

Recombinant human SCO2 protein

Catalog Number: ATGP0867

PRODUCT INFORMATION

Expression system

E.coli

Domain

42-266aa

UniProt No.

O43819

NCBI Accession No.

NP_005129

Alternative Names

Protein SCO2 homolog mitochondrial, Protein SCO2 homolog mitochondrial, SCO1L

PRODUCT SPECIFICATION

Molecular Weight

27.4 kDa (246aa) confirmed by MALDI-TOF

Concentration

0.5mg/ml (determined by Bradford assay)

Formulation

Liquid in. 20mM Tris-HCl buffer (pH 8.0) containing 30% glycerol, 2mM DTT, 200mM NaCl

Purity

> 90% by SDS-PAGE

Tag

His-Tag

Application

SDS-PAGE

Storage Condition

Can be stored at +2C to +8C for 1 week. For long term storage, aliquot and store at -20C to -80C. Avoid repeated freezing and thawing cycles.

BACKGROUND

Description

SCO2 protein belongs to the SCO1/2 family of proteins. Both SCO1 and SCO2 proteins are located on the inner membrane of the mitochondria and plays a crucial role in copper insertion or transport to the active site of cytochrome c oxidase (COX). Defects in SCO2 are the cause of fatal infantile cardioencephalomyopathy with cytochrome c oxidase deficiency (FIC). This disease is characterized by hypertrophic cardiomyopathy, lactic acidosis, and gliosis. Heart and skeletal muscle show reductions in cytochrome c oxidase (COX) activity, whereas liver and fibroblasts show mild COX deficiencies. Recombinant human SCO2 protein, fused to His-tag at N-

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terminus, was expressed in E. coli and purified by using conventional chromatography techniques.

Amino acid Sequence

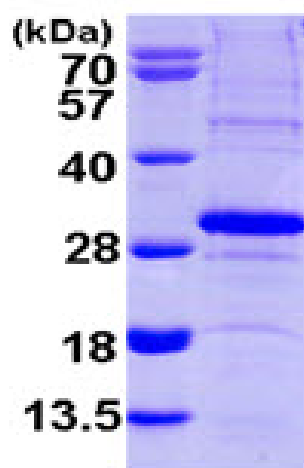
MGSSHHHHHH SSSLVPRGSH MGPAETGGQG QPQGPGLRTR LLITGLFGAG LGGAWLALRA EKERLQQQKR
TEALRQAAVG QGDFHLLDHR GRARCKADFR GQWVLMYFGF THCPDICPDE LEKLVQVVRQ LEAEPGLPPV QPVFITVDPE
RDDVEAMARY VQDFHPRLLG LTGSTKQVAQ ASHSYRVYYN AGPKDEDQDY IVDHSIAIYL LNPDGLFTDY YGRSRSAEQI
SDSVRRHMAA FRSVLS

General References

Meister G., et al. (2001) EMBO J. 20:2304-2314
Talbot K., et al. (1998) Hum. Mol. Genet. 7:2149-2156

DATA

SDS-PAGE



3ug by SDS-PAGE under reducing condition and visualized by coomassie blue stain.

15% SDS-PAGE (3ug)