

产品货号: APRab07182



产品概述 (Summary)

产品名称 (Production Name) Arylsulfatase A Rabbit Polyclonal Antibody

描述 (Description) Rabbit polyclonal Antibody

宿主 (Host) Rabbit

应用 (Application)WB,IHC,ICC/IF,ELISA种属反应性 (Reactivity)Human,Mouse,Rat

产品性能 (Performance)

偶联物 (Conjugation)Unconjugated修饰 (Modification)Unmodified

同种型 (Isotype) IgG

克隆 (Clonality) Polyclonal 形式 (Form) Liquid

Store at 4°C short term. Aliquot and store at -20°C long term. Avoid 存放说明 (Storage)

freeze/thaw cycles.

Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% 储存溶液 (Buffer)

New type preservative N.

纯化方式 (Purification) Affinity purification

免疫原信息 (Immunogen)

基因名 (Gene Name) ARSA

别名 (Alternative Names) ARSA; Arylsulfatase A; ASA; Cerebroside-sulfatase

基因 ID (Gene ID) 410.0

P15289.The antiserum was produced against synthesized peptide derived 蛋白ID (SwissProt ID)

from human ARSA. AA range:251-300

产品应用(Application)

稀释比 (Dilution Ratio) WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:50-1:200,ELISA 1:10000-1:20000

蛋白分子量 (Molecular Weight) 54kDa

研究背景 (Background)

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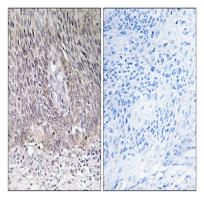


The protein encoded by this gene hydrolyzes cerebroside sulfate to cerebroside and sulfate. Defects in this gene lead to metachromatic leucodystrophy (MLD), a progressive demyelination disease which results in a variety of neurological symptoms and ultimately death. Alternatively spliced transcript variants 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).,

研究领域 (Research Area)

Sphingolipid metabolism;Lysosome;

图片 (Image Data)



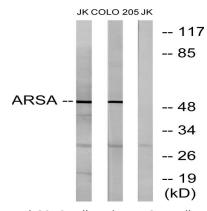
Immunohistochemistry analysis of paraffin-embedded human cervix carcinoma tissue, using ARSA Antibody. The picture on the right is blocked with the synthesized peptide.

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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.

注意事项 (Note)

For research use only.

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