

Espin/DFNB36 Rabbit pAb

Catalog # AP98881

Product Information

Application	IHC-P, IHC-F, IF, E
Primary Accession	B1AK53
Other Accession	B1AK53
Host	Rabbit
Clonality	Polyclonal
Calculated MW	91733
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Espin/DFNB36
Epitope Specificity	601-700/854
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Contains 9 ANK repeats.Contains 1 WH2 domain.
SUBUNIT	Monomer (By similarity). Binds F-actin in a Ca(2+)-resistant fashion (By similarity). Interacts (via N-terminal) with BAIAP2 (via SH3-domain) (By similarity). Interacts with PFN2 (By similarity).
DISEASE	Deafness, autosomal recessive, 36, with or without vestibular involvement (DFNB36) [MIM:609006]: A form of non-syndromic sensorineural hearing loss. Sensorineural deafness results from damage to the neural receptors of the inner ear, the nerve pathways to the brain, or the area of the brain that receives sound information. DFNB36 is characterized by prelingual, profound hearing loss, and vestibular areflexia in some patients. Note=The disease is caused by mutations affecting the gene represented in this entry.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes a multifunctional actin-bundling protein. It plays a major role in regulating the organization, dimensions, dynamics, and signaling capacities of the actin filament-rich, microvillus-type specializations that mediate sensory transduction in various mechanosensory and chemosensory cells. Mutations in this gene are associated with autosomal recessive neurosensory deafness, and autosomal dominant sensorineural deafness without vestibular involvement. [provided by RefSeq, Nov 2009]

Additional Information

Gene ID	83715
Other Names	DFNB36; LP2654; USH1M; ESPN_HUMAN; ESPN; Autosomal recessive deafness type 36 protein; Ectoplasmic specialization protein; espin; deafness, autosomal recessive 36
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:500 0-10000

Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
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Protein Information

Name	ESPN
Synonyms	DFNB36
Function	Multifunctional actin-bundling protein. Plays a major role in regulating the organization, dimension, dynamics and signaling capacities of the actin filament-rich microvilli in the mechanosensory and chemosensory cells (PubMed: 29572253). Required for the assembly and stabilization of the stereociliary parallel actin bundles. Plays a crucial role in the formation and maintenance of inner ear hair cell stereocilia (By similarity). Involved in the elongation of actin in stereocilia (PubMed: 29572253). In extrastricular hair cells, required for targeting MYO3B to stereocilia tips, and for regulation of stereocilia diameter and staircase formation.
Cellular Location	Cytoplasm, cytoskeleton {ECO:0000250 UniProtKB:Q9ET47}. Cell projection, stereocilium. Cell projection, microvillus

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