abcam

Product datasheet

Anti-GBA antibody ab96246

画像数 3

製品の概要

製品名	Anti-GBA antibody
製品の詳細	Rabbit polyclonal to GBA
由来種	Rabbit
アプリケーション	**適用あり:** WB, IHC-P, ICC/IF
種交差性	**交差種:** Human **交差が予測される動物種:** Mouse, Rat, Cow, Pig, Chimpanzee
免疫原	Recombinant fragment corresponding to Human GBA aa 350 to the C-terminus. Database link: **P04062**
ポジティブ・コントロール	HepG2 cell lysate
特記事項	The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing. If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As

製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
バッファー	pH: 7.00 Preservative: 0.025% Proclin 300 Constituents: 79% PBS, 20% Glycerol (glycerin, glycerine)
精製度	Immunogen affinity purified
ポリ/モノ	ポリクローナル
アイソタイプ	IgG

アプリケーション

The Abpromise guarantee **Abpromise保証は、** **次のテスト済みアプリケーションにおけるab96246の使用に適用されます**

アプリケーションノートには、推奨の開始希釈率がありますが、適切な希釈率につきましてはご検討ください。

アプリケーション	Abreviews	特記事項
WB		1/500 - 1/3000. Predicted molecular weight: 60 kDa.
IHC-P		1/100 - 1/1000.
ICC/IF		1/100 - 1/1000.

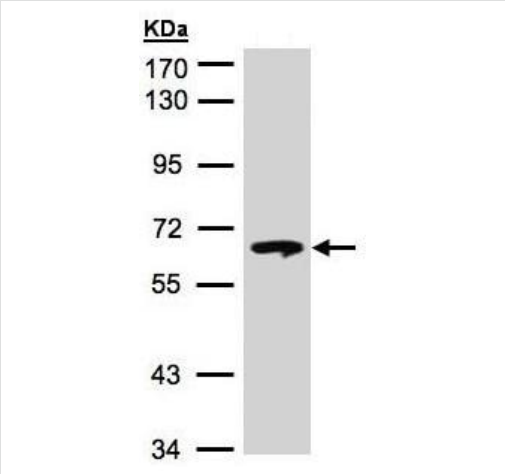
ターゲット情報

関連疾患

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.
配列類似性	Belongs to the glycosyl hydrolase 30 family.
細胞内局在	Lysosome membrane. Interaction with saposin-C promotes membrane association.

画像

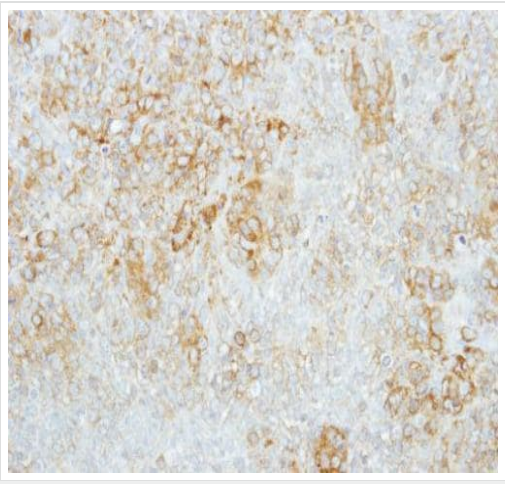


Western blot - Anti-GBA antibody (ab96246)

Anti-GBA antibody (ab96246) at 1/500 dilution + HepG2 whole cell lysate at 30 µg

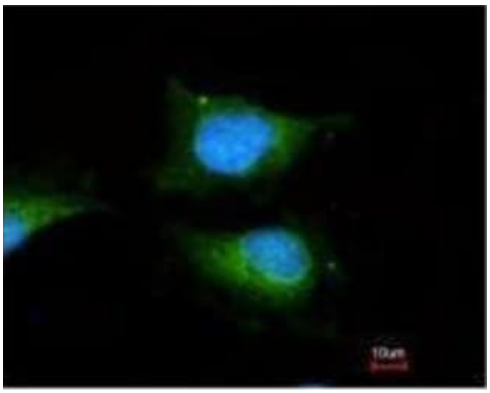
Predicted band size: 60 kDa

7.5% SDS PAGE



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-GBA antibody (ab96246)

Immunohistochemical analysis of paraffin-embedded human lung cancer, using ab96246 at 1:100 dilution.



Immunocytochemistry/ Immunofluorescence - Anti-GBA antibody (ab96246)

Immunofluorescence analysis of methanol-fixed HeLa, using ab96246 at 1:100 dilution. Counterstained with Hoechst 33342 (blue)

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