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Diagnostik & molekulare Diagnostik



Laborgeräte & Service

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Lieferung & Zahlungsart

siehe unsere [Liefer- und Versandbedingungen](#)

Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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BCS1L monoclonal antibody (M01), clone 5F3

Catalog # : H00000617-M01

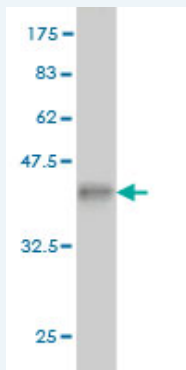
規格 : [100 ug]

[List All](#)

Specification

Product Description:	Mouse monoclonal antibody raised against a partial recombinant BCS1L.
Immunogen:	BCS1L (NP_004319, 320 a.a. ~ 418 a.a) partial recombinant protein with GST tag. MW of the GST tag alone is 26 KDa.
Sequence:	ASTEARIVFMTTNHVDRLDPALIRPGRVDLKEYVGYCSHWQLTQMFQRF YPGQAPSLAENFAEHVLRATNQISPAQVQGYFMLYKNDPVGAIHNAESLR
Host:	Mouse
Reactivity:	Human
Isotype:	IgG1 Kappa

Quality Control Testing: Antibody Reactive Against Recombinant Protein.



Western Blot detection against Immunogen (36.63 KDa) .

Storage Buffer: In 1x PBS, pH 7.4

Storage Instruction: Store at -20°C or lower. Aliquot to avoid repeated freezing and thawing.

MSDS: [Download](#)

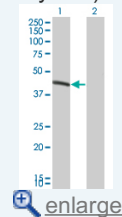
Datasheet: [Download](#)

Publication Reference

- 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.2010.05.009
- 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.

Application Image

Western Blot (Transfected lysate)

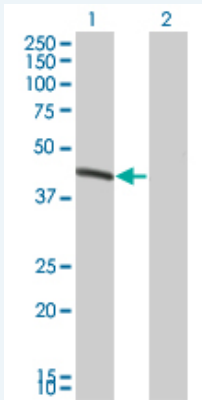


[enlarge](#)

Western Blot (Recombinant protein)

ELISA

Western Blot (Transfected lysate)



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.

 [Protocol Download](#)

Western Blot (Recombinant protein)

 [Protocol Download](#)

ELISA

Gene Information

Entrez GeneID: [617](#)

GeneBank [NM_004328](#)

Accession#:

Protein [NP_004319](#)

Accession#:

Gene Name: BCS1L

Gene Alias: BCS,BCS1,BJS,FLNMS,GRACILE,Hs.6719,PTD,h-BCS

Gene BCS1-like (yeast)

Description:

Omim ID: [124000](#), [256000](#), [262000](#), [603358](#), [603647](#)

Gene Ontology: [Hyperlink](#)

Gene Summary: 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. [provided by RefSeq]

Other Designations: BCS1-like,BCS1-like protein,OTTHUMP00000164147,mitochondrial chaperone BCS1,mitochondrial complex III assembly