

**Day 1; Thursday, 17<sup>th</sup> December**      1      12      17

12:45 Opening Remarks

13:00 Oral Session 1: Mitochondrial DNA Translation, Transcription, and Replication

1      DNA

**O1. Functional dissection of ribosome recycling factor in mammalian mitochondria**

Hiroyuki Morita, Yusuke Nozaki, Masafumi Tsuboi, Takuya Ueda, and Nono Takeuchi  
Graduate School of Frontier Sciences, University of Tokyo

**O2. The knockdown of ERAL1 is involved in assembly of mitochondria ribosome**

Takeshi Uchiumi and Dongchon Kang  
Department of Clinical Chemistry and Laboratory Medicine, Graduated School of Medical Sciences, Kyushu University  
**ERAL1**

**O3. Defect of a novel F-box protein, MUS-10, shows abnormal mitochondrial morphology and short lifespan in *Neurospora crassa***

Kiminori Kurashima, Akihiro Kato, Satoshi Sawada, Shin Hatakeyama, Michael Chae, Shuuitsu Tanaka, and Hirokazu Inoue  
Laboratory of Genetics, Department of Regulatory Biology, Faculty of Science, Saitama University

**F-box**      **MUS-10**

Michael Chae

**O4. Genome wide screen of *Drosophila* dsRNA library for genes involved in mitochondrial DNA maintenance – A study on a candidate gene, DmTTF**

Atsushi Fukuoh<sup>1</sup>, Preet Joers<sup>1</sup>, Susanna Valanne<sup>1</sup>, Mika Rämetsä<sup>1</sup>, Palmiro Cantatore<sup>2</sup>, and Howard T. Jacobs<sup>1</sup>  
<sup>1</sup>Institute of Medical Technology, University of Tampere, Finland  
<sup>2</sup>Dipartimento di Biochimica e Biologia Molecolare “Ernesto Quagliariello”, Università degli Studi di Bari, Italy

**O5. Functional analysis of mitochondrial matrix protease Lon in *Drosophila* Schneider cells**

Yuichi Matsushima<sup>1,2</sup> and Laurie S. Kaguni<sup>1</sup>  
<sup>1</sup>Department of Biochemistry and Molecular Biology, and Center for Mitochondrial Science and Medicine, Michigan State University  
<sup>2</sup>Department of Mental Retardation and Birth Defect Research, National Institute of Neuroscience, NCNP

**Lon**      **mtDNA**  
<sup>1,2</sup> Laurie S. Kaguni<sup>1</sup>

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**O6. Induction of recombination-mediated mitochondrial DNA replication**

Feng Ling<sup>1,2</sup>, Akiko Hori<sup>1,2</sup>, Niu Rong<sup>1,2</sup>, Minoru Yoshida<sup>1,2</sup>, and Takehiko Shibata<sup>3</sup>  
<sup>1</sup>Chem. Genet. Lab., RIKEN  
<sup>2</sup>Chem. Genomics Res. Group, RIKEN Adv. Sci. Inst.  
<sup>3</sup>Distinguished Senior Scientist Lab., RIKEN Adv. Sci. Inst.

<b>ROS</b>						<b>DNA</b>
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**O7. New evidence confirms generation of the mitochondrial bottleneck without the reduction of mtDNA content in early primordial germ cells of mice**

Hiroshi Shitara<sup>1</sup>, Liqin Cao<sup>1,2</sup>, Michihiko Sugimoto<sup>2</sup>, Jun-Ichi Hayashi<sup>3</sup>, Kuniya Abe<sup>2</sup>, and Hiromichi Yonekawa<sup>1</sup>

<sup>1</sup> *The Tokyo Metropolitan Institute of Medical Science (Rinshoken)*

<sup>2</sup> *BioResource Center (BRC) RIKEN Tsukuba Institute*

<sup>3</sup> *University of Tsukuba*

		<b>mtDNA</b>				
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**14:45 Coffee Break**

**15:15 Oral Session 2: Mitochondrial Oxidative Stress**

**2**

**O8. Mitochondrial superoxide anion (O<sub>2</sub><sup>-</sup>) overproduction causes low birthrate and low birth weight in *Tet-mev-1* mice with SDHC V69E**

Takamasa Ishii<sup>1</sup>, Masaki Miyazawa<sup>1</sup>, Kayo Yasuda<sup>1</sup>, Philip S. Hartman<sup>2</sup>, and Naoaki Ishii<sup>1</sup>

<sup>1</sup> *Department of Molecular Life Science, Tokai University School of Medicine*

<sup>2</sup> *Department of Biology, Texas Christian University*

**SDHC V69E**

	1	1	1	Philip S. Hartman <sup>2</sup>	1
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<sup>2</sup> *Department of Biology, Texas Christian University*

**O9. Reactive oxygen species production from the mitochondrial complex II with fumarate reductase activity**

Madhavi P. Paranagama, Kimitoshi Sakamoto, Hisako Amino, Chika Sakai, and Kiyoshi Kita

*Department of Biomedical Chemistry, Graduate School of Medicine, The University of Tokyo*

**O10. Observation of mitochondrial membrane permeability in cells under oxidative stress**

Yoshihiro Ohta, Yoshihiro Matsunomoto, Xiaolei Shi, and Chisako Fujita

*Department of Biotechnology and Life Science, Tokyo University of Agriculture and Technology*

**O11. NSAIDs and acidic environment induce gastric mucosal cellular mitochondrial dysfunction**

Hiroshi Matsui, Tsuyoshi Kaneko, Yumiko N. Nagano, Osamu Shimokawa, Akira Hirayama, and Ichinosuke Hyodo

*Graduate School of Comprehensive Human Sciences, University of Tsukuba*

**NSAID**

**O12. Radiation-induced delayed oxidative stress coupled with mitochondrial morphological change in normal human diploid cells**

Shinko Kobashigawa, Keiji Suzuki, and Shunichi Yamashita

*Life Sciences and Radiation Research, Graduate School of Biomedical Sciences*

**O13. Intervention of mitochondria in radiation carcinogenesis**

Masami Watanabe, Kimiko Watanabe, Genro Kashino, and Keizo Tano

*Laboratory of Radiation Biology, Department of Radiation Life Science, Research Reactor Institute, Kyoto University*

**O14. “Mito cell” move itself**

Tomohiro Nakayama<sup>1,3</sup>, Kazutoshi Nakano<sup>2,3</sup>, and Makiko Osawa<sup>3</sup>

<sup>1</sup>*Matsudo Clinic*

<sup>2</sup>*Nakano Children’s Clinic*

<sup>3</sup>*Department of Pediatrics, Tokyo Women’s Medical University*

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Day 2; Friday, 18<sup>th</sup> December 2 12 18

9:00 Oral Session 3: Mitochondrial Physiology and Dynamics

3

**O15. Physiological role of mitochondrial permeability transition**

Shigeomi Shimizu, Satoko Arakawa, and Ikuko Nakanomyo  
*Department of Pathological Cell Biology, Medical Research Institute, Tokyo Medical and Dental University*  
**permeability transition**

**O16. Role of mitochondrial ubiquitin ligase MITOL in mitochondrial dynamics**

Ryo Yonashiro, Yuya Kimijima, Ayumu Sugiura, and Shigeru Yanagi  
*Laboratory of Molecular Biochemistry, School of Life Sciences, Tokyo University of Life Sciences*  
**MITOL**

**O17. Physiological roles of mitochondrial fission factor Drp1 in mice and cultured cells**

Naotada Ishihara<sup>1</sup>, Masatoshi Nomura<sup>2</sup>, Akihiro Jofuku<sup>3</sup>, Satoshi O. Suzuki<sup>4</sup>, Hidenori Otera<sup>3</sup>, Noboru Mizushima<sup>1</sup>, and Katsuyoshi Mihara<sup>3</sup>  
<sup>1</sup> *Department of Physiology and Cell Biology, Tokyo Medical and Dental University*  
<sup>2</sup> *Departments of* <sup>3</sup> *Medicine and Bioregulatory Science,* <sup>4</sup> *Molecular Biology, Graduate School of Medical Sciences, Kyushu University*

**Drp1**

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**O18. Theoretical considerations of the effect of oxidative phosphorylation defect on neuron-astrocyte communications during stroke-like episodes in MELAS**

Takahiro Iizuka<sup>1</sup>, Junichi Hamada<sup>1</sup>, Fumihiko Sakai<sup>2</sup>, and Hideki Mochizuki<sup>1</sup>  
<sup>1</sup> *Department of Neurology, School of Medicine, Kitasato University*  
<sup>2</sup> *International Headache Center, Shinyurigaoka*

**MELAS**

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10:00 Coffee Break

10:30 Oral Session 4: Mitochondrial Metabolism

4

University of Arkansas for Medical Sciences

**O19. Mitochondrial aldehyde dehydrogenase activity maintains the functional integrity of mitochondria against oxidative stress**

Alexander M. Wolf<sup>1</sup>, Ikuroh Ohsawa<sup>2</sup>, and Shigeo Ohta<sup>1</sup>  
<sup>1</sup> *Department of Biochemistry and Cell Biology,* <sup>2</sup> *The Center of Molecular Hydrogen Medicine, Institute of Development and Aging Sciences, Nippon Medical School*

Alexander M. Wolf <sup>1</sup>	2	1
	1	2

**O20. The Wallerian degeneration slow (*Wld<sup>S</sup>*) gene provides neuroprotective effects against the impairment of mitochondrial electron transport chain in primary cultured mouse cortical neuron**

Shinji Tokunaga<sup>1,2</sup> and Toshiyuki Araki<sup>1,2</sup>

<sup>1</sup> Department of Electrical Engineering and Bioscience, Graduate School of Advanced Science and Engineering Waseda Univ.

<sup>2</sup> Department of Peripheral Nervous System Research, National Institute of Neuroscience, NCNP

*Wld<sup>S</sup>*

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**O21. Roles of adrenomedullin-RAMP2 system in cardiac mitochondrial function**

Takahiro Yoshizawa<sup>1</sup>, Takayuki Sakurai<sup>1</sup>, Akiko Kamiyoshi<sup>1</sup>, Teruhide Koyama<sup>1</sup>, Yuka Shindo<sup>1</sup>, Hayato Kawakami<sup>2</sup>, Hiroki Nakanishi<sup>3</sup>, Ryou Taguchi<sup>3</sup>, and Takayuki Shindo<sup>1</sup>

<sup>1</sup> Department of Organ Regeneration, Shinsu University Graduate School of Medicine

<sup>2</sup> Department of Anatomy, Kyorin University School of Medicine

<sup>3</sup> Department of Metabolome, The University of Tokyo

**AM-RAMP2**

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**O22. Analysis of quinone biosynthesis in tumor microenvironment**

Eriko Tomitsuka and Hiroyasu Esumi

*Cancer Physiol. Project, Natl. Cancer Ctr. Hosp. East*

**O23. Enhanced glycolysis induced by mtDNA mutations does not regulate metastasis**

Osamu Hashizume, Kazuto Nakada, and Jun-ichi Hayashi

*Graduate School of Life and Environmental Sciences, University of Tsukuba*

**mtDNA**

**O24. Reduction of mitochondrial DNA content activates and overexpresses K-Ras4a leading to prostate cancer progression**

Masahiro Higuchi and Cody Cook

*Department of Biochemistry and Molecular Biology, University of Arkansas for Medical Sciences*

**DNA**

Cody Cook

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12:00 Lunch

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13:15 Symposium

## S1. Evolutionary diversity of the mitochondrial genome of Apicomplexa

Kenji Hikosaka<sup>1</sup>, Yoh-ichi Watanabe<sup>2</sup>, Kiyoshi Kita<sup>2</sup>, and Kazuyuki Tanabe<sup>1</sup>

<sup>1</sup>Laboratory of Malariology, Research Institute for Microbial Diseases, Osaka University

<sup>2</sup>Department of Biomedical Chemistry, Graduate School of Medicine, The University of Tokyo

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## S2. Machinery of organellar DNA replication and transcription in malaria parasites

Narie Sasaki

Division of Biological Science, Graduate School of Science, Nagoya University

DNA

## S3. Organellar DNA of the oyster parasite *Perkinsus marinus*

Motomichi Matsuzaki, Isao Masuda, and Kiyoshi Kita

Department of Biomedical Chemistry, Graduate School of Medicine, University of Tokyo

DNA

## S4. Diversity of mitochondrial constituents and functions in eukaryotes: identification and characterization of a novel mitochondrion-related organelle in the enteric protozoan parasite *Entamoeba histolytica*

Fumika Mi-ichi<sup>1</sup>, Mohammad Abu Yousuf<sup>1,2</sup>, Kumiko Nakada-Tsukui<sup>1</sup>, and Tomoyoshi Nozaki<sup>1</sup>

<sup>1</sup>Department of Parasitology, National Institute of Infectious Diseases

<sup>2</sup>Department of Parasitology, Gunma University Graduate School of Medicine

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14:45 Poster Session

: 14:45 - 15:15

: 15:15 - 15:45

## P1. Molecular mechanism of mitochondria autophagy in yeast

Tomotake Kanki<sup>1</sup>, Dongchon Kang<sup>1</sup>, and Daniel J. Klionsky<sup>2</sup>

<sup>1</sup>Department of Clinical Chemistry and Laboratory Medicine, Kyushu University Graduate School of Medical Sciences

<sup>2</sup>Life Sciences Institute and Departments of Molecular, Cellular and Developmental Biology and Biological Chemistry, University of Michigan

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## P2. Knockdown of human TFAM induces aggregation of mitochondrial DNA; a novel function of human TFAM

Katsumi Kasashima, Megumi Sumitani, and Hitoshi Endo

Department of Biochemistry, Jichi Medical University

TFAM

TFAM

DNA

**P3. Utility of the mtGFP-Tg mouse as an analysis tool for mitochondrial morphology**

Midori Shimanuki<sup>1,2</sup>, Jun-Ichi Hayashi<sup>1</sup>, Hiromichi Yonekawa<sup>2</sup>, and Hiroshi Shitara<sup>2</sup>

<sup>1</sup> Graduate School of Life and Environmental Sciences, University of Tsukuba

<sup>2</sup> Laboratory of Mouse Models for Human Heritable Diseases, The Tokyo Metropolitan Institute of Medical Science

**mtGFP-Tg**

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**P4. Development of multi-layered MITO-Porter integrating efficient cytoplasmic delivery system and mitochondrial macromolecule delivery system**

Yuma Yamada, Ryo Furukawa, Yukari Yasuzaki, and Hideyoshi Harashima

Faculty of Pharmaceutical Sciences, Hokkaido University

“ MITO-Porter ”

**P5. Restraint stress induces mitochondrial dysfunction in the liver**

Emiko Kasahara<sup>1,2</sup>, Daisuke Kuratsune<sup>1</sup>, Mika Hori<sup>1,2</sup>, Eisuke F. Sato<sup>1</sup>, Atsuo Sekiyama<sup>1,2</sup>, and Masayasu Inoue<sup>1</sup>

<sup>1</sup> Dept. of Biochemistry & Molecular Pathology, Osaka City University Medical School

<sup>2</sup> Division of Therapeutic Neuropsychiatry, Esaka Hospital

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**P6. Kinetic modeling of the effect of nitric oxide on the mitochondrial respiration, superoxide generation and aging**

Ayako Hamada and Hirohisa Kishino

Department of Agricultural and Environmental Biology, Graduate School of Agriculture and Life Sciences, The University of Tokyo

**P7. Effects of mitochondrial superoxide anion produced by activated A-Raf on neural differentiation**

Masaki Miyazawa, Mika Kirinashizawa, Masashi Maruyama, Takamasa Ishii, Kayo Yasuda, and Naoaki Ishii

Department of Molecular Life Science, Tokai University School of Medicine

**A-Raf**

**P8. Nicotinamide mononucleotide adenylyltransferase (NMNAT) expression in mitochondrial matrix delays Wallerian degeneration**

Toshiyuki Araki and Naoki Yahata

Department of Peripheral Nervous System Research, National Institute of Neuroscience, NCNP

**Nicotinamide mononucleotide adenylyltransferase (NMNAT)**

**P9. Cerebral presynaptic mitochondrial dysfunction in klotho mice is different from natural aging**

Koji Hirata<sup>1,2</sup>, Nataliya Povalko<sup>1</sup>, and Yasutoshi Koga<sup>1</sup>

<sup>1</sup> Dept. Pediatrics and Child Health, Kurume Univ. Sch. of Med.

<sup>2</sup> Fac. Children's Studies, Dept. Children's Studies, Nishikyushu Univ.

## Klotho

<sup>1,2</sup> Nataliya Povalko<sup>1</sup> <sup>1</sup>

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<sup>2</sup>

## P10. Cytokines induce mitochondria related cell-death and its prevention by estrogen in estrogen receptor expressed rheumatoid arthritis synovial fibroblasts

Shigeaki Suenaga<sup>1</sup>, Hiroko P. Indo<sup>1</sup>, Kazuo Tomita<sup>1</sup>, Kosei Ijiri<sup>2</sup>, Setsuro Komiya<sup>2,3</sup>, Takami Matsuyama<sup>4</sup>, Hsiu-Chuan Yen<sup>5</sup>, Masahiro Higuchi<sup>6</sup>, Hirofumi Matsui<sup>7</sup>, Toshihiko Ozawa<sup>8</sup>, and Hideyuki J. Majima<sup>1,3</sup>

*Departments of <sup>1</sup> Oncology; <sup>2</sup> Orthopaedic Surgery; <sup>3</sup> Space Environmental Medicine, <sup>4</sup> Infection and Immunity, Kagoshima University Graduate School of Medical and Dental Sciences*

<sup>5</sup> Graduate Institute of Medical Biotechnology, Chang Gung University

<sup>6</sup> Department of Biochemistry and Molecular Biology, University of Arkansas for Medical Sciences

<sup>7</sup> Division of Gastroenterology, Graduate School of Comprehensive Human Sciences, University of Tsukuba

<sup>8</sup> Department of Health Pharmacy, Yokohama College of Pharmacy

<sup>1</sup> <sup>1</sup> <sup>1</sup> <sup>2</sup> <sup>2,3</sup> <sup>4</sup> Hsiu-Chuan Yen<sup>5</sup> <sup>6</sup>  
<sup>7</sup> <sup>8</sup> <sup>1,3</sup> <sup>1</sup> <sup>2</sup> <sup>3</sup> <sup>4</sup>

<sup>5</sup> Graduate Institute of Medical Biotechnology, Chang Gung University

<sup>6</sup> Department of Biochemistry and Molecular Biology, University of Arkansas for Medical Sciences

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## P11. *In vivo* functional imaging of oxidative stress in the striatum of patients with Parkinson's disease by <sup>62</sup>Cu-ATSM-PET

Masamichi Ikawa<sup>1</sup>, Hidehiko Okazawa<sup>2</sup>, Takashi Kudo<sup>2</sup>, Masaru Kuriyama<sup>1</sup>, Yasuhisa Fujibayashi<sup>2</sup>, and Makoto Yoneda<sup>1</sup>

*<sup>1</sup> Second Department of Internal Medicine (Neurology), Faculty of Medical Sciences, <sup>2</sup> Biomedical Imaging Research Center, University of Fukui*

<sup>62</sup>Cu-ATSM-PET  
<sup>1</sup> <sup>2</sup> <sup>2</sup> <sup>1</sup> <sup>2</sup> <sup>1</sup>  
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## P12. Critical defects in mitochondrial biogenesis dominated by cytochrome c oxidase deficiency: A cell-based diagnostic approach for mitochondrial diseases

Hideyuki Hatakeyama, Kayo Sawa, Shunji Kita, and Yu-ichi Goto

*Dept. Mental Retard. & Birth Def. Res., Natl. Inst. Neurosci., NCNP*

COX

## P13. A novel mutation in the mitochondrial tRNA for tryptophan associated with stroke-like episode, hypertrophic cardiomyopathy, acute pancreatitis, intestinal malabsorption, renal tubular disturbance

Ayako Katayama<sup>1</sup>, Hirofumi Komaki<sup>1</sup>, Ayako Hattori<sup>1</sup>, Yoshiaki Saito<sup>1</sup>, Hiroshi Sakuma<sup>1</sup>, Eiji Nakagawa<sup>1</sup>, Kenji Sugai<sup>1</sup>, Masayuki Sasaki<sup>1</sup>, Hideyuki Hatakeyama<sup>2</sup>, Yu-ichi Goto<sup>2</sup>, and Masato Mori<sup>3</sup>

*<sup>1</sup> Department of Child Neurology, National Center Hospital of Neurology and Psychiatry, NCNP*

*<sup>2</sup> Department of Mental Retardation and Birth Defect Research, National Institute of Neuroscience, NCNP*

*<sup>3</sup> Department of Pediatrics, Jichi Medical University Hospital*



tRNA <sup>Trp</sup> C5541T								mtDNA
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1			3					
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**P14. Granular swollen epithelial cells: a histological and diagnostic marker for mitochondrial nephropathy**

Akimitsu Kobayashi<sup>1</sup>, Yu-ichi Goto<sup>2</sup>, Michio Nagata<sup>3</sup>, and Yutaka Yamaguchi<sup>4</sup>

<sup>1</sup> Division of Kidney and Hypertension, Department of Internal Medicine, The Jikei University School of Medicine

<sup>2</sup> Department of Mental Retardation and Birth Defect Research, National Institute of Neuroscience, NCNP

<sup>3</sup> Department of Pathology, Graduate School of Comprehensive Human Sciences, University of Tsukuba

<sup>4</sup> Department of Pathology, Kashiwa Hospital, The Jikei University School of Medicine

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**P15. Successful cochlear implantation for a case of 11-year-old girl with mitochondrial DNA 625 G>A mutation who showed epilepsy and progressive sensorineural hearing loss**

Akira Sudo<sup>1</sup>, Norihito Takeichi<sup>2</sup>, Kana Hosoki<sup>3</sup>, Kei Murayama<sup>4</sup>, Akira Ohtake<sup>5</sup>, Ichizo Nishino<sup>6</sup>, Hitomi Sano<sup>1</sup>, Naoki Fukushima<sup>1</sup>, and Shinji Saitoh<sup>3</sup>

<sup>1</sup> Department of Pediatrics, Sapporo City General Hospital

Departments of <sup>2</sup> Otorhinolaryngology, <sup>3</sup> Pediatrics, Hokkaido University School of Medicine

<sup>4</sup> Department of Metabolism, Chiba Children's Hospital

<sup>5</sup> Department of Pediatrics, Saitama Medical University

<sup>6</sup> Department of Neuromuscular Research, National Institute of Neuroscience, NCNP

DNA 625G>A									
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**P16. A case of mitochondrial respiratory chain complex I and IV deficiency found by multiple tumor in his liver of an infant with chronic hepatic disorder**

Shigehiro Enkai<sup>1</sup>, Sachi Koinuma<sup>2</sup>, Junko Miyamoto<sup>3</sup>, Yukihiro Hasgawa<sup>3</sup>, Kei Murayama<sup>4</sup>, and Akira Ohtake<sup>5</sup>

<sup>1</sup> Department of Pediatrics, Fussa Hospital

<sup>2</sup> Department of Gastroenterology, National Center for Child Health and Development

<sup>3</sup> Department of Endocrinology, Tokyo Metropolitan Kiyose Children's Hospital

<sup>4</sup> Department of Metabolism, Chiba Children's Hospital

<sup>5</sup> Department of Pediatrics, School of Medicine Saitama Medical University

I						I+IV	1
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**P17. Molecular diagnoses and clinical manifestation of mitochondrial respiratory chain disorders (MRCB) in children**

Kei Murayama<sup>1</sup>, Aya Itoh<sup>2</sup>, Masami Ajima<sup>1</sup>, Yoshitami Sanayama<sup>1</sup>, Ayako Fujinami<sup>3</sup>, Tomoko Tsuruoka<sup>2</sup>, Taro Yamazaki<sup>4</sup>, Hiroko Harashima<sup>4</sup>, Masaki Takayanagi<sup>1</sup>, Masato Mori<sup>5</sup>, and Akira Ohtake<sup>4</sup>

*Departments of <sup>1</sup>Metabolism, <sup>2</sup>Neonatology, Chiba Children's Hospital*

*<sup>3</sup>Department of Pediatrics, Kimitsu Chuo Hospital*

*<sup>4</sup>Department of Pediatrics, Saitama Medical University*

*<sup>5</sup>Department of Pediatrics, Jichi Medical University*

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**15:45 Plenary Lecture 1**

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**PL1. Hydrogen sulfide is a signal molecule as well as a cell protectant from oxidative stress**

Dr. Hideo Kimura

*Department of Molecular Genetics, National Institute of Neuroscience, NCNP, Tokyo, Japan*

**16:25 Plenary Lecture 2**

2

**PL2. Sulfur toxicity in a human mitochondrial disorder**

Dr. Valeria Tiranti

*Unit of Molecular Neurogenetics – Pierfranco and Luisa Mariani Center for the study of Mitochondrial Disorders in Children, IRCCS Foundation Neurological Institute “C. Besta”, Milan, Italy*

**17:05 Plenary Lecture 3**

3

**PL3. The essential roles of mtCRIF1 in mitochondrial translation and oxidative phosphorylation**

Dr. Minh Shong

*Department of Molecular Medicine, Chungnam National University School of Medicine, Daejeon, Korea*

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**18:00 Banquet**

**Capo PELLICANO**

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## Day 3; Saturday, 19<sup>th</sup> December 3 12 19

### 9:00 Oral Session 5: Mitochondrial Diseases part 1

5 1

#### O25. The clinical implication of the endothelial tight junctional disruption in a patient with MELAS

Mihoko Matsuzaki<sup>1,3</sup>, Rieko Takahashi<sup>1</sup>, Tomohiro Nakayama<sup>1</sup>, Kazutoshi Nakano<sup>1</sup>, Makiko Osawa<sup>1</sup>, and Hideaki Oda<sup>2</sup>

*Departments of <sup>1</sup> Pediatrics, <sup>2</sup> Pathology, Tokyo Women's Medical University*

*<sup>3</sup> Nakagawa-no-sato Rehabilitation Center for Disabled and Mentally Retarded Children*

#### MELAS 1

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#### O26. Maternally inherited diabetes and deafness diagnosed at the age of 72

Kengo Maeda, Nobuhiro Ogawa, and Takashi Hisanaga

*Department of Neurology, National Hospital Organization Shiga Hospital*

72

#### O27. Parkin and PINK1 associate with mitochondrial elimination by autophagy

Shigeto Sato<sup>1</sup>, Kahori Shiba<sup>1</sup>, Fumiaki Sato<sup>1</sup>, Sumihiro Kawajiri<sup>1</sup>, Shinji Saiki<sup>1</sup>, Nobutaka Hattori<sup>1</sup>, Noriyuki Matsuda<sup>2</sup>, and Keiji Tanaka<sup>2</sup>

*<sup>1</sup> Department of Neurology, Juntendo University*

*<sup>2</sup> Center Laboratory of Frontier Science, The Tokyo Metropolitan Institute of Medical Science*

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#### O28. Induction of parkinsonism-related proteins in the spinal motor neurons of transgenic mouse carrying a mutant SOD1 gene

Nobutoshi Morimoto, Makiko Nagai, Kazunori Miyazaki, Yasuyuki Ohta, Tomoko Kurata, Yasushi Takehisa, Yoshio Ikeda, Tohru Matsuura, and Koji Abe

*Department of Neurology, Graduate School of Medicine, Dentistry and Pharmaceutical Science, Okayama University*

G93A SOD1-

#### O29. Loss of DJ-1 affects mitochondrial functions

Kimi Sakai<sup>1,2</sup>, Hiroshi Maita<sup>2</sup>, Kazuko Takahashi-Niki<sup>2</sup>, Sanae M. M. Iguchi-Ariga<sup>3</sup>, and Hiroyoshi Ariga<sup>2</sup>

*<sup>1</sup> Grad. Life Sci., <sup>2</sup> Grad. Pharm. Sci., <sup>3</sup> Grad. Agr., Hokkaido Univ.*

**DJ-1**  
12 2 2 3 2  
1 2 3

### 10:15 Coffee Break

### 10:45 Oral Session 6: Mitochondrial Diseases part 2

6 2

## O30. Mitochondrial respiratory chain disorders in neonate

Aya Itoh<sup>1</sup>, Kei Murayama<sup>2</sup>, Masami Ajima<sup>2</sup>, Yoshitami Sanayama<sup>2</sup>, Ayako Fujinami<sup>3</sup>, Tomoko Tsuruoka<sup>2</sup>, Madoka Aizawa<sup>1</sup>, Taro Yamazaki<sup>4</sup>, Hiroko Harashima<sup>4</sup>, Masaki Takayanagi<sup>2</sup>, Masato Mori<sup>5</sup>, and Akira Ohtake<sup>4</sup>

Departments of <sup>1</sup> Neonatology; <sup>2</sup> Metabolism, Chiba Children's Hospital

<sup>3</sup> Department of Pediatrics, Kimitsu Chuo Hospital

<sup>4</sup> Department of Pediatrics, Saitama Medical University

<sup>5</sup> Department of Pediatrics, Jichi Medical University

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 2            5            4  
              1            2  
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## O31. Metabolic analysis of <sup>13</sup>C-labeled respiratory substrates for non-invasive diagnosis of mitochondrial diseases

Masashi Tanaka

Department of Genomics for Longevity and Health, Tokyo Metropolitan Institute of Gerontology

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## O32. Bone marrow transplantation has a risk of transmission of mtDNA mutation

Nataliya Povalko<sup>1</sup>, Koji Hirata<sup>1,2</sup>, and Yasutoshi Koga<sup>1</sup>

<sup>1</sup> Department of Pediatrics and Child Health, Kurume University School of Medicine

<sup>2</sup> Faculty of Children's Studies, Department of Children's Studies, Nishikyushu University

DNA

Nataliya Povalko<sup>1</sup>            1,2            1  
 1  
 2

## O33. Pyruvate therapy in patients with mitochondrial myopathy

Yasutoshi Koga<sup>1</sup>, Nataliya Povalko<sup>1</sup>, Etsuo Naito<sup>2</sup>, and Masashi Tanaka<sup>3</sup>

<sup>1</sup> Department of Pediatrics and Child Health, Kurume University Graduate School of Medicine

<sup>2</sup> Department of Pediatrics, School of Medicine, Tokushima University

<sup>3</sup> Department of Genomics for Longevity and Health, Tokyo Metropolitan Institute of Gerontology

<sup>1</sup> Nataliya Povalko<sup>1</sup>            2            3  
 1  
 2  
 3

## O34. The effect of MCT milk and oil treatment for pyruvate dehydrogenase complex deficiency

Kazutoshi Nakano<sup>1,2,4</sup>, Tomohiro Nakayama<sup>3,4</sup>, and Kaoru Eto<sup>4</sup>

<sup>1</sup> Nakano Children's Clinic

<sup>2</sup> Isesaki-sawa Medical Association Hospital

<sup>3</sup> Matsudo Clinic

<sup>4</sup> Department of Pediatrics, Tokyo Women's Medical University

(PDHC)            MCT  
 1,2,4            3,4            4  
 1  
 2  
 3  
 4

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12:00 Lunch

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**13:15 Exchange Program between the Society and the MCM Supporting Organization**

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**13:45 Workshop: Mitochondrial Diseases Encountered in Various Clinical Departments**

**W1. Pediatrics:**

Masafumi Komaki, *National Center Hospital of Neurology and Psychiatry, NCNP*

**W2. Neurology:**

Takahiro Iizuka, *Kitasato University*

**W3. Otolaryngology:**

Tatsuya Yamasoba, *University of Tokyo*

**W4. Nephrology:**

Hiroshi Shiraga, *Saiseikai Kurihashi Hospital*

**W5. Cardiovascular medicine:**

Tomomi Ide, *Kyushu University Hospital*

**W6. Obstetrics:**

Hiroto Tajima, *Keio University*

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**15:45 Outstanding Oral/Poster Awards**

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**16:00 Closing Remarks**

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