

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

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 [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.

SAMPLE CMS-1500 FORM¹¹

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.

Opfolda Buy-and-Bill

- Confirm that the payer will allow buy-and-bill for Opfolda prior to treatment.
- Medicare does not allow buy-and-bill for Opfolda.¹²

- Box 19:** 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).¹⁰
- Box 21:** Enter appropriate diagnosis code (for example, E74.02 for Pompe disease).²
- 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).
- 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, 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.
- Box 24D – Line 1:** Enter appropriate miscellaneous drug HCPCS code for Pombiliti. For example: J3490/J3590 (Note: check with payer for preferred code).¹⁰ 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.
- 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
- 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
- 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.)^{10,12}
- Box 24G – Lines 1 & 4:** Report 1 unit when billing a miscellaneous drug HCPCS Code.⁶

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.

38		39 CODE		40		41		42		43		44		45		46		47		48		49		50		51		52		53		54		55		56		57		58		59		60		61		62		63		64		65		66		67		68																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																											
a	b	c	d	e	f	g	h	i	j	k	l	m	n	o	p	q	r	s	t	u	v	w	x	y	z	aa	ab	ac	ad	ae	af	ag	ah	ai	aj	ak	al	am	an	ao	ap	aq	ar	as	at	au	av	aw	ax	ay	az	ba	bb	bc	bd	be	bf	bg	bh	bi	bj	bk	bl	bm	bn	bo	bp	bq	br	bs	bt	bu	bv	bw	bx	by	bz	ca	cb	cc	cd	ce	cf	cg	ch	ci	cj	ck	cl	cm	cn	co	cp	cq	cr	cs	ct	cu	cv	cw	cx	cy	cz	da	db	dc	dd	de	df	dg	dh	di	dj	dk	dl	dm	dn	do	dp	dq	dr	ds	dt	du	dv	dw	dx	dy	dz	ea	eb	ec	ed	ee	ef	eg	eh	ei	ej	ek	el	em	en	eo	ep	eq	er	es	et	eu	ev	ew	ex	ey	ez	fa	fb	fc	fd	fe	ff	fg	fh	fi	fj	fk	fl	fm	fn	fo	fp	fq	fr	fs	ft	fu	fv	fw	fx	fy	fz	ga	gb	gc	gd	ge	gf	gg	gh	gi	gj	gk	gl	gm	gn	go	gp	gq	gr	gs	gt	gu	gv	gw	gx	gy	gz	ha	hb	hc	hd	he	hf	hg	hh	hi	hj	hk	hl	hm	hn	ho	hp	hq	hr	hs	ht	hu	hv	hw	hx	hy	hz	ia	ib	ic	id	ie	if	ig	ih	ii	ij	ik	il	im	in	io	ip	iq	ir	is	it	iu	iv	iw	ix	iy	iz	ja	jb	jc	jd	je	jf	jj	jk	jl	jm	jn	jo	jp	jq	jr	js	jt	ju	jv	jw	ka	kb	kc	kd	ke	kf	kg	kh	ki	kj	kk	kl	km	kn	ko	kp	kq	kr	ks	kt	ku	kv	kw	kx	ky	kz	la	lb	lc	ld	le	lf	lg	lh	li	lj	lk	ll	lm	ln	lo	lp	lq	lr	ls	lt	lu	lv	lw	lx	ly	lz	ma	mb	mc	md	me	mf	mg	mh	mi	mj	mk	ml	mm	mn	mo	mp	mq	mr	ms	mt	mu	mv	mw	mx	my	mz	na	nb	nc	nd	ne	nf	ng	nh	ni	nj	nk	nl	nm	nn	no	np	nq	nr	ns	nt	nu	nv	nw	nx	ny	nz	oa	ob	oc	od	oe	of	og	oh	oi	oj	ok	ol	om	on	oo	op	oq	or	os	ot	ou	ov	ow	ox	oy	oz	pa	pb	pc	pd	pe	pf	pg	ph	pi	pj	pk	pl	pm	pn	po	pp	pq	pr	ps	pt	pu	pv	pw	px	py	pz	qa	qb	qc	qd	qe	qf	qg	qh	qi	qj	qk	ql	qm	qn	qo	qp	qq	qr	qs	qt	qu	qv	qw	qx	qy	qz	ra	rb	rc	rd	re	rf	rg	rh	ri	rj	rk	rl	rm	rn	ro	rp	rq	rr	rs	rt	ru	rv	rw	rx	ry	rz	sa	sb	sc	sd	se	sf	sg	sh	si	sj	sk	sl	sm	sn	so	sp	sq	sr	ss	st	su	sv	sw	sx	sy	sz	ta	tb	tc	td	te	tf	tg	th	ti	tj	tk	tl	tm	tn	to	tp	tq	tr	ts	tt	tu	tv	tw	tx	ty	tz	ua	ub	uc	ud	ue	uf	ug	uh	ui	uj	uk	ul	um	un	uo	up	uq	ur	us	ut	uu	uv	uw	ux	uy	uz	va	vb	vc	vd	ve	vf	vg	vh	vi	vj	vk	vl	vm	vn	vo	vp	vq	vr	vs	vt	vu	vv	vw	vx	vy	vz	wa	wb	wc	wd	we	wf	wg	wh	wi	wj	wk	wl	wm	wn	wo	wp	wq	wr	ws	wt	wu	wv	ww	wx	wy	wz	xa	xb	xc	xd	xe	xf	xg	xh	xi	xj	xk	xl	xm	xn	xo	xp	xq	xr	xs	xt	xu	xv	xw	xx	xy	xz	ya	yb	yc	yd	ye	yf	yg	yh	yi	yj	yk	yl	ym	yn	yo	yp	yq	yr	ys	yt	yu	yv	yw	yx	yy	yz	za	zb	zc	zd	ze	zf	zg	zh	zi	zj	zk	zl	zm	zn	zo	zp	zq	zr	zs	zt	zu	zv	zw	zx	zy	zz
42 REV. CD.		43 DESCRIPTION		44 HCPCS / RATE / HIPPS CODE		45 SERV. DATE		46 SERV. UNITS		47 TOTAL		48		49		50		51		52		53		54		55		56		57		58		59		60		61		62		63		64		65		66		67		68																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																			
69 ADMIT DX		70 PATIENT REASON DX		71 PPS CODE		72 ECI		73		74 PRINCIPAL PROCEDURE CODE		75		76 ATTENDING NPI		77 OPERATING NPI		78 OTHER NPI		79 OTHER NPI		80 REMARKS		81 CC		82		83		84		85		86		87		88		89		90		91		92		93		94		95		96		97		98		99																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																																											

Field 46⁵
Report 1 unit when billing a miscellaneous drug HCPCS Code.

Opfolda Buy-and-Bill

- Confirm that the payer will allow buy-and-bill for Opfolda prior to treatment.
- Medicare does not allow buy-and-bill for Opfolda.¹²

Fields 42 & 43^{5,9}
Enter appropriate revenue code and descriptors.
Examples:
• 0250 for general pharmacy
• 0260 for general IV therapy
• 0636 for drugs that require detailed coding

Field 44^{3,4}
Enter appropriate HCPCS coding.
Examples:
• C9399 Unclassified drugs or biologicals (Not all payers accept a C-code for product use in the hospital outpatient setting; therefore, it is important to clarify by payer)
• 96365 Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); initial, up to 1 hour
• +96366 Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); each additional hour (List separately in addition to code for primary procedure)

Field 67 (A-Q)²
Enter appropriate ICD-10-CM diagnosis code(s) as reflected in the patient's medical record (for example, E74.02 for Pompe disease).

Field 80⁴
Payer requirements vary. Examples might include drug name(s), NDC(s), route(s) of administration, dose(s) administered, wastage.

REFERENCES

1. Centers for Medicare and Medicaid Services. Medicare Claims Processing Manual Chapter 25 - **Completing and Processing Form CMS-1450** Data Set. Accessed June 2023 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 June 2023 at: <https://icd10cmtool.cdc.gov/?fy=FY2022>
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>
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 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. <https://med.noridianmedicare.com/web/jfb/topics/claim-submission/submission-errors-solutions/unlisted-procedure-and-noc-codes>
7. Noridian Healthcare Solutions. Jurisdiction F – Medicare Part A. **Revenue Codes**. <https://med.noridianmedicare.com/web/jfa/topics/claim-submission/revenue-codes>
8. Centers for Medicare and Medicaid Services. **CMS-1450**. Accessed June 2023. <https://www.cms.gov/Regulations-and-Guidance/Legislation/PaperworkReductionActof1995/Downloads/CMS-1450.zip>
9. Noridian Healthcare Solutions. **Hospital Revenue Codes**. Accessed June 2023 at: <https://med.noridianmedicare.com/web/jea/topics/claim-submission/revenue-codes>
10. Centers for Medicare and Medicaid Services. Medicare Claims Processing Manual Chapter 26 - **Completing and Processing Form CMS-1500** Data Set. Accessed June 2023 at: <https://www.cms.gov/Regulations-and-Guidance/Guidance/Manuals/Downloads/clm104c26pdf.pdf>
11. Centers for Medicare and Medicaid Services. **CMS-1500**. Accessed June 2023. <https://www.cms.gov/Medicare/CMS-Forms/CMS-Forms/Downloads/CMS1500.pdf>
12. Centers for Medicare and Medicaid Services. **Medicare Prescription Drug Benefit Manual** Chapter 6 – Part D Drugs and Formulary Requirements. Accessed June 2023. <https://www.cms.gov/medicare/prescription-drug-coverage/prescriptiondrugcovcontra/downloads/part-d-benefits-manual-chapter-6.pdf>

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