galsulfase (gal-sul-fase)
Naglazyme

Classification
Therapeutic: replacement enzyme
Pharmacologic: enzymes

Pregnancy Category: B

Indications
Mucopolysaccharidosis VI (MPS IV).

Action
Replaces a deficient enzyme in MPS IV. Without replacement, glycosaminoglycans accumulate resulting in cell, organ and tissue dysfunction. Therapeutic Effects: improved walking and stair climbing.

Pharmacokinetics
Absorption: IV administration results in complete bioavailability.
Distribution: Widely distributed.
Metabolism and Excretion: Unknown.
Half-life: 9 min (after one week of treatment), 26 min (after 24 weeks of treatment).

TIME/ACTION PROFILE (improve exercise parameters)
ROUTE ONSET PEAK DURATION
IV unknown 24 wk unknown

Contraindications/Precautions
Contraindicated in: None.
Use Cautiously in: Febrile or respiratory illness; Pedi: Children <5 yr (safety not established); OB, Lactation: Safety not established.

Adverse Reactions/Side Effects

Interactions
Drug-Drug: None noted.

Route/Dosage
IV (Adults and Children ≥5 yr): 1 mg/kg, once weekly.

NURSING IMPLICATIONS
Assessment
● Assess for infusion reactions (fever, chills/rigors, headache, rash, mild to moderate urticaria); may occur in over half of patients. May also cause nausea, vomiting, elevated BP, retinopathy, pain, malaise, and joint pain. Severe reactions include angioneurotic edema, hypotension, dyspnea, bronchospasm, respiratory distress, anaphylaxis. May occur as late as week 75 of treatment. Patients should receive antihistamines and antipyretics prior to administration to minimize occurrence. Slowing or temporary interruption of infusion and administration of additional antihistamines, antipyretics, and occasionally corticosteroids usually stop symptoms. Most patients are able to complete infusion. Administer subsequent infusions at a slower rate additional prophelial antihistamines and, if reaction is severe, prophelial corticosteroids.

● Assess for sleep apnea, common in patients with MPS VI, prior to therapy. Administration of pretreatment antihistamines may increase risk. Supplemental oxygen and continuous positive airway pressure (CPAP) should be available during infusion.

● Assess vital signs and respiratory status prior to administration. Administration should be delayed in patients with an acute febrile or respiratory illness.

● Monitor for signs and symptoms of spinal/cervical cord compression (back pain, paraparesis, paraplegia, urinary and fecal incontinence). Manage symptomatically.

Potential Nursing Diagnoses
Activity intolerance (Indications)
Deficient knowledge, related to medication regimen (Patient/Family Teaching)

Implementation
● Pretreat with antihistamines with or without antipyretics 30–60 min prior to start of infusion.
IV Administration

• Intermittent Infusion: Diluent: 250 mL bag of 0.9% NaCl
  Determine number of vials to be diluted based on patient’s weight; round to nearest whole vial. Remove
  vials from refrigerator and allow to reach room temperature. Do not use after expiration date. Do not store
  vials in a refrigerator; store in a cool, dry place. Do not freeze. Ensure all vials are at room temperature for
  at least 24 hrs prior to dilution. Do not heat or microwave vials. Solutions are clear to slightly
  opalescent and colorless to pale yellow. A few translucent particles may be present. Do not administer solutions
  that are discolored or contain particulate matter. Withdraw and discard amount equal to galsulfase from a 250 mL bag of
  0.9% NaCl (not necessary if using 100 mL bag in patients susceptible to fluid vol-
  ume overload). Slowly withdraw calculated volume from galsulfase vials noting
  caution to avoid excessive agitation, may denserate and render inactive. Do not use
  a filter needle. Slowly add galsulfase to 0.9% NaCl, avoid agitation. Gently rotate
  infusion bag to ensure distribution, do not shake. Discard unused portions. Use
  diluted solution immediately; storage should not exceed 6 h from time of prep-
  aration to completion of administration. Use PVC containers and administer via
  PVC infusion set with an inline low protein binding 0.2 micrometer filter. NOTE:
  Administer at rate of 6 mL/hr for first hour. Infusion is well tolerated; rate may
  be increased to 80 mL/hr for next 3 hrs. Administer total volume of infusion over
  no less than 4 hrs. If 100 mL bag is used, decrease infusion rate so total volume is
  infused over at least 4 hrs. Infusion time can be extended for up to 20 hr if infusion
  reactions occur.

• Additive Incompatibility: Do not admix or administer with other products.

Patient/Family Teaching

• Inform patient that a Clinical Surveillance Program has been established to better
  understand the variability and progression of MPS VI and to evaluate long-term ef-
  fects of galsulfase. Encourage patients to participate; participation is voluntary and
  may be long term. For information visit www.MPSVI.com/CSP or call 866–906–
  6100.

Evaluation/Desired Outcomes

• Improved walking and stair climbing capacity in patients with MPS VI.

Why was this drug prescribed for your patient?