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AIPL1 Antibody (monoclonal) (M04)

Mouse monoclonal antibody raised against a full-length recombinant AIPL1.
Catalog # AT1078a

Product Information

Application	WB, IHC, IF, E
Primary Accession	Q9NZN9
Other Accession	BC012055
Reactivity	Human
Host	mouse
Clonality	monoclonal
Isotype	IgG2a Kappa
Clone Names	10
Calculated MW	43903

Additional Information

Gene ID	23746
Other Names	Aryl-hydrocarbon-interacting protein-like 1, AIPL1, AIPL2
Target/Specificity	AIPL1 (AAH12055, 1 a.a. ~ 384 a.a) full-length recombinant protein with GST tag. MW of the GST tag alone is 26 KDa.
Dilution	WB~~1:500~1000 IHC~~1:100~500 IF~~1:50~200 E~~N/A
Format	Clear, colorless solution in phosphate buffered saline, pH 7.2 .
Storage	Store at -20°C or lower. Aliquot to avoid repeated freezing and thawing.
Precautions	AIPL1 Antibody (monoclonal) (M04) is for research use only and not for use in diagnostic or therapeutic procedures.

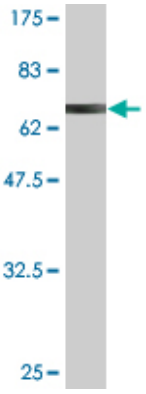
Background

Leber congenital amaurosis (LCA) accounts for at least 5% of all inherited retinal disease and is the most severe inherited retinopathy with the earliest age of onset. Individuals affected with LCA are diagnosed at birth or in the first few months of life with severely impaired vision or blindness, nystagmus and an abnormal or flat electroretinogram. The photoreceptor/pineal -expressed gene, AIPL1, encoding aryl-hydrocarbon interacting protein-like 1, was mapped within the LCA4 candidate region. The protein contains three tetratricopeptide motifs, consistent with nuclear transport or chaperone activity. AIPL1 mutations may cause approximately 20% of recessive LCA.

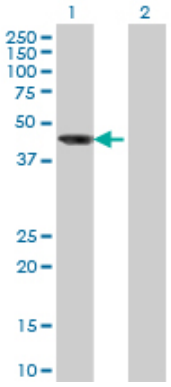
References

The Leber congenital amaurosis protein, AIPL1, is needed for the viability and functioning of cone photoreceptor cells. Kirschman LT, et al. Hum Mol Genet, 2010 Mar 15. PMID 20042464.Differential macular morphology in patients with RPE65-, CEP290-, GUCY2D-, and AIPL1-related Leber congenital amaurosis. Pasadhika S, et al. Invest Ophthalmol Vis Sci, 2010 May. PMID 19959640.Mutations that are a common cause of Leber congenital amaurosis in northern America are rare in southern India. Sundaresan P, et al. Mol Vis, 2009 Sep 4. PMID 19753312.The Leber congenital amaurosis protein AIPL1 functions as part of a chaperone heterocomplex. Hidalgo-de-Quintana J, et al. Invest Ophthalmol Vis Sci, 2008 Jul. PMID 18408180.Identification of mutations in the AIPL1, CRB1, GUCY2D, RPE65, and RPGRIP1 genes in patients with juvenile retinitis pigmentosa. Booij JC, et al. J Med Genet, 2005 Nov. PMID 16272259.

Images

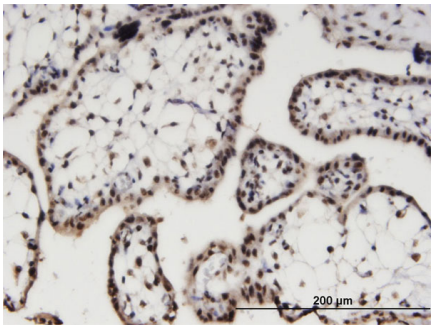


Antibody Reactive Against Recombinant Protein.Western Blot detection against Immunogen (67.98 KDa) .

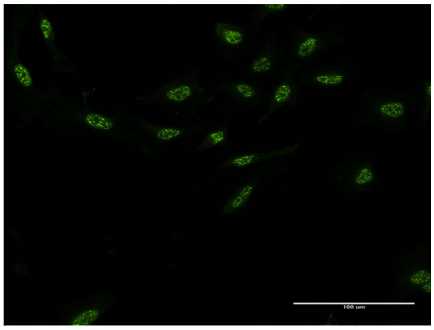


Western Blot analysis of AIPL1 expression in transfected 293T cell line by AIPL1 monoclonal antibody (M04), clone 1E1.

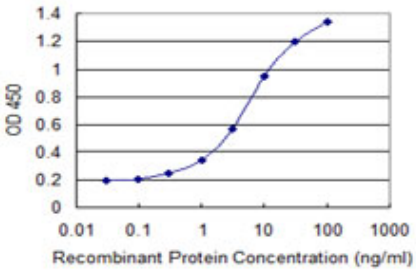
Lane 1: AIPL1 transfected lysate(43.9 KDa).
Lane 2: Non-transfected lysate.



Immunoperoxidase of monoclonal antibody to AIPL1 on formalin-fixed paraffin-embedded human placenta. [antibody concentration 3 ug/ml]



Immunofluorescence of monoclonal antibody to AIPL1 on HeLa cell . [antibody concentration 10 ug/ml]



Detection limit for recombinant GST tagged AIPL1 is 0.1 ng/ml as a capture antibody.