abcam

Product datasheet

epithelial Sodium Channel alpha peptide ab4987

Description

Product name	epithelial Sodium Channel alpha peptide
Animal free	No
Nature	Synthetic

Specifications

Our Abpromise guarantee covers the use of **ab4987** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications	Blocking
Form	Lyophilized
Additional notes	This peptide may be used for neutralization and control experiments with the polyclonal antibody that reacts with this product and endogenous alpha-ENaC, catalog ab3464. Using a solution of peptide of equal volume and concentration to the corresponding antibody will yield a large molar excess of peptide (~ 70-fold) for competitive inhibition of antibody-protein binding reactions.

Preparation and Storage

Stability and Storage	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Store at -20°C or -80°C. Avoid freeze / thaw cycle.
Reconstitution	>95% pure, lyophilized synthetic peptide. Reconstitute with 0.1 ml of distilled water.

General Info

Function	Sodium permeable non-voltage-sensitive ion channel inhibited by the diuretic amiloride. Mediates the electrodiffusion of the luminal sodium (and water, which follows osmotically) through the apical membrane of epithelial cells. Controls the reabsorption of sodium in kidney, colon, lung and sweat glands. Also plays a role in taste perception.
Tissue specificity	Highly expressed in kidney and lung. Detected at intermediate levels in pancreas and liver, and at low levels in heart and placenta. Isoform 1 and isoform 2 predominate in all tissues. Expression of isoform 3, isoform 4 and isoform 5 is very low or not detectable, except in lung and heart.
Involvement in disease	Defects in SCNN1A are a cause of autosomal recessive pseudohypoaldosteronism type 1 (AR-PHA1) [MIM:264350]. PHA1 is a rare salt wasting disease resulting from target organ

	unresponsiveness to mineralocorticoids. There are 2 forms of PHA1: the autosomal recessive form that is severe, and the dominant form which is milder and due to defects in mineralocorticoid receptor. AR-PHA1 is characterized by an often fulminant presentation in the neonatal period with dehydration, hyponatraemia, hyperkalaemia, metabolic acidosis, failure to thrive and weight loss. Note=The degree of channel function impairment differentially affects the renin-aldosterone system and urinary Na/K ratios, resulting in distinct genotype-phenotype relationships in PHA1 patients. Loss-of-function mutations are associated with a severe clinical course and age-dependent hyperactivation of the renin-aldosterone system. This feature is not observed in patients with missense mutations that reduce but do not eliminate channel function. Markedly reduced channel activity results in impaired linear growth and delayed puberty. Defects in SCNN1A are a cause of bronchiectasis with or without elevated sweat chloride type 2 (BESC2) [MIM:613021]; also called cystic fibrosis-like syndrome. BESC2 is a debilitating respiratory disease characterized by chronic abnormal dilatation of the bronchi and other cystic fibrosis-like symptoms in the absence of known causes of bronchiectasis (cystic fibrosis, autoimmune diseases, ciliary dyskinesia, common variable immunodeficiency, foreign body obstruction). Clinical features include subnormal lung function, sinopulmonary infections, chronic productive cough, excessive sputum production, and elevated sweat chloride in some cases.
Sequence similarities	Belongs to the amiloride-sensitive sodium channel (TC 1.A.6) family. SCNN1A subfamily.
Post-translational modifications	Ubiquitinated; this targets individual subunits for endocytosis and proteasome-mediated degradation.
Cellular localization	Apical cell membrane. Apical membrane of epithelial cells.

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