abcam

Product datasheet

Anti-ATP6V0A2 antibody ab96803

5 References 2 图像

概述

产品名称	Anti-ATP6V0A2抗体
描述	兔多克隆抗体to ATP6V0A2
宿主	Rabbit
经测试应用	**适用于:** WB, IHC-P
种属反应性	**与反应:** Human **预测可用于:** Mouse, Rat, Cow
免疫原	Recombinant fragment corresponding to a region within amino acids 156-404 of Human ATP6V0A2 (NP_036595).
阳性对照	293T, A431 and H1299 whole cell lysates; DLD1 xenograft; HeLa, HepG2, MOLT4 and Raji cell lysates
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. Store at -20°C or -80°C. Avoid freeze / thaw cycle.
存储溶液	pH: 7.00 Preservative: 0.01% Thimerosal (merthiolate) Constituents: 79.99% PBS, 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ab96803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. Predicted molecular weight: 98 kDa.
IHC-P		1/100 - 1/500.

靶标

功能 Part of the proton channel of V-ATPases. Essential component of the endosomal pH-sensing machinery. May play a role in maintaining the Golgi functions, such as glycosylation maturation, by controlling the Golgi pH.

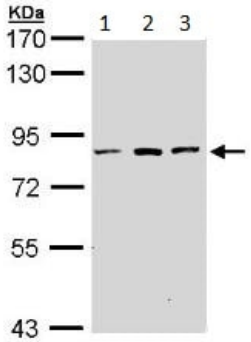
疾病相关 Defects in ATP6V0A2 are the cause of cutis laxa autosomal recessive type 2A (ARCL2A) [MIM:219200]. An autosomal recessive disorder characterized by an excessive congenital skin wrinkling, a large fontanelle with delayed closure, a typical facial appearance with downslanting palpebral fissures, a general connective tissue weakness, and varying degrees of growth and developmental delay and neurological abnormalities. Some affected individuals develop seizures and mental deterioration later in life, whereas the skin phenotype tends to become milder with age. At the molecular level, an abnormal glycosylation of serum proteins is observed in many cases.
Defects in ATP6V0A2 are a cause of wrinkly skin syndrome (WSS) [MIM:278250]. WSS is rare autosomal recessive disorder characterized by wrinkling of the skin of the dorsum of the hands and feet, an increased number of palmar and plantar creases, wrinkled abdominal skin, multiple musculoskeletal abnormalities, microcephaly, growth failure and developmental delay.

序列相似性 Belongs to the V-ATPase 116 kDa subunit family.

翻译后修饰 Phosphorylated upon DNA damage, probably by ATM or ATR.

细胞定位 Cell membrane. Endosome membrane. In kidney proximal tubules, also detected in subapical vesicles.

图片



Western blot - Anti-ATP6V0A2 antibody (ab96803)

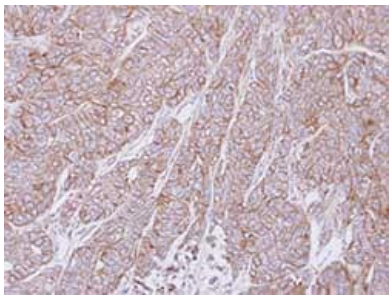
All lanes : Anti-ATP6V0A2 antibody (ab96803) at 1/10000 dilution

Lane 1 : 293T whole cell lysate
Lane 2 : A431 whole cell lysate
Lane 3 : H1299 whole cell lysate

Lysates/proteins at 30 µg per lane.

Predicted band size: 98 kDa

7.5% SDS PAGE



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-ATP6V0A2 antibody (ab96803)

ab96803, at 1/500 dilution, staining ATP6V0A2 in paraffin-embedded DLD1 xenograft by Immunohistochemistry.