CODING AND BILLING GUIDE FOR THE USE OF SOLIRIS® (eculizumab)

In Atypical Hemolytic Uremic Syndrome (Atypical-HUS)

INDICATION

Atypical Hemolytic Uremic Syndrome (aHUS)

Soliris is indicated for the treatment of patients with atypical hemolytic uremic syndrome (aHUS) to inhibit complement-mediated thrombotic microangiopathy.

Limitation of Use

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

SELECT IMPORTANT SAFETY INFORMATION

WARNING: SERIOUS MENINGOCOCCAL INFECTIONS

Life-threatening and fatal meningococcal infections have occurred in patients treated with Soliris and may become rapidly life-threatening or fatal if not recognized and treated early.

- **Comply with the most current Advisory Committee on Immunization Practices (ACIP) recommendations for meningococcal vaccination in patients with complement deficiencies.**
- **Immunize patients with meningococcal vaccines at least 2 weeks prior to administering the first dose of Soliris, unless the risks of delaying Soliris therapy outweigh the risk of developing a meningococcal infection. (See *Serious Meningococcal Infections* for additional guidance on the management of the risk of meningococcal infection).**
- **Vaccination reduces, but does not eliminate, the risk of meningococcal infections. Monitor patients for early signs of meningococcal infections and evaluate immediately if infection is suspected.**

Soliris is available only through a restricted program under a Risk Evaluation and Mitigation Strategy (REMS). Under the Soliris REMS, prescribers must enroll in the program. Enrollment in the Soliris REMS program and additional information are available by telephone: 1-888-SOLIRIS (1-888-765-4747) or at www.solirisrems.com.

Please see additional Important Safety Information on pages 1 and 11-12 and the full Prescribing Information for SOLIRIS® (eculizumab), including Boxed WARNING regarding serious meningococcal infections.

SOLIRIS®
(eculizumab)
Injection for Intravenous Use
300 mg/30 mL vial

US/SOL-a/0046 10/22

Purpose of This Guide and Disclaimer

Alexion Pharmaceuticals, Inc. has developed the SOLIRIS® (eculizumab) Coding and Billing Guide to provide objective and publicly available coding and billing information.

This document is provided for informational purposes only and is not legal advice or official guidance from payers. It is not intended to increase or maximize reimbursement. Alexion does not warrant, promise, guarantee, or make any statement that the use of this information will result in coverage or payment for SOLIRIS, or that any payment received will cover providers' costs. Alexion is not responsible for any action providers take in billing for, or appealing, SOLIRIS claims.

Hospitals and physicians are responsible for compliance with Medicare and other payer rules and requirements and for the information submitted with all claims and appeals. Before any claims or appeals are submitted, hospitals and physicians should review official payer instructions and requirements, should confirm the accuracy of their coding or billing practices with these payers, and should use independent judgment when selecting codes that most appropriately describe the services or supplies provided to a patient.

Please visit www.SOLIRIS.net for additional information or call 1-888-765-4747 to speak with the Alexion OneSource™ Team.

Please see additional Important Safety Information on pages 1 and 11-12 and the full Prescribing Information for SOLIRIS® (eculizumab), including Boxed WARNING regarding serious meningococcal infections.

Coding for SOLIRIS® (eculizumab) in Atypical-HUS

Fiscal Year 2023 Diagnosis Coding Updates

The Centers for Medicare & Medicaid Services (CMS) has updated the ICD-10-CM codes to include two codes specific to the atypical-HUS diagnosis. The following International Classification of Diseases, 10th Revision, Clinical Modification (ICD-10-CM) diagnosis code may be appropriate to describe patients diagnosed with atypical-HUS:

ICD-10-CM Diagnosis Code[1]	D59.39	D59.32
Code Descriptor	**Other hemolytic-uremic syndrome** • Atypical (nongenetic) hemolytic uremic syndrome • Secondary hemolytic-uremic syndrome	**Hereditary hemolytic-uremic syndrome** • Atypical hemolytic uremic syndrome with an identified genetic cause
Appropriate Use	Assign this code when medical record documentation supports that atypical hemolytic uremic syndrome is not further specified as due to a genetic cause	Assign this code when medical record documentation supports that atypical hemolytic uremic syndrome is due to a genetic cause
Coding Instructional Notes[1]	**Code first**, if applicable, any associated: • COVID-19 (U07.1) • complications of kidney transplant (T86.1-) • complications of heart transplant (T86.2-) • complications of liver transplant (T86.4-) **Code also**, if applicable, any associated condition, such as: • hypertensive emergency (I16.1) • malignant neoplasm (C00-C96) • systemic lupus erythematosus (M32.-) **Use additional code**, if applicable, for adverse effect to identify drug (T36-T50 with fifth or sixth character 5)	**Code also**, if applicable: • defects in the complement system (D84.1) • methylmalonic acidemia (E71.120)

IMPORTANT NOTE: Fiscal Year 2022 Diagnosis Coding

The previously active ICD-10-CM code D59.3 (Hemolytic-uremic syndrome) will be replaced by the codes in the above table beginning October 1, 2022.

Coding Tip: Coding atypical-HUS to the highest level of specificity requires 5 characters. Use only valid codes based on medical record documentation to avoid claims processing delays.

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Drug Coding

The following drug-specific Healthcare Common Procedure Coding System (HCPCS) billing code can be reported on medical claim forms to payers:

HCPCS Code[2]	Code Descriptor
J1300	Injection, eculizumab, 10 mg

Some payers may also require the use of HCPCS modifier -RE to indicate SOLIRIS® (eculizumab) was administered in full compliance with the REMS program.

Some payers, including Medicaid, require drugs like SOLIRIS to be billed on medical claims with the product's National Drug Code (NDC) in addition to the HCPCS code. Payers typically require healthcare professionals to use the Health Insurance Portability and Accountability Act (HIPAA)-compliant, 11-digit NDC format[4]:

11-Digit NDC[3,4]	Code Descriptor	Strength
25682-0001-01	SOLIRIS single-use vial	300 mg/30 mL

Please note that payers have different guidance for placement of the NDC on medical claims. Typically, the 11-digit NDC is reported without any dashes or other punctuation.[4]

Some payers may also require a unit of measure (UoM) qualifier. For SOLIRIS, the unit of measure qualifier is mL (milliliter).

Check payer requirements for reporting the NDC and UoM on claims.

Drug Administration Services

Payers may offer separate coverage and reimbursement for drug administration services. The following are possible ICD, 10th Revision, Procedure Coding System (ICD-10-PCS) procedure codes to report the administration of SOLIRIS in inpatient settings:

ICD-10-PCS[5]	Code Descriptor
3E033GR	Introduction of other therapeutic monoclonal antibody into peripheral vein, percutaneous approach
3E043GR	Introduction of other therapeutic monoclonal antibody into central vein, percutaneous approach

The following Current Procedural Terminology (CPT®) codes may be appropriate to report administration of SOLIRIS in physician offices and hospital outpatient facilities. Individual payer policies should be reviewed for reporting requirements.

CPT[6]	Code Descriptor
96365	Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); initial, up to one hour
+ 96366	Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); each additional hour (list separately in addition to primary procedure)
96413	Chemotherapy administration, intravenous infusion technique, up to one hour, single or initial substance/drug
+ 96415	Chemotherapy administration, intravenous infusion technique; each additional hour (list separately in addition to primary procedure)

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Coding for Meningococcal Vaccination

Diagnosis Coding

For an encounter strictly for the vaccination, the diagnosis code for prophylactic vaccination is assigned along with the diagnosis code for aHUS and any other conditions the patient may have.

ICD-10-CM Diagnosis Code[2]	Code Descriptor
Z23	Encounter for immunization

Vaccine Coding

CPT Code[6]	Code Descriptor
90619	Meningococcal conjugate vaccine, serogroups A, C, W, Y, quadrivalent, tetanus toxoid carrier (MenACWY-TT), for intramuscular use
90620	Meningococcal recombinant protein and outer membrane vesicle vaccine, serogroup B (MenB-4C), 2 dose schedule, for intramuscular use
90621	Meningococcal recombinant lipoprotein vaccine, serogroup B (MenB-FHbp), 2 or 3 dose schedule, for intramuscular use
90644	Meningococcal conjugate vaccine, serogroups C & Y and Haemophilus influenzae type b vaccine (Hib-MenCY), 4-dose schedule, when administered to children 6 weeks–18 months of age, for intramuscular use
90733	Meningococcal polysaccharide vaccine, serogroups A, C, Y, W-135, quadrivalent (MPSV4), for subcutaneous use
90734	Meningococcal conjugate vaccine, serogroups A, C, W, Y, quadrivalent, diphtheria toxoid carrier (MenACWY-D) or CRM197 carrier (MenACWY-CRM), for intramuscular use

Vaccine Administration Coding

CPT Code[6]	Code Descriptor
90460	Immunization administration through 18 years of age via any route of administration, with counseling by physician or other qualified healthcare professional; first or only component of each vaccine or toxoid administered
+ 90461	Immunization administration through 18 years of age via any route of administration, with counseling by physician or other qualified healthcare professional; each additional vaccine or toxoid component administered (List separately in addition to code for primary procedure)
90471	Immunization administration (includes percutaneous, intradermal, subcutaneous, or intramuscular injections); 1 vaccine (single or combination vaccine/toxoid)
+ 90472	Immunization administration (includes percutaneous, intradermal, subcutaneous, or intramuscular injections); each additional vaccine (single or combination vaccine/toxoid) (List separately in addition to code for primary procedure)

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Claim Forms

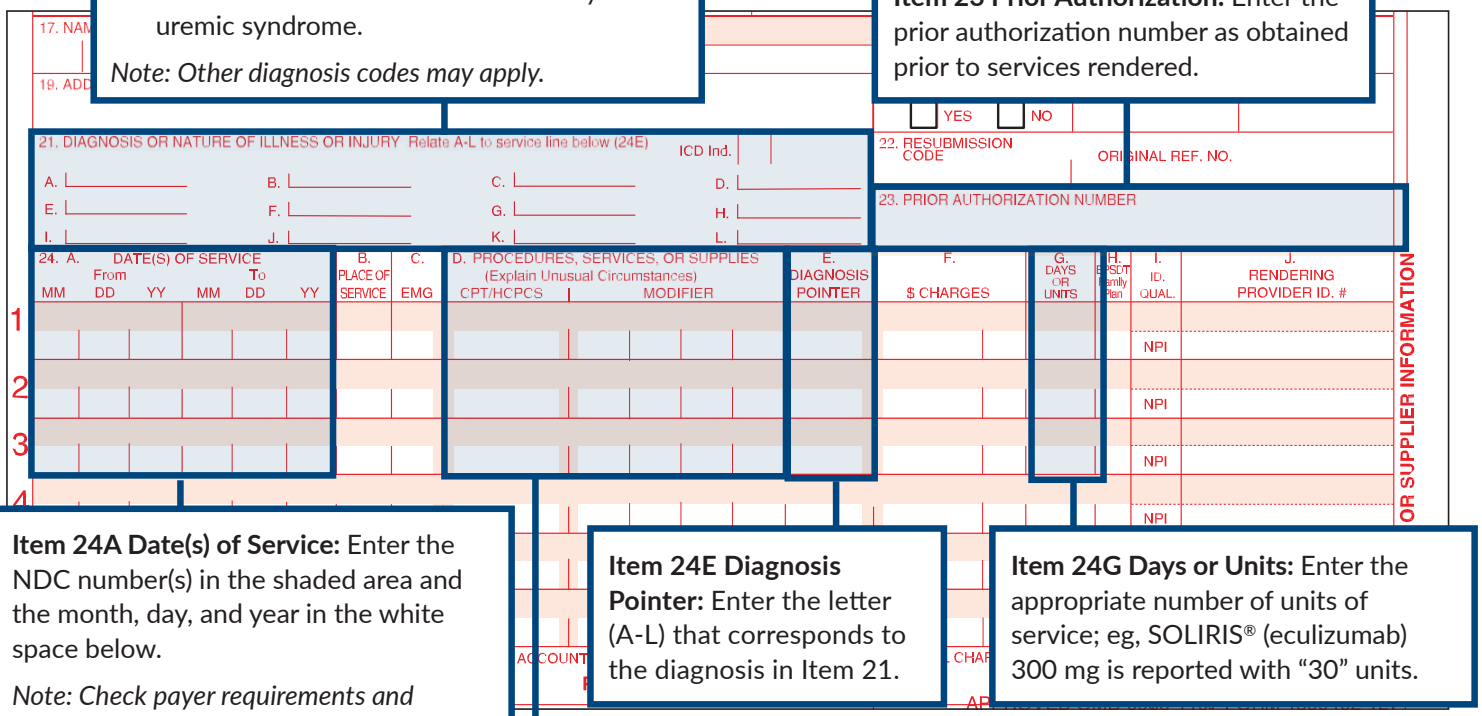
Sample CMS-1500 (or the electronic equivalent 837P): Physician Office

For an example of a completed CMS-1500 form, go to page 7.

Item 21 Diagnosis: Enter the appropriate diagnosis code; eg,
- ICD-10-CM: D59.39 for other hemolytic uremic syndrome.

Note: Other diagnosis codes may apply.

Item 23 Prior Authorization: Enter the prior authorization number as obtained prior to services rendered.



Item 24A Date(s) of Service: Enter the NDC number(s) in the shaded area and the month, day, and year in the white space below.

Note: Check payer requirements and format for reporting NDC.

Item 24E Diagnosis Pointer: Enter the letter (A-L) that corresponds to the diagnosis in Item 21.

Item 24G Days or Units: Enter the appropriate number of units of service; eg, SOLIRIS® (eculizumab) 300 mg is reported with "30" units.

Item 24D Procedures/Services/Supplies: Enter the appropriate HCPCS/CPT codes and modifiers; eg,
- J1300 Injection, eculizumab, 10 mg
- 96365 for drug administration

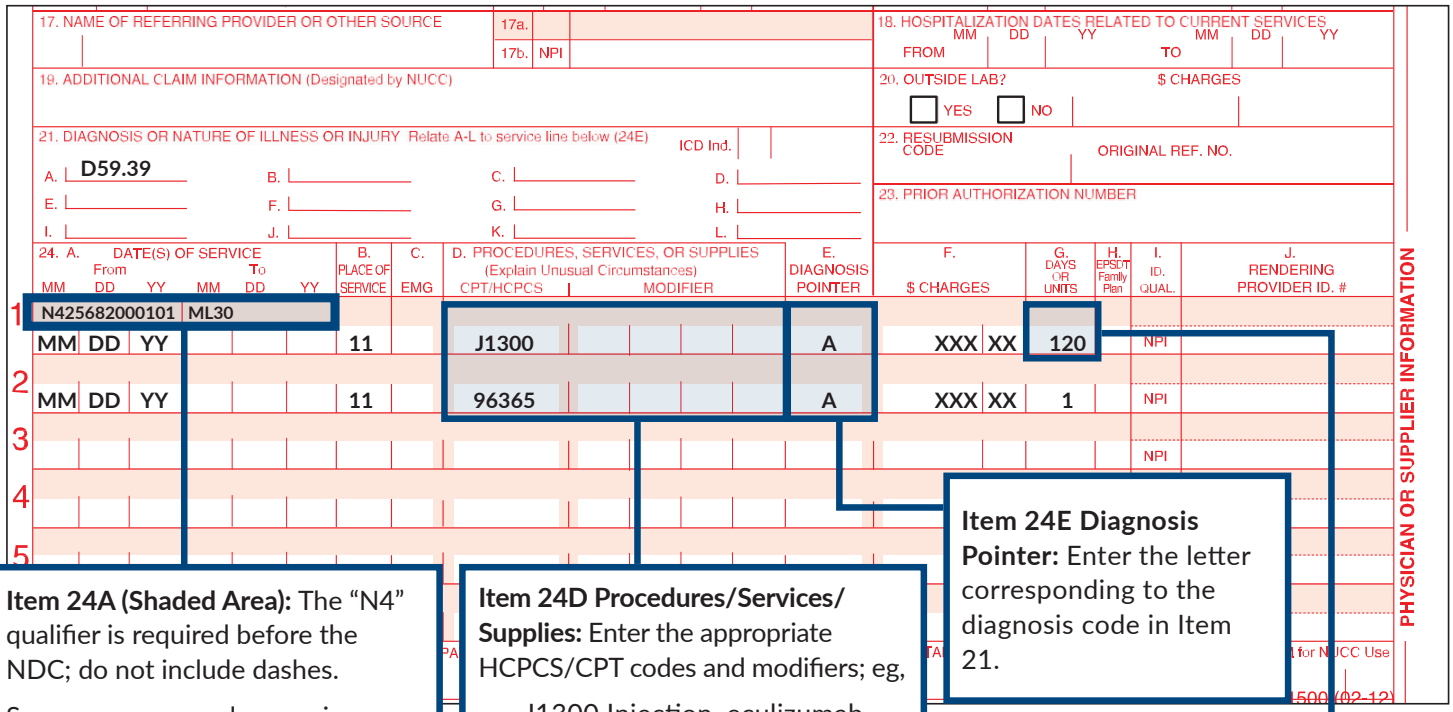
Please see additional Important Safety Information on pages 1 and 11-12 and the full Prescribing Information for SOLIRIS® (eculizumab), including Boxed WARNING regarding serious meningococcal infections.

Sample CMS-1500 (or the electronic equivalent 837P): Physician Office

Example claim form for a SOLIRIS® (eculizumab) IV infusion:

To achieve a SOLIRIS maintenance dose of 1200 mg for a patient 18 years of age and older, the following vials were used:

- 4 single-use 300 mg/30 mL vials (NDC 25682-0001-01)



Item 24A (Shaded Area): The "N4" qualifier is required before the NDC; do not include dashes.

Some payers may also require a Unit of Measure (UoM) for each NDC; eg,

- N425682000101 ML30

Note: Check payer requirements and format for reporting the NDC and UoM.

Item 24D Procedures/Services/ Supplies: Enter the appropriate HCPCS/CPT codes and modifiers; eg,

- J1300 Injection, eculizumab, 10 mg
- 96365 Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); initial, up to 1 hour

Item 24E Diagnosis Pointer: Enter the letter corresponding to the diagnosis code in Item 21.

Item 24G Days or Units: Enter the appropriate number of units for J1300 per 10 mg. For example, enter 120 billing units for 1200 mg.

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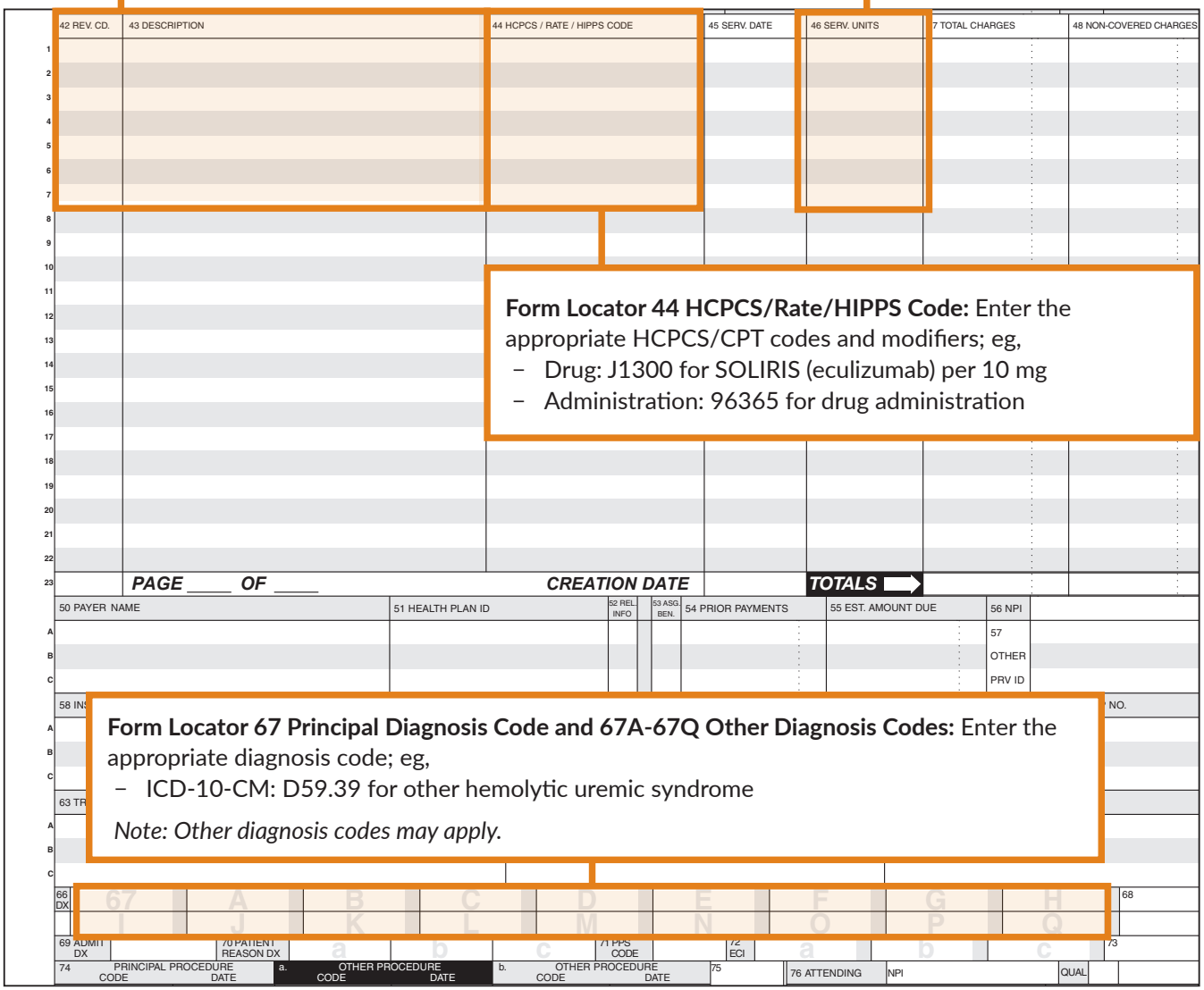
Sample CMS-1450 (UB-04) (or the electronic equivalent 837I): Hospital Outpatient Clinic or Facility

For an example of a completed CMS-1450 form, go to page 9.

Form Locator 42-43 Revenue Code and Description: Enter the appropriate revenue code and description corresponding to the HCPCS code in field 44; eg,
- 0636 for drugs requiring detailed coding
- 0510 for clinic, general

Note: Other revenue codes may apply.

Form Locator 46 Units of Service: Enter the appropriate number of units of service; eg, SOLIRIS 300 mg is reported with "30" units.



Form Locator 44 HCPCS/Rate/HIPPS Code: Enter the appropriate HCPCS/CPT codes and modifiers; eg,
- Drug: J1300 for SOLIRIS (eculizumab) per 10 mg
- Administration: 96365 for drug administration

Form Locator 67 Principal Diagnosis Code and 67A-67Q Other Diagnosis Codes: Enter the appropriate diagnosis code; eg,
- ICD-10-CM: D59.39 for other hemolytic uremic syndrome

Note: Other diagnosis codes may apply.

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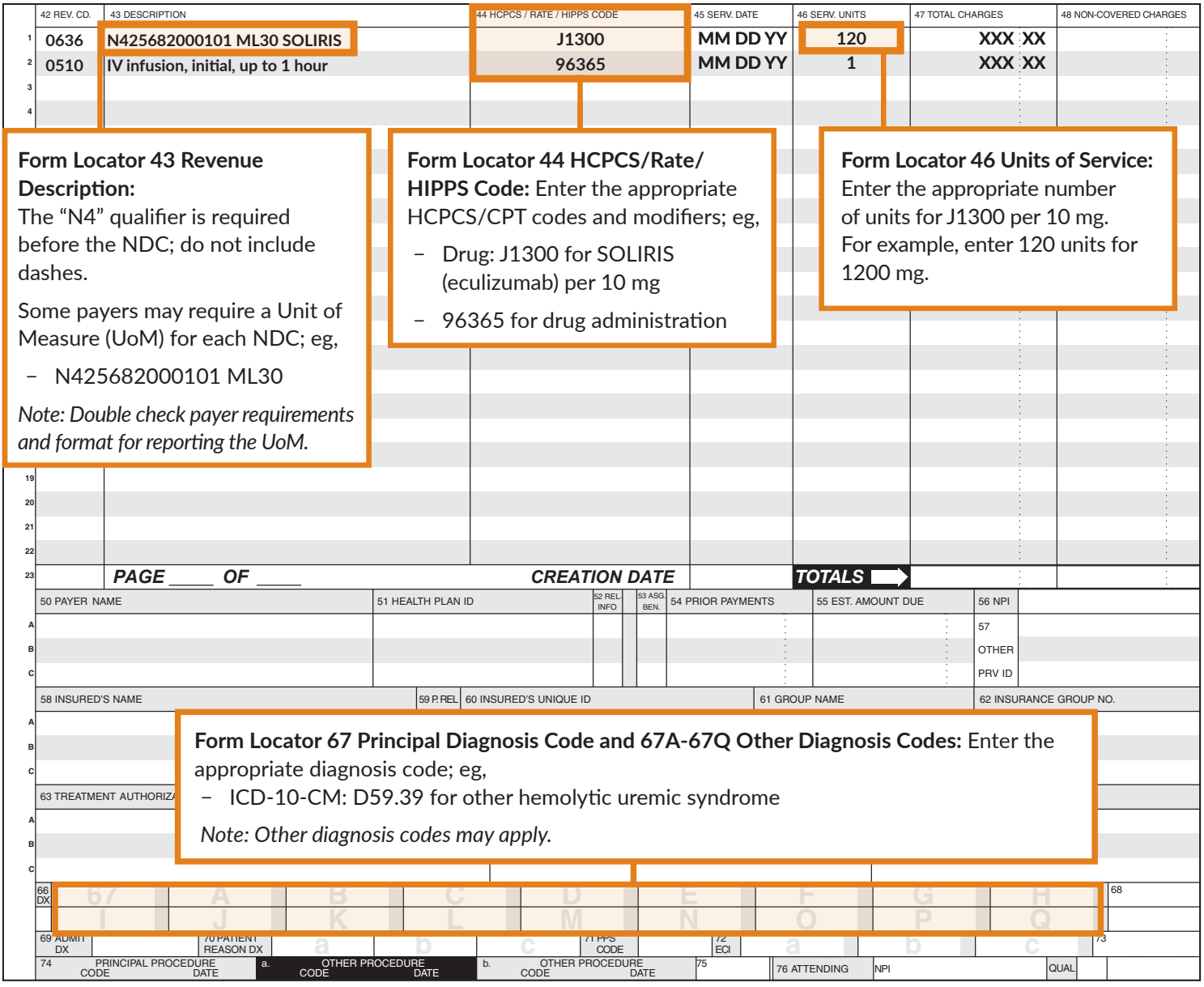
Sample CMS-1450 (UB-04) (or the electronic equivalent 837I): Hospital Outpatient Clinic or Facility

Example claim form for a SOLIRIS® (eculizumab) IV infusion:

To achieve a SOLIRIS maintenance dose of 1200 mg for a patient 18 years of age and older, the following vials were used:

- 4 single-use 300 mg/30 mL vials (NDC 25682-0001-01)



Form Locator 43 Revenue Description:
The "N4" qualifier is required before the NDC; do not include dashes.

Some payers may require a Unit of Measure (UoM) for each NDC; eg,

- N425682000101 ML30

Note: Double check payer requirements and format for reporting the UoM.

Form Locator 44 HCPCS/Rate/HIPPS Code: Enter the appropriate HCPCS/CPT codes and modifiers; eg,

- Drug: J1300 for SOLIRIS (eculizumab) per 10 mg
- 96365 for drug administration

Form Locator 46 Units of Service: Enter the appropriate number of units for J1300 per 10 mg. For example, enter 120 units for 1200 mg.

Form Locator 67 Principal Diagnosis Code and 67A-67Q Other Diagnosis Codes: Enter the appropriate diagnosis code; eg,

- ICD-10-CM: D59.39 for other hemolytic uremic syndrome

Note: Other diagnosis codes may apply.

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Explore Alexion Access Navigator

ALEXION ACCESS NAVIGATOR

Alexion Access Navigator is a dedicated resource website for US Healthcare Professionals and their offices that contains downloadable access and reimbursement materials for SOLIRIS® (eculizumab).

Online: https://alexionaccessnavigator.com

OneSource™ Offers Patient Support

Contact OneSource™

Phone:
1-888-765-4747

Online:
https://alexiononesource.com/

References

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2. Centers for Medicare & Medicaid Services. January 2022 alpha numeric HCPCS file. Updated January 26, 2022. Accessed September 7, 2022. https://www.cms.gov/Medicare/Coding/HCPCSReleaseCodeSets/HCPCS-Quarterly-Update
3. SOLIRIS. Prescribing information. Alexion Pharmaceuticals, Inc.
4. Food and Drug Administration. Future format of the National Drug Code; public hearing; request for comments. *Fed Regist*. 2018;83(152):38666-38668. Accessed September 7, 2022. https://www.federalregister.gov/documents/2018/08/07/2018-16807/future-format-of-the-national-drug-code-public-hearing-request-for-comments
5. Centers for Medicare & Medicaid Services. 2023 ICD-10-PCS code tables and index. Updated May 26, 2022. Accessed September 7, 2022. https://www.cms.gov/medicare/icd-10/2023-icd-10-pcs
6. *2022 CPT Professional*. American Medical Association; 2021. CPT © 2021 American Medical Association. All rights reserved. CPT® is a registered trademark of the American Medical Association.

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SELECT IMPORTANT SAFETY INFORMATION (cont.)

Contraindications

- Patients with unresolved serious *Neisseria meningitidis* infection
- Patients who are not currently vaccinated against *Neisseria meningitidis*, unless the risks of delaying Soliris treatment outweigh the risks of developing a meningococcal infection

Warnings and Precautions

Serious Meningococcal Infections

Risk and Prevention

The use of Soliris increases a patient's susceptibility to serious meningococcal infections (septicemia and/or meningitis).

Vaccinate or revaccinate for meningococcal disease according to the most current ACIP recommendations for patients with complement deficiencies. Immunize patients without a history of meningococcal vaccination at least 2 weeks prior to receiving the first dose of Soliris. If Soliris must be initiated immediately in an unvaccinated patient, administer meningococcal vaccine(s) as soon as possible and provide 2 weeks of antibacterial drug prophylaxis. Discontinue Soliris in patients who are undergoing treatment for serious meningococcal infections.

REMS

Prescribers must counsel patients about the risk of meningococcal infection, provide the patients with the REMS educational materials, and ensure patients are vaccinated with meningococcal vaccine(s).

Other Infections

Serious infections with *Neisseria* species (other than *N. meningitidis*), including disseminated gonococcal infections, have been reported.

Patients may have increased susceptibility to infections, especially with encapsulated bacteria. Additionally, *Aspergillus* infections have occurred in immunocompromised and neutropenic patients. Children treated with Soliris may be at increased risk of developing serious infections due to *Streptococcus pneumoniae* and *Haemophilus influenzae* type b (Hib). Administer vaccinations for the prevention of *Streptococcus pneumoniae* and *Haemophilus influenzae* type b (Hib) infections according to ACIP guidelines. Use caution when administering Soliris to patients with any systemic infection.

Monitoring Disease Manifestations After Soliris Discontinuation

Treatment Discontinuation for aHUS

After discontinuing Soliris, monitor patients with aHUS for signs and symptoms of thrombotic microangiopathy (TMA) complications for at least 12 weeks. In aHUS clinical trials, 18 patients (5 in the prospective studies) discontinued Soliris treatment. TMA complications occurred following a missed dose in 5 patients, and Soliris was reinitiated in 4 of these 5 patients.

Clinical signs and symptoms of TMA include changes in mental status, seizures, angina, dyspnea, or thrombosis. In addition, the following changes in laboratory parameters may identify a TMA complication: occurrence of 2, or repeated measurement of any one of the following: a decrease in platelet count by 25% or more compared to baseline or the peak platelet count during Soliris treatment; an increase in serum creatinine by 25% or more compared to baseline or nadir during Soliris treatment; or, an increase in serum LDH by 25% or more over baseline or nadir during Soliris treatment.

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SELECT IMPORTANT SAFETY INFORMATION (cont.)

Warnings and Precautions (cont.)

Monitoring Disease Manifestations After Soliris Discontinuation (cont.)

Treatment Discontinuation for aHUS (cont.)

If TMA complications occur after Soliris discontinuation, consider reinstitution of Soliris treatment, plasma therapy [plasmapheresis, plasma exchange, or fresh frozen plasma infusion (PE/PI)], or appropriate organ-specific supportive measures.

Thrombosis Prevention and Management

The effect of withdrawal of anticoagulant therapy during Soliris treatment has not been established. Therefore, treatment with Soliris should not alter anticoagulant management.

Infusion-Related Reactions

Administration of Soliris may result in infusion-related reactions, including anaphylaxis or other hypersensitivity reactions. Interrupt Soliris infusion and institute appropriate supportive measures if signs of cardiovascular instability or respiratory compromise occur.

Adverse Reactions

The most frequently reported adverse reactions in aHUS single arm prospective trials (≥20%) are: headache, diarrhea, hypertension, upper respiratory infection, abdominal pain, vomiting, nasopharyngitis, anemia, cough, peripheral edema, nausea, urinary tract infections, pyrexia.

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ALEXION®
AstraZeneca Rare Disease