SunLong Biotech Co.,LTD
Tel: 0086-571-56623320 Fax:0086-571-56623318
E-mail:sales@sunlongbiotech.com
www.sunlongbiotech.com

**[ Marker Primary Antibody ]**

# Rabbit Anti-COX10/FITC Conjugated antibody

**Catalog Number: SL23098R-FITC**

**FOR RESEARCH USE ONLY**

**[ DATASHEET ]**

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| **Host:** | Rabbit |
| **Isotype:** | IgG |
| **Clonality:** | Polyclonal |
| **Target:** | Anti-COX10/FITC |
| **Immunogen:** | KLH conjugated synthetic peptide derived from human COX10 |
| **Purification:** | affinity purified by Protein A |
| **Concentration:** | 1mg/ml |
| **Storage:** | Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 1M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C. |
| **Applications:** | IF=1:100-500<br>not yet tested in other applications.<br>optimal dilutions/concentrations should be determined by the end user. |
| **Reactivity:** | Human,Mouse(predicted:Rat) |
| **Background:** | 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.<br>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 for an asparagine (N204K), is identified to be responsible for cytochrome c oxidase deficiency.<br>In addition, this gene is disrupted in patients with CMT1A (Charcot-Marie- |

Important Note: This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

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| | Tooth type 1A) duplication and with HNPP (hereditary neuropathy with liability to pressure palsies) deletion. [provided by RefSeq, Jul 2008]<br>**Function:** Converts protoheme IX and farnesyl diphosphate to heme O.<br>**Subcellular Location:** Mitochondrion membrane; Multi-pass membrane protein.<br>DISEASE: Mitochondrial complex IV deficiency (MT-C4D) [MIM:220110]: A disorder of the mitochondrial respiratory chain with heterogeneous clinical manifestations, ranging from isolated myopathy to severe multisystem disease affecting several tissues and organs. Features include hypertrophic cardiomyopathy, hepatomegaly and liver dysfunction, hypotonia, muscle weakness, exercise intolerance, developmental delay, delayed motor development and mental retardation. Some affected individuals manifest a fatal hypertrophic cardiomyopathy resulting in neonatal death. A subset of patients manifest Leigh syndrome. Note=The disease is caused by mutations affecting the gene represented in this entry.<br>Leigh syndrome (LS) [MIM:256000]: An early-onset progressive neurodegenerative disorder characterized by the presence of focal, bilateral lesions in one or more areas of the central nervous system including the brainstem, thalamus, basal ganglia, cerebellum and spinal cord. Clinical features depend on which areas of the central nervous system are involved and include subacute onset of psychomotor retardation, hypotonia, ataxia, weakness, vision loss, eye movement abnormalities, seizures, and dysphagia. Note=The disease is caused by mutations affecting the gene represented in this entry.<br>**Similarity:** Belongs to the UbiA prenyltransferase family.<br>**Database links:**<br>**Entrez Gene:** 417329 Chicken<br>**Entrez Gene:** 511440 Cow<br>**Entrez Gene:** 1352 Human<br>**Entrez Gene:** 70383 Mouse<br>**Entrez Gene:** 363617 Rat<br>**Entrez Gene:** 553384 Zebrafish<br>Omim: 602125 Human<br>**SwissProt:** Q12887 Human<br>**SwissProt:** Q8CFY5 Mouse<br>**Unigene:** 462278 Human<br>**Unigene:** 340211 Mouse<br>Important<br>**Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications. |