

Naglazyme (galsulfase) — HCPCS J1458

CARECOST ESTIMATE · BILLING CHEAT SHEET

BioMarin Pharmaceutical 5 mg / 5 mL single-dose vial IV infusion ~4 hours, weekly Reviewed: May 2, 2026 ASP: Q2 2026

HCPCS J1458 1 mg = 1 unit	DOSE 1 mg/kg Weekly · 70 units (70 kg)	MODIFIER JZ/JW Both apply — 5 mg vial waste	ADMIN CPT 96365 + 96366 ×3 (4-hr)	MEDICARE ASP+6% \$513.595 /mg · \$35,952/wk (70kg)
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BOXED WARNING — ANAPHYLAXIS. Life-threatening reactions during or within 24 hr of infusion (shock, respiratory distress, angioedema, throat tightness, hypotension, bronchospasm). Premedication required. HCP admin only with resuscitation equipment immediately available.

CODES & NDC

HCPCS	J1458 — "Galsulfase injection" (permanent, eff. 1/1/2007; pre-2007 used J3490)
NDC	68135-0020-01 (10/11) — N4 qualifier
VIAL	5 mg / 5 mL (1 mg/mL) single-dose — only vial size
FDA APPROVAL	May 31, 2005 (BLA 125117) — orphan drug, ages 5+
BENEFIT	Medical (provider buy-and-bill); some payers permit specialty pharmacy

DOSING & UNIT MATH

- **1 mg/kg actual body weight IV weekly** (lifelong)
- Infusion ~4 hours per FDA label (slow titration)
- No 0.2 micron in-line filter (per label)
- Bill **actual mg administered** as units
- 52 doses/year typical (weekly)

WEIGHT	DOSE	VIALS	UNITS (DRUG+JW)
15 kg child	15 mg	3 (no waste)	15 units, JZ
22 kg child	22 mg	5 (3 mg waste)	22 + JW 3
40 kg adolescent	40 mg	8 (no waste)	40 units, JZ
70 kg adult	70 mg	14 (no waste)	70 units, JZ
73 kg adult	73 mg	15 (2 mg waste)	73 + JW 2

PREMEDICATION CHECKLIST (EVERY INFUSION)

- Antihistamine** (diphenhydramine 1 mg/kg, max 50 mg) PO/IV 30-60 min pre — bill J1200 if IV
- Antipyretic** (acetaminophen 15 mg/kg, max 1 g) PO 30-60 min pre — usually bundled
- Corticosteroid** (methylprednisolone) 30-60 min pre IF prior reaction or asthma — bill J2920/J2930
- Confirm no febrile / respiratory illness today (delay if present)
- Resuscitation equipment + epinephrine ready
- Document drug, dose, route, time, response in record

MPS TREATMENT LANDSCAPE (1 ERT PER SUBTYPE)

MPS	EPONYM	DRUG	HCPCS	DOSE
I	Hurler / Scheie	Aldurazyme	J1931	0.58 mg/kg wkly
II	Hunter	Elaprase	J1743	0.5 mg/kg wkly
IVA	Morquio A	Vimizim	J1322	2 mg/kg wkly
VI	Maroteaux-Lamy	Naglazyme	J1458	1 mg/kg wkly
VII	Sly	Mepsevii	J3397	4 mg/kg q2w

Confirm subtype FIRST. ARSB enzyme assay + ARSB gene mutation. Wrong ERT for wrong subtype = denial + no benefit.

ICD-10 — MPS VI

CODE	FOR
E76.29	Other MPS — primary code for MPS VI (no dedicated code exists)
E76.2	Other MPS (parent) — some payers accept
E76.3	MPS unspecified — avoid , lacks specificity for orphan PA
E76.0	MPS I — NOT for Naglazyme (use Aldurazyme)
E76.1	MPS II — NOT for Naglazyme (use Elaprase)

Pair with organ codes: I35.0 aortic stenosis, I50.x heart failure, J96.9x respiratory failure, H17.x corneal opacity, G91.9 hydrocephalus, R16.2 hepatosplenomegaly.

ADMINISTRATION & MODIFIERS

CODE	WHEN
96365	Therapeutic IV, 1st hour (initial)
96366 ×3	Hours 2, 3, 4 (additional hour)
96367	Sequential premed infusion (if applicable)
96375	Sequential IV push premed
96413	NOT appropriate — galsulfase is ERT, not chemo

JZ/JW required: JZ when weight is exact multiple of 5 kg (no waste); JW on separate line for any partial-vial waste. Document weight, vials drawn, mg given, mg discarded on every claim.

PAYER REQUIREMENTS (MAY 2026)

PAYER	PA	REQUIRED
UnitedHealthcare	Yes	Enzyme + gene confirmation, geneticist/metabolic Rx, baseline 6MWT & uGAG
Aetna	Yes	CPB 0729 LSD ERT criteria, specialist Rx, MPS VI manifestations documented
BCBS plans	Yes	FDA label + AAP guidelines; enzyme + gene confirmation
Medicare	MAC LCD	FDA-labeled use, appropriate dx documentation

Annual renewal: document continued benefit (uGAG, 6MWT, joint ROM, cardiopulmonary stability).

MEDICARE REIMBURSEMENT (Q2 2026)

FIELD	VALUE
ASP + 6%	\$513.595 / mg (effective 4/1 – 6/30/2026)
70 kg weekly dose (70 mg)	\$35,951.65
40 kg weekly dose (40 mg)	\$20,543.80
25 kg weekly dose (25 mg)	\$12,839.88
Annual 70 kg adult (52 doses)	~\$1,869,486
Annual 25 kg child (52 doses)	~\$667,673

SITE OF CARE

SETTING	POS	NOTES
Hospital outpatient	22	Common & preferred — safety + 4-hr duration
Metabolic / genetics center	11/22	Preferred for MPS-experienced clinicians
Ambulatory infusion suite	49	OK after tolerance established (6-12 mo)
Physician office	11	OK if MPS-experienced + emergency capability
Patient home	12	Discouraged — anaphylaxis risk + airway compromise in MPS VI

PATIENT ASSISTANCE — BIOMARIN RARECONNECTIONS

- **Phone: 1-866-906-6100** (Naglazyme Patient Support)
- **Commercial copay:** Naglazyme Copay Assistance Program (excludes Medicare/Medicaid/federal)
- **PAP:** free product for uninsured / underinsured (income-tested)
- **Foundations (Medicare):** PAN, HealthWell, NORD — verify open MPS / LSD funds quarterly
- **Web:** naglazyme.com · biomarin.com/patients

