

## DESCRIPTION

|                                                                                                                                                                                                         |                                                                                         |
|---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-----------------------------------------------------------------------------------------|
| <b>Species Reactivity</b>                                                                                                                                                                               | Human                                                                                   |
| <b>Specificity</b>                                                                                                                                                                                      | Detects human SACS in direct ELISAs.                                                    |
| <b>Source</b>                                                                                                                                                                                           | Polyclonal Sheep IgG                                                                    |
| <b>Purification</b>                                                                                                                                                                                     | Antigen Affinity-purified                                                               |
| <b>Immunogen</b>                                                                                                                                                                                        | <i>E. coli</i> -derived recombinant human SACS<br>Asn4402-Val4579<br>Accession # Q9NZJ4 |
| <b>Conjugate</b>                                                                                                                                                                                        | Alexa Fluor 594<br>Excitation Wavelength: 590 nm<br>Emission Wavelength: 617 nm         |
| <b>Formulation</b>                                                                                                                                                                                      | Supplied 0.2mg/ml in 1X PBS with RDF1 and 0.09% Sodium Azide                            |
| *Contains <0.1% Sodium Azide, which is not hazardous at this concentration according to GHS classifications. Refer to the Safety Data Sheet (SDS) for additional information and handling instructions. |                                                                                         |

## APPLICATIONS

**Please Note:** Optimal dilutions should be determined by each laboratory for each application. [General Protocols](#) are available in the Technical Information section on our website.

**Immunohistochemistry** Optimal dilution of this antibody should be experimentally determined.

## PREPARATION AND STORAGE

|                                |                                                                                                                   |
|--------------------------------|-------------------------------------------------------------------------------------------------------------------|
| <b>Shipping</b>                | The product is shipped with polar packs. Upon receipt, store it immediately at the temperature recommended below. |
| <b>Stability &amp; Storage</b> | Protect from light. Do not freeze. 12 months from date of receipt, 2 to 8 °C as supplied                          |

## BACKGROUND

SACS (Spastic Ataxia of Charlevoix-Saguenay; also DNAJC29 and Sacsin) is a 520-540 kDa, novel cytoplasmic chaperone involved neuronal homeostasis. It is expressed in select cell types, including fibroblasts, skeletal muscle cells, cerebellar granule and Purkinje cells, and multiple CNS neuronal phenotypes. SACS is known to play a key role in protein folding, assisting in the adoption of a stable conformation, possibly through the use of ATP hydrolysis. It appears to collaborate with chaperone Hsp70 family proteins, and provide protection against polypeptides containing polyGlu repeats such as ataxin-1. These repeats tend to form annular structures, causing aggregates involving themselves and accompanying molecular partners. Human SACS is 4579 amino acids (aa) in length. It contains a N-terminal ubiquitin-like domain that associates with proteasomes (aa 9-84), three consecutive SRRs/Sacsin Supradomain Regions (aa 90-2900) that possess ATPase activity, a J-domain that interacts with other chaperones (aa 4306-4393), and a HEPN domain (aa 4451-4567) that binds GTP when SACS is dimerized. SACS contains three utilized Ser phosphorylation sites plus one acetylation site at Lys943. There is one alternate start site at Met751, and a 21 aa peptide that can substitute for either aa 730-750, or aa 812-832. Over aa 4402-4579, human SACS shares 99% aa sequence identity with mouse SACS.

## PRODUCT SPECIFIC NOTICES

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