



COX1 rabbit pAb

Cat No.:ES9123

For research use only

Overview

Product Name	COX1 rabbit pAb
Host species	Rabbit
Applications	WB;ELISA
Species Cross-Reactivity	Human;Mouse
Recommended dilutions	WB 1:500-2000 ELISA 1:5000-20000
Immunogen	Synthesized peptide derived from human protein . at AA range: 380-460
Specificity	COX1 Polyclonal Antibody detects endogenous levels of protein.
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Storage	Store at -20°C . Avoid repeated freeze-thaw cycles.
Protein Name	Cytochrome c oxidase subunit 1 (EC 1.9.3.1) (Cytochrome c oxidase polypeptide I)
Gene Name	MT-CO1 COI COXI MTCO1
Cellular localization	Mitochondrion inner membrane ; Multi-pass membrane protein .
Purification	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen.
Clonality	Polyclonal
Concentration	1 mg/ml
Observed band	56kD
Human Gene ID	4512
Human Swiss-Prot Number	P00395
Alternative Names	
Background	catalytic activity:4 ferrocytochrome c + O(2) + 4 H(+) = 4 ferricytochrome c + 2 H(2)O.,disease:Defects in MT-CO1 are a cause of anemia sideroblastic acquired idiopathic (AISA) [MIM:516030]; a disease characterized by inadequate formation of heme and excessive accumulation of iron in mitochondria.,disease:Defects in MT-CO1 are a





cause of cytochrome c oxidase deficiency (COX deficiency) [MIM:220110]; also called mitochondrial complex IV deficiency. 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.,disease:Defects in MT-CO1 are a cause of Leber hereditary optic neuropathy (LHON) [MIM:535000]. LHON is a maternally inherited disease resulting in acute or subacute loss of central vision, due to optic nerve dysfunction. Cardiac conduction defects and neurological defects have also been described in some patients. LHON results from primary mitochondrial DNA mutations affecting the respiratory chain complexes.,disease:Defects in MT-CO1 are associated with recurrent myoglobinuria [MIM:550500]. Myoglobinuria consists of excretion of myoglobin in the urine.,function:Cytochrome c oxidase is the component of the respiratory chain that catalyzes the reduction of oxygen to water. Subunits 1-3 form the functional core of the enzyme complex. CO I is the catalytic subunit of the enzyme. Electrons originating in cytochrome c are transferred via the copper A center of subunit 2 and heme A of subunit 1 to the bimetallic center formed by heme A3 and copper B.,pathway:Energy metabolism; oxidative phosphorylation.,similarity:Belongs to the heme-copper respiratory oxidase family.,

