

出國報告(出國類別：開會)

五十屆日本小兒神經年會既擴大舉行國際兒童神經
醫學研討會

服務機關：三軍總醫院小兒部

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派赴國家：日本

報告日期：97年7月21日

出國時間：97年5月27日至6月1日

壹、 摘要

2008年於5月29日至6月1日在日本東京舉行第五十屆的日本小兒神經醫學年會既既擴大舉行國際兒童神經醫學研討會。本會議雖為日本小兒神經年但今年為擴大舉辦家學者所積極參與以及相當重視的，涵概的議題舉凡兒童神經相關的疾病皆為研討的議題主要針對兒童中樞 周邊神經的各類病症的現況最新研究、治療方式及研究論文作廣泛而深入的研究與討論。本屆會議大會主席為東京女子醫科大學大澤醫師將此次研討會重點放在神經行為發展學。

本次醫學會討論除了專題演講、口頭論文報告及現場論文展示超過五百篇等多項活動逐一進行外，每個論文皆讓參加活動的學者能更進一步與各相關議題領域的教授、專家即時的討論交換意見。職等此次有幸獲國防部經費補助參與會議並受邀於大會中提出壁報論文報告。藉由參加這次會議，了解他國專家學者的研究成果及趨勢，獲益良多，且對於一個日本當地的年會，如何能擴展成國際型的會議，有其值得學習之處。

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貳、 參加目的

職等目前任職於三軍總醫院小兒部，承國防部之同意及補助出席 50th Japanese Society of Child Neurology and international symposium (第五十屆的日本小兒神經醫學年會暨擴大舉行國際兒童神經醫學研討會) 於 5 月 28 日至 6 月 1 日在日本東京舉行。此次會議延攬世界知名小兒神經科臨床醫師及的工作的相關學術研究領域的學者專家，自有其重要性。此次會議有超過五百多篇的論文報告展出，在小兒神經醫學領域的國際會議而言自有其份量與影響力。

職等於過去九年來對於兒童神經學相關議題有數個研究正持續在進行，也陸續於國內外包含中華民國小兒科醫學會年會、台灣小兒神經醫學會、及國際嬰幼兒癲癇症候群研討會等重要會議內發表口頭或壁報論文展示。今年職等將在基礎研究的自體免疫小鼠腦炎的部分研究成果投稿於本次大會，經由大會委員會遴選後，同意於會議中發表壁報論文報告，此機會實屬不易。職等於整個會期中，一方面學習學者專家有關兒童神經學的研究新知及臨床經驗外，同時發表自己的研究成果與國際人士共同研討及交流，對於職等爾後在臨床、教學及研究工作的方向亦是助益匪淺。

參、 會議過程

在為期四天的議程 整個會議包含二部分的壁報展示在內，依照每日的議程逐一進行，並記載詳細每日會議過程之重要議題如下：

第一日(五月二十八日)：

大會演講以數篇討論兒童神經學之最新進展為開場，包括有 Dr. Takahashi T 發表的調控腦部成長大小的基因與表觀基因參與的機制(genetic and epigenetic mechanisms of brain size alteration)，如近來發現腦部的成長發育中神經起始細胞的增生(neural progenitor cells)由參與細胞週期調節基因(cell cycle regulator gene) p27kip1，來控制神經之發生(neurogenesis)。而相當重要一點是，動物實驗證實暴露在戴奧辛的胚胎會甲基化 p27 而導致腦部無法正常發育，進而產生發育不良的腦，相當讓人震撼的結果。由 Montefiore/Einstein Epilepsy Management Center, Albert Einstein of Medicine and Montofiore Medical Center 的 Dr. Solomo L moshe 演講從生物學角度探討兒童癲癇重積症的新進展(progress in the biological aspects of status epilepticus)genetic basis of our understanding of epileptic syndromes)，關於兒童癲癇重積症對發展中的腦產生的傷害有著 精譬的演說。此外我國還有韓國的小兒神經醫學會理事長，及前任與現任世界小兒神經醫學會理事長，皆有專題演講，關於兒童神經學之最新進展作。

第二日議程(五月二十九日)：

包括有邁阿密兒童醫院的 Dr. Roberto Yuchmanu 演講兒童自閉症病理生理學與治療，與來自英國 Dubowitz Neuromuscular Centre, UCL Institute of Child Health, London 的 Dr. Francesco Muntoni 深入探討了先天性肌肉肌肉失養症的新進展(congenital muscular dystrophy)包括各種細胞外物質(extracellular matrix)蛋白質的基因，如 POMT1, POMT2, POMGNT1, fukutin, FKRP 與 LARGE 與相關疾病的探討， 學到相當多的新知，感到科學進步一日千里。神經免疫學的探討 (advanced neuroimmunology)也被安排此日，如近年熱門的 aquaporin-4 之抗體在眼脊型多發性硬化症(opticospinal multiple sclerosis)的發現之探討，有很多篇的口頭報告與壁報展示；此還有日本各大醫學中心探討學習障礙的研究與治療的新領域(therapeutic innovations in learning disabilities) 與腦性麻痺之復健新進展(cerebral palsy rehabilitation)，讓人相當感興趣的是基因學

與發展問題的探討 (developmental disorder & genetics) 提到了幾個重要發現如 CDKL5 mutation causing an atypical form of Rett syndrome, a clinical study on autism with SHANK3 abnormalities, molecular mechanism of neuron-specific imprinting of Angelman syndrome gene Ube3a 等，都是相當新穎的熱門議題。關於壁報展示，職被安排本日下午，須口頭以英文講述三分鐘令外二分分鐘討論，過程精彩討論熱烈，此外許多國際學者也給予相當多寶貴意見，職亦參觀其他人的壁報展示，也學到相當多的新知。

第三日議程(五月三十日):

包括前任世界小兒神經醫學會理事長，演講治療頑固型癲癇的新進展(treatment for intractable epilepsy)，高雄醫科大學副校長鐘育志教授演講脊髓肌肉萎縮症的新進展 (recent advance in spinal muscular muscular atrophy)，以及東京女子醫科大學 Yasushi Ito y 醫師演講脊髓肌肉萎縮症致病機制的新面觀 (new insights into the pathogenesis of SMA)，此外還有幾個有趣且有意義的議題包括兒童周邊神經肌肉病變(advanced in childhood peripheral neuropathies and myopathies) 與新生兒神經學的熱門課題 包括新生兒腦傷之影像學新進展 (image update in perinatal brain damage) 新生兒腦的保護(topics of the brain protection in neonate)與新生兒醫學對新生兒腦部發展的影響 (impact of recent neonatal medicines on the neurodevelopmental outcomes in human neonates)等，癲癇手術與神經代謝退化疾病也有許多探討其中日本順天堂大學兒科教授 Shinichi Niijima 演講 生長激素對腦代謝的影響 (effect of growth hormone on the brain) 相當新穎。

第四日議程(五月三十一日):

主要為教育演講有外觀異常的臨床疾病(a clinical approach to the dysmorphic child) 腦細胞元移行的分子調節機制 (regulatory mechanism of neuronal migration mediated by the microtubule-associated protein doublecortin and its partners)，以及最近熱門的睡眠與行為障礙 (sleep/behavior disorders)，此外還有影片教學在分析不自主動作與癲癇症的區別，與兒童失張力症診斷與治療演講。

肆.會議心得：

本次醫學會討論除了專題演講、口頭論文報告及現場論文展示超過五百篇等多項活動逐一進行外，每個論文皆讓參加活動的學者能更進一步與各相關議題領域的教授、專家即時的討論交換意見。藉由參加這次會議，且對於一個日本當地的年會，如何能擴展成國際型的會議，有其值得學習之處，獲益良多。

調控腦部成長大小的基因與表觀基因參與的機制(genetic and epigenetic mechanisms of brain size alteration)，國內目前並無兒科領域醫師從事相關研究，但本院婦科癌症研究在此方面已有成果，相信可藉由科部研究交流可以有好的前景。兒童自閉症病理生理學與治療也越來越受到重視，在本次會議再次證實也讓我們對兒心部門更有份使命感。產生先天性肌肉肌肉失養症的原因(congenital muscular dystrophy)包括各種細胞外物質(extracellular matrix)蛋白質的基因，如 POMT1, POMT2, POMGNT1, fukutin, FKRK 與 LARGE 等，也越來越清楚 學到相當多的新知，感到科學進步一日千里。神經免疫學的探討 (advanced neuroimmunology)也被安排此日，如近年熱門的 aquaporin-4 之抗體在眼脊型多發性硬化症(opticospinal multiple sclerosis)的發現之探討，由於 aquaporin-4 此蛋白由日本學者發現，在基礎與臨床結合下，發表很多國際知名雜誌包括自然醫學 (nature medicine) 值得借鏡。

此外大會議程安排與會場便利與舒適，我們在觀摩了一場國際會議，也感受到辦此會議所耗的人力物力是相當龐大，但相對獲得的國際肯定與周邊效益是無法估計。我們可以藉此衡量主客觀因素，積極爭取各種國內外際學術會議的機會，讓三軍總醫院邁向中長程目標前進。

伍. 回單位後報告情形

本次會議職等的研究論文獲大會肯定，並選為第四部份議程的口頭報告討論題目，不但可與專家學者做直接討論外，更代表本部研究已受到國際的肯定，對個人而言更是難得的經驗。已將相關資訊(如附件一)帶回並於單位報告，同仁皆感到興奮。

陸. 建議事項：

國際研討會議所提供的新知為全面性、前瞻性及整合性的，且有機會與研究之學者專家當面交流對談，經由互相討論後，許多新的構想更容易被創造出來。

本次參與國際會議，經由專題演講、專題研討會及論文海報展示等各方面議程，與來自全球各地的學者專家互相討論吸取新知後，對於自己往後的研究工有相當大的助益。同時藉由會議發表自己的論文，促進國際交流，提昇國際地位。

在此次大會中，也讓我們知道自己部份的優勢與很多的不足，對此我們更了解國內相同領域的學者應凝聚共識，加強彼此合作與互動的關係，經由參加國際性學術會議可拓展視野並跟上國際最新進展，讓我們更具競爭力並提昇國內研究水準及國際學術地位，實為值得鼓勵。

此外建議由於補助有限，讓人對參加國際性學術會議意願減低，這是值得思考的事宜。

柒. 參加此會議對單位之貢獻

職等近年來對於兒童神經相關的研究成果已逐漸完成並將陸續發表，此會議除了聽取各國專家意見外，從壁報展示及部份口頭報告討論中，了解職研究已初步獲國際的肯定。職等於會議中獲得許多新的方向以及未來本部可持續進行之研究方向。例如神經表觀基因對嬰幼兒之影響，可以與婦產部賴鴻政 醫師做廣泛的合作並進行相關研究。對於本院研究的整合與能力的提昇，有相當大的幫助。

捌. 附件資料

附件一：回單位後報告之內容

附件二：出國參加會議日程表及議程表


參加第五十屆日本小兒神經醫學會年會暨擴大國際會議 心得報告

報告人 陳錫洲
2008-7-3

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

President: **Makiko Osawa, M.D.**
Professor, Department of Pediatrics, Tokyo Women's Medical University
Date: **May 22(Wed)-31(Sat), 2008**
Venue: **Hotel Nikko Tokyo**

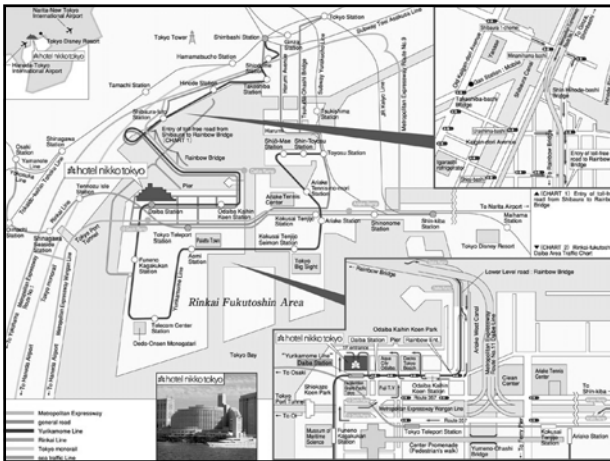
Special Lecturers: **Takeshi Yoro, M.D.**
Makoto Inoue, M.D.



International Symposium Celebrating the 50th JSCN

Invited Speakers:

- Paolo Curatolo (Italy)
- Yong-Seung Hwang (Korea)
- Yuh-Jyh Jong (Taiwan)
- Solomon L. Moshé (U.S.A.)
- Francesco Mantua (U.K.)
- Charles RJC Newton (Kenya + U.K.)
- Robert Ouvrier (Australia)
- Roberto Tuchman (Miami, U.S.A.), etc



May 28 (Wednesday) International Symposium Celebrating the 50th JSCN Part I (13:30-18:00)

- Recent Advances in Child Neurology 1 (13:30-15:00)**
 - Big brains & small brains: genetic and epigenetic mechanisms of brain size alteration (13:30-14:00)
Takao Takahashi (Department of Pediatrics, Keio University School of Medicine, Japan)
 - Recent advance in neurocutaneous syndromes. (tentative) (14:00-14:30)
Paolo Curatolo (Previous President, ICNA, Pediatric Neurosciences, Tor Vergata University of Rome, Italy)
 - Age-related presentations of hereditary peripheral neuropathies in childhood (14:30-15:00)
Robert Ouvrier (President of ICNA - University of Sydney; Division of Neurology, Children's Hospital at Westmead, Australia)
- Recent Advances in Child Neurology 2 (15:10-16:40)**
 - Japanese encephalitis (15:10-15:40)
Yong-Seung Hwang (President of AOCNA, Department of Pediatrics, Seoul National University College of Medicine, Korea)
 - Problems of central nervous system infection in Africa (15:40-16:10)
Charles RJC Newton (Kenya/Tropical Neurosciences and Pediatrics, University College of London, UK)
 - Progress in the biological aspect of status epilepticus (16:10-16:40)
Solomon L. Moshé (Neuroscience & Pediatrics, Albert Einstein College of Medicine, USA)
- Recent advance in Child Neurology 3 (16:50-18:00)**
 - Gene therapy for muscular dystrophy (16:50-17:20)
Shinichi Takada (National Institute of Neuroscience, National Center of Neurology and Psychiatry, Japan)
 - Robot sautechthermal, hibernotonic, control for supporting disabled person (tentative) (17:20-17:55)
Yoshiyuki Sankai (Department of System & Information Engineering, University of Tsukuba, Japan)

Evening seminar I (19:00-21:00)

The second annual meeting of the Japan Association of Child Sleep, chaired by Jun Kohyama (Tokyo Kita Shikai Hoken Hospital)
Satisfactory sleep for children holds promise for the future

Epigenetic in Pediatric Neurology

- Big brains & small brains: genetic and epigenetic mechanisms of brain size alteration (13:30-14:00)
Takao Takahashi (Department of Pediatrics, Keio University School of Medicine, Japan)

Patients with **MCPH5** mutation

C. G. Walsh, P. E. Ozark, K. S. Krishnamoorthy and G. H. Muziklis
nature genetics • volume 32 • october 2002

- Humans with autosomal recessive primary microcephaly (MCPH) show a small but otherwise grossly normal cerebral cortex associated with mild to moderate mental retardation.
- The most common cause of MCPH is homozygous mutation of *ASPM*, the human ortholog of the *Drosophila melanogaster* abnormal spindle gene (*asp*), which is essential for normal mitotic spindle function in embryonic neuroblasts.
- The mapping of the *MCPH5* locus and identification of *ASPM*.

Altered Brain Size Induced by Altered Expression of p27

- p27 Knockout
- p27 Overexpression
- In utero Exposure to Dioxin

[Cell Cycle 5:20, 2314-2318, 15 October 2006]
Extra View

Coupling Cell Cycle Exit, Neuronal Differentiation and Migration in Cortical Neurogenesis

p27^{Kip1} REGULATES RADIAL MIGRATION IN THE CEREBRAL CORTEX

Single Oral Dose of Dioxin Reduces Neuron Number and Cortical Thickness

Dioxins

- ✓ Ubiquitous environmental pollutant
- ✓ Induce p27 expression
- ✓ Disturb cell proliferation
- ✓ *in utero* exposure impairs spatial learning and memory

Summary

- ✓ Neuron number: precisely controlled by cell cycle exit probability (Q).
- ✓ Change in Q: significant change in neuron number and brain size.
- ✓ Brain size constraint: embedded in decision making characteristics of neural progenitor cells.

Recent advance in neurocutaneous syndromes. (tentative) (14:00-14:30)
Paolo Curatolo (Previous President, ICNA, Pediatric Neurosciences, Tor Vergata University of Rome, Italy)

Neurocutaneous Syndromes
Types of skin lesions

- * Decreased skin pigmentation (Tuberous Sclerosis, Hypomelanosis of Ito)
- * Increased skin pigmentation (Epidermal Nevus s., Incontinentia pigmenti, Neurocutaneous Melanosis, Neurofibromatosis type I)
- * Skin tumors/Hamartomas (Neurofibromatosis type 1-2, TSC, Encephalocraniocutaneous Lipomatosis, Epidermal Nevus S)
- * Skin vascular lesions (Atassia-Telangiectasia, Sturge-Weber)
- * Hyperkeratotic/Icthyotic lesions (Sjogren-Larson)
- * Rash-like lesions (Incontinentia pigmenti)
- * Skin laxity (Ehler-Danlos, Progeria, Trichothiodystrophy)

von Hippel Lindau
Genotype-phenotype correlation

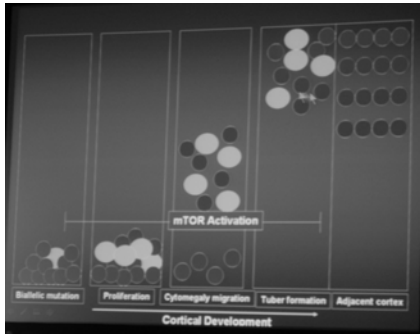
- > TYPE 1: without pheochromocytoma risk
- > TYPE 2: with risk of both hemangioblastoma and pheochromocytoma

- > Type 2 a: low risk of RCC
- > Type 2 b: high risk of RCC

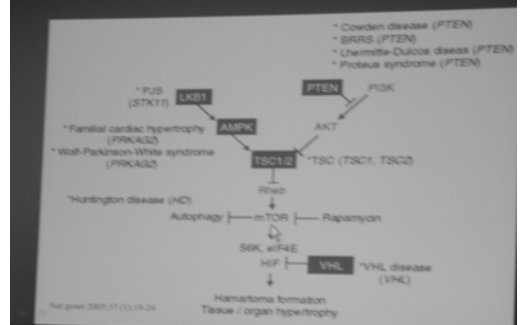
Klippel-Trenaunay

- > capillary malformation of one leg
- > ipsilateral hypertrophy
- > varicose veins

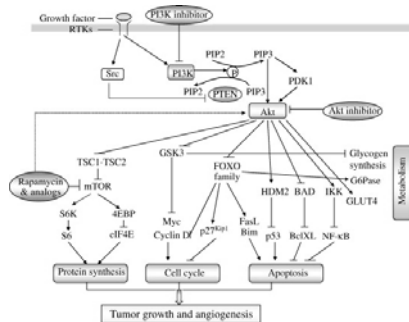
Mammalian target of rapamycin (mTOR)



The molecular link between several inherited human diseases



Biochimica et Biophysica Acta 1784 (2008) 150–158



phosphatidylinositol 3-kinase (PI3K) pathway
 Phosphatase and tensin homologue deleted on chromosome 10 (PTEN) is an antagonist of PI3K. mTOR is a serine/threonine kinase downstream of Akt, which controls protein synthesis, cell cycle progression, cell proliferation, and tumor growth. Rapamycin and its analogues CCI-779, RAD001, and AP-23573 inhibit mTOR activation by binding to FK506-binding protein-12.



Congenital Neuropathies
 Axonal with arthrogryposis

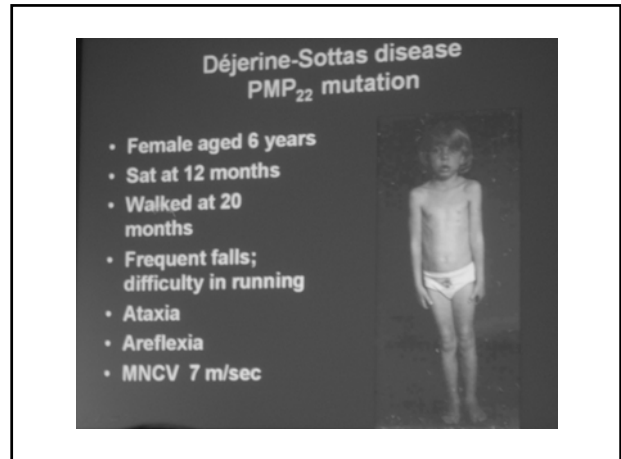
- At birth, scoliosis, hip dislocations, ankylosis of knees, bilateral talipes
- Left tibia and fibula fractures at Caesarean birth
- Marked distal weakness, wasting and hyporeflexia
- MNCVs 13 and 20 m/sec
- Axonal degeneration



Congenital Neuropathies
 Amyelinating

- Severe from birth
- Arthrogryposis common
- High mortality due to breathing and swallowing difficulties
- Severe disability in survivors
- NCV very slow or unrecordable
- Minimal myelin and no onion-bulbs on biopsies
- Molecular biology poorly understood

1. Charcot-Marie-Tooth disease (CMT) is the most common inherited neuropathy. It causes slowly progressive, distally pronounced, symmetric muscle atrophy in patients with age of onset at young adolescence.
2. Originally, CMT was classified using clinical and electrophysiological criteria into demyelinating (type 1), axonal (type 2), and other less frequent forms.
3. More recently, mutations in numerous different genes were found to cause CMT; therefore, the classification was extended by genetic means.
4. In more than 50% of all cases, the genetic defect underlying CMT is an intrachromosomal
5. duplication spanning 1.5Mb on human chromosome 17p12, the locus defining CMT subtype
6. 1A (CMT1A).
7. The gene encoding the peripheral myelin protein of 22kDa (*Pmp22*) is located in the duplicated region and was identified as the responsible disease gene. well.1



How to assess the pathogenicity of mutations in Charcot-Marie-Tooth disease and other diseases?

Andrzej Kochański

J Appl Genet 47(3), 2006, pp. 255-260

Table 1. Mutations in the "CMT genes" whose pathogenicity is unclear

CMT subtype	Mutated gene	Mutation
CMT2A	<i>KIF1B</i>	Q98L
CMT1C	<i>LITAF/SIMPLE</i>	T49M, L122V
CMT2E	<i>NEFL</i>	E7K, I214M
CMT1	<i>PMP22</i>	T118M

Peripheral myelin protein, with a molecular weight of 22 kDa (PMP22), is mainly expressed in Schwann cells in the peripheral nervous system. In the central nervous system, PMP22 expression was detected in motoneurons.

May 29 (Thursday)

Presidential Lecture (9:00-9:30)

How to approach the floppy infant syndrome
Makiko Osawa (Department of Pediatrics, Tokyo Women's Medical University, Japan)

An award ceremony and short lecture from a best publication award-winner (9:30-9:50)

International Symposium Celebrating the 50th JSCN Part II (9:50-11:30)

Opening Address

Makiko Osawa (President of the Congress)

1) Child neurology in Japan (10:05-10:25)

Yuko Fukuyama (Founder of the Japanese Society of Child Neurology)

2) History of child neurology and Japanese contributions to the development of child neurology (10:25-10:50)

Robert Oentner (President of ICNA, University of Sydney, Division of Neurology, Children's Hospital at Westmead, Australia)

3) The Japan's role in child neurology in the AOCN (10:50-11:00)

Yong-Seung Hwang (President of AOCN, Department of Pediatrics, Pediatric Neurology, Seoul National University Children Hospital, Korea)

4) Presentation of awards to JSCN contributors (invitees) (11:00-11:20)

Special Lecture I (11:30-12:20)

Brain mechanisms forming the mind

Makoto Iwata (Department of Neurology, Tokyo Women's Medical University, Japan)

Special Lecture II (12:20-14:30)

Society and developing mind

Takeshi Yoshida (Professor Emeritus, Tokyo University)

Invited Lecture I (14:30-15:30)

Conspital muscular dystrophy

Francesca Muntoni (Department of Pediatrics, Victor Dabowitz Neuromuscular Centre Imperial College London, UK)

Invited Lecture II (15:30-16:30)

Considering our way toward the pathophysiology of autism: clinical models and lessons for treatment

Roberto Tichman (Autism and Related Disorder Program, Miami Children's Hospital Don Marino Center, USA)



Dystroglycan: from biosynthesis to pathogenesis of human disease

Rita Barresi and Kevin P. Campbell*

Journal of Cell Science 119, 199-207

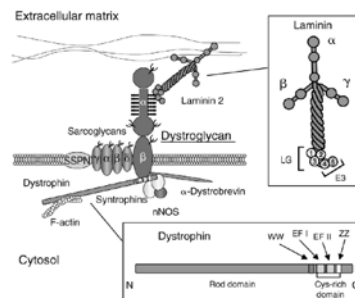
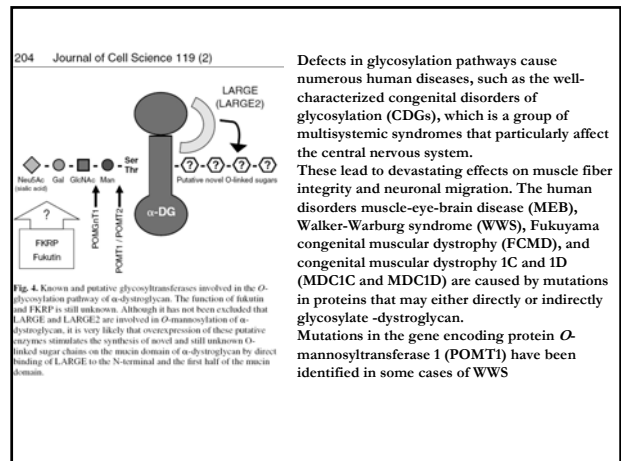
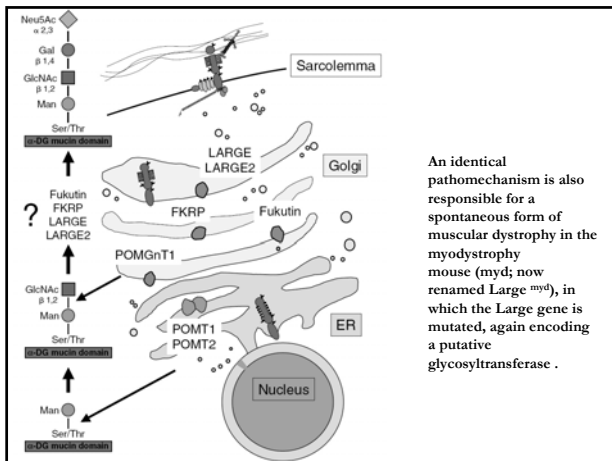
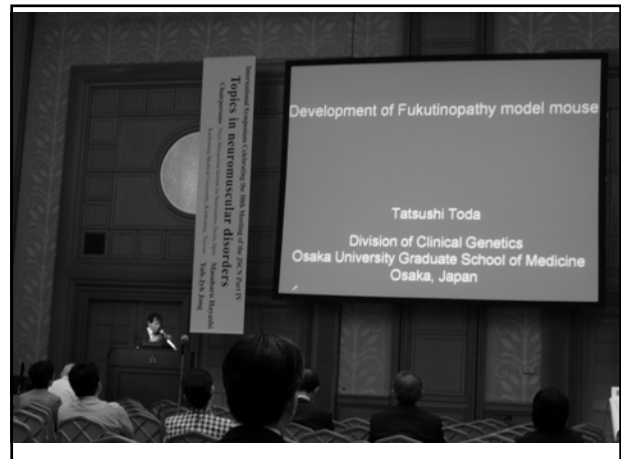


Fig. 2. DGC and dystroglycan ligands. Schematic representation of dystroglycan within the DGC. Structural domains of dystroglycan-binding proteins are represented in the insets. Domains of dystroglycan described in the text are represented. A similar domain structure is found in utrophin. α-Dystrobrevin lacks a WW domain but contains the EF-hand and ZZ domains. Laminins are composed of three distinct chains termed α, β and γ. To date, 5 α, 4 β and 3 γ laminin chains have been identified that can combine to form 15 different isoforms (Hallmann et al., 2005). In the C-terminal ends of the α chains, there are globular G domains (LG 1-5) composed of five similar modules. Dystroglycan binds with high affinity to the LG domains of laminin 1 (α1β1γ1) and 2 (α2β1γ1) α chains (Talis et al., 1999). Antibodies against the C-terminal LG4-LG5 pair of the α1 chain (E3 fragment) have been widely used for organ morphogenesis studies.



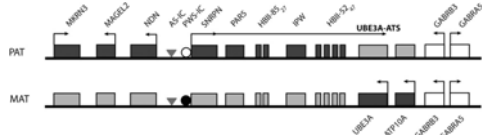
Defects in glycosylation pathways cause numerous human diseases, such as the well-characterized congenital disorders of glycosylation (CDGs), which is a group of multisystemic syndromes that particularly affect the central nervous system. These lead to devastating effects on muscle fiber integrity and neuronal migration. The human disorders muscle-eye-brain disease (MEB), Walker-Warburg syndrome (WWS), Fukuyama congenital muscular dystrophy (FCMD), and congenital muscular dystrophy 1C and 1D (MDC1C and MDC1D) are caused by mutations in proteins that may either directly or indirectly glycosylate α -dystroglycan. Mutations in the gene encoding protein O-mannosyltransferase 1 (POMT1) have been identified in some cases of WWS

Dystroglycanopathies			
	Walker Warburg	Muscle Eye Brain	Fukuyama
- POMT1	Walker Warburg		
- POMT2	Walker Warburg		
- POMGnT1		Muscle Eye Brain	
- Fukutin			Fukuyama
- Large			MDC1D
- FKRP			MDC1C LGMD2I



Biomedicine & Diseases: Review
Molecular epigenetics of Angelman syndrome

Figure 1. Imprinting map of the human chromosome 15q11–q13 AS/PWS region illustrating the status of allele-specific expression in brain



Cell. Mol. Life Sci. Vol. 64, 2007

Biomedicine & Diseases: Review Article 949

Table 1. Molecular classes of AS with methylation status

Class	Chromosome/genetic abnormality	~%	Normal differential methylation
I	15q11–13 deletion	70	No
II	UPD (Uniparental disomy)	5	No
III	ID (Imprinting defect)	5	No
IV	UBE3A mutation	10	Yes
V	Unknown	10	Yes

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Neuron-specific recombination by Cre recombinase inserted into the murine tau locus

Kazuhiro Muramatsu^{a,b}, Yukiko Hashimoto^a, Takefumi Uemura^a, Masataka Kunii^a, Reiko Harada^a, Takashi Sato^a, Akihiro Morikawa^a, Akihiro Harada^{a,*}

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The 50th Meeting of the Japanese Society of Child Neurology

— International Symposium Celebrating the 50th Meeting of the JSCN —

→ Japanese

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Program

May 28 (Wednesday)

International Symposium Celebrating the 50th Meeting of the JSCN Part I (13:30-18:00)

1. Recent Advances in Child Neurology1 (13:35-15:05)

- 1) Big brains & small brains: genetic and epigenetic mechanisms of brain size alteration (13:35-14:05)
Takao Takahashi (Department of Pediatrics, Keio University School of Medicine, Tokyo, Japan)
- 2) Recent advances on neurocutaneous syndromes (14:05-14:35)
Paolo Curatolo (Professor of Pediatric Neurology and Psychiatry, Department of Neuroscience, Tor Vergata University of Rome, Italy)
- 3) Age-related presentations of hereditary peripheral neuropathies in childhood (14:35-15:05)
Robert A Ouvrier (The Institute for Neuromuscular Research, The Children's Hospital at Westmead, Sydney, Australia)

2. Recent Advances in Child Neurology 2 (15:15-16:40)

- 4) Japanese encephalitis in Korea and Asian countries-Can it be under control? (15:15-15:40)
Yong-Seung Hwang (Seoul National University Children's Hospital, Seoul, Korea)
- 5) The impact of infections of the central nervous system on African children (15:40-16:10)
Charles RJC Newton (Kenya Medical Research Institute, Kilifi, Kenya and University College of London, London, UK)
- 6) Progress in the biological aspects of status epilepticus (16:10-16:40)
Solomon L Moshé (Albert Einstein College of Medicine and Montefiore Medical Center New York, NY, USA)

3. Recent Advances in Child Neurology 3 (16:50-17:55)

- 7) Gene therapy for muscular dystrophy (16:50-17:20)
Shinichi Takeda (Department of Molecular Therapy, National Institute of Neuroscience, National Center of Neurology and Psychiatry, Kodaira, Japan)
- 8) Robot suits-Cyberoid - biorobotics, control for supporting disabled persons - (17:20-17:55)
Yoshiyuki Sankai (Department of System & Information Engineering, University of Tsukuba, Tsukuba, Japan)

Evening seminar 1 (19:00-21:00)

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 Collage)
- 5) Noboru Ohki (NoruPro Lights System. Inc)
- 6) Takako Jodoi (Department of Child Dvelopment Kumamoto University Hospital)
- 7) Junko Kawatani (Child Development Sosiology Facutry of Medical and Pharmaceutical Sciences Kumamoto University Graduate School)
- 8) Takashi Ohya (Department of Pediatrics and Child Health, Kurume University School of Medeicine)

May 29 (Thursday)

Presidential Lecture (8:50-9:20)

Clinical Aspects on Myotonic Dystrophy in Childhood

Makiko Osawa (Department of Pediatrics, School of Medicine, Tokyo Women's Medical University)

Presentation of JSCN Awards to Outstanding Young Investigators (9:30-9:50)

International Symposium Celebrating the 50th Meeting of JSCN Part II

Celebration and Ceremoney for the 50th meeting of the JSCN(10:00-11:10)

- 1) Opening Address
Makiko Osawa (President of the 50th JSCN congress)
- 2) History of Child neurology in Japan (10:05)
Yukio Fukuyama (Tokyo Women's Medical University)
- 3) Presentation of Awards to JSCN Contributors (10:23)
- 4) Japanese Contributions To Child Neurology - An International Perspective - (10:33)
Robert Ouvrier (The Institute for Neuromuscular Research, The Children's Hospital at Westmead, Sydney, Australia)
- 5) The Japan's role in child neurology in the AOCNA (10:49)
Yong-Seung Hwang (Seoul National University Children's Hospital, Seoul, Korea)

Special Lecture 1 (11:30-12:30)

Mind viewed from brain

Makoto Iwata (Department of Neurology, Tokyo Women's Medical University, Tokyo, Japan)

Luncheon Seminar 1

- 1) New progress in Treatment of Epilepsy
Akiko Ikeda (Kyoto University)
- 2) Treatment of intractable epilepsies based on the mechanism of action of the antiepileptic drugs
Kenji Sugai (Department of Child Neurology, National Center Hospital for Mental, Nervous and Muscular Disorders, National Center of Neurology and Psychiatry, Tokyo)

Special Lecture 2 (13:30-14:30)

Society and developing mind

Takeshi Yohrou (Tokyo University)

Invited Lecture 1 (14:30-15:30)

Congenital muscular dystrophy

Francesca Muntoni (Dubowitz Neuromuscular Centre, UCL Institute of Child Health, London, UK)

Invited Lecture 2 (15:30-16:30)

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, Miami, FL, USA)

May 30 (Friday)

Morning Seminar 1 (8:00-9:00)

Visually diagnosed epileptic seizures: focal seizures

Chairpersons Tatsuro Izumi (Department of Pediatrics, Oita University)
Masako Sakauchi (Department of Pediatrics, Tokyo Women's Medical University)

Morning Seminar 2 (8:00-9:00)

EEG: an introduction

Chairpersons Yoko Ohtuska (Department of Child Neurology, Okayama University)
Solomon L. Moshe (Albert Einstein Collage of Medicine)

Morning Seminar 3 (8:00-9:00)

Effect of GH treatment on the brain metabolism

Chairpersons Goro Takada (Yonezawa National Hospital)
Shinichi Niijima (Department of Pediatrics, Juntendo University Nerima Hospital)

Didactic Lecture 1 (9:00-9:40)

Surgical management of pediatric epilepsy
Tomokatsu Hori (The Department of Neurosurgery, Tokyo Women's Medical University)

Luncheon Seminar 4 (12:20-13:20)

Challenges toward treatments for neurodegenerative diseases

Chairpersons Yoshikatsu Eto (Department of Pediatrics, the Jikei University School of Medicine)
Shoji Tsuji (Tokyo University)

Luncheon Seminar 5 (12:20-13:20)

Treatment for Intractable Epilepsy

Chairpersons Kazuie Inuma (Ishimaki Red Cross Hospital)
Paolo Curatolo (Professor of Pediatric Neurology and Psychiatry, Department of Neuroscience, Tor Vergata University, Rome, Italy)

Luncheon Seminar 6 (12:20-13:20)

Working women and psychological development of children

Chairpersons Trunekazu Yamano (The Department of Pediatrics, Osaka City University Graduate School of Medicine)
Hisako Watanabe (The Department of Pediatrics, School of Medicine, Keio University)

Luncheon Seminar 7 (12:20-13:20)

Mechanically assisted coughing in patients with neuromuscular disease

Chairpersons Mitsuru Kawai (National Hospital organization Higashisaitama Hospital)
Yuka Ishikawa (Department of Pediatrics, National Hospital Organization, Yakumo Hospital)

Didactic Lecture 2 (15:40-16:20)

"Tailoring" the Best available neuroprotection in High and Low Resource Settings; Bench to bedside, tertiary centres to developing World
Osuke Iwata (Center for Developmental and Cognitive Neuroscience, Department of Pediatrics & Child Health)

International Symposium Celebrating the 50th JSCN Part III (9:00-12:00)

1. Fukutinopathy (9:00-9:40)
Chairpersons 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)
2. Symposium : Phenotypic spectrum of Fukutinopathy (9:50-12:00)
Chairpersons 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 (1)
Francesco Muntoni (Dubowitz Neuromuscular Centre, UCL Institute of Child Health, London, UK)
 - 3) Fukutingene 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)

International Symposium Celebrating the 50th JSCN Part IV (14:30-15:40)

- Topics in neuromuscular disorders
- Chairpersons 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, Kaosiung, Taiwan)
 - 2) New insights into the pathogenesis of SMA
Yasushi Itoh (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)

Didactic Lecture 4 (15:40-16:20)

- Selective dorsal rhizotomy for spasticity in cerebral palsy children
- Chairpersons Kayoko Saito (Institute of Medical Genetics Tokyo Women's Medical University)
Takaomi Taira (Department of Neurosurgery, Tokyo Women's Medical University)

Didactic Lecture 5 (9:00-9:40)

- Understanding and support for developmental disorders with autism
- from the viewpoint of living and working together after adolescence -
- Chairpersons Tsunozuka Yamano (The Department of Pediatrics, Osaka City University Graduate School of medicine)
Masami Sasaki (Department of Pediatrics, Juntendo University Nerima Hospital)

Didactic Lecture 6 (15:40-16:20)

- Epigenetic mechanism regulating neural cell fate determination

Chairpersons Kousaku Ohno (Department of Child Neurology Faculty of Medicine, Tottori University)
Nakashima Kinich (Laboratory of Molecular Neuroscience, Graduate School of Biological Sciences, Nara Institute of Science and Technology)

Didactic Lecture 7 (9:00-9:40)

Cerebral palsy

Chairpersons Tohru Konishi (Nagaoka Ryoikuen)
Kenji Yokochi (Department of Pediatric Neurology, Seirei-Mikatahara General Hospital)

Symposium 1 (9:50-12:00)

Diagnosis and treatment of childhood epilepsy - Expert opinion -

- 1) West syndrome and its related disorders
Katsuhiko Kobayashi (Department of Child Neurology, Okayama University Hospital)
- 2) Lennox syndrome and related epilepsies
Tateki Fujiwara (Shizuoka Institute of Epilepsy and Neurological Disorders)
- 3) Myoclonic epilepsies 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, Landau-Kleffner syndrome, and related syndromes
Akihisa Okumura (Department of Pediatrics, Juntendo University School of Medicine)

Symposium 2 (16:20-18:30)

Hot topics in Neonatal Neurology

- 1) The utility of the amplitude integrated EEG as the brain monitor
Kyoko Hirasawa (The Department of Pediatrics, 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 (The 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)

Symposium 4 (16:20-18:30)

Neurosurgical approach from a perspective of pediatric neurology

Chairpersons Shizuo Ohi (Department of Neurosurgery, the Jikei University School of Medicine)
Hiroaki Date (Chiba Children's Hospital, department of neurosurgery)

- 1) Neurosurgical perspectives of 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 (Division 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)

Symposium 5 (9:50-12:00)

Contribution of Child Neurology toward the Study of Developmental Disabilities

Chairpersons 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)

Symposium 6 (16:20-18:30)

Epigenetics in Neurodevelopmental Diseases

Chairpersons 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 Graduate School of Medicine)
- 3) Regulation of neuronal development by the imprinted gene *Necdin*
Kazuaki Yoshikawa (Lab. of Regulation of Neuronal Development, Institute for Protein Research, Osaka University)
- 4) The chromosome - engineered mouse model of 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)

Symposium 7 (9:50-12:40)

Comprehensive medical care and support for children with severe motor and intellectual disability

Chairpersons Eiji Kitazumi (National Rehabilitation Center for Disabled Children)
Yuji Iwasaki (Tobu Ryoiku Center)

- 1) Advances and problems in treatment of 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 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 the Handicapped)

Evening Seminar 2 (19:00-21:00)

The Committee of the plan macological issues - Concerta® Distribution management committee -

Chairpersons Tasuku Miyajima (Department of Pediatrics, Tokyo Women's Medical University)
Kiatam Hayashi (Tokyo Women's Medical University Yachiyo Medical Center)

- 1) Tasuku Miyajima (Tokyo Medical University 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)

Evening Seminar 3 (19:00-21:00)

Multiple Sclerosis and Neuromyelities Optica in Japan

Chairpersons Toshiro Hara (Department of Pediatrics, Graduate School of Medical Sciences, Kyusyu University)
Kazuo Fujiwara (Department of Multiple Sclerosis Therapeutics, Tohoku University School of Medicine)

Evening Seminar 4: Committee of social activity (19:00-21:00)

Chairpersons Takeo Sugimoto (Biwako Gakuen Medical and Welfare Center)
The preblems 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 Welare 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)

Evening Seminar 5: Botulinum toxin Treatment (19:00-20:00)

Chairpersons 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)

May 31 (Saturday)

Didactic Lecture VIII (9:00-9:40)

Regulation of neuronal migration by doublecortin and its partners
Teruyuki Tanaka (Department of Developmental Medical Sciences, Graduate
School of Medicine, The University of Tokyo)

Morning Seminar 4 (8:00-9:00)

A Clinical Approach to the Dysmorphic Child

Chairpersons Hitoshi Yamamoto (Department of Pediatrics, St.Marianna
University School of Medicine)

Kenjiro Kosaki (Department of Pediatrics, Keio University School of
Medicine)

Morning Seminar 5 (8:00-9:00)

Pediatric neuroimaging diagnosis A to Z

Chairpersons Toshiaki Hashimoto (Naruto University of Education)

Hiroshi Oba (Department of Radiology, Teikyo University School of
Medicine)

Morning Seminar 6 (8:00-9:00)

Visually diagnosed epileptic seizures: generalized seizures

Chairpersons Shunsuke Otawara (Okayama University)

Hirokazu Oguni (Department of Pediatrics, Tokyo Women's Medical
University)

Seminars open to the public (12:00-16:00)

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

1) 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)

2) Introduction to the audience I

Toyojiro Matsuishi (Department of Pediatrics and Child Health, Kurume
University School of Medicine)

3) Introduction to the audience II II

Yoichi Sakakihara(Department of Child Care and Education, Ochanomizu
University)

4) Early risers and early sleepers are healthy both mentally and physically

Kohyama Jun (Tokyo Kita Shakai Hoken Hospital)

5) 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)

6) PTSD and the developing brain

Nobumasa Kato (Department of Psychiatry, Showa University School of
Medicine)

7) As a care-partners for Children who need Special Education

Michiko Hara (Faculty of Education, Gunma University)

8) The prevention of childhood depression

Kayo Inoko(Tokyo Institute of Psychiatry)



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