SunLong Biotech Co.,LTD
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[Primary Antibody]

Rabbit Anti-BSCL2 antibody

Catalog Number: SL6311R

FOR RESEARCH USE ONLY

[DATASHEET]

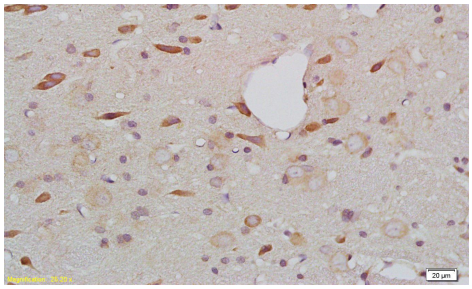
Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Target:	BSCL2
Immunogen:	KLH conjugated synthetic peptide derived from human BSCL2/SPG17: 151-250/398
Purification:	affinity purified by Protein A
Concentration:	1mg/ml
Storage:	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Shipped at 4℃. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.
Applications:	IHC-P=1:100-500 IHC-F=1:100-500 IF=1:100-500 （Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Reactivity:	Rat(predicted:Human,Mouse,Dog,Cow,Horse,Rabbit)
Predicted MW.:	44kDa
Subcellular Location:	cytoplasmic
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.

Important Note: This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

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	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. **Function:** Endoplasmic reticulum membrane; Multi-pass membrane protein. **Tissue Specificity:** Highest expression in brain and testis. DISEASE: Defects in BSCL2 are the cause of congenital generalized lipodystrophy type 2 (CGL2) [MIM:269700]. 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) [MIM:270685]; 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) [MIM:600794]; 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. **Similarity:** Belongs to the seipin family. **Database links:** UniProtKB/Swiss-Prot: Q96G97.3

[VALIDATION IMAGES]



Tissue/cell: rat brain tissue; 4% Paraformaldehyde-fixed and paraffin-embedded;
Antigen retrieval: citrate buffer (1M, 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℃ for 20 min;
Incubation: Anti-BSCL2 Polyclonal Antibody, Unconjugated(SL6311R) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining

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