10320 Camino Santa Fe, Suite G
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WFS1 Polyclonal Antibody
Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP54440

Specification

WFS1 Polyclonal Antibody - Product Information

Application	**WB, IHC-P, IHC-F, IF, ICC, E**
Primary Accession	O76024
Reactivity	**Rat, Dog, Bovine**
Host	**Rabbit**
Clonality	**Polyclonal**
Calculated MW	**97 KDa**
Physical State	**Liquid**
Immunogen	**KLH conjugated synthetic peptide derived from human WFS1**
Epitope Specificity	**791-890/890**
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	**Endoplasmic reticulum; endoplasmic reticulum membrane; multipass membrane protein**
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.
Important Note	**This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.**

Background Descriptions
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 disorder defined by young-onset, non-immune, insulin-dependent diabetes mellitus and progressive optic atrophy (4).

WFS1 Polyclonal Antibody - Additional Information

Gene ID 7466

Other Names
Wolframin, WFS1

Target/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.

Dilution
WB~~1:1000<br \>IHC-P~~N/A<br \>IHC-F~~N/A<br \>IF~~1:50~200<br \>ICC~~N/A<br \>E~~N/A

Format
0.01M TBS(pH7.4), 0.09% (W/V) sodium azide and 50% Glyce

Storage
Store at -20 ℃ 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 ℃.

WFS1 Polyclonal Antibody - Protein Information

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Name WFS1

Function
Participates in the regulation of cellular Ca(2+) homeostasis, at least partly, by modulating the filling state of the endoplasmic reticulum Ca(2+) store (PubMed:16989814). Negatively regulates the ER stress response and positively regulates the stability of V-ATPase subunits ATP6V1A and ATP1B1 by preventing their degradation through an unknown proteasome-independent mechanism (PubMed:23035048).

Cellular Location
Endoplasmic reticulum membrane; Multi-pass membrane protein. Cytoplasmic vesicle, secretory vesicle. Note=Co-localizes with ATP6V1A in the secretory granules in neuroblastoma cell lines

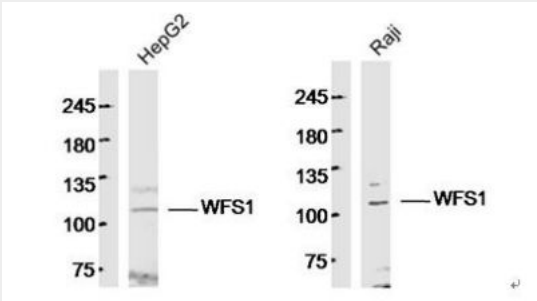
Tissue Location
Highly expressed in heart followed by brain, placenta, lung and pancreas. Weakly expressed in liver, kidney and skeletal muscle. Also expressed in islet and beta-cell insulinoma cell line

WFS1 Polyclonal 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

WFS1 Polyclonal Antibody - Images



Sample:
HepG2 Cell Human Lysate at 40 ug
Raji Cell Human Lysate at 40 ug
Primary: Anti-WFS1 (bs-11272R) at 1/300 dilution
Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution
Predicted band size: 97 kD
Observed band size: 105 kD