液泡膜质子转运 ATP 酶 2 型抗体

产品货号： mlR12761

英文名称： ATP6V0A2

中文名称： 液泡膜质子转运 ATP 酶 2 型抗体

别　　名： a2; A2V ATPase; ARCL; ATP6a2; ATP6N1D; ATP6V0A2; ATPase, H+ transporting, lysosomal V0 subunit a isoform 2; ATPase, H+ transporting, lysosomal V0 subunit a2; Infantile malignant osteopetrosis; J6B7; Lysosomal H(+) transporting ATPase V0 subunit a2; Lysosomal H(+)-transporting ATPase V0 subunit a2; regeneration and tolerance factor; Stv1; TJ6; TJ6M; TJ6s; V ATPase 116 kDa isoform a2; V type proton ATPase 116 kDa subunit a; V type proton ATPase 116 kDa subunit a isoform 2; V-ATPase 116 kDa isoform a2; V-type proton ATPase 116 kDa subunit a isoform 2; Vacuolar proton translocating ATPase 116 kDa subunit a; Vacuolar proton translocating ATPase 116 kDa subunit a isoform 2; Vph1; VPP2_HUMAN; WSS.

研究领域： 肿瘤　细胞生物　信号转导　细胞分化　表观遗传学

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Dog, Pig, Cow, Horse, Sheep,

产品应用： IHC-P=1:400-800 IHC-F=1:400-800 ICC=1:100-500 IF=1:100-500 （石蜡切片需做抗原修复）
not yet tested in other applications.
optimal dilutions/concentrations should be determined by the end user.

分 子 量： 98kDa

细胞定位： 细胞膜

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human ATP6V0A2:1-100/856 <Cytoplasmic>

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： Store at -20 ° C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20° C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 ° C.

PubMed： PubMed

产品介绍： The protein encoded by this gene is a subunit of the vacuolar ATPase (v-ATPase), an heteromultimeric enzyme that is present in intracellular vesicles and in the plasma membrane of specialized cells, and which is essential for the acidification of diverse cellular components. V-ATPase is comprised of a membrane peripheral V(1) domain for ATP hydrolysis, and an integral membrane V(0) domain for proton translocation. The subunit encoded by this gene is a component of the V(0) domain. Mutations in this gene are a cause of both cutis laxa type II and wrinkly skin syndrome. [provided by RefSeq, Jul 2009]

Function:

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.

Subcellular Location:

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

Post-translational modifications:

Phosphorylated upon DNA damage, probably by ATM or ATR.

DISEASE:

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.

Similarity:

Belongs to the V-ATPase 116 kDa subunit family.

SWISS:

Q9Y487

Gene ID:

23545

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic

applications.