

Gamifant (emapalumab-lzsg)
Effective 01/01/2023

Plan	<input checked="" type="checkbox"/> MassHealth (UPPL) <input type="checkbox"/> Commercial/Exchange	Program Type	<input checked="" type="checkbox"/> Prior Authorization <input type="checkbox"/> Quantity Limit <input type="checkbox"/> Step Therapy
Benefit	<input checked="" type="checkbox"/> Pharmacy Benefit <input checked="" type="checkbox"/> Medical Benefit (NLX)		
Specialty Limitations			
Contact Information	Specialty Medications		
	All Plans	Phone: 866-814-5506	Fax: 866-249-6155
	Non-Specialty Medications		
	MassHealth	Phone: 877-433-7643	Fax: 866-255-7569
	Commercial	Phone: 800-294-5979	Fax: 888-836-0730
	Exchange	Phone: 855-582-2022	Fax: 855-245-2134
	Medical Specialty Medications (NLX)		
	All Plans	Phone: 844-345-2803	Fax: 844-851-0882
Exceptions	N/A		

Overview

Gamifant (emapalumab-lzsg) is an interferon gamma (IFN γ) blocking antibody indicated for the treatment of adult and pediatric (newborn and older) patients with primary hemophagocytic lymphohistiocytosis (HLH) with refractory, recurrent or progressive disease or intolerance with conventional HLH therapy.

No PA	Drugs that require PA
	Gamifant® (emapalumab-lzsg)

Coverage Guidelines

Authorization may be reviewed on a case by case basis for members who are new to the plan currently receiving treatment with requested medication excluding when the product is obtained as samples or via manufacturer’s patient assistance programs.

OR

Authorization may be granted for members when **ALL** the following criteria are met, and documentation is provided:

1. The member has a diagnosis primary hemophagocytic lymphohistiocytosis (HLH)
2. Prescriber is a specialist (e.g., hematologist and oncologist) or consult notes from a specialist are provided
3. **ONE** of the following:
 - a. Molecular tests confirming diagnosis of primary HLH
 - b. Presence of at least **5** of the following:
 - i. Fever
 - ii. Splenomegaly
 - iii. Cytopenias defined by 2 of the following: hemoglobin < 9 g/dL, platelets < 100 x 10⁹/L, neutrophils < 1 x 10⁹/L)

- iv. Hypertriglyceridemia (fasting triglyceride \geq 265 mg/dL OR $>$ 3mmol/L) and/or hypofibrinogenemia (\leq to 150 mg/dL)
 - v. Hemophagocytosis in bone marrow, spleen, or lymph nodes
 - vi. Low or absent natural killer (NK) cell activity based on laboratory reference
 - vii. Ferritin \geq 500 mcg/L
 - viii. Soluble CD25 (soluble IL-2 receptor alpha) level \geq 2400 U/mL
4. Member has active disease
 5. Member does not have active infections caused by specific pathogens favored by IFN γ neutralization (e.g., mycobacteria, Histoplasma Capsulatum, Shigella, salmonella, campylobacter, leishmanial infections)
 6. Provider documented of inadequate response, adverse reaction or contraindication to conventional HLH therapy (chemotherapy and/or systemic corticosteroids and/or immunosuppressive therapy) (*See Appendix*)
 7. **ONE** of the following:
 - a. Dexamethasone will be administered concurrently
 - b. Clinical rationale for not using dexamethasone
 8. **ONE** of the following:
 - a. Anticipated HSCT (haematopoietic stem cell transplantation) date
 - b. Member is not a candidate for HSCT
 9. Baseline clinical parameters and laboratory values including presence of fever, presence of splenomegaly, presence of CNS symptoms, hemoglobin, platelets, neutrophils, fasting triglycerides, fibrinogen, D-dimer, presence of hemophagocytosis, NK-cell activity, ferritin, and soluble CD25
 10. Appropriate dosing

Continuation of Therapy

Prescriber provides documentation of **ALL** of the following:

1. Positive response to therapy as evidenced by **ONE** of the following (*evaluate clinical parameters and laboratory values – see appendix for HLH abnormalities*):
 - a. Complete response: normalization of all HLH abnormalities
 - b. Partial response: normalization of \geq 3 HLH abnormalities
 - c. HLH improvement: \geq 3 HLH abnormalities improved by at least 50% from baseline
2. **ONE** of the following:
 - a. Dexamethasone will be administered concurrently
 - b. Clinical rationale for not using dexamethasone
3. **ONE** of the following:
 - a. Anticipated HSCT date
 - b. Member is not a candidate for HSCT

Limitations

1. Initial approvals and reauthorizations will be granted for 6 months.

Drug	Dosing
Gamifant [®] (emapalumab-lzsg) Single-dose vial: 10 mg/2 mL 50 mg/10 mL	<u>Primary Hemophagocytic Lymphohistiocytosis (HLH):</u> 1 mg/kg over on hour twice per week with dose modification based on response up to 10 mg/kg (administer until HSCT or unacceptable toxicity)

100 mg/20 mL	
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Appendix

Conventional HLH Therapies Examples
<p>Conventional HLH therapy regimens include, but are not limited to:</p> <ol style="list-style-type: none"> 1. Etoposide and dexamethasone 2. Methotrexate and hydrocortisone 3. Anti-thymocyte globulin, corticosteroids, cyclosporine, and methotrexate 4. Etoposide, dexamethasone, and cyclosporine 5. Cyclophosphamide, vincristine, and prednisone 6. Cyclophosphamide, doxorubicin, vincristine, and prednisolone (CHOP) 7. Doxorubicin, etoposide, and methylprednisolone (DEP)
HLH Abnormalities and Normalization Values for Recertification
<ol style="list-style-type: none"> 1. No presence of fever 2. No presence of splenomegaly 3. No presence of CNS symptoms 4. Platelets $\geq 100 \times 10^9/L$ 5. Neutrophils $\geq 1 \times 10^9/L$ 6. Ferritin $< 2,000 \text{ mcg/L}$ 7. Fibrinogen $> 1.5 \text{ g/L}$ 8. D-dimer $< 500 \text{ mcg/mL}$ 9. Soluble CD25 $> 2\text{-fold}$ from baseline

References

1. Gamifant® [package insert] Waltham (MA): Sobi Inc; 2020 Jun.
2. FDA approves first treatment specifically for patients with rare and life-threatening type of immune disease [press release on the Internet]. 2018 Nov 20 [cited 2022 Jan 26]. Available from: <https://www.fda.gov/news-events/press-announcements/fda-approves-first-treatment-specifically-patients-rare-and-life-threatening-type-immune-disease>.
3. Hemophagocytic Lymphohistiocytosis (HLH) [database on the Internet]: National Organization of Rare Diseases; 2018 [cited 2022 Jan 26]. Available from: <https://rarediseases.org/rare-diseases/hemophagocytic-lymphohistiocytosis/>.
4. McClain JL. Treatment and prognosis of hemophagocytic lymphohistiocytosis. In Basow DS (Ed). UpToDate [database on the Internet]. Waltham (MA): UpToDate; 2020 May 11 [cited 2022 Jan 26]. Available from: <http://www.utdol.com/utd/index.do>.
5. Daver Naval, McClain K, Allen C, et al. A consensus review on malignancy-associated hemophagocytic lymphohistiocytosis in adults. *Cancer*. 2017 September 01; 123(17): 3229–3240.
6. Lehmsberg K, Nichols KE, Henter JI, et al. Consensus recommendations for the diagnosis and management of hemophagocytic lymphohistiocytosis associated with malignancies. *Haematologica*. 2015 Aug;100(8):997-1004.
7. Locatelli F, Jordan MB, Allen C, Cesaro S, Rizzari C, Rao A, et al. Emapalumab in Children with Primary Hemophagocytic Lymphohistiocytosis *N Engl J Med*. 2020 May 7;382(19):1811-1822.
8. Gamifant® (emapalumab-lzsg) formulary dossier. Sobi Inc, Data on file.

Review History

09/21/22 – Created for Sept P&T; matched MH UPPL.

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