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[Marker Primary Antibody]

Rabbit Anti-BSCL2/HRP Conjugated antibody

Catalog Number: SL6311R-HRP

FOR RESEARCH USE ONLY

[DATASHEET]

Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Target:	Anti-BSCL2/HRP
Immunogen:	KLH conjugated synthetic peptide derived from human BSCL2/SPG17
Purification:	affinity purified by Protein A
Concentration:	1mg/ml
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 1M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
Applications:	IHC-P=1:100-500 IHC-F=1:100-500 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)
Background:	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

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