SANTA CRUZ BIOTECHNOLOGY, INC.

# DBT siRNA (m): sc-142884

## BACKGROUND

DBT (dihydrolipoamide branched chain transacylase), also known as BCATE2 (branched-chain α-keto acid dehydrogenase complex component E2) or E2B, is a 482 amino acid enzyme that catalyzes the overall conversion of α-keto acids to acyl-CoA and CO2. Belonging to the 2-oxoacid dehydrogenase family, DBT has one lipoyl-binding domain and contains multiple copies of three enzymatic components designated branched-chain α-keto acid decarboxylase, lipoamide acyltransferase and lipoamide dehydrogenase. Localized to the mitochondrion matrix, DBT binds one lipoyl cofactor covalently. Defects in the gene that encodes DBT are the cause of maple syrup urine disease type 2 (MSUD2), which is characterized by mental and physical retardation, feeding problems and a maple syrup odor to the urine.

## REFERENCES


1. Edelmann, L., et al. 2001. Maple syrup urine disease: identification and carrier-frequency determination of a novel founder mutation in the Ashkenazi Jewish population. Am. J. Hum. Genet. 69: 863-868.
2. Chang, C.F., et al. 2002. Solution structure and dynamics of the lipoic acid-bearing domain of human mitochondrial branched-chain α-keto acid dehydrogenase complex. J. Biol. Chem. 277: 15865-15873.
3. Csepregi, A., et al. 2003. Characterization of a lipoyl domain-independent B-cell autoepitope on the human branched-chain acyltransferase in primary biliary cirrhosis and overlap syndrome with autoimmune hepatitis. Clin. Dev. Immunol. 10: 173-181.
4. Chang, C.F., et al. 2006. Structure of the subunit binding domain and dynamics of the di-domain region from the core of human branched chain α-ketoacid dehydrogenase complex. J. Biol. Chem. 281: 28345-28353.
5. Flaschker, N., et al. 2007. Description of the mutations in 15 subjects with variant forms of maple syrup urine disease. J. Inherit. Metab. Dis. 30: 903-909.
6. Bogenhagen, D.F., et al. 2008. The layered structure of human mitochondrial DNA nucleoids. J. Biol. Chem. 283: 3665-3675.
7. Quental, S., et al. 2008. Molecular and structural analyses of maple syrup urine disease and identification of a founder mutation in a Portuguese Gypsy community. Mol. Genet. Metab. 94: 148-156.
8. Silao, C.L., et al. 2008. Early diagnosis of maple syrup urine disease using polymerase chain reaction-based mutation detection. Pediatr. Int. 50: 312-314.
9. Gorzelany, K., et al. 2009. Molecular genetics of maple syrup urine disease in the Turkish population. Turk. J. Pediatr. 51: 97-102.


## CHROMOSOMAL LOCATION

Genetic locus: Dbt (mouse) mapping to 3 G1.

## PROTOCOLS

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

## PRODUCT

DBT siRNA (m) 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 µM solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see DBT shRNA Plasmid (m): sc-142884-SH and DBT shRNA (m) Lentiviral Particles: sc-142884-V as alternate gene silencing products.

For independent verification of DBT (m) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-142884A, sc-142884B and sc-142884C.

## 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 µl of the RNAse-free water provided. Resuspension of the siRNA duplex in 330 µl of RNAse-free water makes a 10 µM solution in a 10 µM Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

## APPLICATIONS

DBT siRNA (m) is recommended for the inhibition of DBT expression in mouse 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 µM in 66 µl. 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.

## RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor DBT gene expression knockdown using RT-PCR Primer: DBT (m)-PR: sc-142884-PR (20 µl). 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.