

# Aspartoacylase (F-1): sc-365588

## 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: 55.
4. Online Mendelian Inheritance in Man, OMIM<sup>™</sup>. 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.
8. Bitto, E., et al. 2007. Structure of Aspartoacylase, the brain enzyme impaired in Canavan disease. *Proc. Natl. Acad. Sci. USA* 104: 456-461.

## CHROMOSOMAL LOCATION

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

## SOURCE

Aspartoacylase (F-1) is a mouse monoclonal antibody raised against amino acids 188-279 mapping within an internal region of Aspartoacylase of human origin.

## PRODUCT

Each vial contains 200 µg IgG<sub>1</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

## APPLICATIONS

Aspartoacylase (F-1) 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.

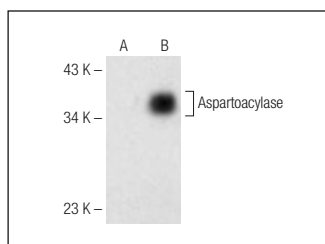
Molecular Weight of Aspartoacylase dimer: 84 kDa.

Positive Controls: Aspartoacylase (h): 293T Lysate: sc-114276, U-87 MG cell lysate: sc-2411 or rat adipose tissue extract.

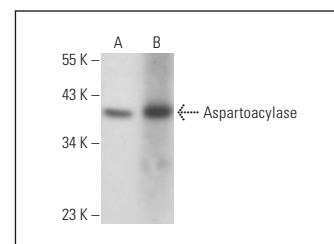
## RECOMMENDED SUPPORT REAGENTS

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<sup>™</sup> Molecular Weight Standards: sc-2035, UltraCruz<sup>®</sup> Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (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<sup>®</sup> Mounting Medium: sc-24941 or UltraCruz<sup>®</sup> Hard-set Mounting Medium: sc-359850.

## DATA



Aspartoacylase (F-1): sc-365588. Western blot analysis of Aspartoacylase expression in non-transfected: sc-117752 (A) and human Aspartoacylase transfected: sc-114276 (B) 293T whole cell lysates.



Aspartoacylase (F-1): sc-365588. Western blot analysis of Aspartoacylase expression in U-87 MG whole cell lysate (A) and rat adipose tissue extract (B).

## 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.