Drugs

Imiglucerase

Targets (1)

IDENTIFICATION

Name	Imiglucerase
Accession Number	DB00053 (BTD00026, BIOD00026)
Type	Biotech
Groups	Approved
Biologic Classification	Protein Based Therapies Recombinant Enzymes
Description	Human Beta-glucocerebrosidase or Beta-D-glucosyl-N-acylsphingosine glucohydrolase E.C. 3.2.1.45. 497 residue protein with N-linked carbohydrates, MW=59.3 kD. Alglucerase is prepared by modification of the oligosaccharide chains of human Beta-glucocerebrosidase. The modification alters the sugar residues at the non-reducing ends of the oligosaccharide chains of the glycoprotein so that they are predominantly terminated with mannose residues.
Protein structure	
Protein chemical formula	$C_{2532}H_{3854}N_{672}O_{711}S_{16}$
Protein average weight	55597.4 Da
Sequences	>DB00053 sequence ARPCIPKSFGYSSVVCVCNATYCDSFDPPTFPALGTFSRYESTRSGRRMELSMGPIQANH TGTGLLLTLQPEQKFQKVKGFGGAMTDAAALNILALSPPAQNLLLKSYFSEEGIGYNIIR VPMASCDFSIRTYTYADTPDDFQLHNFSLPEEDTKLKIPLIHRALQLAQRPVSLLASPWT SPTWLKTNGAVNGKGSLKGQPGDIYHQTWARYFVKFLDAYAEHKLQFWAVTAENEPSAGL LSGYPFQCLGFTPEHQRDFIARDLGPTLANSTHHNVRLLMLDDQRLLLPHWAKVVLTDPE AAKYVHGIAVHWYLDFLAPAKATLGETHRLFPNTMLFASEACVGSKFWEQSVRLGSWDRG MQYSHSIITNLLYHVVGWTDWNLALNPEGGPNWVRNFVDSPIIVDITKDTFYKQPMFYHL GHFSKFIPEGSQRVGLVASQKNDLDAVALMHPDGSAVVVVLNRSSKDVPLTIKDPAVGFL ETISPGYSIHTYLWRRQ Download FASTA Format
Synonyms	Imiglucerasa
External IDs	ISU-302

Prescription Products

Show 10 entries Search

NAME	DOSAGE	STRENGTH	ROUTE	LABELLER	MARKETING START	MARKETING END	
Cerezyme	Powder, for	400 unit	Intravenous	Sanofi	2000-06-14	Not applicable	

NAME	DOSAGE	STRENGTH	ROUTE	LABELLER	MARKETING START	MARKETING END	
Cerezyme	Injection, powder, lyophilized, for solution	40 U/1mL	Intravenous	Genzyme Corporation	1994-05-23	Not applicable	
Cerezyme	Injection, powder, lyophilized, for solution	40 U/1mL	Intravenous	Genzyme Corporation	1994-05-23	Not applicable	
Cerezyme	Powder, for solution	200 unit	Intravenous	Sanofi Genzyme, a Division of Sanofi Aventis Canada Inc	1997-05-01	2016-08-04	

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Categories

- Alimentary Tract and Metabolism
- Enzyme Replacement Therapy
- Enzymes
- Enzymes and Coenzymes
- Gaucher Disease
- Glucosidases
- Glycoside Hydrolases
- Hydrolases
- Hydrolytic Lysosomal Glucocerebroside-specific Enzyme
- Recombinant Proteins

UNII Q6U6J48BWY

CAS number 154248-97-2

PHARMACOLOGY

Indication For the treatment of Gaucher's disease (deficiency in glucocerebrosidase)

Associated Conditions

- Gaucher's Disease Type 1
- Type 3 Gaucher disease

Pharmacodynamics Gaucher disease is characterized by a functional deficiency in Beta-glucocerebrosidase enzymatic activity and the resultant accumulation of lipid glucocerebroside in tissue macrophages which become engorged and are termed Gaucher cells. Gaucher cells are typically found in liver, spleen and bone marrow. This can lead to an enlarged spleen and liver (hepatosplenomegaly). Secondary hematologic sequelae include severe anemia and thrombocytopenia. Injections of imiglucerase into Gaucher disease patients leads to elevated serum levels of the enzyme and reduction in the accumulation of glucocerebroside leading to reduced anemia and thrombocytopenia, reduced spleen and liver size, and decreased cachexia

Mechanism of action Imiglucerase catalyzes the hydrolysis of the glycolipid, glucocerebroside, to glucose and ceramide as part of the normal degradation pathway for membrane lipids.

TARGET	ACTIONS	ORGANISM
(A) Glucocerebroside	other/unknown	Humans

Absorption Not Available

Volume of distribution

- 0.09 to 0.15 L/kg

Protein binding Not Available

Metabolism Not Available

Route of	Not Available

Half life	3.6-10.4 min
Clearance	• 14.5 +/- 4.0 mL/min/kg
Toxicity	Not Available
Affected organisms	Humans and other mammals
Pathways	Not Available
Pharmacogenomic Effects/ADRs	Not Available

INTERACTIONS

Drug Interactions	No interactions found.
Food Interactions	Not Available

REFERENCES

General References

1. Pastores GM, Hughes DA: Gaucher Disease . [PubMed:20301446]

External Links	UniProt	P04062
	Genbank	M16328
	KEGG Drug	D02810
	PubChem Substance	46508744
	ChEMBL	CHEMBL1201632
	PharmGKB	PA164742934
	RxList	RxList Drug Page
	Drugs.com	Drugs.com Drug Page
	Wikipedia	Imiglucerase

ATC Codes

A16AB02 — Imiglucerase
- A16AB — Enzymes
- A16A — OTHER ALIMENTARY TRACT AND METABOLISM PRODUCTS
- A16 — OTHER ALIMENTARY TRACT AND METABOLISM PRODUCTS
- A — ALIMENTARY TRACT AND METABOLISM

AHFS Codes

44:00.00 — Enzymes

FDA label

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CLINICAL TRIALS

Clinical Trials

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PHASE	STATUS	PURPOSE	CONDITIONS	COUNT

PHASE	STATUS	PURPOSE	CONDITIONS	COUNT
2	Unknown	Treatment	Gaucher's Disease	1
3	Recruiting	Treatment	Gaucher's Disease Type 1 / Gaucher's disease type III	1
3	Withdrawn	Not Available	Gaucher's Disease	1
4	Completed	Treatment	Cerebroside Lipidosis Syndrome / Clucocerebrosidase Deficiency Disease / Gaucher Disease, Non-Neuronopathic Form / Gaucher's Disease Type 1 / Glucosylceramide Beta-Glucosidase Deficiency Disease	1
4	Completed	Treatment	Cerebroside Lipidosis Syndrome / Gaucher Disease, Non-Neuronopathic Form / Gaucher Disease, Type 1 / Glucocerebrosidase Deficiency Disease / Glucosylceramide Beta-Glucosidase Deficiency Disease	1
Not Available	Completed	Not Available	Gaucher's Disease	1

Drugs

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PHARMACOECONOMICS

Manufacturers Genzyme corp

Packagers Genzyme Inc.

Dosage forms

Show 10 entries Search

FORM	ROUTE	STRENGTH
Injection, powder, lyophilized, for solution	Intravenous	40 U/1mL
Powder, for solution	Intravenous	200 unit
Powder, for solution	Intravenous	400 unit

Showing 1 to 3 of 3 entries ‹ 1 ›

Prices

Show 10 entries Search

UNIT DESCRIPTION	COST	UNIT
Cerezyme 400 unit vial	1903.2USD	vial
Cerezyme 200 unit vial	951.6USD	vial
Ceredase 80 unit/ml vial	380.64USD	ml

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Patents

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PATENT NUMBER	PEDIATRIC EXTENSION	APPROVED	EXPIRES (ESTIMATED)	
US5549892	No	1996-08-27	2013-08-27	

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PROPERTIES

State Liquid

Experimental Properties

PROPERTY	VALUE	SOURCE
hydrophobicity	-0.168	Not Available

PROPERTY	VALUE	SOURCE
isoelectric point	7.41	Not Available

TAXONOMY

Description	Not Available
Kingdom	Organic Compounds
Super Class	Organic Acids
Class	Carboxylic Acids and Derivatives
Sub Class	Amino Acids, Peptides, and Analogues
Direct Parent	Peptides
Alternative Parents	Not Available
Substituents	Not Available
Molecular Framework	Not Available
External Descriptors	Not Available

TARGETS

1. Glucocerebroside

Kind
Small molecule

Organism
Humans

Pharmacological action
Yes

Actions
Other/unknown

References

1. Pastores GM, Hughes DA: Gaucher Disease . [PubMed:20301446]

Drug created on June 13, 2005 07:24 / Updated on April 13, 2019 20:58

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