Data Sheet (Cat.No.TMAB-03234)

Anti-BSCL2 Polyclonal Antibody

Product Details

Ig Type:	IgG
Reactivity:	Rat (predicted:Human,Mouse,Dog,Cow,Horse,Rabbit)
Molecular Weight:	Theoretical: 44 kDa.
Purification:	Protein A purified

Applications

Verified Activity:	Tissue/cell: rat brain tissue; 4% Paraformaldehyde-fixed and paraffin-embedded; Antigen retrieval: citrate buffer (0.01 M, pH 6.0), Boiling bathing for 15 min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30 min; Blocking buffer (normal goat serum) at 37℃ for 20 min; Incubation: Anti-BSCL2 Polyclonal Antibody, Unconjugated (TMAB-03234) 1: 200, overnight at 4℃, followed by conjugation to the secondary antibody and DAB staining
Application:	IHC-P,IHC-Fr,IF
Recommended	IHC-P: 1:100-500; IHC-Fr: 1:100-500; IF: 1:100-500

Properties

Stability & Storage:	Store at 2℃-8℃ for 1 month. Store at -20℃ or -80℃ for 12 months. Avoid repeated freeze-thaw cycles.
Shipping:	Shipping with blue ice.

Antigen Details

Immunogen:	KLH conjugated synthetic peptide: human BSCL2/SPG17
Antigen Species:	Human
Gene ID:	26580

Research 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.

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