

Product name:	V-ATPase B1 Rabbit Polyclonal Antibody
Cat number:	ABN19733
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human ATP6V1B1. AA range:381-430
Reactivity:	Human,Mouse
Applications:	WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:50-1:200,ELISA 1:5000-1:10000
Molecular Weight:	60kDa
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

Background:

This gene encodes a component of vacuolar ATPase (V-ATPase), a multisubunit enzyme that mediates acidification of eukaryotic intracellular organelles. V-ATPase dependent organelle acidification is necessary for such intracellular processes as protein sorting, zymogen activation, receptor-mediated endocytosis, and synaptic vesicle proton gradient generation. V-ATPase is composed of a cytosolic V1 domain and a transmembrane V0 domain. The V1 domain consists of three A and three B subunits, two G subunits plus the C, D, E, F, and H subunits. The V1 domain contains the ATP catalytic site. The V0 domain consists of five different subunits: a, c, c', c'', and d. Additional isoforms of many of the V1 and V0 subunit proteins are encoded by multiple genes or alternatively spliced transcript variants. This encoded protein is one of two V1 domain B subunit isoforms and is found in disease: Defects in ATP6V1B1 are the cause of distal renal tubular acidosis with deafness (dRTA) [MIM:267300]. Inheritance is autosomal recessive. Patients with recessive dRTA are severely affected, presenting with either acute illness or growth failure at a young age, and bilateral sensorineural deafness. Other features include low serum K(+) due to renal potassium wasting, and elevated urinary calcium. If untreated, this acidosis may result in dissolution of bone, leading to osteomalacia and rickets. Renal deposition of calcium salts (nephrocalcinosis) and renal stone formation commonly occur. domain: The PDZ-binding motif mediates interactions with SLC9A3R1 and SCL4A7. function: Non-catalytic subunit of the peripheral V1 complex of vacuolar ATPase. V-ATPase is responsible for acidifying a variety of intracellular compartments in eukaryotic cells. similarity: Belongs to the ATPase alpha/beta chains family. subcellular location: Endomembrane. subunit: V-ATPase is an heteromultimeric enzyme composed of a peripheral catalytic V1 complex (main components: subunits A, B, C, D, E, and F) attached to an integral membrane V0 proton pore complex (main component: the proteolipid protein). Forms a complex with SLC9A3R1 and SCL4A7. tissue specificity: Expressed in the cochlea and endolymphatic sac.