Mass General Brigham
Health Plan

Complement Inhibitors and Miscellaneous Immunosuppressive Agents
Enjaymo (sutimlimab-jome)
Rystiggo (rozanolixizumab-noli)
Soliris (eculizumab)
Syfovre (pegcetacoplan)
Uplizna (inebilizumab-cdon)
Vyvgart (efgartigimod alfa-fcab)
Vyvgart Hytrulo (efgartigimod alfa and hyaluronidase-qvfc)
Effective 02/18/2025

Plan	☒ MassHealth UPPL ☐Commercial/Exchange	**Program Type**	☒ Prior Authorization ☐ Quantity Limit ☐ Step Therapy
Benefit	☐ Pharmacy Benefit ☒ Medical Benefit		
Specialty Limitations	N/A		
Contact Information	**Medical and Specialty Medications** All Plans | Phone: 877-519-1908 | Fax: 855-540-3693		
	Non-Specialty Medications All Plans | Phone: 800-711-4555 | Fax: 844-403-1029		
Notes	Additional agents from this class are available through the pharmacy benefit. Please see the MassHealth Drug List for coverage and criteria.		

Overview

Enjaymo (sutimlimab-jome) is an immunoglobulin G (IgG), subclass 4 monoclonal antibody that is indicated to decrease the need for red blood cell (RBC) transfusion due to hemolysis in adults with cold agglutinin diseases (CAD).

Rystiggo (rozanolixizumab-noli) is a neonatal Fc receptor blocker indicated for the treatment of gMG in adult patients who are anti-AChR or anti-MuSK antibody positive.

Soliris (eculizumab) is indicated for the treatment of atypical hemolytic uremic syndrome (aHUS), generalized myasthenia gravis (MG), neuromyelitis optica spectrum disorder (NMOSD), and paroxysmal nocturnal hemoglobinuria (PNH).

Syfovre (pegcetacoplan) is indicated for the treatment of geographic atrophy (GA) secondary to age-related macular degeneration (AMD).

Uplizna (inebilizumab-cdon) is a CD19-directed cytolytic antibody that is presumed to be involved in CD19 binding. Following binding, inebilizumab-cdon depletes lymphocytes derived from B-cell lineage.

Vyvgart (efgartigimod alfa-fcab) is indicated for the treatment of gMG in adult patients who are anti- AChR antibody positive.

Vyvgart Hytrulo (efgartigimod alfa and hyaluronidase-qvfc) is a subcutaneous product combination of efgartigimod alfa, a human IgG1 antibody fragment marketed for intravenous use as Vyvgart (efgartigimod alfa), and recombinant human hyaluronidase PH20 (rHuPH20), which is Halozyme's ENHANZE® drug delivery technology to increase permeability of the subcutaneous tissue by depolymerizing hyaluronan, facilitating subcutaneous delivery of biologics.

Coverage Guidelines
Authorization may be granted for members new to the plan who are currently receiving treatment with the requested product 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:

Enjaymo (sutimlimab-jome)
1. Diagnosis of cold agglutinin disease (CAD)
2. Prescriber is a hematologist or consult notes from specialist are provided
3. Member is ≥18 years of age
4. Hb ≤10 g/dL (dated within the last 60 days)
5. Appropriate dosing

Rystiggo (rozanolixizumab-noli)
1. Diagnosis of Generalized Myasthenia Gravis
2. Member is ≥18 years of age
3. Member is AChR or MuSK antibody positive
4. Prescriber is a neurologist or consult notes from a neurology office are provided
5. Inadequate response, adverse reaction, or contraindication to pyridostigmine
6. **ONE** of the following:
    a. **BOTH** of the following:
        i. Member has severe disease requiring faster onset medication
        ii. Inadequate response, adverse reaction or contraindication to IVIG or plasmapheresis with glucocorticoids
    b. Inadequate response or adverse reaction to **TWO** of the following or contraindication to **ALL** of the following immunosuppressant trials:
        i. azathioprine
        ii. cyclosporine
        iii. glucocorticoids (e.g., prednisone)
        iv. mycophenolate
        v. tacrolimus
7. **ONE** of the following:
    a. Inadequate response, adverse reaction, or contraindication to Vyvgart (efgartigimod alfa-fcab) or Vyvgart Hytrulo (efgartigimod alfa and hyaluronidase-qvfc)
    b. Member is MuSK antibody positive
8. Appropriate dosing

Soliris (eculizumab)
Atypical hemolytic-uremic syndrome (aHUS)
1. Diagnosis of atypical hemolytic-uremic syndrome (aHUS)
2. Prescriber is a hematologist or nephrologist or consult notes from specialist are provided

3. Appropriate dosing

Generalized Myasthenia Gravis

1. Diagnosis of Generalized Myasthenia Gravis
2. Member is ≥18 years of age
3. Member is AchR antibody positive
4. Prescriber is a neurologist or consult notes from a neurology office are provided
5. Inadequate response, adverse reaction, or contraindication to pyridostigmine
6. **ONE** of the following:
    a. **BOTH** of the following:
        i. Member has severe disease requiring faster onset medication
        ii. Inadequate response, adverse reaction or contraindication to IVIG or plasmapheresis with glucocorticoids
    b. Inadequate response or adverse reaction to **TWO** of the following or contraindication to **ALL** of the following immunosuppressant trials:
        i. azathioprine
        ii. cyclosporine
        iii. glucocorticoids (e.g., prednisone)
        iv. mycophenolate
        v. tacrolimus
7. Appropriate dosing

Neuromyelitis optica spectrum disorder (NMOSD)

1. Diagnosis of neuromyelitis optica spectrum disorder
2. Documentation of a positive serologic test for anti-aquaporin-4 (AQP4)
3. Member is ≥18 years of age
4. Appropriate dosing

Paroxysmal nocturnal hemoglobinuria (PNH)

1. Appropriate diagnosis
2. Appropriate dosing
3. Member is ≥18 years of age

PLE/CHAPLE (off-label indication)

1. Diagnosis of CD55-deficient protein-losing enteropathy (PLE), or complement hyperactivation, angiopathic thrombosis, and protein-losing enteropathy (CHAPLE) disease
2. Member is ≥2 months of age
3. Prescriber is a specialist in rare genetic or hematologic diseases or consult notes from specialist are provided
4. Results from genetic testing confirming a CD55 loss-of-function mutation
5. Appropriate dosing

Syfovre (pegcetacoplan)

1. Diagnosis of GA secondary to AMD
2. Prescriber is an ophthalmologist
3. Member is ≥ 50 years of age
4. Absence of choroidal neovascularization (CNV or Wet-AMD) in the treatment eye
5. Normal luminance best corrected visual acuity (BCVA) ≥24 letters (20/320 Snellen equivalence)

6. Total GA lesion area ≥2.5 and ≤17.5 mm2, with at least 1 lesion ≥1.25 mm2 if GA is multifocal.
7. Presence of any pattern of hyperautofluorescence in the junctional zone of GA.
8. Requested dosing is 15mg (0.1 mL) once every 25 days to 60 days

Uplizna (inebilizumab-cdon)

1. Diagnosis of neuromyelitis optica spectrum disorder (NMOSD)
2. Documentation of a positive serologic test for anti-aquaporin-4 (AQP4)
3. Member is ≥18 years of age
4. Appropriate dosing

Vyvgart (efgartigimod alfa-fcab)
Vyvgart Hytrulo (efgartigimod alfa and hyaluronidase-qvfc)

1. Diagnosis of Generalized Myasthenia Gravis
2. Member is ≥18 years of age
3. Member is AChR antibody positive
4. Prescriber is a neurologist or consult notes from a neurology office are provided
5. Inadequate response, adverse reaction, or contraindication to pyridostigmine
6. **ONE** of the following:
    a. **BOTH** of the following:
        i. Member has severe disease requiring faster onset medication
        ii. Inadequate response, adverse reaction or contraindication to IVIG or plasmapheresis with glucocorticoids
    b. Inadequate response or adverse reaction to **TWO** of the following or contraindication to **ALL** of the following immunosuppressant trials:
        i. azathioprine
        ii. cyclosporine
        iii. glucocorticoids (e.g., prednisone)
        iv. mycophenolate
        v. tacrolimus
7. Appropriate dosing

Vyvgart Hytrulo (efgartigimod alfa and hyaluronidase-qvfc)

1. Diagnosis of chronic inflammatory demyelinating polyneuropathy (CIDP)
2. Member is ≥ 18 years of age
3. Prescriber is a neurologist or consult notes from neurologist are provided
4. Appropriate dosing
5. **TWO** of the following:
    a. Inadequate response, adverse reaction, or contraindication to immune globulin
    b. Inadequate response, adverse reaction, or contraindication to plasma exchange
    c. **ONE** of the following:
        i. Inadequate response or adverse reaction to glucocorticoids (e.g., budesonide, methylprednisolone, prednisone)
        ii. **BOTH** of the following:
            1. Contraindication to glucocorticoids
            2. Inadequate response or adverse reaction to immunosuppressants (e.g., azathioprine, cyclosporine, cyclophosphamide, mycophenolate mofetil, rituximab)

Continuation of Therapy

aHUS/CIDP/PNH/NMOSD: Reauthorization by physician will infer a positive response to therapy.

AMD (Syfovre):

1. Positive response to therapy
2. Member has not developed nAMD (wet AMD)
3. If requested dosing is ≥ every 60 days, prescriber has assessed using less frequent dosing

CAD/Generalized MG (Soliris)/GPA/MPA: Reauthorization requires physician documentation of a positive response to therapy.

Generalized MG (Vyvgart): Reauthorization by physician will infer a positive response to therapy.

PLE/CHAPLE disease

Medical records documenting **ALL** of the following:

1. Improvement or no worsening of clinical symptoms (e.g., abdominal pain, bowel movements, facial and peripheral edema)
2. **ONE** of the following:
    a. Increase in current serum albumin concentration from baseline serum albumin concentration
    b. Serum albumin concentration stabilized above lower threshold for normal range (≥3.5 g/dL)
3. **ONE** of the following:
    a. Increase in current serum IgG concentration from baseline serum IgG concentration
    a. Serum IgG concentration stabilized above lower threshold for age-adjusted normal range *(See appendix for table of age-adjusted normal serum IgG concentration ranges)*

Limitations

1. Initial approvals will be granted for the following:
    a. aHUS/PNH/NMOSD, PLE/CHAPLE: 1 year
    b. Generalized MG (Rystiggo), CIDP (Vyvgart Hytrulo): 3 months
    c. CAD/Generalized MG (Soliris)/GPA/MPA/AMD (Syfovre): 6 months
    d. Generalized MG (Vyvgart, Vyvgart Hytrulo): 2 months
2. Reauthorizations will be granted for the following:
    a. Generalized MG (Vyvgart, Vyvgart Hytrulo): 2 months
    b. Generalized MG (Rystiggo): 3 months
    c. All other indications: 1 year

Appendix

Soliris: Age-Adjusted Serum IgG Concentration Reference Ranges

The table below shows age-adjusted serum immunoglobulin G (IgG) concentration reference ranges to assist with recertification of Soliris (eculizumab)requests indicated for CHAPLE disease. Based on a brief literature review, there does not appear to be a widely accepted set of age-adjusted IgG reference ranges, and these ranges may vary by laboratory. Therefore, if the provider submits medical records with documented serum IgG laboratory testing, reference ranges specified in the medical records may be acceptable in place of the values in the table below.

Age Group	Age-Adjusted Serum IgG Level Reference Range
≤1 month	251-1051

1-3 months	176-601
4-6 months	172-814
7-12 months	217-1213
13-24 months	424-1051
25-36 months	441-1135
3-5 years	441-1236
6-8 years	633-1280
9-11 years	608-1572
12-16 years	1066-1218
16-18 years	1188-1458
≥18 years	639-1349

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3. Enspryng® [package insert]. South San Francisco (CA): Genentech.; 2020 Aug.
4. Uplizna® [package insert]. Gaithersburg (MD): Viela Bio.; 2021 Jul.
5. Vyvgart® [package insert]. Boston (MA): Argenx US, Inc. 2021 Dec.
6. Enjaymo® [prescribing information]. Waltham (MA): Bioverative USA Inc.; 2022 Feb.
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Review History

02/08/2023 - Reviewed and created for Feb P&T; matched MH UPPL criteria to be in compliance with Masshealth unified formulary requirements. Effective 4/1/23
04/12/23 – Reviewed and updated for P&T. Added GPA/MPA to initial approval durations. Effective 6/5/23.
06/14/23 – Reviewed and updated for P&T. Removed preferred product requirement from Enjaymo, Uplizna, Soliris for requests reviewing under MB. Effective 6/30/23.
09/13/23 – Reviewed and updated for P&T. Enjaymo was updated and now requires members to have received a vaccine against encapsulated bacteria at least two weeks prior to treatment initiation. Due to the Medical Benefit Analysis, a decision was made to update Soliris (eculizumab), Vyvgart (efgartigimod alfa-fcab), Uplizna (inebilizumab-cdon) and Enjaymo (sutimlimab-jome) within this guideline to be managed through medical billing and designated with MB. Effective 10/2/23
2/14/24 – Reviewed and updated for P&T. Added Rystiggo and Vyvgart Hytrulo to policy requiring PA through MB (did not include preferred product requirement). Effective 3/4/24
04/10/24 – Reviewed and updated for P&T. Criteria added for Soliris for off label diagnosis of PLE/CHAPLE. New appendix “Age-Adjusted Serum IgG Concentration Reference Ranges” was created. **Syfovre** (pegcetacoplan 150 mg/mL vial) age criteria updated from ≥ 60 to ≥ 50 years of age. Effective 5/6/24
01/2025 – Reviewed and updated for P&T. Removed Tavneos from MB policy as it is being managed through pharmacy. Removed vaccination requirement throughout policy. Criteria for myasthenia gravis indications now includes severity of disease and trial with IVIG or plasmapheresis with glucocorticoids. Added expanded indication of CIDP for Vyvgart Hytrulo. Effective 2/18/25