Product datasheet

Human Acid sphingomyelinase Antibody Pair - BSA and Azide free (SMPD1) ab253462

重组 RabMAb

2 图像

概述

产品名称	人Acid sphingomyelinase抗体Pair - BSA and Azide free (SMPD1)
检测类型	ELISA set
范围	125 pg/ml - 8000 pg/ml
种属反应性	**与反应:** Human
产品概述	Both capture and detector antibodies are **recombinant rabbit monoclonal antibodies** delivering consistent, specific, and sensitive results. The Antibody Pair can be used to quantify Human Acid sphingomyelinase. BSA and Azide free antibody pairs include **unconjugated** capture and detector antibodies suitable for sandwich ELISAs. The antibodies are provided at an approximate concentration of 1 mg/ml as measured by the protein A280 method. The recommended antibody orientation is based on internal optimization for ELISA-based assays. Antibody orientation is assay dependent and needs to be optimized for each assay type. For additional information on the performance of the antibody pair, see the equivalent SimpleStep ELISA® Kit (**ab277075**), which uses the same antibodies. However, due to differences in their formulation, this antibody pair cannot be used with the consumables provided with our SimpleStep ELISA Kits. Please note that the range provided for the pairs is only an estimation based on the performance of the related product using the same antibody pair. Performance of the antibody pair will depend on the specific characteristics of your assay. We guarantee the product works in sandwich ELISA, but we do not guarantee the sensitivity or dynamic range of the antibody pair in your assay. Download SDS **here**.
经测试应用	**适用于:** Sandwich ELISA
平台	Reagents

性能

存放说明	Store at +4°C. Please refer to protocols.
无载体	是

组件	10 x 96 tests
Human Acid sphingomyelinase Capture Antibody (unconjugated)	1 x 100µg
Human Acid sphingomyelinase Detector Antibody (unconjugated)	1 x 100µg

功能	Converts sphingomyelin to ceramide. Also has phospholipase C activities toward 1,2-diacylglycerolphosphocholine and 1,2-diacylglycerolphosphoglycerol. Isoform 2 and isoform 3 have lost catalytic activity.
疾病相关	Defects in SMPD1 are the cause of Niemann-Pick disease type A (NPDA) [MIM:257200]; also known as Niemann-Pick disease classical infantile form. It is an early-onset lysosomal storage disorder caused by failure to hydrolyze sphingomyelin to ceramide. It results in the accumulation of sphingomyelin and other metabolically related lipids in reticuloendothelial and other cell types throughout the body, leading to cell death. Niemann-Pick disease type A is a primarily neurodegenerative disorder characterized by onset within the first year of life, mental retardation, digestive disorders, failure to thrive, major hepatosplenomegaly, and severe neurologic symptoms. The severe neurological disorders and pulmonary infections lead to an early death, often around the age of four. Clinical features are variable. A phenotypic continuum exists between type A (basic neurovisceral) and type B (purely visceral) forms of Niemann-Pick disease, and the intermediate types encompass a cluster of variants combining clinical features of both types A and B. Defects in SMPD1 are the cause of Niemann-Pick disease type B (NPDB) [MIM:607616]; also known as Niemann-Pick disease visceral form. It is a late-onset lysosomal storage disorder caused by failure to hydrolyze sphingomyelin to ceramide. It results in the accumulation of sphingomyelin and other metabolically related lipids in reticuloendothelial and other cell types throughout the body, leading to cell death. Clinical signs involve only visceral organs. The most constant sign is hepatosplenomegaly which can be associated with pulmonary symptoms. Patients remain free of neurologic manifestations. However, a phenotypic continuum exists between type A (basic neurovisceral) and type B (purely visceral) forms of Niemann-Pick disease, and the intermediate types encompass a cluster of variants combining clinical features of both types A and B. In Niemann-Pick disease type B, onset of the first symptoms occurs in early childhood and patients can survive into adulthood.
序列相似性	Belongs to the acid sphingomyelinase family. Contains 1 saposin B-type domain.
细胞定位	Lysosome.

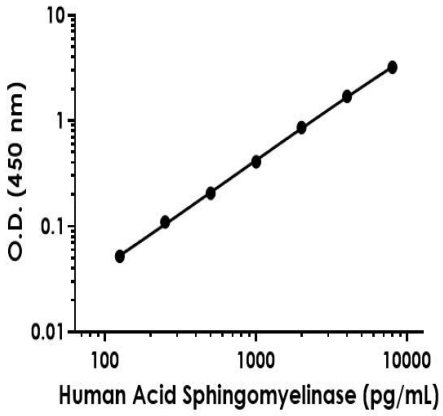
应用

The Abpromise guarantee Abpromise™承诺保证使用ab253462于以下的经测试应用

"应用说明"部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

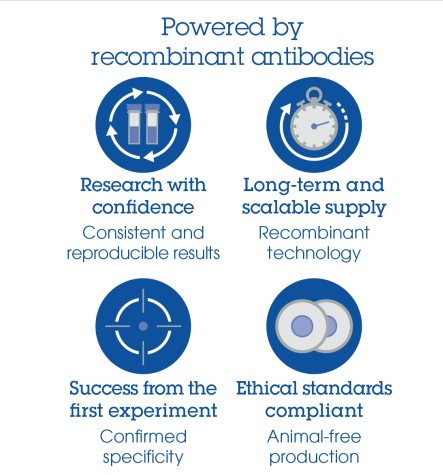
应用	Ab评论	说明
Sandwich ELISA		Use at an assay dependent concentration.

图片



Sandwich ELISA - Human Acid sphingomyelinase Antibody Pair - BSA and Azide free (SMPD1) (ab253462)

Representative standard curve from corresponding SimpleStep ELISA® Kit (**ab277075**), which uses the same antibody pair. For additional information on the performance of pair and kit, refer to the corresponding kit datasheet. Due to differences in the formulation and format of the antibodies in this pair, they cannot be used as substitutes for the antibody components in our SimpleStep ELISA® Kits.



Sandwich ELISA - Human Acid sphingomyelinase Antibody Pair - BSA and Azide free (SMPD1) (ab253462)

To learn more about the advantages of recombinant antibodies see **here**.

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