International Symposium on
Febrile Seizures and Related Conditions

The 11th Annual Meeting of the Infantile Seizure Society

Program & Abstracts

April 10 - 11, 2008
Royal Oak Hotel Spa & Gardens
Otsu-city, Shiga, Japan

Sponsored by Infantile Seizure Society (ISS), Japan
Co-sponsored by Japan Foundation for Neuroscience and Mental Health

Supported by Japan Epilepsy Society
Japan Pediatric Society
Japanese Society of Child Neurology

Contents

Welcome Messages

Dear colleagues;

It is my great pleasure to be hosting the International Symposium on Febrile Seizures and Related Conditions (ISFS) as the 11th Annual Meeting of the Infantile Seizure Society (ISS) in Otsu City, Shiga Prefecture, Japan. ISFS aims to present a comprehensive update of the topic and discussions on such issues as genetics, epidemiology, pathophysiology, imaging, subsequent epilepsy, mesial temporal sclerosis, treatment, and education about febrile seizures and related conditions.

I believe this symposium will be inspiring and fruitful for all participants as many distinguished scientists and physicians in this field are invited from all over the world. It will certainly be a wonderful opportunity for you to exchange recent views and engage in discussions on all aspects of febrile seizures and related conditions - one could say, "from genes and neurons to emergency medicine."

Shiga prefecture is located almost in the center of the Japan archipelago. Located near Kyoto, the nation's capital for 12 centuries, the cultural development of Shiga began early and Shiga has appeared many times on the center stage of Japanese history. Thus, Shiga is endowed with a rich historical and cultural heritage, as well as beautiful nature. All participants of ISFS can take the opportunity to appreciate the spring scenery of Shiga and Kyoto before or after the Symposium.

I look forward to welcoming you to Shiga and I truly hope all of us will have a wonderful time together at the ISS 2008 meeting, deepen friendships, share information with new acquaintances, and advance ourselves for the future of children in the world.

Yoshihiro Takeuchi, MD, PhD
President of ISFS
Professor & Chairman
Department of Pediatrics,
Shiga University of Medical Science

Dear colleagues;

It is our great pleasure to declare that The Infantile Seizure Society will host a significant scientific meeting this spring in Otsu City, Japan, that is, The International Symposium on Febrile Seizures and Related Conditions (ISFS), under the presidency of Dr Yoshihiro Takeuchi, Professor of Pediatrics, Shiga University, Otsu, Japan. The Infantile Seizure Society is a world-oriented organization specializing in the study of seizure problems in infants and young children. Since its birth in 1998, the Society has been hosted serial international symposia on various hot topics every year. The topics dealt in the past include neuro-inflammation and -immunology, West syndrome and epileptic encephalopathies, chromosomal aberrations, neuronal migration disorders, epileptic syndromes and taxonomy, status epilepticus, and biology of seizure susceptibility in developing brain. Invited lecturers and audiences have enthusiastically discussed on these contemporary subjects; the contents of lectures and discussions have been published in special journal issues or even recorded in audio DVD files. Thus past international symposia organized by the Society has been highly evaluated by the circle of relevant researchers, broadly. Now, for the 2008 meeting, febrile seizures and related conditions were chosen as the main theme. This historical topic dramatically revived as one of the central issues of epileptology through numerous new findings and discoveries in recent years. We believe that ISFS 2008 should be an exciting meeting which can't be missed by any clinicians and investigators who are engaging in clinical and bench works in this field. Further, you are fully guaranteed that you will be able to enjoy an essence of and satisfy with a world famous traditional Japanese hospitality while attending at ISFS. Once again, we would like to welcome all relevant colleagues at the ISFS, Otsu, Japan, April 10-11, 2008.

Yukio Fukuyama

Yukio Fukuyama, MD, PhD
Chairperson, Board of Councilors,
Infantile Seizure Society

Organization

ORGANIZING COMMITTEE

[A] GENERAL

Supreme Advisor Yukio FUKUYAMA (Tokyo, Japan)

Chairperson (President) Yoshihiro TAKEUCHI (Otsu, Japan)

Co-chairperson Kenji SUGAI (Kodaira, Japan)

Advisors

Sunao KANEKO (Hirosaki, Japan)
Shunsuke OHTAHARA (Okayama, Japan)
Tatsuya TANAKA (Asahikawa, Japan)
Kazuyoshi WATANABE (Nagoya, Japan)

Comittee Members

Kai-Ping CHANG (Taipei, Taiwan)
Tateki FUJIWARA (Shizuoka, Japan)
Mitsumasa FUKUDA (Matsuyama, Japan)
Shin-ichirou HAMANO (Saitama, Japan)
Shin-ichi HIROSE (Fukuoka, Japan)
Yong-Seung HWANG (Seoul, Korea)
Kazuie IINUMA (Ishinomaki, Japan)
Akio IKEDA (Kyoto, Japan)
Yushi INOUE (Shizuoka, Japan)
Tatsuro IZUMI (Oita, Japan)
Akemi KAKIDA (Niigata, Japan)
Shigeki KAMEYAMA (Niigata, Japan)
Osamu KANAZAWA (Saitama, Japan)
Mitsuhiro KATO (Yamagata, Japan)
Kensuke KAWAI (Tokyo, Japan)
Ryutaro KIRA (Fukuoka, Japan)
Jun KOHYAMA (Tokyo, Japan)
Toyojiro MATSUISHI (Kurume, Japan)
Yoshiya MURASHIMA (Tokyo, Japan)
Toshisaburo NAGAI (Osaka, Japan)
Shin-ichi NIIJIMA (Tokyo, Japan)
HIrokazu OGUNI (Tokyo, Japan)
Akihisa OKUMURA (Tokyo, Japan)
Makiko OSAWA (Tokyo, Japan)
Yoko OTSUKA (Okayama, Japan)
Taisuke OTSUKI (Kodaira, Japan)
Shinji SAITO (Sapporo, Japan)
Yasuhiro SUZUKI (Izumi, Japan)
Satoshi TAKADA (Kobe, Japan)
Takao TAKAHASHI (Tokyo, Japan)
Kazuhiro YAMAKAWA (Wako, Japan)
Tsunekazu YAMANO (Osaka, Japan)
Hideo YAMANOUCHI (Tochigi, Japan)
Hitoshi YAMAMOTO (Kawasaki, Japan)
Yasuko YAMATOGI (Okayama, Japan)

[B] SCIENTIFIC PROGRAM COMMITTEE

Chairpersons Yoshihiro TAKEUCHI (Otsu, Japan), Tomoyuki TAKANO (Otsu, Japan)

Comittee Members

Yukio FUKUYAMA (Tokyo, Japan)
Toyojiro MATSUISHI (Kurume, Japan)
Toshisaburo NAGAI (Osaka, Japan)
Kenji SUGAI (Kodaira, Japan)
Yasuhiro SUZUKI (Osaka, Japan)
Satoshi TAKADA (Kobe, Japan)
Hitoshi YAMAMOTO (Kawasaki, Japan)
Tsunekazu YAMANO (Osaka, Japan)

[C] FUND COMMITTEE AND TREASURER

Chairperson & Treasurer Yoshihiro TAKEUCHI (Otsu, Japan), Shin-ichi NIIJIMA (Tokyo, Japan)

Comittee Members

Yukio FUKUYAMA (Tokyo, Japan)
Toyojiro MATSUISHI (Kurume, Japan)
Toshisaburo NAGAI (Osaka, Japan)
Kenji SUGAI (Kodaira, Japan)
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Hitoshi YAMAMOTO (Kawasaki, Japan)
Tsunekazu YAMANO (Osaka, Japan)
Hideo YAMANOUCHI (Tochigi, Japan)

[D] SPONSORING ORGANIZATIONS

Sponsored: Infantile Seizure Society

Co-sponsored: Japan Foundation for Neuroscience and Mental Health

Supported:
Japan Epilepsy Society
Japan Pediatric Society
Japanese Society of Child Neurology

Head Office

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Tel: +81-77-548-2228 / Fax: +81-77-548-2230
Email: iss2008@belle.shiga-med.ac.jp
URL: http://www.shiga-med.ac.jp/~iss2008/

Registration

Desk for Registration and General Information, located at the B1 floor of Royal Oak Hotel Spa & Gardens, will be opened for the following periods:

April 10th (Thursday) 08:00-19:00
April 11th (Friday) 07:30-19:00

Pre-Registration

Those who completed the registration before March 31, 2008, should go to the Pre-registrant Reception Desk, present his/her Registration Confirmation Sheet to the receptionist and then receive his/ her ready-prepared bag.

On-site Registration

Registration Form should be presented to the reception desk, after filling out its upper part only, together with the fee payment in Japanese yen (cash) of appropriate amount.

The fee rates are defined variably as shown below according to the participant's category.

	2 days participation April 10 & 11	***1 day participation*** April 10 or 11
Symposium		
Japanese colleagues		
ISS* member	21,000 JPY	10,000 JPY
Non-ISS member	24,000 JPY	12,000 JPY
Non-Japanese colleagues		
AOCNA** members	18,000 JPY	10.000 JPY
Non-AOCNA members	24,000 JPY	10,000 JPY
Junior physicians***	18,000 JPY	10,000 JPY
Grand Social Party (April 10)		
Japanese colleagues	5,000 JPY	5,000 JPY
Non-Japanese colleagues	Free	Free

* = Infantile Seizure Society

** = Asian & Oceanian Child Neurology Association

*** = Young physicians graduated from medical school after January, 2002. Students of post-graduate course are also applicable to this category. Copy of official document such as a student's identification or a certificate may be required.

To ISS members:

The members of the Infantile Seizures Society (ISS) are requested to pay his/her annual fees (¥3,000) at the registration desk. Any one who wish to become a member of ISS is requested to fill out the membership application form and pay the 2008 annual fee (¥3,000).

To AOCNA members:

The members of the Asian & Oceanian Child Neurology Association (AOCNA) are requested to contact the AOCNA reception desk, and to confirm his/her correspondence address in the membership roster and status of his/her dues payment. To become a member of AOCNA, please fill out the application form and pay the two years fee for the year 2008 and 2009 (20.00US$) or the whole life fee (100.00US$) in cash.

Don't miss the Grand Social Party

Please get together everybody in the Grand Social Party at the room "ORCHID".

Japanese colleagues are requested to register. Please refer to the "General Information (p.7)".

General Information

Date

April 10th (Thu) - April 11th (Fri), 2008

Venue

Royal Oak Hotel Spa & Gardens
23-1, Kayanoura, Otsu-city, Shiga, 520-2143, Japan
Phone: +81-77-543-0011
URL: http://www.royaloakhotel.co.jp

Official Language

English. No simultaneous translation available.

Social Function

1) Presidential Welcome Reception

Date & Time : Wednesday, April 9th, 19:00 - 20:30
Place : The Room "Le Vent Vert", 2nd floor, Royal Oak Hotel Spa & Gardens
Attire : Casual
Attendance : Limited to the invitees

2) Grand Social Party

Date & Time : Thursday, April 10th, 19:00 - 20:30
Place : The Room "ORCHID", B1 floor, Royal Oak Hotel Spa & Gardens
Attire : Casual
Attendance : Free (Non Japanese colleagues) /5,000 JPY per person (Japanese colleagues)

Official Certificate for Attendance and CME Points

An official certificate for attendance at the ISFS will be delivered to all participants.
To Japanese colleagues, a certificate for authorized CME units will be rewarded by two societies as follows:

Society	Attendance	Authorship	
		Presenter	Co-author
Japan Pediatric Society	5 u	0 u	0 u
Japanese Society of Child Neurology	2 u	2 u	0 u

u=unit

Lunch (Days 1 and 2)

Lunch will be served for participants during the time of Lunch Time & Poster Round (1) and (2) at the dining room, which is adjacent to the place of Oral Presentation in the "ORCHID".

Coffee

Coffee will be served at the B1 floor, beside the PC Preview Center.

Internet

Available in all guest rooms and allows you unlimited access for free while staying on the floor.
Please prepare a LAN cable (10BASE-T) for yourself.
No need for complicated set-up and easy with Plug & Play.

Council Business Meeting

The council business meeting of ISS will be held at the "CATTLEYA", the 1st floor, from 17:30 to 18:30, April 11th after the scientific program is over. The ISS councilors are requested to attend this meeting.

Satellite Business Meeting

The Asian & Oceanian Child Neurology Association (AOCNA) Executive Board Meeting will be held at the "CATTLEYA", the 1st floor, from 14:00 to 18:00, April 9th. The second meeting will be provisionally scheduled at the same room from 19:00, April 11th.

To Make Your Stay Comfortable

Climate

April is the most comfortable season of the year in Otsu area with high temperature being around 20 degree Celsius (68 degree Fahrenheit) and low around 10 degree Celsius (50 degree Fahrenheit).

Currency Exchange

We strongly recommend purchasing yen at Kansai International Airport. This is because you need cash in yen to purchase tickets for ground transportation from the airport to wherever your first destination in Otsu may be.

Voltage and Frequency of Electricity

The voltage is 100 V throughout the country. Frequency is 60 Hz in Otsu area and 50Hz in the Eastern Japan such as Tokyo.

Shopping

Five percent sales tax is applicable to personal purchases. Duty free shops are available at Kansai International Airport.

Gratuity / Tip

Gratuity or "service fee" is included in a bill at most of the descent restaurants. Otherwise they usually do not accept any tips.

Sightseeing Tours

If you wish to have a sightseeing tour during your stay in Japan, we recommend consulting the concierge at the Royal Oak Hotel.

Secretariat

Inquiries on ISFS 2008:

ISFS Secretariat
Yoshihiro TAKEUCHI, MD, PhD
Professor and Chairperson
Department of Pediatrics, Shiga University of Medical Science
Seta-Tsukinowa, Otsu 520-2192, Japan
Phone: +81-77-548-2228 / Fax: +81-77-548-2230
Email: iss2008@belle.shiga-med.ac.jp

Inquiries on Infantile Seizure Society in general:

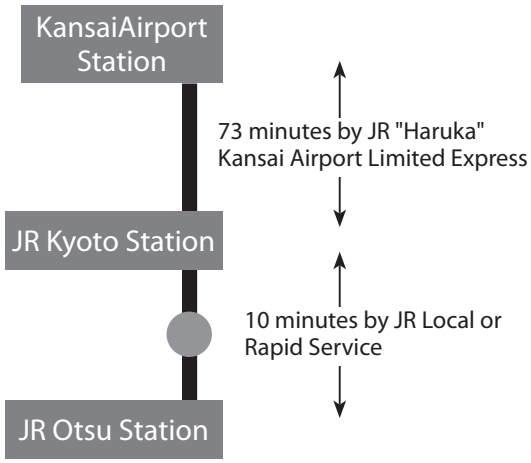
ISS Secretariat
Yukio FUKUYAMA, MD, PhD
c/o Child Neurology Institute
6-12-17-201 Minami-Shinagawa, Shinagawa-ku, Tokyo
140-0004, Japan
Phone: +81-3-3740-7680 / Fax +81-3-3740-0874
Email: yfukuyama@sc4.so-net.ne.jp

ISS / ISFS Website

http:www.iss-jpn.info/

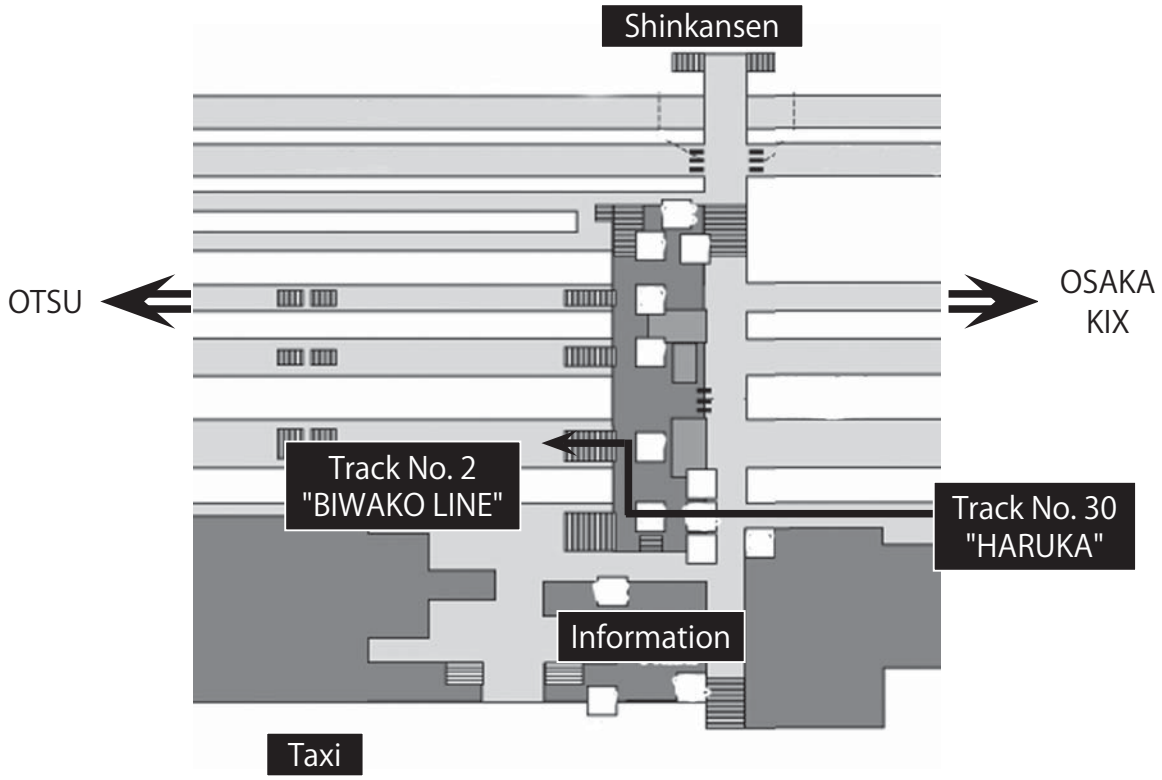
Venue Access

Access to Japan Railway (JR) Otsu Station



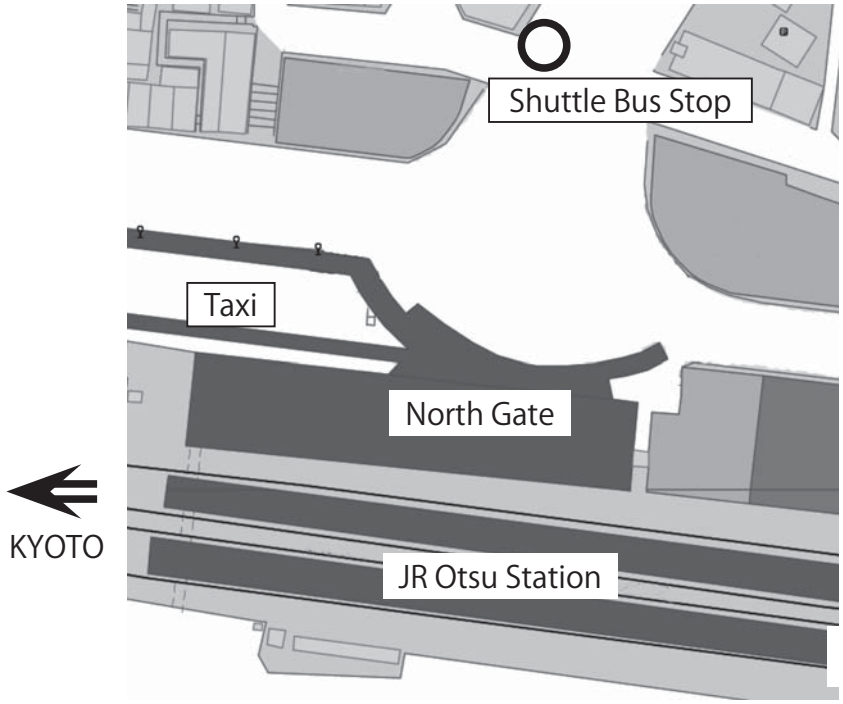
Traveling times do not include the time needed for transferring.

JR Kyoto Station Map



Kansai Airport Limited Express "HARUKA" will arrive in the track No.30 of the Kyoto railway station. Please take the "BIWAKO LINE" of the track No.2, and come to the Otsu station (approx. 10min) where you should get off. In front of the north gate of the Otsu station, please take a taxi or a shuttle bus to the Royal Oak Hotel (approx. 15 min).

JR Otsu Station Map



Shuttle Bus (Free):
April 10: 8:00- every one hour
April 11: 7:00- every one hour

Taxi fee: 2,200 - 2,500 JPY

Instructions for Oral Presentations

1. Keep the time as strictly as possible. Please note that the time frame shown in the program. A bell rings once to indicate one minutes before the end, and twice in sequence to inform the time over.
2. Only one single projection will be available.
3. Every speaker is requested to finish up an arrangement necessary for data projection two hours before the respective presentation at the latest, by contacting the staff of the PC Preview center, located at the B1F.
4. The data has to be presented with USB flash memory or CD-R.
5. Facilities to preview are available at the PC preview center.
6. All presentations should be prepared by "Power Point, after ver. 2002" on Windows system. If your data was prepared by Mac system, the data may deform after its transfer to the Windows system. In this case, please check and correct this possible deformation at the Preview Center. If your Power Point is before ver. 2002, please inform it to the ISFS Secretariat by mail or to the staff of Preview Center at the venue, as early as possible.
7. Each presenter is requested to manipulate the computer placed at the platform during the presentation. If you need help for manipulation, please inform to the ISFS Secretariat at the Preview Center. Your own personal computer is also available to use for presentation.
8. Video tape presentation is not available. If you need to use video records, please transfer them to the computer in a digital form. In this case, it is advised to bring your own personal computer, since the software does not fit often for your record. If you bring your own computer, Mac system is also available.
9. Please remind that the details of the oral presentations will be delivered over a network by audio/video streaming, thereby enabling closed-users to see and hear the audio and video files. In this data streaming, the majority of PC slides will be shown with synchronized oral presentations. It will contain almost all lectures and discussions presented at the ISFS meeting. In this regard, if you have any problems, or any PC slides that you want to delete from this data streaming, please contact the staff of PC Preview Center.

Instructions for Discussion

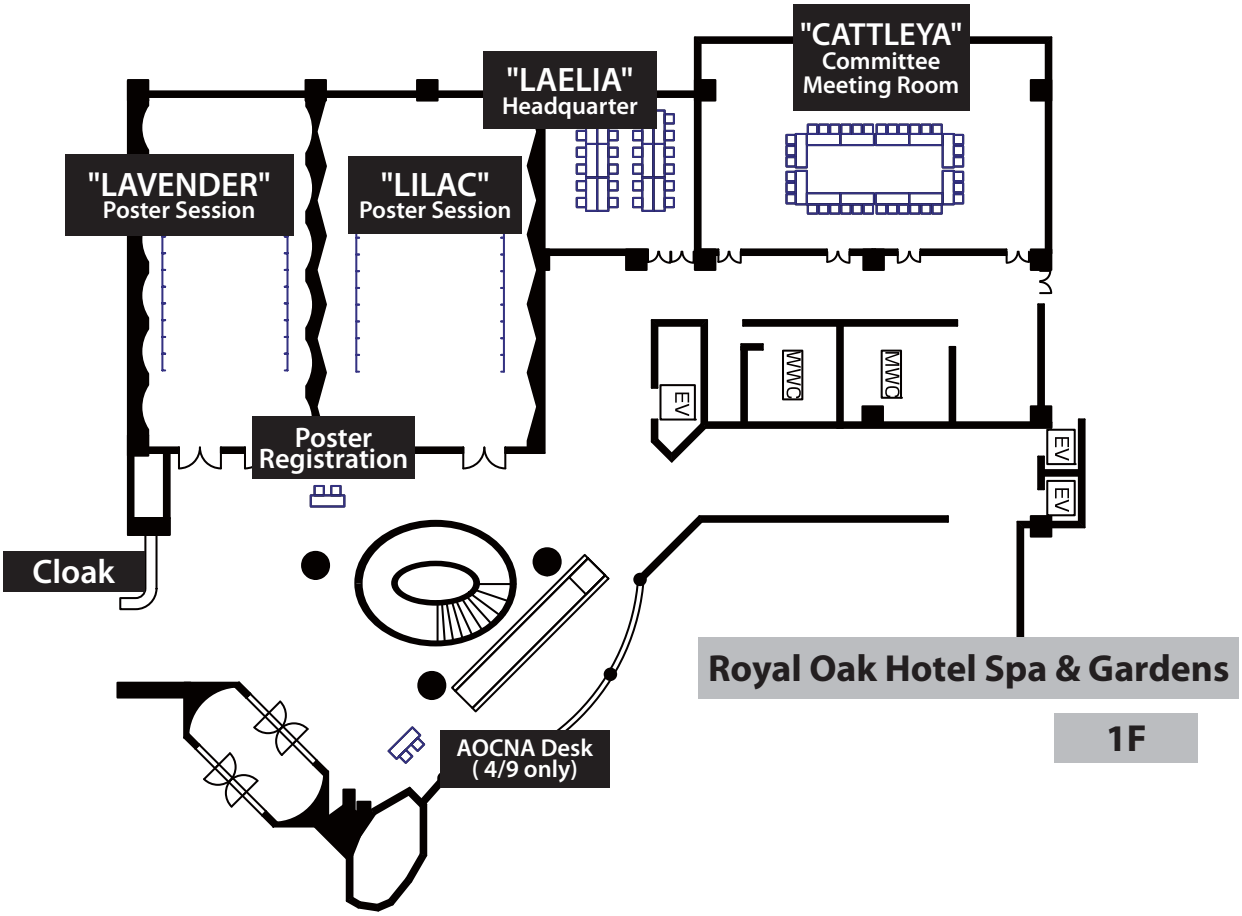
1. Active discussion from the Floor is encouraged as far as the time is availabe.
2. All aspects of discussion session shall be orderd by due consideration of chairpersons.
3. Anyone who wishes to raise a question/discussion is urged to line up befor the microphone stands to save a time, and wait for an order of chairpersons. To begin your discussion, please identify yourself first.

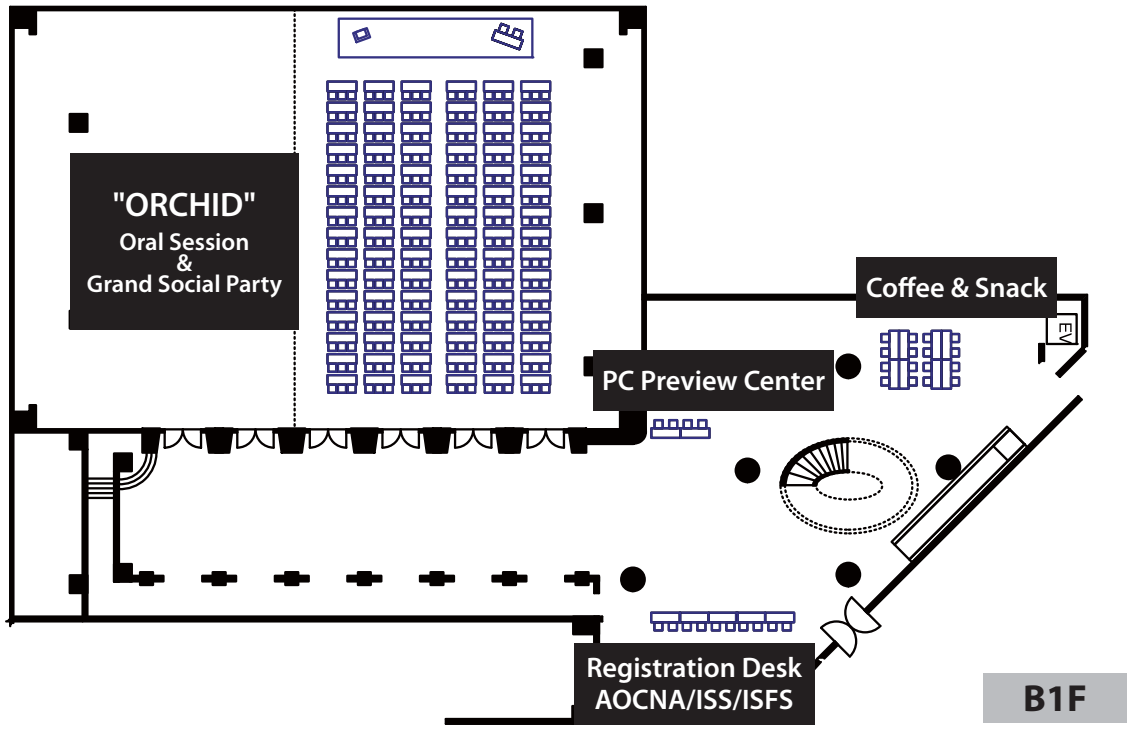
Instructions for Poster Presentations

1. Place: “LILAC” and “LAVENDER“ on the 1st floor.
2. Registration: The presenter is requested to register at the "Poster Reception Desk" on the 1st floor.
3. Pushpins will be provided.
4. All presentations should be posted on the pre-assigned panel by 9:00 am, April 10th.
5. A poster panel has a surface of 90 cm wide and 180 cm high. Top space will be used to place the poster number in a size of 20 cm x 20 cm, pre-fixed by the Secretariat. The title, author's names, and affiliations should be prepared by the presenters in the top space of 70 cm wide and 20 cm high. The other main space of 90 cm wide and 160 cm high is available for the body of poster presentation.
6. Poster round. Presenters are requested to be present at the site of respective posters for discussion during the time of Coffee Break & Poster Presentation (1) or (2). Each presenter is also requested to make a brief oral presentation for 3 minutes, and another 3 minutes will be allowed for questions from the participants. The rooms, the coordinators, and the detailed time schedule for each presentation are as follows.

Coffee Break & Poster Presentation (1) April 10 (Thursday), 14:20 - 15:40			***Coffee Break & Poster Presentation (2)*** April 11 (Friday), 09:30 - 10:50		
Room	LILAC	LAVENDER	Room	LILAC	LAVENDER
Coodinator	Hayashi M	Izumi T	Coordinator	Oguni H	Niijima S
14:30 - 14:36	Poster No.1	Poster No.23	09:40 - 09:46	Poster No.12	Poster No.34
14:36 - 14:42	2	24	09:46 - 09:52	13	35
14:42 - 14:48	3	25	09:52 - 09:58	14	36
14:48 - 14:54	4	26	09:58 - 10:04	15	37
14:54 - 15:00	5	27	10:04 - 10:10	16	38
15:00 - 15:06	6	28	10:10 - 10:16	17	39
15:06 - 15:12	7	29	10:16 - 10:22	18	40
15:12 - 15:18	8	30	10:22 - 10:28	19	41
15:18 - 15:24	9	31	10:28 - 10:34	20	42
15:24 - 15:30	10	32	10:34 - 10:40	21	43
15:30 - 15:36	11	33	10:40 - 10:46	22	44

Floor Plan


"CATTLEYA"
Committee
Meeting Room
"LAELIA"
Headquarter
"LAVENDER"
Poster Session
"LILAC"
Poster Session
MWC
MWC
EV
EV
EV
Poster
Registration
Cloak
AOCNA Desk
(4/9 only)
Royal Oak Hotel Spa & Gardens
1F


"ORCHID"
Oral Session
&
Grand Social Party
Coffee & Snack
EV
PC Preview Center
Registration Desk
AOCNA/ISS/ISFS
B1F

Overview of Daily Program

Wednesday, April 9

14:00 - 18:00
AOCNA
Council Meeting

19:00 - 20:30
Welcome Reception
(Invitees only)

Thursday, April 10

8:00 - 9:00
Registration
Opening Addresses

9:00 - 11:10
Epidemiology
Tsai ML
Vestergaard M
Genetics
Nakayama J
Kira R

11:10 - 12:00
Lunch Time Seminar (1)
Lecture: Neville BG / Chair: Shinnar S

12:00 - 12:40
Lunch Time & Poster Round

12:40 - 14:20
Electrophysiology
Kaila K
Nordli DR

14:20 - 15:40
Coffee Break &
Poster Presentations (1)

15:40 - 16:50
Management & Treatment
Lux AL
Sugai K

16:50 - 18:30
Prolonged FS
Shinnar S
Baram TZ

19:00 - 20:30
Grand Social Party

Friday, April 11

7:30 - 8:00
Registration

8:00 - 9:30
Experimental Approaches
Jiang Q
Dunleavy M
Chang YC

9:30 - 10:50
Coffee Break &
Poster Presentations (2)

10:50 - 12:00
Cytokines
Fukuda M
Heida JG

12:00 - 12:50
Lunch Time Seminar (2)
Lecture: Kubek MJ / Chair: Baram TZ

12:50 - 13:30
Lunch Time & Poster Round

13:30 - 14:30
Encephalopathy
Yamanouchi H
Nagasawa T
Specchio N

14:30 - 16:50
GEFS+
Shike T
Scheffer IE
SMEI
Granata T
Yamakawa K

16:50 - 17:00
Closing Addresses

17:30 - 18:30
ISS Council Meeting

19:00 -
AOCNA Council Meeting
(provisional)

PROGRAM

The 11th Annual Meeting of the Infantile Seizure Society

International Symposium on Febrile Seizures and Related Conditions (ISFS)

Date: April 10th (Thursday) to 11th (Friday)
Venue: Royal Oak Hotel Spa & Gardens
Otsu, Japan

PROGRAM - ORAL PRESENTATIONS

Day 1, APRIL 10 (THURSDAY)

08:00 - 19:00 **REGISTRATION**

08:50 **OPENING ADDRESSES**

08:50 - 08:55
Opening address by Fukuyama Y (Chairperson, Board of Councilors, ISS)

08:55 - 09:00
Opening address by Takeuchi Y (President of ISFS)

09:00 **Epidemiology, Genetics**

Chairpersons: *Matsuishi T (Kurume, Japan)*
Kim DW (Goyang, Korea)

09:00 - 09:20
O 01 **RISK FACTORS FOR SUBSEQUENT EPILEPSY AFTER FEBRILE SEIZURES: A CASE-CONTROL STUDY IN TAIWAN**
Tsai ML[1)], Hung KL[2)] (Taipei[1)2)],Taiwan)

09:20 - 10:10 INVITED
O 02 **RISK AND CONSEQUENCES OF FEBRILE SEIZURES IN DENMARK**
Vestergaard M (Aarhus, Denmark)

10:10 - 10:40 INVITED
O 03 **SEARCHING FOR THE FEBRILE SEIZURE SUSCEPTIBILITY GENES IN JAPANESE**
Nakayama J (Ibaraki, Japan)

10:40 - 11:10 INVITED
O 04 **GENETIC SUSCEPTIBILITY TO FEBRILE SEIZURES: CASE-CONTROL ASSOCIATION STUDIES**
Kira R, Ishizaki Y, Torisu H, Sanefuji M, Yukaya N, Sakai Y, Hara T (Fukuoka, Japan)

11:10 **Lunch Time Seminar (1)**

Chairperson*: Shinnar S (New York, USA)*
Sponsored by Kyowa Hakko Kogyo Co., Ltd, Japan

11:10 - 12:00 INVITED
O 05 **FEBRILE SEIZURES – SEMIOLOGY IN HUMANS AND ANIMAL MODELS. EVIDENCE OF FOCALITY AND HETEROGENEITY**
Neville BGR, Gindner D (London, UK)

12:00 - 12:40 **Lunch Time and Poster Round**

12:40 **Electrophysiology**

Chairpersons: *Nagai T (Osaka, Japan)*
Okumura A (Tokyo, Japan)

12:40 - 13:30 INVITED

O 06 **NEUROBIOLOGICAL AND PHYSIOLOGICAL MECHANISMS UNDERLYING FEVER-RELATED EPILEPTIFORM SYNDROMES**
Kaila K (Helsinki, Finland)

13:30 - 14:20 INVITED

O 07 **THE ROLE OF EEG IN FEBRILE STATUS EPILEPTICUS (FSE)**
Nordli D[1)], Moshé S[2)], Shinnar S[2)], and FEBSTAT team. (Chicago, USA[1)]; New York, USA[2)])

14:20 **Coffee Break and Poster Presentation (1)**

Posters 01 - 11 Coordinator: Hayashi M (Fuchu, Japan)
Posters 23 - 33 Coordinator: Izumi T (Oita, Japan)

15:40 **Management and Treatment**

Chairpersons: *Sugai K (Kodaira, Japan)*
Lux AU (Bristol, UK)

15:40 - 16:00

O 08 **TREATMENT OF FEBRILE SEIZURES: HISTORICAL PERSPECTIVE, CURRENT OPINIONS, AND POTENTIAL FUTURE DIRECTIONS**
Lux AL (Bristol, UK)

16:00 - 16:30 INVITED

O 09 **CURRENT MANAGEMENT OF FEBRILE SEIZURES IN JAPAN: AN OVERVIEW**
Sugai K (Kodaira, Japan)

16:30 - 16:40
Designated Commentary 1; Philippine's Guidelines
Ortiz M (Manila, Philippines)
Designated Commentary 2; Hong Kong's Guidelines
Wong V (Hong-Kong, China)
16:40 - 16:50
General Discussions

16:50 **Prolonged FS**

Chairpersons: *Watanabe K (Nagoya, Japan)*
Neville BGR (London, UK)

16:50 - 17:40 INVITED

O 10 **CONSEQUENCES OF PROLONGED FEBRILE SEIZURES**
Shinnar S (New York, USA)

17:40 - 18:30 INVITED

O 11 **FEVER, FEBRILE SEIZURES AND EPILEPSY: LESSONS FROM THE LABORATORY**
Baram TZ (Irvine, USA)

19:00 - 20:30 **GRAND SOCIAL PARTY**
Venue: (B1F)

Day 2, APRIL 11 (FRIDAY)

07:30 - 08:00 **REGISTRATION**

08:00 **Experimental Approaches**

Chairpersons: *Yamano T (Osaka, Japan)*
Kubek MJ (Indianapolis, USA)

08:00 - 08:20

O 12 **EPILEPTIFORM ACTIVITY EXERTS LONG-LASTING EFFECTS ON NMDAR AND AMPAR SUBUNIT EXPRESSION, DISTRIBUTION AND INTERACTION IN NEOCORTICAL CULTURES**
Jiang Q, Wang JM, Wu Y, Wu XR, Jiang YW (Beijing, China)

08:20 - 08:40

O 13 **A NOVEL MODEL OF PROLONGED SEIZURES IN THE IMMATURE BRAIN**
Dunleavy M[1)], Shinoda S[2)], Schindler C[2)], Bellver-Estelles C[1)], Henshall D[1)]
(Dublin[1)], Ireland; Portland[2)], USA)

08:40 - 09:30 INVITED

O 14 **THE LONG TERM EFFECTS OF FEBRILE SEIZURES ON THE HIPPOCAMPAL NEURONAL PLASTICITY: CLINICAL AND EXPERIMENTAL STUDIES**
Chang YC [1)], Huang CC[2)] (Kaohsiung[1)] and Tainan[2)], Taiwan)

09:30 **Coffee Break and Poster Presentation (2)**

Posters 12 - 22 Coordinator: Oguni H (Tokyo, Japan)
Posters 34 - 44 Coordinator: Niijima S (Tokyo, Japan)

10:50 **Cytokines**

Chairpersons: *Yamanouchi H (Tochigi, Japan)*
Kaila K (Helsinki, Finland)

10:50 - 11:10

O 15 **INFLUENCES OF CYTOKINE ON THE IRRITABILITY OF HYPERTHERMIA-INDUCED SEIZURES IN DEVELOPING RATS**
Fukuda M[1)], Ishizaki Y[2)], Kira R[2)], Suzuki Y [1)], Watanabe S[1)], Morimoto T[3)], Hara T[2)], Ishii E[1)]
(Matsuyama, Japan [1)3)]; Fukuoka, Japan[2)])

11:10 - 12:00 INVITED

O 16 **THE ROLE OF INTERLEUKIN-1β IN EXPERIMENTAL FEBRILE CONVULSIONS**
Heida JG (New York, USA)

12:00 **Lunch Time Seminar (2)**

Chairperson: *Baram TZ (Irvine, USA)*
Sponsored by Pfizer Japan Inc.

12:00 - 12:50 INVITED

O 17 **AGE-DEPENDENT ROLE OF THYROTROPIN-RELEASING HORMONE (TRH) IN SEIZURE MODULATION**
Kubek MJ (Indianapolis, USA)

12:50 - 13:30 **Lunch Time and Poster Round**

13:30 **Encephalopathy**

Chairpersons: *Yamamoto H (Kawasaki, Japan)*
Salonga A (Manila, Philippines)

13:30 - 13:50

O 18 **EARLY BIOMARKER FOR THE DIAGNOSIS OF ACUTE INFANTILE ENCEPHALOPATHY**
Yamanouchi H, Nakajima D, Kuribayashi R, Watabe Y, Imataka G, Arisaka O (Tochigi, Japan)

13:50 - 14:10

O 19 **CAN WE DISTINGUISH ENCEPHALOPATHY FROM FEBRILE SEIZURE CAUSED BY HHV-6 AT THE FIRST SEIZURE?**
Nagasawa T, Hoshino H, Mizuguchi K, Kubota M (Tokyo, Japan)

14:10 - 14:30

O 20 **THE LONG TERM EFFECTS OF FEBRILE SEIZURES ON THE HIPPOCAMPAL NEURONAL PLASTICITY: CLINICAL AND EXPERIMENTAL STUDIES**
Specchio N[1)], Fusco L[1)], Claps D[1)], Cilio MR[1)], Valeriani M[1)], Longo D[2)], Fardello G[2)], Gentile S[3)], Vigevano F[1)] (Rome[1)2)3)], Italy)

14:30 **GEFS+, SMEI**

Chairpersons: *Hirose S (Fukuoka, Japan)*
Guzzetta F (Rome, Italy)

14:30 - 14:50

O 21 **PARTIAL SEIZURES IN PATIENTS WITH SCN1A MUTATION**
Shike T [1)], Fujiwara T [1)], Shimomura J [1)], Kubota Y [1)], Inoue Y [1)], Yamakawa K [2)]
(Shizuoka[1)] and Saitama[2)], Japan)

14:50 - 15:40 INVITED

O 22 **DRAVET SYNDROME OR GEFS+?**
Scheffer IE (Melbourne, Australia)

15:40 - 16:00

O 23 **SMEI: A CLINICAL AND GENETIC STUDY OF 38 ITALIAN PATIENTS**
Granata T[1)], De-Giorgi I[1)], Freri E[1)], Morbi M[1)], Ragona F[1)], Franceschetti S[2)], Zara F[3)], Brazzo D[4)], Veggiotti PA[4)] (Milan[1)2)], Genova[3)] and Pavia[4)], Italy)

16:00 - 16:50 INVITED

O 24 **MOLECULAR BASIS OF SEVERE MYOCLONIC EPILEPSY IN INFANCY**
Yamakawa K (Saitama, Japan)

16:50 **Closing Addresses**

16:50 -
Closing address by Miike T (Chairperson, Board of Trustees, JSCN)
Closing address by Matsuishi T (President, 12th Annual Meeting of ISS, 2009)

PROGRAM - POSTER PRESENTATIONS

Exhibition time: Day 1, April 10, 09:00 - Day 2, April 11, 13:30
Mounting: Day 1: April 10, by 09:00
Take away: Day 2, April 11, 13:30 - 14:00

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Khan NZ[1)], Ferdous S[2)], Banu SH[1)], Afroza S[2)] (Dhaka[1)2)], Bangladesh)

P 02 **FEBRILE CONVULSION : A SURVEY ON 300 CHILDREN**
Karimzadeh P (Tehran, Iran)

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P 04 **A CLINICAL STUDY ON COMPLEX FEBRILE SEIZURES**
Kim DW, Kang JS (Goyang, Korea)

P 05 **CHARACTERIZATION OF THE GENE EXPRESSION PATTERN UPON DOUBLE-STRANDED RNA STIMULATION IN FEBRILE SEIZURES**
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P 10 **EXPRESSION LEVELS OF ICAM-1 AND LFA-1 IN PLASMA AND PBMC OF CHILDREN WITH FEBRILE SEIZURES**
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Abstracts - Oral Presentations

RISK FACTORS FOR SUBSEQUENT EPILEPSY AFTER FEBRILE SEIZURES: A CASE-CONTROL STUDY IN TAIWAN

1

11th ISS

Tsai ML[1], Hung KL[2]
[1] Department of Pediatrics, Cheng Hsin Rehabilitation Medical Center, Taiwan
[2] Department of Pediatrics, Cathay General Hospital, Taipei, Taiwan

Objectives: To examine risk factors for subsequent epilepsy and long-term follow-up after febrile seizures in a tertiary hospital in Taiwan
Methods: Of 530 hospitalized children with diagnosis of fever associated with seizures, 154 children were identified as fulfilling the criteria of febrile seizures (FS). Patients who developed epilepsy and matched controls were analyzed.
Results: The risk factors for afebrile seizures following initial FS were studied in 154 hospitalized children. After follow-up for a mean period of 7 years 2 months, nineteen patients (12.3%) developed subsequent epilepsy. The occurrence of epilepsy was strongly associated with complex features of FS (odds ratio [OR] = 6.52, 95% confidence interval [CI] = 2.41-17.7, P = 0.0002), family history of epilepsy (OR = 5.64, CI = 1.36-20.8, P = 0.02), pre-existing neurodevelopmental abnormalities (OR = 5.66, CI = 1.58-20.3, P = 0.01), and abnormal EEG findings (OR = 3.68, CI = 1.42-9.11, P = 0.02). However, the recurrent FS, sex, family history of FS, age of onset of FS, and long-term prophylaxis of anticonvulsants were not significant factors for subsequent epilepsy.
Conclusions: Our study demonstrated that complex features of FS, family history of epilepsy, and pre-existing neurodevelopmental abnormalities were significant risk factors in developing subsequent epilepsy.

RISK AND CONSEQUENCES OF FEBRILE SEIZURES IN DENMARK

2

11th ISS

Vestergaard M
Department of General Practice, Institute of Public Health, Aarhus University, Aarhus, Denmark

Objectives: To use Danish nationwide registries to evaluate risk factors and prognosis of febrile seizures.
Methods: We have conducted a number of studies within a population-based cohort of 1,6 million children born in Denmark (1977-2004). We constructed the cohort by linking registers on civil service, health, and cause of death. We followed the cohort for up to 28 years with virtually no loss to follow-up.
Results: The etiology of febrile convulsions depends on a genetic susceptibility that can be transmitted through both parents. Preterm birth and intrauterine growth retardation increased the risk of febrile seizures. MMR vaccination increased the risk of febrile seizures for two weeks but the absolute risk was small even in high-risk children. Febrile seizures increased the risk of epilepsy and the risk remained high well into adulthood. The risk was particular high for persons with cerebral palsy, low Apgar scores, or a family history of epilepsy. Complex febrile seizure increased the mortality for two years but the excess mortality was very low. Children with simple febrile seizures had mortality similar to the background population.
Conclusions: Febrile seizure is a common condition with a benign outcome for the vast majority of children. Genes and environmental factors operating in early life seem to play a causal role.

3 SEARCHING FOR THE FEBRILE SEIZURE SUSCEPTIBILITY GENES IN JAPANESE

11th ISS

Nakayama J
Department of Pediatrics, Ibaraki Prefectural University of Health Sciences, Ibaraki, Japan

Febrile seizures (FSs) represent the most common form of childhood seizures. It is recognized that a significant genetic component exists for susceptibility to this type of seizure. FSs affect 2-5% of all children in the Western world and are even more common in Japan, where their prevalence is estimated to be between 6 and 9%. Therefore, it is conceivable that studies of FSs in Japanese are appropriate for genetic analysis. Extensive genetic studies have shown that at least seven loci are responsible for FSs in Caucasians: FEB1 on chromosome 8q13-q21, FEB2 on 19p, FEB3 on 2q23-q24, FEB5 on 6q22-q24, FEB7 on 21q22, FEB8 on 5q31.1-q33.1, and FEB9 on 3p24.2-p23. We also identified two FS loci in Japanese: FEB4 on 5q14-q15 and FEB6 on 18p11. Recently, these two linkage studies have been replicated in some Caucasian families with FSs and epilepsy. We also reported a nonsense mutation of the MASS1/VLGR1 gene, which has been mapped on the FEB4 locus, in a Japanese FS family. In Caucasians, a deletion at 5q14.3 involving MASS1/VLGR1 was reported in a patient with myoclonic epilepsy. Thus, these two FS loci may also be responsible for FSs and/or idiopathic epilepsies in other ethnic groups. Further genetic studies are necessary to confirm this interethnic similarity and variation.

4 GENETIC SUSCEPTIBILITY TO FEBRILE SEIZURES: CASE-CONTROL ASSOCIATION STUDIES

11th ISS

Kira R, Ishizaki Y, Torisu H, Sanefuji M, Yukaya N, Sakai Y, Hara T
Department of Pediatrics, Graduate School of Medical Sciences, Kyushu University, Fukuoka, Japan

Objective: A genetic predisposition to febrile seizures (FS) has long been recognized. The inheritance appears to be polygenic in small families or sporadic cases of FS. To determine whether candidate genes are responsible for the susceptibility to FS, we have performed genetic association studies in FS patients and controls.
Methods: The functional single-nucleotide polymorphisms (SNPs) of genes related to immune response (interleukin (IL) 1B, IL6, IL8, IL10, TNFA, TLR3, TLR7, TLR9, UNC93B1), acute encephalopathy (CPT2), acid-base balance (SLC4A3, SLC9A1, SLC9A3), and neurotransmission (EAAT2, EAAT3, PRIP1) were examined in 249 FS patients (186 simple and 63 complex FS) and 225 controls.
Results: Significant associations between of IL1B -511 SNP and sporadic simple FS ($p = 0.002$), IL10 -592 SNP and simple FS ($p=0.014$), and UNC93B1 SNP (rs308328) and complex FS ($p=0.039$) were found.
Conclusions: These results suggest that IL-1β contributes to the genetic susceptibility to simple FS while IL-10 confers resistance to simple FS. The UNC93B1 gene, the mutations of which were recently reported to confer vulnerability to herpes simplex encephalitis, may be involved in the development of complex FS by direct brain insults.

FEBRILE SEIZURES – SEMIOLOGY IN HUMANS AND ANIMAL MODELS. EVIDENCE OF FOCALITY AND HETEROGENETY

5

11th ISS

Neville BG, Gindner D
UCL Institute of Child Health, Great Ormond Street Hospital and the National Centre for Young People with epilepsy, London, UK

Febrile seizures are defined according to purposes e.g broad pragmatic categorisation; acute management; long term counselling and genetic studies but all would now include prolonged seizures and thus a proportion of complex febrile seizures. The most telling evidence of seizure origin is early semiology but this requires a reasonably homogeneous clinical category.
There is evidence that a proportion of "FS" are provoked reflex asystole with no primary cerebral event with close accordance in semiology between attacks with pain and fever induced triggers.
The exclusion of intracranial infection is primarily for acute management of e.g meningitis but there have been questions about the presence of direct cerebral involvement in FS associated with shigella and malaria. In malaria endemic areas however a strong relationship exists between malaria induced convulsive status epilepticus (CSE) and later temporal lobe seizures.
The evidence radiologically and pathologically for hippocampal involvement in prolonged FS is large. Focality is common in prolonged FS and particularly in CSE with acute CNS infection.
Animal studies of prolonged seizure models have shown limbic seizures at low dose and CSE at high dose.
We studied the early features of short FS and found that 7 of 10 consecutive seizures had definite but subtle focal / temporal lobe symptoms "preceeding" the seizure but only one had an obvious focal seizure. The behaviour of 10 children with a fever (median 39°C) alone showed no temporal lobe features.
The simplest hypothesis to explain all the data seems to be; FS are secondarily generalised medial temporal seizures in which a general epileptogenic stress of fever causes seizure activity in the genetically low threshold hippocampus but the semiology from the undamaged hippocampus differs from that in those with long term temporal lobe damage (to which we are accustomed).

NEUROBIOLOGICAL AND PHYSIOLOGICAL MECHANISMS UNDERLYING FEVER-RELATED EPILEPTIFORM SYNDROMES

6

11th ISS

Kaila K
Department of Biosciences, and Neuroscience Center, University of Helsinki, 00014-Helsinki, Finland

Febrile seizures (FS) are the most common type of convulsive events in children. FS have been extensively studied using animal models, where rat and mice pups are placed in a hyperthermic environment. Such work has largely focused on the consequences rather than on the mechanisms of experimental febrile seizures (eFS). We have recently shown that eFS are preceded by a dramatic rise in the rate of respiration. The consequent respiratory alkalosis affecting the brain and increasing neuronal excitability is a direct cause of the eFS (Schuchmann et al. 2006 Nat Medicine). If a similar mechanism contributes to human FS and other fever-related epileptiform syndromes, a number of factors that have not been previously thought to be involved in their etiology must be considered. These include physiological and pathophysiological factors affecting CO2 chemosensitivity as well as systemic and cellular acid-base regulation. Furthermore, a critical role for brain pH in FS points to novel types of susceptibility genes, which include genes coding pH-sensitive target proteins (e.g. neuronal ion channels) and pH-regulatory proteins. In my talk, I will discuss these novel ideas and putative therapies based on them, and also the possible role of pH in the triggering of epileptiform activity following birth asphyxia.

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11th ISS

THE ROLE OF EEG IN FEBRILE STATUS EPILEPTICUS (FSE)

Nordli D[1)], Moshé S[2)], Shinnar S[2)], and FEBSTAT team.
[1)] Department of Pediatrics, Children's Memorial Hospital, Chicago, USA
[2)] Department of Neurology, Albert Einstein College of Medicine, New York, USA

Objectives: Consequences of Prolonged Febrile Seizures (FEBSTAT) is multi-center prospective study of children with FSE. Detailed analysis of EEG findings obtained within 72 hours of febrile status epilepticus has been an important part of the study.

Methods: Subjects, ages one month to five years, are recruited with 72 hours of FSE. Complete history and physicals are done along with an MRI, viral studies, developmental profile and EEG. EEGs are at least 30 minutes, use the 10-20 system, and read separately by two EEGers. Follow-up imaging, EEG and other evaluations are obtained one year after FSE.

Results: A total of 144 children have been recruited to date. The first 100 acute EEGs have shown focal slowing, attenuation or both are seen in 38%, most often in the temporal region. These findings correlate with the MRI findings of acute increased hippocampal T2-weighted signal and volume loss one year after FSE. Inter-reader reliability is excellent.

Conclusion: In contrast to simple febrile seizures EEG may be useful in children with FSE. In particular, EEG combined with imaging have promise to be useful biomarkers for later development of epilepsy in children with FSE.

8

11th ISS

TREATMENT OF FEBRILE SEIZURES: HISTORICAL PERSPECTIVE, CURRENT OPINIONS, AND POTENTIAL FUTURE DIRECTIONS

LUX AL
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Although most febrile seizures do no harm and two-thirds of initial cases have no witnessed recurrence, the seizures cause much family anxiety, are sometimes prolonged, and in rare cases are the first evidence of important epilepsy syndromes or are implicated in the development of epilepsy with mesial temporal sclerosis in later life. There have been trials of prophylactic treatment with antiepileptic drugs including carbamazepine, diazepam, phenobarbital, phenytoin, and sodium valproate. Several strategies have been employed with these drugs, including continuous secondary prophylaxis, intermittent secondary prophylaxis in response to later episodes of fever, and rescue medication early in the course of further seizures. Another treatment strategy has been using one or more antipyretic agents in early response to fever using agents such as acetaminophen and ibuprofen. Over the years, researchers have identified a variety of clinical, genetic, and environmental risk factors for more severe or prolonged febrile seizures and higher risk of recurrence. The best treatment strategy remains controversial, but the most rational approach would seem to be selective prophylactic treatment of cases that are identified as being at high risk of recurrence, status epilepticus and long-term adverse effects. A better understanding of these risk factors might also lead to the development of more specific treatments.

CURRENT MANAGEMENT OF FEBRILE SEIZURES IN JAPAN: AN OVERVIEW

9
11th ISS

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Management of febrile seizures (FS) includes acute treatment and differential diagnosis of seizures and prevention of recurrent FS. Acute management depends on seizure status and consciousness level on arrival: going home after physical examination when the patient has no seizure and good consciousness; close observation with some laboratory examinations when the patient has no seizure but consciousness disturbance; IV DZP when seizures continue. Among differential diagnosis of FS, central nervous system infections are important. CSF examination and/or CT/MR scan should be obtained if the patient has signs of meningeal irritation or increased intracranial pressure, >one hour consciousness disturbance, partial seizures, >15 minutes seizures, or >one seizure within 24 hours. A proposed guideline for prevention of recurrent FS in Japan indicates risk factors (RF): neurological or developmental abnormalities prior to the onset of FS, atypical FS (partial seizures, >15 minutes prolonged seizures, or recurrent seizures within 24 hours), epilepsies of the parents or siblings, onset of FS under one year, and history of FS in one or both parents. It recommends no medication for <two past FS without RF, prophylactic DZP administration on febrile episodes for prolonged FS or >two FS with >two RF, and chronic medication for FS under 38°C or prolonged FS with failure of DZP prophylaxis.

CONSEQUENCES OF PROLONGED FEBRILE SEIZURES

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11th ISS

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Introduction: Febrile seizures are the most common seizure type in childhood. While simple febrile seizures are benign, there is evidence that prolonged febrile seizures are associated with an increased risk of subsequent epilepsy, particularly temporal lobe epilepsy.
Methods: In a prospective study, we are prospectively enrolling children who present with febrile status epilepticus (>30 min) and performing an MRI, EEG and viral studies within 72 hours. These studies are repeated at one year.
Results: To date we have recruited over 150 children. Mean seizure duration is greater than one hour with approximately two thirds being focal. There is evidence of hippocampal injury (evidenced by acute increased T2 signal) in approximately 25% including 12% with markedly increased T2 signal and 12% with milder signal abnormalities. An additional 10% have other subtle hippocampal abnormalities such as HIMAL. EEG abnormalities including are also common, specifically focal slowing and attenuation. Approximately 30% have evidence of acute viral infection with human herpesvirus 6 or 7. Preliminary analysis of the one year MRIs shows volume loss and persistence of abnormal T2 signal in children with marked hippocampal T2 signal acutely.
Discussion: The data provide evidence that acute hippocampal injury does occur in a significant proportion of children with febrile status epilepticus. Preliminary results also suggest that this injury can be associated with subsequent MTS. Further follow-up is needed to determine long term prognosis in this cohort.
Supported by grant NS 43209 from NINDS.

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FEVER, FEBRILE SEIZURES AND EPILEPSY: LESSONS FROM THE LABORATORY

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Objectives: The study of febrile seizures has been driven, in part, by the association of these seizures with the development of temporal lobe epilepsy. The clinical question of whether long or recurrent febrile seizures cause temporal lobe epilepsy has remained unresolved. The objective of the studies that will be presented is to use mechanistic approaches and answer this question.
Methods: The authors developed a model of prolonged (complex) febrile seizures in immature rat and mouse. This should allow mechanistic examination of the potential causal relationship of febrile seizures and limbic epilepsy. Although the model relied on hyperthermia, the authors found that the hyperthermia provoked the release of endogenous fever mediators including interleukin 1 beta, and thus resembled human febrile seizures.
Results: The seizures were found to evoke epilepsy in a third of affected animals, permitting analysis of the mechanisms of epileptogenesis. To date, alteration of specific ion channels, perhaps driven by genomic effects of fever-related cytokines, as well as changes in endocannabinoid signaling, are candidate mechanisms. Importantly, MRI imaging of animals subsequent to experimental febrile seizures may provide a biomarker for individuals who are at risk for developing temporal lobe epilepsy or cognitive deficits after prolonged febrile seizures.
Conclusions: Prolonged febrile seizures in the animal model provoke epileptogenesis in a subset of individuals. The experimental approach enables discovery of biomarkers and mechanisms that can be employed in the clinic in the evaluation of prolonged febrile seizures in children.

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11th ISS

EPILEPTIFORM ACTIVITY EXERTS LONG-LASTING EFFECTS ON NMDAR AND AMPAR SUBUNIT EXPRESSION, DISTRIBUTION AND INTERACTION IN NEOCORTICAL CULTURES

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Objectives: To explore systematic effects of early epileptiform activity on the formation and function of developing neocortical neurons at the cellular level.
Methods: In vitro model of early-life seizure, total protein extract, biochemical sub-cellular fractionation, immunoblot analysis, Co-immunoprecipitation, immunocytochemistry and confocal microscope analysis.
Results: We found a decrease in expression of total neuronal protein NR2B NMDAR subunits and PSD-95 ($P < 0.05$) shortly after insult (within 24 hours). With the use of cell fractionation, we found that the cellular location of each NMDAR subunit (NR1, NR2A and NR2B), AMPAR subunit GluR1 and GluR2 and PSD-95 changed after the induced eplileptiform activity. Co-IP detection revealed a gradually enhanced interaction between PSD-95 and NR2A compared with NR2B from 7DIV to 21DIV. In addition, early-life seizure-like insults still exert effects on excitatory synapses number, size and distribution.
Conclusions: Epileptiform activity may cumber normal development of neocortical neurons and preserve them at much more naïve period with less matured function. These findings in an in vitro model may inform rodent models of epilepsy, as well as the pathology of seizures in human neocortical development.

A NOVEL MODEL OF PROLONGED SEIZURES IN THE IMMATURE BRAIN

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11th ISS

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Objectives: Experimental models suggest the immature brain is relatively resistant to the damaging effects of prolonged seizures. Nevertheless, some clinical data suggest prolonged seizures in infants can elicit lasting harm. In the present study we describe a novel model of prolonged seizures in rats at P10, which corresponds to a time rats were previously considered invulnerable to harm.
Methods: Seizures were evoked by intraamygdala microinjection of kainic acid. Acute histology was performed at 24-72h. Video-EEG and further histology was performed at adulthood.
Results: Seizures elicited significant unilateral hippocampal cell death within the CA3 and CA1 subfields. No significant injury was detected within contralateral hippocampal subfields. Examination of brain morphology at adulthood revealed a number of hallmarks of hippocampal sclerosis; neuronal loss, gliosis, mossy fiber sprouting and granule cell layer dispersion. Video-EEG recordings at P75 confirmed rats developed spontaneous seizures.
Conclusions: The present study provides evidence that prolonged seizures in neonatal rats can elicit hippocampal injury and this may develop into classical unilateral hippocampal sclerosis and spontaneous seizures, thus providing a platform for the evaluation of neuroprotective and anti-epileptogenic therapies.

THE LONG TERM EFFECTS OF FEBRILE SEIZURES ON THE HIPPOCAMPAL NEURONAL PLASTICITY: CLINICAL AND EXPERIMENTAL STUDIES

14
11th ISS

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Febrile seizures (FS) are the most common seizure disorder in childhood. It is important to delineate whether FS alters long-term neuroplasticity. We conducted a population study in Taiwan and found that school-aged children with prior FS had significantly better scores than controls on achievement test. They also exhibited better mnemonic capacity, more flexible mental processing, better control of distractibility, and higher impulsivity than controls. Multivariate analysis revealed that early-onset FS was the significant risk factor for memory deficits.
We used a heated-air FS paradigm, by which male rat pups were subjected to one, three, or nine brief episodes of FS. Although there was no hippocampal neuronal loss, the adult rats subjected to frequent FS had memory deficits. Their memory deficits correlated with decreased phosphorylation of cAMP response-element binding protein (CREB) and extracellular signal-regulated kinase (pErk)1/2 in the hippocampus. The frequent FS also led to long-term bidirectional modulation in synaptic plasticity and deficits in N-methyl-D-aspartate (NMDA)-dependent Erk1/2 phosphorylation. These findings raise concerns about the long-term cognitive consequences of frequent FS during early brain development.

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11th ISS

INFLUENCES OF CYTOKINE ON THE IRRITABILITY OF HYPERTHERMIA-INDUCED SEIZURES IN DEVELOPING RATS

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Objectives: Cytokines have been indicated to influence the irritability in childhood febrile seizures. We studied the role of IL-1ß, IL-1ra, IL-6, IL-10, and TNF-α in hyperthermia-induced seizures in developing rats.
Methods: Male Lewis rats (21-24 days old) were used. We applied human recombinant cytokines intra-nasally 1h before seizures induced by moist warm air. The brain temperature at the appearance of seizure discharges on EEG, and the latency time from the hyperthermia onset until the appearance of seizure discharges on EEG were measured.
Results: The brain temperature of the IL-1ß group was significantly lower than that of the control, and the brain temperatures of IL-1ra and IL-10 groups were significantly higher than that of the control. The latency times of IL-1ra, IL-6, and IL-10 groups were significantly longer than that of the control. The TNF-α group showed no significant difference.
Conclusions: These results suggest that IL-1ß plays a convulsive, and IL-1ra, IL-6, and IL-10 play an anti-convulsive role in hyperthermia-induced seizures in developing rats.

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THE ROLE OF INTERLEUKIN-1β IN EXPERIMENTAL FEBRILE CONVULSIONS

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Febrile convulsions (FCs) occur in children as a result of fever. Despite their prevalence, the pathophysiology of FCs has remained unclear. Recent evidence from clinical and experimental studies has highlighted a potential role of immune generated products in the genesis of FCs. Of particular interest are the cytokine interleukin-1beta (IL-1β) and its naturally occurring antagonist interleukin-1 receptor antagonist (IL-1ra). Using a novel model of FCs, involving the generation of physiological fever, we investigated the role of the IL-1β/IL-1ra system in the genesis of FCs. FCs were induced in P14 rats. Brain levels of IL-1β and IL-1ra were measured at the onset of FCs and two hours after. Additional animals were given intracerbroventricular (ICV) IL-1β or IL-1ra. Animals with FCs had increased hippocampal and hypothalamic IL-1β compared to equally treated animals without FCs, which was first evident at onset in the hippocampus. There were no differences in IL-1ra levels. ICV IL-1β increased the number of animals with FCs while IL-1ra had an opposite anti-convulsant effect. This suggests that excessive amounts of IL-1β may be involved in the pathophysiology of FCs. The results of these studies, in combination with recent results from others, have further established a role for the IL-1β/IL-1ra system in the genesis of FCs.

AGE-DEPENDENT ROLE OF THYROTROPIN-RELEASING HORMONE (TRH) IN SEIZURE MODULATION

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11th ISS

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Our laboratory was instrumental in characterizing TRH as the first neuropeptide in the extrahypothalamic human CNS. Since then we have focused on the age-dependent role of TRH in the limbic system especially as it relates to seizure modulation. TRH neurons, receptors and specific metabolic enzymes are highly expressed in the amygdala, hippocampus, piriform and entorhinal corticies. Moreover, animal studies have demonstrated that TRH mRNA and TRH are upregulated while TRH receptors are downregulated for some time postictally in specific limbic loci. TRH is also known to have anticonvulsant effects in several animal seizure models. Clinically, it is known to be efficacious in treating infantile spasms, Lennox-Gastaut syndrome, myoclonic seizures, and other refractory generalized and partial seizures. Therefore, we utilized the kindling model of temporal lobe epilepsy to determine if intranasal administration of D,L Polylactide nanoparticles containing TRH (TRH-NPs) could inhibit kindling development (epileptogenesis). Intranasal application of TRH-NPs resulted in a significant reduction in total seizure ADD compared to the control-NPs as kindling progressed, while the number of stimulations required to achieve stage V seizures, and to become fully kindled was significantly greater in TRH-NP treated animals. Additionally, delay to clonus was significantly prolonged while clonus duration was reduced indicating a less severe seizure in TRH-NP treated subjects. Thus, it seems reasonable to suggest that intranasal delivery of sustained-release TRH-NP may be a viable means of therapy for recurrent febrile seizures and febrile status epilepticus in children.

EARLY BIOMARKER FOR THE DIAGNOSIS OF ACUTE INFANTILE ENCEPHALOPATHY

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11th ISS

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Objective: We measured CSF tau protein in patients with acute encephalopathy at the onset to determine whether it could be useful as an early biomarker leading to rapid diagnosis.
Methods: Two clinical categories exhibiting febrile convulsive status epilepticus were highlighted. One was "febrile convulsion status (FCS)", and the other was "acute infantile encephalopathy predominantly affecting the frontal lobes (AIEF)". The manifestations of the latter are similar to FCS at the onset, but regarded as an acute encephalopathy syndrome because of characteristic frontal dysfunctions seen after the onset (Epilepsy Res 2006;70:S263-265). CSF samples were colleted from 15 patients with FCS and 10 with AIEF. Tau protein was measured by ELISA.
Results: The median and mean values of tau protein in CSF taken within 24 hrs were 178 and 178 pg/ml in FCS (n=11). Whereas, they were 736 and 1,553pg/ml in AIEF (n=5), respectively. CSF tau protein taken within 24 hrs from patients with AIEF was significantly higher than those with FCS ($p<0.01$).
Conclusions: CSF tau protein is an early biomarker of AIEF, and its quick measurement may be useful for an early diagnosis of AIEF. (Supported by grants from NCNP, Ministry of Health, Labor and Welfare, Japan)

19 CAN WE DISTINGUISH ENCEPHALOPATHY FROM FEBRILE SEIZURE CAUSED BY HHV-6 AT THE FIRST SEIZURE?

11th ISS

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Background: In Japan, characteristic encephalopathy caused by HHV-6 manifesting as recurrent seizures during the eruptive stage has been reported. No difference in laboratory data and MRI findings has been reported between this encephalopathy and febrile seizure (FS) caused by HHV-6 at the first seizure. Thus, certain predictors for distinguishing between these two conditions are required.
Objectives: To evaluate the duration of fever prior to the first seizure in patients with encephalopathy and FS caused by HHV-6.
Methods: Medical charts of patients with this encephalopathy (n=17) were retrospectively reviewed for determining the duration of fever. Medical charts of patients with FS and exanthema subitum without sequelae (n=64) were also reviewed at our hospital.
Results: Duration of fever was exactly estimated in 12 encephalopathy and 16 FS patients. The mean duration of fever prior to the first seizure was 26.9h (9-43h; median, 28h) in the encephalopathy patients and 10.6h (0.5-31; median, 8.5h) in FS patients. There was a significant difference between the encephalopathy and FS patients. ($p<0.01$). Only two patients (12.5%) with FS showed fever duration longer than 24 h.
Conclusions: The duration of fever prior to the first seizure can help in distinguishing between the encephalopathy and FS caused by HHV-6 at an early stage.

20 SEVERE PARTIAL EPILEPSY AND ENCEPHALOPATHY DUE TO IMMUNE-MEDIATED DISORDER (SPEEDI DISORDER)

11th ISS

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Rationale: To describe the clinical entity characterized by severe epilepsy and severe behavioral and cognitive dysfunctions following an acute febrile seizure disorder resembling encephalitis in previously normal children, probably due to an immune-mediated cerebral disorder.
Methods: We selected, both prospectively and retrospectively, patients with intractable seizures and CNS signs of an ongoing immune-mediated encephalopathy.
Results: We studied 8 patients with a mean age of 11.7+7 yrs. At the age of 6.7+6.2yrs (9m-17.6yrs, median 5 yrs) they presented with fever and stupor associated with frequent partial seizures which developed in epileptic status in seven cases and subsequent seizures in one. EEG at onset showed a diffuse slow activity associated with epileptiform abnormalities involving in all cases the temporal lobe, with extension in the frontal area in one, in parietal and occipital in one and in the central regions in two cases. Viral or bacterial infections have been excluded. Oligoclonal bands were present in four out of six tested patients. Brain MRs were normal at onset and during follow-up in three cases. In the other four MRs showed at the onset T2 and FLAIR bilateral hyperintensity over peri-insular regions in three and over frontal and mesial temporal regions in one. The mean follow-up period was 4.2+2.2 yrs. All patients developed without latency severe partial epilepsy with relapsing remitting course in six. A spectrum from attention deficits to dementia has been observed in association with intermittent and severe behavioral disorder. Treatment with corticosteroids and/or IVIG determined a reduction of seizure number and improvement in competences.
Conclusion: In these patients severe partial epilepsy with a wide spectrum of behavioral and cognitive disorder is associated to signs of an immune-mediated cerebral ongoing process. The immune-mediated process is suggested by inflammatory CSF changes, reinforced by MR changes and sustained by the efficacy of immune-modulating therapy. The striking improvement in some patients after corticosteroids or IVIG, confirm that the recognition of this entity is highly crucial in the outcome.

PARTIAL SEIZURES IN PATIENTS WITH SCN1A MUTATION

21
11th ISS

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Objectives: Mutations of the neuronal voltage–gated sodium channel α subunit 1 gene (SCNIA) have phenotypes from generalized epilepsy with febrile seizures plus (GEFS+) to severe myoclonic epilepsy in infancy (SMEI). We report here patients with predominantly partial seizures.
Methods: 15 patients from 13 families (8 males, 7 females; ages, 4 to 31 years) with predominantly partial seizures were retrospectively investigated for their clinical characteristics and gene mutation.
Results: Age at epilepsy onset was from 2 months to 13 years. The seizure symptoms were partial motor seizure/complex partial seizures occasionally with subsequent uni- or bilateral convulsion. All were fever-sensitive. EEG showed paroxysms in F in 6, CP in 4, CT in 2, O in 1, multifoci in 1 and diffuse in 2. Mental function was delayed mildly in 7 and severely in 3. During the course, 4 patients remitted, and 3 got worse with frequent seizures. The mutations were nonsense in 1, frameshift in 1, missense in 10, and other in 1.
Conclusions: Patients with SCN1A mutation may have predominantly partial seizures. There were some patients who had exclusively partial seizures with poor outcome.

DRAVET SYNDROME OR GEFS+?

22
11th ISS

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The discovery that mutations of the sodium channel alpha 1 subunit gene, *SCN1A*, underlie both simple febrile seizures and Dravet syndrome has been the most important observation in epilepsy genetics to date. The increasing spectrum of phenotypes associated with *SCN1A* mutations makes it difficult to predict outcome; detailed clinical assessment remains essential to interpret *SCN1A* results. *SCN1A* mutations are found in benign phenotypes including classical febrile seizures (FS), and febrile seizures plus (FS+). *SCN1A* mutations typically occur in the setting of the familial epilepsy syndrome of Generalized Epilepsy with Febrile Seizures Plus (GEFS+). In GEFS+, heterogeneous phenotypes are characteristically seen, ranging form mild disorders such as FS and FS+, to severe phenotypes including Myoclonic-Astatic Epilepsy (Doose syndrome) and Dravet syndrome. *SCN1A* mutations in GEFS+ comprise missense mutations.

Dravet syndrome begins with febrile status epilepticus at around 6 months of age, and may be hemiclonic or generalized. Fever is a prominent trigger of further convulsive attacks. 70-80% of children with Dravet syndrome have *SCN1A* mutations. Recently, we identified new *SCN1A* phenotypes including Severe Infantile Multi-Focal Epilepsy and some patients with Cryptogenic Generalized Epilepsy. Mutations in Dravet syndrome arise de novo in 95% cases; 40% are truncation and 40% missense mutations. Copy number variation accounts for 10% of cases negative by conventional mutational analysis. Ongoing work in genotype-phenotype correlation will help in understanding phenotypic variability with *SCN1A* mutations.

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11th ISS

SMEI: A CLINICAL AND GENETIC STUDY OF 38 ITALIAN PATIENTS

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OBJECTIVE: a) to identify the early electroclinical manifestations b) to evaluate cognitive outcome.
METHODS: 38 patients clinically followed and genetically screened by DHPLC and MLPA.
RESULTS: onset (mean age: 6.1 months), was marked by isolated seizure in 17 (during fever in 9) and by epileptic status in 21 (during hyperthermia in 13 patients; EEG was normal in all cases. During the second year of life drug-resistant seizures recurred , mostly triggered by fever, hot bath, intermittent lights; epileptic activity and/or EEG photosensitivity appeared. After the second year of life arrest of psychomotor development was evident in 27 patients, with behaviour disorders in 21. Twenty-two patients carried mutations of SCN1A gene.
CONCLUSION: SMEI tentative diagnosis may be proposed within the first year of life in an infant with febrile status, or recurrent focal seizures and normal EEG. A definite diagnosis is established during the second year given the seizure-favouring factors, EEG photosensitivity and psychomotor slowing. The high seizure frequency and the onset of cognitive impairment relationship support the role of epileptic activity in the final outcome.

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11th ISS

MOLECULAR BASIS OF SEVERE MYOCLONIC EPILEPSY IN INFANCY

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Severe myoclonic epilepsy (SMEI) is caused by mutations of the SCN1A gene that encodes voltage-gated sodium channel alpha-1 subunit. Recently, we generated and characterization of knock-in (KI) mice with an *SCN1A* nonsense mutation that appeared in three independent SMEI patients (Ogiwara et al., *J Neurosci* 27:5903-5914, 2007). The *SCN1A*-KI mice well reproduced the SMEI disease phenotypes. Both homozygous and heterozygous knock-in mice developed epileptic seizures within the first postnatal month. In heterozygous knock-in mice, trains of evoked action potentials in these fast-spiking, inhibitory cells exhibited pronounced spike amplitude decrement late in the burst but not in pyramidal neurons. We further showed that in wild-type mice the Nav1.1 protein is expressed dominantly in axons and moderately in somata of parbalbumin-positive inhibitory interneurons. Our immunohistochemical observations of the Nav1.1 are clearly distinct to the previous studies, and our findings has corrected the view of the Nav1.1 protein distribution. The data indicate that Nav1.1 plays critical roles in the spike output from PV interneurons and further, that the specifically altered function of these inhibitory circuits may contribute to epileptic seizures in the mice. These information should contribute to the understanding of molecular pathomechanism of SMEI and to develop its effective therapies.

Abstracts - Posters

EPIDEMIOLOGY OF FEBRILE SEIZURES AMONG RURAL BANGLADESHI CHILDREN

POSTER 1 11th ISS

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Objectives: To determine the prevalence of febrile seizures among disadvantaged rural children.
Methods: 4005 children aged 2-9 years were screened for functional disabilities in their homes using the Ten Questions (TQ). All screened positive were brought in for neurodevelopmental assessment (NDA).
Results: 400 (10%) children were positive for seizures and 707(17.7%) for =>1 functional deficit (motor, vision, hearing, speech, cognition, behavior, seizure). NDA was done on 1018 children. 222 (5.5%) parents had a 'seizure worry' of whom 181 (4.5%) had infections with the first seizure. 195 (4.9%) had a h/o febrile seizures (FS). Mean age of first occurance of FS was 23 (range 2-74) months, mean number of fits was 3.77 (range 1-24) times, mean duration was 1.37 (range 1-8) minutes. Associated neurodevelopmental impairments were of children with FS included: 53 (25.6%) behavior, 27 (13%) hearing, 21 (10.6%) speech, 13 (6.3%) cognitive, 13 (6.3%) vision, 11 (5.3%) gross motor, 4 (1.9%) fine motor deficits.
Conclusions: Seizures were the commonest neurological disorder found in a population-based survey. Infections were the major precipitating factor for the first seizure. Early recognition of FS and their expeditious management can prevent disability. An algorithmic system of referral from primary to tertiary care is needed.

FEBRILE CONVULSION : A SURVEY ON 300 CHILDREN

POSTER 2 11th ISS

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Background: Febrile convulsion (FC) is the most common convulsive disorder in children , occurring in 2 to 4% of the pediatric population .
Material & Methods: This study was conducted on 300 children with FC at the pediatric Neurology Department of Mofid Children Hospital between June 2001 – June 2007.
Pediatric demographic data including age, sex, type and duration of seizure , the interval between fever onset and occurrence of seizure , family history of FC and epilepsy, Neurodevelopmental delay , abnormal neurologic exam were recorded in questionnaires. Findings were statistically analyzed using T-test and Fisher's exact test.
Results: Recurrence was observed in 62 children (20.5 %) out of the 300, being most common in patients aged less than 18 months 26 children (41.9 %).
Recurrence rates among children with a positive family history of FC was (25.8%) and among children with positive family history of epilepsy was (19.4%) and among children with presence of complex FC was (22 %) respectively.
Recurrence rate among children with neurodevelopmental delay and abnormal neurologic exam was (24.2%). The presence of afebrile seizures in these patients were (10 %). From among those children with "a less than one hour" interval between fever onset and occurrence of seizure recurrence occurred in (71%) while in those with a "more than one hour interval" (17.7 %) experienced recurrence.
In 2.6% of our patients seizure was prior to fever.
Conclusion: Recurrence rate are increased by certain factors including age below one year , positive family history of FC and epilepsy , abnormal neurologic exam and neurodevelopmental delay and less than one hour interval between time of fever onset and occurrence.

POSTER 3 11th ISS

SEIZURES WITH FEVER BEYOND 5 YEARS OF AGE

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Objectives: Febrile seizures (FS) typically occur in children whose age between 2 and 5 years. We investigated the clinical characteristics of the patients experiencing seizures with fever beyond 5 years of age.
Methods: A total of one hundred and one patients (64 males and 37 girls) were included. Medical recordings including detailed history on the previous FS were investigated retrospectively.
Results: The mean onset age of FS was 3.0 years (0.3-8.3 years). Neurological abnormalities were detected in 11 patients (11%). In 16 patient (16%), at least one complex features were noted. Family history of FS or epilepsy was found in 55% (33/60). Among the 80 EEGs performed, abnormalities were observed in 30 recordings. Twenty three patients (23%) experienced their first seizures with fever beyond 5 years of age, in whom, neurological abnormality was more frequent (22% vs. 4%) and family history was less frequent (25% vs. 58%). In 53 patients (52%) whose ages at the last seizure were 6 years or older, mean onset age of FS was younger, and complex features and EEG abnormalities were more frequent.
Conclusions: The patients experiencing seizures with fever beyond 5 years of age may have poor prognosis than the children with typical FS.

POSTER 4 11th ISS

A CLINICAL STUDY ON COMPLEX FEBRILE SEIZURES

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Objectives: Febrile seizures can be classified as simple or complex type, the latter being characterized by increased risk of recurrence of febrile seizures itself and progression to epilepsy. The present study was conducted to delineate the clinical characteristics of complex febrile seizures.
Methods: Between January 2003 to December 2006, 550 children were diagnosed as febrile seizures. Their medical records were retrospectively reviewed for comparison between simple and complex febrile seizures and further depicting clinical findings of complex febrile seizures.
Results: Simple febrile seizures were 432 cases, four-fold larger than complex febrile seizures (118 cases). The causes of their febrile illness were upper airway infection (64.2%), pneumonia (10%), acute gastroenteritis (6.7%), and otitis media (3.6%). We did not find any significant difference between simple and complex febrile seizures in most clinical parameters. Regarding subtypes of complex febrile seizures, prolonged seizures were the most frequent (72.0%), followed by repeated seizures (16.9%) and focal seizures (5.1%).
Conclusions: Although febrile seizures is known as benign, some view it as a basic type of epilepsy caused by fever. An understanding of the natural history and prognosis will enable the physician to reassure the families and provide appropriate counseling and management while avoiding unnecessary diagnostic and therapeutic intervention.

CHARACTERIZATION OF THE GENE EXPRESSION PATTERN UPON DOUBLE-STRANDED RNA STIMULATION IN FEBRILE SEIZURES

POSTER 5 11th ISS

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Objectives: Previously, we showed increased interleukin(IL)-1β production upon double-stranded RNA(dsRNA) stimulation in febrile seizure(FS) patients. To know the genetic background of the immune response, we examined the gene expression upon dsRNA stimulation in FS patients.
Methods: Blood samples were obtained from children with FS and high IL-1β production capacity (n = 4) and control children (n = 2). The isolated leukocytes were stimulated with poly (I:C). RNA was extracted, and differential gene expression was examined using DNA microarray.
Results: On one hour stimulation, Na channel, K channel, and GABA receptor ε subunit(GABRE) were increased in both the FS and the control group. For cytokines, IL-26, IL-11, TNF and chemokines were increased in both groups. On 8 hours stimulation, GABRE and RIPK2 were increased only in the FS group. For cytokines, IL-1α, β and IL-8 were also increased in both groups. In comparison between the FS and the control group, Na channel (ACCN4), K channel (KCNC3), GABRE, RIPK2, TLR4, IL-26, TNF, and caspase-1 were increased in the FS group.
Conclusion: 1. Several cytokines other than IL-1β were also increased in the FS group. 2. Increased expression of RIPK2 and caspase-1 in the FS group might be associated with increased IL-1β production. 3. dsRNA induced expression of several iron channels, which might affect the neuronal excitability.

LACK OF ASSOCIATION BETWEEN A POLYMORPHISM IN SYN2 WITH GENETIC SUSCEPTIBILITY TO FEBRILE SEIZURES IN JAPANESE

POSTER 6 11th ISS

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Objectives: Febrile seizures (FS) are the most common form of childhood convulsions. Genetic factors are known to play an important role in susceptibility to FS. We previously reported that interleukin-1 beta -511 T allele was associated with susceptibility to simple FS of sporadic occurrence (Kira. 2005). Recently, Cavalleri et al reported that a polymorphism in SYN2 gene contributed to the predisposition to FS in several populations (Cavalleri. 2007). Here, we investigated whether SYN2 gene was associated with the development of FS in Japanese.
Methods: We genotyped the single nucleotide polymorphisms in intron 5 of SYN2 genes (rs3773363) in 249 FS patients (186 simple and 63 complex FS) and 225 controls by a TaqMan platform. The p-values of genotype or allele frequencies were calculated by means of chi-square analysis.
Results: The genotype frequencies of rs3773363 (GG, GA, AA) were 0.26, 0.50, 0.24 in controls, 0.33, 0.38, 0.28 in simple, 0.27. 0.40, 0.33 in complex, and 0.32, 0.39, 0.30 in all FS patients. The frequency of the A allele was 0.48 in controls, 0.48 in simple, 0.53 in complex and 0.49 in all FS patients. There were no significant differences between the groups.
Conclusions: These results suggest that SYN2 gene is not associated with the genetic susceptibility to FS in Japanese.

POSTER 7 11th ISS

SEIZURE-INDUCING EFFECT OF HISTAMINE H1 RECEPTOR ANTAGONISTS ON FEBRILE SEIZURES

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Objective: The histaminergic neuron system are known to exert anticonvulsive effect through the histamine H1 receptors. Experimental studies also indicated that H1 receptor antagonists (antihistamines) increase the seizure duration especially during developing period. We examined the seizure-inducing effect of antihistamines in patients with febrile seizures (FS).
Methods: Forty-nine patients with FS were assigned to non-administered or antihistamines administered group. The time between pyrexia and seizure onsets, the duration, the type of seizure, and electroencephalogram (EEG) were assessed.
Results: In the administered group, the seizure showed an earlier onset and longer duration. There were no significant differences in sex, age, cause of pyrexia and the family history of FS / epilepsy between the both groups. There were no significant differences in the type of seizures and the frequency of EEG abnormalities. However, in administered group, localized EEG abnormalities were noted particularly in patients under the age of 3.
Conclusion: Our results indicate that the antihistamines may be involved in prolongation of the seizure duration and in shorten the time between pyrexia and seizure onsets. In the use of antihistamines to infants, the ones which are less likely to pass the blood-brain barrier are more recommended.

POSTER 8 11th ISS

OXIDATIVE DNA DAMAGE IN CHILDREN WITH PROLONGED FEBRILE SEIZURES

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Objectives: Reactive oxygen intermediates can damage various forms of biomolecules, such as DNA, lipids, and proteins in human. Urinary and cerebrospinal fluid (CSF) levels of 8-hydroxydeoxyguanosine (8-OHdG) were measured the relevancy of oxidative stress to the disease in children with prolonged febrile seizures (PFS).
Methods: Urinary and CSF (U&C) 8-OHdG levels were measured in 12 children with PFS and in healthy children. The relationship between U&C levels of 8-OHdG was determined in 5 children where both U&C samples were available. The measurement of urinary 8-OHdG was conducted with an ELISA Kit and CSF 8-OHdG was analyzed by a computer-controlled HPLC analyzer.
Results: The levels of U&C 8-OHdG in children without brain damage associated with PFS showed no significant deference compared to healthy children. However, the levels of U&C 8-OHdG in children with brain damage associated with PFS were significantly higher than that of healthy children. A positive correlation between the levels of U&C 8-OHdG was noted in 5 children with both U&C samples were available.
Conclusions: These results suggested that oxidative stress was strongly related to the acute brain damage in children. We disclosed that U&C 8-OHdG was a useful marker of oxidative stress in children with brain damage associated with PFS.

OXIDATIVE STRESS BIOMARKERS IN FEBRILE SEIZURES

POSTER 9 11th ISS

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Objective: The aim of the present study was to examine the involvement of oxidative stress in febrile seizures.
Methods: Patients with febrile seizures (n=36) and age-matched controls (n=12) were included in this study. Cerebrospinal fluid (CSF) levels of 8-hydroxy-2-deoxyguanosine (8-OHdG), hexanoyl lysine adduct (HEL) and acrolein adduct (ACR) were measured using ELISA techniques.
Results: In 16 of 36 patients with febrile seizures, CSF levels of 8-OHdG were elevated. In addition, there was significant difference of the average levels of 8-OHdG between patients with febrile seizures and controls ($p<0.05$, Mann-Whitney's U test). CSF HEL levels were elevated in 4 of 33 patients with febrile seizures and 2 out of these 4 patients had high levels of CSF 8-OHdG. Another 2 patients with febrile seizures had high levels of CSF ACR. Importantly, elevated 8-OHdG was associated with the frequency of seizures.
Conclusions: Our results suggest that oxidative stress may partly contribute to the development of febrile seizures.

EXPRESSION LEVELS OF ICAM-1 AND LFA-1 IN PLASMA AND PBMC OF CHILDREN WITH FEBRILE SEIZURES

POSTER 10 11th ISS

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Objective: To explore the neuroimmunomodulation mechanism of ICAM-1 and LFA-1 in children with febrile seizures (FS).
Methods: 60 children with FS were divided into simple FS (SFS) group in 30 cases and complex FS (CFS) group in 30 cases, and control group of 30 health children was matched with regard to age and sex. Adopting enzyme-linked immunosorbentassay (ELISA) and flow cytometry (FCM), we measured the levels of ICAM-1 and LFA-1 contained in plasma and peripheral blood mononuclear cell (PBMC) of children with FS and control group.
Results: The plasma levels of soluble ICAM-1 (sICAM-1) in SFS group [(21.54±11.09)ng/ml] and CFS group [(24.34±6.86)ng/ml] were significantly lower than that in the control group [(29.73±12.39)ng/ml]. The plasma soluble LFA-1 (sLFA-1) levels of CFS, SFS and control groups were (12.30±8.04), (12.09±8.83) and (9.51±8.07) ng/ml respectively, but there was no statistical difference between various groups. The PBMC levels of ICAM-1 were significantly higher in SFS group [(29.96±12.31)%] than those in CFS group [(22.50±8.19)%] and control group [(14.21±11.31)%], at same time, there was significantly higher expression in CFS group than in control group. However, the expression levels of LFA-1 was different from the ICAM-1 in the surface of PBMC. Of the three groups, the highest LFA-1 level was the SFS group [(50.89±21.36)%], the lowest LFA-1 level was the CFS group [(34.35±11.45)%] and (41.39±16.30)% was the LFA-1 level of control group in PBMC. Significant difference was found in three groups.
Conclusions: As a kind of costimulatory immunologic molecule under early stress, ICAM-1/LFA-1 participate in leukocytic adherence and cascade reaction of adherence, which results in overexcitation of neurons on base of unadapting to fever stress and induces convulsive seizures. The process of neuronal immunopathology in CFS patients is more complex than that in SFS patients. Therefore, a new idea of neuroprotective therapy may be found by inhibiting the activity of ICAM-1/LFA-1 adhesion in FS.

POSTER 11 11th ISS

DIFFUSION WEIGHTED IMAGE ABNORMALITIES AND GLUCOSE HYPOMETABOLISM IN PATIENTS WITH PROLONGED FEBRILE SEIZURES -PARTIAL VOLUME CORRECTION STUDY-

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Objective: There is continued debate on whether prolonged febrile seizures (PFS) can cause hippocampal damage leading to temporal lobe epilepsy. The objective of this study was to assess metabolic abnormalities of the limbic circuitry using FDG-PET in children who had diffusion weighted image (DWI) abnormalities in hippocampus after PFS.
Methods: DWI showed unilateral hippocampal hyperintensity in three of 12 patients after PFS. We performed FDG-PET in these three patients two years after PFS. Radioactivity in each anatomical area was measured and partial volume correction (PVC) was performed to correct the effect of atrophy by MRI-based geometrical transfer matrix method. Asymmetry index (AI) for the value of each homologous pair was calculated.
Results: FDG-PET revealed hypometabolism in the hippocampus ipsilateral to the side of DWI abnormalities even after PVC in all patients and AIs were 13-25 %.
Conclusions: In children who had DWI abnormalities in hippocampus after PFS, functional abnormality in the limbic system is already seen in the early stage after PFS.

POSTER 12 11th ISS

RISK FACTORS OF UNPROVOKED SEIZURES AFTER ACUTE SYMPTOMATIC SEIZURES IN CHILDREN

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Objectives: Acute symptomatic seizure is defined as a temporary seizure with acute systemic, metabolic, or toxic insult in association with an acute central nervous system insult. We studied the risk factors of unprovoked seizures after acute symptomatic seizure in children.
Methods: We retrospectively reviewed the records of 110 children with acute symptomatic seizures who were admitted between January, 1998 and December, 2003.
Results: We analyzed records of 110 children with acute symptomatic seizures aged from 1 month to 17 years. 24 children had unprovoked seizures (21.8%) after acute symptomatic seizures. Causes in order of frequency were encephalopathy, central nervous system infection, brain tumor, cerebrovascular disease. The risk of unprovoked seizure was significantly greater for those with status epilepticus (68.4%) than without status epilepticus, with partial seizure (64.7%) than generalized seizure. And the risk of unprovoked seizure was strongly associated with abnormal finding of electroencephalogram (79.1%) and neuroimaging (41.6%).
Conclusions: The leading cause of subsequent unprovoked seizure was encephalopathy and age specific incidence was high in the group aged 24-72 months. The risk for subsequent unprovoked seizure was greater for those with partial seizure, status epilepticus, abnormal finding of neuroimaging and electroencephalography.

BINGE DRINKING DURING PREGNANCY AND RISK OF SEIZURES IN CHILDHOOD

POSTER 13 11th ISS

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Objective: We examined whether binge drinking during specific gestational time windows increases the risk of neonatal seizures, epilepsy, and febrile seizures.
Methods: We conducted a population-based cohort study of 80,526 singletons from the Danish National Birth Cohort (DNBC, 1996-2002). We obtained information on binge drinking (intake of five or more drinks on a single occasion) by computer assisted telephone interviews. Children diagnosed with neonatal seizures, epilepsy, or febrile seizures were retrieved from the Danish National Hospital Register.
Results: All binge drinking episodes combined during pregnancy were not associated with an increased risk of seizure disorders, but children exposed to binge episodes between 11 and 16 gestational weeks had an increased risk of neonatal seizures (Incidence Rate Ratio, 3.15; 95% CI, 1.37-7.25) and epilepsy (1.81, 1.13-2.90).
Conclusion: Our finding suggests that a high alcohol intake during pregnancy may have a time specific effect on the risk of some seizure disorders in the offspring.

GLUTARIC ACIDURIA TYPE I WITH ATYPICAL CLINIC FEATURES IN CHILDREN: A CASE REPORT

POSTER 14 11th ISS

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Background: Glutaric aciduria type I(GA I) is an autosomal recessive inborn error of metabolism, due to a deficiency of the enzyme glutaryl-CoA dehydrogenase(GCHD), which gives rise to an accumulation of glutaric and 3-hydroxyglutaric acids. GA I usually presents with an acute encephalitis-like metabolic crisis during infancy, leading to a severe dystonic syndrome. We report a Taiwanese children of GA I with heterozygous mutation in GCHD gene and presented atypical clinic features.
Clinical details: A 14-year-old female, who had intracranial hemorrhage at her infant stage, and had been diagnosed as cerebral palsy, suffered from spastic diplegia, dysarthria, dytonia and choreoathetosis since her childhood. She had neither macrocephaly nor tremor. Neuropsychological testing showed an average intelligence. Urinary organic acids were analyzed by gas chromatography–mass spectrometry confirmed GA I. Brain MRI showed increased signal intensity over the bilateral periventricular white matter and dorsal putamina seen on T2WI and widened bilateral sylvian fissures. No ventriculomegaly. Molecular analysis of the coding region of GCHD gene revealed a compound heterozygosity: a common mutation of IVS 10-2 A-to-C and a novel mutation C-to-A at c873.
Conclusion: The clinical picture and course of glutaric acidemia type I was variable. We suggest systematic organic acids chromatography to children who have acute or progress dystonia combined with basal ganglia abnormalities on MRI.

POSTER 15 11th ISS

FEBRILE SEIZURES IN CHILDREN WITH CEREBRAL PALSY

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Objectives: Although epilepsy frequently develops in children with cerebral palsy (CP), little is known about febrile seizures (FS) in these children. The aim of this study is to describe the prevalence and characteristics of FS in a population of children with CP.
Methods: A retrospective analysis was performed on FS among children with CP who made their first visit to us between 1991 and 2000.
Results: We studied 196 children with CP. Twenty-seven (13.8%) had FS and 86 (43.9%) had epilepsy. There were no differences in the etiology and the type of CP, or the level of gross motor function between the children with FS and those who had neither FS nor epilepsy. Thirteen of the 27 children with FS did not have any afebrile seizures, while the remaining 14 developed epilepsy after FS. Patients with both FS and epilepsy had significantly more complex FS including status epilepticus than those with FS only .
Conclusions: CP was a risk factor not only for epilepsy but also for FS. About half of the children with FS developed epilepsy. Development of complex FS increased the risk for subsequent epilepsy.

POSTER 16 11th ISS

MUTATION OF SODIUM CHANNEL BETA 1 SUBUNIT (SCN1B) IN GEFS+

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Background : The gene encoding the sodium channel beta 1 subunit(SCN1B) was the first gene identified for generalized epilepsy with febrile seizures plus(GEFS+). It was reported that gene locus for GEFS+ exist in chromosome 19q13.1, and has relationship with a 387 C → G mutation in the SCN1B gene. This study is to determine whether there is the 387 C → G mutation in the children with GEFS+ and simple febrile seizures(FS)
Methods : The sample group consisted of 22 patients with GEFS+ and 15 patients with FS who were diagnosed by our department of pediatrics. The control group consisted of 15 children who do not have seizure disorders. Genomic DNA was extracted from peripheral blood and a segment of the SCN1B exon 3 was amplified by PCR technique. Purified PCR products were treated with restriction enzyme, Hin P1. The restriction pattern was analyzed by sequencing analysis.
Results : Sixty eight%(15 of 22) patients with GEFS+ had family history for epilepsy, and epilepsy phenotypes were generalized tonic-clonic seizures in 82%(18 of 22), on the other hand 13.6%(3 of 22) and 4.4%(1 of 22) had absences and atonic seizures respectively. In this study, however we could not observe a 387 C → G mutation of the SCN1B in the children with GEFS+ and febrile seizures.
Conclusion : The gene for GEFS+ may have a heterogenetic characteristics, and there may be racial differences in the mutation frequency. Expanded studies involving large number of different families are required.

POLYMORPHISM OF SODIUM CHANNEL α SUBUNIT TYPE 1 (SCN1A) AND CLINICAL MANIFESTATION OF GENERALIZED EPILEPSY WITH FEBRILE SEIZURE PLUS (GEFS+) SPECTRUM IN CHILDREN

POSTER 17 11th ISS

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Objectives: To know the clinical manifestation and polymorphism of SCN1A of GEFS+ spectrum in children in Indonesia.
Methods: We examined 20 patients of GEFS+ spectrum consisting of 4 patients with febrile seizure plus, 9 patients with GEFS+, 3 patients with severe myoclonic epilepsy borderline (SMEB) and 4 patients of severe myoclonic epilepsy in infancy (SMEI) in Pediatric Department, Sardjito Hospital, Yogyakarta, Indonesia. Polymerase chain reaction was used to identify the polymorphism of the SCN1A gene and was followed by sequencing examination. Sofar, we screened SCN1A in 6 patients.
Result: Twenty patients (16 boys, 4 girls) aged 1 to 12.4 years (mean age 5.7 years). The median age at onset was 1 year. Single Nucleotide Polymorphism (SNP) in the voltage-gated sodium channel subunit gene SCN1A was identified in 3 patients with febrile seizure plus (FS+), GEFS+, and SMEB in the same location. The locations were in intron 2 (c.392+52T>C) and intron 7 (c.1028+21T>C=rs1542484).
Conclusions: SNP locations have actually been identified in other countries in the world. However, they have not been investigated yet in Indonesia.

CLINICAL FEATURES OF GEFS+ AND ITS NEUROLOGICAL OUTCOMES

POSTER 18 11th ISS

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Objectives: The aim of this study was to characterize the clinical and electroencephalographic features as well as the neurological outcome of GEFS+(Generalized Epilepsy with Febrile Seizure Plus).
Methods: We evaluated 55 children with GEFS+ who admitted at the Department of Pediatrics, Kangnam Sacred Heart Hospital from 1993 to 2004. We formed them into two groups by age of first febrile seizure; Group A(<6 years) and Group B(≥ 6 years). We analyzed the clinical features, electroencephalographic findings and neurological outcomes of the subjects.
Results: The mean age of initial febrile seizures of 55 subjects was 3 years and 9 months. Among the 55 subjects, 41 subjects had their initial febrile seizures under 6 years of age; while 14 subjects had them after age 6 years of age. Seventeen of the 55 subjects had family history of convulsion. The mean frequency of convulsion is 4.4 and the type of convulsion was mainly generalized. Nineteen(37.3%) showed abnormal finding on EEG and 23 subjects(41.4%) were treated with antiepileptic drugs(AEDs) for long term prophylaxis. Never the less, there was no subjects of abnormalities in neurological outcomes. The group with initial seizure occurred under 6 years of age had more family history of convulsion, higher frequency of total seizures, febrile seizures, and are administered with AEDs longer than the other group.
Conclusion: In our study of GEFS+, the clinical features and electroencephalogram of tested subjects were various. While there were cases with a possibility of epileptic seizure or in need of long term administration of AEDs, few cases of neurological abnormalities or developmental delay were seen. The group of the initial febrile seizures under 6 years of age revealed more family history of seizure, higher frequency of seizures than the other group.

POSTER 19 11th ISS

A CASE OF CRYPTOGENIC LOCALIZATION RELATED EPILEPSY WITH SCN1A MUTATION SHOWING FREQUENT COMPLEX PARTIAL SEIZURES

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Background: In *SCN1A*, the neuronal voltage-gated sodium channel alfa 1-subunit gene, mutations are reported to have been identified in patients with generalized epilepsy with febrile seizures (FS's) plus and severe myoclonic epilepsy in infancy, etc.
Case: A boy who had been treated with anti-epileptic drugs of VPA, CBZ, CLB, and KBr because of repetitive FS's, had a first episode of sudden loss of consciousness at the age of 5 years. The episodes began with atonic posture and twitching of both palpebrae, continued several minutes to 2 hours, sometimes with automatism, and often ended to sleep. EEG showed frequent spikes in the left frontal and central region in interictal phase and fast rhythm of low amplitude in the occipital region at the onset of the seizure. Addition of PHT and ZNS has decreased frequency and duration of the seizures.
Conclusion: Lack of FS status may exclude the association of temporal sclerosis damaged by FS, and indicates direct association of *SCN1A* with localization related epilepsy.

POSTER 20 11th ISS

A PATIENT WITH RASMUSSEN ENCEPHALITIS AND SCN1A MUTATION

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Case report: In an 11-year old girl with Rasmussen encephalitis that is believed to be an autoimmune disease, mutation was detected in the *SCN1A* gene that encodes the neuronal voltage-gated sodium channel alpha1 subunit.
She had febrile seizures since 1 year 1 month of age, afebrile generalized tonic-clonic seizures since 2 years, and myoclonic seizures since 2 years and 6 months of age. Her development was initially normal but gradually delayed thereafter. At 5 years of age, she frequently had focal motor seizures with asymmetric tonic posturing.
At 8 years of age, right hemiparesis was evident, and atrophy of the left cerebral hemisphere was found in MRI. Her EEG showed left hemisphere-dominant background slowing and spikes.
The R1575C-*SCN1A* mutation was detected in the patient and her healthy father, and it indicated some relationship between the genetic seizure-susceptibility and the immunological abnormality in the pathophysiology of neurological disorders.

SEVERE MYOCLONIC EPILEPSY IN INFANCY (SME) WITH A MILD CLINICAL COURSE

POSTER 21 11th ISS

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Objective: SME, also known as Dravet syndrome, is one of the most severe epileptic syndromes afflicting infants. However, we have experienced borderline SME patients with better seizure and mental outcomes. We investigated this mild form of SME.
Method: The subjects were 17 children (M=7, F=10) meeting some of the SME criteria but who had a milder clinical course than that seen in typical SME. All 17 had drug-resistant, fever-sensitive seizures during the early clinical course. Eleven underwent *SCN1A* mutation analysis. We retrospectively investigated the clinical features and outcomes of these children.
Results: The onset ages, prevalences of fever-sensitivity and seizure refractoriness during infancy and early childhood did not differ from those of typical SME. There were fewer episodes of status epilepticus and fewer polymorphous seizure types than with typical SME and responses to KBr were better in most of our cases. The seizures ultimately decreased markedly in frequency before 10 years of age. Thus, the clinical course was generally milder than that of typical SME, and mental outcomes were also better. Only a mis-sense *SCN1A* mutation was found in 27.2% of our patients.
Conclusion: This distinct group of patients with borderline SME features and better seizure outcomes is between GEFS+ and typical SME in the conceptual framework of the *SCN1A* mutation syndrome.

RISK FACTORS FOR THE PREDICTION OF DRAVET SYNDROME BEFORE ONE YEAR OF AGE

POSTER 22 11th ISS

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Objective: Our aim was to search risk factors to predict Dravet syndrome before the first birthday based on the clinical characteristics of infants and the SCN1A mutation analysis.
Methods: Ninety-six patients who experienced febrile seizures before the age of one were enrolled. The patients were divided into two groups—the Dravet syndrome group (n = 46) and the non-Dravet syndrome group (n = 50). We compared the clinical characteristics before one year of age of the two groups. We undertook the SCN1Amutational analysis.
Results: An age of onset of febrile seizure ≤ 7 months, a total number of seizures ≥ 5, prolonged seizures, hemiconvulsions, partial seizures, myoclonic seizures, and hot water–induced seizures were regarded as significant risk factors for Dravet syndrome. SCN1A missense and truncatedmutations were detected significantly more often in the Dravet syndrome group than in the non-Dravet syndrome group.
Conclusions: These risk factors could help to predict Dravet syndrome before one year of age.

POSTER 23 11th ISS

DURATION OF CONSCIOUSNESS DISTURBANCE AFTER THE FIRST SEIZURE AND RANGE OF MRI LESIONS ARE ASSOCIATED WITH PROGNOSIS OF ACUTE ENCEPHALOPATHY WITH BIPHASIC SEIZURES AND LATE REDUCED DIFFUSION (AESD)

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Purpose: To find risk factor of sequelae of acute encephalopathy with biphasic seizures and late reduced diffusion (AESD). Background: AESD is a newly established disease entity as an encephalopathy syndrome characterized by biphasic clincoradiologic features and often leave sequelae.
Methods: Medical records and MRI were reviewed for three children with AESD consisting of 2 boys and one girl.
Results: One patient achieved a good consciousness improvement within one day after the first seizure lasting for 90 min. DWI-MRI of the patient on day 4 showed high signal intensity only in the subcortical white matter of the bilateral frontal areas. The patient remained mild mental retardation. In contrast, other two showed insufficient improvement of consciousness after the first seizure lasting for 75 and 35 min. DWI-MRI of the two on days 3 and 5 showed high signal intensities in the subcortical white matters of broad areas. Both had severe mental retardation.
Conclusion: The prognosis of AESD may associate with the duration of the consciousness disturbance after the first seizure and the size of MRI lesion.

POSTER 24 11th ISS

CLINICAL, EEG AND MRI FINDINGS ON PROLONGED FEBRILE SEIZURE AND ACUTE ENCEPHALOPATHY IN THE ACUTE STAGES

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Objectives: The differential diagnosis of prolonged febrile seizures (PFS) and acute encepha lopathy (AE) is important. We report clinical, EEG, MRI findings on PFS and AE in the acute stages.
Methods: (1) We prospectively investigated children they were clinically considered to be PFS or AE between November, 2006 and January 2007. (2) Prospectively enrolled AE cases all showed just mild delirium, so we also retrospectively studied severe AE (SAE) cases with status epilepticus.
Results: 10 cases of PFS, 8 cases of AE, 6 cases of SAE were eligible. 2 cases of AE had brief convulsions. All of AE and SAE cases had abnormal EEG findings, mainly included slow waves, while only 30% cases of PFS had abnormal EEG findings. MRI showed diffuse white matter lesion on one of AE and all of SAE cases. Three PFS cases had unilateral hippocampal lesions and they had exactly EEG abnormalities.
Conclusions: Acute phase recordings of EEG and MRI around the same times are useful to differentiate PFS and AE.

TRANSIENT SPLENIAL LESION IN FEBRILE SEIZURE

POSTER 25 11th ISS

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Background: Transient splenial lesions occur in many conditions. Although intramyelinic edema has been suspected to be as an underlying cause, the exact mechanism remains obscure. We present a case of febrile seizure, for which MRI disclosed a transient splenial lesion.
Clinical details: A 6-year-old boy developed a fever of 40 °C following a generalized seizure that lasted for 1 minute. His consciousness soon recovered. Neurological, laboratory and cerebrospinal fluid examination revealed no abnormal finding except elevation of CRP. Thus, febrile seizure was diagnosed. Three days later, diffusion-weighted MRI revealed a mild high-intensity lesion in the splenium of the corpus callosum.
Conclusion: Clinically, this patient was diagnosed as having a febrile seizure, however. MRI revealed a transient splenial lesion, which was also compatible with the mildest form of clinically mild encephalitis/ encephalopathy with a reversible splenial lesion. When a patient develops a febrile seizure and the MRI reveals some transient lesions in the brain, it is uncertain whether the condition is a febrile seizure or the mildest form of acute encephalopathy. Neuroimaging techniques have progressed rapidly and can reveal some abnormal lesions, that are not apparent with normal using conventional CT and MRI. There is no description of neuroimaging findings in febrile seizure. Thus, a definition of febrile seizure including neuroradiological findings is necessary.

A CASE OF KABUKI SYNDROME PRESENTING ACUTE ENCEPHALOPATHY WITH PROLONGED FEBRILE SEIZURES AND LATE REDUCED DIFFUSION (AESD)

POSTER 26 11th ISS

Nishimura A, Kurata K, Shiomi K, Tozawa T, Hasegawa T, Isoda K, Matsui F, Morimoto M
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Objectives: To show the specific clinical course of AESD, we report a girl case of Kabuki Syndrome.
Methods: A 4-year-old girl case with Kabuki Syndrome presented prolonged febrile seizure and followed by the MRI findings of abnormal subcortical white matter. Her clinical course and MRI findings were reviewed.
Results: On admission, her laboratory findings showed hyper-cytokinemia. Steroid pulse therapy was applied. On day2, diffusion weighted (DWI) MRI looked normal, however, on day 6, the DWI showed decreased diffusion in subcortical white mater. On day 50, cerebral atrophy became remarkable, but the reduced diffusion disappeared. No pathogens could be detected.
Conclusions: We must be very careful when we encounter a case with febrile seizure status. 5 or 6 days after the event, subcortical white matter injury become apparent if the diagnosis is AESD. We must conduct a multi-centered clinical research to determine when and what kind of therapeutic interventions we need to lead a better prognosis for this devastating disease.

POSTER
27
11th ISS

SEVERE MYCOPLASMA ENCEPHALITIS PRESENTING AS FEBRILE CONVULSION

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Objectives: Mycoplasma pneumoniae is a common etiologic agent of acute respiratory infection in children. Neurologic manifestations are the most frequent extrapulmonary complications of mycoplasma infection. But the outcomes of mycoplasma encephalitis have been variable and the treatment is still controversial. We report here a fully recovered case of severe mycoplasma encephalitis presenting as febrile convulsion.
Case presentation: A ten year-old boy had admitted due to fever and cough and he was diagnosed as acute pneumonia. His respiratory symptom has progressively worsened. At 10 days after admission, he showed semi-comatose mentality after two attacks of prolonged febrile seizures. The laboratory exam revealed the rising mycoplasma antibody's titer and CSF leukocytosis. Newly developed multifocal T2 hyperintense, T1 shortened, and gyral hyperenhanced cortical gray matter of both cerebral hemispheres with restricted water diffusion was noticed on brain MRI. His mentality had been semi-comatose for near 2 months. But he can attend the elementary school with his peer group after 1 year after the infection.
Conclusions: Mycoplasma encephalitis may be presented as febrile convulsion and the initial clinical manifestations may be very grave. But the results may not be always grave and a remarkable neurological improvement of the infected children can be expected.

POSTER
28
11th ISS

A JAPANESE CASE OF PROGRESSIVE ENCEPHALOPATHY WITH EDEMA, HYPSARRHYTHMIA AND OPTIC ATROPHY(PEHO) SYNDROME

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PEHO syndrome is a rare neurodegenerative syndrome first reported in 1991 in Finnish patients. Subsequently very few cases are reported from other countries. We describe a 3-years-old Japanese boy with hypotonia, profound psychomotor retardation, and severe epilepsy which initially started early in infancy.
He was delivered at 39 weeks with head circumference of 33cm. He had apnea and abnormal movement of extremities in neonate. He was admitted due to status epileptics at the age of 2 months. He had high arched palate, edema of his face and extremities, and clubfoot. The seizures were resistant to multiple anticonvulsants. EEG revealed hypsarrhythmia under the age of one year. Lack of visual fixation with optic atropy was also observed at the age of 8 months.
Serial MRI demonstrated progressive brain atrophy and dysmyelination predominantly in the cerebellum and brainstem.
The diagnosis of PEHO syndrome is supported by the typical symptoms and the lack of abnormalities in laboratory tests including rectal biopsy.

A CASE PRESENTING WITH PROLONGED FEBRILE CONVULSION HAVING NEWLY DEVELOPED HIPPOCAMPAL ABNORMALITIES

POSTER 29 11th ISS

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[2)] Department of Pediatrics, Sang-gye Paik Hospital, Seoul, Korea

Objectives: Mesial temporal sclerosis has been linked to prolonged febrile seizures. We report here a case with newly developed hippocampal abnormalities after prolonged febrile convulsion demonstrated by serial MRI.
Case presentation: A 25 month-old-year boy presented with recurrent tumbling after 10 days of febrile illness. At the first time of ER visit, he had no neurological abnormalities and laboratory exam was normal. But MRI revealed ill-defined T2 prolongation at the right putamen, caudate nucleus and left basal ganglia. The MRS of 5 days after the event showed decreased ill-defined T2 prolongation of the right putamen. After discharge, he suffered febrile illness for 6 days and revisited ER due to two focal, prolonged febrile seizures with right side eyeball deviation and clonic movement of right hand. We obtained repeated MRI after these prolonged febrile seizures. The follow-up MRI revealed abnormal T2 hyperintensity along left hippocampus and the previous T2 change of putamen and basal ganglia has disappeared.
Conclusions: We report here a case with newly developed hippocampal abnormalities after prolonged febrile, focal seizures. Longitudinal follow-up with more cases is needed to demonstrate the clear sequence of febrile seizure and hippocampal sclerosis.

THE CLINICAL CHARACTARIZATION OF FEBRILE STAUTS EPILEPTICUS AND ITS TREATMENT IN INFANCY AND EARLY CHILDHOOD

POSTER 30 11th ISS

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Objectives: The clinical characterization of febrile status epilepticus (FSE) in infancy and early childhood was examined. An early and correct diagnosis might be essential for the treatment and long term prognosis.
Methods: 58 patients in infancy and early childhood, who showed febrile status epilepticus, with fever more than 38.0°C, prolonged for 30 minutes and more, or repeated within 30 minutes without full recovery of consciousness, and had been treated at our hospital between January 1999 and December 2007, were examined.
Results: The range of age was from 1 month to 6 years old, and their mean age and standard deviation was 2.4±1.8 years old. The follow-up period was 3.4±2.0 years. 58 patients showed 73 FSE. They were divided 6 groups, as follows; 1.epilepsy-a.idiopathic epilepsy, 7patients/19FSE, b.symptmatic epilepsy, '7patients/7FSE, 2.acute febrile status epilepticus, 17patients/19FSE, 3.theophylline related FSE, 7patients/8FSE, 4.CNS infections, 12patients/12FSE, 5.Benign infantile convulsion with mild gastroenteritis, 7patients/7FSE-cluster formations, 6.heat stroke, 1patients/1FSE. Recurrence of FSE was limited only in patients with SMEI and acute febrile status epilepticus who was followed by epilepsy with grand mal on awakening. Treatments were essentially consisted of diazepam, chloral hydrate, midazolam, pentobarbiturate and carbamazepine, and their efficacy was depended on the correct diagnosis of basic disorder and the appropriate selection of AED.
Concusions: An appropriate diagnosis might be essential for the efficacious selection of AED.

POSTER 31 11th ISS

CHILDHOOD STATUS EPILEPTICUS IN QUEEN SIRIKIT NATIONAL INSTITUTE OF CHILD-HEALTH (QSNICH): 5-YEAR REVIEW

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Objectives: To determine the prevalence and outcomes of childhood status epilepticus (CSE) in QSNICH.
Methods : The retrospective descriptive study was conducted during Jan.2002 to Dec.2006 for 5 years. The medical records were reviewed as follows: epidemiology, underlying diseases, medications, complications, mortality rates, causes of death.
Results : There were 57 cases with boys 31 cases (54.4%) and girls 26 (45.6%) respectively. The mean age was 53 months old. The underlying diseases were epilepsy 28 (49.1%), stroke 2 (3.7%), febrile seizures 2 (3.7%), and none 16 (28%). The presumed causes of CSE were epilepsy 22 (38.6%), CNS infection 11(19.3%), and first seizures 5 (8.8%). The mortality rate was 17.5% with mean age of 89 months old. The underlying diseases of death were epilepsy 3 (5.3%), SLE 1, NHL 1, sepsis 1, delayed development 1, and none 3 (5.3%).
Conclusions: The prevalence of CSE in QSNICH was 57 cases in 5 years. The most common cause was underlying epilepsy. The mortality rate was 17.5%.

POSTER 32 11th ISS

FEBRILE MYOCLONUS IN CHILDREN IS A BENIGN AND COMMON PHENOMENON

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Objectives: Febrile Myoclonus (FM) is a rarely reported phenomenon. The purpose of this study is to clarify the clinical features of FM.
Methods: Children with FM were analyzed prospectively at Kenwakai Hospital from 1994 to 2006.
Results: Fifty FM episodes of 36 children were observed.; 23 boys and 13 girls, aged 7 to 121 (23.3±19.4) months. In the family history, febrile seizure were noted in 28.1%, and epilepsy in 9.4%.The duration of FM was less than 6 hours in 78%,more than 13 hours in 14%. The times of FM were less than 10 in 62%.Seven cases had febrile seizures in 8 episodes, and 2 cases had fear in 4 episodes. EEG were examined in 16 cases and 2 cases showed epileptic discharges. Of 34 cases followed in 9 to 128 (47.1±35.9) months, recurrences of FM were seen in 10 cases, and epilepsy (BECCT) in one boy.
Conclusion: FM is a benign phenomenon as previous reports, and is more common than is recognized.

MOVEMENT-INDUCED SEIZURES OR PAROXYSMAL KINESIGENIC DYSKINESIA WITH ELECTROGRAPHIC ABNORMALITIES ?

POSTER 33 11th ISS

Kim YO, Woo YJ
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We report a 12-year-old boy who had the attacks of a few minutes occurring 10-20 seconds after such sudden movements as running and hopping. The attacks always involved unilateral limbs independently (right than left) and appeared more severely in upper limb. At the onset of attacks, he complained of weakness and odd sensation in the affected unilateral limbs. Then his head was deviated to the affected side. Coincidently he strongly resisted freeing the susceptible upper limb from involuntary movement by the other hand. During attacks he seemed clumsy although he remained conscious to answer shortly and move to commands. The attacks never occurred spontaneously, passively nor by imaging the motion. Brain MRI and interictal EEG showed no abnormal finding. But interictal SPECT demonstrated decrease in perfusion in both temporal and left parietal lobe. Video EEG showed burst of 2.5-3 Hz general spike and waves during rhythmic hopping with abduction and adduction of both arms, which were attenuated by EMG after stop of hopping due to involuntary movement. A small dosage of carbamazepine was very effective in stopping the attacks. His attacks have the following characteristics sufficient to be clinically diagnosed as paroxysmal kinesigenic choreoathetosis: kinesigenic attacks, dystonic posturing with odd sensations at onset of attacks, maintained consciousness during attacks, and effective carbamazepine in controlling attacks. But video EEG showed abnormal finding such as above during the provoking movement.

SHORT-TERM ANTICONVULSANT PROPHYLAXIS FOR REPEATED FEBRILE SEIZURE DURING THE SAME FEBRILE EPISODE

POSTER 34 11th ISS

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Objective: To compare the efficacy of anticonvulsant prophylaxis for repeated febrile seizure during the same febrile episode
Methodology: The randomized controlled trial was conducted. There were 322 children with febrile seizures hospitalized and were categorized into 3 groups as follows: (1) Oral diazepam 3 mg/kg/dose every 8 hours, (2) Phenobarbital 15 mg/kg/dose IM loading and 5 mg/kg/day orally divided every 12 hours, (3) No anticonvulsant as control. They were observed at least 48 hours for repeated seizure. Data analyses were performed using Chi-square test for categorical data and t-test for continuous data.
Results: There were boys and girls as 196 cases (61%) and 126 cases (39%) respectively. The most common cause of fever was common cold (63.7%). There were 48 cases (15%) with repeated seizure before hospitalization. The repeated seizures during the same febrile episode were 12 cases (3.7%) in diazepam group, 4 cases (3%) in phenobarbital group, and 4 cases (4.6%) in control, which were not statistically different. The odds ratio of diazepam and phenobarbital were 0.87 (95% CI 0.18-4.32) and 0.63 (95% CI 0.13-30.9) respectively. The Number Needed to Treat of diazepam and phenobarbital was 166 and 59 respectively.
Conclusions: Short-term diazepam and phenobarbital for repeated seizure in the same febrile episode were not statistically different compared to supportive care.

POSTER
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11th ISS

SUPPOSITORY DIAZEPAM TO PREVENT A RECURRENCE OF FEBRILE SEIZURES DURING A SINGLE FEBRILE ILLNESS

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Objectives: To assess the efficacy of suppository diazepam for the prevention of the recurrence of febrile seizures during a single febrile illness.
Methods: We studied 203 children with febrile seizures during December 2004 through March 2006. On admission, suppository diazepam was administered between December 2004 and May 2005. Patients between June 2005 and March 2006 were not treated with antiepileptic dugs on admission.
Results: There was a significant difference in the rate of a recurrence of febrile seizures between the children treated with diazepam and without treatment. A recurrence was observed in 2 of 95 children treated with diazepam and in 16 of 108 children without treatment. Among 108 patients without treatment, the median age was 22.8 months in those with a recurrence and 30.6 months in those without a recurrence, suggesting that a younger age was related to a recurrence.
Conclusions: Suppository diazepam after a febrile seizure will reduce the incidence of a recurrence of febrile seizures during the same febrile illness.

POSTER
36
11th ISS

DOES SUPPOSITORY DIAZEPAM PREVENT THE RECURRENCE OF FEBRILE SEIZURES DURING A SINGLE FEBRILE EPISODE? AN EMERGENCY ROOM STUDY

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Objectives: In Japan, prophylactic diazepam treatment is used to prevent febrile seizures (FS), following the guidelines proposed by Fukuyama et al. However, there is no consensus on how to treat emergency room patients in whom FS stop spontaneously. This study clarified the efficacy of suppository diazepam in the emergency room at preventing the recurrence of FS during a single febrile episode.
Methods: We retrospectively studied 465 eligible patients with FS admitted to our emergency room between May 2005 and April 2007. One (group D) received suppository diazepam and the other (group N) did not. Statistical analysis was performed using the Mann-Whitney U-test and chi-square test with $p<0.05$ considered significant.
Results: In groups D and N, FS recurred in 7 (3.7%) out of 187 and 32 (11.5%) out of 287 patients, respectively. There was a significant difference in the rate of recurrence, but no significant differences in the past history, family history, or FS recurrence.
Conclusions: Suppository diazepam after a FS reduces the rate of FS recurrence during a single febrile episode.

HIGH-DOSE LEVETIRACETAM IN REFRACTORY EPILEPSY WITH SCN1A MUTATIONS

POSTER 37 11th ISS

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Objectives: SCN1A mutations may underlie epilepsy syndrome such as generalized epilepsy with febrile seizures plus (GEFS+), and severe myoclonic epilepsy of infancy (SMEI). However, treatment efficacy in epilepsy with particular SCN1A mutations remains unclear.
Methods: Through retrospective review of medical records at Children's Hospital Boston, we identified two refractory epilepsy patients of SCN1A mutations, who responded to high-dose levetiracetam.
Results: Both patients had refractory frrequent generalized seizures, occasional status epilepticus, failed multiple antiepileptic drugs and ketogenic diet.
Patient 1 was a 3 year old girl, with SCN1A testing showed transition from C to T, changing arginine to stop codon (nucleotide position of 3733, codon 1245). Addition of levetiracetam, up to 286 mg/kg/day, resulted in significant decrease in seizures, to only one seizure in 11 months.
Patient 2 was a 6.5 year old girl with, with a history of infantile spasms. SCN1A testing showed heterozygous 31 basepair duplication causing frame shift (nucleotide 1054-1084, codon position 362). Addition of levetiracetam, up to 100 mg/kg/day, resulted in significant decrease in seizures, to three in 3 months.
Conclusions: High dose levetiracetam appeared effective in our cases of SCN1A mutations with refractory epilepsy. Larger scale studies are necessary to further assess reproducibility of the findings and efficacy of levetiracetam in SCN1A epilepsy syndromes.

RESECTIVE SURGERY FOR INTRACTABLE EPILEPSY IN INFANTS -COMPARISON TO ELDER CHILDREN-

POSTER 38 11th ISS

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Objective: Intractable epilepsy in infants is often characterized by its catastrophic nature and significant consequences on development so that early surgical intervention may be necessary although surgical prognosis for these children is not fully investigated.
Methods: Seventy-three consecutive intractable epilepsy children treated by resective epilepsy surgery were divided into four group according to the age at surgery; A (<12mo: n=15), B (1-5y: n=16), C (6-10y: n=17), D (11-15y: n=25) and etiology, surgical procedures, and prognosis were compared. Follow-up period was 1 to 14 years (mean 5.3y). Preoperative evaluation included video-EEG monitoring, PET, MEG and ictal SPECT. Intracranial EEG monitoring was performed in 19 patients (3y-15y, mean:10.2y). Resective procedures included hemispherotomy (9), multilobar resection/disconnection (8), temporal (23), frontal (17), parietal (10), occipital (5), and others (1). Pathological disgnosis was hemimegalencephaly (8), cortical dysplasia (36), tumors and vascular lesions (19), ulegyria (8), hippocampal sclerosis (3), and gliosis (4).
Results: In infants, extensive developmental pathologies (hemimegalencephaly and cortical dysplasia) were often demonstrated (A:82.6%, B:68.8%, C:52.9%, D:32%) and extensive resective procedures (hemispherotomy and multilobar resection/disconnection) were indicated (A:66.7%, B:12.5%, C:11.8%, D:12%). But surgical prognosis of resective surgery was as favorable (Engel class I,II) as other age groups (A:80%, B:75%, C:71%, D:80%).
Conclusion: Surgical prognosis of intractable epilepsy in infants, which often associates extensive developmental pathologies, can be as favorable as elder children by choosing appropriate resective surgical procedures.

POSTER 39 11th ISS

CLINICAL CHARACTERISTICS AND PROGNOSIS OF PATIENTS WITH FEBRILE STATUS EPILEPTICUS

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Objectives: This study is to clarify the incidence and general clinical characteristics and prognosis of patients with febrile status epilepticus(FSE),
Methods: From January 2004 to December 2006, 313 patients(160 boys and 153 girls) who was suffered from febrile seizure, admitted to the department of pediatrics, Eulji University Hospital. They were classified into FSE group(≥30 minutes) and FS group(< 30 minutes) by the duration of seizure. We retrospectively analyzed the patients' medical records for the clinical status at the time of diagnosis and in the follow-up priod after discharge.
Results: Of 313 children with FS, 21 patients (6.6%) experienced seizures longer than 30 minutes. We found no statistically significant difference in etiology, patients'age, prior history of neurological abnormality, family history of epilepsy, seizure type, duration and degree of fever between the two groups. MRI scannings showed negative results of the all patients. In the analysis of after the diagnosis, no death and one patient with mild weakness in the right arm was found among FSE group. Among 292 patients of FS group, no death or neurologia abnormality was found.
Conclusions: Febrile status epilepticus are not rare. The comparative analysis between FSE and FS groups revealed no significant differences on the clinical characteristics. The overall prognosis was also same and benign.

POSTER 40 11th ISS

EXECUTIVE FUNCTION DEFICITS IN CHILDREN WITH HISTORY OF FEBRILE SEIZURES

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Objectives: To determine the prevalence and associated factors of executive function deficits in Thai children with history of febrile seizures
Methods: From May 1st 2004 to April 30th 2005, a cross-sectional study was conducted in Thai children aged 7-15 years who had a history of febrile seizure diagnosed before the age of 6 at the Department of Pediatrics, Ramathibodi Hospital, Bangkok, Thailand. Computerized Wisconsin Card Sorting Test (cWCST) and Wechsler Intelligence Scales for Children-Third edition (WISC-III) were applied to evaluate executive function and intelligence quotient respectively.
Results: 52 children (mean age 10.7 years, SD =1.9) participated in this study. Mean full- scale IQ was 94.0 (SD= 14.4). Prevalence of executive function deficit consisting cognitive flexibility, conceptual level response, and learning to learn, were 23.1, 21.2, and 10.4 respectively. The overall prevalence of any domain dysfunction was 28.9. Seizure recurrence, education level, and the presence of soft neurological signs were associated with the deficits.
Conclusion: Executive function deficit in children with a history of febrile seizure despite having normal IQ is evident in this study. However, further studies in large number and of subjects with a control population is necessary for definitive conclusion.

PROGNOSIS OF THE INDIVIDUALS WITH FEBRILE SEIZURES ASSOCIATED WITH EPILEPTIC DISCHARGE

POSTER 41 11th ISS

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Objective: Epileptic discharge on electroencephalogram (EEG) is occasionally identified in patients with febrile seizures (FS). We studied the prognoses of FS with epileptic discharge.
Method: We recruited 36 patients who had FC with epileptic discharge and been followed up for three years and more after the last seizure. Those who had mental retardation and/or neurological abnormality had been excluded. The patients were classified into three groups; group I (7 with FS only), group II (10 with FS after six years of age and/or one afebrile convulsion) and group III (9 with subsequent epilepsies). The clinical features, EEG and prognoses of each group were reviewed and compared.
Result: Recurrent FS were observed in 25 patients (69.4%) and 9 (25%) had subsequent epilepsies. The seizure phenotypes of groups II and III during the follow-ups were afebrile convulsion, myoclonic and complex partial seizures. The diagnoses given to the patients of group III were benign idiopathic focal epilepsy for 8, among whom 5 were treated as BCECT. Only one had intractable epilepsy with frontal foci on EEG.
Conclusions: The frequencies of recurrent FS and subsequent epilepsies were high in the subjects. However, the subsequent epilepsies were well controlled.

A LONG-TERM FOLLOWING-UP STUDY ON BENIGN CONVULSIONS WITH MILD GASTROENTERITIS

POSTER 42 11th ISS

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Objective: To investigate clinical course of benign infantile convulsions associated with mild gastroenteritis (BICE), and to follow up the outcomes of studied infants.
Method: Long-term follow-ups for 24-40 months and assessments were carried out for 24 patients of BICE, admitted to our hospital between December,2002 to February,2004. The recurrence rate and prognosis were determined by including the cases with tendency of cluster seizures on clinical episodes, and by excluding those with electrolyte disorder, febrile seizures, epilepsy and epileptic syndrome, symptomatic epilepsy and abnormality of neuromotor development.
Results: Of the 24 patients, 4 patient showed small spike sharp in centric area on EEG, and 1 patient external hydrocephalus on CT. During the 24-month-or-more follow-ups, two cases were found to experience another episode due to gastroenteritis, and one to recur twice due to fever Antiepileptic(AEDs) were not administered for the followed-ups and their intelligence developed normally.
Conclusion: Infants with BICE may experience one to two convulsion episodes, and cluster seizures may occur in some cases, but the prognosis is well for all the infants.

POSTER 43 11th ISS

A CASE OF GLUCOSE TRANSPORTER-1 DEFICIENCY SYNDROME WITH EPILEPTIC SEIZURES BUT WITHOUT ATAXIA OR DEVELOPMENTAL DELAY

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Background: GLUT-1 deficiency syndrome is a metabolic disorder caused by a defect in glucose transport across the blood-brain barrier. Many patients present with epileptic seizures, developmental delay, and ataxia. We now report a case of GLUT-1 deficiency syndrome with epileptic seizures but without ataxia or developmental delay.
Clinical Details: A 6-year-old girl presented at age 8 months with generalized tonic-clonic seizures. They were associated with slight fever or exhaustion. EEG/CT/MRI were normal. Valproate was commenced, but her seizures remained intractable. At age 5 years, absence seizures appeared.
Clonazepam was added, but without improvement. Other AEDs also had no effect. At age 6 years, she was hospitalized. Physical and neurological examinations were normal. Since she was obese, she was started on a low-calorie diet. Subsequently, her absence seizures increased before meals, and GLUT-1 deficiency syndrome was suspected. Further examination revealed a low CSF/blood glucose ratio of 0.41. The basic rhythms of EEG were slow before meals, but became normal after. The glucose uptake of erythrocyte was found to be declined, confirming the diagnosis of GLUT-1 deficiency syndrome. After a week of ketogenic diet, she showed almost no epileptic seizures, and improvement in daily activities.
Conclusion: This is a rare case of GLUT-1 deficiency syndrome. The present case showed intractable seizures but no ataxia or developmental delay. There may be other cases of GLUT-1 deficiency with only intractable seizures.

POSTER 44 11th ISS

NEUROPSYCHOLOGICAL FEATURES OF ELEVEN PATIENTS WITH SEVERE MYOCLONIC EPILEPSY (DRAVET SYNDROME): PRELIMINARY RESULTS

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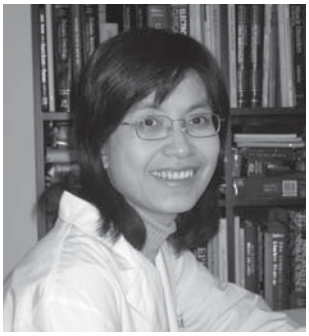
Objective: eleven infants with severe myoclonic epilepsy, admitted to the Unit of Child Neurology and Psychiatry, Catholic University of Rome, from 2000 to 2008, have been serially followed with a full clinical assessment including neuropsychological examination, long-term EEG monitoring, MRI and genetic investigations.
Methods: in six of them final observation was before the age of three (group 1), whereas in the remaining five the end of the follow-up was between 3 years and 8 years and 3 months (group 2). Neuropsychological assessment consisted of Griffiths' scales and a test for the first language; behaviour was evaluated by the Child Behaviour Check-list. Groupe 2 patients underwent also tests of specific cognitive functions.
Results: results show significant differences between the two groups in the sense that final scores of the first group was delayed but within the normal range while those of the second group were below normal limits, suggesting a progressive clinical worsening. Yet, no child present a severe mental retardation. Moreover, language scores were the lowest in group 1, whereas the lowest scores in group 2 regarded eye-hand items. These data are correlated with the evolution of epilepsy.
Conclusions: even though there is an heterogeneous outcome in our sample, in a part of cases a possible continuous developmental deterioration seems related to severity of seizures.

Profiles of Lecturers

Min-Lan TSAI



1

11th ISS

Present Position

Attending Physician of the Department of Pediatrics,
Cheng-Hsin Rehabilitation Medical Center, Taipei, Taiwan

Appointments

1991- 1992:	Research Fellow, Children's Hospital and Medical Center, and Research Fellow, Electroencephalography and Clinical Neurophysiology, University of Washington Medical Center, Seattle, Washington, USA
1992- 1995:	Attending Physician, Department of Pediatrics, Cathay General Hospital
1995- 1998:	Attending Physician, Department of Pediatrics, Show-Chwan Memorial Hospital
1999- 2007:	Senior Attending Physician, Department of Pediatrics, Chang-Hua Christian Hospital
2003- 2007:	Ph.D. student and Master degree in Graduate Program of Neuroscience, Department of Physiology and Pharmacology, University of Western Ontario, London, Ontario, Canada
2008-	Attending physician, Cheng-Hsin Rehabilitation Medical Center, Taipei

Selected publications

1. Tsai ML, Bixia Shen, Leung LS. Seizures induced by $GABA_B$ receptor blockade in early-life induced long-term kindling facilitation and $GABA_B$ receptor hypofunction. Epilespy Res, 2008, accepted.
2. Tsai ML, Leung LS. Decrease of hippocampal $GABA_B$-receptor mediated inhibition after hyperthermia-induced seizures in immature rats. Epilepsia 2006;47:277-287.
3. Tsai ML, Lo HY, Chaou WT. Clinical and electroencephalographic findings in early and late onset benign childhood epilepsy with occipital paroxysms. Brain & Development 2001;23:401-405.
4. Hung KL, Liao HT, Tsai ML. Postinfectious encephalomyelitis: etiologic and diagnostic trends. J Child Neurol 2000;15:666-670.
5. Tsai ML, Hung KL. Topographic mapping and clinical analysis of benign childhood epilepsy with-centrotemporal spikes. Brain & Development 1998;20:27-32.

2

11th ISS

Mogens VESTERGAARD

Present Position

General Practitioner, Låsby, Denmark
Associate professor, Institute of Public Health,
Departments of General Practice and Epidemiology,
Aarhus University, Denmark

Appointments

1996- 1998:	Rotation, Kolding Hospital, Denmark
1999- 2002:	PhD student, Perinatal Epidemiological Research Unit, Aarhus University Hospital
2002- 2003:	Assisting Professor, Institute of Public Health, Department of Epidemiology, Aarhus University Hospital
2004- 2008:	5-years formalized clinical training in Internal Medicine, Geriatrics, Surgery, Obstetrics/Gynecology, Psychiatry, and General Practice for the purpose of specializing as a General Practitioner, Aarhus University Hospital, Denmark
2006-	Associate professor, Institute of Public Health, Departments of General Practice and Epidemiology, Aarhus University, Denmark

Selected publications

1. Vestergaard M, Hviid A, Madsen K, Schendel D, Wohlfahrt J, Thorsen P, Melbye M, Olsen J. MMR vaccination and febrile seizures – evaluation of susceptible subgroups and long-term prognosis. JAMA. 2004. 292: 351-357
2. Vestergaard M, Pedersen CB, Sidenius P, Olsen J, Christensen J. The long-term risk of epilepsy after febrile seizures in susceptible subgroups American Journal of Epidemiology 2007 Apr 15;165(8):911-8.
3. Vestergaard M, Basso O, Henriksen TB, Østergaard J, Olsen J. Risk factors for febrile convulsions. Epidemiology 2002. 13(3): 282-7.
4. Christensen J, Vestergaard M, Sidenius P, Mortensen PB, Agerbo E. Epilepsy and risk of suicide. Lancet Neurology 2007 Aug;6(8):693-8
5. Madsen K, Hviid A, Vestergaard M, Schendel D, Wohlfahrt J, Thorsen P, Olsen J, Melbye M. MMR vaccination and autism–a population based cohort study. NEJM 2002. 347 (19) 1477-82

Junko NAKAYAMA

Present Position

Assistant Professor of the Department of Pediatrics,
Ibaraki Prefectural University of Health Sciences

Appointments

1992-1996 Resident in Department of Pediatrics, University of Tsukuba School of Medicine, Ibaraki, Japan
1996-2000 University of Tsukuba School of Medicine
Awarded the degree of Ph.D. in Doctor of Medical Science
2000-2003 Japan Society for the Promotion of Science (JSPS)
Post-doctoral Fellow in Department of Medical Genetics,
University of Tsukuba School of Medicine, Ibaraki, Japan
2003-2006 Post-doctoral Fellow in Department of Neurology, UCSF, San Francisco, CA
2006- Present position

Selected publications

1. Nakayama J, Arinami T: Molecular Genetics of febrile seizures. Epilepsy Res 70:190-198, 2006.
2. Nakayama J, Yamamoto N, Hamano K, Iwasaki N, Ohta M, Nakahara S, Matsui A, Noguchi E, Arinami T: Linkage and association of febrile seizures to the IMPA2 gene on human chromosome 18. Neurology 63:1803-1807, 2004.
3. Nakayama J, Fu YH, Clark AM, Nakahara S, Hamano K, Iwasaki N, Matsui A, Arinami T, Ptáček LJ: A nonsense mutation of the MASS1 gene in a family with febrile and afebrile seizures. Ann Neurol 52:654-657, 2002.
4. Nakayama J, Hamano K, Iwasaki N, Nakahara S, Horigome Y, Saitoh H, Aoki T, Maki T, Kikuchi M, Migita T, Oto T, Yokouchi Y,Tanaka R, Hasegawa M, Matsui A, Hamaguchi H, Arinami T: Significant evidence for linkage of febrile seizures to chromosome 5q14-q15. Hum Mol Genet 9:87-91, 2000.
5. Nakayama J, Miura M, Honda M, Miki T, Honda Y, Arinami T: Linkage of human narcolepsy with HLA association to chromosome 4p13-q21. Genomics 65:84-86, 2000.

4

11th ISS

Ryutaro KIRA

Present Position

Lecturer of the Department of Pediatrics,
Graduate School of Medical Sciences, Kyushu University

Appointments

1989- 1990: Resident, Fukuoka Children's Hospital
1990- 1991: Resident, Kyushu University Hospital
1991- 1992: Staff Doctor, Miyazaki Prefectural Hospital
1993- 1994: Staff Doctor, Kitakyushu Municipal Wakamatsu Hospital
1994- 1994: Clinical Fellow, Kyushu University Hospital
1994- 1994: Assistant Professor, Department of Pediatrics, Graduate School of Medical Sciences, Kyushu University
2007- present position

Selected publications

1. Ryutaro Kira (corresponding author), Hiroyuki Torisu, Megumi Takemoto, Akihiko Nomura, Yasunari Sakai, Masafumi Sanefuji, Kanji Sakamoto, Shigetaka Matsumoto, Kenjiro Gondo, Toshiro Hara: Genetic susceptibility to simple febrile seizures: interleukin-1beta promoter polymorphisms are associated with sporadic cases. Neurosci Lett 384:239-244, 2005.
2. Yasunari Sakai, Ryutaro Kira (corresponding author), Hiroyuki Torisu, Kenji Ihara, Takashi Yoshiura, Toshiro Hara: Persistent diffusion abnormalities in the brain stem of three children with mitochondrial diseases. AJNR Am J Neuroradiol 27:1924-1926, 2006.
3. Hiroyuki Torisu, Koichi Kusuhara, Ryutaro Kira, Bassuny WM, Yasunari Sakai, Masafumi Sanefuji, Megumi Takemoto, Toshiro Hara: Functional MxA promoter polymorphism associated with subacute sclerosing panencephalitis. Neurology 62:457-460, 2004
4. Vahid Khajoee, Kenji Ihara, Ryutaro Kira (corresponding author), Megumi Takemoto, Hiroyuki Torisu, Yasunari Sakai, Jia Guanjun, Park Myoung Hee, Katsushi Tokunaga, Toshiro Hara: Founder effect of the C9 R95X mutation in Orientals. Hum Genet 112:244-248, 2003.
5. Takehiko Inoue, Ryutaro Kira (corresponding author), Futoshi Nakao, Kenji Ihara, Bassuny WM, Koichi Kusuhara, Kenji Nihei, Kenzo Takeshita, Toshiro Hara: Contribution of the interleukin 4 gene to susceptibility to subacute sclerosing panencephalitis. Arch Neurol 59(5):822-827, 2002.

Brian NEVILLE

5
11th ISS

Present Position

2004- Present
The Prince of Wales's Professor of Childhood Epilepsy
University College London Institute of Child Health,
Great Ormond Street Hospital for Children,
and the National Centre for Young People with Epilepsy, Lingfield

Appointments

1989- 2004:	Professor of Paediatric Neurology
	Institute of Child Health, University of London
	Honorary Consultant, Paediatric Neurologist
	Great Ormond Street Hospital for Sick Children NHS Trust
1973- 1989:	Consultant Paediatric Neurologist and Director of the
	Newcomen Child Development Centre, Guy's Hospital
	Lecturer in Paediatric Research Unit
	United Hospitals Medical School, University of London
1970- 1972:	Senior Registrar
	National Hospital for Nervous Diseases and Great Ormond Street

Selected publications

1. Scott RC, King MD, Gadian DG, Neville BGR, Connelly A. Hippocampal abnormalities after prolonged febrile convulsion: a longitudinal MRI study. Brain 2003, 126 (11): 2551-2557.
2. Neville BGR, Parascandola R, Farrugia R, Felice A. Sepiapterin reductase deficiency: a congenital dopa-responsive motor and cognitive disorder. Brain 2005; 128: 2291-2296.
3. Banu SH, Jahan M, Koli UM, Ferdousi S, Khan NZ, Neville B. Side effects of phenobarbital and carbamazepine in childhood epilepsy: randomized controlled trial. British Medical Journal 2007 Jun; 49 (2): 85.
4. Sandarangani M, Seaton C, Scott AG, Ogutu B, Edwards T, Prins A, Gatakaa H, Idro R, Berkley JA, Peshu N, Neville BGR, Newton CR. Incidence and outcome of convulsive status epilepticus in Kenyan children: a cohort study. Lancet Neurology 2008 Jan, 2: 145-150.
5. Raspall-Chaure M, Neville BG, Scott RC. The medical management of the epilepsies in children: conceptual and practical considerations. Lancet Neurology 2008 Jan, 7 (1): 57-69.

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11th ISS

Kai KAILA

Present Position

Professor of Animal Physiology, University of Helsinki, Finland 1994-
Research Director, Neuroscience Center, University of Helsinki 2008-
Vice-chairman, Finnish Center of Excellence in Molecular and Integrative Neuroscience Research 2008-2013
Vice-chairman, and Co-founder, CORTEX Training Network 2005-
Chairman, and Founder, Finnish Graduate School of Neuroscience (FGSN) 1998-

Previous positions

Head of the Department of Zoology, University of Helsinki 1993 –1994
Head of the Department of Biosciences, University of Helsinki 1995 –1996
Professor of Animal Physiology, University of Helsinki 1994-
Chairman, and Founder, Helsinki Graduate School of Neuroscience (HGSN) 1995-1998
Academy Professor (Academy of Finland) 1996-2006
Vice-chairman, the Academy of Finland Center of Excellence in Molecular Neurobiology 1999-2005
Adjunct Professor, Neuroscience Center, University of Helsinki 2004-2007
Vice-chairman, and Co-founder, CORTEX Training Network 2005-

Selected publications

1. Li H, Khirug S, Cai C, Ludwig A, Blaesse P, Kolikova J, Afzalov R, Coleman SK, Lauri S, Airaksinen MS, Keinänen K, Khiroug L, Saarma M, Kaila K, Rivera C: KCC2 Interacts with the Dendritic Cytoskeleton to Promote Spine Development. Neuron 56: 1019-1033, 2007.
2. Buzsáki G, Kaila K, Raichle M: Inhibition and brain work. Neuron 56: 771-783, 2007.
3. Schuchmann S, Schmitz S, Rivera C, Vanhatalo S, Mackie K, Sipila ST, Voipio J, Kaila K: Experimental febrile seizures are precipitated by a hyperthermia-induced respiratory alkalosis. Nature Med, 12: 817-823, 2006.
4. Vanhatalo S, Kaila K.: Development of neonatal EEG activity: From phenomenology to physiology. Semin Fetal Neonatal Med. 11: 471-478, 2006.
5. Rivera C., Voipio J., Payne J.A., Ruusuvuori E., Lahtinen H., Lamsa K., Saarma M., Kaila K: The K-Cl cotransporter KCC2 renders GABA hyperpolarizing during neuronal maturation. Nature 397: 251-255, 1999.

Douglas R. NORDLI, Jr.

Present Position

Lorna S. and James P. Langdon Chair of Pediatric Epilepsy,
Children's Memorial Hospital, Chicago

Appointments

1990- 1991: Associate Instructor in Neurology, Department of Neurology, Columbia University College of Physicians and Surgeons.
1991- 1999: Assistant Professor of Neurology and Pediatrics. Columbia University College of Physicians and Surgeons.
1994- 1999: Herbert Irving Assistant Professor of Neurology and Pediatrics, Columbia University, New York.
1997- 1999: Director, Pediatric Epilepsy Section, Columbia-Presbyterian Medical Center
1999- 1999: Associate Professor of Clinical Neurology and Clinical Pediatrics, Columbia University, New York
1999- Attending Neurologist, Division of Neurology The Children's Memorial Hospital, Chicago
2000- Lorna S. and James P. Langdon Chair of Pediatric Epilepsy
The Children's Memorial Hospital, Chicago

Selected publications

1. Korff CM, Nordli DR Jr. The clinical-electrographic expression of infantile seizures.Epilepsy Res. 2006 Aug;70 Suppl 1:S116-31.
2. Korff CM, Nordli DR Jr. Epilepsy syndromes undetermined whether focal or generalized in infants. Epilepsy Res. 2006 Aug;70 Suppl 1:S105-9
3. Kim AJ, Kuroda MM, Nordli DR Jr. Abruptly attenuated terminal ictal pattern in pediatrics.J Clin Neurophysiol. 2006 Dec;23(6):532-50.
4. Nordli DR Jr, Korff CM, Goldstein J, Koh S, Laux L, Kelley KR. Cryptogenic late-onset epileptic spasms or late infantile epileptogenic encephalopathy? Epilepsia. 2007 Jan;48(1):206-8.

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Andrew LUX

Present Position

Consultant in Paediatric Neurology, Bristol Royal Hospital for Children, UK
Honorary Clinical Senior Lecturer in Child Health, University of Bristol, UK

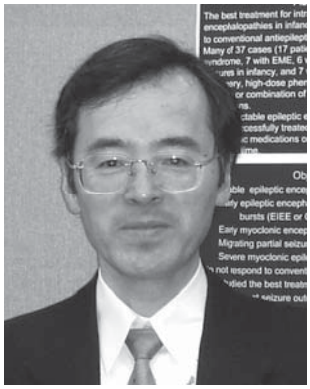
Education and Training

1996- 1997: Postgraduate Student, London School of Hygiene & Tropical Medicine, UK
1997- 1998: Consultant Paediatrician, St Jude Hospital, Vieux Fort, St Lucia, WI
1998- 2001: Clinical Research Fellow, Bath Unit for Research in Paediatrics, UK
2001- 2003: Specialist Registrar in Paediatric Neurology, Bristol Royal Hospital for Children
2003- 2004: Fellow in Paediatric Epilepsy, St Louis Children's Hospital, Missouri, USA
2005- Consultant in Paediatric Neurology, Bristol Royal Hospital for Children, UK

Selected publications

1. Lux, A.L., Edwards, S.W., Osborne, J.P. Responses of local research ethics committees to a study with approval from a multicentre research ethics committee. BMJ 2000; 320:1182-3.
2. Lux, A.L., Edwards, S.W., Hancock, E., O'Callaghan, F., Johnson, T., Kennedy, C.R., Newton, R.W., Verity, C.M., Osborne, J.P. The United Kingdom Infantile Spasms Study comparing vigabatrin with prednisolone or tetracosactide at 14 days: a multicentre, randomised controlled trial. Lancet 2004;364:1773-78.
3. Lux, A.L. and Osborne, J.P. for the West Delphi Group. A proposal for case definitions and outcome measures in studies of infantile spasms and West syndrome: consensus statement of the West Delphi Group. Epilepsia 2004;45:1416-28.
4. Lux, A.L., Edwards, S.W., Hancock, E., Johnson, A.L., Kennedy, C.R., Newton, R.W., O'Callaghan, F.J., Verity, C.M. and Osborne, J.P. The United Kingdom Infantile Spasms Study (UKISS) comparing hormone treatment with vigabatrin on developmental and epilepsy outcomes to age 14 months: a multicentre randomised trial. Lancet Neurology 2005;4:712-7.
5. Lux, A.L., Osborne, J.P. The Influence of Etiology upon Ictal Semiology, Treatment Decisions and Long-Term Outcomes in Infantile Spasms and West Syndrome. Epilepsy Research 2006;70(Suppl1):S77-86.

Kenji SUGAI



Present Position

Physician-in-Chief, Department of Child Neurology,
National Center of Neurology and Psychiatry, Japan

Education

1977 B. Health Sci. The University of Tokyo School of Health Sciences
1981 M.D. The University of Tokyo School of Medicine
1994 Ph.D. Toho University School of Medicine

Postgraduate Training and Professional Experience

1981-1983 Resident in Pediatrics, Kanagawa Children's Medical Center, Yokohama, Japan
1983-1985 Resident in Child Neurology, National Center of Neurology and Psychiatry (NCNP), Tokyo, Japan
1986-1989 Staff physician, Department of Child Neurology, NCNP, Tokyo, Japan
1989-1992 Chief, Department of Pediatrics, Nishi-Kohfu National Hospital, Kohfu, Japan
1992-1993 Section chief, Department of Child Neurology, NCNP, Tokyo, Japan
1993-1994 Research fellow, Department of Neurology, Boston Children's Hospital, Harvard University, Boston, MA, USA
1994-2004 Section chief, Department of Child Neurology, NCNP, Tokyo, Japan
2004-present Physician-in-Chief, Department of Child Neurology, NCNP, Tokyo, Japan

Selected publications

1. Sugai K. Treatment of convulsive status epilepticus in infants and young children in Japan. Acta Neurol Scand 2007: 115 Supple 186; 62-70.
2. Nobutoki T, Sugai K, Fukumizu M, Hanaoka S, Sasaki M. Continuous midazolam infusion for refractory nonconvulsive status epilepticus in children. No To Hattatsu 2005; 37: 369-73.
3. Sugai K. Clobazam as a new antiepileptic drug and clorazepate dipotassium as an alternative antiepileptic drug in Japan. Epilepsia 2004; 45 Suppl 8: 20-5.
4. Sudoh A, Sugai K, Miyamoto T, Mimaki M, Fukumizu M, Hanaoka S, Sasaki M. Non-intravenous high-dose phenobarbital therapy for status epilepticus refractory to continuous infusion of midazolam or pentobarbital: report of three cases. No To Hattatsu 2002; 34: 23-9.
5. Sugai K, Fukuyama Y, Yasuda K, Fujimoto S, Ohtsu M, Ohta H, Ogawa A, Hamano S, Hirano S, Yoshioka H, Ishikawa A, Seki T, Itokazu N, Tawa R. Clinical and pedigree study on familial cases of West syndrome in Japan. Brain Dev 2001; 23: 558-64.
6. Sugai K. Pentobarbital therapy for status epilepticus. Jap J Developmenatl Pharmacology and Therapeutics 1997; 10: 49-52.

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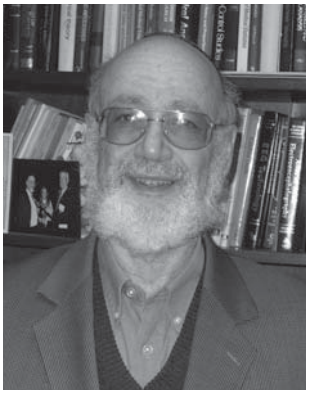
Shlomo SHINNAR



Present Position

Department of Neurology, Pediatrics,
Epidemiology and Population Health and the Comprehensive Epilepsy Management Center, Montefiore Medical Center and the Albert Einstein College of Medicine, Bronx, New York, USA

Tallie Z. BARAM

Dr. Baram is a Professor in the Departments of Pediatrics, Anatomy/ Neurobiology and Neurology at the University of California, Irvine (UCI), where she holds the Danette Shepard endowed chair in Neurological Sciences. Dr. Baram is the Scientific Director of the UCI Epilepsy program, and the founder and Executive Committee chair of the UCI Epilepsy Research Center (EpiCenter). Dr. Baram received a PhD at the Weizmann Institute of Science in Israel, and an MD degree from the University of Miami School of Medicine. She completed her clinical training in Pediatrics and in Child Neurology at Baylor College of Medicine, Houston.

Dr. Baram has focused her research efforts on Seizures and Epilepsies of infants and children, and particularly on Febrile Seizures and Infantile Spasms. Creating suitable immature animal models for these seizures, Dr. Baram's work has led to important discoveries about the mechanisms of infantile Spasms, and about the long-lasting functional effects of experimental prolonged Febrile Seizures on the developing brain. Her work (over 120 peer-reviewed papers) has been published in top journals, and she is co-editor (with Dr. Shinnar) of a popular book on Febrile Seizures.

Dr. Baram has received a number of prizes awards, including the Athalie Clarke Research award, the prestigious American Epilepsy Society Basic Science Research Award and the National Institute of Health Senator Javits Award. She has served on the Executive Board of the American Epilepsy Society, and is currently Executive Trustee of the AES Lennox-Lombroso Research Trust. Baram has organized and contributed to numerous national and international symposia and Investigators Workshops for the Society of Neuroscience, AES, Child Neurology Society and AES and European and Japanese Societies. Dr. Baram is a member of the Professional Advisory Board of the Epilepsy Foundation of America, and chairs the Pediatric Partnership for Epilepsy Research scientific review board. She also has contributed to National Institute of Health, AES, Epilepsy Foundation of America, and CURE study sections, and has devoted considerable effort to mentoring of junior Clinician/Scientists.

Selected Publications:

1. Dube C, Brewster AL, Richichi, C, Zha QQ, Baram TZ. Fever, febrile seizures and epilepsy. Trends Neurosci, 30:490-6; 2007.
2. Richichi C, Brewster AL, Bender RA, Simeone TA, Zha QQ, Yin HZ, Weiss JH, Baram TZ. Mechanisms of seizure-induced 'transcriptional channelopathy' of hyperpolarization-activated cyclic nucleotide gated (HCN) channels. Neurobiol Disease, Nov. 2007
3. Bender RA, Kirschstein T, Kretz O, Brewster AL, Richichi C, Rüschenschmidt C, Shigemoto R, Beck H, Frotscher M, Baram TZ. Localization of HCN1 channels to presynaptic compartments: novel plasticity that may contribute to hippocampal maturation. J Neurosci, 27:4697-706; 2007.
4. Brewster AL, Chen Y, Bender RA, Yeh A, Shigemoto R, Baram TZ. Quantitative analysis and subcellular distribution of mRNA and protein expression of the hyperpolarization-activated cyclic nucleotide-gated channels throughout development in rat hippocampus. Cereb Cortex, 17:702-712; 2007.
5. Dubé C, Richichi C, Bender RA, Chung G, Litt B, Baram TZ. Temporal lobe epilepsy after experimental prolonged febrile seizures: prospective analysis. Brain, 129:911-22; 2006.
6. Dubé C, Vezzani A, Behrens M, Bartfai T, Baram TZ. Interleukin 1$ contributes to the generation of experimental febrile seizures. Ann Neurol, 57:152-5; 2005.
7. Simeone TA, Rho JM, Baram TZ. Single channel properties of hyperpolarization activated cation currents in acutely dissociated rat hippocampal neurons. J Physiol, 568:371-7; 2005.
8. Dubé C, Yu H, Nalcioglu O, Baram TZ. Serial magnetic resonance imaging (MRI) after experimental febrile seizures: altered T2 signal does not signify neuronal death. Ann Neurol, 56:709-14; 2004.
9. Baram TZ. Long-term neuroplasticity and functional consequences of single versus recurrent early-life seizures. Ann Neurol 54; 701-05; 2003.
10. Bender RA, Soleymani SV, Brewster AL, Nguyen ST, Beck H, Mathern GW, Baram TZ. Enhanced expression of a specific hyperpolarization-activated cyclic nucleotide-gated cation channel (HCN) in surviving dentate gyrus granule cells of human and experimental epileptic hippocampus. J Neurosci, 23:6826-36; 2003.
11. Brewster A, Bender RA, Chen Y, Eghbal-Ahmadi M, Dubé C, Baram TZ. Developmental febrile seizures modulate hippocampal gene expression of hyperpolarization-activated channels in an isoform and cell-specific manner. J Neurosci, 22:4591-9; 2002.
12. Dubé C, Chen K, Eghbal-Ahmadi M, Brunson K, Soltesz I, Baram TZ. Prolonged febrile seizures in the immature rat model enhance hippocampal excitability long-term. Ann Neurol, 47:336-44; 2000.
13. Chen K, Baram TZ, Soltesz I. Febrile Seizures in the developing brain result in persistent modification of neuronal excitability in limbic circuits. Nature Medicine, 5:888-94; 1999.
14. Toth Z, Yan XX, Heftoglu S, Ribak CE, Baram TZ. Seizure-induced neuronal injury: vulnerability to febrile seizures in an immature rat model. J Neurosci, 18:4285-94; 1998.

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11th ISS

Qian JIANG

Present Position

PhD student of the Department of Pediatrics, Peking University First Hospital

Appointments

1998- 2003: Peking University Health Science Center, Beijing, China
Medical Bachelor in clinical medicine

2003- Peking University First Hospital, Beijing, China
Ph.D. in epilepsy associated developmental brain injury

Selected publications

1. Q Jiang, JM Wang, XR Wu, YW Jiang. Alterations of NR2B and PSD-95 expression after early-life epileptiform discharges in developing neurons. Int J Dev Neurosci, 2007, 25(3): 165-170
2. Jiang Q, Wang JM, Jiang YW. Synaptic plasticity associated molecules. Journal of International Pathology and Clinical Medicine, 2006, 26(6): 515-518 (review)
3. JM Wang, YW Jiang, HY Cao, LF Yu, T Bo, H Ni, Q Jiang, XR Wu. Long-term effect of early discharge on sEPSC and [Ca2+]i in developing neurons. Neurosci Lett, 2006, 397: 104-109
4. Q Jiang*, JM Wang*, Y Wu, XR Wu, YW Jiang. Epileptiform activity exerts long-lasting effects on AMPAR subunit distribution in neocortical cultures. Int J Dev Neurosci, 2007, under review (*co-first author)
5. Q Jiang, JM Wang, Y Wu, XR Wu, YW Jiang. Changes in NMDAR subunit expression, subcellular distribution and interactions with PSD-95: short and long-lasting effects induced by early-life epileptiform discharges. in revision

Mark DUNLEAVY

Present position

Post Doctorate Research Fellow, Royal College of Surgeons in Ireland, Dublin, Ireland

Appointments

2005- Present position

Selected publications

1. Dunleavy M, Bradford A, O'Halloran KD: Oxidative stress impairs upper airway muscle endurance in an animal model of sleep-disordered breathing.Adv Exp Med Biol. 605:458-62, 2008.
2. O'Sullivan G, Dunleavy M, Hakansson K, Clementi M, Kinsella A, Croke D, Drago J, Fienberg A, Greengard P, Sibley D, Fisone G, Henshall D, Waddington J: Dopamine D1 vs D5 receptor-dependent induction of seizures in relation to DARPP-32, ERK1/2 and GluR1-AMPA signaling. Neuropharm, in press.
3. Murphy B, Dunleavy M, Shinoda S, Schindler C, Meller R, Bellver-Estelles C, Hatazaki S, Dicker P, Yamamoto A, Koegel I, Chu X, Wang W, Xiong Z, Prehn J, Simon R, Henshall D: Bcl-w protects hippocampus during experimental status epilepticus. Am J Pathol. 171:1258-68, 2007.
4. Dunleavy M, Dooley M, Cox D, Bradford A: Chronic intermittent asphyxia increases platelet reactivity in rats.Exp Physiol. 90:411-6, 2005.
5. Martial FP, Dunleavy M, Nolan P, McNicholas WT, O'Regan RG, Bradford A: Simultaneous recording of breathing and respiratory related neuronal activity in the brainstem of conscious rats. Respir Physiol Neurobiol. 145:301-6, 2005.

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11th ISS

Ying-Chao CHANG

Present position

Associate Professor, Department of Pediatrics,
Chang Gung Memorial Hospital-Kaohsiung Medical Center,
Chang Gung University College of Medicine, Taiwan

Appointments

1991-1994:	Pediatric Residency, National Cheng Kung University Hospital, Taiwan
1994-1996:	Fellow in Pediatric Neurology, National Cheng Kung University Hospital, Tainan, Taiwan
1996-1997:	Clinical Research Fellow, Department of Developmental Pediatrics, Kennedy Krieger Institute, Johns Hopkins University, School of Medicine
1997-2002:	Lecturer, Department of Pediatrics, Chang Gung Memorial Hospital, Kaohsiung, Taiwan
2002-2005:	Assistant Professor, Department of Pediatrics, Chang Gung Memorial Hospital, Kaohsiung, Taiwan
2005-	Associate Professor, Department of Pediatrics, Chang Gung Memorial Hospital, Kaohsiung, Taiwan

Selected publications

1. Ying-Chao Chang, Woei-Cherng Shyu, Shinn-Zong Lin, and Hung Li. Regenerative therapy for stroke. Cell Transplantation 2007; 16: 171-181.
2. Ying-Chao Chang, Shun-Fen Tzeng, Lung Yu, A-Min Huang, Hsueh-Te Lee, Chao-Ching Huang, Chien-Jung Ho. Early-Life Fluoxetine Exposure Reduced Functional Deficits after Hypoxic-Ischemia Brain Injury in Rat Pups. Neurobiology of Disease 2006; 24: 101-113.
3. Ying-Chao Chang, Chao-Ching Huang. Perinatal Brain Injury and Regulation of Transcription. Current opinion in Neurology 2006; 19:141-147.
4. Ying-Chao Chang, Chao-Ching Huang. Seizure-induced early plasticity and adverse long-term effects of early-life seizures. Ann Neurology 2004; 56:165-166.
5. Ying-Chao Chang, A-Min Huang, Yu-Min Kuo, Shan-Tair Wang, Yung-Yee Chang, Chao-Ching Huang. Febrile seizures impair memory and hippocampal cAMP response-element binding protein activation. Ann Neurology 2003; 54:706-718.

Mitsumasa FUKUDA

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11th ISS

Present Position

Senior Assistant Professor, Department of Pediatrics,
Ehime University Graduate School of Medicine

Appointments

1991- 1993: Resident in Pediatrics, Ehime University Hospital
1993- 1995: Clinical fellow in Pediatrics, Ehime University Hospital
1995- 1999: Research associate in Pediatrics, Ehime University Hospital
1999- 2004: Chief physician in Pediatrics, Ehime Prefectural Imabari Hospital
2004- 2008: Research associate in Pediatrics, Ehime University Hospital
2008- Present position

Selected publications

1. Fukuda M, Morimoto T, Suzuki Y, Shinonaga C, Ishida Y: Interleukin-6 attenuates Hyperthermia-induced seizures in developing rats. Brain and Development 29: 644-648, 2007.
2. Morimoto T, Fukuda M, Suzuki Y, Kusu M, Kida K: Sequential changes of brain CT and MRI after febrile status epilepticus in a 6 year-old girl. Brain and Development 24: 190-193, 2002.
3. Fukuda M, Morimoto T, Nagao H, Kida K: Clinical study of epilepsy with severe febrile seizures and seizures induced by hot water bath. Brain and Development 19: 212-216,1997.
4. Fukuda M, Morimoto T, Nagao H, Kida K: The effect of GABAergic system activity on hyperthermia-Induced seizures in rats. Developmental Brain Research 104: 197-199, 1997.
5. Morimoto T, Fukuda M, Aibara Y, Nagao H, Kida K: The influence of blood gas changes on hyperthermia-induced seizures in developing rats. Developmental Brain Research 92:77-80, 1996

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James G. HEIDA

Present Position

Postdoctoral Research Associate, Saul R. Korey Department of Neurology, Albert Einstein College of Medicine, Bronx, New York USA

Education and Training

1997- 2001: BSc (honours), Behavioural Neuroscience, Laurentian University Sudbury Ontario Canada

2001- 2006: PhD, Neuroscience, University of Calgary, Calgary Alberta Canada
Supervisor: Dr. Quentin J. Pittman

2006- Present position, Supervisor: Dr. Solomon L. Moshé

Selected publications

1. Heida J.G., Velíšková J., Moshé S.L. (2008). Activation of androgen receptors is involved in sexual differentiation of the substantia nigra pars reticulata seizure-controlling network. Epileptic Disorders IN PRESS.
2. Scantlebury M.H., Heida J.G., Hasson H.J., Velíšková J., Velisek L., Galanopoulou A.S., Moshé S.L. (2007). Age dependant consequences of status epilepticus: animal models. Epilepsia 48 Suppl 2:75-82
3. Heida J.G., Pittman Q.J. (2005). Causal Links between brain cytokines and experimental febrile convulsions in the rat. Epilepsia 46(12); 1906-13
4. Heida J.G., Teskey G.C., Pittman Q.J. (2005). Febrile convulsions induced by the combination of lipopolysaccharide and low dose kainic acid enhance seizure susceptibility not epileptogenesis in rats. Epilepsia 46(12); 1898-905
5. Heida J.G., Boissé L., Pittman Q.J. (2004). Lipopolysaccharide induced febrile convulsions in the rat: short-term sequelae. Epilepsia 45(11); 1317-29.

Michael J. KUBEK

Present Position

Associate Professor of Neurobiology and Medical Neuroscience

Appointments

1975-1978	Postdoctoral Fellow, Center for Endocrinology, Metabolism and Nutrition, Northwestern University School of Medicine
1978-1982	Assistant Professor of Anatomy, Department of Anatomy, Indiana University School of Medicine
1982-1987	Assistant Professor of Anatomy and Neurobiology, Depts. of Anatomy and Psychiatry, Indiana University School of Medicine
1987-Present	Associate Professor of Neurobiology, Medical Neuroscience, Depts. of Anatomy & Cell Biology and Psychiatry, Indiana University School of Medicine

Selected publications

1. Knoblach, S. and Kubek, M.J.: Increases in thyrotropin-releasing hormone (TRH) mRNA expression induced by a model of human temporal lobe epilepsy: Effect of partial and complete kindling. Neuroscience 76(1): 85-95, 1997.
2. Knoblach, S. and Kubek, M.J.: Changes in TRH levels in hippocampal subregions induced by a model of temporal lobe epilepsy: Effect of partial and complete kindling. Neuroscience 76 (1): 97-104, 1997.
3. Kubek, M.J., Shih, T.M. and Meyerhoff, J.L.: Thyrotropin-Releasing Hormone (TRH) is markedly increased in the rat CNS following Soman-induced seizures. Brain Research 747: 328-331, 1997.
4. Kubek, M.J., Liang, D., Byrd, K.E. and Domb, A.J.: Prolonged seizure suppression by a single implantable polymeric- TRH microdisk preparation. Brain Res. 809:189-198, 1998.
5. Kubek, M.J., Ringel, I., and Domb, A.J: Issues elated t2 intranasal delivery of neuropeptides to temporal lobe targets. The Blood Brain Barrier, Drug Delivery and Brain Pathology, pp. 323-350, 2001.
6. Kubek, M.J. and Garg, B.: Thyrotropin-Releasing Hormone in the Treatment of Intractable Epilepsy. Pediatric Neurology 26: 9-17, 2002.
7. Nie, Y., Schoepp, D. D., Klaunig, J.E., Yard, M., Lahiri, D.K. and Kubek, M.J.: Thyrotropin-releasing Hormone (Protirelin) inhibits potassium-stimulated glutamate and aspartate release from hippocampal slices in vitro, Brain Research 1054: 45-54, 2005
8. Kubek,M.J., Yard, M., Lahiri, D.K., and Domb, A.J.: Characterization of Novel Intranasal Sustained-Release Nanoparticles for Delivery of Neuropeptides to the Brain. Nanoparticles for Pharmaceutical Applications. A. Domb, Y. Tabata, and N. V. Ravi Kumar, Eds., American Scientific Publishers, New York pp. 73-84 (2007)
9. Veronesi, M.C., Yard, M., Jackson, J., Lahiri, D.K, and Kubek, M.J.: Thyrotropin-Releasing Hormone (TRH) protects primary fetal rat (E17) cultured neurons against glutamate toxicity. Brain Res. 1128:79-85, 2007
10. Veronesi, M.C., Kubek,D.J. and Kubek, M.J.: Intranasal delivery of the thyrotropin-releasing hormone (TRH) analog attenuates seizures in the amygdala-kindled rat. Epilepsia 48(12):2280-86, 2007
11. Sen, A., Shannon, H.E. and Kubek, M.J.: Analysis of seizure EEG in kindled epileptic rats. Comp. Math. Methods Medicine 8(4):000-000, 2007
12. Veronesi, M.C., Kubek, D.J. and Kubek, M.J.: Isoflurane and halothane exacerbate seizures in amygdala-kindled rats during recovery. Epilepsy Research (in review)

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11th ISS

Hideo YAMANOUCHI

Present Position

Associate Professor of the Department of Pediatrics,
Dokkyo Medical University School of Medicine

Appointments

1991- 1996: Staff, Pediatric Neurologist, Division of Child Neurology, National Center of Neurology and Psychiatry, Kodaira, Japan

1997- 1999: Assistant Professor, Department of Pathology, Gunma University School of Medicine

1999- 2004: Lecturer and Pediatric Neurologist, Department of Pediatrics, Dokkyo Medical University School of Medicine, Mibu, Japan

2004- Associate Professor and Pediatric Neurologist, Department of Pediatrics, Dokkyo Medical University School of Medicine, Mibu, Japan

Selected publications

1. Yamanouchi H, Kawaguchi N, Mori M, Imataka G, Yamagata T, Hashimoto T, Momoi MY, Eguchi M, Mizuguchi M. Acute infantile encephalopathy predominantly affecting the frontal lobes. Pediatr Neurol 2006;34:93-100.
2. Yamanouchi H, Mizuguchi M. Acute infantile encephalopathy predominantly affecting the frontal lobes (AIEF): a novel clinical category and its tentative diagnostic criteria. Epilepsy Res 2006;70:S263-265.
3. Yamanouchi H. Activated remodeling and N-methyl-D-aspartate (NMDA) receptors in cortical dysplasia. J Child Neurol 2005;20:303-307.
4. Yamanouchi H. Imataka G, Nakagawa E, Nitta A, Suzuki N, Hirao J, Suzumura H, Watabe H, Arisawa O, Eguchi M. An analysis of epilepsy with chromosomal abnormalities. Brain Dev 2005;27:370-377.

Tetsuro NAGASAWA

19
11th ISS

Present Position

Staff of the Division of Neurology,
National Center for Child Health and Development

Appointments

1994- 1996: General Residency, Musashino Red Cross Hospital
1996- 1997: Pediatrics Residency, Tokyo Medical and Dental University Hospital
1997- 1999: Pediatrics Residency, Tsuchiura Kyoudo General Hospital
1999- 2001: Residency of Child Neurology, National Center of Neurology and Psychiatry
2001- 2002: Staff of Department of Pediatrics, National Disaster Medical Hospital
2002- Present position

Selected publications

1. Tetsuro Nagasawa, Ikumi Kimura, Yuichi Abe, Akira Oka: HHV-6 encephalopathy with cluster of convulsions during eruptive stage. Pediatric Neurology 36: 61-63, 2007.
2. Tetsuro Nagasawa, Koichi Mizuguchi, Yuichi Abe, Akira Oka: HHV-6 encephalopathy with cluster of convulsions during eruptive stage: A new subtype. Philippine Journal of Neurology 10: 47, 2007.
3. Tetsuro Nagasawa, Ikumi Kimura, Yuichi Abe, Akira Oka:A patient with HHV-6 encephalopathy showing a cluster of convulsions during the eruptive stage and hyperperfusion of the cerebral hemisphere during the acute phase. No To Hattatsu 38: 295-298, 2006. (Japanese)
4. Tetsuro Nagasawa, Yuichi Abe, Manami Honda, Akira Oka, Nobuhito Morota, Kenji Nihei: A patient treated with continuous intrathecal Baclofen therapy effectively for the spasticity of lower extremities. No To Hattatsu 37: 164-165, 2005. (Japanese)
5. Tetsuro Nagasawa, Akira Sudo, Michio Fukumizu, Shigeru Hanaoka, Masayuki Sasaki, Kenji Sugai, Ikuya Nonaka: Congenital monomelic neurogenic discorders. Brain and Development 25: 571-573, 2003.

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11th ISS

Nicola SPECCHIO

Present Position

Full position as Consultant at Division of Neurology,
Bambino Gesù Children Hospital, Roma, Italy

Appointments

2004- 2007	Clinical Fellow at Division of Neurology, Bambino Gesù Children Hospital, Roma.
1999- 2004	University of Bari, attending Department of Neurological and Psychiatric Science - Epilepsy Centre as "post-graduating medical doctor"
2001	Attending the "Neuroscience Unit, Institute of Child Health and Great Ormond Street Hospital for Children, University College London" as research fellow, taking part to the research and clinical activities of the Epilepsy Centre and to the academic meetings of the "National Hospital for Neurology and Neurosurgery, Queen Square, University College London".
2000	Attending King's College Hospital, London as research fellow. University of Bari, attending Department of Neurological and Psychiatric Science as medical doctor and taking part to the clinical, tutorial and researching activities in the 1st Neurologic Clinic.
1997 – 1999	University of Bari, attending Departement of Neurological and Psychiatric Science as student and taking part to the clinical, tutorial and research activities in the 1st Neurologic Clinic.
1996 – 1999	Student's representative in the Faculty of Medicine, University of Bari
1994 – 1996	Institute of Pharmacological Research "Mario Negri", Milano collaboration on a study: "Risk in Epilepsy Study Group".

Selected publications

1. Specchio N, Kahane P, Pasquier B, Tassi L and Guerrini R. Resective surgery for epileptogenic dysembryoplastic neuroepithelial tumor in hemimegalencephaly. Neurology 2005;65:777-778.
2. Fusco L, Specchio N. Non-epileptic paroxysmal manifestations during sleep in infancy and childhood. Neurological Science 2005;26:205-209.
3. Specchio N, Cusmai R, Volkov J, Montaldo P, Vigevano F. Occurrence of a prolonged nonepileptic motor status after a febrile seizure. Epilepsia 2006;47:1079-1081
4. Ferrie CD, Caraballo R, Covanis A, Demirbilek V, Dervent A, Fejerman N, Fusco L, Grunewald RA, Kanazawa O, Koutroumanidis M, Lada C, Livingston JH, Nicotra A, Oguni H, Martinovic Z, Nordli DR Jr, Parisi P, Scott RC, Specchio N, Verrotti A, Vigevano F, Walker MC, Watanabe K, Yoshinaga H, Panayiotopoulos CP. Autonomic status epilepticus in Panayotopoulos syndrome and other childhood and adult epilepsies: a consensus view. Epilepsia 2007;48:1165-1172.
5. Specchio N, Boero G, Michelucci R, Gambardella A, Giallonardo AT, Fattouch J, Di Bonaventura C, de Palo A, Ladogana M, Vigevano F, La Neve A, Specchio LM. Effects of levetiracetam on EEG abnormalities in Juvenile Myoclonic Epilepsy. Epilepsia. 2008 Feb 5 [e-pub ahead of print]

Tatsuhiko SHIKE

Present Position

Pediatrics staff of Shizuoka Institute of Epilepsy and Neurological Disorders

Appointments

1993- 1995: Pediatrics Residency, Fukushima Medical University
1995- 2005: Pediatrics staff, Fukushima medical University Hospital
2005- 2006: Pediatrics Residency, Shizuoka Institute of Epilepsy and Neurological Disorders
2006- Present position

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Ingrid E. SCHEFFER

Present Position

Professor, Departments of Medicine and Pediatrics, University of Melbourne, Australia

Senior Pediatric Neurologist and Director, Children's Epilepsy Program, Austin Health

Senior Pediatric Neurologist, Royal Children's Hospital, Melbourne

Education / Training

INSTITUTION AND LOCATION	DEGREE	YEAR(s)	FIELD OF STUDY
Monash University, Australia	MBBS	1983	Medicine
Royal Australasian College of Physicians	FRACP	1992	Neurology
Austin Hospital Epilepsy Program		1991-4	Epilepsy Fellowship
University of Melbourne, Australia	PhD	1998	Epilepsy Genetics

Positions and Employment

2005- Professor, Departments of Medicine

1999 Chancellor's Prize, University of Melbourne.

1999 Harbison-Higinbotham Research Scholarship.

Honours

1998 Commendation for Premier's Award for Medical Research.

1999 Chancellor's Prize, University of Melbourne.

1999 Harbison-Higinbotham Research Scholarship.

2003 National Association of Research Fellows of NHMRC Post-Doctoral Investigator Award

2007 American Epilepsy Society Clinical Research Recognition Award

Recent relevant publications

1. Harkin LA, McMahon JM, Iona X, Dibbens L, Pelekanos JT, Zuberi SM, Sadleir LG, Andermann E, Gill D, Farrell K, Connolly M, Stanley T, Harbord M, Andermann F, Wang J, Batish SD, Jones JG, Seltzer WK, Gardner A,The infantile epileptic encephalopathy referral consortium, Sutherland G, Berkovic SF, Mulley JC, Scheffer IE. The spectrum of SCN1A-related infantile epileptic encephalopathies. Brain 2007 130;843–852.
2. Jansen FE, Sadleir LG, Harkin LA, Vadlamudi L, McMahon JM, Mulley JC, Scheffer IE, Berkovic SF. Severe myoclonic epilepsy of infancy: The adult phenotype. Neurology 2006 67;2224–2226
3. Scheffer IE, Harkin LA, Grinton BE, Dibbens LM, Turner SJ, Xu R, Jackson G, Adams J, Connellan M, Petrou S, Wellard RM, Briellmann RS, Wallace RH, Mulley JC, Berkovic SF. Temporal lobe epilepsy and GEFS+ phenotypes associated with SCN1B mutations. Brain 2007;130:100–109. Epub October 4 2006
4. Mulley JC, Nelson P, Guerrero S, Dibbens L, Iona X, McMahon JM, Harkin L, Schouten J, Yu S, Berkovic SF, Scheffer IE. A new molecular mechanism for severe myoclonic epilepsy of infancy: exonic deletions of SCN1A. Neurology 2006;67(6):1094–5
5. Berkovic SF, Harkin L, McMahon JM, Pelekanos JT, Zuberi SM, Wirrell EC, Gill DS, Iona X, Mulley JM, Scheffer IE. De-novo mutations of the sodium channel gene SCN1A in alleged vaccine encephalopathy: a retrospective study. Lancet Neurology 2006;5:488-492. Epub 20 April 2006

Tiziana GRANATA

Education

Graduated from Medical School University of Milan Italy 1985
Residency in Child Neurology Medical School University of Pavia Italy 1989

Professional Experience

1985-1991: Post-doc fellowship in Child Neurology at the Neurological Institute C. Besta, Milan
1992-2000 Assistant in Child Neurology at the Neurological Institute C. Besta, Milan
2001- today Associated in Child Neurology at the Neurological Institute C. Besta, Milan

Professional Experties

Epilepsy in childhood, with particular interest in West syndrome and related disorders, epilepsies due to channelopathies, and in epileptic disorders resulting from progressive encephalopathy
Malformation of cortical development
Rasmussen encephalitis and other inflammatory-related epilepsies
Epilepsy and movement disorders
Alternating hemiplegia
Author of more than 50 papers on peer-reviewed, international journals

Selected publications

1. Nabbout R, Gennaro E, Dalla Bernardina B, Dulac O, Madia F, Bertini E, Capovilla G, Chiron C, Cristofori G, Elia M, Fontana E, Gaggero R, Granata T, Guerrini R, Loi M, La Selva L, Lispi ML, Matricardi A, Romeo A, Tzolas V, Valseriati D, Veggiotti P, Vigevano F, Vallee L, Dagna Bricarelli F, Bianchi A, Zara F. Spectrum of SCN1A mutations in severe myoclonic epilepsy of infancy. Neurology. 2003 Jun 24;60(12):1961-7.
2. Vezzani A, Granata T. Brain inflammation in epilepsy: experimental and clinical evidence. Epilepsia. 2005 Nov;46(11):1724-43. Review.
3. Bien CG, Granata T*, Antozzi C, Cross JH, Dulac O, Kurthen M, Lassmann H, Mantegazza R, Villemure JG, Spreafico R, Elger CE. Pathogenesis, diagnosis and treatment of Rasmussen encephalitis: a European consensus statement. Brain. 2005, 128:454-71.
4. Gennaro E, Santorelli FM, Bertini E, Buti D, Gaggero R, Gobbi G, Lini M, Granata T, Freri E, Parmeggiani A, Striano P, Veggiotti P, Cardinali S, Bricarelli FD, Minetti C, Zara F. Somatic and germline mosaicisms in severe myoclonic epilepsy of infancy. Biochem Biophys Res Commun. 2006 Mar 10;341(2):489-93.
5. Striano P, Mancardi MM, Biancheri R, Madia F, Gennaro E, Paravidino R, Beccaria F, Capovilla G, Bernardina BD, Darra F, Elia M, Giordano L, Gobbi G, Granata T, Ragona F, Guerrini R, Marini C, Mei D, Longaretti F, Romeo A, Siri L, Specchio N, Vigevano F, Striano S, Tortora F, Rossi A, Minetti C, Dravet C, Gaggero R, Zara F. Brain MRI Findings in Severe Myoclonic Epilepsy in Infancy and Genotype-Phenotype Correlations. Epilepsia. 2007; 48(6):1092-6.

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Kazuhiro YAMAKAWA

Current Position

Laboratory Head, Laboratory for Neurogenetics,
RIKEN Brain Science Institute, Saitama, Japan.

Education

1980- 1984 B.Sc. Molecular Biology (1984, March),
Faculty of Science, University of Kyoto, Kyoto, Japan.

1989 – 1992 Ph.D. Medical Genetics (1992, May)
Faculty of Medicine, University of Osaka, Osaka, Japan.

1994- 1996 Postdoctoral fellowship,

Research and Professional Experience

1984- 1989 Researcher
Department of Biochemistry, General Institute, Toyobo Co., Ltd., Shiga, Japan.

1989– 1994 Researcher, Department of Biochemistry, Cancer Institute, Tokyo, Japan.
Work supervised by Dr. Yusuke Nakamura

1994- 1996 Research fellow, Department of Medical Genetics, Cedars-Sinai Medical Center,
UCLA School of Medicine.

1996- 1997 Assistant professor-Adjunct, Pediatrics of UCLA School of Medicine.
Research Scientist, Medical Genetics, Cedars-Sinai Reserch Institute

1997- Laboratory Head, Laboratory for Neurogenetics, RIKEN Brain Science Institute

Selected publications

1. Ogiwara I, Miyamoto H, Morita N, Atapour N, Mazaki E, Inoue I, Takeuchi T, Itohara S, Yanagawa Y, Obata K, Furuichi T, Hensch TK, Yamakawa K. (2007) Nav1.1 Localizes to Axons of Parvalbumin-Positive Inhibitory Interneurons: a Circuit Basis for Epileptic Seizures in Mice Carrying an Scn1a Gene Mutation. Journal of Neuroscience 27(22):5903-5914
2. Shukkur EA, Shimohata A, Akagi T, Yu W, Yamaguchi M, Murayama M, Chui D, Takeuchi T, Amano K, Harve Subramhanya K, Hashikawa T, Sago H, Epstein CJ, Takashima A, Yamakawa K. (2006) Mitochondrial dysfunction and tau hyperphosphorylation in Ts1Cje, a mouse model for Down syndrome Human Molecular Genetics 15: 2752-2762.
3. Suzuki T, Delgado-Escueta AV, Aguan K, Alonso ME, Shi J, Hara Y, Nishida M, Numata T, Medina MT, Takeuchi T, Morita R, Bai D, Ganesh S, Sugimoto Y, Inazawa J, Bailey JN, Ochoa A, Jara-Prado A, Rasmussen A, Ramos-Peek J, Cordova S, Rubio-Donnadieu F, Inoue Y, Osawa M, Kaneko S, Oguni H, Mori Y, Yamakawa K (2004) Mutations in EFHC1 cause juvenile myoclonic epilepsy. Nat. Genet. 36: 842-849.
4. Amano K, Sago H, Uchikawa C, Suzuki T, Kotliarova SE, Nukina N, Epstein CJ, Yamakawa K (2004) Dosage-dependent over-expression of genes in the trisomic region of Ts1Cje mouse model for Down syndrome. Hum. Mol. Genet. 13:1333-1340.
5. Kamiya K, Kaneda M, Sugawara T, Mazaki E, Okamura N, Montal M, Makita N, Tanaka M, Fukushima K, Fujiwara T, Inoue Y, Yamakawa K (2004) A nonsense mutation of the sodium channel gene SCN2A in a patient with intractable epilepsy and mental decline. J. Neurosci. 24:2690-2698.

International Symposia in Past 5 Years Organized by Infantile Seizuire Society (ISS), Japan

Year	Theme	Invited Faculties (Japanese excluded)	Publications
2003	Chromosomal Aberrations and Childhood Epilepsies	Andermann E, Battaglia A, Berkovic SF, Biraben A, Laan LEAM, Singhi P, Stephenson JBP, Wang P-J, Williams CA, Zuberi SM	Brain & Development 2005; 27(2): 79-140
2004	Neuronal Migration Disorders and Childhood Epilepsies	Battaglia GS, Crino PB, Curatolo P, Golden JA, Granata T, Guerrini R, Mochida GH, Otsubo H, Woermann FG	Jounal of Child Neurology 2005; 20(4): 273-397
2005	Epileptic Syndromes in Infancy and Early Childhood	Andermann F, Berg AT, Capovilla G, Craiu D, De Vivo DC, Engel J Jr, Fejerman N, Hirsch E, Kasteleijin-Nolst Trenite DGA, Kim DW, Lee WL, Lux AL, Moshe SL, Nordli DR Jr, O'Regan ME, Plouin P, Sankar R, Scheffer I, Scher MS, Specchio N, Wolf P	Epilepsy Reserch 2006; 60: Suppl 1: S1-S279
2006	Status Epilepticus in Infants and Young Children	Banu S, Fusco L, Kalra V, Lee JS, Lux AL, Neville B, Otsubo H, Sanker R, Shinnar S, Specchio N, Wasterlain CG	Acta Neurologica Scandinavica Supplementum 2007; vol. 115 Suppl 186
2007	Biology of Seizure Susceptibility	Arzimanoglou A, Baram TZ, Curatolo P, Goldin AL, Mochida GH, Moshe SL, Otsubo H, Prasad A, Sarnat HB, Schridde U, Staley KJ, Swann JW	Progress in Pediatric Epilepsy Series. John Libbey Eurotext Ltd, Montrouge (France). 2008 (in preparation)

The 12th Annual Meeting of the Infantile Seizure Society (ISS); International Symposium on Epilepsy in Autistic Spectrum Disorders and Related Conditions (ISEASD)

President: Dr. Toyojiro MATSUISHI
Professor and Chairman, Department of Pediatrics & Child Health
Kurume University School of Medicine
Kurume, Fukuoka, Japan

Date: May 9-10, 2009

Venue: Chikusui Hall, Kurume, Fukuoka

Main theme: Epilepsy in Autistic Spectrum Disorders and Related Conditions

Objectives: Autistic spectrum disorders (ASD) are a behaviorally defined set of developmental disorders that result from diverse biologic, genetic, and epigenetic factors. The interaction of genetic and epigenetic factors in ASD has long been a subject of interest among scientists and physicians. Autistic disorder, Asperger syndrome, Rett syndrome, and childhood disintegrative disorder are subsumed under the category of pervasive developmental disorders. The ISEASD aims to present a comprehensive update of the topic and discussions on such issues as genetics, epidemiology, pathophysiology, neuroimaging, subsequent epilepsy, treatment of ASDs and related conditions.

Contact: The 12th ISS Secretariat
c/o Department Pediatrics & Child Health,
Kurume University School of Medicine
67 Asahi-machi, Kurume, Fukuoka 830-0011 Japan
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