Mouse monoclonal to human Deafness dystonia protein 1 [2F11]: IgG

Catalogue No.:	M-831-100
Description:	TIMM8A is a mitochondrial intermembrane chaperone that is involved in the import and insertion of some multi-pass transmembrane proteins into the mitochondrial inner membrane. It also acts as a chaperone-like protein that protects the hydrophobic precursors from aggregation and guide them through the mitochondrial intermembrane space. TIMM8A is probably necessary for normal neurologic development. TIMM8A is a heterohexamer composed of three TIMM8A and three TIMM13 subunits, named soluble 70KDa complex. It associates with the TIMM22 complex. It is located at the mitochondrion inner membrane and is highly expressed in fetal and adult brain, followed by fetal lung, liver and kidney. Defects in TIMM8A cause Mohr-Tranebjaerg syndrome (MTS) also known as dystonia-deafness syndrome (DDS) or X-linked progressive deafnes type 1 (DFN-1). This is a recessive neurodegenerative syndrome characterised by progressive deafness, dystonia, spasticity, dysphagia, mental deterioration, paranoia and cortical blindness. Defects in TIMM8A are also the cause of Jensen syndrom, also known as opticoacoustic nerve atrophy with dementia. This is an X-linked disease characterised by deafness, blindess and muscle weakness.
Batch No.:	See product label
Unit size:	100 ug
Antigen:	Partial recombinant protein of human Deafness dystonia protein 1 (aa 9 to 98) with a GST tag.
Clone:	2F11
Other Names:	Mitochondrial import inner membrane translocase subunit Tim8 A; X-linked deafness dystonia protein; TIMM8A; DDP; DDP1; TIM8A
Accession:	TIM8A_HUMAN
Produced in:	Mouse
Purity:	Protein G purified immunoglobulin
Applications:	This antibody is recommended for WB, IHC and sandwich ELISA. For IHC, a concentration of 3 ug/mL is recommended. Biosensis recommends optimal dilutions/concentrations should be determined by the end user.
Specificity:	Specificity has been confirmed by WB and direct ELISA against the antigen.
Cross-reactivity:	Human. Other species have not been tested.
Form:	Lyophilised from PBS pH 7.2
Reconstitution:	Reconstitute in 100 uL of sterile water. Centrifuge to remove any insoluble material.
Storage:	After reconstitution keep aliquots at -20C for higher stability or at 2-8C with an appropriate antibacterial agent. Glycerol (1:1) may be added for additional stability. Avoid repetitive freeze/thaw cycles.
Expiry Date:	12 months after purchase

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No image

Immunohistochemical detection of Deafness dystonia protein 1 in paraffin-embedded, formalin fixed human liver tissue using mouse monoclonal antibody to Deafness dystonia protein 1, catalogue number M-831-100.

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