

2021 Lecture Series

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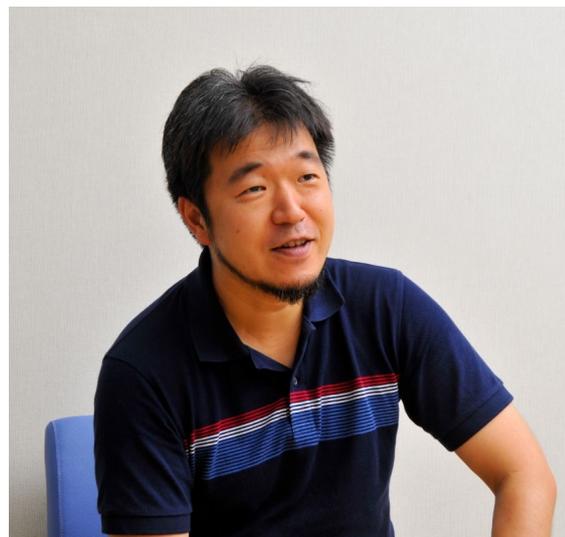
will present a seminar entitled

NGLY1 deficiency and non-lysosomal degradation of glycans -basic science encounters a human genetic disorder

Friday 11 June 2021, 11am

Institute for Glycomics
Lecture Theatre (G26 4.09)

(No food or drink allowed in the lecture theatre)



Abstract

Glycosylation is known to be one of the most prevalent co- and post-translational modification of proteins, affecting the physicochemical/physiological properties of cognate proteins. It is also known that glycans can also occur as a “free”, unconjugated form. Recent studies have clarified that there are multiple pathways involved in the formation of free oligosaccharides (1). For example, intracellular free N-glycans (FNGs), free oligosaccharides structurally related to asparagine-linked (N-linked) glycans, are mainly formed by hydrolysis of oligosaccharides in mammalian cells (2, 3), while they are predominantly formed by the action of the cytoplasmic peptide:N-glycanase (Png1) in budding yeast, *S. cerevisiae* (4). There are also “extracellular” free oligosaccharides found in various animal sera (5) and more recently, we discovered another type of free glycans, i.e. free O-glycans (FOGs) in budding yeast (6). These observations clearly indicate that the major formation mechanism of free glycans are quite distinct between organisms, and they are complicated by diverse formation pathways.

I will also present our most recent progress on developing a therapeutic treatment for NGLY1 deficiency, a rare human genetic disorder bearing mutations in *NGLY1* allele, an ortholog of *PNG1* in human (7-10).

References

1. Harada, Y. et al., *Cell Mol Life Sci* **72**, 2509-2533 (2015)
2. Harada, Y. et al., *J. Biol. Chem.* **288**, 32673-32684 (2013)
3. Harada, Y. et al., *Glycobiology* **25**, 1196-1205 (2015)
4. Hirayama, H., et al., *J. Biol. Chem.* **285**, 12390-12404 (2010)
5. Seino, J., et al., *Glycobiology* **26**, 1072-1085 (2016)
6. Hirayama, H., et al. *J. Biol. Chem.* **294**, 15900-15911 (2019)
7. Fujihira, H., et al. *PLoS Genet.* **13**, e1006696 (2017)
8. Fujihira, H., et al. *BBA Mol. Basis Dis.* **1866**, 165588 (2020)
9. Asahina, M., et al. *Hum. Mol. Genet.* **29**, 1635-1647 (2020)
10. Asahina, M., et al. *Proc. Jpn Acad Ser B* **97**, 89-102 (2021)

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