半乳糖转移酶 7 亚基β1,4 抗体

产品货号： mlR9728

英文名称： B4GALT7

中文名称： 半乳糖转移酶 7 亚基β1,4 抗体

别　　名： B4GAL T7; Beta 1,4 galactosyltransferase 7; Beta 1,4 GalTase 7; Beta4Gal T7; UDP Gal:beta GlcNAc beta 1,4 galactosyltransferase 7; XGALT 1; XGALT1; XGPT1; Xylosylprotein beta 1,4 galactosyltransferase, polypeptide 7; B4GT7_HUMAN.

研究领域： 细胞生物　免疫学　信号转导　细胞周期蛋白　细胞分化　细胞骨架　细胞外基质

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Chicken, Dog, Pig, Cow, Horse, Rabbit, Sheep,

产品应用： WB=1:500-2000 ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 IF=1:50-200 （石蜡切片需做抗原修复）

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

分 子 量： 37kDa

细胞定位： 细胞浆 细胞膜

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human B4GALT7:201-300/327

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： 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 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 ° C.

PubMed： PubMed

产品介绍 background :

β-1,4-galactosyltransferases (β-1,4-Gal-T) are type II membrane-bound glycoproteins that are substrate-specific and function to transfer galactose in a β-1,4 linkage to an acceptor sugar. There are seven members of the β-1,4-Gal-T family, all of which are directed to the golgi apparatus through a hydrophobic sequence at the N-terminus. β-1,4-Gal-T7, also known as B4GALT7 or XGALT1, is a 327 amino acid single-pass type II membrane protein that is expressed at high levels in heart, pancreas and liver. β-1,4-Gal-T7 uses manganese to catalyze the UDP-dependent biosynthesis of glycosphingolipids. The gene encoding β-1,4-Gal-T7 is mutated in Ehlers-Danlos syndrome progeroid type (EDSP), a variant form of Ehlers-Danlos syndrome characterized by progeroid facies, mild mental retardation, short stature, skin hyperextensibility, moderate skin fragility, joint hypermobility principally in digits.β-1,4-galactosyltransferases (β-1,4-Gal-T) are type II membrane-bound glycoproteins that are substrate-specific and function to transfer galactose in a ∫-1,4 linkage to an acceptor sugar. There are seven members of the β-1,4-Gal-T family, all of which are directed to the golgi apparatus through a hydrophobic sequence at the N-terminus. β-1,4-Gal-T7, also known as B4GALT7 or XGALT1, is a 327 amino acid single-pass type II membrane protein that is expressed at high levels in heart, pancreas and liver. β-1,4-Gal-T7 uses manganese to catalyze the UDP-dependent biosynthesis of glycosphingolipids. The gene encoding β-1,4-Gal-T7 is mutated in Ehlers-Danlos syndrome progeroid type (EDSP), a variant form of Ehlers-Danlos syndrome characterized by progeroid facies, mild mental retardation, short stature, skin hyperextensibility, moderate skin fragility, joint hypermobility principally in digits.-1,4-galactosyltransferases (β-1,4-Gal-T) are type II membrane-bound glycoproteins that are substrate-specific and function to transfer galactose in a ∫-1,4 linkage to an acceptor sugar. There are seven members of the β-1,4-Gal-T family, all of which are directed to the golgi apparatus through a hydrophobic sequence at the N-terminus. β-1,4-Gal-T7, also known as B4GALT7 or XGALT1, is a 327 amino acid single-pass type II membrane protein that is expressed at high levels in heart, pancreas and liver. β-1,4-Gal-T7 uses manganese to catalyze the UDP-dependent biosynthesis of glycosphingolipids. The gene encoding β-1,4-Gal-T7 is mutated in Ehlers-Danlos syndrome progeroid type (EDSP), a variant form of Ehlers-Danlos syndrome characterized by progeroid facies, mild mental retardation, short stature, skin hyperextensibility, moderate skin fragility, joint hypermobility principally in digits.

Function:

Required for the biosynthesis of the tetrasaccharide linkage region of proteoglycans, especially for small proteoglycans in skin fibroblasts.

Subcellular Location:

Golgi apparatus, Golgi stack membrane; Single-pass type II membrane protein. Note: Cis cisternae of Golgi stack

Tissue Specificity:

High expression in heart, pancreas and liver, medium in placenta and kidney, low in brain, skeletal muscle and lung.

DISEASE:

Defects in B4GALT7 are the cause of Ehlers-Danlos syndrome progeroid type (EDSP) [MIM:130070]. EDSP is a variant form of Ehlers-Danlos syndrome characterized by progeroid facies, mild mental retardation, short stature, skin hyperextensibility, moderate skin fragility, joint hypermobility principally in digits.

Similarity:

Belongs to the glycosyltransferase 7 family.

SWISS:

Q9UBV7

Gene ID:

11285

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.