FUCA1 (14-J2): sc-100530



The Power to Question

BACKGROUND

FUCA1 (fucosidase, α -L-1, tissue) is a 466 amino acid membrane- and seminal-associated isozyme that is a member of the glycosyl hydrolase 29 family. FUCA1 functions as a homotetramer and is responsible for hydrolyzing and reducing the carbohydrate moieties of glycoproteins in various tissues. Defects in the gene encoding FUCA1 result in fucosidosis, an autosomal recessive disorder caused by an accumulation of fucose-containing glycolipids and glycoproteins. Fucosidosis, a lysosomal storage disease, is characterized by neurologic deterioration, growth retardation, visceromegaly and seizures. Early onset of fucosidosis causes coarse facial features, angiokeratoma corporis diffusum, spasticity, delayed psychomotor development and an unusual spondylometaphyseoepiphyseal dysplasia.

REFERENCES

- Seo, H.C., Willems, P.J. and O'Brien, J.S. 1993. Six additional mutations in fucosidosis: three nonsense mutations and three frameshift mutations. Hum. Mol. Genet. 2: 1205-1208.
- Takeshita, H., Yasuda, T., Nadano, D., Iida, R., Nakanaga, M., Tenjo, E., Sawazaki, K. and Kishi, K. 1994. Genetically polymorphic α-L-fucosidase (FUCA1) isozymes detected in blood plasma. Hum. Genet. 94: 224-230.
- Cragg, H., Williamson, M., Young, E., O'Brien, J., Alhadeff, J., Fang-Kircher, S., Paschke, E. and Winchester, B. 1997. Fucosidosis: genetic and biochemical analysis of eight cases. J. Med. Genet. 34: 105-110.
- 4. Ip, P., Goh, W., Chan, K.W. and Cheung, P.T. 2002. A novel FUCA1 mutation causing fucosidosis in a Chinese boy. J. Inherit. Metab. Dis. 25: 415-416.
- 5. Khunsook, S., Alhadeff, J.A. and Bean, B.S. 2002. Purification and characterization of human seminal plasma α -L-fucosidase. Mol. Hum. Reprod. 8: 221-227.
- Intra, J., Cenni, F. and Perotti, M.E. 2006. An a-L-fucosidase potentially involved in fertilization is present on *Drosophila* spermatozoa surface. Mol. Reprod. Dev. 73: 1149-1158.
- 7. Li, C., Qian, J. and Lin, J.S. 2006. Purification and characterization of α -L-fucosidase from human primary hepatocarcinoma tissue. World J. Gastroenterol. 12: 3770-3775.
- 8. Lin, S.P., Chang, J.H., de la Cadena, M.P., Chang, T.F. and Lee-Chen, G.J. 2007. Mutation identification and characterization of a Taiwanese patient with fucosidosis. J. Hum. Genet. 52: 553-556.
- Venditti, J.J., Donigan, K.A. and Bean, B.S. 2007. Crypticity and functional distribution of the membrane associated α-L-fucosidase of human sperm. Mol. Reprod. Dev. 74: 758-766.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.

CHROMOSOMAL LOCATION

Genetic locus: FUCA1 (human) mapping to 1p36.11.

SOURCE

FUCA1 (14-J2) is a mouse monoclonal antibody raised against recombinant FUCA1 of human origin.

PRODUCT

Each vial contains 100 μg lgG_{2b} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

FUCA1 (14-J2) is recommended for detection of FUCA1 of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for FUCA1 siRNA (h): sc-78583, FUCA1 shRNA Plasmid (h): sc-78583-SH and FUCA1 shRNA (h) Lentiviral Particles: sc-78583-V.

Molecular Weight of FUCA1: 56 kDa.

Positive Controls: K-562 whole cell lysate: sc-2203.

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-lgG κ BP-HRP: sc-516102 or m-lgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz MarkerTM Molecular Weight Standards: sc-2035, UltraCruz[®] Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).

RESEARCH USE

For research use only, not for use in diagnostic procedures.

Santa Cruz Biotechnology, Inc. 1.800.457.3801 831.457.3801 fax 831.457.3801 Europe +00800 4573 8000 49 6221 4503 0 www.scbt.com