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Rabbit Anti-ARSA antibody

SL4006R

Product Name:	ARSA
Chinese Name:	芳基硫酸酯酶A抗体
Alias:	As 2; As2; ASA; metachromatic leucodystrophy; TISP73; arylsulfatase A; AS A; MLD.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,Rabbit,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=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:	45/54kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human ARSA :368-412/507
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:	The protein encoded by this gene hydrolyzes cerebroside sulfate to cerebroside and sulfate. Defects in this gene lead to metachromatic leucodystrophy (MLD), a progressive demyelination disease which results in a variety of neurological symptoms and ultimately death. Alternatively spliced transcript variants have been described for this gene. [provided by RefSeq, Dec 2010]. **Function:**

Hydrolyzes cerebroside sulfate.

Subunit:
Homodimer at neutral pH and homooctamer at acidic pH. Exists both as a single chain of 58 kDa (component A) or as a chain of 50 kDa (component B) linked by disulfide bond(s) to a 7 kDa chain (component C). Interacts with SUMF1.

Subcellular Location:
Lysosome.

Post-translational modifications:
The conversion to 3-oxoalanine (also known as C-formylglycine, FGly), of a serine or cysteine residue in prokaryotes and of a cysteine residue in eukaryotes, is critical for catalytic activity. This post-translational modification is severely defective in multiple sulfatase deficiency (MSD).

DISEASE:
Defects in ARSA are a cause of leukodystrophy metachromatic (MLD) [MIM:250100]. MLD is a disease due to a lysosomal storage defect. It is characterized by intralysosomal storage of cerebroside-3-sulfate in neural and non-neural tissues, with a diffuse loss of myelin in the central nervous system. Progressive demyelination causes a variety of neurological symptoms, including gait disturbances, ataxias, optical atrophy, dementia, seizures, and spastic tetraparesis. Three forms of the disease can be distinguished according to the age at onset: late-infantile, juvenile and adult.
Arylsulfatase A activity is defective in multiple sulfatase deficiency (MSD) [MIM:272200]. A clinically and biochemically heterogeneous disorder caused by the simultaneous impairment of all sulfatases, due to defective post-translational modification and activation. It combines features of individual sulfatase deficiencies such as metachromatic leukodystrophy, mucopolysaccharidosis, chondrodysplasia punctata, hydrocephalus, ichthyosis, neurologic deterioration and developmental delay. Note=Arylsulfatase A activity is impaired in multiple sulfatase deficiency due to mutations in SUMF1. SUMF1 mutations result in defective post-translational modification of ARSA at residue Cys-69 that is not converted to 3-oxoalanine.

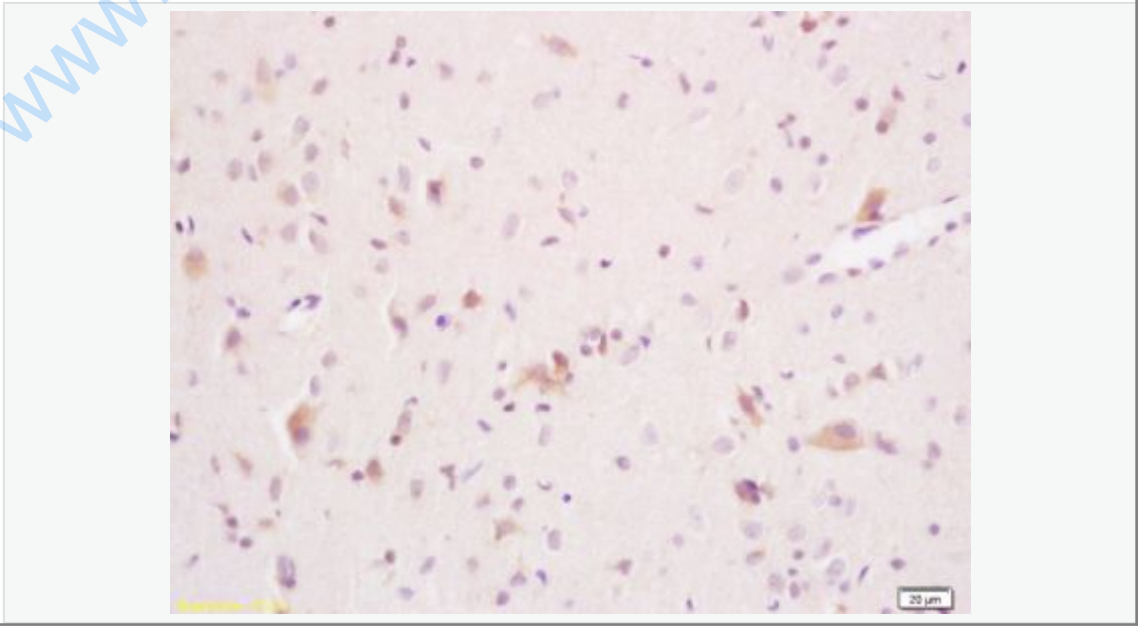
Similarity:
Belongs to the sulfatase family.

SWISS:
P15289

Gene ID:
410

Database links:

Entrez Gene: 410 Human

	Entrez Gene: 11883 Mouse Entrez Gene: 315222 Rat Omim: 607574 Human SwissProt: P15289 Human SwissProt: P50428 Mouse Unigene: 88251 Human Unigene: 620 Mouse Unigene: 23323 Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications. ArsA蛋白是存在于微生物The cell membrane上的一种亚砷酸根阴离子泵的水溶性部分。在亚砷酸根存在的情况下。ArsA具有ATP酶活力。它水解ATP,发生构象变化.芳基硫酯酶A（ArylsulfataseA，ARSA）的缺陷，使溶酶体内脑硫酯水解受阻，沉积于中枢神经系统的白质、周围神经系统及其它内脏组织，导致异染性脑白质营养不良（Metachromatic Leukodystrophy，MLD），他是一种较常见的脑白质营养不良，也是一种最常见的溶酶体病，为常染色体隐性遗传。
Picture:	

	Tissue/cell: rat brain tissue; 4% Paraformaldehyde-fixed and paraffin-embedded; Antigen retrieval: citrate buffer (0.01M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37℃ for 20 min; Incubation: Anti-ARSA Polyclonal Antibody, Unconjugated(SL4006R) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining