

SUBSTRATE REDUCTION THERAPY

PRIOR AUTHORIZATION REQUEST

PRESCRIBER FAX FORM

Only the prescriber may complete this form. This form is for prospective, concurrent, and retrospective reviews.

The following documentation is **REQUIRED**. Incomplete forms will be returned for additional information. For formulary information please visit www.myprime.com. Start saving time today by filling out this form electronically. Visit covermymeds.com to begin using this free service.

What is the priority level of this request?

- Standard review
- Expedited/Urgent review – prescriber certifies that waiting for a standard review could seriously harm the patient’s life, health or ability to regain maximum function

Today’s Date: _____

PATIENT AND INSURANCE INFORMATION

Date of Service (if differs from Today’s Date): _____

Patient Name (First):	Last:	M:	DOB (mm/dd/yyyy):
Patient Address:	City, State, Zip:	Patient Telephone:	
Member ID Number:	Group Number:		

PRESCRIBER/CLINIC INFORMATION

Prescriber Name:	Prescriber NPI#:	Specialty:	Contact Name:
Clinic Name:	Clinic Address:		
City, State, Zip:	Phone #:	Secure Fax #:	

PLEASE ATTACH ANY ADDITIONAL INFORMATION THAT SHOULD BE CONSIDERED WITH THIS REQUEST

Patient’s Diagnosis: <input type="checkbox"/> Gaucher disease type I (GD1) <input type="checkbox"/> Late-onset Pompe disease (acid maltase deficiency [AMD]; glycogen storage disease type II [GSDII]) <input type="checkbox"/> Other (ICD code, plus description): _____	
Medication Requested:	Strength:
Dosing Schedule:	Quantity per Month:
<p>For all requests:</p> <p>1. What is the patient’s weight: _____ (kg)</p> <p>2. Is the patient currently being treated with the requested agent? <input type="checkbox"/> Yes <input type="checkbox"/> No If yes, is the patient currently stable on the requested agent? Please note, chart notes are required..... <input type="checkbox"/> Yes <input type="checkbox"/> No</p> <p>3. Has the patient been treated with the requested agent within the past 90 days (starting on samples is not approvable)?..... <input type="checkbox"/> Yes <input type="checkbox"/> No If yes, is the patient at risk if therapy is changed? <input type="checkbox"/> Yes <input type="checkbox"/> No If yes, please explain risk: _____</p> <p>4. Does the patient have any FDA labeled contraindications to the requested agent?..... <input type="checkbox"/> Yes <input type="checkbox"/> No If yes, please specify contraindications: _____</p> <p>5. Is the prescriber a specialist in the area of the patient’s diagnosis (e.g., neurologist, oncologist), or has the prescriber consulted with a specialist in the area of the patient’s diagnosis? <input type="checkbox"/> Yes <input type="checkbox"/> No</p> <p>6. Is the patient’s age within FDA labeling for the requested indication for the requested agent? <input type="checkbox"/> Yes <input type="checkbox"/> No If no, please provide support for using the requested agent for the patient’s age for the requested indication: _____</p> <p>7. Please list all reasons for selecting the requested agent for the indicated diagnosis, strength, dosing schedule, and quantity over alternatives (e.g., compendia support, journal articles, contraindications, allergies, history of adverse drug reactions to alternatives, lower dose has been tried, information supporting dose over FDA max). Please note, documentation may be required: _____</p> <p>_____</p> <p>_____</p>	
<p>Please continue to the next page.</p>	

Patient Name (First):	Last:	M:	DOB (mm/dd/yyyy):
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For Cerdelga, eliglustat requests:

8. Is the prescriber a specialist in the area of the patient's diagnosis (such as endocrinologist, geneticist), or has the prescriber consulted with a specialist in the area of the patient's diagnosis? Yes No
9. Will the patient be using the requested agent in combination with another substrate reduction therapy agent (e.g., Opfolda, miglustat, Zavesca) for the requested indication? Yes No
- **For Gaucher disease type 1 (GD1) requests:**
10. Does the patient have any neuronopathic symptoms indicative of Gaucher disease type 2 or type 3 [e.g., bulbar signs (e.g., stridor, strabismus, swallowing difficulty), pyramidal signs (e.g., opisthotonos, head retroflexion, spasticity, trismus), oculomotor apraxia, tonic-clonic seizures, myoclonic epilepsy, dementia, ataxia]? Yes No
11. Does the patient have baseline (prior to therapy for the requested indication) glucocerebrosidase enzyme activity of less than or equal to 15% of mean normal in fibroblasts, leukocytes, or other nucleated cells?..... Yes No
If no, has genetic analysis confirmed two pathogenic alleles on the glucocerebrosidase (GBA) gene?..... Yes No
12. Does the patient have ONE of the following clinical presentations at baseline (prior to therapy for the requested indication)? (Select one.)
- Anemia defined as mean hemoglobin (Hb) level below the testing laboratory's lower limit of the normal range based on age and gender
 - Thrombocytopenia (platelet count less than 100,000/microliter on at least 2 measurements)
 - Hepatomegaly
 - Splenomegaly
 - Growth failure (i.e., growth velocity is below the standard mean for age)
 - Evidence of bone disease with other causes ruled out
 - None of the above
13. Is the patient a CYP2D6 extensive metabolizer (EM), intermediate metabolizer (IM), or poor metabolizer (PM) as detected by an FDA-cleared test for determining CYP2D6 genotype? Yes No

For Opfolda requests:

14. Is the prescriber a specialist in the area of the patient's diagnosis (e.g., endocrinologist, geneticist) or has the prescriber consulted with a specialist in the area of the patient's diagnosis? Yes No
15. Will the patient be using the requested agent in combination with another substrate reduction therapy agent (e.g., Cerdelga, eliglustat, Zavesca) for the requested indication? Yes No
16. Will the requested agent be used in combination with Miplyffa (arimoclomol)? Yes No
If yes, please answer the following questions.
- Does the patient have a diagnosis of Niemann-Pick type C disease (NPC)? Yes No
 - Has genetic analysis confirmed a mutation in the NPC1 or NPC2 genes? Yes No
 - Does the patient have disease-related neurological symptoms? Yes No
 - Is the patient's age within Miplyffa FDA labeling for the requested indication? Yes No

• **For late-onset Pompe disease (acid maltase deficiency [AMD]; glycogen storage disease type II [GSDII]) requests:**

17. Has the diagnosis been confirmed by at least ONE of the following? (Select one.)
- Genetic analysis confirms biallelic mutation (two pathogenic variants) in the GAA gene
 - The patient has deficient acid alpha-glucosidase glycogen enzyme activity in dried blood spots, leukocytes, skin fibroblasts, and/or skeletal muscle tissue
 - None of the above
18. Has the patient improved on their current enzyme replacement therapy (ERT)? Yes No
19. Will the requested agent be taken in combination with Pombiliti? Yes No

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Patient Name (First):	Last:	M:	DOB (mm/dd/yyyy):
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For Zavesca, miglustat requests:

20. Is the prescriber a specialist in the area of the patient's diagnosis (e.g., endocrinologist, geneticist), or has the prescriber consulted with a specialist in the area of the patient's diagnosis? Yes No
21. Will the patient be using the requested agent in combination with another substrate reduction therapy agent (e.g., Cerdelga, eliglustat, Opfolda) for the requested indication? Yes No
22. Will the requested agent be used in combination with Miplyffa (arimoclomol)? Yes No
- If yes, please answer the following questions:
- Does the patient have a diagnosis of Niemann-Pick type C disease (NPC)? Yes No
 - Has genetic analysis confirmed a mutation in the NPC1 or NPC2 genes? Yes No
 - Does the patient have disease-related neurological symptoms? Yes No
 - Is the patient's age within Miplyffa FDA labeling for the requested indication? Yes No

• For Gaucher disease type 1 (GD1) requests:

23. Does the patient have any neuronopathic symptoms indicative of Gaucher disease type 2 or type 3 [e.g., bulbar signs (e.g., stridor, strabismus, swallowing difficulty), pyramidal signs (e.g., opisthotonos, head retroflexion, spasticity, trismus), oculomotor apraxia, tonic-clonic seizures, myoclonic epilepsy, dementia, ataxia]? Yes No
24. Does the patient have a baseline (prior to therapy for the requested indication) glucocerebrosidase enzyme activity of less than or equal to 15% of mean normal in fibroblasts, leukocytes, or other nucleated cells? Yes No
- If no, has genetic analysis confirmed two pathogenic alleles on the glucocerebrosidase (GBA) gene? Yes No
25. Is enzyme replacement therapy (ERT) NOT a therapeutic option (e.g., due to allergy, hypersensitivity, poor venous access, or previous ERT failure)? Yes No
26. Does the patient have ONE of the following clinical presentations at baseline (prior to therapy for the requested indication)? (Select one.)
- Anemia defined as mean hemoglobin (Hb) level below the testing laboratory's lower limit of the normal range based on age and gender
 - Thrombocytopenia (platelet count less than 100,000/microliter on at least 2 measurements)
 - Hepatomegaly
 - Splenomegaly
 - Growth failure (i.e., growth velocity is below the standard mean for age)
 - Evidence of bone disease with other causes ruled out
 - None of the above

For brand Zavesca requests:

• Please submit chart notes to support the answers to the following questions:

27. Has the patient tried and had an inadequate response to the generic equivalent (miglustat)? Yes No
28. Was the generic equivalent (miglustat) discontinued due to lack of efficacy or effectiveness, diminished effect, or an adverse event? Yes No
29. Does the patient have an intolerance, or hypersensitivity to the generic equivalent (miglustat) that is not expected to occur with the brand agent? Yes No
30. Does the patient have an FDA labeled contraindication to the generic equivalent (miglustat) that is not expected to occur with the brand agent? Yes No
31. Is the generic equivalent (miglustat) expected to be ineffective based on the known clinical characteristics of the patient and the known characteristics of the prescription drug; OR cause a significant barrier to the patient's adherence of care; OR worsen a comorbid condition; OR decrease the patient's ability to achieve or maintain reasonable functional ability in performing daily activities; OR cause an adverse reaction or cause physical or mental harm? Yes No
32. Is the generic equivalent (miglustat) not in the best interest of the patient based on medical necessity? Yes No
33. Has the patient tried another prescription drug in the same pharmacologic class or with the same mechanism of action as the generic equivalent (miglustat) agent and that prescription drug was discontinued due to lack of efficacy or effectiveness, diminished effect, or an adverse event? Yes No
34. Is support for the use of the requested brand Zavesca over the generic equivalent (miglustat)? Yes No
- If yes, please provide supporting information: _____

Please continue to the next page.

Patient Name (First):	Last:	M:	DOB (mm/dd/yyyy):
For renewal requests: 35. Has the patient had clinical benefit with the requested agent?..... <input type="checkbox"/> Yes <input type="checkbox"/> No			
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