abcam

## Product datasheet

# Recombinant Human SCO1 protein ab151389

### Description

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| **Product name** | Recombinant Human SCO1 protein |
| **Purity** | > 95 % SDS-PAGE.<br>Greater than 95% as determined by reducing SDS-PAGE. |
| **Endotoxin level** | < 1.000 Eu/µg |
| **Expression system** | Escherichia coli |
| **Accession** | O75880 |
| **Protein length** | Protein fragment |
| **Animal free** | No |
| **Nature** | Recombinant |
| **Species** | Human |
| **Sequence** | GSPEFHMGKP LLGGPFSLTT HTGERKTDKD YLGQWLLIYF GFTHCPDVCP EELEKMIQVV DEIDSITTLP DLTPLFISID PERDTKEAIA NYVKEFSPKL VGLTGTREEV DQVARAYRVY YSPGPKDEDE DYIVDHTIIM YLIGPDGEFL DYFGQNKRKG EIAASIATHM RPYRKKS |
| **Predicted molecular weight** | 19 kDa |
| **Amino acids** | 132 to 300 |
| **Tags** | GST tag N-Terminus |

### Specifications

Our **Abpromise guarantee** covers the use of **ab151389** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

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| **Applications** | SDS-PAGE |
| **Form** | Lyophilized |

### Preparation and Storage

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| **Stability and Storage** | Shipped at 4°C. The lyophilized protein is stable for a few weeks at room temperature. Store at -20°C long term.<br>pH: 7.20<br>Constituents: 99% Phosphate Buffer, 0.02% DTT |

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| **Reconstitution** | Lyophilized from a 0.2 µM filtered solution. Always centrifuge tubes before opening. Do not mix by vortex or pipetting. Dissolve the lyophilized protein in 1X PBS. It is not recommended to reconstitute to a concentration less than 100 µg/ml.<br>Reconstituted protein solution can be stored at 4-7°C for 2-7 days. Aliquots of reconstituted samples are stable at < -20°C for 3 months. |

## General Info

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| **Function** | Thought to play a role in cellular copper homeostasis, mitochondrial redox signaling or insertion of copper into the active site of COX. |
| **Tissue specificity** | Predominantly expressed in tissues characterized by high rates of oxidative phosphorylation (OxPhos), including muscle, heart, and brain. |
| **Involvement in disease** | Defects in SCO1 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, excercise intolerance, developmental delay, delayed motor development and mental retardation. A subset of patients manifest Leigh syndrome. |
| **Sequence similarities** | Belongs to the SCO1/2 family. |
| **Cellular localization** | Mitochondrion. |



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