Pombiliti® (cipaglucosidase alfa-atga) + Opfolda® (miglustat) 65 mg capsules

BILLING & CODING GUIDE

INDICATION

POMBILITI in combination with OPFOLDA is indicated for the treatment of adult patients with late-onset Pompe disease (lysosomal acid alpha-glucosidase [GAA] deficiency) weighing ≥40 kg and who are not improving on their current enzyme replacement therapy (ERT).

WARNING: WARNING: SEVERE HYPERSENSITIVITY REACTIONS, INFUSION-ASSOCIATED REACTIONS, AND RISK OF ACUTE CARDIORESPIRATORY FAILURE IN SUSCEPTIBLE PATIENTS

See full prescribing information for complete boxed warning

HYPERSENSITIVITY REACTIONS INCLUDING ANAPHYLAXIS

Appropriate medical support measures, including cardiopulmonary resuscitation equipment, should be readily available. If a severe hypersensitivity reaction occurs, POMBILITI should be discontinued immediately and appropriate medical treatment should be initiated.

INFUSION-ASSOCIATED REACTIONS (IARS)

If severe IARs occur, immediately discontinue POMBILITI and initiate appropriate medical treatment.

RISK OF ACUTE CARDIORESPIRATORY FAILURE IN SUSCEPTIBLE PATIENTS

Patients susceptible to fluid volume overload, or those with acute underlying respiratory illness or compromised cardiac or respiratory function, may be at risk of serious exacerbation of their cardiac or respiratory status during POMBILITI infusion.

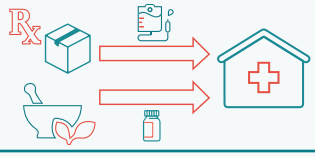
Please see IMPORTANT SAFETY INFORMATION on pages 2–6 and full Prescribing Information, including BOXED WARNING, for POMBILITI and full Prescribing Information for OPFOLDA, also available at PombilitiOpfoldaHCP.com.

Amicus Therapeutics

Amicus has developed this reference guide to assist providers with understanding coding for POMBILITI and OPFOLDA for the approved indications. This guide is provided for informational purposes only. Use of this guide does not guarantee coverage or reimbursement. This information is not intended to substitute for the prescriber's independent medical judgment, and providers are solely responsible for ensuring the accuracy of claims, invoices and documentation submitted to payers. The information in this guide is subject to change and should not be construed as legal advice. Providers should verify all questions, coding and special billing requirements with the payer prior to submission.

POMBILITI and OPFOLDA Acquisition Options

POMBILITI is administered in combination with OPFOLDA. Because POMBILITI is administered intravenously and OPFOLDA is an oral capsule, the process of acquiring and coding for each product could vary based on payer and specific patient benefit plan. Acquisition scenarios include:

SCENARIO A	SCENARIO B	SCENARIO C
		
Provider/facility buys and bills for both POMBILITI and OPFOLDA	Specialty pharmacy dispenses/ships both POMBILITI and OPFOLDA to the provider/facility or patient for administration and the provider only bills the POMBILITI administration fee	Provider/facility buys and bills for POMBILITI and its administration with the specialty pharmacy dispensing/shipping OPFOLDA to the provider/facility or patient

= Specialty Distributor = Specialty pharmacy = Provider/facility = POMBILITI = OPFOLDA

HCPCS Codes[13,18]	Descriptions
POMBILITI: J1203	Injection, cipaglucosidase alfa-atga, 5 mg
OPFOLDA: J1202†	Miglustat, oral, 65mg
G-CODE: G0138‡	Intravenous infusion of cipaglucosidase alfa-atga, including provider/supplier acquisition and clinical supervision of oral administration of miglustat in preparation of receipt of cipaglucosidase alfa-atga. G-codes are national codes assigned by CMS to identify professional healthcare procedures and services that may not have assigned CPT® codes. Not all payers are obligated to adopt this code. Please note Medicare Administrative Contractors (MACs) may require use of G0138 with quantity of OPFOLDA capsules noted in Box 19 (or electronic equivalent). Please confirm with individual MAC for specific billing requirements.

†Please check with payer prior to use of J-Code for OPFOLDA.
‡G-Codes are primarily used for Medicare encounters. For all other payers, the provider/facility should clarify and confirm coding requirements before using the G-Code. CMS assigned G0138 to New Technology APC 1508 [New Technology - Level 8 with a Status Indicator (SI) of S indicating a separate APC payment under Hospital Outpatient Services].[15] Note that G0138 should be billed as 1 unit as it is inclusive of multi-hour infusion of POMBILITI and up to four OPFOLDA capsules.
HCPCS=Healthcare Common Procedure Coding System; CMS=Centers for Medicare & Medicaid Services; CPT® =Current Procedural Terminology. CPT® is a registered trademark of the American Medical Association, 2023.

SELECT IMPORTANT SAFETY INFORMATION

WARNING: SEVERE HYPERSENSITIVITY REACTIONS, INFUSION-ASSOCIATED REACTIONS, and RISK OF ACUTE CARDIORESPIRATORY FAILURE IN SUSCEPTIBLE PATIENTS

HYPERSENSITIVITY REACTIONS INCLUDING ANAPHYLAXIS

Patients treated with POMBILITI have experienced life-threatening hypersensitivity reactions, including anaphylaxis. Appropriate medical support measures, including cardiopulmonary resuscitation equipment, should be readily available during POMBILITI administration. If a severe hypersensitivity reaction (e.g., anaphylaxis) occurs, POMBILITI should be discontinued immediately, and appropriate medical treatment should be initiated. In patients with severe hypersensitivity reaction, desensitization measures to POMBILITI may be considered.

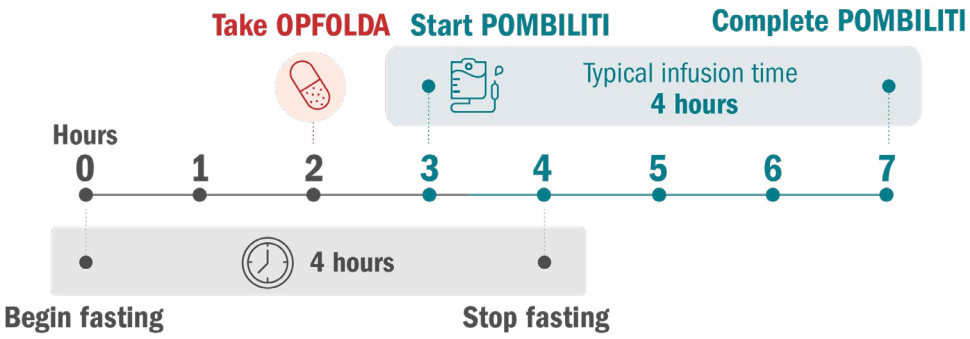
Please see IMPORTANT SAFETY INFORMATION on pages 2–6 and full Prescribing Information, including BOXED WARNING, for POMBILITI and full Prescribing Information for OPFOLDA, also available at PombilitiOpfoldaHCP.com.

Other Codes	Codes	Descriptions
ICD-10-CM (Diagnosis Code)[2]	E74.02	Pompe disease
CPT‡ (Procedure Code)[3]	96365	Intravenous infusion; therapy, prophylaxis, or diagnosis (specify substance or drug); initial, up to 1 hour
	+96366	Each additional hour (list separately in addition to primary procedure code)

J Code Billing Units[18]

POMBILITI		OPFOLDA	
J Code:	J1203 Injection, cipaglucosidase alfa-atga, 5 mg	**J-Code:**	J1202 Miglustat, oral, 65 mg
Billing Units:	5 mg = 1 unit	**Billing Units:**	1 (65) mg capsule = 1 unit
Example:	105 mg single-dose vial = 21 total units per vial	**Example:**	4 count OPFOLDA bottle = 4 units

Treatment Day Schedule[15,16]



SELECT IMPORTANT SAFETY INFORMATION (CONTINUED)

WARNING: SEVERE HYPERSENSITIVITY REACTIONS, INFUSION-ASSOCIATED REACTIONS, and RISK OF ACUTE CARDIORESPIRATORY FAILURE IN SUSCEPTIBLE PATIENTS

HYPERSENSITIVITY REACTIONS INCLUDING ANAPHYLAXIS (CONTINUED)

The risks and benefits of readministering POMBILITI following severe hypersensitivity reaction should be considered. If mild or moderate hypersensitivity reaction occurs, the infusion rate may be slowed or temporarily stopped. Prior to POMBILITI administration, consider pretreating with antihistamines, antipyretics, and/or corticosteroids.

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POMBILITI and OPFOLDA Coding Information (Continued)

POMBILITI (cipaglucosidase alfa-atga) for injection[15]	Carton NDC	Vial NDC	11-Digit NDC§
One (1) 105 mg single-dose vial	71904-200-01	71904-200-01	71904-**0**200-01
Ten (10) 105 mg single-dose vials	71904-200-02	71904-200-01	71904-**0**200-01
Twenty-five (25) 105 mg single-dose vials	71904-200-03	71904-200-01	71904-**0**200-01

OPFOLDA (miglustat) 65 mg capsules[16]	Bottle NDC	11-Digit NDC§
4 count bottle	71904-300-01	71904-**0**300-01
24 count bottle	71904-300-02	71904-**0**300-02
100 count bottle	71904-300-03	71904-**0**300-03

§NDC=National Drug Code. Payer requirements vary. This form is showing a "zero-filled" 11-digit code that meets Health Insurance Portability and Accountability Act (HIPAA) standards. The zero-fill location is indicated in bold.

SELECT IMPORTANT SAFETY INFORMATION (CONTINUED)

WARNING: SEVERE HYPERSENSITIVITY REACTIONS, INFUSION-ASSOCIATED REACTIONS, and RISK OF ACUTE CARDIORESPIRATORY FAILURE IN SUSCEPTIBLE PATIENTS

INFUSION-ASSOCIATED REACTIONS (IARS)

Patients treated with POMBILITI have experienced severe IARs. If severe IARs occur, immediately discontinue the POMBILITI infusion, initiate appropriate medical treatment, and assess the benefits and risks of readministering POMBILITI following severe IARs. If mild or moderate IARs occur regardless of pretreatment, decreasing the infusion rate or temporarily stopping the infusion may ameliorate the symptoms. IARs may still occur in patients after receiving pretreatment.

Patients with an acute underlying illness at the time of POMBILITI infusion may be at greater risk for IARs. Patients with advanced Pompe disease may have compromised cardiac and respiratory function, which may predispose them to a higher risk of severe complications from IARs.

RISK OF ACUTE CARDIORESPIRATORY FAILURE IN SUSCEPTIBLE PATIENTS

Patients susceptible to fluid volume overload, or those with acute underlying respiratory illness or compromised cardiac or respiratory function for whom fluid restriction is indicated may be at risk of serious exacerbation of their cardiac or respiratory status during POMBILITI infusion. More frequent monitoring of vitals should be performed during POMBILITI infusion in such patients.

CONTRAINDICATION

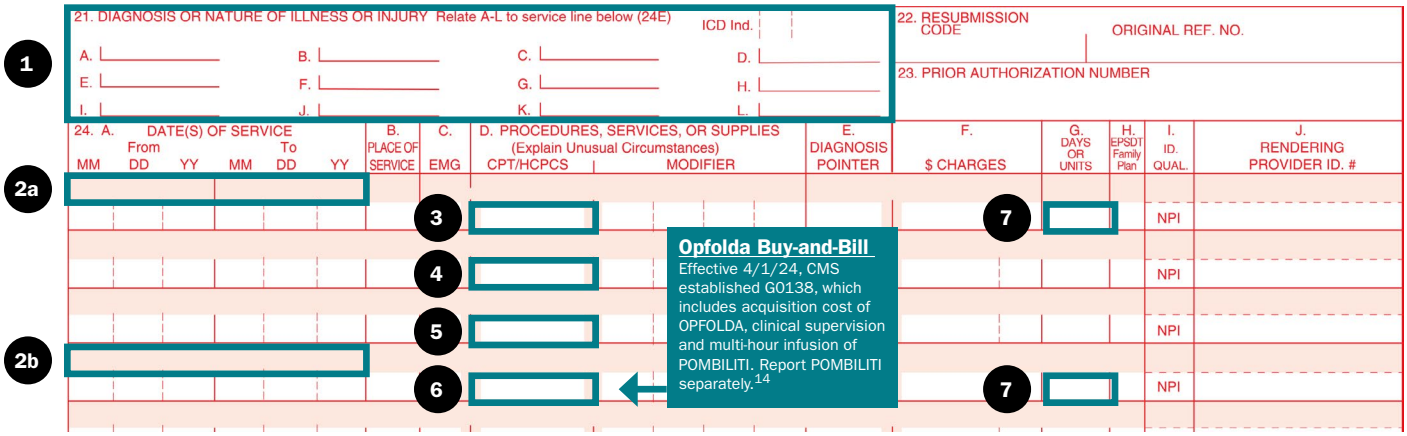
POMBILITI in combination with OPFOLDA is contraindicated in pregnancy.

EMBRYO-FETAL TOXICITY

Based on findings from animal reproduction studies, POMBILITI in combination with OPFOLDA may cause embryo-fetal harm when administered to a pregnant female and is contraindicated during pregnancy. Verify the pregnancy status in females of reproductive potential prior to initiating treatment with POMBILITI in combination with OPFOLDA. Advise females of reproductive potential to use effective contraception during treatment with POMBILITI in combination with OPFOLDA and for at least 60 days after the last dose.

SAMPLE CMS-1500 FORM[11]

This sample form is not intended to be directive and is for informational purposes only. Use of the recommended codes does not guarantee reimbursement. Providers may deem other codes or policies more appropriate and should select the coding options that most accurately reflect payer requirements and services rendered.



1. **Box 21:** Enter appropriate diagnosis code (for example, E74.02 for Pompe disease).[2]
2a. **Box 24A - POMBILITI:** In the shaded area above date of service, enter the NDC preceded by the N4 qualifier and followed by the appropriate abbreviation for the unit of measure (e.g., UN for unit[s]) and the number of units administered (for example, N471904020001UN1400).
2b. **24A - OPFOLDA:** In the shaded area above date of service, enter the NDC preceded by the N4 qualifier and followed by the appropriate abbreviation for the unit of measure (e.g., UN for unit[s]) and the number of units administered (for example, N471904030001). Note that an NDC is required only if a physician-administered drug is billed, so an NDC may not be required for the oral product.
3. **Box 24D – Line 1:** Enter appropriate drug HCPCS code for POMBILITI (for example, J1203.)[13] Note that certain payers may require two lines: one that reflects the amount of drug administered, and a separate line (POMBILITI HCPCS code with a JW-modifier) reflecting the amount of drug wasted. Append modifier JZ to indicate no wastage.[17] This requirement should be clarified by the payer.
4. **Box 24D – Line 2:** Enter CPT® code for intravenous infusion. For example: 96365 Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); initial, up to 1 hour.[3]
   Certain payers may require G0138 to report the infusion of POMBILITI when the practice/facility acquires OPFOLDA. Infusion codes 96365 and +96366 should not be reported when using HCPCS G0138.
5. **Box 24D – Line 3:** Enter CPT® code for each additional infusion hour. For example: +96366 Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); each additional hour (list separately in addition to code for primary procedure).[3]
   Certain payers may require G0138 to report the infusion of POMBILITI when the practice/facility acquires OPFOLDA. Infusion codes 96365 and +96366 should not be reported when using HCPCS G0138.
6. **Box 24D – Line 4:** If accepted by payer, enter oral drug HCPCS code, J1202.
7. **Box 24G – Lines 1 & 4:** Report appropriate number of units.

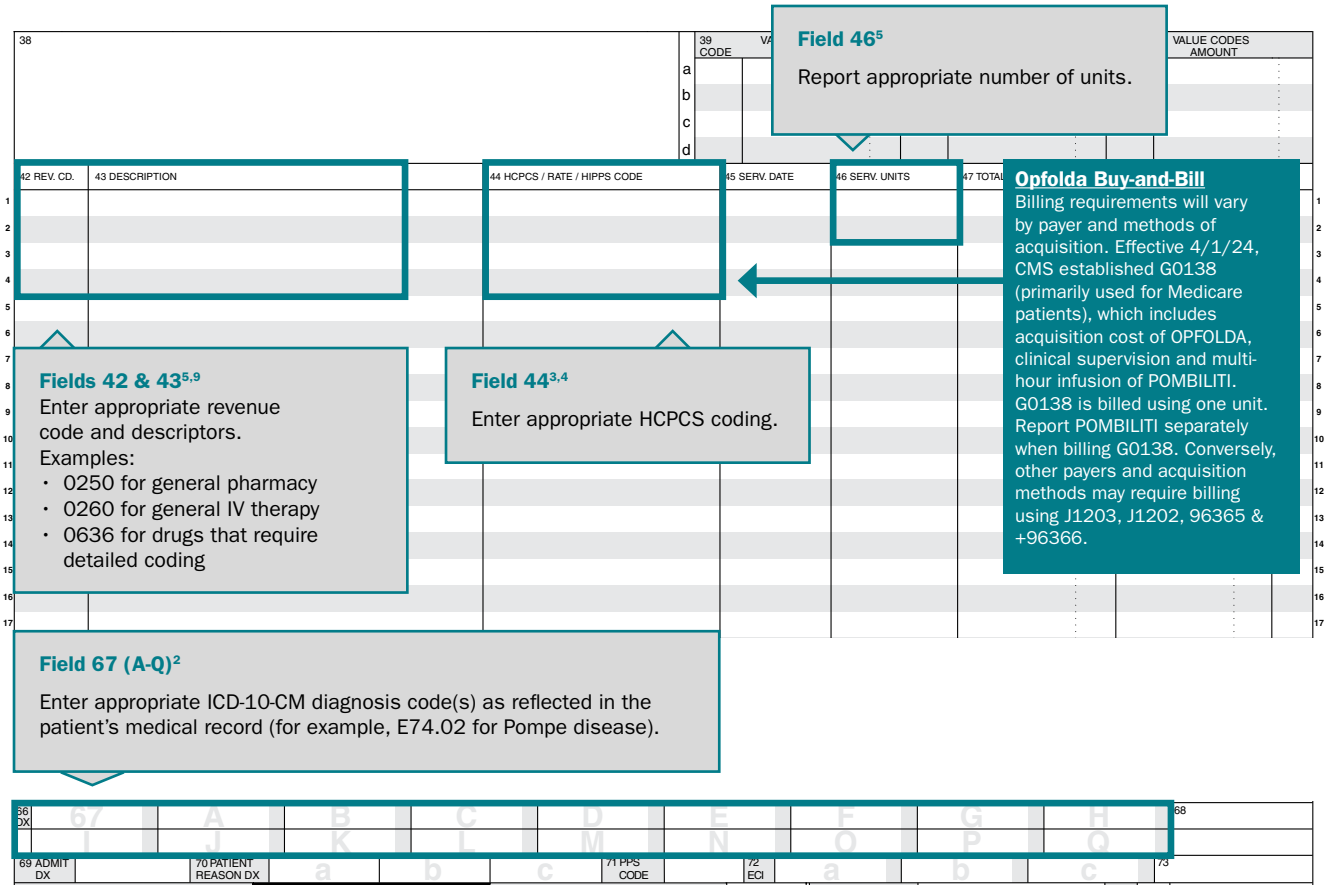
SELECT IMPORTANT SAFETY INFORMATION (CONTINUED)

RISKS ASSOCIATED WITH POMBILITI AND OPFOLDA

POMBILITI and OPFOLDA must be administered in combination.

SAMPLE CMS-1450 FORM[8]

This sample form is not intended to be directive and is for informational purposes only. Use of the recommended codes does not guarantee reimbursement. Providers may deem other codes or policies more appropriate and should select the coding options that most accurately reflect payer requirements and services rendered.



SELECT IMPORTANT SAFETY INFORMATION (CONTINUED)

ADVERSE REACTIONS

The most common adverse reactions (≥5%) reported in the pooled safety population of patients treated with POMBILITI in combination with OPFOLDA in the 3 clinical trials were headache, diarrhea, fatigue, nausea, abdominal pain, and pyrexia.

To report SUSPECTED ADVERSE REACTIONS, contact Amicus Therapeutics at 1-877-4AMICUS or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.

LACTATION

Advise females that breastfeeding is not recommended while on treatment with POMBILITI in combination with OPFOLDA.

REFERENCES

1. Centers for Medicare and Medicaid Services. Medicare Claims Processing Manual Chapter 25 - **Completing and Processing Form CMS-1450** Data Set. Accessed April 2024 at: https://www.cms.gov/Regulations-and-Guidance/Guidance/Manuals/Downloads/clm104c25.pdf
2. Centers for Disease Control and Prevention. **ICD-10-CM Diagnosis Codes**. National Center for Health Statistics – ICD-10-CM. Accessed April 2024 at: https://icd10cmtool.cdc.gov/?fy=FY2024
3. **CPT® 2024 Professional Edition**. Fourth ed. Chicago, IL: American Medical Association; 2023.
4. Centers for Medicare and Medicaid Services. **Medicare Claims Processing Manual** Chapter 17 - Drugs and Biologicals. Accessed April 2024. https://www.cms.gov/Regulations-and-Guidance/Guidance/Manuals/Downloads/clm104c17.pdf
5. Centers for Medicare and Medicaid Services. Palmetto Jurisdictions J and M – Part A. Local Coverage Article; **Billing and Coding: Hospital Outpatient Drugs and Biologicals Under the Outpatient Prospective Payment System (OPPS)** (A55913) Accessed April 2024. https://www.cms.gov/medicare-coverage-database/view/article.aspx?articleid=55913&ver=7&bc=0
6. Noridian Healthcare Solutions. Jurisdiction F – Medicare Part B. **Unlisted and Not Otherwise Classified Code Billing**. Accessed April 2024. https://med.noridianmedicare.com/web/jfb/topics/claim-submission/submission-errors-solutions/unlisted-procedure-and-noc-codes
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14. Centers for Medicare and Medicaid Services. April 2024 **Update of the Hospital Outpatient Prospective Payment System (OPPS).** Transmittal 12552. Accessed April 2024. https://www.cms.gov/files/document/r12552CP.pdf
15. Prescribing Information. **POMBILITI (cipaglucosidase alfa-atga)** for injection, for intravenous use. Accessed April 2024. https://amicusrx.com/pi/pombiliti.pdf
16. Prescribing Information. **OPFOLDA (miglustat)** capsules, for oral use. Accessed April 2024. https://amicusrx.com/pi/opfolda.pdf
17. Centers for Medicare and Medicaid Services. **Discarded Drugs and Biologicals** – JW Modifier and JZ Modifier Policy Frequently Asked Questions. Accessed April 2024. https://www.cms.gov/medicare/medicare-fee-for-service-payment/hospitaloutpatientpps/downloads/jw-modifier-faqs.pdf
18. Centers for Medicare & Medicaid Services. **Healthcare Common Procedure Coding System (HCPCS) Application Summaries and Coding Recommendations.** Fourth Quarter, 2023 HCPCS Coding Cycle. Access April 2024. https://www.cms.gov/files/document/2023-hcpcs-application-summary-quarter-4-2023-drugs-and-biologicals-updated-04/25/2024.pdf

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