Product name:	Ubr1 Rabbit Polyclonal Antibody
Cat number:	ABN19582
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human UBR1. AA range:821-870
Reactivity:	Human,Mouse
Applications:	WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:50-1:200,ELISA 1:10000-1:20000
Molecular Weight:	200kDa
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

Background:

The N-end rule pathway is one proteolytic pathway of the ubiquitin system. The recognition component of this pathway, encoded by this gene, binds to a destabilizing N-terminal residue of a substrate protein and participates in the formation of a substrate-linked multiubiquitin chain. This leads to the eventual degradation of the substrate protein. The protein described in this record has a RING-type zinc finger and a UBR-type zinc finger. Mutations in this gene have been associated with Johanson-Blizzard syndrome. [provided by RefSeq, Jul 2008],developmental stage:Expressed in fetal pancreas.,disease:Defects in UBR1 are a cause of Johanson-Blizzard syndrome (JBS) [MIM:243800]. This disorder includes congenital exocrine pancreatic insufficiency, multiple malformations such as nasal wing aplasia, and frequent mental retardation. Pancreas of individuals with JBS do not express UBR1 and show intrauterine-onset destructive pancreatitis.,domain:The RING-H2 zinc finger is an atypical RING finger with a His ligand in place of the fourth Cys of the classical motif.,function:E3 ubiquitin-protein ligase which is a component of the N-end rule pathway. Recognizes and binds to proteins bearing specific N-terminal residues that are destabilizing according to the N-end rule, leading to their ubiquitination and subsequent degradation. May be involved in pancreatic homeostasis.,pathway:Protein modification; protein ubiquitination.,PTM:Phosphorylated upon DNA damage, probably by ATM or ATR.,similarity:Belongs to the UBR1 family.,similarity:Contains 1 RING-type zinc finger.,similarity:Contains 1 UBR-type zinc finger.,subunit:Interacts with RECQL4.,tissue specificity:Broadly expressed, with highest levels in skeletal muscle, kidney and pancreas. Present in acinar cells of the pancreas (at protein level).,