abcam

Product datasheet

epithelial Sodium Channel alpha peptide ab4987

製品の詳細

製品名	epithelial Sodium Channel alpha peptide
精製度	> 70 % HPLC. Peptides are analyzed by Reverse-Phase HPLC (RP-HPLC) in order to determine purity. Identities are confirmed by MALDI-MS.
Animal free	No
由来	Synthetic

特性

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.

アプリケーション	Blocking
製品の状態	Lyophilized
備考	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.

前処理および保存

保存方法および安定性	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Store at -20°C or -80°C. Avoid freeze / thaw cycle.
再構成	>95% pure, lyophilized synthetic peptide. Reconstitute with 0.1 ml of distilled water.

関連情報

機能	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.
組織特異性	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.
関連疾患	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.
配列類似性	Belongs to the amiloride-sensitive sodium channel (TC 1.A.6) family. SCNN1A subfamily.
翻訳後修飾	Ubiquitinated; this targets individual subunits for endocytosis and proteasome-mediated degradation.
細胞内局在	Apical cell membrane. Apical membrane of epithelial cells.