Day 1 – November 14, 2025

8:45 - 9:00

Opening Remarks
Chair: Hitoshi Osaka (Department of Pediatrics, Jichi Medical University)

9:00 - 10:00

Symposium 1 – Frontiers in Mitochondrial Dynamics and Organelle Interactions
Chairs:
- Naotada Ishihara (Department of Biological Sciences, Graduate School of Science, Osaka University)
- Koji Okamoto (Laboratory of Mitochondrial Dynamics, Graduate School of Frontier Biosciences, Osaka University)

S1-1 **Mitochondrial Dynamics Coordinate Cytoskeletal Remodeling and the Myelin Gene Program in Oligodendrocytes**
Keiko Iwata
Wakayama Medical University

S1-2 **Analysis of the role of ER–Mitochondria contact sites in the degradation of mislocalized proteins**
Shinya Tashiro
Yamagata University

S1-3 **Artificial polypeptides inspired by transmembrane mitochondrial protein for escorting exogenous DNA into the mitochondria**
Keiji Numata
Kyoto University; RIKEN

S1-4 **A Simple Method for Detecting Protein Aggregates in the Mitochondrial Matrix of Vertebrate Cultured Cells**
Yuichi Matsushima
Department of Biological Sciences, Osaka University, [2]Department of Clinical Chemistry and Laboratory Medicine, Kyushu University

10:10 - 11:10

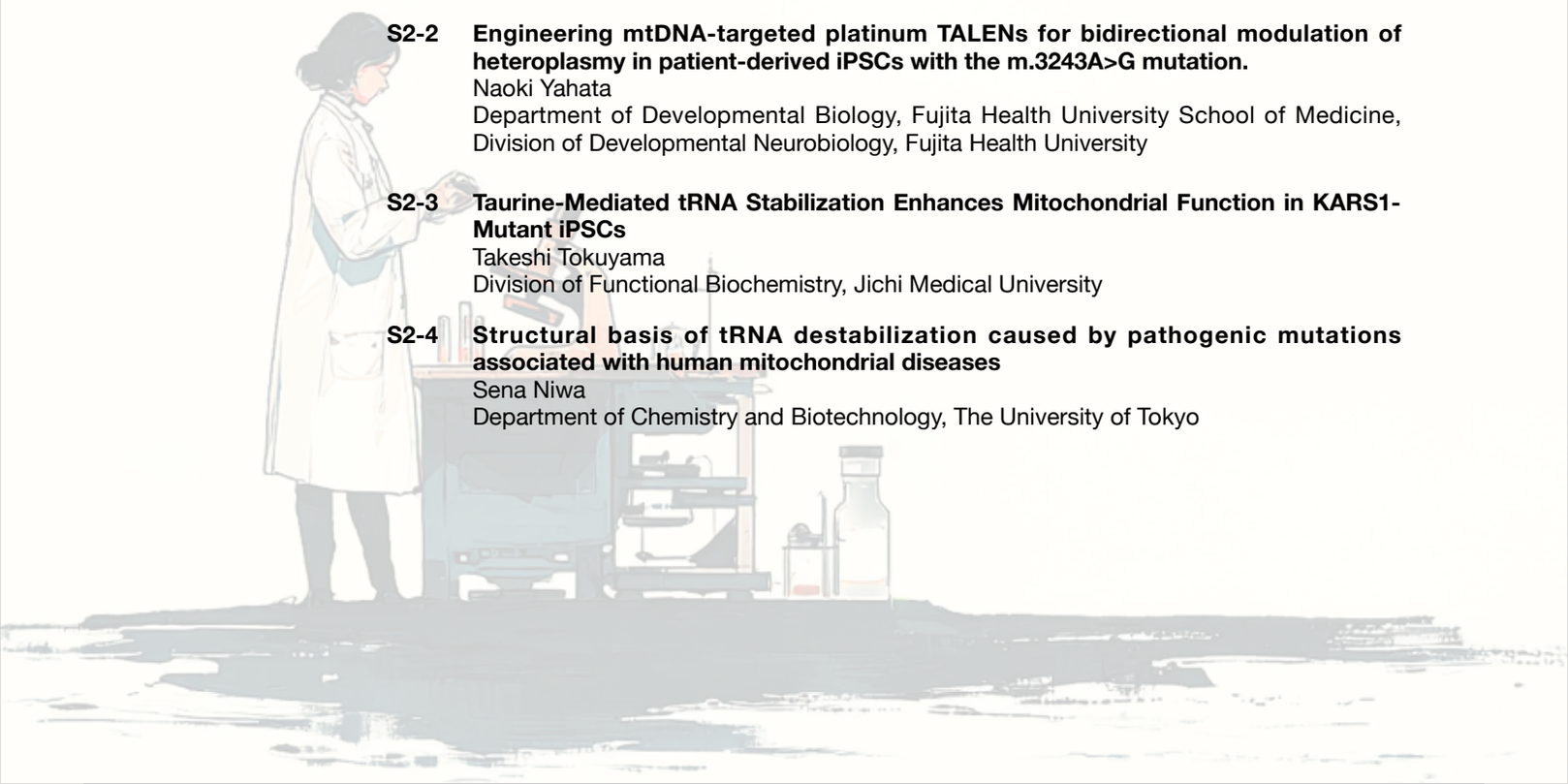
Symposium 2 – Frontiers in Mitochondrial tRNA Metabolism Regulation and Disease Mechanisms
Chairs:
- Fanyan Wei (Department of Modomics Biology and Medicine, Institute of Development, Aging and Cancer (IDAC), Tohoku University)
- Hideki Uosaki (Division of Functional Biochemistry, Department of Biochemistry, JICHI Medical University)

S2-1 **Mitochondrial DNA base editing approaches for generating mouse models**
Haruna Tani
Institute of Development, Aging and Cancer, Tohoku University

S2-2 **Engineering mtDNA-targeted platinum TALENs for bidirectional modulation of heteroplasmy in patient-derived iPSCs with the m.3243A>G mutation.**
Naoki Yahata
Department of Developmental Biology, Fujita Health University School of Medicine, Division of Developmental Neurobiology, Fujita Health University

S2-3 **Taurine-Mediated tRNA Stabilization Enhances Mitochondrial Function in KARS1-Mutant iPSCs**
Takeshi Tokuyama
Division of Functional Biochemistry, Jichi Medical University

S2-4 **Structural basis of tRNA destabilization caused by pathogenic mutations associated with human mitochondrial diseases**
Sena Niwa
Department of Chemistry and Biotechnology, The University of Tokyo



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11:20 - 12:20

Oral Session 1
1 Basic Metabolism & Regulation & Disease Mechanisms & Experimental Models
Chairs:
- Ayumu Sugiura (Graduate school of medicine, Juntendo University)
- Yuhei Araiso (Dept. of Clin. Lab. Sci., Div. of Health Sci., Kanazawa Univ)

O-1 **MICOS/MIB complex conversion contributes to increased cristae density during heart development**
Yuto Ishikawa
Gakushuin University

O-2 **The Effect of Subcellular Localization on AMPK-mediated Phosphorylation of Mff**
Masaki Nakahashi
Department of Biological Sciences, Osaka University

O-3 **Mitochondrial transcription exhibits diverse dynamics depending on the state of the cell.**
Shigeru Matsuda
Dept. Modomics Biology & Medicine IDAC, Tohoku university

O-4 **Novel insights into the regulatory mechanism of energy metabolism by mitochondrial protease ClpXP**
Ko Suzuki
Department of Molecular Biochemistry, Iwate Medical University

O-5 **Relation of mtDNA copy number with cell senescence or cellular organelle**
Katsumi Kasashima
Dep. of Biochemistry, Jichi Medical University

12:30 - 13:30

Luncheon Seminar 1
(Co-hosted by Astellas Pharma Inc., Innovation for NEW HOPE)

Toward a Society Where Cutting-Edge Therapies Reach Patients

Chair: Akira Ohtake (Dept. Clin. Genomics & Pediatr., Saitama Med. Univ. Hosp.)

- Advocacy Department, Astellas Pharma Inc. (10 min)
- Hitoshi Osaka, Department of Pediatrics, Jichi Medical University (25 min)
- Student collaborators of Innovation for NEW HOPE (15 min)

13:40 - 14:40

Oral Session 2
1 Genetic Variants & Pathogenesis & Drug Discovery & Therapeutic Strategies
Chairs:
- Atsuko Okazaki (Department of Diagnostics and Therapeutics of Intractable Diseases, Juntendo University)
- Yoshihito Kishita (Department of Life Science, Kindai University)

O-6 **Deep Intronic Variant-Induced Pseudoexon Inclusion in Undiagnosed Mitochondrial Disease and Therapeutic Rescue by Splicing-Switching ASOs**
Yukiko Yatsuka
Diagnostics and Therapeutics of Intractable Diseases, Juntendo University

O-7 **Leigh Syndrome with Unilateral Brain Lesions at Disease Onset**
Ying Zou
Department of Neurology, Capital Medical University, Beijing, China.

O-8 **Non-coding Spliceosomal snRNA Mutations as a Novel Cause of Mitochondrial Disease: Insights from Systematic WGS Re-analysis**
Kohta Nakamura
Diagnostics and Therapeutics of Intractable Diseases, Juntendo University

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O-9 **Renal Involvement in RRM2B-related Mitochondrial DNA Depletion Syndrome: Review in Japan**
Hisato Aihara
Department of Pediatrics, Juntendo University

O-10 **A Pediatric Case of Homocystinuria Type III with Early Clinical Features Resembling Leigh Encephalopathy**
Tatsunori Itabashi
Department of Pediatrics, Hokkaido University

14:50 - 15:50 **Symposium 3 – New Developments in Diagnosis and Treatment of Pediatric Mitochondrial Diseases**
Chairs:
- Kei Murayama (Diagnostics and Therapeutics of Intractable Diseases, Intractable Disease Research Center, Graduate School of Medicine, Juntendo University)
- Masakazu Mimaki (Department of Pediatrics, Teikyo University School of Medicine)

S3-1 **Clinical Index of Neonatal-onset Mitochondrial Disease**
Taro Nagatomo
Japanese Red Cross Fukuoka Hospital

S3-2 **Leigh syndrome: a well-recognized yet genetically heterogeneous childhood-onset mitochondrial disease**
Erika Ogawa
Pediatrics Department, Tokyo Metropolitan Hiroo Hospital

S3-3 **Nutritional Management in Mitochondrial Disease -Tailoring dietary approaches to address specific metabolic defects-**
Shunsaku Kaji
Department of Pediatrics, Tsuyama Chuo Hospital

S3-4 **Clinical Practice for Hearing Loss in Pediatric Mitochondrial Disease**
Tatsuo Matsunaga
National Institute of Sensory Organs/Medical Genetics Center, National Hospital Organization Tokyo Medical Center

15:50 – 16:50 **Special Lecture**

The Journey from Basic Science to Pharmaceutical Approval in Mitochondrial Disease

Speaker: Shigeo Ohta (Dept. Neurol. Med., Grad. Sch. Med., Juntendo Univ.; Adv. Inst., Nippon Med. Univ.)

16:50 – 18:00 **Evening Seminar**
(Co-hosted by Koinobori Association)

Challenges Emerging from Patients' and Families' Voices: Insights from the Frontline of Mitochondrial Drug Discovery
Chair: Yuichi Goto (Advisor, Koinobori; Special Assistant to the President, NCNP)

Lectures:
- Masashi Suganuma (Representative Director, Koinobori; Chairman, LUCA Science; Director, Suganuma Clinic)
- Taro Inaba (Director, Koinobori; Managing Partner, Remiges Ventures)

Closing Remarks & Presentation of Appreciation
Hitoshi Osaka (Department of Pediatrics, Jichi Medical University)

18:00 – 20:00 **Reception**
4th Floor, Banquet Hall