Datasheet for ABIN7320070

ACO2 Protein (GST tag,His tag)

1 Image

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Overview

Quantity:	100 µg
Target:	ACO2
Origin:	Mouse
Source:	Baculovirus infected Insect Cells
Protein Type:	Recombinant
Purification tag / Conjugate:	This ACO2 protein is labelled with GST tag,His tag.

Product Details

Purpose:	Recombinant Mouse ACO2/Aconitase 2 Protein (His & GST Tag)
Sequence:	Gln 28-Gln 780
Characteristics:	A DNA sequence encoding the mouse ACO2 (Q99KI0) (Gln 28-Gln 780) was fused with the N-terminal polyhistidine-tagged GST tag at the N-terminus.
Purity:	> 90 % as determined by SDS-PAGE
Endotoxin Level:	< 1.0 EU per µg of the protein as determined by the LAL method.

Target Details

Target:	ACO2
Alternative Name:	ACO2/Aconitase 2 (ACO2 Products)
Background:	Background: A homozygous missense mutation was identified in the ACO2 gene (c.124T>G p.Phe414Val) that segregated with HSP complicated by intellectual disability and microcephaly. Lymphoblastoid cell lines of homozygous carrier patients revealed significantly decreased

Target Details

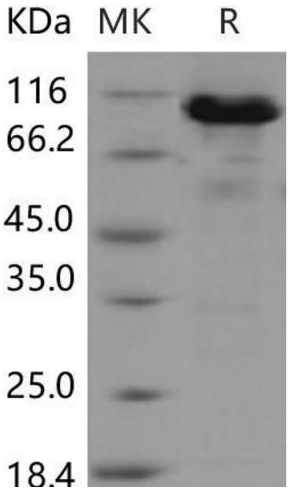
	activity of the mitochondrial aconitase enzyme and defective mitochondrial respiration. ACO2 encodes mitochondrial aconitase, an essential enzyme in the Krebs cycle. Recessive mutations in this gene have been previously associated with cerebellar ataxia. We found homozygous or compound heterozygous missense and frameshift mutations in the gene encoding mitochondrial aconitase (ACO2), a tricarboxylic acid cycle enzyme, catalysing interconversion of citrate into isocitrate. Unlike wild type ACO2, all mutant ACO2 proteins failed to complement the respiratory growth of a yeast aco1-deletion strain. The study shows that autosomal recessive ACO2 mutations can cause either isolated or syndromic optic neuropathy. This observation identifies ACO2 as the second gene responsible for non-syndromic autosomal recessive optic neuropathies and provides evidence for a genetic overlap between isolated and syndromic forms, giving further support to the view that optic atrophy is a hallmark of defective mitochondrial energy supply. Synonym: Aco-2,Aco3,D10Wsu183e
Molecular Weight:	110 kDa
UniProt:	Q99KI0

Application Details

Restrictions:	For Research Use only

Handling

Format:	Lyophilized
Reconstitution:	Please refer to the printed manual for detailed information.
Buffer:	Lyophilized from sterile 50 mM Tris, 100 mM NaCl, 10 % glycerol, 0.5 mM GSH, pH 8.0
Storage:	4 °C,-20 °C,-80 °C
Storage Comment:	Generally, lyophilized proteins are stable for up to 12 months when stored at -20 to -80°C. Reconstituted protein solution can be stored at 4-8°C for 2-7 days. Aliquots of reconstituted samples are stable at < -20°C for 3 months.



Western Blotting

Image 1.