The 50th Annual Meeting of
The Japanese Society of Child Neurology
~International Symposium
Celebrating the 50th Meeting of the JSCN~

PROGRAM

May 28-31, 2008
Hotel Nikko Tokyo
Invitation

Dear colleagues and friends,

I am delighted to announce that the 50th Congress of the Japanese Society of Child Neurology will be held May 28th-31st, 2008, at the Hotel Nikko Tokyo (Daiba Minato-ku, Tokyo 135-8625).

Our society strives to provide support and care for the next generation, even though they do not yet have the right to vote, promote healthy child development and assure better lives for children, suffering from neurological problems, and their families.

This meeting, dating back to July 1961, started as a special meeting in the field focusing on a wide spectrum of neurological abnormalities in children, including developmental and metabolic disorders. It was one of the first research meeting in this field in the world. Meetings have since been held at least once a year with researchers engaging in lively and informative discussions. The society has also played major roles in advancing clinical care, education, and research. The society has now expanded to more than 3,000 members.

The 21st century has been called "The Century of the Brain". At no time in life does the brain change as dramatically as before birth, and these changes continue during childhood, especially in infancy. Development is heralded by remarkable anatomical and physiological changes in the brain. Scientific approaches very different from those of adult neurology are thus required.

Unfortunately, behavioral problems in patients with developmental disabilities lead to social problems, an issue which has recently received considerable attention. As family size has decreased in recent decades, the demand for comprehensive care of individual children has grown. How to best care for these children, to optimize their treatment, is among the most important and significant challenges facing pediatric neurologists in the 21st century.

In this congress, comprehensive presentations and discussions will allow us to explore not only aspects of developmental medicine but also, more broadly, related field including psychosocial issues.

Our long-term goal is to promote research-yielding results which can improve the lives, and possibly even cure the diseases, of children suffering from neurological disorders.

Although this congress is basically a domestic meeting, there will be an international symposium on the first day, as well as lectures and another symposium in English on the second and third days. Abstracts, in English, from anywhere in the world are welcome. There will be a special program for researchers from Asia-Pacific region.

The congress promises to be an exciting and fruitful event for all who attend, providing an opportunity to interact with many distinguished physicians/scientists from all over the world working in the field of pediatric neurology, including developmental medicine. We hope that you will add your knowledge and expertise to this exciting exchange of ideas.

We will do our utmost to make your stay in Tokyo as enjoyable as possible. Please mark your calendar right now and submit your abstracts.

Looking forward to seeing you in Tokyo.

Makiko Osawa, MD, PhD
President, 50th Congress of Japanese Society of Child Neurology
Professor and Chairperson,
Department of Pediatrics,
School of Medicine,
Tokyo Women's Medical University
Hotel Nikko Tokyo
1-9-1 Daiba, Minato-ku, Tokyo 135-8625
TEL+81-3-5500-5500
FAX+81-3-5500-2525
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<th>Jupiter Room 6</th>
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<td>Board of directors 9:00 — 11:00</td>
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<td>General meeting of councilors 11:30 — 13:20</td>
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<td>Opening Address</td>
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<td>15:00</td>
<td>ISC50 JSCN Part 1 13:30 — 18:00 Recent Advances in Child Neurology Chair: Robert Rust Takao Takahashi Chair: Ching-Shiang chi Paolo Curatolo Chair: Masaya Segawa Robert Ouvrier Chair: Wu Xi-Ru Yong-Seung Hwang Chair: Virginia Wong Charles RJC Newton Chair: Eim-Yao Shen Solomon L Moshé Chair: Yoshiyuki Suzuki Shinichi Takeda Chair: Shigehiko Kamoshita Yoshiyuki Sankai</td>
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<td>ES 1 19:00 — 21:00 The Second annual meeting of the Japan Association of Child Sleep Chair: Jun Kohyama</td>
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[Abbreviated word]
- ISC50JSCN (International Symposium Celebrating the 50th Meeting of the JSCN)
- SL (Special Lecture)
- IL (Invited Lecture)
- DL (Didactic Lecture)
- S (Symposium)
- WS (Workshop)
- MS (Morning Educational Seminar)
- LS (Luncheon Seminar)
- ES (Evening Seminar)
- O (Oral)
- P (Poster)
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| 09:00 | Presidential Lecture: Clinical Aspects on Myotonic Dystrophy in Childhood  
9:00 ~ 9:30  
Makiko Osawa  
Chair: Koumei Komugai  
Presentation of JSCN Awards to Outstanding young Investigators  
9:30 ~ 9:50  
Toshiaki Hashimoto |
| 10:00 | ISC50 JSCN Part II  
10:00 ~ 11:00  
Celebration & Ceremony for the 50th Meeting of the JSCN  
Yukio Fukuyama  
Robert A Ouvrier  
Yong-Seung Hwang  
Master and Mistress of Ceremonies: Hirokazu Oguni  
Kyoko Hirasawa |
| 10:30 | SL 1  
11:30 ~ 12:30  
Mind viewed from brain  
Makoto Iwata  
Chair: Terao Shinya |
| 12:00 | SL 1  
12:30 ~ 13:30  
1) New progress in treatment of epilepsy  
Akio Ikeda  
Chair: Tsuyama Tanaka  
2) Treatment of intractable episodes based on the mechanism of action of the antiepileptic drugs  
Kenji Sugai  
Chair: Tsuyama Tanaka |
| 13:00 | SL 2  
13:30 ~ 14:30  
Society and developing mind  
Takeshi Yourou  
Chair: Mariko Momoi |
| 15:00 | IL 1  
14:30 ~ 15:30  
Congenital muscular dystrophy  
Francesca Muntoni  
Chair: Makiko Osawa |
| 16:00 | IL 2  
15:30 ~ 16:30  
Convulsing our way toward the pathophysiology of autism  
~clinical models and lessons for treatment~  
Roberto Tuchman  
Chair: Makiko Kaga |
| 17:00 |  
| 18:00 |  
| 19:00 |  
19:00 ~ 20:00  
Official Social Party  
Awarding party |
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<td>LS 3 12:30 - 13:30 Presentation based on the brain science; Success of our talk at the international conference Hiroshi Otsu &amp; Takanobu Takahashi</td>
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<th>Time</th>
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<th>Developmental disorders (diagnosis)</th>
<th>Neonatology 1</th>
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<td>15:00</td>
<td>14:30 - 15:00 (P-001 - 007) Chair: Takahisa Wada Toshiyuki Yoshimura</td>
<td>14:30 - 15:15 (P-041 - 049) Chair: Fujii Tatsuya Koji Imura</td>
<td>14:30 - 15:15 (P-083 - 091) Chair: Akira Yashahara Yoshiaki Saito</td>
<td>14:30 - 15:05 (P-122 - 128) Chair: Seiichi Sugama Hiroki Iizumi</td>
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<td>16:00</td>
<td>15:05 - 15:50 (P-008 - 016) Chair: Nobutada Tachi Yoshio Nomura</td>
<td>15:15 - 16:05 (P-050 - 059) Chair: Nobuyuki Shinozawa Hitoshi Sakuraba</td>
<td>Developmental disorders (examination 1) 15:05 - 15:45 (P-092 - 097) Chair: Tatsuki Ohno Marika Kurihara</td>
<td>Developmental disorders (examination 2) 15:00 - 16:10 (P-008 - 016) Chair: Hisashi Kuroki Yasutaka Kitajima</td>
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<td>17:00</td>
<td>15:50 - 16:30 (P-017 - 024) Chair: Yasuhiro Takeshima Hirofumi Komaki Vascular disorders 16:05 - 16:45 (P-060 - 067) Chair: Hitomi Saito Akira Oka</td>
<td>Metabolic disorders 2 15:15 - 16:05 (P-050 - 059) Chair: Nobuyuki Shinozawa Hitoshi Sakuraba</td>
<td>Developmental disorders (support) 16:15 - 17:00 (P-104 - 112) Chair: Masaki Ohto Kazuya Iimori</td>
<td>Developmental disorders (treatment) 17:00 - 17:45 (P-113 - 121) Chair: Asayo Ishizaki Hiroyoshi Koide</td>
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<td>Anomaly/Chromosomal disorders 15:50 - 16:50 (P-054 - 059) Chair: Hirofumi Ohashi Hiroshi Tamai</td>
<td>Developmental disorders (treatment) 17:00 - 17:45 (P-113 - 121) Chair: Asayo Ishizaki Hiroyoshi Koide</td>
<td>Brain anomaly 16:15 - 16:55 (P-143 - 150) Chair: Kyoko Ito Tomohide Goto</td>
<td>Infection/Immunology 1 16:45 - 17:25 (P-039 - 047) Chair: Naoyuki Takuma Naoya Hirakata</td>
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<td>Genetics 1 16:50 - 17:50 (P-060 - 065) Chair: Toyohito Matsuhashi Naomichi Kajii</td>
<td>Developmental disorders (treatment) 17:00 - 17:45 (P-113 - 121) Chair: Asayo Ishizaki Hiroyoshi Koide</td>
<td>Sleep/Autonomic nerve 15:35 - 16:15 (P-135 - 142) Chair: Yukara Awaysa Shinji Fujimoto</td>
<td>Infection/Immunology 2 16:45 - 17:25 (P-135 - 142) Chair: Akira Tomoda Mikio Hiraoka</td>
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<td>Anomaly/Chromosomal disorders 15:50 - 16:50 (P-054 - 059) Chair: Hirofumi Ohashi Hiroshi Tamai</td>
<td>Genetic 1 16:50 - 17:50 (P-060 - 065) Chair: Toyohito Matsuhashi Naomichi Kajii</td>
<td>Developmental disorders (treatment) 17:00 - 17:45 (P-113 - 121) Chair: Asayo Ishizaki Hiroyoshi Koide</td>
<td>Infection/Immunology 2 16:45 - 17:25 (P-135 - 142) Chair: Akira Tomoda Mikio Hiraoka</td>
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<td>Genetics 2 17:10 - 17:50 (P-033 - 040) Chair: Yuki Sawaishi Kyotakara Torii</td>
<td>Tumor/Neurosurgery 16:45 - 17:30 (P-068 - 076) Chair: Hisashi Kawamata Sumitomo Yoshimura</td>
<td>Developmental disorders (treatment) 17:00 - 17:45 (P-113 - 121) Chair: Asayo Ishizaki Hiroyoshi Koide</td>
<td>Infection/Immunology 2 16:45 - 17:25 (P-135 - 142) Chair: Akira Tomoda Mikio Hiraoka</td>
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<td>9:00</td>
<td>DL 1: Surgical management of pediatric epilepsy</td>
<td>Tatsuya Tanaka</td>
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<td>ISC50 JSCN Part III (DL 3)</td>
<td>Hideo Sagie</td>
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<td>Fukutinopathy</td>
<td>Tatsuya Kano</td>
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<td>10:00</td>
<td>S 1: Diagnosis and treatment for childhood epilepsy – Expert opinion</td>
<td>Hirokazu Oguni Yoko Ozawa</td>
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<td>ISC50 JSCN Part III (S 3)</td>
<td>Tadayuki Ishihara Masashi Mizuguchi</td>
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<td>Phenotypic Spectrum of Fukutinopathy</td>
<td>Tatsuya Kano</td>
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<td>LS 4: Challenges toward treatments for neurodegenerative diseases</td>
<td>Yoshikatsu Eto</td>
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<td>Kazue Imuma</td>
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<td>Treatment for intractable epilepsy</td>
<td>Shoji Tsuji</td>
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<td>JSCN General assembly</td>
<td>Yoko Ozawa</td>
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<td>ISC50 JSCN Part IV</td>
<td>Yuh-Jyong Masashi Mizuguchi</td>
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<td>Topics in neuromuscular disorders</td>
<td>Keiko Ishihagi</td>
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<td>Masaharu Hayashi Yuh-Jyong</td>
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<td>Keiko Ishihagi</td>
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<td>S 2: Hot Topics in Neonatal Neurology</td>
<td>Shinichi Nijima</td>
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<td>16:20 – 18:30</td>
<td>Masahiro Hayakawa</td>
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<td>Neurosurgical approach from a perspective of pediatric neurology</td>
<td>Shinji Date</td>
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<td>Development/Evaluation</td>
<td>Satoshi Mutsu</td>
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<td>15:30 – 16:30</td>
<td>Hiroshi Yamanouchi</td>
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<td>Epilepsy</td>
<td>Hideo Yamanouchi</td>
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<td>16:40 – 17:40</td>
<td>Tatsueku Yamamoto</td>
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<td>ES2: The Committee of the pharmacological issues – Concerta</td>
<td>Tatsuo Miyajima</td>
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<td>Distribution management committee</td>
<td>Kitami Hayashi</td>
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<td>17:00</td>
<td>ES3: Multiple Sclerosis and Neuromyelitis Optica in Japan</td>
<td>Toshinori Hara</td>
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<td>ES4: The Committee of the social activity</td>
<td>Takanori Sugimoto</td>
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<td>19:00</td>
<td>ES5: The Committee of the pharmacological issues – Boldinium</td>
<td>Takanori Sugimoto</td>
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<td>Treatment – Concerta</td>
<td>Takanori Sugimoto</td>
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<td>20:00</td>
<td>ES6: The Committee of the pharmacological issues – Boldinium</td>
<td>Takanori Sugimoto</td>
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<td>Treatment – Concerta</td>
<td>Takanori Sugimoto</td>
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<td>21:00</td>
<td>ES7: The Committee of the pharmacological issues – Boldinium</td>
<td>Takanori Sugimoto</td>
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<td>Neuroimmunology</td>
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<td>Chair: Yukiko Hirano</td>
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<td>Tatsuo Oya</td>
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<td>Neonatal Imaging/Others</td>
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<td>Shuichi Tsuneishi</td>
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<td>Neonatal Seizures/EEG</td>
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<td>Katsunori Fujii</td>
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<td>Muscular dystrophy 1</td>
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<td>Chair: Kitan Hayashi</td>
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<td>Atsushi Imanuma</td>
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<td>Chair: Koji Usuiji</td>
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<td>Takuya Tanabe</td>
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<td>Encephalitis 2</td>
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<td>Chair: Ryutaro Kira</td>
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<td>Seijiro Aso</td>
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<td>Encephalitis 3</td>
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<td>Chair: Yukihiko Fujita</td>
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<td>Tokyo University</td>
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<td>Encephalitis 4</td>
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<td>Masafumi Matsuo</td>
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<td>Mariko Mazawa</td>
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<td>Yasuhiro Suzuki</td>
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<td>Akemi Tanaka</td>
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<td>Neurinmotor disorders</td>
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<td>Yuichi Oto</td>
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**Poster Presentation**

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<td>8:00 ~ 9:00</td>
<td>A Clinical Approach to the Dysmorphic Child</td>
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<td>Kenjiro Kosaki</td>
<td>Pediatric neurourology: diagnosis A to Z</td>
<td>Hiroshi Oba</td>
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<td>Chair: Hitoshi Yamamoto</td>
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<td>9:00</td>
<td>Regulatory mechanism of neuronal migration mediated by the microtubule-associated protein doublecortin and its partners</td>
<td>Involuntary movement/Epilepsy (PC movie)</td>
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<td>Teruyuki Tomoike</td>
<td>(O–178 ~ 182)</td>
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<td>Chair: Takamori Takahashi</td>
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<td>Kazue Kimura</td>
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<td>10:00</td>
<td>Sleep/Behavior disorders</td>
<td>Epilepsy/Seizures 4</td>
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<td>Chair: Harumi Yoshinaga</td>
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<td>Chair: Jun Kohyama</td>
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**Seminars open to the public**

**How we can help Japanese children to be happy, thoughtful and warm-hearted?**

Chair: Toyoyoshi Matsuishi, Yoichi Sakakibara

**Introduction to the audience I**
Toyoyoshi Matsuishi (Department of Pediatrics and Child Health, Kureme University School of Medicine)

**Introduction to the audience II**
Yoichi Sakakibara (Department of Child Care and Education, Ohshanomizu University)

**Early risers and early sleepers are healthy both mentally and physically**
Kohyama Jun (Tokyo Keita Shokai Hoken Hospital)

1. Challenge of pediatric neurologists to child mental problems: From a clinical and neuroscientific point of view
Shimichiro Nagamizue (Department of Pediatrics and Child Health, Kitasato University School of Medicine)

2. PTNS and the developing brain
Nobumasa Kato (Department of Psychiatry, Showa University School of Medicine)

3. As a care-partners for children who need Special Education
Michiko Hara (Faculty of Education, Gunma University)

4. The prevention of childhood depression
Kayo Inoko (Tokyo Institute of Psychiatry)

5. Closing Remarks Toyoyoshi Matsuishi
Day 1 Room 5 (Apolon)

Opening Address
13:30 ~ 18:00 International Symposium Celebrating the 50th Meeting of the JSCN Part I Recent Advances in Child Neurology 1.
13:35 ~ 14:05 1) Big brains & small brains — genetic and epigenetic mechanisms of brain size alteration —
   Chairman Robert Rust (Department of Neurology, University of Virginia School of Medicine, Charlottesville, VA, USA)
   Takao Takahashi (Department of Pediatrics, Keio University School of Medicine, Tokyo, Japan)
14:05 ~ 14:35 2) Recent advances on neurocutaneous syndromes
   Chairman Ching-Shiang Chi (Department of Pediatrics, Taichung Veterans General Hospital, Taichung, Taiwan)
   Paolo Curatolo (Professor of Pediatric Neurology and Psychiatry, Department of Neuroscience, Tor Vergata University, Rome, Italy)
14:35 ~ 15:05 3) Age-related presentations of hereditary peripheral neuropathies in childhood
   Chairman Masaya Segawa (Segawa Clinic)
   Robert A Ouvrier (The Institute for Neuromuscular Research, The Children's Hospital at Westmead, Sydney, Australia)
   — Intermission —

Recent Advances in Child Neurology 2.
15:15 ~ 15:40 4) Japanese encephalitis in Korea and Asian countries — Can it be under control? —
   Chairperson Wu Xi-Ru (First Teaching Hospital Beijing Medical University, Beijing, China)
   Yong-Seung Hwang (Seoul National University, Children's Hospital, Seoul, Korea)
15:40 ~ 16:10 5) The impact of infections of the central nervous system on African children
   Chairperson Virginia Wong (Department of Pediatrics, Queen Mary Hospital, Hong Kong, China)
   Charles RJC Newton (Kenya Medical Research Institute, Kiliﬁ, Kenya and University College London, London, UK)
16:10 ~ 16:40 6) Progress in the biological aspects of status epilepticus
   Chairperson Ein-Yiao Shen (Department of Pediatrics, Taipei Medical University-Wan Fang Medical Center, Taipei, Taiwan)
   Solomon L Moshé (Albert Einstein College of Medicine and Montefiore Medical Center New York, NY, USA)
   — Intermission —

Recent Advances in Child Neurology 3.
16:50 ~ 17:20 7) Gene therapy for muscular dystrophy
   Chairperson Yoshiyuki Suzuki (International University of Health and Welfare, Ohtawara, Japan)
   Shinichi Takeda (Department of Molecular Therapy, National Institute of Neuroscience, National Center of Neurology and Psychiatry, Kodaira, Japan)
17:20 ~ 17:55 8) Robot suits — Cybernooid, biorobotics, control for supporting disabled persons —
   Chairperson Shigeohiko Kamoshita (President Emeritus, International Medical Center of Japan, Tokyo, Japan)
   Yoshiyuki Sankai (Department of System & Information Engineering, University of Tsukuba, Tsukuba, Japan)

Day 1 Room 6 (Jupiter)

19:00 ~ 21:00 Evening Seminar 1.
   The second annual meeting of the Japan Association of Child Sleep; Satisfactory sleep for children holds promise for the future
   Chairman Jun Kohyama (Tokyo Kita Shakai Hoken Hospital)
   1) Kumi Kato (Molecular Research Center for Children's Mental Development, Osaka University Graduate School of Medicine)
   2) Jun Kohyama (Tokyo Kita Shakai Hoken Hospital)
   3) Yoko Asaka (Kobe University)
   4) Junko Ohnata (Department of Pediatrics, Asahikawa Medical College)
   5) Noboru Ohki (NoruPro Light Systems, Inc)
   6) Takako Jeedoi (Department of Child Development Kumamoto University Hospital)
   7) Junko Kawatani (Child Development Sociology Faculty of Medical and Pharmaceutical Sciences Kumamoto University Graduate School)
   8) Takashi Ohya (Department of Pediatrics and Child Health, Kurume University School of Medicine)
8:50 ～ 9:20  Presidential Lecture
Clinical Aspects on Myotonic Dystrophy in Childhood
Chairman  Koumei Kumagai (chisakihananosono)
Makiko Osawa (Department of Pediatrics, School of Medicine, Tokyo Women's Medical University)

9:30 ～ 9:50  Presentation of JSCN Awards to Outstanding Young Investigators
Chairman Toshiaki Hashimoto (Department of Special Support Education, College of Education, Naruto University of Education)

10:00 ～ 11:30  International Symposium Celebrating the 50th Meeting of JSCN Part II
Master and Mistress of Ceremonies Hirokazu Oguni (Department of Pediatrics, Tokyo Women's Medical University)
Kyoko Hirasawa (Department of Pediatrics, Tokyo Women's Medical University)

10:00  Opening Address Makiko Osawa (President of the 50th JSCN congress)
10:05  1) History of Child Neurology in Japan
  Yukio Fukuyama (Tokyo Women's Medical University)
10:18  2) Introduction of the Program
10:23  Presentation of Awards to JSCN Contributors
10:33  3) The Japan's role in child neurology in the AOCNA
  Yong-Seung Hwang (Seoul National University Children's Hospital, Seoul, Korea)

11:30 ～ 12:30  Special Lecture 1
Mind viewed from brain
Chairman Teruhisa Miike (Department of Child Development, Faculty of Medical and Pharmaceutical Sciences, Kumamoto University Graduate School)
Makoto Iwata (Department of Neurology, Tokyo Women's Medical University, Tokyo, Japan)

12:30 ～ 13:30  Luncheon Seminar 1
1) New progress in Treatment of Epilepsy
Chairman Tatsuya Tanaka (Department of Neurosurgery, Asahikawa Medical College)
Akio Ikeda (Kyoto University)
2) Treatment of intractable epilepsies based on the mechanism of action of the antiepileptic drugs
Chairman Tatsuya Tanaka (Department of Neurosurgery, Asahikawa Medical College)
Kenji Sugai (Department of Child Neurology, National Center Hospital for Mental, Nervous and Muscular Disorders, National Center of Neurology and Psychiatry, Tokyo)

13:30 ～ 14:30  Special Lecture 2
Society and developing mind
Chairperson Mariko Momoi (Jichi Medical University, Department of Pediatrics)
Takeshi Yourou (Tokyo University)

14:30 ～ 15:30  Invited Lecture 1
Congenital muscular dystrophy
Chairperson Makiko Osawa (Department of Pediatrics, Tokyo Women's Medical University)
Francesca Muntoni (Dubowitz Neuromuscular Centre, UCL Institute of Child Health, London, UK)

15:30 ～ 16:30  Invited Lecture 2
Convulsing our way toward the pathophysiology of autism: clinical models and lessons for treatment
Chairperson Makiko Kaga (National Institute of Mental Health, National Center of Neurology and Psychiatry)
Roberto Tuchman (Department of Neurology, Miami Children's Hospital Dan Marino Center, Miami, FL, USA)
12:30 ~ 13:30 Luncheon Seminar 2
   Chair: Wu Xi-Ru (First Teaching Hospital Beijing Medical University, Beijing, China)
   1) Diagnosis and management of seizures in the ICU
      Akihisa Okumura (Department of pediatrics, Juntendo University)
   2) Brain Monitoring Made Easy
      Anita Kharbteng (Clinical Support Manager, NeuroCare, Asia Pacific, VIASYS Healthcare Hong Kong Ltd.)

14:30 ~ 15:30 Learning disabilities
   Chair: Masutomu Miyao, Masao Aihara

O-001 A study of the early detection method of dyslexic children: I. Oral sentence reading task
   * Uchiyama Hitoshi1, Seki Ayumi2, Koeda Tatsuya3
   1) Graduate School of Medical Sciences, Tottori University, Yonago, Japan, 2) Department of Education, Faculty of Regional Sciences, Tottori University, Tottori, Japan, 3) Tottori Medical Center, National Hospital Organization

O-002 A study of the early detection method of dyslexic children: II. Phonological skills
   * Seki Ayumi1, Uchiyama Hitoshi2, Koeda Tatsuya1,2
   1) Department of Education, Faculty of Regional Sciences, Tottori University, Tottori, Japan, 2) Tottori Medical Center, National Hospital Organization, 3) Graduate School of Medical Sciences, Tottori University, Yonago, Japan

O-003 Dual Route Cascaded Model in reading Hiragana and Romaji by phonological reaction time
   * Sugita Katsu1, Sugita Kiyoko1, Fujita Katsunori2
   1) Division of Child Health, Faculty of Education, Chiba University, 2) Department of Health Care and Sports, Faculty of Human Life, Toyo University, 3) Department of Pediatrics, Chiba University

O-004 Phonological Processing in children with Decoding Problems and Problems of Reading Aloud
   * Wakamiya Eiji1, Okumura Tomohito2, Mizuta Mekumi2, Kurimoto Naoko2, Tanaka Keiko3, Tamai Hiroshi1,2
   1) Faculty of Nursing and Rehabilitation, Aino University, Osaka, Japan, 2) LD center, Osaka Medical College, Osaka, Japan, 3) The Department of Pediatrics, Showa Hospital, Amagasaki, Japan, 4) The Department of Pediatrics, Osaja Medical College, Osaka, Japan

O-005 Relationship between Syllable Reading Skill and Oral Text Reading Skill in Developmental Dyslexia
   * Okumura Tomohito1, Kurimoto Naoko1, Mizuta Mekumi1, Tanaka Keiko1, Wakamiya Eiji1, Tamai Hiroshi2
   1) Osaka Medical College, LD Center, Osaka, 2) Osaka Medical College, Department of Pediatrics, Osaka, Japan, 3) Aino University, Osaka, Japan, 4) Syowa Hospital, Hyogo, Japan

O-006 Anterior prefrontal activity associated with emotions: a study with event-related NIRS
   * Hoshi Yoko, Huang Jinghua
   Tokyo Institute of Psychiatry, Integrated Neuroscience Research Team

15:30 ~ 16:30 Cerebral palsy rehabilitation 1
   Chair: Takahiro Nara, Hideo Shimoizumi

O-007 Activation of olfactory cortex in severely-disabled children: A near-infrared spectroscopy study
   * Kobayashi Yasuko1, Omura Kiyoshi1, Kikuchi Toshihiko1, Yashima Takeshi1, Ozaki Hisak1, Tsuchiya Shigeru1
   1) Devison of Pediatric Neurology, Nishitaga National Hospital, Sendai, Japan, 2) Laboratory of Physiology, Faculty of Education, Ibaraki University, Mito, Japan, 3) Department of Pediatrics, Tohoku University School of Medicine, Sendai, Japan
O-008 Clinical implications of cerebellar injury in the extremely premature survivor with cerebral palsy.
* Arai Hiroshi, Nabatame Shin, Kato Yoshimi, Hirai Satori
Department of Pediatric Neurology, Morinomiya Hospital

O-009 Selenium deficiency in patients with severe motor and intellectual disability
* Korematsu Seigo, Shimizu Miki, Anan Aki, Niu Aya, Goto Chika, Sekiguchi Kazuhito, Sato Keisuke,
Suenobu Souichi, Izumi Tatsuro
Department of Pediatrics and Child Neurology, Department of Brain and Nerve Science, Oita University Faculty of Medicine, Oita, Japan

O-010 Comprehensive treatment for severe spasticity
* Nagasawa Tetsuro, Hoshino Hideki, Mizuguchi Koichi, Kubota Masaya,
Morota Nobuhito, Oka Akira
1) Division of Neurology, National Center for Child Health and Development, Tokyo, Japan, 2) Division of Neurosurgery, National Center for Child Health and Development, Tokyo, Japan, 3) Department of Pediatrics, The Tokyo University Hospital, Tokyo, Japan

O-011 Botulinum toxin management of spastic diplegia in childhood
* Nezu Atsu, Takeda Saoko, Ichikawa Kazushi
Yokohama City University Medical Center Children's Medical Center

O-012 Changes of influenza HI-titer after vaccination in profound patients with SMID
* Shinichi Magara, Konishi Tohru, Kinoshita Satoru
1) Nagaoka Ryoikuen, Niigata, Japan, 2) The Department of Pediatrics, Niigata National Hospital, Niigata, Japan

16:30 ～ 17:40  Cerebral palsy rehabilitation 2
Chair: Kazuo Kodama, Hiroshi Ozawa

O-013 The problems of care of patients who have very severe psychophysologic disorders in our hospital
* Takeshita Eri, Otani Ryoko, Itabashi Hisashi, Kita Syunji, Shimamura Keichi, Murakami Nobuyuki, Sakata Ryoichi, Nagai Toshiro
Dokkyo Medical University Koshigaya Hospital, Saitama, Japan

O-014 New QOL Assessment Questionnaire for Persons with SMIDS
* Matsumoto Akiko, Miyazaki Shuji, Hasegawa Sakurako
1) KOBATO GAKUEN Aichi Human Service Center, Aichi, Japan, 2) Institute for Developmental Research, Aichi Human Service Center, Aichi, Japan

O-015 Severely disabled children with respiratory and/or tracheotomy:investigation in 8 prefectures
* Sugimoto Tateo, Tamura Masanori
1) The Ethics Committee, Japan Pediatric Society, Tokyo, Japan, 2) Biwako Gakuen Medical and Welfare Center, Yaw, Japan, 3) Saitama Medical Center, Saitama Medical University, Kawagoe, Japan

O-016 A research for breathing management in Kanagawa
* Sameshima Kiyoko, Takagi Atushi, Thuji Megumi, Osaka Hitoshi, Iai Mizue, Yamashita Sumimasa, Yamada Michiko
Division of Neurology, Kanagawa Children's Medical Center, Yokohama, Japan

O-017 Cooperation between Hospital and Welfare Institute on general care for severely disabled children
* Yamaguchi Fumika, Ishima Atsushi, Kuwashima Katsuko, Kondou Ikuko
1) Ibaraki Disabled Children's Hospital, Mito, Japan, 2) Saitama Medical University, Saitama Medical Center, Kawagoe, Japan

O-018 A survey on teachers idea, about medical cares at school for physically handicapped children
* Miyamoto Yasaku, Yamamoto Hitoshi, Fukuda Mihoko, Murakami Hiroshi, Kamiyama noriko, Hashimoto Syuuji
1) Division of Pediatrics, Kawasaki Municipal Tama Hospital, 2) Department of Pediatrics, St. Marianna University School of Medicine, Kawasaki, Japan
O-019 Community health care program for the handicapped children in the stricken area in Indonesia
* Takada Satoshi 1, Matsuda Nobuko 1, Matsui Gakuyu 1, Yamamoto Akio 1
Sunartini Hapsara 2
1) Faculty of Health Sciences, Kobe University School of Medicine, Kobe, Japan, 2) Department of Pediatrics, Faculty of Medicine, Gadjah Mada University

17:50 ～ 18:30 Developmental disorders & Genetics
Chair: Yasuyuki Niki, Mitsuhiro Kato

O-020 A first Japanese case with CDKL5 mutation causing an atypical form of Rett syndrome
*Takahashi Satoru, Araki Akiko, Ohinata Junko, Suzuki Nao, Tanaka Hajime, Fujieda Kenji
Department of Pediatrics, Asahikawa Medical College, Asahikawa, Japan

O-021 MeCP2-target genes related to Rett syndrome-Toward identification of Autism marker gene-
*Kubota Takeo1, Soutome Masaki1, Itoh Masayuki2, Goto Yu-ichi3, Inazawa Johji 3
1) Department of Epigenetic Medicine, Univ. Yamanashi, Yamanashi, Japan, 2) Department of Mental Retardation and Birth Defect Research, National Institute of Neuroscience, NCNP, Tokyo, Japan, 3) Department of Mol Cytogenetics, MRI, Tokyo Medical Dental University, Tokyo, Japan

O-022 A clinical study on Autism with SHANK3 abnormalities
*Okamoto Nobuhiko1, Uchino Shigeo2
1) Department of Planning and Research, Osaka Medical Center and Research Institute for Maternal and Child Health, Osaka, Japan, 2) Department of Neurochemistry, National Institute of Neuroscience, Kodaira, Tokyo

O-023 Molecular mechanism of neuron-specific imprinting of Angelman syndrome gene Ube3a
*Kishino Tatsuya
Division of Functional Genomics, Center for Frontier Science, Nagasaki University, Nagasaki, Japan

14:30 ～ 15:40 Epilepsy/Seizures 1
Chair: Takehiko Morimoto, Osamu Kanazawa

O-024 Diffusion Weighted Images in children with prolonged febrile seizure, compared with clinical findings
*Yokoi Setsuri1, Tsuji Takeshi2, Nakata Tomohiko2, Kubota Tetsuo2, Maruyama Koichi3, Itomi seiko4, Kato Toru5, Sofue Ayako6, Kajita Mitsuharu6, Okumura Akihisa7, Natsume Jun8
1) Department of Pediatrics, Tosei General Hospital, Seto, Japan, 2) Department of Pediatrics, Okazaki City Hospital, Okazaki, Japan, 3) Department of Pediatrics, Nagoya Graduate University Hospital, Nagoya, Japan, 4) Department of Pediatrics, Anjo Kosei Hospital, Anjo, Japan, 5) Department of Pediatric Neurology, Aichi Colony Central Hospital, Kasugai, Japan, 6) Department of Pediatrics, Nagoya First Red Cross Hospital, Nagoya, Japan, 7) Department of Pediatrics, Nagoya Memorial Hospital, Nagoya, Japan, 8) Department of Pediatrics, Kamo Hospital, Toyota, Japan, 9) Department of Pediatrics, Juntendo University, Tokyo, Japan

O-025 DWI and FDG-PET in patients with prolonged febrile seizures: partial volume correction studies.
*Natsume Jun1, Maruyama Koichi2, Sofue Ayako3, Okumura Akihisa4
1) Department of Pediatrics, Nagoya University School of Medicine, Nagoya, Japan, 2) Department of Pediatrics, Japanese Red Cross Nagoya First Hospital, 3) Department of Child Neurology, Aichi Prefectural Coloney, 4) Department of Pediatrics, Nagoya Memorial Hospital, 5) Department of Pediatrics, Juntendo University

O-026 EEG and MRI findings in patients with acute encephalopathy or prolonged febrile seizure
*Nakata Tomohiko1, Tsuji Takeshi2, Yokoi Setsuri1, Maruyama Kouichi2, Katou Toru3, Kubota Tetsuya4, Sofue Ayako5, Okumura Akihisa1, Natsume Jun4
1) Department of Pediatrics, Nagoya University school of Medicine, Nagoya, Japan, 2) Okazaki City Hospital, 3) Tosei general Hospital, 4) Aichi Prefectural Corony Hospital, 5) Anjo Kosei Hospital, 6) Nagoya Memorial Hospital, 7) Department of Pediatrics, Juntendo University, Tokyo, Japan
O-027  Underlying disease in children with epileptic status
   *Okanishi Tohru1,2, Maegaki Yoshihiro1, Ohno Kousaku1
   1) Division of Child Neurology, Institute of Neurological Sciences, Faculty of Medicine, Tottori University, Yonago, Japan.  2) Department of Pediatrics, Neonatology and Congenital Disorders, Nagoya City University Graduate School of Medical Sciences, Nagoya, Japan

   *Nishiyama Itsuko, Oka Makio, Kobayashi Katsuhiko, Yoshinaga Harumi, Ohtsuka Yoko
   Department of Child Neurology, Okayama University Graduate School of Medicine, Okayama, Japan

O-029  Lack of association between a polymorphism in SYN2 with genetic susceptibility to febrile seizures
   *Ishizaki Yoshito, Kira Ryutaro, Torisu Hiroyuki, Sanefuji Masafumi, Yukaya Naoko, Hara Toshiro
   Department of Pediatrics, Graduate School of Medical Sciences, Kyushu University, Fukuoka, Japan

O-030  Parental psychological reactions to the first febrile seizure in their child
   *Hata Sonoko1,2, Kanemura Hideaki1, Hatakeyama Kazuo1, Sugita Kanji1, Aihara Masao1
   1) Department of Pediatrics, Faculty of Medicine, University of Yamanashi, Yamanashi, Japan.  2) Department of Pediatrics, Yamanashi Red Cross Hospital, Yamanashi, Japan

15:40 ～ 17:00  Epilepsy/Seizures 2

Chair: Kun-Long Hung, Shinichi Hirose

O-031  Efficacy of antiepileptic drugs in 38 patients with SMEI
   *Takahashi Hiroko, Takahashi Yukitoshi, Mine June, Ohotani Sanae, Ohotani Hideyuki, Ikeda Hiroko, Shikis Tatsuhiko, Shimomura Jiro, Kubota Yuhko, Kubota Hidemoto, Shigematsu Hideo, Inoue Yushi, Fujiiwara Tateki
   National Epilepsy Center Shizuoka Institute of Epilepsy and Neurological Disorders

O-032  The clinical effectiveness of CZP and KBr co-therapy for patients with SMEI
   *Okanari Kazuo, Sone Ritsuko, Uchiyama Shinichi, Maeda Tomoki, Sato Keisuke, Izumi Tatsuro
   Department of Pediatrics, Department of Brain and Nerve, Oita University Faculty of Medicine, Oita, Japan

O-033  Do all children with first episode of fever and seizure need a Lumber puncture?
   *Ajit Rayamajhi, Ruby Joshi Bataajo, Chandeshwar Mahaseth
   Department of Pediatrics, National Academy of Medical Sciences, Kathmandu, Nepal

O-034  Levetiracetam adjunctive therapy in children with refractory epilepsy
   *Kun-Long Hung, Ching-Wan Tsai, Chien-Hung Liu, Hung-Tsai Liao
   Department of Pediatrics, Cathay General Hospital, Taipei, Taiwan

O-035  Clinical analyses of 30 patients with tuberous sclerosis complex: relationship of abnormal brain imaging, seizures and cognitive function.
   *Pou-Leng Cheong1, Yi-Ning Su2, Wang-Tso Lee3
   1) Department of Pediatrics, Hsinchu General Hospital, Executive Yuan, Hsinchu, Taiwan.  2) Department of Medical Genetics, National Taiwan University Hospital, Taipei, Taiwan.  3) Department of Pediatrics, National Taiwan University Hospital, Taipei, Taiwan

O-036  Mozart effect on epileptiform discharge in children of epilepsy in Taiwanese
   *Lung-Chang Lin1,2, Rei-Cheng Yang1
   1) Department of Pediatrics, Kaohsiung Medical University Hospital, Kaohsiung Medical University, Kaohsiung, Taiwan.  2) Department of Pediatrics, Kaohsiung Medical University Hospital, Kaohsiung Medical University, Kaohsiung, Taiwan
O-037 Good outcome in Rolandic epilepsy of children with mild cerebral palsy and mental deficiency
* Kanazawa Osamu¹ ², Konishi Kaoru²
¹ Department of Psychiatry, Saitama Medical University, Saitama, Japan, ² Department of Pediatric Neurology, Medical Service Division, Saitama Municipal General Center for Physically, Mentally and Auditorily Handicapped, Saitama, Japan

O-038 Correlation between the prefrontal lobe growth and seizure duration in BECT with behavior disorders
* Kanemura Hideaki, Sugita Kanji, Aihara Masao
Department of Pediatrics, Faculty of Medicine, University of Yamanashi, Yamanashi, Japan

17:00 ~ 18:10 Epilepsy/Seizures 3

O-039 Vigabatrin treatment of infantile spasms due to Tuberous sclerosis.
* Kitai Yukihiro, Morita Yoshiko, Araya Ken, Tominaga Kouji, Nabatame Shin², Shimono Kuriko, Okinaga Takeshi, Oaono Keichii
The department of pediatrics, Osaka University Graduate School of Medicine, Osaka, Japan, ² The hospital of Morinomiya, Osaka, Japan

O-040 Relationship between effect of first ACTH therapy and prognosis in West syndrome
* Okumura Yoshinori, Aiba Hideo, Watanabe Seiji, Hirano Keiko, Hojo Hiroatsu
Department of Pediatric Neurology, Shizuka Children's Hospital, Shizuoka

O-041 Regional cerebral blood flow of West syndrome quantified with 3DSRT
* Hamano Shinichiro¹, Yoshinari Satoshi¹,², Higurashi Norimichi¹,², Tanaka Manabu¹, Minamitani Motoyuki²,³, Kikuchi Kenjiro¹, Koichihiro Reiko¹,², Eto Yoshikatsu²
¹ Division of Neurology, Saitama Children's Medical Center, Saitama, Japan, ² Department of Pediatrics, Jikei University School of Medicine, Tokyo, Japan, ³ Department for Child Health and Human Development, Saitama Children's Medical Center, Saitama, Japan, ⁴ Fukaya Red Cross Hospital, Fukaya, Japan

O-042 The correlation between the prognosis of infantile spasms and the findings of IH-MR spectroscopy
* Imamura Atsushi, Miyajima Hiroko, Matsu Naoki, Ito Reiko, Orii Koji
The Department of Pediatrics, Gifu Prefectural General Medical Center, Gifu, Japan

O-043 Clinical Manifestations of patients with early onset West syndrome and cerebral hypomyelination
* Tohyama Jun¹, Akasaka Noriyuki¹, Saito Naka¹, Osaka Hitoshi², Yamashita Sumimasa³, Maegaki Yishihiro¹, Fukumura Shinoji¹, Takayama Rumiko³, Uematsu Mitsugu³, Haginoya Kazuhiro⁴,⁶
¹ Department of Pediatrics, Nishi-Niigata Chuo National Hospital, Niigata, Japan, ² Division of Neurology, Kanagawa Children's Medical Center, Yokohama, Japan, ³ Division of Child Neurology, Tottori University, Faculty of Medicine, Yonago, Japan, ⁴ Department of Pediatrics, Aomori Prefectural Central Hospital, Aomori, Japan, ⁵ Department of Pediatrics, Tohoku University Graduate School of Medicine, Sendai, Japan, ⁶ Department of Pediatric Neurology, Takuto Rehabilitation Center for Children, Sendai, Japan

O-044 Siblings of holocarboxylase synthetase deficiency with West syndrome
* Hattori Ayako¹, Ando Naoki¹, Kobayashi Satoru¹, Ito Tetsuya¹, Fujimoto Shinji², Ban Kyoko³, Ishikawa Tatsuya³,⁴, Togari Hajime¹
¹ Department of Pediatrics and Neonatology Nagoya City University, Graduate School of Medical Sciences, Nagoya, Japan, ² Tsutsuijigaoka Kodomo Clinic, ³ Department of Pediatrics, Yokkaichi Municipal Hospital, ⁴ Nihon Fukushi University

O-045 Immunologic aspects of West syndrome, through lymphocyte subsets and serum cytokines: 1st report.
* Shihira Takashi, Watanabe Mio
Department of Neurology, Gunma Children's Medical Center, Gunma, Japan
12:30 ～ 13:30  

**Luncheon Seminar 3**

Presentation based on the brain science: Success of our talk at the international conference

Chairman: Takao Takahashi (Department of Pediatrics, School of Medicine, Keio University)

Hiroshi Otsubo (Director of Clinical Neurophysiology Division of Neurology, The Hospital for Sick Children Assistant Professor, Department of Pediatrics, University of Toronto)

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**O-046 Tay-Sachs disease: correlation between structural changes in HexA and phenotypes.**

* Sakuraba Hitoshi

Department of Analytical Biochemistry, Meiji Pharmaceutical University, Tokyo, Japan

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**O-047 Dysregulation of autophagy in GM1-gangliosidosis**

* Nanba Eiji, Higaki Katsumi

Division of Functional Genomics, Research Center for Bioscience and Technology, Tottori University, Yonago, Japan

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**O-048 Disturbances of acetylcholinergic neurons in xeroderma pigmentosum group A**

* Hayashi Masaharu, Ohto Tatsuyuki, Araki Satoshi

1) The Department of Clinical Neuropathology, Tokyo Metropolitan Institute for Neuroscience, Tokyo, Japan.

2) The Department of Pediatrics, University of Tsukuba, School of Medicine, Ibaragi, Japan.

3) The Department of Pediatrics, Tokyo Medical and Dental University, Tokyo, Japan

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**O-049 15 female patients with mutation in pyruvate dehydrogenase E1 alpha subunit and normal activity**

* Naito Etsuo, Shimakawa Seishi, Nishimura Mio, Kotani Yumiko, Kagami Shoji

1) Division of Pediatrics, Tokushima Red Cross Hinomine Medical and Rehabilitation Center.

2) Department of Pediatrics, Institute of Health Biosciences, The University of Tokushima, Graduate School, Tokushima, Japan

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**O-050 Molecular basis of MCT8 deficiency in a Japanese patient**

* Itoh Masatsune, Kakinuma Hiroaki

Department of Pediatrics, Kanazawa Medical University, Ishikawa, Japan

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**O-051 Model mice for mild glycine encephalopathy: abnormal behaviors and vulnerability to ischemic injury**

* Kure Shigeo

Department of Medical Genetics, Tohoku University School of Medicine

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**O-052 Establishment of drug screening systems for Pelizaeus-Merzbacher Disease**

* Osaka Hitoshi, Kurosawa Kenji, Iai Mizu, Yamada Michiko, Yamashita Sumimasa

1) Deivision of Neurology, Kanagawa Childrens Medical Center.

2) Division of Genetics, Kanagawa Childrens Medical Center.

3) Kanagawa Cancer Center Research Institute

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**O-053 Phenotypic characteristic of galactosemia in the post-neonatal age in India**

* Harshuti Shah, Zachary Grinspan

1) Rajvee Child Neuro and orthospine hospital, Ahmedabad, India.

2) Department of child neurology, Columbia University, New York, New York, USA

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15:50 ～ 16:50  

**Anomaly/Chromosomal disorders**

Chair: Hirofumi Ohashi, Hiroshi Tanai

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**O-054 The epidemiological study of lissencephaly using SMID data base**

* Akasaka Noriyuki, Tohyma Jun, Saito Naka, Sasaki Masayuki

1) Department of Pediatrics, Epilepsy center, Nish-Niigata Chuo National Hospital, Niigata, Japan.

2) Department of Child Neurology, National Center Hospital for Mental, Nervous and Muscular Disorders, National Center of Neurology and Psychiatry, Tokyo, Japan
O-055 Exploration of genes related to X-Linked Mental Retardation (XLMR) using MCG X-tiling array
*Honda Shozo1,2, Hayashi Shin1,2, Imoto Issei1,2, Nakagawa Eiji1, Goto Yu-ichi4, Inazawa Johji1,2
1) Department of Molecular Cytogenetics, Medical Research Institute and Graduate School of Biomedical Science, Tokyo Medical and Dental University, Tokyo, Japan, 2) CREST, JST, Saitama, Japan, 3) Division of Child Neurology National Center of Neurology and Psychiatry, Musashino Hospital, Tokyo, Japan, 4) Department of Mental Retardation and Birth Defect Research National Institute of Neuroscience, National Center of Neurology and Psychiatry (NCNP), Tokyo o, Japan

O-056 Application of in-house array-CGH for investigation and diagnosis of congenital genomic disorders
*Hayashi Shin1,2, Honda Shozo1,2, Imoto Issei1,2, Inazawa Johji1,2
1) Department of Molecular Cytogenetics, Medical Research Institute, Tokyo Medical and Dental University, Japan,
2) Core Research for Evolutional Science and Technology (CREST), Japan Science and Technology Agency

O-057 CGH-array Identified Microdeletion Involving 2p15-16.1 in a Patient With Developmental Delay
*Liang Jao-Shwann1, Shimojima Keiko1, Ohno Kouyou2, Sugiura Chitose2, Une Kouji1, Ohno Kousaku1, Yamamoto Toshiyuki1
1) International Research and Educational Institute for Integrated Medical Sciences (IREIIMS), Tokyo Women's Medical University, 2) Division of Child Neurology, Institute of Neurological Sciences, Faculty of Medicine, Tottori University, 3) Une Pediatric and Internal Medical Clinic

O-058 Impact of High Through Put Oligo-array in the field of Pediatric Neurology
*Yamamoto Toshiyuki1, Shimojima Keiko1, Tohya Jun2, Okumura Akihisa1, Maegaki Yoshihiro1, Ogune Hirokazu1
1) International Research and Educational Institute for Integrated Medical Sciences (IREIIMS), Tokyo Women's Medical University, Tokyo, Japan, 2) Department of Pediatrics, Nishi-Niigata Chuo National Hospital, Niigata, Japan, 3) Department of Pediatrics, Juntendo University School of Medicine, Tokyo, Japan, 4) Division of Pediatric Neurology, Institute of Neurological Sciences, Faculty of Medicine, Tottori University, Yonago, Japan, 5) Department of Pediatrics, Tokyo Women's Medical University, Tokyo, Japan

O-059 Maternal uniparental disomy 14 is an important differential diagnosis for floppy infants
*Saitoh Shinji, Yagyu Kazuyori, Sueta Keitaro, Asahina Naoko, Shiraishi Hideaki
Department of Pediatrics, Hokkaido University School of Medicine, Sapporo, Japan

16:50 ~ 17:50 Genetics 1

Chair: Toyojiro Matsuushi, Naofumi Kajii

O-060 Global analysis of CpG methylation status in murine fetal brain prenatally exposed to bisphenol-A
*Yaoi Takeshi1, Itoh Kyoko1, Nakamura Keiko1,2, Fushiki Shinji1
1) Department of Pathology and Applied Neurobiology, Graduate School of Medical Science, Kyoto Prefectural University of Medicine, Kyoto, Japan, 2) Department of Pediatrics, Graduate School of Medical Science, Kyoto Prefectural University of Medicine, Kyoto, Japan

O-061 A patient with benign hereditary chorea with a de novo mutation in the NNX2-1 gene
*Kato Mitsuhiro, Numakura Chikahiko, Nakamura Kazuyuki, Hayasaka Kiyoshi
Department of Pediatrics, Yamagata University School of Medicine, Yamagata, Japan

O-062 Molecular analysis of Japanese CMT Patients using multiplex ligation-dependent probe amplification
*Abe Akiko, Hayasaka Kiyoshi
Department of Pediatrics, Yamagata University Faculty of Medicine, Yamagata, Japan

O-063 Neurodevelopmental abnormalities associated with severe congenital neutropenia
*Ishikawa Nobutsune, Kobayashi Masao
Department of Pediatrics, Hiroshima University Graduate School of Biomedical Sciences, Hiroshima, Japan

O-064 The analysis of CADPS2 splicing variants and the behavioral profile in children with PDD
*Imanishi Hiroyuki, Kitayama Shinji, Matsuo Masafumi
Department of Pediatrics, Kobe University Graduate School of Medicine
O-065  HOXD gene polymorphism and II/IV digit length in the patients with or without autistic disorder
   * Sugie Yoko,  Sugie Hideo¹,  Fukuda Tokiko²,  Osawa Junko,  Suzuki Teruhiko,
   Ohzeki Takehiko
   Department of Pediatrics, Hamamatsu University School of Medicine, Shizuoka, Japan,  2) Department of Pediatrics,
   Medical University

14:30 ～ 15:05  Anomaly/Chromosomal disorders
   Chair: Takahisa Wada, Toshiyuki Yamamoto

P-001  A case of Pitt-Hopkins syndrome
   * Fukumura Shinobu¹,  Tachi Nobutada²
   1) The Department of Pediatrics, Kushiro City General Hospital, Hokkaido, Japan,  2) Sapporo Medical University
   School of Health Sciences, Sapporo, Japan

P-002  A case of congenital vertebral fusion associated with partial trisomy 22
   * Toyoshima Mitsuo,  Yonee Chihiro,  Yotsumata Kazuyuki,  Kawano Yoshifumi
   Department of Pediatrics, Graduate School of Medical and Dental Sciences, Kagoshima University, Kagoshima city, Japan

P-003  A Case of Acrystosostosis with Os odontoidaeum
   * Fukamachi Makoto¹,  Mori Atsuko¹,  Moriiuchi Hiroyuki²,  Fukuda Masafumi³
   1) Department of Pediatrics, Misakaenosono Mutsumi, The Institute for severe intellectual/motor disabled persons,
   Isahaya, Japan,  2) Department of Pediatrics, Nagasaki University School of Medicine

P-004  A sibling of premature chromatid separation syndrome with West syndrome.
   * Omata Taku¹,  Arai Hidee¹,  Tanabe Yuzo¹,²
   1) Department of Neurology, Chiba children's hospital, Chiba, Japan,  2) Soga Pediatric clinic, Chiba, Japan

P-005  A case of translocation between 16p and 17q presenting mental retardation, epilepsy, microphthalmus
   * Inoue Motoko,  Mori Masato,  Yamagita Takanori,  Momoi Mariko
   Jichi Medical University,Department of Pediatrics,Tochigi,Japan

P-006  A Female with Autistic Disorder and 45,X/46,X,idic（Y）
   * Mizuno Seiji¹,  Hirabayashi Yuu¹,  Suzuki Motomasa²,  Maruyama Kouichi²,  Kumagai Toshiyuki²
   1) Central Hospital, Aichi Human Service Center, Kasugai, Japan,  2) Central Hospital, Aichi Human Service Center,
   Kasugai, Japan

P-007  Schizencephaly in Leopard syndrome: a case report
   * Jao-shwann Liangsteven Shinnfong Peng
   1) Departments of Pediatrics, Far Eastern Hospital, Taipei, Taiwan,  2) Departments of Radiology, National Taiwan
   University Hospital, Taipei, Taiwan.

15:05 ～ 15:50  Myopathy 1
   Chair: Nobutada Tachi, Yoshiko Nomura

P-008  Ascorbic acid treatment in Charcot-Marie-Tooth disease type 1A
   * Fujii Katsunori,  Endoh Mamioko,  Tanabe Ryo,  Saito Naoki,  Kohno Yoichi
   Department of Pediatrics, Chiba University graduate School of Medicine, Chiba, Japan

P-009  Pathological analyses of Marinesco-Sjogren syndrome due to SIL1 mutation.
   * Okada Mari,  Noguchi Satoru,  Hayashi Yukiko,  Nonaka Ikuya,  Nishino Ichizo
   Department of Neuromuscular Research, National Institute of Neuroscience, National Center of Neurology and Psychiatry,
   Tokyo, Japan
P-010 Infantile myositis suggest Emery-Dreifuss muscular dystrophy with lamin A/C mutations
* Komaki Hirofumi, Hayashi Yukiko1, Kato Mitsuhiro, Sakuma Hiroshi, Saito Yoshiaki, Nakagawa Eiji, Sugai Kenji, Sasaki Masayuki, Nonaka Ikuya1, Nishino Ichizo2
1) Department of Child Neurology, National Center Hospital for Mental, Nervous and Muscular Disorders, National Center of Neurology and Psychiatry, Tokyo, Japan. 2) Department of Neuromuscular Research, National Institute of Neuroscience, National Center of Neurology and Psychiatry, Tokyo, Japan.

P-011 A case of normokalemic periodic paralysis with persistent muscle weakness
* Terazawa Daisuke, Ori Kenji, Funato Michinori, Teramoto Takahide, Fukao Toshiyuki, Kondou Naomi
Department of Pediatrics, graduate school of medicine, Gifu university, Gifu, Japan

P-012 A case report of a patient with congenital myasthenic syndrome
* Matsuoka Taro
The Department of Pediatrics, Toyonaka Municipal Hospital, Toyonaka, Japan

P-013 Childhood onset MuSK antibody positive myasthenia gravis
* Ozasa Naoko, Mabe Hiroyo, Miike Teruhisa
Department of Child Development, Kumamoto University, Kumamoto, Japan

P-014 A child of ophthalmopregic myasthenia gravis, independent from steroid treatment with tacrolimus
* Kato Takahiro, Ishikawa Aki, Ohya Kazuhiro, Tachi Nobutada
1) The Department of Pediatrics, Sapporo Medical University School of Medicine, Sapporo, Japan, 2) Sapporo Medical University School of Health Science, Sapporo, Japan

P-015 The two cases of myasthenia gravis whose chief symptom were bulbar palsy
* Keiko Nomura, Ozasa Shiro, Mitsui Koichi, Nakamura Kyoko, Kimura Shigemi, Miike Reruhisa
Department of Child Development, Kumamoto University Hospital, Kumamoto, Japan

P-016 Clinical course of childhood myasthenia gravis
* Shiraiishi Kazuhiro, Natori Chieko
The department of neuropaediatrics, Utano hospital, kyoto, Japan

15:50  〜 16:30  Myopathy 2
Chair: Yasuhiro Takeshima, Hirofumi Komaki

P-017 A novel approach to identify DMD patients for exon skipping therapy of dystrophin
* Kimura Shigemi1, Ito Kaori2, Ozasa Shiro1, Nakamura Kyoko1, Nomura Keiko1, Fujii Isao2, Mtsukura Makoto1, Mitsui Kouichi1, Miike Teruhisa1
1) Department of Child Development, Kumamoto University Graduate School, Kumamoto, Japan, 2) Laboratory of Clinical Pharmacology and Therapeutics, Faculty of Pharmaceutical Sciences, Sojo University, Kumamoto, Japan

P-018 A male patient of Duchenne muscular dystrophy with chromosome aberration
* Watanabe Kiyoko1,2, Kawashima Hisashi1, Hoshika Akinori1, Minami Narihiro3, Nishino Ichizo1
1) Department of Child Neurology, Okayama University Hospital, Okayama, Japan, 2) Department of paediatricus, Tokyo medical college hospital, Tokyo, Japan, 3) Musashi Hospital, National center of neurology and psychiatry, Tokyo, Japan

P-019 Administration of aspirin is effective for Duchenne muscular dystrophy?
* Saito Toshio, Shinho Susumu
Division of Neurology, National Hospital Organization Toneyama National Hosp

P-020 Case report of Duchenne type muscular dystrophy with tachycardia, dyspnea and sweating attack
* Nakamura Kyoko, Kimura Shigemi, Ozasa Shirou, Nomura Keiko, Mitsui Kouichi, Miike Teruhisa
Kumamoto University School of Medicine Department of Child Development
P-021 A case of massive bleeding from granulation in a Duchenne muscular dystrophy
* Ozasa Shiro1, Kimura Shigemi1, Nomura Keiko1, Nakamura Kyoko1, Mitsui Koichi1, Miike Teruhisa1
1) Department of Child Development, Faculty of Medical and Pharmaceutical Sciences, Kumamoto University Graduate School, 2) Kezawa Pediatric Clinic, Kumamoto, Japan

P-022 Financial aid for the home-use of a mechanically assisted coughing apparatus in Shiga Prefecture
* Fujii Tatsuya, Miyajima Tomoko, Kumada Tomohiro, Kimura Nobusuke, Mikuni Takayasu
Department of Pediatrics, Shiga Medical Center for Children, Moriyama, Japan

P-023 Dystrophin gene mutations in Duchenne muscular dystrophy and the electrocardiogram abnormalities.
* Takami Yuichi, Awano Hiroyuki, Okizuka Yoh, Oyazato Yoshinobu, Yagi Mariko, Takeshima Yasuhiro, Matsuo Masafumi
Department of Pediatrics, Kobe University Graduate School of Medicine, Kobe, Japan

P-024 Acute cardiac failure in the limb-girdle muscular dystrophy associated with acute gastroenteritis
* Nomura Toshihiko1, Kamimaki Isamu1, Goto Tomohide2, Sakuta Ryoichi3
1) The Department of Pediatrics, National Hospital Organization Saitama National Hospital, Saitama, Japan, 2) Department of Neurology, Tokyo Metropolitan Kiyose Children's Hospital, Tokyo, Japan, 3) Department of Pediatrics, Dokkyo Medical University Koshigaya Hospital, Saitama, Japan

16:30 ~ 17:10 Genetics 2
Chair: Toshiyuki Kumagai, Eiji Nanba

P-025 The analysis of a new oncogene-candidate in a patient with SMA type 3 and medulloblastoma
* Akioyshi Kensuke1, Suenobu Souichi1, Sonoda Kouji1, Maeda Tomoki1, Korematsu Seigo2, Izumi Tatsuro1, Ishikawa Yukitoshi2
1) Department of Pediatrics and child neurology, Oita university, Oita, Japan, 2) Department of pediatrics, National Yukamo hospital, Hokkaido, Japan

P-026 Genetic analysis in mental retardation and expansion of a research resource repository
* Nakagawa Eiji1, Takano Kyoko1, Wada Takahito1, Kubota Takeo2, Kato Mitsuhiro1, Nanba Eiji6, Saitoh Shinji1, Inazawa Johji5, Kurosawa Kenji4, Goto Yuichi1,2
1) Division of Child Neurology, National Center Hospital for Mental, Nervous and Muscular disorders, NCNP, 2) Department of Mental Retardation and Birth Defect Research, National Institute of Neuroscience, NCNP, 3) Division of Clinical and Molecular Genetics, Shinshu University School of Medicine, 4) Interdisciplinary Graduate School of Medical and Engineering, University of Yamanashi, 5) Department of Pediatrics, Faculty of Medicine, Yamagata University, 6) Research Center for Bioscience and Technology, Tottori University, 7) Department of Pediatrics, Hokkaido University Graduate School of Medicine, 8) Medical Research Institute, Tokyo Medical and Dental University, 9) Division of Genetics, Kanagawa Children's Medical Center

P-027 A Case report of a patient with Joubert syndrome due to NPHP1 gene deletion
* Wada Takahito, Hirabayashi Shin-ichi1
1) Department of Medical Genetics, Shinshu University School of Medicine, Matsumoto, Japan, 2) Department of Neurology, Nagano Children's Hospital

P-028 Juvenile Huntington Disease associated with Cerebellar hypoplasia
* Yoshinari Satoshi1,2, Hamano Shin-ichiro1,2, Minamitani Motoyuki1,2, Tanaka Manabu1, Higurashi Norimichi1,2, Eto Yoshikatu1
1) Division of Neurology, Saitama Children's Medical Center, Saitama, Japan, 2) Department of Pediatrics, The Jikei University School of Medicine, Tokyo, Japan, 3) Department for Child Health and Human Development, Saitama Children's Medical Center, Saitama, Japan

P-029 A case of mitochondrial disease who had ptosis,vomiting,fatigability and stroke like episode
* Ozaki Akiko, Takechi Tomoki
Kochi Prefectural Hata Kenmin Hospital
P-030  Effects of Vitamin E on development of three cases with Rett syndrome  
* Ieshima Atsushi, Kondo Ikuko, Kuwajima Katsuko, Yamaguchi Fumika  
Department of Pediatrics, Ibaraki Prefectural Welfare Medical Center for Disabled Children, Mito, Japan

P-031  Variation of exon 1 and promoter of the MECP2 gene in Rett syndrome  
* Ouchida Mamoru1, Yoshinaga Harumi2, Ohmori Iori3, Ohtsuka Yoko4, Oka Eiji5  
1) Department of Molecular Genetics, Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama University, Okayama, Japan, 2) Department of Child Neurology, Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama University, Okayama, Japan, 3) Department of Cellular Physiology, Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama University, Okayama, Japan, 4) Asahigawasou Ryouiku Center Jidouin

P-032  Nedd4-2 variant in a SMEI cohort  
* Kurahashi Hirokazu1,2, Kato Toru1, Kira Ryutaro1, Yasumoto Sawa1, Inoue Takahito1, Hirose Shinichi1  
1) Department of Pediatrics, Fukuoka University, Fukuoka, Japan, 2) Department of Pediatrics, Nagoya University, Nagoya, Japan, 3) Department of Pediatrics, Okayama Municipal Hospital, Okayama, Japan, 4) Department of Pediatrics, Kyushu University, Fukuoka, Japan

17:10 ～ 17:50   Genetics 3
Chair: Yukio Sawaiishi, Kiyotaka Tomiya

P-033  A family of episodic ataxia type 2 - Is reactivity to acetazolamide useful for clinical diagnosis?  
* Nagao Yoshiro, Ohsawa Maki  
Department of Pediatrics, Social Health Insurance Medical Center, Tokyo, Japan

P-034  Molecular analysis of congenital hypomyelination disease  
* Uematsu Mitsugu1, Haginoza Kazuhito1, Fukuyo Naomi1, Wakasawa Keisuke1, Tsuchiya Shigeru1, Kikuchi Atsuo1  
1) Departments of Pediatrics, Tohoku University Graduate School of Medicine, Sendai, Japan, 2) Takuto Rehabilitation Center for Children, Sendai, Japan

P-035  PLP1 Duplication Mechanism of Pelizaeus-Merzbacher Disease Analyzed by CGH-array and Fiber-FISH  
* Shimojima Keiko1, Inoue Takehiko1, Saito Kayoko2, Yamamoto Toshiyuki1  
1) International Research and Educational Institute for Integrated Medical Sciences (IREIMS), Tokyo Women's Medical University, Tokyo, Japan, 2) Division of Child Neurology, Institute of Neurological Sciences, Faculty of Medicine, Tottori University, Tottori, Japan, 3) Institute of Medical Genetics, Tokyo Women's Medical University, Tokyo, Japan

P-036  Development of a simple and highly sensitive mutation screening system by enzyme mismatch cleavage  
* Niida Yo, Tsuji Takanori  
Department of Pediatrics, Kanazawa University Graduate School of Medical Science, Ishikawa, Japan

P-037  A Case of Infantile Alexander Disease Presented with Severe Uncontrollable Epilepsy  
* Endo Mamiko1, Fuji Katsunori2, Tanabe Ryō3, Sawaishi Yukio4, Kawato Jin5, Kouno Youichi5  
1) The Department of Pediatrics, Chiba University School of Medicine, Chiba, Japan, 2) The Department of Pediatrics, Akita University School of Medicine, Akita, Japan, 3) The Department of Pediatrics, Asahi General Hospital, Asahi, Japan

P-038  Examination of the incontinentia pigmenti that developed convulsions for early infancy  
* Abe Shinpei1, Okumura Akihisa1, Hamano Shinichiro2, Tanaka Manabu3, Shihara Takashi1, Tsuru Tomohiko4, Aizaki Koichi1, Toribe Yasuhisa1, Arai Hiroshi4  
1) The Department of Pediatrics, Juntendo University School, Tokyo, Japan, 2) The Division of Neurology, Saitama Children's Medical Center, Saitama, Japan, 3) The Division of Neurology, Gunma Children's Medical Center, Gunma, Japan, 4) The Department of Pediatrics, Matsudo City Hospital, Chiba, Japan, 5) The Division of Pediatric Neurology, Osaka Medical Center and Research Institute, Osaka, Japan, 6) The Division of Pediatric Neurology, Morinomiya Hospital, Osaka, Japan
P-039 Early neurological characteristics of 3 patients in a family with the severe type of MCT8 deficiency
* Sawashita Yuko1, Yanai Tamami1, Watanabe Yasuhiro1, Hirayama Aya2, Makata Masahiro2
1) Department of Pediatrics, Akita University School of Medicine, Akita, Japan, 2) Akita Prefecture Rehabilitation Center for Disabled Children, Akita, Japan

P-040 Long surviving case of osteogenesis imperfecta type II with COL1A2 gene anomaly
* Hachiya Yasuo1, Hayashi Masaharu2, Atsumi So1, Kubota Masaya1
1) The Department of Pediatrics, Tokyo Metropolitan Fuchu Medical Center for the Disabled, Tokyo, Japan, 2) The Department of Clinical Neuropathology, Tokyo Metropolitan Institute for Neuroscience, Tokyo, Japan, 3) The Department of Neurology, National Center for Child Health and Development, Tokyo, Japan

14:30 ~ 15:15 Metabolic disorders 1
Chair: Fujita Tatsuya, Koji Inui

P-041 Autopsy case of MELAS with acute pancreatitis
* Yamashita Sumimasa, Takagi Atsushi, Tsuji Megumi, Samejima Kiyoko, Iai Mizue, Osaka Hitosi, Yamada Midhiko
Division of Child Neurology, Kanagawa Children’s Medical Center, Yokohama, Japan

P-042 A case of Leber disease with mitDNA11778 mutation which became clear after the diagnosis of SCD
* Tomita Sunao1,2, Hoshino Ai1, Hanafusa Yukiko1, Shigetomo Rituko1, Kumada Satoko1, Kurihara Eizi2
1) Neuropediatrics of Tokyo megapololis Neurological Hospital,Tokyo,Japan, 2) Pediatrics of Tokyo Metropolitan Hachioji Children’s Hospital,Tokyo,Japan

P-043 Increase of P1 wave amplitude of visual event related potential in childhood cerebral form of ALD
* Furushima Wakana1,2, Inagaki Masumi1, Gunji Atsuko1, Inoue Yuki1, Kaga Makiko1
1) Department of Developmental Disorders, National Institute of Mental Health, National Center of Neurology and Psychiatry, Tokyo, Japan, 2) Department of Pediatrics, Tokyo Medical and Dental University, Tokyo, Japan

P-044 A case of Canavan disease : long term follow up of a case in Japanese female
* Mizuguchi Koichi1, Hoshino Hideki1, Nagasawa Tetsuro1, Hamaguchi Hiroshi2, Kubota Masaya1
1) Division of Neurology, National Center for Child Health and Development, 2) Tokyo Metropolitan Higashiyamato Medical Center for the Severly Disabled

P-045 A case of obstructive respiratory distress of mucopolysaccaridosis type 2 improved by nppv
* Matsui Shuji, Shiki Toshishi, Taketi Nobuyuki, Tyou Hiroyuki, Funahashi Masuko, Suzuki Yasuyuki
Tokyo Childrens Rehersal Hospital, Tokyo, Japan

P-046 Enzyme replacement therapy in a female case of mucopolysaccaridosis type 2 (Hunter syndrome)
* Sato Tatsuharu1, Honda Ryoko1, Imamura Yoshihiko1, Turu Akira1, Matamoto Tadashi1, Morii Hiroyuki2
1) The Department of Pediatrics, Nagasaki University School of Medicine, Nagasaki, Japan, 2) The Department of Nursing, Nagasaki University School of Health Sciences, Nagasaki, Japan, 3) Nagasaki National Hospital, Nagasaki, Japan

P-047 Histological Evaluation of Hunter Syndrome Patient Brain after Cord Blood Stem Cell Transplantation
* Araya Ken, Kitai Yukihiro, Hoshino Natsuko, Tominaga Koji, Shimono Kuriko, Okinaga Takeshi, Mohri Ikuko, Sakai Norio, Taniike Masako, Ozono Keiichi
Department of Pediatrics, Osaka University Graduate School of Medicine, Osaka, Japan

P-048 A case of Pompe disease (infantile onset form) treated with recombinant human acid alpha-glucosidase
* Akagawa Mie, Awaya Tomonari, Nodomi Seihiro, Shibata Minoru, Yamanaka Yasunari, Kato Takeo, Yorifuji Toru, Nakahata Tatsutoshi
Department of Pediatrics, Graduate School of Medicine, Kyoto University, Kyoto, Japan
P-049 The efficacy of enzyme replacement therapy in a case of glycogen storage type II disease
*Toribe Yasuhisa, Mogami Yukiko, Yanagihara Keiko, Suzuki Yasuhiro
Division of Pediatric Neurology, Osaka Medical Center and Research Institute for Maternal and Child Health, Osaka, Japan

15:15 ~ 16:05 Metabolic disorders 2

Chair: Nobuyuki Shimozawa, Hitoshi Sakuraba

P-050 Mutation analysis and response to riboflavin therapy in Taiwanese MADD patients
*Wen-chen Liang, Ohkuma Aya, Goto Kanako, Hayashi Yukiko K. Yukiko K., Yuh-jiyh Jong
Ichizo Nishino
1) The division of pediatric neurology, Department of pediatrics, Kaohsiung medical university hospital, Kaohsiung,
2) The department of neuromuscular research, National institute of neuroscience, National center of neurology and psychiatry, Tokyo, Japan. 3) The department of neuromuscular research, National institute of neuroscience, National center of neurology and psychiatry, Tokyo, Japan
4) The department of neuromuscular research, National institute of neuroscience, National center of neurology and psychiatry, Tokyo, Japan. 5) The division of pediatric neurology, Department of pediatrics, Kaohsiung medical university hospital, Kaohsiung, Taiwan. 6) The department of neuromuscular research, National institute of neuroscience, National center of neurology and psychiatry, Tokyo, Japan

P-051 Analysis of cerebral molecular species of phosholipids in D-bifunctional protein deficiency
*Saitoh Makiko, Yamashita Sumimasa, Itoh Masayuki, Mizuguchi Masashi
1) Department of developmental medical sciences, Graduate school of Medicine, University of Tokyo, Japan.
2) Department of Neurology, Kanagawa Prefectural Children Medical Center. 3) Department of Mental Retardation and Birth Defect Research, National Institute of Neuroscience, National Center Neurology and Psychiatry

P-052 An infantile case of rapidly progressive vanishing white matter disease during half a year
*Hoshino Ai, Tomita Sunao, Kumada Satoko, Hanafusa Yukiko, Nigo Ayako, Okumura Sayaka, Kurihara Eiji, Yamamoto Toshiyuki
1) The Department of Neuropediatrics, Tokyo Metropolitan Neurological Hospital, Tokyo, Japan. 2) The Department of Pediatrics, Tokyo Metropolitan Fucyu Hospital, Tokyo, Japan. 3) The International Research and Education Institute for Integrated Medical Sciences, Tokyo Wemen's Medical University Hospital. Tokyo, Japan

P-053 Irritability in a patient with Megalencephalic leukoencephalopathy with subcortical cysts
*Amemiya Kaoru, Kashii Hirohumi, Mizuno Yoko, Suzuki Riina, Koide Ayaka, Kubota Masaya, Ide Syuhei
1) Tokyo Metropolitan Hachioji Children's Hospital. 2) TOBU RYOIKU CENTER

P-054 A case with congenital disorders of glycosylation
*Ohto Tatsuyuki, Enokizono Takashi, Iwasaki Yoko, Hirai Naomi, Tanaka Ryuta, Sasaki Masayuki, Ohno Kohsaku, Yuasa Isao, Kamota Tomohiro
1) Department of Pediatrics, Institute of Clinical Medicine, University of Tsukuba, Tsukuba, Japan. 2) Children's Center for Health and Development, Yokohama City Tobu Hospital, Yokohama, Japan. 3) Department of Child Neurology, National Center Hospital for Mental, Nervous and Muscular Disorders, National Center of Neurology and Psychiatry, Kodaira, Tokyo. 4) Division of Child Neurology, Faculty of Medicine, Tottori University, Yonago, Japan. 5) Division of Legal Medicine, Faculty of Medicine, Tottori University, Yonago, Japan

P-055 An autopsy case of congenital disorders of glycosylation with recurrent pulmonary hemorrhage
*Matsuo Muneaki, Sasaki Kazuya, Maeda Toshiyuki, Tajima Daisuke, Ohno Kousaku
1) Department of Pediatrics, Faculty of Medicine, Saga University. 2) Department of Child Neurology, Institute of Neurological Sciences, Faculty of Medicine, Tottori University

P-056 Mass spectrometry for rapid diagnosis of congenital disorders of glycosylation type Ia
*Fujiiwa Shinichi, Okamoto Nobuhiko, Uetake Kimiaki, Onodera Takashi, Yagyu Kazuyori, Sueta Keitaro, Asahina Naoko, Shiraishi Hideaki, Saitoh Shinji
1) Department of Pediatrics, Obihiro Kosei Hospital, Obihiro, Japan. 2) Osaka Medical Center and Research Institute for Maternal and Child Health, Osaka, Japan. 3) Department of Pediatrics, Date Red Cross Hospital, Date, Japan. 4) Department of Pediatrics, Hokkaido University School of Medicine, Sapporo, Japan
The 50th meeting of the Japanese Society of Child Neurology ● Day 2 (May 29)

P-057 The research of Menkes disease with symptoms and treatments
* Ozawa Hiroshi1, Kodama Hiroko1
1) Institution for persons with severe motor and intellectual disabilities, Tokyo, 2) Department of Pediatrics, Teikyo University, Tokyo, Japan

P-058 A case of Methylmalonic acidemia developed symptoms of central nerve system.
* Yoshii Keisuke1,2, Naiki Yasuhiro1, Horikawa Reiko1
1) Division of Endocrinology and Metabolism, National Center for Child Medical Health and Development, Tokyo, Japan, 2) Department of Pediatrics, Tokyo Woman's Medical University Hospital, Tokyo, Japan

P-059 A case of biotin deficiency which was suspected of multiple carboxylase deficiency
* Kakizawa Hiroko1, Itomi Kazuya1, Kobayashi Hironori1, Suzuki Yoiti1, Yamaguchi Seiji2
1) Aichi Children's Hospital Health And Medical Center, Aichi, Japan, 2) Department of Pediatrics, Shimane University Faculty of Medicine, Izumo, Japan, 3) Department of Public Health Chiba University Graduate School of Medicine, Chiba, Japan

16:05 ~ 16:45 Vascular disorders
Chair: Hiromi Sato, Akira Oka

P-060 Transient spasms as early MR angiographic findings in 3 children with convulsive status
* Sugiyama Nobuyoshi, Sasaki Mariko, Yokoyama Jyun-ichi, Miyashita Yoshihiro
Department of Pediatrics, Tokai University School of Medicine, Kanagawa, Japan

P-061 A boy with congenital muscular dystrophy revealed abnormal EEG due to CSVT on steroid therapy
* Harashima Chie, Teduka Junichiro, Ohno Yoichiro
Department of Pediatrics, Fukuoka National Hospital, Fukuoka City, Fukuoka, Japan

P-062 Three Cases of Cerebral Multiple Cavernous Hemangiomas (Cerebral Cavernous Malformations)
* Maniwa Satoshi, Nakano Kousuke
Department of pediatrics, Matsuyama Red Cross Hospital, Matuyama, Japan

P-063 A Case of Anterior Choroidal Artery Infarction Associated with Intracranial ICA Dissection
* Kashii Hirofumi1, Mizuno Yoko1, Amemiya Kaoru1, Suzuki Riina1, Koide Ayaka1, Taniguchi Makoto1, Hayashi Masaharu1, Tanuma Naoyuki1, Miyata Rie2
1) Department of Pediatrics, Tokyo Metropolitan Hachioji Children's Hospital, 2) Department of Neurosurgery, Tokyo Metropolitan Neurological Hospital, 3) The Department of Clinical Neuropathology, Tokyo Metropolitan Institute for Neuroscience

P-064 Reversible cerebral vasoconstriction syndrome with leptomeningeal high signal intensity on MRI
* Yoshioka Seiichiro
Department of Pediatrics, Kusatsu General Hospital, Shiga, Japan

P-065 Three cases of Neurofibromatosis type1 associated with Moyamoya syndrome
* Nakano Kyoko, Niizeki Masae, Yoshikawa Hideto
Department of Neurology, Miyagi Children's Hospital, Sendai, Japan

P-066 A case of the brainstem infarction presented with Claude syndrome
* Fujita Takako, Ninomiya Shinya, Nakamura Noriko, Ideguchi Hiroshi, Inoue Takahito, Yasumoto Sawa, Hirose Sinici
Department of Pediatrics, Fukuoka University School of medicine, Fukuoka, Japan

P-067 Case report : 2 patients with Cutis Marmorata Telangiectasia Congenita
* Gotoh Chika, Akiyoshi Kensuke, Maeda Tomoki, Suenoobu Souichi, Korematsu Seigo, Izumi Taturou
Department of Brain and Nerve Science Division of Pediatrics and Child Neurology, University of Oita, Oita, Japan
16:45 ~ 17:30  Tumor/Neurosurgery
Chair: Hisashi Kawakaki, Sumimasa Yamashita
P-068  Lateral ventricle choroid plexus papilloma of an infant with hypernatremia
* Tanda Koichi1, Ouchi Kazutaka1, Nabeshima Kanae1, Komatsu Hiroshi1, Nakajima Humiaki1, Inoue Yasuo2, Kawarabuki Kentaro3, Shirato Mitsuru2, Hori Koh2
1) The Department of Pediatrics, NHO Maizuru Medical Center, Kyoto, Japan, 2) The Department of Neurosurgery, NHO Maizuru Medical Center, Kyoto, Japan

P-069  A case of ganglioglioma in basal ganglia: rare location and symptom
* Sato Atsushi, Takahashi Kan, Mimaki Masakazu, Saito Makiko, Oka Akira, Mizuguchi Masashi
Department of Pediatrics, Graduate School of Medicine, University of Tokyo

P-070  Noninvasive respiratory management after resection of brainstem tumor: A case report
* Otani Anna
The Department of Pediatrics, Teine Keijinkai Hospital, Hokkaido, Japan

P-071  A case of infatile spasms with hypothalamic hamartoma
* Yano Tamami, Sawaishi Yukio, Takahashi Tsutomu
The Department of Medicine, University of Akita, Akita, Japan

P-072  A girl with ependymoblastoma with sustained arrest of tumor growth by interferon therapy
* Mutoh Kozo, Miki Naoki
Department of Pediatrics, Shimada Municipal Hospital, Shimada, Japan

P-073  Two cases of tuberous sclerosis treated by surgical procedures
* Watanabe Kenji1, Sano Nozomi2, Oosako Yutaka3, Yatusiro Kazutaka3
1) The Department of Pediatrics, Minami Kyushu Hospital, Kagoshima, Japan, 2) Kokubu Seikyou Hospital, 3) Department of Neurosurgery Kagoshima University

P-074  A case of leukoencephalopathy with cerebral atxia after the chemotherapy of acute leukemia (AML)
* Ninomiya Shinya1, Inoue Takahito1, Yonekura Michitaka1, Fujita Takako1, Ihara Yukiko1, Tomonou Yuiko1, Nakamura Noriko1, Ideguchi Hiroshi1, Yasumoto Sawa1, Sakiyama Michiyo2, Hirose Shinichi2
1) The Department of Pediatrics, Fukuoka University, Fukuoka, Japan, 2) National Kyushu Cancer Center, Fukuoka, Japan

P-075  Two cases with hematologic neoplasm manifesting CNS symptom
* Enokizono Taktashi1, Fukushima Takashi1, Ohto Tatsuyuki1, Tanaka Ryuta1, Ohta Masayasu1, Kudo Kazuko2, Kamota Tomohiro3
1) Department of Pediatrics, Institute of Clinical Medicine, University of Tsu kuba, Tsukuba, Japan, 2) Department of Pediatrics, Toride Kyodo General Hospital, Ibaraki, Japan

P-076  A case of desmoid tumor in a Lowe syndrome patient with tracheostomy
* Mitsui Koichi, Ozasa Shiro, Nakamura Kyoko, Nomura Keiko, Kimura Shigemi, Miike Teruhisa
Child Development,Kumamoto University Hospital

17:30 ~ 18:00  Mental disorders
Chair: Akemi Tomoda, Junichi Furusho

P-077  Dexamethasone suppression test to anorexia nervosa
* Arakawa Chikako, Imai Yuki, Endo Ayumi, Kohira Ryutaro, Fujita Yukihiko
Department of Pediatrics Nihon University School of Medicine

P-078  Issues with initial management of hospitalized patients with anorexia nervosa
* Itabashi Hisashi, Takesita Eri, Ootani Ryuko, Koike Makiko, Mazaki Kaoru, Shimamura Keichi, Murakami Nobuyuki, Sakuta Ryouti, Nagai Toshiro
Dokkyo University School of Medicine Kosigaya Hospital, Saitama, Japan
P-079  Malnutrition made adjustment disorder in younger twin but not in older with autistic disorder  
*Nara Chieko*, Yokoyama Hiroyuki, Hirose Mieko, Wakasawa Keisuke, Tsuchiya Shigeru  
1) Department of Pediatrics, Tohoku University School of Medicine, 2) School of Nursing, Yamagata University Faculty of Medicine

P-080  Three cases of social anxiety disorder developed in childhood  
*Yokoyama Hiroyuki*, Hirose Mieko, Nara Chieko, Wakasawa Keisuke, Haginoya Kazuhiro, Tsuchiya Shigeru, Inuma Kazue  
1) School of Nursing, Yamagata University Faculty of Medicine, Yamagata, Japan, 2) Department of Pediatrics, Tohoku University School of Medicine, Sendai, Japan, 3) Takuto Rehabilitation Center for Children, Sendai, Japan, 4) Ishinomaki Red Cross Hospital, Ishinomaki, Japan

P-081  An intractable epilepsy case with repeated forced normalization  
*Hirose Mieko*, Yokoyama Hiroyuki, Haginoya Kazuhiro, Kikuchi Atsuo, Nakayama Tojo, Uematsu Mitsugu, Inuma Kazue, Tsuchiya Shigeru  
1) Department of Pediatrics, Tohoku University Hospital, Sendai, Japan, 2) Department of Nursing, Yamagata university of medicine, Yamagata, Japan, 3) Takuto medical and rehabilitation center, Sendai, Japan, 4) Ishinomaki Red Cross Hospital, Ishinomaki, Japan

P-082  4 patients with Munchausen syndrome by proxy who appealed for neurological symptoms  
*Miwa Mami*, Nakamura Yukiko, Matsuda Hiroo, Bassyo Fumio, Kato Masae  
1) Department of Pediatrics, Kyorin University Hospital, 2) Medical welfare counselor’s office, Kyorin University Hospital, 3) Syukutoku university.

14:30 ～ 15:15  Developmental disorders （diagnosis）  
Chair: Akihiro Yasuhara, Yoshiaki Saito

P-083  The simple judgement of motor difficulty by medical interview and soft neurological signs  
*Kashiwagi Mitsuru*, Hashimoto Ryosaku, Suzuki Shuhei  
Department of Developmental Brain Science Osaka Medical College

P-084  A clinical analysis of schoolchildren with dysgraphia  
*Kawatani Masao*, Nakai Akio, Hiratani Michio  
1) The Department of Pediatrics, Faculty of Medical Sciences, University of Fukui, Fukui, Japan, 2) Hiratani Clinic for Developmental Disorders of Children, Fukui, Japan

P-085  Cognitive function in patients with Williams syndrome  
*Sunahara Mariko*, Inoko Kayo, Osawa Makiko  
1) Dept of Pediatrics, Tokyo Women’s Medical University, Tokyo, Japan, 2) Tokyo institute Psychiatry

P-086  Developmental changes of visuo-spatial abilities and Kanji copying in people with Williams syndrome  
*Nakamura Miho*, Muzuno Seiji, Kumagai Toshiyuki, Matsumoto Akiko  
1) Institute for Developmental Research, Aichi Human Service Center, Kasugai, Aichi, Japan, 2) Central hospital, Aichi Human Service Center, Kasugai, Aichi, Japan, 3) Kobato Gakuen, Aichi Human Service Center, Kasugai, Aichi, Japan

P-087  The developmental processes of maternal attachment in mothers of children with AD/HD  
*Mano Shoko*, Uno Hiroyuki, Horiuchi Fumie  
1) Department of Nursing, Ehime Prefectural University of Health Sciences, Ehime, Japan, 2) Department of Special Support Education, Hyogo University of Teacher Education, Hyogo, Japan, 3) Department of Neuropsychiatry, School of Medicine, Ehime University, Ehime, Japan

P-088  Sleep study of pre/post Summer Treatment Program in children with AD/HD  
*Iwasaki Mizue*, Matsuishi Toyoiro, Iemura Akiko, Oya Takashi, Izuka Chiho, Nakasima Masayuki, Nagamitsu Shinichiro, Yamashita Yushiro  
1) Research Institute of Science and Technology for Society, Japan Science and Technology Agency, 2) Department of Pediatrics and Child Health, Kurume University school of Medicine, Japan
P-089 The fact of adult ADHD
* Nishimaki Atsuko, Miyao Masutomo, Okayama Makiko
Division of Parent-child Psychological Medicine, National Center for Child Health and Development

P-090 A girl of Asperger syndrome diagnosed with the outbreak of anorexia nervosa
* Tomonoh Yuko1,2, Inoue Narito2, Ihara Yukiko1,2, Kanaumi Takeshi1,2, Inoue Tkahito3, Yasumoto Sawa1, Hirose Shinichi1
1) Department of Pediatrics, Nakatsu municipal hospital, Nakatsu, Oita, Japan, 2) Inoue children's clinic, Nakatsu, Oita, Japan

P-091 Comparison of Emotion in Patients with Asperger Syndrome and AD/HD
* Yamashiro Dai, Kanemura Hideaki, Kaga Yoshimi, Aoyagi Kakuro, Sugita Kanji, Aihara Masao
Department of Pediatrics, Faculty of Medicine, University of Yamanashi, Yamanashi, Japan

15:15 ~ 15:45 Developmental disorders (examination 1)
Chair: Tatsuya Ogin, Michiko Sugama

P-092 Developmental Change of Qualitative scores of The Boston Qualitative Scoring System (BQSS)
* Nakano Kousuke1, Ogino Tatsuya2, Watanabe Kiyoko1, Takeuchi Akihito1, Oka Makio1, Ohtsuka Yoko3
1) Department of Pediatrics, Matsuyma Red-Cross Hospital, Matsuyma, Japan, 2) Faculty of Children Studies, Department of Children Studies, Chugokukagakuen University, 3) Department of Child Neurology, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, 4)

P-093 The McGurk effect in developmental disorder
* Morita Kouji, Sakurai Syunsuke, Ichihashi Izumi, Tatsuno Masaru, Itahashi Kazuo
Department of Pediatrics, Showa University School of Medicine, Tokyo, Japan

P-094 Reliability and valiability of the observation of pupillary grating response in normal adults
* Sakai Shinya1, Hirayama Kazumi2, Kato Mitsuhiro3, Saitoh Shinji1, Sakai Naoko1
1) Department of Occupational Therapy, Hokkaido University School of Health Sciences, Sapporo, Japan, 2) Department of Behavioral Neurology and Cognitive Neuroscience, Tohoku University Graduate School of Medicine, Sendai, Japan, 3) Department of Pediatrics, Yamagata University School of Medicine, Yamagata, Japan, 4) Department of Pediatrics, Hokkaido University Graduate School of Medicine, Sapporo, Japan, 5) Midorigaoka Ryoikuen Hospital and Home for Persons with Severe Motor and Intellectual Disabilities, Sapporo, Japan

P-095 The observation of pupillary grating response for children with cerebral visual impairment
* Sakai Shinya1, Hirayama Kazumi2, Kato Mitsuhiro3, Saitoh Shinji1, Sakai Naoko1, Seiwa Chizuru4, Sudo Mutsuko5, Nio Eiko6, Shiraiishi Hideaki4, Takayanagi Masaru8, Kobayashi Yasuko9
1) Department of Occupational Therapy, Hokkaido University School of Health Sciences, Sapporo, Japan, 2) Department of Behavioral Neurology and Cognitive Neuroscience, Tohoku University Graduate School of Medicine, Sendai, Japan, 3) Department of Pediatrics, Yamagata University School of Medicine, Yamagata, Japan, 4) Department of Pediatrics, Hokkaido University Graduate School of Medicine, Sapporo, Japan, 5) Midorigaoka Ryoikuen Hospital and Home for Persons with Severe Motor and Intellectual Disabilities, Sapporo, Japan, 6) Department of Pediatrics, Yamagata prefecture Comprehensive Rehabilitation and Education Center, Kaminoyama, Japan, 7) Department of Pediatrics, Ekok Ryoikuen Hospital and Home for People with Severe Motor and Intellectual Disabilities, Sendai, Japan, 8) Department of Pediatrics, National Sendai Hospital, Sendai, Japan, 9) Department of Pediatrics, Nishitaga National Hospital, Sendai, Japan

P-096 Intervention of Kanji writing in 2 Japanese dyslexic children
* Nakamura Masako1,2, Inagaki Masumi1, Kaga Makiko1
1) Department of Developmental Disorders, National Institute of Mental Health, National Center of Neurology and Psychiatry, 2) Department of Otolaryngology, University of Tokyo
P-097 Developmental changes of rapid automatized naming (RAN) of children in elementary school
* Kobayashi Tomoka1,2, Inagaki Masumi1, Gunji Atsuko1, Yatabe Kiyomi1, Kaga Makiko3, Gotoh Takaaki3, Koike Toshihide1
1) Department of Developmental Disorders, National Institute of Mental Health, NCNP, Kodaira, Japan, 2) Department of Pediatrics, Tokyo Metropolitan Fuchu Medical Center for the Disabled, Tokyo, Japan, 3) Tokyo Gakugei University, Tokyo, Japan.

15:45 ~ 16:15 Developmental disorders (examination 2)
Chair: Hideaki Kanemura, Tohshin Go

P-098 Decreased oxygenated hemoglobin during a CPT task in the prefrontal cortex of children with AD/HD
* Inoue Yuki1, Inagaki Masumi1, Gunji Atsuko1, Shinoda Haruo2, Kaga Makiko3
1) Department of Developmental Disorders, National Institute of Mental Health, National Center of Neurology and Psychiatry, Tokyo, Japan, 2) Faculty of Psychology, Rissho University, Tokyo, Japan

P-099 Neural correlates for processing of situationally discordant behavior; an fMRI study
* Wakusawa Keisuke1,2, Tsuchiya Shigeru1, Yokoyama Hiroyuki1, Kawashima Ryuta2, Sugiura Motoaki4, Sassa Yuko2, Hyeonjeong Jeong2
1) Department of Pediatrics, Tohoku University Graduate School of Medicine, 2) Department of Functional Brain Imaging, Institute of Development, Aging and Cancer, Tohoku University, Sendai, Japan, 3) School of Nursing, Yamagata University Faculty of Medicine, Yamagata, Japan, 4) Department of Cerebral Research, National Institute for Physiological Sciences, Okazaki, Japan

P-100 Characteristic prefrontal activation from visual and auditory stimuli in autism spectrum disorders.
* Narita Naoko1, Narita Masaaki2
1) Institute of Education, Bunkyo University, Koshigaya, Japan, 2) Graduate school of Medicine, Mie University, Tsu, Japan

P-101 MR imaging of Autism-combination of DTI&3D-MRS
* Hanaoka Shigeru
Tokyo Metropolitan Kita Medical Rehabilitation Center, Jonan Branch, Tokyo, Japan

P-102 Frontal lobe's function of autism -analysis of brain 1H-MR Spectroscopy
* Fujii Emiko1, Mori Kenji1, Miyazaki Masahito1, Hashimoto Toshiaki2, Harada Masafumi3
1) Department of Pediatrics, Tokushima University, Tokushima, Japan, 2) Department of Education for the Disabled, Faculty of School Education, Naruto University of Education, Naruto, Tokushima, Japan, 3) Department of Radiologic Technology, School of Health Science, The University of Tokushima, Tokushima, Japan

P-103 Features of sleep spindles recorded in children with pervasive developmental disorders
* Kimura Ikumi1, Miyao Masutomo2
1) Department of child neurology, Metropolitan Tama-Ryoikuen Institution for handicapped children, Tokyo, Japan, 2) Department of developmental psychoneurology, National Center for Child Health and Development

16:15 ~ 17:00 Developmental disorders (support)
Chair: Masaki Ohno, Kazuya Itomi

P-104 The relations between the age at the initial visit and its prognosis on our developmental clinic
* Sawai Chiiro1, Sakue Yuko1, Iwami Mika1, Okada Masako1, Nishikura Noriko1,2, Yoshioka Seiichiro1,2,3, Takano Tomoyuki1, Takeuchi Yoshihiro1
1) Department of Pediatrics, Shiga University of Medical Science, Shiga, Japan, 2) Ishibe Medical Center, Shiga, Japan, 3) Kusatsu General Hospital, Shiga, Japan

P-105 Study of children with the developmental disabilities in Fukuoka
* Gondo Kenjiro1, Takemoto Megumi1, Hanai Toshio1, Shionaga Junko2, Miyazaki Chiaki1
1) The Division of Pediatric Neurology, Fukuoka Children's Hospital, Fukuoka Medical Center for the Disabled, 2) Fukuoka Municipal Welfare Center for The Disabled, 3) Fukuoka-West Rehabilitation Center for Children
P-106 Questionnaire for medical consultation of pervasive developmental disorders to medical institutions  
* Nishimura Satoko  
Department of Pediatrics, Kibougaoka Rehabilitation Center for handicapped children, Gifu, Japan

P-107 Management of the behavior problems of adolescents with HFPDD in their family and community.  
* Maeda Yosuke, Nishimaki Atsuko, Miyao Masutomo  
National Center for Child Medical Health and Development

P-108 Investigation and care of children with pervasive developmental disorder complain of school refusal  
* Miyachi Taishi1, Ishikawa Michiko2, Iguchi Toshiyuki2,3, Imaeda Masayuki2,4, Asai Tomoko2,5  
1) Osaka-hamamatsu joint center for mental development, 2) Department of neonatology and pediatrics nagoya city university medical school, 3) Hoshigaoka maternity hospital, 4) North district care center for disabled children, 5) Nagoya city child welfare center

P-109 Parent's needs on medical emergency of handicapped children  
* Nagai Toshiaburo1, Kubayashi Chika1, Tagawa Tetsuzo2,3, Taniike Masako2,4, Imaishi Hidenori2,5, Arai Hiroshi2,6, Tanabe Takuya2,7, Yabuta Reiko2,8, Tanaka Jyunko2,9, Nishida Masaru2,9  
1) Osaka University, Graduate School of Medicine, Course of Health Science, 2) Osaka Pediatric association, 3) Osaka Kosei Nenkin Hospital, 4) Osaka University, Graduate School of Medicine, 5) Imaishi Children's Clinic, 6) Morinomiya Hospital, Pediatric Neurology, 7) Hirakata City Hospital, Department of Pediatrics, 8) Itami City Hospital, Department of Pediatrics, 9) Tanakakitaumedo Clinic, 10) Hirakataryoikuen

P-110 Analysis of medical information communicated in the field of developmental disorder practices  
* Horiguchi Toshihiro1, Akiyama Chieko2, Kon Kaori1  
1) Department of Social Psychiatry, National Institute of Mental Health, National Center of Neurology and Psychiatry, Tokyo, Japan, 2) Akiyama Children's Clinic, Tokyo, Japan, 3) Kaishundou-Kaori Children's Clinic, Tokyo, Japan

P-111 Support by full time coordinator for patients with tuberous sclerosis  
* Minegishi Machiko, Shimosato Sachiko, Takahashi Takao  
The Department of Pediatrics, Keio University, School of Medicine, Tokyo, Japan

P-112 The assessment of motor and process Skills on schoolchildren with developmenal disorders  
* Hayashi Yuko  
Faculty of Health and Welfare,Prefectural University of Hiroshima,Mihara,Japan

17:00 ～ 17:45       Developmental disorders （treatment）

Chair: Asayo Ishizaki, Hiroshi Koide

P-113 The developmental course of handicapped children with MR and their rehabilitation process  
* Okoshi Yumi, Chou Hiroyuki, Funahashi Masuko, Suzuki Yasuyuki  
Department of Pediatrics, Tokyo Children's Rehabilitation Hospital, Tokyo, Japan

P-114 Music Social Skills Training for Patients with Developmental Disorders  
* Go Tohshin  
Department of Infants' Brain and Cognitive Development, Tokyo Women's Medical University, Tokyo, Japan

P-115 Parent-training program for nursery staff.  
* Tandou Tomoko1, Aoyagi Kakuro2, Hatakeyama Kazuo1, Aihara Masao2  
1) Department of Pediatrics, Akebono medical welfare center, Yamanashi, Japan, 2) Department of Pediatrics, Faculty of Medicine, University of Yamanashi, Yamanashi, Japan

P-116 Roles of pediatric neurologists in special supportive teams for students with developmental disorder  
* Katsumori Hiroshi, Shirakawa Seigo  
Department of Pediatrics, Tokyo Rinkai Hospital, Tokyo, Japan
P-117 Collaboration with school for child in educational needs through consultation by child neurologist
* Nakashima Masayuki, 1, 2, Yamashita Yashiro, 1, Matsuishi Toyoiro 2
1) Department of Neonatology, Maternal and Child's Health Medical Center, St.Mary's Hospital, 2) Department of Pediatrics and Child Health Kurume University School of Medicine

P-118 Problems in the methylphenidate management of attention-deficit-hyperactivity disorder
* Isumi Hiroshi, 1, Kajimoto Madoka, 1, Ichiyama Takashi, 1, Hurukawa Susumu 2
1) Hagi Civil Hospital, Yamaguchi, Japan, 2) Department of Pediatrics Yamaguchi University School of Medicine, Ube, Yamachi, Japan

P-119 A child with ADHD who required individualized program and methylphenidate in summer treatment program
* Yamashita Yushiro, Iizuka Chiho, Ohya Takashi, Nakashima Masayuki, Nagamitsu Shin-ichiro, Matsuishi Toyoiro
Department of Pediatrics and Child Health, Kurume University School of Medicine

P-120 Efficiency of aripiprazole
* Ogawara Sayuri, Miyao Masutomo
National Center for Child Health and Development

P-121 A trial of diagnosis and longitudinal observation of children with autistic disorder
* Nagao Yasuko, 1, Araki Atsushi, 1, Kaneko Kazunari, 2 Kuniyoshi Kyouko, 3 Kusumoto Kenji 4
1) Department of Pediatrics Kansai Medical College, Osaka, Japan, 2) Department of Pediatrics Kansai Medical College, Osaka, Japan, 3) Department of Plastic and Reconstructive surgery Kansai Medical College, Osaka, Japan

14:30 ~ 15:05 Neonatology 1
Chair: Seiichi Sugama, Hiromi Koizumi

P-122 Neurobehavioral outcomes of preterm children born at fewer than GA25 weeks without brain lesions
* Sugama Seiichi
National Center for Child Health and Development, Division of Interdisciplinary Medicine

P-123 3 cases of severe congenital myotonic dystrophy
* Otsuka Harumi, 1 Suzuki Yasuhiro, 1 Iwamatsu Toshiyuki, 1 Tanabe Ryosuke, 1 Mizoguchi Eriko, 1 Sasaki Kao, 1 Saitou Kayoko, 1 Oosawa Makiko, 1
1) Department of Neonatology, Chiba Municipal Kainin Hospital, Chiba, Japan, 2) Department of Pediatrics, Graduate of Medical University of Chiba, Chiba, Japan, 3) Department of Pediatrics, Tokyo Women's Medical University, Tokyo, Japan, 4) Department of Pediatrics, Tokyo Women's Medical University Yachiyo Medical center, Chiba, Japan, 5) Institute of Medical Genetics, Tokyo Women's Medical University, Tokyo, Japan

P-124 Prediction of West syndrome of the infants with HIE by neonatal EEG after 2weeks of age
* Kato Toru, 1, Tii Takeshi, 1, Hayakawa Fumio, 1 Kidokoro Hiroshi, 1 Kubota Tetsuo, 1 Suzuki Motomasa, 1 Maruyama Kichi, 1 Natsume Jun, 1 Okumura Akihisa, 1 Watanabe Kazuyoshi 1
1) Okazaki City Hospital, Department of Pediatrics, Okayama, Japan, 2) Anjo Kosei Hospital, Department of Pediatrics, Anjo, Japan, 3) Aichi Prefectural Colony Central Hospital, Department of Pediatric Neurology, Kasugai, Japan, 4) Nagoya University School of Medicine, Department of Pediatrics, Nagoya, Japan, 5) Juntendo University School of Medicine, Department of Pediatrics, Tokyo, Japan, 6) Aichi Shukutoku University, Aichi, Japan

P-125 Analysis of urinary 6-Sulfatoxyxelatonin and 8-OHdG in low birthweight infants
* Araki Akiko, 1, 2 Shirai Masaru, 1, Ohinata Juunok, 1, Suzuki Nao, 1 Takahashi Satoru, 1 Tanaka Hajime, 1 Oki Jyunichi, 1 Fujieda Kenji 1
1) Department of Pediatrics, Asahikawa Medical College, Asahikawa, Japan, 2) Department of Pediatrics, Asahikawa-Kosei Hospital, Asahikawa, Japan
P-126 Clinical utility of neonatal apnea using bedside apnea analyzer
* Aoyagi Yoko, Zaitsu Ayuko, Mukae Tokutaro, Okada Junichiro, Hirose Akiko, Kanda Hiroshi, Fujino Hiroshi, Maeno Yasuki, Iwata Osuke, Matsuishi Toyojirou
The Department of Pediatrics, University of Kurume, Fukuoka, Japan

P-127 A girl with fetomaternal hemorrhage showing unexpectedly favorable language development.
* Nakamura Kazuyuki, Kato Mitsuhiro, Hayasaka Kiyoshi
The Department of Pediatrics, University of Yamagata, Yamagata, Japan

P-128 Epilepsy in children with PVL
* Fukuda Kunia, Kirino Tomoko, Fujiwara Yumi, Ushida Miyuki, Endo Shoichi
1) The Department of Neurology, Kagawa National Children’s Hospital, Zentju, Japan, 2) The Department of Pediatrics, Kagawa National Children’s Hospital, Zentju, Japan

15:05 ～ 15:35 Drugs
Chair: Susumu Ito, Yasufumi Utsumi

P-129 15 years-old boy with drug-induced hypersensitivity syndrome due to Zonisamide.
* Torio Michiko, Takemoto Megumi, Gondo Kenjiro, Hanai Toshio
Division of pediatric neurology, Fukuoka children’s hospital medical center, Fukuoka, Japan

P-130 A case of influenza encephalopathy with drug induced hypersensitivity syndrome due to phenytoin.
* Irahara Kaori, Aso Seijirou
Japanese RedCross Medical Center, Tokyo, Japan

P-131 A case of DIHS after HIV-6 encephalopathy
* Saiwa Satoshi, Tanaka Rieko1,2
1) The first department of Pediatrics, Japanese red cross society Wakayama medical center, 2) Tosyokai Inada Hospital, Wakayama, Japan

P-132 Drug eruption due to the antiepileptic drugs in children
* Nakamura Yukiko, Miwa Mami, Bessyo Fumio
Department of Pediatrics, Kyorin University, Tokyo, Japan

P-133 The progress report of European Register of Antiepileptic Drugs and Pregnancy (EURAP) in JAPAN
* Ohtani Hideyuki, Tanaka Masaki1,2, Takahashi Yukitoshi1, Inoue Yusu1, Fujiwara Tateki1, Sasagawa Mutsuo1, Mizobuti Masahiro
1) National Epilepsy Center Shizuoka Institute of Epilepsy and Neurological Disorder, Shizuoka Japan, 2) Tanaka Neurology Clinic, kanagawa, Japan, 3) epilepsy Center in Nishi-Niigata Chuo National hospital, Niigata, Japan, 4) Nakamura Memorial Hospital, Sapporo, Japan

P-134 Pediatric Acute Lymphoblastic Leukemia Presenting Tacrolimus related Encephalopathy
* Komatsu Hiroko, Yamashita Yushiro, Iizuka Chijo, Ohya Takashi, Nagamitsu Shinichiro, Inada Hiroko, Matsuishi Toyojirou
Department of Pediatrics and Child Health, Kurume University School of Medicine, Fukuoka, Japan

15:35 ～ 16:15 Sleep/Autonomic nerve
Chair: Yutaka Awaya, Shinji Fujimoto

P-135 The evaluation of sleep, fatigue, and cognition among students and children with school phobia.
* Junko Kawatani1, Joudo Takako2, Tomoda Akemi1, Shirai Seiji1, Miike Teruhisa2
1) Child Development Sociology Faculty of Medical and Pharmaceutical Sciences Kumamoto University Graduate School, 2) Child Development Faculty of Medical and Pharmaceutical Sciences Kumamoto University Graduate School, 3) Department of Child Development Kumamoto University Hospital

P-136 Effects of melatonin on sleep disorders in patients with school phobia.
* Ohinata Junko, Araki Akiko, Suzuki Nao, Takahashi Satoru, Tanaka Hajime, Fujieda Kenji
Department of Pediatrics, Asashikawa Medical College
P-137  CSF orexin-A measurement in pediatric and teenage patients with sleep disorders
* Arai Junko1, Kanbayashi Takashi1, Kubota Hiroaki2, Yano Tamami1, Sawashita Yukio1, Watanabe Yasuhiro2
1) Department of Pediatrics, Chiba Rosai Hospital, Chiba, Japan, 2) Department of Pediatrics, Graduate School of Medicine, Chiba University, Chiba, Japan, 3) Department of Neuropsychiatry, Akita University, School of Medicine, Akita, Japan, 4) Department of Pediatrics, Akita University, School of Medicine, Akita, Japan,

P-138  Triptan for migraine attacks making resistance to analgesic in children: risk and benefit
* Kitamura Shigekazu1, Tatsuoka Yoshihisa2
1) Department of Neurology, Konan Hospital, Kobe, Japan, 2) Tatsuoka Neurology Clinic, Kyoto, Japan

P-139  Prophylactic therapy using valproate sodium for cyclic vomiting syndrome in children
* Hikita Yoshitake1, Kodama Hiroko1, Nakamoto Natsue1, Ogita Koeri2, Amakata Kaori1, Kaneko Sono2, Fujii Yasushi2, Fujita Yasuko1, Yanagawa Yukishige1
1) Department of Pediatrics Teikyo University School of Medicine, Tokyo, Japan, 2) Faculty of Health Sciences Department of Occupational Therapy, Meiji University, Tokyo, Japan

P-140  A 1-year-old girl with comorbid obstructive sleep apnea syndrome and restless legs syndrome
* Kato Kumi1,2, Mohri Ikuko3, Tanikage Masako2
1) The Research Center for Child Mental Development, Osaka University Graduate School of Medicine, Osaka, Japan, 2) Sleep Medical Center, Osaka Kaisei Hospital, Osaka, Japan

P-141  A case of Alice in Wonderland syndrome with vertigo and abnormal behavior.
* Akita Susumu1,2, Miyamoto Yukinobu1
1) The Department of Pediatrics, Sanaikai General Hospital, Saitama, Japan, 2) The Department of Psychiatry, Saitama Children Medical Center, Saitama

P-142  Two cases of acute autonomic neuropathy with local hyperhidrosis
* Arai Hide1, Kubota Hiroaki2, Tanabe Yuzo1, Omata Taku1
1) Department of Neurology, Chiba Children's Hospital, Chiba, Japan, 2) Department of Pediatrics, Chiba Rehabilitation Center, Chiba, Japan, 3) Soga Pediatric clinic, Chiba, Japan

16:15 ～ 16:55   Brain anomaly
Chair: Kyoko Ito, Tomohide Goto

P-143  Two cases of frontal localized hemimegalecephaly: the mildest form of hemimegalecephaly?
* Ono Yoichi1, Tohyama Jun2, Sugai Kenji2, Maegaki Yoshihiro2, Ohno Koosaku1
1) The Child neurology, University of Tottori, Yonago, Japan, 2) Department of Pediatrics, Epilepsy Center, Nishi-Niigata Chuo National Hospital, Niigata Japan, 3) Department of Child Neurology, National Center Hospital for Mental, Nervous and Muscular Disorders, National Center of Neurology and Psychiatry, Tokyo, Japan

P-144  A case of Hemimegalecephaly diagnosed by convulsion
* Nakashima Kentaro1, Tsujikawa Ayako1, Arima Keitaro2, Nakagawa Machiko2, Tsuruta Shio1, Kusakawa Isao2, Hosoya Ryouta1, Ogihara Masaaki2
1) The Department of Pediatrics, St. Luke's International Hospital, Tokyo, Japan, 2) Ogihara clinic, Tokyo, Japan

P-145  3 cases of fetal MRI showed extension of ventricles, hypogenesis of corpus callosum and cerebellum.
* Takahashi Kan1, Satou Atsushi2, Mimaki Masakazu1, Saitou Makiko2, Oka Akira1, Mizuguchi Masashi2
1) Department of Pediatrics, Graduate School of Medicine, The University of Tokyo, Tokyo, Japan, 2) Department of Developmental Medicine, Graduate School of Medicine, The University of Tokyo, Tokyo, Japan

P-146  Management strategy of epilepsy in malformed brains
* Saito Yoshiaki1, Sugai Kenji1, Nakagawa Eiji1, Sakuma Hiroshi1, Komaki Hirofumi1, Sasaki Masayuki1, Otsuki Taisuke2, Ohno Koouyo3, Kondo Akiko3, Maegaki Yoshihiro2, Ohno Koosaku1
1) Department of Pediatric Neurology, National Center Hospital for Mental, Nervous and Muscular Disorders, National Center of Neurology and Psychiatry, Tokyo, Japan, 2) Department of Neurosurgery, National Center Hospital for Mental, Nervous and Muscular Disorders, National Center of Neurology and Psychiatry, Tokyo, Japan, 3) Division of Pediatric Neurology, Faculty of Medicine, Tottori University
P-147 A Case of C (Opitz Trigonocephaly) Syndrome with Brain Anomaly
*Mori Atsuko†, Fukamachi Makoto†, Kaname Tadashi†, Moriuchi Hiroyuki†, Fukuda Masafumi†
1) Department of Pediatrics, Misakaenosono Mutsumi, The Institute for severe intellectual/motor disabled persons, Isahaya, Japan, 2) Department of Pediatrics, Nagasaki University School of Medicine, 3) Department of Medical Genetics, Ryukyu University School of Medicine

P-148 A case of microcephaly with simplified gyral pattern whose EEG shows suppression-burst pattern.
*Nishimura Akira†, Ogami Aya†, Morioka Shigemi†, Hasegawa Tatsushi†, Tozawa Takenori†, Isoda Kenichi†, Matsui Fumihiro†, Morimoto Masafumi†
1) Department of Pediatrics, Kyoto Prefectural University of Medicine, Kyoto, Japan, 2) Department of Pediatrics, Fukuchiyama Municipal Hospital

P-149 Tethered cord in Miller-Dieker syndrome: A new association
*Wen-cheng Changjiao-shwann Liang
Department of Pediatrics, Far Eastern Memorial Hospital, Taipei, Taiwan

P-150 Four case reports with neural tube defects in Mexico- The actual circumstances for prevention
*Onoe Sachiko
Department of Pediatrics, Dr Luis F Nachon Civil hospital, Xalapa, Veracruz, Mexico

16:55 ~ 17:25 Infection/Immunology 1
Chair: Naoyuki Tanuma, Naoya Itokazu

P-151 A 10 year-old boy with polio myelitis like flaccid paralysis in right lower limb
*Hatori Takayuki†, Okubo Takashi†, Hashimoto Kiyoshi†, Fujino Osamu†
1) The Department of Pediatrics, Saitama Red Cross Hospital, Saitama, Japan, 2) The Department of Pediatrics, Nippon Medical School, Tokyo, Japan

P-152 Cerebrospinal fluid markers in a case of myeloradiculitis
*Anzai Yuki†, Hayashi Masaharu†, Miyata Rie†, Tanuma Naoyuki†, Ohya Tatsuo†
1) The Children's Center for Health and Development, Saiseikai Yokohama Eastern Hospital, Yokohama, Japan, 2) The Department of Clinical Neuroscience, Tokyo Metropolitan Institute for Neuroscience, Tokyo, Japan

P-153 An aquaporin-4 antibody positive adolescent girl with PRLES findings
*Yokoi Toshiaki†, Hattori Hideji†, Okano Yoshiyuki†, Tsuruhara Akitoshi†, Matsuoka Osamu†, Yamano Tsunekazu†, Nakano Eiko†, Matsui Masaru†, Tanaka Masami†, Konishi Tetsuro†, Tanaka Keiko†
1) Department of Pediatrics, Osaka City University Graduate School of Medicine, Osaka, Japan, 2) Department of Neurology, Utano National Hospital, Kyoto, Japan, 3) Department of Neurology, Niigata University School of Medicine, Niigata, Japan

P-154 A girl with opticospinal multiple sclerosis positive for anti-aquaporin-4 antibody
*Kodama Haruka, Adou Atsushi, Tanichi Masayo, Sugiura Yuuko, Moriguchi Naohiko
Department of Pediatrics, Sakai Hospital, Kinki University School of Medicine

P-155 A case of pediatric multiple sclerosis with non-convulsive status epilepticus
*Awaya Tomonari†, Kato Takeo†, Daifu Tomoo†, Shibata Minoru†, Yamanaka Yasunari†, Shiraishi Kazuhiro†, Tomiwa Kiyotaka†, Nakahata Tatsutoshi†
1) Department of Pediatrics, Graduate School of Medicine, Kyoto University, Kyoto, Japan, 2) Genetic Counselling and Clinical Research Unit, School of Public Health, Kyoto University, Kyoto, Japan, 3) Department of Pediatrics, National Utano Hospital

P-156 Support system for families with pediatric Multiple Sclerosis
*Hirano Yuiko, Funatsuka Makoto, Kodaira Kayano, Ishigaki Keiko, Nakayama Tomohiro, Osawa Makiko
Department of Pediatrics, Tokyo Women's Medical University, Tokyo, Japan
Infection/Immunology 2

P-157 Decoy Receptor 3 ameliorates experimental autoimmune encephalitis through down regulation of innate and adaptive immunity concomitant with suppression of TH17 cells
* Shyi-Jou Chen1, Yen-Lin Wang2, Su-Feng Wu1, Chih-Chien Wang1, Jen-Hsin Kao1, Pao-Luh Tao1, Chia-Cho Wu1, Chien-Len Liao1, Huey-Kang Syywu2,4
1) Department of Pediatrics, National Defense Medical Center, Taipei 114, Taiwan, 2) Graduate Institute of Life Sciences, National Defense Medical Center, Taipei 114, Taiwan, 3) Department of Microbiology and Immunology, National Chung Cheng University, Cha-Yi 621, Taiwan, 4) Department of Pharmacology, National Defense Medical Center, Taipei 114, Taiwan, 5) Department of Nephrology, National Defense Medical Center, Taipei 114, Taiwan, 6) Department of Microbiology and Immunology, National Defense Medical Center, Taipei 114, Taiwan, 7) Graduate Institute of Medical Sciences, Department of Pharmacology, National Defense Medical Center, Taipei 114, Taiwan

P-158 A patient with juvenile idiopathic arthritis presented recurrent influenza encephalopathy
* Sato Yuko1, Fukuda Tokiko1, Goto Tamako1, Mori Masato1, Yamagata Takanori2, Yotsumoto Sigeru1, Sugie Hideo1, Momoi Mariko1
1) Jichi Medical University, Department of Pediatrics, Tochigi, Japan

P-159 Acute cerebellar ataxia with cerebellar hypoperfusion in SPECT following MR vaccination
* Koyo Ohno1, Kondo Akiko2, Asai Koichi1, Ohno Kousaku2
1) Department of Pediatrics, Shimane Prefectural Central Hospital, Izumo, Japan, 2) Division of Child Neurology, Institute of Neurological Sciences, Faculty of Medicine, Tottori University, Japan

P-160 Four cases of subacute sclerosing panencephalitis performed ribavirin complex therapy.
* Oomi Tsuyoshi1, Tamashiro Kunihito1, Shimura Naohide2
1) University of the Ryukyus Faculty of Medicine, Okinawa, Japan, 2) Takaesu Clinic, Okinawa, Japan

P-161 A case of Rasmussen syndrome with lesions in the cerebral cortex and the contralateral basal ganglia
* Mimaki Masakazu1, Satoh Atsushi1, Takahashi Kan1, Itoh Masayuki2, Takahashi Yukitoshi1, Oka Akira1, Mizuguchi Masashi2
1) The Department of Pediatrics, University of Tokyo, Tokyo, Japan, 2) Department of Mental Retardation and Birth Defect Research, National Institute of Neuroscience, NCPP, Tokyo, Japan, 3) National Epilepsy Center, Shizuoka Institute of Epilepsy and Neurological Disorders, Shizuoka, Japan, 4) Department of Developmental Medical Sciences, Graduate School of Medicine, University of Tokyo, Japan

P-162 A patient with Rasmussen encephalitis and SCN1A mutation
* Kobayashi Katsuhiko1, Ohmori Iori2, Ouchida Mamoru1, Inoue Takashi1, Maegaki Yoshihiro2, Ohtsuka Yoko1
1) Department of Child Neurology, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, and Okayama University Hospital, Okayama, Japan, 2) Department of Cellular Physiology, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama, Japan, 3) Department of Molecular Genetics, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama, Japan, 4) Division of Child Neurology, Institute of Neurological Sciences, Faculty of Medicine, Tottori University, Tottori, Japan
8:00 ~ 9:00  Morning Seminar 1
Visually diagnosed epileptic seizures: focal seizures
Chairman  Tatsuro Izumi（Department of Pediatrics, Oita University）
Masako Sakauchi（Department of Pediatrics, Tokyo Women's Medical University）

9:00 ~ 9:40  Didactic Lecture 1
Surgical management of pediatric epilepsy
Chairman  Tatsuya Tanaka（Department of Neurosurgery, Asahikawa Medical College）
Tomokatsu Hori（The Department of Neurosurgery, Tokyo Women's Medical University）

9:50 ~ 12:00  Symposium 1 Diagnosis and treatment for childhood epilepsy — Expert opinion —
Chairperson  Hirokazu Oguni（Department of Pediatrics, Tokyo Women's Medical University）
Yoko Ohtsuka（Department of Child Neurology Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences）

1) West syndrome and its related disorders
Katsuhiko Kobayashi（Department of Child Neurology, Okayama University Hospital）

2) Lennox-Gastaut Syndrome and related epilepsies
Tateki Fujimura（Shizuoka Institute of Epilepsy and Neurological Disorders）

3) Myoclonic epileptic syndromes
Hirokazu Oguni（Department of Pediatrics, Tokyo Women's Medical University）

4) Topographic diagnosis, pathogenesis and treatment of cryptogenic or symptomatic partial epilepsies
Kenji Sugai（Department of Child Neurology, National Center Hospital for Mental, Nervous and Muscular Disorders, National Center of Neurology and Psychiatry）

5) CSWS, LKS, and related syndromes
Akihisa Okumura（Department of Pediatrics, Juntendo University School of Medicine）

12:20 ~ 13:20  Luncheon Seminar 4
Challenges toward treatments for neurodegenerative diseases.
Chairman  Yoshikatsu Eto（Department of Pediatrics, the Jikei University School of Medicine）
Shoji Tsuji（Tokyo University）

15:40 ~ 16:20  Didactic Lecture 2
"Tailoring" Best Available Neuroprotection in High and Low Resource Settings; Bench to Bedside, Tertiary Centres to Developing World
Chairman  Hitoshi Yamamoto（Department of Pediatrics, St. Marianna University, School of Medicine）
Osuke Iwata（Center for Developmental & Cognitive Neuroscience, Department of Pediatrics & Child Health）

16:20 ~ 18:30  Symposium 2. Hot Topics in Neonatal Neurology
Chairman  Masahiro Hayakawa（Nagoya University Hospital）
Shinichi Niiijima（Department of Pediatrics, Juntendo University Hospital）

1) The utility of the amplitude integrated EEG as the brain monitor
Kyoko Hirasawa（The Department of Paediatrics, Tokyo Women's Medical University）

2) Imaging update in perinatal brain damage
Noriko Aida（Department of Radiology, Kanagawa Children's Medical Center）

3) Topics of the Brain Protection in Neonate. Which has been your good choice, the Brain Hypothermia or the Medicine?
Masaki Shimizu（Division of Neonatology, Saitama Children's Medical Center）

4) Impact of recent neonatal medicines on the neurodevelopmental outcomes in human neonates
Hiroyuki Kidokoro（The Department of Pediatrics, Anjo Kosei Hospital）
19:00 ～ 21:00  Evening Seminar 2
The Committee of the plan macological issues — Concerta® Distribution management committee —
Chairman  Tasuku Miyajima (Department of Pediatrics,  Tokyo Women's Medical University)
Kitami Hayashi (Tokyo Women's Medical University Yachiyo Medical Center)
1) Tasuku Miyajima (Tokyo Medical University Hospital)
2) Tatsuya Koeja (Department of Education Faculty of Regional Sciences Tottori University)
3) Kazuhiko Saito (Kohoduai Hospital, International Medical Center of Japan)
4) Ichiro Sora (Department of Biological Psychiatry, Graduate school of Medicine, Tohoku University)

8:00 ～ 9:00  Morning Seminar 2
EEG:an introduction
Chairperson  Yoko Ohtsuka (Department of Child Neurology, Okayama University)
Solomon L. Moshe (Albert Einstein College of Medicine)

9:00 ～ 9:40  International Symposium Celebrating the 50th Meeting of JSCN Part III
Fukutinopathy
Chairman   Ikuya Nonaka (Musashi Hospital, National Center Hospital for Mental, Nervous and Muscular Disorders, National Center of Neurology and Psychiatry, Tokyo)
Fukuyama congenital muscular dystrophy :An overview
Yukio Fukuyama (Tokyo Women's Medical University)

9:50 ～ 12:00  International Symposium Celebrating the 50th Meeting of JSCN Part III
Phenotypic Spectrum of Fukutinopathy
Chairman  Tadayuki Ishihara (National Hakone Hospital, Tokyo, Japan)
Hideo Sugie (Jichi Medical University, Jichi Children's Medical Center, Japan)
1) Most severe phenotype of Fukutinopathy
Mieko Yoshioka (Department of Pediatric Neurology, Kobe City Pediatric and General Rehabilitation Center for the Challenged)
2) Mild phenotypes in Fukutinopathy
Francesco Muntoni (Dubowitz Neuromuscular Centre, UCL Institute of Child Health, London, UK)
3) Fukutin gene mutations cause dilated cardiomyopathy with minimal muscle weakness
Terumi Murakami (Department of Pediatrics, Tokyo Women's Medical University)
4) Development of Fukutinopathy model mouse
Tatsuhi Toda (Division of Clinical Genetics, Osaka University Graduate School of Medicine)

12:20 ～ 13:20  Luncheon Seminar 5
Treatment for Intractable Epilepsy
Chairman  Kazuei finuma (Ishimaki Red Cross Hospital)
Paolo Curatolo  (Professor of Pediatric Neurology and Psychiatry, Department of Neuroscience, Tor Vergata University, Rome, Italy)
14:30 ～ 15:40  International Symposium Celebrating the 50th Meeting of JSCN Part IV
Topics in neuromuscular disorders
Chairman Masaharu Hayashi（Tokyo Metropolitan Institute for Neuroscience）
Yuh-Jyh Jong（Kaohsiung Medical University, Taiwan）
1) Recent advance in spinal muscular atrophy
Yuh-Jyh Jong（Graduate Institute of Medicine, College of Medicine, Kaohsiung Medical University, Kaohsiung, Taiwan）
2) New insights into the pathogenesis of SMA
Yasushi Ito（Department of Pediatrics, Tokyo Women's Medical University, School of Medicine）
3) Congenital myasthenic syndrome
Keiko Ishigaki（Department of Pediatrics, Tokyo Women's Medical University, School of Medicine）

15:40 ～ 16:20 Didactic Lecture 4
Selective dorsal rhizotomy for spasticity in cerebral palsy children
Chairperson Kayoko Saito（Institute of Medical Genetics Tokyo Women's Medical University）
Takaomi Taira（Department of Neurosurgery, Tokyo Women's Medical University）

16:20 ～ 18:30 Symposium 4
Neurosurgical approach from a perspective of pediatric neurology
Chairman Shizuo Ohi（Department of Neurosurgery, the Jikei University School of Medicine）
Hiroaki Date（Chiba Children’s Hospital, department of neurosurgery）
1) Neurosurgical perspectives in non-accidental head injury child abuse
Hisashi Araki（The Department of Emergency and Critical Care Medicine, Nippon Medical School Hospital）
2) Specific Pathophysiological Features and Therapeutic Aspects in Management of Various types of Cerebrovascular Disease in Children
Rei Tabojin（Department of Neurosurgery, The Jikei University School of Medicine）
3) Operative indication, clinical results and points on intracranial arachnoid cysts
Masakazu Miyajima（Department of Neurosurgery, Juntendo University School of Medicine）
4) Clinical problems of the diagnosis and the strategy of treatment for spina bifida occulta
Jun Kurihara, Hiroshi Nishimoto（Department of Neurosurgery, Saitama Children's Medical Center）
5) Cell transplantation and regenerative therapy for neurological disorders in relation to cerebral ischemia
Isamu Date（Department of Neurological Surgery, Okayama University Graduate School of Medicine）

8:00 ～ 9:00 Morning Seminar 3
Effect of GH treatment on the brain metabolism
Chairman Goro Takada（Yonezawa National Hospital）
Shinichi Nijjima（Department of Pediatrics, Juntendo University Nerima Hospital）

9:00 ～ 9:40 Didactic Lecture 5
Understanding and support for developmental disorders with autism — from the viewpoint of living and working together after adolescence —
Chairman Tsunekazu Yamano（The Department of Pediatrics, Osaka City University Graduate School of Medicine）
Masami Sasaki（Department of Pediatrics, Juntendo University Nerima Hospital）
9:50 ~ 12:00   Symposium 5  Contribution of Child Neurology toward the Study of Developmental Disabilities
Chairman  Hitoshi Hara  (Yokohama Central Nursery Center)
Tatsuya Koeda  (Department of Education Faculty of Regional Sciences Tottori University)
1) Hyperplasia of the brain in autism spectrum disorders: neuroimaging studies
Toshiaki Hashimoto  (Department of Special Support Education, College of Education, Naruto University of Education)
2) Cognitive neuropsychological studies of executive dysfunction in developmental disorders
Yusuke Goto  (Department of Pediatrics, Faculty of Medicine, University of Yamanashi)
3) Behavioral disinhibition of children with Attention-Deficit / Hyperactivity Disorder -from the view of Clinical Neurophysiology-
Shinji Okazaki  (Institute of Disability Sciences, Graduate school of Comprehensive Human Sciences, University of Tsukuba)
4) Psychotherapy for children with developmental disabilities and their parents
Akashi Ishikawa  (Nire-no-kai Children's Clinic)

12:20 ~ 13:20   Luncheon Seminar 6
Working women and psychological development of children
Chairman  Tsunezako Yamano  (The Department of Pediatrics, Osaka City University Graduate School of Medicine)
Hisako Watanabe  (The Department of Pediatrics, School of Medicine, Keio University)

14:30 ~ 15:40   Workshop
Current topics of encephalitis and encephalopathy
Chairman  Hideo Yamauchi  (Department of Pediatrics, Dokkyo Medical University School of Medicine)
Masashi Mizuguchi  (Department of Pediatrics, Tokyo University)
1) Recent topics in acute encephalopathy in childhood
Hideo Yamanouchi  (Department of Pediatrics, Dokkyo Medical University School of Medicine)
2) Clinical findings in acute encephalitis with refractory, repetitive partial seizures  (AERPS)
Yutaka Awaya  (Department of Pediatrics, Seibo International Catholic Hospital)
3) Classification of Influenza encephalopathy
Masashi Shihomi  (Department of Pediatric Emergency Medicine, Osaka City General Hospital)

15:40 ~ 16:20   Didactic Lecture 6
Epigenetic mechanism regulating neural cell fate determination
Chairman  Kousaku Ohno  (Department of Child Neurology Faculty of Medicine, Tottori University)
Kinichi Nakashima  (Laboratory of Molecular Neuroscience, Graduate School of Biological Sciences, Nara Institute of Science and Technology)

16:20 ~ 18:30   Symposium 6
Epigenetics in Neurodevelopmental Diseases
Chairman  Takeo Kubota  (Department of Epigenetic Medicine, University of Yamanashi)
Shinji Fushiki  (Kyoto Prefectural University of Medicine)
1) Overview of Epigenetics in Neurodevelopmental Diseases
Takeo Kubota  (Department of Epigenetic Medicine, University of Yamanashi)
2) Angelman syndrome: diagnosis and treatment of genomic imprinting disorders
Shinji Saitoh  (Department of Pediatrics, Hokkaido University School of Medicine)
3) Regulation of neuronal development by the imprinted gene Ncdin
Kazuki Yoshikawa  (Laboratory of Regulation of Neuronal Development, Institute for Protein Research, Osaka University)
4) The chromosome-engineered mouse model for human chromosome 15q11-13 duplication
Toru Takumi  (Osaka Bioscience Institute)
5) Environmental chemicals may cause epigenomic alterations during brain development
Shinji Fushiki (Department of Pathology and Applied Neurobiology, Kyoto Prefectural University of Medicine Graduate School)

19:00 ~ 21:00  Evening Seminar 3
Multiple Sclerosis and Neuromyelitis Optica in Japan
Chairman  Toshiro Hara (Department of Pediatrics, Graduate School of Medical Sciences, Kyushu University)
Kazuo Fujiwara (Department of Multiple Sclerosis Therapeutics, Tohoku University School of Medicine)

9:00 ~ 9:40  Didactic Lecture 7
Cerebral Palsy
Chairman  Tohru Konishi (Nagaoka Ryokuen)
Kenji Yokochi (Department of Pediatric Neurology, Seirei-Mikatahara General Hospital)

9:50 ~ 12:40  Symposium 7
Comprehensive medical care and support for children with severe motor and intellectual disability
Chairman  Eiji Kitazumi (National Rehabilitation Center for Disabled Children)
Yuji Iwasaki (Toho Ryoiku Center)
1) Advance and problems in treatment to respiratory disorder in children with severe motor and intellectual disability
Eiji Kitazumi (National Rehabilitation Center for Disabled Children)

2) The importance and treatment of laryngotracheal lesions in children with with severe motor and intellectual disabilities
Yuji Mizuno (Department of Pediatrics, National Hospital Organization Fukuokahigashi Medical Center)

3) Surgical problems for neurologically impaired patients with severe deformity-The efficacy of laparoscopic surgery
Osamu Segawa (Division of Pediatric Surgery, Tokyo Women's Medical University)

4) Taking care of dying children with severely neurological disturbed condition : ethics and practicality
Michiko Yamada (Kanagawa Children's Medical Center, Institute of Severe Motor and Intellectual Disabilities)

5) The significance of local medical network for supporting patients with severe motor and intellectual
disabilities;palliative care and QOL at home
Keiko Shishikura (Tomo clinic for handicapped)

12:20 ~ 13:20  Luncheon Seminar 7
Mechanically assisted coughing in patients with neuromuscular disease
Chairman  Mitsuru Kawai (National Hospitai organization Higashisaitama Hospital)
Yuka Ishikawa (Department of Pediatrics, National Hospital Organization, Ykumo Hospital)

14:30 ~ 15:30  Imaging 1
Chair: Hiroaki Shiihara, Satoko Kumada
O-066 Functional Imaging Study of Development of Parenting Brain in Adolescents.
   * Nakai Akio, Matsuki Ken-ichi
   1) Department of Pediatrics, Faculty of Medical Sciences, University of Fukui,  2) Science of Development, Faculty of Education and Regional Studies, University of Fukui

O-067 Diffusion tensor imaging in children with basal ganglia-thalamic lesions
   * Okumura Akihisa, Hayakawa Masahiro, Tsuji Takeshi, Saito Masako, Tanaka Kyoko, Nakazawa Tomoyuki
   1) Department of Pediatrics, Juntendo University School of Medicine, Tokyo, Japan,  2) Department of Pediatrics, Nagoya University Graduate School of Medicine, Nagoya, Japan
O-068 Review of images in 10 cases of Septo-optic dysplasia
* Nuki Megumi, Kuki Chihiro, Kimura Shihoko, Hattori Taeka, Okazaki Shin, Kawashiki Hisashi, Tomiwa Kiyotaka
Department of Pediatric Neurology, Osaka City General Hospital

O-069 Sepsis associated encephalopathy with onset of status epilepticus presenting severe brain edema
* Kondou Akiko, Hujii Yui, Olanishi Touno, Ohno Kouyou, Sugihara Chitose, Inoue Takehiko, Saitou Yoshiaki, Maegaki Yoshihiro, Ohno Kousaku
Division of Child Neurology, Institute for Nurological Sciences, Faculty of Medicine, University of Tottori, Yonago, Japan

O-070 Study of the dissociation between CBF-SPECT and 123Iiomazenil SPECT
* Kuki Ichiro, Kawashiki Hisashi, Hattori Taeka, Nuki Megumi, Kimura Shihoko, Okazaki Shin, Ishikawa Jyunichi, Togawa Masao, Shio Masashi, Tomiwa Kiyotaka
1) Department of Pediatric Neurology, Children's Medical Center, Osaka City General Hospital, Osaka, Japan. 2) Department of Pediatric Emergency Medicine, Children's Medical Center, Osaka City General Hospital, Osaka, Japan. 3) Infection Center, Osaka City General Hospital, Osaka, Japan. 4) Genetic Counselor Coordinator Unit, Kyoto University, Japan

O-071 Non-invasive quantification of lactate by proton MR spectroscopy
* Iwasaki Nobuaki, Tanaka Ryuta, Matsumura Akira, Anno Izumi, Isobe Tomonori, Ohto Tatsuya, Nakayama Jyunuke, Kinugasa Hideyo
1) Department of Pediatrics, Ibaraki Prefectural University of Health Sciences, Ibaraki, Japan. 2) Department of Pediatrics, University of Tsukuba, Ibaraki, Japan. 3) Department of Neurosurgery, University of Tsukuba, Ibaraki, Japan. 4) Department of Radiology, University of Tsukuba, Ibaraki, Japan

15:30 ~ 16:40 Epilepsy 5
Chair: Masaharu Ohfu, Yukitoshi Takahashi

O-072 Prognostic Factors for Epileptic Seizures in SMIDS
* Matsumoto Akiko, Miyazaki Shuji, Nakamura Miho, Kumagai Toshiyuki
1) Kobato Gakuen, Aichi Human Service Center, Aichi, Japan, Institute for Developmental Research, Aichi Human Service Center, Aichi, Japan. 3) Central Hospital, Aichi Human Service Center, Aichi, Japan

O-073 A Long-Term Follow-up Study up to Adulthood in Dravet Syndrome
* Wakai Mari, Endo Fumika, Oka Maikio, Kobayashi Katsuhito, Ohtsuka Yoko
Department of Child Neurology, Okayama University Medical School, Okayama, Japan

O-074 Age dependent change of spike locations
* Yoshinaga Harumi, Ishizaki Yumiko, Inoue Takashi, Kikumoto Kennichi, Ohtsuka Yoko
Department of Child Neurology, Okayama University Precollege School of Medicine, Dentistr, and Pharmaceutical Sciences

O-075 Serial changes of spike localization in childhood partial epilepsy: MEG study
* Saito Naka, Tohyama Jun, Akasaka Noriyuki, Sasagawa Mutsuo, Kameyama Shigeki

O-076 A analysis of childhood epilepsies with a continuous spikes and waves during slow wave sleep (CSWS)
* Fukasawa Tatsuya, Azuma Yoshiteru, Natsume Jun, Ikuta Taketo, Kidokoro Hiroki, Okumura Akira, Negoro Tamiko
1) Department of pediatrics, Nagoya University of Medicine, Aichi, Japan. 2) Department of Pediatrics, Aichi Prefecture Medical Welfare Center of Aoiroti Gakuen, Aichi, Japan. 3) Department of Pediatrics, Anjo Kosei Hospital, Aichi, Japan. 4) Department of Pediatrics, Juntendo University School of Medicine, Tokyo, Japan. 5) Department of human welfare, Okazaki women's junior college, Aichi, Japan
O-077  A clinical study on childhood absence epilepsy preceded by generalized tonic-clonic seizures.
  * Wakamotu Hirokuki\textsuperscript{1}, Fukuda Mitsumasa\textsuperscript{2}, Hayashi Masatoshi\textsuperscript{1}
  1) Department of Pediatrics, Ehime Prefecture Central Hospital, Ehime, Japan,  2) Department of Pediatrics, Ehime University of Medicine,  3) Department of Pediatrics, Uwajima City Hospital

O-078  Follow up study of epileptic discharge in children with focal cortical displasia on frontal lobe
  * Minamitani Motoyuki\textsuperscript{2}, Hamano Shin-ichiro\textsuperscript{2}, Tanaka Manabu\textsuperscript{1}, Yoshinari Satoshi\textsuperscript{2,3}, Higurashi Norimichi\textsuperscript{2,3}, Eto Yoshikatsu\textsuperscript{2}
  1) Department for Child Health and Human Development, Saitama Children's Medical Center, Saitama, Japan,  2) Department of Pediatrics, The Jikei University School of Medicine, Tokyo, Japan,  3) Division of Neurology, Saitama Children's Medical Center, Saitama, Japan

16:40 ~ 17:40  Epilepsy 6

Chair: Eiji Hattori, Akie Miyamoto

O-079  A study with SPM for findings of qualitative FDG-PET in glucose porter 1 deficiency syndrome
  * Azuma Yoshiteru\textsuperscript{1}, Natsume Jun\textsuperscript{2}, Fukasawa Tatsuya\textsuperscript{1}, Negoro Tamiko\textsuperscript{2}, Watanabe Kazuyoshi\textsuperscript{1}, Yanagihara Keiko\textsuperscript{1}
  1) Department of Pediatrics, Nagoya University School of Medicine, Nagoya, Japan,  2) Department of Human Welfare, Okazaki Women's Junior College, Okazaki, Japan,  3) Faculty of Medical Welfare, Aichi Shukutoku University, Nagoya, Japan,  4) Developmental Infectious Diseases, Research Institute, Osaka Medical Center for Maternal and Child Health, Osaka, Japan

O-080  Findings of proton MRS in malformations of cortical development
  * Mori Kenji\textsuperscript{1}, Fujii Emiko\textsuperscript{1}, Mori Tato\textsuperscript{1}, Miyazaki Masahito\textsuperscript{1}, Harada Masafumi\textsuperscript{2}, Kagami Syoju\textsuperscript{1}
  1) Department of Pediatrics, School of Medicine, University of Tokushima, Tokushima, Japan,  2) Department of Radiologic Technology, School of Health Sciences, University of Tokushima, Tokushima, Japan

O-081  The pathology and treatment in three epileptic patients after inactivated influenza vaccination
  * Mine Jun, Takashiki Yukitoshi, Takahashi Hiroka, Ohtani Sanae, Ikeda Hiroko, Yamazaki Etuko, Kubota Yuko, Imai Katsumi, Fujiwara Tateki
  National Epilepsy Center, Shizuoka Institute of Epilepsy and Neurological Disorders

O-082  Neuroanatomical findings of 22q11.2 deletion syndrome with epilepsy.
  * Mori Tatsuo\textsuperscript{1,2}, Mori Kenji\textsuperscript{1}, Fujii Emiko\textsuperscript{1}, Kagami Syoju\textsuperscript{1}, Harada Masafumi\textsuperscript{3}
  1) Department of Pediatrics, Tokushima University School of Medicine, Tokushima, Japan,  2) Takamatsu Red Cross Hospital,  3) Department of Radiology, Tokushima University School o Medicine, Tokushima, Japan

O-083  The effect of theophylline on the susceptibility of hyperthermia-induced seizures in developing rats
  * Fukuda Mitsumasa\textsuperscript{1}, Suzuki Yuka\textsuperscript{1}, Watanabe Syouhei\textsuperscript{1}, Morimoto Takekiko\textsuperscript{1}
  1) Department of Pediatrics, Ehime University Graduate School of Medicine, Ehime, Japan,  2) Ehime Rehabilitation Center for Children, Ehime, Japan

O-084  Expression of cytokines in the rat hippocampus after kainic acid-induced status epilepticus
  * Sakuma Satoru, Yokoishi Toshiaki, Hattori Hideji, Matsuoka Osamu, Yamano Tsunezaku
  Department of Pediatrics, Osaka City University Graduate School of Medicine, Osaka, Japan
19:00 ～ 21:00 Evening Seminar 4: Committee of social activity
Chairman Takeo Sugimoto (Biwako Gakuen Medical and Welfare Center)
The problems of medical and social investigation in severely disturbed children
1) Takeo Sugimoto (Biwako Gakuen Medical and Welfare Center)
2) Noboru Takizawa (The Department of Pediatrics, National Hospital of Toyama)
3) Syuichi Tsubone (Department of Pediatrics and Rehabilitation, Medical and Welfare Center KIZUNA)
4) Kazuhiro Shimokawa (Hachioji Higashi Special School)
5) Kiyokuni Miura (Department of child neurology, Toyota municipal child development center)
6) Tohru Yokoi (Yokoi Clinic)
7) Koichiro Kawashima (Sendai Oushin Clinic)
8) Akira Iida (Nakanagaya Care Clinic)

Day 3 Room 5 (Apolon)

09:00 ～ 09:50 Encephalitis/Encephalopathy 1
Chair: Junichi Takanashi, Hideo Yamanouchi
O-085 Analysis of pathophysiology in acute infantile encephalopathy
  * Hayakawa Fumio1, Tsuji Takeshi1, Kato Toru1, Okumura Akihisa2
  1) The Department of Pediatrics, Okazaki City Hospital, Okazaki, Aichi, Japan, 2) The Department of Pediatrics, Juntendo University, Tokyo, Japan

O-086 Analysis of pathophysiology in acute infantile encephalopathy
  * Tsuji Takeshi1, Hayakawa Fumio1, Kato Toru1, Okumura Akihisa2
  1) The Department of Pediatrics, Okazaki City Hospital, Okazaki, Aichi, Japan, 2) The Department of Pediatrics, Juntendo University, Tokyo, Japan

O-087 Neurological symptoms in mild encephalitis/encephalopathy with a reversible splenial lesion (MERS)
  * Takanashi Jun-ichi1,2, Tada Hiroko1,2, Suzuki Motomasa3, Yamanouchi Hideo3,4, Yoshikawa Hideto5,6
  1) Department of Pediatrics, Kameda Medical Center, Kamogawa, Japan, 2) Segawa Clinic for Pediatric Neurology, Tokyo, Japan, 3) Okazaki City Hospital, Okazaki, Japan, 4) Dokkyo University School of Medicine, Tochigi, Japan, 5) Miyagi Children's Hospital, Sendai, Japan, 6) Research Group for Nervous and Mental Disorders (17A-11) from the Ministry of Health, Labor and Welfare of Japan

O-088 Encephalitis/encephalopathy with a reversible splenial lesion is associated with hyponatremia
  * Tada Hiroko1,2, Takanashi Jun-ichi1,2, Suzuki Motomasa3, Yamanouchi Hideo3,4, Yoshikawa Hideto5,6
  1) Department of Pediatrics, Kameda Medical Center, Kamogawa, Japan, 2) Segawa Clinic for Pediatric Neurology, Tokyo, Japan, 3) Okazaki City Hospital, Okazaki, Japan, 4) Dokkyo University School of Medicine, Tochigi, Japan, 5) Miyagi Children's Hospital, Sendai, Japan, 6) Research Group for Nervous and Mental Disorders (17A-11) from the Ministry of Health, Labor and Welfare of Japan

O-089 Thermolable phenotype of CPT II variations in acute encephalopathy.
  * Kubota Masaya1, Ozawa Hiroshi2, Mizuno Youko1, Kashii Hirohumi1, Amemiya Kaoru1, Suzuki Riina1, Koide Ayaka1, Hoshino Ai1, Yao Dengbing2, Kido Hiroshi1
  1) Division of Neurology, National Center for Child Medical Health and Development, Tokyo, Japan, 2) Shimada Ryoiku Center, 3) Department of Pediatrics, Metropolitan Hachioji Children's Hospital, 4) Department of Pediatric Neurology, Tokyo Metropolitan Neurological Hospital, 5) Division of Enzyme Chemistry, Institute for Enzyme Research, The University of Tokushima, Tokushima, Japan

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09:50 ~ 10:30  Encephalitis/Encephalopathy 2

Chair: Yoshihiro Maegaki, Masashi Mizuguchi

O-090 Clinical features of acute encephalopathy with delirious behavior
* Okumura Akihisa, Kidokoro Hiroyuki, Kubota Tetsuo, Tsuji Takeshi, Suzuki Motomasa, Kato Toru, Natsume Jun, Hayakawa Fumio, Watanabe Kazuyoshi
1) Department of Pediatrics, Juntendo University School of Medicine, Tokyo, Japan, 2) Department of Pediatrics, Anjo Kosei Hospital, Anjo, Japan, 3) Department of Pediatrics, Okazaki City Hospital, Okazaki, Japan, 4) Department of Pediatrics, Nagoya University Graduate School of Medicine, Nagoya, Japan, 5) Faculty of Medical Welfare, Aichi Shukutoku University, Nagoya, Japan

O-091 Clinical Features of Acute Encephalopathy with Bilateral Hemispheric Lesions
* Okumura Akihisa, Kidokoro Hiroyuki, Suzuki Motomasa, Kubota Tetsuo, Saito Masako, Nakazawa Tomoyuki, Tsuji Takeshi, Kato Toru, Natsume Jun, Hayakawa Fumio
1) Department of Pediatrics, Juntendo University School of Medicine, Tokyo, Japan, 2) Department of Pediatrics, Anjo Kosei Hospital, Anjo, Japan, 3) Department of Pediatrics, Okazaki City Hospital, Okazaki, Japan, 4) Department of Pediatrics, Nagoya University Graduate School of Medicine, Nagoya, Japan

O-092 Signal abnormalities of hippocampus of acute encephalopathy in childhood
* Tanaka Manabu, Higurashi Norimichi, Yoshinari Satoshi, Minamitani Motoyuki, Hamano Shin-ichiro
1) Division of Neurology, Saitama Children's Medical Center, Saitama, Japan, 2) Department for Child health and Human Development, Saitama Children's Medical Center, Saitama, Japan

O-093 Prolonged febrile seizures with HHV6 encephalopathy
* Sofue Ayako, Fukasawa Tatsuya
1) Department of pediatrics, Nagoya Memorial Hospital, Nagoya, Japan, 2) Department of pediatrics, Nagoya Memorial Hospital, Nagoya, Japan

10:30 ~ 11:30  Encephalitis/Encephalopathy 3

Chair: Takashi Ichiyama, Masaya Kubota

O-094 Roles of MMP-9 and TIMP-1 in acute encephalopathy following prolonged febrile seizures
* Suegata Naoko, Ichiyama Takashi, Kubota Masaya, Isumi Hiroshi, Tohyama Jun, Furukawa Susumu
1) Department of Pediatrics, Tsudumigaura Handicapped Children's Hospital, Shunan, Japan, 2) Department of Pediatrics, Yamaguchi University Graduate School of Medicine, Ube, Japan, 3) Department of Pediatrics, Tokyo Metropolitan Hachioji Children's Hospital, Hachioji, Japan, 4) Department of Pediatrics, Kawasaki medical School, Kurashiki, Japan, 5) Department of Pediatrics, Nishi-Niigata Chuo National Hospital, Niigata, Japan

O-095 Analysis of cytokines, MMP-9 and TIMP-1 in hemolytic uremic syndrome
* Ichiyama Takashi, Matsushige Takeshi, Kajimoto Madoka, Iyoda Kuniaki, Furukawa Susumu
1) Department of Pediatrics, Yamaguchi University Graduate School of Medicine, Ube, Ipanan, 2) Department of Pediatrics, Hiroshima City Hospital, Hiroshima, Japan

O-096 Early CSF biomarker for the diagnosis of acute encephalophy
* Yamanouchi Hideo, Nakajima Daiku, Kuribayashi Ryota, Watabe Yoshiyuki, Imataka George, Arisaka Osamu
Department of Pediatrics, Dokkyo Medical University School of Medicine, Tochigi, Japan

O-097 Cerebrospinal Tau Protein in acute encephalopathy
* Tanuma Naoyuki, Miyata Rie, Hayashi Masaharu, Kubota Masaya, Takashita Jun-ichi, Okumura Akihisa
1) Department of Pediatrics, Tokyo Metropolitan Fuchu Medical Center for the Disabled, Tokyo, Japan, 2) Department of Pediatrics, Tokyo Kita Shukai Hoken Hospital, Tokyo, Japan, 3) Department of Clinical Neuropathology, Tokyo Metropolitan Institute for Neuroscience, Tokyo, Japan, 4) National Center for Child Health and Development, Tokyo, Japan, 5) Department of Pediatrics, Kameda Medical Center, Kamogawa, Japan, 6) Department of Pediatrics, Juntendo University School of Medicine, Tokyo, Japan
O-098 Autoantibodies against GluR epsilon 2 in pediatric patients with acute limbic encephalitis
* Takahashi Yukitoshi, Yanasaka Etsuko, Nishimura Shigeko, Tsuogae Hisano, Fujisawa Tateki
National Epilepsy Center, Shizuoka Institute of Epilepsy and Neurological Disorders, Shizuoka, Japan

O-099 time course of blood NSE levels in children with brain death
* Mogami Yukiko, Toribe Yasuhisa, Yanagihara Keiko, Suzuki Yasuhiro
Osaka prefectural Hospital Organization, Izumi city, Japan

11:30 ~ 12:10 Encephalitis/Encephalopathy 4
Chair: Masaji Shiom, Shinichiro Hamano

O-100 Therapeutic Hypothermia for Acute Encephalopathy in Childhood: a Pilot Study
* Kawano Go1, Iwata Osuke2, Ohbu Keizo1, Aoki Takeshi1, Akaike Hiroto1, Uematsu Mitsugu1, Shiom Shashas1, Shimono Masayuki1, Senjyu Ayako1, Hamada Hiromi2, Hirano Satoru2, Hirabayashi Shinichi2, Yamanouchi Hideo1, Rinka Hiroshi1, Matsuishi Toyoiro2
1) Division of Pediatrics and Emergency Medicine, St-Mary Hospital, Japan. 2) Department of Pediatrics and Child Health, Kurume University School of Medicine, Japan. 3) Department of Pediatrics Tsukuba Medical Center.

O-101 The efficacy of immunosuppressant therapy for acute encephalopathy
* Takagi Atsushi1, Sameshima Kiyoko1, Tsuji Megumi1, Osaka Hitoshi1, Iai Mizue1, Yamada Michiko1, Nagafuchi Hiroyuki1, Yamashita Sumimasa1
1) Kanagawa Children's Medical Center, Division of Neurology, Kanagawa, Japan, 2) Kanagawa Children's Medical Center, Division of intensive care medicine, Kanagawa, Japan

O-102 Clinical experience of continuous EEG monitoring in pediatric neuro-intensive care
* Maruyama Azusa, Nagase Hiroaki
Kobe Children's Hospital, Hyogo, Japan

O-103 Rehabilitation Approach to Children with Higher Brain Dysfunction after Acute Encephalopathy
* Kurihara Mana2, Kohagizawa Toshitaka1, Yamanauchi Yuko1, Takahashi Kayoko1, Yajima Miki1, Eto Yoshikatsu2
1) Department of Pediatrics, The Kanagawa Rehabilitation Center, Atsugi, Japan, 2) Department of Pediatrics, Jikeikai University School of Medicine, Tokyo, Japan

14:30 ~ 15:30 Development/Evaluation
Chair: Miho Nakamura, Kazue Igarashi

O-104 Developmental changes in visuospatial short-term memory and working memory in healthy children
* Oka Makio1, Takeuchi Akihito1, Morooka Teruko1, Ogino Tatsuya2, Ohtsuka Yoko1
1) Department of Child Neurology, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama, Japan, 2) Faculty of Child Studies, Department of Child Studies, Chugokuakuen University

O-105 The assessment of joint attention in high-risk infants
* Fujii Yasushi1, Amakata Kaori1, Ogita Kaori1, Hikita Toshiyuki1, Kaneko Sono1, Nakamoto Natsue1, Fujita Yasuko1, Yanagawa Yukishige1
1) Department of Pediatrics, Teikyo University School of Medicine, Japan, 2) Department of Occupational Therapy, Mejiro University, Japan

O-106 The prevalence of palm-turned bye-bye and language development for very low birth weight infants.
* Matsui Gakuyu1, Yamamoto Akio1, Asaka Youko1, Takada Satoshi1
1) Kobe University Graduate School of Medicine, 2) Konan Women's University

O-107 Developmental changes of the Color-word matching Stroop task scores in normal children
* Morooka Teruko, Takeuchi Akihito, Oka Makio, Ogino Tatsuya, Ohtsuka Yoko
Department of Child Neurology, Okayama University Hospital, Okayama, Japan
O-108 Early diagnosis of developmental disorders in 3 years old ~The study of checklist of symptoms~
*Tsuda Yoshimi, Hashimoto Toshiaki, Takahara Mitue
The First Faculty, Naruto University of Education, Tokushima, Japan

O-109 Etiology of developmental disorders: prenatal factors
*Kurokawa Toru1, Yokomizo Yuko1, Yukaya Naoko2
1) Department of Pediatrics, Seiiai Rehabilitation Hospital, Ohnojou, Japan, 2) Department of Pediatrics, Kyushu University, Fukuoka

15:30 ~ 16:30 Developmental disabilities (investigation)
Chair: Shinichi Hirabayashi, Takashi Hayashi

O-110 Survey of handicapped Infants admitted to Osaka baby nursery home
*Misaki Takako1, Suehiro Yutaka2
1) The Pediatric Department, Osaka Saiseikai Natatsu Hospital, Osaka, Japan, 2) Osaka baby nursery home, Osaka, Japan

O-111 Study of Child with Developmental problem in Nursery School
*Ishikawa Yukie, Yamamoto Gyousei, Matsumi Gakuyou, Takada Satoshi
Department of Social Welfare Faculty of Human Development and Education

O-112 Early diagnosis of developmental disorders in 3 years old: Survey in Tokushima Prefecture
*Takahara Mitsue, Hashimoto Toshiaki, Tsuda Yoshimi
Department of Special Support Education, Naruto University of Education, Naruto, Japan

O-113 Support system for children with developmental disorders found out by 5 year-old medical checkup
*Miyazaki Masahiro1, Fujii Emiko2, Mori Kenji2, Hashimoto Toshiaki2, Kagami Shoji2
1) Department of Pediatrics, Miyoshi Medical Clinic, Higashikawa, Japan, 2) Department of Pediatrics, The Institute of Health Bioscience, The University of Tokushima Graduate School, Tokushima, Japan, 3) Department of Education for the Handicapped, Naruto University School of Education, Naruto, Japan

O-114 High-functioning pervasive developmental disorders in preschool children: assessment and treatment
*Muraoka Rika1, Koeda Tatsuya2, Ohno Kousaku1
1) Eastern Shimane Rehabilitation Hospital, Matsue, Japan, 2) Department of Education, Faculty of Regional Sciences, Tottori University, Tottori, Japan, 3) Division of Child Neurology, Faculty of Medicine, Tottori University, Yonago, Japan

O-115 Evaluation of prefrontal activity measured by NIRS in children with autism spectrum disorder
*Fukumoto Aya1, Hashimoto Toshiaki2, Tatsuta Youhei1, Fujii Emiko2, Nishimura Mio2, Tsuda Yoshimi2, Mori Kenji2, Miyazaki Masahiro2, Harada Masafumi, Kagami Syojo1
1) Department of Pediatrics, Institute of Health Bioscience The University of Tokushima Graduate School, Tokushima, Japan, 2) Department of Pediatrics Tokushima University School of Medicine, Tokushima, Japan, 3) Department of Special Support Education for the Disabled, Naruto University of Education, Naruto, Tokushima, Japan, 4) Tokushima Red Cross Hinomine Medical Center, 5) Department of Radiography Tokushima University School of Medicine, Tokushima, Japan

16:30 ~ 17:30 Developmental disabilities (support)
Chair: Akinori Hoshika, Jiro Ono

O-116 Problems of Special Support Educations in S City
*Hosoda Nozomi, Miura Hisao, Takei Kenji
Sagamihara Ryuikuen, Institute for Children with Severe Physical & Intellectual Disabilities, Sagamihara, Kanagawa, Japan

O-117 The children with Special Support in Regular Classroom focused on intellectual disabilities
*Yamaguchi Shima2, Takada Satoshi1
1) Faculty of Health Science, University of Kobe, Kobe, Japan, 2) Tarumi Physically handicapped school
O-118  Cooperation with school and rehabilitation center for developmental disturbed children
*Maeda Keiko
The Department of Neuropediatrics, Shizuoka Fukus Center, Shizuoka, Japan

O-119  A Study of Complicated disorder or symptom with Pervasive Developmental Disorder in usual classes
*Furusho Jyunichi1, Matsuzaki Kumiko1, Iwasaki Yoji2, Nakayama Harumi3, Shibata Reiko1, Nemoto Yoshiko4, Kubagawa Tetsuji5, Sone Mie1, Kato Nobumasa4
1) College of Literature, Department of Education, Aoyamagakui University, Tokyo, Japan, 2) Tokyo Metropolitan Tobu Medical Center for Persons with Severe Disabilities, 3) Tokyo Metropolitan Yotsugi Mobile Center for Persons with Severe Disabilities, 4) National Center for Child Health and Development, 5) Ohta General Hospital, 6) Division of Psychiatry, Kawasaki Municipal Hospital, 7) Graduate School, Shirayuri College, 8) Department of Psychiatry, School of Medicine, Showa University

O-120  Study of awareness of school teacher for the behaviors of children with developmental disorders
*Hayashi Takashi
Faculty of Nursing and Nutrition, Yamaguchi Prefectural University, Yamaguchi, Japan

O-121  Present Status and Issues in Transition Services for Children with Mild Developmental Disorders
*Imaeda Masayuki1,2, Funahashi Yoshimi1, Ishikawa Michiko2
1) Nagoya North Habilitation Center for Children, Nagoya, Japan, 2) Department of Pediatrics, Nagoya City University Graduate School of Medicine, Nagoya, Japan

17:30 ～ 18:20  Autism/Others
Chair: Yushiro Yamashita, Satoshi Sanada

O-122  The feature of Autism by KIDS
*Narita Yuri1, Hamano Shin-ichiro2, Kuroda Mai1, Minamitani Noriyuki2, Tanaka Manabu2
1) Division of Psychology, Center for Child Health and Human Development, Saitama Children's Medical Center, Saitama, Japan, 2) Division of Neurology, Saitama Children's Medical Center

O-123  Development of Japanese achievement test as to a screening method of Dyslexia
*Nagao Hideo
Department of Pathology for the handicapped, Faculty of Education, Ehime University, Matsuyama, Japan

O-124  The emotional responses are associated with future reward prediction and decision making
*Hosaka Hiromi1, Aoyagi Kakuro1, Kanemura Hideaki1, Kaga Yoshimi1, Yamashiro Dai1, Gotou Yuusuke1, Tandou Tomoko1, Nakamura Kousuke1, Sugita Kanji1, Aihara Masao1
1) Department of Pediatrics, Faculty of Medicine, University of Yamanashi, Yamanashi, Japan

O-125  Developmental changes of preparatory set and behavior inhibition in the patients with AD/HD
*Noguchi Sayaka, Kaga Yoshimi, Tando Tomoko, Sugita Kanji, Aihara Masao
Department of Pediatrics, Faculty of Medicine, University of Yamanashi, Yamanashi, Japan

O-126  methylphenidate therapy in 198 cases of ADHD or PDD associated with ADHD
*Sugama Michiko, Ishizaki Asayo
Oji Clinic: Division of Medicine The Association of Remedial Teaching for People with Developmental Handicaps, Tokyo, Japan

19:00 ～ 20:00  Evening Seminar 5: Botulinum toxin Treatment
Chairperson Yoko Ohtsuka (Department of Child Neurology Okayama University)
Atsuo Nezu (Children's Medical Centre, Yokohama City University Medical Centre)
1) Yasuhiro Takahashi (Department of Pediatrics, Kyushu Kousei Nenkin Hospital)
2) Masao Adachi (Kakogawa Municipal Hospital)
3) Atsuo Nezu (Children's Medical Centre, Yokohama City University Medical Centre)
**Neuroimmunology**

Chair: Yukiko Hirano, Tatsuo Oya

**O-127 Analysis for 13 patients of myeloradiculopathy**
"Shimono Masayuki, Senju Akiko, Ishii Masahiro, Shiota Naoki
Department of Pediatrics, University of Occupational and Environmental Health, Kitakyushu, Japan

**O-128 Child-onset Multiple Sclerosis (5 cases): Study of Steroid and Interferon Beta 1b Treatment**
"Endo Ayumi, Fujita Yukihiko, Imai Yuki, Haruyama Wakako, Arakawa Chikako, Kohira Ryutarou,
Fuchigami Tatsuo
The Department of Pediatrics, Nihon University School of Medicine, Tokyo, Japan

**O-129 Clinical examination of peripheral facial nerve palsy we had experience**
"Kuroiwa Yuiko, Ishiki Humie, Fukushima Naoya, Takeda Ryoujiyuen, Nanao Kenji, Takahashi Takao
1) Department of Pediatrics, Yamato Municipal Hospital, Kanagawa, Japan, 2) Department of Pediatrics, School of Medicine, Keio University, Tokyo, Japan

**O-130 Clinical diagnosis and therapy analysis of opsoconulus-myoclonus syndrome**
"Xiong Hui, Peng Jing, Zhang Yuehua
Department of Pediatrics, First Hospital, Beijing University, Beijing, China

**Neonatal Imaging/Others**

Chair: Satoshi Kusuda, Tetsuo Kubota

**O-131 Study of periventricular leukomalacia with late-onset circulatory dysfunction of premature infants**
"Kobayashi Satoru, Hattori Ayako, Andou Naoki, Kibe Tetsuya, Koyama Norihisa, Yokochi Kenji, Togari Hajime
1) Department of Pediatrics and Neonatology, Nagoya City University, Graduate School of Medical Sciences, Nagoya, Japan, 2) Department of Pediatrics, Seirei Mikatahara Hospital, Shizuoka, Japan, 3) Department of Pediatrics, Toyohashi Municipal Hospital, Toyohashi, Japan

**O-132 Developmental changes of cerebral hemodynamics in preterm infants**
"Ijichi Sonoko, Nishida Tomoko, Namba Masanori, Kawada Mayumi, Konishi Yukihiko, Okubo Kensuke, Kusaka Takashi, Imai Tadashi, Isobe Kenichi, Ito Susumu
1) Department of Pediatrics Faculty of Medicine, Kagawa University, Kitagun, Japan, 2) Maternal Perinatal Center, Kagawa University, Kitagun, Japan

**O-133 White matter injury in high risk infant**
"Iwata Osuke, Iwata Sachiko, Zaito Ayuko, Aoyagi Yoko, Ishido Yuki, Mukae Tokutaro, Okada Jyunichiro, Hirose Akiko, Kanda Hiroshi, Fujino Hiroshi, Maeno Yasuki, Matsuishi Toyojiro
The Department of Pediatrics, University of Kurume, Fukuoka, Japan

**O-134 Brain imaging of survivors of fetal death of monozygotic twin**
"Suzuki Motomasa, Maruyama Koichi, Kumagai Toshiyuki, Tsuji Takeshi, Kato Toru, Hayakawa Fumio, Okumura Akihisa
1) Department of Pediatric Neurology, Central Hospital, Aichi Welfare Center for Persons with Developmental Disabilities, Kasugai, Japan, 2) Department of Pediatrics, Okayama City Hospital, Okayama, Japan, 3) Department of Pediatrics, Junto University, Tokyo, Japan

**O-135 A case of neonatal cerebral hemorrhagic infarction developed after 1 month without clinical symptoms**
"Zaito Ayuko, Aoyagi Yoko, Kinoshita Masahiro, Ishido Yuki, Mukae Tokutaro, Okada Jyunichiro, Hirose Akiko, Kanda Hiroshi, Fujino Hiroshi, Maeno Yasuki, Iwata Osuke, Matsuishi Toyojiro
1) The Department of Pediatrics, University of Kurume, Fukuoka, Japan

**O-136 Heart-type fatty acid-binding protein as an early predictor of the outcome after birth asphyxia**
"Mitsufuji Nobuto, Kihara Minako
Division of Neonatal Intensive Care Unit, Perinatal Medical Center for Mothers and Children, Department of Pediatrics, Kyoto First Red Cross Hospital
10:40 ～ 11:40  Neonatal Seizures/EEG

Chair: Fumio Hayakawa, Shuichi Tsuneishi

O-137  The longitudinal study of neonatal electroencephalogram in infants with PVL
  "Kodokoro Hiroyuki"1, Kubota Tetsuo1, Kato Toru1, Hayakawa Fumio1.
  Suzuki Motomasa2, Yarayama Koichi2, Okumura Akihisa2, Watanabe Kazuyoshi5
  1) Department of Pediatrics, Anjo Kosei Hospital, 2) Department of Pediatrics, Juntendo University of Medicine, 3) Department of Pediatrics, Okazaki City Hospital, 4) Department of Pediatrics, 5) Faculty of Medical Welfare, Aichi Shukutoku University

O-138  The change of EEG of the patients with anoxic ischemic encephalopathy-using Wavelet analysis-
  "Hirasawa Kyoko"1, Itosamao2, Osawsmaki1
  1) The Department of Pediatrics, Tokyo Women’s Medical University, 2) The Department of Infants’ Brain & Cognitive development Tokyo Women's Medical University, CREST/JST, 3) NICU division, Maternal and Perinatal Center, Tokyo Women's Medical University

O-139  Symptomatic Localized-Related Epilepsy Associated with Periventricular Leukomalacia
  "Kubota Tetsuo"1, Oe Hideyuki1, Kodokoro Hiroyuki1, Suzuki Motomasa2, Maruyama Koichi2, Kato Toru1, Uemura Naoko2, Natsume Jun3, Okumura Akihisa6, Ochi Nobuhiko7
  1) Department of Pediatrics, Anjo Kosei Hospital, Anjo, Japan, 2) Aichi Welfare Center for Persons with Developmental Disabilities, Kasugai, Japan, 3) Department of Pediatrics, Okazaki Municipal Hospital, Okazaki, Japan, 4) Department of Pediatrics, Mitsubushi Nagoya Hospital, Nagoya, Japan, 5) Department of Pediatrics, Nagoya University, Nagoya, Japan, 6) Department of Pediatrics, Juntendo University, Tokyo, Japan, 7) Department of Pediatrics, Daini Aoitori Gakuen, Okazaki, Japan

O-140  The developmental change of the EEG envelop in premature and term infants
  "Saji Ryoya"1, Hirasawa Kyoko2, Ito Osako3, Konishiyukuro1, Taka Gentaro4, Kusuda Satoshio6
  1) Tamagawa University Brain Science Institute, Tokyo, Japan, 2) Tokyo Women’s Medical University, 3) Tokyo Women’s Medical University, 4) The University of Tokyo, 5) CREST/JST, 6) Tokyo Women’s Medical University

O-141  A clinical study of intractable epilepsy with partial seizures from neonatal period
  "Hattori Taeka"1, Kawakami Hisashi1, Nukui Megumi1, Kuki Ichiroh1, Kimura Shihoko1, Okazaki Shin1, Tomiwa Kiyoukata1
  1) The Department of Pediatric Neurology, Osaka City General Hospital, Osaka, Japan, 2) Genetic Counselor Coordinator Unit, Kyoto University, Japan

O-142  long term prognosis of cryptogenic neonatal convulsion
  "Maeda Tomoki"1, Sekiguchi Kazuhito, Akaishi Mutumi, Imai Kazuhide, Izumi Tatsuro
  The Department of Brain and Nerve Science Pediatrics, Oita University, Oita, Japan

11:40 ～ 12:20  Neurosurgery

Chair: Yasuo Aihara, Katsunori Fujii

O-143  Clinical characteristics of pediatric patients with familial moyamoya disease.
  "Mukawa Maki"1, Nairai Tadashi1, Maehara Taketoshi2, Aoyagi Masaru2, Matsushima Yoshihara2, Ohno Kikuo
  Department of Neurosurgery, Tokyo Medical & Dental University, Tokyo, Japan

O-144  Surgical treatment of juvenile patients with moyamoya disease based on hemodynamic measurement.
  "Nairai Tadashi"1, Momose Toshiya1, Inaji Motoki1, Mukawa Maki1, Matsushima Yoshihara2, Ohno Kikuo
  Department of Neurosurgery, Tokyo Medical and Dental University, Tokyo, Japan

O-145  Long term result and indication of neuro-endoscopic surgery
  "Matsukawa Yasuhiko"1, Sakamoto Hiroaki1
  Department of Pediatricneurosurgery, Osaka City General Hospital, Osaka city, Japan
14:30 ～ 15:50  Muscular dystrophy 1

Chair: Yukitatsu Ishikawa, Yukiko Hayashi

O-147  The retrospective study in patients with Juvenile dermatomyositis and polymyositis
* Kishi Takayuki, Hirano Yukiko, Ishigaki Keiko, Murakami Terumi, Suzuki Haruko, Shishikura Keiko, Hirayama Yoshito, Osawa Makiko
Department of Pediatrics, Tokyo Women’s Medical University, Tokyo, Japan

O-148  Clinical and pathological features of Japanese patients with nuclear envelope
* Hayashi Yukiko, Nonaka Ikuya, Nishino Ichizo
Department of Neuromuscular Research, National Institute of Neuroscience, NCNP

O-149  Electron microscopic analysis of myonuclei in EDMD2/LGMD1B
* Young-Eun Park, Y.K. Hayashi, I. Nonaka, I. Nishino
Department of Neuromuscular Research, National Institute of Neuroscience, National Center of Neurology and Psychiatry, Tokyo, Japan

O-150  Clinical, pathological and molecular genetic analysis of a Chinese family with dystrophinopathy
* Xiong Hui, Luo Jing, Wang Xiaozhu
Department of Pediatrics, First Hospital, Beijing University, Beijing 100034, China

15:50 ～ 16:40  Muscular dystrophy 2

Chair: Yoshinobu Otani, Masafumi Matsuo

O-155  Nonsense mutation was successfully revealed by MLPA analysis in a dystrophinopathy case
* Okuzuka Yo, Awano Hiroyuki, Yagi Mariko, Takehina Yasuhiro, Matsuo Masafumi
Department of Pediatrics, Kobe University Graduate School of Medicine, Kobe, Japan

O-156  MLPA analysis of dystrophin gene
* Minami Naruhito, Nishino Ichizo
1) Department of Laboratory Medicine, Musashi Hospital, National Center of Neurology and Psychiatry, Tokyo, Japan,
2) National Institute of Neuroscience, National Center of Neurology and Psychiatry, Tokyo, Japan

O-157  Splicing errors caused by small mutations in the dystrophin gene
* Yagi Mariko, Awano Hiroki, Okuzuka Yo, Takehina Yasuhiro, Matsuo Masafumi
1) Department of Clinical Evaluation of Pharmacotherapy, Kobe University Graduate School of Medicine, Kobe, Japan,
2) Department of Pediatrics, Kobe University Graduate School of Medicine, Kobe, Japan
O-158 Not only Dp71 dystrophin, but beta-dystroglycan is critical to cognitive impairment in DMD patients
* Dai Hongmei1, Itoh Kyoko1, Yaoi Takeshi1, Jinnai Kenji2, Fushiki Shinji1
1) Department of Pathology and Applied Neurobiology, Graduate School of Medical Science, Kyoto Prefectural University of Medicine, Kyoto, Japan, 2) Department of Neurology, National Hospital Organization, Hyogo Chuo Hospital, Sanda, Japan

O-159 Mesenchymal stem cells derived from placenta and chorionic villi for muscular dystrophy therapy.
* Arakawa Reiko1, Kawamichi Yayoi1,2, Aoki Ryoko1, Kondo Eri1, Saito Kayoko1
1) Institute of Medical Genetics, Tokyo Women’s Medical University, Tokyo, Japan, 2) Department of Obstetrics and Gynecology, Tokyo Women’s Medical University, Tokyo, Japan, 3) Support Center for Female Medical Scientists, Tokyo Women’s Medical University, Tokyo, Japan

16:40 ～ 17:50  Pompe disease
Chair: Hiroyuki Ida, Akemi Tanaka

O-160 Important issues in diagnosis of late-onset Pompe disease
* Fukuda Tokiko, Sugie Hideo, Momoi Mariko
Department of Pediatrics, Jichi Medical University, Tochigi, Japan

O-161 Initial efficacy of enzyme replacement therapy for a patient with childhood-onset Pompe disease
* Ishigaki Keiko, Murakami Terumi, Shishikura Keiko, Suzuki Haruko, Hirayama Yoshito, Osawa Makiko
Department of Pediatrics, Tokyo Women's Medical University, Tokyo, Japan

O-162 Enzyme replacement therapy for juvenile Pompe disease
* Tominaga Koji, Morita Yoshiko, Kitai Masahiro, Araya Ken, Shimono Kuriko, Okinaga Takeshi, Sakai Norio, Nagai Toshisaburo, Ozono Keiichi
Department of pediatrics, Osaka university school of medicine, Osaka, Japan

O-163 Lessons from Enzyme Replacement Therapy for Late Onset Pompe disease
* Kobayashi Hiroshi1, Kawai Toshinao1,2, Ohashi Toya1,2, Ida Hiroyuki1,2, Eto Yoshikatsu1,2, Osawa Makiko1
1) Department of Pediatrics, Jikei University school of medicine, Tokyo, Japan, 2) Department of Gene Therapy, Institute of DNA medicine, Jikei university school of medicine, 3) Department of pediatrics, Tokyo Womens Medical University

O-164 Evaluation of enzyme replacement therapy for adult onset Pompe disease
* Oda Eri1, Tanaka Toju1, Kozaki Rika1, Osawa Makiko2, Okuyama Torayuki1,2
1) National Center for Child Health and Development Tokyo Japan, 2) Tokyo Woman's Medical University Tokyo Japan, 3) National Center for Child Health and Development Tokyo Japan

O-165 A follow-up study of enzyme replacement therapy in a patient with glycogen storage disease type II
* Tanaka Akemi, Sawada Tomo, Yamano Tsunekazu
Department of Pediatrics, Osaka City University Graduate Shool of Medicine, Osaka, Japan

O-166 Alglucosidase alfa in Juvenile and Adult Patients with Pompe Disease: Results from a Randomized, Double-Blind, Multicenter, Multinational, Placebo-Controlled Study
* A. van der Ploeg1, P. Clemens2, D. Corzo3, D. Escolar4, J. Florence5, P. Laforet6, S. Lake7, J. Mayhew7, A. Pestronk5, B. Rosenbloom5, A. Skrinar5, M. Wasserstein8
1) Erasmus Medical Center, Rotterdam, NL, 2) University of Pittsburgh, Department of Neurology, Pittsburgh, PA, USA, 3) Genzyme Corporation, Cambridge, MA, USA, 4) Children's National Medical Center, Washington, DC, USA, 5) Washington University?, 6) School of Medicine, St. Louis, MO, USA, 6) Institut de Myologie, Groupe Hospitalier Pitie-Salpetriere, Paris, FR, 7) The Cooperative International Neuromuscular Research Group (CINRG), Washington, DC, USA, 8) Tower Hematology Oncology, Beverly Hills, CA, USA, 9) Mount Sinai School of Medicine, New York, NY, USA?, 10) Tower Hematology Oncology, Beverly Hills, CA, USA, 11) Genzyme Corporation, Cambridge, MA, USA, 12) Mount Sinai School of Medicine, New York, NY, USA?
17:50～18:20  Neuro muscular disorders

Chair: Yoshihiro Takeuchi, Yuichi Goto

O-167  Study on phosphorylation of SMN proteins
  * Aoki Yusuke, Fukao Toshiyuki, Ohnishi Hidenori, Orii Kenji, Kondo Naomi
    Department of Pediatrics, Graduate School of Medicine, Gifu University, Gifu, Japan

O-168  Clinical efficacy of Amantadine for SMAII
  * Haginoya Kazuhiro1, Tanaka Soichiro1, Uematsu Mitsugu1, Fukuyo Naomi2, Watanabe Syu-er1, Niisato Junko1, Onuma Akira1
    1) Takuto Rehabilitation Center for Children, Sendai, Japan, 2) Department of Pediatrics, Tohoku University School of Medicine, Sendai, Japan

O-169  Valproic acid does not always increase SMN2 expressions in fibroblasts in fibroblasts from patient with SMA Type1.
  * Gumadi1, Matsuo Masafumi2, Nishio Hisahide1
    1) Department of Genetic Epidemiology, Kobe University Graduate School of Medicine, Kobe, Japan, 2) Department of Pediatrics, Kobe University Graduate School of Medicine, Kobe, Japan

O-170  Benign infantile mitochondrial myopathy due to reversible cytochrome c oxidase deficiency: a case.
  * Matsushige Takeshi1, Kajimoto Madoka1, Ichiyama Takashi1, Furukawa Susumu1, Sugio Yoko2, Nishino Ichizo3, Goto Yuichi4
    1) Department of Pediatrics, Yamaguchi University School of Medicine, Yamaguchi, Japan, 2) Department of Pediatrics, Yamaguchi University Graduate Medical Center, 3) Department of Neuromuscular Research, National Institute of Neuroscience, National Center of Neurology and Psychiatry, 4) Department of Mental Retardation and Birth Defect Research, National Institute of Neuroscience, National Center of Neurology and Psychiatry

14:30～15:15  Encephalitis I

Chair: Kitami Hayashi, Atsushi Imamura

P-163  Onset related factors of acute encephalopathy with febrile convulsive status epilepticus
  * Inoue Takahito, Kakura Hiroshi, Ideguchi Hiroshi, Fujita Takako, Ishikawa Yukiko, Tomonou Yuko, Ninomiya Shinya, Nakamura Noriko, Yasumoto Sawa, Hirose Shinichi
    The Department of Pediatrics, Fukuoka University, Fukuoka, Japan

P-164  Clinical Analysis of Acute Encephalopathy with Febrile Convulsive Status Epilepticus
  * Takayanagi Masaru1, Kitamura Tarou1, Yamamoto Katsuya1
    1) Department of Pediatrics, Sendai City Hospital, Sendai, Japan, 2) Nankoudai Yamamoto Children’s Clinic, Sendai, Japan

P-165  A case of MRI with acute encephalopathy with febrile convulsive epilepticus like after head injury
  * Kajimoto Madoka1, Ichiyama Takashi1, Suenaga Naoko1, Matsushige Takeshi1, Furukawa Susumu1
    1) Department of Pediatrics, Yamaguchi University Graduate School of Medicine, Ube, Yamaguchi, Japan

P-166  Intractable epilepsy after acute encephalopathy with status epilepticus
  * Kometani Hiroshi, Sakuma Hiroshi, Saito Yoshiaki, Komaki Hirohumi, Nakagawa Eiji, Sugai Kenji, Sasaki Masayuki
    National Center of Neurology and psychiatry, Musash Hospital

P-167  Two cases of acute encephalopathy that developed partial epilepsy without a latent period
  * Koichi Maruyama1, Kondo Yoko2, Itomi Seiko2
    1) Department of Pediatric Neurology, Central Hospital, Aichi Welfare Center for Persons with Developmental Disabilities, 2) Department of Pediatrics, Japan Red Cross Nagoya 1st Hospital
P-168 Temporal characteristics of serum deviation enzymes in AESD.
*Goto Tomohide1, Nomura Toshihiro2, Arima Fujio1, Miyama Sahoko1
1) Department of Neurology, Tokyo Metropolitan Kiyose Children’s Hospital, Tokyo, Japan. 2) Department of Pediatrics, National Hospital Organization Saitama National Hospital, Saitama, Japan. 3) Department of Pediatrics, National Hospital Organization Tokyo Medical Center, Tokyo, Japan

P-169 Neurological sequel associate with influenza encephalopathy
*Goto Tamako, Fujita Hitomi, Suwa Kiyotaka, Mori Masato, Fukuda Tokiko, Yamagata Takanori, Sugie Hideo, Momoi Mariko
Department of Pediatrics, Jichi Medical University, Tochigi, Japan

P-170 Clinical manifestation of hemorrhagic shock and encephalopathy syndrome, a clinical study of 2 cases
*Endo Yusaku1, Suzuki Teruhiko2, Miyamoto Takeshi2, Hirano Kouichi1, Ohzeki Takehiko1
1) The Department of Pediatrics, Hamamatsu University School of Medicine, Hamamatsu, Japan. 2) The Department of Pediatrics, Kosai General Hospital, Kosai, Japan

P-171 A case of peculiar encephalitis/encephalopathy in which high concentration of isoflurane is needed
*Shiraga Hiroshi
National Hospital Organization Okayama Medical Center, Okayama, Japan

15:15 ~ 15:55 Encephalitis 2

Chair: Koji Ushijima, Takuya Tanabe

P-172 Acute encephalitis/encephalopathy with psychiatric symptoms in children
*Miyama Sahoko1, Goto Tomohide1, Goto Yuusuke2, Takuma Yuichi1
1) Department of Neurology, Tokyo Metropolitan Kiyose Children's Hospital, Tokyo, Japan. 2) Department of Pediatrics, University of Yamanashi, School of Medicine, Yamanashi, Japan. 3) Takuma Children's Clinic

P-173 A case of encephalopathy showing unusual speech and transient abnormalities at diffusion MRI.
*Yamamoto Keiichi1, Ozaki Hirohiko1, Takahashi Takao2
1) Department of Pediatrics, Isehara Kyodo Hospital, Isehara, Japan. 2) Department of pediatricians, University of Keio, Tokyo, Japan. 3) Department of Pediatrics, Hiratsuka Kyousai hospital, Hiratsuka, Japan

P-174 A case of non-herpetic acute limbic encephalitis, showing severe onset and good prognosis
*Katagiri Tomoko1, Araki Satoshi1, Yui Takako2, Miyata Rie1, Tanuma Naoyuki1, Hayashi Masaharu1, Takahashi Yukitoshi1
1) The Department of Pediatrics, Tokyo Medical and Dental University, Tokyo, Japan. 2) The Department of Pediatrics, Yokohama City Minato Red Cross Hospital, Yokohama, Japan. 3) The Department of Clinical Neuropathology, Tokyo Metropolitan Institute for Neuroscience, Tokyo, Japan. 4) The Department of Pediatrics, Shizuoka Institute of Epilepsy and Neurological Disorders, Shizuoka, Japan

P-175 A Child with Non-Herpetic Acute Limbic Encephalitis followed by Temporal Lobe Epilepsy as a sequela.
*Araki Atsushi1, Suzukawa Jyunoko1, Kaneko Kazunari2
1) Department of Pediatrics, Kansai Medical University Takii Hospital, Osaka, Japan. 2) Department of Pediatrics, Kansai Medical University Hirakata Hospital, Osaka, Japan

P-176 Acute limbic encephalitis in children
*Sakuma Hiroshi1, Saitoh Yoshiaki1, Komaki Hirofumi1, Nakagawa Eiji1, Sugai Kenji1, Sasaki Masayuki1, Yamanaka Yasunari1
1) Department of Child Neurology, National Center Hospital for Mental, Nervous and Muscular Disorders, National Center of Neurology and Psychiatry, Kodaira, Tokyo, Japan. 2) Department of Pediatrics, University of Kyoto

P-177 Decreased iomazenil uptake in inflammatory focus of non-herpetic acute encephalitis
*Higurashi Norimichi1,2, Hamano Shin-ichiro1, Yoshinari Satoshi1, Tanaka Manabu1, Minamitani Motoyuki2, Eto Yoshikatsu2
1) Division of Neurology, Saitama Children’s Medical Center, Saitama, Japan. 2) Department of Pediatrics, Jikei University School of Medicine, Tokyo, Japan. 3) Division of Child Health and Human Development, Saitama Children's Medical Center, Saitama, Japan
P-178 Clinical observations of five cases with human herpesvirus 6 encephalitis/encephalopathy
* Nishimura Akira1, Sakuma Satoru1, Hattori Eiji1, Takaara Natsuko1, Imamura Takaji1, Asada Minoru1
1) Department of Pediatrics, PL Hospital, Tondabayashi, Japan, 2) Osaka City University Graduate School of Medicine, Department of Pediatrics

P-179 A case of HHV-6 encephalopathy without typical roseola rash
* Hara Keita2, Tanabe Takuya1, Shimakawa Yuichi2, Tamai Hiroshi2
1) Division of pediatrics, Hirakata City Hospital, Osaka, Japan, 2) Department of pediatrics, Osaka Medical College, Osaka, Japan

15:55 ~ 16:35 Encephalitis 3
Chair: Ryutarok Kira, Seijiro Aso

P-180 A case of Herpes simplex virus encephalitis with normal brain MRI
* Tomizawa Naoko, Shinohara Yuki, Suzuki Keiko, Ueda Satoshi, Umezu Ryoji, Sugihara Shigetaka
Department of Pediatrics, Tokyo Women’s Medical University Medical Center East, Tokyo, Japan

P-181 Rhombencephalitis associated with coxsackie virus A16 hand-foot-and-mouth disease
* Sanefuji Masafumi, Kira Ryutarou, Torisu Hirohiko, Ishizaki Yoshito, Yukaya Naoko, Washitou Natsumi, Hara Toshiro
Department of Pediatrics, Graduate School of Medical Sciences, Kyushu University, Fukuoka, Japan

P-182 A case of Acute ecephalitis Associated with Human Parvovirus B19 Presented CPS status
* Yamazaki Sawako1, Tojo Megumu1
1) Department of Pediatrics, Niigata City General Hospital, Niigata, Japan, 2) Department of pediatrics, Hamagumi Ryoiku Center, niigata, Japan

P-183 Case report: Rota virus encephalitis with transient callosal splenium lesion and cerebellar symptoms
* Miyamoto Takeshi1,2, Suzuki Teruhiko2, Endoh Yusaku1, Hirano Kouichi2, Takahashi Yukitoshi1, Ohzeki Takehiko1
1) Department of Pediatrics, Kosai General Hospital, Shizuoka, Japan, 2) Department of Pediatrics, Hamamatsu University School of Medicine, Shizuoka, Japan, 3) Shizuoka Institute of Epilepsy and Neurological Disorders, Shizuoka, Japan

P-184 Transient cerebellar mutism associated with rotavirus cerebellitis in a 4-year-old girl
* Nishioaka Momoko1, Honda Ryoko1, Imamura Yoshihiko1, Turu Akira1, Moriuti Hiroyuki1
1) The Department of Pediatrics, Nagasaki University School of Medicine, Nagasaki, Japan, 2) The Department of Pediatrics, Nagasaki National Hospital, Nagasaki, Japan

P-185 Acute cerebellitis treated by ventricular drainage
* Michishita Takashi, Kouji Toshihiko
Pediatrics, Musashino Red Cross Hospital, Tokyo, Japan

P-186 A case of acute cerebellitis
* Ishida Hiroshi1, Nakajima Seijun1, Yokoi Toshiaki2, Hattori Hideji2, Matsuoka Osamu3, Ichiyama Takashi1, Murakami Seiko1
1) Department of Pediatrics, Izumi Municipal Hospital, Izumi, Osaka, Japan, 2) Department of Pediatrics, Osaka City University Graduate School of Medicine, Osaka, Japan, 3) Department of Pediatrics, Yamaguchi University Graduate School of Medicine,

P-187 Two cases with cerebellar hypoperfusion on brain SPECT
* Koizumi Shinya1, Fuzino Osamu1, Kuwahara Kentarou1, Takagi Atsushi2, Hatori Takayuki2, Fujimatsu Mariko3, Kawakami Yasuhiro4, Fujita Takehisa4, Takaishi Yasuke4, Okada Kazuyoshi4
1) Department of pediatrics, Nippon Medical School Chiba Hokusoh Hospital, Inba, Chiba, Japan, 2) Department of pediatrics, Nippon Medical School Hospital, Tokyo, Tokyo, Japan, 3) Department of pediatrics, Nippon Medical School Musashikosugi Hospital, Kawasaki, Kanagawa, Japan, 4) Department of pediatrics, Nippon Medical School Tamanagayama Hospital, Tama, Tokyo, Japan
16:35 ~ 17:05  Encephalitis 4
Chair: Yukihiko Fujita, Kyomi Hirayasu

P-188  Study for prognosis and characteristics of recent bacterial meningitis
* Murakami Kiyotaka
Nakano Children’s Hospital

P-189  1 year-old girl with Kawasaki disease symptoms at EBV primary infection, merged ADEM with hemorrhage
* Takamoto Megumi, Gondo Kenjiro, Hanai Toshio
Division of pediatric neurology, Fukuoka children’s hospital medical center, Fukuoka, Japan

P-190  Intral EEG in a patient with acute disseminated encephalomyelitis with seizures.
* Isoda Kenichi, Yokoi Kentarou, Matsui Fumihiro, Nishimura Akira, Morimoto Masafumi
Department of Pediatrics, Kyoto Prefectural University of Medicine, Kyoto, Japan

P-191  A case of ADEM treated by methylprednisolone pulse, immunoglobulin, and plasmapheresis.
* Imamura Mari1,2, Maruyama Shinsuke1,2, Toyoshima Mitsuo1, Kawano Yoshifumi2
1) Kagoshima Prefectural Oshima Hospital, Kagoshima, Japan, 2) Department of Pediatrics, Graduate School of
Medical and Dental Sciences, Kagoshima University, Kagoshima city, Japan

P-192  A case of two phases ADEM
* Fujimatsu Mariko, Fujita Takehisa
Department of Pediatrics, Nippon Medical School, Musashikosugi hospital, Kanagawa, Japan

P-193  Cotinal dominant T2 high intensity in a case of acute disseminated encephalomyelitis
* Kitamura Taro1, Takayanagi Masaru1, Yamamoto Katsuya2, Ishii Kiyoshi1
1) Sendai City Hospital, Department of Pediatrics, Sendai, Japan, 2) Nankodai Yamamoto Children’s Clinic, Sendai,
Japan, 3) Sendai City Hospital, Department of Radiology, Sendai, Japan

17:05 ~ 17:50  Encephalitis 5
Chair: Hideo Aiba, Hiroshi Yoshioka

P-194  Two cases with acute encephalitis
* Narita Aya, Ito Masahiro, Tamaki Hismitsu
Department of Pediatrics, Metropolitan Bokutoh Hospital

P-195  Study of electroencephalogram findings in nine cases of fulminant hepatic failure
* Hoshino Hideki, Mizuguchi Koichi, Nagasawa Tetsuro, Kubota Masaya
Division of neurology, National center for child health and development, Tokyo, Japan

P-196  HIE syndrome with biphasic seizures and normal MRI the day before late reduced diffusion
* Takahashi Tsutomu
Saiseikai Utsunomiya Hospital, Tochigi, Japan

P-197  Acquired hypoxic encephalopathy due to cardiac disorders and Higher brain dysfunction.
* Yamauchi Yuko1,2, Kurihara Mana1,2, Kohagizawa Toshitaka1,2, Takahashi Kayoko1,2, Eto Yoshikatsu2
1) Department of Pediatrics, The Kanagawa Rehabilitation Center, Atsugi, Japan, 2) Department of Pediatrics, Jikeikai
University School of Medicine, Tokyo, Japan

P-198  A case report of tuberous sclerosis and acute encephalopathy with persistent severe brain edema
* Terashima Hiroshi, Satoh Atsushi, Takahashi Kan, Mimaki Masakazu, Oka Akira, Mizuguchi Masa
The Department of Pediatrics, University of Tokyo, Tokyo, Japan

P-199  A case of acute encephalopathy with persistent abnormal signal on temporal-occipital lobe in MRI
* Mizuno Yo’s, Kashihi Hirofumi, Amemiya Koro’s, Suzuki Riina, Kubota Masaya’s, Ichiyama Takashi’s, Tanuma
Naoyuki’s, Miyata Rie’s, Hayashi Masaharu’s
1) Tokyo Metropolitan Hachioji Children’s Hospital, Tokyo, Japan, 2) The Department of Pediatrics, University of
Yamaguchi, Yamaguchi, Japan, 3) Tokyo Metropolitan Institute for Neuroscience, Tokyo, Japan
P-200  A case of encephalopathy with extremely localized spikes at the onset
   * Ono Hiroki
   The Department of Pediatrics, Hiroshima Prefectural Hospital, Hiroshima, Japan

P-201  Evaluation of prolonged febrile seizure by the MRI
   * Akaike Hiroto, Kawai Yasuhiro
   Department of pediatrics, Kawasaki Medical School, Okayama, Japan

P-202  Clinical observation of two cases with prolonged brain death-like status
   * Ori Koji1,2, Matsuo Naoki3, Ito Reiko2, Imamura Atsushi2
   1) The Critical Care & Emergency Center, Gifu Prefectural General Medical Center, Gifu, Japan, 2) The Department of Pediatrics, Gifu Prefectural General Medical Center, 3) The Department of Neonatology, Gifu Prefectural General Medical Center

14:30 ~ 15:05  Epilepsy 1
   Chair: Kimio Minagawa, Hideki Horita

P-203  A screening test for early diagnosis of severe myoclonic epilepsy in infancy
   * Ohmori Iori1, Hattori Junri2, Ouchida Mamoru1, Maniwa Satoshi1, Mimaki Yoshinobu1, Miyake Susumu2, Ohtsuka Yoko2
   1) Department of Physiology, Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama University,Okayama, Japan, 2) Department of Child Neurology, Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama University,Okayama, Japan, 3) Department of Molecular Genetics, Graduate School of Medicine, Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama University,Okayama, Japan, 4) Mutsuyama Red Cross Hospital, Pediatrics, Mutsuyama, Japan, 5) Kurashiki Medical Center, Pediatrics, Kurashiki, Japan, 6) Kagawa Prefectural Central Hospital, Pediatrics, Takamatsu, Japan

P-204  Unexpected Death of Severe Myoclonic Epilepsy in Infancy
   * Sokoda Tatsuyuki1,2, Takeo Tomoyuki1, Takeuchi Yoshihiro1, Kumoda Masao1, Fujita Yasuyuki2
   1) The Department of Pediatrics, Shiga University of Medical Science, Shiga, Japan, 2) Biwakogakuen Iryouhukushi Center Kusatsu

P-205  A case of infantile epilepsy with a novel SCN1A mutation
   * Matsumoto Hiroshi1, Nakamura Yasuko1, Takizawa Yuji1, Hirose Shinichir2, Kurahashi Hirokazu2,3
   1) Department of Pediatrics, National Defense Medical College, Tokorozawa, Japan, 2) Department of Pediatrics, Fukuoka University, Fukuoka, Japan, 3) Department of Pediatrics, Nagoya University, Nagoya, Japan

P-206  Infants with diarrhea-related seizures were not associated with SCN1A mutation
   * Wang-Tso LEEWen-Chin Weng1, Yen-Ting Chou1, Shinichi Hirose2
   1) Pediatrics, National Taiwan University Hospital, Taipei, Taiwan, 2) Pediatrics, National Taiwan University Hospital, Taipei, Taiwan, 3) Pediatrics, National Taiwan University Hospital, Taipei, Taiwan, 4) Pediatrics, Fukuoka University, Fukuoka, Japan

P-207  SCN1A gene mutations in human and mouse
   * Ogiwara Ikuro, Yamakawa Kazuhiro
   Lab. Neurogenetics, Brain Science Institute, RIKEN

P-208  The study of syndrome of myoclonic absences
   * Ikeda Hiroko, Kubota Hidemoto, Kubota Yuko, Imi Katsumi, Shimomura Jiro, Shigematsu Hideo, Otani Sanae, Takahashi Hiroko, Mine Jyun, Takahashi Yukitsuki, Fujiwara Tateki
   National Epilepsy Center Shizuoka Institute of Epilepsy and Neurological Disorders

P-209  A patient with myoclonic astatic epilepsy presenting drop attacks induced by the sound stimulation
   * Matsumoto Ayumi, Yamagata Tanakori, Goto Tamako, Sugie Hideo, Momoi Mariko
   Department of Pediatrics, Jichi Medical University, Shimotsuke City, Japan
15:05 ∼ 15:40  Epilepsy 2

Chair: Shuto Yoshikawa, Mariko Maezawa

P-210 Epilepsy in patients with pervasive developmental disorder
* Shimakawa Shuichi¹, Tanabe Takuya², Wakamiya Eiji³, Hara Keita⁴, Tamai Hiroshi¹
1) Department of Pediatrics, Osaka Medical College, Osaka, Japan, 2) Department of Pediatrics, Hirakata municipal hospital, Osaka, Japan, 3) Department of Medical Health, Aino University, Osaka, Japan

P-211 The clinical characterization and differences of epilepsy with schizencephaly and porencephaly
* Shimizu Miki¹,², Maeda Tomoki³, Izumi Tatsuro²
1) Beppu Developmental Medicine and Habilitation Center, Oita, Japan, 2) Department of Brain and Nerve Science, Division of Pediatrics and Child Neurology, Oita University Faculty of Medicine, Oita, Japan

P-212 Clinical course of epilepsy secondary to neonatal hypoglycemia
* Montaser Hesham¹, Maegaki Yoshihiro⁴, Ohno Kousaku¹, Ogura Kaeko³
1) Division of Child Neurology, Institute of Neurological Sciences, Faculty of Medicine, Tottori University, Yonago, Japan, 2) Tohoku University, Sendai, Japan

P-213 Guidelines of driving licenses for persons with epilepsy - questionnaire survey in Hiroshima -
* Iyoda Kuniaki¹, Okazaki Tomio¹, Kishi Takamasa²
1) The Department of Pediatrics, Hiroshima City Hospital, Hiroshima, Japan, 2) The Department of Pediatrics, Hiroshima Memorial Hospital, Hiroshima, Japan

P-214 Atypical case with migrating partial seizures in infancy: good developmental catch-up.
* Ono Nobuyasu, Sugai Kenji, Sakuma Hiroshi, Saitou Yoshiaki, Komaki Hirofumi, Nakagawa Eiji, Sasaki Masayuki
National Center Hospital for Mental, Nervous and Muscular Disorders, National Center of Neurology and Psychiatry, Tokyo, Japan

P-215 Long QT syndrome showing with the cluster of generalized tonic clonic seizures
* Yamamoto Ayumi¹, Yamamoto Go¹, Ohfu Masaharu¹,²
1) Department of Pediatrics, Fukuoka Tokusyuikai Medical Center, Fukuoka, Japan, 2) School of Medicine, Department of Pediatrics, Fukuoka University, Fukuoka, Japan

P-216 A case of sudden unexpected death in Acute Encephalitis with Refractory, Repetitive partial seizure
* Fukuyama Tetsuhiro¹, Ishida Shuichi², Nagaharu Sachiko³, Misawa Yuka¹, Sekiguchi Yukio¹, Inaba Yuu¹, Higuchi Tukasa³, Koike Kenichi¹,², Awaya Yutaka²
1) Pediatrics, University of Shinshu, Matsumoto, Japan, 2) National hospital organization chushinnomatsumoto hospital, Matsumoto, Japan, 3) Advanced emergency and critical care center, Shinshu university hospital, Matsumoto, Japan, 4) Pediatrics, International Catholic Hospital

15:40 ∼ 16:15 Epilepsy 3

Chair: Kouzaburo Aso, Yasuhiro Suzuki

P-217 Visual cognitive function before and after epilepsy surgery for intractable epilepsy in infancy
* Ohashi Tsukasa¹, Nakagawa Eiji¹, Oe Hirokata², Sugai Kenji¹, Sasaki Masayuki¹, Kobayashi Iwao³
1) National Center Hospital for Mental, Nervous and Muscular Disorders, National Center of Neurology and Psychiatry, Tokyo, Japan, 2) National Center Hospital for Mental, Nervous and Muscular Disorders, National Center of Neurology and Psychiatry, Tokyo, Japan, 3) Center for the Research and Support of Educational Practice, Tokyo Gakugei University, Tokyo, Japan

P-218 Analysis of biomarker in an age dependent epileptic encephalopathy
* Yamanaka Gaku, Ishii Chiaki, Soganami Yusuke, Oana Shingo, Watanabe Kiyoko, Watanabe Toshiaki, Kawashima Hisashi, Miyajima Tasuku, Hoshika Akinori
The Department of Pediatrics, Tokyo Medical University, Tokyo, Japan
P-219 Risk for recurrence and outcome after a first unprovoked seizure in infancy
* Chu-Chin Chen, Hsueh-Ting Huang, Pao-Chin Chiu
Department of Pediatrics, Kaohsiung Veterans General Hospital, Kaohsiung city, Taiwan

P-220 Risk factors for treatment resistance in Panayiotopoulos syndrome
* Hirano Yoshiko, Oguni Hirokazu, Funatsuka Makoto, Imai Kaoru, Osawa Makiko
Dept of Pediatrics Tokyo Women’s Medical University, Tokyo, Japan

P-221 A case of Panayiotopoulos syndrome recorded ictal EEG
* Morimoto Kyoko
Pediatrics, Saiseikai Senri Hospital, Osaka, Japan

P-222 Involvement of the frontal foci in CSWS
* Kimura Kazue1, Hachimori Kei1, Nagao Yuri1, Ichikawa Kazushi2, Nezu Atsuo2,
Nomura Yoshiko1, Segawa Masaya1
1) Segawa Neurological Clinic for Children, Tokyo, Japan, 2) Department of pediatrics, Yokohama city university

P-223 An EEG analysis of epileptic syndromes both with diffuse spike and wave bursts and with focal spikes
* Katsumori Hiroshi, Shirakawa Seigo
Department of Pediatrics, Tokyo Rinkai Hospital, Tokyo, Japan

16:15 ~ 16:55 Epilepsy (medical treatment)
Chair: Hideo Nagao, Nobuaki Iwasaki

P-224 A case of malignant migrating partial seizures in infancy: Successful control with phenytoin
* Oyazato Yoshinobu, Nakagawa Taku, Okizuya Yo, Takami Yuichi, Matsuo Masahumi
Department of Pediatrics, Kobe University Graduate School of Medicine, Kobe, Japan

P-225 Efficacy of phenytoin for myoclonic status in progressive myoclonus epilepsies
* Miyahara Ayako, Sakuma Hiroshi, Saitou Yoshiaki, Komaki Hirohumi, Nakagawa Eiji, Sugai Kenji, Sasaki Masayuki
National Center of Neurology and Psychiatry, Musashi Hospital, Tokyo, Japan

P-226 Efficacy of gabapentin for catamenial epilepsy
* Takaishi Yasuko, Fujino Osamu, Kuwabara Kentaro, Okada Kazuyoshi, Fujita Takehisa, Kawakami Yasuhiko, Fujimatsu Moriko, Koizumi Shinya
Department of Pediatrics, Nihon Medical School, Tokyo, Japan

P-227 Clinical efficacy of gabapentin for intractable epilepsy
* Watanabe Yoshihiro1, Ichikawa Kazushi2, Takeshita Saoko2, Nezu Atsuo2
1) Department of Pediatrics, Saiseikai Yokohamashi Nanbu Hospital, Kanagawa, Japan, 2) Children’s Medical Center, Yokohama City University Medical Center, Kanagawa, Japan

P-228 The effectiveness of CBZ in two cases of Wolf-Hirschhorn syndrome with hemi-convulsion status.
* Oana Shingo, Watanabe Kiyoko, Watanabe Yoshiaki, Yamanaka Gaku, Miyajima Tasuku, Hoshika Akinori
Tokyo Medical University, Department of Pediatrics

P-229 The anticonvulsant action of topiramate
* Nagaki Shigeru, Takahashi Rieko, Osawa Makiko
Department of Pediatrics, Tokyo Women’s Medical University, Tokyo, Japan

P-230 The first inning high-dose phenobarbital with benign infantile convulsions with mild gastroenteritis
* Imataka George, Yamanouchi Hideo, Arisaka Osamu
Department of Pediatrics, Dokkyo University School of Medicine
P-231  Three cases of intractable epilepsy which responded to the regular use of diazepam suppository
  * Nigo Ayako1,2, Kumada Satoko1, Hoshino Ai1, Tomita Sunao1, Hanafusa Yukiko1, Kurihara Eiji1, Shimizu Hiroyuki1
  1) The Department of Neuropediatrics, Tokyo Metropolitan Neurological Hospital, Tokyo, Japan. 2) The Department of Pediatrics, Tokyo Metropolitan Fuchu Hospital, Tokyo, Japan. 3) The Department of Neurosurgery, Tokyo Metropolitan Neurological Hospital, Tokyo, Japan

16:55  17:35  Epilepsy 4
Chair: Akiko Matsumoto, Tetsuo Matsuzaka

P-232  The study of the very hemiplegia in symptomatic West syndrome
  * Shioda Mutsuki, Oguni Hirokazu, Ito Yasushi, Osawa Makiko
  Tokyo Women Medical University, Tokyo, Japan

P-233  A female case of West syndrome with remission after multiple hemorrhage
  * Fukuyo Naomi1, Haginoya Kazuhiro1,2, Uematsu Mitsugu1, Tusuchiya Shigeru1
  1) The Department of Pediatrics, School of Medicine, University of Tohoku, Sendai, Japan. 2) Takuto Rehabilitation Center for Children, Sendai

P-234  ACTH therapy for infantile spasms in the children with severe motor and intellectual disabilities
  * Kumada Tomohiro, Mikuni Takayasu, Kimura Nobusuke, Miyajima Tomoko, Fujii Tatsuya
  Department of Pediatrics, Shiga Medical Center for Children, Shiga, Japan

P-235  HIE
  * Tateishi Miho, Nabetani Makoto, Wada Hiroshi, Funato Masahisa
  Yokogawa Children Hospital, Osaka, Japan

P-236  A girl who had nonconvulsive status epilepticus induced by hyperzonisamidemia
  * Moriyama Nobuko1, Kikuchi Hitoshi1, Yamaoka Akiko1, Naoi Takahumi1, Iwasaki Nobuaki2
  1) Department of pediatrics, Ibaraki Children’s Hospital, Mito, Japan. 2) Department of pediatrics, Ibaraki Prefecture University of Health Sciences, Ami, Japan

P-237  Attempt of ketogenic diet in children with intractable epilepsy and congenital hyperlactacidemia
  * Kikuchi Atsu1,2, Uematsu Mitsugu1, Kobayashi Tomoko1, Matsumoto Yoko1, Wakuwasa Keisuke1, Nakayama Tojo1, Fukuyo Naomi1, Haginoya Kazuhiro1, Tuchiya Shigeru1
  1) Department of Pediatrics, Tohoku University School of Medicine, Sendai, Japan. 2) Department of Pediatrics, Ishinomaki Red Cross Hospital, Ishinomaki, Japan. 3) Takuto Rehabilitation Center for Children, Sendai, Japan

P-238  A case of Infantile epileptic encephalopathy treated with intravenous Magnesium
  * Kimura Shihoko, Kawakaki Hisashi, Hattori Taeka, Nukui Megumi, Kuki Ichirou, Okazaki Shin
  Department of pediatric neurology, Osaka City General Hospital, Osaka, Japan

P-239  Low-dose CBZ therapy and IV lidocane therapy for convulsions associated with mild gastroenteritis
  * Shirakawa Seigo, Katsumori Hiroshi
  Department of Pediatrics, Tokyo Rinkai Hospital, Tokyo, Japan

14:30  15:15  Imaging 2
Chair: Yasuhiko Kawakami, Masafumi Morimoto

P-240  Behavioral disorders and regional cerebral blood flow abnormality in Prader-Willi syndrome.
  * Ogura Keiko, Fuji Toshikatsu, Hosokai Yoshiyuki, Abe Nobuhito, Shinohara Mayumi, Mori Etsurou
  Department of Behavioral Neurology and Cognitive Neuroscience, Tohoku University Graduate School of Medicine, Sendai, Japan

P-241  Hemispheric hypoperfusion in a case of Sturge-Weber syndrome without leptomeningeal angioma
  * Yonee Chihiro, Toyoshima Mitsuo, Yoitsumata Kazuyuki, Kawano Yoshifumi
  Department of Pediatrics, Graduate School of Medical and Dental Sciences, Kagoshima University, Kagoshima city, Japan
P-242 Magnetencephalography and functional brain imaging analysis in epilepsy following West syndrome.
* Saeki Keisuke, Nakagawa Eiji, Sakuma Hiroshi, Saitou Yoshiaki, Komaki Hirofumi, Sugai Kenji, Sasaki Masayuki
National Center of Neurology and psychiatry, Tokyo, Japan

P-243 Delayed myelination at the onset of cryptogenic West syndrome
* Takano Tomoyuki, Shibata Masami, Hayashi Anri, Sokoda Tatsuyuki, Sakaue Yuko, Sawai Chihiro, Takeuchi Yoshihiro
Department of Pediatrics, Shiga University of Medical Science, Otsu, Japan

P-244 Three-dimensional ultrasound application in a congenital lipoma of corpus callosum
* Wei-Yuan Huang1,2, Nan-Chang Chiu1, Che-Sheng Ho1, Po-Lei Lee2
1) Department of Pediatric Neurology, Mackay Memorial Hospital, Taipei, Taiwan, 2) Institute of Brain Science, National Yang-Ming University, Taipei, Taiwan

P-245 Near infrared spectroscopy (NIRS) in patients with acute encephalopathy
* Ichikawa Kazushi, Takeshita Saoko, Nezu Atsuo
Institute of Brain Science, National Yang-Ming University, Taipei, Taiwan

P-246 Diffusion tensor image of influenza associated encephalopathy
* Ikeda Mitsu, Okumura Akihisa, Abe Shinpei, Saito Masako, Nakazawa Tomoyuki
Department of Pediatrics, Juntendo University School of Medicine

P-247 A contrast-enhanced MRI study in bacterial meningitis
* Harada Sayaka1, Tsujii Masahiro2, Haruta Tsunekazu1, Ueda Hiroyuki2
1) The Department of Pediatrics, Kobe City General Hospital, Kobe, Japan, 2) The Department of Radiology, Kobe City General Hospital, Kobe, Japan

P-248 A case of a girl with reversible splenium lesion associated with mycoplasma infection
* Tokunaga Yoichi1,2
1) Department of Pediatrics, Kyushu Rosai Hospital, Kitakyushu, Japan, 2) Kokura National Hospital

15:15 ～ 15:50 Imaging 3
Chair: Muneaki Matsuo, Eiji Nakagawa

P-249 Occurrence rate of lesion of central tegmental tract with Perinatal Brain Injury
* Kishino Ai, Sugama Seiichi
interdisciplinary Medicine,National center for Child Health and Developement,Tokyo,Japan

P-250 Clinical study of the 6 cases of ulegria patients.
* Tanabe Ryo1, Fujii Katsunori2, Endo Mamiko3, Maemoto Tatsuo3, Uchikawa Hideki3, Anzai Satoshi4, Yoshiihashi Manabu1, Kohno Yoichi4
1) Department of Pediatrics,Graduate School of Medicine,Chiba University,Chiba,Japan, 2) Department of Pediatrics,Asahi General Hospital,Chiba,Japan

P-251 Hypoxic encephalopathy in radiographic findings with severe anemia
* Kikuchi Kenjiro1, Eto Yoshikatsu2
1) Department of Pediatrics, Aoto Hospital, Jikei University School of Medicine, Tokyo, Japan, 2) Department of Pediatrics, Jikei University School of Medicine

P-252 Posterior Leuenecephalopathy syndrome in children; clinical and neuradiological findings.
* Koichihara Reiko1, Hamano Shinichirou1
1) Department of pediatrics, Fukaya Redcross Hospital, Saitama, Japan, 2) Division of Neurology, Saitama Childrens’ Medical Center
P-253 Diffusion-weighted MRI findings in a patients with medium-chain acyl-CoA dehydrogenase deficiency
*Yamaguchi Yui1, Kira Ryutarō1, Washito Natsumi1, Torisu Hiroyuki1, Sanefuji Masafumi1, Hasegawa Yuki1, Yamaguchi Seiji2, Hara Toshiro1
1) Department of Pediatrics, Graduate School of Medical Sciences, Kyushu University, Fukuoka, Japan, 2) Department of Pediatrics, Faculty of Medicine, Shimane University, Izumo, Japan

P-254 A case of Alagille syndrome with cerebral aneurysm and high T2 signal in the cerebral white matter.
*Shimozato Sachiko, Kosaki Kenjiro, Takahashi Takao
Department of Pediatrics, School of Medicine, Keio University, Tokyo, Japan

P-255 The progress of MRI findings in Lowe syndrome
* Hirayama Aya1, Makuta Masahiro1, Sawaishi Yukio1
1) Akita prefectural Rehabilitation And Nursery Center For Children with Disabilities, Akita, Japan, 2) Department of Pediatrics, Akita University, Akita, Japan

15:50 ~ 16:35 Peripheral nerve
Chair: Masayuki Shimono, Tsuyoshi Okinaga

P-256 Length of myelin internodes infected by adenovirus with wild type and mutated aprataxin cDNA
*Horimoto Yoshitaka1, Kikuchi Shin2, Kozuka Naoki3, Tachi Nobutada4
1) Nishiottaru Hospital, Otaru, Japan, 2) Department of Anatomy 1, School of Medicine, Sapporo Medical University, Sapporo, Japan, 3) Department of Physical Therapy, School of Health Sciences, Sapporo Medical University, Sapporo, Japan, 4) Department of Occupational Therapy, School of Health Sciences, Sapporo Medical University, Sapporo, Japan

P-257 Clinical manifestations of Charcot-Marie-Tooth disease type 1A in childhood
*Saito Naoki1, Fujii Katsunori1, Endo Mamiko1, Tanabe Ryō1, Kaneko Kenichiro2, Kohno Youichi1
1) Department of Pediatrics, Chiba University Graduate School of Medicine, 2) Department of Pediatrics, Juntendo University Urayasu

P-258 Clinical feature of Miller-Fisher Syndrome in children
*Sakakimoto Maiko1, Fujii Katsunori1, Arai Hidee2, Yoshihashi Manabu1, Endo Mamiko1, Omata Taku2, Honda Masakazu2, Ootake Akira1, Tanabe Yugo2, Kohno Yoichi1
1) Department of Pediatrics, Chiba University Graduate, Chiba, Japan, 2) Division of Neurology, Chiba Children's Hospital, Chiba, Japan, 3) Department of Pediatrics, Saitama Medical University, Saitama, Japan

P-259 A case of atypical Guillain-Barre syndrome with myalgia, muscle weakness, and ataxia
*Sato Ikuko1, Wakisawa Keisuke1, Kakisaka Yousuke1, Haginoya Kazuhiro3
1) Department of Pediatrics, Tohoku University School of Medicine, Sendai, Japan, 2) Department of Pediatrics, Kesennuma City Hospital, Kesennuma, Japan, 3) Division of Neurology, Takuto Rehabilitation Center for Disabled Children, Sndai, Japan

P-260 No symptom without pain in a case of acute sensory neuropathy associated with anti-GMI antibody
*Dejima Sunao, Ozaki Nozomu
Department of Pediatrics, Kyoto Min-iren Chuo Hospital

P-261 Acute autonomic and sensori-motor neuropathy caused by erythema infectiosum.
*Hanai Sae1, Sakuma Hiroshi1, Komaki Hieofumi1, Saito Yoshiaki1, Nakagawa Eiji1, Sugai Kenji1, Sasaki Masayuki1, Higurashi Norimichi2, Hamano Shin-ichiro2
1) National Center Hospital for Mental, Nervous and Muscular Disorders, National Center of Neurology and Psychiatry, Tokyo, Japan, 2) Division of Neurology, Saitama Children's Medical Center, Saitama, Japan

P-262 An atypical case of childhood-onset chronic inflammatory demyelinating polineuropath
*Inoue Takehiko1, Saito Yoshiaki1, Maegaki Yoshihiro1, Ohno Kosaku1, Fukuda Chisako2, Tomita Yutaka2
1) Department of Child Neurology, Faculty of Medicine, Tottori University, Yonago, Japan, 2) Department of Pathological Science and Technology, School of Health Science, Faculty of Medicine, Tottori University, Yonago, Japan
P-263 12 years follow up of a case with giant axonal neuropathy harboring novel mutation
* Morita Yoshiko1, Akagi Motohiro2, Mohri Ikuko3, Katho Kumi4, Kitai Yukihiro1, Araya Ken1, Tominaga Koji1, Shimono Kuriko1, Okinaga Tsuyoshi1, Sakai Norio1, Tanaka Misako1, Ozono Keichi1
1) The Department of Pediatrics, Osaka University Graduate School of Medicine, Osaka, Japan, 2) Department of Pediatrics Otemae Hospital, Osaka, Japan, 3) The Research Center for Child Mental Development, Osaka University Graduate School of Medicine, Osaka, Japan

P-264 Effectiveness of gabapentin for neuropathic pain after an episode of hypovolemic shock
* Higuchi Tsukasa, Nagaharu Sachiko, Misawa Yuka, Fukuyama Tetsuhiro, Inaba Yuji, Koike Kenichi
The Department of Pediatrics, Shinshu University School of Medicine, Matsumoto, Japan

16:35 〜 17:10  Morphology/Pathology
Chair: Keiko Shishikura, Tomoyuki Takano
P-265 Effect of hypoxia on the 5HT projection to the spinal motoneurons in neonatal rats
* Tanaka Hajime1, Amами Amami Satoshi2, Takahashi Satoru1, Suzuki Nao1, Araki Akiko1, Ohinata Junko3, Fujieda Kenji1
1) Department of Pediatrics, Asahikawa Medical College, Asahikawa, Japan, 2) Department of Pediatrics, Asahikawa Kosei Hospital, Asahikawa, Japan

P-266 Morphological development of the human abducens nucleus
* Yamaguchi Katsuyuki12
1) Department of Pathology, Dokkyo University School of Medicine, Tochigi, Japan, 2) Department of Pediatrics, Southern Tohoku General Hospital

P-267 Tau-Cre knockin mice mediate highly efficient and specific Cre/loxP recombination in the neuron.
* Muramatsu Kazuhiro12, Ogata Tomomi1, Sawaura Noriko1, Harada Akihiro2, Morikawa Akihiro1
1) Department of Pediatrics and Developmental Medicine, Graduate School of Medicine, Gunma University, Gunma, Japan, 2) Laboratory of Molecular Traffic, Department of Molecular and Cellular Biology, Institute for Molecular and Cellular Regulation, Gunma University

P-268 Dscam1 knockout mice show abnormal central respiratory pattern
* Amano Kenji1, Fujii Morimitsu1, Arata Akiko2, Yamakawa Kazuhiro1
1) Laboratory for Neurogenetics, RIKEN Brain Science Institute, Saitama, Japan, 2) Laboratory for Memory and Learning, RIKEN Brain Science Institute, Saitama, Japan

P-269 Identification of increased HEL- and HNE-adducts in the brain of a mouse model for Down Syndrome
* Ishihara Keichi, Yamakawa Kazuhiro
Laboratory for Neurogenetics, RIKEN Brain Science Institute, Saitama, Japan

P-270 Behavioral Phenotyping of the Ts1Cje mouse: a model for Down syndrome
* Shimohata Atsushi, Yamakawa Kazuhiro
Lab. for Neurogenetics, RIKEN-BSI, Saitama, Japan

P-271 Evolution of early brain development: from a comparative study with chimpanzees in amygdala
* Sakai Tomoko, Mikami Akichika
Department of Behavioral and Brain Sciences, Primate Research Institute, Kyoto University

17:10 〜 17:55  Metabolic/Degenerative disorders
Chair: Kazutoshi Nakano, Hitoshi Osaka
P-272 Two cases of recurrent encephalopathy without apparent mitochondrial DNA abnormalities
* Kohira Ryutarou, Imai Yuki, Enndyo Ayumi, Arakawa Chikako, Fujita Yukihiko, Fuchigami Tatsuo
Nihon University Schhol of Medicine, Tokyo, Japan
P-273  A boy with spinocerebellar ataxia type2 who became symptomatic at 4 years of age
* Kishi Kazuko1, Shibata Naoaki2, Sejima Hitoshi1, Yamaguchi Seiji1, Eda Isema2
1) Department of Pediatrics, Shimane University Faculty of Medicine, Shimane, Japan, 2) Department of Pediatrics, Seibu-Shimane medical welfare center, Simane, Japan

P-274  The patient with neurological type of Wilson disease, onset with microhematuria
* Shimizu Norikazu, Aoki Tsugutoshi
The Second Department of Pediatrics, Toho University School of Medicine, Tokyo, Japan

P-275  A case of H-ABC syndrome with the symptoms of spastic diplegia
* Yoshihiko Hiroshi1, Hiroya Shibuya2, Yoko Saito2, Hayakawa Katsumi2
1) St.Joseph Hospital for people with handicaps,St.Joseph Medical and welfare center, Kyoto, Japan, 2) The Department of Radiology, Kyoto City Hospital, Kyoto, Japan

P-276  Characteristics of Glucose Transporter Type 1 Deficiency Syndrome with a T295M Mutation
* Fujii Tatsuya1, Morimoto Masafumi2, Yoshihiko Hiroshi1, Hiroya Shibuya1, Law Peggy4, Wang Dong5, De Vivo Darryl C5
1) Department of Pediatrics, Shiga Medical Center for Children, Moriyama, Japan, 2) Department of Pediatrics, Kyoto Prefectural University of Medicine, Kyoto, Japan, 3) Yoshihiko Children’s Clinic, Kyoto, Japan, 4) Department of Biochemistry, The Chinese University of Hong Kong, Hong Kong, China, 5) Colleen Giblin Laboratories for Pediatric Neurolgy Research, Department of Neurology, Columbia University, New York, U.S.A

P-277  Improved EEG and symptoms by glucose injection in a child with Glut-1 deficiency syndrome
* Tsurui Satoshi1, Seki Katuyuki1, Ohzeki Takehiro2, Yanagihara Keiko3
1) The Department of Pediatrics, Seirei Numazu Hospital, Numazu, Japan, 2) Hamamatsu University School of Medicine, Department of Pediatrics, Hamamatsu, Japan, 3) Research Institute Osaka Medical Center for Maternal and Child Health, Osaka, Japan

P-278  A case of bilateral striatal necrosis who had progressive dystonia with metabolic acidosis.
* Ishikawa Aki1, Kato Takahiro1, Ohya Kazuhiro1, Tachi Nobutada2, Goto Yuichi2
1) The Department of Pediatrics, Sapporo Medical University School of Medicine, Sapporo, Japan, 2) Sapporo Medical University School of Health Science, Sapporo, Japan, 3) National center of Neurology and Psychiatry, National Institute of Neuroscience, Japan

P-279  The ultrastructural study of the small vessels in patients with alternative hemiplegia of childhood
* Sasaki Masayuki1, Sugai Kenji1, Nakagawa Eiji1, Saito Yoshiaki1, Komaki Hirofumi1, Sakuma Hiroshi1, Arima Kunimasa2
1) The Department of Child Neurology, Musashi Hospital, National Center of Neurology and Psychiatry, Tokyo, Japan, 2) The Department of Clinical Medicine, Musashi Hospital, National Center of Neurology and Psychiatry, Tokyo, Japan

P-280  The case of Mucolipidosis type III with characteristic pathological findings
* Kobayashi Hiroshi1, Fujigasaki Junko2, Fukuda Takahiro1, Sakurai Ken1, Ida Hiroyuki1, Ohashi Toya1, Eto Yoshishigato1
1) The Department of Pediatrics, Jikei University School of Medicine, 2) Department of Neuropathology, Jikei University school of medicine, Tokyo, Japan, 3) Department of Gene Therapy, Institute of DNA Medicine, Jikei University school of medicine, Tokyo, Japan

14:30 ~ 15:10  Development
Chair: Masumi Inagaki, Satoshi Takada

P-281  When dose the handedness appear?
* Shiotani Yuka1, Matuzawa Shigeyuki1, Sawada Akiko1, Yoshida Yumi1, Awaaya Tomonari2, Okada Masako1, Ikeda Hiroko1, Tomiwa Kiyotaka1
1) Japan Science and Technology Agency, Osaka, Japan, 2) Kyoto University Hospital, Kyoto, Japan, 3) National Epilepsy Center, Shizuoka Institute of Epilepsy and Neurological Disorders, Shizuoka, Japan
P-282  Handness in children and their parental wish
  * Shiotani Yuka1, Sawada Akiko1, Matuzawa Shigeyuki1,2, Yoshida Yumi1,
  Awaysa Tomonari1,2, Okada Masako1,2, Ikeda Hiroko1, Tomiwa Kiyotaka1,2
  1) Japan Science and Technology Agency, Osaka, Japan, 2) Kyoto University Hospital, Kyoto, Japan, 3) National
  Epilepsy Center, Shizuoka Institute of Epilepsy and Neurological Disorders, Shizuoka, Japan

P-283  Laterality in infants and handedness of parents-JCS:laterization and development of sociability
  * Matsuzawa Shigeyuki1,2, Shiotani Yuka1, Sawada Akiko1, Yoshida Yumi1,
  Awaysa Tomonari1,2, Okada Masako1,2, Ikeda Hiroko1, Tomiwa Kiyotaka1,2
  1) JST/RISTEX, 2) Kyoto university graduate school of medicine, 3) Shizoka Institute of Epilepsy and Neurological
  Disorders

P-284  Head growth evaluation in early childhood,from JCS :Measurements of physical growth and sociability.
  * Sawada Akiko1, Matsuzawa Sigeyuki1,2, Shiotani Yuka1, Yoshida Yumi1,
  Awaysa Tomonari1,2, Okada Masako1,2, Ikeda Hiroko1, Tomiwa Kiyotaka1,2
  1) Osaka Research Group, Japan Children Study (JST), 2) Kyoto University, Graduate School of Medicine,
  3) National Epilepsy Center, Shizuoka

P-285  A study for medical examination in cohort study-Reevaluation based on diagnosis of 6 years old-
  * Seki Ayumi1,2, Takeuchi Arik01,2, Koeda Tatsuya1,2
  1) Japan Children's Study Group, JST/RISTEX, 2) Department of Education, Faculty of Regional Sciences, Tottori
  University, Tottori, Japan

P-286  Five-year-old health examination in public health center of Namegata-shi Ibaraki prefecture
  * Suzuki Naomitsu
  Department of Pediatrics, Namegata District General Hospital

P-287  Development in visual cognitive function during Childhood using exploratory eye movements
  * Egami Chiyomi1,2,3, Morita Kiichiro1, Ishii Youhei3, Yamashita Yushiro7,
  Matsuishi Toyojiro2
  1) Technical school medical and welfare Ryokuseican, 2) Kurume University School of Medicine, 3) Cognitive and
  Molecular Research Institute of Brain Diseases, Kurume University

P-288  Group therapy for infants with Down Syndrome at Shimada Ryoiku Center
  * Kamiishi Akiko, Ohtaki Ushio, Saski Kyoko, Nakajima Suemi, Ozawa Hiroshi, Ishizuka Takehiro,
  Arimoto Kiyoshi, Kimiya Satoshi
  Shimada Ryoiku Center, Tokyo, Japan

15:10 ～ 15:50  Cerebral palsy Rehabilitation 1
  Chair: Mana Kurihara, Atsushi Ieshima

P-289  Examination of the cerebral paralysis child who was born by low birth weight child or multiple birth
  * Shishido Atsushi1,2, Ariga Masamichi1, Hayakawa Mika1, Imai Masayuki1, Ochiai Yukikatsu1, Etou Yosikatsu2
  1) Department of Pediatrics, Metropolitan Kita Sodairyou Central Department, Tokyo, Japan, 2) Department of
  Pediatrics, The Jikei University School of Medicine, Tokyo, Japan

P-290  Postural reactions of walkable children with cerebral palsy
  * Yamori Yuriko, Yoshida Naoko, Hirota Haruyo, Kanda Toyoko
  The Department of Pediatrics, St. Joseph Medical and Welfare Center, Kyoto, Japan

P-291  The five cases of femoral fracture with cerebral palsy
  * Ando Naoki, Kobayashi Satoru, Hattori Ayako, Ito Tetsuya, Togari Hajime
  Department of Pediatrics, Neonatology and Congenital Disorders, Nagoya City University, Graduates School of Medical
  Sciences, Nagoya, Japan
P-292  Treatment with autologous umbilical cord blood for infantile or childhood cerebral palsy
*Chuan-yu Wang1, Kuan-sheng Chou2, Men-yao Lu3, Kai-hsin Lin4, Dah-chin Yan5, Tzou-yien Lin6
1) Taipei Division of Pediatric Neurology, Chang Gung Children Hospital, Tao-Yuan, Taiwan, 2) Pediatric Hematology Oncology, National Taiwan University Hospital, Taipei, Taiwan, 3) Taipei Division of Pediatric Immunology, Chang Gung Children Hospital, Tao-Yuan, Taiwan, 4) Pediatric Infection, Chang Gung Children Hospital, Tao-Yuan, Taiwan

P-293  Milieu therapy in institution for physically disabled children by the method of parent training
*Nagase Mika1, Kita Michiko2, Takahashi Nagahisa3, Yoneyama Akira4, Kitazumi Eiji5
1) Department of Pediatrics, The National Rehabilitation Center for Disabled Children, 2) Department of Child Psychiatry, The National Rehabilitation Center for Disabled Children

P-294  Social Skills Training for children long-term living in institute for motor disabled children
*Nagase Mika, Nagase Mika, Kita Michiko, Yoneyama Akira, Kitazumi Eiji
Department of Pediatrics, The National Rehabilitation Center for Disabled Children, Tokyo, Japan

P-295  The problems in foster care and welfare system: a case report of abused child with cerebral palsy
*Miyamoto Akiie, Fukuda Ikue, Oka Ryuji, Cho Kazuiko
The Department of Pediatrics, Asahikawa Habilitation Center for Disabled Children, Asahikawa, Japan

P-296  Lifespan related diseases of the adults with severe motor and intellectual disabilities
*Sone Sui, Araki Katsuhito, Kurata Kiyoko
Tokyo Metropolitan Higashiyamato Medical Center for Disabilities, Tokyo, Japan

15:50 ~ 16:20  Cerebral palsy Rehabilitation 2
Chair: Masaru Tatsuno, Tadashi Kitahara

P-297  Education resource for medical care produced by Osaka Medical Association
*Hattori Hideji1, Nagai Toshisaburou2,3, Funato Masahisa1,3, Tagawa Tetsuzo6,7, Negishi Hirokuni1,2, Funato Masahisa1,3
1) Department of Pediatrics, Osaka City University Graduate School of Medicine, 2) Division of Health Sciences, Osaka University Graduate School of Medicine, 3) Department of Pediatrics, Yodogawa Christian Hospital, 4) Department of Pediatrics, Osaka Kousei-Nenkin Hospital, 5) Aijinkai Takatsuki Hospital, 6) Committee of Medical Care for children, Osaka Medical Association

P-298  Investigation of medical care in children or persons with severe motor and intellectual disabilities
*Abe Junko, Nagae Akiko, Fujita Yasuyuki, Kumode Masao
Biwako Gakuen Kusatsu Medical and Welfare Center

P-299  A questionnaire survey to the school doctors about the medical care for the children in their school
*Yokokawa Shinobu1, Nagai Toshisaburou2,3, Ikeda Tomomi1,2, Okuno Hiroko1, Sugiura Keiko1, Takama Satomi1, Tagawa Tetsuzo2,3, Negishi Hirokuni1,2, Hattori Hideji2,3, Funato Masahisa1,3
1) Division of Health Sciences, Osaka University Graduate School of Medicine, Osaka, Japan, 2) Hyogo University, Hyogo, Japan, 3) Department of Pediatrics, Osaka Koseinenkin Hospital, Osaka, Japan, 4) Aijinkai Healthcare Corporation, Osaka, Japan, 5) Department of Pediatrics, Osaka City University Graduate School of Medicine, Osaka, Japan, 6) Department of Pediatrics, Yodogawa Christian, Osaka, Japan, 7) Osaka medical association, Osaka, Japan

P-300  A questionnaire survey on medical-school-education and medical care in Ibaraki
*Ohtoshi Tarō1, Takada Satoshi2, Iwasaki Nobuaki3
1) Division of Occupational Therapy, Faculty of Rehabilitation, Seijoh University, Tokai, Japan, 2) Faculty of Health Science, Kobe University, Kobe, Japan, 3) Department of Pediatrics, Ibaraki Prefectural University of Health Science, Inashiki, Japan

P-301  New degree of medical needs for children or persons with severe motor and intellectual disabilities
*Nagae Akiko1, Abe Junko1, Fujita Yasuyuki2, Kumode Masao2, Yokochi Kenji2
1) Biwako Gakuen Kusatsu Medical Welfare Center, Shiga, Japan, 2) Seirei Ohzora Ryoiku Center
P-302 Present situation of medical practice after adolescence in institution with psychomotor disabilities
  * Otani Ryoko1, Matsui Mihoko1, Sakuta Ryoichi2
  1) Jyohoku Branch Kita Medical and Rehabilitation Center for Disabled, Tokyo, Japan, 2) Department of Pediatrics
  Dokkyo Medical University Koshigaya Hospital, Saitama, Japan

16:20 ～ 17:05 Cerebral palsy Complications
  Chair: Masayuki Sasaki, Yukikatsu Ochiai

P-303 Hyponatremia in patients with severe motor and intellectual disabilities syndrome
  * Gotoh Harumi, Suzuki Ikuko, Maruki Kazuko
  Hikarino-ie Institute, Moro Hospital, Moroyama, Saitama, Japan

P-304 A case of severe anemia and agranulocytosis by copper deficiency during enteral nutrition
  * Nagaharu Sachiko, Inaba Yuji, Misawa Yuka, Fukuyama Tetsuhiro, Higuchi Tsukasa, Koike Kenichi
  The Department of Pediatrics, Shinshu University School of Medicine, Matsumoto, Japan

P-305 Purple glove syndrome in two patients with severe motor and intellectual disabilities
  * Katsura Chiaki, Nakagawa Eiji, Saeki Keisuke, Sakuma Hiroshi, Saito Yoshiaki, Komaki Hirofumi, Sugai Kenji,
  Sasaki Masayuki
  National Center of Neurology and Psychiatry, Musashi Hospital, Tokyo, Japan

P-306 2 cases of Fanconi syndrome with severe motor and intellectual disabilities triggered by infections
  * Niimi Taemi, Ishida Shuichi
  National Hospital Organization Chushinmatsumoto Hospital, Matsumoto, Japan

P-307 Neuroleptic malignant syndrome induced by a combined administration of antiepileptic drugs
  * Kamiya Yuhko1, Nakane Takaya1, Hatakeyama Kazuo2, Nakamura Kousuke2, Noguchi Sayaka1, Sugita Kanji2, Aihara Masao2
  1) National Hospital Organization Kofu Hospital, Yamanashi, Japan, 2) Department of Pediatrics, University of
  Yamanashi, Yamanashi, Japan

P-308 Pancreatitis in patients with severe motor intellectual disability syndrome
  * Suzuki Yume, Mori Masato, Goto Tomako, Yamagata Takanori, Momoi Mariko
  Jichi Medical University, Department of Pediatrics, Tochigi, Japan

P-309 Coagulopathy for vitamin K deficiency in severe mental and motor retardation patients
  * Arai Asako, Sakuma Kei, Saito Yoshiaki, Komaki Hirofumi, Nakagawa Eiji, Sugai Kenji, Sasaki Masayuki
  Department of Child Neurology, National Center Hospital of Mental, Nervous and Muscular Disorders, Tokyo, Japan

P-310 Anemia in the patients receiving enteral nutrition
  * Fujita Hitomi, Suzuki Yume, Goto Tomako, Suwa Kiyotaka, Mori Masato, Yamagata Takanori, Momoi Mariko
  Department, Jichi Medical University

P-311 Academic disability in a severe motor-intellectual disabilities child with cyclic vomiting syndrome
  * Nishimoto Yukihiro
  Department of Pediatrics, Nanki Fukushima Center, Wakayama, Japan

17:05 ～ 17:45 Cerebral palsy/Involuntary movement
  Chair: Kazuo Higuchi, Toyoko Kanda

P-312 Postoperative course of functional posterior rhizotomy
  * Toyama Jun, Touyama Mayumi
  Okinawa Child Development Center, Okinawa, Japan
P-313  Deep brain stimulation for generalized dystonia in a patient with Hallervorden-Spatz disease
*Tachikawa Emiko1, Funatsuka Makoto1, Tamiya Sayaka1, Nakatsukasa Hidetsugu1, Fujii Akiko1,
Kodaira Kayano1, Sakauchi Masako1, Ochiai Takashi1, Taira Takaomi1, Ohsawa Makiko1
1) Pediatric Department, Tokyo Women's Medical University, Tokyo, Japan. 2) Department of Neurosurgery,
Neurological Institute, Tokyo Women's Medical University, Tokyo, Japan

P-314  A case of xeroderma pigmentosum group A which presented laryngeal dystonia and a catatonic state.
*Ideguchi Hiroshi, Ninomiya Sinnya, Ibara Yukiko, Tomonou Yuuko, Hujita Takako, Nakamura Noriko,
Inoue Takahito, Yasumoto Sawa, Hirose Shinti
1) Department of Pediatrics, University of Hukuoka, Hukuoka, Japan

P-315  Percutaneous endoscopic gastrostomy in neurologically disabled children improves quality of life.
*Takekita Saoko1, Hirasawa Kingo1, Yahara Sei1, Ichikawa Kazushi1, Nezu Atsuo1
1) Children Medical Center, Yokohama City University Medical Center, Yokohama, Japan. 2) Gastroenterological
Center, Yokohama City University Medical Center, Yokohama, Japan

P-316  The examination of Home Oxygen Therapy in patients with severe motor and intellectual disabilities
*Matsufuji Hironori, Sueyama Naoko, Nishikawa Miki, Sugio Yoshitsugu
Department of Pediatrics, Tsuzumigaura Handicapped Children's Hospital, Syunan, Japan

P-317  NIV for Chronic Respiratory Failure of Severe motor and intellectual disabilities syndrome
*Kodama Mariko1, Yoneyama Akira1, Murayama Keiko1,2, Hashimi Hiroki1, Nakatani Katutoshi1,
Shinozaki Yuuko1, Anzai Yuki1, Kitazumi Eiji1
1) The National Rehabilitation Center for Disabled Children. 2) Genki Clinic, 3) National Center of Neurology and
Psychiatry Musashi Hospital. 4) Saiseikai Yokohama Eastern Hospital Children's Center for health and developement

P-318  Prevention from aspiration pneumonia with botulinum toxin type A injection to salivary glands
*Soeibjanto Keiji, Sasazuki Momoko, Sakamoto Kei, Aibe Miyuki, Mizuno Yuji
Department of pediatrics, East Fukuoka Medical Center, Fukuoka, Japan

P-319  Treatment of zinc suplementation for severe handicapped people
*Wada Keiko, Tyou Hiroyuki, Funahashi Masuko, Suzuki Yasuyuki
Tokyo childrens rehabilitation hospital, Tokyo, Japan
8:00 ～ 9:00  Morning Seminar 4
A Clinical Approach to the Dysmorphic Child
Chairman：Hitoshi Yamamoto (Department of Pediatrics, St.Marianna University School of Medicine)
Kenjiro Kosaki (Department of Pediatrics, Keio University School of Medicine)

9:00 ～ 9:40  Didactic Lecture 8
Regulatory mechanism of neuronal migration mediated by the microtubule-associated protein doublecortin and its partners
Chairman：Takao Takahashi (Department of Pediatrics, School of Medicine, Keio University)
Teruyuki Tanaka (Department of Developmental Medical Sciences, Graduate School of Medicine, The University of Tokyo)

09:40 ～ 10:50  Sleep/Behavior disorders
Chair: Jun Kohyama, Katsuo Sugita
O-171 Characteristics of night sleep among preterm infants at 12 months of age
* Asaka Yoko1,2, Matsui Gakyou1, Takada Satoshi2
1) Konan Women's University, 2) Kobe University Graduate School of Medicine, Faculty of Health Sciences, Kobe, Japan

O-172 Relationship between sleep problems and behavior problems among primary school children
* Oka Yasunori1,2, Suzuki Shuhei1
1) Japan Somnology Center, Neuropsychiatric Research Institute, Tokyo, Japan, 2) Department of Developmental Brain Science, Osaka Medical College, Osaka, Japan

O-173 The lack of REM sleep induced by cavernoushemangioma in brainstem
* Shimono Kuriko1, Kato Kumi2, Kitai Yoshihiro2, Araya Ken1, Tominaga Kouji1,
Okinaga Takeshi1, Mohri Ikuko1, Iwata Masako2, Ozono Keichi1
1) Department of Pediatrics, Osaka University Graduate School of Medicine, Osaka, Japan, 2) Osaka University Graduate School of Medicine, Department of Mental Health and Environmental Effects Research, Molecular Resarch Center for Child Mental Development

O-174 Melatonin therapy for the developmental disorders with sleep disturbance—the report of the 178 cases
* Ishizaki Asayo, Sugama Michiko, Takeuchi Noriko
Ouji clinic; Division of Medicine, The Association of Remedial Teaching for People with Developmental Handicaps

O-175 Neuronal Antibody in neuropsychiatric disorders
* Hongou Kazuhiwa, Harai Tomomi, Fujiki Yasuko, Miya Kazushi, Kageyama Ryuujii, Tanaka Chiaki, Yagi Shinich, Honma Kazumasu, Miyawaki Toshio, Endou Shouti
Department of Pediatrics Toyama University, Toyama, Japan

O-176 Medication of Oseltamivir to patients with abnormal behaviors associated with influenza infection
* Tanabe Takuya1, Hara Keita1, Tominaga Miwa1, Shimakawa Shuichi1, Tamai Hiroshi2
1) Division of Pediatrics, Hirakata City Hospital, Osaka, Japan, 2) Department of Pediatrics, Osaka Medical College, Osaka, Japan

O-177 Infants with inflicted brain injury present without history of injury, but with neuologic symptom.
* Nagase Hiroaki1, Okuyama Makiko2, Aoki Kazunori1, Maruyama Azusa1
1) Pediatric Neurology, Kobe Children’s Hospital, Kobe, Hyogo, Japan, 2) Department of Psychosocial Medicine, National Center for Child Medical Health and Development, Tokyo, Japan

10:50 ～ 11:00  Closing address
8:00 ～ 9:00  Morning Seminar 5
Pediatric neuroimaging diagnosis A to Z.
Chairman  Toshiaki Hashimoto（Naruto University of Education）
Hiroshi Oba（Department of Radiology, Teikyo University School of Medicine）

09:00 ～ 09:50  Involuntary movement/Epilepsy（PC movie）
Chair: Shuji Wakai, Kazue Kimura
O-178  A case of paroxysmal exertion-induced dyskinesia
* Satou Takatoshi , Itoh Yasushi  , Etou Kaoru , Abe Kazuyo , Nishikawa Aiko , Fujii Akiko , Imai Kaoru , Ogumi Hirokazu , Osawa Makiko
1) Department of Pediatrics, Tokyo Wemen's Medical University, Tokyo, Japan. 2) Department of Pediatrics, Tsuchiya Children’s Hospital

O-179  Haloperidol effective dystonia in 2 patients with tonic cluster formation like movement.
* Sekiguch Kazuhiro , Miyahara Hiroaki , Shimizu Miki , Maeda Tomoki , Akiyoshi Kennsuke , Izumi Tatsuro
Oita university faculty of medicine , oita, japan, Japan

O-180  A case with paroxysmal tonic upgaze of childhood
* Enoki Hideo
Department of Child Neurology, Seirei Hamamatsu General Hospital, Hamamatsu, Japan

O-181  A case of focal-onset periodic spasms cured by ganglioneuroma resection
* Awaya Tomonari , Osada Kazuko , Shibata Minoru , Yamanaka Yasunari , Kato Takeo , Nakahata Tatsutoshi , Tomiwya Kiyotaka
1) Department of Pediatrics, Graduate School of Medicine, Kyoto University, Kyoto, Japan. 2) Genetic Counselling and Clinical Research Unit, School of Public Health, Kyoto University, Kyoto, Japan

O-182  A boy with spasms following tonic seizures without hypsarrhythmia
* Watanabe Toshihide , Minagawa Kimio
Department of Pediatrics, Hokkaido Medical Center for Child Health and Rehabilitation

09:50 ～ 10:50  Epilepsy/Seizures 4
Chair: Harumi Yoshinaga, Toshizaburo Nagai
O-183  Prppofol is effective at epileptic status of 3cases.
* Hosoya Machiko
Pediatrics, Saku Central Hospital, Nagano, Japan

O-184  Effectiveness of gabapentin with intractable partial seizures in childhood and adolescence
* Iwasaki Toshiyuki , Nonoda Yutaka , Takei Kenji , Hosoda Nozomi , Ishii Masahiro
1) Department of Pediatrics, Kitasato University School of Medicine, Kanagawa, Japan. 2) Sagamihara Ryouikuen Institute for Severe Motor and Intellectual Disabilities, Kanagawa, Japan

O-185  Longterm efficacy of gabapentin for intractable epilepsy in childhood
* Okazaki Shin , Kawakami Hisashi , Hattori Taeka , Nukui Megumi , Kuki Ichiro , Kimura Shihoko , Ishikawa Junichiro , Togawa Masao , Shiomi Masashi , Tomiwa Kiyotaka
1) Department of pediatric neurology, Children's Medical Center, Osaka City General Hospital, 2) Department of pediatric emergency medicine, Children's Medical Center, Osaka City General Hospital, 3) Infection Center, Osaka City General Hospital, 4) Geneticcounselor coordinator Unit, Kyoto University

O-186  Efficacy of bromide for refractory epilepsies
* Sugai Kenji , Sakuma Hiroshi , Komaki Hirofumi , Nakagawa Eiji , Sasaki Masayuki
Department of Child Neurology, National Center of Neurology and Psychiatry, Japan
O-187 Reevaluation of the ketogenic diet therapy for childhood intractable epilepsy
  * Oguni Miyako, Oguni Hirokazu, Osawa Makiko, Ito Yasushi, Ito Susumu
  1) Yamawaki Gakuen Junior College, Tokyo, Japan, 2) The Department of Pediatrics, Tokyo Women's Medical University, Tokyo, Japan

O-188 Serum uric acid concentrations in newly diagnosed epileptic children treated with valproate
  * Kishi Takamasa
  Department of Pediatrics, KKR Hiroshima Memorial Hospital, HIroshima, Japan

12:00～16:00 Seminars open to the public
How we can help Japanese children to be happy, thoughtful and warmhearted?

8:00～9:00 Morning Seminar 6
Visually diagnosed epileptic seizures: generalized seizures
Chairman: Shunsuke Otawara (Okayama University)
Hirokazu Oguni (Department of Pediatrics, Tokyo Women's Medical University)

09:00～09:50 Supportive diagnosis (PC movie)
Chair: Takanori Yamagata, Ryouchi Sakuta

O-189 The development of visual function in healthy and high risk infants using the eye tracking system
  * Yamaguchi Fumioka, Hirasawa Kyoko, Tamura Masanori
  1) Saitama Medical University, Saitama Medical Center, Kawagoe, Japan, 2) Tokyo Women's Medical University, Tokyo, Japan

O-190 Dysphagia in Fukuyama congenital muscular disease
  * Mori Yuka, Kibe Tetsuya, Yokochi Kenji
  Department of Pediatrics, Seirei Mikatahara General Hospital

O-191 Familial Marcus Gunn Syndrome
  * Ikeda Kayo, Yanagihara Keiko, Arai Hiroshi
  1) Rinku General Medical Center, 2) Osaka Medical Center and Research Institute for Maternal and Child Health,
  3) Morinomiya Hospital

O-192 A case of localized encephalitis diagnosed by FDG-PET
  * Sekigawa Mariko, Niijima Shinti, Iijima Yoshitaka, Iwasaki Tomohiro, Kamata Ayako, Tahara Kanako,
  Suzuki Ryuyo, Watanabe Naoki, Ootomo Yoshiyuki, Okumura Akihisa, Hayashi Masaharu
  1) Department of Pediatrics, Juntendo University Nerima Hospital, Tokyo, Japan, 2) Department of Pediatrics, Juntendo University,
  3) Department of Clinical Neuropathology, Tokyo dMetropolitan Institute for Neuroscience

O-193 Tele-home-care for Severe Motor and Intellectual Disabilities by Cellular-phone Network
  * Fueki Noboru, Fukuyama Tetsuhiro, Ishida Shuichi, Hirabayashi Shinichi, Hirano Satoru
  1) The Department of Rehabilitation, Nagano Children's Hospital, Nagano, Japan, 2) The Department of Pediatrics,
  University of Shinshu, Nagano, Japan, 3) The Department of Pediatrics, Chushimatsu Hospital, Nagano, Japan,
  4) The Department of Child Neurology, Nagano Childrens Hospital, Nagano, Japan

09:50～10:50 Dystonia (PC movie)
Chair: Akihiko Tateno, Tetsuzo Tagawa

O-194 Epidemiological survey of childhood-onset dystonia in Japan by child neurology specialists
  * Funatsuka Makoto, Kato Ikuko, Osawa Makiko
  Department of Pediatrics, Tokyo Women's Medical University, Tokyo, Japan
O-195  Pteridine analysis in cerebrospinal fluid and plasma for the screening of dopa-responsive dystonia
  "Fujoka Hiroki1, Shintaku Haruo1, Hirabayashi Shinichi2, Yamano Tsunezaku2
  1) Department of Pediatrics, Osaka City University Graduate School of Medicine, 2) Department of Neuropediatrics, Nagano Children's Hospital

O-196  A case of early-onset primary torsion dystonia presenting with cervical dystonia.
  "Kumada Satoko1, Yokochi Fusako2, Taniguchi Makoto3, Okumura Sayaka1,
  Hoshino Ai1, Hanafusa Yukiko1, Tomita Sunao1, Kurihara Eiji1
  1) Department of Neuropediatrics, Tokyo Metropolitan Neurological Hospital, Tokyo, Japan, 2) Department of Neurology, Tokyo Metropolitan Neurological Hospital, Tokyo, Japan, 3) Department of Neurosurgery, Tokyo Metropolitan Neurological Hospital, Tokyo, Japan

O-197  A case of Segawa disease presenting with dysphonia and dropped head
  "Hanafusa Yukiko1, Kumada Satoko1, Yokochi Fusako2, Taniguchi Makoto3,
  Hoshino Ai1, Tomita Sunao1, Kurihara Eiji1, Shintaku Haruo2, Fujioke Hiroki1
  1) Department of Neuropediatrics, Tokyo Metropolitan Neurological Hospital, Tokyo, Japan, 2) Department of Neurology, Tokyo Metropolitan Neurological Hospital, Tokyo, Japan, 3) Department of Neurosurgery, Tokyo Metropolitan Neurological Hospital, Tokyo, Japan, 4) Department of Pediatrics, Osaka City University Graduate School of Medicine, Osaka, Japan

O-198  Low dose levodopa therapy for laryngeal dystonia in xeroderma pigmentosum.
  "Miyata Rie1, Hayashi Masaharu2, Araki Satoshi3, Kohyama Jun1
  1) The Department of Pediatrics, Tokyo Kita Shakai Hoken Hospital, Tokyo, Japan, 2) The Department of Clinical Neuropathology, Tokyo Metropolitan Institute for Neuroscience, Tokyo, Japan, 3) The Department of Pediatrics, Tokyo Medical and Dental University, Tokyo, Japan

O-199  Efficacy of Deep Brain Stimulation for a girl with intractable involuntary movement
  "Sato Ken1, Nakagawa Eiji1, Hanai Sae1, Sakuma Hiroshi2, Komaki Hirofumi1,
  Saito Yoshiaki1, Sugai Kenji1, Sasaki Masayuki1, Nakama Hideyuki2, Otsuki Taisuke2
  1) Department of Child Neurology, National Center Hospital for Mental, Nervous and Muscular Disorders, National Center of Neurology and Psychiatry, Kodaira, Tokyo, Japan, 2) Department of Neurosurgery, National Center Hospital for Mental, Nervous and Muscular Disorders, National Center of Neurology and Psychiatry
International Symposium Celebrating the 50th Meeting of the JSCN Part I

May 28th, Wed

Room 5
First in this talk, the workings of neuronogenesis, i.e., the process of neuron production through proliferation/differentiation of neural progenitor cells (NPCs), and its contribution to the laminar architecture of the neocortex will be reviewed. Principal emphasis will be upon the regulatory mechanisms of the total neuron production during neocortical neuronogenesis, which will ultimately determine the size of the brain: the more neurons produced, the bigger the brain is going to be and vice versa.

In mice, the founder population of NPCs and its progeny execute 11 cell cycles in the course of neuronogenesis. There is a tight correlation between cell cycle of origin and layer destination of newly-born neurons. The most critical parameter of the neuronogenic process is probability of cell cycle exit of NPCs (quiescent fraction or Q). Q is precisely regulated during the course of neuronogenesis and gradually increases from a value of 0 to 1.0, as neuronogenesis proceeds. Thus, each successive cell cycle of NPCs is associated with unique quantitative parameters, both in terms of the value of Q and the destination of the newly born neurons to specific layers of the neocortex. The ontogenetic changes in Q lead to the prediction that relatively few cells will be produced during the early phase of neuronogenesis when deep layer neurons are born and that a surge occurs in neuron production during the later phase when superficial layer neurons are born. According to this model, single founder cell gives rise to approximately 150 projection neurons in the neocortex.

A subtle change in Q occurring at specific stages of neuronogenesis greatly affects the pattern of neocortical neuron production and hence the size of the neocortex. Three different approaches were used to experimentally alter the expression levels of cell cycle regulatory gene p27Kip1 (p27) : 1) p27 knockout mice, in which the superficial layer neurons are over-produced by abnormal decrease in Q, 2) p27 overexpression, in which the superficial layer neurons are decreased by premature increase in Q in the middle phase of neuronogenesis, and 3) in utero exposure to TCDD, in which the deep layer projection neurons are decreased by accelerated nuclear transport of p27 resulting in premature increase of Q in the early phase of neuronogenesis.
Neurocutaneous diseases or syndromes, are a heterogeneous group of congenital disorders with developmental lesions of the skin or subcutaneous tissues and nervous systems, with different clinical presentations, patterns of inheritance and pathological features. Most neurocutaneous syndromes share a tendency to develop hamartomas in many sites of the body. Tuberous Sclerosis Complex is the prototype of multisystemic involvement with hamartomas not only from the brain and skin (ectoderm), but also from the heart and kidneys (mesoderm), and liver (endoderm)(1).

Recently, modern embryology, including the molecular genetic programming of the nervous system and all other organs, has revealed a new interpretation of the clinical and pathological features of these disorders. Many manifestations of "primary neurocutaneous syndromes" may be attributed in large part to abnormal neural crest migration and differentiation. As more genetic information becomes available about the many disorders affecting multiple neural crest derivatives, an integrated morphological and molecular genetic classification of neurocutaneous syndromes could be possible. How defective genes in the neurocutaneous syndromes interact with many other genes during neural crest development is largely unknown (2).

Tuberous Sclerosis Complex continues to be an fascinating area of research for all those interested in molecular genetic programming of brain development (3). Gene mutations in either of the two TSC genes influence neural precursors between weeks 7 and 20 of gestation, disrupting the mTOR pathways, thus resulting in abnormal cell differentiation and dysregulated control of cell size. The discovery of mTOR pathway upregulation in TSC associated tumours, created possibilities of new treatment strategies with mTOR inhibitors. Dysregulation of mTOR may be a common molecular basis, not only for hamartoma syndromes but also for other cellular hypertrophic disorders.

Recent developments from molecular biology are shedding light on the molecular basis of many neurological manifestations of TSC, including epilepsy, autism and cognitive disorders and are leading to a reinterpretation of the neurological phenotypes in molecular terms.

References

Professional experience:
1974: Graduation at Catholic University Medical School, Rome
1974-1978: Postgraduate Education on Child Neurology and Psychiatry, University La Sapienza, Rome
1979-1980: Assistant Professor of Pediatric Neurology, University La Sapienza, Rome
1980-1989: Associate Professor of Pediatric Neurology University La Sapienza, Rome
1990-1994: Professor of Pediatric Neurology, University D'Annunzio, Chieti
1994-present: Professor of Pediatric Neurology and Psychiatry, University of Rome Tor Vergata

Main area of interest and research:
Tuberous Sclerosis Complex
Behavioral Neurology
Epilepsy

Publications:
More than 120 publications on Child Neurology in peer reviewed journals
More than 35 chapters on books in the field of Child Neurology
Editor of 3 books on "Tuberous Sclerosis Complex", "Neurocutaneous Syndromes in Children" and Malformations of Central Nervous System"
Professional Societies Memberships:
International Child Neurology Association (Past-President)
European Pediatric Neurology Society (Executive Board Member 1997-present)
Japanese Child Neurology Society
Child Neurology Society
Society for the Study of Behavioral Phenotype
Chairman of the Section of Child Neurology -International School Neurological Sciences

Editorial Boards:
Journal of Child Neurology (Associate Editor)
European Journal of Pediatric Neurology (Member)
Pediatric Neurology (Member)
Brain & Development (Member)
Journal Pediatric Neurology (Member)
Age-related presentations of hereditary peripheral neuropathies in childhood

Robert A Ouvrier
The Institute for Neuromuscular Research, The Children's Hospital at Westmead, Sydney, Australia

The classification and diagnosis of hereditary peripheral neuropathies in childhood has become extremely complicated. An age-related approach can assist in their correct diagnosis and genetic investigation.

Congenital hereditary polyneuropathies are rare. Two main groups can be distinguished. In one, chronic axonal neuropathies of variable severity often present with arthrogryposis but are non-progressive. Most such cases are sporadic.

A second congenital group consists of amyelinating or hypomyelinating neuropathies. The amyelinating group is usually fatal, and its molecular biological basis is poorly understood. The early onset hypomyelinating neuropathies, characterised by hypotonia, severe motor delay and very slow nerve conduction velocities, are frequently due to mutations of myelin proteins. While they generally result in a chronic, moderately severe outcome, several spectacular examples of recovery are recorded.

Severe infantile axonal neuropathy, which is a variant of spinal muscular atrophy with respiratory disease (SMARD) presents in the first few months of life. It is most commonly caused by mutations of the IGHMBP2 gene and results in death or ventilator dependency within the first year of life. Some phenocopies for which the molecular biological basis is still uncertain are known as SMARD 2.

D'jerine-Sottas syndrome presents in the first two years of life. It is typically associated with proximal and distal weakness, areflexia, ataxia and sensory changes, accompanied by very reduced nerve conduction velocities. Mutations of myelin P zero, PMP22, periaxin and EGR2 are the commonest recognised causes.

HMSN of axonal type commencing in early childhood usually presents between 1 and 5 years of age, and progresses to almost complete loss of voluntary movement below the knees and elbows, by the teenage years. It is most commonly caused by mutations of the mitofusin 2 gene.

While Charcot-Marie-Tooth disease type 1 can be caused by many genes (to be discussed further), the commonest form, CMT type 1A is due to a DNA duplication on chromosome 17, causing an increase in expression of PMP22. It is very commonly symptomatic in the first decade. The axonal (type 2) forms of CMT tend to present later (with the exception of the infantile and early onset forms described above).

NAME: ROBERT ARTHUR OUVRIER
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MARITAL STATUS: Married
Number of Children: Three
QUALIFICATIONS:
1956- Leaving Certificate — Honours in Latin and French
1961- B. Sc(Med). (Hons) — Physiology. University of Sydney
1964- M.B., B.S. (Hons) University of Sydney
1967- M.R.A.C.P.
1972- F.R.A.C.P.
1986- M.D. University of Sydney

AWARDS
Medal of the Order of Australia, 2004

POSITIONS HELD:
1969: Honorary Physician and Fellow in Neurology, Royal Children’s Hospital, Melbourne
1969/71: Fellow in Neurology, University of Kentucky, U.S.A.
1971/72: Fellow in Neurology, John Hopkins University, Baltimore, U.S.A.
1999- Head, Institute for Neuromuscular Research, The Children’s Hospital at Westmead
2001- Petre Foundation Professor of Paediatric Neurology The Children’s Hospital at Westmead
2006- President, International Child Neurology Association

Professor Ouvrier has been an invited overseas guest lecturer on over forty occasions.

His published works include two books [including the only textbook on peripheral nerve diseases in childhood, now in its second edition], thirty book chapters and over one hundred articles on various aspects of child neurology.

His current main research interests include the peripheral neuropathies and screening tests of cognitive function in childhood.
Japanese encephalitis virus is a mosquito-borne member of the genus flavivirus, which causes an estimated 30,000 to 50,000 cases of encephalitis and 10,000 to 15,000 deaths in Asia every year. Pigs can become infected and act as amplifying hosts, bringing the virus closer to human habitats - especially in parts of Asia where pigs are kept near homes. Many mosquito species are potential vectors, but culex species such as Culex trituniorhynchos and C. vishnui, which breed in rice paddies and other dirty water, are especially important. Because the virus and its vectors and hosts are ubiquitous in rural Asia, most of the population is exposed during childhood, as shown by serologic studies, though disease develops in only a small proportion of infected persons. The clinical manifestations range from a nonspecific febrile illness to severe meningoencephalitis, characterized by a reduced level of consciousness, seizures, parkinsonian movement disorders, and acute flaccid paralysis. There is no established treatment for Japanese encephalitis. Typically, 20 to 30 percent of patients with Japanese encephalitis die, and approximately half the survivors have severe neuropsychiatric sequelae. Treatment efforts are directed to controlling both the immediate complications of infection, including seizures and increased intracranial pressure, and the longer-term consequences of neurologic impairment, such as limb contractures and bed sores. The best hope for controlling Japanese encephalitis lies in vaccination. The vaccine's efficacy was demonstrated in large, placebo controlled trials in Taiwan in the 1960s and in Thailand in the 1980s. In developed Asian countries such as Japan, Taiwan, and Korea, where mass vaccination with the inactivated vaccine has been practiced for years, the incidence of Japanese encephalitis has diminished considerably. The vaccine confers no herd immunity because humans are not the primary hosts. The introduction of vaccination into the Expanded Program on Immunization of the World Health Organization has also been associated with reduced disease in Thailand. In Korea, since the last epidemic outbreak in 1982, the yearly incidence of Japanese encephalitis is less than 10 cases per year with no case reported in the year of 1995 to 1997, 2000, 2004 and 2006.
Children living in Africa bear the brunt of the global burden of the infections of the central nervous system (CNS). Over 70% of the *Plasmodium falciparum* and 80% HIV infections occur in Africa, and the incidence of bacterial meningitis is highest on this continent. There is little data on which to estimate the burden of viral infections of the CNS on this region, and other infections such as tetanus that have been eliminated from developed countries still occur. The pathogenesis of malaria, HIV and bacteria infections on the CNS are different. HIV infects the parenchyma, whilst the malaria parasites are confined to the microvessels and bacteria involve the meninges and to a lesser extent cause a vasculitis. These differences produce different patterns of brain damage, although the final common pathways may be similar. These infections are associated with high mortality rates in Africa and the children that survive have high rates of neurological deficits. The proportion of children with more subtle neuro-cognitive sequelae is not well defined, since there is a lack of cultural appropriate tools to assess cognitive function and the long-term follow up is limited. Furthermore the CNS infections often occur in children with delayed development from other causes such as malnutrition or perinatal insults, and this aggravates the impairment caused by these infections. Finally the contribution of these infections to the development of epilepsy is underestimated. The burden of these preventable infections on children is increasingly being recognised, but is still underestimated.

Charles Newton qualified in Cape Town, South Africa, with postgraduate training in Paediatrics in Manchester and London. As a lecturer at University of Oxford, he went out to Kilifi in Kenya, to set up an unit to study severe malaria in African children in 1989. This unit has become one of the leading Tropical Medicine units in the world, ranked first in Africa for malaria research, and third in the world for malaria research. He wrote his doctoral thesis on cerebral malaria. Thereafter he spent 2 years as a Post-doctoral fellow at Johns Hopkins, USA; studying mechanisms of brain damage in central nervous system infections. He returned to Great Ormond Street Hospital, London to complete his training in Paediatric Neurology, where he became a consultant in 1997. In 1998 he was awarded a Wellcome Trust Senior Clinical Fellowship to return to Kilifi, to study CNS infections in children. He became Head of Clinical Research in Kilifi at this time. The Senior Fellowship was renewed in 2003 and 2008 to study Seizures and Epilepsy caused by CNS infections in Kenya. He was promoted to Reader in 2003 and full Professor in 2005.
International Symposium Celebrating the 50th Meeting of the JSCN part I: Recent advances in child neurology

Progress in the biological aspects of status epilepticus

Solomon L. Moshé

Saul R. Korey Department of Neurology, Dominick P. Purpura Department Neuroscience and Department of Pediatrics, Laboratory of Developmental Epilepsy, Montefiore / Einstein Epilepsy Management Center, Albert Einstein College of Medicine and Montefiore Medical Center

Epidemiological studies suggest that early in life, the brain is unusually susceptible to seizures and especially status epilepticus (SE). The increased propensity of the developing brain to experience SE may be related to immaturity of the networks (such as the substantia nigra based network) that can suppress recurrent seizures in adults. To understand the impact of SE on the developing brain, two-way translational studies from the bedside to the animal model are needed. Epidemiologic and outcome-related clinical studies are important to generate hypotheses that can be tested in model systems and then brought back to practice.

There is an ongoing debate on the effects of SE on the brain as a function of age. Most of the studies focus on effects on hippocampal function, based on the notion that SE early in life may lead to the development of temporal lobe epilepsy. However, SE may have more widespread effects that are specific for discrete developmental windows and often sex-related. Understanding the spectrum and progressive nature of SE-induced changes in brain function may have important implications in the design of treatments aimed at disease modification. These treatments can potentially be used over relative short periods thus avoiding the potentially detrimental effects of long-term drug administration.

CV:
Solomon L. Moshé, MD, is professor of Neurology, Neuroscience, and Pediatrics at the Albert Einstein College of Medicine (AECOM). He is also vice chairman of the Department of Neurology as well as director of Child Neurology and Clinical Neurophysiology and the recipient of a Martin A. and Emily L. Fisher fellowship in Neurology and Pediatrics. Dr. Moshe received his medical degree from the National University of Athens School of Medicine, Athens, Greece, in 1972. He trained in pediatrics at the University of Maryland and in neurology and child neurology at Albert Einstein. Dr. Moshe is certified by the American Board of Pediatrics; the American Board of Psychiatry and Neurology, with special qualifications in child neurology and clinical neurophysiology; and the American Board of Clinical Neurophysiology. Since 1979, his research has focused on understanding the mechanisms underlying age- and sex-related differences in epilepsy in humans and animal models.
Duchenne muscular dystrophy (DMD) is a lethal X-linked disorder of striated muscle caused by mutations in the DMD gene. Gene therapy is one of attractive approaches to the treatment of DMD and we paid special attention on a recombinant adeno-associated virus (AAV)-mediated gene transfer and on an exon skipping by antisense Morpholino treatment. It is important to examine therapeutic effects and the safety issue of the approach in larger animal models, such as dystrophic dogs. We recently established a Beagle-based dystrophic dog colony in Japan, CXMD6 (Exp Anim, 52: 93-97, 2003) and the dogs show similar symptoms seen in DMD (Acta Myologica, XXIV: 145-154, 2005; BMC Cardiovasc Disord. 2006;6:47). First, we injected a recombinant AAV2 encoding the LacZ gene into skeletal muscles of normal Beagles. β-galactosidase (β-gal) was expressed only in a few fibers, but instead marked cellular infiltration appeared. Immunosuppressive treatment improved β-gal expression, though the effect was not complete (Gene Ther. 14:1249-1260, 2007). We, then, generated a type 8 recombinant AAV (AAV8) encoding the LacZ gene. Recombinant AAV8 encoding the LacZ gene driven by a CMV promoter was injected into tibialis anterior and extensor carpi ulnaris of normal Beagles. We found more β-gal positive fibers in AAV8-injected canine skeletal muscle than those in AAV2-injected muscle. Moreover, cellular infiltration in AAV8-injected muscle was much less than the AAV2-injected muscles. We, therefore, injected recombinant AAV8 encoding canine micro-dystrophin (c ΔCS1) gene into skeletal muscle of CXMD6 and confirmed the expression of micro-dystrophin. The injection of recombinant AAV8 encoding ΔCS1 through limb perfusion would be a favorable approach to DMD.

Recently, we also tried injection of antisense Morpholino into dystrophic dogs to induce exon skipping of the mutated canine DMD gene in collaboration with Dr. Eric Hoffman's research group (Yokota T, Partridge TA, and Lu QL). Results of ongoing experiments in CXMD6 and a perspective on clinical trials of DMD patients would be presented.

Shin'ichi Takeda received a M.D. from the University of Akita School of Medicine in 1977. He received a Ph.D. in muscle biology from Shinshu University Graduate School in 1981. He is currently Director of Molecular Therapy at National Institute of Neuroscience (2000-). He was previously Research Associate of Neurology at Shinshu University School of Medicine (1985-87) and post-doctoral fellow of Biochemistry at Pasteur Institute (1987-92) and Section Chief of Neuromuscular Disorders (1992-95) and of Molecular Genetics (1995-2000) at National Institute of Neuroscience. He has published more than 100 papers in molecular pathology and molecular therapy of muscular dystrophy, and molecular biology of muscle regeneration and stem cells.
International Symposium Celebrating the 50th Meeting of the JSCN part I: Recent advances in child neurology

Robot suits — cybornoid, biorobotics, control for supporting disabled persons —

Yoshiyuki Sankai
Professor, Head of Cybernetics Lab. (Sanlab), Department of System & Information Engineering, Univ. of Tsukuba
(Visiting Professor, Baylor college of Medicine)

Major Research Projects;
1) Artificial Heart Control Project; New Generation Artificial Heart System
2) Project HAL; Exo-Skeleton type Robot suit named as "Cybersuit" HAL (Hybrid Assistive Limb)
3) Humanoid Control Project

Awards
The 2005 World Technology Award (IT Hardware), etc
International Symposium Celebrating the 50th Meeting of the JSCN Part II

May 29th, Thu.

Room 1〜3
May 29th, Thu.

International Symposium Celebrating the 50th Meeting of the JSCN part II: Celebration & Ceremony for the 50th Meeting of the JSCN

History of Child Neurology in Japan

Yukio Fukuyama
Emeritus Professor, Tokyo Women’s Medical University, Tokyo, Japan

It is rather easy to trace back the history of the Japanese Society of Child Neurology (JSCN) to its birth in 1961, when JSCN was first established under the leadership of the late Prof Tadao Tatsuk. Various documents and archives are rather well stored. Particularly, a monograph "Footnotes of JSCN" of 275 pages was published in 1992, in which relevant informations and materials on various society’s activities are presented. It is worth to note that JSCN is the second oldest child neurology society in the world, only after the National Spastics Society (Oxford) which was first formed in 1959. JSCN grew up rapidly. At the first meeting in 1961, there were 3 special lectures, 46 free papers and about 150 attendees. At the 49th meeting in Osaka, 2007, the scientific program was composed of 75 specially programmed lectures and 459 free papers. As of November 30, 2007, the Society has 3,501 members, 16 working committees and 9 regional sub-societies. Thus, JSCN has been and still is literally the largest CNS in the world.

The situation of child neurology before 1950 remains unclear, however. Extensive review of the official journal of the Japan Pediatric Society since its first issue in 1895 revealed the fact that original / research articles on neurological problems in children were quite scanty. In the Meiji era (1895-1911) there were 16 papers, while there were 21 and 47 papers in the Taisho (1912-1926) and Showa era (1927-1950), respectively, if CNS infections (encephalitis, meningitis, polio, ekiri) or nutritional beriberi were excluded from the count. The subjects studied were varied.

There are two prominent original papers in earlier age, both of which may represent the world oldest records of certain rare neurological diseases in childhood. They are a case report of paroxysmal kinesigenic choreoathetosis by Kure in 1892 and a case report of congenital analgesia with anhidrosis contributed by Nishida et al in 1951.

It would be an interesting task to explore the processes of transition path from an earlier quiet prewar age to the birth of JSCN and the current proliferation of child neurology in Japan.
In this personal view, I describe some of the many important contributions of Japanese physicians and scientists to the development of modern paediatric neurology. Infantile beri-beri was known as *kakke* in Japan from ancient times. It was due to the consumption of polished rice which was lacking in thiamine. Japanese scientists identified the chemical nature of thiamine and saw to the prevention of this disease by diet alterations.

Japanese scientists wrote the first description of Japanese encephalitis in 1871, discovered the virus in 1935 and went on to develop effective vaccines which have (virtually) eradicated the disease from Japan itself and which are utilised throughout South-East Asia.

The description of *moyamoya* in 1955 introduced an important vascular disease causing a wide panorama of presentations. Novel efficacious treatments for revascularisation of the brain resulted and are now implemented worldwide.

Dr Masaya Segawa was the discoverer in 1970 of the condition titled *"Hereditary progressive dystonia with marked diurnal fluctuation"*. Its elucidation has been a beacon for those seeking to find permanent cures for neurodegenerative disorders of childhood. My chance meeting with Dr Segawa resulted in the publication of the first Western cases of Segawa disease.

There are numerous Japanese contributions to the elucidation of metabolic disorders. The names of Ketya Tada and Yoshiyuki Suzuki spring to the forefront. Their work on the amino-acidurias and the gangliosidoses are landmarks.

In the field of muscle disorders, Japanese workers have also made contributions of the highest order. Dr Yukio Fukuyama's description of the condition which universally bears his name is a striking example. Miyoshi's description of the myopathy due to *dysferlin* mutations, Dr. Nonaka's myopathy and Dr Makiko Osawa's many papers on myasthenia and muscular dystrophy are other examples of Japanese leadership in the study of muscle disease.

In epilepsy, the description of Ohtahara's syndrome and numerous other epileptic syndromes, the EEG studies of Dr. Watanabe, the bibliography of the epilepsies, together with international conferences on the causes of seizure disorders are examples of outstanding activity in this field.

Apart from clinical activity, Japan has been very active in international affairs in the field of child neurology. There have been two Japanese Presidents of the International Child Neurology Association—Drs. Yukio Fukuyama and Yoshiyuki Suzuki. Drs. Makiko Osawa and Masaya Segawa are current members of the Executive Board. Yukio Fukuyama was the founding father of the Asian-Oceanian Child Neurology Association, founded in 1983.

In the publishing arena, the journal *Brain and Development* has found an important place in the international paediatric neurology literature.

Japanese child neurologists have been very important contributors to key areas in the development of child neurology. At this celebration of the 50th birthday of the Japanese Child Neurology Society and the 80th birthday of Yukio Fukuyama, it is so appropriate that these two vital organisms, so inextricably linked over the past 50 years, should take enormous pride in the fruitfulness of their symbiotic association.
International Symposium Celebrating the 50th Meeting of the JSCN part II: Celebration & Ceremony for the 50th Meeting of the JSCN

The Japan’s role in child neurology in the AOCNA

Yong-Hee Hwang
Seoul National University Children's Hospital, Seoul, Korea

Since the year of 1983, when the first congress of Asian and Oceanian Child Neurology Association (AOCNA) was held in Taipei under the presidency of the late Professor Yu-Zen Shen, the congress has been held once in every 2 or 4 years. The latest 9th congress was held in Cebu, Philippine, 2007 under presidency of Professor Aida Salonga. As Professor Fukuyama, the emeritus president of AOCNA, wrote in the recent AOCNA Newsletter, the past nine congresses were all extremely successful and owing to enormous efforts and devotion of respective presidents, every congress had been well attended by colleagues region-wide. Excellent scientific program presented advanced knowledge and information on child neurology to those attended.

Regarding the Japanese Society of Child Neurology, it is one of the oldest and largest societies of child neurology specialty in the world, having 3,300 members currently. Among the 930 members from 24 countries of AOCNA, Japanese members are 240 counting more than 25% of total members. Also there are so many great doctors, scholars or professors of child neurology in Japan and many of them devoted themselves for the development of AOCNA including Professor Fukuyama, the emeritus president of AOCNA, and longtime secretary-treasurer Professor Sakakihara.

AOCNA, however, need to be changed to upgrade the organization, network construction and to solve the discrepancy of status of child neurology between the membership countries. To achieve these goals, Japanese members of AOCNA will be highly appreciated for their support and contribution as the largest membership country.

CURRICULUM VITAE

NAME IN FULL : Yong-Seung Hwang
DATE OF BIRTH : October 27, 1950

Seoul National University College of Medicine
M.D. 1971–1975
Seoul National University Majoring in Medicine, Pediatrics
Ph. D. 1978–1983

POSTDOCTORAL TRAINING
1975–1976 Intern at Seoul National University Hospital, Seoul, Korea
1976–1980 Resident in Pediatrics at Seoul National University Hospital, Seoul, Korea
1985–1987 Special Fellow in Pediatric Neurology at University of Minnesota Hospital, USA

POSITION HELD
1983–1985 Instructor in Pediatrics
1985–1990 Assistant Professor in Pediatrics
1990–1995 Associate Professor in Pediatrics
1995– Professor (subspecialty: Pediatric Neurology) at Seoul National University College of Medicine & Seoul National University Children's Hospital, Seoul, Korea
1999–2002 Vice President of Korean Epilepsy Society
2000–2006 Vice President of Korean Child Neurology Society
2002–2005 President of Korean Epilepsy Society
2005– Honorary President of Korean Epilepsy Society
2006– President of Korean Child Neurology Society
2006– President of Asia Oceania Child Neurology Association and 10th Asia Oceania Congress of child Neurology
50th Meeting of the JSCN
Invited Lecture

May 29th, Thu.

Room 1〜3
Muscular dystrophies are clinically and genetically heterogeneous. Limb girdle (LGMD) and Duchenne (DMD) muscular dystrophies are largely due to defects of proteins associated with the sarcolemma; defective extracellular matrix proteins is at the basis of common forms of congenital muscular dystrophies (CMD) such as merosin deficient CMD (laminin 2 deficiency) and the Ullrich variant (collagen VI deficiency). More recently a novel pathogenic mechanism has been identified, the abnormal glycosylation of alpha dystroglycan. This is a peripheral membrane protein which undergoes complex glycosylation steps before being able to interact with extracellular matrix proteins such as laminins, agrin, perlecan and neurexin. Mutations in six genes (POMT1, POMT2, POMGNT1, fukutin, FKRP and LARGE) have so far being identified in patients with reduced glycosylation of alpha dystroglycan. While initially an apparent good correlation between mutated gene and resulting phenotypes was suggested (with Walker Warburg syndrome (WWS) associated with mutations in POMT1 and POMT2, and Fukuyama muscular dystrophy associated with fukutin mutations, and Muscle Eye Brain disease associated with POMGNT1 mutations) it has recently been demonstrated that allelic mutations in each of these 6 genes can result in a wide spectrum of clinical syndromes ranging from severe and fatal conditions with associated structural brain involvement (WWS) to childhood or adult onset LGMDs. The severity of the phenotype appears therefore not related to which gene is primarily mutated, but how severely the mutation affects the glycosylation of alpha-dystroglycan. Despite the improved knowledge, a significant proportion of CMD children do not have mutations in any of the known genes, suggesting further heterogeneity.

Present title: Professor of Paediatric Neurology, Institute of Child Health, University College of London. FRCPCH, FMedSci
Director, the Dubowitz Neuromuscular Centre, Institute of Child Health, University College, London UK
1. Previous Appointments:
1993–2007
Lecturer (1993) , Senior Lecturer (1994) , Reader (1996) and Honorary Consultant in Paediatric Neurology; Royal Postgraduate Medical School, Hammersmith Hospital London, UK
Clinical and Research Director (1996) , Hammersmith Hospital Neuromuscular Unit, Department of Paediatrics & Neonatal Medicine, London UK
Professor in Paediatric Neurology (1998), Imperial College London (formerly Royal Postgraduate Medical School)
Head of the Nationally Commissioned (NSCAG) Centre for Congenital Muscular Dystrophies (2001), Hammersmith Hospital, Du Cane Road, London
August 2005–November 2007: as above but as part of the Division of Medicine
Autism and epilepsy are heterogeneous developmental disorders associated with many diverse etiologies and pathologies. The severity of impairments and the variety of symptoms associated with autism spectrum disorders or with epilepsy syndrome reflects focal or global, structural or functional dysfunction of neuronal networks. Social communication impairments are the core clinical features and dimension that best differentiates autism from other developmental disorders of brain function. Epilepsy co-exists with autism in approximately one-third of children with either an autism spectrum disorder or with an epilepsy syndrome. Epilepsy, seizures, EEG abnormalities, and regression can identify a subset of individuals within the larger spectrum of autism that can further genetic studies in autism. Clinical models in which social impairments and epilepsy co-exist such as tuberous sclerosis complex, Rett syndrome and epileptic encephalopathies illustrate the complex relationship between autism and epilepsy. The role of minicolumns, specific genetic and epigenetic variations, synaptic receptors, neurotransmitters or modulators, and the organization of widespread functional networks have all been hypothesized as contributing to the co-occurrence of social communicative deficits and epilepsy and to the pathophysiology of autism spectrum disorders. There are developmental time windows crucial to cognitive and behavioral outcomes and early recognition of social communicative deficits is essential for maximizing the potential of children with common neurological disorders such as autism and epilepsy.

Comprehensive management of children with autism or epilepsy requires a multidisciplinary i.e., pharmacological, behavioral and educational approach that addresses the specific cognitive, communicative, and behavioral problems of this group of children.

Dr. Tuchman is the director of Autism and Related Disorder Programs at Miami Children’s Hospital Dan Marino Center and the director of Developmental and Behavioral Neurology at Miami Children’s Hospital. He was the Founding Director of the Miami Children’s Hospital Dan Marino Center for children with developmental disorders serving as its executive medical director from its start in 1998 thru 2001. Dr. Tuchman is an Associate Professor of Neurology at University of Miami Miller School of Medicine, Miami Children’s Hospital. He earned his M.D. from New York University School of Medicine and is certified by the American Board of Pediatrics and the American Board of Psychiatry and Neurology with Special Qualification in Child Neurology. He is a fellow of both the American Academy of Pediatrics and the American Academy of Neurology. Dr. Tuchman has published and lectured nationally and internationally on the topic of neurodevelopmental disorders including autism, ADHD, epilepsy and learning disorders. He is the co-editor of Autism: A neurological disorder of early brain development, which was published by Mac Keith Press in association with the International Child Neurology Association (ICNA) , in 2006.
50th Meeting of the JSCN
Luncheon Seminar

May 29th, Thu.

Room 4
LS 2
Diagnosis and Management of Seizures in the ICU
Akihisa Okumura

A SOLUTION TO THE NICU CHALLENGE
Neurodevelopmental disabilities and clinically silent seizures are estimated to affect more than 16% of the NICU patient population.
CLINICAL APPLICATIONS
The NicoletOne Monitor allows for better evaluation of cerebral function, faster time to treatment and better prognostic information which results in quality care for the NICU patient. Can we afford to leave out any vital sign?

Seizures
Seizure incidence is higher in neonates than in any other age group – a majority occurring in the first week. These seizures often have no or very subtle clinical manifestations. EEG is the only way to confirm seizure activity.

Treatment of Seizures
The deterioration effect of on going seizure activity can be slowed or prevented with clinical intervention. The effects of intervention require close monitoring through EEG.

Burst Suppression
This crucial EEG pattern provides significant information on brain development in the premature infant. With Burst count and Interburst interval measurements, the neonatologist is able to extract useful information on brain development.

Brain function recovery and development of the brain
EEG monitoring is ideal for providing prognostics values such as background activity, burst suppression and sleep/wake cycles. These can easily be recognized by NICU staff, in the aEEG trend.

Hypoxic Ischemic Encephalopathy (HIE) and Intra Ventricular Hemorrhage (IVH)
These serious conditions can be diagnosed and monitored at earlier stages to allow for a “therapeutic window” by monitoring brain function with EEG.

CHALLENGING CLINICAL CONDITIONS
The NicoletOne Monitor enables NICU staff to address specific and important neurological hazards through brain function monitoring of:
. Preterm babies and/or Low Apgar score and treatment
. Asphyxia
. Intubated infants
. Rhythmic movements, pedaling, chewing or ocular movements that might indicate

subtle seizures
. Epileptic diagnosis
. Sepsis
. Hypothermia

Nicolet One Monitor offers EFFICIENT, BETTER CARE: COMPREHENSIVE CAPABILITIES
. Easy-to-use protocols reduce set-up time for different patient groups. Users can modify pre-configured protocols or create custom settings.
. Innovative touch-screen interface simplifies training and daily operation.
. Wall-mounted or cart-based designed for the increasing demands of a busy NICU.
. Continuous impedance checks monitor signal quality and indicate which electrodes need attention.
. Alerts are attached to major events, and automatic notifications can be made by sound, pager, email or SMS.
. Network connectivity allows for easy export of data for further analysis and research.
. All necessary raw EEG data is stored for complete neurological diagnosis
. With more channels, the NicoletOne Monitor tests more regions of the brain, identifies focal activity and performs a full range of EEG functions without the use of additional equipment.
. Monitors other functions such as EKG, respiration and temperature, allowing observation of sleep patterns.
. Remote analysis for experts to review patient data without actually visiting the NICU
50th Meeting of the JSCN
Morning Educational Seminar

May 30th, Fri.

Room 2
Despite the emergence of innovative imaging techniques, the EEG still is the premier tool to determine brain physiology in patients. In this presentation, I will discuss the basics of EEG: how it is recorded, rules of polarity and how to localize discharges, the differences between bipolar and referential montages, the identification of artifacts, the basic EEG rhythms, and how EEG patterns can be modified by age and sleep state. I will explain how EEG patterns can be used to identify background abnormalities that may be specific for cortical and or subcortical grey matter dysfunction, white matter injury or diffuse insults. Time permitting I will show various epileptiform discharges and periodic and periodic or pseudoperiodic discharges. Finally I will propose EEG classification schemes that can be easily understood by the referring physicians and may accurately guide other diagnostic interventions, treatment decisions and prognosis.

Solomon L. Moshe, MD, is professor of Neurology, Neuroscience, and Pediatrics at the Albert Einstein College of Medicine (AECOM). He is also vice chairman of the Department of Neurology as well as director of Child Neurology and Clinical Neurophysiology and the recipient of a Martin A. and Emily L. Fisher fellowship in Neurology and Pediatrics. Dr. Moshe received his medical degree from the National University of Athens School of Medicine, Athens, Greece, in 1972. He trained in pediatrics at the University of Maryland and in neurology and child neurology at Albert Einstein. Dr. Moshe is certified by the American Board of Pediatrics; the American Board of Psychiatry and Neurology, with special qualifications in child neurology and clinical neurophysiology; and the American Board of Clinical Neurophysiology. Since 1979, his research has focused on understanding the mechanisms underlying age-related and sex-related differences in epilepsy in humans and animal models.
International Symposium Celebrating the 50th Meeting of the JSCN Part III

May 30th, Fri.

Room 2
Fukuyama congenital muscular dystrophy (FCMD) is a unique autosomal recessively inherited condition characterized by a combination of systemic progressive muscular dystrophy (PMD), severe cerebro-cerebellar cortical malformation in association with or without retinal derangement of very early onset. FCMD is the second most prevalent type of PMD in Japan, while it is extremely rare outside Japan. Historically, the first half of the 20th century was a dark age in terms of CMD. The category of CMD was practically absent in most international classifications of PMD until 1986, when MIM (McKusick) first enlisted FCMD as a disease entity. In the midst of this dark age, that is, in 1960, we first reported a series of 15 cases of CMD and advocated that this will represent a completely new disease entity. Key clues to the correct diagnosis were derived from keen clinical observation, application of then newly developed diagnostic procedures, including muscle biopsy, needle EMG and determination of serum CK activities. Another important factor was an exhaustive review of pertinent literatures. Through the above approaches combined, we could reach to our conviction that we are dealing with an entirely new type of CMD.

Clinical features of typical FCMD patients mimic those of the floppy infant syndrome of infancy and later, after age 2, resemble those of congenital multiple arthrogryposis. Clinical onset may be at any time during the first 9 months after birth, but it is most often between 3–6 months, sparing the neonatal period, with some exceptions. Development of gross, but not fine, motor movements delays, but its progressive deterioration is hardly recognizable until 6–7 years of age. As a general rule, the maximum motor function attainable is “sit and slide on the floor”, never able to stand or walk with or without support for whole life. Language expression remains practically absent, but there will be no problem in social interaction nor behavioral problems. Global IQ ranges between 40–60 in most cases. Life expectancy is shorter than that of Duchenne patients, but they can survive up to age 20 or more, not rarely. Outside the above “core” group, there are two extremes; the one being the severe type of neonatal onset (never able to keep sitting), and the other the mild type with walking ability, though of limited degree.

Recent molecular genetic studies revealed that there is a rather good genotype–phenotype correlation among FCMD patients; the severe type appeared to be compound heterozygotes for the founder mutation and another mutation. In recent years, CMD research became so active and productive worldwide, that more than 15 clinico–genetic subtypes of CMD has been delineated. In addition, the latest studies revealed that fukutin gene mutation have a far broader clinical spectrum than thought before, ranging from the WWS phenotype to the mildest LGMD2I. Considering these circumstances, it is imperatively important to re–define what is the distinguishing clinical phenotype of FCMD.
Fukuyama-type congenital muscular dystrophy (FCMD), Walker-Warburg syndrome (WWS), and muscle-eye-brain disease are clinically similar autosomal recessive disorders characterized by congenital muscular dystrophy (CMD), cobblestone lissencephaly, and eye anomalies. WWS is the most severe syndrome of the group. During the study about clinical variation within sibs in FCMD, we found in one family the elder brother presented typical clinical findings of FCMD, while the younger brother showed hydrocephalus, cephalocele and retinal detachment at birth which were more consistent with WWS than FCMD. Both sibs were compound heterozygotes of FCMD. This shows the clinical spectrum of FCMD is much broader than previously presumed. Combined heterozygotes between a founder mutation and nonsense or missense mutations generally have a more severe phenotype than individuals homozygous for the founder mutation of FCMD. A Turkish CMD patient with hydrocephalus, buphthalmus, and cataracts was referred to us. His parents were first cousins, and their first son is unaffected. After birth this patient was supported by mechanical ventilation and died on the 10th day. Neuropathological examination showed agyric hemispheres with polymicrogyria in several cortical segments and severe cortical disorganization in other segments. CMD was also seen, with variation in fiber size, fibrosis, and fat replacement. Immunohistochemical analysis showed greatly reduced staining for α-dystroglycan but normal immunoreactivity for β-dystroglycan in the skeletal muscle membrane. Sequence analysis of the patient’s DNA identified a homozygous 1 bp insertion mutation in exon 5 of the fukutin gene. This mutation causes a frameshift, resulting in a premature termination at codon 157. Both parents and the brother were heterozygous for this mutation. This is the first case worldwide in which a fukutin mutation has been found outside the Japanese population. Later, a homozygous nonsense mutation in exon 4 of the fukutin gene was identified in another Turkish patient with WWS phenotype. The Japanese FCMD patients carrying at least one copy of a founder mutation in the noncoding region may produce a lower level of mature fukutin than normal and generate a relatively mild FCMD phenotype, while the homozygous nonsense mutations within the coding region identified in two Turkish patients are predicted to cause a total loss of fukutin activity and are likely produce a more severe phenotype which closely resembles WWS.

CV:
1965 M.D. from Kyoto University, School of Medicine, Kyoto, Japan
1977 Doctor of Medical Science from Department of Pediatrics, Kyoto University, School of Medicine
1966–67 Resident, Department of Pediatrics, Kyoto University, School of Medicine
1967–72 Staff, Department of Pediatrics, Kyoto National Hospital, Kyoto
1972–78 Chief, Department of Pediatrics, Utano National Hospital, Kyoto
1978–81 Staff, Department of Pediatrics, Kyoto University, School of Medicine, Kyoto
1980–81 Visiting Associate, Department of Molecular, Cellular and Developmental Biology, University of Colorado, Boulder, USA
1981–82 Assistant Professor, Department of Pediatrics, Kyoto University, School of Medicine, Kyoto
1982–99 Chief, Department of Pediatrics, Kobe General Hospital, Kobe
1999– Director, Kobe City Pediatric and General Rehabilitation Center for the Challenged, Kobe
International Symposium Celebrating the 50th Meeting of the JSCN Part III: Phenotypic Spectrum of Fukutinopathy

Mild phenotypes in Fukutinopathy

Francesco Muntoni
Dubowitz Neuromuscular Centre, UCL Institute of Child Health, London, UK

Defects in glycosylation of alpha-dystroglycan are associated with several forms of muscular dystrophy, often characterised by congenital onset and severe structural brain involvement, collectively known as dystroglycanopathies. Fukuyama congenital muscular dystrophy (FCMD), due to mutations in the putative glycosyltransferase fukutin, is the second most common form of muscular dystrophy in Japan and is invariably associated with mental retardation and structural brain defects.

Objectives: We intended to determine if mutations in fukutin could be responsible for dystroglycanopathies outside Japan. Methods: The fukutin gene was studied in a cohort of patients with abnormal dystroglycan expression in muscle. Patients were included in the study irrespective of their severity or associated brain involvement.

Results: We identified pathogenic fukutin mutations in three patients belonging to 2 Caucasian families. Affected children shared a limb girdle muscular dystrophy (LGMD) phenotype, were all ambulant and had marked elevation of serum CK, normal intelligence and brain structure on magnetic resonance imaging (MRI). In 2 families a marked and sustained response to corticosteroid administration was observed.

Interpretation. Our data suggest that fukutin mutations occur outside Japan and can be associated with much milder phenotypes than FCMD. These data significantly expand the spectrum of the phenotype associated with fukutin mutations, to include a novel form of LGMD that we propose to name LGMD2L.

Present title: Professor of Paediatric Neurology, Institute of Child Health, University College of London. FRCPCH, FMedSci
Director, the Dubowitz Neuromuscular Centre, Institute of Child Health, University College, London UK
1. Previous Appointments:
1993–2007 Lecturer (1993), Senior Lecturer (1994), Reader (1996) and Honorary Consultant in Paediatric Neurology; Royal Postgraduate Medical School, Hammersmith Hospital London, UK
Clinical and Research Director (1996), Hammersmith Hospital Neuromuscular Unit, Department of Paediatrics & Neonatal Medicine, London UK
Professor in Paediatric Neurology (1998), Imperial College London (formerly Royal Postgraduate Medical School)
Head of the Nationally Commissioned (NSCAG) Centre for Congenital Muscular Dystrophies (2001), Hammersmith Hospital, Du Cane Road, London
August 2005–November 2007: as above but as part of the Division of Medicine
International Symposium Celebrating the 50th Meeting of the JSCN Part III: Phenotypic Spectrum of Fukutinopathy

Fukutin gene mutations cause dilated cardiomyopathy with minimal muscle weakness

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Department of Pediatrics, Tokyo Women’s Medical University, Tokyo, Japan1
Department of Neuromuscular Research, National Institute of Neuroscience, National Center of Neurology and Psychiatry, Kodaira, Japan

A group of disorders due to altered glycosylation of α-dystroglycan (α-DG), namely, α-dystroglycanopathy (α-DG), is clinically characterized by a combination of muscular dystrophy, structural brain anomaly, and ocular involvement. Fukuyama type congenital muscular dystrophy (FCMD) is the most common form of α-DG in Japan, and the patients carry the founder mutation of 3kb retrotransposonal insertion in the fukutin gene (FKTN), homozygously or heterozygously. Clinically, FCMD is characterized by severe congenital muscular dystrophy associated with mental retardation due to brain malformation. Most patients canspeak only less than 20 meaningful words with no sentence formation. The peak motor function is seen from the age of 2 to 8 years, and their maximal motor ability is usually unassisted sitting or sliding on the buttocks. A number of the patients never acquire head control. Few patients can obtain independent ambulation, but would soon lose this ability. The prognosis is poor with their mean life span of less than 20 years. The FCMD patients with a compound heterozygous mutation of 3kb insertion and missense mutation often show more severe clinical features compared with the patients with a homozygous 3kb insertion mutation. Reportedly, there has been only two non-Japanese patients harboring null mutation in FKTN gene in both alleles. Both were Turkish boys whose clinical features were quite severe resembling Walker-Warburg syndrome, which include generalized hypotonia, hydrocephaly, bilateral ocular abnormalities, and cataracts. They died during early infancy. Here we report on six Japanese patients in four un related families with a compound heterozygous FKTN mutation presenting with dilated cardiomyopathy. All six patients show no muscle weakness until adulthood without any mental retardation. Our findings expand the phenotypic spectrum of FKTN mutations from severe congenital muscular dystrophy to dilated cardiomyopathy with mildest limb girdle muscular dystrophy.

CV:
Terumi Murakami
Present Academic & Hospital Appointments:
Assistant Professor,
Department of Pediatrics, Tokyo Women’s Medical University, Tokyo, Japan
Education
Tokyo Women’s Medical University, Tokyo, Japan, M.D. 1995 – 2001
Tokyo Women’s Medical University Graduate School, Tokyo, Japan, Ph.D. 2003 – 2007
Postgraduate Training
Resident in Pediatrics
Department of Pediatrics, Tokyo Women’s Medical University Hospital, Tokyo, Japan, 2001 – 2003
Research fellow
Department of Neuromuscular Research, National Institute of Neuroscience National Center of Neurology and Psychiatry, Tokyo, Japan, 2004 – 2006

S 97
Abnormal glycosylation of α-dystroglycan by mutations in determined or putative glycosyltransferases causes congenital muscular dystrophies called dystroglycanopathy such as Fukuyama congenital muscular dystrophy (FCMD). Because aberrant glycosylation reduces the laminin-binding activity of α-dystroglycan, functional restoration of α-dystroglycan is thought to be a potential target for therapeutic strategies of α-dystroglycanopathy. There are no naturally occurring mice carrying mutations in the fukutin gene. Fukutin knockout mice led to lethality at embryonic day 7, suggesting that fukutin is essential for early embryonic development. Chimeric mice generated using embryonic stem cells targeted for both fukutin alleles developed severe muscular dystrophy, with the selective deficiency of α-dystroglycan and laminar disorganization of the cortical structures. Next, to understand molecular pathogenesis of dystroglycanopathy and develop a therapeutic strategy, we developed model mice for FCMD which carry the retrotransposon insertion in the 3’ UTR of the fukutin gene. Histological examination revealed that the FCMD mice do not show typical signs of muscular dystrophy. Although glycosylation of α-dystroglycan in the FCMD mice skeletal muscle was reduced, reactivity against a monoclonal antibody IIH6 was still detectable. Solid-phase laminin binding assays demonstrated that more than 50% laminin binding activity was present in the FCMD mice skeletal muscle. After adenoviral LARGE gene transfers, we observed increases of glycosylation and laminin binding activity in FCMD mice skeletal muscle. Taken together, our data demonstrate that only a partial restoration of glycosylation is able to suppress the progression of muscular dystrophy and provide in vivo evidence that the LARGE gene transfer is effective strategy to restore hypoglycosylation of α-dystroglycan.
50th Meeting of the JSCN
Luncheon Seminar

May 30\textsuperscript{th}, Fri.

Room 2
Treatement for intractable epilepsy

Paolo Curatolo
Professor of Pediatric Neurology and Psychiatry, Department of Neuroscience, Tor Vergata University, Rome, Italy

Most patients affected by epilepsy can be controlled with pharmacotherapy, but at least 20% of patients with epilepsy suffer from refractory seizures which cannot be controlled even by proper treatment. The definition of refractory epilepsy or its remission differs from study to study, and it is difficult to compare different studies.

Several AEDs are concomitantly used in treatment for refractory epilepsy, but it has been reported that patients who remained drug resistant after the first AED are less likely to achieve remission due to the subsequent treatment.

In the past 15 years, new antiepileptic drugs successively appeared in European countries and in the United States. The new drugs are generally characterized by the fact that they have the efficacy similar to that of conventional AEDs and have superior safety and tolerability with few interactions with other drugs. In Europe and the U.S., prescriptions of new antiepileptic drugs have progressively increased and they have become indispensable for epilepsy treatment. In add-on therapy, a combination of an existing drug and a new drug is used more commonly than a combination of existing drugs. In selection of additional drugs, it is important to consider the profiles of side effects, or the potential of interactions and efficacy. Although some new drugs have new mechanisms of actions, there is currently no evidence that combination of drugs with different mechanisms has higher efficacy and safety.

The increased number of treatment options led to the implementation of treatments, aiming at the improvement of patients’ Quality of Life (QOL) as well as control of seizures. QOL of children affected by intractable epilepsy is influenced not only by the status of seizure control but also mental functions and drug side effects.

Professional experience:
1974: Graduation at Catholic University Medical School, Rome
1974-1978: Postgraduate Education on Child Neurology and Psychiatry, University La Sapienza, Rome
1979-1980: Assistant Professor of Pediatric Neurology, University La Sapienza, Rome
1980-1989: Associate Professor of Pediatric Neurology
University La Sapienza, Rome
1990-1994: Professor of Pediatric Neurology, University D’Annunzio, Chieti
1994-present: Professor of Pediatric Neurology and Psychiatry, University of Rome Tor Vergata

Main area of interest and research:
Tuberous Sclerosis Complex
Behavioral Neurology
Epilepsy

Publications:
More than 120 publications on Child Neurology in peer reviewed journals
More than 35 chapters on books in the field of Child Neurology
Editor of 3 books on “Tuberous Sclerosis Complex” , “Neurocutaneous Syndromes in Children” and Malformations of Central Nervous System”

Professional Societies Memberships:
International Child Neurology Association (Past-President)
European Pediatric Neurology Society (Executive Board Member 1997-present)
Japanese Child Neurology Society
Child Neurology Society
Society for the Study of Behavioral Phenotype
Chairman of the Section of Child Neurology, International School Neurological Sciences

Editorial Boards:
Journal of Child Neurology (Associate Editor)
European Journal of Pediatric Neurology (Member)
Pediatric Neurology (Member)
Brain & Development (Member)
Journal Pediatric Neurology (Member)
International Symposium Celebrating the 50th Meeting of the JSCN Part IV

May 30th, Fri.

Room 2
Spinal muscular atrophy (SMA) is an autosomal recessive motor neuron disease characterized by degeneration of the anterior horn cells of spinal cord which leads to muscular paralysis and muscular atrophy. Clinically, SMA is categorized as type I (severe), type II (intermediate) and type III (mild), according to the age of onset and clinical severity. It occurs in approximately 1 in 10,000 live birth, and has a carrier frequency of 1 in 50. SMA is caused by homozygous deletion or mutations of the telomeric copy of the survival motor neuron (SMN1) gene on chromosome 5q13. All SMA patients carry at least one copy of a nearly identical SMN2 gene. However, a critical nucleotide change (C to T) at the 6th position of exon 7 in SMN2 results in about 80% of exon 7 in the majority of SMN2 messenger RNA (mRNA), lacking exon 7, thus producing a low level of functional SMN protein. This alteration, but not other variations in the SMN genes, affects the splicing pattern, and thus, causes the loss of an SF2/ASF-dependent exonic splicing enhancer or creates a heterogeneous nuclear ribonucleoprotein (hnRNP) A1-dependent exonic splicing silencer. Currently pursued therapeutic strategies for SMA include induction of SMN2 gene expression, modulation of splicing of SMN2-derived transcripts, stabilization of SMN protein, neuroprotection of SMN deficit neurons, and SMN1 gene replacement.

In recent years, sodium butyrate, phenylbutyrate, valproic acid (VPA), suberoylanilide hydroxamic acid (SAHA), benzamide M344, trichostatin A (TSA), gabapentin, albuterol, salbutamol, riluzole, hydroxyurea, aclarubicin, polyphenol botanical compound, and 5-(N-ethyl-N-isopropyl)-amiloride (EIPA) have been used in SMA-like mice, SMA patients’ fibroblasts, lymphoid cell lines or SMA patients, with some showing effective results either in terms of elevation of SMN2 expression in cell lines or improving muscle strength, lung function, have been increasing SMN2 gene expression in SMA patients. Clinical trials in SMA present a unique set of challenges, including the development of meaningful outcome measures and disease biomarkers. Discoveries regarding the genetics and pathogenesis of SMA have identified potential targets for pharmacotherapy, raising hope that better treatments will eventually be developed.

Dr. Yuh-Jyh Jong is the Professor of Graduated Institute of Medicine, College of Medicine, and Vice President of Kaohsiung Medical University, Kaohsiung, Taiwan. He received his MD and MS degrees and his specialty training in Pediatrics & Pediatric Neurology (Chair: Prof. Chi-Hsiun Chiang) at the Kaohsiung Medical University and D.M.Sci. degree in Tokyo Women's Medical University (Chair: Prof. Yukio Fukuyama), Japan. In 1985~1986, he had been a research fellow in the field of neuromuscular diseases at the Department of Pediatrics (Chair: Prof. Yukio Fukuyama, Neuromuscular Division Chief.: Prof. Makiko Osawa), Tokyo Women's Medical University and Department of Ultrastructural Research (Chair: Prof. Ikuya Nonaka), National Institute of Neuroscience, NCNP, Tokyo, Japan. Professor Jong has been a pioneer in the field of pediatric neuromuscular diseases in Taiwan, and he is actively involved in basic and translational research in spinal muscular atrophy (SMA) and neuromuscular diseases for past 20 years. In 2000, their research team had published the first mouse model of SMA in Nature Genetics and subsequently proved the novel ideas that SMA may become one of the first treatable inherited diseases in humans by using drugs which affect transcriptional activation and correction of the splicing of a copy gene in PNAS.
Spinal muscular atrophy (SMA) is an autosomal-recessively inherited disorder characterized by loss of motoneurons, in the spinal cord and lower brain stem, associated with neurogenic muscular atrophy. SMA is divided into three clinical types; Werdnig-Hoffmann disease (type 1), intermediate form (type 2) and Kugelberg-Welander disease (type 3). The gene responsible for most SMA, the survival motor neuron (SMN) gene, was identified in 1995 on human chromosome 5q13. Uncertainty persists regarding the pathological differences in clinical severity between SMA type 1 and milder forms of this disease. Furthermore, the precise mechanisms underlying the variable age at onset of the three types of SMA, which have a common genetic error, are unknown. Programmed cell death (PCD) may explain the pathogenesis of SMA type 1, but whether the mechanism of motoneuron death in SMA type 2 or 3 is associated with the reactivation or persistence of PCD remains to be undetermined. Some pathogenesis of SMA is still unknown. To elucidate the mechanism and timing of motoneuron death in SMA, we immunohistochemically analyzed the localizations of stress-related proteins, apoptosis-related proteins, and SMN protein in motoneurons using autopsied spinal cords of subjects, from fetuses to patients who had survived into their late teens. We also conducted an extensive literature search and the findings are discussed herein, with reference to our own experience.

Neuropathological analyses were performed on spinal cords obtained at autopsy from 2 fetuses with molecular genetically confirmed SMA, 9 SMA patients meeting clinical criteria [Munsat, 1992], six of whom had molecular genetically confirmed diagnoses, and 13 age-matched control individuals with neither neurological nor neuropathological disorders. Fully informed consent to perform these studies was obtained from family members. The main postnatal pathology of SMA was severe loss and degeneration of motoneurons in the spinal cord. Degenerating balloononed neurons (BNs) were filled with phosphorylated neurofilaments. Both BNs and remaining motoneurons diminished with aging. More motoneurons had survived in the spinal cord of a patient with SMA type 2 who had lived into the late teens than in an adolescent with type 1 SMA. Immature control motoneurons during the fetal period morphologically resembled the BNs in SMA or chromatolytic neurons. We speculate that motoneuron maturation is disrupted in SMA. Immature motoneurons that failed to form networks may be eliminated by PCD, and the severity of SMA may depend on the number of mature motoneurons surviving into the postnatal period. It is possible that a few immature motoneurons survive as BNs after birth but are incapable of prolonged survival. Our observations that SMN protein was as clearly detectable within the embryonal and postnatal cytoplasm of remaining motoneurons in SMA cases as in controls and that there were more GFAP positive cells in the anterior roots (glial bundles) than in the anterior horn in SMA specimens, suggest ‘dying-back’ axonopathy as the primary pathological change in SMA. This form of axonopathy would be initiated in the terminal axons of motoneurons in the spinal cord and their cell bodies would then undergo retrograde degeneration. Neither immunohistochemical analysis of other apoptosis-related proteins nor TUNEL analysis showed any significant difference between controls and SMA cases, and we detected no TUNEL-positive apoptotic neurons.

Our hypothesis does not contradict the evidence of PCD involvement in the development of SMA and might even explain the differences in maximum motor ability among the three types of SMA. We must reconsider SMA as an inborn defect rather than a postnatal degenerative disorder. This would open new treatment possibilities, including the embryonic therapy.

Education:
1993  Graduated from Wakayama Medical University

Professional background:
1993-1995 Resident in Tokyo Women's Medical University
1995-  Medical staff
2001-  Assistant professor
Congenital myasthenic syndromes (CMS) constitute a group of rare heterogeneous neurotransmission disorders caused by genetic defects of molecules at the neuromuscular junction (NMJ). Myasthenic symptoms, such as hypotonia, ptosis, fatigability and muscle weakness, usually manifest before age 2 years. These symptoms fluctuate and worsen with exertion and stress. A family history of similar symptoms supports a congenital origin. CMS is classified into presynaptic, synaptic and postsynaptic forms, according to the primary defect site. Progress in genetic analysis of CMS has allowed identification of mutations in nine genes encoding NMJ proteins. In the majority of cases, CMS primarily affects postsynaptic function and is the result of mutations located in the muscle AChR subunit genes. CMS patients have been reported in many parts of the world, but there have been only a few reports of adult CMS patients with acetylcholinesterase (AChE) deficiency and slow channel syndrome (SCS) in Japan. We previously reported a Japanese CMS patient with AChR deficiency who was diagnosed during childhood. The patient was a 13-year old Japanese boy with severe myasthenic symptoms since infancy. Ptosis had manifested at 5 months, a nasal voice at 2 years, of age. The diagnosis of sero-negative myasthenia gravis (MG) prompted thymectomy and immunosuppressive therapies including steroids and FK506. However, his clinical symptoms gradually worsened and he was ultimately limited to wheelchair activity. Genetic analyses for AChR, Musk, Rapsyn and collagenQ (ColQ) of AChE were negative. At 11 years of age, electron and confocal microscopic analyses of endplates showed severe deficiency of AChR, confirming the diagnosis of CMS with AChR deficiency. After establishing the CMS diagnosis, we stopped all immunosuppressive therapies. Instead, we he was treated with Ubretide and 3,4-diaminopyridine after obtaining informed consent.

We occasionally encounter problems in diagnosing CMS. During the neonatal or infantile period, symptomatic hypotonia is not specific. The onsets of some cases are delayed, occasionally until adolescence or even adult life. Since we do not have extensive experience with this syndrome, there is a possibility of missing these patients.

CMS should be considered not only in diagnosing sero-negative MG, but also in cases with hypotonia or muscle weakness of unknown cause. Making a definitive diagnosis requires special biopsy techniques allowing the NMJ to be sampled. Genetic analysis is the most reliable means of establishing the diagnosis, though mutations are detected in only one-third of patients. Even when the diagnosis of CMS has been confirmed, treatment can often be challenging. Quinidine produces dramatic results in treating SCS. However, there is essentially no effective therapy for most CMS cases, since these disorders are due to congenital molecular defects involving the NMJ. AChE inhibitors are sometimes effective for CMS with a positive Tensilon test. In refractory cases, 3,4-diaminopyridine is reportedly effective, though this drug has not been approved as a medication in Japan. We still face numerous problems in managing CMS patients.

Education:
Tokyo Women’s Medical University
Date of graduation: March 1998
Postgraduate course of Tokyo Women’s Medical University
Date of graduation: March, 2002
Ph.D in Pediatrics – Study of Congenital myasthenic syndrome
Extracurricular Education:
Studied in Department of Ultrastructructural Research, National Center of Neurology and Psychiatry, National Institute of from April to September in 2001
Studied in Columbia University and Case Western Reserve University in the USA in March, 2005 sent by Ministry of Education, Culture, Sports, Science and Technology
Work Experience:
Resident, Department of Pediatrics, Tokyo Women’s Medical University, Tokyo, Japan, 1998 – 2002
Assistant Professor: Department of Pediatrics, Tokyo Women’s Medical University, Tokyo, Japan, 2002 to the present time
Oral presentation

May 29\textsuperscript{th}, Thu.
Room 5, 6

May 30\textsuperscript{th}, Fri.
Room 6
O-033 Do all children with first episode of fever and seizure need a Lumbar puncture?

Ajit Rayamajhi, Ruby Joshi Bataajo, Chandeshwar Mahaseth
Department of Pediatrics, National Academy of Medical Sciences

Background: Prospective cross-sectional study was conducted for 1 year recruiting all children between 6–60 months with first episode of fever with seizure to find out whether younger children with first episode of fever and seizure had more chance of meningitis than febrile seizure than older children. Methodology: Those with CSF cells ≥5/mm³, protein ≥40 mg% and sugar ≤2/3 of blood sugar and/or positive Gram’s stain and/or bacterial culture were diagnosed meningitis and without, febrile seizure.

Results: Of the 175 included, male/female was 2.24, age 6–12 months 30%, 12–18 25%, >18 45%. Meningitis was 17% (6–12 months 57%, 12–18 30%, >18 13%; bacteria in culture 4.5%) and febrile seizure 83%. Regarding febrile seizure, family history was in 4%, cough 40% and rhinorrhea 13%. All culture positive meningitis was in 6–12 months and none had signs of meningeal irritation. Meningitis was significantly associated with vomiting, altered sensorium, bulging fontanel, signs of meningeal irritation, fever ≥72 hours, >3 seizures, seizure duration ≥15 minutes and focal seizure. Complex febrile seizure had sensitivity 67%, specificity 79% and signs of meningeal irritation sensitivity 20%, specificity of 97% diagnosing meningitis.

Conclusion: LP should be considered in all children 6–12 months presenting first time with fever and seizure.

（特别 Travelar Award）

O-034 Levetiracetam adjunctive therapy in children with refractory epilepsy

Kun-Long Hung, Ching-Wan Tsai, Ching-Wan Liu, Hung-Tsai Liao
Department of Pediatrics, Cathay Hospital, Taipei, Taiwan

[Background] To evaluate the efficacy and tolerability of levetiracetam (LEV), as adjunctive therapy in children with drug-resistant epilepsy.

[Method] Children with intractable seizures despite appropriate treatment were placed on LEV as add-on therapy at our institute during the past two years (Oct. 2005 to Oct. 2007). A retrospective analysis of medical information of these patients including age, six, seizure type, EEG findings, dose of LEV and its efficacy and adverse effect was conducted.

[Results] A total of 28 patients, aged 1 to 24 years, were enrolled. Males were 18 and females 10. The seizure types in semiology were partial with or without secondary generalization in 17 (60.7%), generalized in 8 (28.6%) and mixed in 3 (10.7%). The mean dose of LEV was 30.8 mg/kg/day (range 10–80). The mean period of follow-up was 12.3 months. Over 50% seizure reduction was observed in 11 patients (39.3%) including 2 seizure free. Five patients dropped out due to no effect in 4 and seizure increase in 1. Adverse effects were noted as somnolence in 4, poor appetite in 1 and behavior change in another 1.

[Conclusion] LEV was effective as adjunctive therapy for children with intractable epilepsy, either partial or generalized. It was well tolerated with minimal adverse effects.
O-035 Clinical analysis of 30 patients with tuberous sclerosis complex: relationship of abnormal brain imaging, seizures and cognitive function

Pou-leng Cheong¹, Yi-Ning Su², Wang-Tso Lee³
Department of Pediatrics, Hsinchu General Hospital, Executive Yuan, Hsinchu, Taiwan¹, Department of Medical Genetics, National Taiwan University Hospital, Taipei, Taiwan², Department of Pediatrics, National Taiwan University Hospital, Taipei, Taiwan

[Background] Tuberous sclerosis complex (TSC) is a genetic disorder with multisystem hamartomatous involvement, inherited as autosomal dominant but 60–70% sporadic. Cognitive impairment and behaviour problems (autism, ADHD, and learning disabilities) are common in patients with TSC. This study is to identify risk factors for their cognitive impairment.

[Methods] Medical records of patients aged below 18 years with TSC admitted to a tertiary medical center from 1993 to 2007 were reviewed. Mutational analysis of TSC1 and TSC2 genes were performed using denaturing high performance liquid chromatography (DHPLC) and direct sequencing. Statistical analysis was done using Mann–Whitney test and Chi-square or Fisher’s exact test.

[Results] 30 patients (male: female: 12:18) were included (median onset age: 17 months). The patients with anticonvulsant monotherapy (p: 0.01), focal epileptiform discharges in the electroencephalograms (p: 0.05) or absence of recurrent seizures under anticonvulsant (p: 0.01) may have better cognitive function. 13 (43%) patients have mental retardation. The presence of >10 cortical subcortical tubers (p: 0.118), infantile spasm (p: 0.264), or positive family history (p: 0.42) do not show significance in cognitive impairment. Mutational analyses show 5 TSC1 and 9 TSC2 mutations. The median onset age in TSC1 patients and TSC2 patients is 24 months and 6 months respectively, with no significant difference in disease severity.

[Conclusions] Cognitive outcome in the TSC is associated with the presence of refractory seizures, but not related to the number of cortical/subcortical tubers.
(JSCN Award for Asian Young Investigator)

O-036 Mozart effect on epileptiform discharge in children of epilepsy in Taiwanese

Lung-Chang Lin, Rei-Cheng Yang
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[Background] Music has a long history in healing physical and mental illness. The Mozart effect was initially reported by Rauscher and colleagues in 1993. They found the performance scores were 9 point higher in Mozart-listening group than silence or relax groups. Later, the beneficial influence of Mozart music on Parkinson’s disease, senile dementia, and attention-deficit hyperactivity disorder were reported. Few reports also demonstrated that patients with seizures exposed to Mozart’s music can significantly decrease the interictal epileptiform discharge of rolandic seizure. However, whether the Mozart’s music is also effective in other type of music remains unclear.

[Methods] Fourteen children with epilepsy, nine partial complex, two generalized tonic-clonic, two absence, and one rolandic, were exposed to Mozart’s sonata K.448 and then instructed to silence. At the same time, electroencephalogram was examined and the frequencies of epileptiform discharge were counted.

[Results] Twelve of fourteen patients with different seizure types who exposed to Mozart’s sonata K.448 revealed average decrease by 41.18 ± 26.73% in interictal spike frequencies. There was no significant difference among the patients of various seizure types.

[Conclusions] Our result suggests that the Mozart’s sonata K.448 is also effective in the decrease of interictal spike in the Taiwanese children with epilepsy.
(JSCN Award for Asian Young Investigator)
O-053  Phenotypic characteristic of galactosemia in the post-neonatal age in India

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[Summary] Classical Galactosemia is an inborn error of metabolism, which manifests in neonatal age group. Treatment with galactose free diet in neonatal age group, leads to symptom free outcome. In developing countries, lack of compulsory neonatal screening programme and lack of awareness leads to delay in diagnosis resulting in permanent neurological handicap. [Abstract] Classical Galactosemia is an inborn error of galactose metabolism caused by a deficiency of the enzyme Galactose-1-phosphate uridyl transferase leading to significant neurological impairment. In Indian population, incidence, phenotypic characteristics are less known. The present study shows the current phenotypes in Indian population with striking findings because of delayed diagnosis associated with neurological impairment. They were followed up for one year or more with Galactose free diet. We describe a cohort of 21 patients diagnosed by assay of galactose-1-phosphate uridyl transferase. Age at diagnosis was 6 months to 14 years with male preponderance. 20 patients demonstrated delay in developing motor milestones. 18 had hypotonia and 2 had hypertonia. 4 had extra pyramidal movements. 13 patients failed to develop language, 5 had dysarthria 9 patients had epilepsy. 9 patients had neurobehavioral problems, 16 patients had a history of prolonged neonatal jaundice with or without sepsis. Of 7 patients who underwent MRI, five had abnormalities. The EEG tracings of 9 patients showed epileptiform abnormalities. All patients of Galactosemia maintained on galactose free diet were followed for 1 year or more but there was no significant neurological improvement. Our observation reminds us of the severe consequences of treatable metabolic disorders due to delayed diagnosis and futility of galactose free diet for neurological outcome once the damage has set in.

O-057  New Microdeletion Syndrome Involving 2p15-16.1 in a Patient With Developmental Delay identified by CGH-array

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[Introduction] A newly recognized microdeletion syndrome of 2p15-16.1 has been reported by Rajcan-Separovic et al. (2007). Their reported two patients showed deletions with 4.5 Mb and 5.7 Mb, respectively. We encountered a patient with a deletion of this region. [Patient] A 4 years and 6 months girl was referred to our institution. There is no consanguinity and no family history. The girl was born with birth weight of 2020g as the first child of healthy parents. She had small stature, microcephaly, optic nerve atrophy and bilateral metatarsus abductus. Her dysmorphic facial expression consisted of hypertelorism, epicanthal folds, and low set ears. She had attention deficit hyperkinetic behavior and neurological examination showed (central) hypotonia. She had severely delayed development with DQ of 20. Brain radiological examination revealed bilateral perisylvian cortical dysplasia. Since her karyotype was normal, CGH-array analysis using agilent 105A was performed, and microdeletion of 2p15-16.1 has been identified. The deletion size was confirmed by FISH. The patient’s deletion was de novo as her parental genetic study did not show same deletion of this region. [Discussion] The smallest 3 Mb deletion of 2p15-16.1 was identified in a patient with dysmorphic facial expression and severe developmental delay. This is the third report of this deletion syndrome.
O-130 Clinical diagnosis and therapy analysis of opsoclonus–myoclonus syndrome

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[Abstract] Objective: Clinical manifestations of opsoclonus–myoclonus syndrome (OMS) children were summarized and analyzed and the clinical features and therapeutic approaches of OMS were investigated in order to improve its diagnosis and management. Methods: Clinical information on features and management of 6 cases of OMS inpatients being followed up from 2006 to 2007 were collected and analyzed. Results: Among the 6 cases, one was male and the other five were female. The age at the onset ranged from 12 to 26 months old (average 21 months). Four of them had parainfectious history. The visit presenting symptoms were opsoclonus, myoclonus, ataxia, sleep disturbances and behavioural problems in 6 cases. Urinary VMA was positive in 1 case. Abdomen B ultrasound showed a mild hepatomegaly in 4 cases. The EEG presented abnormal in 3 cases which showed slow background activity. Epileptiform discharges were found in none of the patients. MRI showed a high signal in medial longitudinal fasciculus and tectospinal tract on T2-weighted image in 1 case. Computerized tomography found L3–4 arachnoid cysts in 1 case and the others were normal. ACTH was given in all patients and was effective in all during acute stage. 2 cases were relapsed during follow-up stage. Conclusion: OMS is a rare neurological condition with opsoclonus, myoclonus, ataxia, sleep disturbances and behavioral problems, which might relapse easily and had adverse neurological outcome. ACTH therapy is effective in management of OMS.

O-148 Muscular dystrophies associated with nuclear envelope proteins

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Mutations in the genes encoding nuclear envelope proteins of lamin A-C (LMNA) and emerin (EMD) cause muscular dystrophies with life-threatening cardiac involvement. We examined clinical and pathological features of muscular dystrophies associated with these nuclear envelope proteins. Patients who suspected to have Emery–Dreifuss muscular dystrophy (EDMD) or limb girdle muscular dystrophy (LGMD) were analyzed. Skeletal muscles were examined using a set of histochemical and immunohistochemical analyses and electron microscopic observation. Mutation screening for EMD and LMNA was also performed.

We identified 33 patients in 27 families carrying a mutation in LMNA (laminopathy), and 20 patients in 18 families with a mutation in EMD (emerinopathy). In both genes, mutations were widely distributed through the genes however LMNA p. R453W mutation was frequently found in 7/27 (26%) families. Clinically, laminopathy patients showed earlier onset of the disease starting by muscle symptoms. In addition, calf hypertrophy was often seen. Cardiac symptoms appeared later. Both laminopathy and emerinopathy patients are clinically presented as EDMD, LGMD, or their intermittent forms. Pathologically, nuclear abnormalities are prominent together with the dystrophic changes. Muscular dystrophy associated with nuclear envelope proteins is not a rare disorder. Early diagnosis and careful follow up is necessary to avoid sudden cardiac death.
O-149  Electron microscopic analysis of myonuclei in edmd2/lgmd1b

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[Background] Autosomal dominant Emery-Dreifuss muscular dystrophy (EDMD2) and limb-girdle muscular dystrophy type 1B (LGMD1B) are caused by mutations in the gene encoding nuclear envelope protein, lamin A/C (LMNA). We examined detailed morphological changes of myonuclei in EDMD2 LGMD1B muscles.

[Methods] Using skeletal muscles from four patients with EDMD2 LGMD1B, histochemical, immunohistochemical and electron microscopic (EM) analyses were performed.

[Results] All muscle specimens showed mild dystrophic features with increased number of myonuclei with variable size. From longitudinal aspect of semithin sections, chained nuclei were frequently seen in subsarcaclemmal regions. On EM, 20% of myonuclei were fragmented or segmented into several particles in a line, which were mostly located in the periphery of myofibers. Besides, many myonuclei were irregular in shape revealing saw-tooth and serpentine features. Chromatin was disorganized in about 50% of myonuclei. Heterochromatin was scanty in amount, and partially absent from nuclear periphery. Euchromatin was less condensed. Specifically, various kinds of vacuoles were found inside or just beside myonuclei. Importantly, nuclei of satellite cells also showed scanty heterochromatin.

[Conclusion] In skeletal muscles from the patients with EDMD2 LGMD1B, myonuclei were significantly altered in their shape and chromatin, but even with these abnormalities myofibrils were well preserved in arrangement. Vacuoles, related to the degenerating or fragmented myonuclei are suspected to have roles in nuclear disruption. In addition, altered nuclei of satellite cells may reflect reduced capacity of regeneration in damaged muscle cells.

(JSCN Award for Asian Young Investigator)

O-150  Clinical, pathological and molecular genetic analysis of a Chinese family with dystrophinopathy

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[Abstract] Objective To analyze and determine the clinical, molecular pathology and genetic features of a Chinese family with dystrophinopathy. [Methods] Clinical data of the proband and his family members were collected. Immunohistochemistry staining was perfomed on muscular biopsy tissues with anti-merosin, emerin and the N, C and central rod domains of dystrophin. Genomic DNA was extracted using standard procedures from the peripheral blood leukocytes. Multiplex ligation-dependent probe amplification (MLPA) was used to test DMD gene to determine the ways and sites of genetic mutation, and analyze the relationships between genotype and phenotype. [Results] Patients from this family were diagnosed muscular dystrophy clinically, present atypical and serious manifestations although the immunohistochemistry analysis for the proband exhibited partial loss of dystrophin staining, and positive expression with merosin and emerin. Further test with MLPA detected the loss of exons 45～54 in DMD gene in the proband, while his mother had heterozygotic loss in exons 45～54. [Conclusions] The losses of exons 45~54 in the proband are all derived from his mother, who is a genetic mutation carrier with normal phenotype. He was diagnosed dystrophinopathy. At the same time, his partial loss of dystrophin is not paralleled to the out-of-frame mutation of the gene and his severe clinical manifestations. Abnormal expression of DMD is the pathological basis for dystrophinopathy phenotype. Its clinical outcome depends not only on the degree of the protein expression, but also on the function of the sites where the DMD gene loss occurs.
O-151 Clinical and molecular pathological analysis of congenital muscular dystrophy–20 cases

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[Abstract] Background The aim of this study was to find out the clinical diagnosis, immunohistochemistry features and follow-up information of 20 cases of The congenital muscular dystrophies (CMD). [Methods] Clinical manifestations and immunohistochemistry features of biopsied muscle specimens using anti-laminin α 2 (merosin), dystrophin, collagen VI, α-dystroglycan (α-DG) and β-dystroglycan antibodies were summarized and analyzed. [Results] All of the 20 patients were presented at birth or during the first six months of life with muscle weakness, hypotonia, contractures, and feeding difficulties or respiratory problems. Hematoxylin–eosin staining of skeletal muscles showed typical character of CMD. Twelve of the 20 patients were clinically diagnosed MDC1A (muscular dystrophy, congenital, 1A) and abnormalities of the white matter in these 12 cases was shown through T2-weighted magnetic resonance imaging of the brain. However, muscle biopsy was performed in six cases out of the 12 and merosin-stain was negative in immunohistochemistry which might be due to primary merosin deficiency. Eight cases were merosin-stain positive among which four were clinically diagnosed VI type collagenopathy and two of them had muscle biopsy. Immunohistochemistry staining showed collagen VI-stain negative. Another 3 cases had hypoglycosylation of α-DG with eye disorder and brain structural abnormality. The one case left was diagnosed RSMD1 (rickety spine muscular dystrophy type 1). [Conclusions] There are two types of CMD in these 20 cases. MDC1A is more common. "Alpha-dystroglycanopathy" and collagenopathy can be seen in merosin-positive cases. (JSCN Award for Asian Young Investigator)

O-166 Alglucosidase alfa in juvenile and adult patients with Pompe disease: results from a randomized, double-blind, multicenter, multinational, placebo-controlled study

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[Introduction:] Pompe disease (also known as acid maltase deficiency and glycogenosis type 2) is an autosomal recessive, metabolic myopathy caused by a deficiency of lysosomal acid α-glucosidase (GAA), an enzyme that degrades intralysosomal glycogen. Total or near total GAA deficiency leads to a rapidly progressive phenotype, with disease manifestations in infancy and early lethality from cardiac and respiratory failure. Partial deficiency results in later onset of symptoms (childhood to adulthood) and relentless progression to wheelchair dependency and respiratory failure. [Methods:] For study inclusion patients needed to be ≥8 years of age, ambulatory, free of invasive ventilation, and with quantifiable respiratory and lower extremity muscle weakness. Patients were randomized 2:1 to biweekly alglucosidase alfa (Myozyme®) 20 mg kg IV or placebo for 78 weeks at 8 centers in the US and Europe. Distance walked in the six minute walk test (6MWT) and % predicted forced vital capacity (FVC) were co-primary endpoints. [Results:] 90 patients (45M:45F; 93% Caucasian; age range 10–70 years) were enrolled. Mean baseline ± SD 6MWT distance was 327.4 ± 128.0 meters (50.1% predicted) and mean baseline FVC ± SD was 54.6 ± 14.8% predicted. At last evaluation (78 weeks), estimated mean absolute differences ± SD of 28.1 ± 13.1 meters in 6MWT distance (p=0.03) and 3.4 ± 1.2% in % predicted FVC (p=0.003) were observed in favor of alglucosidase alfa vs. placebo. Frequency of infusion associated reactions were comparable between alglucosidase alfa and placebo (approximately 25%). Three patients in the alglucosidase alfa treatment group experienced hypersensitivity reactions, two of whom discontinued treatment. All evaluable patients receiving alglucosidase alfa (n=59) developed IgG antibodies to rhGAA (mean time to seroconversion: 5.6 weeks). A trend toward decreasing IgG titers was observed over time (Median peak titer: 6,400, range 200 to 819,200. Median last titer (week 78): 1,600, range 0 to 819,200). [Conclusions:] In this long-term, first placebo-controlled study of rhGAA conducted in juveniles and adults with Pompe disease, alglucosidase alfa was shown to improve walking and pulmonary outcomes when compared to placebo. Follow-up data collection continues under an open-label extension study.
O-169 Valproic acid does not always increase SMN 2 expressions in fibroblasts from patient with SMA type 1

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Background: Most spinal muscular atrophy (SMA) patients lack SMN1, but their clinical severity is modified by the presence of a highly homologous gene, SMN2. Recently, valproic acid (VPA) has been regarded as a potential treatment for SMA patients because it was shown to increase SMN2 expressions. However, VPA effect varies among different studies.

Methods: In this study, we determined SMN2 transcript amounts in the fibroblast from a patient with SMA type I. The patient lacked SMN1 and carried two copies of SMN2. For time dependency experiment, we extracted RNA from the fibroblasts incubating with 500 μM of VPA for 0, 2, 4, 8, 16 and 24 hours and performed quantitative RT-PCR. For dose dependency experiment, we did the same procedures using the fibroblasts incubating with 0.5, 5, 50, 500 and 1000 μM of VPA for 16 hours.

Results: Time dependency or dose dependency experiment did not show any significant alteration in total, full-length (FL) and exon 7 deficient (Δ7) transcript amounts or Δ7/FL ratio in the VPA–treated fibroblasts, either.

Conclusion: In our study, VPA did not increase SMN2 expressions in fibroblasts from our patient, suggesting that VPA is not effective in this case. Quantitative RT-PCR of SMN2 transcripts may be used to predict the effect of VPA treatment in SMA.

(JSCN Award for Asian Young Investigator)
Poster presentation

May 29\textsuperscript{th}, Thu.
30\textsuperscript{th}, Fri.
Room Orion
P-07 Schizencephaly in Leopard syndrome — a case report —

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[Background] LEOPARD syndrome (LS) is an autosomal dominant disorder with multiple lentigines, congenital cardiac abnormalities, ocular hypertelorism, and retardation of growth. Deafness and genital abnormalities are less frequently found. Neurological deficit in LS included mental retardation, sensorineural deafness, oculomotor defect and seizures. Central nervous system anomaly was rarely reported in this disease. We report a patient of LEOPARD syndrome, who presented with hypertelorism, multiple lentigines and hypertrophic cardiomyopathy (HCM), has schizencephaly.

[Clinical details] The male proband was the second born child of healthy, unrelated parents. His face was characterized by ocular hypertelorism. Small, dark brown, irregularly shaped macules that varied in size had been observed on the trunk since infancy. Echocardiography demonstrated hypertrophic cardiomyopathy. Brain stem auditory evoked potentials test revealed a moderate hearing impairment. His developmental milestone was delayed. He developed epileptic seizures at 1-year- and 6-month of age. Brain magnetic resonance imaging revealed a wide, open cleft at the right parietal area. Open-lip schizencephaly associated with focal pachygyria is suggested. He had severe psychomotor retardation. Mutation analysis of this patient and his mother revealed a Y279C mutation in the PTPN11 gene.

[Conclusion] Partial agenesis of corpus callosum and Chiara I malformation have been reported in LS. The presence of schizencephaly associated with LS observed in our patient may represent, if not merely coincidental, a previously unrecognized brain malformation associated with LEOPARD syndrome. In patients with LEOPARD syndrome, brain imaging studies are suggested as part of the diagnostic work-up of LS.

(JSCN Award for Asian Young Investigator)

P-050 Mutation analysis and response to riboflavin therapy in Taiwanese MADD patients

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[Background] Multiple acyl-coenzyme dehydrogenase deficiency (MADD), or glutaric aciduria type II, is caused by defects in electron transfer flavoprotein or ETF-ubiquinone oxidoreductase (ETF:QO). This disorder can show neonatal onset form with or without congenital anomalies or later-onset milder form. Recently, riboflavin treatment has been known to be effective in MADD patients with mutations in the gene for ETF:QO (ETFDH).

[Methods] Four patients in 3 Taiwanese families with later-onset MADD were analyzed. Clinical information was reviewed according to their medical record. Mutation screening of ETFDH was done, together with other related genes, by using genomic DNA.

[Results] We identified mutations in ETFDH in all 4 patients; a compound heterozygous mutation of c.250G>A (p.A84T) and c.524G>T (p.A175L) in two sisters, a homozygous mutation of c.250G>A (p.A84T) in one patient, and a compound heterozygous mutation of c.250G>A (p.A84T) and c.380T>A (p.L127H) in one patient who died in one episode of severe metabolic acidosis at the age of 10. Riboflavin treatment to the other 3 patients resulted in improvement of muscle weakness with no more metabolic crisis.

[Conclusions] Later-onset MADD can show muscle weakness with lethal attack of metabolic problem. Since MADD patients with ETFDH mutations are responsive to riboflavin therapy, riboflavin treatment should be started as soon as possible together with genetic analysis.

(JSCN Award for Asian Young Investigator)
P-149 Tethered cord in Miller-Dieker syndrome — a new association —

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Introduction: Miller-Dieker syndrome is a disorder of neuronal migration caused by deletion of chromosome 17p13.3. It was characterized by classic lissencephaly and typical facial features. The association with tight filum terminale in Miller-Dieker syndrome was reported in previous study. We reported a new patient with tethered cord.

Patient: A 15 years old boy with the diagnosis of Miller-Dieker syndrome was regularly followed up at out-patient department because of intractable epilepsy and spastic posture that made him bed-ridden. Brain magnetic resonance imaging (MRI) disclosed lissencephaly, and chromosome study revealed 17p13.3 deletion. He was admitted due to perirenal fascitis complicated with left flank abscess. Lumbar MRI demonstrated tight filum terminale with tethered cord and urodynamic study revealed neurogenic bladder. Empyema of left lower pleural space developed after antibiotic treatment and surgical drainage for left flank abscess. He received video assisted thoracoscopic surgery for drainage but bronchopleural fistula complicated the course. He was repeatedly admitted due to recurrent pneumonia thereafter. He was expired after an episode of pneumonia with respiratory failure.

Conclusion: Tethered cord may be a new association in Miller-Dieker syndrome. Spinal MRI should be suggested in patients with Miller-Dieker syndrome to rule out tethered cord and early surgical intervention can avoid neurogenic bladder and related complications.

P-157 Decoy Receptor 3 ameliorates experimental autoimmune encephalitis through down regulation of innate and adaptive immunity concomitant with suppression of TH17 cells

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Decoy receptor 3 (DCR3), known as secreted form of TNFR superfamily, exhibits numerous immuno-modulated functions. Here we demonstrate intrathecal (IT) administration of DCR3 significantly ameliorates MOG-induced experimental autoimmune encephalomyelitis (EAE) in C57BL/6 mice despite of inflammatory cells infiltration into the spinal cord. Recently IL-17?producing T cell (TH-17) was highlighted mediating inflammatory pathology in certain autoimmune diseases. During EAE experiment, we observed less TNF-α, IFN-γ, IL-17 mRNA expression from CNS in mice treated IT DCR3 than control. We detected significant lower levels of IL-17 and higher levels of IL-4 from MOG-specific stimulation of splenocytes obtained from IT DCR3 treated mice than control. Ex vivo, less expression of IFN-γ on CD4 lymphocytes and especially TH17 cells was revealed in CNS lymphocytes from IT-DCR3 treated mice than control. Alternatively, we examined higher expression of IL-4 mRNA and more IL-4 producing CD4 T cells in the CNS of IT DCR3. DCR3 inhibited antigen-specific T cell proliferation. We finally proved DCR3 manipulated splenocytes preserves immune-modulated ability to reduce disease severity in EAE through adoptive transfer. Taken together, DCR3 suppresses inflammatory response and amends encephalogenic Th17 cells that at least in part attributes to either directly residential counteraction of DCR3 on CNS inflammatory process, or of DCR3 manipulated T cells introduce immuno-modulated effects on EAE. These data provide a therapeutic potential of DCR3 in human CNS autoimmune disease, such as multiple sclerosis.
P-206 Infants with diarrhea-related seizures were not associated with SCN1A mutation

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Background:
De novo mutations of the sodium channel gene SCN1A were found in patients with alleged vaccine encephalopathy. Infants with diarrhea were frequently associated with benign seizures resemblance to severe myoclonic epilepsy of infancy for which such mutations have been identified. In the present study, we investigate whether SCN1A mutation is also associated with benign seizures in patients with diarrhea.

Methods:
We retrospectively studied 15 patients with benign seizures associated with diarrhea. We reviewed the relation of seizures with diarrhea. Genomic DNA was extracted from the blood with the informed consent from the parents. Mutations in SCN1A were identified by PCR amplification with subsequent sequencing.

Results:
Of those 15 patients with benign seizures associated with diarrhea, no mutation of SCN1A was found. It indicate that SCN1A mutation may not be responsible for the benign associated with diarrhea.

Conclusion:
Benign seizures associated with diarrhea were not associated with SCN1A mutation. Other pathogenic mechanisms should be searched in the future.

P-212 Clinical course of epilepsy secondary to neonatal hypoglycemia

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Objectives: To report on clinical course of epilepsy in patients with epilepsy secondary to neonatal hypoglycemia.

Subjects: 9 patients with neonatal hypoglycemia and epilepsy were studied through reviewing their medical records retrospectively.

Results: The mean onset age of epilepsy was 2 years 7 months and mean follow-up period was 13 years. Initial seizure types were generalized convulsion in 6 patients, hemiconvulsion in 2, and versive seizure in 1. Most patients showed symptoms of occipital lobe seizures in their follow-up. All patients had status epilepticus. Seizures frequency was maximum during infancy and early childhood and decreased thereafter in all but one. One patient had surgical resection of the occipital lesion because of intractable seizures at the age of 16. MRI revealed cortical atrophy and T2 prolongation occipitally in 3 patients, parieto–occipitally in 1 patient, fronto–occipitally in 2 patients, diffuse white matter volume loss in 1 patient, and hippocampal atrophy in 1 patient. EEGs showed parieto–occipital spikes in 8 patients and multifocal spikes in 1 patient whose MRI revealed diffuse white matter lesion.

Conclusion: This study indicates that epilepsy secondary to neonatal hypoglycemia is intractable during infancy and early childhood with frequent status epilepticus but tends to decrease in older ages in most cases.
P-219  Risk for recurrence and outcome after a first unprovoked seizure in infancy

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Most children with single unprovoked seizure do not experience recurrence. However, there is limited data when the study confined to infants. This study assessed the risk for recurrence and outcome after an unprovoked seizure in this special population.

Between January 2002 and December 2005, all patients aged 29 days to 24 months, experiencing their first unprovoked seizure, were enrolled. Age of onset, sex, any prior febrile seizures, family history of epilepsy, seizure characteristics, etiology and EEG findings were collected. We recorded the recurrent time of the 2nd seizure if any and individualized management was given. At the end, we reevaluated the neurodevelopmental status and the use of antiepileptic drugs. All factors were analyzed for the recurrence and the chronic AED therapy

There were 87 infants, 71/87 (81.6%) experienced recurrence. The recurrent rates in idiopathic, cryptogenic, and symptomatic groups were all high: 38/53 (71.7%) , 9/9 (100%) , and 24/25 (96.0%) . 51/71 were on chronic AED therapy; only 6 from idiopathic group. 12/38 from idiopathic group were classified as cryptogenic after follow up.

Recurrence in infancy is high. Etiology is a significant risk factor for recurrence and chronic AED therapy. Without chronic AED therapy, a substantial number in idiopathic group could outgrow the seizures. Nearly 100% in cryptogenic/ symptomatic group experienced recurrence and required long-term AED therapy. About one third of the infants originally classified as idiopathic were revised to be cryptogenic during follow up.

P-244  Three-dimensional ultrasound application in a congenital lipoma of corpus callosum

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Background
Three-dimensional (3D) ultrasound (US) offers a new methodology for serial intracerebral structure and volume evaluation. Intracranial lipoma is a rare anomaly and usually localizes in the midline without symptom but occasionally is associated with seizure and mental dysfunction. We reported a baby with lipoma of corpus callosum (LoCC) that was found at fetal stage and the tumor volume was followed subsequently by 3D US.

Clinical details
The female infant was born to a 29-year-old mother at 38 weeks of gestation via normal delivery with a birth weight of 3148g. The pregnancy had been unremarkable until the 24th week when fetal US revealed a band form brain tumor over corpus callosum. The tumor was evaluated by US and MRI in utero and after birth. LoCC was diagnosed by hyperechoic signal in US and increased intensity on T1WI and T2WI in MRI. 3D US were done in gestation 34th week, 1st day, 6th week and 6th month of age to evaluate the shape and size of the tumor. Tumor volume increased gradually along splenium and inter-hemispheric space with circumvented vessels but no neurological deficit.

Conclusions
3D US provides an economic, convenient way to demonstrate the changing shape and volume of LoCC. Under increasing tumor size, LoCC may still have good outcome.
P-292 Treatment with autologous umbilical cord blood for infantile or childhood cerebral palsy

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Background: Despite new development in obstetrics and newborn medical care, the incident of hypoxic ischemic encephalopathy (HIE) in newborns is still high. There is no effective therapy at present, because the developing brain could not form integrated myelin sheath (induced decreased volume of white matter) and the white matter was damaged at the HIE condition. In addition, cortical lesions with neuronal loss and abnormal gliosis were also found. Treatment with autologous umbilical cord blood (UCB) for infantile cerebral palsy can be considered a safe procedure without concerns of graft rejection, which showed excellent therapeutic effect in this case.

Objectives and Methods: A boy with the diagnosis of infantile cerebral palsy induced by threatened presentation and HIE was given the treatment of autologous umbilical cord blood infusion intravenously under informed consent from his parents. We analyzed the outcome for any neurologic improvements by evaluation of his motor function skill by Prectal’s, Vojta’s methods, as well as Gross Motor Function Classification System (GMFCS). Brain sonography, MRI, EEG and laboratory data were examined before and after UCB treatment.

Results: The motor function skill evaluated by GMFCS showed level V before UCB (age 1Y4M) and level III after UCB (age 1Y7M), and the estimated developing ages by Prectal’s and Vojta’s methods is approximately 5 M/O before, and 8–9 M/O after UCB treatment. All the laboratory data were within normal ranges including values of T Cells and B Cells, and T Cell subset analysis before and after UCB treatment. Brain sonography before treatment revealed: (a) bilateral periventricular leukomalacia, Gr.3, (b) suspect mild cortical atrophy, (c) mild lateral ventriculomegaly, and (d) left subpialcystal cyst. No obvious changes were noted after treatment. Brain MRI before treatment showed: (a) periventricular leukomalacia, Gr.3 (late stage), and (b) brain atrophy. No prominent changes were found after treatment. EEG showed diffuse cortical dysfunction especially over bilateral central-temporal-occipital areas before UCB treatment, but near-normal record after UCB treatment.

Conclusion: (1) We treated a case with cerebral palsy with autologous umbilical cord infusion without any side effects, and with proceeding neurological development with concordance to his developmental age. (2) Autologous umbilical cord blood infusion may become an option for treatment of cerebral palsy or similar diseases in the near future.