10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

BCS1L Antibody (monoclonal) (M01)

Mouse monoclonal antibody raised against a partial recombinant BCS1L.
Catalog # AT1287a

Product Information

Application	WB
Primary Accession	Q9Y276
Other Accession	NM_004328
Reactivity	Human
Host	mouse
Clonality	monoclonal
Isotype	IgG1 Kappa
Clone Names	5F3
Calculated MW	47534

Additional Information

Gene ID	617
Other Names	Mitochondrial chaperone BCS1, h-BCS1, BCS1-like protein, BCS1L, BCS1
Target/Specificity	BCS1L (NP_004319, 320 a.a. ~ 418 a.a) partial recombinant protein with GST tag. MW of the GST tag alone is 26 KDa.
Dilution	WB~~1:500~1000
Format	Clear, colorless solution in phosphate buffered saline, pH 7.2 .
Storage	Store at -20°C or lower. Aliquot to avoid repeated freezing and thawing.
Precautions	BCS1L Antibody (monoclonal) (M01) is for research use only and not for use in diagnostic or therapeutic procedures.

Background

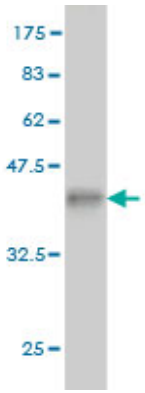
This gene encodes a homolog of the S. cerevisiae bcs1 protein which is involved in the assembly of complex III of the mitochondrial respiratory chain. The encoded protein does not contain a mitochondrial targeting sequence but experimental studies confirm that it is imported into mitochondria. Mutations in this gene are associated with mitochondrial complex III deficiency and the GRACILE syndrome. Two alternatively spliced transcripts encoding the same protein have been described.

References

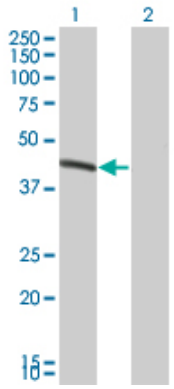
1.Characterization of complex III deficiency and liver dysfunction in GRACILE syndrome caused by a BCS1L mutation.Kotarsky H, Karikoski R, Morgelin M, Marjavaara S, Bergman P, Zhang DL, Smet J, van Coster R,

Fellman F.Mitochondrion (2010), doi:10.1016/ j.mito.2010.05.0092.Pathogenic mutations in the 5' untranslated region of BCS1L mRNA in mitochondrial complex III deficiency.Gil-Borlado MC, Gonzalez-Hoyuela M, Blazquez A, Garcia-Silva MT, Gabaldon T, Manzanares J, Vara J, Martin MA, Seneca S, Arenas J, Ugalde C.Mitochondrion. 2009 Sep;9(5):299-305. Epub 2009 Apr 21.

Images



Antibody Reactive Against Recombinant Protein.Western Blot detection against Immunogen (36.63 KDa) .



Western Blot analysis of BCS1L expression in transfected 293T cell line by BCS1L monoclonal antibody (M01), clone 5F3.

Lane 1: BCS1L transfected lysate(47.534 KDa).
Lane 2: Non-transfected lysate.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.