

# 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 alphaglucosidase [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.





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.

Product Cod	les						
	POMBILITI HCPCS Code	Description			OPFOLDA HCPCS Code*	Description	
In-office	J3490/J3590		assified drug/ assified 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 p	ayer prior to use of J Code for OPFOLDA.	
Other Codes		Codes	Description				
ICD-10-CM (Diagnosis Code)		E74.02	Pompe disease				
CPT <sup>†</sup> (Procedure Code)		96365	Intravenous infusion; therapy, prophylaxis, or diagnosis (specify substance or drug); initial, up to 1 hour				
		+96366	Each additional	Each additional hour (list separately in addition to primary procedure code)			

#### **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.





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- <b>0</b> 200-01				
Ten (10) 105 mg single-dose vials	71904-200-02	71904-200-01	71904- <b>0</b> 200-01				
Twenty-five (25) 105 mg single-dose vials	71904-200-03	71904-200-01	71904- <b>0</b> 200-01				
OPFOLDA (miglustat) 65 mg capsules	В	11-Digit NDC‡					
4 count bottle	719	71904- <b>0</b> 300-01					
24 count bottle	719	71904- <b>0</b> 300-02					
100 count bottle	719	71904- <b>0</b> 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 <a href="https://www.fda.gov/medwatch">www.fda.gov/medwatch</a>.

#### **LACTATION**

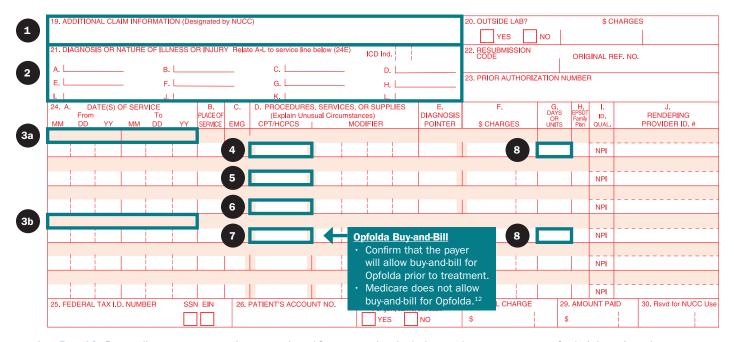
Advise females that breastfeeding is not recommended while on treatment with POMBILITI in combination with OPFOLDA.





# SAMPLE CMS-1500 FORM<sup>11</sup>

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.



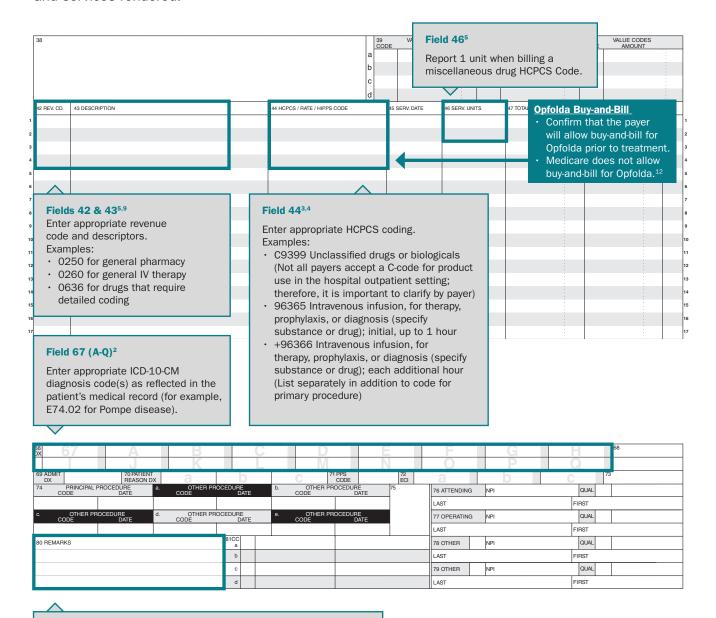
- 1. <u>Box 19:</u> Depending on payer requirements, box 19 may need to include: product name, route of administration, dose administered, amount wasted, and product NDCs (be sure to use vial NDC, not carton NDC).<sup>10</sup>
- 2. Box 21: Enter appropriate diagnosis code (for example, E74.02 for Pompe disease).<sup>2</sup>
- 3a. 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, N471904030001UN1400).
- **3b.** <u>24A OPFOLDA:</u> 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, N471904020001). 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.
- 4. <u>Box 24D Line 1:</u> Enter appropriate miscellaneous drug HCPCS code for Pombiliti. For example: J3490/J3590 (Note: check with payer for preferred code). Note that even when billing a miscellaneous HCPCS code, certain payers may require two lines: one that reflects the amount of drug administered, and a separate line (miscellaneous HCPCS code with a JW-modifier) reflecting the amount of drug wasted. This requirement should be clarified by the payer.
- 5. <u>Box 24D Line 2:</u> 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.<sup>3</sup>
- 6. <u>Box 24D Line 3:</u> 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).<sup>3</sup>
- 7. Box 24D Line 4: If accepted by payer, enter miscellaneous oral drug HCPCS code. For example: J8499 Prescription drug, oral, nonchemotherapeutic, NOS. (Note: Check with payer to ensure that they will allow providers to buy-and-bill Opfolda utilizing HCPCS prior to treating patient. Medicare does not allow buy-and-bill for Opfolda.)<sup>10,12</sup>
- 8. Box 24G Lines 1 & 4: Report 1 unit when billing a miscellaneous drug HCPCS Code.6





# SAMPLE CMS-1450 FORM<sup>8</sup>

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.



## Field 80<sup>4</sup>

Payer requirements vary. Examples might include drug name(s), NDC(s), route(s) of administration, dose(s) administered, wastage.

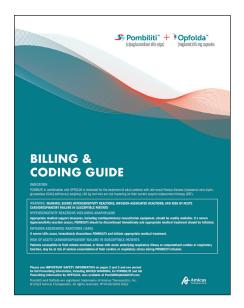




# REFERENCES

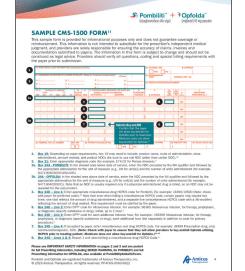
- Centers for Medicare and Medicaid Services. Medicare Claims Processing Manual Chapter 25 Completing and Processing Form CMS-1450 Data Set. Accessed June 2023 at: <a href="https://www.cms.gov/Regulations-and-Guidance/Guidance/Manuals/Downloads/clm104c25.pdf">https://www.cms.gov/Regulations-and-Guidance/Guidance/Manuals/Downloads/clm104c25.pdf</a>
- Centers for Disease Control and Prevention. ICD-10-CM Diagnosis Codes. National Center for Health Statistics ICD-10-CM. Accessed June 2023 at: <a href="https://icd10cmtool.cdc.gov/?fy=FY2022">https://icd10cmtool.cdc.gov/?fy=FY2022</a>
- 3. CPT® 2022 Professional Edition. Fourth ed. Chicago, IL: American Medical Association; 2021.
- 4. Centers for Medicare and Medicaid Services. **Medicare Claims Processing Manual** Chapter 17 Drugs and Biologicals. Accessed June 2023. https://www.cms.gov/Regulations-and-Guidance/Guidance/Manuals/Downloads/clm104c17.pdf
- 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 June 2023. 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 June 2023. <a href="https://med.noridianmedicare.com/web/jfb/topics/claim-submission/submission-errors-solutions/unlisted-procedure-and-noc-codes">https://med.noridianmedicare.com/web/jfb/topics/claim-submission/submission-errors-solutions/unlisted-procedure-and-noc-codes</a>
- 7. Noridian Healthcare Solutions. Jurisdiction F Medicare Part A. **Revenue Codes**. <a href="https://med.noridianmedicare.com/web/jfa/topics/claim-submission/revenue-codes">https://med.noridianmedicare.com/web/jfa/topics/claim-submission/revenue-codes</a>
- 8. Centers for Medicare and Medicaid Services. **CMS-1450**. Accessed June 2023. <a href="https://www.cms.gov/Regulations-and-Guidance/Legislation/PaperworkReductionActof1995/Downloads/CMS-1450.zip">https://www.cms.gov/Regulations-and-Guidance/Legislation/PaperworkReductionActof1995/Downloads/CMS-1450.zip</a>
- 9. Noridian Healthcare Solutions. **Hospital Revenue Codes**. Accessed June 2023 at: <a href="https://med.noridianmedicare.com/web/jea/topics/claim-submission/revenue-codes">https://med.noridianmedicare.com/web/jea/topics/claim-submission/revenue-codes</a>
- Centers for Medicare and Medicaid Services. Medicare Claims Processing Manual Chapter 26 Completing and Processing Form CMS-1500 Data Set. Accessed June 2023 at: <a href="https://www.cms.gov/Regulations-and-Guidance/Guidance/Manuals/Downloads/clm104c26pdf.pdf">https://www.cms.gov/Regulations-and-Guidance/Guidance/Manuals/Downloads/clm104c26pdf.pdf</a>
- 11. Centers for Medicare and Medicaid Services. **CMS-1500**. Accessed June 2023. <a href="https://www.cms.gov/Medicare/CMS-Forms/">https://www.cms.gov/Medicare/CMS-Forms/</a> <a href="https://www.cms.gov/Medicare/CMS-Forms/">CMS-Forms/</a> <a href="https://www.cms.gov/Medicare/CMS-Forms/">CMS-Forms/</a> <a href="https://www.cms.gov/Medicare/CMS-Forms/">CMS-Forms/</a> <a href="https://www.cms.gov/Medicare/CMS-Forms/">CMS-Forms/</a> <a href="https://www.cms.gov/Medicare/CMS-Forms/">CMS-Forms/</a> <a href="https://www.cms.gov/Medicare/CMS-Forms/">https://www.cms.gov/Medicare/CMS-Forms/</a> <a href="https://www.cms.gov/Medicare/CMS-Forms/">https://www.cms.gov/Medicare/CMS-Forms/</a> <a href="https://www.cms.gov/Medicare/CMS-Forms/">Lower and Common and C
- 12. Centers for Medicare and Medicaid Services. **Medicare Prescription Drug Benefit Manual** Chapter 6 Part D Drugs and Formulary Requirements. Accessed June 2023. <a href="https://www.cms.gov/medicare/prescription-drug-coverage/prescri













FURTHER

