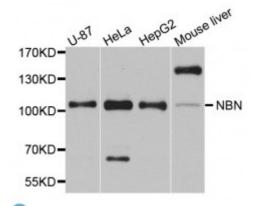


Anti-NBN Antibody



Description

Mutations in this gene are associated with Nijmegen breakage syndrome, an autosomal recessive chromosomal instability syndrome characterized by microcephaly, growth retardation, immunodeficiency, and cancer predisposition. The encoded protein is a member of the MRE11/RAD50 double-strand break repair complex which consists of 5 proteins. This gene product is thought to be involved in DNA double-strand break repair and DNA damage-induced checkpoint activation.

Model STJ114898

Host Rabbit

Reactivity Human, Mouse

Applications WB

Immunogen Recombinant fusion protein containing a sequence corresponding to amino

acids 435-754 of human NBN (NP_002476.2).

Gene ID 4683

Gene Symbol NBN

Dilution range WB 1:500 - 1:1000

Tissue Specificity Ubiquitous, Expressed at high levels in testis

Purification Affinity purification

Note For Research Use Only (RUO).

Protein Name Nibrin Cell cycle regulatory protein p95 Nijmegen breakage syndrome protein

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Molecular Weight 84.959 kDa

Clonality Polyclonal

Conjugation Unconjugated

Isotype IgG

Formulation PBS with 0.02% sodium azide, 50% glycerol, pH7.3.

Storage Instruction Store at -20C. Avoid freeze / thaw cycles.

Database Links HGNC:7652OMIM:114480Reactome:R-HSA-2559586

Alternative Names Nibrin Cell cycle regulatory protein p95 Nijmegen breakage syndrome protein

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Function Component of the MRE11-RAD50-NBN (MRN complex) which plays a

critical role in the cellular response to DNA damage and the maintenance of chromosome integrity, The complex is involved in double-strand break (DSB) repair, DNA recombination, maintenance of telomere integrity, cell cycle checkpoint control and meiosis, The complex possesses single-strand endonuclease activity and double-strand-specific 3'-5' exonuclease activity, which are provided by MRE11, RAD50 may be required to bind DNA ends and hold them in close proximity, NBN modulate the DNA damage signal sensing by recruiting PI3/PI4-kinase family members ATM, ATR, and probably DNA-PKcs to the DNA damage sites and activating their functions, It can also recruit MRE11 and RAD50 to the proximity of DSBs by an interaction with the histone H2AX, NBN also functions in telomere length maintenance by generating the 3' overhang which serves as a primer for telomerase dependent telomere elongation, NBN is a major player in the control of intra-S-phase checkpoint and there is some evidence that NBN is involved in G1 and G2 checkpoints, The roles of NBS1/MRN encompass DNA damage sensor, signal transducer, and effector, which enable cells to maintain DNA integrity and genomic stability, Forms a complex with RBBP8

Cellular Localization Nucleus

Post-translational Phosphorylated by ATM in response of ionizing radiation, and such

phosphorylation is responsible intra-S phase checkpoint control and telomere

to link DNA double-strand break sensing to resection, Enhances AKT1 phosphorylation possibly by association with the mTORC2 complex,

maintenance,

Modifications