MADISON NEUROLOGY LLC

608 403 0000

Neurology Care 7 days a Week. Because Your Brain Matters.

Lumizyme® / Myozyme®

Alglucosidase alfa

COMPLETE CLINICAL PROTOCOL: PRE-TREATMENT CHECKLIST • ORDER SET • FOLLOW-UP • HOLD/DISCONTINUE CRITERIA

GENERIC NAME	alglucosidase alfa
BRAND	LUMIZYME® / MYOZYME®
MEDICATION CLASS	Enzyme replacement therapy (recombinant acid alpha-glucosidase)
PRESENTATION	50 mg single-dose vial for reconstitution and dilution
ROUTE	IV infusion over approximately 4 hours with stepwise rate escalation, every 2 weeks
US INDICATION (FDA)	Pompe disease (acid alpha-glucosidase deficiency). Lumizyme: patients ≥1 year (late-onset forms in the US label); Myozyme: infantile-onset Pompe disease.

Class context.
Boxed warnings: life-threatening anaphylaxis and severe allergic/immune-mediated reactions, and risk of acute cardiorespiratory failure in susceptible infantile-onset patients with compromised cardiac/respiratory function — infusion sites must be immediately prepared to manage anaphylaxis, and high-risk patients need appropriate support available throughout.

Section 1 — Pre-Treatment Checklist

A. Required screening labs

- ☐ Confirmed acid alpha-glucosidase deficiency (enzyme assay ± genotyping).
- ☐ Baseline cardiorespiratory status — infantile-onset and severely compromised patients need intensive support immediately available during infusions.
- ☐ Baseline motor/respiratory function for response tracking (6-minute walk, FVC where age-appropriate).
- ☐ Anti-rhGAA IgG antibody baseline — titers affect efficacy and reaction risk; CRIM status is assessed in infantile-onset disease (CRIM-negative infants usually need immune-tolerance induction managed by the metabolic specialist).
- ☐ Pregnancy test when relevant.

B. Vaccinations

☐ No label restrictions; routine schedules per age.

C. Clinical assessments & counseling

☐ Counsel on infusion-associated reactions (during or within hours after) and on delayed immune-mediated reactions.

☐ Verify emergency readiness (epinephrine, airway support) before EVERY course in high-risk patients.

D. Exclusion / hold criteria — do not start

CRITERION	RATIONALE / REQUIRED ACTION
Prior anaphylaxis to alglucosidase without a specialist desensitization plan	Do not administer.
Acute cardiorespiratory decompensation	Stabilize first; infuse only with appropriate support.

Section 2 — Order Set

Premedication

☐ Optional: antihistamine ± antipyretic ± corticosteroid before infusion — standard after any prior infusion-associated reaction.

Dosing schedule

Standard dose	20 mg/kg IV every 2 weeks.
Infusion	Approximately 4 hours with stepwise rate escalation per label; extend the duration in patients with reaction history.
Reaction rule	Interrupt for mild–moderate reactions; resume at a slower rate once resolved; STOP for severe reactions.

Standing orders — every infusion

☐ Vital signs at baseline, each rate change, and completion; risk-appropriate post-infusion observation.

☐ Document reactions meticulously — they drive premedication and rate decisions at subsequent visits.

Missed or delayed dose

Resume as soon as feasible at the every-2-week interval.

Section 3 — Follow-Up Protocol

ITEM	FREQUENCY / TIMING	ACTION / THRESHOLD
Anti-rhGAA IgG antibodies	Periodically per label/specialist	Rising/sustained high titers can track with reduced response or reactions — specialist review.

Motor and respiratory function	Every 6–12 months	Continuation documentation.
Infusion-reaction history	Reviewed at every visit	Escalating reactions → premedication/rate revision or specialist reevaluation.
Immune-mediated events (nephrotic syndrome, skin, other)	Vigilance at every visit	Specialist evaluation.

Section 4 — Pause vs. Stop Criteria

A. Pause or delay (resume when resolved)

CRITERION	REQUIRED ACTION
Moderate infusion reaction	Interrupt; resume slower once resolved.
Intercurrent cardiorespiratory compromise	Defer and stabilize.

B. Discontinue permanently

CRITERION	REQUIRED ACTION
Anaphylaxis	Stop; rechallenge only under specialist desensitization protocols.
Immune-mediated end-organ toxicity attributed to therapy	Stop; specialist review.
High sustained antibody titers with loss of clinical response	Specialist review of continuing ERT.

Section 5 — Notes & Practical Points

☐ Scheduling note for infantile-onset CRIM-negative patients: immune-tolerance induction runs alongside early ERT under the metabolic specialist — infusion scheduling must accommodate it.

Prepared November 2026 by Madison Neurology LLC.

This document is an educational compilation and does not reproduce the complete prescribing information. Verify every element against the current FDA label and applicable REMS requirements before clinical use.

- Current U.S. prescribing information via NIH DailyMed — https://dailymed.nlm.nih.gov/dailymed/
- FDA Drugs@FDA database — https://www.accessdata.fda.gov/scripts/cder/daf/
- FDA drug safety and availability updates — https://www.fda.gov/drugs/drug-safety-and-availability

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Questions about infusion referrals: call 608 403 0000. Neurology Care 7 days a Week. Because Your Brain Matters.