



CURRICULUM VITAE
NICHOLAS J. MARAGAKIS, M.D.

Current Appointments

Assistant Professor
Department of Neurology
Johns Hopkins University

Personal Data

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Education and Training

B.S.	1989	University of Utah, Biology
M.D.	1994	University of Utah
Medicine Internship	1994-95	University of Utah
Neurology Residency	1995-1998	Johns Hopkins University
Neuromuscular Fellowship	1998-1999	Johns Hopkins University

Professional Experience (in chronological order)

1999-Current Assistant Professor, Department of Neurology, Johns Hopkins University

RESEARCH ACTIVITIES

Publications

- Peer-reviewed original research articles

1. Coffeen CM, McKenna CE, Koeppen AH, Plaster NM, **Maragakis N**, Mihalopoulos J, Schwankhaus JD, Flanigan KM, Gregg RG, Ptacek LJ, Fu YH. Genetic localization of an autosomal dominant leukodystrophy mimicking chronic progressive multiple sclerosis to chromosome 5q31. *Hum Mol Genet* 2000 Mar 22;9(5):787-9

2. Irwin RP, **Maragakis NJ**, Rogawski MA, Purdy RH, Farb DH, and Paul SM. - Pregnenolone Sulfate Augments N-Methyl-D-Aspartate Receptor Mediated Increases In Intracellular Calcium In Cultured Rat Hippocampal Neurons. *Neuroscience Letters*, 141 (1992) 30-34

3. David S. Howland, Jian Liu, Yijin She, Beth Goad, **Nicholas J. Maragakis**, Benjamin Kim, Jamie Erickson, John Kulik, Lisa DeVito, George Psaltis, Louis J. DeGennaro, Don W. Cleveland, and Jeffrey D. Rothstein. -

Focal loss of the glutamate transporter EAAT2 in a transgenic rat model of SOD1 mutant-mediated amyotrophic lateral sclerosis (ALS). *PNAS* 2002. 99(3) 1604-09.

4. **Maragakis NJ**, Jackson M, Ganel R, Rothstein JD. - Topiramate protects against motor neuron degeneration in organotypic spinal cord cultures but not in G93A SOD1 transgenic mice. *Neurosci Lett.* 2003 Feb 27;338(2):107-10.

5. Kerr DA, Llado J, Shamlott MJ, **Maragakis NJ**, Irani DN, Crawford TO, Krishnan C, Dike S, Gearhart JD, Rothstein JD. Human embryonic germ cell derivatives facilitate motor recovery of rats with diffuse motor neuron injury. *J Neurosci.* 2003 Jun 15;23(12):5131-40.

6. Ueki T, Tanaka M, Yamashita K, Mikawa S, Qiu Z, **Maragakis NJ**, Hevner RF, Miura N, Sugimura H, Sato K. A novel secretory factor, Neurogenesis-1, provides neurogenic environmental cues for neural stem cells in the adult hippocampus. *J. Neurosci.* 2003 Dec 17;23(37):11732-40.

7. **Nicholas J. Maragakis**, Joerg Dietrich, Victor Wong, Haipeng Xue, Margot Mayer-Proschel, Mahendra S. Rao, Jeffrey D. Rothstein. - Glutamate transporter expression and function in human glial progenitors. *Glia.* 2004 Jan 15;45(2):133-43.

8. **Nicholas J. Maragakis**, Margaret Dykes-Hoberg, Jeffrey D. Rothstein. Altered Expression of the Naturally Occurring Alternatively Spliced Glutamate Transporter EAAT2b in Neurologic Disease. *Ann Neurol.* 2004 Apr;55(4):469-77.

9. Darman J, Backovic S, Dike S, **Maragakis NJ**, Krishnan C, Rothstein JD, Irani DN, Kerr DA.

Viral-induced spinal motor neuron death is non-cell-autonomous and involves glutamate excitotoxicity.

J Neurosci. 2004 Aug 25;24(34):7566-75.

10. Llado J, Haenggeli C, **Maragakis NJ**, Snyder EY, Rothstein JD. Neural stem cells protect against glutamate-induced excitotoxicity and promote survival of injured motor neurons through the secretion of neurotrophic factors. *Mol Cell Neurosci.* 2004 Nov;27(3):322-31.

11. **Nicholas J. Maragakis**, Mahendra S. Rao, Jeronia Llado, Victor Wong, Haipeng Xue, Andrea Pardo, Joseph Herring, Douglas Kerr, Carol Coccia, and Jeffrey D. Rothstein. Glial Restricted Precursors Protect Against Chronic Glutamate Neurotoxicity of Motor Neurons in Vitro. *Glia.* 2005 Apr 15;50(2):145-59.

12. Llado J, Haenggeli C, Pardo A, Wong V, Benson L, Coccia C, Rothstein JD, Shefner JM, **Maragakis NJ**. Degeneration of respiratory motor neurons in the SOD1 G93A transgenic rat model of ALS. *Neurobiol Dis.* 2005 Aug 3 (published online).

13. Raquelli Ganel, Tony Ho, **Nicholas J. Maragakis**, Mandy Jackson, Joseph P. Steiner, and Jeffrey D. Rothstein
Selective up-regulation of the glial Na⁺-dependent glutamate transporter GLT1 by a neuroimmunophilin ligand results in neuroprotection. *Neurobiol Dis.* (in press)

- Review articles

1. **N.J. Maragakis** and J.D. Rothstein. -Glutamate Transporters in Neurologic Disease. *Archives of Neurology, Vol. 58, March 2001*

2. Patel SA, **Maragakis NJ**. - Amyotrophic lateral sclerosis: pathogenesis, differential diagnoses, and potential interventions. *J Spinal Cord Med.* 2002 Winter;25(4):262-73.

3. **Nicholas J. Maragakis** and Jeffrey D. Rothstein. Glutamate Transporters: Animal Models to Neurologic disease. *Neurobiol Dis.* 2004 Apr;15(3):461-73.

- Book Chapters

1. **Maragakis, NJ** and Nestor Galvez-Jimenez. Epidemiology and pathogenesis of amyotrophic lateral sclerosis. In: *UpToDate, Rose, BD (Ed), UpToDate (in press), Wellesley, MA, 2005.*

2. **Nicholas J. Maragakis** and Richard Kimball. Amyotrophic Lateral Sclerosis. *Current Therapy in Neurological Diseases 7th edition (in press) eds. Johnson, Griffin, McArthur*

3. **Nicholas J. Maragakis** and Jeffrey D. Rothstein Amyotrophic Lateral Sclerosis (ALS): Idiopathic and Inherited.. *Neurobiology of Disease (in press) ed. Gilman.*

Extramural Sponsorship (current, pending, previous)

Previous

NIH-KO8 NS002131 The Glutamate transporter EAAT4 in Purkinje Cell biology.

PI : N. Maragakis 9/30/99-8/31/04

The purpose of this study is to evaluate the biology of EAAT4 using organotypic cerebellar cultures, and in vivo manipulations of EAAT4 expression to determine the normal role for EAAT4 in cerebellar synaptic biology and potential role for the protein in cerebellar degenerative disease.

MDA Stem cell transplantation in animal models of ALS

PI: N. Maragakis 1/01/01-12/30/03

This study was designed to use glial restricted precursors for transplantation in the mutant SOD1 mouse model of ALS. The focus was a cell line which overexpresses the glutamate transporter GLT1 and its potential to reduce glutamate neurotoxicity in the SOD1 mouse model.

Current

NIH-R01 Stem Cell Therapy for Motor Neuron Disease

P.I.: J.D. Rothstein, Collaborator: N.J. Maragakis 12/1/02-11/30/07

Effort 50%

The purpose of this study is to identify properties of stem cells, particularly astroglial precursors, which may make them attractive tools for transplantation. The study utilizes both in vitro analyses of motor neuron death through a glutamate mediated pathway as well as in vivo analysis following stem cell transplantation into the mutant SOD1 mouse.

ALS Association/Packard Center for ALS Research: Mutant SOD1-derived glial and neuronal restricted precursors as tools for understanding cell autonomy in ALS

PI: N.J. Maragakis 8/1/04-7/31/07 Effort 5%

The purpose of this study is to use stem cells to understand the relationship between astrocytes and neurons in the pathology of mutant SOD1 motor neuron disease.

Mass General Hospital: Safety and Dose Escalating Study of Oral Sodium Phenylbutyrate in Subjects with ALS

PI: N.J. Maragakis 1/1/05 – 12/31/05 Effort 2%

This is a phase I of this study is to establish the safety of the histone deacetylase inhibitor sodium phenylbutyrate in patients with ALS.

EDUCATIONAL ACTIVITIES

Teaching

- Classroom instruction (course title, dates, role)

Neuropathology Course (2nd year medical students), Jan 2003-current, Laboratory

Instructor

Neuropathology Course (2nd year medical students), 2003-current, Lecturer

“Neuromuscular Disorders: Diseases of the Motor Unit”

Neurology Clerkship (Bimonthly 1999-current), “Numbness and Weakness”

Clinical Neurophysiology Lecture (yearly), “Approach to Myopathies”

Clinical Neurophysiology Lecture (2002), “EMG findings in Myopathy”

Neurology Resident Lecture (2004), “ALS Diagnosis and Management”

Neurology Resident Lecture (yearly), “Inflammatory Myopathies”

Neurology Resident Lecture (yearly), “Cramps and Fasciculations”

Neuropathology Slide Reading (yearly), “Vacuolar Myopathies”

- Clinical instruction (service, dates, role)
General Neurology (Inpatient service, yearly, attending physician)
General Neurology (Consult Service, yearly, attending physician)
Neuromuscular Consult Service, yearly 1999-current, attending physician)

Mentoring (pre- and post-doctoral)

- Advisees
Victor Wong, M.S., 8/01-5/04, B.S., M.S. Biotechnology and M.S. Physiology., M.S. Biotechnology during time in my laboratory.
Richard Lee, 2003-current, Johns Hopkins University undergraduate student
Andrew Keller, 6/04-9/04, Davidson College undergraduate student
Andrea Pardo, M.D., 7/04-current, postdoctoral fellow
Takatoshi Ueki, M.D. 8/03-4/05, postdoctoral fellow
Angelo Lepore, Ph.D., 11/05-current, postdoctoral fellow

Editorial Activities

- Journal peer review activities Annals of Neurology, Journal of Neuroscience, Neurobiology of Disease, Neuron Glia Biology, Neuroscience

CLINICAL ACTIVITIES

Certification

- Medical, other state/government licensure
Physician and Surgeon, State of Maryland, Exp. Sept. 30, 2007, #D0053872
- Boards, other specialty certification
American Board of Psychiatry and Neurology (2000)

Service Responsibilities

- Neurology/Neuromuscular Clinic, Physician, 8 hours/month
- Electromyography and Nerve Conduction Studies, Physician, 16 hours/month

ORGANIZATIONAL ACTIVITIES

Institutional Administrative Appointments

- Johns Hopkins University Department of Neurology Residency Selection Committee(2005)
- Johns Hopkins University Department of Neurology Appointment and Promotions Committee (2005)
- Department of Neurosciences Implementation Team (2005)

Professional Societies

- Society for Neuroscience, Member
- American Academy of Neurology, Member
- Packard Center for ALS Research at Johns Hopkins(2000-current), Investigator
- ALS Research Group (2004-current), Member
- Northeast ALS Consortium (2004-current), Member

RECOGNITION

Awards, Honors

- Wintrobe Award (1994) The outstanding senior medical student at the University of Utah School of Medicine
- The Outstanding Intern in Internal Medicine Award at the University of Utah (1995)
- Jay Slotkin Award for Excellence in Research(1998) The Johns Hopkins University Department of Neurology

Invited Talks, Panels

1. "Ataxia Due to Loss of the Purkinje-Cell Specific Glutamate Transporter EAAT4"(1997), American Neurological Association Plenary Session)
2. Diagnostic and Management Issues in Neuromuscular Disease (Nov 2000), Osler Medical Service Conference
3. "Stem Cell Therapy for ALS", Dec. 14, 2000, Neurology Grand Rounds, Johns Hopkins University.
4. "Stem Cell Therapy for ALS", Apr 12, 2001, MDA ALS Support Group Meeting, Baltimore, MD
5. "Stem Cell Strategies in ALS", Dec. 5, 2001, Neurology Grand Rounds, Walter Reed Hospital, Washington, D.C.
6. "ALS Research Update", Apr 2002, MDA ALS Support Group Meeting, Baltimore, MD.
7. "Glial Restricted Precursor Transplantation in Models of Motor Neuron Disease", Oct 4, 2002, Department of Pharmacology, University of Montana
8. American Neurological Association Plenary Session-Acute and Chronic Neural Injury Alters Splicing of Functional Glutamate transporters (2003)
9. "Glial Restricted Precursor Transplantation in Models of Motor Neuron Disease", Sept 2003, Neurology Grand Rounds, Vanderbilt University.
10. "ALS: Current Management and Future Directions", Dec. 11, 2003, Medicine Grand Rounds, Franklin Square Hospital, Baltimore, Maryland.
11. "ALS: Current Management and Future Directions, Dec 11, 2003, MDA ALS Support Group Meeting, Baltimore, MD.
12. "Stem Cells for Motor Neuron Disease: Tools for Discovery and Therapeutics", Feb 27, 2004, Univ. California San Francisco, Department of Neurology.
13. "ALS Management", Aug 2004, MDA ALS Support Group Meeting, Baltimore, MD
14. "A Prospective Randomized Trial of Resistance and Endurance Exercise in ALS" (Sept 2004), Northeast ALS Consortium Meeting, Boston, MA.
15. "The Potential Role for Exercise in Slowing ALS Progression", Apr. 30, 2005. Intermountain MDA Meeting, Salt Lake City, Utah.
16. "Restless Legs Syndrome", May 18,05, Tumulty Rounds, Johns Hopkins University Department of Medicine
17. "Trial of Resistance and Endurance Exercise in ALS", Sept 2005, Northeast ALS Consortium Meeting, Boston MA.