

PA Criteria	Criteria Details						
Covered Uses (FDA approved indication)	Pombiliti is a hydrolytic lysosomal glycogen-specific enzyme indicated, in combination with Opfolda (an enzyme stabilizer) 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).						
Exclusion Criteria	Must not be used in combination with another ERT (such as Lumizyme or Nexvzyme).						
Required Medical Information	Medical records supporting the request must be provided, including the following: <ol style="list-style-type: none"> 1. Patient's current weight. 2. For initial coverage: Confirmation of diagnosis by enzyme assay or genetic testing. 						
Age Restriction	Must be at least 13 years of age.						
Prescriber Restrictions	Must be prescribed by or in consultation with a specialist for the condition (such as genetic and metabolic specialists, neurologist, cardiologist, pediatrician).						
Coverage Duration	One year initial; two years reauthorization. Dose will be approved according to the FDA- approved labeling or within accepted standards of medical practice. Must be used in combination with Opfolda. For reauthorization, must also have documented response to therapy evidenced by improvement or stabilization in the condition (such as improved or stable muscle strength, motor function, cardiac involvement, FVC, and/or 6MWT).						
Other Criteria/Information	Refer to the Gold Coast Health Plan Medicare Part B Reference and Summary of Evidence document. <table border="1" data-bbox="495 1161 1513 1306"> <thead> <tr> <th>HCPCS</th> <th>Description</th> <th>Billing Units/How Supplied</th> </tr> </thead> <tbody> <tr> <td>J1203</td> <td>Pombiliti (cipaglicosidase-alfa)</td> <td>Billing unit: 5 mg 105 mg SDV</td> </tr> </tbody> </table>	HCPCS	Description	Billing Units/How Supplied	J1203	Pombiliti (cipaglicosidase-alfa)	Billing unit: 5 mg 105 mg SDV
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STATUS	DATE REVISED	REVIEW DATE	APPROVED/REVIEWED BY	EFFECTIVE DATE
Created	3/26/2025	3/26/2025	Dawn Shojai, PharmD, Senior Pharmacy Benefit Consultant (PSG)	N/A
Approved	N/A	5/15/2025	Pharmacy & Therapeutics (P&T) Committee	5/15/2025