Format: Abstract

Biochim Biophys Acta. 2017 Jul 12. pii: S0304-4165(17)30214-3. doi: 10.1016/j.bbagen.2017.06.024. [Epub ahead of print]

Glycosylation with ribitol-phosphate in mammals: New insights into the O-mannosyl glycan.

Manya H[1], Endo T[2].

Author information

Abstract

BACKGROUND: O-mannosyl glycans have been found in a limited number of glycoproteins of the brain, nerves, and skeletal muscles, particularly in α-dystroglycan (α-DG). Defects in O-mannosyl glycan on α-DG are the primary cause of a group of congenital muscular dystrophies, which are collectively termed α-dystroglycanopathy. Recent studies have revealed various O-mannosyl glycan structures, which can be classified as core M1, core M2, and core M3 glycans. Although many dystroglycanopathy genes are involved in core M3 processing, the structure and biosynthesis of core M3 glycan remains only partially understood.

SCOPE OF REVIEW: This review presents recent findings about the structure, biosynthesis, and pathology of O-mannosyl glycans.

MAJOR CONCLUSIONS: Recent studies have revealed that the entire structure of core M3 glycan, including ribitol-5-phosphate, is a novel structure in mammals; its unique biosynthetic pathway has been elucidated by the identification of new causative genes for α-dystroglycanopathies and their functions.

GENERAL SIGNIFICANCE: O-mannosyl glycan has a novel, unique structure that is important for the maintenance of brain and muscle functions. These findings have opened up a new field in glycoscience. These studies will further contribute to the understanding of the pathomechanism of α-dystroglycanopathy and the development of glycotherapeutics. This article is part of a Special Issue entitled Neuro-glycoscience, edited by Kenji Kadomatsu and Hiroshi Kitagawa.

KEYWORDS: Glycan biosynthesis; Glycosyltransferase; Muscular dystrophy; O-glycosylation; O-mannosyl glycan; Ribitol-5-phosphate; α-dystroglycanopathy

PMID: 28711406 DOI: 10.1016/j.bbagen.2017.06.024

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