GENERIC NAME	BRAND NAME	JCODE	COMMENTS
ANIFROLUMAB-FNIA	SAPHNELO	J0491	

CAMS PA Guideline

GUIDELINES FOR COVERAGE

INITIAL CRITERIA: Must meet all the following:
1. Prescriber is a rheumatologist
2. Patient is at least 18 years of age
3. Patient has diagnosis of moderate to severe SLE WITHOUT active lupus nephritis
4. Patient with failure, intolerance, or contraindication to at least 2 of the following immunosuppressants: hydroxychloroquine, azathioprine, mycophenolate mofetil, mycophenolic acid, cyclophosphamide
5. Patient with failure, intolerance, contraindication to Benlysta (belimumab)

If above criteria are met, approve x 1 year
If above criteria are not met, do not approve

RENEWAL CRITERIA: Must meet the following criteria:
1. Provider attests to improvement in condition.

If renewal criteria is met, approve x 1 year
If renewal criteria are not met, do not approve

RATIONALE
Per plan.

FDA APPROVED INDICATIONS
Treatment of adult patients with moderate SLE without active lupus nephritis

REFERENCES

Creation Date: 01/2023
Effective Date: 02/2023
Reviewed Date:
Revised Date:

CABOTEGRAVIR (APRETUDE)

GENERIC NAME	BRAND NAME	JCODE	COMMENTS
Cabotegravir	Apretude	J3490	Apretude only – for PrEP, NOT Cabenuva or Vocabria

CAMS GUIDELINES FOR COVERAGE
NOT APPLICABLE FOR MEDICARE PART B ENROLLEES

INITIAL CRITERIA: Must meet both of the following:
1. Ordered by Infectious Diseases provider
2. Patient is age 12 years or older AND patient has experienced adverse events with, or has a contraindication to, both oral preexposure prophylaxis (PrEP) medications [i.e. Truvada (emtricitabine/tenofovir disiproxil fumarate) and Descovy (emtricitabine/tenofovir alafenamide)]

If met, approve x 8 months (4 doses).
If not met, do not approve. Suggest oral PrEP (generic emtricitabine/tenofovir disiproxil fumarate preferred).

RENEWAL CRITERIA: Must meet the following:
1. Patient has NOT missed 2 or more scheduled administrations of Apretude by 7 or more days.

If renewal criteria are met, approve x 12 months.
If initial criteria are not met, do not approve.

RATIONALE

1. Oral PrEP agents after safe and effective options to prevent HIV infection.
2. Although Apretude offers greater relative risk reduction compared to oral PrEP drugs, 59 – 87 patients need to receive Apretude over the course of 3 years to prevent one additional case of HIV infection.

FDA APPROVED INDICATIONS

1. **Preexposure prophylaxis of HIV infection in patients at least 12 years of age.**

REFERENCES

Creation Date: 01/2023
Effective Date:
Reviewed Date:
Revised Date:

GENERIC NAME	BRAND NAME	J-CODE	COMMENTS
Eculizumab	Soliris	J1300	

GUIDELINES FOR COVERAGE

TRANSPLANT CRITERIA[&]:

INITIAL AND NEW MEMBER CRITERIA: Must have all the following:

A. Medication must be prescribed by a transplant provider.
B. Request is for treatment of complement mediated antibody rejection of transplanted organ

If met, approve for 6 weeks
If not met, do not approve

RENEWAL CRITERIA:

C. Must meet the following step therapy based on dosing frequency:
    1. If q 2 week frequency is requested, then patient must have trialed and failed, or has an intolerance or contraindication to, ravulizumab-cwvz (Ultomiris)
        If met, then approve for duration of treatment plan.
        If not met then do not approve must try raviluzumab.

    2. If q 4 week or greater frequency is requested, then approve for duration of treatment plan.
        If met, approve for duration of treatment plan.
        If not met, do not approve.

ONCOLOGY CRITERIA:

INITIAL AND NEW MEMBER CRITERIA: Must have one of the following indications and meet applicable diagnosis-specific criteria in A or B:

A. Request is for treatment of aHUS (atypical hemolytic uremic syndrome): Patient must have trialed and failed, or has an intolerance or contraindication to, ravulizumab-cwvz (Ultomiris)
B. Request is for treatment of PNH (paroxysmal nocturnal hemoglobinuria): Patient must have trialed and failed, or has an intolerance or contraindication to, ravulizumab-cwvz (Ultomiris), and pegcetacoplan (Empaveli) [after trial of Ultomiris (ravulizumab/eculizumab)].

If initial criteria are met, approve for duration of treatment plan.
If Initial Criteria are not met, do not approve.

RENEWAL CRITERIA:
If patient is currently on eculizumab and tolerating, authorize for duration of treatment plan.
If not met, do not approve (see alternatives above).

NEUROLOGY CRITERIA:

INITIAL CRITERIA: Must meet all General criteria in section A, plus applicable diagnosis-specific criteria in either section B or C:

A – General criteria for all neurology-related requests
B – Diagnosis-specific criteria for Neuromyelitis Optica Spectrum Disorder (NMOSD/NMO)

CAMS ECULIZUMAB (SOLIRIS)

C – Diagnosis-specific criteria for Treatment-Refractory Generalized Myasthenia Gravis (gMG)

A. General criteria for all neurology-related requests: Must meet all the following:
   1. The patient is 18 years or older
   2. At the time of request, the patient does not have a serious unresolved Neisseria meningitidis infection
   3. Patient has either received meningococcal vaccine 14 days prior to eculizumab start, or the patient will receive appropriate prophylaxis until 14 days after meningococcal vaccination
   4. Patient must have one of the following diagnoses: Neuromyelitis Optica Spectrum Disorder (NMOSD/NMO) or Treatment-Refractory Generalized Myasthenia Gravis (gMG)

B. Neuromyelitis Optica Spectrum Disorder (NMOSD/NMO): Must meet all the following:
   1. Medication must be prescribed by a neurologist
   2. Diagnosis of NMO/NMOSD and not MS
   3. Seropositive for anti-AQP4-antibody
   4. The patient has a history of at least 1 relapse in the last 12 months or two relapses in the last 2 years
   5. Member must have experienced either:
      a. A severe***** breakthrough relapse while on rituximab[¥] or biosimilar for at least 6-months at recommended NMO dosing****** not attributed to rapid steroid withdrawal or discontinuation
      b. Recurrent breakthrough relapse after 6-month trial of rituximab[¥] or its biosimilar at recommended NMO dosing****** in combination with maximum tolerated doses of either mycophenolate mofetil[¥] or azathioprine[¥]
   6. Patient must have tried and failed or have contraindication to Uplizna (nonformulary) or Enspryng (PA required)

C. Treatment-Refractory Generalized Myasthenia Gravis (gMG): Must meet all the following:
   1. Diagnosis of gMG
   2. Evaluated by a neuromuscular/electrophysiology - electromyography (EMG) fellowship-trained neurologist from CPMG or affiliated network with appropriate referral, if needed
   3. Positive serologic test for anti-acetylcholine receptor (AChR) antibodies[§]
   4. No history of thymic neoplasms or a thymectomy within 12 months prior to treatment initiation
   5. Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class II to IV
   6. Myasthenia Gravis-Specific Activities of Daily Living scale (MG-ADL) total score of at least 6 at baseline
   7. Patient is currently taking chronic corticosteroid with pyridostigmine as prescribed unless there is an intolerance or contraindication to one or both
   8. The patient has tried and failed, or has contraindication to, at least ONE of the following oral disease modifying therapies, and ALL of the following infused disease modifying therapies
      ORAL
      a. azathioprine[¥], 2 mg/kg daily, for at least 9-12 months
      b. cyclophosphamide[¥], mycophenolate mofetil[¥], cyclosporine[¥], methotrexate[¥]) for at least 6-9 months

INFUSED

a. Efgartigimod (Vyvgart)
b. Rituximab or its biosimilar
c. Chronic IVIG

If initial criteria are met, authorize for duration and quantity of therapy plan.
If initial criteria are not met, do not approve.

RENEWAL CRITERIA
Must meet indication-specific criteria below:

A. For Treatment of Neuromyelitis Optica Spectrum Disorder: Must meet all the following:
   1. Patient is responding positively to therapy including, but not limited to, improvement or stabilization in any one of the following parameters: frequency of relapse, pain, fatigue, motor function, progression of symptoms, or visual acuity.

B. For Treatment-Refractory Generalized Myasthenia Gravis: Must meet all the following:
   1. The patient has at least a 2-point improvement in the Myasthenia Gravis-Specific Activities of Daily Living scale (MG-ADL) from baseline
   2. Patient has not had a dose increase or add-on of immunosuppressive therapies (IST) since starting eculizumab (Soliris), and has either discontinued, reduced, or maintained the same dose of immunosuppressive therapies (IST) started prior to eculizumab.

If renewal criteria are met, authorize for duration and quantity of therapy plan.
If Initial Criteria are not met, do not approve.

RATIONALE

Eculizumab (Soliris) is a humanized monoclonal antibody that blocks terminal complement activation. Eculizumab (Soliris) was the first FDA-approved agent treatment for AQP4 antibody-positive NMOSD. Inebilizumab (Uplizna), an anti-CD19 monoclonal antibody, was the second to be approved, followed by Satralizumab (Enspryng). With the exception of satralizumab, a subcutaneous injection, all other agents are infused intravenously. Eculizumab for NMOSD was studied in a randomized, double-blinded, placebo-controlled phase III time-to-event trial over 4 years, Prevention of Relapses in Neuromyelitis Optica (PREVENT). One hundred percent of the patients were positive for presence of anti-AQP4 antibodies. Baseline annualized relapse rate for the placebo group was 2.7 and for eculizumab was 1.94. The majority of patients in both the eculizumab (78%) and placebo (72%) groups continued on prior immunotherapy including glucocorticoids, azathioprine, and mycophenolate mofetil. A significantly smaller proportion of patients in the eculizumab group experienced an adjudicated relapse compared with the placebo group (3% vs 43%). The adjudicated annualized relapse rate for placebo was .35 events per year and .02 event per year for eculizumab group. Change from baseline in disability measures including EDSS, modified Rankin scale, and Hauser Ambulation Index did not significantly differ between the two groups. There are some criticisms of this study including that 109 of 143 patients were able to continue on with other immunosuppressants. In addition to the active treatment group, the placebo group showed decent relapse reduction at end of study. The efficacy for the placebo group was likely related to the use of self-reported relapses at baseline, changes in relapse evaluation by adjudication committee during the study. Also imaging evidence of relapse was not needed and patients censored from the study at time of relapse. There is no data that eculizumab is more effective or safer than current standards of treatment. Evidence for maintenance therapy of NMOSD is with off - label immunosuppressive therapies including rituximab (or biosimilar), mycophenolate mofetil, and azathioprine with prednisone when necessary is clinically significant also. Treatment with these agents was associated with significant reductions in annualized relapse rates in the range of 72%-88%. The KP ETSP treatment guidelines recommend that

rituximab +/- oral immunosuppressants be used prior to the newer agents. Off-label use of rituximab is supported by a CMS-approved Compendia resource. LexiComp categorizes NMOSD (relapse prevention) as an off-label use for rituximab based on Level of Evidence [B,G].

Treatment – Refractory Myasthenia Gravis

Ensure appropriate criteria are used for the management of requests for eculizumab (Soliris) according to the approved FDA indication and national Kaiser Permanente treatment guidelines. Data are limited to one Phase 3 study of a specific subpopulation of patients with generalized myasthenia gravis who have demonstrated treatment failure or intolerance to either 2 or more ISTs or 1 IST plus 12 months of treatment with either IVIG or PE. Given the limited clinical trial data and lack of long-term safety data, exceedingly judicious prescribing and monitoring of therapy are warranted. The KP ETSP treatment guidelines recommend that rituximab +/- oral immunosuppressants be used prior to the newer agents. Off-label use of rituximab is supported by a CMS-approved Compendia resource. LexiComp categorizes off-label rituximab for use in Myasthenia Gravis (refractory) with level of evidence [B,G] and Micromedex categorizes off-label use of rituximab for Myasthenia Gravis (refractory); Strength of recommendation Adult, Class1. Strength of evidence, Adult Category B.

FDA APPROVED INDICATIONS:
Atypical Hemolytic Uremic Syndrome (aHUS)
Paroxysmal Nocturnal Hemoglobinuria (PNH)
Neuromyelitis Optica Spectrum Disorder
Refractory Myasthenia gravis

[&] OFF-LABELED INDICATIONS:
Complement mediated antibody rejection of renal transplant
Complement mediated antibody rejection of heart transplant

APPENDIX

*Examples of severe breakthrough relapse include but are not limited to:

- Hospitalization for neurological deficits from NMOSD relapse (e.g., muscle weakness that affects both legs (paraparesis); muscle weakness that affects one side of body such as left arm and left leg (hemiparesis); muscle weakness that affects all four limbs (quadriparesis)
- Optic neuritis severity (hand motion only or worse) confirmed by an ophthalmologist

**NMO dosing for rituximab or biosimilar requires a minimum of 1000mg at a fixed interval of every 6 months dosing

[¥] Peer-Reviewed Evidence-Based and CMS Compendia Approved Therapies: rituximab, azathioprine, mycophenolate

[§]Positive antibody status does NOT include anti-muscle-specific receptor tyrosine kinase (MuSK) or anti-low-density lipoprotein receptor-related protein (LRP4) antibodies.

REFERENCES

1. Pittock et al. Eculizumab in Aquaporin-4-positive Neuromyelitis Optica Spectrum Disorder. New England Journal of Medicine. 2019, 281. 614-625
2. Cree BA, Lamb S, Morgan K, Chen A, Waubant E, Genain C. An open label study of the effects of rituximab in neuromyelitis optica. Neurology. 2005;64(7):1270-1272.

3. Jacob A, Weinshenker BG, Violich I, et al. Treatment of neuromyelitis optica with rituximab: retrospective analysis of 25 patients. Arch Neurol. 2008;65(11):1443-1448.
4. Mandler RN, Ahmed W, Dencoff JE. Devic's neuromyelitis optica: a prospective study of seven patients treated with prednisone and azathioprine. Neurology. 1998;51(4): 1219-1220.
5. Costanzi C, Matiello M, Lucchinetti CF, et al. Azathioprine: tolerability, efficacy, and predictors of benefit in neuromyelitis optica. Neurology. 2011;77(7):659-666.
6. Elsone L, Kitley J, Luppe S, et al. Long-term efficacy, tolerability and retention rate of azathioprine in 103 aquaporin-4 antibody-positive neuromyelitis optica spectrum disorder patients: a multicentre retrospective observational study from the UK. Mult Scler. 2014;20(11):1533-1540.
7. Jacob A, Matiello M, Weinshenker BG, et al. Treatment of neuromyelitis optica with mycophenolate mofetil: retrospective analysis of 24 patients. Arch Neurol. 2009;66(9): 1128-1133.
8. Huh SY, Kim SH, Hyun JW, et al. Mycophenolate mofetil in the treatment of neuromyelitis optica spectrum disorder. JAMA Neurol. 2014;71(11):1372-1378.
9. Kim SH, Huh SY, Lee SJ, Joung A, Kim HJ. A 5-year follow-up of rituximab treatment in patients with neuromyelitis optica spectrum disorder. JAMA Neurol. 2013;70(9):1110-1117.
10. Zéphir H, Bernard-Valnet R, Lebrun C, et al. Rituximab as firstline therapy in neuromyelitis optica: efficiency and tolerability. J Neurol. 2015;262(10):2329-2335
11. Papp V, Illes Z, Magyari M, et al. Nationwide prevalence and incidence study of neuromyelitis optica spectrum disorder in Denmark. Neurology 2018;91:e2265-e2275. doi:10.1212/WNL.0000000000006645
12. KP Drug Information Services, California Regions. Guideline Update: Eculizumab (Soliris) for Myasthenia Gravis, Last reviewed 5.9.2018
13. Howard JF Jr, Utsugisawa K, Benatar M, et al. Safety and efficacy of eculizumab in anti-acetylcholine receptor antibody-positive refractory generalized myasthenia gravis (REGAIN): a phase 3, randomized, double-blind, placebo-controlled, multicentre study. Lancet Neurol. 2017, Oct 20. pii: S1474-4422(17)30369-1.
14. Soliris (eculizumab) [package insert]. Alexion Pharmaceuticals, Inc. New Haven, CT. November 2020.
15. Rituximab. Lexicomp Online, Pediatric and Neonatal Lexi-Drugs Online. Waltham, MA: UpToDate, Inc.; 1/27/23. https://online.lexi.com. Accessed January 31, 2023.
16. Rituximab. Micromedex® (electronic version). Merative, Ann Arbor, Michigan, USA. Available at: https://www.micromedexsolutions.com/ (cited: January 31, 2023).
17. Jehn U, Altuner U, Pavenstade H et al. First Report on Successful Conversion of Long-Term Treatment of Recurrent Atypical Hemolytic Uremic Syndrome with Eculizumab to Ravulizuma in a Renal Transplant Patient. Transpl Int. 2022;35:10846.
18. Rondeau E, Scully M, Ariceta G et al. The long-acting C5 inhibitor, ravulizumab, is effective and safe in adult patients with atypical hemolytic uremic syndrome naïve to complement inhibitor treatment. Kidney International. 2020;97:1287-1296.

Creation Date: 10/2019
Effective Date: 10/2023
Reviewed Date: 09/2023

CAMS ECULIZUMAB (SOLIRIS)

Revised Date: 09/2023

GENERIC NAME	BRAND NAME	J-CODE	COMMENTS
Efgartigimod alfa-fcab	Vyvgart	J3490	

GUIDELINES FOR COVERAGE

INITIAL CRITERIA: Must meet all of the following:
1. The patient is 18 years or older
2. Evaluated by a neuromuscular/electrophysiology - electromyography (EMG) fellowship-trained neurologist from CPMG or affiliated network with appropriate referral, if needed
3. Patient must be diagnosed with Generalized Myasthenia Gravis (gMG)
4. Positive serologic test for anti-acetylcholine receptor (AChR) antibodies[§]
5. No history of thymic neoplasms or a thymectomy within 3 months prior to treatment initiation
6. Myasthenia Gravis Foundation of America (MGFA) Clinical Classification Class II to IV
7. Myasthenia Gravis-Specific Activities of Daily Living scale (MG-ADL) total score of at least 5 at baseline (with >50% MG-ADL non-ocular related)
8. Patient is currently taking chronic corticosteroid with pyridostigmine as prescribed unless there is an intolerance or contraindication to one or both
9. The patient has tried and failed, or has contraindication to at least ONE of the following disease modifying therapies and BOTH of the following infused disease modifying therapies
    ORAL
        a. azathioprine[¥], 2 mg/kg daily, for at least 9-12 months
        b. cyclophosphamide[¥], mycophenolate mofetil[¥], cyclosporine[¥], methotrexate[¥]) for at least 6-9 months
    INFUSED
        a. Rituximab or its biosimilar
        b. Chronic IVIG

If Initial Criteria are met, authorize for duration and quantity of therapy plan.
If Initial Criteria are not met, do not approve.

RENEWAL CRITERIA: Must meet 1 and either 2 or 3 below:
1. Request is at least 50 days from the first day of the previous efgartigimod treatment cycle
2. This request is for the first renewal of efgartigimod: the patient has at least a 2-point improvement in the Myasthenia Gravis-Specific Activities of Daily Living scale (MG-ADL) from baseline
3. This request is for a patient who has received 2 or more cycles of efgartigimod: the patient has maintained a stable MG-ADL score over the last 12 months

If renewal criteria are met, authorize for duration and quantity of therapy plan.
If Initial Criteria are not met, do not approve.

RATIONALE
Efgartigimod alpha-fcab injection (Vyvgart) is a neonatal FC receptor blocker that blocks recycling and increased degeneration of autoantibodies in generalized Myasthenia Gravis. While it is the first agent to

be FDA-approved with this novel mechanism of action, it is the second drug to be FDA approved for the treatment of gMG.
Data is limited to one, small (n=167) 26-week Phase 3 study of patients with generalized myasthenia gravis, the majority of whom were acetylcholine receptor positive (77%), white, and MGFA class III. Fifty-seven percent of patients had undergone a thymectomy with differential rates in the efgartigimod group (70%) compared to placebo (43%). Though the mean time since thymectomy was 10.84 (SD 9.0) years, patients could enroll as early as 3 months post-thymectomy.

All patients were required to be on a stable dose of at least one treatment for gMG (ie, acetylcholinesterase inhibitors, corticosteroids, or NSISTs). At baseline 71% of percent of patients were on a steroid, 61% were on a non-steroidal immunosuppressant therapy and 51% were on both treatments. Nineteen percent were on no treatment. Additionally, patients were allowed to have trialed rituximab or eculizumab but just not within 6 months of study enrollment.

Efgartigimod infusion was found to be more effective than placebo for the primary outcome of percentage of Myasthenia Gravis-Activities of Daily Living (MG-ADL) responders at week 8 (odds ratio [OR] 4.95; 95% confidence interval [CI], 2.21 to 11.53; P<0.001; absolute risk reduction [ARR] 38% / number needed to treat [NNT] 3).[1] Responders were defined as patients with a two or more point reduction in the MG-ADL total score compared to baseline that was maintained for four consecutive weeks, with the first reduction occurring no later than one week after the last infusion of the product after 4 weeks of initial treatment. Approximately, 70% of patients experienced the minimum point reduction in the MG-ADL to be classified as a responder with a clinically significant change.

*Adverse reactions occurring in 10% or more of patients treated with efgartigimod, and more frequently than placebo, are respiratory tract infections, headache and urinary tract infections. Serious adverse reactions occurred in 5% of efgartigimod patients versus 8% in the placebo group. Efgartigimod transiently reduces IgG levels and should not be given if the patient has an active infection and immunization with live-attenuated or live vaccines is not recommended during treatment.
who have demonstrated treatment failure or intolerance to either 2 or more ISTs or 1 IST plus 12 months of treatment with either IVIG or therapeutic plasma exchange. Given the limited clinical trial data and lack of long-term safety data, exceedingly judicious prescribing and monitoring of therapy are warranted. The KP ETSP treatment guidelines recommend that rituximab +/- oral immunosuppressants be used prior to the newer agents. Off-label use of rituximab is supported by a CMS-approved Compendia resource. LexiComp categorizes off-label rituximab for use in Myasthenia Gravis (refractory) with level of evidence [B,G] and Micromedex categorizes off-label use of rituximab for Myasthenia Gravis (refractory); Strength of recommendation Adult, Class1. Strength of evidence, Adult Category B.

FDA APPROVED INDICATIONS

APPENDIX

*Examples of severe breakthrough relapse include but are not limited to:

- Hospitalization for neurological deficits from NMOSD relapse (e.g., muscle weakness that affects both legs (paraparesis); muscle weakness that affects one side of body such as left arm and left leg (hemiparesis); muscle weakness that affects all four limbs (quadriparesis)
- Optic neuritis severity (hand motion only or worse) confirmed by an ophthalmologist

**NMO dosing for rituximab or biosimilar requires a minimum of 1000mg at a fixed interval of every 6 months dosing

¥ Peer-Reviewed Evidence-Based and CMS Compendia Approved Therapies: rituximab, azathioprine, mycophenolate

[§]Positive antibody status does NOT include anti-muscle-specific receptor tyrosine kinase (MuSK) or anti-low-density lipoprotein receptor-related protein (LRP4) antibodies.

REFERENCES

1. Pittock et al. Eculizumab in Aquaporin-4-positive Neuromyelitis Optica Spectrum Disorder. New England Journal of Medicine. 2019, 281. 614-625
2. Cree BA, Lamb S, Morgan K, Chen A, Waubant E, Genain C. An open label study of the effects of rituximab in neuromyelitis optica. Neurology. 2005;64(7):1270-1272.
3. Jacob A, Weinshenker BG, Violich I, et al. Treatment of neuromyelitis optica with rituximab: retrospective analysis of 25 patients. Arch Neurol. 2008;65(11):1443-1448.
4. Mandler RN, Ahmed W, Dencoff JE. Devic's neuromyelitis optica: a prospective study of seven patients treated with prednisone and azathioprine. Neurology. 1998;51(4): 1219-1220.
5. Costanzi C, Matiello M, Lucchinetti CF, et al. Azathioprine: tolerability, efficacy, and predictors of benefit in neuromyelitis optica. Neurology. 2011;77(7):659-666.
6. Elsone L, Kitley J, Luppe S, et al. Long-term efficacy, tolerability and retention rate of azathioprine in 103 aquaporin-4 antibody-positive neuromyelitis optica spectrum disorder patients: a multicentre retrospective observational study from the UK. Mult Scler. 2014;20(11):1533-1540.
7. Jacob A, Matiello M, Weinshenker BG, et al. Treatment of neuromyelitis optica with mycophenolate mofetil: retrospective analysis of 24 patients. Arch Neurol. 2009;66(9): 1128-1133.
8. Huh SY, Kim SH, Hyun JW, et al. Mycophenolate mofetil in the treatment of neuromyelitis optica spectrum disorder. JAMA Neurol. 2014;71(11):1372-1378.
9. Kim SH, Huh SY, Lee SJ, Joung A, Kim HJ. A 5-year follow-up of rituximab treatment in patients with neuromyelitis optica spectrum disorder. JAMA Neurol. 2013;70(9):1110-1117.
10. Zéphir H, Bernard-Valnet R, Lebrun C, et al. Rituximab as firstline therapy in neuromyelitis optica: efficiency and tolerability. J Neurol. 2015;262(10):2329-2335
11. Papp V, Illes Z, Magyari M, et al. Nationwide prevalence and incidence study of neuromyelitis optica spectrum disorder in Denmark. Neurology 2018;91:e2265-e2275. doi:10.1212/WNL.0000000000006645
12. KP Drug Information Services, California Regions. Guideline Update: Eculizumab (Soliris) for Myasthenia Gravis, Last reviewed 5.9.2018
13. Howard JF Jr, Utsugisawa K, Benatar M, et al. Safety and efficacy of eculizumab in anti-acetylcholine receptor antibody-positive refractory generalized myasthenia gravis (REGAIN): a phase 3, randomized, double-blind, placebo-controlled, multicentre study. Lancet Neurol. 2017, Oct 20. pii: S1474-4422(17)30369-1.
14. Soliris (eculizumab) [package insert]. Alexion Pharmaceuticals, Inc. New Haven, CT. November 2020.
15. Vyvgart (efgartigimod) [package insert]. Argenx US, Inc. Boston, MA. April 2022

Creation Date: 5/2022
Effective Date: 9/2022
Reviewed Date: 3/2023
Revised Date: 3/2023

CAMS - LEQVIO (Inclisiran)

GENERIC NAME	BRAND NAME	J-CODE	COMMENTS
INCLISIRAN	LEQVIO	J3490	Nonformulary

CAMS GUIDELINES FOR COVERAGE

INITIAL CRITERIA: Must meet all the following:

1. Patient must have a diagnosis of atherosclerotic cardiovascular disease (ASCVD) with a clinical event^^ or a diagnosis of heterozygous familial hypercholesterolemia (HeFH)
2. Patient must:
    a. have been taking atorvastatin 80 mg or rosuvastatin 40 mg daily or statin therapy at the maximally tolerated dose for at least 30 days prior to LDL lab
    OR
    b. have an absolute contraindication to statin therapy (active, decompensated liver disease; nursing female, pregnancy, or plans to become pregnant; hypersensitivity reaction; documented history of CPK >10x ULN or rhabdomyolysis attributed to a statin and not explained by a drug interaction, fall, or prolonged immobility)
    OR
    c. be statin-intolerant as defined by the National Lipid Association Statin (NLA) Intolerance Panel**
3. Patient must have been taking ezetimibe for at least 30 days prior to LDL lab or have a contraindication or intolerance to ezetimibe
4. Patient must have a contraindication or intolerance to a PCSK9 inhibitor (i.e. evolocumab (Repatha) or alirocumab (Praluent))
5. Patient must have a current LDL level drawn within the last 6 months of greater than or equal to one of the following:
    a. 55 mg/dL for ASCVD at very high risk defined as multiple ASCVD events^^ or 1 ASCVD event and 2 or more high risk conditions (age ≥ 65 years, familial hypercholesterolemia, diabetes, HTN, eGFR 15-59, current smoking)
    b. 70 mg/dL for ASCVD not at very high risk
    c. 100 mg/dL for HeFH

If initial criteria are met, approve x 12 months.
If criteria are not met, do not approve.

RENEWAL CRITERIA
Patient's LDL must have decreased by at least 20% after starting the Leqvio (inclisiran).

If met, approve for the duration of the order.
If criteria are not met, do not approve.

---

^^Includes: MI, ACS, CAD with intervention (e.g. PCI, stent, CABG), ischemic noncardioembolic stroke, PAD with intervention (e.g. stent, surgery); Excludes: High CAC score, AAA, CAD finding on diagnostic cath without MI/ACS/intervention, CAD equivalents (e.g. DM, CKD), primary prevention patients regardless of CV risk score
** Inability to tolerate at least 2 statins, with at least one at the lowest starting daily dose

Created: 3/2022
Effective:
Reviewed: 3/2023
Revised: 3/2023