

Biotin Anti-GBA antibody [EPR5142] ab201496

リコンビナント **RabMAb**

画像数 2

製品の概要

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|--------------|---|
| 製品名 | Biotin Anti-GBA antibody [EPR5142] |
| 製品の詳細 | Biotin Rabbit monoclonal [EPR5142] to GBA |
| 由来種 | Rabbit |
| 標識 | Biotin |
| アプリケーション | 適用あり: IHC-P |
| 種交差性 | 交差種: Human |
| 免疫原 | Synthetic peptide. This information is proprietary to Abcam and/or its suppliers. |
| ポジティブ・コントロール | IHC-P: Normal human thyroid tissue. |
| 特記事項 | <p>This product is a recombinant monoclonal antibody, which offers several advantages including:</p> <ul style="list-style-type: none"> - High batch-to-batch consistency and reproducibility - Improved sensitivity and specificity - Long-term security of supply - Animal-free production <p>For more information see here.</p> <p>Our RabMAb[®] technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to RabMAb[®] patents.</p> |

製品の特性

| | |
|--------|--|
| 製品の状態 | Liquid |
| 保存方法 | 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. |
| バッファー | <p>pH: 7.40</p> <p>Preservative: 0.02% Sodium azide</p> <p>Constituents: 30% Glycerol (glycerin, glycerine), 1% BSA, PBS</p> |
| 精製度 | Protein A purified |
| ポリ/モノ | モノクローナル |
| クローン名 | EPR5142 |
| 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ab201496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 |
|----------|-----------|--|
| IHC-P | | 1/50 - 1/100. Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol. |

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.

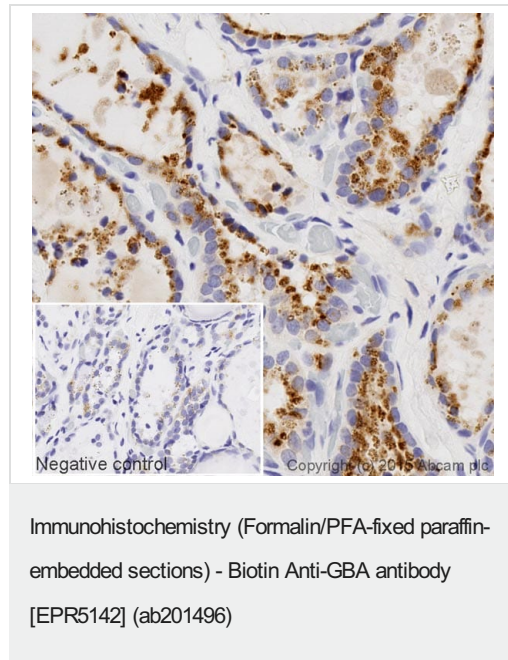
配列類似性

Belongs to the glycosyl hydrolase 30 family.

細胞内局在

Lysosome membrane. Interaction with saposin-C promotes membrane association.

画像







IHC image of GBA staining in a section of formalin-fixed paraffin-embedded normal human thyroid*, performed on a Leica Bond system using the standard protocol B. The section was pre-treated using heat mediated antigen retrieval with sodium citrate buffer (pH6, epitope retrieval solution 1) for 20 mins. The section was then incubated with ab201496 at 1/50 dilution, for 15 mins at room temperature and detected using an HRP conjugated ABC system. DAB was used as the chromogen. The section was then counterstained with haematoxylin and mounted with DPX.

The inset negative control image is taken from an identical assay without primary antibody.

For other IHC staining systems (automated and non-automated) customers should optimize variable parameters such as antigen retrieval conditions, primary antibody concentration and antibody incubation times.

*Tissue obtained from the Human Research Tissue Bank, supported by the NIHR Cambridge Biomedical Research Centre.

Why choose a recombinant antibody?

- **Research with confidence**
Consistent and reproducible results
- **Long-term and scalable supply**
Recombinant technology
- **Success from the first experiment**
Confirmed specificity
- **Ethical standards compliant**
Animal-free production

Biotin Anti-GBA antibody [EPR5142] (ab201496)

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