

Rabbit Anti-ARSA/AP Conjugated antibody

SL4006R-AP

Product Name	Anti-ARSA/AP
Chinese Name	碱性磷酸酶 (AP) 标记的芳基硫酸酯酶 A 抗体
Alias	As 2; As2; ASA; metachromatic leucodystrophy; TISP73; arylsulfatase A; ASA; MLD.
Research Area	immunology Neurobiology
Immunogen Species	Rabbit
Clonality	Polyclonal
React Species	Mouse,Rat(predicted:Human,Dog,Pig,Cow,Horse,Rabbit) WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500
Applications	not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight	47/54kDa
Form	Lyophilized or Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from human ARSA (368-412aa)
Lsotype	IgG
Purification	affinity purified by Protein A
Storage Buffer	1M TBS(pH7.4) with 1% BSA, 3% Proclin300 and 50% Glycerol. 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 1M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
Storage	
Product Detail	background: 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.

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.芳基硫酸酯酶 A (ArylsulfataseA, ARSA) 的缺陷,使溶酶体内脑硫酸酯水解受阻,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。