abcam

Product datasheet

HRP Anti-GBA antibody [EPR5142] ab200856

KO VALIDATED Recombinant RabMAb

3 Images

Overview

Product name	HRP Anti-GBA antibody [EPR5142]
Description	HRP Rabbit monoclonal [EPR5142] to GBA
Host species	Rabbit
Conjugation	HRP
Tested applications	**Suitable for:** WB
Species reactivity	**Reacts with:** Human
Immunogen	Synthetic peptide. This information is proprietary to Abcam and/or its suppliers.
Positive control	WB: Saos-2 and SH-SY5Y whole cell lysates.
General notes	This product is a recombinant monoclonal antibody, which offers several advantages including: - High batch-to-batch consistency and reproducibility - Improved sensitivity and specificity - Long-term security of supply - Animal-free production For more information **see here**. Our RabMAb® technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to **RabMAb® patents**.

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C. Avoid freeze / thaw cycle. Store In the Dark.
Storage buffer	pH: 7.40 Preservative: 0.1% Proclin 300 Solution Constituents: 30% Glycerol (glycerin, glycerine), 1% BSA, PBS
Purity	Protein A purified
Clonality	Monoclonal
Clone number	EPR5142
Isotype	IgG

Applications

The Abpromise guarantee Our **Abpromise guarantee** covers the use of ab200856 in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
WB		1/5000. Detects a band of approximately 60 kDa (predicted molecular weight: 60 kDa).

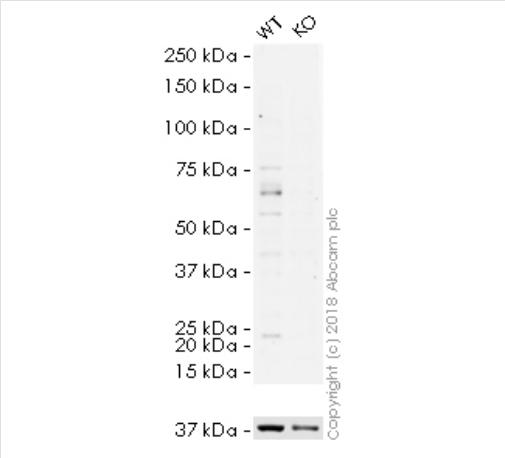
Target

Involvement in disease

Defects in GBA are the cause of Gaucher disease (GD) [MIM:230800]; also known as glucocerebrosidase deficiency. GD is the most prevalent lysosomal storage disease, characterized by accumulation of glucosylceramide in the reticulo-endothelial system. Different clinical forms are recognized depending on the presence (neuronopathic forms) or absence of central nervous system involvement, severity and age of onset.
Defects in GBA are the cause of Gaucher disease type 1 (GD1) [MIM:230800]; also known as adult non-neuronopathic Gaucher disease. GD1 is characterized by hepatosplenomegaly with consequent anemia and thrombopenia, and bone involvement. The central nervous system is not involved.
Defects in GBA are the cause of Gaucher disease type 2 (GD2) [MIM:230900]; also known as acute neuronopathic Gaucher disease. GD2 is the most severe form and is universally progressive and fatal. It manifests soon after birth, with death generally occurring before patients reach two years of age.
Defects in GBA are the cause of Gaucher disease type 3 (GD3) [MIM:231000]; also known as subacute neuronopathic Gaucher disease. GD3 has central nervous manifestations.
Defects in GBA are the cause of Gaucher disease type 3C (GD3C) [MIM:231005]; also known as pseudo-Gaucher disease or Gaucher-like disease.
Defects in GBA are the cause of Gaucher disease perinatal lethal (GDPL) [MIM:608013]. It is a distinct form of Gaucher disease type 2, characterized by fetal onset. Hydrops fetalis, in utero fetal death and neonatal distress are prominent features. When hydrops is absent, neurologic involvement begins in the first week and leads to death within 3 months. Hepatosplenomegaly is a major sign, and is associated with ichthyosis, arthrogryposis, and facial dysmorphism.
Note=Perinatal lethal Gaucher disease is associated with non-immune hydrops fetalis, a generalized edema of the fetus with fluid accumulation in the body cavities due to non-immune causes. Non-immune hydrops fetalis is not a diagnosis in itself but a symptom, a feature of many genetic disorders, and the end-stage of a wide variety of disorders.
Defects in GBA contribute to susceptibility to Parkinson disease (PARK) [MIM:168600]. A complex neurodegenerative disorder characterized by bradykinesia, resting tremor, muscular rigidity and postural instability. Additional features are characteristic postural abnormalities, dysautonomia, dystonic cramps, and dementia. The pathology of Parkinson disease involves the loss of dopaminergic neurons in the substantia nigra and the presence of Lewy bodies (intraneuronal accumulations of aggregated proteins), in surviving neurons in various areas of the brain. The disease is progressive and usually manifests after the age of 50 years, although early-onset cases (before 50 years) are known. The majority of the cases are sporadic suggesting a multifactorial etiology based on environmental and genetic factors. However, some patients present with a positive family history for the disease. Familial forms of the disease usually begin

	at earlier ages and are associated with atypical clinical features.
Sequence similarities	Belongs to the glycosyl hydrolase 30 family.
Cellular localization	Lysosome membrane. Interaction with saposin-C promotes membrane association.

Images



Western blot - HRP Anti-GBA antibody [EPR5142] (ab200856)

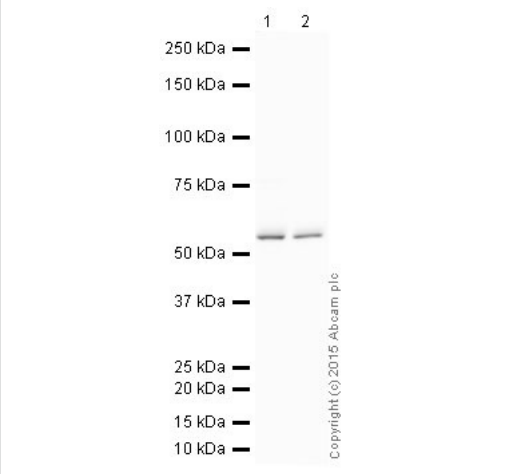
All lanes : HRP Anti-GBA antibody [EPR5142] (ab200856) at 1/10000 dilution

Lane 1 : Wild-type HAP1 whole cell lysate
Lane 2 : GBA knockout HAP1 whole cell lysate

Lysates/proteins at 20 µg per lane.

Predicted band size: 60 kDa

ab200856 was shown to specifically react with GBA in wild-type HAP1 cells as signal was lost in GBA knockout cells. Wild-type and GBA knockout samples were subjected to SDS-PAGE. Ab200856 and **ab184095** (Mouse monoclonal [mAbcam 9484] to GAPDH - Loading Control (Alexa Fluor® 680) loading control) were incubated overnight at 4°C at 10000 dilution and 1/1000 dilution respectively. The loading control was imaged using the Licor Odyssey CLx prior to blots being developed with ECL technique.



Western blot - HRP Anti-GBA antibody [EPR5142] (ab200856)

All lanes : HRP Anti-GBA antibody [EPR5142] (ab200856) at 1/5000 dilution

Lane 1 : Saos 2 (Human epithelial-like osteosarcoma cell line) Whole Cell Lysate
Lane 2 : SHSY-5Y (Human neuroblastoma cell line) Whole Cell Lysate

Lysates/proteins at 10 µg per lane.

Developed using the ECL technique.

Performed under reducing conditions.

Predicted band size: 60 kDa
Observed band size: 60 kDa

Exposure time: 1 minute

This blot was produced using a 4-12% Bis-tris gel under the MOPS buffer system. The gel was run at 200V for 50 minutes before being transferred onto a Nitrocellulose membrane at 30V for 70 minutes. The membrane was then blocked for an hour using 3% milk before being incubated with ab200856 overnight at 4°C. Antibody binding was visualised using ECL development solution **ab133406**.

HRP Anti-GBA antibody [EPR5142] (ab200856)

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