

Tafazzin (JK-2): sc-100870

BACKGROUND

Tafazzin protein is a single-pass membrane protein that is abundant in cardiac and skeletal muscle, where it influences mitochondrial structure. There are various isoforms associated with Tafazzin, most of which are ubiquitous. Isoforms with hydrophobic N-terminal domains are membrane anchored, whereas the short isoforms that lack a hydrophobic leader sequence may exist as cytoplasmic proteins. The isoforms that lack the N-terminal domain are not found in cardiac or skeletal muscle, rather they are located in fibroblasts and leukocytes. Mutations in the Tafazzin gene are associated with various diseases, including dilated cardiomyopathy (DCM), hypertrophic DCM, endocardial fibroelastosis, left ventricular noncompaction (LVNC) and Barth syndrome (BTHS), a severe inherited disorder marked by neutropenia, cardiac and skeletal myopathy and short stature.

REFERENCES

- Gu, Z., Valianpour, F., Chen, S., Vaz, F.M., Hakkaart, G.A., Wanders, R.J. and Greenberg, M.L. 2003. Aberrant cardiolipin metabolism in the yeast taz1 mutant: a model for Barth syndrome. *Mol. Microbiol.* 51: 149-158.
- Schlame, M., Kelley, R.I., Feigenbaum, A., Towbin, J.A., Heerdt, P.M., Schieble, T., Wanders, R.J., DiMauro, S. and Blanck, T.J. 2003. Phospholipid abnormalities in children with Barth syndrome. *J. Am. Coll. Cardiol.* 42: 1994-1999.
- Lu, B., Kelher, M.R., Lee, D.P., Lewin, T.M., Coleman, R.A., Choy, P.C. and Hatch, G.M. 2004. Complex expression pattern of the Barth syndrome gene product Tafazzin in human cell lines and murine tissues. *Biochem. Cell Biol.* 82: 569-576.
- Testet, E., Laroche-Traineau, J., Noubhani, A., Coulon, D., Bunoust, O., Camougrand, N., Manon, S., Lessire, R. and Bessoule, J.J. 2005. Ypr140wp, "the yeast Tafazzin", displays a mitochondrial lysophosphatidylcholine (lyso-PC) acyltransferase activity related to triacylglycerol and mitochondrial lipid synthesis. *Biochem. J.* 387: 617-626.
- Xu, Y., Sutachan, J.J., Plesken, H., Kelley, R.I. and Schlame, M. 2005. Characterization of lymphoblast mitochondria from patients with Barth syndrome. *Lab. Invest.* 85: 823-830.
- Brandner, K., Mick, D.U., Frazier, A.E., Taylor, R.D., Meisinger, C. and Rehling, P. 2005. Taz1, an outer mitochondrial membrane protein, affects stability and assembly of inner membrane protein complexes: implications for Barth syndrome. *Mol. Biol. Cell* 16: 5202-5214.

CHROMOSOMAL LOCATION

Genetic locus: TAZ (human) mapping to Xq28; Taz (mouse) mapping to X A7.3.

SOURCE

Tafazzin (JK-2) is a mouse monoclonal antibody raised against recombinant Tafazzin of human origin.

PRODUCT

Each vial contains 100 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

Tafazzin (JK-2) is recommended for detection of Tafazzin of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for Tafazzin siRNA (h): sc-61637, Tafazzin siRNA (m): sc-61638, Tafazzin shRNA Plasmid (h): sc-61637-SH, Tafazzin shRNA Plasmid (m): sc-61638-SH, Tafazzin shRNA (h) Lentiviral Particles: sc-61637-V and Tafazzin shRNA (m) Lentiviral Particles: sc-61638-V.

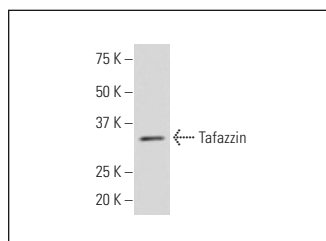
Molecular Weight of Tafazzin: 34 kDa.

Positive Controls: human uterus myoma tissue extract or SW-13 cell lysate: sc-24778.

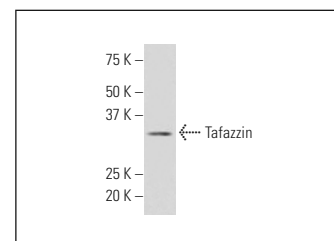
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).

DATA



Tafazzin (JK-2): sc-100870. Western blot analysis of Tafazzin expression in SW-13 whole cell lysate.



Tafazzin (JK-2): sc-100870. Western blot analysis of Tafazzin expression in human uterus myoma tissue extract.

SELECT PRODUCT CITATIONS

- Chao, H., Anthonymuthu, T.S., Kenny, E.M., Amoscato, A.A., Cole, L.K., Hatch, G.M., Ji, J., Kagan, V.E. and Bayir, H. 2018. Disentangling oxidation/hydrolysis reactions of brain mitochondrial cardiolipins in pathogenesis of traumatic injury. *JCI Insight* 3: e97677.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.