# **PROGRAM**

# EIGHTY EIGHTH ANNUAL MEETING OF THE AMERICAN ASSOCIATION OF NEUROPATHOLOGISTS

**JUNE 21-24, 2012** 

# THE PALMER HOUSE HILTON

CHICAGO, ILLINOIS

This activity is sponsored by the American Association of Neuropathologists

For additional information about the accreditation of this program, please contact the AANP office at 440-793-6565 or via email at aanpoffice @gmail.com

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### AMERICAN ASSOCIATION OF NEUROPATHOLOGISTS

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### **OFFICERS**

Raymond A. Sobel, MD, Stanford University School of Medicine, President
John M. Lee, MD, PhD, Loyola University, Vice-President
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Karen M. Weidenheim, MD, Montefiore Medical Center (Member-at-Large)

### **ARCHIVIST**

Michael N. Hart, MD, University of Wisconsin School of Medicine

### OFFICIAL JOURNAL

Journal of Neuropathology and Experimental Neurology
Raymond A. Sobel, Editor
Barbara J. Crain, Associate Editor
Jeffrey A. Golden, Associate Editor
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### **DIAGNOSTIC SLIDE SESSION**

Anthony T. Yachnis, MD, *University of Florida Medical College*, Moderator Mark L. Cohen, MD, *University Hospitals Case Medical Center*, Manager

### **COMMITTEES**

### **AWARDS COMMITTEE**

Maria Beatriz S. Lopes, MD, Chair Adekunle M. Adesina, MD, PhD Ada Baisre, MD Rudy J. Castellani, MD Mark T. Curtis, MD, PhD Ivana Delalle, MD, PhD Marc R. Del Bigio, MD. PhD David W. Ellison, MD, PhD Matthew P. Frosch, MD, PhD Kymberly A. Gyure, MD Brent T. Harris, MD, PhD Craig Horbinski, MD, PhD Alexander Judkins, MD Bradley M. Miller, MD, PhD C. Ryan Miller, MD, PhD Peter Pytel, MD

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### PROGRAM COMMITTEE

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Kymberly A. Gyure, MD
Marta Margeta, MD, PhD
Amyn M. Rojiani, MD, PhD
Shahriar M. Salamat, MD, PhD
Julie Schneider, MD
Suash Sharma, MD

### **PROFESSIONAL AFFAIRS**

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Eileen H. Bigio, MD
Daniel J. Brat, MD, PhD
William F. Hickey, MD
John M. Lee, MD
William C. McDonald, MD
Brian E. Moore, MD
Robert E. Mrak, MD, PhD
Kathy L. Newell, MD
William H. Yong, MD
Marie Rivera Zengotita, MD

### **EDUCATION COMMITTEE**

John M. Lee, MD, PhD, Chair Mark Cohen, MD Kar-Ming Fung, MD, PhD William F. Hickey, MD Maria Martinez-Lage, MD Suzanne Z. Powell, MD C. Harker Rhodes, MD, PhD Tarik Tihan, MD, PhD Jane Uyehara-Lock, MD Charles L. White, III, MD

# INTERNATIONAL SOCIETY OF NEUROPATHOLOGY COUNCILORS

Joseph E. Parisi, MD Arie Perry, MD Clayton A. Wiley, MD, PhD Anthony T. Yachnis, MD

### **TARGET AUDIENCE**

The educational design of AANP's Annual Meeting addresses the needs of physicians and scientists in the field of neuropathology who are involved in the diagnosis and/or treatment of patients with neurological disorders.

### STATEMENT OF NEED

The purpose of this activity shall be to advance the knowledge of new techniques, scientific findings, treatments, and the practice and teaching of neuropathology. The practice of neuropathology is understood to include the diagnosis of diseases of the nervous system, and teaching and training in the science and practice of neuropathology

### **LEARNING OBJECTIVES**

Upon completion of this activity, participants should be able to:

- Discuss recent technical advances that enhance the diagnostic accuracy of neuropathology
- Review new information on the pathogenesis of neurological diseases that can be incorporated into the teaching of neuropathology to trainees and colleagues
- Identify recent trends in the incidence and epidemiology of neurological diseases in which neuropathologists impact therapeutic decisions and public health

### **DISCLAIMER**

Participants have an implied responsibility to use the newly acquired information to enhance patient outcomes and their own professional development. The information presented is this activity is not meant to serve as a guideline for patient management. Any procedures, medications, or other courses of diagnosis or treatment discussed in this activity should not be used by clinicians without evaluation of patient conditions and possible contraindications on dangers in use, review of any applicable manufacturer's product information, and comparison with recommendations of other authorities.

### **CME CREDIT**

### Physician Accreditation Statement

The American Association of Neuropathologists is accredited by the Accreditation Council for Continuing Medical Education (ACCME) to provide continuing medical education for physicians.

### Physician Credit Designations

The American Association of Neuropathologists designates this live educational activity for a maximum of 24.25 *AMA PRA Category 1 Credit(s)*<sup>TM</sup>. Physicians should only claim credit commensurate with the extent of their participation in the activity.

### Instructions to Receive Credit:

In order to receive credit for this activity, the participant must complete the CME credit application in the registration packet and return it to the American Association of Neuropathologists office at:

American Association of Neuropathologists C/o Peggy Harris 25373 Tyndall Falls Drive Olmsted Falls, Ohio 44138

The chart below details the maximum number of credit hours a physician can earn for each educational activity being certified for *AMA PRA Category 1 Credit*<sup>TM</sup> at this year's Annual Conference.

Activity	CME Credit Hours
Special Course	7
Scientific Sessions	8
Korey Lecture	1
DeArmond Lecture	1
Parisi Lecture	1
Diagnostic Slide Session	3
Presidential Symposium	3.25
Total	24.25

### **DISCLOSURE INFORMATION:**

### **Disclosure of Commercial Support:**

This activity is supported educational grants from Teva Neurosciences and The National Multiple Sclerosis Society. "In-kind" support through the donation of microscopes is being provided by Olympus.

### Disclosure of Unlabeled Use:

This educational activity may contain discussion of published and/or investigational uses of agents that are not indicated by the FDA. The American Association of Neuropathologists does not recommend the use of any agent outside of the labeled indications.

The opinions expressed in the educational activity are those of the faculty and do not necessarily represent the views of any organization associated with this activity. Please refer to the official prescribing information for each product for discussion of approved indications, contraindications and warnings.

### Disclosure of Conflict of Interest:

The American Association of Neuropathologists requires instructors, planners, managers and other individuals who are in a position to control the content of this activity to disclose any real or apparent conflict of interest they may have as related to the content of this activity. All identified conflicts of interest are thoroughly vetted by AANP for fair balance, scientific objectivity of studies mentioned in the materials or used as the basis for content, and appropriateness of patient care recommendations. Complete disclosure information will be provided to learners on-site.

The **Planners and Managers** reported the following financial relationships or relationships to products or devices they or their spouse/life partner have with commercial interests related to the content of this CME activity:

Name of Planner or Manager	Reported Financial Relationship
Eileen H. <b>Bigio</b> , Northwestern University	Nothing to Disclose
Feinberg School of Medicine	
Daniel J. Brat, Emory University School of	Nothing to Disclose
Medicine	
Rudy J. Castellani, University of Maryland	Nothing to Disclose
Elizabeth J. Cochran, Medical College of	Nothing to Disclose
Wisconsin	
Mark L. Cohen, University Hospitals Case	Nothing to Disclose
Medical Center	
James D. <b>Dollar</b> , Carolinas Pathology Group	Nothing to Disclose
Kar-Ming Fung, University of Oklahoma	Nothing to Disclose
Murat Gokden, University of Arkansas	Nothing to Disclose
Kymberly A. <b>Gyure</b> , West Virginia University	Nothing to Disclose

The *Planners and Managers* reported the following financial relationships or relationships to products or devices they or their spouse/life partner have with commercial interests related to the content of this CME Activity

Name of Planner or Manager	Reported Financial Relationship	
Robert F. <b>Hevner</b> , University of Washington	Consultant for the Allen Institute of Brain Science	
William F. Hickey, Dartmouth Medical School	Nothing to Disclose	
John M. Lee, Loyola University	Consultant/Reviewer For Up to Date on CJD	
	articles; Holds group patents with Cornelli	
	Consulting	
David N. <b>Louis</b> , Massachusetts General Hospital	Nothing to Disclose	
William C. McDonald, Abbott Northwestern	Nothing to Disclose	
Hospital		
Marta <b>Margeta</b> , University of California San	Nothing to Disclose	
Francisco		
Maria Martinez-Lage, University of Pennsylvania	Nothing to Disclose	
Brian E. <b>Moore</b> , Memorial Medical Center	Nothing to Disclose	
Steven A. <b>Moore</b> , University of Iowa	Nothing to Disclose	
Robert E. Mrak, University of Toledo	Nothing to Disclose	
Kathy L. <b>Newell</b> , University of Kansas Medical Center	Nothing to Disclose	
George <b>Perry</b> , University of Texas at San	Consultant/Independent Contractor for Neurotez;	
Antonio	Stock Shareholder with Curaxis	
Suzanne <b>Powell</b> , The Methodist Hospital	Nothing to Disclose	
C. Harker <b>Rhodes</b> , Dartmouth-Hitchcock Medical	Nothing to Disclose	
Center		
Marie <b>Rivera-Zengotita</b> , University of Florida Medical College	Nothing to Disclose	
Amyn Rojiani, Medical College of Georgia	Nothing to Disclose	
Shahriar M. Salamat, University of Wisconsin	Nothing to Disclose	
Hospital		
Julie <b>Schneider</b> , Rush University Medical Center	Consultant for Eli Lilly co. and Avid	
	Radiopharmaceuticals	
Suash <b>Sharma</b> , Medical College of Georgia	Nothing to Disclose	
Raymond A. <b>Sobel</b> , Stanford University School of Medicine	Nothing to Disclose	
Anat <b>Stemmer-Rachamimov</b> , Massachusetts General Hospital	Nothing to Disclose	
Tarik <b>Tihan</b> , University California San Francisco	Nothing to Disclose	
Jane <b>Uyehara-Lock</b> , Kaiser Foundation Hospital	Nothing to Disclose	
Karen M. Weidenheim, Montefiore Medical	Nothing to Disclose	
Center		
Charles L. White III, University of Texas	Nothing to Disclose	
Southwestern Medical Center		
Anthony T. <b>Yachnis</b> , University of Florida Medical College	Nothing to Disclose	
William H. Yong, University of California Los	Grant/Research Support for Genentech, Amgen,	
Angeles	and Tocagen; Stock shareholder with Pfizer	
	-	

The *faculty* reported the following financial relationships or relationships to products or devices they or their spouse/life partner have with commercial interests related to the content of this CME Activity

Name of Faculty/Presenter	Reported Financial Relationship
Adekunle <b>Adesina</b> , Baylor College of Medicine	Nothing to Disclose
Homa Adle-Biassette, INSERM	Nothing to Disclose
Safa <b>Al-Sarraj</b> , Kings College Hospital, London	Nothing to Disclose
Derick <b>Aranda</b> , University of California San	Nothing to Disclose
Francisco	ŭ
Leonidas <b>Arvanitis</b> , Rush University	Nothing to Disclose
Krystof <b>Bankiewicz</b> , University of California San Francisco	Nothing to Disclose
Thomas <b>Beach</b> , Banner Health	Nothing to Disclose
,	
Joseph <b>Berger</b> , University of Kentucky	Consultant/Independent Contractor for Amgen, Bayer, Biogen Idec, Genentech, Millenium, Eisai, Genzyme, Novartis, and Pfizer; Grant/Research Support for PML Consortium; Honoraria from Bayer and Biogen Idec; Speaker's Bureau for Bayer and Biogen Idec
Juan M. Bilbao, Sunnybrook Hospital	Nothing to Disclose
T. Bourne, University of Virginia Health System	Nothing to Disclose
Robert H. <b>Brown</b> , Jr. University of Massachusetts	Consultant/Independent Contractor for Biogen Idec; Grant/Research Support for NINDS, PZALS, Project ALS; other Royalty from McGraw Hill
Nigel <b>Cairns</b> , Washington University School of Medicine	Nothing to Disclose
H. Brent <b>Clark</b> , University of Minnesota Medical School	Nothing to Disclose
Kenneth <b>Clark</b> , University of Pittsburgh Medical Center	Nothing to Disclose
Misti <b>Coronel</b> , Thomas Jefferson University Hospital	Nothing to Disclose
Stephen <b>DeArmond</b> , University of California San Francisco	Nothing to Disclose
Marc <b>Del Bigio</b> , University of Manitoba	Nothing to Disclose
Ivana <b>Delalle</b> , Boston University School of Medicine	Nothing to Disclose
Dennis <b>Dickson</b> , Mayo Clinic	Nothing to Disclose
Mohamed El-Hag, University Case Medical	Nothing to Disclose
Center David <b>Ellison</b> , St. Jude Children's Research Hospital	Nothing to Disclose
Ferechte <b>Encha-Razavi</b> , Necker-Enfants Malades Hospital	Nothing to Disclose
Michelle Madden <b>Felicella</b> , University of California San Francisco	Nothing to Disclose
Amanda <b>Fisher-Hubbard</b> , University of Michigan	Nothing to Disclose
Bernardino <b>Ghetti</b> , Indiana University	Consultant with Bayer Pharma AG
Jeffrey <b>Golden</b> , Brigham and Women's Hospital	Nothing to Disclose
James <b>Hackney</b> , University of Alabama	Nothing to Disclose  Nothing to Disclose
Leslie <b>Hamilton</b> , University of Calgary	I NOTHING TO DISCUSE
May Htwe <b>Han</b> , Stanford University School of Medicine	Nothing to Disclose

The *faculty* reported the following financial relationships or relationships to products or devices they or their spouse/life partner have with commercial interests related to the content of this CME Activity

Name of Faculty/Presenter	Reported Financial Relationship
John <b>Hedreen</b> , Harvard Brain Tissue Resource	Nothing to Disclose
Center	-
Annie Hiniker, University of California San	Nothing to Disclose
Francisco	
Cheng-Ying <b>Ho</b> , Johns Hopkins University	Nothing to Disclose
Thomas <b>Huebner</b> , University of Maryland	Nothing to Disclose
Jason <b>Huse</b> , Memorial Sloan-Kettering Cancer	Nothing to Disclose
Center	
Cristiane M. Ida, Mayo Clinic	Nothing to Disclose
Mark <b>Jentoft</b> , Mayo Clinic	Nothing to Disclose
Hannah Kinney, Children's Hospital Boston	Nothing to Disclose
Barry Kosofsky, Cornell University	Nothing to Disclose
Julia Kofler, University of Pittsburgh	Nothing to Disclose
Naomi <b>Kouri</b> , Mayo Clinic	Nothing to Disclose
Michael Lawlor, Medical College of	Nothing to Disclose
Wisconsin/Children's Hospital of Wisconsin	
Jian Yi Li, North Shore-LIJ Health System	Nothing to Disclose
Rong Li, University of Alabama at Birmingham	Nothing to Disclose
Seth <b>Love</b> , University of Bristol	Nothing to Disclose
Jian-Qiang <b>Lu</b> , University of Alberta	Nothing to Disclose
Claudia Lucchinetti, Mayo Clinic	Nothing to Disclose
Marta Margeta, University of California San	Nothing to Disclose
Francisco	
Ann McKee, Boston University School of	Nothing to Disclose
Medicine	N. d Di . l
Joshua Menke, Mayo Clinic	Nothing to Disclose
C. Miller, University of North Carolina	Nothing to Disclose
Steven A. Moore, University of Iowa	Nothing to Disclose
David <b>Munoz</b> , St. Michael's Hospital, University of Toronto	Nothing to Disclose
Jill Murrell, Indiana University School of Medicine	Nothing to Disclose
Peter <b>Nelson</b> , University of Kentucky	Nothing to Disclose
Ho Keung <b>Ng</b> , The Chinese University of Hong	Nothing to Disclose
Kong	Nothing to Disclose
Michael <b>Norenberg</b> , University of Miami School of Medicine	Nothing to Disclose
Adriana <b>Olar</b> , The Methodist Hospital	Nothing to Disclose
Brent Orr, The Johns Hopkins School of	Nothing to Disclose
Medicine	
Dan Perl, Uniformed Services Health Science	Nothing to Disclose
Center	
George <b>Perry</b> , The University of Texas at San	Consultant/Independent Contractor for Neurotez;
Antonio	Stock Shareholder with Curaxis
Tracie <b>Pham</b> , University of California Los	Nothing to Disclose
Angeles	
Joanna <b>Phillips</b> , University of California San Francisco	Nothing to Disclose
David <b>Pisapia</b> , Columbia University	Nothing to Disclose

The *faculty* reported the following financial relationships or relationships to products or devices they or their spouse/life partner have with commercial interests related to the content of this CME Activity

Phillip G. <b>Popovich</b> , Ohio State University	Nothing to Disclose
Medical Center	
Dushyant <b>Purohit</b> , Mount Sinai School of	Nothing to Disclose
Medicine	
Peter <b>Pytel</b> , University of Chicago	Nothing to Disclose
Jiang <b>Qian</b> , Albany Medical Center	Nothing to Disclose
Veena Rajaram, Children's Memorial	Nothing to Disclose
Hospital/Northwestern University	
Shannon <b>Risacher</b> , Indiana University School of	Nothing to Disclose
Medicine	-
Fausto Rodriguez, Johns Hopkins University	Nothing to Disclose
Amyn <b>Rojiani</b> , Georgia Health Sciences	Nothing to Disclose
University, Medical College of Georgia	
Kathryn <b>Saatman</b> , University of Kentucky	Nothing to Disclose
A. Dessa <b>Sadovnick</b> , VCHA-UBC Hospital	Nothing to Disclose
Walter <b>Schulz-Schaeffer</b> , University Medical	Consultant with Bayer Healthcare
Center Göttingen	·
Leroy <b>Sharer</b> , New Jersey Medical School	Nothing to Disclose
Susan Staugaitis, Cleveland Clinic	Nothing to Disclose
Kimberly Stogner-Underwood, Virginia	Nothing to Disclose
Commonwealth University	_
Mario Suvà Massachusetts General Hospital	Nothing to Disclose
Masaki <b>Takao</b> , Tokyo Metropolitan Geriatric	Nothing to Disclose
Hospital	
Bruce D. <b>Trapp</b> , The Cleveland Clinic Foundation	Consultant/Independent Contractor for Teva,
	Biogen Idec, and Renova Neural;
	Grant/Research support for NIH, MS Society,
	and Third Frontier; Honoraria from EMD Serono,
	Merck Serono, and Teva; Speakers Bureau for
	EMD Serono and Teva
Sriram Venneti, University of Pennsylvania	Nothing to Disclose
Ruben Vidal, Indiana Alzheimer Disease Center	Nothing to Disclose
Christopher William, Massachusetts General	Nothing to Disclose
Hospital	
Kum Thong <b>Wong</b> , University of Malaya	Nothing to Disclose
Xiongwei <b>Zhu</b> , Case Western Reserve University	Nothing to Disclose

### **GENERAL INFORMATION**

Hotel: The Palmer House Hilton 17 East Monroe Street Chicago, IL 60603

Phone: 312-726-7500

### ALL MEETING SESSIONS WILL BE HELD AT THE PALMER HOUSE HILTON

*All* platform presentations and general sessions (Special Lectures, Korey Lecture, DeArmond Lecture, Parisi Lecture, Business Meetings, Diagnostic Slide Session, and Presidential Symposium) will be held in the **Adams Ballroom** of the hotel on the sixth floor.

All poster sessions will be held in Exhibit Hall on the fourth floor.

### PRE-REGISTRATION PICK-UP

Attendees pre-registered and pre-paid for the Special Course and/or Meeting will have their name badge, course syllabus, program booklets, June 2012 issue of JNEN with abstracts, reception ticket(s) and registration receipt ready for pick-up at the AANP Registration Desk, located in the Adams Pre-function area of the hotel on the sixth floor. On-site registration and additional tickets for the Annual Reception will be available at the Desk.

### **REGISTRATION DESK**

Location	Adams Ballroom Pre-Function	Area
Time	Wednesday, June 20	6:30 pm – 9:00 pm
Time	Thursday, June 21	6:30 am - 12 noon
		6:30 pm – 9:00 pm
	Friday, June 22	7:00 am - 12 noon
		5:30 pm – 6:00 pm
	Saturday, June 23	7:00 am - 12 noon

### PLEASE, wear your name badge!

Your name badge is *required for admittance* to any function of the Association, including the Special Course, all Friday, Saturday and Sunday sessions, and the Friday evening reception.

### **NOTES to PRESENTERS**

### Platform Presenters (PowerPoint)

### Please include in your presentation a conflict of interest slide.

All platform presentations will be held in either the **Adams or Monroe Ballrooms** of the hotel. All general sessions (Special Lectures, Korey Lecture, DeArmond Lecture, Parisi Lecture, Business Meetings, Diagnostic Slide Session, and Presidential Symposium) will be held in the **Adams Ballroom**.

Presenters should use PowerPoint for their presentation.

All PowerPoint presentations will be transferred onto a show computer prior to the start time of each session. Each room will be equipped with a lectern, audience microphones, central computer (loaded with MS Office XP), LCD/Data projector, screens and a laser pointer.

### Special Notes for PowerPoint presenters:

- Each speaker must bring his/her PowerPoint presentation on a disc (CD-ROM) or USB memory stick.
- Please title the presentation with your name (name.ppt).
- Macintosh users, be sure to save your presentation as .ppt (your name.ppt). If the ".ppt" extension is not present in the file name, the file will not be recognized by the PC computer.
- Label your disc with your name, session name, time, and day of presentation. Your presentation will be transferred onto the show computer for each session by the technician. Please make sure your presentation is in its final form, since once loaded onto the show computer, no changes can be made.
- Please take your disc or memory stick to the room in which you will be presenting, Adams or Monroe Ballrooms, at one of the times indicated below. It is your responsibility to get your file to the AV staff prior to your presentation.
- The AV staff will be available to load your file onto the computer during scheduled evening and
  morning times, or during session breaks. These will be the <u>only</u> times available to you to load and
  test your presentation.

**Schedule for Loading PowerPoint Presentations** 

Load show computer in Adams or Monroe Ballroom		
Thursday, June 21	7:00 am - 7:45 am	
	10:30 am - 11:00 am	
	3:00 pm – 3:30 pm	
Friday, June 22	7:00 am - 7:45 am	
	10:00 am - 10:30 am	
	4:00 pm – 4:30 pm	
Saturday, June 23	7:00 am – 7:45 am	
	10:00 am - 10:30 am	
	4:00 pm – 4:30 pm	
Sunday, June 24	7:00 am - 7:45 am	

- If you are presenting in a morning session, it is preferable to check in the previous day. Sameday presentations may be loaded in the morning prior to session start time, but since this time necessarily is limited, you are encouraged to have your presentation loaded on the evening preceding your talk. Presenters at the evening Diagnostic Slide Session also will be able to submit their files on Saturday evening in the Adams Ballroom from 6:30-7:45 pm.
- To avoid time delays and potential problems with your presentation, you will **not** be allowed to use your own computer, although you may bring your laptop as a backup.

### **Notes to Poster Presenters**

**Both** poster sessions will be held in **Exhibit Hall** on the fourth floor. Approximately half the posters will be displayed all day Friday and the remainder all day Saturday. Posters should be up by 8:00 am on the morning of your presentation and taken down by 6:30 pm the same day. The poster board size is 8 ft wide x 4 ft high. Please plan your poster to be at least a few inches smaller in each direction. The poster board surface and construction should accommodate either Velcro or push pins.

To encourage interaction with interested attendees, authors must be present at their posters for discussion/questions during morning or afternoon refreshment breaks, at the following designated times:

	Fri June 22 Authors Present at:	Sat June 23 Authors Present at:
EVEN Numbered Poster	10:00 - 10:30 am	4:00 – 4:30 pm
ODD Numbered Poster	4:00 – 4:30 pm	10:00 - 10:30 am

### **MICROSCOPE VIEWING ROOM**

Multi-headed microscopes will be available in the *Medinah Parlor* on the sixth floor of the hotel.

Location	Medinah Parlor	Medinah Parlor	
Time	Thursday, June 21	7:00 am - 5:30 pm	
	Friday, June 22	7:00 am - 5:30 pm	
	Saturday June 23	7:00 am - 5:30 pm	

### **BUSINESS MEETING**

Location	Adams Ballroom	Adams Ballroom		
Time	Friday, June 22	11:45 am - 12:45 pm		
	Saturday June 23	11:45 am – 12:45 pm		

The awards for *Meritorious Contributions to Neuropathology* will be presented on Saturday, June 23

# **SPECIAL MEETINGS BY INVITATION ONLY**

Date	Meeting	Time/Location
Thurs	Executive Council Meeting	6:00 pm
June 21		Grant Park Parlor, Sixth Floor
Fri	Education Committee Meeting	7:00 am
June 22		Grant Park Parlor, Sixth Floor
	Trainee Luncheon	11:45 pm – 2:00 pm
		The Price Room, Fifth Floor
	Awards Committee Meeting	5:30 pm – 6:30 pm
	_	Grant Park Parlor, Third floor
Saturday	JNEN Editorial Board Meeting	7:00 am – 8:00 am
June 23		The Price Room, Fifth Floor
	NP Program Directors Meeting	1:00 pm – 2:00 pm
		Grant Park Parlor, Sixth Floor
	Awards Committee Meeting	6:00 pm 7:30 pm
		Grant Park Parlor, Sixth Floor
	Professional Affairs	6:00 pm – 8:00 pm
		The Spire Parlor, Sixth Floor
	Presidential Reception	6:30 pm – 8:00 pm
		The Price Room, Fifth Floor
Sun	Founders Breakfast	7:00 am – 8:00 am
June 24		Grant Park Parlor, Sixth Floor

### **ABSTRACTS**

Abstracts of the papers presented in the program are published in the June 2012 issue of the *Journal of Neuropathology and Experimental Neurology*.

### ANNUAL RECEPTION

The annual reception will be held 6:30 to 8:30 pm, Friday in the Mezzanine Lobby. Registrants and guests of the AANP are welcome to attend. There will be a cash bar. Additional tickets are \$20 each for guests of AANP attendees, and may be purchased at the time of registration or at the door. Several "prizes" will be awarded to trainees.

Location	Mezzanine Lobby	
Time	Friday, June 22	6:30 pm – 8:30 pm

### **SPONSORS and DONORS**

This meeting is sponsored in part by generous contributions from several sponsors and donors. Please visit their displays and exhibits in the Adams Ballroom Pre-Function area.

Location	Exhibit Hall, Fourth Floor	Exhibit Hall, Fourth Floor	
Time	Thursday, June 23	12:00 pm – 5:30 pm	
	Friday, June 24	7:00 am - 5:30 pm	
	Saturday June 25	7:00 am - 5:30 pm	

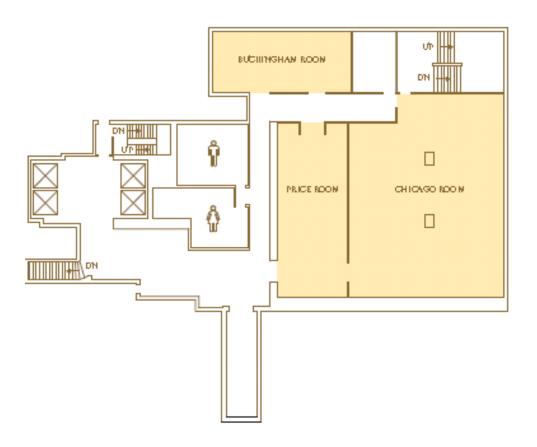
### **MEETING EXHIBITORS**

- Histobiotec LLC
- Olympus America, Inc.
- Wolters Kluwers Health

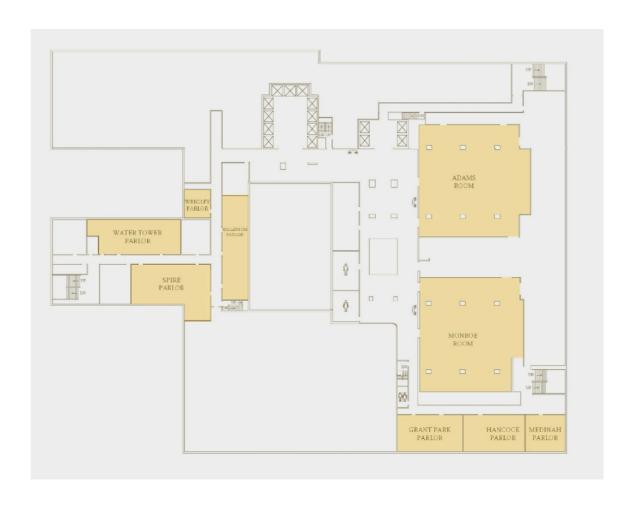
### **RECEPTION PRIZE CONTRIBUTORS**

- Wolters Kluwers Health
- Elsevier Inc.

# The Palmer House Hilton Floor Plan Fifth Floor



# The Palmer House Hilton Floor Plan Sixth Floor



### **PROGRAM and SCIENTIFIC SESSIONS**

### **SPECIAL COURSE:**

Location	Adams Ballroom	
Date/Time	Thursday, June 21	8:00 am - 5:00 pm
	Acquired Neuropathology in a (	Changing World
		Director: Raymond A. Sobel, MD

### **PLATFORM PRESENTATIONS**

· — · · · · · · · · · · · · · · · · · ·			
Location	Adams Ballroom and Monroe Ballroom		
Date/Time	Friday, June 22	8:00 am – 4:00 pm	
	Saturday, June 23	8:00 am – 4:00 pm	

### **POSTER PRESENTATIONS**

Location	Exhibit Hall, Fourth Floor	
Date/Time	Friday, June 22	8:00 am - 6:30 pm
	Saturday, June 23	8:00 am - 6:30 pm

### MATTHEW T. MOORE LECTURE

Location	Adams Ballroom	Adams Ballroom	
Date/Time	Friday, June 22	10:30 am - 11:30 am	
	Pathogenesis of ALS		
		Robert H. Brown, Jr. MD	
		University of Massachusetts, Worcester, MA	

### **DEARMOND LECTURE**

Location	Adams Ballroom		
Date/Time	Friday, June 22		4:30 pm – 5:30 pm
	Novel MRI-based Platform for Efficient Gene Delivery to the Brain		
	Krystof Bankiewicz, MD, PhD		
		University of Ca	lifornia San Francisco, San Francisco, CA

### SAUL R. KOREY LECTURE

Location	Adams Ballroom	
Date/Time	Saturday, June 23	10:30 am - 11:30 am
		Why the Brain Fails when the Astrocyte Ails
		Michael Norenberg, MD
		University of Miami School of Medicine

### **DIAGNOSTIC SLIDE SESSION**

Location	Adams Ballroom	
Date/Time	Saturday, June 23	8:00 pm -11:00 pm

### PRESIDENTIAL SYMPOSIUM

Location	Adams Ballroom	
Date/Time	Sunday, June 24	8:00 am - 12 noon
	Current Topics in Multiple Sclerosis	

# **MEETING AT A GLANCE**

THURSDAY June 21, 2012			
	Adams Ballroom		
8:00 am - 5:15 pm	8:00 am - 5:15 pm   SPECIAL COURSE		
	Acquired Neuropathology in a Changing World		

### (Abstract Numbers in Italics)

		FRIDAY June 22	, 2012
	Adams Ballroom	Monroe Ballroom	Exhibit Hall, Fourth Floor
8:00 - 10:00 am	Platform 1 Tumors- I	Platform 2 Neurodegenerative – Alzheimer's Disease	
	#1 - 8	#9 - 16	
10:00 - 10:30 am		IMENT BREAK	
10:30 - 11:30 am		<b>MOORE LECTURE</b> s Ballroom	
	Pathoge	enesis of ALS	
		Robert H. Brown, Jr. MD Massachusetts, Worcester, MA	
11:45 - 12:45 pm		SS MEETING I s Ballroom	All Posters
12:45 - 2:00 pm	L	UNCH	
	Adams Ballroom	Monroe Ballroom	Friday June 22 <sup>nd</sup> and Saturday June 23 <sup>rd</sup>
2:00 - 4:00 pm	Platform 3 Demyelinating/Pediatric Neuropathology/ Development	Platform 4 Neurodegenerative: Other	10:00 – 10:30 am 4:00 - 4:30 pm
	#17-24	#25 -32	
4:00 – 4:30 pm	REFRESHMENT BREAK		
4:30 – 5:30 pm	DEARMOND LECTURE Adams Ballroom		
		n for Efficient Gene Delivery to e Brain	
	University of California Sal	Krystof Bankiewicz, MD, PhD on Francisco, San Francisco, CA	

6:30 - 8:30 pm ANNUAL RECEPTION: Mezzanine Lobby

# **MEETING AT A GLANCE**

### (Abstract Numbers in Italics)

		SATURDAY J	une 23, 2012
	Adams Ballroom	Monroe Ballroom	Exhibit Hall, Fourth Floor
8:00 -	Platform 5	Platform 6	
10:00 am	Muscle/Nerve/Other	Pediatric Pediatric Tumor/ Developmental and Pediatric Neuropathology	
	#97 - 104	#105-112	
10:00 - 10:30 am		HMENT BREAK	
10:30 -		REY LECTURE	
11:30 am	Adan	ns Ballroom	
	Why the Brain Fail.	s when the Astrocyte Ails	
		Michael Norenberg, MD	
		ni School of Medicine, Miami, FL	
11:45 -		SS MEETING II	
12:45 pm		ns Ballroom	
12:45 - 2:00pm	L	UNCH	
2.000111	Adams Ballroom	Monroe Ballroom	
2:00 -	Platform 7	Platform 8	
4:00 pm	Tumors II	Neurodegenerative – Other II	All Posters
	#113-120	#121-128	Friday June 22 <sup>th</sup> and Saturday June 23 <sup>th</sup> 10:00 – 10:30 am
4:00 - 4:30 pm	REFRESHMENT BREAK		4:00 - 4:30 pm
4:30 - 5:00 pm		<b>ial Lecture</b> ns Ballroom	
	History of Chicago Neuropathology		
	John M. Lee, MD, PhD		
	Loyola University, Maywood, IL		
5:00 -	What Every Neurop	athologist Needs to Know:	
5:15 pm	NIA-AA Revised Guidelines for the Diagnosis of Alzheimer's Disease		
	7 IIZIICIII	ici 3 Discusc	
	Julie Schneider, MD		
F 00	Rush University Medical Center		
5:00 - 5:30 pm	What Every Neurop IDH1 and its Practica	athologist Needs to Know: I Utility in Glioma Diagnosis	
0.50 piii	IDITI GIIG ROTTGOROG	, and the second	
		David N. Louis, MD	
		Massachusetts General Hospital	
8:00 - 11:00 pm		C SLIDE SESSION as Ballroom	
50 pin	, tdull		

SUNDAY June 24, 2012		
Adams Ballroom		
8:00 am - 12:00 pm	PRESIDENTIAL SYMPOSIUM	
	Current Topics in Multiple Sclerosis	

# THURSDAY, June 21, 2012

## SPECIAL COURSE

# **Acquired Neuropathology in a Changing World**

Director: Raymond A. Sobel, MD

# Adams Ballroom

8:00 am	Welcome and CME Pretest
	Raymond A. Sobel, MD
	Stanford University School of Medicine, Stanford, CA
8:15 am – 9:00 am	Emerging Epidemic Viral Encephalitides with a Special Focus on
	Henipaviruses
	Kum Thong Wong, MBBS, MPath FRCPath
	University at Malaya, Malaysia
9:00 am – 9:45 am	Neuropathology in a Changing World: AIDS Neuropathology – 30 Year Perspective
	Leroy Sharer, MD
0.45	New Jersey Medical School, Newark, NJ
9:45 am – 10:30 am	Progressive Multifocal Leukoencephalopathy in the Monoclonal Antibody Era
	Joseph Berger, MD
10:20 am 11:00 am	University of Kentucky Medical Center, Lexington, KY
10:30 am – 11:00 am	REFRESHMENT BREAK
11:00 am – 11:45 am	Acquired Perinatal Brain Injury
	Hannah Kinney, MD
11.15	Children's Hospital, Boston, MA
11:45 am - 12:30 pm	Neuropathological Effects of Perinatal Cocaine Exposure
	Barry Kosofsky, MD, PhD
	Cornell University, New York, NY
12:30 pm - 1:30 pm	LUNCH
1:30 pm – 2:15 pm	Immune Responses in Spinal Cord Injury
	Phillip G. Popovich, PhD
0.45	Ohio State University Medical Center, Columbus, OH
2:15 pm – 3:00 pm	Experimental Traumatic Brain Injury
	Kathryn Saatman, PhD
2:00 pm 2:20 pm	University of Kentucky, Lexington, KY
3:00 pm - 3:30 pm	REFRESHMENT BREAK
3:30 pm – 4:15 pm	Traumatic Brain Injury, Shell Shock, and Posttraumatic Stress Disorder in the
	Military – Past, Present, and Future
	Daniel Perl, MD Uniformed Services Health Science Center, Bethesda, MD
4:15 pm – 5:00 pm	Chronic Traumatic Encephalopathy
7.10 piii – 3.00 piii	Ann McKee, MD
	Boston University School of Medicine, Bedford, AM

### **FRIDAY, JUNE 22, 2012**

# TRAINEE LUNCHEON: FORMULA FOR SUCCESS

(Not Offered for CME Credit)

# 11:45 pm – 2:00 pm - Price Room, Fifth Floor

Introduction
Eileen Bigio, MD
Northwestern University Feinberg School of Medicine, Chicago, IL
Academic Neuropathology
Matthew P. Frosch, MD, PhD
Massachusetts General Hospital, Boston, MA
Academic NP to Big Pharma Employee to Consulting to Biotech
Meredith Halks-Miller, MD
Pathwork Diagnostics, Redwood City, CA
Private Practice NP
Steve Dubner, MD
Midwest Pathology Associates, Leawood, KS
SAMS and Re-certification
Daniel J. Brat, MD, PhD
Emory University School of Medicine, Atlanta, GA
Job Fair
Michael Lawlor, MD, PhD
Medical College of Wisconsin, Milwaukee, WI

# SATURDAY, JUNE 23, 2012

### **SPECIAL LECTURES**

(Not Offered for CME Credit)

### **Adams Ballroom**

4:30 pm –	History of Chicago Neuropathology
5:00 pm	John M. Lee, MD, PhD Loyola University, Maywood, IL
5:00 pm – 5:15 pm	What Every Neuropathologist Needs to Know: NIA-AA Revised Guidelines for the Diagnosis of Alzheimer's Disease
	Julie Schneider, MD Rush University Medical Center
5:15 pm – 5:30 pm	What Every Neuropathologist Needs to Know: IDH1 and its Practical Utility in Glioma Diagnosis
	David N. Louis, MD Massachusetts General Hospital

# **SUNDAY, JUNE 24, 2012**

# PRESIDENTIAL SYMPOSIUM

# **Current Topics in Multiple Sclerosis**

### **Adams Ballroom**

Adams Balliooni				
8:00 am - 8:05 am	Introduction			
	Raymond A. Sobel, MD			
	Stanford University School of Medicine, Stanford, CA			
8:05 am - 9:00 am	Parisi Lecture			
	Neuronal Damage in Multiple Sclerosis			
	Bruce D. Trapp, PhD			
	The Cleveland Clinic Foundation, Cleveland, OH			
9:00 - 9:45 am	Sphingosine-1-phosphate signaling in MS "Oriental Medicine to			
	Immune Modulation"			
	May Htwe Han, MD, PhD			
	Stanford University School of Medicine, Stanford, CA			
9:45 am - 10:30 am	AANP Award Presentations and Refreshment Break			
10:30 am - 11:15	The Changing Epidemiology of MS			
am	A. Dessa Sadovnick, PhD			
	VCHA-UBC Hospital, Vancouver, BC			
11:15 am – 12:00	How MS could be an Acquired Disease			
pm	Raymond A. Sobel, MD			
	Stanford University School of Medicine, Stanford, CA			
12:00 pm	INSTALLATION OF NEW OFFICERS AND ADJOURNMENT			

Platform 1: Tumors I

Chairpersons: David Louis, MD and Craig Horbinski, MD, PhD

	1	Gene Expression Profiling on Matched Neurofibroma / MPNST Pairs
8:00- 8:15		Peter Pytel, Kammi Henriksen, Thomas Krausz, Thomas Stricker
8:15- 8:30	2	Decreased 5hmC is Associated with Neural Progenitor Phenotype in Normal
0.10	-	Brain and Shorter Survival in Malignant Glioma
		Brent Orr, Sidney Kimmel, Charles Eberhart, Jessica Hicks, William Nelson, Srinivasan
		Yegnasubramanian
8:30- 8:45	3	Patterns of PDGFRA Copy Number Gain in High-Grade Adult and Pediatric
		Astrocytomas
		Derick Aranda, David Ellison, Alexander Judkins, Sidney Croul, Gelareh Zadeh, Sabine
		Mueller, Shengmei Zhou, Roxanne Marshall, Daphne Haas-Kogen, Joanna Phillips, Arie Perry
8:45- 9:00	4	GBM with Oligodendroglial Differentiation has Better Clinical Outcome but
		Similar Biological Markers Compared with other GBMs
		Safa Al-Sarraj, Ross Laxton, Lawrence Doey, Sergey Popov, Alexa Jury, Chris Jones, Lucy
		Brazil, Gill Sadler, Ronald Beaney, Ranj Bhangoo, Richard Gullan, Chris Chandler, Naomi
		Sibtain, Istvan Bodi, Andrew King, Keyoumars Ashkan
9:00- 9:15	5	Histology Trumps Apparent 1p/19q Codeletion in Glioblastomas
		Kenneth Clark, Marina Nikiforova, Ronald Hamilton, Craig Horbinski
9:15- 9:30	6	Unique Subset of Adult and Pediatric Astrocytoma Exhibit Increased
		Expression of Microglia/Macrophage Genes
		Jane Engler, Aaron Robinson, Ivan Smirnov, J. Graeme Hodgson, Nalin Gupta, C. James,
		Annette Molinaro, Joanna Phillips
9:30- 9:45	7	Biology, Immunophenotype, and Molecular Genetics of Central Nervous
		System Lymphomas
		Amanda Fisher-Hubbard, Catherine Dixon, Lili Zhao, Bryan Betz, Megan Lim, Sandra Camelo-
		Piragua
9:45- 10:00	8	Primary Central Nervous System Lymphomas in Immunocompetent Patients-
		North Shore-Long Island Jewish Health System Experience
		Jian Yi Li, Xinmin Zhang, Hua Qiang Chen, Peter Farmer, Mansoor Nasim, Alexis
		Demopoulos, Craig Devoe, Tulika Ranjan, Mark Eisenberg, Michael Schulder, Chengpeng Bi

10:00 - 10:30 am REFRESHMENT BREAK

10:30 – 11:30 am Matthew T. Moore Lecture

Pathogenesis of ALS Robert H. Brown, Jr., MD

University of Massachusetts, Worcester, MA

11:45 am – Business Meeting I (Adams Ballroom) 12:45 pm

12:45 – 2:00 pm Lunch

FRIDAY, JUNE 22, 2012 Monroe Ballroom 8:00 am - 2:00 pm

### Platform 2: Neurodegenerative - Alzheimer Disease Chairpersons: Kathy Newell, MD and Tessa E. Hedley -Whyte, MD

8:00- 8:15	9	Review and Diagnostic Application of the NIA-AA Guidelines for
		Neuropathological Assessment of Alzheimer's Disease
		Dushyant Purohit, Nirmala Batheja, Mona Lisa Thybulle, Maria Maroukian, Mary Sano
8:15- 8:30	10	Alzheimer's disease associated Genes MS4A4A and MS4A6A are Expressed in
		Microglia and Downregulated by Classical activation
		Julia Kofler, Stephanie Bissel, Mark Stauffer, Clayton Wiley, Geoffrey Murdoch
8:30- 8:45	11	Impaired Mitochondrial Biogenesis Contributes to Mitochondrial Dysfunction in
		Alzheimer's Disease
		Xiongwei Zhu, Baiyang Shen, Hyoung-gon Lee, Gemma Casadesus, George Perry, Xinglong
		Wang
8:45- 9:00	12	Endothelin-1, Oxidative Stress and Vascular Dysfunction in Alzheimer's
		Disease
		Seth Love and Jen Palmer
9:00- 9:15	13	Ultrastructure of Ubiquitin-positive, TDP-43-negative Neuronal Inclusions in
		C9ORF72-linked FTD/AL
		Wen-Lang Lin, Dennis Dickson, Kevin Bieniek, Mariely DeJesus-Hernandez, Nicola
		Rutherford, Matthew Baker, Neill Graff-Radford, Kevin Boylan, Rosa Rademakers
9:15- 9:30	14	Early Visual System Synaptic Plasticity Defects in Alzheimer Disease Model
		Transgenic Mice
		Christopher William, Mark Andermann, Glenn Goldey, Demetris Roumis, R Clay Reid, Carla
		Shatz, Mark Albers, Matthew Frosch, Bradley Hyman
9:30- 9:45	15	Increased Expression of Synaptic Regulators Accompanies Preserved
		Cognitive Status in Early Alzheimer's Disease Pathology
		Ivana Delalle, Patricia Kao, Meredith Banigan, Charles Vanderburg, Ann McKee, Peter Polgar,
		Sudha Seshadri
9:45- 10:00	16	Hypertrophy of Hippocampal CA1 Neurons in Asymptomatic Alzheimer's
		Disease in the Oldest Old 90+ Study
		Zhihong Guo, Maria Corrada, Gay Rudow, Alena Savonenko, Diego Iacono, Claudia Kawas,
		Juan Troncoso

10:00 - 10:30 am REFRESHMENT BREAK

**10:30 – 11:30 am Matthew T. Moore Lecture** (Adams Ballroom)

Pathogenesis of ALS Robert H. Brown, Jr., MD

University of Massachusetts, Worcester, MA

11:45 am – 12:45 pm **Business Meeting I** (Adams Ballroom)

12:45 – 2:00 pm Lunch

### Platform 3: Demyelinating/Pediatric Neuropathology/Development Chairpersons: Marc Del Bigio, MD, PhD and Joseph Parisi, MD

2:00- 2:15	17	X-Linked Hydrocephalus Spectrum: Evidence for Related Clinical Entities with Unknown Molecular Bases. Review of 140 Cases  Homa Adle-Biassette, Catherine Fallet-Bianco, Anne-Lise Delezoide, INSERM U676; Nicole
		Drouot, Pascale Marcorelles, Pascale Saugier-Veber, Annie Laquerriere
2:15- 2:30	18	Differential Effects of a Polyalanine Tract Expansion in Arx on Neural
		Development and Gene Expression
		Jeffrey Golden, MacLean Nasrallah, Ginam Cho, Jacqueline Simonet, Mary Putt, Kunio Kitamura
2:30- 2:45	19	Retrospective Review of Autopsies on Persons with Known in utero Alcohol
		Exposure
		Marc Del Bigio and Payam Pahlavan
2:45- 3:00	20	Rosenthal Fibers in Neuromyelitis Optica
		Yong Guo, James Goldman, Bogdan Popescu, Charles Howe, Joseph Parisi, Vanda Lennon,
0.00.0.15		Claudia Lucchinetti
3:00- 3:15	21	Aquaporin-4 Immunohistochemistry Aids Pathological Diagnosis of Neuromyelitis Optica Spectrum Disorder
		Mark Jentoft, Yong Guo, Joseph Parisi, Bogdan Popescu, Vanda Lennon, Sean Pittock,
		Caterina Giannini, Lucchinetti Claudia
3:15-3:30	22	Atrophy of Fornix is Associated with Hippocampal Demyelination in Multiple Sclerosis Patients
		Susan Staugaitis, Elizabeth Fisher, Kathryn Easley, Ansi Chang, Robert Fox, Ranjan Dutta,
		Bruce Trapp
3:30- 3:45	23	Apoptotic Profiles Correlate With GFAP Loss and Microglial Activation in
		Electrolyte-Induced-Demyelination
		Amyn Rojiani, Kimberly Smith, Robert Kersting, Mumtaz Rojiani
3:45- 4:00	24	pStat3 Immunoreactivity Surrounding Multiple Sclerosis Lesions
		Jian-Qiang Lu, Fabrizio Giuliani, Gregg Blevins, Christopher Power, V. Wee Yong

### 4:00 - 4:30 pm REFRESHMENT BREAK

### 4:30 – 5:30 pm DeArmond Lecture

Novel MRI-based Platform for Efficient Gene Delivery to the Brain Krystof Bankiewicz, MD, PhD University of California San Francisco, San Francisco, CA

# 6:30 – 8:30 pm Annual Reception Mezzanine Lobby

FRIDAY, JUNE 22, 2012 Monroe Ballroom 2:00 pm - 5:30 pm

Platform 4: Neurodegenerative: Other

Chairpersons: Mathew Frosch, MD, PhD and Bernardino Ghetti, MD

2:00- 2:15	25	Neocortical Amyloid-ß and Tau Pathology in Patients with Cerebral Amyloid Angiopathy Thomas Huebner, Justin Slavin, Rupal Mehta, Rudy Castellani
2:15- 2:30	26	Amyloid Deposition in Human Wild-type Amyloid Precursor Protein YAC Transgenic Mice with the Psen1-L166P Knock-in Mutation
		Ruben Vidal, Neeraja Sammeta, Holly Garringer, Kumar Sambamurti, Leticia Miravalle, Bruce Lamb, Bernardino Ghetti
2:30- 2:45	26	Detection of ß-Amyloid by Florbetaben PET: Histopathological Verification in a Global Phase 3 Clinical Trial Walter Schulz-Schaeffer, Anja Hoffmann, Osama Sabri, John Seibyl, Hiroyasu Akatsu, Masaki Takao, Thomas Beach, Shigeo Murayama, Bernardino Ghetti, James Ironside, James Leverenz, Katrin Roth, Cornelia Reininger, Marwan Sabbagh
2:45- 3:00	28	In Alzheimer's Disease, Braak Stages V and VI Differ Considerably from Each Other Peter Nelson and Janna Neltner
3:00- 3:15	29	Mutations in FTL Lead to Neurodegeneration by Causing a Gain-of-toxic Function and a Loss-of-normal Function of Ferritin Ruben Vidal, Martin Baraibar, Ana Barbeito, Holly Garringer, Thomas Hurley, Barry Muhoberac, Bernardino Ghetti
3:15-3:30	30	Preventing PrPC Synthesis in Mice with Adeno-Associated Viral Vectors AAV2, AAV6 and AAV9 Stephen DeArmond, Misol Ahn, Krystyna Bajsarowicz, Abby Oehler, Krystof Bankiewicz
3:30- 3:45	31	New Observations in Gerstmann-Sträussler-Scheinker Disease Associated with the PRNP A117V Mutation  Masaki Takao, Salvatore Spina, Jill Murrell, Francine Epperson, Bradley Glazier, Martin Farlow, Frederick Unverzagt, Andrew Saykin, Karmen Yoder, Shannon Risacher, Vladimir Kepe, Jorge Barrio, Bernardino Ghetti
3:45- 4:00	32	Spinocerebellar Ataxia 26: Neuropathological Findings and an Association with a Mutation in Eukaryotic Elongation Factor 2 H Brent Clark, Katherine Hekman, Guo-Yun Yu, Christopher Gomez

### 4:00 - 4:30 pm REFRESHMENT BREAK

**4:30 – 5:30 pm DeArmond Lecture** (Adams Ballroom)

Novel MRI-based Platform for Efficient Gene Delivery to the Brain

Krystof Bankiewicz, MD, PhD

University of California San Francisco, San Francisco, CA

6:30 – 8:30 pm Annual Reception Mezzanine Lobby

# Poster Session I:

(1401.0	nered for Civic Credit)
33	Autopsy Findings in a Case of Langerhans Cell Histiocytosis-associated
	Neurodegeneration
	Randall Woltjer, and Garth Warren
34	Extensive Pontine and Extrapontine Myelinolysis in a Patient with X-linked Myopathy
	with Excessive Autophagy
	Bojana Mitrovic, Amer Ghavanini, Berge Minassian, David Munoz
35	Fatal Bactrim-Associated Hypersensitivity In A Patient With Auto-Immune Neuro-
	Vasculitis
	Anne Lee, Alexander Diaz de Villalvilla, Nicole Alexander, Deena Kuruvilla, Mohammad Mahboob, Jeffrey
	Rogg, Molly Tracy, Shamlal Mangray, Suzanne DeLa Monte
36	Graft Versus Host Disease of the Brain Following Allogeneic Stem Cell Transplant for
	Myelodysplastic Syndrome
	Janet Yoo, Lindsay Simon, Zoe Wang, Lawrence Kenyon
37	Lymphoplasmacytic Hypophysitis: An IgG4-related Disease?
	Declan McGuone, Anat Stemmer-Rachamimov, John Stone, Vikram Deshpande
38	MS with Abundant Spinal Cord and Cortical Demyelination, but Rare Cerebral White
	Matter Demyelination
	Megan Hendrickson, Ansi Chang, Susan Staugaitis, Kathryn Easley, Lidia Sviderskaya, Robert Fox,
	Bruce Trapp
39	Neuromyelitis Optica Spectrum Disorder (NMOSD) with Predominant Active Cerebral
	Involvement: A Case Report
	Patrick Malafronte, Nicole DeSimone, Donna Graves, Charles White, Dennis Burns
40	Necrotizing Myelopathy in a Young Subject. Neuromyelitis Optica Variant? Case Report
	Miguel Riudavets, Naomi Arakaki, Correale Jorge, Fernanda Diaz, Gustavo Sevlever
41	Association between Cortical Malformation and Medically Intractable Epilepsy in
	Children with Sturge-Weber Syndrome
	Li Chen, Anna Pinto, Annapurna Poduri, Mustafa Sahin, Sanjav Prabhu, Masanori Takeoka, Hart Lidov
42	Meningioangiomatosis: Review of Five Cases
	Tracie Pham, Kristina Takahashi, Jason De Jesus, William Yong, Gary Mathern, Harry Vinters
43	Neuropathologic Findings in the Fetal Alcohol Syndrome (FAS): A Rare Case Study of
	an Adult Brain
	Catherine Stoos, Laura Nelsen, Amy Elliot, Kathryn Schissler, Hannah Kinney
44	Neuropathologic Insights into the X-Linked Leukodystrophy Pelizaeus-Merzbacher
	Disease
	Jeremy Laukka, Kathryn Lovell, Anders Sima, Skoff Robert, John Kamholz
45	Neuropathology of 22q11 Deletion Syndrome (22q11DS) in an Infant
	Kathryn McFadden, Peter Wu, Geoffrey Murdoch
46	Orbitocranial Intravascular Papillary Endothelial Hyperplasia and Microphthalmia: A
	Case Report
	Jose Bonnin, Chie-Schin Shih, Richard Burgett, Joel Boaz, Chang Yueh Ho
47	Revisiting the Neuropathology of Late Infantile Neuronal Ceroid Lipofuscinosis in the
	Molecular Age
40	Leslie Hamilton, Aneal Khan, Ismail Mohamed, Jeffrey Joseph
48	Searching for a Possible CSF Biomarker of Sudden and Unexpected Infant Death
	Ingvar Rognum, Elisabeth Haas, Keith Hyland, David Paterson, Robin Haynes, Kevin Broadbelt, Brian
40	Harty, Henry Krous, Hannah Kinney
49	Sensory Ganglion Cells in the Third Nerve: An Anatomical Curiosity
	Douglas Miller

# FRIDAY, JUNE 22, 2012 Exhibit Hall, Fourth Floor

# Poster Session I Continued:

50	Peters' Anomaly with Multiple Congenital Malformations: Light and Electron
	Microscopic Study of a Case
	Jason Wells, Cathy Housman, Joel Weinstein, David Liang, Charles Specht
51	Cerebral Vascular Abnormalities in Signal Transducer and Activator of Transcription 3
	(STAT3) Deficiency: A Neuropathological
	Francoise Gray, Marie-Olivia Chandesris, Capucine Picard, Soraya Taleb, Kim-Thanh Ong, Arshid
	Azarine, Alain Fischer
52	Eclampsia-Associated Double-Simultaneous Supra- and Infratentorial Fatal
	Hypertensive Intracerebral Hemorrhages
	Mohammad Mahboob, Sonja Chen, Anne Lee, Daniel Aghion, Adetokunbo Oyelese, William Martland,
50	Suzanne de la Monte
53	Intraventricular Cavernous Angioma With Superficial Siderosis
E A	James Hackney, Cheryl Palmer, James Markert, L. Nabors  SUR1 Protein Expression in Human Stroke
54	Rupal Mehta, Svetlana Ivanova, Cigdem Tosun, Rudy Castellani, Volodymyr Gerzanich, J Simard
55	Alzheimer Disease Associated with the I229F PSEN1 Mutation: PiB and Neuropathologic
55	Studies
	Bernardino Ghetti, Jill Murrell, Karmen Yoder, Shannon Risacher, Bradley Glazier, Martin Farlow,
	Frederick Unvezagt, Kathy Newell, Andrew Saykin
56	Clinical and Neuropathologic Heterogeneity in PSEN1 A431E
	Jill Murrell, Rupal Mehta, Eileen Bigio, Nigel Cairns, Elizabeth Cochran, Darren Gitelman, Elizabeth Head,
	James Leverenz, Wayne Poon, Masaki Takao, Sandra Weintraub, John Ringman, Bernardino Ghetti
57	Early-Onset Alzheimer Disease with the G209E PSEN1 Mutation: A Neuropathologic
	Study
	Jose Bonnin, Jill Murrell, Martin Farlow, Bradley Glazier, Tatiana Foroud, Bernardino Ghetti
58	Eliminating Background Signal with ELISA Analysis of Murine Beta-Amyloid
	Mitesh Patel, Ottavio Arancio, Andrew Teich
59	Neuropathologic Assessment of ADNI Participants: the Essential Role of the
	Neuropathology Core
00	Nigel Cairns, Lisa Taylor-Reinwald, John Morris  Neuropathological Expression of the PSEN1 I229F Mutation in 2 Family Members
60	Kathy Newell, Jill Murrell, Cynthia Gouvion, Francine Epperson, Bernardino Ghetti
61	Neuropathology of Early-Onset Familial Alzheimer Disease Associated with PSEN1
01	Y115C Mutation
	Andrea Wiens, Salvatore Spina, Jill Murrell, Martin Farlow, Ann Hake, Francine Epperson, Rose
	Richardson, Brenda Dupree, Jose Bonnin, Bernardino Ghetti
62	oAß Induces mRNA Dysregulation in the 5XFAD Mouse Model of Alzheimer's Disease
_	and Rat Frontal Cortex Organotypic Cultures
	Celia Williams, Chiara Ferrari, Sarah Herrman, Anu Ramachandran, Amit Rajaram, Carol Miller
63	Poly(ADP-ribose) Polymerase-1 Expression in Alzheimer's Disease
	Jianying Zeng, Jenny Libien, Fatima Shaik, Olga Krasnozhen, A. Iván Hernández
64	Reclassification of "Not Classifiable" Cases with AD Pathology According to NIA-AA
	Guidelines for ADNC
	Kyung-Hwa Lee, Qinwen Mao, Rakhee Ganti, Eileen Bigio
65	The Neuropathology of Alzheimer Disease in the Setting of Chronic Traumatic
	Encephalopathy
	Thor Stein, Victor Alvarez, Ann McKee
66	The Use of Digital Pathology and Image Analysis to Rapidly Quantitate Alzheimer's
	Disease Neuropathologic Changes
	Janna Neltner, Stephanie Denison, Ela Patel, Sonya Anderson, Peter Nelson

# Poster Session I Continued:

67	Characterization of Perivascular ABeta and Tau Immunoreactivity in Association with
	Cerebral Amyloid Angiopathy
	Tracie Pham, Emad Farag, Spencer Tung, Kristina Takahashi, Eric Chu, Harry Vinters
68	C9ORF72 Expansion in Hippocampal Sclerosis Reaffirms its Classification Within the
	Spectrum of Frontotemporal Dementias
	Olga Pletnikova, Kelly Sloane, Bryan Traynor, Barbara Crain, Juan Troncoso, Peter Rabins, Chiadi
	Onyike
69	Case Report: Novel Gly141X SOD1 Mutation in Familial Frontotemporal Dementia and Amyotrophic Lateral Sclerosis
	Masataka Nakamura, Melissa Murray, Wen-Lang Lin, Monica Castanedes-Casey, Neill Graff-Radford,
	Kevin Boylan, Mariely DeJesus-Hernandez, Nicola Rutherford, Rosa Rademakers, Dennis Dickson
70	Cytoprotective Chemical Screens in a Yeast Model for α-synucleinopathies
, 0	Pavan Auluck and Susan Lindquist
71	Disease-specific Neuronal inclusions in c9FTD/ALS with the Ubiquitin-binding Proteins
	Ubiquilin 2 and P62/Sequestosome-1
	Kevin Bieniek, Melissa Murray, Wen-Lang Lin, Mariely DeJesus-Hernandez, Nicola Rutherford, Matthew
	Baker, Monica Castanedes-Casey, Neill Graff-Radford, Kevin Boylan, Rosa Rademakers, Dennis Dickson
72	Frontotemporal Dementia And Motor Neuron Disease Associated With C9ORF72
	Mutation: A New Family
	Salvatore Spina, Martin Farlow, Frederick Unverzagt, Jill Murrell, Bernardino Ghetti
73	Hereditary Diffuse Leukoencephalopathy With Spheroids: Clinicopathologic Study Of
	Three Cases
7.4	Salvatore Spina, Jill Murrell, Frederick Unverzagt, Martin Farlow, Bernardino Ghetti
74	Retrograde Trafficking Defects Enhance α-Synuclein Toxicity in a Yeast Model
75	Pavan Auluck and Susan Lindquist,  A Rare Case of Secondary Gliosarcoma with Extracranial Metastases
73	Richa Dawar, Nikhil Khushalani, Andrew Fabiano, Jingxin Qiu
76	Aggressive Histologic Features in Angiocentric Glioma: Clinical Correlation
, 0	Sakir Gultekin and Mark Hiken
77	Assessment of RNA Extraction Methods for the Real-Time PCR Analysis of the
	BRAF:KIAA1549 Fusion Gene in Pilocytic Astrocytoma
	Francesca Brett, Patrick Buckley, Joanne O'Sullivan
78	BMP Signaling Reduces Proliferation and Upregulates p57 mRNA in Oncogenic Murine
	Neural Stem Progenitor Cells
	Laura Hover, Philip Owens, Alex Munden, Ty Abel
79	Differential Expression of Ketone Metabolizing Enzymes in Malignant Gliomas:
	Implication for Ketogenic Diet Therapy
00	Howard Chang, Lawrence Olson, Kenneth Schwartz
80	Ganglion Cells in Low-Grade Circumscribed/Cystic Astrocytic Tumors: Possible
	Implication in Classification and Prognosis L. Chimelli, VG Moreira, N Canedo
81	Genetic and Pathologic Evolution of Secondary Gliosarcoma
01	Kari-Elise Codispoti, Stacy Mosier, Ming-Tseh Lin, Robert Ramsey, Fausto Rodriguez
82	Metabotropic Glutamate Receptor Type 5 in Human Glioma
52	Hilary Nickols, Jerri Rook, Nellie Byun, P. Conn
83	Multiply Recurrent Glioblastoma in a Young Woman with Lynch Syndrome: Complete Autopsy
	Examination with Clinical Correlation
	Hidehiro Takei, Nitin Tandon, Clark Sitton, Jay-Jiguang Zhu, Meenakshi Bhattacharjee

# FRIDAY, JUNE 22, 2012 Exhibit Hall, Fourth Floor

# Poster Session I Continued:

	Mutated lossitrate Dehydrogenese 4 (IDH 4) Immunerasetive Clicklestoms with IDH 4
84	Mutated Isocitrate Dehydrogenase-1 (IDH-1) Immunoreactive Glioblastoma with IDH-1
	Negative 'PNET- like' Elements
0.5	Marie Rivera-Zengotita, Kelly Devers, Anthony Yachnis
85	Oligosarcoma: Further Evidence for Mesenchymal Metaplastic Change in Neoplastic
	Oligodendrocytes P. C. "
	Michal Raz, Deborah Blumenthal, Zvi Ram, Dov Soffer
86	Oncocytic Ependymoma: A Case Report
87	Veena Rajaram, Corey Bregman, Wendy Stellpflug, Tomita Tadanori, Jason Fangusaro  Pilot Study of Digital Image Analysis as a Way to Predict Likelihood of 1p/19q Codeletion
07	in Diffuse Gliomas
	Meggen Walsh, Fuyong Xing, Janna Neltner, Lin Yang, Craig Horbinski
88	Pituicytoma With Gelsolin Amyloid Deposition
00	Cristiane Ida, Mark Jentoft, Xiaoling Yan, Joseph Parisi, Ahmet Dogan, Kalman Kovacs, Bernd
	Scheithauer
89	TGFß Signaling in Glioma Microenvironment Inhibits Engraftment and Differentiation of
00	Oncogenic Neural Stem/Progenitor Cells
	Sabah Ghazi, Alex Munden, Laura Hover, Ty Abel
90	TGF-ß Signaling Promotes Astrocytic Differentiation in Normal and Oncogenic Neural
00	Stem/Progenitor Cells
	Sabah Ghazi, Alex Munden, Laura Hover, Ty Abel
91	Recurrent Desmoplastic Non-infantile Ganglioglioma in Late Adulthood
	Jo Elle Peterson, Andreana Rivera, Hidehiro Takei, Suzanne Powell
92	Synchronous Dysembryoplastic Neuroepithelial Tumor and Extraventricular
	Neurocytoma in a 14-year-old with Epilepsy
	Ian White, Sarah Martin, Jody Smith, Eyas Hattab
93	Aggressive Cerebellar Neuroepithelial Tumor in a 10 Year Old Boy
	Hannes Vogel, Michael Edwards, Paul Fisher, Sonia Partap, Yoon-Jae Cho
94	Anaplastic Meningioma with Single Cell Infiltration and Loss of E-Cadherin
	Immunoreactivity
	Brett Danielson, David Munoz, Jason Karamchandani
95	Are NF2-Associated Meningiomas Morphologically or Immunohistochemically
	Distinguishable from Sporadic Ones?
	Andrea Wiens and Eyas Hattab
96	Arrest Defect 1 (ARD1) Protein Expression in Primary Central Nervous System
	Lymphomas (PCNSLs)
07	Jessica Levesque, Christine Sheehan, Jeffrey Ross, Jiang Qian
97	Atypical Teratoid Rhabdoid Tumor of the Pineal Region in an Eleven-week Old Infant with a Germline INI1 Mutation
	Leili Mirsadraei, Tom Davidson, Whitney Pope, Kristanapol Boon-Unge, Jason Hauptman, Jorge Lazareff,
	Harry Vinters, Negar Khanlou, William Yong
	1 - 7

### Platform 5: Muscle/Nerve/Other Chairpersons: Juan Bilbao, MD and Michael Lawlor, MD, PhD

8:00- 8:15	98	Clinical Utility of LC3 and p62 Immunohistochemistry in Diagnosis of Drug-induced Autophagic Vacuolar Cardiomyopathy
		Brianne Daniels, Rodney McComb, Bret Mobley, S. Gultekin, Han Lee, Marta Margeta
8:15- 8:30	99	Satellite Cell Depletion and Dysfunction Correlates with Disease Progression in
		Severe Murine Myotubularin Deficiency
		Michael Lawlor, Matthew Alexander, Marissa Viola, Hui Meng, Vandana Gupta, Norio
		Motohashi, Richard Manfready, Cynthia Hsu, Ping Huang, Romaine Joubert, Anna Buj-Bello,
		Louis Kunkel, Alan Beggs, Emanuela Gussoni
8:30- 8:45	100	Evaluation of Calpain-3 In Patients with Limb-Girdle Muscular Dystrophy
		Steven McGaughey, Terese Nelson, Mary Cox, Steven Moore
8:45- 9:00	101	Diagnosis of Mitochondrial Disorders Using a Tissue-based Mitochondrial
		Immunofluorescence Assay - A Five Case Illustration
		Adekunle Adesina, Vidya Mehta, Benjamin Ellezam, Dmitriy Niyazov, Tim Lotze, Lee-Jun
		Wong, Fernando Scaglia
9:00- 9:15	102	Ultrastructural Pathology and Functional Deficits Can Be Reversed Following
		Enzyme-Replacement in Myotubularin Deficient Mice
		Michael Lawlor, Dustin Armstrong, Marissa Viola, Hui Meng, Anna Buj-Bello, Cynthia Hsu,
		Christopher Pierson, Martin Childers, Robert Grange, Jeffrey Widrick, Alan Beggs
9:15- 9:30	103	Novel Beta-Tropomyosin Mutation (TPM2) Causes Distal Core-Rod Myopathy
		And Trismus-Pseudocamptodactyly With Nemaline Rods
		Steven Moore, Brian Moore, Seth Love, Jun Li, Michael Lopez, Maja von der Hagen, Angela
		Huebner, Thomas Winder, Thomas Cullup, Hossam Abdel-Salam, Fazeel Siddiqui, Anirban
0.00.0.45	404	Majumdar, Peter Lunt, James Dowling
9:30- 9:45	104	Clinical Utility of LC3 and p62 Immunohistochemistry in Diagnosing the
		Spectrum of T-cell Mediated Inflammatory Myopathies
		Annie Hiniker, Brianne Daniels, Han Lee, Marta Margeta
9:45- 10:00	105	Cross-sectional Versus Flat-mount DSAEK Specimen Preparations: Quantitative
		Differences and Association with Visual Acuity
		T. Bourne, David Shiple, Leslie Olsakovsky

10:00 - 10:30 am REFRESHMENT BREAK

10:30 – 11:30 am Saul Korey Lecture

Why the Brain Fails when the Astrocyte Ails

Michael Norenberg, MD

University of Miami School of Medicine, Miami, FL

11:45 am – Business Meeting II (Adams Ballroom) 12:45 pm

12:45 – 2:00 pm Lunch

SATURDAY, JUNE 23, 2012 Monroe Ballroom 8:00 am - 2:00 pm

### Platform 6: Pediatric Tumor/Developmental and Pediatric Neuropathology Chairpersons: Harry Vinters, MD and David Ellison, MD, PhD

8:00- 8:15	106	Cobblestone-Lissencephaly Encompasses 3 Subtypes Correlated to Genes of
0.00 0.10	100	Alpha-dystroglycanopathies
		Louise Devisme, Céline Bouchet, Marie Gonzalès, Sandrine Vuillaumier, Tania Attié-Bitach,
		Nathalie Seta, Ferechte Encha-Razavi
0.45 0.00	407	
8:15- 8:30	107	Low Density of Layer IV Neurons in Prefrontal Cortex in Autism
0.00.0.15	100	John Hedreen
8:30- 8:45	108	Splice Site GFAP Mutation in Alexander Disease
		Rong Li, Cheryl Palmer, Michael Brenner
8:45- 9:00	109	Diffuse Leptomeningeal Glioneurinal Tumor, a Newly Recognized Pediatric
		Neoplasm
		Veena Rajaram, Jessica Stern, Maura Ryan, Tord Alden, Stewart Goldman, Rishi Lulla
9:00- 9:15	110	Disseminated Oligodendroglial-like Leptomeningeal Tumor of Childhood - A
		Distinctive Indolent Neoplasm
		Fausto Rodriguez, Arie Perry, Marc Rosenblum, Sherry Krawitz, Charles Eberhart, Peter
		Burger
9:15- 9:30	111	RAF Gene Abnormalities Define Subsets of Pediatric Low-grade Gliomas and
		Glioneuronal Tumors
		David Ellison, Ruth Tatevossian, Ibrahim Qaddoumi, Bo Tang, James Dalton, Sheila Shurtleff,
		Chandanamali Punchihewa, Wilda Orisme, Geoffrey Neale, Amar Gajjar, Suzanne Baker,
		Denise Sheer
9:30- 9:45	112	Identification of Biologically Relevant Targets in Pilocytic Astrocytoma by
		MicroRNA Profiling
		Cheng-Ying Ho, Eli Bar, Caterina Giannini, Matthias Karajannis, David Zagzag, Charles
		Eberhart, Fausto Rodriguez
9:45- 10:00	113	Deregulation of PI3K/AKT Pathway Is Frequent in Pilocytic Astrocytoma with
0.10 10.00	' '	Anaplastic Features
		Adriana Olar, Diep Tran, Vidya Mehta, Benjamin Ellezam, Carrie Mohila, Gerald Campbell,
		Suzanne Powell, Gregory Fuller, Kenneth Aldape, Adekunle Adesina

10:00 - 10:30 am REFRESHMENT BREAK

10:30 – 11:30 am Saul Korey Lecture

Why the Brain Fails when the Astrocyte Ails Michael Norenberg, MD

University of Miami School of Medicine, Miami, FL

11:45 am – Business Meeting II (Adams Ballroom) 12:45 pm

12:45 – 2:00 pm Lunch

### Platform 7: Tumors II

Chairpersons: Catherina Giannini, MD, PhD and Ryan Miller, MD, PhD

2:00- 2:15	114	Genomic Gains Acquired During Glioblastoma Progression Obscure Driver-
		Specific Signatures Present in Low-grade Astrocytomas
		C. Miller, Mark Vitucci, Ryan Bash, Ralf Schmid
2:15- 2:30	115	Chromatin Landscape Analysis to Identify the Core Transcriptional Regulatory
		Network of Glioblastoma Cancer Stem Cells
		Mario Suvà, Esther Rheinbay, Andrew Chi, David Louis, Bradley Bernstein
2:30- 2:45	116	Study of Genetic and Epigenetic Alterations in Paediatric Glioblastomas
		Chitra Sarkar, Prerana Jha, Irene Pia Patrick, Kumaravel Somasundaram, Pankaj Pathak,
		Mehar Sharma, Vaishali Suri, Ashish Suri
2:45- 3:00	117	IDH Mutation and Neuroglial Developmental Features Define Distinct
		Subclasses of Lower-Grade Diffuse Astrocytic Glioma
		Daniel Gorovets, Kasthuri Kannan, Ronglai Shen, Edward Kastenhuber, Timothy Chan, Jason
		Huse
3:00- 3:15	118	Patterns of Repressive Histone 3 Lysine 9 Trimethylation (H3K9me3) in
		Isocitrate Dehydrogenase Mutant and Wild Type Gliomas
		Sriram Venneti, Michelle Madden, Thomas Coyne, Joanna Phillips, Jason Huse, Chao Lu,
		Tarik Tihan, Lisa Sullivan, Mariarita Santi, Alexander Judkins, Craig Thompson, Arie Perry
3:15-3:30	119	Ki-67 on the Web: A Browser-Based Biomarker Web Application for Analysis of
		the Ki-67 Proliferation Index
		James Hackney, Jonas Almeida, Sean Wilkerson, Benjamin Hill
3:30- 3:45	120	Genetic Profiling of Orbital and Optic Nerve Meningiomas by Single-Nucleotide
		Polymorphism-Based Array Analysis
		Cheng-Ying Ho, Stacy Mosier, Charles Eberhart, Denise Batista, Fausto Rodriguez
3:45- 4:00	121	Hsa-miR-383 and its Target Peroxiredoxin 3 (PRDX3) Have Major Roles Controlling Cell
		Growth in Medulloblastoma
		Ho Keung Ng

### 4:00 - 4:30 pm REFRESHMENT BREAK

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5:15 - 5:30 pm

History of Chicago Neuropathology John M. Lee, MD, PhD Loyola University, Maywood, IL

### 5:00 – 5:15 pm What Every Neuropathologist Needs to Needs to Know

NIA-AA Revised Guidelines for the Diagnosis of Alzheimer's Disease Julie Schneider, MD Rush University Medical Center, Chicago, IL

# 30 pm What Every Neuropathologist Needs to Needs to Know

IDH1 and its Practical Utility in Glioma Diagnosis David N. Louis, MD Massachusetts General Hospital, Boston, MA Platform 8: Neurodegenerative: Other II

Chairpersons: Nigel Cairns, PhD and Eileen Bigio, MD

Chairpers	ions: i	Nigel Cairns, PhD and Elleen Biglo, MD
2:00- 2:15	122	Latent Trait Analysis Identifies Genetic Determinants of Glial Tau Pathology in
		Progressive Supranuclear Palsy
		Dennis Dickson, Daniel Serie, Melissa Murray, Mariet Allen, Nilufer Ertekin-Taner, Julia Crook
2:15- 2:30	123	Parkinson-Related LRRK2 Mutations Result in Impaired Mitochondrial
		Dynamics and Function via Direct Interaction with DLP1
		George Perry, Michael Yan, Hisashi Fujioka, Jun Liu, Amy Wilson-Delfosse, Shu Chen,
		Gemma Casadesus, Xiongwei Zhu, Xinglong Wang
2:30- 2:45	124	Amyloid Imaging with [11C] PiB PET in Gerstmann-Sträussler-Scheinker
		Disease
		Shannon Risacher, Karmen Yoder, Andrew Saykin, Gary Hutchins, Qi-Huang Zheng, Jill
		Murrell, Bradley Glazier, Francine Epperson, Martin Farlow, Bernardino Ghetti
2:45- 3:00	125	Poly(ADP-ribose) Accumulates in FTLD-TDP and a Cuprizone-treated GRN-/-
		Mouse
		Mingqiang Xie, Hua Shen, Sheng-Kwei Song, Nigel Cairns
3:00- 3:15	126	Genetic, Clinical, And Pathological Associations Of TDP43pathy With
		Cerebellar P62 Inclusions (TDP43 Plus)
		David Munoz, Ekaterina Rogaeva, Lorne Zinman, Beverly Young, Mario Masellis, Sandra
		Black, Janice Robertson, Julia Keith, Juan Bilbao
3:15-3:30	127	Analyses of the c9ORF72 Expanded Repeat in a Series of Autopsy Cases
		Jill Murrell, Bernardino Ghetti, Martin Farlow, Francine Epperson, Edward Huey, Jordan
		Grafman, Gregory Jicha, Salvatore Spina
3:30- 3:45	128	Feasibility Study of Needle Core Biopsy of the Submandibular Gland for the
		Diagnosis of Parkinson's Disease
		Thomas Beach, Jose Hidalgo, Jonette Henry-Watson, Geidy Serrano, Monica Mariner, Lucia
		Sue, Marwan Sabbagh, Holly Shill, Charles Adler
3:45- 4:00	129	FTDP-17 with MAPT Exon 13 mutations: Comparison of Neuropathologic
		Features of Gly389Arg to a Novel mutation, Glu372Gly
		Naomi Kouri, Joseph Parisi, Ronald Petersen, Matthew Baker, Rosa Rademakers, Dennis
		Dickson

### 4:00 - 4:30 pm REFRESHMENT BREAK

### 

History of Chicago Neuropathology John M. Lee, MD, PhD Loyola University, Maywood, IL

### 5:00 – 5:15 pm What Every Neuropathologist Needs to Needs to Know

NIA-AA Revised Guidelines for the Diagnosis of Alzheimer's Disease *Julie Schneider, MD* 

Rush University Medical Center, Chicago, IL

### 5:15 – 5:30 pm What Every Neuropathologist Needs to Needs to Know

IDH1 and its Practical Utility in Glioma Diagnosis

David N. Louis, MD

Massachusetts General Hospital, Boston, MA

### Poster Session II:

(INOL O	nered for Civic Credit)
130	Biopsy Pathology of HIV-2 Encephalitis: Case Report
	Pedro Ciarlini, Brian Wood, Joshua Klein, Jennifer Lyons, Danny Milner, Richard Philips, Martin Schutten,
	Timothy Henrich, Jennifer Johnson, Dana Gabuzda, Umberto De Girolami, Rebecca Folkerth
131	Cerebral Phaeohyphomycosis: A Report of Two Cases
	Fahad Bafakih, Cara Sedney, P. Lasala, Kymberly Gyure
132	Immunohistochemistry Using IDH1 Mutant-specific Antibody in Progressive Multifocal
	Leukoencephalopathy - Report of a Case
	Kimberly Stogner-Underwood, Knarik Arkun, Christine Fuller
133	Oncogenic Brain Metazoan Parasitic Infection
	Douglas Miller, Angela Spurgeon, Gabor Oroszi, Qing-Qing Ding, Marshall Cress, Tomoko Tanaka
134	Progressive Neuronal Pathology Associated with Latent Herpes Simplex Virus Infection
	in Mice
	Tibor Valyi-Nagy, Sandor Dosa, Karla Castellanos, Sarolta Bacsa, Eva Gagyi, Krisztian Kovacs,
	Bernadett Kormos, Klara Valyi-Nagy
135	Rabies Encephalitis of Bat-Strain Type in a 63 Year Old Man: Failure of the Milwaukee
	Protocol
	Declan McGuone, Eyal Kimchi, Anne Neilan, Gregory Robbins, Matthew Frosch, E. Tessa Hedley-Whyte
136	Hypopituitarism and Diabetes Insipidus in a Patient with Wegener Granulomatosis
.00	Pedro Ciarlini, Garni Barkhoudarian, Edward Laws, Li Chen, Umberto De Girolami
137	Whipple's Disease Masquerades as Dementia with Lewy Bodies: A Case Report
.07	Kyle Hurth, Robert Schmidt, Rawan Tarawneh, Nupur Ghoshal, Tammie Benzinger, David Clifford,
	Michael Geschwind, John Morris, James Galvin, Nigel Cairns
138	A Rare Case Of Monocular Visual Loss Caused By An Isolated Neurosarcoidosis
	Rocky Adams and Jeffrey Sosnowski
139	ECRG4 and its Product Augurin in Human Choroid Plexus and Hypothalamus:
	Implications for CSF Homeostasis and Fluid Balance
	Edward Stopa, Miles Miller, Ryan Rossi, Jasmine Chukwueke, Ibrahem Salloum, Sonia Podvin, John
	Donahue, Xitong Dang, Ana Gonzalez, Brian Eliceiri, Conrad Johanson, Andrew Baird
140	Immunoglobulin Lambda Light Chain Deposition Disease Presenting as a Periventricular
	White Matter Lesion
	Joshua Menke, Mark Jentoft, Ahmet Dogen, James Avent, Dylan Miller, Caterina Giannini
141	Lyophilization of Brain Tumor Biospecimens Significantly Limits DNA and RNA
	Degradation from Freeze-thaw Cycles
	Sergey Mareninov, Linda Liau, Desiree Sanchez, Ryan Wilson, Harry Vinters, Negar Khanlou, Tracie
	Pham, Paul Mischel, Timothy Cloughesy, William Yong
142	Point by Point Histopathological Correlation of Subfield Hippocampus in Identical
	Planes From high Field 7T MRI
	Kant Matsuda, William Wu, Ke Zhang, David Zagzag, Odet Gonen
143	National Brain and Tissue Resource for Parkinson's Disease and Related Disorders
	Thomas Beach, Charles Adler, Holly Shill, John Caviness, Marwan Sabbagh, Lucia Sue, Douglas Walker,
	LihFen Lue, Geidy Serrano, Alex Roher, Joseph Hentz, Brittany Dugger
144	Danon disease in a Chinese Family with Atypical Presentation
•	Amanda Kan and Sophelia Chan
145	Histopathological Abnormalities are Progressive in Myotonic Dystrophy Type 2
	Sijie Wang, Kevin Felice, Qian Wu
146	In-Frame Mutation in the WW-CR Dystrophin Domain: Case with DMD Phenotype and
	Relatively Preserved Dystrophin Expression
	Divisha Raheja, Cathy Housman, Anthony Giordano, Matthew Wicklund, Charles Specht
147	Myofibrillar Myopathy with Large Cores: A Case Report
17/	Sarah Martin, Steven Moore, Cynthia Bodkin, Eyas Hattab
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### SATURDAY, JUNE 23, 2012 Exhibit Hall, Fourth Floor

### Poster Session II Continued:

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148	Novel Homozygous Stop Mutation in AlphaB Crystallin (CRYAB:p.Ser135*) Causes Fatal Congenital Form of Myofibillar Myopathy
	Steven Moore, Thomas Winder, Terese Nelson, Mary Cox, Christine Reyes, Caroline Tesi-Rocha, Taeun Chang
149	Significance of Negative Sural Nerve Biopsy in Neurolymphomatosis
143	Kritsanapol Boon-Unge, Anthony Verity, Perry Shieh, Nader Pouratian, Negar Khanlou
150	Trabecular Myopathy: Clinical-Pathologic Correlation and Ultrastructural Analysis in
100	Five Cases
	Tracie Pham, M. Verity, Jennifer Yi, Kritsanapol Boon-Unge, William Yong, Perry Shieh, Harry Vinters,
	Jason Peragallo, Joseph Demer, Negar Khanlou
151	Tau Phosphorylation and Truncation at D421 Precedes Danish Amyloid Deposition in
	Mice Expressing Mutant BRI2 and TauP301S
	Holly Garringer, Jill Murrell, Neeraja Sammeta, Leticia Miravalle, Bernardino Ghetti, Ruben Vidal
152	A Case of Globular Glial Tauopathy Presenting Clinically as Alzheimer's Disease
102	Karen SantaCruz, Susan Rottunda, Joyce Meints, Eileen Bigio, J. McCarten
153	Abundant Tau Accumulation in Gerstmann-Sträussler-Scheinker Disease Associated
100	with the PRNP P102L-129M Mutation
	Masaki Takao, Mariko Takada, Katsuhisa Ogata, Mikiya Suzuki, Yoji Yoshida, Ban Mihara, Shinji Ito,
	Akane Nogami, Sayaka Funabe, Hiroyuki Hatsuta, Mitsuru Kawai, Shigeo Murayama, Takayuki Haga,
	Tetsuyuki Kitamoto
154	Brown-Vialetto-Van Laere Syndrome: A Case Report with Immunohistochemistry for
	C20orf54
	Patrick Malafronte, Irene Castaneda-Sanchez, H Clark, Charles White, Kimmo Hatanpaa
155	Disruption of the Cerebellar Module Causes the Main Clinical Phenotype in Hereditary
	Ataxia
	Arnulf Koeppen, Sarah Collins, R Ramirez, Peter Bauer
156	Immunohistochemical Study and Western blotting of Tauopathy in ALS/ Parkinson
	Dementia Complex(PDC), Kii, Japan
	Satoru Morimoto, Yasumasa Kokubo, Masato Hasegawa, Shigeki Kuzuhara, Shigeo Murayama
157	Juvenile-onset Tauopathy with Parkinsonism and Chronic Pancreatitis in Two Brothers:
	A Clinical, Genetic, and Autopsy Study
	Leonidas Arvanitis, Elizabeth Berry-Kravis, Christopher Goetz, Jamie Jacobsohn, Christian Kubisch,
	Elizabeth Cochran
158	Neuronal TDP-43-Positive Inclusions in the Spinal Cord Distinguish ALS from FTLD-TDP
450	Marla Gearing, Deborah Cooper, Jonathan Glass
159	Neuropathologic Findings in Myotonic Dystrophy Type 2 Marie Rivera-Zengotita, Guang-bin Xia, Kelly Devers, Burt Martha, Tetsuo Ashizawa, Anthony Yachnis
160	Neuropathology of Rapidly Progressing Parkinsonism and Dystonia in a P102L PRNP
100	Mutation Carrier
	Bernardino Ghetti, Chizoba Umeh, Piyush Kalakoti, Michael Greenburg, Pierluigi Gambetti, Zoltan Mari
161	Niemann-Pick Disease Type C Associated with 2 Mutations in the NPC1 Gene
101	Kathy Newell, Russell Swerdlow, Bernardino Ghetti
162	Pick disease with Severe Involvement of the Brainstem and Glia
102	Jose Bonnin, Andrea Wiens, Jill Murrell, Martin Farlow, Bernardino Ghetti, Frederick Unverzagt
163	Prion Disease in Virginia: A Demographic and Phenotypic Analysis of Cases from 2003
103	to 2012
	Heather Sumner, Ignazio Cali, Janis Blevins, M. Beatriz Lopes
164	Prion Protein Deposition in the Outer Plexiform Layer of the Retina in Gerstmann-
104	Sträussler-Scheinker Disease
	Andrea Wiens, Martin Farlow, Jill Murrell, Francine Epperson, Rose Richardson, Pedro Piccardo, Jose
	Bonnin, Bernardino Ghetti
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### SATURDAY, JUNE 23, 2012 Exhibit Hall, Fourth Floor

### Poster Session II Continued:

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165	Progressive Supranuclear Palsy-like Tauopathy in Gerstmann-Straussler-Scheinker
	Syndrome: Report of a Case
	Thomas Huebner, Marian Lamonte, Rupal Mehta, Rudy Castellani
166	Rapid Onset Dystonia-Parkinsonism Associated with the I758S ATP1A3 Mutation: A
	Neuropathologic Study of Two Affected Siblings
	Bernardino Ghetti, Allison Brashear, Matthew Hagen, Kathleen Sweadner
167	Bilateral Cranial Nerve VII-VIII Malignant Peripheral Nerve Sheath Tumors Mimicking
	Neurofibromatosis Type 2
	Kathy Newell and Ania Pollack
168	Case Report: Solitary Intracranial Plasmacytoma Comprised of Atypical Plasma Cells: A
	Diagnostic Challenge on Frozen Sections
	Yunguang Liu and Jingxin Qiu,
169	Central Nervous System Desmoplastic Small Round Cell Tumor. Case Report
	Miguel Riudavets, Naomi Arakaki, Elisabeth Rushing, Ana Lia Taratuto, Gustavo Sevlever
170	Choristoma of the Trochlear Nerve in a 5-Month-Old Child: Clinical Features and Light
	Microscopic Study of a Case
	Meghan Riley, Karmaine Millington, Mark Dias, Arabinda Choudhary, Charles Specht
171	EBV-associated Lymphoproliferative Disorder in a Dermatomyositis Patient with
	Clinicoradiological Follow-up at 15 months
	Matthew Cykowski, Kar-Ming Fung, Eduardo De Sousa, David Parham, Lichao Zhao, Ethan Stolzenberg
172	FOXG1 Modulation Suggests Possible Role in Deregulation of Neuronal Differentation
	and Poor Survival in Medulloblastoma Model
	Adekunle Adesina, Girarrd Courteau, Vidya Mehta, Xiao-Nan Li
173	Further Evidence of the Non Germinal Center Phenotype and Likely Origin of Primary
	Central Nervous System Lymphoma
	Alicia Hirzel, Cristina Vincentelli, Diana Morlote, Amilcar Castellanos-Sanchez
174	Germinoma with Anaplastic Features and Atypical Presentation as a Large Cerebral
	Hemispheric Mass
	Jennifer Ross and Adekunle Adesina
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177	Immunohistochemical Diagnosis of Silent Subtype III Pituitary Adenoma
470	Derek Mathis, Aaron Tauer, Charles White, Kimmo Hatanpaa
178	Incidental Dural-based Marginal Zone B-cell Lymphoma in a Fibrous Meningioma
470	Sarah Martin, Hasan Khalidi, Eyas Hattab
179	Light Microscopic and Ultrastructural Findings in a Case of Intraventricular
	Schwannoma
100	Jason Wells, Omar Zalatimo, Cathy Housman, Mark lantosca, Charles Specht  Loss of Expression of Dendritic Cell-Specific Transcript (DC-SCRIPT) in Brain Metastatic
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181	Low Grade Malignant Peripheral Nerve Sheath Tumor in Cervical Spine with Mature
	Skeletal Muscle Differentiation Ming Zhang Michael Wegyer, Jogyir Khurana Ahir Mukhariaa
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	Neuroepithelial Tumors  Knarik Arkun and Christine Fuller
105	Neurocytoma and Intraventricular Haemorrhage
185	Francesca Brett
186	Neuroendocrine Carcinoma of the Pineal Parenchyma: The First Reported Case
100	Qinwen Mao, Kyung-Hwa Lee, James Chandler, Eileen Bigio
187	Paraganglioma Arising in a Mature Teratoma of the Ovary
107	Leili Mirsadraei, Yuki Takasumi, Nora Ostrzega, Tracie Pham, Jian Yu Rao, William Yong
188	Pediatric Malignant Mixed Glial / Primitive Neuroectodermal Tumor. The Importance of
100	Molecular Analysis
	Christine Fuller, Kimberly Stogner-Underwood, Asadullah Khan, R. Graham, Knarik Arkun
189	Perineural Spread of Squamous Cell Carcinoma to the Brainstem: A Complex Clinical
	Presentation and Delayed Diagnosis
	Sarah Martin, Aaron Kamer, Todd Vogel, Troy Payner, Jose Bonnin
190	Plasmablastic Lymphoma with MYC Rearrangement Involving the CNS: A Case Report
	Adriana Doldan, Carlos Acevedo, Xianyuan Song, Peter Shen
191	Primary Histiocytic Sarcoma of the Brain: A Case Report and Review of the Literature
	Tammy Tyree, Peter Nakaji, Stephen Coons
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	Regional Children's Hospital
	Andrea Wiens and Eyas Hattab
193	Strong Desmin Expression in a Congenital Desmoplastic Infantlie Astrocytoma
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	Sarah Alghamdi, Amilcar Castellano-Sanchez, Carole Brathwaite, Taiyo Shimizu, Ziad Khatib, Sanjiv
	Bhatia
194	Tumoral Bing-Neel Syndrome Presenting as a Cerebellar Mass
	Jingxin Qiu, Robert Fenstermaker, Andrew Fabiano

# American Association of Neuropathologists

Endowed Lectureships Meritorious Awards Presidential Symposium

### The Parisi Lecture

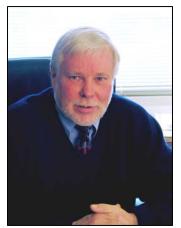
The *Parisi Lecture* was established with a generous endowment from Teva Pharmaceuticals in 2007. Teva Neuroscience, a subsidiary of Teva Pharmaceuticals, is devoted to the study and development of products and services that address the health management needs of people in the field of neurology. One of the focal points of their efforts is multiple sclerosis.

The lecture was named the Parisi Lectureship in honor of one of the American Association of Neuropathologists' exceptional members, Dr. Joseph E. Parisi. He has published seminal neuropathological studies on a wide range of diseases affecting the nervous system, with particular focus on neurodegenerative diseases and multiple sclerosis. He has held virtually every office of the Society, including President, and has served on several AANP committees. In 2006, his dedication and generosity were recognized with the Award for Meritorious Contributions to Neuropathology. He is considered by many the heart and soul of the association and a man worth emulating.

We are pleased to have Bruce D. Trapp, PhD join our list of distinguished speakers.

2008	Claudia	The Spectrum of CNS Inflammatory
	Lucchinetti	Demyelinating Diseases: From Pathology
		to Pathogenesis
2009	Hans Lassmann	Inflammation Induced Mitochondrial
		Injury: A Major Mechanism of
		Neurodegeneration
2010	Joseph Dalmau	Autoimmune Synaptic Encephalitis
2011	Steven S. Scherer	Molecular Pathologies at the Nodes of
		Ranvier
2012	Bruce D. Trapp	Neuronal Damage in Multiple Sclerosis

### 2012 PARISI LECTURE Neuronal Damage in Multiple Sclerosis Bruce D. Trapp, PhD



**Dr. Bruce D. Trapp** is Chairman of the Department of Neurosciences at the Lerner Research Institute, Cleveland Clinic and Professor of Neurosciences at Case Western Reserve University.

Dr. Trapp received his Ph.D. from Loyola University Stritch School of Medicine in Chicago, IL. He received postdoctoral training at the National Institutes of Health (NIH), Bethesda, MD and then was appointed Assistant and subsequently Associate Professor of Neurology at the Johns Hopkins University School of Medicine in Baltimore. He joined the Cleveland Clinic as Chairman of the Department of Neurosciences in 1994.

He is the recipient of the Jordi Folch-Pi Award from the American Society of Neurochemistry, The Weil Award from the American Association of

Neuropathologists, the Harry Weaver Neuroscience Scholar Award from the National Multiple Sclerosis Society (NMSS), the Jacob Javits Award in Neuroscience from the National Institute of Neurological Disorders and Stroke, the John Dystel Prize for MS Research from the American Academy of Neurology and the National Multiple Sclerosis Society, the Stephen C. Reingold award from the NMSS, the Scientific Achievement Award in Basic Science and the Award for Excellence in Science from the Cleveland Clinic and Dr Trapp is a Fellow of the AAAS.

Dr. Trapp's research investigates the cause of neurological disability in multiple sclerosis patients, cellular mechanism of brain repair in neurodegenerative diseases, and the molecular biology of myelination in the

central and peripheral nervous systems. He is internationally known for his work on mechanisms of neurodegeneration and repair in multiple sclerosis and has published over 200 peer-reviewed articles and over 35 book chapters

### **Abstract**

Multiple Sclerosis (MS), an inflammatory-mediated demyelinating disease of the central nervous system (CNS), affects more than 2.5 million people worldwide. Although traditionally considered a white matter disease, grey matter demyelination and associated neuronal pathology play significant roles in the pathogenesis of permanent neurological disability in MS patients and may cause the cognitive dysfunction found in over 50% of MS patients. Brain imaging studies have correlated white matter lesion load and brain atrophy with cognitive dysfunction in MS patients. A subgroup of cognitively impaired MS patients, however, have low white matter lesion load and minimal physical disability, raising the possibility that hippocampal demyelination may cause memory dysfunction in MS patients. Recent imaging studies have correlated altered hippocampal magnetic resonance imaging (MRI) measures and increased hippocampal atrophy with memory dysfunction in MS patients. Demyelination has been detected in 53% to 79% of postmortem MS hippocampi. This presentation will summarize cellular and molecular changes in demyelinated hippocampi from postmortem MS brains. Compared to hippocampi from control brains or myelinated hippocampi from MS brains, demyelinated hippocampi had minimal neuronal loss but significant decreases in synaptic density. Hippocampal demyelination negatively impacts the expression of neuronal molecules involved in axonal transport, synaptic integrity, glutamate homeostasis, synaptic plasticity and memory/learning. The molecular changes observed in demyelinated hippocampi were not detected in demyelinated MS motor cortex or in hippocampi from Alzheimer's disease (AD) brains. These data support the concept that myelin is essential for normal hippocampal function and that the neuronal genes regulated by myelination reflect the specialized functions of different neuronal subpopulations.

- Explain the role of grey matter demyelination in the pathogenesis of MS.
- Describe how myelination helps regulate neuronal gene expression and synaptic connectivity.
- Review the pathogenesis of cognitive dysfunction in individuals with MS.

### The DeArmond Lecture

he DeArmond lecture was established in recognition of Stephen J. DeArmond's excellent leadership and organization of the scientific program for the 2006 International Congress of Neuropathology. This successful meeting garnered significant support intended for the future advancement of the mission of the American Association of Neuropathologists. To continue these intended goals and recognize Dr. DeArmond's contributions, the American Association of Neuropathologists has honored him by establishing the *DeArmond Lecture*. Dr. DeArmond is a leading authority on prion disease, where his work has been fundamental in demonstrating mechanisms of transmission and routes to therapeutics. The DeArmond Lecture focuses on honoring those making major advances in the field of neurodegeneration and aging with a particular emphasis on translating these findings to patient care.

We are pleased to have Krystof Bankiewicz, MD, PhD join our list of distinguished speakers.

2008	Virginia M. –Y. Lee	TDP-43, A New Class of Proteinopathies in Neurodegenerative
		Diseases
2009	Rudy Tanzi	Decoding Alzheimer's Disease Gene by Gene
2010	Todd Golde	Alzheimer's Disease: Models and Therapeutics
2011	Beverley L. Davidson	Emerging Therapies for Neurogenetic Diseases
2012	Krystof Bankiewicz	New Therapies for Parkinson Disease

### 2012 DEARMOND LECTURE Novel MRI-based Platform for Efficient Gene Delivery to the Brain Krystof Bankiewicz, MD, PhD



**Dr. Krystof Bankiewicz** holds position of the Kinetics Foundation Endowed Chair in Translational Research, Professor in Residence in Neurosurgery and Neurology at University of California San Francisco. Dr. Bankiewicz is also Vice Chair for Research in the Department of Neurosurgery and Director of Translational NeuroTherapy Center at UCSF. Dr. Bankiewicz is an inventor of numerous patents, and has published more than 160 peer-reviewed research articles. Dr. Bankiewicz has considerable experience in supervising multi-investigator translational programs. He is a Principal Investigator on several multi-center, multi-investigator grants. He has supervised a total of 30 post-doctoral fellows, and manages a core research group of 20 individuals including technicians, post-doctoral fellows, and a senior scientists.

Dr. Bankiewicz received his MD degree from Jagiellonian University in Crakow and his PhD and DSc degrees from the Institute of Neurology and Psychiatry in Warsaw, Poland. After his residency and an appointment as Assistant Professor with the Post-graduate Medical Center in Warsaw, he received a Fogarty Fellowship and became a Visiting Fellow and then Visiting Associate Scientist with the Surgical Neurology Branch of the NINDS at the NIH in Bethesda, Maryland. There, he became Chief of the Central Nervous System (CNS) Implantation Unit in 1991. Shortly afterward, he came to California to serve successively as Chief of Preclinical Studies with the Somatix Therapy Corporation in Alameda, the Director of the Division of CNS Implantation and Regeneration with The Parkinson's Institute in Sunnyvale, and from 1994-1998 a Visiting Scientist with the Laboratory for Functional Imaging of the Lawrence Berkeley National Laboratory. From 1997-2001 he returned to the NIH as Acting Chief of the Molecular Therapeutics Section of NINDS. Since 1998 he has been a Professor in Residence of Neurosurgery and Neurology, Principal Investigator with the Movement Disorders Research Program and the Brain Tumor Research Center at the University of California San Francisco.

Throughout his career, Dr. Bankiewicz has maintained a strong focus on the development of translational approaches to gene and cell replacement therapies, and has displayed a remarkable ability to synthesize several individual technologies into powerful new approaches to the treatment of such serious diseases as brain cancer and neurodegenerative disorders of the brain, including Parkinson's, Huntington's, Alzheimer's diseases and pediatric neurotransmitter deficiency and lysosomal storage disorders. Dr. Bankiewicz was instrumental at every

stage of the Phase-1 clinical trials for AAV2-hAADC and AAV2-hGDNF gene therapy, resolving technical and scientific issues with respect to filing an IND applications with the FDA, and also in recruiting a clinical team to undertake the clinical trials. He continues to champion the development of novel therapeutic strategies to treat these important diseases. The recently established Translational NeuroTherapy Center at UCSF, of which he is the first Director, is a logical extension of Dr. Bankiewicz's efforts to close the gap between bench and bed side by engaging academia, industry, NIH and non-for profit organizations in a joint effort in the clinical development of novel therapeutics for brain disorders.

### Abstract:

Gene transfer technology can correct genetic mutations in the brain. Neuro gene delivery via direct intrapranchymal injections of adeno-associated viral (AAV) vectors is a locally administered treatment that requires accurate delivery to maximize safety and efficacy. The large volume and convoluted architecture of the human brain is a considerable barrier to translating small animal findings into efficacious clinical procedures. Too little target coverage and the treatment risks being ineffective. Conversely, excessive distribution or off-target gene delivery increases the possibility for unexpected adverse effects. Optimal viral vector delivery into the brain is challenging and brain distribution of viral vectors is uncertain. To address this issue we developed viral vector delivery system that permits direct MRI monitoring of vector distribution within the brain in real-time. This significant advance allows for the first time to adjust parameters of vector infusion while delivering gene therapy, giving surgeon full control over gene transfer technology.

To allow for precise intracerebral delivery of biologics for therapy of neurological disease we also developed a skull-mounted aiming device (SmartFrame) and integrated software platform (ClearPoint) for interventional MRI guided placement of deep brain stimulators. In anticipation of upcoming gene therapy clinical trials in brain disorders we adapted this device for real-time convection enhanced delivery (RCD) of therapeutics via a custom designed infusion cannula. Based on real-time MRI data, this system allows selection of brain targets, provides instructions for cannula insertion along a chosen trajectory, and permits visual monitoring of infusions.

Subsequent to our discovery that AAV2 vectors undergo anterograde transport along thalamocortical projections resulting in transduction of cortical neurons, we analyzed properties of several AAV serotypes and evaluated their potential for correcting genetic deficit in the brain via axonal transport. Combination of RCD and axonal transport may allow for predictable gene transfer over large cortical and sub-cortical regions of a human brain.

Our advanced gene delivery system is currently tested for delivery of therapeutic genes in Parkinson's (PD), Huntington's (HD) and Nieman-Pick, AADC deficiency in children and brain tumors. Data will be provided to demonstrate promises and challenges in successful clinical translation of gene transfer technology for CNS disorders.

- Describe technological innovations that permit monitoring of drug delivery to the brain in real time
- Describe axonal transport pathways of viral vectors that will facilitate gene therapy of brain diseases

### The Saul R. Korey Lectureship-a Brief History

he *Korey Lectureship* was established by Dr. Robert D. Terry in honor of Dr. Saul R. Korey, the founder and first Chair of the Department of Neurology at Albert Einstein College of Medicine. Dr. Korey's vision of an interdisciplinary approach to the study of neurological diseases by basic and clinical scientists has inspired generations of colleagues and trainees. Dr. Terry, a close collaborator and colleague of Dr. Korey, was the first recipient of the prestigious *Potamkin Prize for Pick's and Alzheimer's Disease* in 1988, in recognition of his seminal observations of the pathological changes in Alzheimer disease. Dr. Terry generously contributed a portion of the prize funds to endow the *Korey Lectureship*, to be administered by the American Association of Neuropathologists, with the lecturer to be chosen annually by the president.

Dr. Terry has summarized the qualities of the Korey lecturer as someone who has "... been an active member of the Association...a working MD or MD/PhD neuropathologist...responsible for diagnostic work as well as teaching and research. The lecture should be aimed at the members of the Association, and the lecturer might well serve as a role model for younger members."

We are pleased to have Michael Norenberg, MD, join our list of distinguished speakers.

<u>Year</u> 1989	Lecturer Nicholas K. Gonatas	Title MG-60, a Novel Sialoglycoprotein of Medial Cisternae of the Neuronal Golgi Apparatus: Implications and Applications	<u>Year</u> 1998 1999	Lecturer Sandra H. Bigner William F. Hickey	Title Molecular Genetics of Medulloblastoma Key Participants in the Initiation of Inflammation in the Central Nervous System
1990	Henry M. Wisniewski	Amyloidosis in Alzheimer's Disease and the Spongiform Encephalopathies	2000	Mary E. Case	Neuropathology and Forensic Pathology: A Natural Synergism
1991	Robert D. Terry	Alzheimer's Disease as Seen by a Lucky Morphologist	2001	Paul H. Kleihues James E.	Molecular Biology of Brain Tumors Astrocytes, Intermediate
1992	Henry deF Webster	Formation and Regeneration of Myelin	2002	Goldman	Filaments, Cellular Stress and Neuropathology
1993	Kunihiko Suzuki	Molecular Genetics of Tay-Sachs and Related Disorders: The Legacy of	2003	Samuel K. Ludwin	Pathology and Pathogenesis in Multiple Sclerosis
1004	<b>N</b> 7 <b>T</b>	Saul Korey	2004	James M.	The Road Not Taken
1994	No Lecture	XIIth International Congress (Toronto)	2005	Powers Bernardino	Deciphering Hereditary
1995	Blas Frangione	Amyloid Genes and Chaperones in Alzheimer Disease	2003	Ghetti	Presenile Dementias: Neuropathology at the Crossroads of
1996	Floyd Gilles	The 3R's of Neuro- oncology – Recording,	2006	Donna M.	Neuropsychiatry and Molecular Genetics Molecular Mechanisms of
1997	Donald L. Price	Reliability and Reporting The Role of Neuropathologists in the Analyses of Models of Neurodegenerative Disease	2000	Ferriero	Hypoxic-Ischemic Injury in the Developing Nervous System

<u>Year</u>	<u>Lecturer</u>	<u>Title</u>			
2007	Dennis W.	Neuropathology and	Year	<u>Lecturer</u>	<u>Title</u>
	Dickson	Genetics of Parkinsonism	2010	Peter C.	A Long-Term Perspective
2008	David N.	Brain Tumor		Burger	on Pediatric CNS Tumors
	Louis	Classification: Little Steps	2011	Hans H.	Protein Aggregate
		and Big Jumps		Goebel	Myopathies
2009	Stephen J.	Mechanisms of	2012	Michael	Astrocyte Pathobiology
	DeArmond	Neurodegeneration in		Norenberg	
		Prion Disease			

### 2012 SAUL R. KOREY LECTURE Why the Brain Fails when the Astrocyte Ails Michael D. Norenberg, MD

Orgininating from the Neuronal Plasma Membrane



Michael D. Norenberg obtained his undergraduate degree from Trinity College and his medical degree from the University of Rochester. He served in the military as a general medical officer (US Air Force) before completing his Anatomic Pathology training and Neuropathology Fellowship (under Lowell W. Lapham) at the University of Rochester. He then joined the medical faculty at the University of Colorado and subsequently took an academic position at the University of Miami where he currently serves as Director of Neuropathology. He holds appointments in the Departments of Pathology and Biochemistry and Molecular Biology. Throughout his academic career Dr. Norenberg has been associated with the Veterans Administration where he has carried out his research activities. His research work has focused on the normal function of astrocytes and their role in CNS disorders, particularly in hepatic encephalopathy. He has also investigated factors involved in the pathogenesis of central pontine myelinolysis, and in mechanisms involved in the brain edema/astrocyte swelling following neurotrauma. In 2010 Dr. Norenberg received the William S. Middleton Award from the

Department of Veterans Affairs, the highest honor presented by the VA Research and Development Office in recognition of outstanding scientific contribution and achievement in the areas of biomedical and bio-behavioral research related to the health care of veterans. In 2012 he was inducted into the prestigious Association of American Physicians.

### Abstract

Astrocytes are the most common cell constituents in the CNS, making up approximately 50% of the volume of human brain. They traditionally have been known as supportive cells that respond to destructive injuries (reactive gliosis) and form most of the primary neoplasms of the CNS. Among their well accepted functions include the regulation of extracellular levels of K<sup>+</sup>, pH, glutamate, water transport, involvement in the glutamate-glutamine cycle, maintenance of the blood-brain barrier, regulation of cerebral blood flow, production of growth/trophic factors, energy metabolism, and antioxidant properties, among others. As astrocytes are electrically "silent" or "inactive", the notion that they may play a key role in neurotransmission had never been seriously considered. It was not until the 1990's when investigators began to explore the possibility that astrocytes could indeed be activated and could communicate with neurons through elevations in intracellular calcium and the subsequent release of gliotransmitters, leading to the concept of the tripartite synapse. These new findings have resulted in a major paradigm shift whereby astrocytes now are believed to play crucial roles in the modulation of neurotransmission and synaptic plasticity. In parallel with these developments, a view has evolved that in many neurological conditions astrocytes are injured during the very early phase of the disease process, and that failure

of astrocytes to carry out their critical functions results detrimental effects on other neural cells. In other words, rather than simply "reacting" to neuronal or oligodendroglial injury, it is very likely that "ailing" astrocytes resulting from the initial insult, cause or contribute to the disease process itself. A major focus of this presentation will be to review the role of astrocytes in the pathogenesis of major neurological disorders.

- Review newer aspects of structure and functions of astrocytes
- Discuss the various means by which astrocytes may contribute to CNS dysfunction
- Explain the research that supports that astroglial dysfunction is at the core of many/most neurological disorders

### Matthew T. Moore Distinguished Lecture

### **Pathogenesis of ALS**

Robert H. Brown, Jr., MD University of Massachusetts, Worcester, MA

[Materials to be distributed at the Meeting]

### **Awards for Meritorious Contributions to Neuropathology**

he *Award for Meritorious Contributions to Neuropathology* recognizes a member who has made significant contributions to the advancement of knowledge in neuropathology and provided service to the American Association of Neuropathologists. Each recipient of the award is nominated by the president, in conjunction with the nominating committee and with the approval of the executive council.

The qualities of outstanding scientific achievement and service are embodied in this year's recipients, Drs. William W. Schlaepfer and Leroy R. Sharer. They join the rich roster of distinguished former award recipients.

Year	Recipient		
1959	Armando Ferraro	Year	Recipient
	Arthur Weil	1995	Amico Bignami
1960	Joseph H. Globus		Asao Hirano
	George B. Hassin	1996	Pasquale A. Cancilla
1968	Abner Wolf		Franz Seitelberger
	Paul I. Yakovlev	1997	Henryk M. Wisniewski
	Harry M. Zimmerman	1998	Richard L. Davis
1970	Webb E. Haymaker		Wolfgang Zeman
1971	James W. Kernohan	1999	Lucy B. Rorke
1972	George A. Jervis	2000	William R. Markesbery
1979	Raymond D. Adams	2001	John J. Kepes
	David Cowen		Henry de Forest Webster
	Matthew T. Moore	2002	Dikran S. Horoupian
1981	Richard Lindenberg		Fusahiro Ikuta
1983	Orville T. Bailey		Kurt A. Jellinger
1984	Margaret Murray	2003	Bernardino F. Ghetti
1985	Kenneth M. Earle	2004	Michael N. Hart
	Nathan Malamud	2005	E. Tessa Hedley-Whyte
	Leon Roizin		Suzanne S. Mirra
1986	Martin G. Netsky	2006	Joseph E. Parisi
1987	No Award Presented		Jeannette J. Townsend
1988	Edward P. Richardson, Jr.	2007	James M. Powers
	F. Stephen Vogel		Cedric S. Raine
1989	Lucien J. Rubinstein	2008	Kinuko Suzuki
	Robert D. Terry		Margaret G. Norman
1991	Lysia K. S. Forno	2009	Peter C. Burger
1992	John Moossy		Pierluigi Gambetti
	Gabriele M. ZuRhein		Nicholas K. Gonatas
1993	Peter W. Lampert	2010	Stephen J. DeArmond
	Elias E. Manuelidis		Samuel K. Ludwin
1994	Murray B. Bornstein	2011	William W. Schlaepfer
	Samuel P. Hicks		Leroy R. Sharer
	Lowell W. Lapham	2012	

### Awards for Meritorious Contributions to Neuropathology

## 2011 AWARD RECIPIENTS Bernd W. Scheithauer, MD and Donald L. Price, MD



Bernd Walter Scheithauer was one of those robust positive professional and personal constants in the field of neuropathology for several decades during which he made meritorious contributions to the field. He was a member of our Association for over 34 years and was also a senior and dedicated charter member of the Association's Diagnostic Slide Session. He was known personally by numerous members of the Association and, by the vast majority of pathologists, neurosurgeons, and neurologists for his significant and diverse contributions to surgical neuropathology. However, his impact goes far beyond the incredibly impressive number and extensive diversity of his published work, including approximately 700 peer-reviewed publications and nearly 90 book chapters and books over a 35 year career. Dr.

Scheithauer was a spirited icon of surgical neuropathology as an international luminary whose passionate affection of neuropathology was a legendary inspiration to both colleagues and trainees. For Dr. Scheithauer, there was never a conflict between work and pleasure time, because he derived so much pleasure from his work.

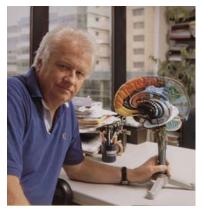
Dr. Scheithauer graduated from Humboldt State University in Arcata, CA in 1969. He then headed south to Loma Linda University School of Medicine where his prodigious talent for pathology was already becoming evident. Following graduation, he headed north in 1974 to Stanford University for the next five years where he completed for his post-graduate studies in Anatomic Pathology, Neuropathology, and Surgical Pathology at a time when the Pathology Department was experiencing an exciting and golden era. Drs. Richard Kempson and Ronald Dorfman were setting new standards for the practice of surgical pathology and Lucien Rubinstein was energetically applying his superb talents to teaching innovative diagnostic neuropathology with a combination of superbly performed histochemistry in the British tradition and pioneering state-of-the-art immunohistochemistry. It was during this time that GFAP immunohistochemistry was first used for systematically analyzing surgical pathology specimens. The breadth of the Rubinstein training program also included hallmark in vitro studies of brain stromal-glial tumor interactions using long-term primary organotypic cultures. energetically embraced these intellectual riches of the neuropathology training program with the distinguished visiting neuropathologists, numerous talented trainees, and the incredible array of the consultation cases that were the bases of subsequent hallmark clinicopathologic correlations. Even during his training, he published studies which advanced our understanding of subependymomas, the complex cerebral medulloepitheliomas, and meningeal mesenchymal chondrosarcomas. It was during these formative years that he developed his keen attention to both histopathologic and relevant clinical details, while embracing and respecting how basic research, enhanced current methods and the application of new techniques could enhance his studies.

Dr. Scheithauer joined the faculty in the Department of Laboratory Medicine and Pathology at the Mayo Clinic and College of Medicine as an instructor in 1979. His entire professional career ensued at the Mayo Clinic and College of Medicine where he held the positions of Section Head of Surgical Pathology and Professor with Master's Faculty Privileges in Neuroscience. He described both new clinicopathologic entities and refined criteria for established tumor types, ranging glioneuronal neoplasms, unusual gliomas, subependymal giant cell tumors, and solitary fibrous tumors and mesenchymal tumors arising within neuraxis to a variety of tumors arising from cells of the peripheral nerve sheath. He was a principal investigator on 1 and a co-investigator on 10 extramurally funded research grants. In this perspective, Dr. Scheithauer took the traditions of Lucien Rubinstein and advanced the field of modern surgical neuropathology both in the breadth of study and the application of novel biomarkers in collaboration with numerous neuroscientists. He effectively bridged diagnostic surgical pathology and neuropathology and, in this way, created an important legacy for future neuropathology trainees.

The field of pituitary pathology was remarkably enhanced by Dr. Scheithauer. He made hallmark contributions with his strong and long-standing collaborators and friends, Drs. Kalman Kovacs, Eva Horvath and Ricardo Lloyd. His work led to the better characterization of pituitary adenomas by meticulous morphological, immunohistochemical and ultrastructural studies of these tumors with correlation to the clinical behavior and biological features. These included pituitary adenomas producing GH, PRL and glycoprotein hormones, silent subtype 3 adenomas, Crooke's cell adenoma, Spindle-cell oncocytoma of the adenohypophysis, and pituitary blastomas. He contributed to the fundamental understanding of the pathogenesis and biological behavior of several pituitary tumors, and proposed biomarkers that may serve as prognostic factors of biologic behavior. Finally, he was one of the initial proponents of a new classification of pituitary tumors based on the morphofunctional features of these tumors.

Dr. Scheithauer truly enjoyed teaching neuropathology to fellows and visiting scholars and used his extensive case files to produce exciting projects for them. He mentored over 100 visiting clinical scholars and 6 fellows over a period of 19 years. Through his dedication to teaching, he has provided the future of neuropathology with talented trainees, the majority of whom are now highly productive members of our Association. In addition, Dr. Scheithauer was well-known to practicing surgical pathologists and neuropathologists as an articulate and informative speaker. He delivered over 90 invited lectures, including named lectureships, gave over 70 presentations at international meetings, and over 50 presentations and slide conferences at national meetings. He was especially proud of his award of the Lucien J. Rubinstein Visiting Lectureship in Neuropathology at the University of Virginia. In addition to a meritorious record of publications, Dr. Scheithauer contributed to our field by his review board membership and ad hoc reviewing activities for 17 journals, including our Association journal. In addition to our Association, he was an active member of over ten professional societies, including the Arthur Purdy Stout Society of Surgical Pathologists, the Canadian Association of Neuropathologists, the United States and Canadian Academy of Pathology (and the Paleopathology Club), the Pituitary Pathology Group, the Schwann Society, and the Society for Ultrastructural Pathology.

The world community of neuropathology lost an extraordinary contributor, energetic personality, and devoted friend with the death of Bernd Walter Scheithauer. His meritorious contributions live on to enrich the field of neuropathology and our Association.



**Donald Lowell Price** was born in Stamford, Connecticut. He began his academic career at Wesleyan University in Connecticut where he studied English literature and received a Bachelor of Arts. Then, he entered the Albany Medical School from where he graduated in 1961. After two years of internal medicine training at the New England Medical Center, he became a neurology resident at the Massachusetts General Hospital. Following two years as Staff Neurologist at the National Naval Medical Center in Bethesda, Dr. Price returned to Boston as a Fellow in Neuropathology under the mentorship of Dr. E. P. Richardson at the Massachusetts General Hospital. Subsequently, he worked in cell and molecular biology in the laboratory of Dr. Keith Porter at Harvard. In 1970, Dr. Price became an Assistant Professor of Neuropathology at the Boston

City Hospital. The next year, Dr. Price moved to Baltimore to join the faculty of the Johns Hopkins University School of Medicine as Associate Professor and become the Founding Director of the Neuropathology Laboratory, where he would continue his work until retiring in 2010. After retiring, Dr. Price became Emeritus Professor of Pathology. His successful career at Hopkins was extremely productive, both in research, as well as in teaching and mentoring of several generations of neuropathologists and neurobiologists. He made major contributions to the understanding of a variety of human diseases, including peripheral neuropathies and several neurodegenerative diseases including Alzheimer's disease, Parkinson's disease, and amyotrophic lateral sclerosis. At Hopkins, Dr. Price's talent, energy, and scientific contributions were recognized with a Professorship in 1978. His achievements were also internationally recognized by the scientific community and received multiple honors. He became President of the American Association of Neuropathologists in 1989 and of the Society for Neuroscience in 2000. In 1998, he was elected to the Institute of Medicine. The author of more than 500 research publications, during the "Decade of the Brain" (1990-2000), he was ranked among the top ten neuroscientists as authors of high-impact papers by Science Watch.

Dr. Price's initial research work focused on the biology and pathobiology of motor neurons, in particular the mechanisms of degeneration and regeneration following axotomy. Subsequently, his research interest expanded to include age-associated neurodegenerative diseases, especially Alzheimer's disease. This endeavor required not only observations of human brains, but also the study of animal models that would allow a more direct insight into pathogenesis. In the early 80s, the Price laboratory made seminal contributions to the understanding of Alzheimer's, including the identification of lesions of cholinergic neurons in the basal forebrain. In 1984, Johns Hopkins was awarded one of the original NIH-funded Alzheimer's Disease Research Centers under the direction of Dr. Price. His research of naturally occurring animal diseases, in parallel to human neuropathologic studies, yielded important information on motor neuron diseases. Equally important were the studies of aged Rhesus monkeys with memory deficits and AD-like neuropathology which paved the way for the approaches to be used later on the analyses of transgenic models. In the 1990s, the Price laboratory embraced the development of genetically-engineered mouse models of disease. These animal models allowed Hopkins' researchers to make seminal observations on the cell and molecular biology of APP and amyloidogenesis, SOD mutations and motor neuron diseases, as well as the mechanism of trinucleotide expanded repeats in Huntington's disease. These animal models, which were made available to the research community and became the standard models in many laboratories nationwide and abroad, are now being used in experimental therapeutic studies. Over the past several decades, the work of Dr. Price has been well recognized and supported by program projects, R01s (LEAD award) and training grants from the NIH, as well as grants from the Adler Foundation, Metropolitan Life Foundation, BMS Foundation and other private sources

The combined backgrounds in medicine, neurology, neuropathology, and cell biology gave Dr. Price a fairly unique perspective on research that, in addition to propelling him into an outstanding scientific career, made him a most sought after advisor to scientific societies, research foundations, the National Institutes of Health, U.S. Congress panels, and the scientific boards of pharmaceutical and biotechnology companies.

As impressive as his research work is, a major legacy of Dr. Price is his contribution to training and mentoring innumerable neuropathologists, neurologists, and basic neuroscientists who have become leaders at academic institutions throughout the country. After retirement, Dr. Price has continued mentoring and guiding graduate

students and neuropathology trainees with great devotion, and keeps coming to the lab regularly to stay abreast of current studies and to stimulate us all into new research endeavors.

It is impossible to talk with Dr. Price for more than few minutes and not notice his deep interest in and love of classic literature and music. Many scientific conversations with him easily stray into Shakespeare or Dante; paper and grant reviews have often in the background the music of Wagner or Puccini. But there is more to Dr. Price than science, writing, and music. He is an avid swimmer and his competitive spirit also took him into running and triathlons.

These remarks would be incomplete without mentioning the immense devotion of Dr. Price for his family, including Helen his wife, their three children, all of them in medical practice including one neuropathologist, and seven grandchildren.

Donald L. Price, M.D., has been a leader and major force in contemporary neuropathology and a tireless advocate of amalgamating basic neurobiology with pathology and neurology. He is a most worthy recipient of the Award for Meritorious Contributions to Neuropathology of the American Association of Neuropathologists 2012.

### AANP PRESIDENTIAL SYMPOSIUM Sunday, 24 June 2011

### **Current Topics in Multiple Sclerosis**

8:00 am – 8:05 am	Introduction
	Raymond A. Sobel, MD
	Stanford University School of Medicine, Stanford, CA
8:05 am - 9:00 am	Parisi Lecture
	Neuronal Damage in Multiple Sclerosis
	Bruce D. Trapp, PhD
	The Cleveland Clinic Foundation, Cleveland, OH
9:00 – 9:45 am	Sphingosine-1-phosphate signaling in MS "Oriental Medicine to Immune
	Modulation"
	May Htwe Han, MD, PhD
	Stanford University School of Medicine, Stanford, CA
9:45 am – 10:30 am	AANP Award Presentations and Refreshment Break
10:30 am – 11:15 am	The Changing Epidemiology of MS
	A. Dessa Sadovnick, PhD
	VCHA-UBC Hospital, Vancouver, BC
11:15 am – 12:00 pm	How MS could be an Acquired Disease
	Raymond A. Sobel, MD
	Stanford University School of Medicine, Stanford, CA
12:00 pm	INSTALLATION OF NEW OFFICERS AND ADJOURNMENT

### 2012 PRESIDENTIAL SYMPOSIUM

Sphingosine-1-phosphate Signaling in MS "Oriental Medicine to Immune Modulation" May Htwe Han, MD, PhD Stanford University School of Medicine, Stanford, CA



May H. Han is an Assistant Professor and a clinician-Scientist in the Department of Neurology and Neurological Sciences at Stanford University School of Medicine. She received her medical degree in Myanmar and completed her Neurology residency at the University of Washington in Seattle. She did a translational Fellowship in Neuroimmunology at Stanford University with Dr. Lawrence Steinman. In 2009, she joined the Stanford University School of Medicine's Neurology and Neuroligical Sciences Department and the Stanford Hospital and Clinic's Multiple Sclerosis Center.

Dr. Han's research focuses on identification of biomarkers and therapeutic targets in multiple sclerosis and other demyelinating diseases of the central nervous system. She utilizes a systems biology approach, using proteomics and transcriptomics, with the goal of identifying biomarkers to monitor disease activity and to understand protective molecules that are present during neuroinflammation. Her lab studies patient samples and tests their hypotheses in animal models and cellular and biochemical assays, with the ultimate goal of applying the knowledge directly to patient care.

### **Abstract**

Multiple sclerosis (MS) is an autoimmune de-myelinating disease that damages the central nervous system, affecting over one million young adults worldwide<sup>1</sup>. FTY-720 (Fingolimod) was recently approved by the FDA as the first orally bio-available, first-line therapy for treatment of relapsing remitting MS<sup>2</sup>. FTY-720 is a biomimetic of the endogenous, bioactive signaling lipid sphingosine-1-phosphate (S1P). S1P signals extracellularly through five G-protein coupled receptors (S1P<sub>1-5</sub>), and FTY-720 treatment modifies this pathway. FTY-720's mechanism of action currently centers around the concept of "functional antagonism" where agonist-like binding of drug to receptor leads to internalization and degradation of the signaling complex. This internalization event is viewed as critical in the "functional antagonism" paradigm, and previous studies have shown that C-terminal mutation/deletion of S1Pr<sub>1</sub> inhibits receptor internalization<sup>3</sup>. Proteomic analysis of active MS brain lesions identified C-terminal peptides of S1Pr<sub>1</sub> found to be phosphorylated on serine 351. To understand the function of S1Pr<sub>1</sub> signaling by means of post-translational modifications, we utilized the S1Pr<sub>1</sub> S5A/S5A mouse, which is phosphorylation defective on its C-terminal tail and has altered S1Pr<sub>1</sub> signaling kinetics as a consequence<sup>4</sup>. We induced experimental autoimmune encephalomyelitis (EAE) in these mice and found that S1Pr<sub>1</sub><sup>S5A/S5A</sup> mice experienced more severe disease, higher titers of inflammatory cytokines, and greater numbers of CNS infiltrating immune cells compared to wild type. Administration of FTY-720 to S1Pr<sub>1</sub>SSA/SSA mice induced within presymptomatic EAE demonstrated that S1Pr<sub>1</sub>S5A/S5A were refractory to treatment as evidenced by inflammatory cytokine production. Our studies demonstrate the importance of C-terminal S1Pr<sub>1</sub> phosphorylation in the context of neuroinflammation and FTY-720 treatment. These investigations hold the potential to elucidate the mechanism of action of FTY-720 and stratification for potential side effects resulting from therapy.

- Describe how systems biology is elucidating the pathology and pathogenesis of MS
- Explain the bases for new therapies in MS
- Explain the role of lipid second messenger signaling in MS

### 2012 PRESIDENTIAL SYMPOSIUM

### The Changing Epidemiology of MS

A. Dessa Sadovnick, PhD VCHA-UBC Hospital, Vancouver, BC



**Dr. Sadovnick** was born in Montreal and obtained degrees from McGill University (B.Sc., Honors Genetics; M.Sc., Human Genetics) and the University of British (Ph.D., Genetics).

Dr. Sadovnick is a Professor in the Department of Medical Genetics and the Faculty of Medicine, Division of Neurology, UBC. She is the Director of the Western Pacific Regional Research and Training Center for Multiple Sclerosis, established by the MS Society of Canada.

Dr. Sadovnick is the Principal Investigator of several multicenter Canadian and International collaborative research projects on Multiple Sclerosis, pediatric Multiple Sclerosis and Dementia.

Dr. Sadovnick was one of the developers of the M.Sc.Genetic Counseling Training Program at UBC, has served as co-director and is now on the advisory board. She has published extensively (over 280 articles in peer-review journals) and serves as a reviewer for a wide variety of medical journals and grant review panels. She is often an invited speaker at Canadian, American and

International (Europe, Asia, South America, Russia, Asia) scientific meetings. Dr. Sadovnick is a member of several National and International advisory groups and task forces for both MS and dementia. She is currently involved in Canadian-China initiatives in both MS and AD, the Canada-wide monitoring system for MS and the BC outcomes registry for MS therapeutics outcomes.

### **Abstract**

The exact etiology of multiple sclerosis (MS) remains unclear but genes, environment and the interactions thereof are critical to disease susceptibility in both adults and children. Genetic epidemiological studies have clearly shown that the excess of MS among biological relatives, i.e. familial aggregation, is due to DNA sharing rather than a common intrafamilial environment. However, even among identical (monozygotic) twins who share virtually 100% of their DNA, the MS recurrence risk is only about 35%. Thus, environment and epigenetics cannot be ignored. Further complicating the transmission of MS susceptibility within families is the observed gender effect which has been replicated in many studies. There appears to be a maternal effect through both unaffected mothers and relatives of unaffected mothers. Furthermore, migration and mixed mating studies have been shown to alter an individual's risk to develop MS.

Genome-wide association studies (GWAS) have identified many potential susceptibility genes for MS but to date, none have surpassed the magnitude of the influence of HLA. Nevertheless, even the function of the HLA genotype is not straight-forward and can be influenced in many ways.

This talk will update our 2012 knowledge on the genetics and epidemiology of MS and will also identify future directions of research that are needed.

- Describe the relative roles of genes & environment & the interactions thereof in the pathogenesis of MS
- Identify possible "windows of opportunity" for intervention aimed to prevent the Clinical onset or alter the course of MS
- Review how these findings can answer questions raised by patients and family members in the clinical setting

### 2012 PRESIDENTIAL SYMPOSIUM

"How MS Could be an Acquired Disease"

Raymond A. Sobel, MD

Palo Alto VA Health Care System and Stanford University School of Medicine, Stanford, CA



**Raymond Sobel, MD** is a graduate of Stanford University and received his MD from the University of California San Francisco. He received Anatomic and Neuropathology residency training at UC Davis, UCSF and Stanford. He then did a fellowship in Immunopathology and subsequently stayed on the faculty at the Massachusetts General Hospital, Harvard Medical School. In 1992, he returned to California where he is a neuropathologist at the Palo Alto VA Health Care System and Professor of Pathology (Neuropathology) at Stanford. He has authored or coauthored 183 peer-reviewed articles in national and international journals and 6 book chapters, including the Demyelinating Diseases chapter in the 8<sup>th</sup> edition of *Greenfield's Neuropathology*. He is currently on the Editorial Boards of the *Journal of Neuroimmunology, Brain Pathology*, and has been the Editor-in-Chief of *The Journal of Neuropathology and Experimental Neurology* since 2007.

Dr. Sobel's research has primarily addressed inflammatory responses in the CNS, particularly as they relate to CNS infections, multiple sclerosis (MS) and the MS animal model, experimental autoimmune encephalomyelitis.

#### **Abstract**

The causes and fundamental nature of MS are not understood; it likely has autoimmune as well as neurodegenerative components. Edward Rubenstein, MD, Professor Emeritus, Department of Medicine, Stanford University School of Medicine has advanced the hypothesis that the naturally-occurring non-protein imino acid proline homologue Azetidine-2-carboxylic acid (Aze) is an environmental agent that contributes to MS pathogenesis. Aze is abundant in sugar beets and other edible plants. Sugar from beets is not a source of Aze consumption by humans, but after the sucrose is extracted from sugar beets, the molasses residue, which is rich in Aze, is commonly fed to livestock as a sweetener to entice them to eat their requisite grasses and grains. Over the past 200 years, (i.e. since sugar beet agriculture, which originated in Europe, became widespread), Aze entered the human food chain in milk, dairy products and meat in large amounts. The dietary misincorporation of Aze in place of proline into myelin proteins in utero or in early postnatal life, (i.e. during the period of major brain and spinal cord myelination), could contribute to unstable myelin and/or oligodendrocyte (OGC) injury. Additionally, Aze-containing proteins might affect the development of the immune repertoire and be viewed by the immune system as, "altered" or "non-self", thereby provoking autoimmune responses. This presentation will highlight the possible relationships of dietary Aze to MS history, dynamic epidemiology, pathogenesis and neuropathology. Results of ongoing studies on the effects of Aze in the induction of proteotoxic stress in OGC precursor cells (OPC) in vitro and effects of Aze feeding in mice will be presented.

The Aze hypothesis confronts numerous disparate and mysterious features of MS, including its history, epidemiology, and cellular and molecular pathogenetic mechanisms. It raises the possibility that this acquired environmental agent might enhance MS risk or lesion progression in genetically prone individuals. If it is validated, it could have major effects on the diagnosis, treatment and ultimately, the prevention of MS.

- Describe the Aze Hypothesis
- Identify the historical and epidemiological features of MS that the Aze Hypothesis addresses
- Explain how Aze has entered the human food chain
- Describe how Aze may affect mechanisms of immunity and myelin injury in MS patients

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