

# ADSL siRNA (h): sc-72457

## 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 adenylosuccinate 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

1. 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.
2. Race, V., et al. 2000. Clinical, biochemical and molecular genetic correlations in adenylosuccinate lyase deficiency. *Hum. Mol. Genet.* 9: 2159-2165.
3. 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.
5. 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.
6. 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.

## CHROMOSOMAL LOCATION

Genetic locus: ADSL (human) mapping to 22q13.1.

## PRODUCT

ADSL siRNA (h) is a pool of 3 target-specific 19-25 nt siRNAs designed to knock down gene expression. Each vial contains 3.3 nmol of lyophilized siRNA, sufficient for a 10  $\mu$ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see ADSL shRNA Plasmid (h): sc-72457-SH and ADSL shRNA (h) Lentiviral Particles: sc-72457-V as alternate gene silencing products.

For independent verification of ADSL (h) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-72457A, sc-72457B and sc-72457C.

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) for detailed protocols and support products.

## STORAGE AND RESUSPENSION

Store lyophilized siRNA duplex at -20° C with desiccant. Stable for at least one year from the date of shipment. Once resuspended, store at -20° C, avoid contact with RNases and repeated freeze thaw cycles.

Resuspend lyophilized siRNA duplex in 330  $\mu$ l of the RNase-free water provided. Resuspension of the siRNA duplex in 330  $\mu$ l of RNase-free water makes a 10  $\mu$ M solution in a 10  $\mu$ M Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

## APPLICATIONS

ADSL siRNA (h) is recommended for the inhibition of ADSL expression in human cells.

## SUPPORT REAGENTS

For optimal siRNA transfection efficiency, Santa Cruz Biotechnology's siRNA Transfection Reagent: sc-29528 (0.3 ml), siRNA Transfection Medium: sc-36868 (20 ml) and siRNA Dilution Buffer: sc-29527 (1.5 ml) are recommended. Control siRNAs or Fluorescein Conjugated Control siRNAs are available as 10  $\mu$ M in 66  $\mu$ l. Each contain a scrambled sequence that will not lead to the specific degradation of any known cellular mRNA. Fluorescein Conjugated Control siRNAs include: sc-36869, sc-44239, sc-44240 and sc-44241. Control siRNAs include: sc-37007, sc-44230, sc-44231, sc-44232, sc-44233, sc-44234, sc-44235, sc-44236, sc-44237 and sc-44238.

## GENE EXPRESSION MONITORING

ADSL (C-11): sc-365623 is recommended as a control antibody for monitoring of ADSL gene expression knockdown by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) or immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500).

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG $\kappa$  BP-HRP: sc-516102 or m-IgG $\kappa$  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) Immunofluorescence: use m-IgG $\kappa$  BP-FITC: sc-516140 or m-IgG $\kappa$  BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

## RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor ADSL gene expression knockdown using RT-PCR Primer: ADSL (h)-PR: sc-72457-PR (20  $\mu$ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

## RESEARCH USE

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