abcam

Product datasheet

Anti-GAA antibody ab102815

★★★★★ 1 Abreviews　画像数 2

製品の概要

製品名	Anti-GAA antibody
製品の詳細	Rabbit polyclonal to GAA
由来種	Rabbit
アプリケーション	**適用あり:** WB
種交差性	**交差種:** Mouse, Human
免疫原	Recombinant full length protein within Human GAA aa 1-1000. The exact immunogen sequence used to generate this antibody is proprietary information. If additional detail on the immunogen is needed to determine the suitability of the antibody for your needs, please **contact** our Scientific Support team to discuss your requirements.
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. Store at -20°C or -80°C. Avoid freeze / thaw cycle.
バッファー	pH: 7.4 Constituent: PBS
精製度	Protein A 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ab102815の使用に適用されます**

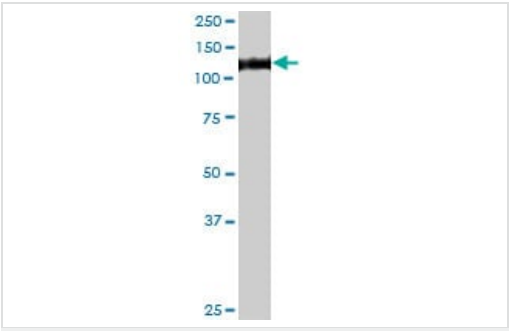
アプリケーションノートには、推奨の開始希釈率がありますが、適切な希釈率につきましてはご検討ください。

アプリケーション	Abreviews	特記事項
WB	★★★★★ (1)	1/500 - 1/1000. Predicted molecular weight: 105 kDa.

ターゲット情報

機能	Essential for the degradation of glygogen to glucose in lysosomes.
関連疾患	Defects in GAA are the cause of glycogen storage disease type 2 (GSD2) [MIM:232300]; also called acid alpha-glucosidase (GAA) deficiency or acid maltase deficiency (AMD). GSD2 is a metabolic disorder with a broad clinical spectrum. The severe infantile form, or Pompe disease, presents at birth with massive accumulation of glycogen in muscle, heart and liver. Cardiomyopathy and muscular hypotonia are the cardinal features of this form whose life expectancy is less than two years. The juvenile and adult forms present as limb-girdle muscular dystrophy beginning in the lower limbs. Final outcome depends on respiratory muscle failure. Patients with the adult form can be free of clinical symptoms for most of their life but finally develop a slowly progressive myopathy.
配列類似性	Belongs to the glycosyl hydrolase 31 family. Contains 1 P-type (trefoil) domain.
翻訳後修飾	The different forms of acid glucosidase are obtained by proteolytic processing. Phosphorylation of mannose residues ensures efficient transport of the enzyme to the lysosomes via the mannose 6-phosphate receptor.
細胞内局在	Lysosome. Lysosome membrane.

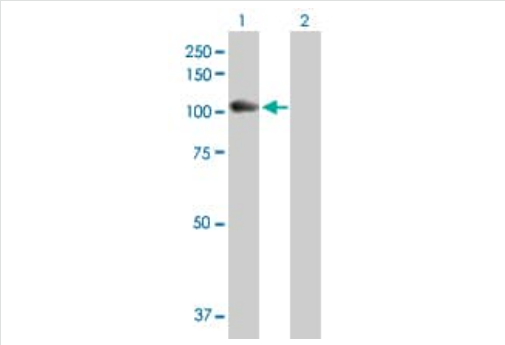
画像



Western blot - Anti-GAA antibody (ab102815)

Anti-GAA antibody (ab102815) at 1/500 dilution + Mouse intestine lysate at 50 µg

Predicted band size: 105 kDa



Western blot - Anti-GAA antibody (ab102815)

All lanes : Anti-GAA antibody (ab102815) at 1/500 dilution

Lane 1 : GAA transfected 293T cell line

Lane 2 : Non transfected 293T cell line

Lysates/proteins at 25 µg per lane.

Predicted band size: 105 kDa

Our Abpromise to you: Quality guaranteed and expert technical support

- Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours
- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit **https://www.abcam.co.jp/abpromise** or contact our technical team.

Terms and conditions

- Guarantee only valid for products bought direct from Abcam or one of our authorized distributors