

Produktinformation



Forschungsprodukte & Biochemikalien
Zellkultur & Verbrauchsmaterial
Diagnostik & molekulare Diagnostik
Laborgeräte & Service

Weitere Information auf den folgenden Seiten! See the following pages for more information!



Lieferung & Zahlungsart siehe unsere Liefer- und Versandbedingungen

Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

SZABO-SCANDIC HandelsgmbH

Quellenstraße 110, A-1100 Wien T. +43(0)1 489 3961-0 F. +43(0)1 489 3961-7 <u>mail@szabo-scandic.com</u> www.szabo-scandic.com

SANTA CRUZ BIOTECHNOLOGY, INC.

Aspartoacylase (h): 293T Lysate: sc-170145



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 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, 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.
- Kaul, R., et al. 1994. Canavan disease: mutations among Jewish and non-Jewish patients. Am. J. Hum. Genet. 55: 34-41.
- 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/
- 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.
- 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 μ l per lane.

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

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.