

Product Name: Arylsulfatase A Rabbit Polyclonal Antibody
Catalog #: APRab07182

Summary

Production Name	Arylsulfatase A Rabbit Polyclonal Antibody
Description	Rabbit Polyclonal Antibody
Host	Rabbit
Application	WB,IHC-P,IF-P,IF-F,ICC/IF,ELISA
Reactivity	Human,Mouse,Rat

Performance

Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
Buffer	Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type preservative N.
Purification	Affinity purification

Immunogen

Gene Name	ARSA
Alternative Names	ARSA; Arylsulfatase A; ASA; Cerebroside-sulfatase
Gene ID	410.0
SwissProt ID	P15289.The antiserum was produced against synthesized peptide derived from human ARSA. AA range:251-300

Application

Dilution Ratio	WB 1:500-1:2000, IHC-P 1:100-1:300, ELISA 1:20000, IF-P/IF-F/ICC/IF 1:50-200
Molecular Weight	54kDa

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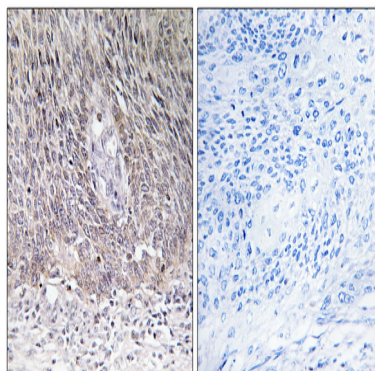
Background

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], catalytic activity: A cerebroside 3-sulfate + H₂O = a cerebroside + sulfate., cofactor: Binds 1 magnesium ion per subunit., disease: Arylsulfatase A activity is defective in multiple sulfatase deficiency (MSD) [MIM:272200]. MSD is a disorder characterized by decreased activity of all known sulfatases. MSD is due to defects in SUMF1 resulting in the lack of post-translational modification of a highly conserved cysteine into 3-oxoalanine. It combines features of individual sulfatase deficiencies such as metachromatic leukodystrophy, mucopolysaccharidosis, chondrodysplasia punctata, hydrocephalus, ichthyosis, neurologic deterioration and developmental delay., 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., function: Hydrolyzes cerebroside sulfate., online information: Arylsulfatase A entry, PTM: 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)., similarity: Belongs to the sulfatase family., 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).

Research Area

Sphingolipid metabolism; Lysosome;

Image Data

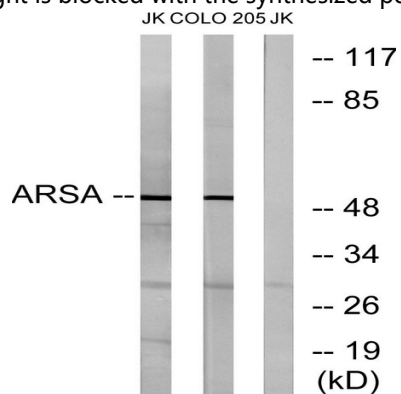


Immunohistochemistry analysis of paraffin-embedded human cervix carcinoma tissue, using ARSA Antibody. The picture on

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the right is blocked with the synthesized peptide.



Western blot analysis of lysates from Jurkat and COLO cells, using ARSA Antibody. The lane on the right is blocked with the synthesized peptide.

Note

For research use only.