Reference number
2482-A

SPECIALTY GUIDELINE MANAGEMENT

SOLIRIS (eculizumab) ENHANCED SGM

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered covered benefits provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications

A. Paroxysmal nocturnal hemoglobinuria (PNH) to reduce hemolysis
B. Atypical hemolytic uremic syndrome (aHUS) to inhibit complement-mediated thrombotic microangiopathy

Limitations of Use: Soliris is not indicated for the treatment of patients with Shiga toxin E. Coli related hemolytic uremic syndrome (STEC-HUS).

All other indications are considered experimental/investigational and are not covered benefits.

II. REQUIRED DOCUMENTATION

Submission of the following information is necessary to initiate the prior authorization review for new requests for treatment of:

A. Atypical hemolytic uremic syndrome: ADAMTS 13 level
B. Paroxysmal nocturnal hemoglobinuria: deficiency of glycosylphosphatidylinositol (GPI)-anchored proteins, flow cytometry used to show results of GPI-APs deficiency

III. CRITERIA FOR INITIAL APPROVAL

A. Atypical hemolytic uremic syndrome

Authorization of 6 months may be granted for treatment of atypical hemolytic uremic syndrome not caused by Shiga toxin when all of the following criteria are met:

1. ADAMTS 13 activity level above 5%
2. Absence of Shiga toxin

B. Paroxysmal nocturnal hemoglobinuria

Authorization of 6 months may be granted for treatment of paroxysmal nocturnal hemoglobinuria when all of the following criteria are met:

1. Deficiency of glycosylphosphatidylinositol-anchored proteins (GPI-APs)
2. Flow cytometry is used to demonstrate GPI-APs deficiency

IV. CONTINUATION OF THERAPY

A. Atypical hemolytic uremic syndrome

Authorization of 12 months may be granted to all members (including new members) requesting continuation of therapy provided they meet all initial authorization criteria and demonstrate a positive response to therapy (e.g., normalization of LDH levels, platelet counts).

© 2018 CVS Caremark. All rights reserved.

This document contains confidential and proprietary information of CVS Caremark and cannot be reproduced, distributed or printed without written permission from CVS Caremark. This document contains prescription brand name drugs that are trademarks or registered trademarks of pharmaceutical manufacturers that are not affiliated with CVS Caremark.

B. Paroxysmal nocturnal hemoglobinuria
Authorization of 12 months may be granted to all members (including new members) requesting continuation of therapy provided they meet all initial authorization criteria and demonstrate a positive response to therapy (e.g., improvement in hemoglobin levels, normalization of LDH levels).

IV. REFERENCES

1. Soliris [package insert]. New Haven, CT: Alexion Pharmaceuticals, Inc.; October 2017.
2. Loirat C, Fakhouri F, Ariceta G, et al. An international consensus approach to the management of atypical hemolytic uremic syndrome in children. *Pediatr Nephrol*. Published online: April 11, 2015.
3. Parker CJ. Management of paroxysmal nocturnal hemoglobulinuria in the era of complement inhibitory therapy. *Hematology.* 2011; 21-29.
4. Sanders D, Wolfe G, Benatar M et al. International consensus guidance for management of myasthenia gravis. *Neurology*. 2016; 87 (4):419-425.
5. Jaretzki A, Barohn RJ, Ernstoff RM et al. Myasthenia Gravis: Recommendations for Clinical Research Standards. *Ann Thorac Surg*. 2000;70: 327-34.
6. Hillmen P, Young NS, Schubert J, et al. The complement inhibitor eculizumab in paroxysmal nocturnal hemoglobinuria. *NEJM.* 2006;335:1233-43.
7. Howard JF, Utsugisawa K, Benatar M. Safety and efficacy of eculizumab in anti-acetylcholine receptor antibody-positive refractory generalized myasthenia gravis (REGAIN); a phase 3, randomized, double-blind, placebo-controlled, multicenter study. *Lancet Neurol*. 2017 Oct 20. http://dx.doi.org/10.1016/S1474-4422(17)30369-1Ingenix HCPCS Level II, Expert 2011.

© 2018 CVS Caremark. All rights reserved.

This document contains confidential and proprietary information of CVS Caremark and cannot be reproduced, distributed or printed without written permission from CVS Caremark. This document contains prescription brand name drugs that are trademarks or registered trademarks of pharmaceutical manufacturers that are not affiliated with CVS Caremark.