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Rabbit Anti-CYB5R3 antibody

SL12162R

Product Name:	CYB5R3
Chinese Name:	细胞色素b5还原酶3抗体
Alias:	B5R; Cyb5r3; Cytochrome b5 reductase 3; Cytochrome b5 reductase; DIA1; Diaphorase 1; Diaphorase-1; NADH cytochrome b5 reductase 3; NADH-cytochrome b5 reductase 3 membrane-bound form; NADH-cytochrome b5 reductase 3 soluble form; NB5R3_HUMAN; OTTHUMP00000028761; OTTHUMP00000198435; OTTHUMP00000198574; OTTHUMP00000198662; OTTHUMP00000198665.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Dog,Pig,Cow,Horse,Sheep,
Applications:	WB=1:500-2000ELISA=1:500-1000 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	34kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human CYB5R3:101-200/301
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
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:	CYB5R3 is a 301 amino acid protein encoded by the human gene CYB5R3. CYB5R3 belongs to the flavoprotein pyridine nucleotide cytochrome reductase family and has two naturally occuring isoforms. Isoform 1 is anchored to the cytoplasmic side of the endoplasmic reticulum membrane and mitochondrion outer membrane, while isoform 2

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is the soluble form found in erythrocytes. CYB5R3 is involved in the desaturation and elongation of fatty acids, cholesterol biosynthesis, drug metabolism and, in erythrocytes, methemoglobin reduction. A serine residue at position 117 seems to only be found in persons of African origin. The allele frequency is 0.23 in African Americans. It is not found in Caucasians, Asians, Indo-Aryans or Arabs. This difference seems to have no effect on the enzyme activity. Defects in CYB5R3 are the cause of hereditary methemoglobinemia (HM). There are three forms of this disease: type 1 (HM1), in which the enzyme is only deficient in erythrocytes with a mild cyanosis; type 2 (HM2), in which the enzyme is completely deficient; and type 3 (HM3), where the deficiency is seen in all blood cells. Type 2 is a severe form accompanied by mental retardation and neurological impairment.

Function:
Desaturation and elongation of fatty acids, cholesterol biosynthesis, drug metabolism, and, in erythrocyte, methemoglobin reduction.

Subunit:
Component of a complex composed of cytochrome b5, NADH-cytochrome b5 reductase (CYB5R3) and MOSC2 (By similarity).

Subcellular Location:
Endoplasmic reticulum membrane. Mitochondrion outer membrane and Cytoplasm. Produces the soluble form found in erythrocytes.

Tissue Specificity:
Isoform 2 is expressed at late stages of erythroid maturation.

DISEASE:
Defects in CYB5R3 are the cause of methemoglobinemia CYB5R3-related (METHB-CYB5R3) [MIM:250800]. A form of methemoglobinemia, a hematologic disease characterized by the presence of excessive amounts of methemoglobin in blood cells, resulting in decreased oxygen carrying capacity of the blood, cyanosis and hypoxia. There are two types of methemoglobinemia CYB5R3-related. In type 1, the defect affects the soluble form of the enzyme, is restricted to red blood cells, and causes well-tolerated methemoglobinemia. In type 2, the defect affects both the soluble and microsomal forms of the enzyme and is thus generalized, affecting red cells, leukocytes and all body tissues. Type 2 methemoglobinemia is associated with mental deficiency and other neurologic symptoms.

Similarity:
Belongs to the flavoprotein pyridine nucleotide cytochrome reductase family. Contains 1 FAD-binding FR-type domain.

SWISS:
P00387

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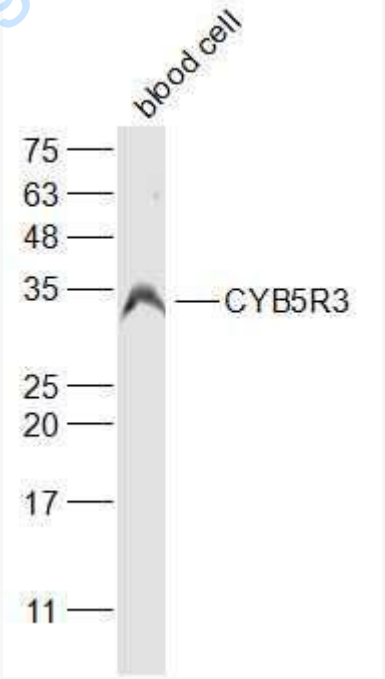
	Gene ID: 1727 **Database links:** Entrez Gene: 1727Human Entrez Gene: 25035Rat Omim: 613213Human SwissProt: P00387Human SwissProt: P20070Rat Unigene: 561064Human Unigene: 35994Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	 Sample: Blood(Mouse) Cell Lysate at 30 ug

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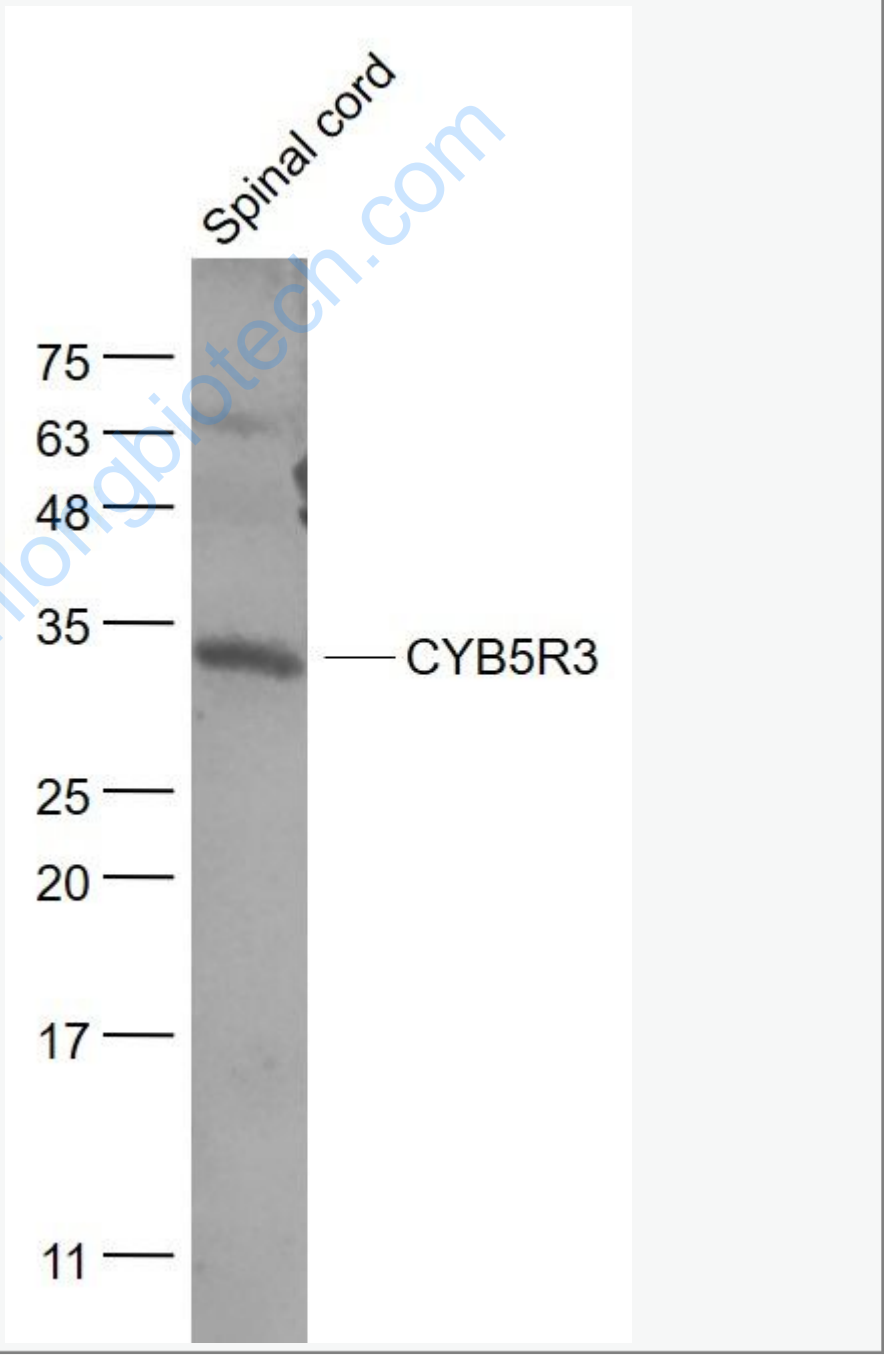
Primary: Anti-CYB5R3 (SL12162R) at 1/1000 dilution

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

Predicted band size: 34 kD

Observed band size: 34 kD



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	Sample: Spinal cord (Mouse) Lysate at 40 ug Primary: Anti- CYB5R3 (SL12162R) at 1/1000 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 34 kD Observed band size: 34 kD

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