Subject: **Eculizumab (Soliris)**

Effective Date: **September 11, 2007**

Department(s): **Utilization Management**

Policy: Eculizumab (Soliris) (**HCPCS J1300**), a monoclonal antibody complement inhibitor, is reimbursable under Plans administered by QualCare, Inc. under the circumstances enumerated in this Policy.

Objective: To assure proper and consistent reimbursement and to delineate criteria for coverage of a specific therapeutic agent.

Procedure:

A. For use in treating paroxysmal nocturnal hemoglobinuria (PNH) (ICD-**9 283.2;** ICD-**10 D59.5**), the physician ordering Soliris must document the following information:

1. The data supporting the diagnosis of PNH, by flow cytometry documentation of PNH AND at least one of the following:
   a. Evidence of acquired hemolysis (negative direct Coombs test)
   b. Evidence of intravascular hemolysis, including at least one of the following:
      i. Hemoglobinemia
      ii. Hemoglobinuria
      iii. Hemosiderinuria
   c. Granulocytopenia and/or thrombocytopenia in the presence of an elevation in the reticulocyte count

d. Venous thrombosis
e. Aplastic anemia with evidence of acquired hemolysis (negative direct Coombs test) or thrombosis
f. Myelodysplastic syndrome (refractory anemia variant)
g. Episodic dysphagia or abdominal pain with evidence of intravascular hemolysis

2. Complications of PNH experienced by this patient
3. The patient's transfusion requirement (if any)
4. All prior therapy
5. The reasons why this specific patient should receive Soliris, apart from the fact that he/she has PNH
6. The individual has received or will receive a meningococcal vaccine in conjunction with eculizumab therapy

B. For the treatment of **atypical** hemolytic uremic syndrome (**ICD-9-ICD-10-no code specific to the atypical subset**), documentation of all of the following:

1. Presence of microangiopathic hemolytic anemia.
2. C3 complement level below the lower limit of normal for the reporting laboratory.
3. Shiga-toxin producing E. coli infection has been ruled out.
4. Blood activity level of the ADAMTS-13 protein of >5 % OR a trial of plasma exchange did not result in clinical improvement (to exclude thrombotic thrombocytopenic purpura)
5. The individual has received or will receive a meningococcal vaccine in conjunction with eculizumab therapy

C. For the treatment of recurrent post-renal transplant atypical hemolytic uremic syndrome and the individual has received or will receive a meningococcal vaccine in conjunction with eculizumab therapy.

D. For the prevention of recurrent atypical hemolytic uremic syndrome in a renal transplant recipient who has **either** an identified mutation in the CFH, CF1, C3 or CFB genes, **or** has had a previous post-transplant episode of atypical hemolytic uremic syndrome, and the individual has received or will receive a meningococcal vaccine in conjunction with eculizumab therapy

NOTE: Soliris is **not** reimbursable for typical hemolytic uremic syndrome, shiga toxin E. coli related hemolytic uremic syndrome, or any other indication as it is considered experimental, investigational, or unproven.

E. Anti-acetylcholine receptor antibody positive refractory generalized myasthenia gravis (**ICD-10 G70.00, G70.01**) with documentation of the following:
Positive serologic test for anti-acetylcholine receptor(AChR) antibody, treatment failure over 1 year or more with either two immunosuppressive agents (e.g. azathioprine, mycophenolate, cyclosporin), OR one immunosuppressive agent and required chronic plasma exchange or intravenous immunoglobulin. Treatment failure consists of continued difficulty breathing or swallowing, or functional disability causing discontinuation of physical activity. Initial authorization for this indication will be for 12 weeks.

F. All requests for Soliris must be reviewed by the medical director before this preparation can be authorized.

<u>FDA Recommended Dosing</u>
Patients must be administered a meningococcal vaccine at least two weeks prior to initiation of Soliris therapy and revaccinated according to current medical guidelines for vaccine use.
PNH:
600 mg weekly for the first 4 weeks, followed by 900 mg for the fifth dose 1 week later, then 900 mg every 2

weeks thereafter. Soliris should be administered at the recommended dosage regimen time points, or within two days of these time points.

aHUS:
For patients 18 years of age and older, eculizumab therapy consists of 900 mg weekly for the first 4 weeks, followed by 1200 mg for the fifth dose 1 week later, then 1200 mg every 2 weeks thereafter.

For individuals under 18 years of age Soliris is administered according to body weight as shown in the the following table:

Body Weight	Induction	Maintenance
≥ 40 kg	900 mg weekly x 4doses	1200 mg at week 5; then 1200 mg every 2 weeks
30 kg to <40 kg	600 mg weekly x 2 doses	900 mg at week 3; then 900 mg every 2 weeks
20 kg to < 30 kg	600 mg weekly x 2 doses	600 mg at week 3; then 600 mg every 2 weeks
10 kg to < 20kg	600 mg weekly x 1 dose	300 mg at week 2; then 300 mg every 2 weeks
5 kg to < 10kg	300 mg weekly x 1 dose	300 mg at week 2; then 300 mg every 3 weeks

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*Consistent with Summary Plan Description (SPD). If there is discordance with the SPD, provisions of the SPD take precedence.