SZABO SCANDIC

Part of Europa Biosite

Produktinformation

- Forschungsprodukte & Biochemikalien
- Zellkultur & Verbrauchsmaterial
- Diagnostik & molekulare Diagnostik
- Laborgeräte & Service

Weitere Information auf den folgenden Seiten!
See the following pages for more information!

Lieferung & Zahlungsart

siehe unsere Liefer- und Versandbedingungen

Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

SZABO-SCANDIC HandelsgmbH

Quellenstraße 110, A-1100 Wien

T. +43(0)1 489 3961-0

F. +43(0)1 489 3961-7

mail@szabo-scandic.com

www.szabo-scandic.com

linkedin.com/company/szaboscandic

Nordic MUbio

Mouse anti alpha-Glucosidase

Catalogue number: **MUB0707P**

Clone	43G7
Isotype	IgG1
Product Type	Primary Antibodies
Units	0.1 mg
Host	Mouse (Balb/c)
Species reactivity	Human
Application	Immunoblotting Immunohistochemistry (paraffin)

Distributors

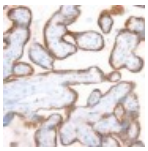
For Purchasing Information, please contact your local distributor

Find Distributor

Background

Lysosomal α -glucosidase, like all other lysosomal enzymes of which the 'life-cycle' has been studied, is synthesized as a large precursor that is processed to mature forms of lower molecular mass. Acid α -glucosidase Catalyzes the hydrolysis of the α1 -> 4 and α1 -> 6 glucosidic linkages in glycogen and the α1 -> 4 glucosidic linkage in maltose and artificial substRates likep-nitrophenyl- α -glucoside. The enzyme is deficient in patients with Glycogenosis Type II (Pompe's disease). Pompe disease (also called Glycogen storage disease type II (GSD II) or acid maltase deficiency) is an autosomal recessive metabolic disorder which damages muscle and nerve cells throughout the body. It is caused by an accumulation of glycogen in the lysosome due to deficiency of the lysosomal acid alpha-glucosidase enzyme. It is the only glycogen storage disease with a defect in lysosomal metabolism. The build-up of glycogen causes progressive muscle weakness (myopathy) throughout the body and affects various body tissues, particularly in the heart, skeletal muscles, liver and nervous system. There are exceptions but levels of alpha-glucosidase determines the type of GSD II an individual may have. More alpha glucosidase present in the individuals muscles means symptoms occur later in life and progress more slowly. GSD II is broadly divided into two

Figure 1 Immunohistochemistry on paraffin-embedded sections of human placenta.



onset forms based on the age symptoms occur: Infantile-onset form is usually diagnosed at 4-8 months; muscles appear normal but are limp and weak preventing them from lifting their head or rolling over. As the disease progresses heart muscles thicken and progressively fail. Without treatment death usually occurs due to heart failure and respiRatory weakness.Late/later onset form occurs later than one to two years and progresses more slowly than Infantile-onset form. One of the first symptoms is a progressive decrease in muscle strength starting with the legs and moving to smaller muscles in the trunk and arms, such as the diaphragm and other muscles required for breathing. RespiRatory failure is the most common cause of death. Enlargement of the heart muscles and rhythm disturbances are not significant features but do occur in some casesThe disease is caused by a mutation in a gene (acid alpha-glucosidase: also known as acid maltase) on long arm of chromosome 17 at 17q25.2-q25.3 (base pair 75,689,876 to 75,708,272). The number of mutations described is currently (in 2010) 289 with 67 being non-pathogenic mutations and 197 pathogenic mutations. The remainder are still being evaluated for their association with disease.

Source
43G7 is a Mouse monoclonal IgG1 antibody derived by fusion of Mouse myeloma cells with spleen cells from a Balb/cHeA Mouse hyperimmunized with purified acid alpha-glucosidase from Human placenta.

Product
Each vial contains 100 ul 1 mg/ml purified monoclonal antibody in PBS containing 0.09% sodium azide.

Applications
43G7 is useful for paraffin-embedded tissues and Western blots. Optimal antibody dilution should be determined by titration.

Specificity
Reacts with the two major bands with apparent molecular weights of 76 000 and 70 000, the band with a molecular weight of about 94000 and minor bands with apparent molecular weights of less than 67 000, when analyzing a-glucosidase isolated from placenta by polyacrylamide gel electrophoresis in the presence of SDS and a reducing agent.

Storage

Store at 4°C, or in small aliquots at -20°C.

References

1. John Hilkens, Joseph M. Tager, Femke Buijs, Betty Brouwer-Kelder, Gerda M. Van Thienen, Frans P.W. Tegelaers and Jo Hilgers (1981): Monoclonal antibodies anti Human acid t~-glucosidase, Biochimica et Biophysica Acta, 678 (1981) 7-11.

Caution

This product is intended FOR RESEARCH USE ONLY, and FOR TESTS IN VITRO, not for use in diagnostic or therapeutic procedures involving humans or animals. This product contains sodium azide. To prevent formation of toxic vapors, do not mix with strong acidic solutions. To prevent formation of potentially explosive metallic azides in metal plumbing, always wash into drain with copious quantities of water. This datasheet is as accurate as reasonably achievable, but Nordic-MUbio accepts no liability for any inaccuracies or omissions in this information.