BlueCross BlueShield
Federal Employee Program

SOLIRIS (eculizumab)
BKEMV* (eculizumab-aeeb)
EPYSQLI* (eculizumab-aagh)

*These medications are included in this policy but are not available on the market as of yet

RATIONALE FOR INCLUSION IN PA PROGRAM

Background

Soliris and its biosimilars are complement inhibitors indicated for the treatment of patients with paroxysmal nocturnal hemoglobinuria (PNH), atypical hemolytic uremic syndrome (aHUS), generalized myasthenia gravis (gMG), and neuromyelitis optica spectrum disorder (NMOSD). Soliris and its biosimilars are humanized monoclonal IgG antibodies that binds to complement protein C5, preventing cleavage into C5a and C5b. Blocking the formation of C5b inhibits the subsequent formation of terminal complex C5b-9 or MAC. Terminal complement-mediated intravascular hemolysis is a key clinical feature of paroxysmal nocturnal hemoglobinuria (PNH), blocking the formation of membrane attack complex (MAC) results in stabilization of hemoglobin and a reduction in the need for RBC transfusions. Impairment of complement activity regulation leads to uncontrolled complement activation in atypical hemolytic uremic syndrome (aHUS) (1-4).

Regulatory Status

FDA-approved indications: Soliris and its biosimilars are complement inhibitors indicated for: (1-3)

1. The treatment of patients with paroxysmal nocturnal hemoglobinuria (PNH) to reduce hemolysis.
2. The treatment of patients with atypical hemolytic uremic syndrome (aHUS) to inhibit complement-mediated thrombotic microangiopathy.
    a. Limitation of Use: Soliris and its biosimilar are not indicated for the treatment of patients with Shiga toxin E. coli related hemolytic uremic syndrome (STEC-HUS).
3. The treatment of generalized myasthenia gravis (gMG) in adult and pediatric patients six years of age and older who are anti-acetylcholine receptor (AchR) antibody positive.
4. The treatment of neuromyelitis optica spectrum disorder (NMOSD) in adult patients who are anti-aquaporin-4 (AQP4) antibody positive.

The International Consensus Guidance for Management of Myasthenia Gravis recommends the

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use of chronic IVIG and immunosuppressants (5).

Soliris and its biosimilars include a boxed warning citing the risk of life-threatening and fatal meningococcal infections. Additionally, all patients must be vaccinated with a meningococcal vaccine at least 2 weeks prior to receiving their first dose (1-3).

Soliris and its biosimilars are available only through a restricted program under a Risk Evaluation and Mitigation Strategy (REMS). Under the REMS, prescribers must enroll in the program (1-3).

In addition, Soliris and its biosimilars have warnings regarding infusion-related reactions and using caution when administering to patients with any other systemic infection (1-3).

The safety and effectiveness of Soliris and its biosimilars for the treatment of PNH and NMOSD in pediatric patients less than 18 years of age have not been established. The safety and effectiveness of Soliris and its biosimilars for the treatment of gMG in pediatric patients less than 6 years of age have not been established. The safety and effectiveness of Soliris and its biosimilars for the treatment of aHUS have been established in pediatric patients (1-3).

Summary

Soliris and its biosimilars are complement inhibitors indicated for the treatment of patients with paroxysmal nocturnal hemoglobinuria (PNH), atypical hemolytic uremic syndrome (aHUS), generalized myasthenia gravis (gMG) and neuromyelitis optica spectrum disorder (NMOSD). Soliris and its biosimilars include a boxed warning citing the risk of life-threatening and fatal meningococcal infections. Soliris and its biosimilars are not indicated for the treatment of patients with Shiga toxin E. coli- related hemolytic uremic syndrome (STEC-HUS). Soliris and its biosimilars are available only through a restricted program under a Risk Evaluation and Mitigation Strategy (REMS). In addition, Soliris and its biosimilars have warnings regarding infusion-related reactions and using caution when administering to patients with any other

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systemic meningococcal infections. The safety and effectiveness of Soliris and its biosimilars for the treatment of PNH and NMOSD in pediatric patients less than 18 years of age have not been established. The safety and effectiveness of Soliris and its biosimilars for the treatment of gMG in pediatric patients less than 6 years of age have not been established. The safety and effectiveness of Soliris and its biosimilars for the treatment of aHUS have been established in pediatric patients (1-3).

Prior authorization is required to ensure the safe, clinically appropriate, and cost-effective use of Soliris and its biosimilars while maintaining optimal therapeutic outcomes.

References

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5. Sanders DB, Wolfe GI, Benatar M, et al. International consensus guidance for management of myasthenia gravis: Executive summary. *Neurology*. 2016; 87(4):419. Epub 2016 Jun 29.