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Rabbit Anti-PQBP1 antibody

SL11751R

Product Name:	PQBP1
Chinese Name:	多谷氨酰胺Binding protein1抗体
Alias:	38 kDa nuclear protein containing a WW domain; Mental retardation, X linked 55; MRX55; MRXS3; MRXS8; Npw38; Nuclear protein containing WW domain 38 kD; Polyglutamine binding protein 1; Polyglutamine tract binding protein 1; Polyglutamine tract-binding protein 1; Polyglutamine-binding protein 1; PQBP 1; PQBP-1; PQBP1; PQBP1_HUMAN; RENS1; SHS; Sutherland Haan X linked mental retardation syndrome.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,Rabbit,Sheep,
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:	30kDa
Cellular localization:	The nucleus
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human PQBP1:185-265/265
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:	Polyglutamine(Q) tract binding protein-1 (PQBP-1) is a transcription repressor that associates with polyglutamine tract-containing transcription regulators and causative

genes for neurodegenerative disorders. Hepta- and di-amino acid repeat sequences rich in polar residues are essential for PQBP-1 to interact with polyglutamine tract-containing proteins (i.e. huntingtin, androgen receptor and Brain-2). PQBP-1 contains a WWP/WW domain that binds proline-rich motifs and a C2 domain that can influence Ca2+-dependent phospholipid signaling. PQBP-1 localizes to the nucleus and is present in neurons throughout the brain, with abundant levels in hippocampus, cerebellar cortex and olfactory bulb. The human PQBP-1 gene maps to chromosome Xp11.23.

Function:
May suppress the ability of POU3F2 to transactivate the DRD1 gene in a POU3F2 dependent manner. Can activate transcription directly or via association with the transcription machinery. May be involved in ATXN1 mutant-induced cell death. The interaction with ATXN1 mutant reduces levels of phosphorylated RNA polymerase II large subunit.

Subunit:
nteracts with POU3F2/Brn-2, ATXN1, TXNL4A, HTT and AR.Interaction with ATXN1 correlates positively with the length of thepolyglutamine tract. Interacts with RNA polymerase II large subunitin a phosphorylation-dependent manner. Forms a ternary complex withATXN1 mutant and phosphorylated RNA polymerase II.

Subcellular Location:
Nucleus. Co-localized with POU3F2. Co-localized with ATXN1 in nuclear inclusion bodies.

Tissue Specificity:
Widely expressed with high level in heart, skeletal muscle, pancreas, spleen, thymus, prostate, ovary, small intestine and peripheral blood leukocytes.

DISEASE:
Defects in PQBP1 are the cause of Renpenning syndrome 1 (RENS1) [MIM:309500]; also known as Sutherland-Haan X-linked mental retardation syndrome (SHS) or X-linked mental retardation syndromes MRXS3/MRXS8/MRX55. The clinical features are mental retardation, microcephaly, short stature, and small testes. The craniofacies tends to be narrow and tall with upslanting palpebral fissures, abnormal nasal configuration, cupped ears, and short philtrum. The nose may appear long or bulbous, with overhanging columella. Less consistent manifestations include ocular colobomas, cardiac malformations, cleft palate, and anal anomalies. RENS1 is more frequently in males than in females where little or no expression is found.

Similarity:
Contains 1 WW domain.

SWISS:
O60828

	Gene ID: 10084 **Database links:** Entrez Gene: 10084Human Entrez Gene: 54633Mouse Omim: 300463Human SwissProt: O60828Human SwissProt: Q91VJ5Mouse Unigene: 534384Human Unigene: 14616Mouse **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.