

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
Formulation	Lyophilized from a 0.2 µm filtered solution in PBS with Trehalose. See Certificate of Analysis for details. *Small pack size (-SP) is supplied either lyophilized or as a 0.2 µm filtered solution in PBS.

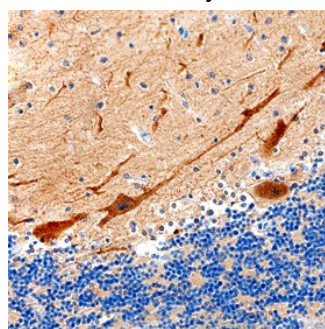
APPLICATIONS

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.

	Recommended Concentration	Sample
Immunohistochemistry	5-15 µg/mL	See Below

DATA

Immunohistochemistry



SACS in Human Brain. SACS was detected in immersion fixed paraffin-embedded sections of human brain (cerebellum) using Sheep Anti-Human SACS Antigen Affinity-purified Polyclonal Antibody (Catalog # AF8014) at 3 µg/mL overnight at 4 °C. Before incubation with the primary antibody, tissue was subjected to heat-induced epitope retrieval using Antigen Retrieval Reagent-Basic (Catalog # CTS013). Tissue was stained using the Anti-Sheep HRP-DAB Cell & Tissue Staining Kit (brown; Catalog # CTS019) and counterstained with hematoxylin (blue). Specific staining was localized to Purkinje neurons. View our protocol for [Chromogenic IHC Staining of Paraffin-embedded Tissue Sections](#).

PREPARATION AND STORAGE

Reconstitution	Sterile PBS to a final concentration of 0.2 mg/mL.
Shipping	The product is shipped at ambient temperature. Upon receipt, store it immediately at the temperature recommended below. *Small pack size (-SP) is shipped with polar packs. Upon receipt, store it immediately at -20 to -70 °C
Stability & Storage	Use a manual defrost freezer and avoid repeated freeze-thaw cycles. <ul style="list-style-type: none"> • 12 months from date of receipt, -20 to -70 °C as supplied. • 1 month, 2 to 8 °C under sterile conditions after reconstitution. • 6 months, -20 to -70 °C under sterile conditions after reconstitution.

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.