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PHOX2B Rabbit pAb

Catalog # AP99483

Product Information

Application	WB, IHC-P, IHC-F, IF, E
Primary Accession	Q99453
Other Accession	Q99453
Predicted	Rat, Pig, Human, Mouse, Rabbit, Chicken, Dog, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	31621
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human PHOX2B
Epitope Specificity	101-200/314
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the paired homeobox family. Contains 1 homeobox DNA-binding domain.
SUBUNIT	Interacts with TRIM11
DISEASE	Defects in PHOX2B are a cause of congenital central hypoventilation syndrome (CCHS) [MIM:209880]; also known as congenital failure of autonomic control or Ondine curse. Most mutations consist of 5-10 alanine expansions in the poly-Ala region from amino acids 241-260. CCHS is a rare disorder characterized by abnormal control of respiration in the absence of neuromuscular or lung disease, or an identifiable brain stem lesion. A deficiency in autonomic control of respiration results in inadequate or negligible ventilatory and arousal responses to hypercapnia and hypoxemia. CCHS is frequently complicated with neurocristopathies such as Hirschsprung disease that occurs in about 16% of CCHS cases. Defects in PHOX2B are the cause of susceptibility to neuroblastoma type 2 (NBLST2) [MIM:613013]. A common neoplasm of early childhood arising from embryonic cells that form the primitive neural crest and give rise to the adrenal medulla and the sympathetic nervous system.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Phox2a (also designated Arix1) and Phox2b are closely related, paired-homeodomain transcription factors that are necessary for neuronal differentiation throughout the developing sympathetic, parasympathetic and enteric ganglia. All enteric nervous system cells evolve from the neural crest, and all cells that are undifferentiated initially express Phox2b. The cells that begin to differentiate along a neuronal lineage continue to express Phox2b, and begin to express Phox2a. Phox2b is required for the differentiation of all central and nonperipheral noradrenergic centers in the brain. In contrast, Phox2a controls only the differentiation of the main noradrenergic center of the brain, the locus coeruleus. Both Phox2a and Phox2b are crucial for the regulation of endogenous tyrosine hydroxylase and dopamine-beta hydroxylase, which are transiently expressed in neural crest cells. In addition, Phox2 proteins are sufficient to promote sympathetic neuron generation. The

gene which encodes Phox2a maps to human chromosome 11q13.3-q13.4.

Additional Information

Gene ID	8929
Other Names	CCHS; NBLST2; NBPhox; PMX2B; Dilp1; Px2b; PHX2B_HUMAN; PHOX2B; Neuroblastoma Phox (NBPhox); PHOX2B homeodomain protein; Paired-like homeobox 2B; PHX2B_MOUSE; paired like homeobox 2B; paired mesoderm homeobox 2b
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name	PHOX2B
Synonyms	PMX2B
Function	Involved in the development of several major noradrenergic neuron populations, including the locus coeruleus. Transcription factor which could determine a neurotransmitter phenotype in vertebrates. Enhances second-messenger-mediated activation of the dopamine beta- hydrolase and c-fos promoters, and of several enhancers including cAMP- response element and serum-response element.
Cellular Location	Nucleus {ECO:0000255|PROSITE-ProRule:PRU00108}.
Tissue Location	Expressed in neuroblastoma, brain and adrenal gland

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