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Recombinant Human Arylsulfatase A / ARSA Protein (His tag)

产品货号: TP08068

第三版

别名:MLD

描述: Arylsulfatase A (ARSA) is synthesized as a 52KDa lysosomal enzyme. It is a member of the sulfatase family that is required for the lysosomal degradation of cerebroside-3-sulfate, a sphingolipid sulfate ester and a major constituent of the myelin sheet. Arylsulfatase A is activated by a required co- or posttranslational modification with the oxidation of cysteine to formylglycine. Metachromatic leukodystrophy (MLD) is a lysosomal storage disease in the central and peripheral nervous systems with severe and progressive neurological symptoms caused by the deficiency of Arylsulfatase A. Deficiency of this enzyme is also found in apparently healthy individuals, a condition for which the term pseudodeficiency is introduced. ARSA forms dimers after receiving three N-linked oligosaccharides in the endoplasmic reticulum, and then the dimers are transported to the Golgi where they receive mannose 6-phosphate recognition markers. And thus, ARSA is transported and delivered to dense lysosomes in a mannose 6-phosphate receptor-dependent manner. It has been shown that within the lysosomes, the ARSA dimers can oligomerize to an octamer in a pH-dependent manner. The ARSA deficiency leads to metachromatic leucodystrophy (MLD), a lysosomal storage disorder associated with severe and progressive demyelination in he central and peripheral nervous system. Additionally, the serum level of arylsulfatase A might be helpful in diagnosis of lung and central nervous system cancer.

配方:PBS

分子量:53 kDa

序列: Met 1-Ala 507

纯度:> 95% by HPLC

浓度:

内毒素:<1.0 EU per 1 ug of protein (determined by LAL method)

存储: +4 ° C 保存 (1-2 周). 长期保存在-20 ° C或者-70 ° C. 避免反复冻融.