

BCKDHB FISH Probe

Catalog # FA0171 Size 200 uL

Specification

Product Description	Made to order FISH probes for identification of gene amplification using Fluorescent In Situ Hybridization Technique. (Technology).
Origin	Human
Source	Genomic DNA
Reactivity	Human
Notice	We strongly recommend the customer to use FFPE FISH PreTreatment Kit 1 (Catalog #: KA2375 or KA2691) for the pretreatment of Formalin-Fixed Paraffin-Embedded (FFPE) tissue sections.
Regulation Status	For research use only (RUO)
Supplied Product	DAPI Counterstain (1500 ng/mL) 250 uL
Storage Instruction	Store at 4°C in the dark.

Applications

- Fluorescent In Situ Hybridization (Cell)

[Protocol Download](#)

Gene Info — BCKDHB

Entrez GeneID	594
Gene Name	BCKDHB
Gene Alias	E1B, FLJ17880, dJ279A18.1
Gene Description	branched chain keto acid dehydrogenase E1, beta polypeptide

Omim ID [248600 248611](#)

Gene Ontology [Hyperlink](#)

Gene Summary

Branched-chain keto acid dehydrogenase is a multienzyme complex associated with the inner membrane of mitochondria, and functions in the catabolism of branched-chain amino acids. The complex consists of multiple copies of 3 components: branched-chain alpha-keto acid decarboxylase (E1), lipoamide acyltransferase (E2) and lipoamide dehydrogenase (E3). This gene encodes the E1 beta subunit, and mutations therein have been associated with maple syrup urine disease (MSUD), type 1B, a disease characterized by a maple syrup odor to the urine in addition to mental and physical retardation, and feeding problems. Alternative splicing at this locus results in transcript variants with different 3' non-coding regions, but encoding the same isoform. [provided by RefSeq]

Other Designations

2-oxoisovalerate dehydrogenase beta subunit|E1b-beta subunit of the branched-chain complex|OTTHUMP00000018048|OTTHUMP00000018049|branched chain alpha-ketoacid dehydrogenase E1-beta subunit|branched chain keto acid dehydrogenase E1 beta polypeptide|branched chain keto acid dehydrogenase E1 beta subunit

Pathway

- [Metabolic pathways](#)
- [Valine](#)

Disease

- [Maple syrup urine disease](#)
- [Tobacco Use Disorder](#)