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FGGY Rabbit pAb

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Catalog # AP54596

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q96C11
Reactivity	Rat, Mouse
Predicted	Dog, Human, Rabbit, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	59993
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human FGGY
Epitope Specificity	151-250/551
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
DISEASE	Defects in FGGY are associated with sporadic amyotrophic lateral sclerosis (ALS) [MIM:105400]. Amyotrophic lateral sclerosis is a neurodegenerative disorder affecting upper and lower motor neurons and resulting in fatal paralysis. Sensory abnormalities are absent. Death usually occurs within 2 to 5 years. The etiology of amyotrophic lateral sclerosis is likely to be multifactorial, involving both genetic and environmental factors.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	FGGY is a 551 amino acid member of the FGGY kinase family that exists as four isoforms which are produced by alternative splicing events. Expressed in lung, kidney, small intestine, liver and fetal brain, FGGY is encoded by a gene that maps to chromosome 1 and, when mutated, is associated with sporadic amyotrophic lateral sclerosis (ALS). ALS is a neurodegenerative disorder that affects motor neurons and results in fatal paralysis, usually within 2 to 5 years after initial diagnosis. Chromosome 1, on which the gene encoding FGGY is located, is the largest human chromosome, spanning about 260 million base pairs and making up 8% of the human genome. There are about 3,000 genes on chromosome 1, many of which are associated with genetic diseases, including Hutchinson-Gilford progeria, familial adenomatous polyposis, Stickler syndrome, Gaucher disease and Usher syndrome.

Additional Information

Gene ID	55277
Other Names	FGGY carbohydrate kinase domain-containing protein, D-ribulokinase FGGY, 2.7.1.47, FGGY {ECO:0000303|PubMed:27909055}

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	FGGY {ECO:0000303|PubMed:27909055}
Function	Catalyzes ATP-dependent phosphorylation of D-ribulose at C-5 to form D-ribulose 5-phosphate. Postulated to function in a metabolite repair mechanism by preventing toxic accumulation of free D-ribulose formed by non-specific phosphatase activities. Alternatively, may play a role in regulating D-ribulose 5-phosphate recycling in the pentose phosphate pathway. Can phosphorylate ribitol with low efficiency.
Tissue Location	Expressed in kidney, lung and small intestine and to a lower extent in liver and detected in cerebrospinal fluid (at protein level).

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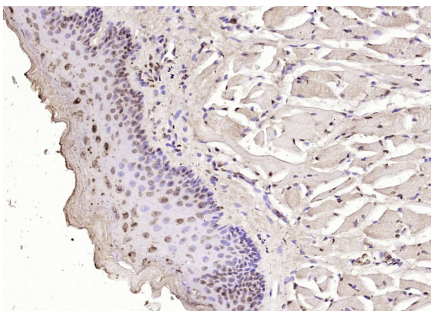
Images



Paraformaldehyde-fixed, paraffin embedded (rat tongue); 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; Antibody incubation with (FGGY) Polyclonal Antibody, Unconjugated (AP54596) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.