## bs-9331R

## [ Primary Antibody ]

# NHLRC1 Rabbit pAb

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DATASHEET -

Host: Rabbit Isotype: IgG

Clonality: Polyclonal

GeneID: 378884 SWISS: Q3SYB1

Target: NHLRC1

Immunogen: KLH conjugated synthetic peptide derived from human NHLRC1:

221-320/395.

**Purification:** affinity purified by Protein A

Concentration: 1mg/ml

Storage: 0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50%

Glycerol.

Shipped at 4°C. Store at -20°C for one year. Avoid repeated

freeze/thaw cycles.

Background: Progressive myoclonic epilepsy type 2 (EPM2), also called Lafora disease, is an autosomal recessive disease characterized by grand mal seizures and/or myoclonus at about 15 years of age. Rapid and severe mental deterioration follows, often with psychotic features. Survival is less than 10 years after onset. Starch-like, endoplasmic reticulum-associated polyglucosans, called Lafora bodies, can be observed in brain, muscle, liver and heart. One cause of Lafora disease is due to mutations in NHLRC1, the gene encoding Malin. Forty-nine different mutations in NHLRC1 have been shown to cause EPM2. Malin, also called NHL repeat-containing protein 1, is a single subunit E3 ubiquitin ligase, containing 6 NHL repeats and 1 RING-type zinc finger. Malin's RING domain is responsible for its ability to mediate ubiquitination. Malin interacts with and polyubiquitinates Laforin, a protein also implicated in EPM2. Malin localizes to the endoplasmic reticulum and, to a lesser extent, in the nucleus. Malin is expressed in brain, cerebellum, spinal cord, medulla, heart, liver, skeletal muscle and pancreas.

Applications: WB (1:500-2000)

Reactivity: Rat (predicted: Human,

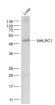
Mouse, Rabbit)

Predicted 42 kDa

MW.:

Subcellular Cytoplasm , Nucleus

### VALIDATION IMAGES -



Sample: Liver(Rat) Lysate at 40 ug Primary: Anti-NHLRC1 (bs-9331R) at 1/1000 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 42 kD Observed band size: 52 kD