

Trifunctional enzyme subunit beta, mitochondrial Polyclonal Antibody

Catalog No: #42383

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

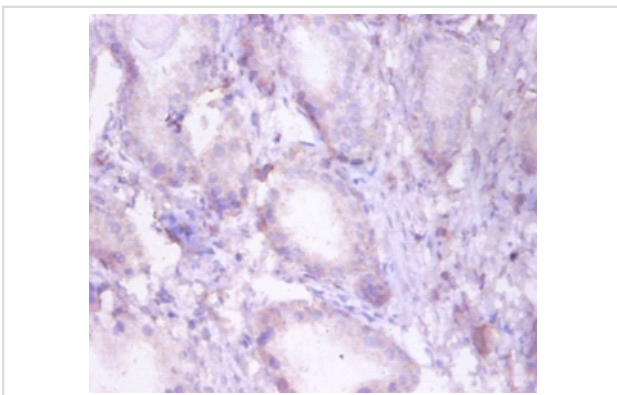
Description

Product Name	Trifunctional enzyme subunit beta, mitochondrial Polyclonal Antibody
Host Species	Rabbit
Clonality	Polyclonal
Purification	Caprylic Acid Ammonium Sulfate Precipitation purified
Applications	IHC
Species Reactivity	Hu
Specificity	The antibody detects endogenous level of total Trifunctional enzyme subunit beta, mitochondrial polyclonal antibody.
Immunogen Type	protein
Immunogen Description	Recombinant human Trifunctional enzyme subunit beta, mitochondrial protein
Target Name	Trifunctional enzyme subunit beta, mitochondrial
Other Names	P55084
Accession No.	Swiss-Prot#: P55084
Uniprot	P55084
GeneID	3032;
Formulation	Preservative: 0.03% Proclin 300 Constituents: 50% Glycerol, 0.01M PBS, PH 7.4
Storage	Store at -20°C

Application Details

Immunohistochemistry: 1:20 - 1:200

Images



Immunohistochemical analysis of paraffin-embedded human prostate using #42383 at dilution of 1:50 .

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.

References

[1] "Structural analysis of cDNAs for subunits of human mitochondrial fatty acid beta-oxidation trifunctional protein." Kamijo T., Aoyama T., Komiyama A., Hashimoto T. *Biochem. Biophys. Res. Commun.* 199:818-825(1994) [2] "Genomic and mutational analysis

Note: This product is for in vitro research use only