



Rabbit Anti-SLC19A3 antibody

SL8702R

Product Name:	SLC19A3
Chinese Name:	溶质载体家族19成员3抗体
Alias:	Solute carrier family 19 member 3; Thiamine transporter 2; thTr 2; THTR2.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	56kDa
Cellular localization:	The cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human SLC19A3:261-360/496<Extracellular>
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	Preservative: 15mM Sodium Azide, Constituents: 1% BSA, 0.01M PBS, pH 7.4
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	This gene encodes a ubiquitously expressed transmembrane thiamine transporter that lacks folate transport activity. Mutations in this gene cause biotin-responsive basal ganglia disease (BBGD); a recessive disorder manifested in childhood that progresses to chronic encephalopathy, dystonia, quadriplegia, and death if untreated. Patients with BBGD have bilateral necrosis in the head of the caudate nucleus and in the putamen. Administration of high doses of biotin in the early progression of the disorder

eliminates pathological symptoms while delayed treatment results in residual paraparesis, mild mental retardation, or dystonia. Administration of thiamine is ineffective in the treatment of this disorder. Experiments have failed to show that this protein can transport biotin. Mutations in this gene also cause a Wernicke's-like encephalopathy.[provided by RefSeq, Jan 2010]

Function:

Mediates high affinity thiamine uptake, probably via a proton anti-transport mechanism. Has no folate transport activity.

Subcellular Location:

Plasma membrane.

Tissue Specificity:

Widely expressed but most abundant in placenta, kidney and liver.

DISEASE:

The disease is caused by mutations affecting the gene represented in this entry. Disease description: An autosomal recessive metabolic disorder characterized by episodic encephalopathy, often triggered by febrile illness, presenting as confusion, seizures, external ophthalmoplegia, dysphagia, and sometimes coma and death. If untreated, encephalopathies can result in permanent dystonia. Brain imaging may show characteristic bilateral lesions of the basal ganglia.

Similarity:

Belongs to the reduced folate carrier (RFC) transporter (TC 2.A.48) family.

SWISS:

Q9BZV2

Gene ID:

80704

Database links:

[Entrez Gene: 80704](#) Human

[Omid: 606152](#) Human

[SwissProt: Q9BZV2](#) Human

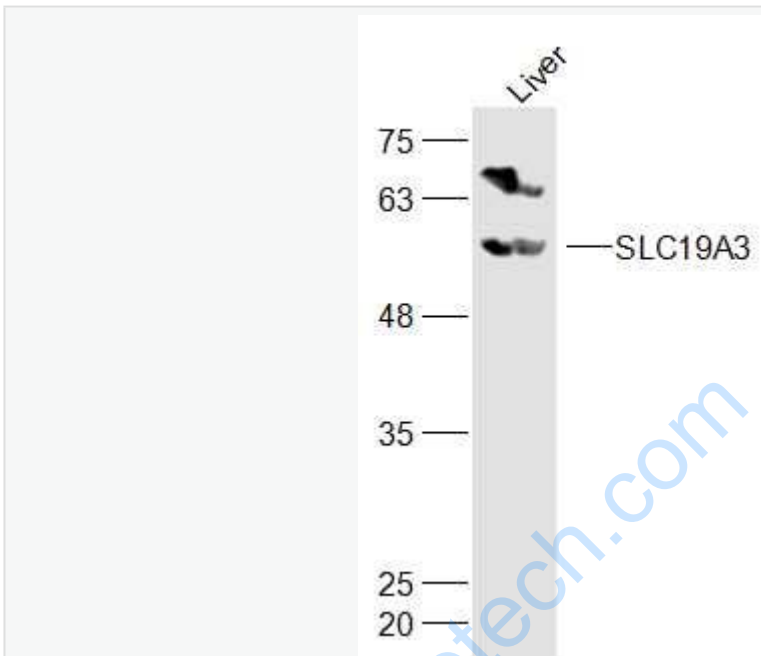
[Unigene: 221597](#) Human

Important Note:

This product as supplied is intended for research use only, not for use in human,

therapeutic or diagnostic applications.

Picture:



Sample:

Liver (Mouse) Lysate at 40 ug

Primary: Anti-SLC19A3 (SL8702R) at 1/300 dilution

Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

Predicted band size: 56 kD

Observed band size: 56 kD