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

In Atypical Hemolytic Uremic Syndrome (Atypical-HUS)

INDICATION

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

SOLIRIS, a complement inhibitor, increases the risk of serious infections caused by Neisseria meningitidis [*see Warnings and Precautions (5.1)*]. Life-threatening and fatal meningococcal infections have occurred in patients treated with complement inhibitors. These infections may become rapidly life-threatening or fatal if not recognized and treated early.

- **Complete or update vaccination for meningococcal bacteria (for serogroups A, C, W, Y, and B) at least 2 weeks prior to the first dose of SOLIRIS, unless the risks of delaying SOLIRIS therapy outweigh the risk of developing a serious infection. Comply with the most current Advisory Committee on Immunization Practices (ACIP) recommendations for vaccinations against meningococcal bacteria in patients receiving a complement inhibitor. *See Warnings and Precautions (5.1)* for additional guidance on the management of the risk of serious infections caused by meningococcal bacteria.**
- **Patients receiving SOLIRIS are at increased risk for invasive disease caused by *Neisseria meningitidis*, even if they develop antibodies following vaccination. Monitor patients for early signs and symptoms of serious meningococcal infections and evaluate immediately if infection is suspected.**

Because of the risk of serious meningococcal infections, SOLIRIS is available only through a restricted program under a Risk Evaluation and Mitigation Strategy (REMS) called ULTOMIRIS and SOLIRIS REMS [*see Warnings and Precautions (5.2)*].

Please see additional Important Safety Information on pages 1 and 11-12 and accompanying full Prescribing Information for SOLIRIS, including Boxed WARNING regarding serious and life-threatening or fatal meningococcal infections.

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

US/SOL-a/0046 V4 04/2024

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 accompanying full Prescribing Information for SOLIRIS, including Boxed WARNING regarding serious and life-threatening or fatal meningococcal infections.

Coding for SOLIRIS® (eculizumab) in Atypical-HUS

Diagnosis Coding

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)

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.

Drug Coding

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

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

Please see additional Important Safety Information on pages 1 and 11-12 and accompanying full Prescribing Information for SOLIRIS, including Boxed WARNING regarding serious and life-threatening or fatal meningococcal infections.

The following HCPCS modifiers may be required for SOLIRIS® (eculizumab), as applicable:

Modifier[2]	Description	Commercial Requirement	Medicare Requirement
JZ	Zero drug amount discarded/not administered to any patient	Varies by payer	Y
JG	Drug or biological acquired with 340B drug pricing program discount, reported for informational purposes	N	Y
RE	Furnished in full compliance with FDA-mandated risk evaluation and mitigation strategy (REMS)	Y	Y
TB	Drug or biological acquired with 340B drug pricing program discount, reported for informational purposes for select entities	N	Y

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[a]	Chemotherapy administration, intravenous infusion technique, up to one hour, single or initial substance/drug
+ 96415[a]	Chemotherapy administration, intravenous infusion technique; each additional hour (list separately in addition to primary procedure)

a. Billing highly complex administration codes (96413 and 96415) requires the provider in the medical record to document the complexity beyond what is required for therapeutic infusions (96365 and 96366).[7]

Please see additional Important Safety Information on pages 1 and 11-12 and accompanying full Prescribing Information for SOLIRIS, including Boxed WARNING regarding serious and life-threatening or fatal meningococcal infections.

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[1]	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
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
90749	Unlisted vaccine/toxoid

Vaccine Administration Coding

CPT Code[6]	Code Descriptor
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)

Please see additional Important Safety Information on pages 1 and 11-12 and accompanying full Prescribing Information for SOLIRIS, including Boxed WARNING regarding serious and life-threatening or fatal meningococcal infections.

Claim Forms

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

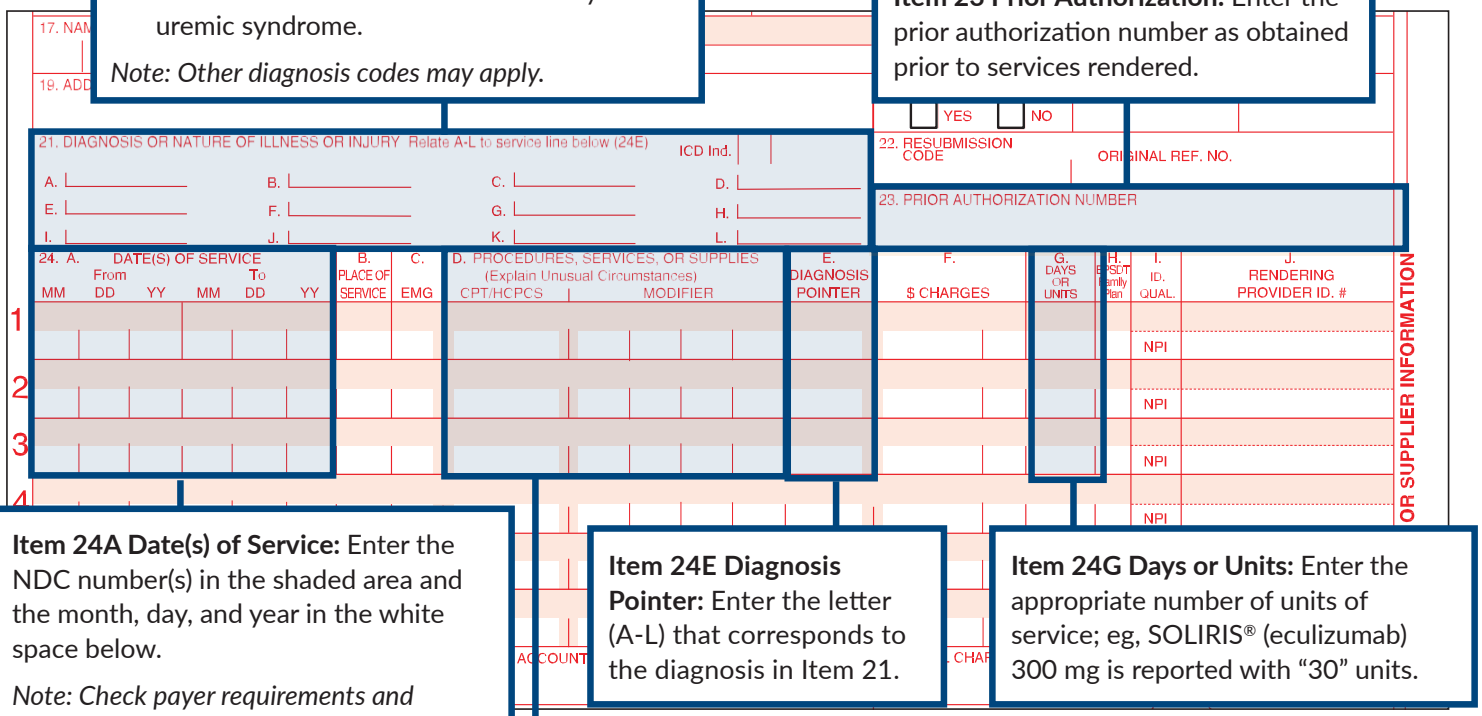
For an example of a completed CMS-1500 form, go to page 7.

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 CPT/HCPCS codes and modifiers, eg,

- Drug: **J1300** Injection, eculizumab, 10 mg
- Applicable modifiers:
  - **JZ** Zero drug amount discarded/not administered to any patient
  - **RE** Furnished in full compliance with FDA-mandated risk evaluation and mitigation strategy (REMS)
- Administration: **96365** for IV infusion

Note: Some payers may provide specific guidance.

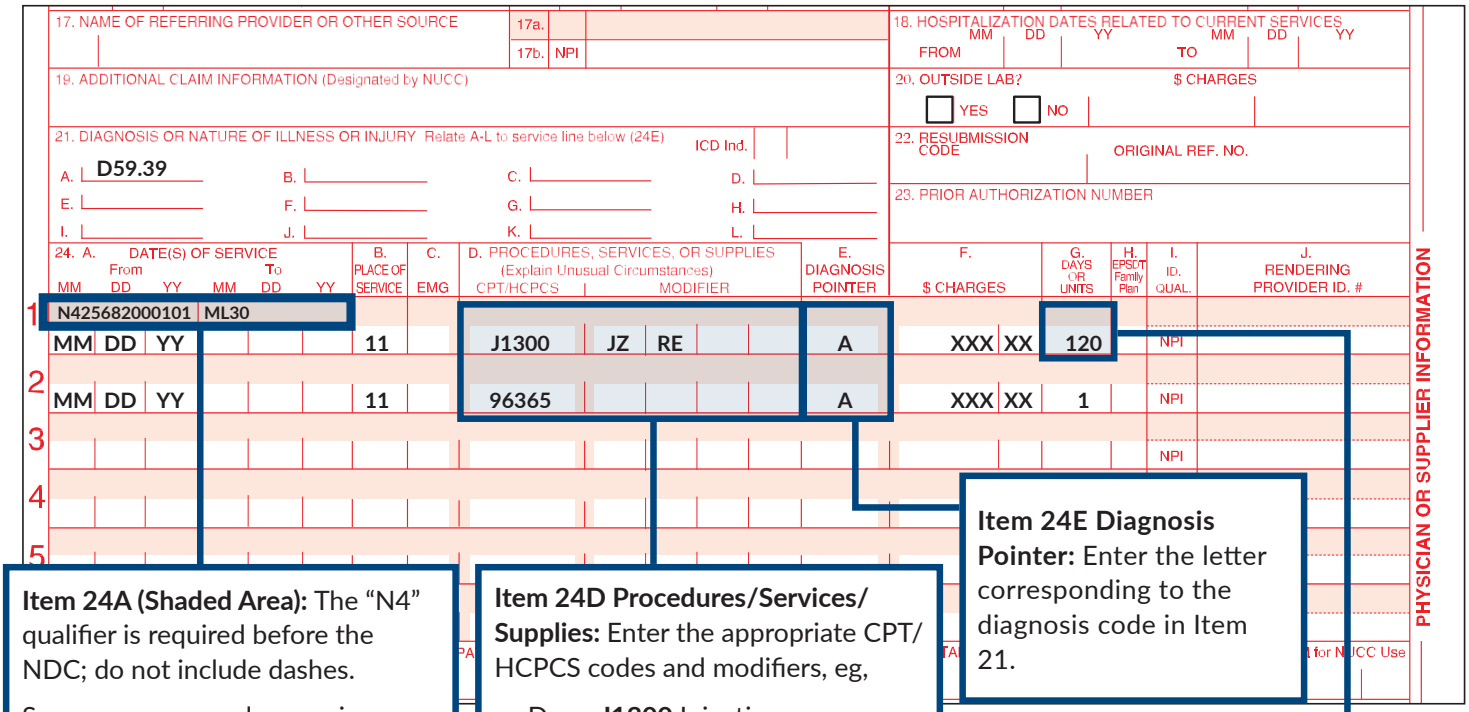
Please see additional Important Safety Information on pages 1 and 11-12 and accompanying full Prescribing Information for SOLIRIS, including Boxed WARNING regarding serious and life-threatening or fatal meningococcal infections.

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 CPT/ HCPCS codes and modifiers, eg,

- Drug: **J1300** Injection, eculizumab, 10 mg
- Applicable modifiers:
  - **JZ** Zero drug amount discarded/not administered to any patient
  - **RE** Furnished in full compliance with FDA-mandated risk evaluation and mitigation strategy (REMS)
- Administration: **96365** for IV infusion

Note: Some payers may provide specific guidance.

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.

Please see additional Important Safety Information on pages 1 and 11-12 and accompanying full Prescribing Information for SOLIRIS, including Boxed WARNING regarding serious and life-threatening or fatal meningococcal infections.

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

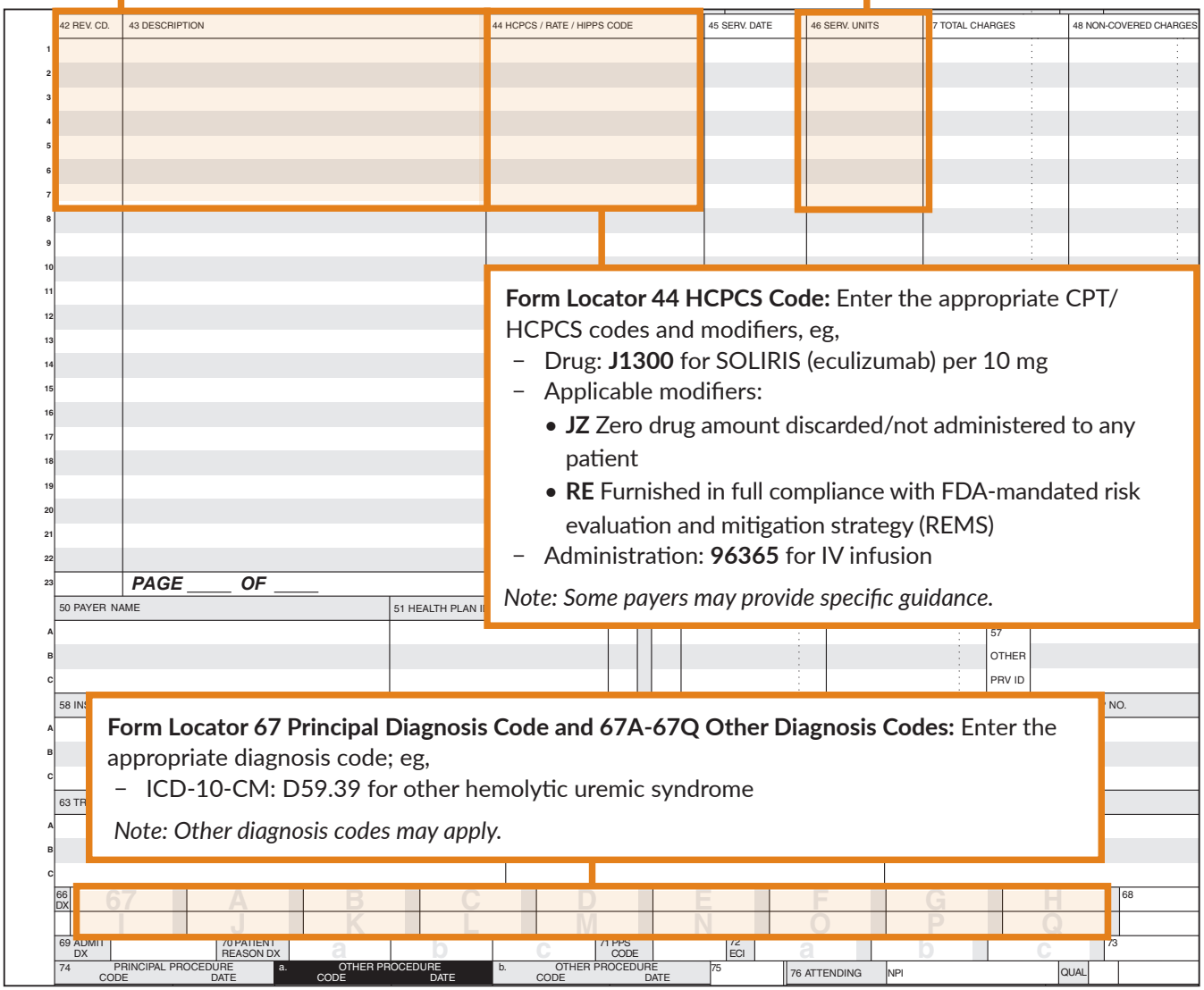
For an example of a completed CMS-1450 form, go to page 9.

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® (eculizumab) 300 mg is reported with "30" units.



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

- Drug: **J1300** for SOLIRIS (eculizumab) per 10 mg
- Applicable modifiers:
  - **JZ** Zero drug amount discarded/not administered to any patient
  - **RE** Furnished in full compliance with FDA-mandated risk evaluation and mitigation strategy (REMS)
- Administration: **96365** for IV infusion

Note: Some payers may provide specific guidance.

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:

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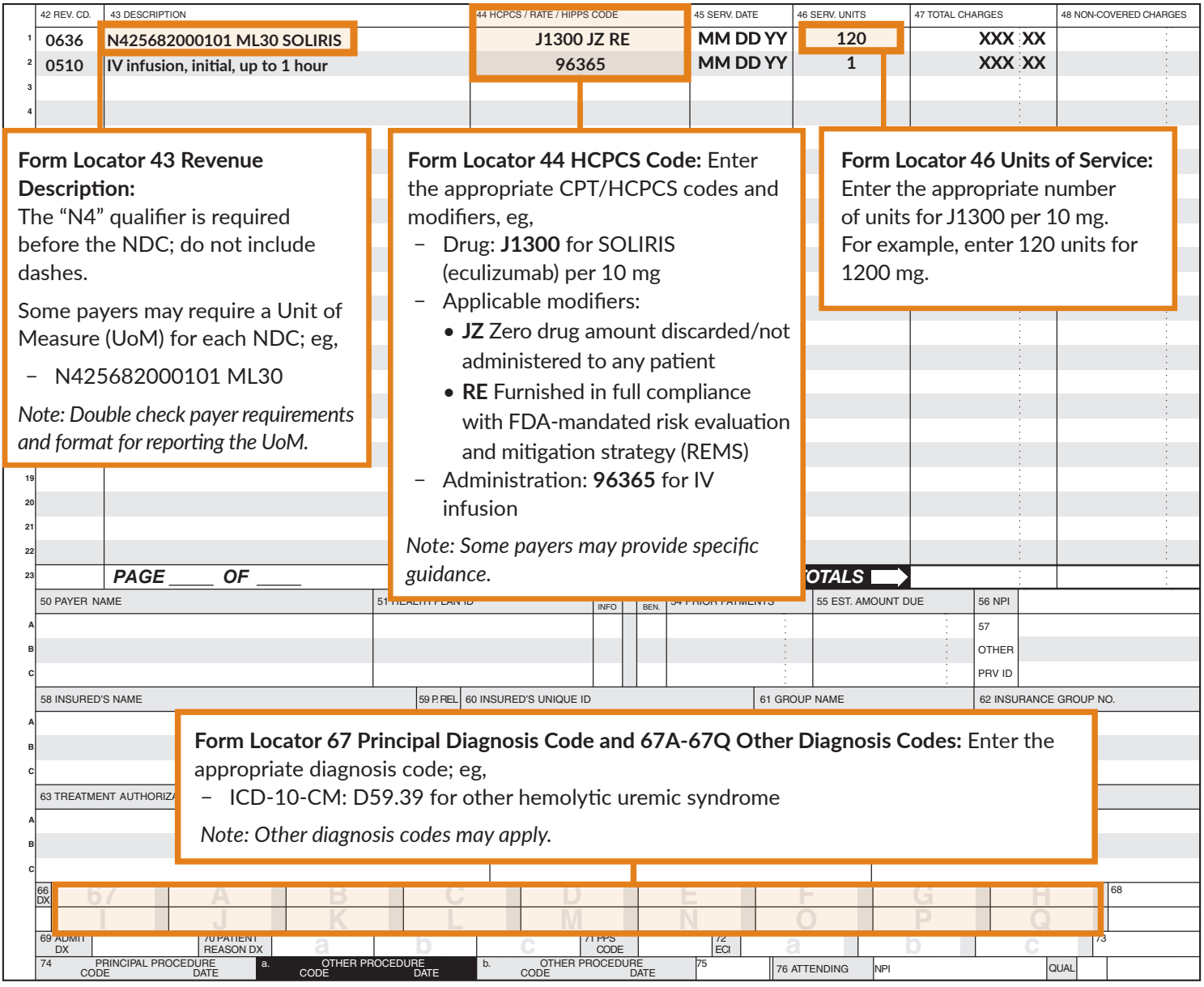

	42 REV. CD.	43 DESCRIPTION	44 HCPCS / RATE / HIPPS CODE	45 SERV. DATE	46 SERV. UNITS	47 TOTAL CHARGES	48 NON-COVERED CHARGES
1	0636	N425682000101 ML30 SOLIRIS	J1300 JZ RE	MM DD YY	120	XXX XX	
2	0510	IV infusion, initial, up to 1 hour	96365	MM DD YY	1	XXX XX	
3							
4							

Please see additional Important Safety Information on pages 1 and 11-12 and accompanying full Prescribing Information for SOLIRIS, including Boxed WARNING regarding serious and life-threatening or fatal meningococcal infections.

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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5. Centers for Medicare & Medicaid Services. 2024 ICD-10-PCS Conversion Table. Updated December 19, 2023. Accessed May 5, 2024. https://www.cms.gov/medicare/coding-billing/icd-10-codes/2024-icd-10-pcs
6. American Medical Association. *CPT 2024 Professional Edition*. AMA; 2023. All rights reserved. CPT® is a registered trademark of the American Medical Association.
7. Centers for Medicare & Medicaid Services. Billing and coding: complex drug administration coding (A58527). November 26, 2020. Updated April 1, 2024. Accessed May 10, 2024. https://www.cms.gov/medicare-coverage-database/details/article-details.aspx?articleId=58532&Cntrctr=365&ContrVer=1&CntrctrSelected=365*1&DocType=Active

Please see additional Important Safety Information on pages 1 and 11-12 and accompanying full Prescribing Information for SOLIRIS, including Boxed WARNING regarding serious and life-threatening or fatal meningococcal infections.

SELECT IMPORTANT SAFETY INFORMATION (cont.)

CONTRAINDICATIONS

- SOLIRIS is contraindicated for initiation in patients with unresolved serious *Neisseria meningitidis* infection.

WARNINGS AND PRECAUTIONS

Serious Meningococcal Infections

SOLIRIS, a complement inhibitor, increases a patient's susceptibility to serious, life-threatening, or fatal infections caused by meningococcal bacteria (septicemia and/or meningitis) in any serogroup, including non-groupable strains. Life-threatening and fatal meningococcal infections have occurred in both vaccinated and unvaccinated patients treated with complement inhibitors.

Revaccinate patients in accordance with ACIP recommendations considering the duration of therapy with SOLIRIS. Note that ACIP recommends an administration schedule in patients receiving complement inhibitors that differs from the administration schedule in the vaccine prescribing information.

If urgent SOLIRIS therapy is indicated in a patient who is not up to date with meningococcal vaccines according to ACIP recommendations, provide the patient with antibacterial drug prophylaxis and administer meningococcal vaccines as soon as possible. Various durations and regimens of antibacterial drug prophylaxis have been considered, but the optimal durations and drug regimens for prophylaxis and their efficacy have not been studied in unvaccinated or vaccinated patients receiving complement inhibitors, including SOLIRIS. The benefits and risks of treatment with SOLIRIS, as well as those associated with antibacterial drug prophylaxis in unvaccinated or vaccinated patients, must be considered against the known risks for serious infections caused by *Neisseria meningitidis*.

Vaccination does not eliminate the risk of serious meningococcal infections, despite development of antibodies following vaccination.

Closely monitor patients for early signs and symptoms of meningococcal infection and evaluate patients immediately if infection is suspected. Inform patients of these signs and symptoms and instruct patients to seek immediate medical care if these signs and symptoms occur. Promptly treat known infections. Meningococcal infection may become rapidly life-threatening or fatal if not recognized and treated early. Consider interruption of SOLIRIS in patients who are undergoing treatment for serious meningococcal infection, depending on the risks of interrupting treatment in the disease being treated.

ULTOMIRIS and SOLIRIS REMS

Due to the risk of serious meningococcal infections, SOLIRIS is available only through a restricted program called ULTOMIRIS and SOLIRIS REMS.

Prescribers must enroll in the REMS, counsel patients about the risk of serious meningococcal infection, provide patients with REMS educational materials, assess patient vaccination status for meningococcal vaccines (against serogroups A, C, W, Y, and B) and vaccinate if needed according to current ACIP recommendations two weeks prior to the first dose of SOLIRIS. Antibacterial drug prophylaxis must be prescribed if treatment must be started urgently and the patient is not up to date with both meningococcal vaccines according to current ACIP recommendations at least two weeks prior to the first dose of SOLIRIS. Patients must receive counseling about the need to receive meningococcal vaccines and to take antibiotics as directed, the signs and symptoms of meningococcal infection, and be instructed to carry the Patient Safety Card with them at all times during and for 3 months following SOLIRIS treatment.

Further information is available at www.UltSolREMS.com or 1-888-765-4747.

Other Infections

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

SOLIRIS blocks terminal complement activation; therefore, patients may have increased susceptibility to infections, especially with encapsulated bacteria, such as infections with *Neisseria meningitidis* but also *Streptococcus pneumoniae*, *Haemophilus influenzae*, and to a lesser extent, *Neisseria gonorrhoeae*. 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 recommendations. Patients receiving SOLIRIS are at increased risk for infections due to these organisms, even if they develop antibodies following vaccination.

Please see additional Important Safety Information on pages 1 and 11-12 and accompanying full Prescribing Information for SOLIRIS, including Boxed WARNING regarding serious and life-threatening or fatal meningococcal infections.

SELECT IMPORTANT SAFETY INFORMATION (cont.)

WARNINGS AND PRECAUTIONS (cont.)

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.

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. In clinical trials, no patients experienced an infusion-related reaction which required discontinuation of SOLIRIS. 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 the aHUS single arm prospective trials (≥20%) were: headache, diarrhea, hypertension, upper respiratory infection, abdominal pain, vomiting, nasopharyngitis, anemia, cough, peripheral edema, nausea, urinary tract infections, pyrexia.

To report SUSPECTED ADVERSE REACTIONS contact Alexion Pharmaceuticals, Inc. at 1-844-259-6783 or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.

Please see accompanying full prescribing information for SOLIRIS, including Boxed WARNING regarding serious and life-threatening or fatal meningococcal infections.

ALEXION®
AstraZeneca Rare Disease