PROGRAM

EIGHTY SEVENTH
ANNUAL MEETING
OF THE
AMERICAN ASSOCIATION
OF
NEUROPATHOLOGISTS

JUNE 23-26, 2011

SHERATON SEATTLE HOTEL

SEATTLE, WASHINGTON

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 216-368-3671 or via email at aanp@case.edu

CONTENTS

AMERICAN ASSOCIATION OF NEUROPATHOLOGISTS

Office of Secretary-Treasurer
C. Harker Rhodes, MD, PhD
Pathology Department
Dartmouth Hitchcock Medical Center
Borwell 530W, DHMC
1 Medical Drive
Lebanon, NH 03756
Phone: 603-650-7744

Administrative Assistant – Peggy Harris
Phone: 216-368-3671
Email: aanp@cwru.edu
Home page http://www.neuropath.org

OFFICERS

Steven A. Moore, MD, PhD, *University of Iowa College of Medicine*, President
Arie Perry, MD, *University of California San Francisco*, Vice-President
Raymond A. Sobel, MD, *Stanford University School of Medicine*, President Elect
John M. Lee, MD, PhD, *Loyola University*, Vice-President Elect
Elizabeth J. Cochran, MD, *Medical College of Wisconsin*, VP for Professional Affairs
C. Harker Rhodes, MD, PhD, *Dartmouth Hitchcock Medical Center*, Secretary-Treasurer
Charles L. White III, MD, *University of Texas Southwestern Medical Center*, Assistant Secretary-Treasurer

OTHER EXECUTIVE COUNCIL MEMBERS

David N. Louis, MD, *Massachusetts General Hospital*, (Past President)
Jeffrey A. Golden, MD, *Children's Hospital of Philadelphia* (Past President)
George Perry, PhD, *University of Texas at San Antonio* (Past President)
Robert F. Hevner, MD, PhD, *University of Washington* (Member-at-Large)
Raymond A. Sobel, MD, *Stanford University School of Medicine* (JNEN)
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
Eileen S. Healy, Managing Editor
E-mail jnen@pathology.wisc.edu
Home page: http://www.jneuropath.com

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
Anne F. Buckley, MD, PhD
Mark T. Curtis, MD, PhD
Suzanne M. de la Monte, MD, MPH
Marc R. Del Bigio, MD, PhD
John E. Donahue, MD
David W. Ellison, MD, PhD
Kymberly A. Gyure, MD
Kimmo Hatanpaa, MD, PhD
Brent T. Harris, MD, PhD
Craig Horbinski, MD, PhD
Alexander Judkins, MD
Hannah C. Kinney, MD
Kathy L. Newell, MD
Bradley M. Miller, MD, PhD
Ravi Raghavan, MD

MEMBERSHIP COMMITTEE
Leslie A. Bruch, MD, Chair
Christine E. Fuller, MD
Murat Gokden, MD
Rodney D. McComb, MD
Cheryl A. Palmer, MD
Peter Pytel, MD

NOMINATING COMMITTEE
David N. Louis, MD, Chair
Barbara J. Crain, MD, PhD
Jeffrey A. Golden, MD
George Perry, PhD

PROGRAM COMMITTEE
Amyn M. Rojiani, MD, PhD, Chair
Sandra L. Cottingham, MD, PhD
James Dollar, MD
Murat Gokden, MD
Kymberly A. Gyure, MD
Eyas M. Hattab, MD
Marta Margeta, MD, PhD
Shahriar M. Salamat, MD, PhD
Suash Sharma, MD
Anat Stemmer Rachaminov, MD

PROFESSIONAL AFFAIRS
Elizabeth J. Cochran, MD, Chair
Rebecca D. Folkerth, MD
Murat Gokden, MD
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
Suzanne Z. Powell, 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
Suzanne Z. Powell, MD
C. Harker Rhodes, MD, PhD
Tarik Tihan, MD, PhD
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 this activity addresses the needs of physicians and scientist in the field of neuropathology involved in the diagnosis and/or treatment of patients with neurological disorders.

NEEDS STATEMENT

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

.

OVERALL EDUCATIONAL OBJECTIVES

After participating in AANP's 2011 Annual Meeting learners should be able to:

- Describe general and specific techniques for evaluating muscle biopsies.
- Define criteria for establishing a specific diagnosis in a variety of genetic and acquired myopathies.
- Explan the pathogenesis of major subcategories of neuropathic and myopathic disorders.

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 26.25 *AMA PRA Category 1 Credits™* . 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
Case Western Reserve University
2103 Cornell Road, WRB 5101
Cleveland, Ohio 44106

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™* at this year's Annual Conference.

Activity	CME Credit Hours
Special Course	7
Scientific Sessions	10 (Platform = 8; Posters=2)
Korey Lecture	1
DeArmond Lecture	1
Parisi Lecture	1
Diagnostic Slide Session	3
Presidential Symposium	3.25
Total	**26.25**

DISCLOSURE INFORMATION:

Disclosure of Commercial Support:
This activity is supported by an educational grant from Teva Neurosciences. "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 do 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
Elizabeth J. **Cochran**, Medical College of Wisconsin	None
Mark L. **Cohen**, University Hospitals Case Medical Center	None
Sandra L. **Cottingham**, Spectrum Health Hospitals	None
Barbara J. **Crain**, Johns Hopkins University	None
James D. **Dollar**, Carolinas Pathology Group	None
Rebecca D. **Folkerth**, Brigham and Woman's Hospital/ Children's Hospital of Boston	None
Kar-Ming **Fung**, University of Oklahoma	None
Murat **Gokden**, University of Arkansas	None
Jeffrey A. **Golden**, Children's Hospital of Philadelphia	None

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 Faculty/Presenter	**Reported Financial Relationship**
Kymberly A. **Gyure**, West Virginia University	None
Eyas M. **Hattab**, Clarian Pathology Laboratory	Speaker's Bureau: Eisai, Inc.
Robert F. **Hevner**, University of Washington	None
William F. **Hickey**, Dartmouth Medical School	None
John M. **Lee,** Loyola University	Consultant/Reviewer For Up to Date on CJD articles; Holds group patents with Cornelli Consulting
David N. **Louis**, Massachusetts General Hospital	None
William C. **McDonald**, Abbott Northwestern Hospital	None
Marta **Margeta**, University of California San Francisco	None
Brian E. **Moore**,Memorial Medical Center	None
Steven A. **Moore**, University of Iowa	None
Robert E. **Mrak**, University of Toledo	None
Kathy L. **Newell**, University of Kansas Medical Center	None
Arie **Perry**, University of California San Francisco	None
George **Perry**, University of Texas at San Antonio	None
Suzanne **Powell**, The Methodist Hospital	None
C. Harker **Rhodes**, Dartmouth-Hitchcock Medical Center	None
Marie **Rivera-Zengotita**, University of Florida Medical College	None
Amyn **Rojiani**, Medical College of Georgia	None
Shahriar M. **Salamat**, University of Wisconsin Hospital	None
Suash **Sharma**, Medical College of Georgia	None
Raymond A. **Sobel,** Stanford University School of Medicine	None
Anat **Stemmer Rachaminov**, Massachusetts General Hospital	None
Tarik **Tihan**, University California San Francisco	None
Karen M. **Weidenheim**, Montefiore Medical Center	None
Charles L. **White III,** University of Texas Southwestern Medical Center	None
Anthony T. **Yachnis**, University of Florida Medical College	None

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

Ty **Abel**, Vanderbilt University Medical Center	None
Homa **Adle-Biassette**, INSERM	None
Fahad **Alghamdi**, Vancouver General Hospital	None
Sarah **Alghamdi**, Mount Sinai Medical Center	None
Dana **Altenburger**, University of California Los Angeles	None

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**
Bijan **Ameri**, Brain Tumor Translational Resource	None
Christina **Appin**, Emory University School of Medicine	None
Derick **Aranda**, University of California San Francisco	None
Knarik **Arkun**, Montefiore Medical Center	None
Pavan **Auluck**, Massachusetts General Hospital	None
Jennifer **Baccon**, Penn State Hershey Medical Center	None
Jilin **Bai**, University of Connecticut School of Medicine	None
Michael **Barnes**, University of California San Francisco	None
Thomas **Beach**, Banner Sun Health Research Institute	Research support: Eli Lily, GE, Bayer
Jennifer **Bennett**, Penn State, Hershey Medical Center	None
Juan **Bilbao**, Sunnybrook Hospital, University of Toronto	None
Daniel **Brat**, Emory University	None
Kevin **Broadbelt**, Children's Hospital Boston	None
Steven **Carroll**, University of Alabama at Birmingham	None
Rudy **Castellani**, University of Maryland	None
Howard **Chang**, Sparrow Health System	None
Stephen **Coons**, Barrow Neurological Institute	Global Media Advisory Board
John **Crary**, Columbia University	None
Sidney **Croul**, University of Toronto	None
Jane **Cryan**, Beaumont Hospital, Dublin, Ireland	None
Matthew **Cykowski**, Oklahoma University Health Sciences Center	None
Christian **Davidson**, Massachusetts General Hospital	None
Stephen **DeArmond**, University of California San Francisco	None
Jeremy **Deisch**, University of Texas Southwestern Medical Center	None
Marc **Del Bigio**, University of Manitoba	None
Laura **Denham**, Loma Linda University Medical Center	None
Dennis **Dickson**, Mayo Clinic	None
John **Donahue**, Rhode Island Hospital	None
Andrew **Dwork**, New York State Psychiatric Institute, Columbia University	Loan of equipment and Software – Olympus and Visiopharn.
Benjamin **Ellezam**, The Methodist Hospital	None
David **Ellison**, St. Jude Children's Research Hospital	None
Jennifer **Eschbacher**, Barrow Neurological Institute/St. Joseph's Hospital and Medical Center	None

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**
Mary **Fowkes**, Mount Sinai Medical Center	None
Morgan **Freret**, Stanford University	None
Shintaro **Fukushima**, National Cancer Center Hospital	None
Marla **Gearing**, Emory University	None
Bernardino **Ghetti**, Indiana University	Consultant: Bayer
Murat **Gokden**, University of Arkansas	None
Peter **Gould**, CHA Hôpital de l'Enfant-Jésus	None
Kymberly **Gyure**, West Virginia University	None
Terri **Haddix**, Stanford University Medical Center	None
Brian **Harding**, Children's Hospital of Philadelphia	None
Brent **Harris**, Georgetown University School of Medicine	None
Keith **Harrison**, The Children's Hospital of Alabama	None
Cynthia **Hawkins**, The Hospital for Sick Children	None
John **Hedreen**, Harvard Brain Tissue Resource Center	None
Emily **Herndon**, University of Texas Southwestern Medical Center at Dallas	None
Annie **Hiniker**, University of California San Francisco	None
Craig **Horbinski**, University of Kentucky	None
Byron **Huff**, University of North Carolina	None
Jason **Huse**, Memorial Sloan-Kettering Cancer Center	None
Cristiane **Ida**, Mayo Clinic	None
Mircea **Iftinca**, University of calgary	None
Mark **Jentoft**, Mayo Clinic College of Medicine	None
Jeffrey **Joseph**, University of Calgary	None
Nancy **Joseph**, University of California San Francisco	None
Keith **Josephs**, Mayo Clinic	None
Gordana **Juric-Sekhar**, University of Washington	None
Spyridon **Karadimas**, Toronto Western Research Institute Krembil Neuroscience Research Center	None
Soma **Karak**, Hartford Hospital	None
Syed **Kazmi**, University of Nebraska Medical Center	None
Julia **Keith**, University of Toronto, Sunnybrook Health Sciences Centre	None
Negar **Khanlou**, University of California Los Angeles	None
Bette **Kleinschmidt-DeMasters**, University of Colorado Anschutz Medical Campus	None
Keyla **Kleyser-Sugrue**, Hartford Hospital	None
Georgios **Klironomos**, University of Patras	None
Arnulf **Koeppen**, VA Medical Center	None
Julia **Kofler**, University of Pittsburgh	None
Hans **Kretzschmar**, Institut für Neuropathologie Universität München	None

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**
Michael **Kyle**, Loma Linda University Medical Center	None
Boleslaw **Lach**, McMaster University	None
Facundo **Las Heras**, University of Chile Clinical Hospital	None
Michael **Lawlor**, Children's Hospital Boston, Harvard Medical School	Patent Pending for use of a drug in cong. Myopathy, Collaborator- Acceleron Pharma Inc.
Edward **Lee,** University of Pennsylvania	None
Han **Lee**, University of California San Francisco	None
Kyung-Hwa **Lee**, Northwestern University Feinberg School of Medicine	None
Min-Cheol **Lee**, Chonnam National University Medical School	None
Hart **Lidov**, Children's Hospital Boston	None
Wen-Lang **Lin**, Mayo Clinic	None
Jian-Qiang **Lu**, University of Alberta	None
Fabiana **Lubieniecki**, Pediatric Hospital JP Garrahan	None
Michelle **Madden Felicella**, University of California San Francisco	None
James **Mandell**, University of Virginia	None
Qinwen **Mao**, Northwestern University	None
Sergey **Mareninov**, Brain Tumor Translational Resource	None
Marta **Margeta**, University of California San Francisco	None
Stefanie **Marquez**, University of Southern California	None
Sarah **Martin**, Indiana University School of Medicine	None
Maria **Martinez-Lage**, University of Pennsylvania	None
Kant **Matsuda**, New York University	None
Ann **McKee**, Bedford VAMC/Boston University School of Medicine	None
Janet **McNaughton**, Orlando Health	None
Jean **Michaud**, Children's Hospital of Eastern Ontario and University of Ottawa	None
C. **Miller**, University of North Carolina	None
Leili **Mirsadraei**, Brain Tumor Translational Resource	None
Hajime **Miyata**, Research Institute for Brain and Blook Vessels - Akita	None
Bret **Mobley**, Vanderbilt University	None
Carrie **Mohila**, University of Virginia	None
Edwin **Monuki**, University of California Irvine	None
Michela **Morbin**, IRCSS Foundation Istituto Neurologico 'Carlo Besta'	None
Sverre **Mörk**, Haukeland University Hospital	None
Shigeo **Murayama**, Tokyo Metropolitan Geriatric Hospital & Institute of Gerontology	None

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**
Masataka **Nakamura**, Kansai Medical University	None
Peter **Nelson**, University of Kentucky	None
Janna **Neltner**, University of Kentucky	None
Hilary **Nickols**, Vanderbilt University	None
José **Otero**, University of California San Francisco	None
Cheryl **Palmer**, University of Alabama at Birmingham	None
Maryam **Pezhouh**, University of Minnesota	None
Rolf **Pfannl**, Tufts Medical Center	None
Clyde **Phelix**, University of Texas at San Antonio	None
Joanna **Phillips**, University of California San Francisco	None
Christopher **Pierson**, Nationwide Children's Hospital and Ohio State University	None
Edward **Plowey**, University of Pittsburgh Medical Center	None
Jocelyn **Posthumus**, University of Virginia Health System	None
Peter **Pytel**, University of Chicago	None
Shakti **Ramkissoon**, Brigham and Women's Hospital	None
Miguel **Riudavets**, FLENI	None
Fausto **Rodriguez**, John Hopkins University	None
Amyn **Rojiani**, Georgia Health Sciences University, Medical College of Georgia	None
Jennifer **Ross**, Harris County Institute of Forensic Sciences	None
Desiree **Sanchez**, University of California Los Angeles	None
Mariarita **Santi**, Children's Hospital of Philadelphia	None
Harvey **Sarnat**, University of Calgary	None
Bernd W. **Scheithauer**, Mayo Clinic	None
Matthew **Schniederjan**, Emory University School of Medicine	None
Yuan **Shan**, Moffitt Cancer Center	None
Suash **Sharma**, Medical College of Georgia	None
Mark **Smethurst**, Mount Sinai Medical Center	None
Matija **Snuderl**, Massachusetts General Hospital	None
Charles **Specht**, Penn State Hershey Medical Center	None
Thor **Stein**, Massachusetts General Hospital	None
Anat **Stemmer-Rachamimov**, Massachusetts General Hospital	None
Kimberly **Stogner-Underwood**, Virginia Commonwealth University Health System	None
Ethan **Stolzenberg**, University of Oklahoma Health Sciences Center	None
Yasuo **Sugita**, Kurume University School of Medicine	None
Mario **Suvà** Massachusetts General Hospital	None

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

Masaki **Takao**, Tokyo Metropolitan Geriatric Hospital	None
Hidehiro **Takei**, The Methodist Hospital	None
Jantima **Tanboon**, Siriraj Hospital	None
Ana Lia **Taratuto** Institute for Neurological Research FLENI	None
Dimitri **Trembath**, University of North Carolina	None
Vinata **Vedam-Mai**, McKnight Brain Institute	None
Sriram **Venneti**, University of Pennsylvania	None
Cristina **Vincentelli**, Emory University	None
Nguyen **Vo**, LSUHSC	None
Aaron **Wagner**, University of California Los Angeles	None
W. **Waldman**, The Ohio State University	None
Garth **Warren**, Oregon Health and Sciences University	None
Keith **Wharton**, Biogen Idec	Employee and Stockholder – Biogen Idec
Andrea **Wiens**, Indiana University School of Medicine	None
Clayton **Wiley**, University of Pittsburgh Medical Center	None
Christopher **William**, Massachusetts General Hospital	None
Jon **Wilson**. Louisiana State University Health Sciences Center Shreveport	None
Zhenggang **Xiong**, Tulane University	None
Gang **Xu**, Children's Hospital Boston and Harvard Medical School	None
Anthony **Yachnis**, University of Florida College of Medicine	None
Stephen **Yip**, BC Cancer Agency	None
Jianying **Zeng**, Suny Downstate Medical Center	None

GENERAL INFORMATION

Hotel: Sheraton Seattle Hotel
1400 Sixth Avenue
Seattle, WA 98101
USA

Phone: 206-621-9000

ALL MEETING SESSIONS WILL BE HELD AT THE SHERATON SEATTLE HOTEL

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 **Metropolitan Ballroom** of the hotel on the third floor.

All poster sessions will be held in **Metropolitan Pre-function** area on the third 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 2011 issue of JNEN with abstracts, reception ticket(s) and registration receipt ready for pick-up at the AANP Registration Desk, located in the Metropolitan Pre-function area of the hotel on the third floor. On-site registration and additional tickets for the Annual Reception will be available at the Desk.

REGISTRATION DESK

Location	**Metropolitan Foyer/Pre-Function Area**	
Time	Wednesday, June 22	6:30 pm – 9:00 pm
Time	Thursday, June 23	6:30 am - 12 noon 6:30 pm – 9:00 pm
	Friday, June 24	7:00 am - 12 noon 5:30 pm – 6:00 pm
	Saturday, June 25	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 **Metropolitan Ballroom A or B** 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 **Metropolitan 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, Metropolitan Ballroom A or B, 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 *only* times available to you to load and test your presentation.**

Schedule for Loading PowerPoint Presentations

Load show computer in *Metropolitan A or B*	
Thursday, June 23	6:45 am - 7:45 am 5:30 pm - 6:30 pm
Friday, June 24	6:45 am - 7:45 am 6:00 pm - 7:00 pm
Saturday, June 25	6:45 am – 7:45 am 6:30 pm – 7:45 pm (Congress Room, 4th Floor)
Sunday, June 26	6:45 am - 7:45 am

- **If you are presenting in a morning session, it is preferable to check in the previous day**. Same-day 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 Congress Room on the fourth floor 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 **Metropolitan Pre-Function Area.** 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 6 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.

As a requirement to offer AMA PRA Category 1 Credit™ and 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 24 ***Authors Present at:***	**Sat June 25** ***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 ***Queen Anne Room*** on the third floor of the hotel.

Location	**Queen Anne Room**	
Time	Thursday, June 23	7:00 am - 5:30 pm
	Friday, June 24	7:00 am - 5:30 pm
	Saturday June 25	7:00 am - 5:30 pm

BUSINESS MEETING

Location	**Metropolitan Ballroom B**	
Time	Friday, June 24	11:45 am - 12:45 pm
	Saturday June 25	11:45 am – 12:45 pm

The awards for ***Meritorious Contributions to Neuropathology*** will be presented on Saturday, June 25

SPECIAL MEETINGS BY INVITATION ONLY

Date	**Meeting**	**Time/Location**
Thurs June 23	Executive Council Meeting	6:00 pm Issaquah Room, third floor of the hotel
Fri June 24	Education Committee Meeting	7:00 am Wallingford Room, third floor of the hotel
	Trainee Luncheon	12:45 pm – 2:00 pm Issaquah Room, third floor of the hotel
	Awards Committee Meeting	5:30 pm – 6:30 pm Medina Room, third floor of the hotel
Saturday June 25	JNEN Editorial Board Meeting	7:00 am – 8:00 am Issaquah Room, third floor of the hotel
	NP Program Directors Meeting	1:00 pm – 2:00 pm Medina Room, third floor of the hotel
	Awards Committee Meeting	6:00 pm 7:30 pm Medina Room, third floor of the hotel
	Professional Affairs	6:00 pm – 8:00 pm Leschi Room, Third floor of the hotel
	Presidential Reception	6:30 pm – 8:00 pm Issaquah Room, third floor of the hotel
Sun June 26	Founders Breakfast	7:00 am – 8:00 am Medina Room, third floor of the hotel

ABSTRACTS

Abstracts of the papers presented in the program are published in the June 2011 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 Metropolitan Ballroom. 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	**Metropolitan Ballroom**	
Time	Friday, June 24	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 Kirkland room on the third floor.

Location	**Kirkland Room**	
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

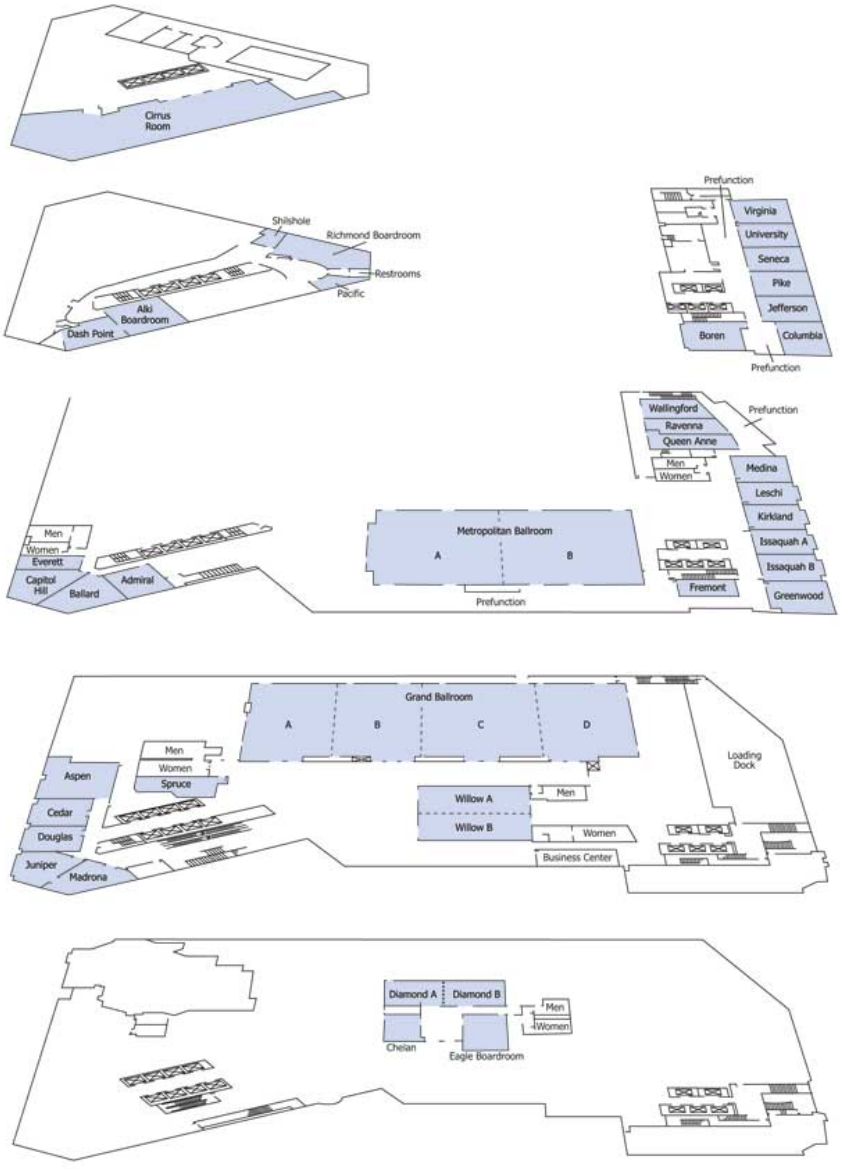
MEETING EXHIBITORS

- Eisai
- Olympus America, Inc.
- Wolters Kluwers Health

RECEPTION PRIZE CONTRIBUTORS

- Wolters Kluwers Health

Sheraton Seattle Hotel Floor Plan



PROGRAM and SCIENTIFIC SESSIONS

SPECIAL COURSE:

Location	**Metropolitan Ballroom**	
Date/Time	Thursday, June 23	8:00 am - 5:00 pm
	Common Problems in Muscle Biopsy Diagnostics Directors: Steven A. Moore, MD, PhD and Rodney D. McComb, MD	

PLATFORM PRESENTATIONS

Location	**Metropolitan Ballroom A & B**	
Date/Time	Friday, June 24	8:00 am – 4:00 pm
	Saturday, June 25	8:00 am – 4:00 pm
	Saturday, June 25 - Bio/Brain Banking	4:30 am – 5:30 pm

POSTER PRESENTATIONS

Location	**Metropolitan Pre-Function Area**	
Date/Time	Friday, June 24	8:00 am – 6:30 pm
	Saturday, June 25	8:00 am – 6:30 pm

PARISI LECTURE

Location	**Metropolitan Ballroom**	
Date/Time	Friday, June 24	10:30 am - 11:30 am
	Molecular Pathologies at the Nodes of Ranvier Steven S. Scherer, MD, PhD *University of Pennsylvania, Philadelphia, PA*	

DEARMOND LECTURE

Location	**Metropolitan Ballroom**	
Date/Time	Friday, June 24	4:30 pm – 5:30 pm
	Emerging Therapies for Neurogenetic Diseases Beverley L. Davidson, PhD *University of Iowa, Iowa City, IA, USA*	

SAUL R. KOREY LECTURE

Location	**Metropolitan Ballroom**	
Date/Time	Saturday, June 25	10:30 am - 11:30 am
	Protein Aggregate Myopathies Hans H. Goebel, MD *Johannes Gutenberg-University Medical Center, Mainz, Germany*	

DIAGNOSTIC SLIDE SESSION

Location	**Metropolitan Ballroom**	
Date/Time	Saturday, June 25	8:00 pm -11:00 pm

PRESIDENTIAL SYMPOSIUM

Location	**Metropolitan Ballroom**	
Date/Time	Sunday, June 26	8:00 am – 12 noon
	"Dystroglycan-Related Congenital Muscular Dystrophies"	

MEETING AT A GLANCE

THURSDAY June 23, 2011	
	Metropolitan Ballroom
8:00 am - 5:15 pm	*SPECIAL COURSE* *Common Problems in Muscle Biopsy Diagnostics*

(Abstract Numbers in Italics)

FRIDAY June 24, 2011			
	Metropolitan Ballroom A	Metropolitan Ballroom B	Metropolitan Pre-Function Area
8:00 - 10:00 am	Platform 1 Tumors- Glial *#1 - 8*	Platform 2 Neurodegenerative – Alzheimer's Disease *#9 - 16*	All Posters CME Credit offered for participation at the following times: Friday June 24th and Saturday June 25th 10:00 – 10:30 am 4:00 - 4:30 pm
10:00 - 10:30 am	*REFRESHMENT BREAK*		
10:30 - 11:30 am	*PARISI LECTURE* *Metropolitan Ballroom* *Molecular Pathologies at the Nodes of Ranvier* Steven S. Scherer, MD, PhD *University of Pennsylvania, Philadelphia, PA*		
11:45 - 12:45 pm	*BUSINESS MEETING I* *Metropolitan Ballroom*		
12:45 - 2:00 pm	*LUNCH*		
	Metropolitan Ballroom A	Metropolitan Ballroom B	
2:00 - 4:00 pm	Platform 3 Inflammation/Infection/ Myelin *#17-24*	Platform 4 Neurodegenerative: FTD/Lewy Body/Parkinson *#25 -32*	
4:00 – 4:30 pm	*REFRESHMENT BREAK*		
4:30 – 5:30 pm	*DEARMOND LECTURE* *Metropolitan Ballroom* *Emerging Therapies for Neurogenetic Diseases* Beverley L. Davidson, PhD *University of Iowa, Iowa City, IA, USA*		

6:30 - 8:30 pm *ANNUAL RECEPTION:*
Metropolitan Ballroom

MEETING AT A GLANCE

(Abstract Numbers in Italics)

<table>
<tr><th colspan="4">SATURDAY June 25, 2011</th></tr>
<tr><td></td><td>Metropolitan Ballroom A</td><td>Metropolitan Ballroom B</td><td>Metropolitan Pre-Function Area</td></tr>
<tr><td>8:00 - 10:00 am</td><td>Platform 5
Muscle and Nerve

#97 - 104</td><td>Platform 6
Tumors

#105-112</td><td rowspan="11">All Posters
CME Credit offered for participation at the following times:

Friday June 24th and Saturday June 25th
10:00 – 10:30 am
4:00 - 4:30 pm</td></tr>
<tr><td>10:00 - 10:30 am</td><td colspan="2">REFRESHMENT BREAK</td></tr>
<tr><td>10:30 - 11:30 am</td><td colspan="2">SAUL KOREY LECTURE
Metropolitan Ballroom

Protein Aggregate Myopathies

Hans H. Goebel, MD
Johannes Gutenberg-University Medical Center, Mainz, Germany</td></tr>
<tr><td>11:45 - 12:45 pm</td><td colspan="2">BUSINESS MEETING II
Metropolitan Ballroom</td></tr>
<tr><td>12:45 - 2:00pm</td><td colspan="2">LUNCH</td></tr>
<tr><td></td><td>Metropolitan Ballroom A</td><td>Metropolitan Ballroom B</td></tr>
<tr><td>2:00 - 4:00 pm</td><td>Platform 7
Developmental/Pediatric

#113-120</td><td>Platform 8
Neurodegenerative - Other

#121-128</td></tr>
<tr><td>4:00 - 4:30 pm</td><td colspan="2">REFRESHMENT BREAK</td></tr>
<tr><td>4:30 - 5:30 pm</td><td colspan="2">BIO/BRAIN BANKING SESSION
Metropolitan Ballroom A</td></tr>
<tr><td>8:00 - 11:00 pm</td><td colspan="2">DIAGNOSTIC SLIDE SESSION
Metropolitan Ballroom</td></tr>
</table>

SUNDAY June 26, 2011	
	Metropolitan Ballroom
8:00 am - 12:00 pm	*PRESIDENTIAL SYMPOSIUM* *"Dystroglycan-Related Congenital Muscular Dystrophies"*

THURSDAY, June 23, 2011

SPECIAL COURSE

Common Problems in Muscle Biopsy Diagnostics

Directors: Steven A. Moore, MD, PhD and Rodney D. McComb, MD

Metropolitan Ballroom

8:00 am	Welcome and CME Pretest *Steven A. Moore, MD, PhD* *University of Iowa, Iowa City, IA* *and* *Rodney D. McComb,MD* *University of Nebraska, Omaha, NE*
8:15 am – 9:00 am	Muscle Biopsy Interpretation: The Basics *Reid Heffner, MD* *University at Buffalo, Buffalo, NY*
9:00 am – 9:45 am	Neuropathic Aspects of Muscle Pathology *Hannes Vogel, MD* *Stanford University, Palo Alto, CA*
9:45 am – 10:30 am	Distal Myopathies: Using Muscle MRI for Diagnostic Testing *Bjarne Udd, MD, PhD* *University of Tampere, Finland*
10:30 am – 11:00 am	*REFRESHMENT BREAK*
11:00 am – 11:45 am	Pathological Characterization of Acquired Immune and Inflammatory Myopathies *Alan Pestronk, MD* *Washington University, St. Louis, MO*
11:45 am – 12:30 pm	Ragged Red Fibers and Beyond: Mitochondrial Disease and Other Metabolic Myopathies *Kurenai Tanji,PhD* *Columbia University, New York, NY*
12:30 pm - 1:30 pm	*LUNCH*
1:30 pm – 2:15 pm	Muscular Dystrophies: Integrating the Muscle Biopsy with Molecular Diagnostics *Steven A. Moore, MD, PhD* *University of Iowa, Iowa City, IA*
2:15 pm – 3:00 pm	Floppy Infants and Arthrogryposis *Caroline Sewry, PhD* *Dubowitz Neuromuscular Centre, London, UK*
3:00 pm - 3:30 pm	*REFRESHMENT BREAK*
3:30 pm – 4:15 pm	Autophagic Vacuolar Myopathies *Rodney D. McComb, MD* *University of Nebraska, Omaha, NE*
4:15 pm – 5:00 pm	Training Issues for Muscle Pathology *Elizabeth Cochran, MD* *Medical College of Wisconsin, Milwaukee, WI* *and* *Suzanne Powell, MD* *The Methodist Hospital, Houston, TX*

FRIDAY, JUNE 24, 2011

TRAINEE LUNCHEON: FORMULA FOR SUCCESS
(Not Offered for CME Credit)

12:45 pm – 2:00 pm - Issaquah Room

Formula for Success in the Clinical/Translational Career Track *Arie Perry, MD* *University of California San Francisco*
Formula for Success in the Basic Science Career Track *Thomas Montine, MD, PhD* *University of Washington, Seattle, WA*
Job Fair

SUNDAY, JUNE 26, 2011

PRESIDENTIAL SYMPOSIUM

Dystroglycan-Related Congenital Muscular Dystrophies

Metropolitan Ballroom

8:00 am – 8:05 am	Introduction and Overview of CMDs *Steven A. Moore, MD, PhD* *University of Iowa, Iowa City, IA*
8:05 am - 9:00 am	***Matthew T. Moore Distinguished Lecture*** Mechanistic and Molecular Insights into the Pathogenesis of Glycosylation-Deficient Muscular Dystrophy *Kevin P. Campbell, PhD* *University of Iowa, Iowa City, IA*
9:00 – 9:45 am	Brain Development is Sugar-Coated: The Emerging Story of the Cobblestone Cortical Malformations *William B. Dobyns, MD* *University of Washington, Seattle, WA*
9:45 am – 10:30 am	AANP Award Presentations and Refreshment Break
10:30 am – 11:15 am	Neuronal Migration Abnormalities in Dystroglycanopathies and Related Disorders *Robert F. Hevner, MD, PhD* *University of Washington, Seattle, WA*
11:15 am – 12:00 pm	Peripheral Nerve Laminin and Dystroglycan – The Basis for Neuropathies in Congenital Muscular Dystrophies *Steven A. Moore, MD, PhD* *University of Iowa, Iowa City, IA*
12:00 pm	*INSTALLATION OF NEW OFFICERS AND ADJOURNMENT*

FRIDAY, JUNE 24, 2011
Metropolitan Ballroom A
8:00 am – 2:00 pm

Platform 1 Tumors - Glial
Chairpersons: Christine E. Fuller, MD and Tarik Tihan, MD, PhD

8:00- 8:15	1	**Integrated Analysis of Necrosis, Angiogenesis, and Gene Expression in Glioblastoma** Daniel Brat, Lee Cooper, Carlos Moreno, Candace Chisolm, Christina Appin, David Gutman, Jun Kong, Tahsin Kurc, Joel Saltz
8:15- 8:30	2	**Genome-Wide Chromatin Landscape of Glioblastoma Cancer Stem Cells Highlights an Aberrant Network of Transcription Factors** Mario Suvà, Andrew Chi, Esther Rheinbay, Hiro Wakimoto, Miguel Rivera, Ozgur Oksuz, Samuel Rabkin, Robert Martuza, David Louis, Bradley Bernstein
8:30- 8:45	3	**Correlates of Mass Spectrometric Measurements of Gadolinium in Gliomas Undergoing Fluorescence Guided Resection** Brent Harris, Pablo Valdes, Ziev Moses, Anthony Kim, Carolyn Niu, Marco Brantsch, Brian Wilson, Keith Paulsen, David Roberts
8:45- 9:00	4	**BRAF-KIAA1549 Fusion Predicts Better Clinical Outcome in Pediatric Low Grade Astrocytoma** Cynthia Hawkins, Erin Walker, Nequesha Mohamed, Iris Fried, Nada Jabado, Uri Tabori
9:00- 9:15	5	**BRAF Alterations in Primary Glial and Glioneuronal Neoplasms of the Central Nervous System: Clinicopathologic Correlations** Fausto Rodriguez, Alex Lin, Susan Williams, Matthias Karajannis, David Zagzag, Peter Burger, Eric Raabe, Kenneth Cohen, Charles Eberhart, Eli Bar
9:15- 9:30	6	**Diffuse Astrocytomas can Arise from either GFAP+ Astrocytes or Subventricular Zone (SVZ) Neural Stem/Progenitor Cells** C. Miller, Ryan Bash, Mark Vitucci, Andrea Werneke, Ralf Schmid
9:30- 9:45	7	***In-vivo* and Immediate Ex-vivo Confocal Laser Endomicroscopy for Intraoperative Histopathological Imaging of Brain Tumors** Jennifer Eschbacher, Nikolay Martirosyan, Nader Sanai, Kris Smith, Mark Preul, Stephen Coons, Peter Nakaji, Robert Spetzler
9:45- 10:00	8	**The Impact of IDH1 and IDH2 Mutations on Progression-Free and Overall Survival in Low Grade Gliomas** Sidney Croul, Christiane Knobbe, Nesrin Sabha, Andreas von Deimling, Abhijit Guha

10:00 - 10:30 am ***REFRESHMENT BREAK***

10:30 – 11:30 am ***Parisi Lecture***
Molecular Pathologies at the Nodes of Ranvier
Steven S. Scherer, MD, PhD
University of Pennsylvania, Philadelphia, PA

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

12:45 – 2:00 pm ***Lunch***

FRIDAY, JUNE 24, 2011
Metropolitan Ballroom B
8:00 am – 2:00 pm

Platform 2 Neurodegenerative – Alzheimer Disease
Chairpersons: Shigeo Murayama, MD, PhD and Elizabeth Cochran, MD

8:00- 8:15	9	**Differences in Clinical and Neuropathological Concepts of Alzheimer's Disease at NIA Alzheimer's Disease Centers 2005-2010** Thomas Beach, Leslie Phillips, Sarah Monsell, Walter Kukull
8:15- 8:30	10	**Towards a Novel Therapeutic Strategy for Alzheimer Disease (AD) Based on Modified Aß Peptides.** Michela Morbin, Giulia Mazzolelni, Andrea Uggetti, Laura Colombo, Marco Gobbi, Matteo Salvalglio, Giuseppe Di Fede, Marcella Catania, Mario Salmona, Fabrizio Tagliavini
8:30- 8:45	11	**Intraneuronal APP, not Free Aß Peptides in 3xTg-AD Mice: Implications for Tau Versus Aß Mediated Alzheimer Neurodegeneration** Edward Lee, Matthew Winton, Eveline Sun, Margaret Wong, Susan Leight, Bin Zhang, John Trojanowski, Virginia Lee
8:45- 9:00	12	**Ocular Dominance Plasticity Defect in Alzheimer Disease Model Mice** Christopher William, Carla Shatz, Matthew Frosch, Bradley Hyman
9:00- 9:15	13	**Dementia without AD in Advanced Aging: Many Diseases, Much Confusion** Peter Nelson
9:15- 9:30	14	**Striatal Plaque Density Predicts Braak Tangle Stage and Clinicopathological AD: Implications for Amyloid Imaging** Thomas Beach, Lucia Sue, Monica Mariner, Kim Yantos, Leslie Souders, Glenn Chiarolanza, Jonette Henry-Watson, Geidy Serrano, Marwan Sabbagh, Douglas Walker
9:30- 9:45	15	**Characterization of Human Brains Resilient to ß-Amyloid and Tau Pathologies** Thor Stein, Oriol Dols, Thomas Scotton, Bibiana Da Rocha-Souto, Serrano-Pozo Alberto, Frosch Matthew, Growdon John, Hyman Bradley, Gomez-Isla Teresa
9:45- 10:00	16	**Relationship Between Apolipoprotein E Genotype and Blood-Brain Barrier Permeability** John Donahue, Junghyun Kim, Jill Dennis, Michelle Reyzer, Richard Caprioli

10:00 - 10:30 am ***REFRESHMENT BREAK***

10:30 – 11:30 am ***Parisi Lecture***
Molecular Pathologies at the Nodes of Ranvier
Steven S. Scherer, MD, PhD
University of Pennsylvania, Philadelphia, PA

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

12:45 – 2:00 pm ***Lunch***

FRIDAY, JUNE 24, 2011
Metropolitan Ballroom A
2:00 pm – 6:30 pm

Platform 3: Inflammation/Infection/Myelin
Chairpersons: Susan Staugaitis, MD, PhD and G. Wayne Moore, MD

2:00 – 2:15	17	**Paradoxical *in vivo* versus *in vitro* Expression of YKL-40 by Macrophages and Astrocytes** Clayton Wiley, Julia Kofler, Adam Starkey, Guoji Wang, Dafna Bonneh-Barkay
2:15- 2:30	18	**MiRNA Expression Profiles of Classically and Alternatively Activated Human Adult Microglia** Julia Kofler, Stephanie Bissel, Uma Chandran, Mark Stauffer, Clayton Wiley
2:30- 2:45	19	**Automated Identification of Myelin in Paraffin Sections** Andrew Dwork
2:45- 3:00	20	**Myelin Loss Despite Normal Oligodendroglial Density in Autosomal Dominant Leukodystrophy with Duplication of the LMNB1 Gene** Michael Barnes, Shu-Ting Lin, Ying-Hui Fu, Louis Ptácek, Eric Huang
3:00- 3:15	21	**Central Nervous System Demyelination Following Hepatitis A and DTaP Vaccinations Clinically Mimicking High-grade Glioma** Sarah Martin, Joel Boaz, Mandy Harris, Eyas Hattab
3:15- 3:30	22	**Neuroprotection Provided by IV Transplanted Human Fetal Cerebral Endothelial Cells in an Experimental Brain Ischemia Model** Min-Cheol Lee, Kyung-Hwa Lee, Hyung-Seok Kim, Seung U Kim
3:30- 3:45	23	**Antenatal and Perinatal Coxsackie B4 Brain Infection: Case Series** Marc Del Bigio, Jennifer Hunt
3:45- 4:00	24	**Neurosurgical Presentation of Inflammatory Demyelinating Disease: Clinical, Neuroimaging and Neuropathologic Correlation** Suash Sharma, Mitzi Williams, Nicole DeMers-Riddle, Pushpa Allam-Nandyala, James Crownover, Steven Brem, Reed Murtagh, Cargyle Allen, Samuel Macomson, Amyn Rojiani

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

4:30 – 5:30 pm ***DeArmond Lecture***
Emerging Therapies for Neurogenetic Diseases
Beverley L. Davidson, PhD
University of Iowa, Iowa City, IA

6:30 – 8:30 pm ***Annual Reception***
Metropolitan Ballroom

FRIDAY, JUNE 24, 2011
Metropolitan Ballroom B
2:00 pm – 6:30 pm

Platform 4: Neurodegenerative: FTD/Lewy Body/Parkinson
Chairpersons: Hans Kretzschmar, MD, PhD and Joseph E. Parisi, MD

2:00 – 2:15	25	**Mitochondrial Fragmentation Mediates 1-methyl-4-phenylpyridinium Toxicity in Neurons: Implication for Parkinson Disease** Xiongwei Zhu, Bo Su, Yuan Gao, Rudy Castellani, George Perry, Mark Smith, Xinglong Wang
2:15- 2:30	26	**Hierarchical Cluster Analysis of Cortical Pathology Suggests Pathologic Heterogeneity of Dementia in Parkinson's Disease** Dennis Dickson, Carolyn Orr, Melissa Murray, Rajesh Pahwa, Kelly Lyons, Samuel Goldman, William Langston, Zbigniew Wszolek, Ryan Uitti, Neill Graff-Radford, Tanis Ferman
2:30- 2:45	27	**Causal Therapy of Prion and Parkinson´s Disease with Novel Inhibitors of Protein Aggregation** Hans Kretzschmar, Jens Wagner, Sergey Ryazanaov, Andrei Leonov, Johannes Levin, Song Shi, Catharina Prix, Martin Groschup, Christian Griesinger, Armin Giese
2:45- 3:00	28	**Overexpression of Mutant LRRK2 in Cultured Cortical Neurons is Associated with Excitotoxic Neurodegeneration** Edward Plowey, Jon Johnson, Charleen Chu
3:00- 3:15	29	**Ultrastructural Localization of FUS/TLS in Neuronal Intermediate Filament Inclusion Disease** Wen-Lang Lin, Dennis Dickson
3:15- 3:30	30	**Light and Ultrastructural Analysis of FUS Immunoreactivity in Neuronal Intermediate Filament Inclusion Disease** Keith Josephs, Wen-Lang Lin, Joseph Parisi, Neil Graff-Radford, Ronald Petersen, Dennis Dickson
3:30- 3:45	31	**Neurodegeneration with Brain Iron Accumulation and Lewy Body Disease with Mutations in a Novel Mitochondrial Membrane Protein** Hans Kretzschmar, Sigrun Roeber, Gabor Kovacs, Herbert Budka, Wolfgang Feiden, Howard Hurtig, John Trojanowski, Wouter Kamphorst, Monika Hartig, Thomas Meitinger, Holger Prokisch
3:45- 4:00	32	**The relationship between Mitochondrial Dysfunction and Alpha-Synuclein in a Yeast Model of Alpha-Synucleinopathies** Pavan Auluck, Susan Lindquist

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

4:30 – 5:30 pm ***DeArmond Lecture***
Emerging Therapies for Neurogenetic Diseases
Beverley L. Davidson, PhD
University of Iowa, Iowa City, IA

6:30 – 8:30 pm ***Annual Reception***
Metropolitan Ballroom

FRIDAY, JUNE 24, 2011
Metropolitan Pre-Function Area

Poster Session I:
(CME Credit is offered only during scheduled times that authors are present for discussion)

33	**Astroblastoma: A Rare Clinicopathological Entity** Derick Aranda, Andrew Parsa, Tarik Tihan, Soonmee Cha
34	**MIB-1 Labeling Index (MLI) Estimation Using Reference Images in Gliomas** Soma Karak, Xianyuan Song, Srinivas Mandavilli
35	**Dysembryoplastic Neuroepithelial Tumor-Useful Diagnostic Features and Importance of Radiographic Correlation** Knarik Arkun, William Gomes, Patrick LaSala, Ira Abbott, Christian Keller, James Goodrich, Karen Weidenheim
36	**Adult-Onset Angiocentric Glioma in the Mesial Temporal Lobe** Hajime Miyata, Masae Ryufuku, Taku Ochiai, Kaku Niimura, Tomokatsu Hori
37	**Cerebellar Low Grade Astrocytomas With Diffuse Growth Pattern** Cristiane Ida, Fausto Rodriguez, Jesse Voss, Brooke Mc Cann, Sarah Jenkins, Kevin Halling,Caterina Giannini
38	**Next Generation Sequencing of Oligodendroglioma – A Work in Progress** Stephen Yip, Yaron Butterfield, Olena Morozova, Michael Blough, Jennifer Chan, Alexandra Maslova, Suganthi Chittaranjan, J Greg Cairncross, Marco Marra
39	**Malignant Transformation of Clear Cell Ependymoma: A Case Report** Mark Smethurst, Alexis Demopoulos, Susan Morgello
40	**Anaplastic Astrocytomas with Abundant Rosenthal Fibers in Elderly Patients** Yasuo Sugita, Yousuke Okada, Yoshihiko Nakamura, Koichi Ohshima, Mizuhiko Terasaki
41	**Papillary Tumor of the Pineal Region: Ultrastructural Study of a Case** Matthew Cykowski, Eric Wartchow, Gary Mierau, Ethan Stolzenberg, Mary Kay Gumerlock, Kar-Ming Fung
42	**Stem Cell Marker Expression and Genetic Mutations in Glioblastoma: A Comparison of Primary and Recurrent Tumors** Christina Appin, Matthew Schniederjan, Erwin Van Meir, Gena Mastrogianakis, Daniel Brat
43	**Immunohistochemical Analysis of Actinin-4 in Malignant Gliomas** Shintaro Fukushima
44	**Supratentorial Extension of Diffuse Intrinsic Pontine Glioma (DIPG)** Terri Haddix, Michelle Monje, Marilyn Masek, Morgan Freret, Paul Fisher, Hannes Vogel
45	**Atypical Teratoid/Rhabdoid Tumor Arising in a Ganglioglioma** Bette Kleinschmidt-DeMasters, Diane Birks, Todd Hankinson, Marc Rosenblum
46	**Immunohistochemical Staining Profile of Seventeen Subependymal Giant Cell tumors** Dana Altenburger, Alexander Bottini, Spencer Tung, Harry Vinters
47	**The Role of Metabotropic Glutamate Receptor Allosteric Modulators in the Investigation and Treatment of Gliomas** Hilary Nickols, P Conn
48	**Clonal 8q Amplification Detected in a Multicentic Anapastic Astrocytoma by array CGH** Michelle Madden Felicella, Jill Hagenkord, Shera Kash, Martin Powers, Mitchel Berger, Arie Perry
49	**Anaplastic Oligodendroglioma in a Patient with Neurofibromatosis Type I** Ronnie Self, Liqiong Liu, John Gauchiani, Arthur Ulm, Zhenggang Xiong
50	**pStat3 Immunoreactivity in Glioblastomas and Atypical Teratoid/Rhabdoid Tumors** Jian-Qiang Lu, Beverly Wilson, Lothar Resch, Jeffrey Pugh, Vivek Mehta
51	**Mutant Isocitrate Dehydrogenase 1 Immunohistochemistry: Not Useful in the Diagnosis of Low Grade Diffuse Astrocytomas in NF1** Anat Stemmer-Rachamimov, Sandra Camelo-Piragua, Scott Plotkin

Poster Session I Continued:

(CME Credit is offered only during scheduled times that authors are present for discussion)

52	**Pathology of "Pseudoprogression" in a Phase II Study of PPX, TMZ, and RT for Newly Diagnosed High-Grade Gliomas** John Donahue, Suriya Jeyapalan, Marc Goldman, Heinrich Elinzano, Jerrold Boxerman, Thomas DiPetrillo, Howard Safran
53	**p53 Deletion Synergizes with Oncogenic K-ras in a Transgenic and Orthotopic Transplant Model of Giant Cell Glioblastoma** Ty Abel, Sabah Ghazi, Ping Li, Harold Moses, Michelle Stark
54	**Gliosarcoma Arising in an Oligodendroglioma (Oligosarcoma)** Annie Hiniker, Arie Perry
55	**Quantum Dot Probes for Glioma Cell Detection** Cody Weston, Achuthamangalam Madhankumar, Jennifer Baccon, Michael Glantz, James Connor
56	**Utility of Spectral Image Analysis For Prediction of 1p / 19q Deletion Status in Gliomas** Kimberly Stogner-Underwood, Colleen Jackson-Cook, Carleton Garrett, Hope Richard, Catherine Dumur, Christine Fuller
57	**Is Polysomy of Chromosome 1p and/or 19q in Oligodendrogliomas Prognostically Significant?** Andrea Wiens, Karen Johnson, Liang Cheng, Eyas Hattab
58	**Orbital Glioma with Focal Rhabdoid Morphology** Andrea Wiens, Richard Burgett, Mitesh Shah, Eyas Hattab, Jose Bonnin
59	**Determination of RB Mutation Status in Glioblastoma and Correlation with Data From The Cancer Genome Atlas** Patricia Goldhoff, Ivan Smirnov, C. James, Arie Perry, Joanna Phillips
60	**Ependymosarcoma: Light and Electron Microscopic Study of a Case** Kael Mikesell, Cathy Housman, Jonas Sheehan, Charles Specht
61	**Massive Gliosis of the Retina - A Müller Cell Proliferation or an Intraocular Ependymoma?** Dana Timek, Cathy Housman, Michael Wilkinson, Charles Specht
62	**Mitotic Counts and the Accuracy of Tumor Grading** Jeffrey Joseph
63	**Primary Diffuse Leptomeningeal Gliomatosis – a Case Study** Maria Martinez-Lage, Zissimos Mourelatos
64	**Characterization of R132H Mutant Form of Isocitrate Dehydrogenase 1 and Olig2 Expression Patterns in Glioblastoma Variants** Nancy Joseph, Michelle Madden, Joanna Phillips, Arie Perry
65	**Deep White Matter Venous Infarction in Infants: A Neuroradiologic-Neuropathologic Correlation** Christopher Pierson, Jerome Rusin
66	**Unusual Low-grade Neuroepithelial Neoplasm With Pseudopapillary Growth Pattern and Perivascular Collagen Deposition** Nguyen Vo, Eduardo Gonzalez-Toledo, Shashikant Patil, Jon Wilson
67	**Tissue Factor Expression in Glioblastoma does not Correlate with Clinical Development of Venous Thromboembolism** Carrie Mohila, Stephen Cook, Kaanchan Gangal, Dawit Aregawi, Nigel Key, M. Lopes, David Schiff
68	**Glioblastoma Occurring at the Site of a Previous Medulloblastoma Following a Five Year Remission Period** Sarah Martin, Daniel Brat, Annette Douglas-Akinwande, Eyas Hattab
69	**Locus Ceruleus Neurofibrillary Degeneration in Patients with High Braak Stage Pathology but Relative Cognitive Preservation** Maryam Pezhouh, Gang Chentree, Joyce Meints, Laura Hemmy, Michael Lee, Karen SantaCruz

FRIDAY, JUNE 24, 2011
Metropolitan Pre-Function Area

Poster Session I Continued:
(CME Credit is offered only during scheduled times that authors are present for discussion)

70	**Neurogenesis in Alzheimer's Disease Neocortex** Stefanie Marquez, Li Zhou, Celia Williams, Carol Miller
71	**Biochemical Analysis of ß-amyloid and Tau in Tangle-Only Dementia** John Crary, Ismael Santa-Maria Perez, Aya Haggiagi, Jean Paul Vonsattel, Michael Shelanski
72	**Distinctive Patterns of Tau and TDP-43 in a Former Professional Football Player and Marine as Compared to 3 Siblings** Ann McKee, Thomas Montine, Victor Alvarez, Aimee Schantz, Eileen Steinbart, Thomas Bird
73	***In vivo* and *In Silico* Evidence: Hippocampal Cholesterol Metabolism Decreases with Aging and Increases with Alzheimer Disease** Clyde Phelix, Sandra Siedlak, Xiongwei Zhu, George Perry
74	**Embryonic Stem Cell-Derived Choroid Plexus Epithelial Cells: A Potential Therapeutic for Alzheimer's Disease** Edwin Monuki, Momoko Watanabe, Lauren Davies, Chi-Yeh Chung, Jaymin Kathiriya
75	**FAM76B, a Novel PGRN Interacting Protein Screened by Yeast Two-Hybrid System** Qinwen Mao, Lihong Chai, Shunjun Wang, Xiaojing Zheng, Eileen Bigio, Haibin Xia
76	**Japanese Consortium for Research in Motor Neuron Disease and Frontotemporal Dementia** Shigeo Murayama, Yuko Saito, Masaki Takao, Hiroyuki Hatsuta, Jun Shimizu, Tameko Kihira, Yasumasa Kokubo, Haruhiko Akiyama, Kinuko Suzuki, Masato Hasegawa
77	**Neuroanatomical Distribution of Tau Pathology in a Case of Frontotemporal Dementia Associated with the MAPT P301L Mutation** Bernardino Ghetti, Jill Murrell, Jose Bonnin, Matthew Hagen, Alberto Espay, Michael Keys, Clarissa Rentz
78	**Diffuse TDP-43 Pathology in a case of Hereditary Spastic Paraparesis with a NIPA1/SPG6 Mutation** Maria Martinez-Lage, Laura Molina-Porcel, Dana Falcone, Leo McCluskey, Vivianna Van Deerlin, Virginia Lee, John Trojanowski
79	**Blastomycosis Presenting as a Pontine Mass Lesion** Cheryl Palmer, Paula Province, Hassan Fathallah-Shaykh, Barton Guthrie, James Hackney
80	**Aspergillus Granuloma – A Rare Pseudoneoplasm of the CNS** Maria Martinez-Lage, Sriram Venneti, Melandee Brown, James Schuster, Zissimos Mourelatos
81	**Quantitation of JC Virus in a Case of Progressive Multifocal Leukoencephalopathy (PML) Caused by Selective Immune Deficiency** Keith Wharton, Catherine Quigley, Marian Themeles, Jing Wei, Alex Buko, Carl Reid, Chao Sun, John Carulli, Susan Goelz, Susan Staugaitis, Robert Fox
82	**Chronic Granulomatous Herpes Encephalitis In A 10-Year-Old Boy With Clinically Intractable Epilepsy** Keith Harrison
83	**Progressive Multifocal Leukoencephalopathy Secondary to Natalizumab with Cortical Involvement and Negative CSF Evaluation** Andrea Wiens, Jose Bonnin, Scott Shapiro, Monica Mazda, Riley Snook
84	**Bacterial Abscess With Diffuse Sterile Leptomeningeal Calcific "Sugar-Coating" Xanthogranulomatous Hypophysitis Occurring in Association with Pituitary Adenoma: A Report of Two Cases** Nguyen Vo, James Traylor, Wilson Jon
85	**Xanthogranulomatous Hypophysitis Occurring in Association with Pituitary Adenoma: A Report of Two Cases** Cara Sedney, Fahad Bafakih, Charles Rosen, Kymberly Gyure

Poster Session I Continued:

(CME Credit is offered only during scheduled times that authors are present for discussion)

86	**Fulminant Demyelinating Process Presenting as Wernicke Encephalopathy in a Non-Alcoholic Patient** Murat Gokden, Dittana Phoncharoensri
87	**Intimal Thickening of Meningeal Arteries after Serial Corticectomies for Rasmussen's Encephalitis** Aaron Wagner, Kristina Biado, Madeline Schwarz, Alexander Bottini, Gary Mathern, Harry Vinters
88	**Radiologic-Pathologic-Proteomics Correlation. High Field 7T MRI and Multiplex Tissue Immunoblotting of Autopsy Brains** Kant Matsuda, Joon-Yong Chung, Kris Ylaya, Masaki Fukunaga, Tie-Qiang Li, Elisabeth Rushing, Stephen Hewitt
89	**Pituitary Blastoma: An Entity** Bernd Scheithauer, Eva Horvath, Cheri Deal, Yves Robitaille, Kalman Kovacs
90	**Collicular Hemorrhages; Metronidazole Toxicity or Wernicke's Encephalopathy ?** Chia-Ling Phuah, Hart Lidov
91	**Pathologic Findings In Sudden Unexpected Death In Epilepsy (SUDEP): A Ten Year Retrospective Review** Janet McNaughton, Jan Garavaglia, Gary Pearl
92	**Going Green in the Neuropathology Laboratory- A Review of Long-Term Room Temperature Biospecimen Storage** Leili Mirsadraei, Jerry Lou, Sergey Mareninov, William Yong
93	**What Constitutes Adequate Histologic Validation for Tumor Presence in a Frozen Glioblastoma Biospecimen?** Desiree Sanchez, Jason De Jesus, Andrew Kay, Bob Shafa, Negar Khanlou, Harry Vinters, William, Yong
94	**Ideal Warm Ischemia Times for Minimizing RNA Expression and Protein Phosphorylation Changes- a Literature Review** Bijan Ameri, William Yong
95	**Lyophilization as a Brain Tumor Preservation Method for Long-Term Storage at Ambient Temperature** Sergey Mareninov, Andrew Kay, Desiree Sanchez, Leili Mirsadraei, Jerry Lou, Bob Shafa, Isaac Yang, Tracie Pham, Aaron Wagner, Harry Vinters, William Yong
96	**ß-Amyloid Precursor Protein Immunoreactivity in Infant Brains: A Detailed Look at Pattern of Injury** Jocelyn Posthumus, Carrie Mohila, Virginia Richards, Deborah Kay, Maria Lopes

Platform 5: Muscle and Nerve
Chairpersons: Peter Pytel, MD and Christopher Pierson, MD, PhD

8:00 –8:15	97	**Expanding Histopathological and Clinical Spectrum of RYR1 Associated Myopathies** Mariarita Santi, Kristen Perkins, Amanda Kan, Ying Hu, Livija Medne, Diana Bharrucha, Perry Shieh, Carsten Bönnemann
8:15 –8:30	98	**Double Null Mice Lacking Dysferlin and Myoferlin Show a Muscular Dystrophy Phenotype** Peter Pytel, Alexis Demonbreun, Kieran Devaux, Manuel Alvarez, Elizabeth McNally
8:30 –8:45	99	**Amyloid Myopathy Associated with Dysferlinopathy: Comparison with Light Chain Amyloidosis** Syed Kazmi, Rodney McComb, Gokden Murat, Simone Spuler, J. Americo Fernandes Filho, Kent Huston, Betul Gundogdu, Steven Moore
8:45 –9:00	100	**Modeling the Human C205T Mutation in Murine Mtm1 Results in Exon 4 Skipping and a Less Severe Myotubular Myopathy Phenotype** Christopher Pierson, Ashley Dulin-Smith, Ashley Durban, Jordan Marshall, Andrew Snyder, Jordan Gladman, Dawn Chandler, Anna Buj-Bello, Michael Lawlor, James Dowling, Alan Beggs
9:00 –9:15	101	**Distribution and Extent of Weakness Is Dependent on Fiber Type and Mutation in Murine Models of Myotubularin Deficiency** Michael Lawlor, Marissa Viola, Jeffrey Widrick, Robert Grange, Anna Buj-Bello, Christopher Pierson, Alan Beggs
9:15 –9:30	102	**Immunohistochemistry for LC3 as a Diagnostic Marker for Drug-Induced Autophagic Myopathy** Han Lee, Brianne Daniels, Eduardo Salas, Andrew Bollen, Jayanta Debnath, Marta Margeta
9:30 –9:45	103	**Patterns of TDP-43 Immunoreactivity in Selected Inflammatory Myopathies** Jeremy Deisch, Dennis Burns
9:45 –10:00	104	**Clinical and Myopathological Manifestations of Filaminopathy in a Newly Identified Pedigree** Juan Bilbao, Lev Goldfarb, Beverley Young, Kester Kong, Sandra Cohen

10:00 - 10:30 am ***REFRESHMENT BREAK***

10:30 -11:30 am ***Saul Korey Lecture***
Protein Aggregate Myopathies
Hans H. Goebel, MD
Johannes Gutenberg-University Medical Center, Mainz, Germany

11:45 am – 12:45 pm ***Business Meeting II*** *(Metropolitan Ballroom)*
Presentation of awards for Meritorious Contributions to Neuropathology

12:45 – 2:00 pm ***Lunch***

SATURDAY, JUNE 25, 2011
Metropolitan Ballroom B
8:00 am – 2:00 pm

Platform 6: Tumors
Chairpersons: Alexander Judkins, MD and Cynthia Hawkins, MD, PhD

8:00 –8:15	105	**Molecular Subgroups In Medulloblastoma** David Ellison, James Dalton, Dan Brat, Arie Perry, William Yong, Mehmet Kocak
8:15 –8:30	106	**Prospective Comparison of FISH versus PCR-based Microsatellite LOH in the Evaluation of Gliomas for 1p/19q Codeletion** Craig Horbinski, Marina Nikiforova, Jonathan Hobbs, Kathy Cieply, Ronald Hamilton
8:30 –8:45	107	**Dissecting the Role of PTEN in Astrocytoma Invasion using Genetically-Engineered Mouse Models** Byron Huff, Ryan Bash, Natalie Karpinich, Ralf Schmid, C. Miller
8:45 –9:00	108	**Glioneuronal Tumor with Neuropil-Like Islands (GTNI) is Characterized by IDH1 R132H Mutations** Jason Huse, Khedoudja Nafa, Marc Ladanyi, Cyrus Hedvat, Marc Rosenblum
9:00 –9:15	109	**Identification of Driver Mutations Promoting Tumorigenesis in a Transgenic Mouse Model of Neurofibroma-MPNST Progression** Steven Carroll, Syed Kazmi, Stephanie Byer, Nicole Brossier, William Grizzle, Fady Mikhail
9:15 –9:30	110	**Dye-Enhanced Multimodal Confocal Imaging as a Novel Approach to Intraoperative Diagnosis of Brain Tumors** Matija Snuderl, Sameer Sheth, Dennis Wirth, Polina Ogas, Churl-Su Kwon, William Curry, Matthew Frosch, Anna Yaroslavsky
9:30 –9:45	111	**Malignant Rhabdoid Tumors Express Stem Cell Factors, which Relate with the Expression of EZH2 and Id Proteins** Sriram Venneti, Paul Le, Daniel Martinez, Sharon Xie, Lisa Sullivan, Lucy Rorke-Adams, Bruce Pawel, Alexander Judkins
9:45 –10:00	112	**Integrative Pathogenomic Analysis of Primary Human Glioblastoma Cell Line Library Reveals Novel Subclass Distinctions** Shakti Ramkissoon, Karl Olausson, Matthews Theisen, Justin Craig, Malika Hayashi, Sam Haidar, Cecile Maire, Keith Ligon

10:00 - 10:30 am ***REFRESHMENT BREAK***

10:30 -11:30 am ***Saul Korey Lecture***
Protein Aggregate Myopathies
Hans H. Goebel, MD
Johannes Gutenberg-University Medical Center, Mainz, Germany

11:45 am – 12:45 pm ***Business Meeting II*** *(Metropolitan Ballroom)*
Presentation of awards for Meritorious Contributions to Neuropathology

12:45 – 2:00 pm ***Lunch***

SATURDAY JUNE 25, 2011
Metropolitan Ballroom A
2:00 pm – 6:00 pm

Platform 7: Developmental/Pediatric
Chairpersons: Jean Michaud, MD and Edwin S. Monuki, MD, PhD

2:00 – 2:15	113	**The Application of Layer Specific Immunohistochemistry to Hemimegalencephay** Aaron Wagner, Nicole Yin, Alexander Bottini, Gary Mathern, Harry Vinters
2:15- 2:30	114	**GABAA Receptor Abnormalities in the Medullary Serotonergic System in the Sudden Infant Death Syndrome** Kevin Broadbelt, David Paterson, Richard Belliveau, Felicia Trachtenberg, Elizabeth Haas, Henry Krous, Hannah Kinney
2:30- 2:45	115	**Persistence Of Apoptotic Debris In Brain-Specific Rac1-Conditional Knockouts: Evidence For A Macroglial Clearance Function** James Mandell, Daniel Heffron, Jennifer Sokolowski
2:45- 3:00	116	**Mechanisms of Sensorineural Hearing Loss Related to Congenital Infection by the CMV in Human** Natacha Teissier, Anne-Lise Delezoide, Suonavy Khung-Savatovsky, Bettina Bessiere, Anne-Elizabeth Mas, Pierre Gressens, Thierry Van Den Abbeele, Homa Adle-Biassette
3:00- 3:15	117	**Mitochondrial Enzyme Histochemistry Demonstrates Hypermetabolic Neurons in Paediatric Epileptic Foci** Harvey Sarnat, Laura Flores-Sarnat, Walter Hader
3:15- 3:30	118	**GABAA Receptor Binding in the Cerebral Cortex in Periventricular Leukomalacia (PVL)** Gang Xu, Kevin Broadbelt, Robin Haynes, Rebecca Folkerth, Felicia Trachtenberg, Joseph Volpe, Hannah Kinney
3:30- 3:45	119	**Diffuse Astrocytomas of the Childhood Spinal Cord: Correlation between Incidence and Presence of a Neural Precursor Cell Type** Morgan Freret, Michelle Monje, Marilyn Masek, Paul Fisher, Hannes Vogel, Philip Beachy
3:45- 4:00	120	**Is Hemimegalencephaly a Fetal Tauopathy?** Harvey Sarnat, Laura Flores-Sarnat, Peter Crino, Walter Hader, Luis Bello-Espinosa

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

4:30 – 5:30 pm ***Bio/Brain Banking Session***
Metropolitan Ballroom A

8:00 – 11:00 pm ***Diagnostic Slide Session***
Metropolitan Ballroom

SATURDAY JUNE 25, 2011
Metropolitan Ballroom B
2:00 pm – 6:00 pm

Platform 8: Neurodegenerative - Other
Chairpersons: Robert E. Mrak, MD, PhD and Brent T. Harris, MD, PhD

2:00 – 2:15	121	**Treatment of Prion Disease in Brain Aggregate Cultures Predicts Results *in vivo*** Stephen DeArmond, Krystyna Bajsarowicz, Misol Ahn
2:15- 2:30	122	**Pathologic Examination of Hippocampal Sclerosis (N=106 Cases and 1,004 Controls) in the Aging Population** Janna Neltner, Frederick Schmitt, Yushun Lin, Erin Abner, Gregory Jicha, Ela Patel, Paula Thomason, Charles Smith, Karen Santacruz, Linda Van Eldik, Peter Nelson
2:30- 2:45	123	**Comparative Study: Tauopathy and TDP-43 Proteinopathy in Amyotrophic Lateral Sclerosis and Chronic Traumatic Encephalopathy** Kyung-Hwa Lee, Qinwen Mao, Nailah Siddique, Lisa Kinsley, Manjari Mishra, Katherine Gasho, Teepu Siddique, Eileen Bigio
2:45- 3:00	124	**The Spectrum of Neuropathology in Patients Dying with Amyotrophic Lateral Sclerosis (ALS) – A Clinicopathologic Study** Marla Gearing, Deborah Cooper, Nichole Costa, Jonathan Glass
3:00- 3:15	125	**Neuropathologic Findings in 32 Nondemented Elderly Subjects** Mark Jentoft, Joseph Parisi, Dennis Dickson, Kris Johnson, Bradley Boeve, David Knopman, Ronald Petersen
3:15- 3:30	126	**The Cerebellum in Friedreich's Ataxia** Arnulf Koeppen, Ashley Davis, Jennifer Morral
3:30- 3:45	127	**Loss of Layer Pre-Alpha Entorhinal Neurons in Huntington Disease and Controls: Age, Braak Stage, and Vonsattel Grade** John Hedreen
3:45- 4:00	128	**Nrf2 Signaling Modulates Neuronal NMDA Receptor Currents in a Glia-Dependent Manner** Junghyun Hahn, Xianhong Wang, Marta Margeta

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

4:30 – 5:30 pm ***Bio/Brain Banking Session***
Metropolitan Ballroom A

8:00 – 11:00 pm ***Diagnostic Slide Session***
Metropolitan Ballroom

Poster Session II:
(CME Credit is offered only during scheduled times that authors are present for discussion)

129	**Sensory Perineuritis: Reappraisal of the Entity** Negar Khanlou, Tracie Pham, Perry Shieh, M. Verity
130	**Nemaline Rods-like Structures Presented in Colchicine-Induced Myopathy: A Case Report and Literature Review** Jilin Bai, Mark Belsky, Daniel Menkes, Qian Wu
131	**Non-Diagnostic Muscle Biopsies in Late Onset Pompe's Disease** Ana Lia Taratuto, Alberto Dubrovsky, Jose Corderi
132	**Myopathy with Atypical Inclusions in a Child with Progressive Hypertonia, Scoliosis and Respiratory Failure** Benjamin Ellezam, Tim Lotze, Chester Brown, Adekunle Adesina
133	**Fatal Hydroxychloroquine-Induced Skeletal and Cardiac Myopathy** Bret Mobley, Morteza Azimian, James Atkinson
134	**Unusual Morphological Changes in Skeletal Muscle Biopsy of Children with Drop-head Syndrome due to LMNA Mutations** Fabiana Lubieniecki, Maria Soledad Monges, Maria Saccoliti, Susana Quijano-Roy, Pascale Richard, Norma Romero, Ana Lia Taratuto
135	**Responses to Treatment are Dependent on Fiber Type, Muscle, and Mutation in Murine Models of Myotubularin Deficiency** Michael Lawlor, Marissa Viola, Rachel Edelstein, Christopher Pierson, Anna Buj-Bello, Jennifer Lachey, Jasbir Seehra, Alan Beggs
136	**Lymphoid Follicles in a Gastrocnemius Biopsy** Derek Mathis, Jeffrey Elliott, Jack Raisanen, Kimmo Hatanpaa, Dennis Burns, Charles White, III, Emily Herndon
137	**Granulomatous Inflammation and Myasthenia Gravis - is there a Connection ?** Michael Kyle, Gordon Peterson, Bryan Tsao, Ravi Raghavan
138	**Juvenile Xanthogranulomas of the Nervous System: A Report of Two Cases and Review of the Literature** Jeremy Deisch, Rajankumar Patel, Korgun Koral, Sandy Cope-Yokoyama
139	**Silent Clivial Pituitary Adenoma** Knarik Arkun, Adam Sandler, Patrick Lasala, Yvonne Lui, Sophia Rodriguez, Christian Keller, Karen Weidenheim
140	**Solitary Fibrous Tumour of the Central Nervous System** Jane Cryan, Michael Jansen, Joanna Ti, Patrick Buckley, Irfan Yousef, Clare Faul, Francesca Brett, Ray Stallings, Michael Farrell
141	**Primary Burkitt Lymphoma of CNS: A Case Report and Literature Review** Fahad Alghamdi, Christopher Gillis, Brian Toyota, Bijal Patel, Manraj Heran, Stephen Yip, Adam Smith, Laurie Sehn, Kenneth Berean, Robert O'Connor, G. Moore
142	**Giant Cell Tumor of Bone with Intradural Extension** Fahad Bafakih, Vijayalakshmi Rajasekaran, Cara Sedney, Jeffery Hogg, Kymberly Gyure
143	**Myoepithelial Neoplasm Arising From Pleomorphic Adenoma of the Lacrimal Gland: A Case Report** Sarah Martin, Andrea Wiens, Richard Burgett, Eyas Hattab
144	**Primary Intracranial Leiomyosarcoma in a Young Adult with Hodgkin's Lymphoma** Hidehiro Takei, Benjamin Ellezam, Andreana Rivera
145	**Intraventricular Collision Tumor of Metastatic Lung Adenocarcinoma to Atypical Meningioma, a Case Report** Keyla Kleyser-Sugrue, Srinivas Mandavilli, Xianyuan Song
146	**Analysis of Invasive and Non-invasive Meningiomas** Sidney Croul, Fateme Salehi, Shahrzad Jalali, Kelly Burrell, Gelareh Zadeh
147	**Simultaneous Intrasellar Meningioma and Pituitary Adenoma Discovered at Autopsy: Case Report and Review of the Literature** Jennifer Ross, Alex John, Glenn Sandberg, Luis Sanchez

SATURDAY, JUNE 25, 2011
Metropolitan Pre-Function Area

Poster Session II Continued:
(CME Credit is offered only during scheduled times that authors are present for discussion)

148	**DNET-like Tumor of the Septum Pellucidum: Part of an Expanding Category of Rosette-Forming Glioneuronal Tumor** Matthew Schniederjan, Cristina Vincentelli, Daniel Brat
149	**Primary Intracranial Atypical Ewing's Sarcoma-Peripheral Primitive Neuroectodermal Tumor: A Case Report and Review Literature** Jantima Tanboon, Bunphot Sitthinamsuwan, Tewajetsada Paruang, Paul Thorner
150	**Diffuse Low Grade Leptomeningeal Glio-Neuronal Tumor not an Oligodendrogliomatosis: Clinic-Pathologic Features of 2 Cases** Sarah Alghamdi
151	**Giant Pituitary Adenomas: Pathologic-Radiographic Correlations and Lack of Role for p53 and MIB-1 Labeling** Bette Kleinschmidt-DeMasters, Helen Madsen, Thomas Borges, Aaron Knox, Katherine Michaelis, Mei Xu, Kevin Lillehei, Margaret Wierman
152	**The Role of the Inhibitor of Growth-4 (ING-4), a NF-KappaB Regulatory Molecule, in Astrocytoma's Cells Invasion** Georgios Klironomos, Spyridon Karadimas, Georgios Gatzounis, Eleni Papadaki
153	**Infantile Hemangiopericytomas in Children** Garth Warren, Sakir Gultekin
154	**Dura-Based High-Grade Large B-Cell Lymphoma without Systemic Involvement 4 Years after Diagnosis and Successful Therapy** Sverre Mörk, Ellen Berget, Lars Helgeland, Anne Lehmann, Olav Vintermyr
155	**Primary Leptomeningeal Sarcomatosis: Case Report With Immunohisotchemical Analysis** Janet McNaughton, Carlos Pardo, Gary, Pearl
156	**Primary Intracranial Dural Based Ewing's Sarcoma In An Elderly Woman -Case Report** Knarik Arkun, Patrick Lasala, Jack Farinhas, Christian Keller, Karen Weidenheim
157	**Granular Cell Tumor of the Trigeminal Nerve** Dimitri Trembath, Michael Solle, Adam Zanation
158	**Spinal Neurocytoma in a 6-year-old Boy Presenting with Hydrocephalus and Diffuse Leptomeningeal Dissemination** Andrea Wiens, Aaron Kamer, Joel Boaz, Jose Bonnin
159	**Diffusely Infiltrative Plexiform Schwannoma in Two Patients with Neurofibromatosis Type 2** Christian Davidson, Scott Plotkin, Anat Stemmer-Rachamimov
160	**Papillary Tumor of Pineal Region (PTPR): Clinico-pathological Findings and Follow-up in Three Pediatric Cases** Fabiana Lubieniecki, Silvana Sandrone, Daniel Alderete, Ana Lia Taratuto
161	**Central Nervous System Involvement of Inflammatory Myofibroblastic Tumor (IMT) in Two Children** Fabiana Lubieniecki, Silvana Sandrone, Jessica Lopez Marti, Dora Diaz, Javier Gonzalez Ramos, Maria Teresa Garcia de Davila
162	**Primary Central Nervous System Lymphoma in an Adolescent: A Rare Presentation of Acquired Immunodeficiency Syndrome (AIDS)** Juanita Evans, Robert Greiner, Mark Iantosca, Charles Specht
163	**Hybrid Schwannoma/Perineurioma of the VIII cranial nerve. Case Report** Facundo Las Heras, Anat Stemmer-Rachamimov
164	**Primary Intracerebral Histiocytic Sarcoma in a 53-year Old Woman** Cristina Vincentelli, Matthew Schniederjan, Stephen Hunter
165	**Embryonal Tumor with Abundant Neuropil and True Rosettes (ETANTR)** Laura Denham, Alexander Zouros, Arie Perry, Ravi Raghavan

Poster Session II Continued:
(CME Credit is offered only during scheduled times that authors are present for discussion)

166	**Two Cases of Atypical Teratoid Rhabdoid Tumor of the Spinal Cord** Ethan Stolzenberg, Matthew Cykowski, Ryan Rahhal, Timothy Mapstone, Mary Gumerlock, Kalliopi Petropoulou, Kar-Ming Fung
167	**Metastatic Hepatocellular Carcinoma to Brain and Skull** Yuan Shan, Jessa Kresak
168	**Angiofibromyxoma of the Falx Cerebri** Rolf Pfannl, Safain Mina, James Kryzanski
169	**Inflammatory Cerebral Amyloid Angiopathy in a Cognitively Intact Young Subject** Miguel Riudavets, Naomi Arakaki, Marcelo Schultz, Ana Lia Taratuto, Gustavo Sevlever
170	**Brain Biopsy of Susac's Syndrome: A Case Report** Howard Chang
171	**Joubert Syndrome: Neuropathological Update** Gordana Juric-Sekhar, Daniel Doherty, Robert Hevner
172	**Neuropathological Homology in True Galloway Mowat Syndrome** Julia Keith, Victoria Fabian, Peter Walsh, Adrian Charles, Yves Robitaille
173	**A Six-Week Old Male with Haddad Syndrome: Clinical, Genetic, and Pathological Evaluation** José Otero, Olivier Danhaive, Maria Cilio, Eric Huang, David Rowitch
174	**Asperger's Syndrome with Unusual Cerebral Pathology: Case Report and Literature Review** Liqiong Liu, Van Vo, Marcus Ware, Zhenggang Xiong
175	**Infantile Sialic Acid Storage Disease (ISSD): Neuropathology of two Inuit Patients with the novel c.526-2a>G Mutation** Jean Michaud, Matthew Lines, Michael Geraghty, Berivan Baskin, David Grynspan, Elizabeth Nizalik
176	**Spinal Muscular Atrophy, not Only a Motor Neuron Disorder: Phenotype-Genotype Correlation** Brian Harding, Sabrina Yum, Wendy Chung, Richard Finkel
177	**Atypical Posterior Reversible Encephalopathy with Underlying Multilobar Cortical Dysplasia in an Older Patient** Stephen Saikali, Martin Savard, Steve Jodoin, Myriam Pétrin, Peter Gould
178	**Neuropathology of Schinzel-Giedion Syndrome** Boleslaw Lach, Jeorge Arredondo, Jackie Bourgeois
179	**The Pathologic Findings Of An 18q22.2q23 Loss And Its Relationship To Other 18q Terminal Deletions** John Fortune, Jennifer Eschbacher, Stephen Coons
180	**Neuropathology of PEHO-Like Syndrome** Mircea Iftinca, Ross McLeod, Jeff Joseph
181	**Cystic Atypical Choroid Plexus Papilloma with a Markedly Elevated Mitotic Rate in an Infant** Jennifer Bennett, Arabinda Choudhary, Narasimha Jatavallabhula, Mark Dias, Jennifer Baccon
182	**The Neuropathology of Mucopolysaccharidosis Type VII (Sly Disease): A Case Report and Review of the Literature** Kelly Devers, Marie River-Zengotita, Charles Williams, Anthony Yachnis
183	**Septo-Optic Dysplasia with Auricular Duplication and Transposition of the Great Vessels: A Case Report** Mark Smethurst, Mary Fowkes
184	**Brain Abnormalities Associated with Glycogen Storage Disease, Type IV (Glycogen Branching Enzyme Deficiency), a Case Report.** Mary Fowkes

Poster Session II Continued:
(CME Credit is offered only during scheduled times that authors are present for discussion)

185	**Propionic Acidemia: Neuropathologic Features and Review of the Literature** Stephen Wilhem, Hannah Coulson, Suash Sharma, David Flannery, Amyn Rojiani
186	**Smurf2 Accumulates in TDP-43 Positive Cytoplasmic Inclusions in Spinal Cord but not in Hippocampus of Sporadic ALS** Masataka Nakamura, Satoshi Kaneko, Hidefumi Ito, Shinya Asayama, Kengo Fujita, Reika Wate, Hirofumi Kusaka
187	**Presence of Somatic Dendrites and Reduction of Apical Dendrites of Purkinje Cells in Spinocerebellar Ataxia Type 31** Masaki Takao, Bernardino Ghetti, Tatsuru Mihara, Kinya Ishikawa, Aya Tokumaru, Sayaka Funabe, Hiroaki Kimura, Ban Mihara, Masaki Fujita, Kinuko Suzuki, Shigeo Murayama
188	**Anti-Yo-Associated Paraneoplastic Cerebellar Degeneration without Associated Malignancy** Carrie Mohila, T. Bourne
189	**Atypical Protein Kinase C in Down Syndrome (DS): Preliminary Neuropathological Observations** Jianying Zeng, Charles Shao, Chandrakant Rao, Jenny Libien, Suzanne Mirra
190	**Endothelial-Reactive Antibodies in Susac's Syndrome** W. Waldman, Deborah Knight, Cynthia Magro, Martin Lubow, Robert Rennebohm, John Susac
191	**FTLD Pathology in Adult Polyglucosan Body Disease** Kyung-Hwa Lee, Qinwen Mao, Manjari Mishra, Katherine Gasho, Darren Gitelman, Charles White, Dennis Burns, Kimmo Hatanpaa, Hasan Akman, Salvatore DiMauro, Eileen Bigio
192	**Collagenous Fibrosis: A Rare Tissue Reaction of Deep Brain Stimulation (DBS)** Vinata Vedam-Mai, Michael Ullman, Michael Okun, Anthony Yachnis

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 Steven S. Scherer, MD, PhD join our list of distinguished speakers.

2008	Claudia Lucchinetti	The Spectrum of CNS Inflammatory 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

2011 PARISI LECTURE
Molecular Pathologies at the Nodes of Ranvier
Steven S. Scherer, MD, PhD

Steven S. Scherer is a Professor of Neurology at the University of Pennsylvania. He received his B.S. (1977), as well as his M.D. and Ph.D. (1985) from the University of Michigan. His Ph.D. advisor was Dr. Stephen S. Easter. He did an internship in internal medicine (1985-86), and a residency in neurology (1986-1989) at the Hospital of the University of Pennsylvania. He was a Charles A. Dana fellow at the University of Pennsylvania from 1989-1991, in the laboratory of Dr. John Kamholz. He joined the faculty in 1991, obtaining the rank of Professor in 2001, and served as Vice Chair for Research.

The diagnosis, treatment, and pathogenesis of peripheral neuropathies are the focus of Dr. Scherer's professional life, as documented in more than 110 original research papers and 50 reviews, chairing the first NIH conference on Peripheral Neuropathy, and serving on the Boards of the Peripheral Nerve Society and the Charcot-Marie-Tooth Association. He is a co-investigator in the Rare Diseases Clinical Research Network – Inherited Neuropathies Consortium.

Dr. Scherer, along with his colleagues and students, have contributed to our knowledge of how the "molecular architecture" of myelinated axons relates to their ability to conduct action potentials. The talk will review the molecules that form myelinated axons, highlighting examples of molecular components that play essential roles in the function of myelinated axons.

Abstract

Schwann cells and oligodendrocytes make the myelin sheaths of the PNS and CNS, respectively. Their myelin sheaths are structurally similar, consisting of multiple layers of specialized cell membrane that spiral around axons. Both kinds of myelin sheaths mainly consist of compact myelin, which is mostly composed of lipids as well as a few unique proteins. PNS myelin sheaths also have prominent regions of non-compact myelin, in which tight junctions, gap junctions, and adherens junctions join apposed layers of the myelin sheath. The nodal regions of the PNS and CNS are similarly organized. The nodal axolemma contains high concentrations of voltage-dependent Na^+ and K^+ channels that are linked to the spectrin cytoskeleton by ankyrinG. The paranodal membrane contains septate-like junctions, which are comprised of both glial and axonal components. The juxtaparanodal and internodal regions lack distinct morphological specializations, but the apposed axonal and glial membranes have distinct molecular components that are probably organized by *trans* molecular interactions.

Learning objectives

At the end of the Lecture attendees should be able to:

- Distinguish the molecular distinctions between "compact" myelin and the "non-compact" myelin that is found in incisures and paranodes.
- Distinguish the molecular distinctions of the axonal membrane at the nodes of Ranvier, paranodes, and juxtaparanodes.
- Cite that the nodal membrane contains voltage-gated Na^+ channels (mainly Nav1.6) as well as K^+ channels Kv7.2 and Kv7.3 channels; both kinds of channels are localized there by the cytoskeletal adaptor protein, ankyrinG.

The DeArmond Lecture

The 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 Beverley L. Davidson, 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

2011 DEARMOND LECTURE
Emerging Therapies for Neurogenetic Diseases
Beverley L. Davidson, PhD

Beverly L. Davidson holds the Roy J. Carver Biomedical Chair in Internal Medicine, and is Professor in Internal Medicine, Neurology, and Physiology & Biophysics. She is also Vice Chair for Research in the Department of Internal Medicine, Director of the Gene Transfer Vector Core and Associate Director for the Center for Gene Therapy. She currently manages a research team that includes research scientists, postdoctoral fellows, graduate students and undergraduates.

Dr. Davidson received her Bachelor of Science degree in Biology from Nebraska Wesleyan University and her Ph.D. in Biological Chemistry from the University of Michigan. After a postdoctoral fellowship she was Research Investigator and subsequently Assistant Professor at the University of Michigan. In 1994, she was recruited to the University of Iowa. Dr. Davidson is a member of several editorial boards, is a member of the American Association for the Advancement of Science, American Federation for Clinical Research (Midwest Section), American Society for Neuroscience, American Society for Gene Therapy, and the American Society for Microbiology. Dr. Davidson serves on the Board of Directors (Treasurer) for the American Society for Gene Therapy and is past Co-Director of the Iowa Biosciences Advantage Program.

Dr. Davidson's research is focused on inherited genetic diseases that cause central nervous system dysfunction, with a focus on (1) recessive, childhood onset neurodegenerative disease, in particular the lysosomal storage diseases such as the mucopolysaccharidoses and Battens disease; and (2) dominant genetic diseases for example, the CAG repeat disorders, Huntington's disease and spinocerebellar ataxia type I. Professor Davidson's research in inherited brain disorders and developing novel therapies is nationally and internationally recognized, and has been funded from the NIH since 1994. In 2007, she was named a Fellow by the American Association for the Advancement of Science, received a University of Iowa Regents Award for Faculty Excellence, and was named a University of Iowa Carver Research Program of Excellence. In 2008 she was an Iowa Women of Innovation Nominee for Research Innovation and Leadership. In 2009, Dr. Davidson received the Mathilde Solowey Award, National Institutes of Health, and was named a Member, Electorate Nominating Committee, Medical Sciences, AAAS (2009-12). In 2011 Dr. Davidson gave the Distinguished Alumni Lecture at the University of Michigan, in the Department of Biological Chemistry.

Abstract
This lecture will discuss inherited CNS disorders and present recent advances in therapeutic strategies. The late infantile neuronal liposfuscinoses are an example of a fatal, recessively inherited childhood onset disease due to a deficiency in the lysosomal enzyme tripeptidyl peptidase I (TPP1), a non-membrane bound lysosomal hydrolase. In culture, cells restored to 5-10% of normal enzyme levels are alleviated of their storage disease, indicating that enzyme replacement to the CNS need not be 100%. And because non-membrane bound hydrolases can be processed through the secretary pathway, genetic correction of a small percentage of cells can provide sufficient protein for enzymatic correction of many cells. I will present data demonstrating the utility of genetically correcting distinct cell types in the brain for disease therapy. Once corrected, these cells serve subsequently as enzyme secretion depots, providing for widespread correction of CNS phenotypes in TPP1 deficient models.

Huntington's disease (HD) is one of nine dominantly inherited, polyglutamine repeat expansion diseases, with only symptomatic therapies to date. Dominantly inherited disorders present the challenge of removing a toxic, gain of function product, in contrast to gene replacement. We recently exploited the RNA interference machinery to reduce expression of mutant huntingtin in mice models, with dramatic improvements in phenotypes. Recent data show that modifications to the inhibitory RNAs targeting huntingtin retain efficacy in rodents, and are safe in nonhuman primates using behavioral, histological and molecular readouts. These preclinical programs exemplify the challenges and progress in the development of gene therapy for brain diseases.

Learning objectives
At the end of the Lecture attendees should be able to:

- Compare the proteins responsible for the various subtypes of neuronal lipofuscinoses.
- Explain the biological properties of non-membrane bound hydrolases that permit an enzyme replacement approach to therapy in late infantile neuronal lipofuscinosis.
- Recall how RNA interference may be utilized as a therapy for Huntington's disease and similar trinucleotide repeat diseases caused by polyglutamine repeat expansion.

The Saul R. Korey Lectureship–a Brief History

The *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 Dr. Hans H. Goebel join our list of distinguished speakers.

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

Year	Lecturer	Title
2007	Dennis W. Dickson	Neuropathology and Genetics of Parkinsonism
2008	David N. Louis	Brain Tumor Classification: Little Steps and Big Jumps
2009	Stephen J. DeArmond	Mechanisms of Neurodegeneration in Prion Disease Orgininating from the Neuronal Plasma Membrane
2010	Peter C. Burger	A Long-Term Perspective on Pediatric CNS Tumors
2011	Hans H. Goebel	Protein Aggregate Myopathies

2011 SAUL R. KOREY LECTURE
Protein Aggregate Myopathies
Hans H. Goebel, MD

Hans H. (Hilmar) Goebel was born in Breslau/Silesia (now Wroclaw/Poland) and graduated from High School in l956. This High School diploma entitled him - as all the other graduates of German High Schools (Gymnasium) - to enroll in whichever subject at whichever German university without any entry exam or selective process. He studied medicine at the universities of Bonn, Heidelberg and (West)Berlin (the Free University), this kind of mobility among Geman medical students now being another defunct custom. After two years of rotating internship he took up residency in Anatomic Pathology at the Free University of (West)Berlin where, in the absence of any neuropathology training, he slowly drifted towards neuropathological aspects in that institution. When he became aware of the essentiality of formal training in neuropathology he left for New York City to start specialty training at Bellevue Hospital/New York University under the guidance of Drs. Irwin Feigin, Humberto Craviot, and Gleb Budzilovich. Two years later he moved to the Division of Neuropathology at Indiana University Medical Center in Indianapolis. There, Drs. Wolfgang Zeman and Jans Muller further trained him with special emphasis on lysosomal and neuromuscular disorders until he succeeded in passing the Board in Neuropathology. Back in Europe, he became a senior staff member at the Department of Neuropathology at the Georg August University of Goettingen. The final period of his official professional career he spent as Head of Neuropathology at the Johannes Gutenberg University in Mainz. After official retirement, he became an itinerant consultant in myopathology at various German neuromuscular labs and abroad. Since his times in the US he has been a member of the AANP.

Abstract
Protein Aggregate Myopathies (PAM) belong to the group of Protein Aggregate Disorders, sometimes termed Conformation Disorders, which also entail Protein Aggregate Encephalopathies (PAE) and Protein Aggregate Neuropathies. PAM may occur as hereditary or sporadic conditions, often appearing later in life. They are marked myopathologically by the formation and deposition of intracellular proteins within muscle fibers containing, in hereditary forms, the disease-specific mutant protein due to mutations in its respective gene and many other coaggregated proteins. Extralysosomally impaired degradation of the mutant proteins, perhaps, forming a seed to further aggregation of additional proteins, may also involve the ubiquitin-proteasome system and autophagy, the latter seen in muscle fibers as often present rimmed vacuoles. Inherited forms of PAM encompass myofibrillar myopathies, distal myopathies, hereditary inclusion body myopathies, and certain

congenital myopathies. Myofibrillar myopathies, the main group of PAM, are desminopathy, alpha B crystallinopathy, myotilinopathy, filaminopathy, ZASPopathy and BAG 3 myopathy, valosin-containing myopathy forming a nosological link between PAM and PAE because the abnormal proteins form aggregates in muscle fibers and nerve cells. Inclusion body myositis is a sporadic PAM. While these PAM are marked by impaired extralysosomal degradation of many proteins, thus suggesting a defect in catabolism, others, of earlier onset and with apparently mutant proteins aggregating only, such as actin filament aggregate myopathy due to mutations in the ACTA 1 gene and myosin storage myopathy due to mutations in the MYH 7 gene may represent anabolic forms of PAM where development, integration and maturation of defective proteins in myofibers are impaired. Whenever the structural integrity of sarcomeres and myofibrils may be implicated, such as in cores, target lesions, caps, reducing bodies and ragged red fibers, protein aggregation may occur, adding the respective conditions to the class of PAM. Sporadic protein aggregation within muscle fibers may also be induced by drugs, such as emetin, griseofulvin, and elinafide offering models to investigate the formation and degradation of proteins within muscle fibers on a non-hereditary basis. PAM are an emerging group of neuromuscular conditions increasingly appearing in the myopathological diagnostic practice and, hence, deserving our attention.

Learning objectives
At the end of the Lecture attendees should be able to:

- Explain the concept of Protein Aggregate Disorders
- Cite the different forms of Protein Aggregate Myopathies
- Discuss the neuropathological features of protein aggregation in muscle fibers
- Distinguish between catabolic and anabolic forms of Protein Aggregate Myopathies

Awards for Meritorious Contributions to Neuropathology

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

Awards for Meritorious Contributions to Neuropathology

2011 AWARD RECIPIENTS

William W. Schlaepfer, MD and Leroy R. Sharer, MD

William W. Schlaepfer, MD trained in pathology and neuropathology at the Yale Medical School where he conducted a doctoral thesis on Alzheimer type II astrocytes during experimental ammonium intoxication in rats under the tutelage of Elias E. Manuelidis. He completed his training at the Max-Planck-Institut fur Psychiatrie in Munich, Germany, a Mecca for classical neuropathology. While in Munich, he became interested in the ultrastructure of peripheral nerve and the nature of the axonal cytoskeleton. He continued his experimental studies on peripheral nerve as staff neuropathologist at Cornell and Washington University medical schools where he introduced teased fiber analyses of nerve biopsies for differentiating demyelinating and axonal neuropathies and for semi-quantitative assessment of Wallerian degeneration.

Key insight into the nature of Wallerian degeneration derived from observations that the granular disintegration of axonal cytoskeleton was simulated when frozen sections of rat peripheral nerve were placed in Ringer's solution, but not in isotonic saline. These observations led to a collaborative study with Richard Bunge in the Anatomy Department of Washington University showing that transected neurites of cultured dorsal root ganglia do not undergo Wallerian degeneration when calcium ions are removed from the media. Subsequent studies showed that granular disintegration of axonal cytoskeleton and fragmentation of myelin on teased fiber preparation could be prevented in transected and excised nerve when bathed in media lacking calcium ions.

In pursuit of the calcium story, Dr Schlaepfer utilized a Research Career Development Award to spend a year in the laboratory of Peter Baker in Cambridge, England. The neurophysiology labs at Cambridge were world-renown for Nobel laureate personages and their landmark discoveries of ion movements during nerve conduction in the giant axons of squid. Peter Baker was their leading authority on the movements of calcium. Dr Schlaepfer was, indeed, a small fish in a very big pond. Nevertheless, his work at the Marine Laboratories in Plymouth, England, was able to define high-affinity, low-capacity and low-affinity, high-capacity calcium ion binding systems in axoplasm.

Upon returning to the states, Dr Schlaepfer began structural, biochemical and immunochemical studies on isolated and purified neurofilaments that led to the identification of the neurofilament triplet proteins. The simultaneous loss of the triplet proteins during calcium-induced granular disintegration of neurofilaments provided reassurance to a highly controversial issue at the time. Subsequent studies identified and characterized the calcium-activated proteases that mediate neurofilament proteolysis and are triggered by excesses of calcium in the axoplasm.

His entry in the emerging world of molecular biology began as member of the Path A and, later, Neuro B NIH Study Sections, as he became exposed to novel grants studying DNA and RNA. At the time, he was fortunate that his RO1 grant was designated a Jacob Javits award, providing an extended period for boot-strapping his learning of new concepts and reorienting his studies in a molecular direction. He began probing trans-acting factors and cis-acting elements that regulate neurofilament gene transcription and, later, the stability of neurofilament RNA. The latter studies led to the discovery that expression of a small fragment of untranslated neurofilament RNA could reproduced selective degeneration of motor neurons with aggregates of cognate RNA-binding protein and alterations in NF expression in degenerating motor neurons of transgenic mice. This discovery was one of the earliest and most convincing demonstrations of the central role that RNA plays in motor

neuron degeneration. Provocative at that time, it is now a widely accepted concept that underlies current, exciting research efforts in understanding molecular mechanisms of neuronal degeneration. Furthermore Dr. Schlaepfer's studies underscored the complexity of selective neuronal degeneration in neurodegenerative disease, in support of the concepts of systems (or network) biology whereby neuronal homeostasis is maintained by complex but unique interactions of macromolecules that have evolved in and around different subsets of neurons and can be selectively disrupted by alterations of widely expressed gene products.

Along the way, Dr Schlaepfer ascended the academic ladder with his 1979 appointment to Professor of Pathology at the University of Pennsylvania, which was also the year that he joined Neuropathology at PENN. In 1987, he served as President of the American Association of Neuropathologists. While always attendant to his research laboratory, he shared fully in the teaching, training and service of academic neuropathology. He was rewarded by many close and fruitful interactions with scores of very talented residents and fellows that he trained over the years. These were very happy occasions when he could share his enthusiasm for pathology while exploring the meaning of change and the role of observable changes towards gaining insight into pathogenesis of disease.

He serves as a role model for many of us that aspire to follow in his steps: an almost 50-year record of continuous grant funding, running an active lab that spawned such major discoveries, and at the same time excelling in day-to-day surgical neuropathology and teaching the next generation of academic neuropathologists; a very tough act to follow indeed!

And now, even in the complex world of systems biology, he remains a strong advocate for the primacy of pathology for uncovering and dissecting the intricate interactions in tissues and findings key interactions that can modify disease.

Zissimos Mourelatos, MD

Leroy R. Sharer, MD, has been a member of the AANP for over 30 years. After graduating from Cornell University Medical School, he pursued training in Neuropathology with Robert Porro at Cornell and Philip Duffy and colleagues, including David Cowan and Abner Wolf, at Columbia. In 1981, he joined Dr. Eun-Sook (Lucia) Cho at New Jersey Medical School (NJMS), University of Medicine and Dentistry of New Jersey (UMDNJ), where he is Professor of Pathology and Director of Neuropathology. Dr Sharer's research has concentrated on the effects of HIV-1 on the Central Nervous System. He has been a continuously NIH funded investigator in this field for almost 25 years. Sixty two of his more than 90 papers and chapters relate to AIDS, particularly the associated encephalopathy in children. His papers from the mid 1980s on this subject have been cited almost 300 times each. His long-term collaborators in this field, Leon Epstein and David Volsky, speak highly of his attention to detail and his expertise as a neuropathologist. Dr Epstein noted that their collaboration was highly productive as their approaches to science were complementary although different; Leroy extremely methodical and much more cautious. Dr Volsky has relied on Leroy as his 'neuropathology *alter ego*' even with mice. Leroy, together with his mentor, colleague and friend for more than 40 years, Lucy Cho, have made major contributions not only to the teaching and clinical programs at NJMS and the pathology of HIV-1 but also to the field of multiple sclerosis.

In addition to his well-recognized contributions to the understanding of the CNS effects of HIV-1 and SIV-1, he has served our Association in many capacities, including Vice-President in 2005, the Awards Committee, Program Committee and Manager of the Diagnostic Slide Session (DSS) for 13 years. This is the role the majority of our members associate with him. He is the person who carefully composed the minutes (diagnoses and discussions) for the DSS sessions and dutifully collected the money for the slide boxes as well as offering diagnoses from the floor. Leroy reformed the financial affairs of the DSS and was meticulous in his record keeping. The Moderators of the DSS during his tenure, Tessa Hedley-Whyte and Anthony Yachnis, are immensely grateful for his efforts and guidance. Leroy is a member of the editorial board of the *Journal of Neuropathology and Experimental Neurology,* and continues as a founding editorial board member of the *Journal of Neurovirology.* Leroy is the sole elected president of The Neuroplex, the New York Society of Neuropathology, following his role in its incorporation in 2001. In this capacity he established the highly successful annual Marius Valsamis Lecture in 2005 in memory of our Association's archivist.

Leroy is a valued teacher receiving many high ratings from students and residents including the Excellence in Teaching Award from the NJMS. He has chaired many important faculty committees including the Institutional Review Board, the Faculty Affairs Committee and Faculty Committee on Appointments and Promotions. Dr Cho notes that he is a 'truly wonderful colleague' whose 'dedicated working habits and humble way' of interacting with people make him a desirable leader for tasks requiring diplomacy and tact.

Dr Sharer serves on the Board of Trustees of his church and the Board of Directors of the Momentum Project, an AIDS outreach/nutrition organization, and is currently Chair of their IRB. Leroy has a delightful dry sense of humor and enjoys travel, photography, music and song (chant).

For all of these reasons, Dr Leroy Sharer richly deserves the Award for Meritorious Contributions to Neuropathology of the American Association of Neuropathologists 2011.

AANP PRESIDENTIAL SYMPOSIUM
Sunday, 26 June 2011

Dystroglycan-Related Congenital Muscular Dystrophies

8:00 am – 8:05 am	Introduction and Overview of CMDs *Steven A. Moore, MD, PhD* *University of Iowa, Iowa City, IA*
8:05 am - 9:00 am	***Matthew T. Moore Distinguished Lecture*** Mechanistic and Molecular Insights into the Pathogenesis of Glycosylation-Deficient Muscular Dystrophy *Kevin P. Campbell, PhD* *University of Iowa, Iowa City, IA*
9:00 am – 9:45 am	Brain Development is Sugar-Coated: The Emerging Story of the Cobblestone Cortical Malformations *William B. Dobyns, MD* *University of Washington, Seattle, WA*
9:45 am – 10:30 am	*AANP AWARD PRESENTATIONS AND REFRESHMENT BREAK*
10:30 am - 11:15 am	Neuronal Migration Abnormalities in Dystroglycanopathies and Related Disorders *Robert F. Hevner, MD, PhD* *University of Washington, Seattle, WA*
11:15 am -12:00 pm	Peripheral Nerve Laminin and Dystroglycan – The Basis for Neuropathies in Congenital Muscular Dystrophies *Steven A. Moore, MD, PhD* *University of Iowa, Iowa City, IA*
12:00 pm	*INSTALLATION OF NEW OFFICERS AND ADJOURNMENT*

2011 PRESIDENTIAL SYMPOSIUM

Dystroglycan-Related Congenital Muscular Dystrophies

Steven A. Moore, MD, PhD
University of Iowa, Iowa City, IA

After graduating from Purdue University, Steve completed his M.D. and Ph.D. degrees at Indiana University School of Medicine. Both his anatomic pathology residency and neuropathology fellowship training were at the University of Iowa, where he joined the faculty in 1986. Over the first 10 to 15 years of his career, Steve participated in a general neuropathology practice and focused his research on bioactive lipids in the cerebral microcirculation. In the late 1990s he transitioned to a focus on muscular dystrophies and neuromuscular disease diagnostics. Currently, Steve is Co-Director of the Iowa Wellstone Muscular Dystrophy Cooperative Research Center where he has primary responsibility for a Muscle Tissue and Cell Culture Repository and carries out specialty diagnostic testing not available in clinical laboratories. He operates a muscle biopsy referral service and assists with the interpretation of molecular genetic testing for several of the more common muscular dystrophies. Steve serves on the editorial boards of the *Journal of Neuropathology and Experimental Neurology* and *Brain Pathology*. In collaborative basic science research with Dr. Kevin Campbell and others, Steve has studied the role of dystroglycan in brain development and the biology of peripheral nerve myelination.

Overview of Congenital Muscular Dystrophies

The congenital muscular dystrophies (CMDs) are a genetically and phenotypically heterogeneous group of rare neuromuscular and developmental diseases. Original descriptions of CMD began appearing in the literature with the publication of "Three cases of myopathy, infantile type" by Batten in 1903 and the description of a child who very likely had merosin-deficient CMD by Batten in 1909. Seminal papers describing the major forms of CMD include Ullrich 1930, Walker 1942, Fukuyama et al., 1960, Santavouri et al. 1977, Warburg 1978, Williams et al. 1984, Dobyns et al. 1985, and Tomé et al., 1994. Since the discovery and publication in 1995 that *LAMA2* mutations are responsible for merosin-deficient CMD, around fifteen additional CMD genes have been identified (see Table 1, next page). Although the clinical severity varies across a broad spectrum, most CMD patients are severely impaired by muscle weakness and joint contractures, with or without accompanying devastating brain or eye malformations. Life expectancy ranges from only a few months in Walker-Warburg syndrome (WWS) to a few decades in milder forms of Ullrich CMD (UCMD).

The explosion of genetic information that has occurred since 1995 has led to the precise diagnosis of many children with CMD, yet it has also opened a new era of complexity in recognizing and classifying CMD patients (see Table 1). This is because the clinical spectrum of disease associated with nearly every CMD gene has expanded – and continues to expand as genetic diagnostic testing becomes increasingly available and clinicians become more familiar with these diseases. Thus, close cooperation among clinicians, clinical laboratory physicians, and basic scientists is critical for efficient and accurate diagnosis.

Table 1. Congenital muscular dystrophies – classic scheme (allelic variations not included).

Classification	Locus	Gene	Protein	Clinical	Diagnostic testing	Primary refs
Extracellular Matrix Proteins						
Collagen VI disorders						
Ullrich CMD	21q22	*COL6A1*	collagen VI α1	prox. contractures, distal hyperextensibility; autosomal dominant and recessive forms	muscle biopsy[1], fibroblast cultures[2], sequencing DNA or cDNA	Pan 2003
	21q22	*COL6A2*	collagen VI α2			Camacho Vanegas 2001
	2q37	*COL6A3*	collagen VI α3			Demir 2002
Merosin Deficiency						
MDC1A	6q22-23	*LAMA2*	laminin 2	severe cong. hypotonia, contractures white matter MRI changes and peripheral neuropathy	muscle bx[1], sequencing DNA	Helbling-Leclerc 1995
MDC1B	1q42	unknown	unknown		linkage	Muntoni 1998
Glycosyltransferases and Glycosyltransferase-like Proteins						
Dystroglycanopathies (Secondary Dystroglcanopathies[3])						
MDC1C	19q13	*FKRP*	fukutin-related protein	CMD with mild CNS involvement	muscle biopsy[1], fibroblast cultures[4], sequencing DNA, POMT or POMGnT1 activity assays	Brockington 2001
MDC1D	22q12	*LARGE*	LARGE	CMD		Longman 2003
Walker-Warburg syndrome (WSS)	9q34	*POMT1*	POMT1	severe cong. hypotonia, hydrocephalus, type II lissencephaly, cerebellar hypoplasia, microphthamia		Beltran-Valero 2002
Walker-Warburg syndrome (WSS)	14q24	*POMT2*	POMT2			van Reeuwijk 2005
Muscle-eye-brain disease (MEB)	1p32	*POMGnT1*	POMGnT1	similar to, but milder than WWS		Yoshida 2001
Fukuyama CMD	9q31-33	*FCMD*	fukutin	similar to MEB		Kobayashi 1998
Other Established or Emerging Congenital Muscular Dystrophies[6]						
Integrin α7 deficiency	12q13	*INTA7*	integrin α7	congenital myopathy	sequencing DNA	Hayashi 1998
Selenoprotein N1 deficiency	1p36	*SEPN1*	selenoprotein N1	rigid spine syndrome	sequencing DNA	Moghadaszadeh 2001
Marinesco-Sjogren syndrome (MSS)	5q31	*SIL1*	nucleotide exchange factor SIL1	MSS - cerebellar ataxia with cerebellar atrophy, early-onset cataracts, mental retardation, hypotonia, and muscle weakness	sequencing DNA	Senderek 2005; Anttonen 2005
Laminopathy	1q21	*LMNA*	lamin A/C	CMD with features of Emery-Dreifuss muscular dystrophy (EDMD)	sequencing DNA	Mercuri 2004
Plectin – epidermolysis bullosa simplex (EBS)	8q24	*PLEC1*	plectin 1	EBS with muscular dystrophy	skin biopsy[5], sequencing DNA	Smith 1996

1 – Muscle biopsies are evaluated primarily by immunostaining of cryosections. Western blotting may provide additional diagnostic information.

2 – Fibroblast cultures (most often derived from skin) are used for collagen VI biosynthesis assays that involve immunostaining and western blotting. In addition, cultures are a source of mRNA for cDNA sequencing.

3 – The secondary dystroglycanopathies are particularly difficult to place into a single classification scheme. For each responsible gene there are many allelic clinical presentations. For simplicity, the table links genes to clinical diseases according to the seminal publications for each gene. An alternate scheme is to combine genotype and phenotype into a single classification, e.g., POMT1-WWS for Walker-Warburg syndrome caused by POMT1 mutations, and FKRP-WWS for Walker-Warburg syndrome caused by FKRP mutations.

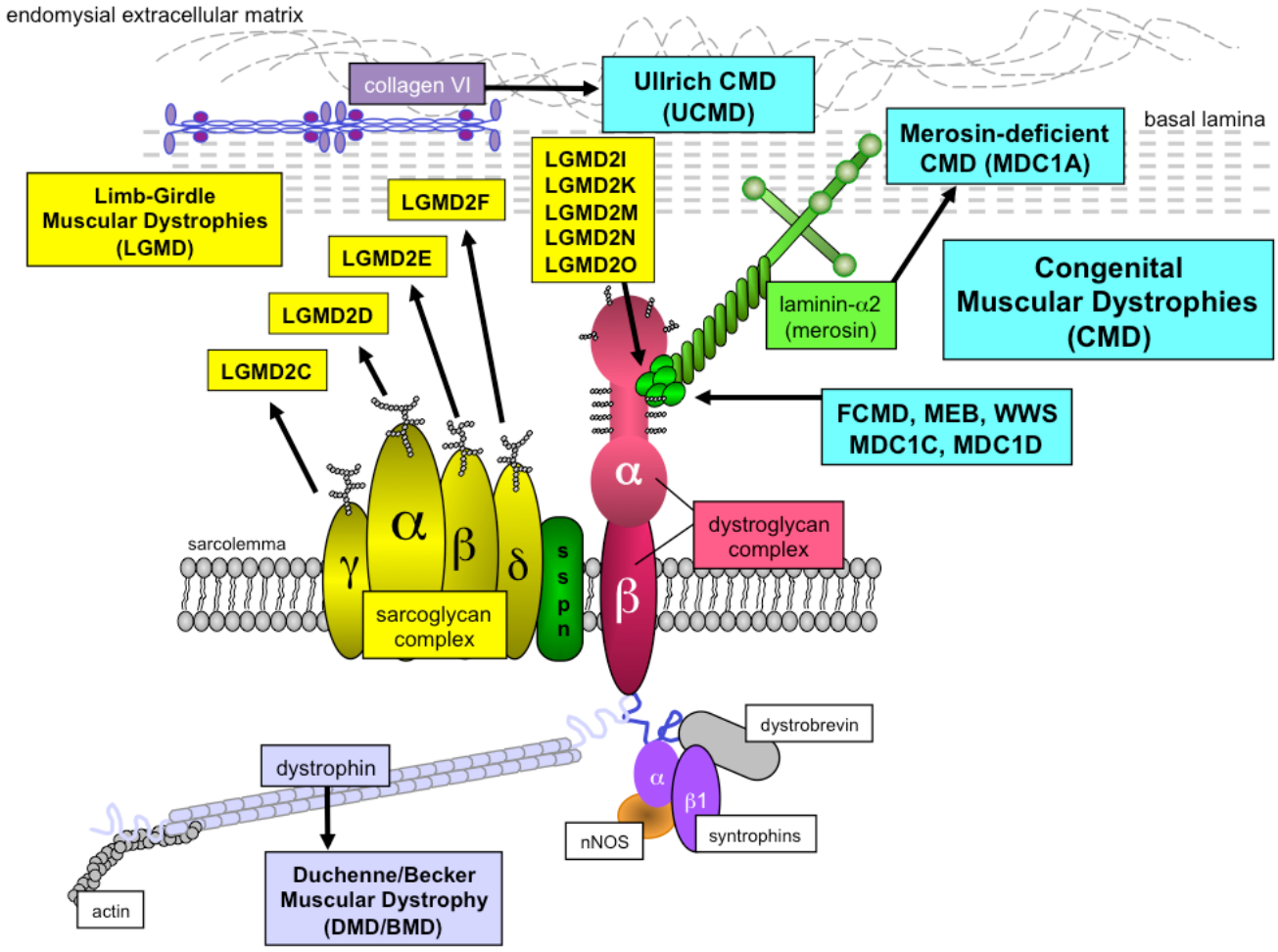
4 – Fibroblast cultures (most often derived from skin) may be used for glycosyltransferase assays such as POMT or POMGnT1 activity assays.

5 – Skin biopsies are evaluated by immunostaining of cryosections and by electron microscopy, which make it possible to classify the epidermolysis bullosa subtypes.

6 – Two additional CMD genes have been presented in the context of national meetings or workshops, but have not yet been formally published. Integrin α9 (INTA9) appears to be responsible for a French Canadian dystrophy clinically similar to the collagen VI disorders. The allelic variations due to mutations in the ryanodine receptor 1 gene (RYR1) appears to include CMD.

The CMD subtypes best characterized to date are due to deficiencies in merosin (MDC1A), abnormal glycosylation of alpha-dystroglycan (the secondary dystroglycanopathies – WWS, MEB, FCMD, MDC1C, and MDC1D), and abnormalities in collagen VI (UCMD). The gene mutations responsible for these dystrophies all lead to defects in either the skeletal muscle extracellular matrix (collagen VI and laminin 2) or the linkage of these (and other) extracellular matrix proteins to the sarcolemma. The latter category is represented by the CMDs in which alpha-dystroglycan is hypoglycosylated. Figure 1 illustrates the large number of muscular dystrophies that are associated with the dystrophin-glycoprotein complex, merosin (laminin 2), or collagen VI, and the relationships among these proteins. Allelic variants have been described for many of these dystrophies and the clinical phenotypes continue to expand. For example, UCMD and Bethlem myopathy are caused by mutations in any of the three collagen VI genes, with classic UCMD and Bethlem myopathy representing the severe and mild ends, respectively, of a spectrum of clinical severity (Lampe and Bushby 2004). Allelic variations are especially complex among the secondary dystroglycanopathies, where mutations in any of six glycosyltransferases or glycosyltransferase-like genes can result in clinical severity ranging from the WWS to the LGMD phenotype (Godfrey et al., 2007). A single LGMD patient with dystroglycan (DAG1) mutations was recently described (Hara et al., 2011).

Figure 1. ***Muscular dystrophies caused by abnormalities in proteins of the skeletal muscle dystrophin-glycoprotein complex, merosin (laminin 2), or collagen VI.*** *All of these proteins are important for linkage of the extracellular matrix to cytoskeletal and sarcomeric (contractile) proteins.*



Peripheral Nerve Laminin and Dystroglycan – The Basis for Neuropathies in Congenital Muscular Dystrophies

Merosin (laminin 2) and dystroglycan are also expressed in peripheral nerves where they comprise one of the major linkages between Schwann cells and their basement membranes. The dystroglycan-laminin linkage as well as integrin-laminin linkages have been demonstrated to be important for various aspects of peripheral nerve development, primarily radial sorting and myelination. The biology of these interactions has been extensively studied in mice using spontaneous mutant mice and several genetically engineered mouse models. The peripheral neuropathies that develop in these mice will be presented as the standard of comparison for similar neuropathies that almost undoubtedly occur in congenital muscular dystrophy patients, either MDC1A or one of the dystroglycanopathies. To date, neuropathies in these patients are either poorly characterized or unexplored. Future successful therapies for skeletal muscle disease are likely to expose neuropathic clinical phenotypes.

Learning objectives
At the end of the session, the learner should be able to:

- Enumerate the major subtypes of congenital muscular dystrophy.
- Describe the major phenotypes seen in patients with dystroglycanopathies.
- Identify potential mechanisms by which merosin or dystroglycan abnormalities may lead to peripheral neuropathy.

2011 PRESIDENTIAL SYMPOSIUM

Matthew T. Moore Distinguished Lecture

Mechanistic and Molecular Insights into the Pathogenesis of Glycosylation-Deficient Muscular Dystrophy

Kevin P. Campbell, PhD
Howard Hughes Medical Institute
University of Iowa, Iowa City, IA

Dr. Campbell is the Roy J. Carver Biomedical Research Chair in Molecular Physiology and Biophysics, Head of the Department of Molecular Physiology and Biophysics, and Director of the Senator Paul D. Wellstone Muscular Dystrophy Cooperative Research Center at The University of Iowa. Dr. Campbell also holds joint appointments as a professor in the Departments of Neurology and Internal Medicine. In 1989, he was appointed an investigator with the Howard Hughes Medical Institute (HHMI), and in 2009 he received his fourth five-year renewal with HHMI.

Dr. Campbell is internationally recognized for his fundamental contributions to muscular dystrophy research and developing strategies to treat muscular dystrophy. For the past 20 years, Dr Campbell and his colleagues have actively investigated the molecular pathogenesis of muscular dystrophy. Dr. Campbell's research is funded by a variety of organizations, and the scientific accomplishments of his laboratory have resulted in numerous publications in peer-reviewed journals. In addition to receiving many awards for his outstanding research, Dr. Campbell is an elected member of the Institute of Medicine, the National Academy of Sciences, and the American Academy of Arts and Sciences. In 2009 Campbell was the recipient of the March of Dimes Prize in Developmental Biology for his pioneering research on cell mechanisms involved in muscular dystrophy.

Dr. Campbell earned his Bachelor of Science degree in physics from Manhattan College, his master's degree from the University of Rochester School of Medicine and Dentistry, and his PhD in Biophysics from the Department of Radiation Biology and Biophysics at the University of Rochester. He completed his postdoctoral studies in the laboratory of Dr David MacLennan at the Banting and Best Department of Medical Research, University of Toronto, before moving to Iowa in 1981.

Abstract

Muscular dystrophies are genetic diseases characterized by weakness and progressive degeneration of skeletal muscle. The transmembrane protein dystroglycan, which is ultimately cleaved into an α and a β component, is a key link between the cytoskeleton and extracellular-matrix proteins that bear laminin globular domains (e.g., laminin, agrin, and neurexin). The mucin domain of α-dystroglycan is modified with numerous O-linked oligosaccharides that are essential for its normal function as an extracellular-matrix receptor in various tissues, including skeletal muscle and brain. Maturation of α-dystroglycan to its laminin-binding form requires a novel biosynthetic pathway involving phosphorylation on O-mannosyl glycans and that LARGE is crucial for further modification of phosphorylated O-mannosyl glycans on α-dystroglycan. Hypoglycosylation of α-dystroglycan and a consequent reduction of α-dystroglycan binding to extracellular matrix proteins are observed in patients with the Walker–Warburg syndrome, muscle–eye–brain disease, Fukuyama-type congenital muscular dystrophy, congenital muscular dystrophy types 1C and 1D, and limb-girdle muscular dystrophy 2I. To date, genes encoding six putative and known glycosyltransferases (POMT1, POMT2, POMGnT1, LARGE, FKTN, and FKRP) have been shown to be responsible for approximately 50% of cases of these diseases, which are collectively referred to as secondary dystroglycanopathies and which often feature brain abnormalities

as well as muscular dystrophy. Despite recent advances in our understanding of the molecular mechanisms underlying secondary dystroglycanopathies, it remains unclear whether dystroglycan is the only target of these enzymes or whether other substrates contribute to the pathogenesis of these diseases. We recently report a missense mutation in the dystroglycan-encoding gene, *DAG1,* in a patient with limb-girdle muscular dystrophy and cognitive impairment. Our studies revealed that this substitution interferes with LARGE-dependent maturation of phosphorylated O-mannosyl glycans on α-dystroglycan and leads to the disease-causing defect of α-dystroglycan binding to laminin.

Learning Objectives:
At the end of the lecture, the attendee should be able to:

- Describe the structure and function of dystroglycan in skeletal muscle and brain.
- Review genetics of various dystroglycanopathies.
- Explain the molecular pathogenesis of glycosylation-deficient muscular dystrophy.

2011 PRESIDENTIAL SYMPOSIUM

Brain Development Is Sugar-Coated: The Emerging Story of the Cobblestone Cortical Malformations

William B. Dobyns, MD
University of Washington, Seattle, WA

Dr. Dobyns is a physician-scientist who studies the nature and causes of human developmental disorders. While best known for studies of human brain malformations, his work has expanded to include autism, mental retardation, early childhood "developmental" epilepsy, and other complex developmental disorders that combine features of autism, mental retardation, epilepsy and more neurological deficits. He is trained and Board Certified in both Medical Genetics and Child Neurology, and leads a Developmental Disorders Research Consortium in Seattle.

Abstract

This talk will review the complete spectrum of cobblestone brain malformations both with and without congenital muscular dystrophy, relating them to both genes and pathogenesis.

Learning Objectives:
At the end of the lecture, the attendee should be able to:

- Recognize the spectrum of cortical and cerebellar malformations associated with cobblestone-type brain malformations.
- Describe the link between defective glycosylation, alpha-dystroglycan function, the pial basement membrane, and the brain malformation.
- Explain the differences between cobblestone malformations with and without muscular dystrophy.

2011 PRESIDENTIAL SYMPOSIUM
Neuronal Migration Abnormalities in Dystroglycanopathies and Related Disorders

Robert F. Hevner, MD, PhD
University of Washington, Seattle, WA

Dr. Robert F. Hevner is Professor of Neurological Surgery and Pathology at the University of Washington, Principal Investigator at Seattle Children's Research Institute, and Neuropathologist at Seattle Children's Hospital. Dr. Hevner's clinical neuropathology practice focuses on pediatric neuropathology. His research focuses on neurogenesis in the cerebral cortex and cerebellum, and the molecular basis of brain malformations. He has contributed over 80 original articles, reviews and chapters to the literature in this field. His laboratory pioneered the use of layer-specific markers as tools for studying malformations of cortical development, and elucidated the functions of several transcription factors in brain development and adult neurogenesis. Dr. Hevner has received a number of prestigious awards for his work, including the 1997 Weil Award for Neuropathology Research from AANP, and the 2005 Kurt Jellinger Prize in Neuropathology from Acta Neuropathologica. Dr. Hevner serves on the Executive Council of the AANP, and on editorial boards of several national and international journals, including Neuroscience, Journal of Neuroscience, Acta Neuropathologica, Brain Pathology, and JNEN. Dr. Hevner was chair of the 2009 Autism Research Program Review Panel sponsored by the Department of Defense, is a regular appointed member of the NIH Study Section on Neurodifferentiation, Plasticity, and Regeneration (NDPR), and has served on several other NIH review panels as well.

Abstract
Neuronal migration disorders are a category of cerebral malformations related by the abnormal positioning of neurons, including such entities as lissencephaly, polymicrogyria, and periventricular heterotopia. Progress in research on many fronts, from cortical neurogenesis to human genetics, has generated new insights and understanding of the pathogenesis and underlying features of such disorders. Progress has been particularly evident in the analysis of cerebral malformations caused by dystroglycanopathies, a group of disorders in which biological activity of the dystrophin-glycoprotein complex is deficient, typically leading to "cobblestone" malformations of the cortex and other brain structures, histologically related to defects or gaps in the glia limitans. Such defects permit neuronal "overmigration" past the glia limitans, and into or through the leptomeninges. Recent studies of dystroglycanopathies in humans and in rodent models have demonstrated the importance of interactions between radial glia and the basement membrane at the pial surface of the brain, and have furthermore revealed patterns of neuronal laminar disorganization, as well as axonal and dendritic structures in dystroglycanopathies. Immunohistochemical studies using layer-specific cortical neuron markers have also revealed the neuronal composition of polymicrogyria, classic lissencephaly, and periventricular heterotopia. Using the dystroglycanopathies as a focal point, this presentation will also compare defects among the neuronal migration disorders, and discuss evolving views of these disorders in light of the recent progress.

Learning Objectives
At the end of the lecture, the attendee should be able to:

- Compare the cerebral cortical malformations in dystroglycanopathies, polymicrogyria, classic lissencephaly, and periventricular heterotopia.
- Describe the role of the dystrophin-glycoprotein complex in brain development.
- Discuss how layer-specific markers have been used to understand malformations of cortical development.

Author Index