



# SZABO SCANDIC

Part of Europa Biosite

## Produktinformation



Forschungsprodukte & Biochemikalien



Zellkultur & Verbrauchsmaterial



Diagnostik & molekulare Diagnostik



Laborgeräte & Service

Weitere Information auf den folgenden Seiten!  
See the following pages for more information!



### Lieferung & Zahlungsart

siehe unsere [Liefer- und Versandbedingungen](#)

### Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

### SZABO-SCANDIC HandelsgmbH

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## BCKDHB FISH Probe

Catalog # : FA0171

規格 : [ 200 uL ]

List All

### Specification

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

### Application Image

Fluorescent In Situ Hybridization (Cell)

### Applications

Fluorescent In Situ Hybridization (Cell)

 [Protocol Download](#)

### Gene Information

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** 2-oxoisovalerate dehydrogenase beta subunit,E1b-beta subunit of the  
**Designations:** branched-chain  
complex,OTTHUMP00000018048,OTTHUMP00000018049,branched  
chain alpha-ketoacid dehydrogenase E1-beta subunit,branched chain  
keto acid dehydrogenase E1 beta polypeptide,branche

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#### **Gene Pathway**

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Metabolic pathways Valine, leucine and isoleucine degradation

#### **Related Disease**

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Maple Syrup Urine Disease Maple syrup urine disease Tobacco Use Disorder

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