bs-6311R **[Primary Antibody]**

BSCL2 Rabbit pAb

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DATASHEET

Host: Rabbit **Isotype:** IgG

Clonality: Polyclonal

GeneID: 26580

Target: BSCL2

Immunogen: KLH conjugated synthetic peptide derived from human BSCL2/SPG17: 151-250/398.

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: Defects in BSCL2 are the cause of congenital generalized lipodystrophy type 2 (CGL2) . Congenital generalized lipodystrophy is an autosomal recessive disorder characterized by a near absence of adipose tissue, extreme insulin resistance, hypertriglyceridemia, hepatic steatosis and early onset of diabetes.
Defects in BSCL2 are the cause of spastic paraplegia type 17 (SPG17) ; also known as Silver spastic paraplegia syndrome. Spastic paraplegia is a neurodegenerative disorder characterized by a slow, gradual, progressive weakness and spasticity of the lower limbs. SPG17 is characterized by prominent amyotrophy of the hand muscles, the presence of mild to severe pyramidal tract signs, and spastic paraplegia.
SPG17 is a motor neuron disease overlapping with distal spinal muscular atrophy type 5. Defects in BSCL2 are a cause of distal hereditary motor neuropathy type 5 (HMN5); also known aS distal hereditary motor neuropathy type V (DSMAV). HMN5 is an autosomal dominant disorder characterized by degeneration of motor nerve fibers, predominantly in limb distal regions.

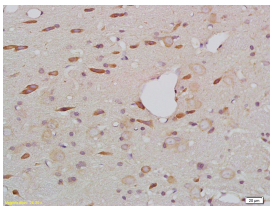
Applications: **IHC-P** (1:100-500)
IHC-F (1:100-500)
IF (1:100-500)

Reactivity: Rat (predicted: Human, Mouse, Rabbit, Cow, Dog, Horse)

Predicted MW.: 44 kDa

Subcellular Location: Cytoplasm

VALIDATION IMAGES



Tissue/cell: rat brain tissue; 4% Paraformaldehyde-fixed and paraffin-embedded; 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-BSCL2 Polyclonal Antibody, Unconjugated(bs-6311R) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining