

# Aspartoacylase (D-11): sc-377308

## BACKGROUND

Aspartoacylase, also known as ASPA, ACY2 or ASP, is a 313 amino acid protein that is expressed in liver, lung and kidney tissue, as well as in skeletal muscle and in cerebral white matter. Existing as a homodimer, Aspartoacylase functions to catalyze the deacetylation of N-acetylaspatic acid (NAA) (a protein whose hydrolysis is crucial to maintenance of intact white matter) to produce acetate and L-aspartate. Defects in the gene encoding Aspartoacylase are the cause of Canavan disease (CAND), which is a rare neurodegenerative condition that is characterized by white matter vacuolization and demyelination, resulting in a spongy deterioration of brain tissue. CAND is generally characterized by atonia of neck muscles, hypotonia, hyperextension of legs and flexion of arms, blindness, severe mental retardation, megaloccephaly and death.

## REFERENCES

1. Kaul, R., et al. 1993. Cloning of the human aspartoacylase cDNA and a common missense mutation in Canavan disease. *Nat. Genet.* 5: 118-123.
2. Kaul, R., et al. 1994. Canavan disease: mutations among Jewish and non-Jewish patients. *Am. J. Hum. Genet.* 55: 34-41.
3. Olsen, T.R., et al. 2002. Two novel aspartoacylase gene (ASPA) missense mutations specific to Norwegian and Swedish patients with Canavan disease. *J. Med. Genet.* 39: e55.
4. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 608034. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
5. Le Coq, J., et al. 2006. Characterization of human aspartoacylase: the brain enzyme responsible for Canavan disease. *Biochemistry* 45: 5878-5884.
6. Hershfield, J.R., et al. 2006. Aspartoacylase is a regulated nuclear-cytoplasmic enzyme. *FASEB J.* 20: 2139-2141.
7. Hershfield, J.R., et al. 2007. Mutational analysis of aspartoacylase: implications for Canavan disease. *Brain Res.* 1148: 1-14.

## CHROMOSOMAL LOCATION

Genetic locus: ASPA (human) mapping to 17p13.2; Aspa (mouse) mapping to 11 B4.

## SOURCE

Aspartoacylase (D-11) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 241-270 within an internal region of Aspartoacylase of human origin.

## PRODUCT

Each vial contains 200 µg IgM kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-377308 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

## APPLICATIONS

Aspartoacylase (D-11) is recommended for detection of Aspartoacylase of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for Aspartoacylase siRNA (h): sc-93596, Aspartoacylase siRNA (m): sc-141303, Aspartoacylase shRNA Plasmid (h): sc-93596-SH, Aspartoacylase shRNA Plasmid (m): sc-141303-SH, Aspartoacylase shRNA (h) Lentiviral Particles: sc-93596-V and Aspartoacylase shRNA (m) Lentiviral Particles: sc-141303-V.

Molecular Weight of Aspartoacylase monomer: 38 kDa.

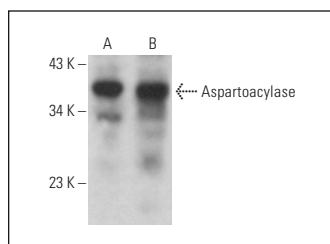
Molecular Weight of Aspartoacylase dimer: 84 kDa.

Positive Controls: mouse adipose tissue extract: sc-395042 or rat adipose tissue extract.

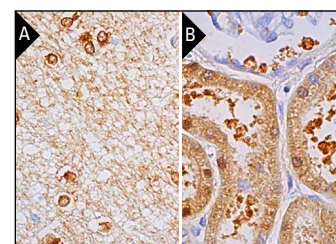
## RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein L-Agarose: sc-2336 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

## DATA



Aspartoacylase (D-11): sc-377308. Western blot analysis of Aspartoacylase expression in rat adipose (A) and mouse adipose (B) tissue extracts.



Aspartoacylase (D-11): sc-377308. Immunoperoxidase staining of formalin fixed, paraffin-embedded human cerebral cortex tissue showing nuclear and cytoplasmic staining of glial cells and neuropil staining (A) and human kidney tissue showing cytoplasmic and nuclear staining of cells in tubules (B). Blocked with 0.25X UltraCruz® Blocking Reagent: sc-516214. Detection reagents used: m-IgGκ BP-B: sc-516142 and ImmunoCruz® ABC Kit: sc-516216.

## STORAGE

Store at 4° C, \*\*DO NOT FREEZE\*\*. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

## RESEARCH USE

For research use only, not for use in diagnostic procedures.