THE FOURTH ANNUAL HIGHLIGHTS OF THE RESIDENT AND FELLOW SECTION: 2011
A REPRESENTATIVE COLLECTION OF PREVIOUSLY PUBLISHED ARTICLES

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   Mitchell S.V. Elkind

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Meet the Resident & Fellow Editors of Neurology
And learn how you can contribute to the journal
Monday, April 11, 2011, 7:30-9:00 p.m.
At the Residents & Fellows Career Forum
ANNOUNCEMENT

Neurology® Resident and Fellow Section Writing Award

The winner of the 2011 Award is Amy Gelfand, MD

Right Brain: “We were all once ‘fixed and dilated’” published in Neurology on November 16, 2010;75:1851-1852.

Dr. Gelfand will be honored at the 2011 AAN awards luncheon
See page 32 of this Highlights booklet for the award-winning article.

The Neurology® Resident and 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 and 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 2012 and will be awarded for a paper published in 2011.

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

Welcome to this fourth edition of the Highlights of the Resident and Fellow Section of Neurology!

The Highlights of the Resident and Fellow Section provides representative examples of some of the finest articles written for the Section by neurology trainees and educators in the past year.

The Resident and Fellow Section (RFS) has served the trainee readership of the journal and provided a forum for resident writing since January 2004. The Section is trainee-run: a nationally representative team of 12 residents and fellows, each of whom serves 3 years, has responsibility for reviewing, editing, and publishing articles of interest to trainees. Section members also write articles, but manuscript submissions also come from hundreds of trainees, program directors, and educators around the world. Photographs and brief biographies of the Resident and Section Editorial Team follow this introduction.

Publications of the RFS have grown tremendously over the years. The number of submissions to the section has increased dramatically (from 12 in 2004 to 309 in 2010), and the quality of published manuscripts has improved (represented by our current acceptance rate of about 38%). We published 98 manuscripts in 2010, our highest number to date.

The Section has several different subsections, represented by the articles in this booklet. These include Emerging Subspecialties in Neurology, Clinical Reasoning, Right Brain, Child Neurology, Pearls and Oy-sters, International Issues, Education Research and Initiatives, Teaching Neuroimages (including both static images and videos), and Book Reviews. The descriptions of the subsections appear before each sample article.

The group also initiates and develops other unique projects, including podcasts, weekly electronic communications, an annual writing award, Mystery Cases, and new subsection ideas. Podcasts related to articles published in the RFS began in December 2007, for example, and weekly E-Pears, or email “pears,” have been sent to residents nationwide since July 2008. An archive of E-pears can be found at www.aan.com/go/education/residents/epear. The first annual RFS writing award was awarded in April 2009. Our first Mystery Case was published in August 2009. Our website was launched in 2010.

The Group is strongly supported by Neurology’s Editor-in-Chief, Dr. Robert Gross, the Associate Editors, the journal staff, the American Academy of Neurology, and the publishers Lippincott Williams and Wilkins. 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.

We anticipate further developments for the RFS in the future, limited only by the imagination of the students, residents, fellows, and others who are interested in neurology education. We hope to start our Journal Club soon, and also to have residents become involved in the peer review of manuscripts submitted to the journal. Through these efforts, we hope that the Section can play a role in helping trainees to meet requirements for core competencies related to practice-based learning and improvement, communication skills, and professionalism.

We welcome submission of manuscripts for the Resident and Fellow Section, and author instructions can be found at www.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 Mitchell Elkind or Kathy Pieper at kpieper@neurology.org.

We hope you enjoy this special fourth edition of Highlights of the Resident and Fellow Section of Neurology!

Mitchell S. V. Elkind, MD, MS, FAAN
Resident and Fellow Section Editor
Section Editor
Resident & Fellow Section

Michael E.V. Elkind, MD, MS, FAAN

Dr. Elkind graduated from Harvard Medical School in 1982, interned at Brigham and Women’s Hospital, and completed neurology residency at Massachusetts General Hospital. He then obtained a Master’s degree in Epidemiology from Columbia University while doing his clinical stroke fellowship. Currently, Dr. Elkind is an Associate Professor of Neurology and Epidemiology at Columbia University in the Division of Stroke and the Associate Chair for Clinical Research and Training. His research is focused on inflammatory and infectious biomarkers in stroke risk prediction, as well as acute stroke therapy. Dr. Elkind is a Principal Investigator of 7 NINDS-independent investigator awards. These include NeuDAPT (Neuroprotection with Statin Therapy for Acute Recovery Trial), a clinical trial evaluating short-term high-dose statin therapy in acute stroke; Levels of Inflammatory Markers in the Treatment of Stroke (LIMST), a multicenter blood biomarker study among lacunar stroke patients participating in the SPS3 trial; and the Northern Manhattan Study, a prospective cohort study of stroke risk factors. He is the former Neurology Residency Program Director at Columbia University Medical Center, and is a fellow of the American Academy of Neurology and the American Neurological Association.

Editorial Team, Resident & Fellow Section

James Berry, MD
James Berry is a neurovascular clinical research fellow at the Massachusetts General Hospital Neurology Clinical Trials Unit studying ALS. He earned his MD and MPH from Northwestern University, during which time he spent a year as a Doris Duke Clinical Research Fellow at UCSF. He completed his neurology residency at the Harvard Partners Neurology Residency where he served as chief resident. His specific interests include biomarker identification and clinical trials in ALS.

Stacey L. Cloud, MD, PhD
Stacey L. Cloud is currently a neurology resident at the University of California, San Francisco. She completed her medical degree at the University of Pennsylvania and her neurology training at the Mayo Clinic. Her research interests include the relationship between inflammation and neurological disease, especially in vascular cognitive impairment and Alzheimer’s disease. She is also interested in government policy as it applies to neurological disease.

Sheng-Han Kuo, MD
Sheng-Han Kuo completed his medical degree at National Taiwan University in Taipei, Taiwan, and his neurology residency at Baylor College of Medicine in Houston, Texas. He is currently a Movement Disorders Fellow at Columbia University in New York City. His research interest focuses on the basic science aspects of essential tremor.

Sarah Song, MD
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Jeffrey Hasty is currently a neurology resident at UCSF. He completed his medical degree at Stanford University and his neurology residency at the University of California, San Francisco. His research interests include neurocritical care, acute stroke, and the intersection of technology and medicine.

Deputy Section Editor

Ryan Dooreman, MD
Dr. Dooreman graduated from the University of Michigan School of Medicine in 2004. He completed neurology residency at Northwestern University and fellowship training in movement disorders at Indiana University School of Medicine. He serves as a liaison between the Rutgers New Jersey Medical School Neurology Residency Clinical Fellow Working Group and the Neurology Residency Program. He is a member of the American Academy of Neurology. He serves as a member of the Neurology Podcasting Committee.
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.

Resident & Fellow Section
Section Editor
Mitchell S. V. Etkind, MD, MS

Clinical Reasoning:
A 21-year-old woman with right eye swelling and bruising

SECTION 1
A 21-year-old woman presented with right eye swelling and bruising. Several weeks prior to admission, she had bifrontal headaches associated with a “whooshing” pulsation in her right ear. She went to a local emergency department where CT scan of the brain was read as negative. One week later, she noticed that her right eye was turning toward her nose. She denied diplopia. On the day of admission, she awoke and noticed bruising under her right eye accompanied by swelling. She also had blurry vision.

Her physical examination was remarkable for right eye proptosis, conjunctival injection, and mild proptosis with ecchymosis above and below the eye (figure 1). She was unable to abducted her right eye past midline, visual acuity was 20/30 bilaterally, and her visual fields were intact. There was an ocular bruit auscultated over the right eye.

Questions for consideration:
1. What other history is important to obtain?
2. What is the differential diagnosis?

Figure 1
Initial external eye findings

Right eye proptosis with proptosis, conjunctival injection, and ecchymosis surrounding the right eye.

From the Departments of Neurology (I.M.R., C.R., V.S., R.S.M.) and Radiology (D.S., J.P.-S.), Columbia Presbyterian Medical Center, New York, NY.

Disclosure: Author disclosures are provided at the end of the article.
SECTION 2
The patient denied any history of trauma. Her past medical history was significant for Ehlers-Danlos type IV, complicated by bowel rupture 6 years prior, necessitating subtotal colectomy and colostomy. She also had chronic migraines and corrective surgery for amblyopia as a child. Family history was significant for her mother who died at age 35 of splenic rupture and maternal grandmother who died at age 43 of subarachnoid hemorrhage. The differential diagnosis included thyroid ophthalmopathy, tumor, infection, inflammation (e.g., Tolosa-Hunt), and carotid cavernous fistula or fistula.

Question for consideration:
1. What further testing should be considered?

SECTION 3
Magnetic resonance angiography revealed enlarged superior ophthalmic veins bilaterally, right greater than left, with an increased prominence in the right cavernous sinus (figure 2, A and B). The patient was diagnosed with carotid cavernous fistula. She was sent to the ophthalmology clinic where her intraocular pressures (IOP) were 22 mm Hg in the right eye and 12 mm Hg in the left eye. She was started on timolol for increased IOP. Transcranial Doppler revealed accelerated flow and reduced resistance in the right internal carotid siphon.

Questions for consideration:
1. What are the treatment options for a patient with a direct carotid cavernous fistula?
2. How does her diagnosis of Ehlers-Danlos affect her treatment?

GO TO SECTION 4

SECTION 4
Because the patient had Ehlers-Danlos type IV, she had a high potential mortality rate from angiography alone. Therefore, management decisions had to be made very carefully, with extensive interdisciplinary discussions involving the family. Digital angiography showed complete seal of right internal carotid artery (ICA) flow into the cavernous sinus (figure 3). There was filling of the right middle cerebral artery and anterior cerebral artery via the anterior and posterior communicating arteries. Options for treating the direct carotid cavernous fistula included transarterial coiling via the ICA, transvenous coiling through the superior or inferior petrosal sinus, or ophthalmic approach via the ophthalmic vein. In addition, because of the high rate of treatment complications, the conservative option of observation alone was also considered. The last alternative was discarded because she would likely develop progressive visual loss without intervention. The carotid cavernous fistula was treated with transarterial endovascular coil occlusion extending from the clinoid to the petrosus internal carotid artery segment, occluding the fistula between the ICA and the cavernous sinus. Immediately after the procedure, IOP in the right eye normalized to 10 mm Hg. The edema and erythema resolved within 1 day, and she was able to adduct her right eye 10 degrees past midline (figure 4).

DISCUSSION
Carotid cavernous fistulas are classified as direct or indirect.1 Direct fistulas are characterized by carotid arterial blood shunting directly into the venous cavernous sinus, whereas indirect fistulas take place when another vessel connects the internal or external carotid to the cavernous sinus. Indirect fistulas may occur between the meningeval branches of the ICA and cavernous sinus, between the meningeval branches of the external carotid artery and cavernous sinus, or both. Fistulas most commonly occur secondary to trauma, but may happen spontaneously in patients with diseases that weaken the vessel wall, including Ehlers-Danlos.2,3

Ehlers-Danlos type IV is an autosomal dominant disorder, associated with easy bruising, thin skin, and spontaneous rupture of bowel, the uterus, or arteries. The disease is caused by mutation of COL3A1, which encodes type III collagen.3 Cultured skin fibroblasts from patients are shown to secrete reduced type III procollagen, which confirms the diagnosis of Ehlers-Danlos. Since arterial vessel walls have impaired collagen formation, this predisposes patients to cerebrovascular complications including arterial dissection, rupture, and aneurysms.4,5 Although neurologic sequelae arise in a minority of patients, it is a major cause of morbidity and mortality in this disease.5

Symptoms of carotid cavernous fistulas are more prominent with patients who have a direct fistula. Patients develop conjunctivitis and chemosis because of high-pressure arterial blood flow in the cavernous sinus transmitted to the ophthalmic veins and conjunctival vessels. Additional symptoms include pulsating exophthalmos from venous congestion in the orbit and decreased visual acuity from optic nerve ischemia. Because the fistula involves the cavernous sinus, cranial nerves III, IV, V1, V2, VI, sympa-

![MRI (A) and magnetic resonance angiography (B) showing mild proptosis of the right eye and enlarged ophthalmic veins (R > L), enlarged cavernous sinus (R > L), with multiple fistulous vessels connecting the right and left cavernous sinus.](image)

![Digital angiography](image)

![Magnified lateral view of the fistula at the posterior petrous portion of the right internal carotid artery (ICA) (dark arrowhead) with massively dilated superior ophthalmic vein (white arrow). Total steal of the ICA (white arrowhead) into the fistula is seen. (B) Lateral view of the right vertebral artery injection (dark arrow) shows flow through the posterior communicating artery (white arrow) with back filling to the level of the ophthalmic artery. Small dark arrowhead. A coil cast is demonstrated just below the ophthalmic artery extending into cavernous and petrous portion of the ICA (white arrowheads).](image)
thetic, and parasympathetics may be affected. Oph-
thalmoplegia may be secondary to cranial nerve palsies from increased pressure in the cavernous sinus or because of engorged extracranial muscles. Sensory changes in the V1 and V2 distribution may also be seen. Palsy and miosis occur because of parasympa-
thetic involvement. The presence of an ocular bruit is characteristic of carotid cavernous fistula. Transcran-
ial Doppler of the distal ICA via the ophthalmic or temporal window shows high-velocity and low-
resistance flow characteristic of a direct shunt from artery to vein. Venous distension and rupture may lead to intracranial hemorrhage.7

Imaging can aid in the diagnosis of carotid cav-
ernous fistula. CT and MRI may both show propto-
sis of the affected eye, and CT may detect a bony fracture if trauma is the cause. CT and magnetic reso-
nance imaging can diagnose a carotid cavernous fistula by showing ophthalmic vein and cavernous sinus expansion, but it is difficult to distinguish be-
 tween direct and indirect fistulas by these modalities because the resolution generally does not allow visu-
alization of small vessels connecting the carotid artery and the cavernous sinus. The gold standard is digital angiography, which can demonstrate both the anat-
omy of the fistula as well as the extent of collateral flow from the contralateral hemisphere, which is im-
portant if the treatment requires occlusion of the in-
 volved ICA.8

Repair of a carotid cavernous fistula is indicated if visual compromise is compromised, IOP is > 60 mm Hg, there is retrograde cortical venous drainage from the cavernous sinus causing the patient to be at high risk for intracranial hemorrhage, or neurological symptoms are unbearable for the patient. Indirect fistulas are more likely to close without intervention by sponta-
nous thrombosis because of the lower pressure flow. Some studies show that repeated, intermittent ca-
rotid vasoconstriction performed by patients may be used to successfully close these fistulas.9

For direct fistulas, repair is usually achieved through closure of the fistula by endovascular tech-
niques. The transarterial approach involves directing a catheter from the femoral artery to the internal ca-
rotid, and then placing detachable coils in the arterial tear to close off the fistula. Complete occlusion of the ICA at the site of the fistula may be necessary, but this can only be accomplished safely if the contralat-
eral ICA or PCA provides adequate collateral flow to supply the ipsilateral hemisphere.10 A balloon test occlusion can be performed prior to ICA takedown to determine adequacy of collateral flow. The tran-
verse venous approach is reserved for patients with dissec-
tions or stenosis of the carotid artery that would make transarterial approach difficult. In the tran-
verse arterial approach, a catheter is placed into the femo-
ral vein and threaded through the inferior petrosal vein to the cavernous sinus. This approach can be chal-
 lenging, as the opening of the inferior petrosal into the cavernous sinus is small. Another transvenous approach involves directly inserting a catheter through the superior ophthalmic vein into the cav-
ernous sinus.8 In some cases, direct surgical closure may be warranted.11

In the general population, angiography carries a risk of 2.5% morbidity, but in patients with Ehlers-
Danlos type IV, angiography has been associated with morbidity rates of 36%–67% because of the increased risk of carotid dissection, arterial perfora-
tion, and hematoma.10–14 Both the transarterial and transvenous approaches carry these high risks because of the fragility of the vessels. Arteries in these patients are also often more tortuous, making it difficult to navigate a catheter through the vasculature.4

DISCLOSURE
Dr. Reff, Dr. Neugut, Dr. Rahbani, and Dr. Smoker report no disclosures.
Dr. Philis-Tsim等相关 papers are adjustable (with medical device
including catheter, Systems and methods for determining a temperature differential using temporal-scaled magnetic resonance imaging; Syst-
tems and methods for imaging a blood vessel using temperature-scaled magnetic resonance imaging; Systems and methods for determining war-
more rate using temperature-scaled magnetic resonance imaging; Syst-
tems and methods for determining a cardiovascular parameter using temperature-scaled magnetic resonance imaging; Systems and methods for endovascular therapy; System for autonomous robotic navigation; En-

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OPINION

These manuscripts will 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 and Fellow Section Editor before submission to inquire about the interest in specific topics.

Opinion:
Interventional vascular neurology
An ability acquired through training

Simultaneously published in the Journal of Stroke and Cerebrovascular Disease and the Journal of Neurosurgery,1-12 “Performance and training standards for endovascular ischemic stroke treatment” by Meyers et al. proposes guidelines for training requirements to achieve cognitive and technical qualifications in interventional treatment of acute stroke patients, thus ensuring quality of care and safety.

Recommendations for cognitive training include at least 6 months of documented training in cerebrovascular disease within an ACGME approved residency program (neurology, radiology, or neurosurgery) and additional experience in a 3-year endovascular surgical neuroangiography fellowship. The cognitive part of the latter training was undefined and will likely vary among fellowships.

Technical requirements focus on the minimum quantity of cases. They were defined as documented training in 100 diagnostic cerebral arteriograms, training in intracranial microcatheter and microguidewire navigation (or 30 cases as primary operator with supervision by a neurointerventionalist), and 10 endovascular stroke procedures.

A grandfathering option was proposed for all endovascular specialties not specifically trained and not yet credentialed in the clinical neurosciences, and recommends 6 months of cognitive training and documentation of supervised cerebral angiography in the aforementioned amount. No recommendation was made as to how the cognitive training curriculum should be structured (i.e., full-time vascular neurology fellowship, part-time participant on stroke services, directed readings).

Because stroke care is tailored to the individual patient, it makes sense that skilled care be available from physicians trained in the clinical neurosciences. As interest in and utilization of endovascular techniques increases by clinicians from different specialties, such as interventional cardiologists, radiologists, and vascular surgeons, these training guidelines provide a useful document for understanding what is regarded as the minimum requirements by various neurologic experts analogous to the stringent training and credentialing guidelines for acute coronary interventions by interventional cardiologists. Credentialing committees have an obligation to maintain recognized accreditation standards and to be aware of recommendations endorsed by national organizations most directly involved in acute stroke care. How this will translate into practice remains to be determined.

As a stroke neurologist and current fellow in interventional neuroradiology (also called endovascular surgical neuroangiography), I believe that 6 months of cognitive training in cerebrovascular diseases is a bare minimum to acquire the skills needed to take care of an acute endovascular stroke patient. Stroke is a complex disease and decisions made in the angiography suite at the time of the event require an understanding of the pathology, likely outcome, possible complications, and safety of the applied drugs and devices used in the cerebral vasculature. A comprehensive stroke team approach—such as 24/7 access to noninterventional neurologists and neurosurgeons in a comprehensive stroke center—is helpful in the evaluation of acute ischemic stroke patients for selection for interventional treatment. Any neurointerventionalist can take advantage of and benefit from the expertise and knowledge of such a team, where it exists. Ultimately, however, the neurointerventionalist must be comfortable making these judgments independently. Cognitive training leads to superior performance; for example, cognitive training of inexperienced interventionalists has been shown to improve the quality of the endovascular outcome on virtual reality endovascular simulators.

During the past 18 months of my neuroendovascular training, I have come to appreciate that acute stroke recanalization is one of the most challenging procedures of my training. The typical stroke patient is elderly and has a difficult vascular tree. This poses technical challenges and might test the neuro-
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CHILD NEUROLOGY SECTION

The Child Neurology Section in the Resident and 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 neuropathology 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.

RESIDENT & FELLOW SECTION

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Child Neurology: Hereditary spastic paraplegia in children

Because the medical literature on hereditary spastic paraplegia (HSP) is dominated by descriptions of adult case series, there is less emphasis on the genetic evaluation in suspected pediatric cases of HSP. The differential diagnosis of progressive spastic paraplegia strongly depends on the age at onset, as well as the accompanying clinical features, possible abnormalities on MRI, and family history. In order to develop a rational diagnostic strategy for pediatric HSP cases, we performed a literature search focusing on presenting signs and symptoms, age at onset, and genotype. We present a case of a young boy with a REEP1 (SPG31) mutation.

CASE REPORT A 4-year-old boy presented with progressive walking difficulties from the time he started walking at the age of 12 to 13 months. His family history was significant for minimal gait abnormalities with onset after age 35, occurring in the patient’s mother, maternal grandfather, and maternal aunt; none of them had ever sought medical attention. Neurologic examination revealed a mildly spastic gait and marked lower limb hypertonia with bilateral Babinski signs present. Vibration perception was reduced at the ankles. Neurologic examination of the patient’s mother and maternal aunt revealed subtle gait abnormalities with bilateral Babinski signs present.

MRI of the brain and spinal cord and general metabolic screening revealed no abnormalities. Diagnostic genetic testing in both the patient and his mother revealed a pathogenic mutation (c.417 + 1 G>T) in REEP1 (SPG31) which causes a pure HSP. Mutations in ATPL (SPG3A) and SPAST (SPG4) had previously been excluded.

DISCUSSION HSP is a genetically and clinically heterogeneous group of disorders in which the main clinical feature is progressive lower limb spasticity secondary to pyramidal tract dysfunction. HSP is classified as pure if neurologic signs are limited to the lower limbs (although urinary urgency and mild impairment of vibration perception in the distal lower extremities may occur). In contrast, complicated forms of HSP display additional neurologic and MRI abnormalities such as ataxia, more significant peripheral neuropathy, mental retardation, or a thin corpus callosum. HSP may be inherited as an autosomal dominant, autosomal recessive, or X-linked disease. Over 40 loci and nearly 20 genes have already been identified. An autosomal dominant transmission is observed in 70% to 80% of all cases and typically results in pure HSP.

Spastic paraplegia is a common problem in the daily practice of pediatric neurologists, generally caused by acquired brain disorders such as perinatal asphyxia or infections early in life resulting in cerebral palsy. In addition, there is a long list of more rare disorders to consider when confronted with spastic paraplegia including structural, infectious, demyelinating, and metabolic disorders (table). Only in a small minority of cases does HSP underlie the spastic syndrome. Many patients with childhood-onset HSP are mistakenly diagnosed with cerebral palsy. In children with spastic paraplegia in whom no acquired cause can be identified, HSP should be considered. A positive family history aids with the diagnosis. Our case illustrates the importance of neurologic examination of family members who may be mildly affected.

Since the medical literature on HSP is dominated by adult case series, it is difficult to decide how the genetic evaluation should be structured when a child is suspected to have HSP. In order to develop a rational diagnostic strategy for HSP in children, we per-

GLOSSARY

ATP1A3 (ATPase 1; 85CL2) – Bearden; Sell congenital lipodystrophy 2 (autosomal recessive); KIF2A – kinesin family member 5A; L1CAM – L1 cell adhesion molecule; NAPA1 – non-protein coding-François-Willemse syndrome region protein 1; LP1S5 – proteinase-activated protein 1; REEF3 – receptor expression enhancing protein 1; SPAST – spastic; SPG3 – spastic paraplegia gene; ZFYVE20 – zinc finger FYVE domain-containing protein 25 (spastin).

From the Departments of Neurology (S.T.d.B., B.P.C.v.d.W.) and Pediatric Neurology (M.A.A.P.W.), Disorders Center for Brain, Cognition, and Behavior, Radboud University Nijmegen Medical Center, Nijmegen; and Department of Neurology (H.P.H.K.), University Medical Center Goteborg, Goteborg, the Netherlands.

Disclosure: Author disclosures are provided at the end of the article.
formed a literature search focusing on presenting signs and symptoms at onset, and genotype. We also share some of our personal experiences from a clinicogenetic database, as our institution has served as a tertiary referral center for Dutch HSP patients for over 2 decades.

Characteristics. In the medical literature, symptom onset before age 18 has been documented in many HSP cases, particularly in the complicated forms, which show a clear overlap with many metabolic disorders and leukodystrophies. In a series of 23 children with HSP, 15 of 23 (65%) were reported to have a complicated (mostly recessively inherited) HSP, compared to 8 of 23 (35%) with a pure HSP.5

In our HSP database, an early age at symptom onset (prior to age 18) was found in 72 of 175 (41%) patients, with a heterogenous genetic background: 47 of 72 (65%) autosomal dominant cases, 12 of 72 (17%) autosomal recessive cases, and 13 of 72 (18%) sporadic cases. GI tract difficulties were the presenting symptom in 81%, with an age mean of 8 years. A complicated phenotype was present in 25%. Of these 72 early-onset HSP patients, at least 20 (28%) had been placed in childhood to a pediatrician or pediatric neurologist. Prior reviews have provided in-depth descriptions and overviews of all known HSP forms.1–2 In this article, we focus on the most prevalent (>5 families described) forms of HSP with a possible childhood onset.

Autosomal dominant pure HSP, ATLI (SPG2), is a pure form of HSP, comparable to SPG4, and is almost never seen after age 20 years. It is the most frequent cause of HSP (twice as frequent as SPAST), with onset before age 10 years.4 Therefore, ATLI, which encodes atlatin (a dynamin-like GTPase), is the first gene candidate that should be tested in patients with a suspected pure autosomal dominant or sporadic HSP with symptom onset before age 10.\(^5\)

SPAST (SPG4). SPG4 is the most prevalent, mostly pure form of HSP with a variable age at onset, varying from infancy through over 70 years of age. SPAST encodes spastin with microtubule-stabilizing activity, necessary for axonal transport. In a large study of 172 SPAST patients, approximately 50% had an age of onset before 20 years.8 In our SPAST cohort, comparable figures were found, with walking difficulties presenting at a mean age of 7.5 years (range 1–18 years) in this young-onset group. Onset in infancy is unusual. After the description of 5 patients from 1 family,8 we identified an additional 5 patients from 6 families with symptom onset in infancy. Until that point, such a young onset had been described only in association with co-dominant mutations (genetic modifiers) in the SPAST gene. Therefore, SPAST is the second candidate gene that should be tested in patients with a pure HSP with symptom onset before age 10, after ATLI. With an onset of symptoms between 10 and 20 years, both genes should be tested.

NPAS4 (SPG6), KIF5A (SPG10), and SPG12. SPG6 causes a pure HSP, occasionally with a childhood onset, but more commonly with onset of symptoms in the late teen age to early adult years.9 SPG10 and SPG12 both lead to an early-onset pure HSP. All 3 are described in fewer than 10 families.10 REEP3 (SPG3), SPG3, a pure form of HSP, shows a variable age at onset, with an onset before 20 years in 71% of cases.10 REEP3 encodes the mitochondrial protein receptor expression-enhancing protein 1. REEP1 mutations were found in 8.2% of pure autosomal dominant HSP patients (of all ages), in whom ATLI or SPAST mutations had been excluded.11

We encountered 5 SPG31 patients who presented before age 18 years, with gait abnormalities and foot deformities at a mean age of 4 years. REEP1 mutations can cause a pure HSP in children, but should only be tested after SPAST and ATLI mutations have been ruled out.

Complicated forms of HSP, LIGAM (SPG1) and PLP1 (SPG2). These are both X-linked and complicated forms of HSP, which may be tested in boys with mental retardation and other clinical features.12,13 SPG2, an autosomal recessive cause, causes a spastic paraplegia in combination with cerebellar ataxia, cerebellar atrophy, optic atrophy, and peripheral neuropathy. Age at onset varies from 40 to 42 years in the literature, but mostly adult cases have been reported. SPG1 and ZFYVE26 (SPG9). SPG11 is the most frequent form of autosomal recessive HSP with onset typically in childhood (age range 1.5–21 years). It is characterized by a thin corpus callosum, mild leukoencephalopathy, mild mental retardation, and peripheral neuropathy. A comparable autosomal recessive syndrome is SPG19 (Kjellin syndrome), with additional cerebellar signs, maculopapulosis, and onset between 5 and 19 years. ROCA2 (SPG5), SPG17 (Silver syndrome) has a variable age at onset. Distal amyotrophy affecting upper extremities more than lower extremities accompanies the spastic paraplegia. Inheritance is autosomal dominant.

Four SPG7 patients in our cohort, with onset between 10 and 16 years, presented with weakness of the upper extremities greater than in the lower extremities, and 3 of the 4 had foot or hand deformities. Genetic testing in children. A formal diagnosis provides a prognosis, prevents additional burdensome and potentially costly diagnostic evaluation, may facilitate the prevention of complications, and allows for potential inclusion in clinical trials. In addition, a genetic diagnosis allows for genetic counseling with regard to the recurrence risk within the family. Ethical, social, and financial considerations, as well as written or verbal informed consent from the parents, according to established guidelines, are necessary before genetic testing in children.
The Resident and Fellow Section is a primarily online feature that serves the resident and fellow readership. Residents and fellows are expected to be the primary authors for most submissions, but those highly involved in graduate medical education (e.g., program directors) may also contribute submissions on appropriate topics. Submissions for all article categories should be no more than 1,500 words; permission for longer articles will be needed from the editors. The number of references should be 10 or less and one to two tables or figures may be incorporated. The topic must be mentioned in the cover letter of the submission. Potential article topics include: teaching, ethics, practice, career choices, residency training, editorial, international education, research, historical, opinion, book review, training videos, or teaching NeuroImages.

Teaching NeuroImages has the same requirements as NeuroImages but is especially valuable to the trainee audience and will be published in the online Resident and Fellow Section. Queries and comments should be addressed to Mitchell Elkind, MD, MS, FAAN, or Kathy Pieper at kpieper@neurology.org.

DISCLOSURE
Dr. de Be is an expert in disclosures. Dr. van der Wantingen has served as Movement Disorders Section Editor for Movement Disorders (Intel Electronic), and receives/received research support from Ipsen Pharmaceuticals, the European Union (FP7-Program), the Prions Brains Fund, and the Dutch Brain Foundation. Dr. Kneier serves on scientific advisory boards for the Hereditary spastic Nederlander and Prions Brains Fund. He has received speaker honoraria from Pfizer Inc., and receives research support from the Radboud University Nijmegen Medical Centre, the Netherlands. Dr. Wijesinghe reports no disclosures.

REFERENCES

General Submission Instructions
The Resident and Fellow Section is a primarily online feature that serves the resident and fellow readership. Residents and fellows are expected to be the primary authors for most submissions, but those highly involved in graduate medical education (e.g., program directors) may also contribute submissions on appropriate topics. Submissions for all article categories should be no more than 1,500 words; permission for longer articles will be needed from the editors. The number of references should be 10 or less and one to two tables or figures may be incorporated. The topic must be mentioned in the cover letter of the submission. Potential article topics include: teaching, ethics, practice, career choices, residency training, editorial, international education, research, historical, opinion, book review, training videos, or teaching NeuroImages. Teaching NeuroImages has the same requirements as NeuroImages but is especially valuable to the trainee audience and will be published in the online Resident and Fellow Section. Queries and comments should be addressed to Mitchell Elkind, MD, MS, FAAN, or Kathy Pieper at kpieper@neurology.org.
PEARLS AND OY-STERS

Pearls and Oy-sters focuses on fundamental clinical neurology. Each article should address a specific niche of neurologic 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 neurologic disease, such as localization, elaboration of a differential diagnosis, evaluation, or treatment. The article should 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.

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PEARSAL AND OY-STERS

The trigeminal autonomic cephalalgias (TACs) are a group of primary headaches that are characterized by unilaterality of pain, a relatively short duration of symptoms, and associated ipsilateral cranial autonomic symptoms, such as Horner syndrome, lacrimation, and nasal congestion. Incidence is rare when compared to other primary headache disorders but diagnosis (and, more importantly, treatment) can prove to be a challenge even when presented with a typical clinical presentation. The TACs are listed in the International Classification of Headache Disorders (ICHD-II) under their own section and include the following:1

1. Cluster headache (CH)
2. Paroxysmal hemicrania (PH)
3. Short unilateral neuralgiform headache with conjunctival injection and tearing/cranial autonomic symptoms (SUNCT/SUNA)

See the table for a summary of treatment options.

CLUSTER HEADACHE

Pearl: All cluster headaches need to be treated with abortive, transitional, and preventive therapies.

Oy-sters: The average time it takes for a patient with CH to be correctly diagnosed is 6.6 years. The average number of physicians seen prior to correct diagnosis is 4, and the average number of incorrect diagnoses prior to a diagnosis of CH is also 4.1

CH has a very typical clinical presentation and for this reason, the aforementioned “oy-sters” is unacceptable as patients suffer needlessly. Cluster sufferers will attest to thoughts of suicide, as the pain is extremely severe, and CH is often dubbed a “suicide headache.”

CH comes in 2 epidemiologic forms. Episodic cluster, the more common form, is characterized by attacks that occur daily during a cluster period; the period of attacks generally lasting 1–3 months, and followed by months or even years of remission before recurring. In chronic cluster, attacks occur for more than 1 year without remission, or with remissions lasting less than 1 month.1

Attacks are strictly unilateral with associated ipsilateral cranial autonomic features, and can be discerned from migraine by 2 key factors: 1) attack duration is less than the 4 hour minimum duration of a migraine attack according to ICHD-II criteria (CH attacks last 15–180 minutes); and 2) restlessness is present in CH attacks. Migraine attacks are accompanied by avoidance of movement; CH attacks by pacing and other manifestations of agitation.

When an attack of CH or another TAC occurs, the posterior hypothalamic is activated, causing a disruption in the connections for sleep. Thus, CH attacks frequently wake patients out of sleep.1 Finally, the clinician should not be distracted by migrainous symptoms—photophobia can occur in 91% and phonophobia in 89% of CH attacks, and nausea occurs frequently in cluster as well.4

All CH sufferers need more than one acute treatment for attacks in case one fails. Because of the abruptness of the attack and its severity, as well as the short duration, oral medications should be avoided—they are too slow.

An extremely effective treatment to terminate a cluster attack is 6 mg of subcutaneous sumatriptan, achieving relief in 74% of patients within 15 minutes of onset vs placebo, and this formulation and dosage is Food and Drug Administration-approved for cluster.5 Sumatriptan nasal spray at 20 mg has also been shown to be effective in randomized controlled trials, and 5 mg zolmitriptan nasal spray has also demonstrated efficacy vs placebo.5

All patients should be given a portable 100% oxygen tank for acute treatment, and O2 becomes the first line therapy if triptans are contraindicated. A non-rebreather mask is used at a flow rate of 7–15 L/min for 20 minutes and can be repeated safely without issue.6 Other effective abortive treatments include IV, IM, or subcutaneous dihydroergotamine, possibly intranasal lidocaine, and blockade of the ipsilateral greater occipital nerve.2,3,6

Many patients will present with a sense that a cluster cycle or period is beginning, and will know

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Disclosure: Author disclosures are provided at the end of the article.
ABSTRACT

Hemiracia continua is a unilateral headache with similar autonomic symptoms but is constant and responds well to indomethacin.

Opioid headache can be mimicked by HCs, particularly in the elderly.

HC is a continuous, unilateral, moderate level side-locked headache with severe exacerbations of variable duration manifesting at least one ipsilateral cranial autonomic symptom. HC is probably much more common than originally thought and often misdiagnosed.

Cervicogenic headache can mimic almost any headache, especially a TAC, and should be considered when there is lack of response to treatment. Unilaterality of pain, radiation from the neck, and failure to meet ICHD-II criteria for primary headache suggest cervicogenic headache as an alternative diagnosis to a TAC. Occipital nerve blocks can be helpful in cervicogenic headache, but physical therapy for maintenance, although Sjaastad et al. suggested controlled C2–3 blocks for diagnosis.

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 and Fellow Section Editor before submission to inquire about the need for an article on a particular topic.

Emerging Subspecialties in Neurology:

Career opportunities available to graduates of neurology residency programs continue to grow. In contrast to traditional neurology subspecialty practice that is patient-centered and disease specific, neurology hospitalists or “neurohospitalists” specialize in the care of patients admitted to the hospital with a wide array of neurological disorders. This subspecialty has emerged in parallel with mounting pressures on office-based neurologists to see larger outpatient volumes and the increasing complexity of inpatient neurologic care.1-4 Neurohospitalists are uniquely positioned to provide high-quality care at a time when many neurologists have limited or abandoned emergency coverage because of reduced reimbursement and increased litigation risk.5,6 The purpose of this article is to define what neurohospitalists do as professionals, discuss training and employment trends, and provide a future outlook on neurohospitalist practice.

NEUROHOSPITALIST PRACTICE Neurohospitalists are best defined as “site-based” subspecialists dedicated to providing and improving inpatient neurologic care. In contrast to the traditional model of an office-based neurologist concurrently delivering inpatient care, the neurohospitalist is free of outpatient responsibility and provides on-site availability for urgent evaluations and administration of time-sensitive therapies.1 Neurohospitalists evaluate and treat a multitude of conditions including altered mental status, acute stroke, seizure disorders, nervous system cancer, headaches, and neuromuscular respiratory failure. Neurohospitalists may also diagnose and comanage patients with critical illness polyneuropathy/myopathy, coma, complications of solid-organ transplantation, or increased intracranial pressure. The scope of neurohospitalist practice encompasses the entire spectrum of inpatient care including prevention and management of medical, surgical, and psychiatric complications of neurologic disease. Effective communication with outpatient and postacute care providers to ensure smooth and effective transitions of care is an additional responsibility of the neurohospitalist. Experience and competency with end-of-life issues is requisite as some acute and progressive neurologic disorders culminate with the need to provide palliative care. Commitment to improving inpatient systems of care, responsible utilization of resources, and implementing evidence-based practice in the inpatient setting are likely to become defining characteristics of the neurohospitalist.

Neurohospitalists have been successfully integrated into several practice settings. A neurohospitalist may complement a private practice group by providing hospital coverage during the day while allowing office-based neurologists uninterrupted time to see patients and perform office-based procedures. The availability of the neurohospitalist minimizes disruptions to the office schedule and likely improves both patient and physician satisfaction. In some models, night and weekend coverage is equitably shared among the group. In larger practices, continuous hospital coverage is provided by a core group of neurohospitalists that complement the clinical practice. Some neurohospitalists are on call for a 24-hour cycle every few days, while others cover up to a week straight (24/7) with resident, fellow, or mid-level provider support. Neurohospitalists may also provide value in academic institutions where limitations on resident work hours have reduced house officer availability for continuous clinical coverage.

NEUROHOSPITALIST TRAINING AND EMPLOYMENT A recent survey designed to define the current scope of neurohospitalist practice found that 79% of respondents had post-residency training or certification, 75% of which was in vascular neurology.7 Respondents were equally split between academic and private practice and 8% considered themselves full-time neurohospitalists. Neurohospitalist income varied depending on geography, but ranged from $150,000 to $450,000 per year with a median around $220,000. The survey highlights the current diversity in background and practice models among neurohospitalists. A definitive paradigm for neurohospitalist training has yet to be established. A limited number of
INTERNATIONAL ISSUES

More than 85 percent of the world’s population lives in low- and middle-income countries, where the burden of neurologic disease is the largest. Relatively little is known, however, about patients and practitioners of neurology in most countries. This section aims to explore international issues in neurology education. We welcome manuscripts describing international educational exchanges, personal rotations and experiences in low- and middle-income countries, and work by neurology trainees from around the globe. Descriptions of notable differences in training between countries are of interest. Inclusion of practical information regarding how interested residents might get involved in international programs would also be of use.

REFERENCES
International Issues: On the localization of saintly neurology: A neurology elective in India

Armed with the knowledge that I had passed my Bachelor of Medicine and Surgery examinations, I left for my elective in neurology in India. Little did I know that the next 10 weeks would be some of the most inspirational weeks of my medical training.

I landed in the lush modern utopia of Chandigarh (figure 1), the capital of the 2 states Punjabi and Haryana. Chandigarh displays meticulous urban planning with Le Corbusier’s sectorial design. It was also the first Indian city to become smoke-free in public places. I visited Shri Nek Chand’s Rock Garden (figure 2) on my first day; a remarkable 18-acre space devoid of flowers but full, instead, of sculptures made from recycled bricks. The innovative, yet humble, Shri Nek Chand is a radical: he was recycling on a large scale in the late 1950s, well before the wheely-bin culture took off in the West.

My medical experience began with wearing my hitherto unused long-sleeved white coat, banned in the United Kingdom, owing to the bare-below-the-elbows infection-control policy practiced there. I am now able to appreciate my course tutors’ comments of how useful the coat pockets are for lugging medical equipment, pocket textbooks, and jottings. I shall miss wearing the coat, which was cleaned regularly, and allowed for easy identification of the doctor among the mass of patients.

The neurology department at the Postgraduate Institute of Medical Education and Research (PGIMER, colloquially called “The Pig”) by the residents comprises only 5 consultants, about 20 senior residents (sitting the DIM examination in neurology), and a handful of junior residents (sitting the MD examination in Internal Medicine). PGIMER has recently published the largest series of patients with ophthalmoplegic migraines in the world literature1 and their Molecular Laboratory has published encouraging results on the herb, Brahmi, purported to be an Ayurvedic memory enhancer. Their mouse model of amnesia has shown that it improves parameters of learning and memory.2 The doctors serve a large population from all walks of life; between 2008 and 2009, the Outpatient Department (OPD) saw 1,319,973 patients and there were 56,078 admissions to the ward. These patients flock from the northern states of India to this government, tertiary-level institution. Health care is offered free here at the point of delivery, although equipment (e.g., hammer puncture kits) and most treatment (e.g., IV immunoglobin) must be procured and paid for by the patient. However, certain inpatient prophylactic medication, such as low molecular weight heparin, can be administered free of charge.

Stimulants and concentration were essential qualities each resident tirelessly worked 12-hour days, by asking whether the patient has any problems breaking chapsis with the fingers or, in the lower limbs, by asking whether their chappals slip off their feet. Furthermore, the latter information is refined by checking for a sensory overlay: Do the chappals slip off with or without the patient’s knowledge? The clinical examination is impressively thorough. It is what I had dreamed neurology should be like, and the degree of involvement of the residents in their examination testifies to the encyclopedic knowledge that they have. They do not have the luxury of being able quickly to check UpToDate, or, dare I say, Wikipedia, online, since there is no computer in sight. The temptation to look at the scribbles made by previous practitioners, many of whom are not medically qualified, is huge, but the residents are immune from any bias. Next, we move rooms with the patient, and the resident presents the findings to a consultant (who is also seeing new patients). After reviewing the analysis, any investigations, and perhaps even repeating aspects of the history-taking and examination, the consultant takes the time to teach us about the nuances of the condition. The consultants are therefore very much in the front line and in tune with the residents’ academic needs. The residents are conscious of the costs of the various brands of medication and tailor their prescription accordingly. Management plans are written in duplicate, both on the file that is retained by the hospital and also on a summary card that is kept with the patient. A diagnosis, even if it is provisional, is always offered to the patient in writing. The patient is told where to purchase the medication and roughly how much it would cost, so that he or she is not abused by the vast array of pharmacies in the local bazaars.

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Disclosures: Author disclosures are provided at the end of the article.


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Disclosures: Author disclosures are provided at the end of the article.

The West, such experiences are invaluable in one’s personal development. As I embark on my academic foundation-year job in neurosciences, I am sure that the wisdom that has been passed to me here will enhance my undergraduate knowledge and experience. Together, this will improve the care that I can offer to my patients. My overseas friends are to be saluted for working in extraordinarily challenging circumstances and for not deviating from their goal to serve their country. For them, there are no greener pastures.

ACKNOWLEDGMENT
The author thanks his mentor Professors Fushakur and Lal of PGIMER, and Professor Rehmat of AIIMS, for making his experience memorable and enjoyable; the Medical Council of India, Ministry of External Affairs, and Ministry of Health and Family Welfare for granting permission to undertake an elective in India; and the British High Commission for facilitating this Professor Rehmat, Oxford, for his advice on early drafts of this article; and Professor Sanchez, Stenhouse, without whose initial support this review would never have been produced.

REFERENCES

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Figure 3 International Stroke Conference, All India Institute of Medical Sciences, New Delhi

The touch technique being practiced here, yet line-related infections are seldom reported. Junior residents perform lumbar punctures at the bedside and central lines are effortlessly placed without ultrasound guidance. My mentor supervises me as I appear lost in the milieu of activity and covers the mouth of an unconscious patient, who has oral candida and military shadowing on his chest radiograph, while I perform fundoscopy. Twice daily there is a consultant ward round where the 20 or so neurologic emergencies are reviewed. I am asked to localize the lesion that is provoking drowsy nystagmus in my patient. In a patient who should have an external plantar response based on her clinical presentation I am shown the plethora of methods other than that of Babinski (absent in this patient) of how to elicit the important plantar reflex; Chaddock’s and Oppenheim’s signs are present. I was able to contrast this to my observations back home, where a significant proportion of plantar responses (elicited by only the method of Babinski) are recorded as equivocal or mute. Neurologists at this institute have both the knowledge and the passion to elicit all relevant signs and will not be satisfied unless their clerking documents a coherent account.

Finally, what happens on the wards! The floors are continuously being cleaned and the nurses and doctors help one another to give the best care to the patients. A consultant notifies that a trainee nurse is giving an IM injection in the wrong quadrant of the buttck and he steps to teach her how it should be done. I am shown how to perform chest physiotherapy and the dietician teaches the residents about nutrition issues in patients who have had a stroke. The senior residents, remarkably, perform nerve biopsies and sometimes even percutaneous tracheostomies. We workup patients ready for the teaching ward rounds. Each of us takes our turn in the fitting line as we are asked questions about our diagnostic formulation and the justification for our chosen investigations. I am quizzed on whether creatine kinase levels alone can be used to measure the response of polymyositis to steroids, and on the latest evidence for optic nerve fenestration in idiopathic intracranial hypertension. A resident is counselled by a senior about the awful practice of Nazi doctors and is encouraged to refer to the eye-of-tiger MRI appearance, not as suggestive of Hallervorden-Spatz disease, but instead of pontocerebellar-kinese–associated neurodegeneration. Certain cases are promoted to a more in-depth discussion in the seminar room. Refreshments are plentiful and the setting for the majority of us is relaxed. Nevertheless, 3 senior residents sit in a line and are asked to localize in the neuretis every symptom the patient complains of. The approach taken is that of Sir Gordon Holmes, who seems to have found a new home here in India.

I have 2 weeks left here in India, and have now moved to the All India Institute of Medical Sciences (AIIMS) (figure 3) in fast-paced and sweltering New Delhi. A recently published elective experience at AIIMS by an American neurology resident aptly depicts the liveliness of this busy center, which nurtures the academic neurologists of tomorrow. The campus is beautiful but daunting in size. I have been given a spectacularly warm welcome by the neurology department and have not been allowed to pay for lunch while in the presence of the team. The hospital is air-conditioned and the wards are similar in design to those at Oxford. The neurosurgeons are doing deep brain stimulation for the treatment of movement disorders and the interventional neuroradiologists are experienced in carotid stenting for symptomatic internal carotid artery stenosis. AIIMS also runs and coordinates worldwide collaborative clinical trials and is currently taking part in stem cell trials for ischemic strokes and Parkinson disease (PD). April is PD Awareness month and the Movement Disorders Unit hosted a well-attended educational morning for its patients and their carers. The chief guest was the retired cricketer, Bishan Singh Bedi, of the Indian spin quartet. I was impressed by the patient appreciation in group therapy and their understanding of their disease. The neurologists take pride in empowering their patients with holistic, yet simple, information and provide practical suggestions on how to live a full life. In the United Kingdom, this rewarding aspect of management has largely been delegated to nurse specialists.

I shall miss working in India very much and have been lucky to have had this unique opportunity to see so many interesting patients in such a short time. With the reduction in training that we are seeing in...
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.

I was the child neurology resident on service when we were asked to consult on a 27-week premature baby transferred to the intensive care nursery. The baby had developed *Escherichia coli* meningitis at 1 week of life. The referring pediatrics team described him as having “fixed and dilated” pupils with a “burst suppression” pattern on EEG. They appeared to have given up hope for the baby’s recovery, telling the transport team, “I’m so glad you have an ethics team at your hospital that can help facilitate withdrawal of care.”

I immediately felt the need to protect the baby as I listened to him being described in these terms. I was 26 weeks pregnant at the time. The baby was a gestational peer to my own baby, and I felt he was being profoundly misunderstood. I knew that developing in a darkened uterus, my baby’s pupils were also “fixed and dilated” and his brain wave activity was also “burst suppressed” in appearance. At that age, all babies’ are.

Just like the neocortex, the brainstem develops over the course of gestation. As neurologists, we tend to think of brainstem function as essential to life. However, many core brainstem functions, such as the regulation of breathing and a gag reflex to protect the airway, are unnecessary during fetal life. These abilities mature only in late gestation in preparation for birth. A pupillary response to light, not yet needed in a darkened womb, is almost invariably absent prior to 30 weeks’ gestation. The pupils rest comfortably at 4 mm until then.

Cortical electrical activity also develops and evolves over the course of gestation. At 24 weeks’ gestation, cortical neurons have just barely completed their long migration to the brain’s surface. A pattern of continuous brain wave activity is not present in all behavioral states until at least 35 weeks. Periods of electrical discontinuity punctuated by brief bursts of electrical activity—what might be called “burst suppression” in a more mature brain—i.e. how the premature brain burns along. Pediatric epileptologists describe this normal premature EEG tracing as “trace discontinuous” or “appropriately discontinuous for gestational age,” not “burst suppression.”

Certain labels are emotionally laden in medicine—technological euphemisms for an unusual but clearly intended deeper truth about a patient’s condition. “Fixed and dilated” and “burst suppression” are certainly among these, connecting severe neurologic dysfunction in an adult, or even a full-term infant. But the place no longer in describing the neurologic status of a very premature baby.

The nurses in our neonatal intensive care unit spoke in hushed voices, with eyes lowered, around the baby. Those labels had marked him in their minds as a baby they should take care not to get too attached to. I was grateful that his parents did not overhear their discussions.

As neurologist consultants, we tried to undo the harm this inappropriate labeling had done. He was not a “fixed and dilated baby,” but rather a premature baby with a developmentally appropriate neurologic examination. Further, his EEG was appropriate for gestational age. However, the power of these phrases, and the associated perception of the baby’s condition, persisted. The pediatric residents continued to write “pupils fixed and dilated” in their daily progress notes despite our suggestion that they simply describe what they see: “pupils 4 mm and not reactive.” The nurses would ask us, “But isn’t he in burst suppression? How could he ever recover?” I began to learn how the power of certain neurologic terms, once uttered, biased all subsequent information to the contrary.

Working in the neonatal intensive care unit while pregnant was challenging. There was the understandable fear that my baby might suffer from any of these same conditions. But I found it more frightening to imagine that my healthy baby, if born too early like this baby, could be mistakenly perceived as so neurologically damaged as to be beyond hope. Our role as neurologists in reframing this baby’s prognosis felt far more important to me than our traditional role.
diagnosing neurologic injury. Just as it is often said in law that it is better to let a guilty person go free than to wrongfully imprison an innocent one, it seemed to me a worse medical error to assign neurologic injury where there was none than to miss it where there was. For days his team had talked about him and approached his care as if he were practically brain dead. I wonder how much time, thought, and attention he did not receive because his doctors and nurses had been led astray by the inaccurate use of neurologic terms. Thankfully, the baby continued to do well regardless and after several days of observation returned to the referring hospital to complete his course of IV antibiotics.

Health care providers are often attuned to the subtleties of the language used within their narrow spheres, but these can be lost in translation for others less familiar. As neurologists, for example, we are usually cognizant of how clinical context affects the interpretation of key phrases. "Pupils fixed and dilated" may simply mean the patient has just returned from the ophthalmologist’s office. "Burst suppression" may have been intentionally induced for the treatment of status epilepticus. However, for our colleagues outside of neurology, these phrases may be assumed to be synonymous with neurologic devastation. In most circumstances, avoiding the use of coded euphemisms is probably best. When used by others, our responsibility must be to ensure they are interpreted in the proper neurologic context.

DISCLOSURE
Dr. Geffen sits on the Neurology® Resident & Fellow Section editorial team.

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: A new system for reducing patient nonattendance in residents’ clinic

ABSTRACT
Background: Patient nonattendance in neurology and other sub-specialty clinics is closely linked to longer waiting times for appointments. We developed a new scheduling system for residents’ clinic that reduced average waiting times from >4 months to <3 weeks. The purpose of this study was to compare nonattendance for clinics scheduled using the new model (termed “rapid access”) vs those scheduled using the traditional system.

Methods: In the rapid access system, nonestablished (new) patients are scheduled on a first-come, first-served basis for appointments that must occur within 2 weeks of their telephone request. Nonattendance for new patient appointments (cancellations plus no-shows) was compared for patients scheduled under the traditional vs the rapid access scheduling systems. Nonattendance was compared for periods of 6, 12, and 18 months following change in scheduling system using the \( x^2 \) test and logistic regression.

Results: Compared to the traditional scheduling system, the rapid access system was associated with a 90% reduction in nonattendance over 18 months [64% (812/2,161 scheduled visits) vs 51% (526/1,059 scheduled visits); \( p < 0.0001 \)]. In logistic regression models, appointment waiting time was a major factor in the relation between rapid access scheduling and nonattendance. Demographics, diagnoses, and likelihood of scheduling follow-up visits were similar between the 2 systems.

Conclusions: A new scheduling system that minimizes waiting times for new patient appointments has been shown to be effective in substantially reducing nonattendance in our neurology residents’ clinic. This rapid access system should be considered for implementation and will likely enhance the patient educational experience for trainees in neurology. Neurology\(^{7}\) 2010;74:a34-a36

Outpatient training is a critical component of neurology residency programs.1-3 Studies of neurology residents’ clinics have emphasized the need to optimize competency, and have suggested that interventions, such as changes in timing of continuity clinics, may improve experiences for both patients and residents.4,5 Nonattendance, defined as patient-initiated cancellation of an appointment or a no-show, represents an area for improvement.6 Reducing nonattendance in residents’ clinics is important for increasing the number and diversity of new patients for evaluation and longitudinal follow-up.

Nonattendance in neurology and other sub-specialty clinics has been consistently linked to longer waiting times for appointments.\(^{6,10}\) Studies emphasize the strength of this association across a variety of cultures and health care systems.\(^{6,10}\) In one study of neurology practices in Ireland, factors associated with nonattendance included male sex, age <50 years, urban home address, referral from emergency departments, and >2-month waiting time for the appointment.7 While this relatively long waiting time was associated with greater than double the nonattendance rate, studies in other specialties have shown that waiting times of even 1–2 weeks can make a dramatic difference in nonattendance.\(^{6,10}\)

In our US-based academic neurology program, we developed a new scheduling system for residents’ clinic that reduced average waiting times for appointments from 4 months to <3 weeks. The purpose of this study was to compare rates of nonattendance for clinics scheduled using the new model (termed “rapid access”) vs those scheduled using the traditional system.

METHODS Under the traditional system for neurology residents’ clinic scheduling at the Hospital of the University of Pennsylvania (HUP), new patients requested appointments by telephone and were given next available appointments. Appointments were scheduled to occur no more than 1 month later, and clinics were intentionally overbooked to minimize effects of cancellations and no-shows. Patients received phone call reminders 2 business days before appointments.

In July 2006, the new scheduling system, rapid access, was implemented. Using this system, the waiting time for new patient appointments is designed to be <3 weeks. Appointments become available each Monday at 8:30 am and are filled on a first-come, first-served basis. Once appointments are filled, patients are instructed to call back the following Monday morning. Those who do not receive appointments after calling 3 Mondays in a row are automatically scheduled. Patients receive phone calls reminders prior to appointments.

Data were compiled using EDS. Numbers of scheduled new patient visits and proportions not arriving for appointments (nonattendance) were determined for 6, 12, and 18 months prior to and following implementation of the new system. Primary analyses were based on the 18-month data; additional follow-up time also allowed us to determine nonattendance at 36 months. Nonattendance was defined as either a patient-initiated cancellation or a no-show. Proportion of visits with nonattendance were compared using the \( x^2 \) test. Logistic regression models were used to assess the relation between traditional vs rapid-access scheduling and nonattendance, accounting for age and appointment waiting time. Type I error was \( p < 0.05 \).

RESULTS The table presents demographic factors and nonattendance. Age, gender, racial/ethnic distributions, and diagnoses did not differ between the scheduling systems. The most common diagnoses were headache (20%), epilepsy (19%), and neuro-muscular disease (16%). On average, 46% of patients received appointments at the time of their first call to schedule. Average waiting time for new patient appointments were lower for the rapid access system (table). Nonattendance was substantially reduced using the rapid access system (64% vs 31% at 18 months, \( p < 0.0001 \), \( x^2 \) test). Proportions of patients who returned for at least one follow-up visit did not differ between rapid access and traditional scheduling (table).

Nonattendance did not change between 6 and 36 months following implementation of the new system (table). The association between rapid access scheduling and reduced nonattendance was explained by appointment waiting time, as demonstrated by stepwise logistic regression. At 18 months after implementation of the rapid access system, the odds ratio in favor of a visit being rapid access if the patient arrived was 4.1 (95% confidence interval 3.4–4.9, \( p < 0.0001 \), for accounting age). Accounting for appointment waiting time, the corresponding odds ratio was 1.3 (95% confidence interval 0.97–1.8, \( p = 0.08 \)), indicating that waiting time is a factor in the relation between scheduling system and nonattendance.

DISCUSSION Implementation of a system that limits its waiting times to ≤3 weeks to schedule a new pa-
tient appointment has substantially reduced patient nonattendance in our residents’ clinic. Our findings are novel in providing data on nonattendance for a US-based academic neurology practice, and are particularly unique in describing an intervention designed to improve the outpatient experience for neurology residents and their patients.

The rapid access system was developed based on investigations of outpatient clinics in neurology and other subspecialties; these have demonstrated a strong association between longer waiting times and nonattendance.1–18 In one report of a neurology clinic, there was a 50% difference in nonattendance for waiting times >2 months (32%) vs ≤2 months (17%).1 Smaller differences were found for ≥1 vs <1 week in otolaryngology, pulmonary medicine, and obstetrics/gynecology studies (30%–37% nonattendance for >1 week vs 24%–27% for ≤1 week, p < 0.001 for all studies).1–18 We chose a 2-week maximum waiting time for rapid access since it is a practical period for nonurgent scheduling in a medical subspecialty.1–18

Introduction of the new system was not associated with any measurable changes in demographics or diagnoses. Furthermore, patients in both groups were equally likely to return for a follow-up visit. Forty-six percent received an appointment following their first call, and patients were automatically accommodated if they called for 3 consecutive weeks. These numbers are encouraging for an academic program that by definition is limited by the number of resident physicians and by the need to balance patient care excellence with the educational experience. Differences in nonattendance, however, were greater in our study (31% for ≥3 weeks vs 64% for longer waiting times) compared to values in the literature.1–18 This finding is likely due to the fact that our investigation compared nonattendance for 2 different cohorts, and examined nonattendance for periods before and after a change in scheduling system. Although demographics are similar between traditional vs rapid access cohorts, other differences may exist that were not measured.

Our data demonstrate that a new rapid access scheduling system should be considered by neurology training programs for reducing waiting times and patient nonattendance. Such changes will likely improve patient satisfaction, clinical outcomes, and educational experiences in residents’ clinics.

AUTHOR CONTRIBUTIONS
Statistical analysis was conducted by Dr. Laura J. Bailer.

DISCLOSURE
Dr. Price reports no disclosures. Dr. Bailer served on a scientific advisory board for Regen Med; served as a consultant and received speaker honoraria from Bogen Idaho and Bayer Schering Pharma; and has received research support from the NIH (1K23 TR081836 JP2) and from the National MS Society. Dr. Gabry serves on the editorial board of Neurology® and the Journal of Neuro-Ophthalmology as a consultant for Medtronic, Inc.; serves on a speaker’s bureau for Bogen Idaho; and has received speaker honoraria from Teva Pharmaceutical Industries Ltd. and Bogen Idaho.

REFERENCES

TEACHING NEUROIMAGES
Teaching Neuroimages are interesting, previously unpublished photomicrographs, patient photographs, neuroradiologic images, or other pictorial material. They should be particularly clear examples of established observations intended for the trainee audience.
Teaching NeuroImages: Middle cerebral artery aneurysm rupture presenting as pure acute subdural hematoma

Figure 1 Funduscopic photograph

(A) Subhyaloid hemorrhage from another patient, and axial CT scan (B, C) shows right-sided subdural hematoma with no subarachnoid or intraventricular blood (black arrow) and subhyaloid hemorrhage (gray arrow). Photograph reprinted with permission from www.neurosophotology.ca.

A previously well 33-year-old man with no history of trauma or substance abuse presented with poor right eye visual acuity, somnolence, and vomiting several hours after sudden onset of severe, persistent headache. Examination revealed only a right relative afferent papillary defect and subretinal blood on funduscopy (Terson syndrome, figure 1A). Hunt and Hess grade was 3.

CT showed right subdural and subhyaloid hemorrhages (figure 1, B and C). Angiography revealed a right middle cerebral artery aneurysm (figure 2).

Aneurysm rupture rarely presents as pure acute subdural hematoma. Proposed mechanisms involve direct aneurysm rupture into subdural space, from tangential adherence to dura, or rupture through subarachnoid space by a superificial or high-pressure bleed. Terson syndrome refers to intraocular hemorrhage with aneurysm rupture. Proposed pathophysiology includes retinal venous bleeding from stasis secondary to increased intracranial pressure, or from blood forced into the subarachnoid space and then along the optic nerve sheath into the periretinal space under pressure.

REFERENCES

Coronal (A) and sagittal (B) views demonstrating bilobed aneurysm at the middle cerebral artery bifurcation. There was no evidence of an arteriovenous dural fistula.

Teaching NeuroImages: Long-term outcome of untreated Rasmussen syndrome

Figure 2 Digital subtraction right carotid angiography

A 13-year-old boy diagnosed with Rasmussen syndrome as a toddler presented with worsening seizures. History revealed onset of left-sided focal seizures in a healthy 18-month-old child. Epilepsia partialis continua was refractory to anticonvulsants and immunotherapy. MRI showed inflammatory changes (figure, A). Hemispherectomy and neurology follow-up were declined. Off anticonvulsants, seizures plateaued and development regressed to a nonverbal encephalopathy with hemiparesis. The latest MRI shows end-stage atrophy (figure, B and C). Rasmussen syndrome is a progressive, focal autoimmunencephalitis of unknown etiology that leads to intractable epilepsy, cognitive decline, and hemiparesis. Immunotherapy or hemispherectomy is recommended, so long-term neuroimaging of untreated patients is rare.

REFERENCES
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E-Pearls are listed only on our website and sent out on a weekly basis to Residents and Fellows. They are composed to be read and absorbed within a few minutes. The editorial team of the Resident & Fellow Section invites E-Pearl submissions. The length should be 85 words or less and include one reference, if applicable. Please submit your E-Pearl to Ryan Overman at rtoverman@iupui.edu.

PERIPHERAL NEUROPATHIES IN CLINICAL PRACTICE

edited by Steven Herskowitz, Stephen Sela, and Herbert Schaumburg. 408 pp., Oxford University Press, 2010, $125

Peripheral neuropathies are often challenging to neurology residents and general neurologists. In addition, most neurology residencies focus more on inpatient rather than outpatient neurology, which results in less exposure to peripheral neuropathies. This makes the recently published Peripheral Neuropathies in Clinical Practice, part of the Contemporary Neurology Series, a much welcomed book.

Peripheral Neuropathies in Clinical Practice is an updated version of Disorders of Peripheral Nerves, published in 1992. While the previous version of the text focused on the pathology of neuropathies, the current version is more clinically based. The book covers peripheral neuropathies only, and does not include central disorders of the motor neuron unit, such as motor neuron diseases and infectious polio-like syndromes, or neuromuscular junction disorders.

The first 4 chapters of the book cover basic concepts of pathophysiology and evaluation techniques for patients with peripheral neuropathies. There is a later chapter that presents a dozen clinical scenarios followed by differential diagnoses, laboratory results, electrophysiological evaluation, and final diagnosis. These illustrative cases are an excellent introduction to understanding the clinical reasoning behind clinical neuropathies. The next 12 chapters cover the different neuropathies, from immune-mediated to metabolic and toxic, hereditary, neoplastic, infectious, and those associated with systemic disease. The last 3 chapters describe the focal neuropathies,plexopathies, and hyperexcitability disorders. The text was written by 3 expert clinicians who not only refer to the latest literature—making the content up-to-date—but also to their own experience when the literature is insufficient or inconclusive. The book, although comprehensive, remains concise. There are numerous tables, drawings, and diagrams as well as several colored pathology plates of high quality. For example, chapter 3 consists of several tables outlining the differential diagnosis of neuropathies. These tables would be useful when trying to quickly formulate a differential diagnosis in a patient with, for example, a peripheral neuropathy and a concomitant myopathy.

The authors successfully present a practical approach to peripheral neuropathies. Whereas there are several other books on the subject—some of them covering motor neuron diseases and disorders of neuromuscular junction—this text is concise and well-written, making it readable cover-to-cover in a couple of weeks. The numerous tables and figures help integrate the knowledge and form a quick reference when evaluating a patient with a peripheral neuropathy. The book is affordable and small, making it easy to carry during the neuromuscular rotation or in the clinic. However, if a more thorough understanding of electrophysiology is required, one should consider a different text. I enjoyed reading Peripheral Neuropathies in Clinical Practice and would recommend it to both trainees and practicing neurologists.

Reviewed by Chafic Karam, MD

Disclosure: Dr. Karam serves on the editorial team for the Neurology® Resident & Fellow Section and is training in the same institution where Dr. Stephen Sela, one of the authors of the book, is a teacher.

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Motor movements in brain death

A variety of spontaneous and reflexive movements may be seen in patients with brain death. The most common are myoclonus-like finger jerks. Others often seen are undulating toe flexion and triple flexion response. The most dramatic of these movements is the classic "Lazarus sign," which includes flexion of the arms at the elbow, adduction of the shoulders, lifting of the arms, dystonic posturing of the hands, and crossing of the hands. Neurologists must be able to identify these movements and recognize that their presence does not preclude the determination of brain death.

Reference


Submitted by Aamir Hussain, M.D.

Disclosure: Dr. Hussain has nothing to disclose.

Atypical facial pain in smokers

Unilateral facial pain can be a rare presenting symptom of nonmetastatic lung carcinoma. The pain is most often described as an ache located in the ear. The majority also have a history of tobacco smoking. The suspected mechanism is referred pain from tumor invasion or compression of the vagus nerve. This pain can occur early in the course of malignancy and can occasionally even precede the appearance of a neoplasm on chest radiographs. As neurologists, it is important to be familiar with the clinical features of this syndrome in order to choose appropriate diagnostic testing in smokers with atypical facial pain.


Reference

Submitted by Aamir Hussain, M.D.

Disclosure: Dr. Hussain has nothing to disclose.

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