Annual Highlights of the Resident & Fellow Section: 2017

A Representative Collection of Previously Published Articles

From the article: Mystery Case: Neurocutaneous melanosis with diffuse leptomeningeal malignant melanoma in an adult
Navigating Your Career: ALL ABOARD!

Experiential Learning Area

Looking for useful advice for every stage of your career development? One-on-one mentoring and small-group sessions and a lively presentation stage will offer something for everyone: medical students, residents, fellows, junior faculty, senior faculty, and advanced practice providers.

- Learn from successful neurologists and advanced practice providers on how to establish and maintain effective partnerships
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- Hone your skills in conflict resolution, giving and receiving feedback, and interviewing

Attend the Faculty and Trainee Reception and Meet the Neurology Resident & Fellow Section Editors and Editorial team

Monday, April 24 • 6:00 p.m.–9:00 p.m.
BCEC Level 3 Ballroom East/West

Experience a unique place for undergraduate and graduate attendees to network with peers, find information about residency programs, on pursuing fellowships and/or careers in neurology academics, research, or practice, and get recognized for their scholarships/awards.
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(A) Large congenital nevus on trunk extending over back and thighs. (B) Noncontrast CT brain shows hydrocephalus. (C) MRI brain axial T1-weighted image shows hyperintensity along sulcal spaces (arrows) and (D) postcontrast enhanced image shows diffuse meningeal enhancement along sulcal spaces (arrows). Sagittal noncontrast T1-weighted image of spine shows hyperintensity on the dorsal aspect of the cord (arrows in G). Following contrast administration, sagittal fat-suppressed T1-weighted spine MRI shows diffuse hyperintensity in the subarachnoid space suggestive of enhancement (H). (E) Papanicolaou and (F) May-Grünwald–stained CSF cytospin show malignant cells (arrows) with cytoplasmic melanin pigment, vesicular nuclei, and prominent macronucleoli (×400). Mitotic figures are seen (E, ×400). These malignant cells are immunopositive for HMB-45 (F, ×400).
Dr. Roy Strowd, Assistant Professor of Philosophy first at Cornell University and then at Princeton University, before graduating from Oxford University before coming to the United States to study philosophy. After attending the University of Heidelberg Medical School and University of Kentucky, Lexington. He has a strong interest in neurogenetics, particularly epilepsy genetics and is an avid blogger on Beyond www.epilepsygenetics.net

Ilena George, MD
Ilena George is a neurology resident at Yale University. Originally from New York City, she graduated from Stanford University with a degree in English Literature and completed her MD at New York University. Prior to graduating medical school, she spent two years at the NIH researching neuroimaging in multiple sclerosis. She plans to pursue neuroimmunology fellowship after residency.

Rachel Gottlieb-Smith, MD
Rachel Gottlieb-Smith is a child neurology resident at the Children’s Hospital of Philadelphia. She graduated from Harvard College with a concentration in Biochemical Sciences and a secondary concentration in Psychology. She received her MD from the Johns Hopkins University School of Medicine. She is passionate about medical education, particularly curricular development and medical education research and scholarship.

Ittai Bushlin, MD
Ittai Bushlin is a Child Neurology fellow at Oregon Health and Science University. He completed a BA in brain and cognitive sciences at the University of Rochester before spending two years researching mechanisms of synaptic vesicle trafficking at the NIH. He then attended Mount Sinai School of Medicine in New York City, where he obtained an MD and PhD, his research focused on changes in opioid and cannabinoid receptor signaling during neuropathic pain. He completed his pediatric training at Oregon Health and Science University. His current interests include pediatric epilepsy, pediatric stroke and neurogenetics. He will also complete a Clinical Pediatric Epilepsy fellowship at Boston Children’s Hospital starting in 2018.

Aravind Ganesh, MD
Aravind Ganesh is a senior neurology resident at the University of Calgary. He is currently pursuing a DPhil in clinical neurosciences on a Rhodes scholarship at Oxford’s Centre for Prevention of Stroke and Dementia. He is proficient in digital health technology, having cofounded SnapDr, a mHealth group that builds point-of-care tools to optimize guideline-based care and patient education.

Michael Fara, MD, PhD
Michael Fara is a neurology resident at New York University. He grew up in England, where he graduated from Oxford University before coming to the United States to study philosophy. After receiving his PhD from Princeton University, he was an Assistant Professor of Philosophy first at Cornell University and then at Princeton University, before discovering a new interest in medicine. He received his MD from Weill Cornell Medical College in New York City, and now plans to pursue a fellowship in vascular neurology.
Jen Vermillion, MD

Jen Vermillion is a child neurology resident at University of Rochester Medical Center in Rochester, NY. She obtained her bachelors degree in Molecular and Cellular Biology from Vanderbilt University in Nashville, TN. She obtained her MD from University of Rochester School of Medicine and Dentistry where she also did research on Battie’s Disease. After residency, she will be doing a fellowship in Experimental Therapeutics with a focus in Pediatric Movements Disorders at the University of Rochester.

Joseph Kleinman, MD

Jonathan T. Kleinman is a Neurocritical Care Fellow at the University of California, Los Angeles. He obtained his bachelors degree from The Johns Hopkins University in Cellular and Molecular Neuroscience. Prior to medical school, he performed research focused on the neural correlates of apnea and unilateral spatial neglect in acute stroke patients. He received his medical degree from Stanford University, where his research focused on advanced MR imaging of blood flow in transient ischemic attack, and multimodal MR imaging in hyperacute/ischemic stroke.

Khaled Moussawi, MD, PhD

Khaled Moussawi is currently a postdoctoral fellow at the NIH and a Behavioral Neurology and Neuropsychiatry fellow at Johns Hopkins Medicine. He attended the American University of Beirut in Lebanon for his undergraduate studies. He then completed his MD and PhD degrees at the Medical University of South Carolina and Adult Neurology residency at Partners Neurology (MGH and BWH). His research focused on advance MR imaging of blood flow in transient ischemic attack, and multimodal MR imaging in hyperacute/ischemic stroke hemorrhage.

Kristen Lindgren, MD, PhD

Kristen Lindgren is a third-year child neurology resident at Massachusetts General Hospital. She obtained her BA from Wesleyan University and MD/PhD from Boston University School of Medicine where she examined the neurobiological substrates of language in autism. Her clinical and research interests include autism and other related developmental disorders, neuroimaging, genetics, and medical education.

Pouya Khankhanian, MD

Pouya Khankhanian is an adult neurology resident at the University of Pennsylvania in Philadelphia, and a researcher at the Center for Neuroengineering and Therapeutics. He graduated with a degree in applied mathematics from the University of California at Berkeley before getting his medical degree at the University of California, San Francisco. His research interests include bio-statistics, genetics and genomics, and multiple projects under the umbrella of “big data research”, with focuses in Multiple Sclerosis and Epilepsy. My free time is spent either in the great outdoors, fostering pets, or home-brewing beers and wines.

James Siegler, MD

James Siegler is a vascular neurology research fellow and instructor at UPenn. He graduated from The Johns Hopkins University with degrees in neuroscience and history of medicine prior to completing his MD at Tulane. While a resident, he built and continues to produce a weekly educational audio podcast for neurologists and trainees called BrainWaves which can be found on iTunes (https://bit.ly/2cM33fZ). He has an associated blog at http://brainwaves.me/.

Sarah Flanagan Wesley , MD, PhD

Sarah Flanagan Wesley is a neuroimmunology fellow at Yale School of Medicine. She graduated from the Royal College of Surgeons in Ireland, serving as President of the RCS Society of Neuroscience, and completed neurology residency at Mount Sinai Beth Israel in New York where she was chief resident. She has contributed to other educational publications, including First Aid for the USMLE. Her research interests are in the areas of immune-mediated disorders of the central nervous system and transnational research in multiple sclerosis.

Steve O’Donnell, MD

Steve O’Donnell attended the University of Rochester where he studied political science and economics. Having no intention to go into medicine, he worked for four years in jobs that included fundraising for a presidential campaign, a non-profit public health organization, and an Emergency Medical Technician. It was the latter experience that pushed him towards medical school and an unexpected return to the University of Rochester where he obtained his MD. He is currently at the University of Utah and will finish his residency in adult neurology in 2017 and then will pursue a fellowship in vascular neurology at the University of Washington.

Andrew Sas, MD, PhD

Andrew Sas is the sports neurology clinical fellow and traumatic brain injury research fellow at the University of Michigan. He completed his BS in Biology at Dickinson College. He then attended the Medical University of South Carolina where he completed his MD and PhD studying neuroimmunology. He completed his neurology residency at the University of Michigan. His research interests include clinical care and translational research in the area neuroimmunology of traumatic brain injury and sports neurology.

Emer McGrath, MD, PhD, MRCP

Emer McGrath is an adult Neurology resident at Massachusetts General Hospital and Brigham and Women’s Hospital Partners Neurology program. She completed her MD and PhD at the National University of Ireland, Galway. She trained in clinical epidemiology and biostatistics during her PhD, focusing on the epidemiology of stroke and atrial fibrillation. Her research focuses on the clinical epidemiology of stroke, vascular disease and dementia. Her clinical interests include cognitive and vascular Neurology.

Michael White, MD

Michael White graduated from Lake Forest College in 2007 and received his MD from the University at Buffalo in 2012. He completed his medicine internship and neurology residency at Washington University in St. Louis and is currently a neuro-oncology fellow at the Massachusetts General Hospital and Dana-Farber Cancer Institute. His current research focuses on utilizing single-cell and cell-free DNA analysis to better understand the clonal evolution of CNS lymphoma.
The mission of the Resident & Fellow Section (RFS) is to keep our readers up to date on issues related to training and career considerations as well as support the development of lifelong learning skills. RFS is trainee-run by an editorial team of more than 20 neurology residents and fellows with the responsibility for reviewing, editing, and publishing articles. Residents are selected annually through a competitive process that attracts dozens of applicants and each will serve a three-year term. Past editorial team members have gone on to other important editorial activities, at Neurology® and elsewhere, and they have found the experience a formative part of their careers. Photographs and brief biographies of the current Resident & Fellow Section editorial team can be found in this Highlights booklet.

The history of the RFS began in 2004 when the Resident & Fellow “Page” was launched in Neurology as a forum for trainees and educators to publish articles on topics relevant to residency and fellowship. Initially lead by Robert “Berch” Griggs, then the editor-in-chief of Neurology, and Karen Johnston, associate editor, the article types included clinical reviews and education research projects, as well as commentaries on practice, ethics, teaching, history, and international training experiences. During its first decade, with the guidance of Mitch Elkind, the “Page” had grown to a “Section,” with articles appearing weekly, projects like Podcasts and the Writing Award, and a growing team of editorial members. Dr. John Millichap, a former editorial team member and the new RFS Section Editor, assumed leadership of the section from Dr. Elkind in 2015. He is joined by Deputy Section Editor Roy Strowd, another former editorial team member.

Over the years, the RFS has also introduced several subsections, which focus on (1) clinical neurologic education, such as Clinical Reasoning; Pearls & Oy-sters, Child Neurology, and Teaching NeurolImages (including both static images and videos); (2) graduate medical training, such as International Issues and Education Research and Initiatives; and (3) career issues, such as Emerging Subspecialties in Neurology. In addition, a Right Brain section includes creative writing, Mystery Cases engage readers in interactive discussion of critical aspects of clinical neurology, and Media and Book Reviews provide valuable analyses of textbooks, eBooks, Apps, and other resources for neurologists. Descriptions of these subsections appear before each sample article in this Highlights booklet.

Other unique projects developed during the past decade include podcasts (beginning in 2007), weekly E-Pearls (2008), an annual Writing Award (first given in 2009), our website (launched in 2010), the Journal Club (2011). Our ongoing Call for Authors program, in which trainees throughout the world have the opportunity to sign up to write articles on selected topics, was launched in January 2012. In 2012, we also began making all Teaching NeurolImages available as teaching slides. In 2014, we completed our first research project, initiated by editorial team members, to explore the role of mentored peer review of journal articles as a way of teaching evidence-based medicine and peer review skills to residents. The project, funded through an American Academy of Neurology Education Research Grant, involved residents at nine residency programs throughout the country, and the results were presented at the AAN and other national meetings. In 2015, Luca Bartolini, editorial team member of the RFS, developed his original idea for “Practice Current: An interactive exchange on controversial topics” in collaboration with the editors of Neurology® Clinical Practice (NCP). This is a brand new section of NCP that aims to identify and discuss difficult clinical scenarios and diseases with conflicting or insufficient evidence regarding diagnosis or treatment.

Over the past year, we had several successes; including a Clinical Reasoning book of previously published cases compiled to provide an educational resource for trainees and program directors and the launch of the RFS blog on the journal website. Another major accomplishment in 2015-2016 was the establishment a RFS mentor-mentee program designed to pair new RFS team members with recent graduates of the section. This pilot program is designed to guide new trainees in the review and editorial process, providing early mentoring on how to approach and conduct a peer review, and help create mentor teams with recently graduated team members. In its first year, the program was a great success with 10 mentor-mentee pairs and is now a standing component of the RFS team member training. In years to come, we hope that this program may serve as a structured model for bringing new, young peer reviewers into the process, even outside the RFS itself.

The accomplishments of the first decade of the RFS are accompanied by visions for the future. Plans are already in motion for several projects. To engage new residents and help bring them into the writing and publishing processes in the RFS, a RFS welcome program of resources and information about the section has been developed for residency programs. Other projects in development include “Journal Editing 101,” a formal three-year curriculum for the editorial team members and an update to the RFS website. Neurology recognizes that the future of the journal, and the future of the field of neurology itself, depends on the interest and commitment of its readers and writers. This journal is one of the most important records of our profession, and current trainees are the profession’s most valuable resource.

The RFS has been strongly supported by Neurology’s current Editor-in-Chief Robert A. Gross, Executive Editor Patty Baskin, editorial staff, the AAN, and the publishers Wolters Kluwer. In particular, staff members Kathy Pieper, Sandi Moriaty, and Robert Witherow have provided continual assistance and encouragement without which the section could not have survived.

We welcome submission of manuscripts for the Resident & Fellow Section, and author instructions can be found at Neurology.org. Papers submitted for this section will undergo the same thorough peer review process as all Neurology submissions, and it is anticipated they will reflect the same high level of quality. It is further expected that manuscripts published in the section will carry the same academic weight, whether on-line or in print, as papers published elsewhere in Neurology. We also continue to welcome input from our readers, including program directors and other educators, on features that will be most valuable.

Questions and comments should be addressed to John Millichap, Roy Strowd, or Kathy Pieper at kpieper@neurology.org. We hope you enjoy this year’s edition of the Highlights of the RFS!
Top 10 Ways for Program Directors to Use the *Neurology* Resident & Fellow Section

Aravind Ganesh, Khaled Moussawi, John J. Millichap, Roy E. Strowd

Visit the Resident & Fellow Section (RFS) website at Neurology.org/site/feature/index.xhtml to access the features below:

1. The “Clinical Reasoning” subsection presents clinical cases of significant educational value or originality. The goal is to systematically work through a patient case focusing on the patient presentation, exam, and diagnostic investigation. A question-answer format is utilized to provide real-time feedback, which helps to sharpen the resident’s clinical reasoning skills. This subsection can be the basis for an educational conference, a morning report, or can be a great starting point for incoming residents to hone their neurologic skills.

2. The “Mystery Case” subsection provides two or three multiple-choice questions inspired by a relevant teaching case published by the RFS. Residency programs are invited to incorporate these into their curricula and join in the friendly mystery case competition. Respondents can track their performance over time and compare their answers with others utilizing the new online format.

3. Each “Teaching NeuroImage” has a supplemental PowerPoint slide set available for download from the Neurology website. These can be used for group presentations or for a rapid review of illustrative or unique imaging findings. Similarly, each “e-Pearl” on the RFS website offers succinct learning points about a neurological presentation, which is ideal for rapid review.

4. “Journal Club” articles provide critical appraisals of recent articles published in Neurology with a focus on research methodology. The format is ideal for guiding discussions at Journal Club meetings.

5. The “Emerging Subspecialties in Neurology” subsection offers valuable new ideas and viewpoints for residents considering different career options. The RFS website provides a link to the AAN Fellowship Directory.

6. The “Media and Book Reviews” section evaluates written and electronic resources for residents, assisting them in finding the best resources to complement their training. In addition to traditional texts, the RFS reviews neurology apps and other electronic media. This is also a great way for residents to review new books or electronic media which they are provided free as a reviewer!

7. The “Right Brain” subsection allows residents to exercise their “write” brain by composing narratives, poems, or other expressions of their experiences as clinicians.

8. The “Education Research” subsection reports high-quality research on educational topics, including surveys of program directors and residents, as well as studies on educational interventions. Program directors and residents alike will enjoy the novel ways residents find to improve education in residency and beyond. These articles are also a helpful resource for rising chief residents who are exploring new approaches to resident education.

9. Many residents are interested in scholarly activities but may not know how to start. Program directors can help residents get involved by encouraging them to write for the RFS! Refer to the ‘Call for Authors’ page on the website for ideas to jump-start the writing process. All published articles are considered for the Annual Resident & Fellow Writing Award.

10. Follow the RFS on Facebook: Join our group entitled ‘American Academy of Neurology Residents and Fellows.’ For further digital access to RFS content, download the Neurology app onto your iPad/iPhone®, listen to the weekly Neurology podcast which includes the E-Pearl of the week, and follow Neurology Twitter for updates. Help spread the word!
Announcement

Neurology Resident & Fellow Section Writing Award

The winners of the 2017 Award are:

Elan L. Guterman, MD, Brian Yurgionas, MD, MS, and Alexandra B. Nelson, MD, PhD.
For their article: Pearls & Oy-sters: Episodic ataxia type 2: Case report and review of the literature

See page 43 of this Highlights book.

The Neurology Resident & Fellow Section Writing Award is intended to recognize the extraordinary writing abilities of those currently in training in neurology. Eligible manuscripts will include any submitted to and published in the Neurology Resident & Fellow Section, whether online or in print. Submissions on any topic of interest to trainees and in any subcategory of the section will be eligible. The main criteria for selection will be educational value, novelty, depth of exposition, and clarity of writing. At least one author of the manuscript must be currently in a neurology residency program or in fellowship training in one of the neurological subspecialties. All authors will be considered equal recipients of the award in order to recognize and encourage collaborative work among trainees. The next award will be announced in early 2018 and will be awarded for a paper published in 2017.

No formal application process is required. All manuscripts submitted to the section will be considered. Manuscripts should be submitted online at Neurology.org. Please direct any questions to kpieper@neurology.org.

PAST RECIPIENTS

2016 Award Winner
Emerging Subspecialties in Neurology: Telestroke and teleneurology
Sunil A. Mutgi, MD; Alicia M. Zha, MD; and Reza Behrouz DO
Neurology June 2, 2015, 84:22 e191-e193

2015 Award Winner
Clinical Reasoning: An unusual cause of transverse myelitis?
Pavan Bhargava, MD, and Rodger J. Elble, MD, PhD
Neurology February 11, 2014, 82: e46-e50

2014 Award Winner
Right Brain: A reading specialist with alexia without agraphia: Teacher interrupted
Jason Cuomo, MA; Murray Flaster, MD, PhD; and José Biller, MD
Neurology January 7, 2014, 82:e5-e7

2013 Award Winner
Clinical Reasoning: A 55-year-old woman with vertigo: A dizzying conundrum
Daniel R. Gold and Stephen G. Reich
October 23, 2012, 79:e146-e152

2012 Award Winner
Child Neurology: Brachial plexus birth injury: What every neurologist needs to know
Christina B. Pham, Johannes R. Kratz, Angie C. Jelin, and Amy Gelfand
Neurology August 16, 2011, 77:695-697

2011 Award Winner
Right Brain: We were all once ‘fixed and dilated’
Amy Gelfand, MD
November 16, 2010, 75: 1851-1852
June 14, 2016: Cruciate Paralysis

First described by Bell in 1970, cruciate paralysis denotes selective bilateral upper extremity paralysis, without involvement of the lower extremities. The neuroanatomical basis is the selective involvement of the corticospinal fibers of the upper extremity decussating in the rostral pyramids, at the cervicomedullary junction. The corticospinal fibers of the lower extremities decussate more caudally, which explains the relative sparing of the lower extremities. The lower cranial nerves may be involved, causing respiratory insufficiency, which helps differentiate cruciate paralysis from central cord syndrome. Reported etiologies include trauma, compressive tumors, congenital malformations, vascular, demyelinating and metabolic etiologies (eg diabetes). The prognosis depends on the cause with better outcome seen following early surgical decompression for compressive lesions.

References

Submitted by Sunil Munakomi, MD, Senior Resident, Department of Neurosurgery, College of Medical Sciences—Kathmandu University, Nepal.
Dr. Munakomi reports no disclosures.

November 28, 2016: Reversible Lesion of the Corpus Callosum

The boomerang sign, also known as mild encephalitis/encephalopathy with a reversible splenial (MERS) lesion, or reversible splenial lesion syndrome (RESLES), is a finding of isolated diffusion restriction without contrast enhancement in the central portion of the splenium. It's a non-specific finding described in seizures, antiepileptic withdrawal, neuroleptic malignant syndrome, hemicrania continua, high altitude cerebral edema, herbicide toxicity, metabolic derangements (i.e. hyponatremia, hypoglycemia) and certain infections. Most patients are children and young adults. Symptoms are nonspecific without a clear disconnection syndrome. Radiographic findings resolve within one month.

References

Submitted by Asya Izraelit Wallach, MD, Neurology Resident, New York University School of Medicine, New York, NY.
Dr. Wallach reports no disclosures.
Child Neurology

The Child Neurology section in the Resident & Fellow Section of Neurology focuses on contemporary educational issues in child neurology. The goal of the section is to provide up-to-date reviews on important topics in child neurology that are relevant to all neurologists, both adult and child, particularly those still in their training. Examples include management of acute stroke in children, childhood demyelinating disease, neuroimaging in metabolic disorders, and the neurobiology of autism. Each piece will begin with a patient case, followed by a brief discussion about the differential diagnosis and a detailed discussion about the topic of focus. Submissions are welcome from residents and fellows in either child or adult neurology. Ideally, submissions will include the patient case as well as the discussion, but submission of timely review articles without an accompanying case will also be considered. In this situation, the editors of this section may supply an appropriate patient case.
Child Neurology:  
Two sisters with dystonia and regression

PLA2G6-associated neurodegeneration

CLINICAL CASE, PART 1 A 19-month-old girl presented for neurologic consultation for delayed walking. She rolled at 6 months, sat unsupported at 8 months, but never walked independently. She babbled only. Her examination was notable for slightly decreased bulk in her legs, mild truncal hypotonia, and decreased deep tendon reflexes. She had difficulty pulling to stand and could only walk with support. She exhibited a steppage gait with hyperextension of her knees, exaggerated lifting of her feet, and out-turning of her ankles. Birth history was unremarkable and parents were not consanguineous. A head CT performed at 13 months for mild head trauma was normal.

The initial diagnostic workup was directed at causes of gait abnormality and developmental delay. Initial metabolic screening labs, including serum lactate/pyruvate, amino acids, creatine phosphokinase, carnitine, lipid panel, and coenzyme Q10 profile, were normal. MRI of the spine was normal. At age 23 months, brain MRI showed new mild to moderate cerebellar atrophy and minimal brainstem volume loss (figure).

By age 26 months, the patient developed pain and dystonia in her legs. On examination, she had striatal toes (spontaneous extensor plantar response without fanning of the toes) and continued decreased deep tendon reflexes. She could no longer stand, although she still crawled. Repeat brain MRI at 29 months showed progressive cerebellar volume loss.

By age 31 months, she developed a mild spastic quadripareis and continued to have significant painful dystonic posturing of lower extremities. Developmental regression continued. She was no longer able to sit independently or crawl and only made occasional sounds. She developed bilateral optic atrophy and intermittent left esotropia.

Differential diagnosis. Dystonia is a movement disorder characterized by sustained or intermittent muscle contractions causing abnormal, often repetitive, movements, postures, or both.1 The differential diagnosis for dystonia in children is broad (table e-1 on the Neurology® Web site at Neurology.org) but can be narrowed by presence of other neurologic manifestations. Dystonia associated with cerebellar atrophy and developmental delay or regression is concerning for an inherited neurodegenerative process. Etiologies to consider include neurodegeneration with brain iron accumulation (NBIA), neuronal ceroid lipofuscinosis, pontocerebellar hypoplasia, Rett syndrome, Wilson disease, and other mitochondrial diseases.

CLINICAL CASE, PART 2 Genetic testing results included normal karyotype and interstitial duplication of 129 kb of DNA at 17q21.31, likely benign, on chromosomal microarray. The Cerebellar/Pontocerebellar Hypoplasia Sequencing Panel (University of Chicago Genetic Services Laboratory, 2013) detected no pathogenic variants. Copper and ceruloplasmin were normal. Mitochondrial DNA (mtDNA) Deletion/Duplication and mtDNA Common Mutation Panels (Cincinnati Children’s Molecular Genetics Laboratory, 2014) showed no abnormalities.

Continued hyporeflexia and pain, and new intermittent episodes of leg flushing, raised concern for peripheral neuropathy. This, in conjunction with regression, dystonia, and MRI findings, prompted sending the NBIA Sequencing Panel (University of Chicago Genetic Services Laboratory, 2014) at age 33 months. The panel showed 2 mutations in the PLA2G6 gene (PLA2G6 c.1674del and PLA2G6 c.2370T>G), both pathogenic variants previously described in PLA2G6-related disorders.2,3 Based on the phenotype (clinical history, neurologic examination, and neuroimaging), the patient was diagnosed with the infantile neuroaxonal dystrophy (INAD) subtype of PLA2G6-associated neurodegeneration (PLAN). At the time of diagnosis the patient had 2 younger siblings, one of whom was displaying signs of developmental regression. Given the patient’s family history, her classic presentation, and the identification of 2 previously reported pathogenic variants, parental genetic testing was not thought necessary. Genetic counseling was provided.

The patient was prescribed docosahexaenoic acid (DHA) 250 mg per day. During the 18 months following diagnosis, she continued to have dystonia,
tight heel cords, and lower extremity pain, which were treated symptomatically with baclofen, trihexyphenidyl, gabapentin, diazepam, and tramadol. After video swallow study showed silent aspiration, a gastrostomy tube was placed. She developed epilepsy with occasional generalized tonic-clonic seizures. Sleep study showed brief central and obstructive sleep apnea, and she was placed on overnight oxygen. Her last brain MRI, performed at age 33 months, showed stable volume loss in the cerebellum and brainstem, subtle nonspecific hyperintense T2/fluid-attenuated inversion recovery signal in the supratentorial white matter, but no radiographic evidence of brain iron accumulation (figure). Palliative services were engaged, and the patient expired at age 4 years from respiratory causes.

The patient’s younger sister developed motor delays by age 1 year. By age 2, the younger sister could not walk independently, displayed a steppage gait when supported, and spoke only 2 recognizable words. Over the next few months, she regressed in motor and language skills and developed dystonia in her lower extremities. A brain MRI showed cerebellar volume loss similar to her sister’s. Given the similar phenotype, she was given a presumptive diagnosis of PLAN.

**DISCUSSION** PLAN, also known as NBIA2, is a rare autosomal recessive disorder first reported by Seitelberger in 1952 and later clinically defined by Aicardi and Castelein in 1979. The incidence of PLAN is unknown. It falls in the broader disease category of NBIA. NBIA is a heterogeneous group of rare diseases characterized by progressive extrapyramidal symptoms, intellectual impairment, and excessive iron deposition in the brain, especially the globus pallidus. The 2 most common NBIA diseases are pantothenate kinase–associated neurodegeneration (PKAN, also known as NBIA1), which accounts for approximately 50% of all NBIA cases, and PLAN, which accounts for about 20%. Because of the rarity of NBIA, knowledge of the clinical characteristics, response to treatment, and prognosis are based mostly on case series.

PLAN has 3 distinct clinical phenotypes. The first, often referred to as *infantile neuroaxonal dystrophy* or INAD, typically presents between 6 months and 3 years of age with neurodevelopmental arrest and then devastating regression in all domains. Motor symptoms include early truncal hypotonia, limb dystonia, and eventual development of spastic quadriplegia. Many children develop an axonal-type sensorimotor neuropathy with hyporeflexia and paresthesias. Cerebellar ataxia commonly but not invariably develops. Early visual disturbances due to optic atrophy occur in the form of strabismus, nystagmus, and eventual blindness. Seizures occur in up to 17% of patients. Most children with INAD die before age 10 years.

A second phenotype referred to as atypical neuroaxonal dystrophy presents between early childhood and the end of the second decade. It has a slower progression than INAD and presents with heterogeneous clinical features including language difficulties, autism spectrum disorder, eye movement abnormalities, spastic quadriplegia, and progressive dystonia and dysarthria.

A third phenotype called *PLA2G6*-related dystonia-parkinsonism has been described. Patients with this phenotype present between childhood and the third decade of life and experience dystonia, bradykinesia, rigidity, and marked cognitive decline.

In contrast, the more well-known PKAN typically presents in the first decade of life with gait disturbance, dystonia, rigidity, and dysarthria. Slow progression leads to loss of ambulation within 15 years.
PKAN is not typically associated with ataxia or peripheral neuropathy, and visual disturbances occur primarily due to pigmentary retinopathy.6

Brain MRI shows cerebellar atrophy in virtually all well-established PLAN cases. Other neuroimaging findings are more variable. Diffuse T2 white matter hyperintensities, thinning of corpus callosum, and thinning of the optic nerves and chiasm are commonly seen. Unlike PKAN, in which the majority of patients will have the classic “eye of the tiger” sign in globus pallidus, the neuroradiologic evidence of iron deposition in PLAN is much more variable. Many patients with advanced PLAN never have clear radiographic evidence of brain iron accumulation. The gold standard for diagnosis of PLAN used to be demonstration of dystrophic axonal spheroids in nerve and conjunctival biopsies. Now most diagnoses are confirmed through the detection of mutations in the PLA2G6 gene.

The PLA2G6 gene, located on chromosome 22q13, encodes the protein iPLA2-beta, which is a subunit of the calcium-independent phospholipase A_2 enzyme. Phospholipase A_2 is important in the synthesis of free fatty acids and lysophospholipids. A recent study demonstrated that loss of normal PLA2G6 activity leads to elevated mitochondrial lipid peroxidation, mitochondrial dysfunction, and subsequent mitochondrial membrane abnormalities.8 Mouse models of PLAN reveal decreased incorporation of DHA into the brain.9 DHA is a precursor of anti-inflammatory neuroprotectins, and decreased brain DHA metabolism may increase vulnerability to neuro-inflammation. However, it remains unclear how these disturbances lead to brain iron accumulation or the clinical findings seen in PLAN. There is not a clear correlation between specific PLA2G6 gene mutations and clinical presentation.

The standard of care for PLAN is supportive care and symptomatic treatment of dystonia, spasticity, and epilepsy. There is no disease-specific treatment for PLAN. Some current clinical trials are evaluating treatment of NBIA with iron chelators,10 but most only enroll patients with PKAN. Some centers have treated individual patients with supplementary DHA, although efficacy is unclear and results have not been published. Multinational consortia will be needed to enroll sufficient numbers of patients for any treatment trials of this rare disease.

CONCLUSION PLAN is a form of NBIA that should be considered in any child with developmental regression, especially when associated with dystonia, truncal hypotonia, peripheral neuropathy, ataxia, visual disturbance, and/or cerebellar atrophy. The lack of radiographic evidence of iron accumulation, especially early in disease course, should not dissuade clinicians from including this rare neurodegenerative disease in their differential.

AUTHOR CONTRIBUTIONS
Robert B. Blake, MD: conceptualized, drafted, and revised the manuscript for intellectual content. Donald L. Gilbert, MD: provided significant revision of manuscript for intellectual content. Mark B. Schapiro, MD: conceptualized, drafted, and revised the manuscript for intellectual content.

STUDY FUNDING
No targeted funding reported.

DISCLOSURE
R. Blake reports no disclosures relevant to the manuscript. D. Gilbert has received honoraria from the Tourette Syndrome Association, Centers for Disease Control and Prevention, the American Academy of Pediatrics, and the Hong Kong Society of Child Neurology and Developmental Pediatrics; has received book royalties from Elsevier. He has received compensation for expert testimony for the U.S. D.O.J. D.V.I.C. program. Dr. Gilbert has received research support from the NIH (NIMH, NINDS). He has received funding for work as a clinical trial site investigator from Ecopipam Pharmaceuticals (clinical trial, Tourette Syndrome) and EzyDel (clinical trial, Ataxia Telangiectasia). M. Schapiro reports no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

REFERENCES
Clinical Reasoning

Clinical Reasoning focuses on case presentations with the aim of developing clinical reasoning skills among trainees. Appropriate cases for publication would include uncommon presentations of common neurological disorders and also typical presentations of more exotic disorders. The emphasis of the case presentation should be on generating a sound, thorough differential diagnosis; logically arriving at the correct diagnosis; and thoughtfully discussing the teaching-points of the case. Cases discussed in the section should utilize data presented serially in two to four segments that could be opened sequentially by the reader, allowing them to challenge themselves by thinking through the differential diagnosis or treatment options at each step. The manuscript should indicate where each break would occur, with specific questions for the reader to consider as they work their way through the case. The final section should provide the experienced clinician’s discussion (or resident author’s literature review). Ideally the individual sections will also include visually presented data, such as radiology, EEG, EMG, or other studies. See published samples as examples.
Clinical Reasoning: Heart to swallow

SECTION 1
A 55-year-old woman was admitted to our intensive care department after intoxication with lithium. Her medical history was relevant for bipolar disorder for which she received medical treatment with lithium, haloperidol, and citalopram. In the week before admission, she had developed a clinical picture of gastroenteritis with diarrhea and vomiting, which resulted in dehydration and a marked deterioration in kidney function. During the last days before admission, she had become progressively lethargic and had developed dysarthria as well as postural tremors of the extremities. The tremors were coarse and irregular, most clearly present in the hands with a frequency of approximately 8 Hz. At admission, she opened her eyes spontaneously, localized pain, and showed normal verbal responses (E4M5V5). Physical examination showed severe dysarthria, ataxia, and no focal neurologic deficits. Reflexes were normal. Laboratory investigation revealed a de novo elevated creatinine level (220 μmol/L, normal 49–90) corresponding to an estimated glomerular filtration rate of 22 mL/min/1.73 m² (normal >60) and a lithium level that was 5.8 mmol/L (target value 0.6–0.8 mmol/L). ECG at admission was normal except for a prolonged QTc interval of 533 milliseconds (normal <450).

Questions for consideration:
1. What is the causal relationship between the observed symptoms and lithium and creatinine levels?
2. What is the treatment of this disorder?
SECTION 2
Potential explanations for altered consciousness and tremors include intoxications (lithium, serotonergic, or antiepileptic drugs) and alcohol withdrawal. The tremor described here has the typical characteristics of a lithium tremor.1 These symptoms, in combination with elevated lithium levels led to a diagnosis of severe lithium neurotoxicity due to hypovolemia-induced renal failure. Since the excretion of lithium is almost uniformly renal, acute lithium toxicity may be initiated by a loss of renal function. Patients with lithium intoxication often develop gastrointestinal symptoms, e.g., nausea and vomiting. If these symptoms are severe, dehydration and decreased renal function may develop. This impairs the ability to excrete lithium and exacerbates lithium toxicity. Therapeutic intervention should focus on rehydration and the removal of lithium from the body. Restoration of electrolyte and water balance by rehydration in hypovolemic patients with lithium toxicity is mandatory to maintain or restore kidney function and maximize lithium clearance. Removal of lithium should be achieved by discontinuation of the drug as well as extracorporeal removal by means of hemodialysis or continuous veno-venous hemofiltration (CVVH). Lithium is readily dialyzable since it is nonprotein bound and has a low molecular weight and a small volume of distribution. Usually one session of hemodialysis or 24 hours of CVVH is sufficient, although some experts advise to continue dialysis after normal (<1 mmol/L) levels of lithium have been achieved to prevent a rebound effect.2

In our patient, treatment was initiated with rehydration as well as CVVH and the patient was admitted to our intensive care unit (ICU). CVVH was continued for 24 hours, which resulted in a decrease of lithium levels toward therapeutic levels. However, severe lethargy as well as neurologic sequelae persisted for 14 days despite normalization of lithium levels.

Question for consideration:
1. Should a different diagnosis be considered in view of the persistence of neurologic symptoms?
SECTION 3

Neurologic complications of lithium intoxication usually develop later in the clinical course because of the relatively slow absorption in the CNS. Furthermore, they may persist despite removal of lithium by hemodialysis or hemofiltration. This "syndrome of irreversible lithium effectuated neurotoxicity" (SILENT) is characterized by prolonged neurologic and neuropsychiatric sequelae, which may persist for months. Demyelination at multiple sites in the CNS has been suggested to be the cause and can sometimes be observed on MRIs as focal white matter abnormalities. In our patient, in addition to her neurologic symptoms, episodic symptomatic bradycardias and atrioventricular (AV) blocks associated with hypotension were observed during her stay at the ICU. Upon careful analysis, we observed a close relationship of bradycardias with swallowing. The patient denied ever having symptomatic bradycardias or presyncopal complaints when swallowing before the current episode of lithium intoxication. A representative ECG after swallowing is shown in the figure; also see the video on the Neurology® Web site at Neurology.org (which was recorded relatively late in the clinical course). These bradycardias persisted during the first week of admission but eventually diminished and disappeared. After 2 weeks of admission, her neurologic symptoms slowly improved as well and she was discharged toward a psychiatric care facility for rehabilitation, reinstitution, and optimization of medical therapy for her bipolar disorder.

Questions for consideration:

1. What is the phenomenon observed in the figure and the video?

2. What is the presumed pathophysiology of this phenomenon and what is its relation to lithium intoxication?
SECTION 4

Our patient had episodes of swallow-induced transient bradycardia. As bradycardia was not severe enough to cause loss of consciousness, these episodes do not fulfill the criteria for syncope. Nonetheless, episodes of swallow-induced bradycardia are characteristic for “swallow syncope,” a rare syndrome that belongs to the reflex syncope syndromes. The pathophysiology of swallow syncope is incompletely understood but likely involves a vagal reflex that is initiated by activation of the glossopharyngeal nerve during swallowing. Efferent impulses lead to the sinoatrial node (right vagus nerve) or the AV node (left vagus nerve) and may lead to various types of paroxysmal bradyarrhythmias. The importance of vagal pathways in this reflex is stressed by studies in which pretreatment with atropine or other anticholinergic drugs is effective in preventing swallow-induced bradycardia. A recent review of all 80 published cases of swallow syncope showed that the majority of cases (62%) had underlying cardiac or gastrointestinal disease, although a substantial minority of patients did not have any underlying pathology. Treatment of the syndrome may involve implantation of a permanent pacemaker. In patients in whom quality of life is severely affected by recurrent syncopal events, this treatment is usually effective.

DISCUSSION

Herein, we report a case of a patient who had been chronically treated with lithium who developed severe neurologic as well as cardiac symptoms due to an intoxication of lithium, which was caused by prerenal kidney insufficiency-related reduced elimination of lithium. Lithium salts have been used for the treatment of psychosis and bipolar disorder since the 19th century. Although effective, it has a narrow therapeutic index. This is illustrated by the fact that a majority of patients chronically treated with lithium experience at least one episode of toxicity during their course of treatment. Patients may be relatively asymptomatic despite very high serum concentrations, and severe clinical toxicity may develop despite lithium concentrations in the therapeutic range. Therefore, the diagnosis and treatment of this syndrome should rely on a combination of clinical symptoms as well as drug levels.

Despite rapid normalization of plasma lithium levels, neurologic symptoms persisted for several weeks, which is consistent with the SILENT syndrome as discussed above. Potential neurologic symptoms include lethargy and coma, ataxia, confusion or agitation, and neuromuscular excitability.7

In addition to neurologic symptoms, she developed cardiac toxicity: she presented with a markedly prolonged QTc time, which did not lead to rhythm disturbances initially and normalized rapidly after plasma lithium levels were corrected. Of note, however, during her stay at the ICU, she developed symptomatic sinus node bradycardias as well as AV blocks that were provoked by swallowing. Cardiac toxicity may cause changes in the ECG. Although arrhythmias are rare, prolonged QTc intervals and bradycardia have been reported. Whereas swallow-induced bradycardia has been described previously, provocation of this syndrome by lithium toxicity has not. Lithium exerts its actions through the alteration of sodium transport in neurons, which increases intraneuronal metabolism and reduces stores of catecholamines. In the heart, lithium is a potent blocker of cardiac sodium channels. This lithium-related blockade of sodium channels can unmask conduction abnormalities in the heart such as conduction delays as well as Brugada syndrome although other mechanisms of lithium-associated bradyarrhythmias have been described as well.11 Plans were made to perform extensive autonomic nervous system tests in our patient to evaluate the effects of vagal maneuvers on her symptoms and heart, but unfortunately these were hampered by her persistent neurologic and psychiatric symptoms. By the time her neuropsychiatric status improved, her episodes of swallow syncope had resolved completely. Finally, we aimed to exclude whether genetic defects in ion channels contributed to the observed clinical phenomena. Recent molecular insights have defined a molecular basis for sinoatrial and AV node dysfunctions and several mutations in the genes encoding for cardiac sodium channels have been described that contribute to these dysfunctions.12 To analyze whether such mutations may have had a role in our patient’s symptoms, we performed genetic testing of our patient and tested for mutations in 48 genes (next-generation sequencing arrhythmia panel, http://amsterdamgenomedx.com) that are associated with arrhythmias but were unable to find genetic polymorphisms that predispose to arrhythmias in this specific case. We postulate that lithium provoked episodes of swallow-induced bradycardia in our patient due to the combined effects of swallowing-induced vagal efferent activity and an increased susceptibility to bradycardia caused by lithium.

AUTHOR CONTRIBUTIONS

Dr. van Westerloo: concept and design of the report, data accrual, wrote the manuscript. Dr. Barge-Schaapveld: analysis of genetics, critical revision of the manuscript for important intellectual content. Dr. Bikker: performed DNA analysis, critical revision of the manuscript for important intellectual content. Dr. van Noorden: critical revision of the manuscript for important intellectual content. Dr. Tannemaat: performed DNA analysis, critical revision of the manuscript for important intellectual content. Dr. van Westerloo: concept and design of the report, data accrual, cowrote the manuscript.

STUDY FUNDING

No targeted funding reported.

DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.
REFERENCES
Education Research

As the central mission of *Neurology*, education is a top priority. This is a section for interventional educational studies, as well as more traditional educational research, such as surveys. This section will examine the way neurologists not only practice, but also the way we teach and approach education. Neurologists have traditionally been respected, perhaps above all other specialties, for their scholarship and teaching. Educational issues will therefore continue to be at the center of the mission of *Neurology*.
Education Research: Neurology resident education
Trending skills, confidence, and professional preparation

ABSTRACT
Objective: To survey US-trained graduating neurology residents who are American Academy of Neurology members, in an effort to trend perceived quality and completeness of graduate neurology education.

Methods: An electronic survey was sent to all American Academy of Neurology members graduating from US neurology residency programs in the Spring of 2014.

Results: Of 805 eligible respondents, 24% completed the survey. Ninety-three percent of adult neurology residents and 56% of child neurology residents reported plans to pursue fellowship training after residency. Respondents reported a desire for additional training in neurocritical care, neuro-oncology, neuromuscular diseases, botulinum toxin injection, and nerve blocks. There remains a clear deficit in business training of neurology residents, although there was notable improvement in knowledge of coding and office management compared to previous surveys.

Discussion: Although there are still areas of perceived weakness in neurology training, graduating neurology residents feel generally well prepared for their chosen careers. However, most still pursue fellowship training for reasons that are little understood. In addition to certain subspecialties and procedures, practice management remains deficient in neurology training and is a point of future insecurity for most residents. Future curriculum changes should consider resident-reported gaps in knowledge, with careful consideration of improving business training.
neurology fellows (as such trainees are classified by certain institutions). Regardless of institutional nomenclature, all child neurology trainees are referred to as residents in this report. Members were e-mailed approximately 1 month before the expected graduation date by the chair of the Consortium of Neurology Residents and Fellows with a link to the online survey as well as information on a sweepstakes in which two $500 gift cards would be raffled for incentive. Two additional reminder e-mails were sent to nonrespondents over the following 2 weeks. Data collection closed after 4 weeks.

A statistical comparison of responses from those who had completed adult neurology residencies (n = 168) and child neurology residencies (n = 27) was completed. Significant differences between groups are mentioned where relevant in this article. Overall survey analysis combined adult and child neurology residents. Comparisons between adult and child neurology respondents were tested for significance with $\chi^2$ tests, and longitudinal differences between survey responses, where relevant, were tested for significance with analysis of variance or $t$ tests.

**RESULTS**

The survey response rate was 24.2% (195/805), which included 168 adult neurology residents and 27 child neurology residents. The margin of error for all respondents was ±6.1% (95% confidence interval). Demographics of survey respondents are shown in table 1. There were no differences between respondents and nonrespondents regarding age or sex.

**Fellowship trends and subspecialty training.** Eighty-eight percent of respondents reported plans to pursue fellowship, 44% in their home institution and 56% elsewhere. Broken down by residency type, significantly more adult neurology residents plan to pursue fellowship than child neurology residents (93% vs 56%, $p < 0.0001$). Subspecialties in which respondents ultimately plan to practice are shown in table 2. When asked to mark all areas in which they plan to specialize, respondents most often selected general neurology (25%), epilepsy (21%), and vascular neurology (19%). Forty-four percent of respondents selected multiple areas of specialization; when those individuals were then asked to identify a single

<table>
<thead>
<tr>
<th>Demographic characteristics of the survey population</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Demographic characteristics</strong></td>
</tr>
<tr>
<td>Age, y, mean (SD)</td>
</tr>
<tr>
<td>Sex, %</td>
</tr>
<tr>
<td>Male</td>
</tr>
<tr>
<td>Female</td>
</tr>
<tr>
<td>Neurology residency, %</td>
</tr>
<tr>
<td>Adult</td>
</tr>
<tr>
<td>Child</td>
</tr>
</tbody>
</table>

There were no significant differences between age or sex among survey respondents and nonrespondents. Data were missing for age in 15% of respondents and 16% of nonrespondents. Data were missing for sex in 4% of respondents and 4% of nonrespondents.

Because of respondents choosing more than one response option, total percent sums to more than 100%.

Table 2  Planned subspecialty practice of 192 residents graduating in 2014

<table>
<thead>
<tr>
<th>Subspecialty</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>General neurology</td>
<td>24.5</td>
</tr>
<tr>
<td>Epilepsy</td>
<td>20.8</td>
</tr>
<tr>
<td>Vascular neurology</td>
<td>18.8</td>
</tr>
<tr>
<td>Movement disorders</td>
<td>14.1</td>
</tr>
<tr>
<td>Clinical neurophysiology</td>
<td>14.1</td>
</tr>
<tr>
<td>Neurohospitalist</td>
<td>11.5</td>
</tr>
<tr>
<td>Neurophysiology</td>
<td>10.9</td>
</tr>
<tr>
<td>Neurocritical care</td>
<td>9.9</td>
</tr>
<tr>
<td>Neuromuscular medicine</td>
<td>9.9</td>
</tr>
<tr>
<td>Child neurology</td>
<td>9.4</td>
</tr>
<tr>
<td>Headache medicine</td>
<td>8.3</td>
</tr>
<tr>
<td>Neuro-oncology</td>
<td>6.8</td>
</tr>
<tr>
<td>Neuroimmunology and multiple sclerosis</td>
<td>5.2</td>
</tr>
<tr>
<td>Behavioral neurology</td>
<td>4.7</td>
</tr>
<tr>
<td>Sleep medicine</td>
<td>3.6</td>
</tr>
<tr>
<td>Infectious disease and neurovirology</td>
<td>2.6</td>
</tr>
<tr>
<td>Traumatic brain injury</td>
<td>2.6</td>
</tr>
<tr>
<td>Neurogenetics</td>
<td>2.6</td>
</tr>
<tr>
<td>Endovascular and interventional neurology</td>
<td>2.1</td>
</tr>
<tr>
<td>Neuro-ophthalmology</td>
<td>2.1</td>
</tr>
<tr>
<td>Neuroimaging</td>
<td>2.1</td>
</tr>
<tr>
<td>Neuromuscular pathalogy</td>
<td>2.1</td>
</tr>
<tr>
<td>Pain medicine</td>
<td>2.1</td>
</tr>
<tr>
<td>Autonomic disorders</td>
<td>1.6</td>
</tr>
<tr>
<td>Geriatric neurology</td>
<td>1.6</td>
</tr>
<tr>
<td>Sports neurology</td>
<td>1.6</td>
</tr>
<tr>
<td>Neuro-otology</td>
<td>1</td>
</tr>
<tr>
<td>Neuroepidemiology</td>
<td>1</td>
</tr>
<tr>
<td>Palliative neurology</td>
<td>0.5</td>
</tr>
<tr>
<td>Neural repair and rehabilitation</td>
<td>0.5</td>
</tr>
<tr>
<td>Global health</td>
<td>0.5</td>
</tr>
<tr>
<td>Unsure</td>
<td>0.5</td>
</tr>
</tbody>
</table>

Primary specialty, child and general neurology were most prevalent (both 13%) followed by neurocritical care and vascular neurology (both 8%). When asked the open-ended question of whether additional exposure to any topics could have better prepared respondents for their careers, neurocritical care, neuro-oncology, and neuromuscular disease were most frequently listed.

**Procedure skills and diagnostic test interpretation.** Extent of experience with various procedures is shown in table 3. Of note, many respondents
reported a desire for additional residency training in botulinum toxin injections (66%), nerve blocks (51%), vagus nerve stimulator programming (33%), and deep brain stimulation programming (33%). Adult neurology residents reported significantly more experience than child neurology residents in performing one or more instances of IV tissue plasminogen activator administration (98% vs 70%, \( p < 0.0001 \)). However, child neurology residents reported significantly more experience than adult neurology residents in performing one or more procedures of intrathecal pump management (13% vs 9%, \( p < 0.0001 \)) and vagus nerve stimulator programming (78% vs 28%, \( p < 0.0001 \)).

Resident-reported experience with interpreting diagnostic studies is summarized in table 4. The majority (88%) of all respondents thought that they were either “very prepared” or “somewhat prepared” to perform the procedures that may be expected of them in their upcoming jobs or fellowships. Child neurology residents reported significantly less experience than adult neurology residents performing or interpreting EMG and nerve conduction studies (71% vs 90%, respectively, \( p = 0.04 \)).

Business training. Reported business training during residency is trended over time in the figure. While the majority of respondents thought that they were “not at all well” prepared or “not very well” prepared for competency in billing (63%), coding (63%), contract negotiations (80%), office management (80%), malpractice insurance (84%), or relative value units (79%), there were still significant improvements across serial surveys in preparation for coding (\( p = 0.003 \)) and office management (\( p = 0.01 \)). Furthermore, between 2011 and 2014, there was a significant improvement in level of preparedness for billing (\( p = 0.048 \)). Fifty-five percent of all respondents reported no business training during residency, 26% reported informal, extracurricular training, and 19% reported business management training as a part of their residency curriculum. On the whole, 53% of respondents reported feeling either “very unprepared” or “somewhat unprepared” for practice management tasks that may be expected of them in their subsequent jobs, with only 6% feeling “very prepared” for these tasks. When asked what particular educational resources the AAN could develop to help residents and early-career members with business management, respondents’ fill-in-the-blank responses most commonly mentioned billing and coding.

Student loans. Graduating residents were asked about indebtedness from student loans and plans for repayment. These questions were optional, and only 22 responses were received. More than half of those who responded reported \( \geq \$100,000 \) in debt, with nearly one-third reporting \( \geq \$200,000 \). Almost one-third of respondents reported no specific plan for paying off their debt, while the others were equally split between fast and slow payoff plans.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Performed more than once</th>
<th>Performed once</th>
<th>Observed only</th>
<th>No training</th>
<th>Not applicable to residency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lumbar puncture</td>
<td>97.9</td>
<td>1.1</td>
<td>0.5</td>
<td>0.5</td>
<td>0</td>
</tr>
<tr>
<td>IV tPA</td>
<td>92.5</td>
<td>1.6</td>
<td>4.8</td>
<td>0.5</td>
<td>0.5</td>
</tr>
<tr>
<td>Botulinum toxin</td>
<td>39.2</td>
<td>16.4</td>
<td>33.3</td>
<td>9.5</td>
<td>1.6</td>
</tr>
<tr>
<td>Local anti-inflammatory or anesthetic</td>
<td>35.7</td>
<td>13.5</td>
<td>15.7</td>
<td>29.2</td>
<td>5.9</td>
</tr>
<tr>
<td>Nerve blocks</td>
<td>28.5</td>
<td>9.7</td>
<td>23.1</td>
<td>32.8</td>
<td>5.9</td>
</tr>
<tr>
<td>VNS program</td>
<td>22.7</td>
<td>11.9</td>
<td>36.8</td>
<td>25.9</td>
<td>2.7</td>
</tr>
<tr>
<td>DBS program</td>
<td>11.2</td>
<td>8</td>
<td>49.5</td>
<td>25</td>
<td>6.4</td>
</tr>
<tr>
<td>IT pump management</td>
<td>5.4</td>
<td>4.3</td>
<td>38.4</td>
<td>37.3</td>
<td>14.6</td>
</tr>
<tr>
<td>Muscle/nerve biopsy</td>
<td>4.3</td>
<td>4.3</td>
<td>33.2</td>
<td>49.2</td>
<td>9.1</td>
</tr>
<tr>
<td>Lumbar drain placement</td>
<td>3.2</td>
<td>5.4</td>
<td>24.2</td>
<td>48.9</td>
<td>18.3</td>
</tr>
<tr>
<td>IA thrombolysis</td>
<td>1.1</td>
<td>0.5</td>
<td>64.2</td>
<td>21.9</td>
<td>12.3</td>
</tr>
<tr>
<td>Intravascular coiling</td>
<td>1.1</td>
<td>0</td>
<td>46.7</td>
<td>34.4</td>
<td>17.8</td>
</tr>
<tr>
<td>Angioplasty and stenting</td>
<td>0.5</td>
<td>1.6</td>
<td>51.9</td>
<td>25.1</td>
<td>20.9</td>
</tr>
<tr>
<td>Embolization</td>
<td>0.5</td>
<td>0.5</td>
<td>43.2</td>
<td>36.2</td>
<td>19.5</td>
</tr>
</tbody>
</table>

Table 3 Extent of resident experience with therapeutic or invasive neurologic procedures

Abbreviations: DBS = deep brain stimulator; IA = intra-arterial; IT = intrathecal; tPA = tissue plasminogen activator; VNS = vagus nerve stimulator.

Data are percentages. Number of respondents for each category is between 180 and 189.
DISCUSSION While nearly 90% of graduating neurology residents believe they are prepared to perform the procedures necessary for their subsequent jobs, the rate of fellowship training remains high. Understanding the influences on this trend may provide important insight into the value and perceived needs of resident training, as well as the future of the neurology workforce. While education may be affected by systemic elements such as work hour restrictions and imbalance of inpatient and outpatient rotations, external factors such as board certification requirements and financial incentives likely impose equal pressure for trainees to seek fellowship training. Demonstrative of this, the American Board of Psychiatry and Neurology mandates 1 year of neurophysiology fellowship training to achieve eligibility for neurophysiology board certification. Not only is this certification prestigious but it also may be required as a condition of employment in various centers, and ultimately could even be required to bill for certain procedures. This latter point may be especially important for the 25% of respondents who will ultimately work in general neurology, of whom 81% reported plans for a fellowship first. Therefore, while curriculum changes may raise the bar for procedural exposure and competency, like the Neurology Milestones by the Neurology Residency Review Committee of the ACGME, their effect on postresidency training may be tempered by external incentives.

It is of interest that more than 50% of respondents reported a desire for more training on botulinum toxin injections and nerve blocks. This likely reflects the growing number of uses for these procedures, including in headache neurology, movement disorders, pain management, and neuro-oncology.

The following table presents the frequency of interpretation or performance of diagnostic studies reported by graduating neurology residents. The data are presented as percentages, with the number of responses for each category ranging between 181 and 186.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Interpreted more than once</th>
<th>Interpreted once</th>
<th>Observed interpretation</th>
<th>No training</th>
<th>Not applicable to training</th>
</tr>
</thead>
<tbody>
<tr>
<td>EEG</td>
<td>96.8</td>
<td>0</td>
<td>3.2</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>MRI</td>
<td>95.1</td>
<td>0.5</td>
<td>4.3</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>CT</td>
<td>94</td>
<td>0</td>
<td>4.9</td>
<td>1.1</td>
<td>0</td>
</tr>
<tr>
<td>EMG/NCS</td>
<td>86</td>
<td>1.6</td>
<td>12.4</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Angiogram</td>
<td>60.2</td>
<td>1.7</td>
<td>26.5</td>
<td>9.9</td>
<td>1.7</td>
</tr>
<tr>
<td>Evoked potentials</td>
<td>31.7</td>
<td>9.8</td>
<td>31.7</td>
<td>26.2</td>
<td>0.5</td>
</tr>
<tr>
<td>PET</td>
<td>25.7</td>
<td>8.2</td>
<td>37.2</td>
<td>24</td>
<td>4.9</td>
</tr>
<tr>
<td>TCD</td>
<td>24.5</td>
<td>7.6</td>
<td>40.8</td>
<td>25</td>
<td>2.2</td>
</tr>
<tr>
<td>PSG</td>
<td>21.9</td>
<td>11.5</td>
<td>41</td>
<td>24</td>
<td>1.6</td>
</tr>
<tr>
<td>Carotid US</td>
<td>18.5</td>
<td>7.1</td>
<td>39.7</td>
<td>31</td>
<td>3.8</td>
</tr>
<tr>
<td>fMRI</td>
<td>13.1</td>
<td>6</td>
<td>41.1</td>
<td>36.1</td>
<td>3.8</td>
</tr>
<tr>
<td>IOM</td>
<td>9.9</td>
<td>4.9</td>
<td>42.3</td>
<td>39.6</td>
<td>3.3</td>
</tr>
<tr>
<td>Other*</td>
<td>1.7</td>
<td>1.7</td>
<td>10.3</td>
<td>41.4</td>
<td>44.8</td>
</tr>
</tbody>
</table>

Abbreviations: EMG/NCS = electromyography/nerve conduction study; fMRI = functional MRI; IOM = intraoperative monitoring; PSG = polysomnogram; TCD = transcranial Doppler; US = ultrasound.

Data are percentages. Number of responses for each category is between 181 and 186. *SISCOM (subtraction ictal SPECT coregistered to MRI), DaT (dopamine transporter) scan, optical coherence tomography, video nystagmogram.

The percentage of residents feeling either “very well” or “somewhat well” prepared for business management topics is shown in the figure. Questions on all 3 survey iterations were presented in a 4-point Likert scale design. Significant improvement was noted in the categories of coding (p = 0.003) and office management (p = 0.011) when trended with 2007 and 2011, and in the category of billing preparedness when compared to 2011 (p = 0.048). The 2014 responses for meaningful use of electronic medical records were compared to 2011 responses for medical record use, because of category similarity. EMR = electronic medical record; RVUs = relative value units.
disorders, autonomic neurology, pain management, and rehabilitative neurology (both with and without Food and Drug Administration indications). Given that these are office-based procedures, and offer rapid therapeutic benefit to patients and potential revenue to practitioners, adding them to core competencies during residency training may be useful.

This survey revisited aspects of business management training important to residents’ future careers. Although modest, there have been significant improvements in preparation for various business topics, including coding, billing, and office management, presumably reflecting increased exposure. The fact that more than two-thirds of respondents believe they are well trained for meaningful use of electronic medical records likely reflects not only the relatively young age and technologic savvy of respondents, but also focused hospital-required training. This was also the first AAN resident survey in which respondents were asked about competency in the use of relative value units, and less than 20% reported feeling either “very well” or “somewhat well” prepared to use these. While this number is quite low, it is in line with deficiencies in business training in general.

An important consideration in graduate medical education is trainee student loan indebtedness, and how this relates to training and career plans. The current survey provides the first look into this issue in graduating neurology residents. Although there were relatively few responses, the data nonetheless highlight the large amount of student debt held by graduating neurology residents and the lack of specific planning toward paying this off. This topic should be explored more thoroughly in future survey iterations, as well as how debt correlates with fellowship and career plans, and overall trainee and neurologist satisfaction.

The survey response rate of 24% was low compared to roughly 50% response rates in prior iterations of this survey. This reflects an ongoing trend toward lower response rates within all survey-based research through the AAN and may stem, in part, from survey fatigue. This was the first time the survey was incentivized with a drawing for two $500 gift cards, which did not succeed in bringing response rates to 2007 or 2011 levels; future surveys may consider a small prize for each respondent rather than random drawings for large prizes. While the number of child neurology respondents was much smaller than adult neurology respondents (14% vs 86%, respectively), this is reflective of the 12% of AAN members who self-describe as child neurology practitioners according to the 2015 AAN Member Insights Report, an unpublished report based on AAN member database information. However, the low absolute number of child neurology respondents limits the interpretability of population-specific data.

It is worth noting, though, that the relative stability of various answers across serial surveys and the 6% margin of error on the present survey suggest that the results are nonetheless generalizable across AAN member graduating neurology resident cohorts.

The neurology resident survey will need to evolve as requirements and trends in education change. Possible future topics include the following: (1) develop an understanding of how student loan indebtedness influences financial, personal, and professional choices of residents after training; (2) understand the perceived need for trainees to obtain fellowship training if they plan to pursue a career in general neurology; and (3) determine whether residency training programs are providing instruction about reducing variance in the care of patients with similar neurologic problems within a practice or health system, in line with priorities designated by the Affordable Care Act and the Institute of Medicine Report on the Future of Graduate Medical Training in the United States. In addition, evaluating both specific fellowships pursued and subspecialty career plans may improve interpretation of the trends. Longitudinal follow-up of survey respondents may also be helpful in order to reevaluate perceived training deficiencies and strengths after entering neurology careers. The AAN performs qualitative studies of early-career neurologists, which serve to better capture the benefits and incentives for fellowship training. The data from these groups may provide additional insights into the preparation of future surveys.

AUTHOR CONTRIBUTIONS
Design of study: Jordan, Mayans, Schneider, Adams, Engstrom. Analysis and interpretation of data: Jordan, Mayans, Schneider, Adams, Khawaja. Drafting of the manuscript: Jordan, Adams. Critical revision of the manuscript for important intellectual content: Jordan, Mayans, Schneider, Adams, Khawaja, Engstrom.

ACKNOWLEDGMENT
The authors thank Cheryl Alementi, Lucy Persaud, the members of the AAN Graduate Education and Member Research Subcommittees and Consortium of Neurology Residents and Fellows, the AAN Member Insights Department, as well as all respondents of our survey.

STUDY FUNDING
No targeted funding reported.

DISCLOSURE

REFERENCES
Emerging Subspecialties in Neurology

These manuscripts will review the history and development of emerging subspecialties in neurology, including fields such as pain medicine, headache, neurocritical care, interventional neurology, and others. The focus should be on educating residents with a possible interest in this subspecialty. Those interested in writing these manuscripts should contact the Resident & Fellow Section editor before submission to inquire about the need for an article on a particular topic.
Emerging Subspecialties in Neurology: Pediatric stroke and cerebrovascular disorders

In 1895, a neuropathologist at the University of Vienna observed that “a large number of cases of infantile cerebral palsy is caused by the same factors that bring about the majority of cases of cerebral paralysis of adults: by tearing, embolism, and thrombosis of cerebral vessels.” While Sigmund Freud would go on to become better known for other endeavors, his observation that children can have strokes paved the way for the development of the emerging subspecialty of pediatric stroke and cerebrovascular disorders.

**PEDIATRIC STROKE: CLINICAL ASPECTS** Stroke, ischemic and hemorrhagic, is estimated to occur in at least 2–3 per 100,000 children annually, and 1 in 4,000 neonates. This makes pediatric strokes relatively common among serious pediatric neurologic conditions. However, recognition by both parents and providers is often delayed due in part to lack of awareness of stroke as a disorder affecting children. While children show remarkable resiliency and potential for recovery, 20%–40% of children die after their stroke, and up to 80% experience poststroke disability compounded over decades of remaining life expectancy.

Causes of stroke in children range from infectious and inflammatory arteriopathies to cardiogenic embolism to genetic and acquired thrombophilias and vascular abnormalities. Sites of pediatric stroke care delivery range from the emergency room and intensive care unit to rehabilitation centers and outpatient multidisciplinary clinics. The field is therefore inherently multidisciplinary, offering neurologists the opportunity to work together with numerous specialists across fields and settings. This interdisciplinary richness and variety appeal to many who choose the subspecialty.

**CAREERS IN PEDIATRIC STROKE** Pediatric stroke neurology is now a career option, one that offers significant rewards. The excitement of being on the cutting edge of a new field of discovery provides opportunities for challenging and stimulating intellectual and research pursuits. Pediatric stroke specialists will follow neonates and children with stroke from the onset of acute stroke through diagnosis and recovery, affording both the excitement of acute management and the fulfillment of long-term relationships with patients and families. Some pediatric stroke specialists will attend on both the pediatric and adult stroke services in the hospital, including sharing in both call pools. Others will see pediatric stroke patients as a portion of their clinical efforts, devoting other inpatient or outpatient time to critical care neurology or other general pediatric neurology. Many pediatric stroke neurologists also devote time to basic, translational, or clinical research. The parallel emergence of neuropsychologists, nurse practitioners, and rehabilitation therapists focused on pediatric stroke provides opportunities for a team approach in both clinical care and research. Finally, children with stroke and their parents have formed strong collaborations with health practitioners in this field, providing partnership in advocacy for children with stroke (see Resources).

**HISTORY OF THE FIELD** Until the 1990s, there was little forward movement in pediatric stroke. Then, neurologists and hematologists convened the first pediatric stroke interest groups and symposia at the Institute of Child Health, London, United Kingdom. The field was defined as interdisciplinary and collaborative from the outset. To overcome the challenge of studying a relatively uncommon condition, hematologist Maureen Andrew and neurologist Gabrielle deVeber founded the Canadian Pediatric Ischemic Stroke Registry in 1992 and the “1-800-NOCLOTS” telephone consultation database in 1994, gathering data on thousands of cases. This ultimately led to the 2003 creation of the International Pediatric Stroke Study (IPSS), a database currently approaching 5,000 cases of pediatric stroke from 215 centers in 54 countries around the world (figure). The IPSS has produced 22 publications, and multiple NIH-funded studies. The Pediatric Stroke Outcome Measure, developed in 1995, enables cross-center comparisons of outcomes similar to adult measures such as the modified Rankin Scale. A pediatric version of the National Institutes of Health Stroke Scale (NIHSS), the...
PedNIHSS, was validated in 2011. Three sets of guidelines have been published by the Royal College of Physicians, the American Heart Association/American Stroke Association, and the American College of Chest Physicians to summarize existing evidence and provide recommendations for diagnosis and management of pediatric stroke. Clinical and translational research related to pediatric stroke is booming; in the last 15 years, there have been over 1,500 publications. Finally, centers of excellence are developing analogous to adult primary and comprehensive stroke centers, with at least 17 centers now prepared to diagnose and treat pediatric stroke based upon standardized protocols. Acute and long-term treatment trials in pediatric stroke are underway.

**FELLOWSHIP TRAINING** Pediatric stroke has emerged not only as a field of practice but also as a dedicated training pathway. Before dedicated training programs existed, current leaders in the field trained in adult neurology, pediatric neurology, or hematology. A pediatric stroke fellowship was established in 1998 at Toronto Children’s Hospital, and other programs have followed, responding to the need for future pediatric stroke neurologists to master an understanding of pediatric stroke and individualized application of emerging guidelines and standards of care in addition to knowledge of the rapidly expanding literature. Dedicated training offers fellows not only depth of experience based on caring for a sufficient volume of pediatric stroke cases, but also breadth, seeing many unique disorders associated with pediatric stroke that are unlikely to be encountered in regular residency training. Fellows gain experience in providing acute stroke care within complex systems, leading “stroke codes” under the supervision of attending stroke specialists. As fellowship training generally includes both adult and pediatric clinical experience, stroke fellows also gain experience with stroke syndromes in adults and with models of stroke care delivery in established adult stroke centers. Depending on the training program, some pediatric stroke fellows are eligible for Vascular Neurology board certification by the American Board of Psychiatry and Neurology. Finally, fellowships offer the opportunity for mentored research experience to launch a career in academic medicine.

Multiple training pathways are open to trainees interested in pediatric stroke. At present, 5 academic centers in North America offer clinical and research fellowships in pediatric stroke and cerebrovascular disorders on an ongoing basis: The Hospital for Sick...
Children in Toronto, Children’s Hospital of Philadelphia, Boston Children’s Hospital, the University of Calgary, and the University of Colorado. Additional centers including the University of California San Francisco, Johns Hopkins University, and the University of Texas–Houston have accepted child neurology trained applicants to their vascular neurology fellowships accredited by the American College of Graduate Medical Education (ACGME); only ACGME-accredited programs confer eligibility for board certification. Other trainees may choose to pursue established fellowships in adult or pediatric critical care or neurocritical care, a related emerging subspecialty with an emphasis on acute management of neurologic emergencies.9 Other centers with pediatric stroke expertise may be willing to customize a program for interested trainees.

While the length and composition of pediatric stroke training programs vary, all typically include a combination of inpatient and outpatient clinical experience in pediatric stroke and related disorders. Most have some clinical time devoted to adult stroke. Additional clinical exposures that programs may provide include neurosurgery, interventional neuroradiology, hematology, rehabilitation, and critical care. Some programs include formal training in research methodology. To further trainee education, programs typically include teaching conferences and journal clubs, including with the adult stroke fellows from sister programs. Pediatric stroke trainees may also become involved with clinical education for other neurologists through consultations or formal teaching opportunities, as well as with interprofessional education. Pediatric stroke trainees benefit from exposure to the state of the field and the larger pediatric stroke community through attendance and presentations at conferences. As the number of pediatric stroke trainees grows, collaboration across training programs would provide further opportunities to enhance training. Perhaps in the future there may even be a board certification in pediatric vascular neurology.

EXPLORING THE FIELD Neurology trainees and medical students interested in learning more about the field of pediatric stroke should feel welcomed and encouraged to explore this career path and consider dedicated training. Clinical rotations at institutions with pediatric stroke specialists and a high volume of pediatric stroke patients offer an excellent introduction to the field. Research projects in topics related to pediatric stroke are an opportunity to become familiar with and contribute to the burgeoning literature. Finally, attendance at sessions related to pediatric stroke at annual conferences including the International Stroke Conference, Child Neurology Society, and American Academy of Neurology offer an up-to-date overview of the field and its exciting new developments. The IPSS holds investigator meetings in conjunction with the International Stroke Conference and Child Neurology Society annual meetings.

FUTURE DIRECTIONS The future of the field of pediatric stroke is bright, with increasing workforce demand for pediatric stroke clinicians and many opportunities for further research. Clinical trials in stroke treatment are urgently needed and increasingly feasible given the increasing capacity of pediatric centers to deliver acute stroke care. Such treatments will require increased cross-disciplinary collaboration. In parallel, new technologies are being explored to bring subspecialty expertise including acute stroke care into the community: telestroke systems for adult patients have enabled rapid, remote assessment and treatment, and this is an important area of future development for children with symptoms of stroke.10 Finally, understanding the mechanisms of pediatric stroke is increasingly possible through advanced neuroimaging, genetic testing, and other areas of scientific progress. Trainees who are excited by the prospect of a multidisciplinary, collaborative, and rapidly evolving field of clinical care and research will find pediatric stroke and cerebrovascular disorders to be a rewarding subspecialty.

ADDITIONAL ONLINE RESOURCES

• International Pediatric Stroke Study: app3.ccb.sickkids.ca/cstrokedstudy/
• American Heart Association/American Stroke Association Stroke in Children: strokeassociation.org/STROKEORG/AboutStroke/StrokeInChildren/Stroke-In-Children_UCM_308543_SubHomePage.jsp
• Pediatric National Institutes of Health Stroke Scale (PedNIHSS) (data supplement to Ichord et al.5): stroke.ahajournals.org/content/suppl/2011/02/17/STROKEAHA.110.607192.DC1/607192_ONLINE_SUPPLEMENT.pdf
• International Alliance for Pediatric Stroke: iapediatricstroke.org
  Also see local organizations: iapediatricstroke.org/organizations.aspx
• World Pediatric Stroke Organization: http://www.worldpediatricstrokeassociation.org/
• Canadian Paediatric Stroke Support Association: cpssa.org
• Children’s Hemiplegia and Stroke Association: chasa.org
• StroKidz: strokidz.com

ADVOCACY AND SUPPORT ORGANIZATIONS (SELECTED)
AUTHOR CONTRIBUTIONS
Dr. Bernson-Leung drafted the manuscript. Dr. deVeber critically revised the manuscript for intellectual content. Both approve the manuscript in its final form.

STUDY FUNDING
No targeted funding reported.

DISCLOSURE
M.E. Bernson-Leung is a former member of the Resident & Fellow Section and a fellow in the Stroke and Cerebrovascular Disorders program at Boston Children’s Hospital. G.A. deVeber is the director of the Canadian Pediatric Ischemic Stroke Registry and the International Pediatric Stroke Study and creator of the Pediatric Stroke Outcome Measure. Go to Neurology.org for full disclosures.

REFERENCES
**Neurology Journal Club**

Neurology Journal Club submissions are structured evaluations of recent Neurology research articles. The aim is to enhance the training of residents and fellows by instruction in the critical appraisal of medical literature. Residents or fellows interested in submitting a Neurology Journal Club article should review the e-Publication Ahead of Print articles at Neurology.org/content/early/recent for the most recently published material and email Neurology with their selection for prior approval. Selections will aim to represent the major categories of research methodology over the course of a three-year residency cycle. Submissions should be timely and are requested no longer than four weeks following the original e-publication date of the subject article. These Journal Club critiques, written by neurology residents and fellows with faculty supervision, should follow a specific outline and contain subtitles for background and significance, hypothesis and design, methods, results, and interpretation. Rather than a critical correspondence or editorial, this feature will highlight methods for the critical appraisal of medical literature. This online feature could be used as an adjunct to traditional institutional journal clubs and promote discussion among neurologists, including trainees and those in practice.
Journal Club: Depression before and after diagnosis with amyotrophic lateral sclerosis

The prevalence of depression among individuals with amyotrophic lateral sclerosis (ALS) is reported to be up to 44%, depending on the assessment methodology. Depression negatively affects quality of life in ALS, and psychological stress is related to poorer survival among individuals with ALS.2

This Journal Club article reports on a study from Roos et al.3 who have calculated the association between ALS and depression, before and after ALS diagnosis. The study provides an elegant example of epidemiologic methods and has important implications for clinical practice, clinical trial design, and public health policy.

HYPOTHESIS AND DESIGN What is the risk of developing depression following diagnosis with ALS? What is the risk of developing depression before ALS is diagnosed? To answer these important epidemiologic questions, Roos et al. performed a population-based nested case-control study using administrative data from the Swedish national health and population registers. The use of a nested case-control study is appropriate here because it is efficient (not all members of the parent cohort need to be examined). This design also reduces selection bias because cases and controls are sampled from the same parent population that is fully enumerated; therefore, control selection can be truly random.

METHODS Cases. ALS cases were defined as an individual with at least one inpatient or outpatient hospital visit at which ALS was recorded as a diagnosis in the Patient Register, using International Classification of Diseases (ICD) codes (ICD-10 G12.2, ICD-9 335C, ICD-8 348.0, ICD-7 356.1). The first hospital visit for ALS was used as the date of ALS diagnosis. The study included cases that were diagnosed between July 2005 and December 2010.

Controls. For each case, 5 controls who were free of an ALS diagnosis on the date of selection were randomly selected from the study base and were individually matched to the controls on year of birth, sex, and region of residence. This individual matching allows for a more efficient control of these sociodemographic characteristics.

Depression. Depression was ascertained using outpatient ICD codes (ICD-10 F32, F33, F341), and antidepressant use was ascertained from prescription information within the Swedish Prescribed Drug Registers. Users of antidepressants were defined as individuals with ≥2 dispenses of antidepressants.

Statistical analysis. To examine the association between depression and subsequent risk of ALS, conditional logistic regression models were used to estimate odds ratios and 95% confidence intervals (CIs). Conditional logistic regression allows for the matched design to be factored into the analysis. To examine the association of ALS with subsequent risk of depression, conditional Cox regression (survival analysis) was used to estimate hazard ratios and corresponding 95% CIs. Both approaches (conditional logistic regression and survival analysis) can be used to control for multiple confounders (such as educational level and socioeconomic status as in this study) simultaneously.

RESULTS A total of 1,752 ALS cases and 8,760 controls were included in this study. Depression (either defined as a clinical diagnosis or by antidepressant use) was associated with a higher risk of ALS. Namely, within 1 year after depression diagnosis, there was a 3.6-fold increased risk of ALS (95% CI 2.2–5.8). The risk of ALS decreased as the interval between depression and subsequent ALS increased. Specifically, after more than 3 years following depression diagnosis, the risk of ALS was 0.9 (95% CI 0.6–1.4). A similar pattern was observed for antidepressant use. In examining ALS and the subsequent risk of depression, patients with ALS had a higher risk of depression diagnosis in the year after ALS diagnosis (hazard ratio 6.7, 95% CI 3.9–11.5) that declined in the second year after diagnosis. Further adjustments for education and socioeconomic status (occupation) did not appreciably alter any of the noted effect sizes.
Roos et al. have brought to attention the risk of depression before and after diagnosis with ALS, and some of the implications of these data:

1. Depression may be a prodromal symptom of ALS.
2. Symptoms of depression may overlap with those of cognitive impairment, leading to potential misclassification of these 2 diagnoses.
3. Depression might be a result of psychological distress experienced by patients with ALS between the first symptom onset and the final diagnosis.
4. Depression may be a reaction to the ALS diagnosis.
5. This study may have importance for clinical trial design. Stress and depression may affect study enrollment. Although this has not been examined specifically in an ALS population, ALS depression could factor into enrollment success and retention in clinical trials.

Strengths of this study. There are several strengths of this study related to its study design and research question:

1. This is the largest study regarding number of cases of ALS depression (before and after diagnosis) to date.
2. The cases come from a defined and completely enumerated population allowing control selection to be completely random and thus free from selection bias.
3. Data were collected prospectively and therefore free from recall bias.
4. The exposure and outcome data are collected independently and uniformly for the entire population, thus avoiding biases inherent in pulling information from different sources.
5. An important strength of this study is the ascertainment of cases of ALS using hospital discharge data. Bias could result if the diagnostic accuracy of ALS was low. A study of 2,650 cases in the Danish National Patient Register showed a high validity of ICD codes for capturing ALS diagnoses. While slight bias toward the null could occur with a small number of incorrectly identified cases, bias away from the null would only occur if the misidentification of cases was strongly related to the exposure of interest—which would likely not be the case here since the case identification procedures were blind to exposure status.
6. Another important strength of this study is the ascertainment of depression using both a clinical diagnosis and antidepressant dispenses (≥2).

Study limitations. Weaknesses of the study are as follows:

1. There were a limited number of confounding factors examined in this study. As is generally an issue with the use of administrative data, limited clinical information was available. For example:
   a. A potential confounder is frontotemporal dementia, especially for ALS associated with C9ORF72 expansion mutations. C9 mutations can lead to psychiatric symptoms years before presentation of ALS or frank frontotemporal dementia.
   b. Other factors not examined in the present study are pain and head trauma, each of which is associated with both depression and ALS.
   c. They did not capture cigarette smoking. Although the evidence is not completely consistent, there is some evidence for smoking as a risk factor for ALS. To assess whether confounding by cigarette smoking may be occurring using administrative data, other outcomes could be used either as proxies for smoking or for use in a negative control outcome approach, as is done in other studies using registry data.
   d. This study did not differentiate familial from sporadic ALS. It is possible that some exposures may not affect risk of ALS among subjects with a familial form of ALS. However, familial forms of ALS only account for 5% to 10% of all ALS cases.
2. There were no data before start of the different registries. The absence of data from before a given registry started could lead to exposure misclassification to the extent that a study subject was exposed (e.g., took antidepressants) before the start of the registry. However, any such exposure misclassification would be expected to be nondifferential and therefore, if anything, bias true associations toward the null.

Despite these concerns, is the calculated risk relevant to current neurologic practice? Indeed, the most consistently established nongenetic risk factors for ALS are age and male sex, and both were accounted for in this study. This, in combination with the other major strengths of this study, make this an important contribution to ALS epidemiology and ALS clinical practice.

AUTHOR CONTRIBUTIONS
Dr. Cragg: drafting/revising the manuscript, interpretation of data. Dr. Seals: drafting/revising the manuscript, interpretation of data. Dr. Cashman: drafting/revising the manuscript, interpretation of data. Dr. Weisskopf: drafting/revising the manuscript, interpretation of data.

STUDY FUNDING
Dr. Cragg is an ALS Canada Tim E. Noel postdoctoral fellow.
DISCLOSURE
The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

REFERENCES
Mystery Case

Interesting teaching cases submitted to the Resident & Fellow Section are chosen by the editors to be published under the new Mystery Case subcategory. The *Neurology* editorial office disseminates a teaser through social media before the case is published. This usually includes a short description of the case, video or partial figure, and one to three questions. Responses are compiled and then published with the full case.
**Mystery Case: Neurocutaneous melanosis with diffuse leptomeningeal malignant melanoma in an adult**

**SECTION 1**

A 35-year-old man presented with a history of headache, vomiting, and visual blurring of 6 months’ duration. Two months after the onset of symptoms, he developed behavioral changes in the form of irrelevant talking. He was then seen at another center where the clinical possibility of tuberculous meningitis was considered. His CSF findings at this time were 4 cells, all lymphocytes, protein 221 mg/dL (2.21 g/L), and glucose 59 mg/dL (3.3 mmol/L). A non-contrast CT head showed hydrocephalus (figure, B). He was treated with antitubercular treatment (ATT) and oral dexamethasone. He improved transiently but worsened again and presented to us with increasing headaches, recurrent vomiting, deterioration in vision, alteration in behavior, psychosis, and bilateral lower limb weakness of 10 days’ duration.

General physical examination revealed the presence of a large nevus over the trunk and back in a bathing suit distribution, which he had since birth (figure, A). The lesion had been considered as a birthmark and had never been biopsied. The patient was conscious and oriented with intermittent behavior changes in form of restlessness and agitation.

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From the Departments of Neurology (R.B., V.K., D.V., K.P.), Cytopathology (A.K., S.M.), Neuroradiology (A.G.), and Oncology (S.B.), All India Institute of Medical Sciences, New Delhi, India.

Go to Neurology.org for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.
There were no meningeal signs. His visual acuity was finger counting at 1 meter in both eyes and fundus examination revealed bilateral papilledema. Ocular movements and other cranial nerve examinations were normal. He had paraparesis with proximal more than distal weakness and attenuated lower limb reflexes. The plantar response was flexor. Sensory examination was normal. There were no cerebellar signs.

**Questions for consideration:**
1. What is the differential diagnosis?
2. What will be the next step for a definitive diagnosis?

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**SECTION 2**

The patient’s history of headache, vomiting, and papilledema were suggestive of raised intracranial pressure. Lack of fever, no CSF pleocytosis, high protein, mild hydrocephalus, nonresponsiveness to ATT, and worsening neurologic status raised the possibility of an alternative diagnosis of raised intracranial pressure and meningeal disease other than tubercular meningitis. Since examination revealed a large congenital nevus, the possibility of either a malignant melanoma with intracranial dissemination or neurocutaneous melanosis with malignant meningitis was strongly considered. For an etiologic workup, brain imaging, lumbar puncture, and a skin biopsy were performed.

A noncontrast CT scan of the brain showed hydrocephalus (figure, B). Noncontrast T1-weighted MRI brain (figure, C) and spine (figure, G) revealed diffuse hyperintensity along sulcal spaces of bilateral cerebral hemispheres (arrows) and spinal cord surface (arrows). Postgadolinium T1-weighted brain MRI revealed leptomeningeal and pachymeningeal enhancement (figure, D, arrows) and fat-suppressed postgadolinium T1-weighted MRI spine showed diffuse enhancement along the spinal subarachnoid space (figure, H, arrows). Lumbar puncture revealed a high opening pressure of 300 mm of CSF. There was no pleocytosis, protein was 339 mg/dL (3.39 g/L), and glucose was 26 mg/dL (1.44 mmol/L) (corresponding blood glucose 104 mg/dL [5.77 mmol/L]). CSF cultures for bacteria, fungi, and viruses were negative. CSF for cryptococcal antigen, acid-fast bacilli, GeneXpert MTB/RIF, and PCR for *Mycobacterium tuberculosis* were negative. CSF cytology (figure, E and F) revealed several atypical cells with moderate to abundant cytoplasm and large vesicular nuclei with prominent macronucleoli. Many of these cells contained melanin pigment in their cytoplasm. Binucleated cells were seen. Frequent mitotic figures were identified. On immunocytochemistry, these malignant cells were immunopositive for S-100, HMB-45, and Melan-A, confirming their melanocytic nature. The skin biopsy showed features of pigmented intradermal nevus but no evidence of malignancy was observed.

Based on clinical, CSF, and MRI findings, the patient was diagnosed with neurocutaneous melanosis (NCM) with diffuse leptomeningeal malignant melanomatosis (leptomeningeal melanoma). A ventriculoperitoneal shunt was placed owing to worsening hydrocephalus and headaches. He was started on definitive treatment for NCM in the form of oral temozolomide (dose 200 mg/m² for 5 days) every 4 weeks. A recent assessment on follow-up showed worsened neurologic status and no clinical improvement.

**Questions for consideration:**
1. What is NCM and malignant melanoma? Are they the same entities?
2. How does one differentiate NCM from infective meningitis?
SECTION 3
NCM is a noninherited disorder and was first described by Rokitansky in 1861 and the term NCM was coined by Von Bogaert in 1948. This is one of the pigmented disorders of the nervous system that include focal, diffuse, benign, or malignant pathologies, either arising primarily within the nervous system (neurocutaneous melanosis, meningeal melanocytoma, primary leptomeningeal melanomatosis, and melanoma) or disseminating from a systemic lesion. Our patient fulfilled the revised criteria of NCM laid down by Kadonaga and Frieden which include a large nevus (>20 cm in adults, 6–9 cm in infants) or multiple nevi (at least 3) in association with CNS melanosis or melanoma, no evidence of a cutaneous melanoma except in patients with no evidence of brain melanoma, and no evidence of meningeal melanoma except in patients without cutaneous malignant lesions.

NCM is thought to represent a phakomatosis, which possibly results from an abnormality in the development of neural crest–derived melanocyte precursors, or melanoblasts of the skin and leptomeninges; a form of neuroectodermal dysplasia. Malignant transformation in the CNS occurs in about 40%–50% of cases of NCM, with grave outcomes. It has been proposed that melanin-containing cells are either derived from neural crest–derived nerve sheath precursor cells and migrate to skin via the nerves, paraspinal ganglia, blood vessels, and adnexal tissue or they are neural crest cells that have been genetically or phenotypically altered as melanin-containing cells. One hypothesis suggests that in patients with NCM, the migration occurs along the pia-arachnoid during meningeal invagination of blood vessels during the formation of perivascular spaces.

NCM has been reported to manifest most commonly in childhood, generally within the first 2 years of life, and less commonly in adults. So far, only 12 adult cases have been described in the literature, of which only 3 had leptomeningeal melanocyte infiltration. The condition can be asymptomatic or symptomatic in its presentation and no sex or racial predilection is reported. The diagnosis in adults may be delayed or missed owing to late onset of symptoms. The neurologic presentation in these cases can be variable depending upon the age, site, and extent of involvement, with features of obstructive hydrocephalus, headache, vomiting, neuropsychiatric manifestations, seizures, ataxia, intracerebral hemorrhage, or, less commonly (about 20%), spinal cord involvement with arachnoiditis, syringomyelia, or radiculopathy. Imaging using MRI and CT scan can reveals features of NCM. MRI is the optimal method of imaging. Leptomeningeal disease characteristically presents as hyperintensity on unenhanced T1-weighted images, usually hypointense on T2-weighted sequences, hyperintense on fluid-attenuated inversion recovery images, and shows diffuse enhancement following gadolinium injection. However, melanin deposits without signal abnormality on unenhanced MRI may also be seen. The imaging features are believed to be derived from the paramagnetic effect of the melanin, which shortens the T1 and T2 relaxation times, possibly reflecting interactions of unpaired electrons in melanin with water protons. The leptomeningeal involvement in NCM is different from that seen in primary melanoma of the meninges. In NCM, the dura mater is not typically affected, but involvement of the cerebral parenchyma, choroid plexus, and ventricular ependyma has been observed.

Although histopathology with positive meningeal biopsy is required for definitive diagnosis, a positive CSF cytology can help confirm the diagnosis. Our patient had a positive CSF cytologic specimen for malignant cells confirming the diagnosis. Anti-melanoma antibody (HMB-45) positivity (as seen in CSF cytology in our case) is observed in primary melanocytic lesions of the nervous system. Treatment options for patients with NCM are restricted to radiotherapy, chemotherapy, or CSF diversion strategies. Prognosis remains poor and therapies offer little benefit. The overall prognosis of diffuse leptomeningeal disease limits survival to less than 3 years.

This rare case suggests that adult patients presenting with neurologic symptoms and benign congenital dermal nevi may harbor NCM and herald a malignant leptomeningeal disease. Careful investigation and a high index of suspicion are mandatory for a correct diagnosis.

AUTHOR CONTRIBUTIONS
Rohit Bhatia: manuscript editing and writing. Vijay Kataria: manuscript writing and editing. Deepthi Vihla: manuscript editing. Aanchal Kakkar: cytology review, manuscript editing. Kameshwar Prasad: manuscript editing. Sandeep Mathur: cytology review, manuscript writing and editing. Ajay Garg: radiology review, manuscript editing. Sameer Bakhshi: oncology review and manuscript editing.

STUDY FUNDING
No targeted funding reported.

DISCLOSURE
The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

REFERENCES

MYSTERY CASE RESPONSES
The Mystery Case series was initiated by the Neurology® Resident & Fellow Section to develop the clinical reasoning skills of trainees. Residency programs, medical student preceptors, and individuals were invited to use this Mystery Case as an educational tool. Responses were solicited through a group e-mail sent to the American Academy of Neurology Consortium of Neurology Residents and Fellows and through social media.

Of the 14 respondents to this mystery case, many suggested potential diagnoses, but only 4 respondents first described the cutaneous and neuroimaging findings presented, which would be a key step in working through an uncommon case of this nature. Of these 4 respondents, 2 correctly identified the patient’s cutaneous abnormality as a large congenital nevus, 2 identified the neuroimaging finding of hydrocephalus, and 3 (75%) identified the presence of meningeal hyperintensity and enhancement. Of all the respondents, 6 (43%) offered the correct diagnosis of NCM. Another 36% suggested metastatic melanoma, including metastasis to the meninges, or another neurocutaneous syndrome like primary leptomeningal melanoblastosis. Indeed, this patient did have a diffuse leptomeningeal melanoma in addition to the congenital NCM.

This patient was initially misdiagnosed with tuberculous meningitis and the skin finding was overlooked. This case highlights the enduring relevance of appropriate exposure of the patient during examination as part of the diagnostic process. NCM with leptomeningeal disease is important to consider in patients who present with neurologic symptoms and dermal nevi.

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Thank You R&F Team!
The RFS Editors and editorial staff would like to thank the following team members for their assistance in the compilation of this Highlights Booklet: Jennifer Vermilion, James Siegler, and Malik Adil.
Opinion and Special Articles

These articles provide timely opinions about important areas in neurology education and training. Relevant topics include medical student teaching, training requirements, work/life balance, board certification, and directions in education. Seeking the assistance of senior faculty members is often useful. Those interested in writing these manuscripts should contact the Resident & Fellow Section editor before submission to inquire about the interest in specific topics.
Opinion and Special Articles: “Physician debtor”

ABSTRACT
The increasing cost of attending medical school has contributed to increasing physician indebtedness. The burden of medical school debt has implications for physician career choice, professional satisfaction, and burnout. This opinion discusses the impact of physician indebtedness, the importance of improving debt awareness among neurology trainees, and program- and policy-level solutions to the debt crisis. Neurology® 2016;86:e29–e31

Wilt thou heal others thou thyself full of sores?
—Plutarch

Federally backed US student loan debt principal surpassed $1 trillion in 2013.1 As of 2014, the American Association of Medical Colleges (AAMC) reports median medical school debt at public and private universities to be $170,000 and $200,000, respectively, placing the average medical school graduate in the top 2%ile among all student debt holders.2 According to published AAMC data, between 1998 and 2010 the compounded annual growth rate in the cost of attendance was 4.5% and 5.7% for public and private medical schools, respectively. The graduates shoulder the burden of this cost escalation disproportionately as a deferred liability. Medical education debt levels have increased substantially in comparison to prior decades. Also according to published AAMC data, mean medical educational debt in 1978 when adjusted for inflation was approximately $49,000 (2015 US dollars). By 1998, mean debt had increased to approximately $125,000 (2015 US dollars). The median debt will be lower than mean debt reported here, reflecting the sensitivity of the mean to outliers. There are legitimate concerns about the sustainability of medical student debt in the United States. Medical student debt has been shown to impact a variety of factors ranging from timing of major life decisions3 and specialty choice (and by extension, patient access to care).4 This brief opinion discusses the current state of physician indebtedness, its relevance to neurology trainees and training programs, and some policy- and program-level solutions.

CURRENT STATE
The cost of attending medical school is typically financed through the Department of Education. Loans can be consolidated through the Federal Direct Program and interest rates currently range from 3.4% to 7.9%.5 Assuming AAMC debt figures, the median annual interest expense for servicing this debt is approximately $11,560 and $13,600 for public and private university graduates, respectively. Historically, all student loan interest payments were tax deductible at the federal level, until this benefit was repealed by the Tax Reform Act of 1986. The Taxpayer Relief Act of 1997 restored the student loan interest deduction but with an annual maximum allowable deduction of $1,000. Subsequent legislation increased the maximum allowable annual deduction to where it stands today: $2,500. Furthermore, student debt holders become ineligible to claim the student loan interest deduction at incomes of approximately $75,000 for single filers and $150,000 for joint filers.

MEDICAL EDUCATION DEBT AND SPECIALTY CHOICE
Does educational debt affect choice of medical specialty? The AAMC cites data that suggest that medical education debt does not have an influence on subsequent specialty choice.6 A closer examination of the literature reveals the difficulty determining the influence of debt on the complex process of specialty choice. In one analysis, a 4-variable logistic regression of 5 years of medical school (2 state, 1 private) graduates, educational debt failed to predict specialty choice as a binary outcome: primary care or not primary care. The second study cited was the 1999 questionnaire of 4,500 US women physicians. In this survey, medical school graduation debt failed to predict primary care vs non–primary care specialty choice outcome as a dichotomous variable. Fewer than half of the survey respondents in this study graduated after 1980 and only 5% of those graduates reported educational debt in excess of $100,000 ($178,000, inflation adjusted to

From the Department of Neurology, Mayo Clinic, Rochester, MN.
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particularly relevant for neurology as a specialty.

TAXATION ON PAID INTEREST? The current student loan interest federal tax deduction appears inadequate when compared to contemporary levels of medical student indebtedness. An example illustrates this case. Assume a resident physician with adjusted gross income of $50,000 annually servicing consolidated loans totaling a principal of $170,000 under a modified repayment scheme such as income-based repayment (IBR) or income-contingent repayment (ICR). The IBR and ICR debt management programs defer the large financing costs of educational debt during residency by modifying current monthly payments. Therefore, annual paid interest expense on this debt would total approximately $5,000 annually (and unpaid interest is capitalized). The maximum allowable deduction of $2,500 is reached, leaving $2,500 in additional paid interest that cannot be deducted. The resident is taxed on paid interest. For this resident physician, filing single with tax bracket of 25%, these unrealized deductions cost approximately $700 annually. Following completion of training, physicians are generally ineligible to claim the student loan interest deduction (excluded for incomes greater than $160,000). With a 10-year repayment schedule of $185,000 in principal, a conservative estimate of total interest expense is $100,000. The true financing cost would likely be higher because of interest capitalization after residency and graded repayment schemes. The physician cannot deduct any paid interest. At a 28% tax bracket, this amounts to an extra tax bill of at least $28,000 over the term of loan in this best-case scenario. The true economic cost of the inability to fully deduct student loan interest and the associated finance expense of contemporary educational debt levels are not fully realized until the opportunity cost is appreciated. Because this cost is absorbed at the beginning of a physician’s career, there is a substantial loss of lifetime earnings. Based on current AAMC data for median educational debt, a conservative estimate of financing costs for medical education is approximately $100,000. Assume a 35-year career with this figure accruing at an interest rate of 5% compounded annually. At retirement, this initial amount would be worth over a half million dollars without any further contribution to the principal. The income lost to taxation on paid interest itself alone would be worth over $150,000 under this conservative estimate. A contrarian opinion might view these concerns as negligible, citing low medical school admission rates that reflect large, competitive applicant pools, extremely low educational debt default rates for medical school graduates, and published salary data that support physicians being among the highest paid professionals in America. While valid, these mitigating factors should not justify continued inflation of medical school tuition, ignore the growing long-term indebtedness of mid-career physicians, nor excuse a continuation of unfair tax policy.

POLICY REMEDY Indebted resident physicians and young attending physicians should be aware of recent legislation introduced in the 114th Congress addressing student loan interest and debt repayment. The first bill, HR 509, The Student Loan Interest Deduction Act of 2015, seeks to double the maximum allowable federal student loan interest deduction to $5,000 for single filers and $10,000 for joint filers where both parties have student loan debt. The second bill, HR 1352, Student Loan Borrowers’ Rights Act of 2015, outlines broad expansions in loan management and repayment assistance. Three key items in this legislation are (1) the exclusion from taxable income any forgiven debt (both principal and interest); (2) the removal of educational debt from the list of loans that are not dischargeable in bankruptcy; and (3) amending the Public Service Loan Forgiveness program to forgive 50% of the balance of eligible loans after 60 payments. It is critically important that all medical students, residents, and young indebted physicians familiarize themselves with this legislation and contact their legislative offices to voice support of these bills’ passage.

EDUCATING TRAINEES ON THE RISKS AND MANAGEMENT OF STUDENT DEBT Medical students and resident physicians are likely well aware of the magnitude of their indebtedness. However, they may not appreciate the impact of indebtedness on their nonfinancial well-being, or be prepared to manage the financial implications of indebtedness.
The exploding array of medical knowledge and clinical skills that trainees are expected to master during the period of medical training often crowds out other areas of professional interest. Recognizing the numerous competing demands for trainees’ time, undergraduate and graduate medical training programs should seek to educate students and residents on the implications of accumulation of debt, strategies to manage indebtedness, and tools to recognize the nonfinancial impact of high levels of student debt. The resources required to integrate this education into existing programs could be considerable, possibly requiring internal or external financial expertise, and would likely require recognition and support at the institutional level.

DISCUSSION
There are no signs that physician indebtedness is improving, and in fact there are concerns regarding GME funding and physician burnout that could exacerbate the problem. Undergraduate and graduate medical education leadership should be aware of these factors, and develop strategies to help physicians manage debt early in their careers. Currently, policy solutions exist but will require physician and resident advocacy to enact meaningful change.

AUTHOR CONTRIBUTIONS
Eugene L. Scharf: drafted and revised the manuscript, design and conceptualization. Lyell K. Jones, Jr.: drafted and revised the manuscript, design and conceptualization.

STUDY FUNDING
No targeted funding reported.

REFERENCES

DISCLOSURE
The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.
Pearls & Oy-sters

“Pearls and Oy-sters” is a feature focusing on fundamental clinical neurology. Each article addresses a specific niche of neurological disease and provide expertise in the form of clinical insights and tips, i.e., “pearls,” as well as advice for avoiding mistakes, or “oy-sters.” The author may choose to address a particular facet of the approach to neurological disease such as localization, elaboration of a differential diagnosis, evaluation, or treatment. These articles concentrate on what may be found in a textbook and/or provide what textbooks cannot, in the form of knowledge rendered from clinical experience. The target audience consists of those in training; however, the subject matter should be of interest to all in the world of clinical neurology.
Pearls & Oy-sters: Episodic ataxia type 2
Case report and review of the literature

PEARLS

• Episodic ataxia type 2 (EA2) is an autosomal dominant calcium channelopathy caused by a mutation in CACNA1A.

• Spells are characterized by ataxia, which may be accompanied by vertigo, diplopia, dysarthria, and generalized weakness.

• Between spells, patients often demonstrate persistent nystagmus.

• Acetazolamide and 4-aminopyridine are reported to decrease severity and frequency of spells.

OY-STERS

• Patients may not report a family history of episodic ataxia or familial hemiplegic migraine.

• Patients who initially have episodic symptoms may develop a progressive ataxia syndrome.

• Coexisting functional disorder may make evaluation more difficult.

CASE REPORT

A 38-year-old man presented to our clinic for evaluation of increasingly frequent spells of gait instability, diplopia, vertigo, and dysarthria. These spells began at 12 years of age. Initially, he experienced isolated periods of vertigo without accompanying symptoms. He told no family or friends of his problem at the time. Two years later, the symptoms progressed, manifesting as semistereotyped episodes characterized by a shock-like sensation radiating from his neck down his back followed by binocular diplopia, vertigo, gait instability, mental clouding, and nausea lasting 2–8 hours. Later, he experienced dysarthria during spells. Interictally, he returned to his baseline except for a persistent mild diplopia.

The patient concealed his symptoms for many years. His mother first witnessed a spell when he was 27 years old. She found him disoriented and imbalanced. He was carried to the emergency department, where he received diazepam for anxiety, and his symptoms as usual resolved within 1–2 hours. He continued to use diazepam for future spells, which physicians variably diagnosed as seizures, anxiety attacks, TIA, a functional disorder, or malingering (table 1). He was empirically treated with phenytoin without improvement. He underwent closure of a patent foramen ovale for possible recurrent TIA. Ultimately, he was diagnosed with a functional disorder, and no further workup was conducted for 10 years. The frequency of these spells varied throughout his life, occurring daily, monthly, or even yearly. Then, following the unexpected death of a family member 2 months before our initial evaluation, his symptoms became nearly continuous. He had no family history of episodic neurologic symptoms, gait impairment, migraines, or epilepsy. He had a history of remote methamphetamine use and ongoing tobacco and marijuana use. He had normal motor and social development, but language and cognitive development were delayed. His speech was incomprehensible as a child, prompting childhood speech therapy and special education through 8th grade, when he dropped out of school.

During physical examination, he did not experience a typical spell. Much of his neurologic examination was elaborated, requiring several attempts with coaching to accurately perform routine tests of function unrelated to his symptoms. For the majority of his neurologic testing, he ultimately performed the maneuvers without deficit over a short time interval. Notable findings on his examination included bilateral esophorias, hypometric saccades with catch-up bilaterally, bilateral gaze-evoked nystagmus, mild dysthria, and overshoot with finger follow bilaterally. While seated he had truncal titubation, and on gait testing he had a wide-based stance and significant difficulty with tandem walking. Gait and posture were markedly irregular and variable, presumed to represent a functional overlay on what was likely a mildly abnormal gait. Finally, his cognitive evaluation showed mild to moderate impairment in frontal-executive, episodic memory, visuospatial, and language tasks. His Montreal Cognitive Assessment score was 23 out of 30.

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Go to Neurology.org for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.
Laboratory evaluation had previously excluded metabolic, endocrine, nutritional, infectious, autoimmune, and paraneoplastic etiologies of ataxia. His CSF revealed a normal protein level, leukocyte count, immunoglobulin G index, and no unique oligoclonal bands. Brain MRI the year before our evaluation was reported as normal although in retrospect, mild midline cerebellar atrophy was likely present. Cervical and thoracic MRI showed no spinal cord abnormalities. He did not undergo EMG. Genetic testing for episodic ataxia type 2 (\textit{CACNA1A}) demonstrated a heterozygous, single base pair deletion in \textit{CACNA1A} causing a pathologic frameshift mutation. He was started on acetazolamide for symptomatic therapy.

**DISCUSSION** The first published clinical description of EA2 dates back to 1946 while the molecular basis of the disorder was identified in 1996 with the discovery of mutations in the P/Q type voltage-gated calcium channel \textit{CACNA1A}. It is an autosomal dominant disorder and the most common inherited episodic ataxia, though the prevalence is estimated at less than 1 in 100,000 with high though incomplete penetrance.\(^1,2\)

Clinically, the disorder is characterized by ataxic spells lasting hours to days, often with interictal nystagmus; however, there is considerable variation in disease presentation.\(^3\) The spells may be characterized by isolated ataxia or, as seen in our patient, a broader range of symptoms, often localizing to the brainstem. Associated features include generalized and hemiplegic weakness, migraine, intellectual disability, dystonia, and seizures. Indeed, there is a broad range of neurocognitive deficits associated with \textit{CACNA1A} mutations, although until recently this has been largely unexplored.\(^3\) While initially an episodic disorder, some patients will develop a secondarily progressive ataxia, as in our patient. Notable triggers include physical and emotional stress. Disease onset is typically between 5 and 20 years of age.\(^3\) It is clinically differentiated from other episodic ataxias through a variety of features including age at onset, spell duration, interictal nystagmus, and genetic locus (table 2).\(^2,4,5\)

Genetically, the disorder has been linked to \textit{CACNA1A}, which encodes the pore-forming subunit of the P/Q-type voltage-gated calcium channel.\(^1\) The P/Q channel is expressed throughout the CNS, but is most densely expressed in cerebellar Purkinje cells and granule layer neurons. It is found principally on presynaptic terminals and plays a key role in synaptic transmission.\(^6\) Over 80 different pathologic mutations have been identified to date, and they typically result in premature truncation of the protein via nonsense or frameshift mutations though a number of missense mutations have also been found to be pathologic. Based on these observations, EA2 is believed to be characterized by loss of P/Q channel function in the cerebellum. This hypothesis is supported by electrophysiologic studies that demonstrate that EA2-associated mutations result in loss of or diminished channel function when expressed in vitro.\(^7\) Research

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Differential diagnosis for episodic ataxia</th>
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<td><strong>Central causes</strong></td>
<td><strong>Peripheral causes</strong></td>
</tr>
<tr>
<td>TIAs</td>
<td>Benign paroxysmal positional vertigo</td>
</tr>
<tr>
<td>Migraine</td>
<td>Labyrinthitis</td>
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<tr>
<td>Presyncope</td>
<td>Meniere disease</td>
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<tr>
<td>Multiple sclerosis</td>
<td>Acoustic neuroma</td>
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<tr>
<td>Toxic (alcohol, antiepileptics)</td>
<td>Perilymphatic fistula</td>
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<tr>
<td>Seizures</td>
<td>Immune-mediated inner ear disease</td>
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<td>Chiari type 1 malformation</td>
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<td>Atlantoaxial abnormalities</td>
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<td>Paroxysmal dyskinesias</td>
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<td>Functional disorders</td>
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<tr>
<th>Table 2</th>
<th>Clinical features of the episodic ataxias (EA)</th>
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<tbody>
<tr>
<td><strong>Gene/chromosomal location</strong></td>
<td><strong>Age at onset</strong></td>
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<tr>
<td>EA1</td>
<td>KCNA1</td>
</tr>
<tr>
<td>EA2</td>
<td>CACNA1A</td>
</tr>
<tr>
<td>EA3</td>
<td>Chromosome 1q42</td>
</tr>
<tr>
<td>EA4</td>
<td>Unknown</td>
</tr>
<tr>
<td>EA5</td>
<td>CACNB4</td>
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<tr>
<td>EA6</td>
<td>SLC1A3</td>
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also suggests that the mutation may exert a dominant negative effect on the wild-type channel by interfering with proper channel folding and trafficking rather than simple haploinsufficiency. None of these explanations, however, clearly accounts for the unmistakably episodic nature of the disease. 

Interestingly, mutations in \textit{CACNA1A} have also been identified in familial hemiplegic migraine type 1 and spinocerebellar ataxia type 6 (SCA6). Thus, EA2 is allelic with these disorders and there is recognized clinical overlap. Familial hemiplegic migraine is reliably associated with a wide range of missense mutations. In SCA6, a CAG repeat expansion in the C-terminus of the gene is believed to cause cerebellar degeneration. These and other mechanisms of disease in P/Q-type channel dysfunction have been well reviewed previously. 

Our patient was found to have a novel single base pair deletion (3,535C) leading to a frameshift mutation in exon 20 of \textit{CACNA1A} resulting in a premature stop codon and aberrant mRNA likely destined for nonsense mediated decay. He has no family history of the disease, though nonpaternity was not explored. He has 3 children, all of whom are under age 20 and are currently healthy with no neurologic symptoms, including migraine and intellectual disability. 

Therapeutically, acetazolamide responsiveness is a hallmark of the disease. Though there are patients who do not have symptomatic benefit, an estimated 50%–75% of patients report improvement in episode severity and frequency with acetazolamide doses ranging from 250 to 1,000 mg daily. Therapeutic benefit was seen in our patient. He developed nephrolithiasis while on acetazolamide 1,000 mg/d and discontinued the medication; however, after his genetic diagnosis, the medication was restarted at a lower dose with close clinical monitoring. For those who cannot tolerate acetazolamide, the potassium channel blocker 4-aminopyridine at doses of 5 mg 3 times daily has also been shown to decrease episode frequency and improve quality of life in a randomized controlled trial of 10 patients.

Finally, there are no earlier reports of functional disorders among patients with EA2. Comorbid functional disorders create a diagnostic challenge. In patients with psychogenic nonepileptic seizures, time to correct diagnosis is estimated to be 7 years from onset. Our patient’s experience would suggest that equally lengthy diagnostic delays are possible in EA2 when comorbid functional disorder is present. Importantly, this case also highlights the fact that functional overlay can lead to premature diagnostic closure after the symptoms are labeled as such. Improved recognition and understanding of episodic ataxias is necessary. EA2 must be considered for any patient with episodes of ataxia lasting hours, although shorter and longer attacks do occur, and more rarely in patients who present with a progressive ataxia syndrome.

\textbf{AUTHOR CONTRIBUTIONS}

Dr. Guterman: drafting of manuscript. Dr. Yurgionas: drafting of manuscript. Dr. Nelson: critical revision of the manuscript for important intellectual content.

\textbf{STUDY FUNDING}

No targeted funding reported.

\textbf{DISCLOSURE}

E. Guterman and B. Yurgionas report no disclosures relevant to the manuscript. A. Nelson is funded by NIH grant 5K08NS081001, serves as the Richard and Shirley Cahill Endowed Chair in Parkinson’s Disease Research, and has received honoraria for speaking at the American Academy of Neurology annual meeting and Stanford Research Institute International. Go to Neurology.org for full disclosures.

\textbf{REFERENCES}

Residency Training

These manuscripts will address issues related to residency training, including educational initiatives, programs, opinions, and other topics related to neurology education and training. Relevant topics could include work hours and sleep deprivation, the role of neurocritical care or outpatient neurology in training, quality assurance initiatives, incorporation of evidence-based neurology into training, medical student teaching, work/life balance, and others. Seeking the assistance of senior faculty members is often useful.
Residency Training:
Work engagement during neurology training

ABSTRACT
Objective: Work engagement, defined as a positive, fulfilling, work-related state of mind that is characterized by vigor, dedication, and absorption, can ameliorate patient care and reduce medical errors. The purpose of this cross-sectional study was to investigate work engagement among neurology residents in the region of Attica, Greece.

Methods: In total, 113 residents participated in this study. Demographic and work-related characteristics, as well as emotional exhaustion and personality traits (neuroticism), were examined via an anonymous questionnaire. Work engagement was measured by the Utrecht Work Engagement Scale.

Results: The study sample had a mean age of 34.6 ± 3.6 years, ranging from 26 to 45 years. Sixty-two (54.9%) participants were women and 45 (39.8%) were married. After adjusting for sex, emotional exhaustion, and neuroticism, the main factors associated with work engagement were autonomy and chances for professional development.

Conclusions: Providing more chances for trainees’ professional development as well as allowing for and supporting greater job autonomy may improve work engagement during neurology training.

GLOSSARY
EE = emotional exhaustion; EWTD = European Working Time Directive; JD-R = Job Demands-Resources; UWES = Utrecht Work Engagement Scale.

Work engagement is an important indicator of occupational well-being for both employees and organizations. Work engagement is characterized by a positive motivational state of dedication (i.e., being strongly involved in one’s work and experiencing a sense of inspiration, enthusiasm, and challenge), vigor (i.e., experiencing high levels of energy and mental resilience while working), and absorption (i.e., being fully concentrated and happily engrossed in one’s work). Work engagement can ameliorate patient care and reduce medical errors.

Work engagement is considered to be the positive opposite of another core concept in the field of organizational behavior—burnout (i.e., a state of emotional exhaustion [EE], depersonalization, and cynicism toward work, and reduced professional efficacy in response to chronic stressors at work). However, whereas burnout has been extensively studied across different medical specialties in order for investigators to identify its determinants, work engagement has been relatively neglected.

The evidence regarding the antecedents and consequences of work engagement can be organized in an overall model. According to the Job Demands–Resources (JD-R) model, job resources such as social support from colleagues and supervisors, chances for professional development, performance feedback, and job autonomy can help employees to achieve work goals, reduce job demands (e.g., workload, emotional demands), and stimulate personal growth. Job resources can start a motivational process that may lead to work engagement and consequently to higher performance. Apart from work-related characteristics, personality factors may influence work engagement and can also shape the way employees perceive their work environment and react to it.

The purpose of this cross-sectional study was to study work engagement among neurology trainees.
METHODS Procedure and participants. Details about the procedure and the participants have been published. In summary, all 131 neurology trainees of the wider area of Athens working in 18 hospitals were invited to participate in the study. The study protocol was approved by the local ethics committee.

Measures. Demographic characteristics included sex, age, and marital status. Work-related characteristics included stage of training, compliance with the European Working Time Directive (EWTD), and 8 specific JD-R characteristics, based on the JD-R model and assessed via a 31-item validated questionnaire (autonomy, opportunities for professional development, support from colleagues, supervisor support, workload, intellectual demands, emotional demands, work–home demands interference). Each of these items was rated on a 5-point Likert scale, ranging from never to always.

EE, considered as the main aspect of burnout and denoting a sense of being depleted of one’s emotional and physical resources, was assessed via the 9-item EE scale from the Maslach Burnout Inventory. EE reflects the stress dimension of burnout and may be considered the most obvious manifestation of this complex syndrome. Neuroticism (a fundamental personality trait characterized by one’s tendency to experience negative affect such as anxiety, fear, sadness, worry, and tension) was examined via a 12-item scale, based on the NEO–Five Factor Inventory. Work engagement was measured with the Utrecht Work Engagement Scale (UWES).

Statistical analysis. A database was developed using the statistical software package SPSS (version 16.0 for Macintosh; Chicago, IL). Descriptive statistics were examined for each variable. Correlations between the UWES score and baseline continuous variables were examined using Spearman correlations. Mann-Whitney U was used to compare UWES scores between groups. Where statistically significant correlations or differences were found, these variables were entered in a multiple linear regression model in order to examine the relationships between these independent variables and the UWES score (set as the continuous dependent variable). Level of statistical significance was set at 0.05.

RESULTS Full data were available for 113 trainees (response rate 86.3%). Table 1 summarizes the demographic and work-related characteristics of the study sample. Regarding demographic characteristics, the univariate analysis showed that the UWES score did not correlate with age, while the UWES score did not differ significantly between married and unmarried participants. However, women showed higher UWES scores compared to men (64.1 ± 15.8 vs 53.4 ± 23.9, Mann-Whitney U, Z = −2.493, p = 0.013).

Regarding work-related characteristics, the univariate analysis showed that the UWES score did not correlate with the stage of training, the latter defined as the remaining months to complete training, while the UWES scores did not differ significantly between trainees exceeding the EWTD limit and trainees who did not. Correlations between the UWES score and the JD-R variables showed that UWES was significantly correlated with supervisor support (Spearman ρ = 0.326, p < 0.001), home–work demands interference (Spearman ρ = −0.245, p = 0.009), autonomy (Spearman ρ = 0.442, p < 0.001), and chances for professional development (Spearman ρ = 0.597, p < 0.001), but not with social support, workload, or intellectual and emotional demands. Regarding psychological characteristics, the UWES score correlated significantly with both the EE (Spearman ρ = −0.546, p < 0.001) and the neuroticism score (Spearman ρ = −0.249, p = 0.008).

Multivariate linear regression analysis was conducted to find the significant variables associated with work engagement. The following independent variables were entered into the model: sex, EE, neuroticism, supervisor support, home–work demands interference, autonomy, and chances for professional development. Table 2 shows that the only variables that appeared to have unstandardized regression coefficients significantly different from zero in predicting work engagement were sex, EE, autonomy, and chances for professional development. According to the standardized β values, EE and chances for professional development were the most potent variables related to work engagement. All variables in the model accounted for 49.8% of the total UWES variability, which is considered to be high.

DISCUSSION This cross-sectional study involved all neurology trainees of the wider area of Athens, Greece. The novelty of our study is that it was designed to identify factors related to work engagement during neurology residency training.

An interesting finding in our study was that female neurology trainees were more engaged at work compared to men. Contrary to our findings, a recent study on work engagement in psychiatry residents found that men were more engaged at work compared to women. Moreover, in studies investigating work engagement in medical related fields, such as nurses, no significant difference between the 2 sexes has been found. Similarly, although

Table 1 Characteristics of the total sample of the study

<table>
<thead>
<tr>
<th>Demographics</th>
<th>Total sample (n = 113)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, y, mean (SD)</td>
<td>34.6 (3.6)</td>
</tr>
<tr>
<td>Male sex, n (%)</td>
<td>51 (45.1)</td>
</tr>
<tr>
<td>Marital status, n (%)</td>
<td></td>
</tr>
<tr>
<td>Single</td>
<td>67 (59.3)</td>
</tr>
<tr>
<td>Married</td>
<td>45 (39.8)</td>
</tr>
<tr>
<td>Divorced</td>
<td>1 (0.9)</td>
</tr>
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</table>

<table>
<thead>
<tr>
<th>Work-related characteristics</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Months remaining to complete training, mean (SD)</td>
<td>18.1 (10.8)</td>
</tr>
<tr>
<td>Working hours per day (not including on-call duties), mean (SD)</td>
<td>7.1 (1.1)</td>
</tr>
<tr>
<td>On-calls per month, mean (SD)</td>
<td>4.7 (1.7)</td>
</tr>
<tr>
<td>Days-off per month, mean (SD)</td>
<td>2.4 (1.7)</td>
</tr>
</tbody>
</table>
status has been associated with work engagement in cancer workers including nurses, radiation therapists, allied health, and medical staff. These findings suggest that demographic characteristics may possibly play a different role in work engagement in diverse study populations and settings.

Our finding that the UWES score did not differ significantly between trainees exceeding the EWT limit and trainees who did not is in line with previous research, but stands in contrast to those reported by other studies that have found that hours worked per week is positively associated with work engagement in registered nurses. Again, job-related and study population characteristics may contribute to variations between study results.

After having adjusted for sex, EE, and neuroticism, we showed that the main factors related to work engagement were autonomy and chances for professional development. Both these variables are considered to be job resources and both may help trainees to become skilled specialists. Autonomy provides young doctors with the necessary independence—appropriate to their level of expertise—in managing a patient clinically. Similarly, opportunities for professional development may increase work engagement. Interestingly, this variable has not only been the strongest factor positively related to work engagement, but it has also been found to be the most significant attenuation determinant of burnout. The direct characteristics of the residents’ mentors (including the structure of supervision and the time spent with mentees) were not evaluated in this study. Future research might explore their role in achieving work engagement.

No significant relationship was found between work engagement and neuroticism. This can be explained by the strong relationship between neuroticism and another independent variable introduced in the regression analysis, exhaustion. We acknowledge as a limitation of our study the fact that we have not examined the role of additional personality traits (e.g., extraversion, conscientiousness) or low-order individual factors (e.g., optimism, self-efficacy) that may function as personal resources. Future studies need to examine the role of these factors in achieving work engagement.

Given that work engagement is characterized by a high level of energy and strong identification with one’s work, it was not unexpected that a strong and significant negative relationship was found between work engagement and EE. The present study emphasized the inherently motivational qualities of engagement that foster residents’ willingness to dedicate their efforts and abilities to the work task.

Our results shed light on some significant parts of training in neurology, where, apart from gaining clinical experience, trainees are in need of having autonomy in their daily practice, along with the appropriate supervision. Open-minded clinical teachers and mentors should help residents to professionally develop further skills and have more chances for professional development. Organizational-level interventions may focus on increasing job resources by redesigning the work environment or through training. Improving work engagement is important for both young doctors and patients, as the latter will receive better quality of care.

AUTHOR CONTRIBUTIONS
Panagiotis Zis: drafting/revising the manuscript, study concept and design, data collection, statistical analysis, accepts responsibility for conduct of research and final approval. Artemios K. Artemiadis: drafting/revising the manuscript, data collection, accepts responsibility for conduct of research and final approval. Fotios Anagnostopoulos: drafting/revising the manuscript, study concept and design, accepts responsibility for conduct of research and final approval.

ACKNOWLEDGMENT
The authors thank Dr. Seraphim Karavas, Dr. Maria Lykourou, Dr. Sophia Xiroou, Dr. Andromachi Roussopoulou, Dr. Ermoni Papageorgiou, Dr. Elena Bakola, Dr. Panagiotis Iliopoulos, and Dr. Ioannis Stavropoulos for their contribution to data collection, and all the participants in the study.

STUDY FUNDING
No targeted funding reported.

<table>
<thead>
<tr>
<th>Predictors</th>
<th>B</th>
<th>SE</th>
<th>p</th>
<th>t</th>
<th>p</th>
<th>95% CI for B*</th>
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</thead>
<tbody>
<tr>
<td>Sex</td>
<td>-6.082</td>
<td>2.931</td>
<td>-0.148</td>
<td>-2.075</td>
<td>0.040</td>
<td>-11.895 to -0.270</td>
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<tr>
<td>Emotional exhaustion</td>
<td>-0.458</td>
<td>0.157</td>
<td>-0.267</td>
<td>-2.924</td>
<td>0.004</td>
<td>-0.769 to -0.148</td>
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<tr>
<td>Neuroticism</td>
<td>-0.317</td>
<td>0.246</td>
<td>-0.099</td>
<td>-1.288</td>
<td>0.201</td>
<td>-0.806 to 0.171</td>
</tr>
<tr>
<td>Home-work demands interference</td>
<td>0.132</td>
<td>0.313</td>
<td>0.031</td>
<td>0.422</td>
<td>0.674</td>
<td>-0.489 to 0.754</td>
</tr>
<tr>
<td>Supervisor support</td>
<td>0.607</td>
<td>0.318</td>
<td>0.142</td>
<td>1.908</td>
<td>0.059</td>
<td>-0.024 to 1.237</td>
</tr>
<tr>
<td>Autonomy</td>
<td>1.559</td>
<td>0.572</td>
<td>0.205</td>
<td>2.725</td>
<td>0.008</td>
<td>0.425 to 2.693</td>
</tr>
<tr>
<td>Chances for professional development</td>
<td>2.020</td>
<td>0.564</td>
<td>0.289</td>
<td>3.582</td>
<td>0.001</td>
<td>0.902 to 3.138</td>
</tr>
</tbody>
</table>

Abbreviation: CI = confidence interval.
*Unstandardized coefficients.
*Standardized coefficients.
*p < 0.05.
REFERENCES
Right Brain

Right Brain is a feature devoted to the relationship between neurology and the medical humanities, with submissions either written by trainees or with a focus on the experience of the trainee. Appropriate submissions include articles, commentaries, and reflections on the interaction between neurology and history, literature, ethics, theology, sociology, anthropology, philosophy, poetry, theater, film, fine arts, or the media. Right Brain also will publish original works of fiction, poetry, and reflection written by residents and fellows relating to the practice of neurology or neurology training.
Right Brain: 
Home is where the heart is

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Home. Few 4-letter words can be more haunting, and at times, more impossible, to a physician. “I want to go home.” These were the words that were on 3-second repeat 3 days before my 66-year-old patient died in the intensive care unit (ICU). Whispering in a rumbling murmur, barely audible, my patient’s voice was still fervent in its desire to connect, to inform those in his company of his wish.

For days, we did not know what was wrong with him, or perhaps we knew what was wrong but did not want to admit it. He had chronic lymphocytic leukemia and had already undergone transplant and subsequent rejection. But now he presented with multiorgan failure without an identifiable cause. Antibiotics were not working, dialysis only temporized his progression, and diagnostic tests did not reveal a culprit. However, at the end of a long ICU day, our team knew that he was a patient that we could not cure.

His family huddled in his room, holding vigil over their patriarch, already reminiscing over his ability to make each of them feel that he or she was the most special person in his life. He was loved, adored by his family, coworkers, and friends. He always gave of himself, making it all the more difficult to grapple with the impending loss.

As physicians, we diagnose and treat numerous maladies in our patients, often ignoring the global history of present illness before us. When we encounter a complicated case, we roll up our sleeves, and we systematically dissect their ailments on subway and car rides home. The difficult cases keep us up at night. We wonder if we are missing something.

Yet what we do not often ponder in these late-night contemplations are the human beings in front of us, their hopes, their fears, and most importantly, their perspective. More than goals of care, patients bring to the hospital their view of their underlying illness, often radically different than our own summation.

They may see their complaint as serious, as minor, or at times, may wholly ignore significant symptomatology. They may come to the doctor or to the hospital quickly or may linger at home until the end stage of their illness, when bedrest fails their body. As physicians, our role is to negotiate the muddy waters between often divergent views of their best interest. But more difficult is to align ourselves with their own intuition, ignoring our years of training and deferring to their years of training in the study of their own bodies.

Like a mechanic, we admit the patient to work him or her up and hopefully return the patient as good as new, or as good as baseline, to return to a life already in progress. We may know what is wrong with his or her body, or if not, we can run diagnostics to find the broken part. But what happens if in the process of diagnosis, the patient suffers?

Often, suffering is specifically what the patient seeks to avoid, and is often the reported reason for not coming to a hospital. Largely, the reason for this decision-making algorithm on the part of the patient deals with the fact that the hospital, and in this case, the ICU, is remarkably unlike home. There is no familiar wallpaper, no nostalgic scent emanating from the kitchen, and no neighborhood noises that remind the patient of his or her surroundings. Instead, the patient’s space is invaded by a seemingly ongoing onslaught of unfamiliar staff, inharmonious beeping from throughout the room, and cutaneous bodily injury in the form of fingersticks, blood draws, and IV lines. It is therefore no wonder that many patients avoid the hospital at all costs.

Each time I entered my patient’s room, I wondered what I could say to him, to his wife, to his 2 adult children who lingered at his side. My response, with all I could muster, was “You cannot go home.” Home is where the heart is, or where his heart should have been. But his heart was sustained by 3 bags of pressors; it would not survive the ambulance ride home.

Initially, our team’s goal was to identify the cause of his deterioration, to repair the problem, and to send him on his way. Yet, as time progressed, my goal became synergistic with his own: to send him home as soon as possible, regardless of the eventual clinical outcome.

I wondered if all we had done to extend his life in the hospital was truly caring for him. What are a few
more days in the ICU compared to a day at home? What is the metric conversion of home to ICU? How many high-decibel ICU days equal the silence of home?

I encountered another patient in the neurologic ICU, a 36-year-old man with a newly diagnosed brainstem glioma, who presented for resection. He was just at the beginning of his oncologic journey, and had many options at his disposal. His resection was largely successful with no radiologically visible disease present in his pons. However, he encountered numerous postoperative complications, including significant dysphagia and ventilator dependence.

Given that he was just 36 years old with an essentially incurable disease, our team’s and the patient’s goal was to discharge him home to enjoy as much time as possible with his family. He and his family decided to pursue tracheostomy and percutaneous gastrostomy in order to fulfill this goal. Yet his course was complicated by persistent tube feed aspiration, requiring multiple revisions to his gastrostomy, nearly fatal pneumonitis, and another month in the hospital. He eventually was discharged to a skilled nursing facility. Whether he would improve with subsequent radiation and chemotherapy was not of immediate concern; we knew more inpatient time would not improve his quality of life.

In the case of my first patient, it was not entirely clear to me that we were caring for him. He knew that he had precious few days left, and so did we. He endured a barrage of tests, lines, and treatments meant to care for him, but this diagnostic workup was futile in the end.

In retrospect, I wonder if every member of his health care team—nurses, technicians, medical students, physicians—would have said that he never should have come to the hospital. I imagine that the patient also felt that way. What we gave him was not the gift of life; we did not provide more days, but instead, arguably less quality time with his family. Whatever home represented to him, his ICU room was not that.

In the case of my second patient, we actively tried to send him home right from the first moments of his admission. Though his course of complications was not unexpected given his underlying diagnosis, it is conceivable that he may have chosen not to even undergo the first surgery if only to avoid the subsequent prolonged hospitalization. If he could have seen the future at the beginning of the journey, what future would he have chosen for himself?

These cases are a mere example of numerous instances where personal removal and distance from a difficult health care scenario provides critical benefit. In this case, I wonder, is ICU care really caring? And if not, when should we stop caring, or rather start caring for our patients, sending them home, greatly daring?

As physicians, our own vulnerability is often the barrier to achieving our patients’ goals. Our role in these complex health care decision algorithms is to use our experience to transform patients’ objectives into reality. Our own need for validation in arriving at the correct diagnosis is often at odds with a patient’s desire to return home. Thus, the most appropriate use of our time in caring for our patients may be merely to return to the beginning and ask, “What brings you to the hospital and what do you hope to achieve from your stay?” Receiving answers to these simple questions allows us to formulate a plan to achieve these goals, whether it means imminent discharge or reasonable workup in the framework that they provided.

Empowering our patients to formulate their own plan of care can also restore control at a point in their lives when much of their control has been lost. And placing our patients in control may allow us to let go of that critical diagnosis, not to breathe an inward sigh of defeat for not finding the answer, but rather to reap joy by focusing our attention on the centerpiece of the case: the patient. As providers, deferring to the vision of our patients provides comfort, satisfaction, and reassurance that whatever the clinical outcome, our patients chose for themselves how to spend the rest of their days.

Home. Few 4-letter words are now as meaningful to me. We must treat not the illness, but the patient. As I contemplate the oath that launched my career as a physician, to do no harm, I will always remember: a spoonful of home is sometimes just what the doctor ordered.

STUDY FUNDING
No targeted funding reported.

DISCLOSURE
E. Noch reports no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.
Teaching NeuroImages

Teaching NeuroImages are interesting, previously unpublished photomicrographs, patient photographs, neuroradiologic images, or other pictorial material. They are clear examples of established observations intended for the trainee audience. Educational videos may also be submitted under this category (Teaching Video NeuroImages). Teaching NeuroImages and Teaching Video NeuroImages now feature accompanying ‘Teaching Slides.’ These slides are available online with the article as a teaching tool for trainees and program directors.
Hemigeographic tongue following an acute ischemic stroke

We present 2 patients with a hemigeographic tongue following a left acute hemispheric stroke: a 70-year-old man (figure 1, A–B) and a 43-year-old man (figure 2, A–B). Trigeminal trophic syndrome (TTS) is an unusual complication of trigeminal injury that causes a neuropathic disorder with ulceration of the nasal ala.1,2 Nerve section and brainstem stroke have been described as TTS cause.1,2 However, the presence of a not previously described hemigeographic tongue following a hemispheric acute stroke points toward a central trigeminal disturbance, probably related to a cortical connection lesion. This suggests a complex mechanism in TTS in which supranuclear lesions should also be included.

AUTHOR CONTRIBUTIONS
Montserrat G. Delgado: study concept and design. Sergio Calleja: critical revision of manuscript for intellectual content.

STUDY FUNDING
No targeted funding reported.

DISCLOSURE
The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

REFERENCES
Figure 2  Hemigeographic tongue and MRI

(A) Hemigeographic tongue (white arrow). (B) Cranial MRI shows a lenticular ischemic stroke (white arrow).

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