

Product name:	COX10 Rabbit Polyclonal Antibody
Cat number:	ABN09265
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human COX10. AA range:98-147
Reactivity:	Human,Rat,Mouse
Applications:	WB 1:500-1:2000,ELISA 1:5000-1:20000
Molecular Weight:	49kDa
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:

Cytochrome c oxidase (COX), the terminal component of the mitochondrial respiratory chain, catalyzes the electron transfer from reduced cytochrome c to oxygen. This component is a heteromeric complex consisting of 3 catalytic subunits encoded by mitochondrial genes and multiple structural subunits encoded by nuclear genes. The mitochondrially-encoded subunits function in electron transfer, and the nuclear-encoded subunits may function in the regulation and assembly of the complex. This nuclear gene encodes heme A:farnesyltransferase, which is not a structural subunit but required for the expression of functional COX and functions in the maturation of the heme A prosthetic group of COX. This protein is predicted to contain 7-9 transmembrane domains localized in the mitochondrial inner membrane. A gene mutation, which results in the substitution of a lysine residue, is a cause of cytochrome c oxidase deficiency (COX deficiency) [MIM:220110]. COX deficiency is a clinically heterogeneous disorder. The clinical features are ranging from isolated myopathy to severe multisystem disease, with onset from infancy to adulthood. Defects in COX10 are a cause of Leigh syndrome (LS) [MIM:256000]. LS is a severe neurological disorder characterized by bilaterally symmetrical necrotic lesions in subcortical brain regions. Converts protoheme IX and farnesyl diphosphate to heme O. Belongs to the ubiA prenyltransferase family.