MADISON NEUROLOGY LLC

608 403 0000

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

Fabrazyme®

Agalsidase beta

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

GENERIC NAME	agalsidase beta
BRAND	FABRAZYME®
MEDICATION CLASS	Enzyme replacement therapy (recombinant alpha-galactosidase A)
PRESENTATION	5 mg and 35 mg vials for reconstitution and dilution
ROUTE	IV infusion every 2 weeks; slow initial rate (no more than 0.25 mg/min) with stepwise escalation as tolerated
US INDICATION (FDA)	Fabry disease in patients ≥2 years of age.

Class context.
Labeled warnings include anaphylaxis and severe hypersensitivity — maintain emergency readiness at every infusion. Infusion reactions are common early and usually managed with premedication plus rate reduction rather than discontinuation.

Section 1 — Pre-Treatment Checklist

A. Required screening labs

☐ Confirmed Fabry diagnosis (alpha-galactosidase A activity ± GLA genotype).

☐ Baseline organ assessment: renal function/eGFR, cardiac status (Fabry cardiomyopathy), and pain/quality-of-life measures.

☐ Pregnancy test when relevant.

B. Vaccinations

☐ No label restrictions.

C. Clinical assessments & counseling

- ☐ Counsel that early infusion reactions are common and usually manageable with rate reduction and premedication — this framing prevents early abandonment.
- ☐ Document baseline neuropathic pain and GI symptom burden for response tracking.

D. Exclusion / hold criteria — do not start

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

Section 2 — Order Set

Premedication

- ☐ Antipyretic (e.g., acetaminophen) and antihistamine before infusion — standard early in therapy; relax per reaction history.

Dosing schedule

Standard dose	1 mg/kg IV every 2 weeks.
Rate	Initial infusion no faster than 0.25 mg/min; escalate stepwise per label as tolerance is established (subsequent infusions may run faster).
Reaction rule	Slow or interrupt for reactions; resume at a reduced rate once resolved.

Standing orders — every infusion

- ☐ Vital signs during infusion; premedicate consistently early in therapy.
- ☐ Document reactions to inform the premedication/rate plan.

Missed or delayed dose

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

Section 3 — Follow-Up Protocol

ITEM	FREQUENCY / TIMING	ACTION / THRESHOLD
Anti–agalsidase IgG antibodies	Periodically — seroconversion is common, especially in males	High/sustained titers with waning response → specialist review.
Renal function	Every 6–12 months	Disease- and therapy-tracking.
Cardiac status	Per cardiologist schedule	Fabry cardiomyopathy surveillance.
Infusion-reaction review	Every infusion	Escalation → revise plan.

Section 4 — Pause vs. Stop Criteria

A. Pause or delay (resume when resolved)

CRITERION	REQUIRED ACTION
Moderate infusion reaction	Interrupt; resume slower.
Intercurrent acute illness	Defer as appropriate.

B. Discontinue permanently

CRITERION	REQUIRED ACTION
Anaphylaxis or severe recurrent reactions uncontrolled by premedication and rate reduction	Stop; specialist rechallenge only.

Section 5 — Notes & Practical Points

☐ Antibody-positive males with attenuated response may warrant a mechanistic switch (e.g., pegylated ERT) — a specialist decision the infusion data (reaction log, trough timing) should inform.

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.