

## **Product Name: SACS Rabbit Polyclonal Antibody**

Catalog #: APRab17571

For research use only.

#### **Summary**

**Description** Rabbit polyclonal Antibody

**Host** Rabbit

Application IHC,ICC/IF

Reactivity Human, Mouse
Conjugation Unconjugated
Modification Unmodified

**Isotype** IgG

ClonalityPolyclonalFormLiquidConcentration1mg/ml

**Storage** Aliquot and store at -20°C (valid for 12 months). Avoid freeze/thaw cycles.

**Shipping** Ice bags

Buffer Liquid in PBS containing 50% glycerol, and 0.02% New type preservative N.

**Purification** Affinity purification

#### **Application**

**Dilution Ratio** IHC 1:50-1:300,ICC/IF 1:50-1:200

Molecular Weight 503kDa

# **Antigen Information**

Gene Name SACS

Alternative Names KIAA0730
Gene ID 26278.0
SwissProt ID 09NZJ4

**Immunogen** Synthesized peptide derived from human protein . at AA range: 4291-4340

## **Background**

This gene encodes the sacsin protein, which includes a UbL domain at the N-terminus, a DnaJ domain, and a HEPN domain at the C-terminus. The gene is highly expressed in the central nervous system, also found in skin, skeletal muscles and at low levels in the pancreas. This gene includes a very large exon spanning more than 12.8 kb. Mutations in this gene result in autosomal

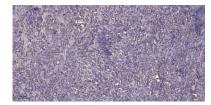


recessive spastic ataxia of Charlevoix-Saguenay (ARSACS), a neurodegenerative disorder characterized by early-onset cerebellar ataxia with spasticity and peripheral neuropathy. The authors of a publication on the effects of siRNA-mediated sacsin knockdown concluded that sacsin protects against mutant ataxin-1 and suggest that "the large multi-domain sacsin protein is able to recruit Hsp70 chaperone action and has the potential to regulate the effects of other ataxia proteins" (Parfitt et al., PubMed: 19208651).disease:Defects in SACS are the cause of autosomal recessive spastic ataxia of Charlevoix-Saguenay (ARSACS) [MIM:270550]. ARSACS is an early onset neurodegenerative disease with high prevalence in the Charlevoix-Saguenay-Lac-Saint-Jean region of Quebec. It is characterized by absent sensory-nerve conduction, reduced motor-nerve velocity and hypermyelination of retinal-nerve fibers.,function:May function in chaperone-mediated protein folding.,similarity:Contains 1 HEPN domain.,similarity:Contains 1 J domain.,tissue specificity:Highly expressed in the central nervous system. Also found in skeletal muscle and at low levels in pancreas.,

#### **Research Area**

Epigenetics and Nuclear Signaling; Transcription; Domain Families; HLH / Leucine Zipper; Leucine Zipper; Signal Transduction; Protein Trafficking; Chaperones; Other Chaperones; Metabolism; Pathways and Processes; Mitochondrial Metabolism; Mitochondrial markers

### **Image Data**



Immunohistochemical analysis of paraffin-embedded human Colon cancer. 1, Antibody was diluted at 1:200 (4° overnight) . 2, Tris-EDTA,pH9.0 was used for antigen retrieval. 3,Secondary antibody was diluted at 1:200 (room temperature, 45min) .

Web: https://www.enkilife.com E-mail: order@enkilife.com techsupport@enkilife.com Tel: 0086-27-87002838