**Glycopathology in muscular dystrophy**

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*O*-Mannosylation is an uncommon type of glycosylation in mammals but is important in muscle and brain development. We have identified and characterized glycosyltransferases, protein *O*-mannose β1,2-*N*-acetylglucosaminyltransferase (POMGnT1) and protein *O*-mannosyltransferase 1 (POMT1) and its homolog, POMT2 are involved in *O*-mannosyl glycan synthesis. Then POMGnT1 is found to be responsible for muscle-eye-brain disease (MEB) and POMT1/2 are for Walker-Warburg syndrome (WWS). MEB and WWS are congenital muscular dystrophies with brain malformation and structural eye abnormalities. Recent data suggest that aberrant glycosylation of α-dystroglycan is also the cause of other four muscular dystrophies. These all are named α-dystroglycanopathies because these are caused by incomplete-glycosylation of α-dystroglycan. Here I focus on protein *O*-mannosylation that is catalyzed by POMT1/2. Recently, we show that POMT1 forms a complex with POMT2, and the complex possesses protein *O*-mannosyltransferase activity, indicating that POMT1 and POMT2 associate physically and functionally *in vivo*. All mutations found in the *POMT1* gene of patients with WWS lead to great reduction of protein *O*-mannosyltransferase activity, although all POMT1 mutants coprecipitated with POMT2. These results indicate that the mutant POMT1s found in WWS patients could form hetero-complexes with POMT2 but that such complexes are insufficient for enzymatic activity. Possible regulatory mechanism of *O*-mannosylation will be discussed.