bs-6723R

[Primary Antibody]

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PNPLA6/NTE Rabbit pAb

- DATASHEET -

Host: Rabbit Isotype: IgG

Clonality: Polyclonal

GeneID: 10908 **SWISS:** Q8IY17

Target: PNPLA6/NTE

Immunogen: KLH conjugated synthetic peptide derived from human PNPLA6:

74-170/1366.

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: Phospholipase B that deacylates intracellular phosphatidylcholine

(PtdCho), generating glycerophosphocholine (GroPtdCho). This deacylation occurs at both sn-2 and sn-1 positions of PtdCho. Its specific chemical modification by certain organophosphorus (OP)

compounds leads to distal axonopathy.

Tissue specificity; Expressed in brain, placenta, kidney, neuron and

skeletal muscle.

Involvement in diseaseDefects in PNPLA6 are the cause of spastic paraplegia autosomal recessive type 39 (SPG39); also known as NTE-related motor neuron disorder (NTEMND). Spastic paraplegia is a neurodegenerative disorder characterized by a slow, gradual, progressive weakness and spasticity of the lower limbs. Rate of progression and the severity of symptoms are quite variable. Initial symptoms may include difficulty with balance, weakness and stiffness in the legs, muscle spasms, and dragging the toes when walking. In some forms of the disorder, bladder symptoms (such as incontinence) may appear, or the weakness and stiffness may spread to other parts of the body. SPG39 is associated with a motor axonopathy affecting upper and lower limbs and resulting in progressive wasting of distal upper and lower extremity muscles.

Applications: IHC-P (1:100-500)

IHC-F (1:100-500) **IF** (1:100-500)

Reactivity: Rat (predicted: Human,

Mouse, Pig, Cow, Chicken,

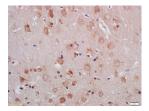
Dog, Horse)

Predicted MW.: 150 kDa

14144..

Subcellular Location: Cell membrane ,Cytoplasm

VALIDATION IMAGES



Tissue/cell: rat brain tissue; 4%
Paraformaldehyde-fixed and paraffinembedded; Antigen retrieval: citrate buffer (0.01M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37°C for 20 min; Incubation: Anti-PNPLA6 Polyclonal Antibody, Unconjugated(bs-6723R) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining