abcam

Product datasheet

Anti-OCRL antibody ab125917

3 图像

概述

产品名称	Anti-OCRL抗体
描述	兔多克隆抗体to OCRL
宿主	Rabbit
经测试应用	**适用于:** WB, IHC-P, ICC/IF
种属反应性	**与反应:** Human
免疫原	Recombinant fragment corresponding to Human OCRL aa 591-857. Database link: **Q01968**
阳性对照	293T and A431 cell lysates, A431 cells and Human Breast ca tissue.
常规说明	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

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	pH: 7.00 Preservative: 0.01% Thimerosal (merthiolate) Constituents: 1.21% Tris, 0.75% Glycine, 20% Glycerol (glycerin, glycerine)
纯度	Immunogen affinity purified
克隆	**多克隆**
同种型	IgG

应用

The Abpromise guarantee **Abpromise™承诺保证使用ab125917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		1/500 - 1/3000. Detects a band of approximately 104 kDa.
IHC-P		1/100 - 1/500. Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol. citrate buffer-10mM
ICC/IF		1/100 - 1/500.

靶标

功能 Converts phosphatidylinositol 4,5-bisphosphate to phosphatidylinositol 4-phosphate. Also converts inositol 1,4,5-trisphosphate to inositol 1,4-bisphosphate and inositol 1,3,4,5-tetrakisphosphate to inositol 1,3,4-trisphosphate. May function in lysosomal membrane trafficking by regulating the specific pool of phosphatidylinositol 4,5-bisphosphate that is associated with lysosomes.

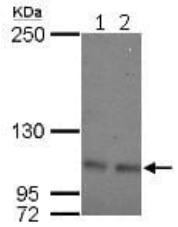
组织特异性 Brain, skeletal muscle, heart, kidney, lung, placenta and fibroblasts.

疾病相关 Defects in OCRL are the cause of Lowe oculocerebrorenal syndrome (OCRL) [MIM:309000]. It is an X-linked multisystem disorder affecting eyes, nervous system, and kidney. It is characterized by hydrophthalmia, cataract, mental retardation, vitamin D-resistant rickets, aminoaciduria, and reduced ammonia production by the kidney. Ocular abnormalities include cataract, glaucoma, microphthalmos, and decreased visual acuity. Developmental delay, hypotonia, behavior abnormalities, and areflexia are also present. Renal tubular involvement is characterized by impaired reabsorption of bicarbonate, amino acids, and phosphate. Musculoskeletal abnormalities such as joint hypermobility, dislocated hips, and fractures may develop as consequences of renal tubular acidosis and hypophosphatemia. Cataract is the only significant manifestation in carriers and is detected by slit-lamp examination.
Defects in OCRL are the cause of Dent disease type 2 (DD2) [MIM:300555]. DD2 is a renal disease belonging to the 'Dent disease complex', a group of disorders characterized by proximal renal tubular defect, hypercalciuria, nephrocalcinosis, and renal insufficiency. The spectrum of phenotypic features is remarkably similar in the various disorders, except for differences in the severity of bone deformities and renal impairment. Characteristic abnormalities include low-molecular-weight proteinuria and other features of Fanconi syndrome, such as glycosuria, aminoaciduria, and phosphaturia, but typically do not include proximal renal tubular acidosis. Progressive renal failure is common, as are nephrocalcinosis and kidney stones.

序列相似性 Belongs to the inositol-1,4,5-trisphosphate 5-phosphatase type II family.
Contains 1 Rho-GAP domain.

细胞定位 Endosome. Also found on macropinosomes.

图片



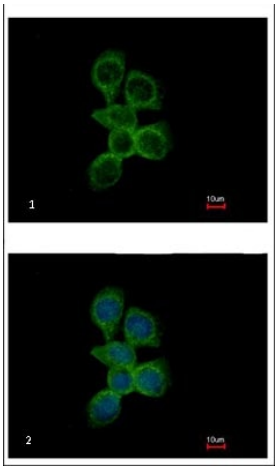
Western blot - Anti-OCRL antibody (ab125917)

All lanes : Anti-OCRL antibody (ab125917) at 1/500 dilution

Lane 1 : 293T whole cell lysate
Lane 2 : A431 whole cell lysate

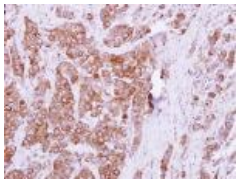
Lysates/proteins at 30 µg per lane.

Gel concentration:5%



Immunocytochemistry/ Immunofluorescence - Anti-OCRL antibody (ab125917)

ab125917, at 1/500, staining OCRL in Human A431 cells (methanol-fixed) by immunofluorescence (panel 1) and co-stained with Hoechst 33342 (panel 2).



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-OCRL antibody (ab125917)

ab125917, at 1/250, staining OCRL in Human Breast ca tissue by immunohistochemistry [Paraffin Embedded Tissues (IHC-P)].

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