

DESCRIPTION

Species Reactivity	Human
Specificity	Detects human SACS in direct ELISAs.
Source	Polyclonal Sheep IgG
Purification	Antigen Affinity-purified
Immunogen	<i>E. coli</i> -derived recombinant human SACS Asn4402-Val4579 Accession # Q9NZJ4
Conjugate	Alexa Fluor 488 Excitation Wavelength: 488 nm Emission Wavelength: 515-545 nm
Formulation	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

Shipping	The product is shipped with polar packs. Upon receipt, store it immediately at the temperature recommended below.
Stability & Storage	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.

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