

Product datasheet

Anti-beta glucuronidase (GUSB) antibody ab88567

2 图像

概述

产品名称	Anti-beta glucuronidase (GUSB)抗体
描述	小鼠多克隆抗体to beta glucuronidase (GUSB)
宿主	Mouse
经测试应用	适用于: WB
种属反应性	与反应: Human
免疫原	Full length Human beta glucuronidase (GUSB) protein (NP_000172.1)
阳性对照	Human liver tissue lysate and beta glucuronidase (GUSB) transfected 293T cell lysate.

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
存储溶液	Preservative: None Constituents: 1X PBS, pH 7.2
纯度	Protein A purified
克隆	多克隆
同种型	IgG

应用

Our [Abpromise guarantee](#) covers the use of **ab88567** in the following tested applications.

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

应用	Ab评论	说明
WB		Use a concentration of 1 µg/ml. Predicted molecular weight: 75 kDa.

靶标

功能	Plays an important role in the degradation of dermatan and keratan sulfates.
疾病相关	Defects in GUSB are the cause of mucopolysaccharidosis type 7 (MPS7) [MIM:253220]; also known as Sly syndrome. MPS7 is an autosomal recessive lysosomal storage disease characterized by inability to degrade glucuronic acid-containing glycosaminoglycans. The phenotype is highly variable, ranging from severe lethal hydrops fetalis to mild forms with survival into adulthood. Most patients with the intermediate phenotype show hepatomegaly, skeletal anomalies, coarse facies, and variable degrees of mental impairment. Note=Mucopolysaccharidosis type 7 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.
序列相似性	Belongs to the glycosyl hydrolase 2 family.
翻译后修饰	N-linked glycosylated with 3 to 4 oligosaccharide chains.
细胞定位	Lysosome.

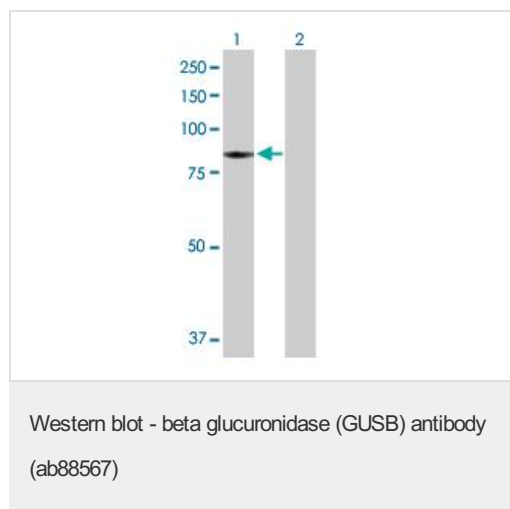
图片



Anti-beta glucuronidase (GUSB) antibody (ab88567) at 1 µg/ml + human liver tissue lysate at 50 µg

Predicted band size: 75 kDa

Observed band size: 60 kDa



All lanes : Anti-beta glucuronidase (GUSB) antibody (ab88567) at 1 µg/ml

Lane 1 : beta glucuronidase (GUSB) transfected 293T cell lysate

Lane 2 : Non transfected 293T cell lysate

Lysates/proteins at 25 µg per lane.

Predicted band size: 75 kDa

Observed band size: 85 kDa

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