ISCU Lysate – Tissue Lysate

PRODUCT OVERVIEW

Product Name	ISCU, Lysate
Full Product Name	Human ISCU Protein Lysate
Product Gene Name	ISCU lysate
Product Synonym Names	ISCU; 2310020H20Rik; HML; hnifU; ISU2; NIFU; NIFUN
Host	Human
Form/Format	Lyophilized
Preparation and Storage	Store at -20 degree C Once reconsituted, aliquot and store any remaining material at-20 degree C to avoid repeated freeze-thaw cycles.
Molecular Weight	16.7kDa (154aa), confirmed by MALDI-TOF
OMIM	phenotype 611911
3D Structure	ModBase 3D Structure for Q9H1K1
Related Product Information for ISCU lysate	Human ISCU Protein Lysate 20ug is provided as a Lyophilized powder. This is a Recombinant Protein Lysate produced in HEK293T cells. This protein lysate can be reconsituted using SDS Sample Buffer.
Other Notes	Small volumes of ISCU lysate vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.
Searchable Terms for ISCU purchase	MBS8413714 is a lysate targeting ISCU. Also known as Human ISCU Protein Lysate. Host: Human.

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for ISCU. It may not necessarily be applicable to this product.

NCBI GI #	56699456
NCBI GeneID	23479
NCBI Accession #	NP_998760
UniProt Primary Accession #	Q9H1K1
UniProt Secondary Accession #	Q6P713; Q99617; Q9H1K2
UniProt Related Accession #	Q9H1K1
GenBank Nucleotide #	NM_213595
NCBI Official Full Name	iron-sulfur cluster assembly enzyme ISCU, mitochondrial isoform 2
NCBI Official Synonym Full Names	iron-sulfur cluster assembly enzyme
NCBI Official Symbol	ISCU
NCBI Official Synonym Symbols	HML; ISU2; NIFU; NIFUN; hnifU; 2310020H20Rik
UniProt Protein Name	Iron-sulfur cluster assembly enzyme ISCU, mitochondrial
UniProt Synonym Protein Names	NifU-like N-terminal domain-containing protein; NifU-like protein

Protein Family	Iron-sulfur cluster assembly enzyme
UniProt Gene Name	ISCU
UniProt Synonym Gene Names	NIFUN
UniProt Entry Name	ISCU_HUMAN
NCBI Protein Information	iron-sulfur cluster assembly enzyme ISCU, mitochondrial
NCBI Summary for ISCU	This gene encodes a component of the iron-sulfur (Fe-S) cluster scaffold. Fe-S clusters are cofactors that play a role in the function of a diverse set of enzymes, including those that regulate metabolism, iron homeostasis, and oxidative stress response. Alternative splicing results in transcript variants encoding different protein isoforms that localize either to the cytosol or to the mitochondrion. Mutations in this gene have been found in patients with hereditary myopathy with lactic acidosis. A disease-associated mutation in an intron may activate a cryptic splice site, resulting in the production of a splice variant encoding a putatively non-functional protein. A pseudogene of this gene is present on chromosome 1. [provided by RefSeq, Feb 2016]
UniProt Comments for ISCU	ISCU: Involved in the assembly or repair of the [Fe-S] clusters present in iron-sulfur proteins. Binds iron. Defects in ISCU are the cause of myopathy with exercise intolerance Swedish type (MEIS); also known as myopathy with deficiency of succinate dehydrogenase and aconitase or myoglobinuria due to abnormal glycolysis or hereditary myopathy with lactic acidosis (HML). This autosomal recessive metabolic disease is characterized by lifelong severe exercise intolerance, in which minor exertion causes fatigue of active muscles, shortness of breath, and cardiac palpitations in association with lactic acidosis. The biochemical phenotype is characterized by a deficiency in mitochondrial iron-sulfur proteins and impaired muscle oxidative metabolism. Belongs to the NifU family. 2 isoforms of the human protein are produced by alternative splicing. Protein type: Mitochondrial Chromosomal Location of Human Ortholog: 12q24.1 Cellular Component: mitochondrion; mitochondrial matrix; cytoplasm; cytosol; nucleus Molecular Function: protein binding; iron ion binding; iron-sulfur cluster binding; protein complex scaffold Biological Process: iron-sulfur cluster assembly; nitrogen fixation Disease: Myopathy With Lactic Acidosis, Hereditary
Research Articles on ISCU	**1.** We have shown that ASO treatment diminished aberrant splicing and increased ISCU protein levels in both patient fibroblasts and patient myotubes in a concentration dependent fashion. Upon ASO treatment, levels of SDHB in patient myotubular cell lines increased to levels observed in control myotubular cell lines
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