

**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.

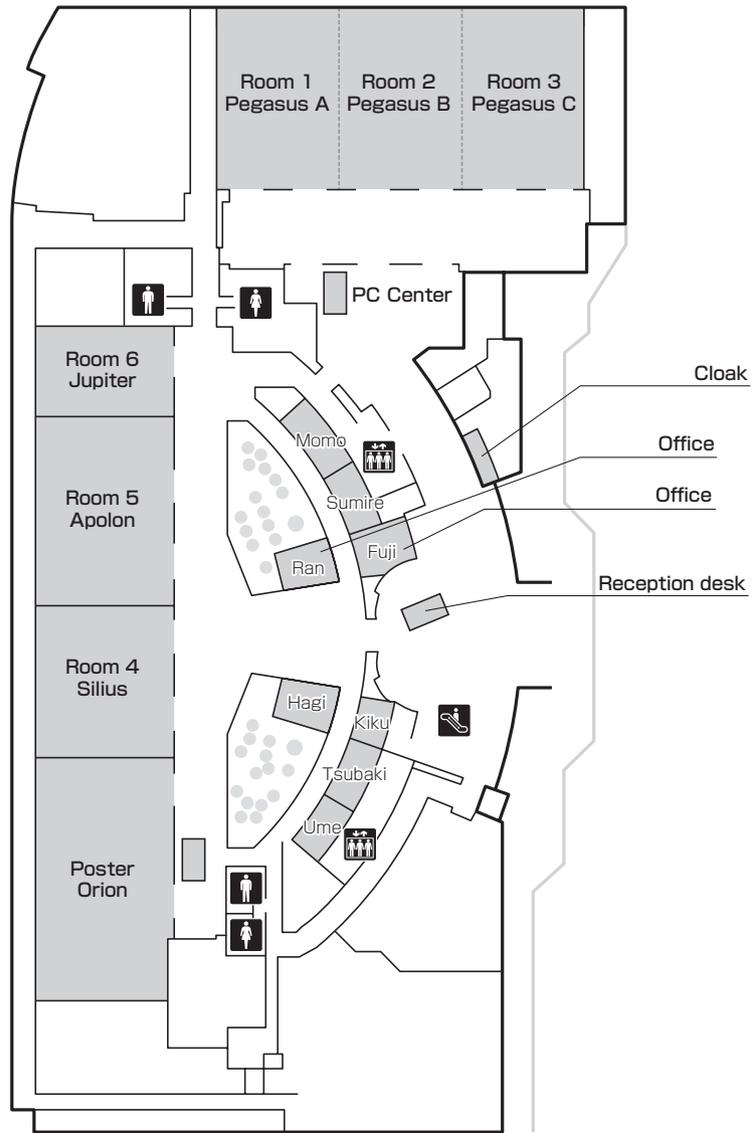
We will do our utmost to make your stay in Tokyo as enjoyable as possible. Please mark your calendar right now and submit your abstracts.

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

INFORMATION

Hotel Nikko Tokyo
1F



■ Hotel Nikko Tokyo
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	Apolon Room 5	Jupiter Room 6	Orion Poster Presentation	
8:00				
9:00		Board of directors 9:00 ~ 11:00		
10:00				
11:00				
12:00	General meeting of councilors 11:30 ~ 13:20	Board of directors (new) 11:40 ~ 12:00		
13:00				
14:00	Opening Address ISC50 JSCN Part I 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:Ein-Yiao Shen Solomon L Moshé Chair:Yoshiyuki Suzuki Shinichi Takeda Chair:Shigehiko Kamoshita Yoshiyuki Sankai		Poster tack up	
15:00				
16:00				
17:00				
18:00				
19:00				
20:00		ES 1 19:00 ~ 21:00 The Second annual meeting of the Japan Association of Child Sleep Chair: Jun Kohyama		
21:00				

- [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)

Pegasus A + B + C			Silius	Apolon
Room 1	Room 2	Room 3	Room 4	Room 5
8:00				
9:00	Presidential Lecture Clinical Aspects on Myotonic Dystrophy in Childhood 8:50 ~ 9:20 Makiko Osawa Chair: Koumei Kumagai			
10:00	Presentation of JSCN Awards to Outstanding young Investigators 9:30 ~ 9:50 Chair: Toshiaki Hashimoto			
11:00	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			
12:00	SL 1 11:30 ~ 12:30 Mind viewed from brain Makoto Iwata Chair: Teruhisa Miike			
13:00	LS 1 12:30 ~ 13:30 1) New progress in treatment of epilepsy Akio Ikeda 2) Treatment of intractable epilepsies based on the mechanism of action of the antiepileptic drugs Kenji Sugai Chair: Tatsuya Tanaka		LS 2 12:30 ~ 13:30 1) Diagnosis and management of Seizures in the ICU Akihisa Okumura 2) Brain monitoring made easy Anita Kharbteng Chair: Wu Xi-Ru	
14:00	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		Learning disabilities 14:30 ~ 15:30 (O-001~ 006) Chair: Masutomo Miyao Masao Aihara	Epilepsy/Seizures 1 14:30 ~ 15:40 (O-024~ 030) Chair: Takehiko Morimoto Osamu Kanazawa
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		Cerebral palsy rehabilitaion 1 15:30 ~ 16:30 (O-007~ 012) Chair: Takahiro Nara Hideo Shimoizumi	Epilepsy/Seizures 2 15:40 ~ 17:00 (O-031~ 038) Chair: Kun-Long Hung Shinichi Hirose
17:00			Cerebral palsy rehabilitaion 2 16:30 ~ 17:40 (O-013~ 019) Chair: Kazuo Kodama Hiroshi Ozawa	Epilepsy/Seizures 3 17:00 ~ 18:10 (O-039~ 045) Chair: Kazuhiro Haginoya Toshio Hanai
18:00			Developmental disorders& Genetics 17:40 ~ 18:20 (O-020~ 023) Chair: Yasuyuki Niki Mitsuhiro Kato	
19:00				
20:00	19:00 ~ Offical Social Party Awarding party			

	Jupiter Room 6	Orion Poster Room			
8:00					
9:00		Poster Presentation			
10:00					
11:00					
12:00					
13:00					
13:00	LS 3 12:30 ~ 13:30 Presentation based on the brain science; Success of our talk at the international conference Hiroshi Otsubo Chair: Takao Takahashi				
14:00					
15:00	Metabolic/Degenerative disorders 1 14:30 ~ 15:50 (O-046 ~ 053) Chair: Hiroko Kodama Tatsuro Izumi	Anomaly/Chromosomal disorders 14:30 ~ 15:05 (P-001 ~ 007) Chair: Takahisa Wada Toshiyuki Yamamoto	Metabolic disorders 1 14:30 ~ 15:15 (P-041 ~ 049) Chair: Fujii Tatsuya Koji Inui	Developmental disorders (diagnosis) 14:30 ~ 15:15 (P-083 ~ 091) Chair: Akihiro Yasuhara Yoshiaki Saito	Neonatology 1 14:30 ~ 15:05 (P-122 ~ 128) Chair: Seiichi Sugama Hirochi Koizumi
16:00	Anomaly/Chromosomal disorders 15:50 ~ 16:50 (O-054 ~ 059) Chair: Hirofumi Ohashi Hiroshi Tamai	Myopathy 1 15:05 ~ 15:50 (P-008 ~ 016) Chair: Nobutada Tachi Yoshiko Nomura	Metabolic disorders 2 15:15 ~ 16:05 (P-050 ~ 059) Chair: Nobuyuki Shimozawa Hitoshi Sakuraba	Developmental disorders (examination 1) 15:15 ~ 15:45 (P-092 ~ 097) Chair: Tatsuya Ogino Michiko Sugama	Drugs 15:05 ~ 15:35 (P-129 ~ 134) Chair: Susumu Ito Yasufumi Utsumi
17:00	Genetics 1 16:50 ~ 17:50 (O-060 ~ 065) Chair: Toyojiro Matsuishi Naofumi Kajii	Myopathy 2 15:50 ~ 16:30 (P-017 ~ 024) Chair: Yasuhiro Takeshima Hirofumi Komaki	Vascular disorders 16:05 ~ 16:45 (P-060 ~ 067) Chair: Hiromi Sato Akira Oka	Developmental disorders (examination 2) 15:45 ~ 16:15 (P-098 ~ 103) Chair: Hideaki Kanemura Tohshin Go	Sleep/Autonomic nerve 15:35 ~ 16:15 (P-135 ~ 142) Chair: Yutaka Awaya Shinji Fujimoto
18:00		Genetics 2 16:30 ~ 17:10 (P-025 ~ 032) Chair: Toshiyuki Kumagai Eiji Nanba	Tumor/Neurosurgery 16:45 ~ 17:30 (P-068 ~ 076) Chair: Hisashi Kawawaki Sumimasa Yamashita	Developmental disorders (support) 16:15 ~ 17:00 (P-104 ~ 112) Chair: Masaki Ohno Kazuya Itomi	Brain anomaly 16:15 ~ 16:55 (P-143 ~ 150) Chair: Kyoko Ito Tomohide Goto
19:00		Genetics 3 17:10 ~ 17:50 (P-033 ~ 040) Chair: Yukio Sawaishi Kiyotaka Tomiwa	Mental disorders 17:30 ~ 18:00 (P-077 ~ 082) Chair: Akemi Tomoda Junichi Furusho	Developmental disorders (treatment) 17:00 ~ 17:45 (P-113 ~ 121) Chair: Asayo Ishizaki Hiroyoshi Koide	Infection/Immunology 1 16:55 ~ 17:25 (P-151 ~ 156) Chair: Naoyuki Tanuma Naoya Itokazu
20:00					Infection/Immunology 2 17:25 ~ 17:55 (P-157 ~ 162) Chair: Ayako Muto Mikio Hiraiwa

		Pegasus A + B + C			Silius	Apolon
		Pegasus A	Pegasus B	Pegasus C	Room 4	Room 5
		Room 1	Room 2	Room 3		
8:00		MS 1 8:00 ~ 9:00 Visually diagnosed epileptic seizures: focal seizures Masako Sakauchi Chair: Tatsuro Izumi	MS 2 8:00 ~ 9:00 EEG—an introduction— Solomon L. Moshé Chair: Yoko Otsuka	MS 3 8:00 ~ 9:00 Effect of GH treatment on the brain metabolism Shinichi Nijima Chair: Goro Takada		
9:00		DL 1 9:00 ~ 9:40 Surgical management of pediatric epilepsy Tomokatsu Hori Chair: Tatsuya Tanaka	ISC50 JSCN Part III (DL 3) 9:00 ~ 9:40 Fukutinopathy Yukio Fukuyama Chair: Ikuya Nonaka	DL 5 9:00 ~ 9:40 Understanding and support for developmental disorders with autism—from the viewpoint of living and working together after adolescence— Masami Sasaki Chair: Tsunekazu Yamano	DL 7 Cerebral Palsy 9:00 ~ 9:40 Kenji Yokochi Chair: Toru Konishi	Encephalitis/Encephalopathy 1 9:00 ~ 9:50 (O-085 ~ 089) Chair: Junichi Takanashi Hideo Yamanouchi
10:00		S 1 9:50 ~ 12:00 Diagnosis and treatment for childhood epilepsy – Expert opinion – Chair: Hirokazu Oguni Yoko Otsuka	ISC50 JSCN Part III (S 3) 9:50 ~ 12:00 Phenotypic Spectrum of Fukutinopathy Chair: Tadayuki Ishihara Hideo Sugie	S 5 9:50 ~ 12:00 Contribution of Child Neurology toward the Study of Developmental Disabilities Chair: Hitoshi Hara Tatsuya Koeda	S 7 9:50 ~ 12:00 Comprehensive medical care and support for children with severe motor and intellectual disability Chair: Eiji Kitazumi Yuji Iwasaki	Encephalitis/Encephalopathy 2 9:50 ~ 10:30 (O-090 ~ 093) Chair: Yoshihiro Maegaki Miyabi Mizuguchi
11:00						Encephalitis/Encephalopathy 3 10:30 ~ 11:30 (O-094 ~ 099) Chair: Takashi Ichiyama Masaya Kubota
12:00						Encephalitis/Encephalopathy 4 11:30 ~ 12:10 (O-100 ~ 103) Chair: Masaji Shiomi Shinichiro Hamano
13:00		LS 4 12:20 ~ 13:20 Challenges toward treatments for neurodegenerative diseases Shoji Tsuji Chair: Yoshikatsu Eto	LS 5 12:20 ~ 13:20 Treatment for intractable epilepsy Paolo Curatolo Chair: Kazuie Inuma	LS 6 12:20 ~ 13:20 Working women and psychological development of children Hisako Watanabe Chair: Tsunekazu Yamano	LS 7 12:20 ~ 13:20 Mechanically assisted coughing in patients with neuromuscular disease Yuka Ishikawa Chair: Mitsuru Kawai	
14:00		JSCN General assembly 13:30 ~ 14:30				
15:00			ISC50 JSCN Part IV 14:30 ~ 15:40 Topics in neuromuscular disorders Yuh-Jyh Jong Yasushi Ito Keiko Ishigaki Chair: Masaharu Hayashi Yuh-Jyh Jong	WS 14:30 ~ 15:40 Current topics of encephalitis and encephalopathy Chair: Hideo Yamanouchi Masashi Mizuguchi	Imaging 1 14:30 ~ 15:30 (O-066 ~ 071) Chair: Hiroaki Shiihara Satoko Kumada	Development/Evaluation 14:30 ~ 15:30 (O-104 ~ 109) Chair: Miho Nakamura Kazue Igarashi
16:00		DL 2 15:40 ~ 16:20 Bench to Bedside, Tertiary Centres to Developing World: "Tailoring" Best Available Neuroprotection in High and Low Resource Settings Osuke Iwata Chair: Hitoshi Yamamoto	DL 4 15:40 ~ 16:20 Selective dorsal rhizotomy for spasticity in cerebral palsy children Takaomi Taira Chair: Kayoko Saito	DL 6 15:40 ~ 16:20 Epigenetic mechanism regulating neural cell fate determination Kinichi Nakashima Chair: Kousaku Ohno	Epilepsy 5 15:30 ~ 16:40 (O-072 ~ 078) Chair: Masaharu Ohfu Yukitoshi Takahashi	Developmental disabilities (investigation) 15:30 ~ 16:30 (O-110 ~ 115) Chair: Shinichi Hirabayashi Takashi Hayashi
17:00		S 2 16:20 ~ 18:30 Hot Topics in Neonatal Neurology Chair: Masahiro Hayakawa Shinichi Nijima	S 4 16:20 ~ 18:30 Neurosurgical approach from a perspective of pediatric neurology Chair: Shizuo Ohi Hiroaki Date	S 6 16:20 ~ 18:30 Epigenetics in Neurodevelopmental Diseases Chair: Tateo Kubota Shinji Fushiki	Epilepsy 6 16:40 ~ 17:40 (O-079 ~ 084) Chair: Eiji Hattori Akie Miyamoto	Developmental disabilities (support) 16:30 ~ 17:30 (O-116 ~ 121) Chair: Akinori Hoshika Jiro Ono
18:00						Autism/Others 17:30 ~ 18:20 (O-122 ~ 126) Chair: Yushiro Yamashita Satoshi Sanada
19:00		ES2 19:00 ~ 21:00 The Committee of the pharmacological issues – Concerta® Distribution management committee – Chair: Tasuku Miyajima Kitami Hayashi		ES3 19:00 ~ 20:00 Multiple Sclerosis and Neuromyelitis Optica in Japan Chair: Toshiro Hara	ES4 19:00 ~ 21:00 The Committee of the social acitivity Chair: Takeo Sugimoto	ES5 19:00 ~ 20:00 The Committee of the pharmacological issues –Botulinum Toxin Treatment– Chair: Yoko Otsuka Atsuo Nezu
20:00						
21:00						

	Jupiter	Orion			
	Room 6	Poster Room			
8:00					
9:00	Neuroimmunology 9:00 ~ 9:40 (O-127 ~ 130) Chair: Yukiko Hirano Tatsuo Oya				
10:00	Neonatal Imaging/Others 9:40 ~ 10:40 (O-131 ~ 136) Chair: Satoshi Kusuda Tetsuo Kubota				
11:00	Neonatal Seizures/EEG 10:40 ~ 11:40 (O-137 ~ 142) Chair: Fumio Hayakawa Shuichi Tsuneishi	Poster Presentation			
12:00	Neurosurgery 11:40 ~ 12:20 (O-143 ~ 146) Chair: Yasuo Aihara Katsunori Fujii				
13:00					
14:00					
15:00	Muscular dystrophy 1 14:30 ~ 15:50 (O-147 ~ 154) Chair: Yukitatsu Ishikawa Yukiko Hayashi	Encephalitis 1 14:30 ~ 15:15 (P-163 ~ 171) Chair: Kitami Hayashi Atsushi Imamura	Epilepsy 1 14:30 ~ 15:05 (P-203 ~ 209) Chair: Kimio Minagawa Hideki Horita	Imaging 2 14:30 ~ 15:15 (P-240 ~ 248) Chair: Yasuhiko Kawakami Masafumi Morimoto	Development 14:30 ~ 15:10 (P-281 ~ 288) Chair: Masumi Inagaki Satoshi Takada
		Encephalitis 2 15:15 ~ 15:55 (P-172 ~ 179) Chair: Koji Ushijima Takuya Tanabe	Epilepsy 2 15:05 ~ 15:40 (P-210 ~ 216) Chair: Shuto Yoshikawa Mariko Maezawa	Imaging 3 15:15 ~ 15:50 (P-249 ~ 255) Chair: Muneaki Matsuo Eiji Nakagawa	Cerebral palsy Rehabilitation 1 15:10 ~ 15:50 (P-289 ~ 296) Chair: Mana Kurihara Atsushi Ieshima
16:00	Muscular dystrophy 2 15:50 ~ 16:40 (O-155 ~ 159) Chair: Yoshinobu Otani Masafumi Matsuo	Encephalitis 3 15:55 ~ 16:35 (P-180 ~ 187) Chair: Ryutarō Kira Seiji Aso	Epilepsy 3 15:40 ~ 16:15 (P-217 ~ 223) Chair: Kouzaburo Aso Yasuhiro Suzuki	Peripheral nerve 15:50~16:35 (P-256 ~ 264) Chair: Masayuki Shimono Tsuyoshi Okinaga	Cerebral palsy Rehabilitation 2 15:50 ~ 16:20 (P-297 ~ 302) Chair: Masaru Tatsuno Tadashi Kitahara
17:00	Pompe disease 16:40 ~ 17:50 (O-160 ~ 166) Chair: Hiroyuki Ida Akemi Tanaka	Encephalitis 4 16:35 ~ 17:05 (P-188 ~ 193) Chair: Yukihiko Fujita Kyomi Hirayasu	Epilepsy (medical treatment) 16:15 ~ 16:55 (P-224 ~ 231) Chair: Hideo Nagao Nobuaki Iwasaki	Morphology/Pathology 16:35 ~ 17:10 (P-265 ~ 271) Chair: Keiko Shishikura Tomoyuki Takano	Cerebral palsy Complications 16:20 ~ 17:05 (P-303 ~ 311) Chair: Masayuki Sasaki Yukikatsu Ochiai
	Neuromuscular disorders 17:50 ~ 18:20 (O-167 ~ 170) Chair: Yoshihiro Takeuchi Yuichi Goto	Encephalitis 5 17:05 ~ 17:50 (P-194 ~ 202) Chair: Hideo Aiba Hiroschi Yoshioka	Epilepsy 4 16:55 ~ 17:35 (P-232 ~ 239) Chair: Akiko Matsumoto Tetsuo Matsuzaka	Metabolic/Degenerative disorders 17:10 ~ 17:55 (P-272 ~ 280) Chair: Kazutoshi Nakano Hitoshi Osaka	Cerebral palsy Involuntary movement 17:05 ~ 17:45 (P-312 ~ 319) Chair: Kazuo Higuchi Toyoko Kanda
18:00					
19:00					
20:00					
21:00					

	Pegasus A	Pegasus B	Pegasus C
	Room 1	Room 2	Room 3
8:00	<p>MS 4 8:00 ~ 9:00 A Clinical Approach to the Dysmorphic Child Kenjiro Kosaki Chair: Hitoshi Yamamoto</p>	<p>MS 5 8:00 ~ 9:00 Pediatric neuroimaging diagnosis A to Z Hiroshi Oba Chair: Toshiaki Hashimoto</p>	<p>MS 6 8:00 ~ 9:00 Visually diagnosed epileptic seizures: generalized seizures Hirokazu Oguni Chair: Shunsuke Ohtahara</p>
9:00	<p>DL 8 9:00 ~ 9:40 Regulatory mechanism of neuronal migration mediated by the microtubule-associated protein doublecortin and its partners Teruyuki Tanaka Chair: Takao Takahashi</p>	<p>Involuntary movement/Epilepsy (PC movie) 9:00 ~ 9:50 (O-178 ~ 182) Chair: Shuji Wakai Kazue Kimura</p>	<p>Supportive diagnosis (PC movie) 9:00 ~ 9:50 (O-189 ~ 193) Chair: Takanori Yamagata Ryouichi Sakuta</p>
10:00	<p>Sleep/Behavior disorders 9:40 ~ 10:50 (O-171 ~ 177) Chair: Jun Kohyama Katsuo Sugita</p>	<p>Epilepsy/Seizures 4 9:50 ~ 10:50 (O-183 ~ 188) Chair: Harumi Yoshinaga Toshisaburo Nagai</p>	<p>Dystonia (PC movie) 9:50 ~ 10:50 (O-194 ~ 199) Chair: Akihiko Tateno Tetsuzo Tagawa</p>
11:00	<p>Closing address 10:50 ~ 11:00</p>		
12:00		<p>Seminars open to the public How we can help Japanese children to be happy, thoughtful and warmhearted? 12:00 ~ 16:00 Chair: Toyojiro Matsuishi, Yoichi Sakakibara</p>	
13:00		<p>How we can help Japanese children to be happy, thoughtful and warm-hearted? Makiko Osawa (President of the Congress, Department of Pediatrics, Tokyo Women's Medical University)</p>	
14:00		<p>Introduction to the audience I Toyojiro Matsuishi (Department of Pediatrics and Child Health, Kurume University School of Medicine)</p> <p>Introduction to the audience II Yoichi Sakakihara (Department of Child Care and Education, Ochanomizu University)</p>	
15:00		<p>1. Early risers and early sleepers are healthy both mentally and physically Kohyama Jun (Tokyo Kita Shakai Hoken Hospital)</p> <p>2. Challenge of pediatric neurologists to child mental problems: From a clinical and neuroscientific point of view Shinichiro Nagamitsu (Department of Pediatrics and Child Health, Kurume University School of Medicine)</p>	
16:00		<p>3. PTSD and the developing brain Nobumasa Kato (Department of Psychiatry, Showa University School of Medicine)</p> <p>4. As a care-partners for Children who need Special Education Michiko Hara (Faculty of Education, Gunma University)</p> <p>5. The prevention of childhood depression Kayo Inoko (Tokyo Institute of Psychiatry)</p>	
17:00		<p>Closing Remarks Toyojiro Matsuishi</p>	
18:00			

Day 1 Room 5 (Apolon)

Opening Adress

**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, HongKong, 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
Chairman 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
Chairman 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—Cyberoid, biorobotics, control for supporting disabled persons—
Chairman Shigehiko 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 Ohinata (Department of Pediatrics, Asahikawa Medical College)
 - 5) Noboru Ohki (NoruPro Light Systems, Inc)
 - 6) Takako Jodoi (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)

Day 2 Room 1~3 (Pegasus)

8:50 ~ 9:20 Presidential Lecture

Clinical Aspects on Myotonic Dystrophy in Childhood

Chairman Koumei Kumagai (chiisakihananosono)

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

10:23 Presentation of Awards to JSCN Contributors

10:33 2) Japanese Contributions To Child Neurology – An International Perspective –

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

10:49 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)

Day 2 Room 4 (Silius)

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: Masutomo Miyao, Masao Aihara

O-001 A study of the early detection method of dyslexic children : I. Oral sentence reading task

* Uchiyama Hitoshi¹, Seki Ayumi^{2,3}, Koeda Tatsuya^{2,3}

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 Ayumi^{1,2}, Uchiyama Hitoshi³, Koeda Tatsuya^{1,2}

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 Katsuo¹, Sugita Kiyoko², Fujii Katsunori³

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 Hiroshi^{2,4}

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 Keiko⁴,
Wakamiya Eiji³, 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 Hisaki², Tsuchiya Shigeru³

1) Division 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 Tetsuro¹, Hoshino Hideki¹, Mizuguchi Koichi¹, Kubota Masaya¹, Morota Nobuhito², Oka Akira^{1,3}
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 Atsuo, Takeshita 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 Magara¹, Konishi Tohru¹, Kinoshita Satoru²
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 psychophysiologic disorders in our hospital**
* Takeshita Eri, Otani Ryoko, Itabashi Hisashi, Kita Syunji, Shimamura Keiichi, Murakami Nobuyuki, Sakuta Ryoichi, Nagai Toshiro
Dokkyo Medical University Koshigaya Hospital, Saitama, Japan
- O-014 New QOL Assessment Questionnaire for Persons with SMIDS**
* Matsumoto Akiko¹, Miyazaki Shuji¹, Hasegawa Sakurako²
1) KOBATO GAKUENN Aichi Human Service Center, Aichi, Japan, 2) Institute for Developmental Research, Aichi Human Service Center, Aichi, Japan
- O-015 Severely disabled children with respirator and/or tracheotomy: investigation in 8 prefectures**
* Sugimoto Tateo^{1,2}, Tamura Masanori^{1,3}
1) The Ethics Committee, Japan Pediatric Society, Tokyo, Japan, 2) Biwako Gakuen Medical and Welfare Center, Yasu, 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^{1,2}, Ieshima Atsushi¹, Kuwashima Katsuko¹, Kondou Ikuko¹
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 Yusaku^{1,2}, Yamamoto Hitoshi², Fukuda Miho², Murakami Hiroshi², Kamiyama noriko², Hashimoto Syuji²
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 Satoshi¹, Matsuda Nobuko¹, Matsui Gakuyou¹, Yamamoto Akio¹, Sunartini Hapsara²

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-ichi², Inazawa Johji³

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

Day 2 Room 5 (Apolon)

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 Tomohiko³, Kubota Tetsuo⁴, Maruyama Koichi⁵, Itomi seiko⁶, Kato Toru², Sofue Ayako⁷, Kajita Mitsuharu⁸, Okumura Akihisa⁹, Natsume Jun³

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 Jun^{1,2}, Maruyama Koichi³, Sofue Ayako⁴, Okumura Akihisa⁵

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 Colony, 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 Setsuri³, Maruyama Kouichi⁴, Katou Toru², Kubota Tetsuya⁵, Sofue Ayako⁶, Okumura Akihisa⁷, Natsume Jun¹

1) Department of Pediatrics, Nagoya University school of Medicine, Nagoya, Japan, 2) Okazaki City Hospital, 3) Tousei 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 Tohru^{1,2}, 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

O-028 An Epidemiological Study of Children with Status Epilepticus in Okayama (2003-2005)

* 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 Ryutarō, 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 Sonoko^{1,2}, 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 Hiroka, Takahashi Yukiotoshi, Mine June, Ohotani Sanae,

Ohotani Hideyuki, Ikeda Hiroko, Shike Tatsuhiko, Shimomura Jiro, Kubota Yuhko, Kubota Hidemoto, Shigematsu Hideo, Inoue Yushi, Fujiwara 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 Tatsuhiro

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 Lumbar 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 Lin^{1,2}, Rei-Cheng Yang^{1,2}

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 Osamu^{1,2}, 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

Chair: Kazuhiro Haginoya, Toshio Hanai

O-039 Vigabatrin treatment of infantile spasms due to Tuberous sclerosis.

* Kitai Yukihiko, Morita Yoshiko, Araya Ken, Tominaga Kouji, Nabatame Shin², Shimono Kuriko, Okinaga Takeshi, Oaono Keiichi

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, Shizuoka Children's Hospital, Shizuoka

O-041 Regional cerebral blood flow of West syndrome quantified with 3DSRT

* Hamano Shinichiro¹, Yoshinari Satoshi^{1,2}, Higurashi Norimichi^{1,2}, Tanaka Manabu¹, Minamitani Motoyuki^{2,3}, Kikuchi Kenjiro², Koichihara Reiko^{1,4}, 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 1H-MR spectroscopy

* Imamura Atsushi, Miyajima Hiroko, Matsuo 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 Sumimasa², Maegaki Yishihiro³, Fukumura Shinobu⁴, Takayama Rumiko⁴, Uematsu Mitsugu⁵, Haginoya Kazuhiro^{5,6}

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 holocoboxylase synthetase deficiency with West syndrome

* Hattori Ayako¹, Ando Naoki¹, Kobayashi Satoru¹, Ito Tetsuya¹, Fujimoto Shinji²,

Ban Kyoko³, Ishikawa Tatsuya^{1,4}, Togari Hajime¹

1) Department of Pediatrics and Neonatology Nagoya City University, Graduate School of Medical Sciences, Nagoya, Japan, 2) Tsutsujigaoka Kodomo Clinic, 3) Department of Pediatrics, Yokkaichi Municipal Hospital, 4) Nihonn Fukushi University

O-045 Immunologic aspects of West syndrome, through lymphocyte subsets and serum cytokines: 1st report.

* Shiihara Takashi, Watanabe Mio

Department of Neurology, Gunma Children's Medical Center, Gunma, Japan

Day 2 Room 6 (Jupiter)

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)

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

* Sakuraba Hitoshi

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

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

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

* Hayashi Masaharu¹, Ohto Tatsuyuki², Araki Satoshi³

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

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

* Naito Etsuo¹, Shimakawa Seishi¹, Nishimura Mio¹, Kotani Yumiko², Kagami Shoji²

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

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

* Itoh Masatsune, Kakinuma Hiroaki

Department of Pediatrics, Kanazawa Medical University, Ishikawa, Japan

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

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

* Osaka Hitoshi^{1,3}, Kurosawa Kenji², Iai Mizue¹, Yamada Michiko¹, Yamashita Sumimasa¹

1) Division of Neurology, Kanagawa Childrens Medical Center, 2) Division of Genetics, Kanagawa Childrens Medical Center, 3) Kanagawa Cancer Center Research Institute

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

* Harshuti Shah¹, Zachary Grinspan²

1) Rajvee child Neuro and orthospine hospital, Ahmedabad, India, 2) Department of child neurology, Columbia University, Newyork, Newyork, USA

15:50 ~ 16:50 Anomaly/Chromosomal disorders

Chair: Hirofumi Ohashi, Hiroshi Tamai

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

* Akasaka Noriyuki¹, Tohyama Jun¹, Saito Naka¹, Sasaki Masayuki²

1) Department of Pediatrics, Epilepsy center, Nishi-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 Shozo^{1,2}, Hayashi Shin^{1,2}, Imoto Issei^{1,2}, Nakagawa Eiji³, Goto Yu-ichi⁴, Inazawa Johji^{1,2}
 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, Musashi Hospital, Tokyo, Japan, 4) Department of Mental Retardation and Birth Defect Research National Institute of Neuroscience, National Center of Neurology and Psychiatry (NCNP), Tokyo, Japan
- O-056 Application of in-house array-CGH for investigation and diagnosis of congenital genomic disorders**
 * Hayashi Shin^{1,2}, Honda Shozo^{1,2}, Imoto Issei^{1,2}, Inazawa Johji^{1,2}
 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 Kouji³, Ohno Kousaku², 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, Tohyama Jun², Okumura Akihisa³, Maegaki Yoshihiro⁴, Oguni Hirokazu⁵
 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 Matsuishi, Naofumi Kajii
- O-060 Global analysis of CpG methylation status in murine fetal brain prenatally exposed to bisphenol-A**
 * Yaoi Takeshi¹, Itoh Kyoko¹, Nakamura Keiko^{1,2}, 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 NKX2-1 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 Hideo², Fukuda Tokiko², Osawa Junko, Suzuki Teruhiko, Ohzeki Takehiko

Department of Pediatrics, Hamamatsu University School of Medicine, Shizuoka, Japan, 2) Department of Pediatrics, Medical University

Day 2 Poster Room (Orion)

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 Acrodysostosis with Os odontoideum

* Fukamachi Makoto¹, Mori Atsuko¹, Moriuchi Hiroyuki², Fukuda Masafumi¹

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 Yuzo^{1,2}

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, Yamagata 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 Yuu², 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 Liang Steven Shinnforng 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 Mamiko, 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 Hirofumi¹, Hayashi Yukiko², Kato Mitsuhiro³, Sakuma Hiroshi¹, Saito Yoshiaki¹, Nakagawa Eiji¹, Sugai Kenji¹, Sasaki Masayuki¹, Nonaka Ikuya^{1,2}, 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, 3) Department of Pediatrics, Yamagata University, Yamagata, Japan
- P-011 A case of normokalemic periodic paralysis with persistent muscle weakness**
 * Terazawa Daisuke, Orii 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 Takahiro¹, Ishikawa Aki¹, Ohya Kazuhiro¹, Tachi Nobutada²
 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 Teruhisa
 Department of Child Development, Kumamoto University Hospital, Kumamoto, Japan
- P-016 Clinical course of childhood myasthenia gravis**
 * Shiraishi Kazuhiro, Natori Chieko
 The department of neuropediatrics, 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 Makoto², 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 Kiyoko^{1,2}, Kawashima Hisashi¹, Hoshika Akinori¹, Minami Narihito³, Nishino Ichizo³
 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, Shinno 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) Ikezawa 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 Toshihiro¹, Kamimaki Isamu¹, Goto Tomohide², Sakuta Ryoichi³

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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

* Akiyoshi Kensuke¹, Suenobu Souichi¹, Sonoda Kouji¹, Maeda Tomoki¹,

Korematsu Seigo¹, Izumi Tatsuro¹, Ishikawa Yukitoshi²

1) Department of pediatrics and child neurology, Oita university, Oita, Japan, 2) Department of pediatrics, National Yakumo hospital, Hokkaido, Japan

P-026 Genetic analysis in mental retardation and expansion of a research resource repository

* Nakagawa Eiji¹, Takano Kyoko², Wada Takahito³, Kubota Takeo⁴, Kato Mitsuhiro⁵, Nanba Eiji⁶,

Saitoh Shinji⁷, Inazawa Johji⁸, Kurosawa Kenji⁹, Goto Yuichi^{1,2}

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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 Takahito¹, Hirabayashi Shin-ichi²

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P-028 Juvenile Huntington Disease associated with Cerebeller hypoplasia

* Yoshinari Satoshi^{1,2}, Hamano Shin-ichiro^{1,2}, Minamitani Motoyuki^{2,3}, Tanaka Manabu¹,

Higurashi Norimichi^{1,2}, Eto Yoshikatu²

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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 Yoko², Oka Eiji⁴
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P-032 Nedd4-2 variant in a SMEI cohort

* Kurahashi Hirokazu^{1,2}, Kato Toru³, Kira Ryutarō⁴, 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, Okazaki Municipal Hospital, Okazaki, Japan, 4) Department of Pediatrics, Kyushu University, Fukuoka, Japan

17:10 ~ 17:50 Genetics 3

Chair: Yukio Sawaishi, Kiyotaka Tomiwa

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, Haginoya Kazuhiro², Fukuyo Naomi¹, Wakusawa 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 Takehiko², Saito Kayoko³, Yamamoto Toshiyuki¹
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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, Fujii Katsunori¹, Tanabe Ryo¹, Sawaishi Yukio², Kawato Jin³, Kouno Youichi¹
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 Manabu², Shiihara Takashi³, Tsuru Tomohiko⁴, Aizaki Koichi⁴, Toribe Yasuhisa⁵, Arai Hiroshi⁶
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P-039 Early neurological characteristics of 3 patients in a family with the severe type of MCT8 deficiency
 * Sawaishi Yukio¹, Yano Tamami¹, Watanabe Yasuhiro¹, Hirayama Aya², Makuta 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 Masaya³
 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: Fujii 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 mtDNA11778 mutation which became clear after the diagnosis of SCD
 * Tomita Sunao^{1,2}, Hoshino Ai¹, Hanafusa Yukiko¹, Shigetomo Rituko¹, Kumada Satoko¹, Kurihara Eizi¹
 1) Neuropediatrics of Tokyo megalopolis 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 Wakana^{1,2}, 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 Higashiyama Medical Center for the Severely Disabled

P-045 A case of obstructive respiratory distress of mucopolysaccharidoses type 2 improved by nppv
 * Matsui Shuji, Shiiki Toshihide, Takethi Nobuyuki, Tyou Hiroyuki, Funahashi Masuko, Suzuki Yasuyuki
 Tokyo Childrens Rehabilitation Hospital, Tokyo, Japan

P-046 Enzyme replacement therapy in a female case of mucopolysaccharidoses type 2 (Hunter syndrome) .
 * Sato Tatsuharu¹, Honda Ryouko¹, Imamura Yoshihiko¹, Turu Akira³, Matumoto Tadasi², Moriuti 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 Yukihiko, 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, Noudomi Seishiro, 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^{1,2}, Ohkuma Aya², Goto Kanako², Hayashi Yukiko K. Yukiko K.², Yuh-jyh Jong¹, Ichizo Nishino²

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P-051 Analysis of cerebral molecular species of phospholipids in D-bifunctional protein deficiency

* Saitoh Makiko¹, Yamashita Sumimasa², Itoh Masayuki³, Mizuguchi Masashi¹

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P-052 A infantile case of rapidly progressive vanishing white matter disease during half a year

* Hoshino Ai¹, Tomita Sunao¹, Kumada Satoko¹, Hanafusa Yukiko¹, Nigo Ayako², Okumura Sayaka², Kurihara Eiji¹, Yamamoto Toshiyuki³

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P-053 Irritability in a patient with Megalencephalic leukoencephalopathy with subcortical cysts

* Amemiya Kaoru¹, Kashii Hirohumi¹, Mizuno Yoko¹, Suzuki Riina¹, Koide Ayaka¹, Kubota Masaya¹, Ide Syuhei²

1) Tokyo Metropolitan Hachioji Children's Hospital, 2) TOBU RYOIKU CENTER

P-054 A case with congenital disorders of glycosylation

* Ohto Tatsuyuki¹, Enokizono Takashi¹, Iwasaki Yoko², Hirai Naomi¹, Tanaka Ryuta¹, Sasaki Masayuki³, Ohno Kohsaku⁴, Yuasa Isao⁵, Kamota Tomohiro¹

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P-055 An autopsy case of congenital disorders of glycosylation with recurrent pulmonary hemorrhage

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P-056 Mass spectrometry for rapid diagnosis of congenital disorders of glycosylation type Ia

* Fujiwara Shinichi¹, Okamoto Nobuhiko², Uetake Kimiaki¹, Onodera Takashi³, Yagy Kazuyori⁴, Sueta Keitaro⁴, Asahina Naoko⁴, Shiraishi Hideaki⁴, Saitoh Shinji⁴

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P-057 The research of Menkes disease with symptoms and treatments

* Ozawa Hiroshi¹, Kodama Hiroko²

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 Keisuke^{1,2}, 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 Hironori², Suzuki Yoiti³, Yamaguchi Seiji²

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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 Jun-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 Makoto², Hayashi Masaharu³, Tanuma Naoyuki³, Miyata Rie³

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 Sinichi

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P-067 Case report : 2 patients with Cutis Marmorata Telangiectasia Congenita

* Gotoh Chika, Akiyoshi Kensuke, Maeda Tomoki, Suenobu Souichi, Korematsu Seigo, Izumi Taturou

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16:45 ~ 17:30 Tumor/Neurosurgery

Chair: Hisashi Kawawaki, Sumimasa Yamashita

P-068 Lateral ventricle cholid plexus papilloma of an infant with hypernatremia

* Tanda Koichi¹, Ouchi Kazutaka¹, Nabeshima Kanae¹, Komatsu Hiroshi¹, Nakajima Humiaki¹, Inoue Yasuo², Kawarabuki Kentaro², 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

* Ohtani Anna

The Department of Pediatrics, Teine Keijinkai Hospital, Hokkaido, Japan

P-071 a case of infantile 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 Nozomi¹, Oosako Yutaka², Yatusiro Kazutaka³

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P-074 A case of leukoencephalopathy with cereberal atxia after the chemotherapy of acute leukemia (AML)

* Ninomiya Shinya¹, Inoue Takahito¹, Yonekura Michitaka¹, Fujita Takako¹,

Ihara Yukiko¹, Tomonou Yuko¹, Nakamura Noriko¹, Ideguchi Hiroshi¹, Yasumoto Sawa¹, Sakiyama Michiyo², Hirose Shinichi¹

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 Masayasu², Kudo Kazuko¹, Kamota Tomohiro¹

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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 Ryutaru, Fujita Yukihiro

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 Ryouko, Koike Makiko, Mazaki Kaoru, Shimamura Keiichi,

Murakami Nobuyuki, Sakuta Ryouiti, Nagai Tosirou

Dokkyo University School of Medicine Kosigaya Hospital, Saitama, Japan

P-079 Maltreatment made adjustment disorder in younger twin but not in older with autistic disorder

* Nara Chieko¹, Yokoyama Hiroyuki², Hirose Mieko¹, Wakusawa Keisuke¹,
Tuchiya Shigeru¹

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P-080 Three cases of social anxiety disorder developed in childhood

* Yokoyama Hiroyuki¹, Hirose Mieko², Nara Chieko², Wakusawa Keisuke²,
Haginoya Kazuhiro³, Tuchiya Shigeru², Iinuma Kazuie⁴

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P-081 An intractable epilepsy case with repeated forced normalization

* Hirose Mieko¹, Yokoyama Hiroyuki², Haginoya Kazuhiro^{1,3}, Kikuchi Atsuo^{1,4},
Nakayama Tojo¹, Uematsu Mitsugu¹, Iinuma Kazuie⁴, Tsuchiya Shigeru¹

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P-082 4 patients with Munchausen syndrome by proxy who appealed for neurological symptoms

* Miwa Mami¹, Nakamura Yukiko¹, Matsuda Hiroo^{1,3}, Bessyo Fumio¹, Kato Masae²

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 Ryusaku, Suzuki Shuhei

Department of Developmental Brain Science Osaka Medical College

P-084 A clinical analysis of schoolchildren with dysgraphia

* Kawatani Masao¹, Nakai Akio¹, Hiratani Michio²

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P-085 Cognitive function in patients with Williams syndrome

* Sunahara Mariko¹, Inoko Kayo², Osawa Makiko¹

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P-086 Developmental changes of visuo-spatial abilities and Kanji copying in people with Williams syndrome

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P-087 The developmental processes of maternal attachment in mothers of children with AD/HD

* Mano Shoko¹, Uno Hiroyuki², Horiuchi Fumie³

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P-088 Sleep study of pre/post Summer Treatment Program in children with AD/HD

* Iwasaki Mizue¹, Matsuishi Toyojiro², Iemura Akiko², Oya Takashi², Iizuka Chiho², Nakasima Masayuki²,
Nagamitsu Shinichiro², Yamashita Yushiro²

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P-089 The fact of adult ADHD

* Nishimaki Atsuko, Miyao Masutomo, Okuyama 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 Yuko^{1,3}, Inouwe Nario², Ihara Yukiko^{1,3}, Kanaumi Takeshi^{1,3}, Inoue Tkahito³, Yasumoto Sawa³, Hirose Shinichi³
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P-091 Comparison of Emotion in Patients with Asperger Syndrome and AD/HD

* Yamashiro Dai, Kanemura Hideaki, Kaga Yoshimi, Aoyagi Kakurou, 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 Ogino, Michiko Sugama

P-092 Developmental Change of Qualitative scores of The Boston Qualitative Scoring System (BQSS)

* Nakano Kousuke¹, Ogino Tatsuya², Watanabe Kiyoko³, Takeuchi Akihito³, Oka Makio³, Ohtsuka Yoko³
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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 variability of the observation of pupillary grating response in normal adults

* Sakai Shinya¹, Hirayama Kazumi², Kato Mitsuhiro³, Saitoh Shinji⁴, Sakai Naoko⁵
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 Shinji⁴, Sakai Naoko⁵, Seiwa Chizuru⁶, Sudo Mutsuko⁷, Nio Eiko⁷, Shiraishi Hideaki⁴, Takayanagi Masaru⁸, Kobayashi Yasuko⁹
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P-096 Intervention of Kanji writing in 2 Japanese dyslexic children

* Nakamura Masako^{1,2}, 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 Tomoka^{1,2}, Inagaki Masumi¹, Gunji Atsuko¹, Yatabe Kiyomi¹, Kaga Makiko¹, Gotoh Takaaki³, Koike Toshihide³

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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 Makiko¹

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 Keisuke^{1,2}, Tsuchiya Shigeru¹, Yokoyama Hiroyuki³, Kawashima Ryuta², Sugiura Motoaki⁴, Sassa Yuko², Hyeonjeong Jeong²

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P-100 Characteristic prefrontal activation by 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 Chihiro¹, Sakaue Yuko¹, Iwami Mika¹, Okada Masako¹, Nishikura Noriko^{1,2}, Yoshioka Seiichirou^{1,3}, 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 Chiaki³

1) The Division of Pediatric Neurology, Fukuoka Children's Hospital Medical Center, 2) Fukuoka Municipal Welfare Center for The Disabled, 3) Fukuoka-West Rehabilitation Center for Children

- P-106 Questionnaire for medical consultaion of pervasive developmental disorders to medical institutions**
 * Nishimura Satoko
 Department of Pediatrics, Kibougaoka Rehabilitaiton 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 Taishi^{1,2}, Ishikawa Michiko², Iguchi Toshiyuki^{2,3}, Imaeda Masayuki^{2,4},
 Asai Tomoko^{2,5}
 1) Osaka-hamamatsu joint center for mental development, 2) Department of neonatology and pediatrics nagoya city university medical school, 3) Hoshigaoka matanity 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 Toshisaburo¹, Kubayashi Chika¹, Tagawa Tetsuzo^{2,3}, Taniike Masako^{2,4},
 Imaishi Hidenori^{2,5}, Arai Hiroshi^{2,6}, Tanabe Takuya^{2,7}, Yabuta Reiko^{2,8}, Tanaka Jyunko^{2,9}, Nishida Masaru^{2,9}
 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) Tanakakitaumeda Clinic, 10) Hirakataryoikuen
- P-110 Analysis of medical information communicated in the field of developmental diorder practices**
 * Horiguchi Toshihiro¹, Akiyama Chieko², Kon Kaori³
 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) Kaishundo-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 deveopmental disorders**
 * Hayashi Yuko
 Faculty of Health and Welfare, Prefectural University of Hiroshima, Mihara, Japan
- 17:00 ~ 17:45 Developmental disorders (treatment)**
Chair: Asayo Ishizaki, Hiroyoshi 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 Kakurou¹, 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 Yushiro², Matsuishi Toyojiro²
 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 Hiroshi¹, Kajimoto Madoka², Ichiyama Takashi², Hurukawa Susumu²
 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 Toyojiro
 Department of Pediatrics and Child Health, Kurume University School of Medicine
- P-120 Efficiency of aripiprazole**
 * Ogasawara 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 Yasuko¹, Araki Atsushi¹, Kaneko Kazunari², Kuniyoshi Kyouko³, Kusumoto Kenji³
 1) Department of Pediatrics Kansai Medical Collage, Osaka, Japan, 2) Department of Pediatrics Kansai Medical College, Osaka, Japan, 3) Department of Plastic and Reconstructive surgery Kansai Medical Collage, 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 sever congenital myotonic dystrophy**
 * Otsuka Harumi¹, Suzuki Yasuhiro¹, Iwamatsu Toshiyuki¹, Tanabe Ryou², Mizoguchi Eriko³, Sasaki Kaori⁴, Saitou Kayoko^{3,5}, Oosawa Makiko³
 1) Department of Neonatology, Chiba Municipal Kaihin 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 2 weeks of age**
 * Kato Toru¹, Tuji Takeshi¹, Hayakawa Fumio¹, Kidokoro Hiroshi², Kubota Tetsuo², Suzuki Motomasa³, Maruyama Koichi³, Natsume Jun⁴, Okumura Akihisha⁵, Watanabe Kazuyoshi⁶
 1) Okazaki City Hospital, Department of Pediatrics, Okazaki, 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-Sulfatoxymelatonin and 8-OHdG in low birthweight infants**
 * Araki Akiko^{1,2}, Shirai Masaru², Ohinata Jyunko¹, Suzuki Nao¹, Takahashi Satoru¹, Tanaka Hajime¹, Oki Jyunichi², Fujieda Kenji¹
 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, Zaito Ayuko, Mukae Tokutaro, Okada Junichiro, Hirose Akiko, Kanda Hiroshi, Fujino Hiroshi, Maeno Yasuki, Iwata Osuke, Matsuishi Toyojiro
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 Kuniaki¹, Kirino Tomoko², Fujiwara Yumi², Ushida Miyuki², Endo Shoichi¹
1) The Department of Neurology, Kagawa National Children's Hospital, Zentuji, Japan, 2) The Department of Pediatrics, Kagawa National Children's Hospital, Zentuji, 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 HHV-6 encephalopathy

* Saida Satoshi¹, Tanaka Rieko^{1,2}
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 Hideyuki¹, Tanaka Masaki^{1,2}, Takahashi Yukitoshi¹, Inoue Yusi¹, Fujiwara Tateki¹, Sasagawa Mutsuo³, Mizobuti Masahiro⁴
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 Chiho, Ohya Takashi, Nagamitsu Shinichiro, Inada Hiroko, Matsuishi Toyojiro
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, Joudoi Takako², Tomoda Akemi¹, Shiraishi Seiji³, 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

* Ariei Junko¹, Kanbayashi Takashi³, Kubota Hiroaki², Yano Tamami⁴, Sawaishi Yukio⁴, Watanabe Yasuhiro⁴

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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 Toshiyuki¹, Kodama Hiroko¹, Nakamoto Natsue^{1,2}, Ogita Kaori¹, Amakata Kaori¹, Kaneko Sono¹, Fujii Yasushi¹, Fujita Yasuko¹, Yanagawa Yukishige¹

1) Department of Pediatrics Teikyo University School of Medicine, Tokyo, Japan, 2) Faculty of Health Sciences Department of Occupational Therapy, Mejiro University, Tokyo, Japan

P-140 A 1-year-old girl with comorbid obstructive sleep apnea syndrome and restless legs syndrome

* Kato Kumi^{1,2}, Mohri Ikuko¹, Taniike Masako¹

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.

* Akitani Susumu^{1,2}, Miyamoto Yukinobu¹

1) The Department of Pediatrics, Sanaikai general Hospital, Saitama, Japan, 2) The Department of Psychiatry, Saitama Children Medical Center, Saitama, Japan

P-142 Two cases of acute autonomic neuropathy with local hyperhidrosis

* Arai Hidee¹, Kubota Hiroaki², Tanabe Yuzo^{1,3}, 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 hemimegalencephaly: the mildest form of hemimegalencephaly?

* Ono Yoichi¹, Tohyama Jun², Sugai Kenji³, Maegaki Yoshihiro¹, Ohno Kousaku¹

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 Hemimegalencephaly diagnosed by convulsion

* Nakashima Kentaro¹, Tsuchioka Ayako¹, Arima Keitaro¹, Nakagawa Machiko¹,

Tsuruta Shio¹, Kusakawa Isao¹, Hosoya Ryouta¹, Ogihara Masaaki²

1) The Department of Pediatrics, St. Luke's International Hospital, Tokyo, Japan, 2) Ogihara clinic, Tokyo, Japan

P-145 3cases of fetal MRI, showed extension of ventricles, hypogenesis of corpus callosum and cerebellum.

* Takahashi Kan¹, Satou Atsushi¹, Mimaki Masakazu¹, Saitou Makiko², Oka Akira¹, Mizuguchi Masashi²

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P-146 Management strategy of epilepsy in malformed brains

* Saito Yoshiaki^{1,3}, Sugai Kenji¹, Nakagawa Eiji¹, Sakuma Hiroshi¹, Komaki Hirofumi¹, Sasaki Masayuki¹, Otsuki Taisuke², Ohno Kouyo³, Kondo Akiko³, Maegaki Yoshihiro³, Ohno Kousaku³

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- P-147 A Case of C (Opitz Trigenocephaly) Syndrome with Brain Anomaly**
 * Mori Atsuko¹, Fukamachi Makoto¹, Kaname Tadashi³, Moriuchi Hiroyuki², Fukuda Masafumi¹
 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 Akira¹, Ogami Aya², Morioka Shigemi¹, Hasegawa Tatsushi¹, Tozawa Takenori¹, Isoda Kenichi¹, Matsui Fumihiko¹, Morimoto Masafumi¹
 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 Takayuki¹, Okubo Takashi¹, Hashimoto Kiyoshi², Fujino Osamu²
 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 Yuki¹, Hayashi Masaharu², Miyata Rie², Tanuma Naoyuki², Ohya Tatsuo¹
 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 Toshiaki¹, Hattori Hideji¹, Okano Yoshiyuki¹, Tsuruhara Akitoshi¹, Matsuoka Osamu¹, Yamano Tsunekazu¹, Nakano Eiko², Matsui Masaru², Tanaka Masami², Konishi Tetsuro², Tanaka Keiko³
 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, Taniuchi 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 Tomonari¹, Kato Takeo¹, Daifu Tomoo¹, Shibata Minoru¹, Yamanaka Yasunari¹, Shiraishi Kazuhiro³, Tomiwa Kiyotaka², Nakahata Tatsutoshi¹
 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 Yukiko, Funatsuka Makoto, Kodaira Kayano, Ishigaki Keiko, Nakayama Tomohiro, Osawa Makiko
 Department of Pediatrics, Tokyo Women's Medical University, Tokyo, Japan

17:25 ~ 17:55 Infection/Immunology 2

Chair: Ayako Muto, Mikio Hiraiwa

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 Wang,² Su-Feng Wu³, Chich-Chien Wang,¹ Jen-Hsin Kao^{2,4}, Pao-Luh Tao⁴, Chia-Cho Wu⁵, Chien-Len Liao⁶, Huey-Kang Sytwu^{2,6,7}

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 Takanori¹, 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, Shiroma 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 Yukitoshi³, Oka Akira¹, Mizuguchi Masashi⁴

1) The Department of Pediatrics, University of Tokyo, Tokyo, Japan, 2) Department of Mental Retardation and Birth Defect Research, National Institute of Neuroscience, NCNP, 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 Mamoru³, Inoue Takushi¹, Maegaki Yoshihiro⁴, Ohtsuka Yoko¹

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Day 3 Room 1 (Pegasus A)

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 Fujiwara (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 Nijima (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 Unibersity Hospital)
- 2) Tatsuya Koeda (Department of Education Faculty of Regional Sciences Tottori University)
- 3) Kazuhiko Saito (Kohnodai Hospital, International Medical Center of Japan)
- 4) Ichiro Sora (Department of Biological Psychiatry, Graduate school of Medicine, Tohoku University)

Day 3 Room 2 (Pegasus B)

8:00 ~ 9:00 Morning Seminar 2

EEG:an intoduction

Chairperson Yoko Ohtsuka (Deperntment 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
Tatsushi Toda (Division of Clinical Genetics, Osaka University Graduate School of Medicine)

12:20 ~ 13:20 Luncheon Seminar 5

Treatment for Intractable Epilepsy

Chairman Kazuie Inuma (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)

Day 3 Room 3 (Pegasus C)

8:00 ~ 9:00 Morning Seminar 3

Effect of GH treatment on the brain metabolism

Chairman Goro Takada (Yonezawa National Hospital)

Shinichi Nijima (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 Tsunekazu 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 (AERRPS)

Yutaka Awaya (Department of Pediatrics, Seibo International Catholic Hospital)

3) Classification of Influenza encephalopathy

Masashi Shiomi (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 Necdin

Kazuaki 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)

Day 3 Room 4 (Silius)

9:00 ~ 9:40 Didactic Lecture 7

Cerebral Palsy
 Chairman Tohru Konishi (Nagaoka Ryoikuen)
 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 (Tobu 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 Hosupital organization Higashisaitama Hospital)
 Yuka Ishikawa (Department of Pediatrics, National Hospital Organization, Yakumo 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 Akio¹, Matsuki Ken-ichi²

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 Akihisa¹, Hayakawa Masahiro², Tsuji Takeshi², Saito Masako¹, Tanaka Kyoko¹, Nakazawa Tomoyuki¹

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

* Nukui Megumi, Kuki Ichiro, Kimura Shihoko, Hattori Taeka, Okazaki Shin, Kawawaki 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 Yuji, Okanishi Touru, Ohno Kouyou, Sugiura Chitose, Inoue Takehiko, Saitou Yoshiaki, Maegaki Yoshihiro, Ohno Kousaku
Division of Child Neurology, Institute for Neurological Sciences, Faculty of Medicine, University of Tottori, Yonago, Japan

O-070 Study of the dissociation between CBF-SPECT and 123I iomazenil SPECT

* Kuki Ichiro¹, Kawawaki Hisashi¹, Hattori Taeka¹, Nukui Megumi¹, Kimura Shihoko¹, Okazaki Shin¹, Ishikawa Jyunichi², Togawa Masao², Shiomi Masashi³, Tomiwa Kiyotaka^{1,4}
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 Nobuaki¹, Tanaka Ryuta², Matsumura Akira³, Anno Izumi⁴, Isobe Tomonori³, Ohto Tatsuyuki², Nakayama Jyunko¹, Kinugasa Hideyo¹
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 SMDS

* Matsumoto Akiko^{1,2,3}, Miyazaki Shuji¹, Nakamura Miho², Kumagai Toshiyuki³
1) Kobato Gakuenn, 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 Katsuhiro, 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 Takushi, Kikumoto Kennichi, Ohtsuka Yoko
Department of Child Neurology, Okayama University Graduate School of Medicine, Dentistry, and Pharmaceutical Sciences

O-075 Serial changes of spike localization in childhood partial epilepsy: MEG study

* Saito Naka¹, Tohyama Jun¹, Akasaka Noriyuki¹, Sasagawa Mutsuo², Kameyama Shigeki³
1) Department of Pediatrics, Nishi-niigata-chuo-National hospital, 2) Department of psychiatry, Nishi-niigata-chuo-National hospital, 3) Department of neurosurgery, Nishi-niigata-chuo-National hospital

O-076 A analysis of childhood epilepsies with a continuous spikes and waves during slow wave sleep (CSWS)

* Fukasawa Tatsuya¹, Azuma Yoshiteru¹, Natsume Jun¹, Ikuta Taketo², Kidokoro Hiroyuki³, Okumura Akihisa⁴, Negoro Tamiko⁵
1) Department of pediatrics, Nagoya University of Medicine, Aichi, Japan, 2) Department of Pediatrics, Aichi Prefecture Medical Welfare Center of Aotiori 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.

* Wakamoto Hiroyuki¹, Fukuda Mitsumasa², Hayashi Masatoshi³

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 dysplasia on frontal lobe

* Minamitani Motoyuki^{1,2}, Hamano Shin-ichiro^{2,3}, Tanaka Manabu³, Yoshinari Satoshi^{2,3}, Higurashi Norimichi^{2,3}, Eto Yoshikatsu²

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 transporter 1 deficiency syndrome

* Azuma Yoshiteru¹, Natsume Jun¹, Fukasawa Tatsuya¹, Negoro Tamiko², Watanabe Kazuyoshi³, Yanagihara Keiko⁴

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 Kenji¹, Fujii Emiko¹, Mori Tatu¹, Miyazaki Masahito¹, Harada Masafumi², Kagami Syouji¹

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, Takahashi 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 Neuroradiological findings of 22q11.2 deletion syndrome with epilepsy.

* Mori Tatsuo^{1,2}, Mori Kenji¹, Fujii Emiko¹, Kagami Syouji¹, Harada Masafumi³

1) Department of Pediatrics, Tokushima University School of Medicine, Tokushima, Japan, 2) Takamatu Red Cross Hospital, 3) Department of Radiology, Tokushima University School of Medicine, Tokushima, Japan

O-083 The effect of theophylline on the susceptibility of hyperthermia-induced seizures in developing rats

* Fukuda Mitsumasa¹, Suzuki Yuka¹, Watanabe Syouhei¹, Morimoto Takehiko²

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, Yokoi Toshiaki, Hattori Hideji, Matsuoka Osamu, Yamano Tsunekazu

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 Tsuneishi (Department of Pediatrics and Rehabilitation, Medical and Welfare Center KIZUNA)
- 4) Kazuhiro Shimokawa (Hachiojihigashi 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-ichi^{1,6}, Tada Hiroko^{1,2}, Suzuki Motomasa³, Yamanouchi Hideo^{4,6}, Yoshikawa Hideto^{5,6}

- 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 Hiroko^{1,2}, Takanashi Junichi^{1,6}, Suzuki Motomasa³, Yamanouchi Hideo^{4,6}, Yoshikawa Hideto^{5,6}

- 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 Thermolabile phenotype of CPT II variations in acute encephalopathy.

* Kubota Masaya¹, Ozawa Hiroshi², Mizuno Youko³, Kashii Hirohumi³, Amemiya Kaoru³, Suzuki Riina³, Koide Ayaka³, Hoshino Ai⁴, Yao Dengbing⁵, Kido Hiroshi⁵

- 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

09:50 ~ 10:30 Encephalitis/Encephalopathy 2

Chair: Yoshihiro Maegaki, Masashi Mizuguchi

O-090 Clinical features of acute encephalopathy with delirious behavior

* Okumura Akihisa¹, Kidokoro Hiroyuki², Kubota Tetsuo², Tsuji Takeshi³,
Suzuki Motomasa³, Kato Toru³, Natsume Jun⁴, Hayakawa Fumio³, Watanabe Kazuyoshi⁵

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O-091 Clinical Features of Acute Encephalopathy with Bilateral Hemispheric Lesions

* Okumura Akihisa¹, Kidokoro Hiroyuki², Suzuki Motomasa³, Kubota Tetsuo²,
Saito Masako¹, Nakazawa Tomoyuki¹, Tsuji Takeshi³, Kato Toru³, Natsume Jun⁴, Hayakawa Fumio³

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O-092 Signal abnormalities of hippocampus of acute encephalopathy in childhood

* Tanaka Manabu¹, Higurashi Norimichi¹, Yoshinari Satoshi¹, Minamitani Motoyuki², Hamano Shin-ichiro¹

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 Ayako¹, Fukasawa Tatsuya²

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

* Suenaga Naoko^{1,2}, Ichiyama Takashi², Kubota Masaya³, Isumi Hiroshi⁴, Tohyama Jun⁵, Furukawa Susumu²

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 Takashi¹, Matsushige Takeshi¹, Kajimoto Madoka¹, Iyoda Kuniaki²,
Furukawa Susumu¹

1) Department of Pediatrics, Yamaguchi University Graduate School of Medicine, Ube, Japan, 2) Department of Pediatrics, Hiroshima City Hospital, Hiroshima, Japan

O-096 Early CSF biomarker for the diagnosis of acute encephalopathy

* Yamanouchi Hideo, Nakajima Daisuke, 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 Naoyuki¹, Miyata Rie², Hayashi Masaharu³, Kubota Masaya⁴,
Takanashi Jun-ichi⁵, Okumura Akihisa⁶

1) Department of Pediatrics, Tokyo Metropolitan Fuchu Medical Center for the Disabled, Tokyo, Japan, 2) Department of Pediatrics, Tokyo Kita Shakai 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, Yanasaki Etsuko, Nishimura Shigeko, Tsuogae Hisano, Fujiwara 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 Shiomi, Shinichiro Hamano

O-100 Therapeutic Hypothermia for Acute Encephalopathy in Childhood: a Pilot Study

* Kawano Go¹, Iwata Osuke², Ohbu Keizo¹, Aoki Takeshi³, Akaike Hiroto⁴,
Uematsu Mitsugu⁵, Shiomi Masashi⁶, Shimono Masayuki⁷, Senjyu Ayako⁷, Hamada Hiromi⁸, Hirano Satoru⁹,
Hirabayashi Shinichi⁹, Yamanouchi Hideo¹⁰, Rinka Hiroshi⁶,
Matsuishi Toyojiro²

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, 4) Department of Pediatrics Kawasaki Medical School Hospital, 5) Department of Pediatrics Tohoku University Hospital, 6) Osaka City General Hospital, 7) Department of Pediatrics University of Occupational and Environmental Health, 8) Nikko Memorial Hospital, 9) Nagano Children Hospital, 10) Department of Pediatrics Dokkyo Medical University

O-101 The efficacy of immunosuppressant therapy for acute encephalopathy

* Takagi Atsushi¹, Sameshima Kiyoko¹, Tsuji Megumi¹, Osaka Hitoshi¹, Iai Mizue¹,
Yamada Michiko¹, Nagafuchi Hiroyuki², 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 Mana^{1,2}, Kohagizawa Toshitaka^{1,2}, Yamauchi Yuko^{1,2}, Takahashi Kayoko^{1,2}, 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 Children Studies, Department of Children Studies, Chugokugakuen University

O-105 The assessment of joint attention in high-risk infants

* Fujii Yasushi¹, Amakata Kaori¹, Ogita Kaori¹, Hikita Toshiyuki¹, Kaneko Sono¹, Nakamoto Natsue^{1,2},
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 Gakuyou¹, Yamamoto Akio¹, Asaka Youko^{1,2}, 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, Seiai 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 Takako^{1,2}, Suehiro Yutaka^{1,2}
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

* Ishioka Yuki, Yamamoto Gyousei, Matsui 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 Masahito^{1,2}, Fujii Emiko², Mori Kenji², Hashimoto Toshiaki³, Kagami Shoji²
1) Department of Pediatrics, Miyoshi Medical Clinic, Higashikagawa, 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

* Hiraiwa Rika¹, Koeda Tatsuya^{1,2}, Ohno Kousaku³
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 Toshiaki^{2,3}, Tatsuta Youhei³, Fujii Emiko², Nishimura Mio^{2,4}, Tsuda Yoshimi^{2,3}, Mori Kenji², Miyazaki Masahito², Harada Masafumi⁵, Kagami Syoji^{1,2}
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 Ryouikuen, 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 Shima^{1,2}, 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 Fukusi Center, Shizuoka, Japan

O-119 A Study of Complicated disorder or symptom with Pervasive Developmental Disorder in usual classes

* Furusho Jyunichi^{1,8}, Matsuzaki Kumiko¹, Iwasaki Yuji², Nakayama Harumi³,

Shibata Reiko⁴, Nemoto Yoshiko⁵, Kubagawa Tetsuji⁶, Sone Mie⁷, Kato Nobumasa⁸

1) College of Literature, Department of Education, Aoyamagakuin University, Tokyo, Japan, 2) Tokyo Metropolitan Tobu Medical Center for Persons with Severe Disabilities, 3) Tokyo Metropolitan Yotsugi Medical Center for 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 Masayuki^{1,2}, 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 Kakurou¹, Kanemura Hideaki¹, Kaga Yoshimi¹, Yamashiro Dai¹, Gotou Yuusuke¹, Tando 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) Yasuhiko 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)

Day 3 Room 6 (Jupitar)

09:00 ~ 09:40 Neuroimmunology

Chair: Yukiko Hirano, Tatsuo Oya

O-127 Analysis for 13 Patients of Myeloradiculopathy.

* Shimono Masayuki, Senju Ayako, 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 Tatuo
The Department of Pediatrics, Nihon University School of Medicine, Tokyo, Japan

O-129 Clinical examination of peripheral facial nerve palsy we had experience

* Kuroiwa Yuko¹, Ishiki Humie¹, Fukushima Naoya¹, Takeda Ryoujyun¹, Nanao Kenji¹, Takahashi Takao²
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 opsoclonus-myoclonus syndrome

* Xiong Hui, Peng Jing, Zhang Yuehua
Department of Pediatrics, First Hospital, Beijing University, Beijing, China

09:40 ~ 10:40 Neonatal Imaging/Others

Chair: Satoshi Kusuda, Tetsuo Kubota

O-131 Study of periventricular leukomalacia with late-onset circulatory dysfunction of premature infants

* Kobayashi Satoru¹, Hattori Ayako¹, Andou Naoki¹, Kibe Tetsuya², Koyama Norihisa³, Yokochi Kenji²,
Togari Hajime¹
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 Sonoko¹, Nishida Tomoko¹, Namba Masanori¹, Kawada Mayumi¹,
Konishi Yukihiko¹, Okubo Kensuke¹, Kusaka Takashi², Imai Tadashi¹, Isobe Kenichi¹, Itoh Susumu¹
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, Zaitzu 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 Motomasa¹, Maruyama Koichi¹, Kumagai Toshiyuki¹, Tsuji Takeshi², Kato Toru², Hayakawa Fumio²,
Okumura Akihisa³
1) Department of Pediatric Neurology, Central Hospital, Aichi Welfare Center for Persons with Developmental Disabilities, Kasugai, Japan, 2) Department of Pediatrics, Okazaki City Hospital, Okazaki, Japan, 3) Department of Pediatrics, Juntendo University, Tokyo, Japan

O-135 A case of neonatal cerebral hemorrhagic infarction developed after 1 month without clinical symptoms

* Zaitzu Ayuko¹, Aoyagi Yoko¹, Kinoshita Masahiro¹, Ishido Yuki¹, Mukae Tokutaro¹, Okada Jyunichiro¹,
Hirose Akiko¹, Kanda Hiroshi¹, Fujino Hiroshi¹, Maeno Yasuki¹, Iwata Osuke¹, Matsuishi Toyojiro¹
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

* Kidokoro Hiroyuki¹, Kubota Tetsuo¹, Kato Toru³, Hayakawa Fumio³,
Suzuki Motomasa⁴, Yaruyama Koichi⁴, 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 Kyoko¹, Ito Masako², Kusuda Satoshi³, Osawa Makiko¹

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 Tetsuo¹, Oe Hideyuki¹, Kidokoro Hiroyuki¹, Suzuki Motomasa²,
Maruyama Koichi², Kato Toru³, Uemura Naoko⁴, Natsume Jun⁵, 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, Mitsubishi 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 Ryoya¹, Hirasawa Kyoko², Ito Masako³, Konishi Yukuo³, Taga Gentaro^{4,5},
Kusuda Satoshi⁶

1) Tamagawa University Brain Science Institute, Tokyo, Japan, 2) Tokyo Womens's Medical University, 3) Tokyo
Womens's Medical University, 4) The University of Tokyo, 5) CREST/JST, 6) Tokyo Womens's Medical University

O-141 A clinical study of intractable epilepsy with partial seizures from neonatal period

* Hattori Taeka¹, Kawawaki Hisashi¹, Nukui Megumi¹, Kuki Ichiroh¹, Kimura Shihoko¹, Okazaki Shin¹, Tomiwa
Kiyotaka²

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, 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, Nariai Tadashi, Maehara Taketoshi, Aoyagi Masaru, Matsushima Yoshiharu, 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.

* Nariai Tadashi, Momose Toshiya, Inaji Motoki, Mukawa Maki,
Matsushima Yoshiharu, Ohno Kikuo
Department of Neurosurgery, Tokyo Medical and Dental University, Tokyo, Japan

O-145 Long term result and indication of neuro-endoscopic surgery

* Matsusaka Yasuhiro, Sakamoto Hiroaki
Department of Pediatricneurosurgery, Osaka City General Hospital, Osaka city, Japan

O-146 Treatment strategy for infant intracranial cyst (s) with callosal agenesis.

* Ochi Satoko¹, Yoshifuzi Kazuhisa¹, Takahashi Yoshio²

1) Dept. of Neurosurgery, Hokkaido Medical Center for Child Health and Rehabilitation, 2) Tomakomai Neurosurgical Hospital, Pediatric neurosurgery

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 envelopathy

* 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

O-151 Clinical and molecular pathological analysis of congenital muscular dystrophy-20 cases

* Xiong Hui, Jing Luo, Yun Yuan

Department of Pediatrics, First Hospital, Peking University, Beijing 100034, China

O-152 Merosin-negative congenital muscular dystrophy; Sequential study of MRI and electrophysiology

* Fujii Yuji¹, Sugiura Titose¹, Fukuda Chisako², Maegaki Yoshihiro¹, Ohno Kousaku¹

1) Division of Child Neurology, Institute of Neurological Sciences, Faculty of Medicine, Tottori University

O-153 Fukuyama congenital muscular dystrophy :Rhabdomyolysis induced by Viral Infection

* Murakami Terumi, Ishigaki Keiko, Sato Takatoshi, Osawa Makiko

Department of Pediatrics, Tokyo Women's Medical University

O-154 A Windows program ""RISCALW"" for risk calculation in carrier with Duchenne muscular dystrophy

* Ishikawa Yukitoshi, Ishikawa Yuka

Department of Pediatrics, Yakumo National Hospital, National Hospital Organization, Yakumo, Japan

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

* Okizuka Yo, Awano Hiroyuki, Yagi Mariko, Takehima Yasuhiro, Matsuo Masafumi

Department of Pediatrics, Kobe University Graduate School of Medicine, Kobe, Japan.

O-156 MLPA analysis of dystrophin gene

* Minami Narihiro¹, Nishino Ichizo²

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 Mariko¹, Awano Hiroyuki², Okizuka Yo², Takeshima Yasuhiro², Matsuo Masafumi²

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 Yayoi^{1,2}, Aoki Ryoko¹, Kondo Eri^{1,3}, 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 Toshinao^{1,2}, Ohashi Toya^{1,2}, Ida Hiroyuki^{1,2}, Eto Yoshikatsu^{1,2}, Osawa Makiko³

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, Oosawa Makiko², Okuyama Torayuki^{1,2}

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 School 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. Lake³, J. Mayhew⁷, A. Pestronk⁵, B. Rosenbloom⁸, A. Skrinar³, M. Wasserstein⁹

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, 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 Neuromuscular 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 Mitsugu², Fukuyo Naomi²,
Watanabe Syu-ei¹, 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 from patient with SMA Type1.

* Gunadi¹, Matsuo Masafumi², Nishio Hisahide^{1,2}

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 Grand 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

Day 3 Poster room (Orion)

14:30 ~ 15:15 Encephalitis 1

Chair: Kitami Hayashi, Atsushi Imamura

P-163 Onset related factors of acute encephalopathy with febrile convulsive status epileptics

* Inoue Takahito, Kakura Hiroya, Ideguchi Hiroshi, Fujita Takako, Ihara 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 Epileptics

* Takayanagi Masaru¹, Kitamura Tarou¹, Yamamoto Katsuya²

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, Musashi 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 Fujiyo³, 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 Teruhiko¹, 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 Yuichi³

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 Hirohiko³, Takahashi Takao²

1) Department of Pediatrics, Isehara kyodo hospital, Isehara, Japan, 2) Department of pediatrics, 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 Rie³, Tanuma Naoyuki³, Hayashi Masaharu³, Takahashi Yukitoshi⁴

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 Jyunko¹, 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 Yasunari²

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 Norimichi^{1,2}, Hamano Shin-ichiro¹, Yoshinari Satoshi¹, Tanaka Manabu¹, Minamitani Motoyuki³, 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 Satoru², Hattori Eiji², Takaura Natsuko¹, Imamura Takuji¹, 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 Keita¹, Tanabe Takuya¹, Shimakawa Syuichi², 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: Ryutaro Kira, Seijiro Aso

P-180 A case of Herpes simplex virus encephalitis with normal brain MRI

* Tomizawa Naoko, Shinohara Yuki, Suzuki Keiko, Ueda Satoshi, Umezaki 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 Ryutaro, Torisu Hiroyuki, 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 Megumu²

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 Takeshi^{1,2}, Suzuki Teruhiko², Endoh Yusaku², Hirano Kouichi²,

Takahashi Yukitoshi³, Ohzeki Takehiko²

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

* Nishioka Momoko¹, Honda Ryouko¹, Imamura Yoshihiko¹, Turu Akira², 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 Osamu², Ichiyama Takashi³, 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, Fujino Osamu¹, Kuwahara Kentarou², Takagi Atsushi², Hatori Takayuki², Fujimatsu Mariko³, Kawakami Yasuhiko⁴, Fujita Takehisa³, Takaishi Yasuko², Okada Kazuyoshi²

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: Yukihiro 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**
* Takemoto Megumi, Gondo Kenjiro, Hanai Toshio
Division of pediatric neurology, Fukuoka children's hospital medical center, Fukuoka, Japan
- P-190 Ictal EEG in a patient with acute disseminated encephalomyelitis with seizures.**
* Isoda Kenichi, Yokoi Kentarou, Matsui Fumihiko, 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 Mari^{1,2}, Maruyama Shinsuke^{1,2}, Toyoshima Mitsuo², 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 Cortical dominant T2 high intensity in a case of acute disseminated encephalomyelitis**
* Kitamura Taro¹, Takayanagi Masaru¹, Yamamoto Katsuya², Ishii Kiyoshi³
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 Hisamitsu
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 HHE 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 Yuko^{1,2}, Kurihara Mana^{1,2}, Kohagizawa Toshitaka^{1,2}, Takahashi Kayoko^{1,2}, 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 Yoko¹, Kashii Hirofumi¹, Amemiya Kaoru¹, Suzuki Riina¹, Kubota Masaya¹, Ichiyama Takashi², Tanuma Naoyuki³, Miyata Rie³, Hayashi Masaharu³
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 Hiroaki

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

* Orii Koji^{1,2}, 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 Mamoru³, Maniwa Satoshi⁴, Mimaki Yoshinobu⁵, Miyake Susumu⁶, 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) Matsuyama Red Cross Hospital, Pediatrics, Matsuyama, 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 Tatsuyuki^{1,2}, Takano Tomoyuki¹, Takeuchi Yoshihiro¹, Kumode Masao², 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 Shinichi², Kurahashi Hirokazu^{2,3}

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 Ikuo, 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 Jirou, Shigematsu Hideo, Otani Sanae, Takahashi Hiroka, Mine Jyun, Takahashi Yukitoshi, 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 Takanori, 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 Keita², 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 Miki^{1,2}, Maeda Tomoki², 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 Yoshihiro¹, Ohno Kousaku¹, Ogura Kaeko²

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 Masaharu^{1,2}

1) Department of Pediatrics, Fukuoka Tokusuyukai 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 Shuuichi², Nagaharu Sachiko¹, Misawa Yuka¹, Sekiguchi Yukio³,

Inaba Yuuji¹, Higuchi Tukasa¹, Koike Kenichi^{1,4}, Awaya Yutaka⁴

1) Pediatrics, University of Shinshu, Matsumoto, Japan, 2) National hospital organization chushinmatsumoto 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 Chiako, Suganami Yusuke, Oana Shingo, Watanabe Kiyoko, Watanabe Tishiaki, 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, Okizuka 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 Mariko, 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 Ayako^{1,2}, Kumada Satoko¹, Hoshino Ai¹, Tomita Sunao¹, Hanafusa Yukiko¹, Kurihara Eiji¹, Shimizu Hiroyuki³

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 Kazuhiro^{1,2}, Uematsu Mitsugu¹, Tsuchiya 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
Yodogawa Christian 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 Atsuo^{1,2}, Uematsu Mitsugu¹, Kobayashi Tomoko¹, Matsumoto Yoko¹,
Wakusawa Keisuke¹, Nakayama Tojo¹, Fukuyo Naomi¹, Haginoya Kazuhiro³,
Tsuchiya 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, Kawawaki 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 lidocaine 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 Kaeko, Fujii Toshikatsu, Hosokai Yoshiyuki, Abe Nobuhito, Shinohara Mayumi, Mori Etsuro
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, Yotsumata Kazuyuki, Kawano Yoshifumi

Department of Pediatrics, Graduate School of Medical and Dental Sciences, Kagoshima University, Kagoshima city, Japan

- P-242 Magnetoencephalography 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 Huang^{1,2}, 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**
 * Ikeno Mitsuru, 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, Tsuji Masahiro¹, 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 Yoichi^{1,2}
 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 Development, Tokyo, Japan
- P-250 Clinical study of the 6 cases of ulegyria patients.**
 * Tanabe Ryo¹, Fujii Katsunori¹, Endo Mamiko¹, Maemoto Tatsuo², Uchikawa Hideki¹, Anzai Satoshi¹, Yoshihashi Manabu¹, Kohno Yoichi¹
 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 Leucoencephalopathy syndrome in children; clinical and neuroradiological findings.**
 * Koichihara Reiko¹, Hamano Shinichirou²
 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 Ryutaro¹, Washito Natsumi¹, Torisu Hiroyuki¹, Sanefuji Masafumi¹, Hasegawa Yuki², 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 Yukio²
 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) Nishiotaru 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 Ryo¹, 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 Masakazu³, Ootake Akira³, Tanabe Yuzo², 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 Ikuko^{1,2}, Wakusawa Keisuke¹, Kakisaka Yousuke¹, Haginoya Kazuhiro³
 1) Department of Pediatrics, Tohoku University School of Medicine, Sendai, Japan, 2) Department of Pediatrics, Kesenuma City Hospital, Kesenuma, Japan, 3) Division of Pediatric Neurology, Takuto Rehabilitation Center for Disabled Children, Snedai, Japan
- P-260 No symptom without pain in a case of acute sensory neuropathy associated with anti-GM1 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 polyneuropathy**
 * Inoue Takehiko¹, Saito Yoshiaki¹, Maegaki Yoshihiro¹, Ohno Kousaku¹, 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 Kumi³, Kitai Yukihiko¹, Araya Ken¹, Tominaga Koji¹, Shimono Kuriko¹, Okinaga Tsuyoshi¹, Sakai Norio¹, Taniike Masako³, Ozono Keiichi¹
 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, Amamiya Satoshi², Takahashi Satoru¹, Suzuki Nao¹, Araki Akiko¹, Ohinata Junko¹, 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 Katsuyuki^{1,2}
 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 Kazuhiro^{1,2}, 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 Morimitsu², 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 Keiichi, 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, Enndo Ayumi, Arakawa Chikako, Fujita Yukihiko, Fuchigami Tatsuo
 Nihon University School of Medicine, Tokyo, Japan

- P-273 A boy with spinocerebellar ataxia type2 who became symptomatic at 4 years of age**
 * Kishi Kazuko¹, Shibata Naoaki¹, Sejima Hitoshi¹, Yamaguchi Seiji¹, Eda Isematu²
 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**
 * Yoshida Naoko¹, Hirota Haruyo¹, Kanda Toyoko¹, Yamori Yuriko¹, Yoshida Shoko², 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, Yoshioka Hiroshi³, Ho Yuan-yuan⁴,
 Law Peggy Py⁴, 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) Yoshioka Children's Clinic, Kyoto, Japan, 4) Department of Biochemistry, The Chinese University of Hong Kong, Hong Kong, China, 5) Colleen GIBLIN Laboratories for Pediatric Neurology 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, Sekii Katuyuki¹, Ohzeki Takehiko², 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 Yuichi³
 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 Mucopolipidosis type III with characteristic pathological findings**
 * Kobayashi Hiroshi^{1,3}, Fujigasaki Junko², Fukuda Takahiro², Sakurai Ken^{1,3}, Ida Hiroyuki^{1,3}, Ohashi Toya^{1,3}, Eto Yoshikatsu^{1,3}
 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 Shigeyuki^{1,2}, Sawada Akiko¹, Yoshida Yumi¹, Awaya Tomonari^{1,2}, Okada Masako^{1,2}, Ikeda Hiroko³, Tomiwa Kiyotaka^{1,2}
 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-282 Handness in children and their parental wish

* Shiotani Yuka¹, Sawada Akiko¹, Matuzawa Shigeyuki^{1,2}, Yoshida Yumi¹,
Awaya Tomnari^{1,2}, Okada Masako^{1,2}, Ikeda Hiroko³, Tomiwa Kiyotaka^{1,2}

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:lateralization and development of sociability

* Matsuzawa Shigeyuki^{1,2}, Shiotani Yuka¹, Sawada Akiko¹, Yoshida Yumi¹,
Awaya Tomonari^{1,2}, Okada Masako^{1,2}, Ikeda Yuko³, Tomiwa Kiyotaka^{1,2}

1) JST/RISTEX, 2) Kyoto university graduate school of medicine, 3) Shizuoka 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 Shigeyuki^{1,2}, Shiotani Yuka¹, Yoshida Yumi¹,
Awaya Tomonori^{1,2}, Okada Masako¹, Ikeda Hiroko³, Tomiwa Kiyotaka^{1,2}

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 Ayumi^{1,2}, Takeuchi Ariko^{1,2}, Koeda Tatsuya^{1,2}

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 Chiyomi^{1,2,3}, Morita Kiichiro³, Ishii Youhei³, Yamashita Yushiro²,
Matuishi Toyojiro²

1) Technical school medical and welfare Ryokuseikan, 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, Sasaki 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 Atsushi^{1,2}, 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, Graduate 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 Lu², Kai-hsin Lin², Dah-chin Yan³, Tzou-yien Lin⁴

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 Nagahisa¹, Yoneyama Akira¹, Kitazumi Eiji¹

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

* Takahashi Nagahisa, 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 Akie, Fukuda Ikue, Oka Ryuji, Cho Kazuhiko

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 Higashiyamoto 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 Toshisaburo^{2,6}, Funato Masahisa^{3,6}, Tagawa Tetsuzo^{4,6}, Negishi Hirokuni^{5,6}

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 Toshisaburo^{1,7}, Ikeda Tomomi^{1,2}, Okuno Hiroko¹, Sugiura Keiko¹, Takama Satomi¹, Tagawa Tetsuzo^{3,7}, Negishi Hirokuni^{4,7}, Hattori Hideji^{5,7}, Funato Masahisa^{6,7}

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 cooperation and medical care in Ibaraki

* Ohtoshi Taro¹, 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 Yasuyuki¹, Kumode Masao¹, 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 Ryoko^{1,2}, 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 Yuhko^{1,2}, Nakane Takaya¹, Hatakeyama Kazuo², Nakamura Kousuke¹,
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 Tamako, 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 Tamako, 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 Fukushi 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 Takashi², Taira Takaomi², 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, Ihara Yukiko, Tomonou Yuuko, Hujita Takako, Nakamura Noriko, Inoue Takahito, Yasumoto Sawa, Hirose Shiniti
1) Department of Pediatrics, University of Hukuoka, Hukuoka, Japan
- P-315 Percutaneous endoscopic gastrostomy in neurologically disabled children improves quality of life.**
* Takeshita Saoko¹, Hirasawa Kingo², Yahara Sei², 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, Suenaga 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 Keiko^{1,2}, Hasumi Hiroki¹, Nakatani Katutosi¹, Shinozaki Yuuko³, Anzai Yuki⁴, 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 development
- P-318 Prevention from aspiration pneumonia with botulinum toxin type A injection to salivary glands**
* Soebijanto Keiji, Sasazuki Momoko, Sakamoto Kei, Aibe Miyuki, Mizuno Yuji
Department of pediatrics, East Fukuoka Medical Center, Fukuoka, Japan
- P-319 Treatment of zinc supplementation for severe handicapped people**
* Wada Keiko, Tyou Hiroyuki, Funahashi Masuko, Suzuki Yasuyuki
Tokyo childrens rehabilitation hospital, Tokyo, Japan

Day 4 Room 1 (Pegasus A)

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 Yoko^{1,2}, Matsui Gakuyou², 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 Yasunori^{1,2}, Suzuki Shuhei²

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 cavernous hemangioma in brainstem

* Shimono Kuriko¹, Kato Kumi², Kitai Yoshihiro¹, Araya Ken¹, Tominaga Kouji¹,

Okinaga Takeshi¹, Mohri Ikuko², Taniike Masako², Ozono Keiichi¹

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 Research 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

Uji clinic; Division of Medicine, The Association of Remedial Teaching for People with Developmental Handicaps

O-175 Neuronal Antibody in neuropsychiatric disorders

* Hongou Kazuhisa, Harai Tomomi, Fujiki Yasuko, Miya Kazushi, Kageyama Ryuuji, Tanaka Chiaki,

Yagi Shinichi, Honnma Kazumasa, 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 Shuichi², 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 neurologic 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

Day 4 Room 2 (Pegasus B)

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 Takatoshi¹, Itou Yasushi^{1,2}, Etou Kaoru¹, Abe Kazuyo¹, Nishikawa Aiko¹,

Fujii Akiko¹, Imai Kaoru¹, Oguni Hirokazu¹, Osawa Makiko¹

1) Department of Pediatrics, Tokyo Women'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 Kazuhito, Miyahara Hiroaki, Shimizu Miki, Maeda Tomoki, Akiyoshi Kennosuke, Izumi Tatsuro
Oita university faculty of medicine, oita, 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 Tomonari¹, Osada Kazuko¹, Shibata Minoru¹, Yamanaka Yasunari¹, Kato Takeo¹,

Nakahata Tatsutoshi¹, Tomiwa Kiyotaka²

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

Pediatirics, Saku Central Hospital, Nagano, Japan

O-184 Effectiveness of gabapentin with intractable partial seizures in childhood and adolescence

* Iwasaki Toshiyuki¹, Nonoda Yutaka¹, Takei Kenji^{1,2}, Hosoda Nozomi^{1,2}, Ishii Masahiro¹

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 Shin¹, Kawawaki Hisashi¹, Hattori Taeka¹, Nukui Megumi¹, Kuki Ichirou¹,

Kimura Shihoko¹, Ishikawa Junichi², Togawa Masao², Shiomi Masashi³,

Tomiwa Kiyotaka^{1,4}

1) Department of pediatric neurology, Children's Medical Center, Osaka City General Hospital, 2) Department of pediatric emergencymedicine, Children's Medical Center, Osaka City General Hospital, 3) Infection Center, Osaka City General Hospital, 4) Geneticcounselor coordinator Unit, Kyoto Univercity

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 Miyako², Oguni Hirokazu², Osawa Makiko², Ito Yasushi², Ito Susumu²

1) Yamawaki Gakuenn 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?

Day 4 Room 3 (Pegasus C)

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, Ryouichi Sakuta

O-189 The development of visual function in healthy and high risk infants using the eye tracking system

* Yamaguchi Fumika¹, Hirasawa Kyoko², Tamura Masanori¹

1) Saitama Medical University, Saitama Medical Center, Kawagoe, Japan, 2) Tokyo Women's Medical University, Tokyo, Japan

O-190 Disphagia 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 Kayo¹, Yanagihara Keiko^{1,2}, Arai Hiroshi^{1,3}

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 Mariko¹, Niiijima Shiniti¹, Iijima Yoshitaka¹, Iwasaki Tomohiro¹, Kamata Ayako¹, Tahara Kanako¹, Suzuki Ryuyo¹, Watanabe Naoki¹, Ootomo Yoshiyuki¹, Okumura Akihisa², Hayashi Masaharu³

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 Noboru¹, Fukuyama Tetsuhiro², Ishida Shuichi³, Hirabayashi Shinichi⁴,

Hirano Satoru⁴

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, Chushinmatsumoto 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**
* Fujioka Hiroki¹, Shintaku Haruo¹, Hirabayashi Shinichi², Yamano Tsunekazu¹
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 Haruo⁴, Fujioka Hiroki⁴
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 Hiroshi¹, 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

International Symposium Celebrating the 50th Meeting of the JSCN part I : Recent advances in child neurology

Big brains & small brains — genetic and epigenetic mechanisms of brain size alteration —

Takao Takahashi

Pediatrics, Keio University School of Medicine, Tokyo, Japan

First in this talk, the workings of neurogenesis, 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 neurogenesis, 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 neurogenesis. There is a tight correlation between cell cycle of origin and layer destination of newly-born neurons. The most critical parameter of the neurogenetic process is probability of cell cycle exit of NPCs (quiescent fraction or Q). Q is precisely regulated during the course of neurogenesis and gradually increases from a value of 0 to 1.0, as neurogenesis 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 neurogenesis 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 neurogenesis 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 neurogenesis, 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 neurogenesis.

EDUCATION

1976-1978 B.S. Keio University Undergraduate Course

1978-1982 M.D. Keio University School of Medicine

SPECIALIZED FIELD

General Pediatrics, Pediatric Neurology, Developmental Neurobiology

ACADEMIC APPOINTMENTS:

2002-present Professor and Chairman, Department of Pediatrics, Keio University School of Medicine

1999-2002 Associate Professor, Department of Pediatrics, Keio University School of Medicine

1996-1999 Assistant Professor, Department of Pediatrics, Keio University School of Medicine

HOSPITAL APPOINTMENTS:

1994-1996 Senior Clinical Fellow, Department of Pediatrics, Keio University School of Medicine

1994-present Assistant in Neurology, Massachusetts General Hospital, Harvard Medical School

1992-present Instructor in Neurology, Harvard Medical School

1988-1994 Research Fellow, Developmental Neurobiology, Massachusetts General Hospital, Harvard Medical School

1984-1988 Clinical Fellow, Department of Pediatrics, Keio University School of Medicine

1982-1984 Resident, Department of Pediatrics, Keio University School of Medicine

HONORS AND AWARDS

1992 Charles A. King Trust Medical Foundation. Young Investigator Award

1995 Keio Medical Award, Cell cycle kinetics in mouse neocortical development

1996 The Senator Jacob Javits Award of Excellence (Co-investigator at Harvard Medical School)

MEMBERSHIPS

Japanese Pediatric Society

Japanese Society of Child Neurology

Japanese Epilepsy Society

Society for Neuroscience (USA)

Child Neurology Society (USA)

International Child Neurology Society

EDITORIAL BOARDS:

2003- Managing Editor, Brain and Development

International Symposium Celebrating the 50th Meeting of the JSCN part I : Recent advances in child neurology

Recent advances on neurocutaneous syndromes

Paolo Curatolo

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

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 hamarthomas in many sites of the body. Tuberous Sclerosis Complex is the prototype of multisystemic involvement with hamarthomas 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 hamarthritis 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

- 1) Curatolo P., Riva D., Eds (2006). Neurocutaneous Syndromes in Children, John Libbey Eurotext, Paris
- 2) Sarnat H.B., Curatolo P., Eds (2008). Malformations of the nervous system, Elsevier, Amsterdam
- 3) Curatolo P., Ed (2003). Tuberous Sclerosis Complex: from

basic science to clinical phenotypes, Mac Keith Press, London

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)

May 28th, Wed.

International Symposium Celebrating the 50th Meeting of the JSCN part I : Recent advances in child neurology

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.

Djerine-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

NATIONALITY: Australian

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

Chevalier de l'Ordre de la Légion d'Honneur, Republic of France, 2001.

Chevalier de l'Ordre National du Mérite, Republic of France, 1996.

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.

1972: Staff Physician in Neurology, Royal Alexandra Hospital for Children, Sydney.

1978- 1998: Head, T Y Nelson Department of Neurology and Neurosurgery, Royal Alexandra (New Children's) Hospital for Children, Sydney.

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.

International Symposium Celebrating the 50th Meeting of the JSCN part I : Recent advances in child neurology

Japanese encephalitis in Korea and Asian countries — Can it be under control? —

Yong-Seung Hwang

Seoul National University Children's Hospital, Seoul, Korea

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 tritaeniorhynchus* 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.

CURRICULUM VITAE

NAME IN FULL : Yong-Seung Hwang

DATE OF BIRTH : October 27, 1950

PRESENT ADDRESS : 31-601, Hyundai Apt. Abgujeong Dong, Gangnam Gu, Seoul, Korea 135-110

EDUCATIONAL BACKGROUND

Seoul National University M.D. 1971-1975

College of Medicine

Postgraduate Course Ph. D. 1978-1983

Seoul National University

Majoring in Medicine, Pediatrics

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, U S A

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

May 28th, Wed.

International Symposium Celebrating the 50th Meeting of the JSCN part I : Recent advances in child neurology

The impact of infections of the central nervous system on African children

Charles RJC Newton

Kenya Medical Research Institute, Kilifi, Kenya and University College London, London, United Kingdom.

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.

International Symposium Celebrating the 50th Meeting of the JSCN part I : Recent advances in child neurology

Gene therapy for muscular dystrophy

Shinichi Takeda

Department of Molecular Therapy, National Institute of Neuroscience, National Center of Neurology and Psychiatry, Kodaira, Japan

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, CXMD_J (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* Δ CS1) gene into skeletal muscle of CXMD_J and confirmed the expression of micro-dystrophin. The injection of recombinant AAV8 encoding Δ CS1 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 CXMD_J 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 — cybournoid, biorobotics, control for supprting disabled persons —

Yoshiyuki Sankai

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

Major Research Projects;

- 1) Areificial 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 Talatsu. 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.

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

Japanese contributions to child neurology — an international perspective —

Robert A Ouvrier

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

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 Keiya 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.

May 29th, Thu.

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-Seung 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 congress 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.

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135-110

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, U S A

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

IL 1

Congenital muscular dystrophy

Francesco Muntoni

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

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 neuexin. Mutations in six genes (POMT1, POMT2, POMGNT1, fukutin, FKR1 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

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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

IL 2

**Convulsing our way toward the pathophysiology of autism
— clinical models and lessons for treatment —**

Roberto Tuchman

Department of Neurology, Miami Children's Hospital Dan Marino Center

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 prognostic 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

Brain monitoring made easy

Anita Kharbteng

Clinical Support Manager, VIASYS Healthcare

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

May 30th, Fri.

MS 2

EEG — an introduction —

Solomon L Moshé

Albert Einstein College of Medicine and Montefiore Medical Center

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. 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. Moshé 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. Moshé 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.

**International Symposium Celebrating the
50th Meeting of the JSCN Part III**

May 30th, Fri.

Room 2

May 30th, Fri.

International Symposium Celebrating the 50th Meeting of the JSCN part III: Fukutinopathy (Didactic Lecture 3)

Fukuyama congenital muscular dystrophy — an overview —

Yukio Fukuyama

Emeritus Professor, Tokyo Women's Medical University, Tokyo, Japan

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 / 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 mimick 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 / 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.

International Symposium Celebrating the 50th Meeting of the JSCN Part III : Phenotypic Spectrum of Fukutinopathy

Most severe phenotype of Fukutinopathy

Mieko Yoshioka

Department of Pediatric Neurology, Kobe City Pediatric and General Rehabilitation Center for the Challenged, Kobe, Japan

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 sibships 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

May 30th, Fri.

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

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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

Terumi Murakami¹, Yukiko Hayashi², Ikuya Nonaka², Makiko Osawa¹, Ichiro Nishino²

Department of Pediatrics, Tokyo Women's Medical University, Tokyo, Japan¹

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A group of disorders due to altered glycosylation of α -dystroglycan (α -DG), namely, α -dystroglycanopathy (α -DGP), 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 -DGP in Japan, and the patients carry the founder mutation of 3kb retrotransposal 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 can speak 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 unrelated 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

May 30th, Fri.

International Symposium Celebrating the 50th Meeting of the JSCN Part III : Phenotypic Spectrum of Fukutinopathy

Development of Fukutinopathy model mouse

Tatsushi Toda

Division of Clinical Genetics, Osaka University Graduate School of Medicine, Suita, Japan

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 retrotransposal 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 30th, Fri.

Room 2

LS 5

Treatment 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

International Symposium Celebrating the 50th Meeting of the JSCN Part IV: Topics in neuromuscular disorders

Recent advance in spinal muscular atrophy

Yuh-Jyh Jong

Graduate Institute of Medicine, College of Medicine, Kaohsiung Medical University, Kaohsiung, Taiwan

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), suberoyl anilide 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.

International Symposium Celebrating the 50th Meeting of the JSCN Part IV: Topics in neuromuscular disorders

New insights into the pathogenesis of SMA

Yasushi Ito

Department of Pediatrics, Tokyo Women's Medical University, School of Medicine, Tokyo, Japan

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 ballooned 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

International Symposium Celebrating the 50th Meeting of the JSCN Part IV: Topics in neuromuscular disorders

Congenital myasthenic syndrome

Keiko Ishigaki

Department of Pediatrics, Tokyo Women's Medical University, School of Medicine, Tokyo, Japan

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 Ultrastructural 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

Research associate (Post doctoral stay), INSERM 523, Institut de Myologie, Groupe Hospitalier Pitie-Salpetriere, Universite Paris VI, Paris, France, 2000-2001

Assistant Professor: Department of Pediatrics, Tokyo Women's Medical University, Tokyo, Japan, 2002 to the present time

Oral presentation

May 29th, Thu.
Room 5, 6

May 30th, 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 rhinorrhoea 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 >72hours, >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.

(特別 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, sex, 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

Department of Pediatrics, Kaohsiung Medical University Hospital, Kaohsiung Medical University, Kaohsiung, Taiwan

【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 $41.18 \pm 26.73\%$ 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

Harshuti Shah, MBBS, MD (ped) I.P.T.M. (child Neuro)

Zachary Grinspan, MD

Rajvee Child Neuro and orthospine hospital¹, Department of child neurology, Columbia University, Newyork, Newyork, USA²

【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

Jao-Shwann Liang¹, Keiko Shimojima¹, Koyo Ohno², Chitose Sugiura², Yukiharu Une³, Kousaku Ohno², Toshiyuki Yamamoto¹

¹International Research and Educational Institute for Integrated Medical Sciences (IREIIMS), Tokyo Women's Medical University, Tokyo, Japan

²Division of Child Neurology, Institute of neurological Sciences, Faculty of Medicine, Tottori University, Yonago, Japan

³Une Clinic, Onomichi, Japan

【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

Xiong Hui, Peng Jing, Zhang Yuehua
Department of Pediatrics, First Hospital, Beijing University

[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

Yukiko K. Hayashi, Park Young-Eun, Ikuya Nonaka, Ichizo Nishino
Department of Neuromuscular Research, National Institute of Neuroscience, National Center of Neurology and Psychiatry Tokyo, Japan

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

Young-Eun Park, Y. K. Hayashi, I. Nakano, I. Nishino
Department of Neuromuscular Research, National Institute of Neuroscience, National Center of Neurology and Psychiatry
Tokyo, Japan

【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 subsarcolemmal 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

Xiong Hui, Luo Jing, Wang Xiaozhu, et al.
Department of Pediatrics, First Hospital, Beijing University,
Beijing 100034, China

【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 performed 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 heterozygous 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

Xiong Hui, Jing Luo, Yun Yuan, et al
Department of Pediatrics, First Hospital, Peking University University, Beijing 100034, China

[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 $\alpha 2$ (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 (rigid 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 \pm SD 6MWT distance was 327.4 \pm 128.0 meters (50.1% predicted) and mean baseline FVC \pm SD was 54.6 \pm 14.8% predicted. At last evaluation (78 weeks), estimated mean absolute differences \pm SD of 28.1 \pm 13.1 meters in 6MWT distance (p=0.03) and 3.4 \pm 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 μ M 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 μ M 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 ($\Delta 7$) transcript amounts or $\Delta 7$ /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 29th, Thu.

30th, Fri.

Room Orion

**P-007 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 Chiari 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 fasciitis 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 resembling 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 indicates that SCN1A mutation may not be responsible for the benign association 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. Seizure 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 subependymal 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.