abcam

Product datasheet

Anti-GBE1 antibody ab103133

1 Abreviews　画像数 2

製品の概要

製品名	Anti-GBE1 antibody
製品の詳細	Rabbit polyclonal to GBE1
アプリケーション	**適用あり:** WB
種交差性	**交差種:** Mouse, Human **交差が予測される動物種:** Horse
免疫原	Synthetic peptide conjugated to KLH, corresponding to a region within internal sequence amino acids 534-564 of Human GBE1.
ポジティブ・コントロール	HL-60 cell lysate; Mouse liver tissue lysate

製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Store at 4°C (up to 6 months). Store at -20°C long term.
バッファー	Preservative: 0.09% Sodium Azide Constituents: PBS
精製度	Immunogen affinity purified
特記事項（精製）	ab103133 is purified through a protein A column, followed by peptide affinity purification.
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

Our Abpromise guarantee covers the use of **ab103133** in the following tested applications.

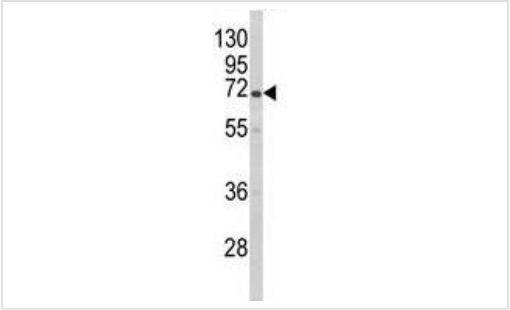
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

アプリケーション	Abreviews	特記事項
WB		1/100 - 1/500. Predicted molecular weight: 80 kDa.

ターゲット情報

機能	Required for sufficient glycogen accumulation. The alpha 1-6 branches of glycogen play an important role in increasing the solubility of the molecule and, consequently, in reducing the osmotic pressure within cells.
組織特異性	Highest levels found in liver and muscle.
パスウェイ	Glycan biosynthesis; glycogen biosynthesis.
関連疾患	Defects in GBE1 are the cause of glycogen storage disease type 4 (GSD4) [MIM:232500]; also known as Andersen disease. GSD4 is a metabolic disorder characterized by the accumulation of an amylopectin-like polysaccharide. The typical clinical manifestation is liver disease of childhood, progressing to lethal hepatic cirrhosis. Most children with this condition die before two years of age. However, the liver disease is not always progressive. No treatment apart from liver transplantation has been found to prevent progression of the disease. There is also a neuromuscular form of GSD4 that varies in onset (perinatal, congenital, juvenile, or adult) and severity. Note=Neuromuscular perinatal glycogen storage disease type 4 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 GBE1 are the cause of adult polyglucosan body disease (APBD) [MIM:263570]. APBD is a late-onset, slowly progressive disorder affecting the central and peripheral nervous systems. Patients typically present after age 40 years with a variable combination of cognitive impairment, pyramidal tetraparesis, peripheral neuropathy, and neurogenic bladder. Other manifestations include cerebellar dysfunction and extrapyramidal signs. The pathologic hallmark of APBD is the widespread accumulation of round, intracellular polyglucosan bodies throughout the nervous system, which are confined to neuronal and astrocytic processes.
配列類似性	Belongs to the glycosyl hydrolase 13 family.

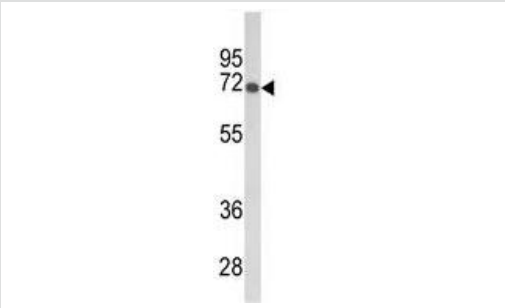
画像



Western blot - GBE1 antibody (ab103133)

Anti-GBE1 antibody (ab103133) at 1/100 dilution + HL-60 cell lysate at 35 µg

Predicted band size : 80 kDa



Western blot - GBE1 antibody (ab103133)

Anti-GBE1 antibody (ab103133) at 1/100 dilution + Mouse liver tissue lysate at 35 µg

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