

Trifunctional enzyme subunit beta, mitochondrial Polyclonal Conjugated Antibody

Catalog No: #C42383

Orders: order@signalwayantibody.comSupport: tech@signalwayantibody.com

Package Size: #C42383-AF350 100ul #C42383-AF405 100ul #C42383-AF488 100ul

#C42383-AF555 100ul #C42383-AF594 100ul #C42383-AF647 100ul

#C42383-AF680 100ul #C42383-AF750 100ul #C42383-Biotin 100ul

Description

Product Name	Trifunctional enzyme subunit beta, mitochondrial Polyclonal Conjugated Antibody
Host Species	Rabbit
Clonality	Polyclonal
Species Reactivity	Hu
Specificity	The antibody detects endogenous level of total Trifunctional enzyme subunit beta, mitochondrial polyclonal antibody.
Immunogen Description	Recombinant human Trifunctional enzyme subunit beta, mitochondrial protein
Conjugates	Biotin AF350 AF405 AF488 AF555 AF594 AF647 AF680 AF750
Other Names	P55084
Accession No.	Swiss-Prot#:P55084
Uniprot	P55084
GeneID	3032;
Excitation Emission	AF350: 346nm/442nm AF405: 401nm/421nm AF488: 493nm/519nm AF555: 555nm/565nm AF594: 591nm/614nm AF647: 651nm/667nm AF680: 679nm/702nm AF750: 749nm/775nm
Formulation	0.01M Sodium Phosphate, 0.25M NaCl, pH 7.6, 5mg/ml Bovine Serum Albumin, 0.02% Sodium Azide
Storage	Store at 4°C in dark for 6 months

Application Details

Suggested Dilution:

AF350 conjugated: most applications: 1: 50 - 1: 250

AF405 conjugated: most applications: 1: 50 - 1: 250

AF488 conjugated: most applications: 1: 50 - 1: 250

AF555 conjugated: most applications: 1: 50 - 1: 250

AF594 conjugated: most applications: 1: 50 - 1: 250

AF647 conjugated: most applications: 1: 50 - 1: 250

AF680 conjugated: most applications: 1: 50 - 1: 250

AF750 conjugated: most applications: 1: 50 - 1: 250

Background

Defects in HADHB are a cause of trifunctional protein deficiency (TFP deficiency) [MIM:609015]. The clinical manifestations are very variable and include hypoglycemia, cardiomyopathy and sudden death. Phenotypes with mainly hepatic and neuromyopathic involvement can also be distinguished. Biochemically, TFP deficiency is defined by the loss of all three enzyme activities of the TFP complex.

Note: This product is for in vitro research use only