

PLACEHOLDER

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 and 3 and see pocket for full Prescribing Information, including **BOXED WARNING**, for POMBILITI and full Prescribing Information for OPFOLDA, also available at PombilitiOpfoldaHCP.com.

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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 Coding Information

Product Codes

	POMBILITI HCPCS Code	Description		OPFOLDA HCPCS Code*	Description
In-office	J3490/J3590	Unclassified drug/ unclassified biologic	+	J8499*	Prescription drug, oral, non- chemotherapeutic, not otherwise specified
Hospital Outpatient	C9399	Unclassified drugs or biologicals (Medicare hospital outpatient setting only)		C9399*	Unclassified drugs or biologicals (Medicare hospital outpatient setting only)
*Please check with payer prior to use of J Code for OPFOLDA.					

Other Codes

	Codes	Description
ICD-10-CM (Diagnosis Code)	E74.02	Pompe disease
CPT† (Procedure Code)	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)

†CPT® – Current Procedural Terminology. CPT® is a registered trademark of the American Medical Association, 2019.

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. 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.

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.

Please see **IMPORTANT SAFETY INFORMATION** on pages 2 and 3 and see pocket for full Prescribing Information, including **BOXED WARNING**, for POMBILITI and full Prescribing Information for OPFOLDA, also available at PombilitiOpfoldaHCP.com.

POMBILITI and OPFOLDA Coding Information (Continued)

POMBILITI (cipaglucosidase alfa-atga) for injection	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	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

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.

RISKS ASSOCIATED WITH POMBILITI AND OPFOLDA

POMBILITI and OPFOLDA must be administered in combination.

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.

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SAMPLE CMS-1450 FORM⁸

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.




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Please see **IMPORTANT SAFETY INFORMATION** on pages 2 and 3 and see pocket for full Prescribing Information, including **BOXED WARNING**, for **POMBILITI** and full Prescribing Information for **OPFOLDA**, also available at PombilitiOpfoldaHCP.com.

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POMBILITI and OPFOLDA Coding Information (Continued)			
POMBILITI (cyclosporine) all-in-one injection	Curien CDC	Vital CDC	11-digit NDC
One (1) 100 mg single-dose vial	71904-2000-01	71904-2000-01	71904-0200-01
ten (10) 100 mg single-dose vials	71904-2002-02	71904-2002-02	71904-0200-02
Twenty (20) 100 mg single-dose vials	71904-2003-03	71904-2003-03	71904-0200-03
OPFOLDA (meprobamate) 650 mg capsules	Bottles NDC		11-digit NDC
4 count bottle	71904-3000-01	71904-0300-01	71904-0300-01
24 count bottle	71904-3002-02	71904-0302-02	71904-0300-02
100 count bottle	71904-3003-03	71904-0303-03	71904-0300-03
<small> Pombiliti is a registered trademark of Curien, LLC. NDC (National Drug Code) is a unique numerical code of 11 digits used for billing, National Healthcare Provider and Accountability, and Offsets collection. The product's location is indicated in boldface. </small>			
SELECTING APPROPRIATE SAFETY INFORMATION (CONTINUED) WARNING: SEVERE HYPERTENSIVE REACTIONS, HYPOKALEMIC ASSOCIATED REACTIONS, AND RISK OF ACUTE RENAL IMPAIRMENT IN SUSCEPTIBLE PATIENTS			
RISK OF ACUTE RENAL IMPAIRMENT IN SUSCEPTIBLE PATIENTS Patients susceptible to fluid volume overload, or those with acute underlying respiratory distress or compromised cardiac or respiratory function for whom fluid restriction is indicated may be at risk of acute exacerbation of their cardiac or respiratory condition during POMBILITI infusion. More frequent monitoring of status should be performed during POMBILITI infusions in such patients.			
CONTRAMICATION POMBILITI in combination with OPFOLDA is contraindicated in pregnancy.			
EMPHATIC TOLCICLY Based on findings from animal reproductive studies, POMBILITI in combination with OPFOLDA may cause embryofetal harm when administered to a pregnant female in combination with cyclosporine. Verify the pregnancy status is females of reproductive potential prior to initiating treatment with POMBILITI in combination with OPFOLDA. Advise females of reproductive potential of the effective contraindication during treatment with POMBILITI in combination with OPFOLDA for at least 90 days after the last dose.			
RISKS ASSOCIATED WITH POMBILITI AND OPFOLDA POMBILITI and OPFOLDA must be administered in combination.			
ADVERSE REACTIONS The most common adverse reactions (≥5%) reported in the pooled safety analysis of patients treated with POMBILITI in combination with OPFOLDA in the 13 clinical trials were headache, dizziness, fatigue, nausea, abdominal pain, and pruritus.			
FOR USPECTED ACUTE RENAL IMPAIRMENT, CONTACT AMICUS THERAPEUTICS AT 1-877-AMICUS-1 or 1-800-FDA-1088 or www.fda.gov/medwatch.			
LACTATION Amicus females that breastfeeding is not recommended while on treatment with POMBILITI in combination with OPFOLDA.			
Please see IMPORTANT SAFETY INFORMATION on pages 2 and 3 and see product label for Prescribing Information, including BLACK BOX WARNING, for POMBILITI and see Prescribing Information for OPFOLDA, also available at PombilitiAmicus.com.			
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NOT FOR EXTERNAL USE

2. DOSAGE AND ADMINISTRATION

2.2 Recommended Dosage and Administration

POMBELITI must be administered in combination with Opdiva (see Figure 1) for the dosing timeline. If the Opdiva dose is missed, POMBELITI should not be administered. Refer to the Opdiva Prescribing Information for Opdiva dosage and administration recommendations.

Prior to POMBELITI administration, consider pre-treating with anti-inflammatories, antipruritics and/or anti-emetics (see *Guidance and Recommendations* 3.1.2.2.1). If premedication was used with previous systemic rechallenge therapy (TRT), prior to POMBELITI administration, pre-treat with anti-inflammatories, anti-emetics, and/or anti-nausea agents.

The recommended dosage of POMBELITI is 20 mg/kg (of actual body weight) administered every other week as an intravenous infusion over approximately 4 hours.

Start POMBELITI in combination with Opdiva 2 weeks after the last TRT dose.

Initiate the POMBELITI infusion approximately 1 hour after oral administration of Opdiva. If the POMBELITI infusion cannot be started within 3 hours of oral administration of Opdiva, reschedule initiation of Opdiva and POMBELITI at least 24 hours after Opdiva was last taken. If Opdiva and POMBELITI are both missed, that treatment with Opdiva as soon as possible.

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Figure 1. Dosing Timeline

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Please see IMPORTANT FACT INFORMATION on pages 2 and 3 and see point for full Processing Information, including BOXED WARNING, for POMBALLI and N6 Processing Information for OPFOLDA, this includes all POMBALLI and OPFOLDA. Pomballi and Opfolda are registered trademarks of Amicus Therapeutics, Inc. © 2022 Amicus Therapeutics. All rights reserved. P016101-001-0002

6