

# ABAT (h3): 293T Lysate: sc-159887

## BACKGROUND

In the central nervous system GABA ( $\gamma$ -aminobutyric acid) functions as the main inhibitory transmitter by increasing a  $Cl^-$  conductance that inhibits neuronal firing. ABAT (4-aminobutyrate aminotransferase), also known as GABAT (GABA aminotransferase), L-AIBAT or (S)-3-amino-2-methylpropionate transaminase, is a 500 amino acid mitochondrial matrix protein belonging to the class-III pyridoxal-phosphate-dependent aminotransferase family, which catabolizes GABA into succinic semialdehyde. Existing as a homodimer, ABAT binds pyridoxal phosphate as a cofactor and is expressed in liver, brain, pancreas, kidney, placenta and heart. The gene encoding ABAT maps to human chromosome 16p13.2, and defects in ABAT are the cause of GABA-AT deficiency, which is characterized by hypotonia, hyper-reflexia, psychomotor retardation, lethargy, EEG abnormalities and refractory seizures.

## REFERENCES

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- Jaeken, J., et al. 1984.  $\gamma$ -aminobutyric acid-transaminase deficiency: a newly recognized inborn error of neurotransmitter metabolism. *Neuropediatrics* 15: 165-169.
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- Osei, Y.D., et al. 1995. Screening and sequence determination of a cDNA encoding the human brain 4-aminobutyrate aminotransferase. *Gene* 155: 185-187.
- Medina-Kauwe, L.K., et al. 1999. 4-Aminobutyrate aminotransferase (GABA-transaminase) deficiency. *J. Inher. Metab. Dis.* 22: 414-427.
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## CHROMOSOMAL LOCATION

Genetic locus: ABAT (human) mapping to 16p13.2.

## PRODUCT

ABAT (h3): 293T Lysate represents a lysate of human ABAT transfected 293T cells and is provided as 100  $\mu$ g protein in 200  $\mu$ l SDS-PAGE buffer.

## APPLICATIONS

ABAT (h3): 293T Lysate is suitable as a Western Blotting positive control for human reactive ABAT antibodies. Recommended use: 10-20  $\mu$ l per lane.

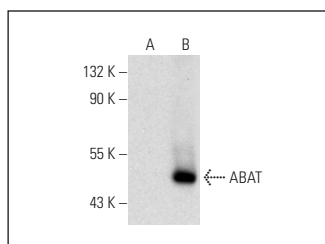
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

ABAT (B-12): sc-393769 is recommended as a positive control antibody for Western Blot analysis of enhanced human ABAT expression in ABAT 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-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.

## DATA



ABAT (B-12): sc-393769. Western blot analysis of ABAT expression in non-transfected: sc-117752 (A) and human ABAT transfected: sc-159887 (B) 293T whole cell lysates.

## STORAGE

Store at  $-20^{\circ}$  C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

## RESEARCH USE

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

## PROTOCOLS

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