

BSCL2 Rabbit pAb

Catalog # AP98582

Product Information

Application	IHC-P, IHC-F, IF
Reactivity	Rat
Predicted	Human, Mouse, Rabbit, Dog, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	44 KDa
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human BSCL2/SPG17
Epitope Specificity	151-250/398
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the seipin family.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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.

Additional Information

Gene ID	26580
Other Names	BSCL2_HUMAN; BSCL2; Bernardinelli-Seip congenital lipodystrophy type 2 protein; BSCL2_MOUSE; Bernardinelli-Seip congenital lipodystrophy type 2 protein homolog; Gng3lg; BSCL2_RAT; HMN5; HMN5C; HMND13; PELD; SPG17; 2900097C17Rik; BSCL2 lipid droplet biogenesis associated, seipin; spastic paraplegia 17 (Silver syndrome); Berardinelli-Seip congenital lipodystrophy 2 (seipin); BSCL2, seipin lipid droplet biogenesis associated
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
Name	BSCL2 Rabbit pAb
Function	Endoplasmic reticulum membrane; Multi-pass membrane protein.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.