

Olipudase alfa-rpcp (Xenpozyme) Induction Followed by Maintenance Every 2 Weeks

olipudase alfa-rpcp (XENPOZYME) Dose escalation regimen Induction followed by maintenance 3 mg/kg IV every 14 days

Protocol Types

Infusion Treatment 1, Infusion Treatment 2, Infusion Treatment 3, Infusion Treatment 4

Patient name: _____ DOB: _____

Please include: ☐ Patient face sheet ☐ Insurance authorization (if other than Medicaid or non-Advantage Medicare) Diagnosis: _____

Review: Every 358 days

Orders

	Interval	Defer Until	Duration
--	----------	-------------	----------

Provider Communication

☒ Provider communication	Every visit		Until discontinued
Until discontinued Starting when released Until Specified			
Severe hypersensitivity (ie, anaphylaxis, angioedema) and infusion-associated reactions have been reported with olipudase alfa-rpcp. Ensure appropriate medical monitoring and support measures are available during treatment.			

Treatment should be in direct consultation with a physician knowledgeable in the management of acid sphingomyelinase deficiency (ASMD).

Pregnancy status should be verified in all patients with reproductive potential prior to treatment start. Those who may become pregnant should use effective contraception during therapy and for 14 days after the last dose of olipudase alfa-rpcp.

Labs

☒ hCG, serum, qualitative	Every visit		Until discontinued
Status: Once, Expected: S, Clinic Collect, Blood, Venous, Blood, AH INTERNAL LAB (BEAKER), Routine			
Inclusion conditions: Patient could become pregnant			

☒ Comprehensive metabolic panel	Every visit		Until discontinued
Status: Once, Expected: S, Clinic Collect, Blood, Venous, Blood, AH INTERNAL LAB (BEAKER), Routine			

Nursing Orders

☒ Treatment conditions	Every visit		Until discontinued
Until discontinued Starting when released Until Specified			
HOLD treatment and contact provider for AST/ALT greater than 2x ULN.			

Pre-Medications

☒ AH PREMEDS OPTIONAL ACETAMINOPHEN 650 / DIPHENHYDRAMINE 50 PO / LORATADINE 10 PO / METHYLPREDNISOLONE IV 40 (OFFSET 0 MIN)			
☐ acetaminophen (Tylenol) tablet 650 mg	Every visit		Until discontinued
650 mg, oral, Once, Starting at treatment start time			

	Interval	Defer Until	Duration
☐ diphenhydrAMINE (BENADryl) 50 mg 50 mg, oral, Once, Starting at treatment start time Give diphenhydramine if patient has transportation. Give either loratadine or diphenhydrAMINE, NOT both.	Every visit		Until discontinued
☐ loratadine (Claritin) tablet 10 mg 10 mg, oral, Once as needed, allergies, if diphenhydramine is not given, Starting at treatment start time, Until Discontinued Give loratadine if patient does NOT have transportation. Give either loratadine or diphenhydrAMINE, NOT both.	Every visit		Until discontinued
☐ methylPREDNISolone sod suc (PF) (SOLU-Medrol) injection 40 mg 40 mg, intravenous, Administer over: 2 Minutes, Once, Starting at treatment start time	Every visit		Until discontinued

Therapeutic Medications

☒ Provider communication For patients with a BMI ≤ 30 kg/m ² , use actual body weight. For patients with BMI > 30 kg/m ² , use the following adjusted body weight for dosage calculation: adjusted body weight = (actual height in meters) ² $\times$ 30.	Every visit		Until discontinued
☒ olipudase alfa (Xenpozyme) infusion 0.1 mg/kg 0.1 mg/kg, intravenous, Once, Starting 30 minutes after treatment start time Start infusion at 20 ml/hr. If no infusion reaction after 20 minutes, increase rate to 60 ml/hr and maintain until infusion complete.	Once		1 treatment
☒ olipudase alfa (Xenpozyme) infusion 0.3 mg/kg 0.3 mg/kg, intravenous, Once, Starting 30 minutes after treatment start time Start infusion at 3.33 ml/hr. After 20 minutes at this rate, if no infusion reaction is observed, increase to 10 mL/hr. After an additional 20 minutes, if no infusion reaction is observed, increase to 20 mL/hr. After an additional 20 minutes, if no infusion reaction is observed, increase to 33.33 mL/hr and maintain until infusion is complete.	Every 14 days	S+14	2 treatments
☒ olipudase alfa (Xenpozyme) infusion 0.6 mg/kg 0.6 mg/kg, intravenous, Once, Starting 30 minutes after treatment start time Start infusion at 3.33 ml/hr. After 20 minutes at this rate, if no infusion reaction is observed, increase to 10 mL/hr. After an additional 20 minutes, if no infusion reaction is observed, increase to 20 mL/hr. After an additional 20 minutes, if no infusion reaction is observed, increase to 33.33 mL/hr and maintain until infusion is complete.	Every 14 days	S+42	2 treatments
☒ olipudase alfa (Xenpozyme) infusion 1 mg/kg 1 mg/kg, intravenous, Once, Starting 30 minutes after treatment start time Start infusion at 3.33 ml/hr. After 20 minutes at this rate, if no infusion reaction is observed, increase to 10 mL/hr. After an additional 20 minutes, if no infusion reaction is observed, increase to 20 mL/hr. After an additional 20 minutes, if no infusion reaction is observed, increase to 33.33 mL/hr and maintain until infusion is complete.	Once	S+70	1 treatment
☒ olipudase alfa (Xenpozyme) infusion 2 mg/kg 2 mg/kg, intravenous, Once, Starting 30 minutes after treatment start time Start infusion at 3.33 ml/hr. After 20 minutes at this rate, if no infusion reaction is observed, increase to 10 mL/hr. After an additional 20 minutes, if no infusion reaction is observed, increase to 20 mL/hr. After an additional 20 minutes, if no infusion reaction is observed, increase to 33.33 mL/hr and maintain until infusion is complete.	Once	S+84	1 treatment
☒ olipudase alfa (Xenpozyme) infusion 3 mg/kg 3 mg/kg, intravenous, Once, Starting 30 minutes after treatment start time Start infusion at 3.33 ml/hr. After 20 minutes at this rate, if no infusion reaction is observed, increase to 10 mL/hr.	Every 14 days	S+98	Until discontinued

	Interval	Defer Until	Duration
After an additional 20 minutes, if no infusion reaction is observed, increase to 20 mL/hr. After an additional 20 minutes, if no infusion reaction is observed, increase to 33.33 mL/hr and maintain until infusion is complete.			
Emergency Medications			
☒ Nursing communication STAT, Until discontinued Starting when released If hypersensitivity reaction occurs, stop infusion IMMEDIATELY and CALL provider.	PRN		Until discontinued
☒ Vital Signs STAT, Every 15 min Starting when released If a hypersensitivity reaction occurs, perform vital signs every 15 minutes for one hour, then every 30 minutes for 2 hours.	PRN		Until discontinued
☒ Nursing communication Once Starting when released If hypersensitivity reaction occurs: - Elevate head of bed 30 degrees and place patient's head at 30 degree angle if possible - Start 2 liters oxygen per nasal cannula and maintain airway - Change and maintain IV line with 1000 mL 0.9% Normal Saline	PRN		Until discontinued
☒ sodium chloride 0.9 % infusion 30 mL/hr, intravenous, Continuous PRN, KVO, Starting when released, for 1 day	PRN		Until discontinued
☒ albuterol 2.5 mg /3 mL (0.083 %) nebulizer solution 2.5 mg 2.5 mg, nebulization, Every 4 hours PRN, Hypersensitivity reaction, Starting when released, for 1 day	PRN		Until discontinued
☒ albuterol HFA 90 mcg/act inhaler 2 puff 2 puff, inhalation, Every 4 hours PRN, Hypersensitivity reaction, Starting when released, for 1 day SHAKE WELL	PRN		Until discontinued
☒ acetaminophen (Tylenol) tablet 650 mg 650 mg, oral, Once as needed, fever, GREATER THAN or EQUAL TO 38 C, Starting when released, for 1 dose For mild/moderate reaction: flushing, rash, pruritus	PRN		Until discontinued
☒ diphenhydramine (BENADryl) injection 25 mg 25 mg, intravenous, Administer over: 2 Minutes, Once as needed, Hypersensitivity reaction, Starting when released, for 1 dose For mild/moderate reaction: flushing, rash, pruritus	PRN		Until discontinued
☒ diphenhydramine (BENADryl) injection 50 mg 50 mg, intravenous, Administer over: 2 Minutes, Once as needed, Hypersensitivity reaction, Starting when released, for 1 dose For severe reaction: respiratory distress, stridor, wheezing, hypotension (SBP LESS THAN 100 mm Hg)	PRN		Until discontinued
☒ hydrocortisone sodium succinate (Solu-CORTEF) injection 100 mg 100 mg, intravenous, Once as needed, Hypersensitivity reaction, Starting when released, for 1 dose For mild/moderate reaction: flushing, rash, pruritus - OR - For severe reaction: respiratory distress, stridor, wheezing, hypotension (SBP LESS THAN 100 mm Hg)	PRN		Until discontinued
☒ EPINEPHrine (EpiPen;Adrenalin) injection solution 0.3 mg 0.3 mg, intramuscular, Once as needed, Hypersensitivity reaction, Starting when released, for 1 dose For severe reaction: respiratory distress, stridor, wheezing, hypotension (SBP LESS THAN 100 mm Hg)	PRN		Until discontinued

Appointment Requests

☒ Infusion Appointment Request Status: Once, Expected: S, Sched. Duration: 90 minutes, Sched. Tolerance: Schedule appointment at most 3 days before or at most 3 days after	Once		1 treatment
☒ Infusion Appointment Request	Every 14 days	S+14	2 treatments

	Interval	Defer Until	Duration
Status: Once, Expected: S, Sched. Duration: 90 minutes, Sched. Tolerance: Schedule appointment at most 3 days before or at most 3 days after			
☒ Infusion Appointment Request	Every 14 days	S+42	2 treatments
Status: Once, Expected: S, Sched. Duration: 90 minutes, Sched. Tolerance: Schedule appointment at most 3 days before or at most 3 days after			
☒ Infusion Appointment Request	Once	S+70	1 treatment
Status: Once, Expected: S, Sched. Duration: 90 minutes, Sched. Tolerance: Schedule appointment at most 3 days before or at most 3 days after			
☒ Infusion Appointment Request	Once	S+84	1 treatment
Status: Once, Expected: S, Sched. Duration: 90 minutes, Sched. Tolerance: Schedule appointment at most 3 days before or at most 3 days after			
☒ Infusion Appointment Request	Every 14 days	S+98	Until discontinued
Status: Once, Expected: S, Sched. Duration: 90 minutes, Sched. Tolerance: Schedule appointment at most 3 days before or at most 3 days after			

*Provider signature	Provider printed name	*Date	Time
---------------------	-----------------------	-------	------

*Provider phone#

***Denotes field that must be completed by healthcare worker**