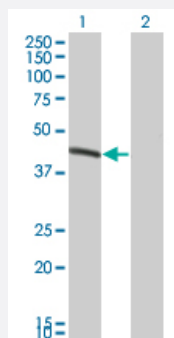


BCS1L monoclonal antibody (M01), clone 5F3

Catalog # H00000617-M01

Size 100 ug

Applications

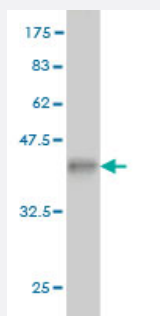


Western Blot (Transfected lysate)

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.



Western Blot detection against Immunogen (36.63 KDa).

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	ASTEARIVFMTTNHVDRLDPALIRPGRVDLKEYVGYCSHWQLTQMFQRFYPGQAPSLAENFAEHVLRATNQISPAQVQGYFMLYKNDPVGAIHNAESLR
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.

Applications

- 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 Info — BCS1L

Entrez GeneID

[617](#)

GeneBank Accession#

[NM_004328](#)

Protein Accession#

[NP_004319](#)

Gene Name

BCS1L

Gene Alias

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

Gene Description

BCS1-like (yeast)

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

Publication Reference

- [The mitochondrial peptidase, neurolysin, regulates respiratory chain supercomplex formation and is necessary for AML viability.](#)

Mirali S, Botham A, Voisin V, Xu C, St-Germain J, Sharon D, Hoff FW, Qiu Y, Hurren R, Gronda M, Jitkova Y, Nachmias B, MacLean N, Wang X, Arruda A, Minden MD, Horton TM, Kornblau SM, Chan SM, Bader GD, Raught B, Schimmer AD.

Science Translational Medicine 2020 Apr; 12(538):eaaz8264.

Application: WB-Tr, Human, OCI-AML2 cells

- [Molecular genetic investigations identify new clinical phenotypes associated with BCS1L-related mitochondrial disease.](#)

Oláhová M, Berti CC, Collier JJ, Alston CL, Jameson E, Jones SA, Edwards N, He L, Chinnery PF, Horvath R, Goffrini P, Taylor RW, Sayer JA.

Human Molecular Genetics 2019 Aug; [Epub].

Application: WB, Human, Fibroblasts, Skeletal muscles

- [Cellular pathophysiological consequences of BCS1L mutations in mitochondrial complex III enzyme deficiency.](#)

Maria Moran, Lorena Marin-Buena, M Carmen Gil-Borlado, Henry Rivera, Alberto Blazquez, Sara Seneca, Maria Vazquez-Lopez, Joaquin Arenas, Miguel A Martin, Cristina Ugalde.

Human Mutation 2010 Aug; 31(8):930.

Application: WB-Ce, Human, Human fibroblasts

- [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 Aug; 10(5):497.

Application: WB-Ce, WB-Ti, Human, Human fibroblasts, Livers

- [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.

Application: WB-Ce, Human, Human fibroblasts