

COX6B1 Antibody, Biotin conjugated



PACO25379

Product Information

Size:

50ug

Reactivity:

Human

Source:

Rabbit

Isotype:

IgG

Applications:

ELISA

Recommended dilutions:**Protein Background:**

Defects in COX6B1 are a cause of mitochondrial complex IV deficiency (MT-C4D) [MIM:220110]; also known as cytochrome c oxidase deficiency. 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. A subset of patients manifest Leigh syndrome.

Gene ID:

COX6B1

Uniprot

P14854

Synonyms:

Cytochrome c oxidase subunit 6B1 (Cytochrome c oxidase subunit VIb isoform 1) (COX VIb-1), COX6B1, COX6B

Immunogen:

Recombinant Human Cytochrome c oxidase subunit 6B1 protein (1-86AA).

Storage:

Preservative: 0.03% Proclin 300. Constituents: 50% Glycerol, 0.01M PBS, PH 7.4

Product Images

N/A

N/A