MADISON NEUROLOGY LLC

608 403 0000

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

Nexviazyme®

Avalglucosidase alfa-ngpt

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

GENERIC NAME	avalglucosidase alfa-ngpt
BRAND	NEXVIAZYME®
MEDICATION CLASS	Enzyme replacement therapy (enhanced M6P-targeted recombinant acid alpha-glucosidase)
PRESENTATION	100 mg single-dose vial for reconstitution and dilution
ROUTE	IV infusion every 2 weeks; duration escalates/stepwise per label (initial infusions longer, may shorten as tolerated)
US INDICATION (FDA)	Late-onset Pompe disease in patients ≥1 year of age.

Class context.
Boxed warning: severe hypersensitivity reactions INCLUDING ANAPHYLAXIS, and infusion-associated reactions — the same emergency-readiness requirements as alglucosidase apply.

Section 1 — Pre-Treatment Checklist

A. Required screening labs

☐ Confirmed late-onset Pompe diagnosis (enzyme assay ± genotype).

☐ Baseline motor/respiratory function (6-minute walk, FVC).

☐ Prior ERT history: document alglucosidase use, dose, duration, and reactions — most adults starting avalglucosidase are switching.

☐ Pregnancy test when relevant.

B. Vaccinations

☐ No label restrictions.

C. Clinical assessments & counseling

☐ Hypersensitivity/infusion-reaction counseling; verify anaphylaxis readiness.

☐ Document the switch rationale when transitioning from alglucosidase.

D. Exclusion / hold criteria — do not start

CRITERION	RATIONALE / REQUIRED ACTION
Prior anaphylaxis to avalglucosidase without a specialist rechallenge plan	Do not administer.

Section 2 — Order Set

Premedication

☐ Consider antihistamine, antipyretic, and/or corticosteroid before infusion — label-supported mitigation, particularly with reaction history.

Dosing schedule

Standard dose	20 mg/kg IV every 2 weeks.
Infusion	Gradual escalation per label; initial infusions run longer and may shorten as tolerated.
Reaction rule	Interrupt/slow for mild–moderate; stop for severe reactions.

Standing orders — every infusion

☐ Monitor during and after infusion; document reactions to inform future premedication.

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
Motor/respiratory function	Every 6–12 months	Continuation documentation.
Infusion-reaction review	Every infusion	Escalating reactions → revise premedication/rates.
Antibody development	Per specialist when response plateaus	High titers → specialist review.

Section 4 — Pause vs. Stop Criteria

A. Pause or delay (resume when resolved)

CRITERION	REQUIRED ACTION
Moderate infusion reaction	Interrupt, then resume slower.
Intercurrent cardiorespiratory illness	Defer.

B. Discontinue permanently

CRITERION	REQUIRED ACTION
Anaphylaxis or severe immune-mediated reaction	Stop; specialist rechallenge only.

Section 5 — Notes & Practical Points

☐ Switches from alglucosidase should be scheduled as a clean handoff at the routine 2-week interval — no washout needed, but document the transition date.

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

Nexviazyme is a registered trademark of its respective owner; referenced here for identification only.

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