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% BSA 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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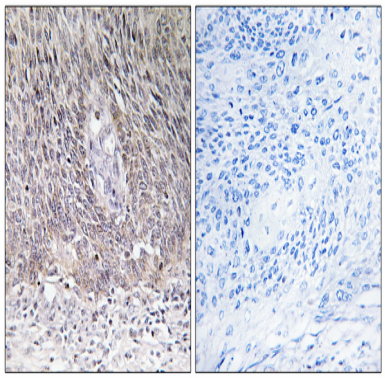
Background

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 + H(2)O = 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 the right is blocked with the synthesized peptide.

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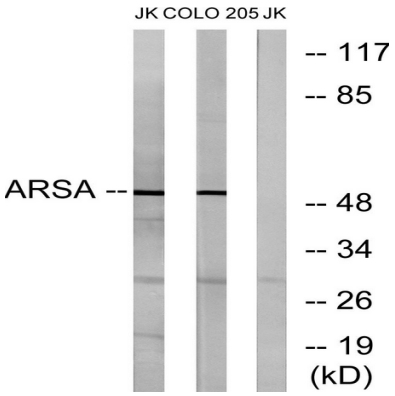

Western blot analysis of lysates from Jurkat and COLO cells, using ARSA Antibody. The lane on the right is blocked with the synthesized peptide.

Note

For research use only.