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LRP2 Rabbit pAb

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Catalog # AP94003

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	A2ARV4
Reactivity	Mouse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	519208
Physical State	Liquid
Immunogen	Recombinant mouse LRP2 protein
Epitope Specificity	4551-4660/4660
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Membrane; Single-pass type I membrane protein. Membrane, coated pit.
SIMILARITY	Belongs to the LDLR family.Contains 17 EGF-like domains.Contains 36 LDL-receptor class A domains.Contains 37 LDL-receptor class B repeats.
SUBUNIT	Binds plasminogen, extracellular matrix components, plasminogen activator-plasminogen activator inhibitor type I complex, apolipoprotein E-enriched beta-VLDL, lipoprotein lipase, lactoferrin, CLU/clusterin and calcium. Forms a multimeric complex together with a receptor-associated protein (RAP). Binds to ankyrin-repeat family A protein 2 (ANKRA2). Interacts with LRP2BP.
DISEASE	Defects in LRP2 are the cause of Donnai-Barrow syndrome (DBS) [MIM:222448]; also known as facio oculoacousticorenal syndrome (FOAR syndrome). DBS is a rare autosomal recessive disorder characterized by major malformations including agenesis of the corpus callosum, congenital diaphragmatic hernia, facial dysmorphology, ocular anomalies, sensorineural hearing loss and developmental delay. The FOAR syndrome was first described as comprising facial anomalies, ocular anomalies, sensorineural hearing loss, and proteinuria. DBS and FOAR were first described as distinct disorders but the classic distinguishing features between the 2 disorders were presence of proteinuria and absence of diaphragmatic hernia and corpus callosum anomalies in FOAR. Early reports noted that the 2 disorders shared many phenotypic features and may be identical. Although there is variability in the expression of some features (e.g. agenesis of the corpus callosum and proteinuria), DBS and FOAR are now considered to represent the same entity.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The protein encoded by this gene, low density lipoprotein-related protein 2 (LRP2) or megalin, is a multi-ligand endocytic receptor that is expressed in many different tissues but primarily in absorptive epithilial tissues such as the kidney. This glycoprotein has a large amino-terminal extracellular domain, a single transmembrane domain, and a short carboxy-terminal cytoplasmic tail. The extracellular ligand-binding-domains bind diverse macromolecules

including albumin, apolipoproteins B and E, and lipoprotein lipase. The LRP2 protein is critical for the reuptake of numerous ligands, including lipoproteins, sterols, vitamin-binding proteins, and hormones. This protein also has a role in cell-signaling; extracellular ligands include parathyroid horomones and the morphogen sonic hedgehog while cytosolic ligands include MAP kinase scaffold proteins and JNK interacting proteins. Recycling of this membrane receptor is regulated by phosphorylation of its cytoplasmic domain. Mutations in this gene cause Donnai-Barrow syndrome (DBS) and facio-oculoacoustico-renal syndrome (FOAR).[provided by RefSeq, Aug 2009].

Additional Information

Gene ID	14725
Other Names	Low-density lipoprotein receptor-related protein 2, LRP-2, Glycoprotein 330, gp330, Megalin, Lrp2
Target/Specificity	Absorptive epithelia, including renal proximal tubules.
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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.

Protein Information

Name	Lrp2
Function	Multiligand endocytic receptor. Acts together with CUBN to mediate endocytosis of high-density lipoproteins (PubMed:10766831). Mediates receptor-mediated uptake of polybasic drugs such as aprotinin, aminoglycosides and polymyxin B (By similarity). In the kidney, mediates the tubular uptake and clearance of leptin (PubMed:22841573). Also mediates transport of leptin across the blood-brain barrier through endocytosis at the choroid plexus epithelium (By similarity). Endocytosis of leptin in neuronal cells is required for hypothalamic leptin signaling and leptin-mediated regulation of feeding and body weight (PubMed:24825475). Mediates endocytosis and subsequent lysosomal degradation of CST3 in kidney proximal tubule cells (PubMed:17462596). Mediates renal uptake of 25-hydroxyvitamin D3 in complex with the vitamin D3 transporter GC/DBP (PubMed:10052453). Mediates renal uptake of metallothionein-bound heavy metals (By similarity). Together with CUBN, mediates renal reabsorption of myoglobin (By similarity). Mediates renal uptake and subsequent lysosomal degradation of APOM (By similarity). Plays a role in kidney selenium homeostasis by mediating renal endocytosis of selenoprotein SEPP1 (PubMed:18174160). Mediates renal uptake of the antiapoptotic protein BIRC5/survivin which may be important for functional integrity of the kidney (PubMed:23825075). Mediates renal uptake of matrix metalloproteinase MMP2 in complex with metalloproteinase inhibitor TIMP1 (PubMed:28659595). Mediates endocytosis of Sonic hedgehog protein N-product (ShhN), the active product of SHH (By similarity). Also mediates ShhN transcytosis (By similarity). In the embryonic neuroepithelium, mediates endocytic uptake and degradation of BMP4, is required for correct SHH localization in the ventral neural tube and plays a role in patterning of the ventral telencephalon (PubMed:15623804). Required at the onset of

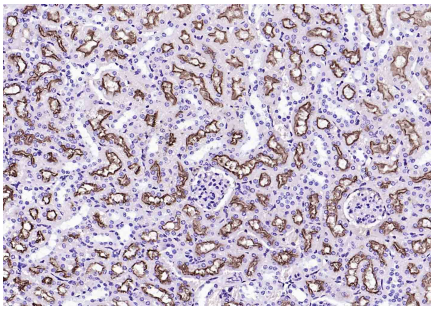
	neurulation to sequester SHH on the apical surface of neuroepithelial cells of the rostral diencephalon ventral midline and to control PTCH1-dependent uptake and intracellular trafficking of SHH (PubMed:22340494). During neurulation, required in neuroepithelial cells for uptake of folate bound to the folate receptor FOLR1 which is necessary for neural tube closure (PubMed:24639464). In the adult brain, negatively regulates BMP signaling in the subependymal zone which enables neurogenesis to proceed (PubMed:20460439). In astrocytes, mediates endocytosis of ALB which is required for the synthesis of the neurotrophic factor oleic acid (By similarity). Involved in neurite branching (PubMed:20637285). During optic nerve development, required for SHH-mediated migration and proliferation of oligodendrocyte precursor cells (PubMed:22354480). Mediates endocytic uptake and clearance of SHH in the retinal margin which protects retinal progenitor cells from mitogenic stimuli and keeps them quiescent (PubMed:26439398). Plays a role in reproductive organ development by mediating uptake in reproductive tissues of androgen and estrogen bound to the sex hormone binding protein SHBG (PubMed:16143106). Mediates endocytosis of angiotensin-2 (By similarity). Also mediates endocytosis of angiotensin 1-7 (By similarity). Binds to the complex composed of beta-amyloid protein 40 and CLU/APOJ and mediates its endocytosis and lysosomal degradation (By similarity). Required for embryonic heart development (PubMed:26822476). Required for normal hearing, possibly through interaction with estrogen in the inner ear (PubMed:17846082).
Cellular Location	Apical cell membrane; Single-pass type I membrane protein. Endosome lumen {ECO:0000250|UniProtKB:P98158}. Membrane, coated pit. Cell projection, dendrite. Cell projection, axon. Note=Localizes to brush border membranes in the kidney. In the endolymphatic sac of the inner ear, located in the lumen of endosomes as a soluble form {ECO:0000250|UniProtKB:P98158}
Tissue Location	In the inner ear, strongly expressed in the marginal cells of the stria vascularis (at protein level) (PubMed:17846082). In the female reproductive tract, expressed on the luminal side of the uterine epithelium (at protein level) (PubMed:16143106). In the adult brain, expressed in ependymal cells of the lateral ventricles where expression is restricted to the ependyma that faces the stem cell niche (at protein level) (PubMed:20460439) Expressed in neurons throughout the brain including in the hippocampus, limbic cortices and cerebellum (at protein level) (PubMed:20637285). In the developing optic nerve, expressed exclusively in astrocytes at 14.5 dpc, 16.5 dpc and 18.5 dpc (at protein level) (PubMed:22354480)

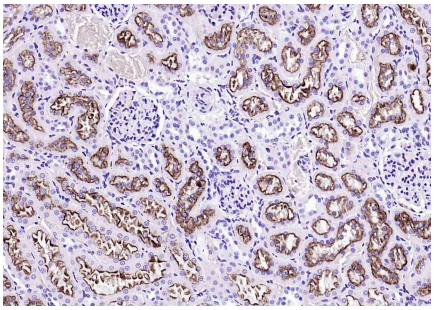
Background

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

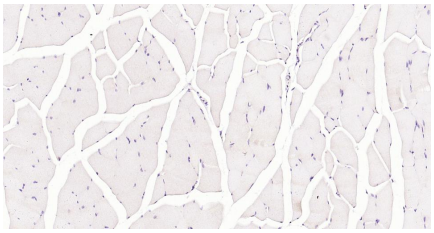
Images

Paraformaldehyde-fixed, paraffin embedded (mouse kidney); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Incubation with (LRP2) Polyclonal Antibody, Unconjugated (AP94003) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.

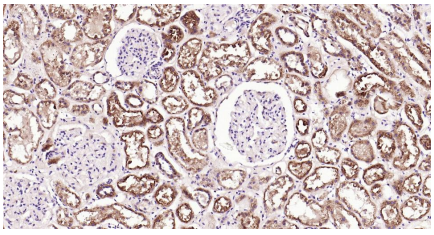




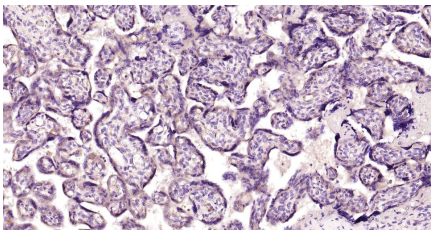
Paraformaldehyde-fixed, paraffin embedded (rat kidney); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Incubation with (LRP2) Polyclonal Antibody, Unconjugated (AP94003) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.



(Negative control) Paraformaldehyde-fixed, paraffin embedded Rat Skeletal muscle; Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15 min; Antibody incubation with LRP2 Polyclonal Antibody, Unconjugated (AP94003) at 1:200 overnight at 4°C, followed by conjugation to the SP Kit (Rabbit, SP-0023) and DAB (C-0010) staining.



Paraformaldehyde-fixed, paraffin embedded Human Kidney; Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15 min; Antibody incubation with LRP2 Polyclonal Antibody, Unconjugated (AP94003) at 1:200 overnight at 4°C, followed by conjugation to the SP Kit (Rabbit, SP-0023) and DAB (C-0010) staining.



Paraformaldehyde-fixed, paraffin embedded Human Placenta; Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15 min; Antibody incubation with LRP2 Polyclonal Antibody, Unconjugated (AP94003) at 1:200 overnight at 4°C, followed by conjugation to the SP Kit (Rabbit, SP-0023) and DAB (C-0010) staining.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.