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# Rabbit Anti-BSCL2 antibody

SL6311R

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| **Product Name:** | BSCL2 |
| **Chinese Name:** | 先天性脂肪代谢障碍蛋白2抗体(常染色体显性遗传痉挛性截瘫17) |
| **Alias:** | Bernardinelli Seip congenital lipodystrophy 2; Bernardinelli Seip congenital lipodystrophy type 2 protein; Bernardinelli-Seip congenital lipodystrophy type 2 protein; BSCL 2; BSCL2; BSCL2_HUMAN; GNG3LG; HMN 5; HMN5; MGC4694; Seipin; Spastic paraplegia 17 (autosomal dominant); Spastic paraplegia 17 (Silver syndrome); Spastic paraplegia 17; Spastic paraplegia with amyotrophy of hands and feet (Silver syndrome); Spastic paraplegia with amyotrophy of hands and feet; SPG 17; SPG17. |
| **Organism Species:** | Rabbit |
| **Clonality:** | Polyclonal |
| **React Species:** | Human,Mouse,Rat,Dog,Cow,Horse,Rabbit, |
| **Applications:** | WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=1:100-500(Paraffin sections need antigen repair)<br>not yet tested in other applications.<br>optimal dilutions/concentrations should be determined by the end user. |
| **Molecular weight:** | 44kDa |
| **Cellular localization:** | cytoplasmic |
| **Form:** | Lyophilized or Liquid |
| **Concentration:** | 1mg/ml |
| **immunogen:** | KLH conjugated synthetic peptide derived from human BSCL2/SPG17:151-250/398 |
| **Lsotype:** | IgG |
| **Purification:** | affinity purified by Protein A |
| **Storage Buffer:** | 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol. |
| **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 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C. |
| **PubMed:** | PubMed |
| **Product Detail:** | 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.

**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.

**SWISS:**
Q96G97

**Gene ID:**
26580

**Database links:**

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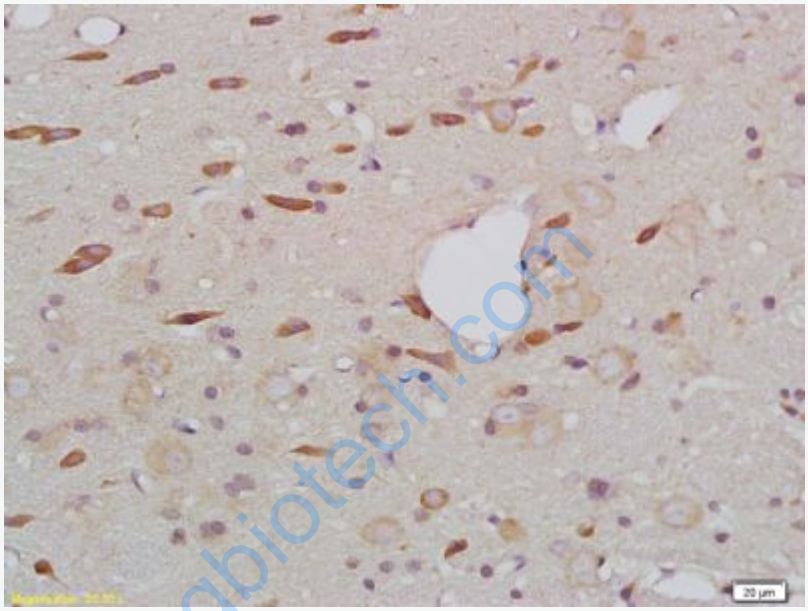
| | UniProtKB/Swiss-Prot: Q96G97.3<br><br>**Important Note:**<br>This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications. |
| **Picture:** |  <br>Tissue/cell: rat brain tissue; 4% Paraformaldehyde-fixed and paraffin-embedded; Antigen retrieval: 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℃ for 20 min;<br>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 |