abcam

Product datasheet

# Anti-GCH1 antibody [EPR27331-30] ab307507

重组 RabMAb

13 图像

## 概述

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| **产品名称** | Anti-GCH1抗体[EPR27331-30] |
| **描述** | 兔单克隆抗体[EPR27331-30] to GCH1 |
| **宿主** | Rabbit |
| **经测试应用** | **适用于:** WB, IHC-P, IP<br>**不适用于:** Flow Cyt (Intra),ICC/IF or IHC-Fr |
| **种属反应性** | **与反应:** Mouse, Rat, Human |
| **免疫原** | Synthetic peptide. This information is proprietary to Abcam and/or its suppliers. |
| **阳性对照** | WB: Mouse liver, Mouse hypothalamus, Rat liver, Rat hypothalamus, SH-SY5Y, Neuro-2a, N9 and PC-12 lysates. IHC-P: Human colon, Human pancreas, Mouse midbrain, Mouse stomach, Rat colon and Rat pancreas tissues. IP: Neuro-2a cell. |
| **常规说明** | This product is a recombinant monoclonal antibody, which offers several advantages including:<br>- High batch-to-batch consistency and reproducibility<br>- Improved sensitivity and specificity<br>- Long-term security of supply<br>- Animal-free production<br>For more information **see here**.<br>Our RabMAb® technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to **RabMAb® patents**. |

## 性能

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| **形式** | Liquid |
| **存放说明** | Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term. Avoid freeze / thaw cycle. |
| **存储溶液** | pH: 7.2<br>Preservative: 0.01% Sodium azide<br>Constituents: 40% Glycerol (glycerin, glycerine), 0.05% BSA, 59% PBS |
| **纯度** | Protein A purified |
| **克隆** | 单克隆 |
| **克隆编号** | EPR27331-30 |

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|---|---|
| **同种型** | IgG |

## 应用

**The Abpromise guarantee** **Abpromise™承诺保证使用ab307507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 |
|---|---|---|
| **WB** | | 1/1000. Predicted molecular weight: 28 kDa. |
| **IHC-P** | | 1/500 - 1/1000. Perform heat mediated antigen retrieval with Tris/EDTA buffer pH 9.0 before commencing with IHC staining protocol. |
| **IP** | | 1/30. |

**应用说明** Is unsuitable for Flow Cyt (Intra),ICC/IF or IHC-Fr.

## 靶标

**功能** Positively regulates nitric oxide synthesis in umbilical vein endothelial cells (HUVECs). May be involved in dopamine synthesis. May modify pain sensitivity and persistence. Isoform GCH-1 is the functional enzyme, the potential function of the enzymatically inactive isoforms remains unknown.

**组织特异性** In epidermis, expressed predominantly in basal undifferentiated keratinocytes and in some but not all melanocytes (at protein level).

**通路** Cofactor biosynthesis; 7,8-dihydroneopterin triphosphate biosynthesis; 7,8-dihydroneopterin triphosphate from GTP: step 1/1.

**疾病相关** Defects in GCH1 are the cause of GTP cyclohydrolase 1 deficiency (GCH1D) [MIM:233910]; also known as atypical severe phenylketonuria due to GTP cyclohydrolase I deficiency;. GCH1D is one of the causes of malignant hyperphenylalaninemia due to tetrahydrobiopterin deficiency. It is also responsible for defective neurotransmission due to depletion of the neurotransmitters dopamine and serotonin. The principal symptoms include: psychomotor retardation, tonicity disorders, convulsions, drowsiness, irritability, abnormal movements, hyperthermia, hypersalivation, and difficulty swallowing. Some patients may present a phenotype of intermediate severity between severe hyperphenylalaninemia and mild dystonia type 5 (dystonia-parkinsonism with diurnal fluctuation). In this intermediate phenotype, there is marked motor delay, but no mental retardation and only minimal, if any, hyperphenylalaninemia.
Defects in GCH1 are the cause of dystonia type 5 (DYT5) [MIM:128230]; also known as progressive dystonia with diurnal fluctuation, autosomal dominant Segawa syndrome or dystonia-parkinsonism with diurnal fluctuation. DYT5 is a DOPA-responsive dystonia. Dystonia is defined by the presence of sustained involuntary muscle contractions, often leading to abnormal postures. DYT5 typically presents in childhood with walking problems due to dystonia of the lower limbs and worsening of the dystonia towards the evening. It is characterized by postural and motor disturbances showing marked diurnal fluctuation. Torsion of the trunk is unusual. Symptoms are alleviated after sleep and aggravated by fatigue and excercise. There is a favorable response to L-DOPA without side effects.

**序列相似性** Belongs to the GTP cyclohydrolase I family.

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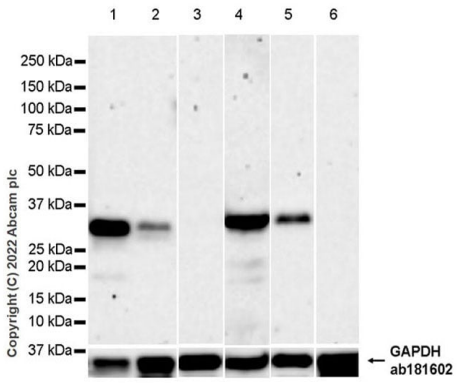
| **翻译后修饰** | Phosphorylated by casein kinase II at Ser-81 in HAECs during oscillatory shear stress; phosphorylation at Ser-81 results in increased enzyme activity. |
| **细胞定位** | Cytoplasm. Nucleus. |

## 图片




Western blot - Anti-GCH1 antibody [EPR27331-30] (ab307507)

**All lanes :** Anti-GCH1 antibody [EPR27331-30] (ab307507) at 1/1000 dilution

**Lane 1 :** Mouse liver tissue lysate 20 µg
**Lane 2 :** Mouse hypothalamus tissue lysate 20 µg
**Lane 3 :** Mouse skeletal muscle tissue lysate 20 µg
**Lane 4 :** Rat liver tissue lysate 20 µg
**Lane 5 :** Rat hypothalamus tissue lysate 20 µg
**Lane 6 :** Rat skeletal muscle tissue lysate 20 µg

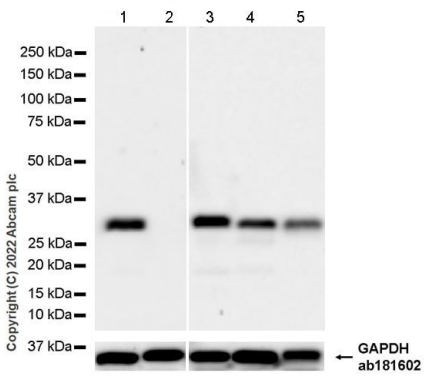
**Secondary**
**All lanes :** Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated (**ab97051**) at 1/100000 dilution

**Predicted band size:** 28 kDa
**Observed band size:** 27 kDa

Blocking and diluting buffer and concentration: 5% NFDM/TBST

Negative control: skeletal muscle (PMID:22798524)

Exposure time: 180 seconds.




Western blot - Anti-GCH1 antibody [EPR27331-30] (ab307507)

**All lanes :** Anti-GCH1 antibody [EPR27331-30] (ab307507) at 1/1000 dilution

**Lane 1 :** SH-SY5Y (human neuroblastoma epithelial cell) whole cell lysate 20 µg
**Lane 2 :** K562 (human chronic myelogenous leukemia lymphoblast) whole cell lysate 20 µg
**Lane 3 :** Neuro-2a (mouse neuroblastoma neuroblast) whole cell lysate 20 µg
**Lane 4 :** N9 (mouse microglia) whole cell lysate 20 µg
**Lane 5 :** PC-12 (rat adrenal gland pheochromocytoma) whole cell lysate 20 µg

**Secondary**
**All lanes :** Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated (**ab97051**) at 1/100000 dilution

**Predicted band size:** 28 kDa
**Observed band size:** 27 kDa

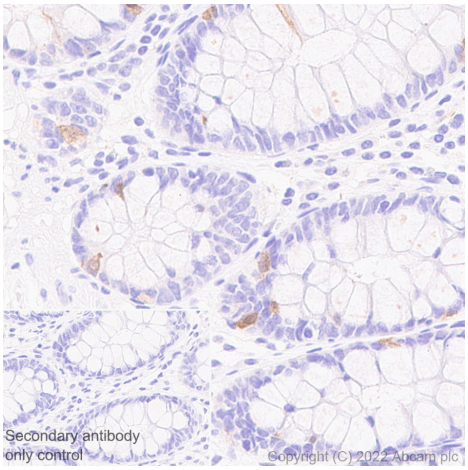
Blocking and diluting buffer and concentration: 5% NFDM/TBST

Negative control: K562 (PMID:32778843)

Exposure time:

Lanes 1-2: 37 seconds

Lanes 3-5: 10 seconds.

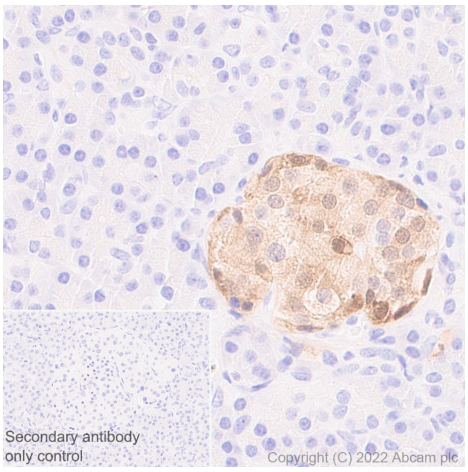



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-GCH1 antibody [EPR27331-30] (ab307507)

Immunohistochemical analysis of paraffin-embedded Human colon tissue labeling GCH1 with ab307507 at 1/500 (1.044 µg/ml) followed by a LeicaDS9800 (Bond™ Polymer Refine Detection) was used at Ready to use dilution. Positive staining on scattered cells in human colon (PMID: 22753274). The section was incubated with ab307507 for 30 mins at room temperature. The immunostaining was performed on a Leica Biosystems BOND® RX instrument. Counterstained with Hematoxylin.

Secondary antibody only control: Secondary antibody is LeicaDS9800 (Bond™ Polymer Refine Detection) was used at Ready to use dilution.

Heat mediated antigen retrieval was performed with Tris-EDTA buffer (pH 9.0, Epitope Retrieval Solution2) for 20 mins.

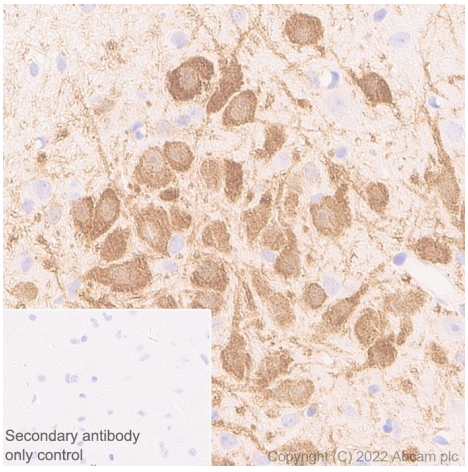



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-GCH1 antibody [EPR27331-30] (ab307507)

Immunohistochemical analysis of paraffin-embedded Human pancreas tissue labeling GCH1 with ab307507 at 1/500 (1.044 µg/ml) followed by a LeicaDS9800 (Bond™ Polymer Refine Detection) was used at Ready to use dilution. Positive staining on human pancreatic islets. The section was incubated with ab307507 for 30 mins at room temperature. The immunostaining was performed on a Leica Biosystems BOND® RX instrument. Counterstained with Hematoxylin.

Secondary antibody only control: Secondary antibody is LeicaDS9800 (Bond™ Polymer Refine Detection) was used at Ready to use dilution.

Heat mediated antigen retrieval was performed with Tris-EDTA buffer (pH 9.0, Epitope Retrieval Solution2) for 20 mins.

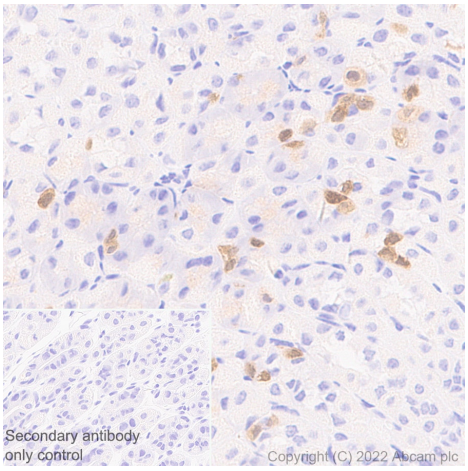



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-GCH1 antibody [EPR27331-30] (ab307507)

Immunohistochemical analysis of paraffin-embedded Mouse midbrain tissue labeling GCH1 with ab307507 at 1/1000 (0.522 µg/ml) followed by a LeicaDS9800 (Bond™ Polymer Refine Detection) was used at Ready to use dilution. Positive staining on neurons of mouse midbrain (PMID: 10907721). The section was incubated with ab307507 for 30 mins at room temperature. The immunostaining was performed on a Leica Biosystems BOND® RX instrument. Counterstained with Hematoxylin.

Secondary antibody only control: Secondary antibody is LeicaDS9800 (Bond™ Polymer Refine Detection) was used at Ready to use dilution.

Heat mediated antigen retrieval was performed with Tris-EDTA buffer (pH 9.0, Epitope Retrieval Solution2) for 20 mins.

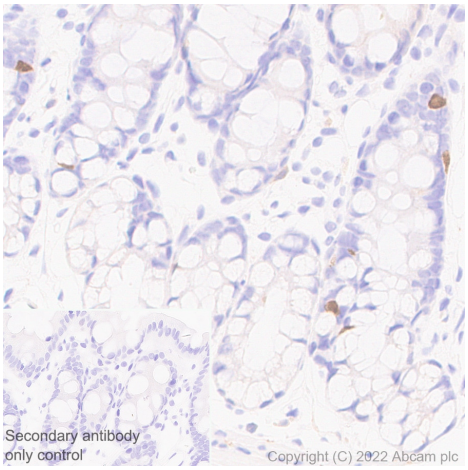



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-GCH1 antibody [EPR27331-30] (ab307507)

Immunohistochemical analysis of paraffin-embedded Mouse stomach tissue labeling GCH1 with ab307507 at 1/1000 (0.522 µg/ml) followed by a LeicaDS9800 (Bond™ Polymer Refine Detection) was used at Ready to use dilution. Positive staining on scattered cells in mouse stomach. The section was incubated with ab307507 for 30 mins at room temperature. The immunostaining was performed on a Leica Biosystems BOND® RX instrument Counterstained with Hematoxylin.

Secondary antibody only control: Secondary antibody is LeicaDS9800 (Bond™ Polymer Refine Detection) was used at Ready to use dilution.

Heat mediated antigen retrieval was performed with Tris-EDTA buffer (pH 9.0, Epitope Retrieval Solution2) for 20 mins.

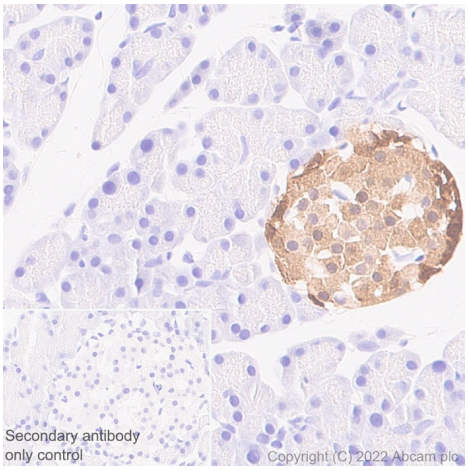



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-GCH1 antibody [EPR27331-30] (ab307507)

Immunohistochemical analysis of paraffin-embedded Rat colon tissue labeling GCH1 with ab307507 at 1/1000 (0.522 µg/ml) followed by a LeicaDS9800 (Bond™ Polymer Refine Detection) was used at Ready to use dilution. Positive staining on scattered cells in rat colon. The section was incubated with ab307507 for 30 mins at room temperature. The immunostaining was performed on a Leica Biosystems BOND® RX instrument. Counterstained with Hematoxylin.

Secondary antibody only control: Secondary antibody is LeicaDS9800 (Bond™ Polymer Refine Detection) was used at Ready to use dilution.

Heat mediated antigen retrieval was performed with Tris-EDTA buffer (pH 9.0, Epitope Retrieval Solution2) for 20 mins.

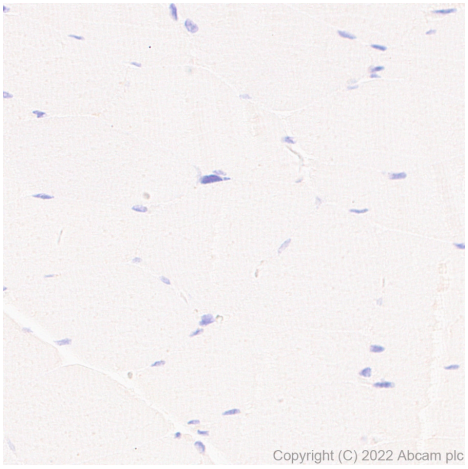



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-GCH1 antibody [EPR27331-30] (ab307507)

Immunohistochemical analysis of paraffin-embedded Rat pancreas tissue labeling GCH1 with ab307507 at 1/1000 (0.522 µg/ml) followed by a LeicaDS9800 (Bond™ Polymer Refine Detection) was used at Ready to use dilution. Positive staining on rat pancreatic islets. The section was incubated with ab307507 for 30 mins at room temperature. The immunostaining was performed on a Leica Biosystems BOND® RX instrument. Counterstained with Hematoxylin.

Secondary antibody only control: Secondary antibody is LeicaDS9800 (Bond™ Polymer Refine Detection) was used at Ready to use dilution.

Heat mediated antigen retrieval was performed with Tris-EDTA buffer (pH 9.0, Epitope Retrieval Solution2) for 20 mins.



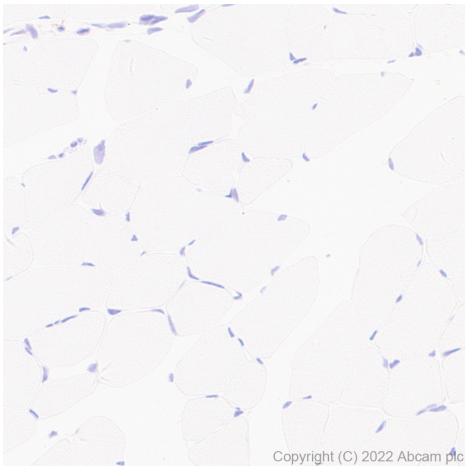

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-GCH1 antibody [EPR27331-30] (ab307507)

Immunohistochemical analysis of paraffin-embedded Human skeletal muscle tissue labeling GCH1 with ab307507 at 1/500 (1.044 µg/ml) followed by a LeicaDS9800 (Bond™ Polymer Refine Detection) was used at Ready to use dilution.

Negative control: No staining on human skeletal muscle.The section was incubated with ab307507 for 30 mins at room temperature. The immunostaining was performed on a Leica Biosystems BOND® RX instrument. Counterstained with Hematoxylin.

Secondary antibody only control: Secondary antibody is LeicaDS9800 (Bond™ Polymer Refine Detection) was used at Ready to use dilution.

Heat mediated antigen retrieval was performed with Tris-EDTA buffer (pH 9.0, Epitope Retrieval Solution2) for 20 mins.



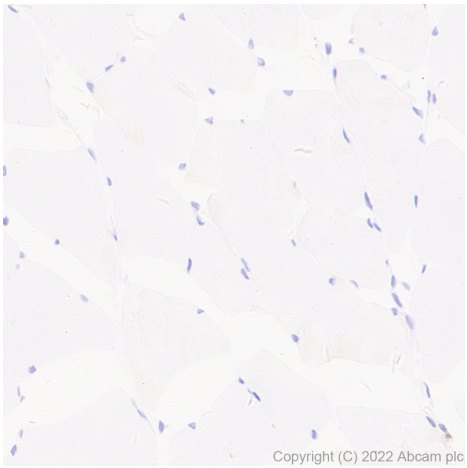

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-GCH1 antibody [EPR27331-30] (ab307507)

Immunohistochemical analysis of paraffin-embedded Mouse skeletal muscl tissue labeling GCH1 with ab307507 at 1/1000 (0.522 µg/ml) followed by a LeicaDS9800 (Bond™ Polymer Refine Detection) was used at Ready to use dilution.

Negative control: No staining on mouse skeletal muscle.The section was incubated with ab307507 for 30 mins at room temperature. The immunostaining was performed on a Leica Biosystems BOND® RX instrument. Counterstained with Hematoxylin.

Secondary antibody only control: Secondary antibody is LeicaDS9800 (Bond™ Polymer Refine Detection) was used at Ready to use dilution.

Heat mediated antigen retrieval was performed with Tris-EDTA buffer (pH 9.0, Epitope Retrieval Solution2) for 20 mins.



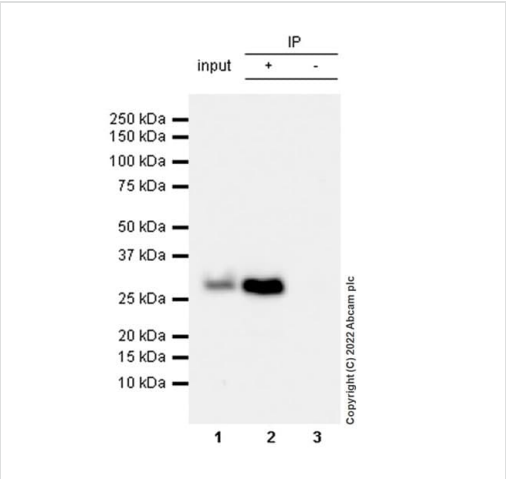

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-GCH1 antibody [EPR27331-30] (ab307507)

Immunohistochemical analysis of paraffin-embedded Rat skeletal muscle tissue labeling GCH1 with ab307507 at 1/1000 (0.522 µg/ml) followed by a LeicaDS9800 (Bond™ Polymer Refine Detection) was used at Ready to use dilution.

Negative control: No staining on rat skeletal muscle. The section was incubated with ab307507 for 30 mins at room temperature. The immunostaining was performed on a Leica Biosystems BOND® RX instrument. Counterstained with Hematoxylin.

Secondary antibody only control: Secondary antibody is LeicaDS9800 (Bond™ Polymer Refine Detection) was used at Ready to use dilution.

Heat mediated antigen retrieval was performed with Tris-EDTA buffer (pH 9.0, Epitope Retrieval Solution2) for 20 mins.




Immunoprecipitation - Anti-GCH1 antibody [EPR27331-30] (ab307507)

GCH1 was immunoprecipitated from 0.35 mg Neuro-2a (mouse neuroblastoma neuroblast) whole cell lysate 10 µg with ab307507 at 1/30 dilution (2µg in 0.35mg lysates). Western blot was performed on the immunoprecipitate using ab307507 at 1/1000 dilution. VeriBlot for IP secondary antibody(HRP)(**ab131366**) was used at 1/5000 dilution.

Lane 1: Neuro-2a (mouse neuroblastoma neuroblast), whole cell lysate 10 µg

Lane 2: ab307507 IP in Neuro-2a whole cell lysate

Lane 3: Rabbit monoclonal IgG (**ab172730**) instead of ab307507 in Neuro-2a whole cell lysate

Blocking and dilution buffer and concentration: 5% NFDM/TBST.

Exposure time: 6 seconds.


Anti-GCH1 antibody [EPR27331-30] (ab307507)

**Please note:** All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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