SANTA CRUZ BIOTECHNOLOGY, INC.

Aspartoacylase (h): 293T Lysate: sc-114276

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-acetylaspartic 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 demeylination, 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, megalocephaly 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.
8. Bitto, E., et al. 2007. Structure of aspartoacylase, the brain enzyme impaired in Canavan disease. Proc. Natl. Acad. Sci. USA 104: 456-461.
9. Le Coq, J., et al. 2008. Examination of the mechanism of human brain Aspartoacylase through the binding of an intermediate analogue. Biochemistry 47: 3484-3492.

CHROMOSOMAL LOCATION

Genetic locus: ASPA (human) mapping to 17p13.2.

PRODUCT

Aspartoacylase (h): 293T Lysate represents a lysate of human Aspartoacylase transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

APPLICATIONS

Aspartoacylase (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive Aspartoacylase antibodies. Recommended use: 10-20 µl per lane.

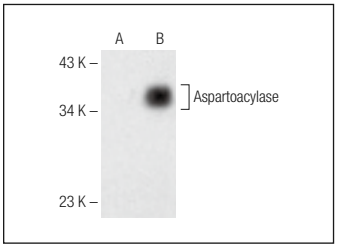
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

Aspartoacylase (F-1): sc-365588 is recommended as a positive control antibody for Western Blot analysis of enhanced human Aspartoacylase expression in Aspartoacylase transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

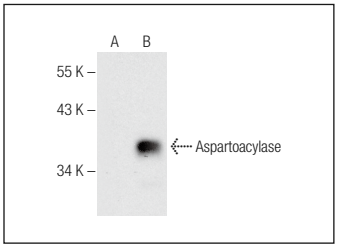
RECOMMENDED SUPPORT REAGENTS

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.

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 (D-11): sc-377308. Western blot analysis of Aspartoacylase expression in non-transfected: sc-117752 (**A**) and human Aspartoacylase transfected: sc-114276 (**B**) 293T whole cell lysates.

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.