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Rabbit Anti-WFS1 antibody

SL11272R

Product Name:	WFS1
Chinese Name:	Wolfram综合征蛋白1抗体
Alias:	DFNA14; DFNA38; DFNA6; DIDMOAD; WFRS; WFS; Wolfram syndrome 1 (wolframin); Wolfram syndrome; WOLFRAMIN; WFS1_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Cow,Horse,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	97kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human WFS1:791-890/890
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	Wolfram syndrome protein (WFS1) is an 890 amino acid protein that contains a cytoplasmic N-terminal domain, followed by nine-transmembrane domains and a luminal C-terminal domain. WFS1 is predominantly localized to the endoplasmic reticulum (ER) (1) and its expression is induced in response to ER stress, partially through transcriptional activation (2,3). Research studies have shown that mutations in the WFS1 gene lead to Wolfram syndrome, an autosomal recessive neurodegenerative

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	disorder defined by young-onset, non-immune, insulin-dependent diabetes mellitus and progressive optic atrophy (4). **Function:** WFS1 is a novel component of Wolfram syndrome, a rare form of juvenile diabetes. WFS1 plays an important role in maintaining homeostasis of the endoplasmic reticulum (ER) in the pancreas. It is normally up-regulated during insulin secretion, whereas inactivation of the protein can cause ER stress. Chronic ER stress is a major involvement in Wolfram syndrome. **Subcellular Location:** Endoplasmic reticulum; endoplasmic reticulum membrane; multipass membrane protein **Tissue Specificity:** Highly expressed in heart followed by brain, placenta, lung and pancreas. Weakly expressed in liver, kidney and skeletal muscle. Also expressed in islet and beta-cell insulinoma cell line. **DISEASE:** Defects in WFS1 are the cause of Wolfram syndrome type 1 (WFS1) [MIM:222300]. A rare autosomal recessive disorder characterized by juvenile diabetes mellitus, diabetes insipidus, optic atrophy, deafness and various neurological symptoms. Defects in WFS1 are the cause of deafness autosomal dominant type 6 (DFNA6) [MIM:600965]; also called non-syndromic sensorineural deafness autosomal dominant type 14 (DFNA14) or non-syndromic sensorineural deafness autosomal dominant type 38 (DFNA38). DFNA6 is a form of 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. DFNA6 is a low-frequency hearing loss in which frequencies of 2000 Hz and below are predominantly affected. Many patients have tinnitus, but there are otherwise no associated features such as vertigo. Because high-frequency hearing is generally preserved, patients retain excellent understanding of speech, although presbycusis or noise exposure may cause high-frequency loss later in life. DFNA6 worsens over time without progressing to profound deafness. Defects in WFS1 are the cause of Wolfram-like syndrome autosomal dominant (WFSL) [MIM:614296]. A disease characterized by the clinical triad of congenital progressive hearing impairment, diabetes mellitus, and optic atrophy. The hearing impairment, which is usually diagnosed in the first decade of life, is relatively constant and alters mainly low- and middle-frequency ranges. **SWISS:** O76024 **Gene ID:** 7466

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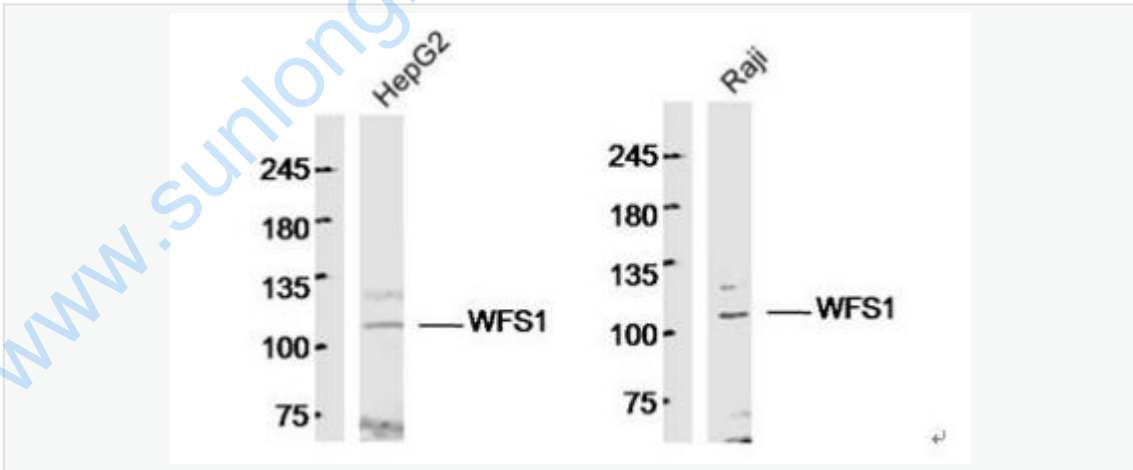
	Database links: Entrez Gene: 7466Human Entrez Gene: 22393Mouse Entrez Gene: 83725Rat Omim: 606201Human SwissProt: O76024Human SwissProt: P56695Mouse Unigene: 518602Human Unigene: 20916Mouse Unigene: 229139Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	 Sample: HepG2 Cell（Human）Lysate at 40 ug Raji Cell（Human）Lysate at 40 ug Primary: Anti-WFS1 (SL11272R) at 1/300 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

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	Predicted band size: 97 kD Observed band size: 105 kD

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