDICALLY NECESSARY	Click Here

Immune Globulin (intravenous [IVIG]: Bivigam, Carimune NF, Flebogamma DIF, Gammagard Liquid, Gammagard S/D, Gammaked, Gammaplex, Gamunex-C, Octagam, Privigen and Thymoglobulin; subcutaneous [SCIG]: Cuvitru, Hizentra and HyQvia) is a product that will only be considered for coverage under the **medical** benefit when the following criteria are met:

Members must be clinically diagnosed with one of the following disease states and meet their individual criteria as stated.
Limitations: SCIG are only indicated for primary humoral immunodeficiency.

AUTOIMMUNE BULLOUS DISEASE

For **initial** authorization:

1. Member has contraindications to, failure of (refractory to), or significant side effects from systemic corticosteroids or immunosuppressive treatment (e.g., azathioprine, cyclophosphamide, mycophenolate mofetil); AND
2. Member has dermatologic condition, as indicated by **one** or more of the following:
   a) Bullous pemphigoid;
   b) Epidermolysis bullosa acquisita;
   c) Linear IgA bullous dermatosis;
   d) Mucous membrane (cicatricial) pemphigoid;
   e) Pemphigoid gestationis;
   f) Pemphigus foliaceus;
   g) Pemphigus vulgaris.
3. **Dosage allowed:** Please see dosage and administration information in individual drug package insert.

If member meets all the requirements listed above, the medication will be approved for 6 months.

For **reauthorization**:
1. Chart notes have been provided that show the member has shown improvement of signs and symptoms of disease; AND
2. Documentation of titration to the minimum dose and frequency needed to maintain a sustained clinical effect is provided with chart notes.

If member meets all the reauthorization requirements above, the medication will be approved for an additional 12 months.

B-CELL CHRONIC LYMPHOCYTIC LEUKEMIA (CLL)

For **initial** authorization:
1. IVIG is prescribed for prophylaxis of bacterial infections; AND
2. Member has a history of recurrent sinopulmonary infections requiring intravenous antibiotics or hospitalization, AND
3. Member has a pretreatment serum IgG level <500 mg/dL (Copy of laboratory report with pre-treatment serum IgG level must be provided with chart notes).
4. **Dosage allowed:** Please see dosage and administration information in individual drug package insert.

If member meets all the requirements listed above, the medication will be approved for 6 months.

For **reauthorization**:
1. A reduction in the frequency of bacterial infections has been demonstrated since initiation of IVIG therapy.

If member meets all the reauthorization requirements above, the medication will be approved for an additional 6 months.

CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY (CIDP)

For **initial** authorization:
1. Member has moderate to severe functional disability; AND
2. Electrodiagnostic studies are consistent with multifocal demyelinating abnormalities (Pre-treatment electrodiagnostic studies (electromyography [EMG] or nerve conduction studies [NCS] provided with chart notes); AND
3. Member has elevated CSF protein (Pre-treatment cerebrospinal fluid (CSF) analysis when available).
4. **Dosage allowed:** Please see dosage and administration information in individual drug package insert.

If member meets all the requirements listed above, the medication will be approved for 3 months.

For **reauthorization**:
1. Member has significant improvement in disability and maintenance of improvement since initiation of IVIG therapy; AND
2. In those who are clinically stable and receiving long-term treatment (i.e., more than 1 year), the dose has been tapered and/or treatment withdrawn to determine whether continued treatment is necessary; AND
3. IVIG is being used at the lowest effective dose and frequency.

If member meets all the reauthorization requirements above, the medication will be approved for an additional 12 months.

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DERMATOMYOSITIS OR POLYMYOSITIS

For **initial** authorization:

1. Diagnosis established by clinical features (e.g., proximal weakness, rash), elevated muscle enzyme levels, electrodiagnostic studies (EMG/NCS), and muscle biopsy (when available); supportive diagnostic tests include autoantibody testing and muscle imaging (e.g., MRI), AND
2. Standard first-line treatments (corticosteroids or immunosuppressants) have been tried but were unsuccessful or not tolerated; OR
3. Member is unable to receive standard first-line therapy because of a contraindication or other clinical reason.
4. **Dosage allowed:** Please see dosage and administration information in individual drug package insert.

If member meets all the requirements listed above, the medication will be approved for 3 months.

For **reauthorization**:

1. Member has significant improvement in disability and maintenance of improvement since initiation of IVIG therapy.

If member meets all the reauthorization requirements above, the medication will be approved for an additional 12 months.

FETAL/NEONATAL ALLOIMMUNE THROMBOCYTOPENIA (F/NAIT)

For **initial** authorization:

1. Member is a newborn, and thrombocytopenia persists after transfusion of antigen-negative compatible platelet; OR
2. Member is pregnant and has diagnosis of F/NAIT with **one** or more of the following:
   a) Family history of disease;
   b) Platelet alloantibodies found on screening;
   c) Previously affected pregnancy.
3. **Dosage allowed:** Please see dosage and administration information in individual drug package insert.

If member meets all the requirements listed above, the medication will be approved for 6 months.

For **reauthorization**:

3. Medication will not be reauthorization for continuous use.

GUILLAIN-BARRE SYNDROME (GBS)

For **initial** authorization:

1. Physical mobility is severely affected such that member requires an aid to walk; AND
2. IVIG therapy will be initiated within 2 weeks of symptom onset.
3. **Dosage allowed:** Please see dosage and administration information in individual drug package insert.

If member meets all the requirements listed above, the medication will be approved for 2 months.

For **reauthorization**:

1. Medication will not be reauthorization for continuous use.

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IDIOPATHIC THROMBOCYTOPENIC PURPURA (IMMUNE THROMBOCYTOPENIA)

For **initial** authorization:

1. Initial therapy (Member diagnosed with ITP within the past 3 months):
   a) Children (< 18 years of age):
      i) Significant bleeding symptoms (mucosal bleeding or other moderate/severe bleeding); OR
      ii) High risk for bleeding* (see Appendix A); OR
      iii) Rapid increase in platelets is required* (e.g., surgery or procedure);
   b) Adults (≥ 18 years of age):
      i) Platelet count < 30,000/mcL; OR
      ii) Platelet count < 50,000/mcL and significant bleeding symptoms, high risk for bleeding or rapid increase in platelets is required*; AND
      iii) Corticosteroid therapy is contraindicated and IVIG will be used alone or IVIG will be used in combination with corticosteroid therapy.
2. Chronic/persistent ITP (≥ 3 months from diagnosis) or ITP unresponsive to first-line therapy (i.e., corticosteroids):
   a) Platelet count < 30,000/mcL; OR
   b) Platelet count < 50,000/mcL and significant bleeding symptoms, high risk for bleeding* or rapid increase in platelets is required*; AND
   c) Relapse after previous response to IVIG or inadequate response/intolerance/contraindication to corticosteroid or anti-D therapy.
3. Adults with refractory ITP after splenectomy:
   a) Platelet count < 30,000/mcL; OR
   b) Significant bleeding symptoms.
4. ITP in pregnant women: authorization through delivery may be granted to pregnant women with ITP if any **one** or more of the following:
   a) Any bleeding during pregnancy;
   b) Platelet count less than 10,000/mm^3 ($10x10^9$/L) at any time during pregnancy;
   c) Platelet count between 10,000/mm^3 ($10x10^9$/L) and 30,000/mm^3 ($30x10^9$/L) in second or third trimester.
5. **Dosage allowed:** Please see dosage and administration information in individual drug package insert.

***** *The member's risk factor(s) for bleeding (see Appendix A) or reason requiring a rapid increase in* platelets must be provided.

If member meets all the requirements listed above, the medication will be approved for 1 months for initial therapy, or for 6 months for chronic/persistent ITP or for adults with refractory ITP after splenectomy.

For **reauthorization**:

1. Medication will not be reauthorization for continuous use.

KAWASAKI SYNDROME

For **initial** authorization:

1. Pediatric member with Kawasaki syndrome.
2. **Dosage allowed:** Please see dosage and administration information in individual drug package insert.

If member meets all the requirements listed above, the medication will be approved for 1 months.

For **reauthorization**:
1. Medication will not be reauthorization for continuous use.

KIDNEY TRANSPLANT

For **initial** authorization:
1. Medication is used for prophylaxis or treatment of acute kidney rejection in conjunction with concomitant immunosuppression (e.g., cyclosporine, mycophenolate mofetil, and corticosteroids).
2. **Dosage allowed:** Please see dosage and administration information in individual drug package insert.

If member meets all the requirements listed above, the medication will be approved for 12 months.

LAMBERT-EATON MYASTHENIC SYNDROME (LEMS)

For **initial** authorization:
1. Member has diagnosis of LEMS and steroids and other immunosuppressive treatments do not control symptoms.
2. **Dosage allowed:** Please see dosage and administration information in individual drug package insert.

If member meets all the requirements listed above, the medication will be approved for 12 months.

For **reauthorization**:
1. Member has significant improvement in disability and maintenance of improvement since initiation of IVIG therapy.

If member meets all the reauthorization requirements above, the medication will be approved for an additional 12 months.

MULTIFOCAL MOTOR NEUROPATHY (MMN)

For **initial** authorization:
1. Member has weakness without objective sensory loss in 2 or more nerves; AND
2. Electrodiagnostic studies (electromyography [EMG]) are consistent with motor conduction block; AND
3. Normal sensory nerve conduction studies provided in chart notes.
4. **Dosage allowed:** Please see dosage and administration information in individual drug package insert.

If member meets all the requirements listed above, the medication will be approved for 3 months.

For **reauthorization**:
1. Member has significant improvement in disability and maintenance of improvement since initiation of IVIG therapy.

If member meets all the reauthorization requirements above, the medication will be approved for an additional 12 months.

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MYASTHENIA GRAVIS

For **initial** authorization:

1. Member has Neonatal Myasthenia Gravis; OR
2. Member is adult and has worsening weakness including an increase in any of the following symptoms: diplopia, ptosis, blurred vision, difficulty speaking (dysarthria), difficulty swallowing (dysphagia), difficulty chewing, impaired respiratory status, fatigue, and limb weakness. Acute exacerbations include more severe swallowing difficulties and/or respiratory failure; OR
3. Member is adult and medication used for pre-operative management (e.g., prior to thymectomy).
4. **Dosage allowed:** Please see dosage and administration information in individual drug package insert.

Note: Immune Globulin must not be used for maintenance therapy.

If member meets all the requirements listed above, the medication will be approved for 1 month.

For **reauthorization**:

1. Medication will not be reauthorization for continuous use.

PARVOVIRUS B19-INDUCED PURE RED CELL APLASIA (PRCA)

For **initial** authorization:

1. Member has parvovirus B19-induced PRCA.
2. **Dosage allowed:** Please see dosage and administration information in individual drug package insert.

If member meets all the requirements listed above, the medication will be approved for 6 months.

For **reauthorization**:

1. Medication will not be reauthorization for continuous use.

PRIMARY IMMUNODEFICIENCY

For **initial** authorization:

Member must have **one** of the following diagnoses:

1. Severe combined immunodeficiency (SCID) or congenital agammaglobulinemia (e.g., X-linked or autosomal recessive agammaglobulinemia):
   a) Diagnosis confirmed by genetic or molecular testing; OR
   b) Pretreatment IgG level < 200 mg/dL; OR
   c) Absence or very low number of T cells (CD3 T cells < 300/microliter) or the presence of maternal T cells in the circulation (SCID only);
2. Wiskott-Aldrich syndrome, DiGeorge syndrome, or ataxia-telangiectasia (or other non-SCID combined immunodeficiency):
   a) Diagnosis confirmed by genetic or molecular testing (if applicable); AND
   b) History of recurrent bacterial infections (e.g., pneumonia, otitis media, sinusitis, sepsis, gastrointestinal); AND
   c) Impaired antibody response to pneumococcal polysaccharide vaccine (see Appendix B);
3. Common variable immunodeficiency (CVID):
   a) Member is 4 years of age or older; AND
   b) Other causes of immune deficiency have been excluded (e.g., drug induced, genetic disorders, infectious diseases such as HIV, malignancy); AND
   c) Member's pretreatment IgG level < 500 mg/dL or ≥ 2 SD below the mean for age; AND
   d) Member has a history of recurrent bacterial infections; AND
   e) Member has impaired antibody response to pneumococcal polysaccharide vaccine documented in chart notes (see Appendix B);

4. Hypogammaglobulinemia (unspecified), IgG subclass deficiency, selective IgA deficiency, selective IgM deficiency, or specific antibody deficiency:
   a) Member has a history of recurrent bacterial infections; AND
   b) Member has impaired antibody response to pneumococcal polysaccharide vaccine (see Appendix B)
   c) Member has ANY of the following pre-treatment laboratory findings:
      i) Hypogammaglobulinemia: IgG < 500 mg/dL or ≥ 2 SD below the mean for age;
      ii) Selective IgA deficiency: IgA level < 7 mg/dL with normal IgG and IgM levels;
      iii) Selective IgM deficiency: IgM level < 30 mg/dL with normal IgG and IgA levels;
      iv) IgG subclass deficiency: IgG1, IgG2, or IgG3 ≥ 2 SD below mean for age assessed on at least 2 occasions; normal IgG (total) and IgM levels, normal/low IgA levels;
      v) Specific antibody deficiency: normal IgG, IgA and IgM levels.
5. Other predominant antibody deficiency disorders must meet a), b), and c) i) in section 4. above.
6. Other combined immunodeficiency must meet criteria in section 2. above.
7. **Dosage allowed:** Please see dosage and administration information in individual drug package insert. Note: Gammagard Liquid, Gamunex-C, and Gammaked may be administered intravenously or subcutaneously for primary immunodeficiency.

If member meets all the requirements listed above, the medication will be approved for 12 months.

For **reauthorization**:

1. A reduction in the frequency of bacterial infections has been demonstrated since initiation of IVIG therapy, AND
2. IgG trough levels are monitored at least yearly and maintained at or above the lower range of normal for age (when applicable for indication), OR
3. The prescriber will re-evaluate the dose of IVIG and consider a dose adjustment (when appropriate).

If member meets all the reauthorization requirements above, the medication will be approved for an additional 12 months.

PROPHYLAXIS OF BACTERIAL INFECTIONS IN BMT/HSCT RECIPIENTS

For **initial** authorization:

1. Member is BMT/HSCT recipient; AND
2. IVIG is prescribed for prophylaxis of bacterial infections; AND
3. Either of the following:
   a) IVIG is requested within the first 100 days post-transplant; OR
   b) Member has a pretreatment serum IgG < 400 mg/dL.
4. **Dosage allowed:** Please see dosage and administration information in individual drug package insert.

If member meets all the requirements listed above, the medication will be approved for 6 months.

For **reauthorization**:

1. Reduction in the frequency of bacterial infections has been demonstrated since initiation of IVIG therapy and documented in chart notes.

If member meets all the reauthorization requirements above, the medication will be approved for an additional 6 months.

PROPHYLAXIS OF BACTERIAL INFECTIONS IN HIV-INFECTED PEDIATRIC PATIENTS

For **initial** authorization:

1. Member with HIV infection and is 18 years of age or younger; AND
2. IVIG is prescribed for **primary** prophylaxis of bacterial infections and pretreatment serum IgG < 400 mg/dL; OR
3. IVIG is prescribed for **secondary** prophylaxis of bacterial infections with ALL of the following:
   a) History of recurrent bacterial infections (> 2 serious bacterial infections in a 1-year period);
   b) Member is not able to take combination antiretroviral therapy;
   c) Antibiotic prophylaxis was tried but was not effective (e.g., trimethoprim-sulfamethoxazole).
4. **Dosage allowed:** Please see dosage and administration information in individual drug package insert.

If member meets all the requirements listed above, the medication will be approved for 6 months.

For **reauthorization**:

1. Reduction in the frequency of bacterial infections has been demonstrated since initiation of IVIG therapy and documented in chart notes.

If member meets all the reauthorization requirements above, the medication will be approved for an additional 6 months.

STIFF-PERSON SYNDROME

For **initial** authorization:

1. Medication is used for treatment of stiff-person syndrome in members who have experienced an inadequate response or intolerance, or have a contraindication to first-line therapy such as a benzodiazepine (e.g., diazepam) and/or baclofen.
2. **Dosage allowed:** Please see dosage and administration information in individual drug package insert.

If member meets all the requirements listed above, the medication will be approved for 6 months.

For **reauthorization**:

1. Medication will not be reauthorization for continuous use.

CareSource considers Immune Globulin (intravenous [IVIG]: Bivigam, Carimune NF, Flebogamma DIF, Gammagard Liquid, Gammagard S/D, Gammaked, Gammaplex, Gamunex-C, Octagam, Privigen, Thymoglobulin; subcutaneous [SCIG]: Cuvitru, Hizentra, HyQvia) not medically necessary for the treatment of the following disease states based on a lack of robust clinical controlled trials showing superior efficacy compared to currently available treatments:

Acquired hemophilia	Myocarditis, acute
Adrenoleukodystrophy	Neonatal sepsis, prevention
Alzheimer's disease	Neonatal sepsis, treatment
Amyotrophic lateral sclerosis (ALS)	Ocular myasthenia
Antiphospholipid antibody syndrome (APS) in pregnancy	Paraneoplastic cerebellar degeneration, sensory neuropathy, or encephalopathy

Asthma, non-steroid dependent	Pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections (PANDAS)
Atopic dermatitis	POEMS syndrome
Autism spectrum disorders	Postinfectious cerebellar ataxia
Autoimmune liver disease	Postoperative sepsis
Autoimmune neutropenia	Pseudomembranous colitis
Campylobacter species-induced enteritis	Respiratory syncytial virus (RSV) lower respiratory tract infection
Cerebral infarctions with antiphospholipid antibodies	Rheumatic fever, acute
Chronic fatigue syndrome	Sjogren's syndrome
Demyelinative brain stem encephalitis	Spontaneous recurrent abortions, prevention
Demyelinating neuropathy associated with monoclonal IgM	Systemic lupus erythematosus
Dilated cardiomyopathy	Urticaria, chronic
HIV infection or prophylaxis	Vasculitides and antineutrophil antibody syndromes
HTLV-1-associated myelopathy	Routine prophylaxis of Measles, Varicella, and Rubella
Idiopathic dysautonomia, acute	Treatment of Measles, Varicella, and Rubella
Inclusion body myositis	
Isolated IgA deficiency	
Isolated IgG4 deficiency	
Lumbosacral or brachial plexitis	

DATE	ACTION/DESCRIPTION
11/15/2017	New policy for Immune Globulin created. Diagnoses associate with impatient life-threatening therapies were removed. Diagnoses of CIDP, Dermatomyositis or Polymyositis, ITP, MMN, Primary Immunodeficiency and Stiff-Person Syndrome got criteria expanded. Diagnosis of Acquired red cell aplasia was revised to PRCA with criteria. Length of coverage and reauthorization length were added.

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Effective date: 11/29/2017
Revised date: 11/14/2017

APPENDICES

Appendix A: Examples of Risk Factors for Bleeding (not all inclusive)

- Undergoing a medical or dental procedure where blood loss is anticipated
- Comorbidity (e.g., peptic ulcer disease, hypertension)
- Mandated anticoagulation therapy
- Profession or lifestyle predisposes patient to trauma (e.g., construction worker, fireman, professional athlete)

Appendix B: Impaired Antibody Response to Pneumococcal Polysaccharide Vaccine

- Age 6 years and older: antibody levels are not ≥ 1.3 mcg/mL for at least 70% of serotypes in the vaccine
- Age 2 to 5 years: antibody levels are not ≥ 1.3 mcg/mL for at least 50% of serotypes in the vaccine
- Not established for children less than 2 years of age