VIMIZIM® (elosulfase alfa) Fact sheet

Indication

VIMIZIM® (elosulfase alfa) is indicated for patients with Mucopolysaccharidosis type IVA (MPS IVA; Morquio A syndrome).[1]

Dosage

The recommended dosage for VIMIZIM is 2 mg/kg once every week, given intravenously over a minimum range of 3.5 to 4.5 hours, based on infusion volume. Pretreatment with antihistamines with or without antipyretics is recommended 30 to 60 minutes prior to start of infusion.[1]

How Supplied

VIMIZIM is supplied as a concentrated solution for infusion (1 mg/mL) requiring dilution. One 5-mL vial contains 5 mg of VIMIZIM.[1]

Storage

Store VIMIZIM under refrigeration at 2°C to 8°C (36°F to 46°F). Do not freeze or shake. Protect from light.

Diluted VIMIZIM should be used immediately. If immediate use is not possible, diluted VIMIZIM may be stored for up to 24 hours at 2°C to 8°C (36°F to 46°F) followed by up to 24 hours at 23°C to 27°C (73°F to 81°F) during administration. Discard any unused product.[1]

Shipping and Delivery

VIMIZIM is available directly from BioMarin. BioMarin Customer Service hours of operation are Monday through Friday, 7:00 AM to 6:00 PM (CST). To secure overnight delivery, place your order with BioMarin Customer Service by 2:00 PM (CST).

Contact BioMarin Customer Service at 1-866-274-0606 for more information on ordering VIMIZIM.

Returned Goods

VIMIZIM is a nonreturnable product, except in cases of BioMarin shipping error or product defect. BioMarin reserves the right to review return requests on a case-by-case basis. All returns require prior authorization from BioMarin. Call BioMarin Customer Service at 1-866-274-0606 to request return authorization. A copy of the BioMarin Returned Goods Policy is available upon request.

Commonly Used Billing Codes

The adjacent table contains common billing codes that may be used when filing claims for VIMIZIM. **These codes are provided for informational purposes only; please contact your individual payers to determine appropriate codes and billing requirements.**

NDC 68135-100-01

Rx only.

Commonly used billing codes–updated November 2016

ICD-10-CM[2]	E76.210	Morquio A mucopolysaccharidoses
NDC	68135-100-01	5.0 mg elosulfase alfa, per 5 mL
HCPCS[3]	J1322	Injection, elosulfase alfa, per 1.0 mg
CPT-4[4]	96365	Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); initial, up to 1 hour
	96366	Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); each additional hour (list separately in addition to code for primary procedure)
REVENUE[5]	258	Pharmacy: IV solutions
	261	IV therapy: infusion pump
	262	IV therapy: IV therapy, pharmacy services
	263	IV therapy: IV therapy/drug/supply/delivery
	631	Drugs require specific ID: single source drug
	636	Drugs require specific ID: drugs requiring detailed coding

Please see Important Safety Information, including boxed warning, on reverse.

Visit vimizim.com for more information.

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IMPORTANT SAFETY INFORMATION

Life-threatening anaphylactic reactions have occurred in some patients during VIMIZIM® (elosulfase alfa) infusions. Anaphylaxis, presenting as cough, erythema, throat tightness, urticaria, flushing, cyanosis, hypotension, rash, dyspnea, chest discomfort, and gastrointestinal symptoms (e.g., nausea, abdominal pain, retching, and vomiting) in conjunction with urticaria, have been reported to occur during VIMIZIM infusions, regardless of duration of the course of treatment. Closely observe patients during and after VIMIZIM administration and be prepared to manage anaphylaxis. Inform patients of the signs and symptoms of anaphylaxis and have them seek immediate medical care should symptoms occur. Patients with acute respiratory illness may be at risk of serious acute exacerbation of their respiratory compromise due to hypersensitivity reactions, and require additional monitoring.

Due to the potential for anaphylaxis, appropriate medical support should be readily available when VIMIZIM is administered and for an appropriate period of time following administration. In clinical trials, cases of anaphylaxis occurred as early as 30 minutes from the start of infusion and up to three hours after infusion, and as late into treatment as the 47th infusion.

In clinical trials, hypersensitivity reactions have been observed as early as 30 minutes from the start of infusion but as late as six days after infusion. Frequent symptoms of hypersensitivity reactions (occurring in more than 2 patients) included anaphylactic reactions, urticaria, peripheral edema, cough, dyspnea, and flushing.

Because of the potential for hypersensitivity reactions, administer antihistamines with or without antipyretics prior to infusion. Management of hypersensitivity reactions should be based on the severity of the reaction and include slowing or temporary interruption of the infusion and/or administration of additional antihistamines, antipyretics, and/or corticosteroids for mild reactions. However, if severe hypersensitivity reactions occur, immediately stop the infusion of VIMIZIM and initiate appropriate treatment.

Consider the risks and benefits of re-administering VIMIZIM following a severe reaction.

Patients with acute febrile or respiratory illness at the time of VIMIZIM infusion may be at higher risk of life-threatening complications from hypersensitivity reactions. Careful consideration should be given to the patient's clinical status prior to administration of VIMIZIM and consider delaying the VIMIZIM infusion.

Sleep apnea is common in MPS IVA patients. Evaluation of airway patency should be considered prior to initiation of treatment with VIMIZIM. Patients using supplemental oxygen or continuous positive airway pressure (CPAP) during sleep should have these treatments readily available during infusion in the event of an acute reaction, or extreme drowsiness/sleep induced by antihistamine use.

Spinal or cervical cord compression (SCC) is a known and serious complication of MPS IVA and may occur as part of the natural history of the disease. In clinical trials, SCC was observed both in patients receiving VIMIZIM and patients receiving placebo. Patients with MPS IVA should be monitored for signs and symptoms of SCC (including back pain, paralysis of limbs below the level of compression, urinary and fecal incontinence) and given appropriate clinical care.

All patients treated with VIMIZIM 2 mg/kg once per week in the placebo-controlled trial developed anti-drug antibodies. The relationship between the presence of neutralizing antibodies and long-term therapeutic response or occurrence of anaphylaxis or other hypersensitivity reactions could not be determined.

VIMIZIM should be used during pregnancy only if the potential benefit justifies the potential risk to the fetus. It is not known if VIMIZIM is present in human milk. Exercise caution when administering VIMIZIM to a nursing mother. There is a Morquio A Registry that collects data on pregnant women and nursing mothers with MPS IVA who are treated with VIMIZIM. Contact MARS@BMRN.com for information and enrollment.

Safety and effectiveness in pediatric patients below 5 years of age have not been established and are currently being evaluated.

In clinical trials, the most common adverse reactions (>10%) occurring during infusion included pyrexia, vomiting, headache, nausea, abdominal pain, chills, and fatigue. The acute reactions requiring intervention were managed by either temporarily interrupting or discontinuing infusion, and administering additional antihistamine, antipyretics, or corticosteroids.

To report SUSPECTED ADVERSE REACTIONS contact BioMarin Pharmaceutical Inc. at 1-866-906-6100, or FDA at 1-800-FDA-1088 or go to www.fda.gov/medwatch.

Please see accompanying full Prescribing Information, including boxed warning, or visit www.VIMIZIM.com.

References: 1. Vimizim [package insert]. Novato, CA: BioMarin Pharmaceutical Inc; 2014. **2.** Centers for Disease Control and Prevention. ICD-10-CM tabular list of diseases and injuries. ftp://ftp.cdc.gov/pub/Health_Statistics/NCHS/Publications/ICD10CM/2017/ICD10CM_FY2017_Full_PDF.zip. Accessed November 30, 2016. **3.** Centers for Medicare and Medicaid Services. HCPCS release and code sets. http://www.cms.gov/Medicare/Coding/HCPCSReleaseCodeSets/Downloads/2016-Annual-Alpha-Numeric-HCPCS-File.zip. Accessed November 30, 2016. **4.** CodeManager® cpt® code/relative value search. Chicago, IL: American Medical Association; 2016. https://ocm.ama-assn.org/OCM/CPTRelativeValueSearch.do. Accessed November 30, 2016. **5.** Centers for Medicare and Medicaid Services. Current procedural terminology, fourth edition ("CPT®"). http://www.cms.gov/apps/ama/license.asp?file=/Medicare/Medicare-Fee-for-Service-Payment/HospitalOutpatientPPS/Downloads/CMS-1613-P-Revenue-Code-Crosswalk.zip. Accessed November 30, 2016.

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