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|--------------------------|----------------------------------------------------------------------------------------------|
| <b>Product name:</b>     | SACS Rabbit Polyclonal Antibody                                                              |
| <b>Cat number:</b>       | ABN17571                                                                                     |
| <b>Conjugate:</b>        | Unconjugated                                                                                 |
| <b>Size:</b>             | 100µL                                                                                        |
| <b>Clone:</b>            | Polyclonal                                                                                   |
| <b>Concentration:</b>    | 1mg/ml                                                                                       |
| <b>Host:</b>             | Rabbit                                                                                       |
| <b>Isotype:</b>          | IgG                                                                                          |
| <b>Immunogen:</b>        | Synthesized peptide derived from human protein . at AA range: 4291-4340                      |
| <b>Reactivity:</b>       | Human, Mouse                                                                                 |
| <b>Applications:</b>     | IHC 1:50-1:300, ICC/IF 1:50-1:200                                                            |
| <b>Molecular Weight:</b> | 503kDa                                                                                       |
| <b>Purification:</b>     | Affinity purification                                                                        |
| <b>Form:</b>             | Liquid                                                                                       |
| <b>Buffer:</b>           | Liquid in PBS containing 50% glycerol, and 0.02% New type preservative N.                    |
| <b>Storage:</b>          | Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles. |

**Background:**

This gene encodes the saccin 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 saccin knockdown concluded that saccin protects against mutant ataxin-1 and suggest that "the large multi-domain saccin 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.