

## Rabbit Polyclonal Anti-Usherin antibody

Catalog Number: USH-121AP

Lot Number:

### General Information

<b>Product</b>	Usherin Antibody
<b>Description</b>	Usherin isoform A Antibody C-epitope
<b>Accession #</b>	Uniprot: O75445 NCBI: NP_009054.5
<b>Verified Applications</b>	CM, ELISA, ICC, IF, IHC, IP, WB
<b>Species Cross Reactivity</b>	Human
<b>Host</b>	Rabbit
<b>Immunogen</b>	Synthetic peptide taken within amino acid region 150-180 on Usher Type 2A protein.
<b>Alternative Nomenclature</b>	dJ1111A8.1 antibody, US1 antibody, USH1A antibody, Usher syndrome type Ia protein antibody, Usherin antibody

### Physical Properties

<b>Quantity</b>	100 µg
<b>Volume</b>	200 µl
<b>Form</b>	Affinity Purified Immunoglobulins
<b>Immunoglobulin &amp; Concentration</b>	0.62 mg/ml IgG in antibody stabilization buffer
<b>Determinant</b>	C-epitope
<b>Storage</b>	Store at -20°C for long term storage.

### Recommended Dilutions

<b>DOT Blot</b>	1:4,000
<b>ELISA</b>	1:4,000
<b>Immunocytochemistry</b>	1:250
<b>Immunofluorescence</b>	1:250
<b>Immunohistochemistry</b>	1:250
<b>Immunoprecipitation</b>	1:200
<b>Western Blot</b>	1:500

## Related Products

## Catalog #

<b>BIOTIN-Conjugated</b>	USH.121-BIOTIN
<b>FITC-Conjugated</b>	USH.121-FITC
<b>Antigenic Blocking Peptide</b>	P-USH.121
<b>Western Blot Positive Control</b>	PC-USH
<b>N-epitope</b>	USH-101AP
<b>Mid Region epitope</b>	USH-112AP

## Overview:

Usher syndrome, an autosomal recessive heterogeneous disorder, is the most common cause of deafness and blindness in adults and affects 3-6% children born with hearing impairment. The affected individuals have sensory-neural deficiencies at birth and subsequently develop progressive retinitis pigmentosa (RP). Three forms of Usher syndrome have been characterized, Usher Type I, II and III which can be distinguished based on severity of hearing loss and vestibular involvement. Type I patients are profoundly deaf while type II (most common form of Usher syndrome) patients are mildly deaf but have normal vestibular responses (1). Ush2A gene has been mapped on a 1 mega base pair interval between marker D1S474 and AFM144XF2. The Usher 2A gene expresses an extra cellular matrix protein that resembles unconventional Myosin and do not appear to have any un-conventional functional correlations. The human Usher Syndrome 2A (Ush2A) protein has 1272 amino acid (Approximate MW 153kDa) with several functional domains (Laminin type EGF like domain; Laminin-Type epidermal growth factor like domain and fibronectin type 3 like domain). The protein has a single transmembrane domain that anchor the protein to the cell membrane, the rest of the protein stays outside the cell.

The Ush2A protein is approximately 153kDa (1272 amino acids) with multiple conserved domains that is mainly expressed in RPEs. The Anti-Usher selective antibodies were generated against three regions: N-epitope, Mid-Region and C-epitope. These epitope are unique to Usherin protein. The polyclonal antibody strongly labels a 186kDa protein in RPE cell extracts. Anti-Ush2A-selective antibodies are also available in affinity-purified form for confocal, Western blotting and immunocytochemical analyses. Antibodies can be conjugated with fluorescent probes or secondary enzymes for an extra charge. Western blot positive control in ready-to-use SDS sample buffer and antigenic blocking peptides are available.

## References:

1. Judy JD., Weston M. D., Yao S. F., Hoover D. M. et. Al., Mutation of a gene encoding a protein with extracellular matrix motifs in Usher syndrome Type IIa. *Science* 280, 1753-1757; 1998.
2. Farooqui, S. M., Brock. W. J., A. Hamdi., Prasad. C. (1991) *J. Neurochem.* 57, 1363-1369.

\* For users who may require large amounts of the products listed above, please inquire about bulk material discounts.

This Product is for Research Use Only and is NOT intended for use in humans or clinical diagnosis.