RECORDATI RARE DISEASES
Focused on the Few®

Enjaymo®
sutimlimab-jome
Injection for intravenous use
1100 mg/22 mL

ENJAYMO Billing and Coding Guide for Reimbursement

Please see Important Safety Information on pages 3-4, as well as full Prescribing Information.

USING THIS BILLING AND CODING GUIDE

The codes referenced in this document may be used to communicate services rendered when filing claims for ENJAYMO.

These codes are being provided for informational purposes only and should be verified, as codes may change.

The provision of billing codes does not constitute reimbursement or legal advice. Providers are solely responsible for ensuring the accuracy of billing submissions to any payer.

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INDICATION AND IMPORTANT SAFETY INFORMATION

INDICATION

ENJAYMO® (sutimlimab-jome) is indicated for the treatment of hemolysis in adults with cold agglutinin disease (CAD).

IMPORTANT SAFETY INFORMATION

CONTRAINDICATIONS

ENJAYMO is contraindicated in patients with known hypersensitivity to sutimlimab-jome or any of the inactive ingredients.

WARNINGS AND PRECAUTIONS

Serious Infections Including Those Caused by Encapsulated Bacteria

- ENJAYMO, a proximal classical complement C1s inhibitor, increases susceptibility to serious infections, including those caused by encapsulated bacteria, eg, *Neisseria meningitidis* (any serogroup, including non-groupable strains), *Streptococcus pneumoniae*, and *Haemophilus influenzae* type B.
- Life-threatening and fatal infections with encapsulated bacteria have occurred in both vaccinated and unvaccinated patients treated with complement inhibitors.
- Serious infections (bacterial and viral) were reported in 15% (10/66) of patients receiving ENJAYMO in the two phase 3 trials. These infections included urinary tract infection with sepsis, respiratory tract infection, pneumonia, otomastoiditis, and skin infections. One patient (1.5%) died due to *Klebsiella pneumoniae*.
- Complete or update vaccination against encapsulated bacteria at least 2 weeks prior to administration of the first dose of ENJAYMO, according to the most current ACIP recommendations for patients receiving a complement inhibitor.
- If urgent ENJAYMO therapy is indicated in a patient who is not up to date on their vaccine(s), administer as soon as possible.
- Vaccination does not eliminate the risk of serious encapsulated bacterial infections. Closely monitor patients for early signs and symptoms of serious infection and evaluate patients immediately if an infection is suspected.
- If ENJAYMO treatment is administered to patients with active systemic infections, monitor closely for signs and symptoms of worsening infection. Some infections may become rapidly life-threatening or fatal if not recognized and treated promptly. Inform patients of these signs and symptoms and steps to be taken to seek immediate medical care.
    - Consider interruption of ENJAYMO treatment in patients who are undergoing treatment for serious infection.
    - Consider patients' immune status when initiating treatment with ENJAYMO.

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IMPORTANT SAFETY INFORMATION (cont'd)

WARNINGS AND PRECAUTIONS (cont'd)

Infusion-Related Reactions

- Administration of ENJAYMO may result in infusion-related reactions. In the two phase 3 trials, 29% (19/66) of patients treated with ENJAYMO experienced infusion-related reactions. One patient permanently discontinued ENJAYMO due to an infusion-related reaction.
- Monitor patients for infusion-related reactions and interrupt if a reaction occurs.
- Discontinue ENJAYMO infusion and institute appropriate supportive measures if signs of hypersensitivity reactions, such as cardiovascular instability or respiratory compromise, occur.

Risk of Autoimmune Disease

- Based on its mechanism of action, ENJAYMO may potentially increase the risk for developing autoimmune diseases such as systemic lupus erythematosus (SLE). Development of SLE has been associated with inherited classical complement deficiency.
- In clinical trials, 4.5% (3/66) of patients developed a relapse or worsening of previously diagnosed autoimmune disease.
- Monitor ENJAYMO patients for signs and symptoms and manage medically.

Recurrent Hemolysis After ENJAYMO Discontinuation

- If treatment with ENJAYMO is interrupted, closely monitor patients for signs and symptoms of recurrent hemolysis, eg, elevated levels of total bilirubin or lactate dehydrogenase (LDH) accompanied by a decrease in hemoglobin, or reappearance of symptoms such as fatigue, dyspnea, palpitations, or hemoglobinuria. Consider restarting ENJAYMO if signs and symptoms of hemolysis occur after discontinuation.

ADVERSE REACTIONS

- The most common adverse reactions in the CADENZA trial (Part A) (incidence ≥18%) are rhinitis, headache, hypertension, acrocyanosis, and Raynaud's phenomenon. The most common adverse reactions in the CARDINAL trial (incidence ≥25%) are urinary tract infection, respiratory tract infection, bacterial infection, dizziness, fatigue, peripheral edema, arthralgia, cough, hypertension, and nausea.

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CODING SUMMARY

Diagnosis

The ICD-10-CM diagnosis code for cold autoimmune hemolytic anemia (CAD) is D59.12.[1]

ICD-10-CM Code	
D55-D59	Hemolytic anemias
D59	Acquired hemolytic anemias
D59.1	Other autoimmune hemolytic anemias
D59.12	Cold autoimmune hemolytic anemia*

*ICD-10 code D59.12 can also be referred to as chronic cold hemagglutinin disease, cold agglutinin disease, cold agglutinin hemoglobinuria, cold type (primary) (secondary) (symptomatic) autoimmune hemolytic anemia, and cold type autoimmune hemolytic disease.

National Drug Code (NDC)

ENJAYMO has a 10-digit NDC code displayed on its packaging. In most cases, this should be converted to an 11-digit NDC code for billing purposes.[2] Below are both NDC codes for ENJAYMO.

10-digit NDC	NDC 80203-347-01
11-digit NDC	NDC 80203-0347-01
How supplied	One 1100 mg/22 mL (50 mg/mL) single dose vial per carton[3]

Payer requirements for NDC use and format may vary. Please contact each payer for specific coding policies.

ICD-10-CM, International Classification of Diseases, Tenth Revision, Clinical Modification.

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CPT® Codes

CPT® codes are used to describe the procedures that may be performed on a patient and/or how a drug or supply being billed was administered.[4] CPT codes commonly associated with the administration of IV-infused monoclonal antibodies like ENJAYMO are listed below. Confirm preferred coding policy with payer prior to administration whenever possible.

Primary	
96365	Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); initial; up to 1 hour
96366	Each additional hour (list separately in addition to code for primary procedure; report 96366 for infusion intervals of greater than 30 minutes beyond 1-hour increments)*

*Per CMS guidelines, if the incremental amount of infusion time is 30 minutes or less, the time is not to be billed separately. Note that some payers may require reporting the actual number of minutes on the claim.

HCPCS Procedure Codes

HCPCS codes are assigned by CMS and are used by Medicare and most private payers to describe products administered in a physician's office or hospital setting.[5] As part of the standardized Level II HCPCS coding system, ENJAYMO has been issued a permanent J code, which went into effect on October 1, 2022, and is listed below. All claims made as of October 1 should utilize the permanent J code to facilitate reimbursement.

Note that utilization of the permanent J code does not impact the ability of ENJAYMO to maintain transitional pass-through payment status under the Medicare Hospital Outpatient Prospective Payment System (OPPS). As always, the coding system is not a methodology for making coverage or payment determinations. The existence of a HCPCS code does not imply coverage; it implies only that the product may be reimbursed if covered. Coding should be verified with each patient's health plan prior to submitting a claim.

Permanent J Code	Description
J1302	Intravenous injection, sutimlimab-jome, 10 mg

The permanent J code replaces all temporary codes and should be used for both inpatient and outpatient billing as of October 1, 2022.

JW modifier: Providers and suppliers are required to report the JW modifier on Part B drug claims for discarded drugs and biologicals. Also, providers and suppliers must document the amount of discarded drugs or biologicals in Medicare beneficiaries' medical records.[6]

Place of Service Codes

Because ENJAYMO can be administered in various settings (infusion center, physician office, patient's home if deemed clinically appropriate by the prescribing physician), it is important to populate a claim with the appropriate 2-digit place-of-service (POS) code.[7] Always verify the preferred POS codes for your patient's health plan before submitting a claim.

CMS, Centers for Medicare & Medicaid Services; CPT®, Current Procedural Terminology; HCPCS, Healthcare Common Procedure Code System; IV, intravenous.

IMPORTANT SAFETY INFORMATION (cont'd)

WARNINGS AND PRECAUTIONS

Serious Infections Including Those Caused by Encapsulated Bacteria

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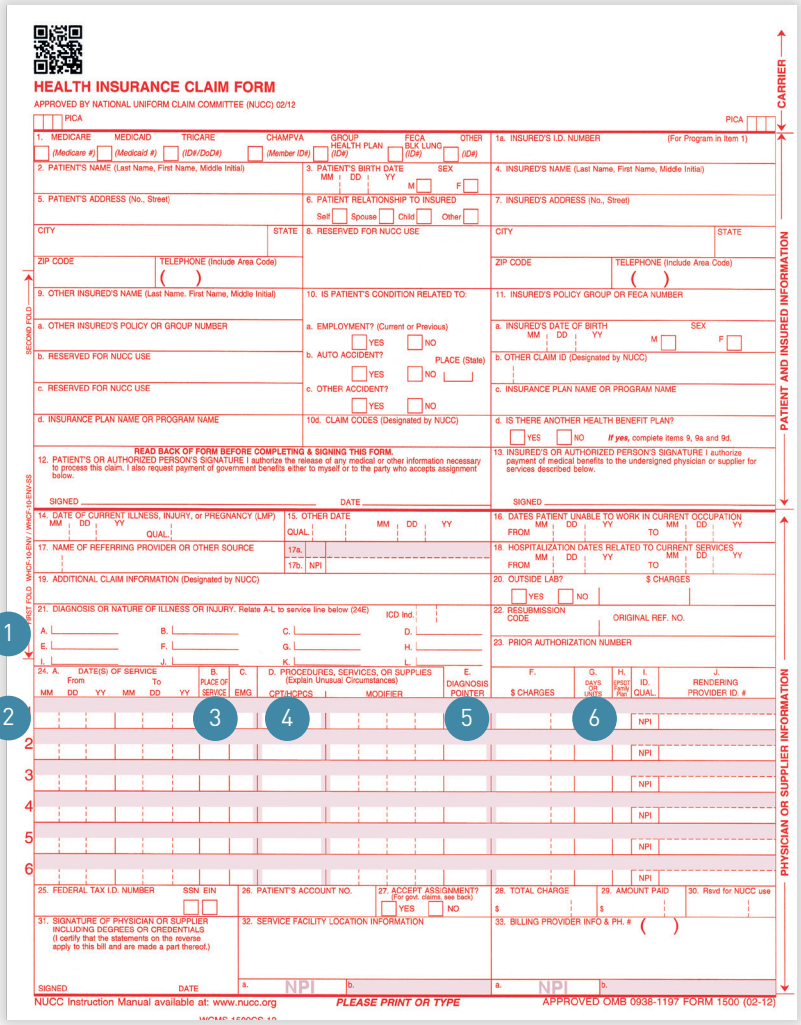
SAMPLE CMS REIMBURSEMENT FORMS[8,9]

These sample claim forms are intended for use only as a reference. Reimbursement codes are subject to continual change. Please confirm the accuracy of the codes you use to bill for the prescribed medications with each payer.

1. **ITEM 21:** Enter the appropriate ICD-10-CM diagnosis codes
2. **ITEM 24A:** Enter the date of service for each procedure. Include NDC information, if required, in the shaded areas above each date
3. **ITEM 24B:** Enter appropriate place of service code (office, infusion center, etc)
4. **ITEM 24D:** Include payer-required details such as HCPCS (J code), CPT codes and modifiers
5. **ITEM 24E:** Enter the diagnosis code reference letter or number from Box 21 that relates to the date of service and the services or procedures performed that are entered on that same line under 24D
6. **ITEM 24G:** Enter the billing units, using the conversion of 10 mg ENJAYMO = 1 billing unit



This sample form is for informational purposes only.

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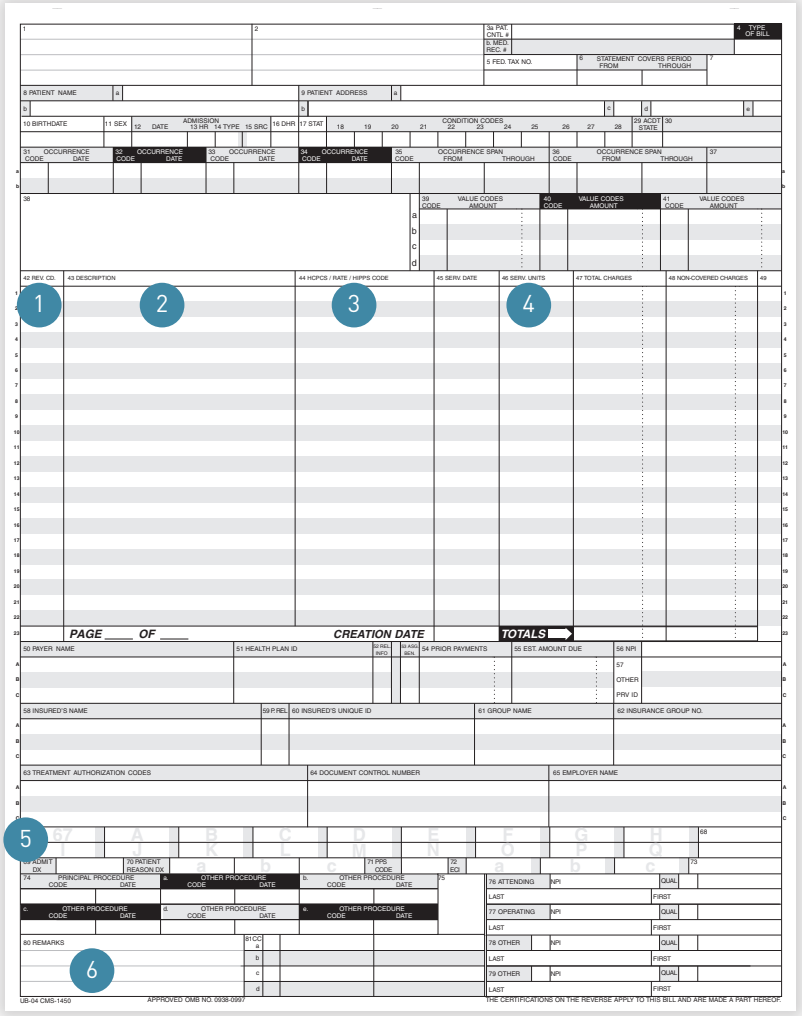
SAMPLE CMS REIMBURSEMENT FORMS (cont'd)

1. **Field 42:** Enter the 4-digit revenue code that best describes the service provided, in accordance with hospital billing policy
2. **Field 43:** Enter the corresponding description of service (eg, IV therapy)
3. **Field 44:** Include payer-required details such as relevant HCPCS and CPT codes
4. **Field 46:** Enter the billing units, using the conversion of 10 mg ENJAYMO = 1 billing unit
5. **Field 66:** Enter the appropriate ICD-10-CM diagnosis codes
6. **Field 80:** Provide any required detailed information such as drug name, total dosage and strength, method of administration, and 11-digit NDC (attach separately if needed)



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DOSING AND ADMINISTRATION OF ENJAYMO

ENJAYMO can be administered undiluted or diluted[3]

ENJAYMO is for intravenous infusion only and should be administered weekly for the first 2 weeks of treatment and every other week thereafter. The recommended dosage of ENJAYMO is based on body weight.

Infusion rates for undiluted ENJAYMO

Body-Weight Range	Dose (mg)	Number of ENJAYMO Vials Needed	ENJAYMO Volume	Maximum Infusion Rate
≥39 kg to <75 kg	6500	6	130 mL	130 mL/hour*
≥75 kg	7500	7	150 mL	150 mL/hour*

*Patients with cardiopulmonary disease should receive the infusion over 120 minutes.

Infusion rates for diluted ENJAYMO

Body-Weight Range	Dose (mg)	Number of ENJAYMO Vials Needed	ENJAYMO Volume	Volume of NaCl Diluent	Total Volume	Maximum Infusion Rate
39 kg to <70 kg	6500	6	130 mL	370 mL	500 mL	250 mL/hour
70 kg to <75 kg	6500	6	130 mL	370 mL	500 mL	500 mL/hour*
≥75 kg	7500	7	150 mL	350 mL	500 mL	500 mL/hour*

*Patients with cardiopulmonary disease should receive the infusion over 120 minutes.

Vaccination note: Complete or update vaccination against encapsulated bacteria at least 2 weeks prior to administration of the first dose of ENJAYMO, according to the most current ACIP recommendations for patients receiving a complement inhibitor.

Once removed from refrigeration, allow the ENJAYMO infusion solution to adjust to room temperature (59°F to 77°F). In-line infusion warmers may be used.

For initial ENJAYMO infusion, monitor the patient for at least 2 hours following completion for signs or symptoms of an infusion and/or hypersensitivity reaction. Monitor the patient for one hour following completion of subsequent infusions for signs or symptoms of an infusion reaction.

IMPORTANT SAFETY INFORMATION (cont'd)

WARNINGS AND PRECAUTIONS (cont'd)

Serious Infections Including Those Caused by Encapsulated Bacteria (cont'd)

- Life-threatening and fatal infections with encapsulated bacteria have occurred in both vaccinated and unvaccinated patients treated with complement inhibitors.

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ADDITIONAL BILLING AND CODING CONSIDERATIONS

Reimbursement considerations

ENJAYMO is designed to be prepared and administered by a health care provider. The drug costs are expected to be covered under the Medicare Part B benefit. Please refer to the individual patient's plan to determine any applicable coverage requirements. The specifics of coverage may vary by payer.

When filing a claim

It is recommended that ENJAYMO coverage be confirmed with all payers prior to patient administration, as patient benefits vary among payers and by plans.

Some payers also have policies that may affect coverage for ENJAYMO. These include:

- Site of care: Some payers may have coverage rules that restrict certain providers from delivering infusion therapies.
- Network providers: Some payers have exclusive contracts with in-network or participating providers to provide infusion therapies. These may include contracts for coverage in physician offices and outpatient settings or to specialty pharmacies that provide drugs and biologics to the provider.
- Prior authorization: Many plans may require providers to obtain prior authorization (eg, medical necessity) to begin a course of treatment. Check with the payer to determine their process, requirements, and method for requesting authorization.

Documenting necessity

As a new medication used to treat a rare disease, some insurers may not be familiar with ENJAYMO, and may require additional documentation to process either a prior authorization or a claim upon receipt. Examples of documentation that can be required include:

- Statement of medical necessity from the attending physician
- ENJAYMO Prescribing Information available at **www.ENJAYMOhcp.com**
- Documentation of ENJAYMO FDA approval
- Details on the patient's case history, previous therapy, and clinical course

FDA, Food and Drug Administration.

IMPORTANT SAFETY INFORMATION (cont'd)

WARNINGS AND PRECAUTIONS (cont'd)

Serious Infections Including Those Caused by Encapsulated Bacteria (cont'd)

- Serious infections (bacterial and viral) were reported in 15% (10/66) of patients receiving ENJAYMO in the two phase 3 trials. These infections included urinary tract infection with sepsis, respiratory tract infection, pneumonia, otomastoiditis, and skin infections. One patient (1.5%) died due to *Klebsiella pneumoniae*.
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SUPPORT SERVICES

ENJAYMO Patient Solutions supports patients from the start and throughout treatment

PATIENT EDUCATION SERVICES

Patient Education Services may be able to provide patients with education services, reimbursement services, and related material.

FINANCIAL ASSISTANCE PROGRAMS

ENJAYMO Financial Assistance Programs may be able to help with the cost of treatment. Access to ENJAYMO at no cost may be available to eligible patients who are uninsured or underinsured. Co-pay assistance may be available for out-of-pocket co-pay or coinsurance costs related to ENJAYMO prescriptions or infusion costs.*

For more information on ENJAYMO Patient Solutions, and to download the ENJAYMO Patient Enrollment Form, visit **www.ENJAYMOhcp.com**

*For co-pay or coinsurance program, not valid for ENJAYMO prescriptions covered by or submitted for reimbursement under Medicare, Medicaid, VA, DoD, TRICARE® or similar federal or state programs including any state pharmaceutical assistance programs. Not valid where prohibited by law. Savings may vary depending on patient's out-of-pocket costs. Upon registration, patient will receive all program details.

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ORDERING INFORMATION

To order ENJAYMO:

ENJAYMO is available through the following authorized specialty distributors:

ASD Healthcare:
800-746-6273; asdhealthcare.com

Besse Medical:
800-543-2111; besse.com

Cardinal Health Specialty Distribution:
877-453-3972; specialtyonline.cardinalhealth.com

McKesson Specialty (MSCD):
800-482-6700; oncology.mckessonspecialtyhealth.com

McKesson Plasma & Biologics (MPB):
877-625-2566; connect.mckesson.com

Oncology Supply:
800-633-7555; oncologysupply.com

ENJAYMO is also available through **CVS Specialty Pharmacy:**
844-287-1298; cvsspecialty.com

REFERENCES

1. Centers for Disease Control and Prevention. ICD-10-CM tabular list of diseases and injuries. Accessed September 12, 2022. https://ftp.cdc.gov/pub/Health_Statistics/NCHS/Publications/ICD10CM/2022/icd10cm-tabular-2022-April-1.pdf#:~:text=ICD-10-CM%20TABULAR%20LIST%20of%20DISEASES%20and%20INJURIES%20
2. Blue Cross Blue Shield of Illinois. National Drug Code (NDC) Billing Guidelines. February 2019.
3. ENJAYMO. Prescribing Information.
4. Lee K. Current Procedural Terminology (CPT) code. SearchHealthIT. 2020. Accessed September 9, 2022. https://searchhealthit.techtarget.com/definition/Current-Procedural-Terminology-CPT
5. American Academy of Professional Coders. What is HCPCS? Reviewed October 2019. Accessed April 5, 2022. https://www.aapc.com/resources/medical-coding/hcpcs.aspx
6. Centers for Medicare & Medicaid Services. Medicare Program JW Modifier: Drug/Biological Amount Discarded/Not Administered To Any Patient Frequently Asked Questions. August 2016.
7. Centers for Medicare & Medicaid Services. Place of Service Codes for Professional Claims. Updated September 2021. Accessed September 6, 2022. https://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/PhysicianFeeSched/Downloads/Website-POS-database.pdf
8. Centers for Medicare & Medicaid Services. Health Insurance Claim Form. OMB-0938-1197 FORM 1500. February 2012. Accessed September 12, 2022. https://www.cms.gov/Medicare/CMS-Forms/CMS-Forms/Downloads/CMS1500.pdf
9. Centers for Medicare & Medicaid Services. Sample Form CMS-1450. Accessed September 9, 2022. https://www.cms.gov/Regulations-and-Guidance/Legislation/PaperworkReductionActof1995/PRA-Listing-Items/CMS-1450

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ENJAYMO: the first and only approved treatment for hemolysis in adults with cold agglutinin disease (CAD)

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