

東京大学 グローバル COE 特別セミナー

東京大学大学院 理学系研究科 生物化学専攻

演者 : Catherine Florentz 博士

Université de Strasbourg

演題 : Unexpected structural and functional properties of human mitochondrial tRNAs and aminoacyl-tRNA synthetases. Relationships with neurodegenerative disorders.

日時 : 平成 23 年 3 月 2 日 (水) 14:00~15:00

場所 : 東京大学理学部 3 号館 3F 303 号室

Mitochondria are best known for their prominent role in cellular energetics. The genome of human mitochondria encodes for 13 proteins only, all sub-units of respiratory chain complexes and accordingly actors of the ATP synthesis process. The mitochondrial (mt) protein synthesis machinery is formed by RNAs encoded by the mt genome (2 rRNAs, 22 tRNAs) and all other factors encoded by the nuclear genome, and imported. These include a full set of aminoacyl-tRNA synthetases. Over more than 20 years, numerous point mutations in mt-tRNA genes have been correlated to a variety of neuromuscular and neurodegenerative disorders, and more recently, mutations in nuclear genes for translation factors have also been revealed as causative of disorders, leading to an enlarged family of “mitochondrial translation disorders”.

Recent progress on fundamental knowledge on structure/function relationships in mammalian mt aminoacylation systems will be presented. A comparative analysis with bacterial counterparts will highlight unexpected specific properties of mt systems. Molecular impacts of pathology-related mutations in both tRNA and synthetases will be discussed.

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