

## Goat Anti-USH1C / Harmonin Antibody

Peptide-affinity purified goat antibody Catalog # AF2136a

#### **Specification**

## **Goat Anti-USH1C / Harmonin Antibody - Product Information**

**Application** WB, IHC **09Y6N9 Primary Accession** 

Other Accession NP 710142, 10083, 72088 (mouse), 308596

Human

Reactivity Predicted Mouse, Rat, Cow

Goat Clonality **Polyclonal** 

Concentration 100ug/200ul Isotype laG Calculated MW 62211

## Goat Anti-USH1C / Harmonin Antibody - Additional Information

# **Gene ID 10083**

#### **Other Names**

Harmonin, Antigen NY-CO-38/NY-CO-37, Autoimmune enteropathy-related antigen AIE-75, Protein PDZ-73, Renal carcinoma antigen NY-REN-3, Usher syndrome type-1C protein, USH1C, AIE75

Host

0.5 mg lgG/ml in Tris saline (20mM Tris pH7.3, 150mM NaCl), 0.02% sodium azide, with 0.5% bovine serum albumin

# **Storage**

Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

#### **Precautions**

Goat Anti-USH1C / Harmonin Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

#### **Goat Anti-USH1C / Harmonin Antibody - Protein Information**

#### Name USH1C

# **Synonyms** AIE75

#### **Function**

Anchoring/scaffolding protein that is a part of the functional network formed by USH1C, USH1G, CDH23 and MYO7A that mediates mechanotransduction in cochlear hair cells. Required for normal



development and maintenance of cochlear hair cell bundles (By similarity). As part of the intermicrovillar adhesion complex/IMAC plays a role in brush border differentiation, controlling microvilli organization and length. Probably plays a central regulatory role in the assembly of the complex, recruiting CDHR2, CDHR5 and MYO7B to the microvilli tips (PubMed:<a href="http://www.uniprot.org/citations/24725409" target="\_blank">24725409</a>, PubMed:<a href="http://www.uniprot.org/citations/26812018" target=" blank">26812018</a>).

#### **Cellular Location**

Cytoplasm, cytosol. Cytoplasm, cytoskeleton. Cell projection, microvillus Note=Colocalizes with F-actin (By similarity). Detected at the tip of cochlear hair cell stereocilia (By similarity). Enriched in microvilli of the intestinal brush border (PubMed:24725409, PubMed:32209652) {ECO:0000250|UniProtKB:Q9ES64, ECO:0000269|PubMed:24725409, ECO:0000269|PubMed:32209652}

#### **Tissue Location**

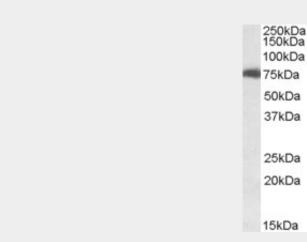
Expressed in small intestine, colon, kidney, eye and weakly in pancreas. Expressed also in vestibule of the inner ear

## Goat Anti-USH1C / Harmonin Antibody - Protocols

Provided below are standard protocols that you may find useful for product applications.

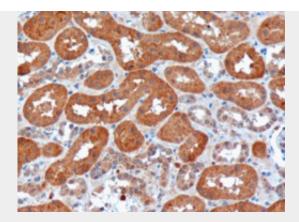
- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- Cell Culture

#### Goat Anti-USH1C / Harmonin Antibody - Images



AF2136a (0.1  $\mu$ g/ml) staining of HEK293 lysate (35  $\mu$ g protein in RIPA buffer). Primary incubation was 1 hour. Detected by chemiluminescence.





AF2136a (10  $\mu$ g/ml) staining of paraffin embedded Human Kidney. Microwaved antigen retrieval with citrate buffer pH 6, HRP-staining. (Data have been obtained with AF2136a from a previous batch).

# Goat Anti-USH1C / Harmonin Antibody - Background

This gene encodes a scaffold protein that functions in the assembly of Usher protein complexes. The protein contains PDZ domains, a coiled-coil region with a bipartite nuclear localization signal and a PEST degradation sequence. Defects in this gene are the cause of Usher syndrome type 1C and non-syndromic sensorineural deafness autosomal recessive type 18. Multiple transcript variants encoding different isoforms have been found for this gene.

#### **Goat Anti-USH1C / Harmonin Antibody - References**

Personalized smoking cessation: interactions between nicotine dose, dependence and quit-success genotype score. Rose JE, et al. Mol Med, 2010 Jul-Aug. PMID 20379614.

The structure of the harmonin/sans complex reveals an unexpected interaction mode of the two Usher syndrome proteins. Yan J, et al. Proc Natl Acad Sci U S A, 2010 Mar 2. PMID 20142502. Microarray-based mutation analysis of 183 Spanish families with Usher syndrome. Jaijo T, et al. Invest Ophthalmol Vis Sci, 2010 Mar. PMID 19683999.

Assembling stable hair cell tip link complex via multidentate interactions between harmonin and cadherin 23. Pan L, et al. Proc Natl Acad Sci U S A, 2009 Apr 7. PMID 19297620.

UMD-USHbases: a comprehensive set of databases to record and analyse pathogenic mutations and unclassified variants in seven Usher syndrome causing genes. Baux D, et al. Hum Mutat, 2008 Aug. PMID 18484607.