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Lieferung & Zahlungsart

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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](https://www.linkedin.com/company/szaboscandic) 

Mpi (m): 293T Lysate: sc-121729

BACKGROUND

Mpi (mannose phosphate isomerase), also known as PMI (phosphomannose isomerase) or PMI1, is a 423 amino acid zinc metalloenzyme belonging to the mannose-6-phosphate isomerase type 1 family, and is expressed in all tissues, more abundantly in heart, brain and skeletal muscle. A steady supply of D-mannose derivatives, which are required for most glycosylation reactions, is maintained by Mpi. Localized to the cytoplasm, Mpi utilizes zinc as a cofactor and catalyzes the interconversion of fructose-6-phosphate and mannose-6-phosphate. Mutations in the gene encoding Mpi lead to congenital disorder of glycosylation type 1B (CDG1B), also designated carbohydrate-deficient glycoprotein syndrome type 1b (CDGS1B), which is characterized by protein-losing enteropathy. Congenital disorders of glycosylation are metabolic deficiencies in glycoprotein biosynthesis that usually results in severe mental and psychomotor retardation.

REFERENCES

1. Proudfoot, A.E., et al. 1996. *In vivo* and *in vitro* folding of a recombinant metalloenzyme, phosphomannose isomerase. *Biochem. J.* 318: 437-442.
2. Jaeken, J., et al. 1998. Phosphomannose isomerase deficiency: a carbohydrate-deficient glycoprotein syndrome with hepatic-intestinal presentation. *Am. J. Hum. Genet.* 62: 1535-1539.
3. Nihues, R., et al. 1998. Carbohydrate-deficient glycoprotein syndrome type 1b. Phosphomannose isomerase deficiency and mannose therapy. *J. Clin. Invest.* 101: 1414-1420.
4. Schollen, E., et al. 2000. Genomic organization of the human phosphomannose isomerase (MPI) gene and mutation analysis in patients with congenital disorders of glycosylation type 1b (CDG-1b). *Hum. Mutat.* 16: 247-252.
5. de Lonlay, P., et al. 2001. A broad spectrum of clinical presentations in congenital disorders of glycosylation I: a series of 26 cases. *J. Med. Genet.* 38: 14-19.
6. Schollen, E., et al. 2002. DHPLC analysis as a platform for molecular diagnosis of congenital disorders of glycosylation (CDG). *Eur. J. Hum. Genet.* 10: 643-648.
7. Vuillaumier-Barrot, S., et al. 2002. Protein losing enteropathy-hepatic fibrosis syndrome in Saguenay-Lac St-Jean, Quebec is a congenital disorder of glycosylation type 1b. *J. Med. Genet.* 39: 849-851.
8. de Lonlay, P. and Seta, N. 2009. The clinical spectrum of phosphomannose isomerase deficiency, with an evaluation of mannose treatment for CDG-1b. *Biochim. Biophys. Acta* 1792: 841-843.
9. Higashidani, A., et al. 2009. Exogenous mannose does not raise steady state mannose-6-phosphate pools of normal or N-glycosylation-deficient human fibroblasts. *Mol. Genet. Metab.* 96: 268-272.

CHROMOSOMAL LOCATION

Genetic locus: Mpi (mouse) mapping to 9 B.

PRODUCT

Mpi (m): 293T Lysate represents a lysate of mouse Mpi transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

Mpi (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive Mpi antibodies. Recommended use: 10-20 µl per lane.

Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

RESEARCH USE

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

PROTOCOLS

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