ASHBi SEMINAR

Neural organoids to model molecular defects of patients with accelerated ageing and neurodegeneration

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Date Thursday, 28 March 2024

Time 16:00 – 17:00 [JST]

Venue Conference Room Onsite Only*

B1F, Faculty of Medicine Bldg. B

*Register via the right QR code

Abstract

We study Cockayne syndrome, a rare genetic disease characterized by precocious ageing and neurodegeneration, the cause of which is generally attributed to a DNA repair defect. Our data point to mechanisms that imply oxidative and nitrosative stress and lead to mitochondrial dysfunction and transcriptional reprogramming. In the absence of reliable animal models, we have generated neural organoids by reprogramming cells of patients with diverse clinical severity, to investigate neurological defects in a context of accelerated ageing.

Crochemore C, *et al.* Epigenomic signature of accelerated ageing in progeroid Cockayne syndrome. *Aging Cell* (2023) 22(10): e13959

Crochemore C, *et al.* Reactive species in progeroid syndromes and aging-related processes". *Antioxidant and Redox Signaling.* (2022) 37 (1-3) 208-228.

Crochemore C, *et al.* CSB promoter downregulation via histone H3 hypoacetylation is an early determinant of replicative senescence. *Nature Communications.* (2019) 10(1): 5576.

Chatre L, *et al.* Reversal of mitochondrial defects with CSB-dependent serine-protease inhibitors in patient cells of the progeroid Cockayne syndrome. *Proc. Natl. Acad. Sci. U.S.A.* (2015) 112 (22): E2910-2919

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