## SANTA CRUZ BIOTECHNOLOGY, INC.

# ADSL (D-12): sc-86274



## BACKGROUND

ADSL (adenylosuccinate lyase), also known as AMPS, ASL or ASASE, is a 484 amino acid protein that is involved in both purine biosynthesis and in the formation of adenosine monophosphate (AMP) from inosine monophosphate. Expressed ubiquitously, ADSL catalyzes two key reactions in AMP biosynthesis, namely the removal of a fumarate from succinylaminoimidazole carboxamide (SAICA) ribotide to give aminoimidazole carboxamide ribotide (AICA) and the subsequent removal of fumarate from adenylosuccinate to yield AMP. Defects in the gene encoding ADSL are the cause of adenylosuccinase deficiency (ADSL deficiency), an autosomal recessive disorder characterized by epilepsy, growth retardation and muscular wasting. Multiple isoforms of ADSL exist due to alternative splicing events.

#### REFERENCES

- Kmoch, S., et al. 2000. Human adenylosuccinate lyase (ADSL), cloning and characterization of full-length cDNA and its isoform, gene structure and molecular basis for ADSL deficiency in six patients. Hum. Mol. Genet. 9: 1501-1513.
- Race, V., et al. 2000. Clinical, biochemical and molecular genetic correlations in adenylosuccinate lyase deficiency. Hum. Mol. Genet. 9: 2159-2165.
- Tabucchi, A., et al. 2001. Determination, activity and biological role of adenylosuccinate lyase in blood cells. Biomed. Pharmacother. 55: 277-283.
- 4. Marie, S., et al. 2002. Mutation of a nuclear respiratory factor 2 binding site in the 5' untranslated region of the ADSL gene in three patients with adenylosuccinate lyase deficiency. Am. J. Hum. Genet. 71: 14-21.
- Edery, P., et al. 2003. Intrafamilial variability in the phenotypic expression of adenylosuccinate lyase deficiency: a report on three patients. Am. J. Med. Genet. A 120A: 185-190.
- Sivendran, S., et al. 2004. Two novel mutant human adenylosuccinate lyases (ASLs) associated with autism and characterization of the equivalent mutant *Bacillus subtilis* ASL. J. Biol. Chem. 279: 53789-53797.
- Lee, P., et al. 2007. Expression, purification, and characterization of stable, recombinant human adenylosuccinate lyase. Protein Expr. Purif. 51: 227-234.
- 8. Online Mendelian Inheritance in Man, OMIM™. 2009. Johns Hopkins University, Baltimore, MD. MIM Number: 608222. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/

#### CHROMOSOMAL LOCATION

Genetic locus: ADSL (human) mapping to 22q13.1; Adsl (mouse) mapping to 15 E1.

#### SOURCE

ADSL (D-12) is an affinity purified rabbit polyclonal antibody raised against a peptide mapping within an internal region of ADSL of human origin.

## **STORAGE**

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

#### PRODUCT

Each vial contains 100  $\mu g$  IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-86274 P, (100  $\mu$ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## **APPLICATIONS**

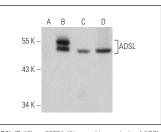
ADSL (D-12) is recommended for detection of ADSL of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2  $\mu$ g per 100-500  $\mu$ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution 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 ADSL siRNA (h): sc-72457, ADSL siRNA (m): sc-140888, ADSL shRNA Plasmid (h): sc-72457-SH, ADSL shRNA Plasmid (m): sc-140888-SH, ADSL shRNA (h) Lentiviral Particles: sc-72457-V and ADSL shRNA (m) Lentiviral Particles: sc-140888-V.

Molecular Weight of ADSL: 52 kDa.

Positive Controls: ADSL (h3): 293T Lysate: sc-170309, Jurkat whole cell lysate: sc-2204 or HL-60 whole cell lysate: sc-2209

#### DATA



ADSL (D-12): sc-86274. Western blot analysis of ADSL expression in non-transfected 293T: sc-11752 (**A**), human ADSL transfected 293T: sc-170309 (**B**), Jurkat (**C**) and HL-60 (**D**) whole cell lysates.

#### **RESEARCH USE**

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

#### **PROTOCOLS**

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

MONOS Satisfation Guaranteed