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TMEM70 Antibody

Catalog # ASC11085

Product Information

Application	WB, IF, E, IHC-P
Primary Accession	Q9BUB7
Other Accession	NP_060336, 34147498
Reactivity	Human, Mouse, Rat
Host	Rabbit
Clonality	Polyclonal
Isotype	IgG
Calculated MW	28969
Concentration (mg/ml)	1 mg/mL
Conjugate	Unconjugated
Application Notes	TMEM70 antibody can be used for detection of TMEM70 by Western blot at 1 g/mL. Antibody can also be used for immunohistochemistry starting at 5 g/mL. For immunofluorescence start at 20 g/mL.

Additional Information

Gene ID	54968
Other Names	Transmembrane protein 70, mitochondrial, TMEM70
Target/Specificity	TMEM70;
Reconstitution & Storage	TMEM70 antibody can be stored at 4℃ for three months and -20℃, stable for up to one year. As with all antibodies care should be taken to avoid repeated freeze thaw cycles. Antibodies should not be exposed to prolonged high temperatures.
Precautions	TMEM70 Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	TMEM70 (HGNC:26050)
Function	Scaffold protein that participates in the c-ring assembly of mitochondrial ATP synthase (F(1)F(0) ATP synthase or complex V) by facilitating the membrane insertion and oligomer formation of the subunit c/ATP5MC1 through its interaction (PubMed:31652072, PubMed:32275929, PubMed:33359711, PubMed:33753518). Therefore, participates in the early stage of mitochondrial ATP synthase biogenesis and also protects subunit c/ATP5MC1 against intramitochondrial proteolysis (PubMed:18953340, PubMed:20937241, PubMed:31652072, PubMed:33359711). In addition, binds the mitochondrial proton-transporting ATP synthase complexes I and may play a role in the stability of its membrane-bound subassemblies

(PubMed:32275929).

Cellular Location	Mitochondrion inner membrane; Multi-pass membrane protein. Note=Mostly located within the inner cristae membrane
Tissue Location	Lower expressed in the heart than in the liver (at protein level).

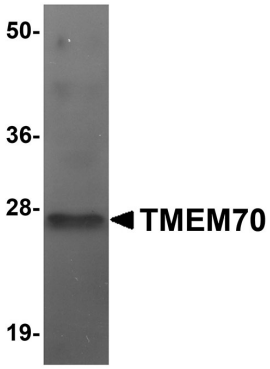
Background

TMEM70 Antibody: TMEM70 is a recently identified mitochondrial protein that is thought to play a role in the biogenesis of the ATP synthase in higher eukaryotes. Mutations in this gene result in early neonatal onset of hypotonia, hypertrophic cardiomyopathy, lactic acidos and 3-methylglutaconic aciduria (3-MGC-uria), and usually cause death within the first six weeks of life, although some patients survive much longer. Little is known of the role of TMEM70, but it is conserved across multicellular eukaryotic organisms. It contains a conserved DUF1301 domain and two putative transmembrane regions.

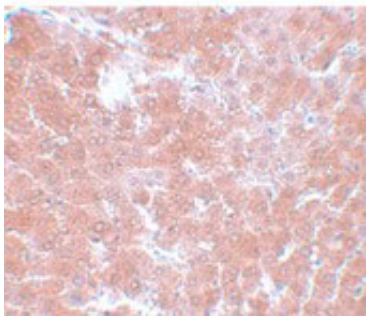
References

Cizkova A, Stranecky V, Mayr JA, et al. TMEM70 mutations cause isolated ATP synthase deficiency and neonatal mitochondrial encephalocardiomyopathy. Nat. Genet.2008; 11:1288-90.
Honzik T, Tesarova M, Mayr JA, et al. Mitochondrial encephalocardio-myopathy with early neonatal onset due to TMEM70 mutation. Arch. Dis. Child.2010; 95:296-301.
Houstek J, Kmoch S, and Zeman J. TMEM70 protein - a novel ancillary factor of mammalian ATP synthase. Biochim. Biophys. Acta2009; 1787:529-32.

Images

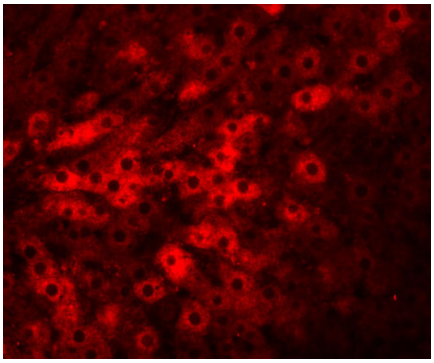


Western blot analysis of TMEM70 in human liver tissue lysate with TMEM70 antibody at 1 µg/mL.



Immunohistochemistry of TMEM70 in rat liver tissue with TMEM70 antibody at 5 µg/mL.

Immunofluorescence of TMEM70 in rat liver tissue with TMEM70 antibody at 20 µg/mL.



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