



Defending the solo and small practice neurologist

James N. Goldenberg, MD: I have great respect and concern for the solo and small practice neurologist in our shifting health care landscape. Jones and Evans¹ offer a comprehensive overview of the options available to sustain a practice of any size. My concern is that the focus of the article is on the quantitative analysis of the business. Although important, these strategies are like patching holes in the bottom of a leaky bucket. The greater problem is that the fee-for-service spigot is running dry. With a mandate to shift to value-based medicine,^{2,3} the emphasis should be on opening new spigots that are well supplied. It is affiliation and alignment with a value-based entity that will allow neurologists to regain stable financial footing. I commend the authors for reviewing alignment strategies, although I don't think they should be discussed in the context of selling a practice. There are many ways to affiliate or align and remain an independent practitioner and business owner. As we educate ourselves, I look forward to a more elaborate discussion of these important strategies.

Medical Specialists of the Palm Beaches, Atlantis, FL.

Disclosures: The author reports no disclosures.

Author Responds: Elaine C. Jones, MD: As the practice of health care changes more dramatically than when Medicare was established in 1965, solo and small neurology practices may be struggling more than ever. In our article¹ we tried to outline ways to monitor and improve the “bottom line” in the current fee-for-service model of practice. As Dr. Goldenberg rightly points out, there are other ways to maintain an independent practice and not have to sell out. As health care reform moves forward and reimbursement is based on Merit-Based Incentive Payment Systems (MIPS) and alternative payment models (APMs), neurologists will need to focus on collaboration with other value-based entities to remain successful. It will be vital to share our experiences about successes and failures as we develop these collaborations. The American Academy of Neurology is working on many resources about these new models as well. As Ringel points out, “[i]ntegrated models of care have the potential to meet the needs of patients and will allow neurologists to focus their time more efficiently and effectively.”⁴ We look forward to the conversation.

Bristol, RI.

Disclosures: The author reports no disclosures.

1. Jones EC, Evans DA. Defending the solo and small practice neurologist. *Neuro Clin Pract* 2015;5:158–163.
2. Better, Smarter, Healthier: In Historic Announcement, HHS Sets Clear Goals and Timeline for Shifting Medicare Reimbursements From Volume to Value [press release]. Washington, DC: U.S. Department of Health & Human Services. Available at: <http://www.hhs.gov/news/press/2015pres/01/20150126a.html>. Accessed June 10, 2015.
3. Major Health Care Players Unite to Accelerate Transformation of U.S. Health Care System [press release]. Available at: <http://www.hcttf.org/releases/2015/1/28/major-health-care-players-unite-to-accelerate-transformation-of-us-health-care-system>. Accessed June 10, 2015.
4. Ringel SP. The practice of neurology: looking ahead by looking back. *Neurology* 2015;84:2086–2091.

Dystonia: Five new things

Stella Marousi, MD, Richard Grünwald, DPhil: We read with interest the article “Dystonia: Five new things” by Berman and Jinnah.¹ The authors describe evidence that dystonia is a complex network disorder arising from the dysfunction of one or more parts of the brain, including the basal ganglia, thalami, cortex, cerebellum, and pontine and dentate nuclei. We

would like to stress that muscle spindles and their distorted proprioceptive feedback to central processing structures also form a crucial part of the pathophysiologic network described above.²

We have demonstrated that idiopathic focal dystonias are underpinned by abnormalities in genetically determined physical properties of muscle spindles, which produce aberrant fatigue-induced proprioceptive signals.³ This genetic predisposition possibly interacts with other related genetic changes (i.e., *TORIA*, *THAPI*) that have only partial penetrance in order to produce the dystonic phenotype.⁴ Moreover, it has previously been shown that, apart from its peripheral effects, botulinum toxin may also work by inducing central “cortical molding” through modulation of the afferent muscle spindle inputs.^{2,5} Thus, the primary abnormality in idiopathic focal dystonia may not lie exclusively in the brain. Although science has thrown some light onto the pathophysiology of dystonia, the scene is much easier to interpret if the key player is acknowledged.

Royal Hallamshire Hospital, Sheffield, UK.

Disclosures: The authors report no disclosures.

Authors Respond: **Brian D. Berman, MD, MS, H.A. Jinnah, MD, PhD:** We would like to thank Marousi and Grünwald for responding to our article¹ and for pointing out that the entire network involved in the pathogenesis of the many different types of dystonia is not yet fully delineated. We are aware of the evidence that peripheral mechanisms, including abnormal proprioceptive feedback from muscle spindles, may contribute to dystonia. It is interesting to note that this sensory information has direct connections to the cerebellum, so it is not hard to imagine that faulty source information from spindle afferents and faulty processing within the cerebellum could both cause dystonia. More research is needed to determine whether peripheral mechanisms are causal or consequential, whether they play a central role in all forms of dystonia or only some subtypes, and how they might be exploited to treat dystonia.

University of Colorado Denver (BDB), Aurora, CO; and Emory University (HAJ), Atlanta, GA.

Disclosures: B.D. Berman serves on the medical advisory boards for the Benign Essential Blepharospasm Research Foundation and the National Spasmodic Torticollis Association; has received funding for travel to conferences from Parkinson Study Group, American Neurological Association, Movement Disorder Society, Dystonia Medical Research Foundation, and Benign Essential Blepharospasm Research Foundation; serves on the editorial board of *Journal of Neurology and Neurophysiology*; and receives research support from the NIH, Dystonia Coalition, Dystonia Medical Research Foundation, Colorado Translational Research Imaging Center, University of Colorado Center for Neuroscience, The Dana Foundation, and the Benign Essential Blepharospasm Research Foundation. H.A. Jinnah serves on the scientific advisory boards for Cure Dystonia Now, the Dystonia Medical Research Foundation, Tyler’s Hope for a Dystonia Cure, the Lesch-Nyhan Syndrome Children’s Research Foundation, and Lesch-Nyhan Action France; has received funding for travel or speaker honoraria from Bachmann-Strauss Dystonia & Parkinson’s Foundation, Dystonia Medical Research Foundation, National Spasmodic Torticollis Association, Tyler’s Hope, and Cure Dystonia Now; serves as a consultant for Psyadon Pharmaceuticals and Medtronic, Inc.; provides botulinum toxin injections as a clinical service; has received research support from Psyadon Pharmaceuticals, Merz Pharmaceuticals, Ipsen Pharmaceuticals, NIH/National Institute of Neurological Disorders and Stroke, Emory Neurosciences Initiative, Atlanta Clinical and Translational Institute, Emory University Research Council, Bachmann-Strauss Dystonia & Parkinson’s Foundation, Dystonia Medical Research Foundation, Lesch-Nyhan Syndrome Children’s Research Foundation, Dystonia Coalition, Benign Essential Blepharospasm Research Foundation, and Cure Dystonia Now; and is principal investigator for the Dystonia Coalition, which receives the majority of its support through NIH grant NS065701 from the Office of Rare Diseases Research in the National Center for Advancing Translational Science and National Institute of Neurological Disorders and Stroke. The Dystonia Coalition also receives additional material or administrative support from industry sponsors (Allergan Inc., Ipsen Biopharm, Medtronic Inc., and Merz Pharmaceuticals) as well as private foundations (The American Dystonia Society, The Bachmann-Strauss Dystonia and Parkinson Foundation, BeatDystonia, the Benign Essential Blepharospasm Foundation, Dystonia Europe, Dystonia Ireland, the Dystonia Medical Research Foundation, The Dystonia Society, The Foundation for Dystonia Research, the National Spasmodic Dysphonia Association, and the National Spasmodic Torticollis Association).

1. Berman BD, Jinnah HA. Dystonia: five new things. *Neurol Clin Pract* 2015;5:232–240.
2. Kanovsky P, Rosales RL. Debunking the pathophysiological puzzle of dystonia—with special reference to botulinum toxin therapy. *Parkinsonism Relat Disord* 2011;17(suppl 1):S11–S14.
3. Frima N, Rome SM, Gruüewald RA. The effect of fatigue on abnormal vibration induced illusion of movement in idiopathic focal dystonia. *J Neurol Neurosurg Psychiatry* 2003;74:1154–1156.
4. Frima N, Nasir J, Gruüewald RA. Abnormal vibration-induced illusion of movement in idiopathic focal dystonia: an endophenotypic marker? *Mov Disord* 2008;23:373–377.
5. Walsh R, Hutchinson M. Molding the sensory cortex: spatial acuity improves after botulinum toxin treatment for cervical dystonia. *Mov Disord* 2007;22:2443–2446.

First seizure management: I can see clearly now?

Allan Krumholz, MD, Shlomo Shinnar, MD, PhD, Jacqueline A. French, MD, Gary S. Gronseth, MD, Samuel Wiebe, MD: We appreciate Drs. Cole and Cascino's thoughtful comments and provocative insights¹ regarding our recently published first seizure management guideline.² We concur that even the American Academy of Neurology's (AAN's) rigorous guideline process has limitations, but caution not to make the perfect the enemy of the good.

Practice guidelines are meant to systematically summarize the best evidence relevant to specific clinical questions. Sometimes guidelines can make recommendations that favor one treatment approach over another.^{3,4} Practice guidelines are not meant to replace clinical judgment. Rather, guidelines highlight when good judgment is needed because of limitations in the evidence. This guideline and its earlier companion⁵ review the evidence on reducing short- and long-term seizure recurrence risk, but this may be different from the consequences inherent in a recurrent seizure. This is why the guideline emphasizes the clinician's role in helping patients individually weigh those risks and values.¹ That is the practice of medicine, and guidelines are not supposed to replace it but rather to inform good clinical decision making.^{3,4} AAN guidelines are highly regarded and used by neurologists,³ and growing access to them on modern digital media make them more available to all medical providers and to patients. This is also good.

Maryland Epilepsy Center (AK), Baltimore; Albert Einstein College of Medicine (SS), Yeshiva University, Bronx, NY; New York University Comprehensive Epilepsy Center (JAF), New York; University of Kansas School of Medicine (GSG), Kansas City, KS; and University of Calgary (SW), Alberta, Canada.

This letter is copublished in *Neurology*[®] and *Neurology*[®] *Clinical Practice*.

Disclosures: A. Krumholz serves on the editorial board for *Clinical EEG and Neuroscience* and has received royalties from *UpToDate*. S. Shinnar has served on scientific advisory boards for Acorda, Questcor, and Upsher-Smith; has received royalties for *Febrile Seizures* and honoraria from Questcor, UCB, and Upsher-Smith; has received research funding from the National Institute of Neurological Disorders and Stroke and the Citizens United for Research in Epilepsy Foundation; and has given expert testimony. J.A. French has served as a consultant for Acorda, Biotie, Eisai Medical Research, GlaxoSmithKline, Impax, Johnson & Johnson, LCGH, Marinus, Novartis, Pfizer, Sunovion, SK Life Science, Supernus Pharmaceuticals, UCB, Upsher-Smith, and Vertex; has received grants from Eisai Medical Research, Epilepsy Research Foundation, Epilepsy Study Consortium, Epilepsy Therapy Project, Lundbeck, Pfizer, and UCB; and is president of the Epilepsy Study Consortium. All consulting is done on behalf of the Consortium, and fees are paid to the Consortium. New York University receives salary support from the Consortium. G.S. Gronseth reports no disclosures. S. Wiebe has received research funding from the Alberta Heritage Medical Research Foundation, the Canadian Institutes for Health Research, the M.S.I. Foundation of Alberta, and the Hotchkiss Brain Institute of the University of Calgary.

Authors Respond: Andrew J. Cole, MD, Gregory D. Cascino, MD: We appreciate Dr. Krumholz and his team's hard work on the guidelines² and their thoughtful response to our commentary.¹ While the perfect may be the enemy of the good, the good is not perfect. Nonetheless, guidelines take on the mantle of standard of care and are sometimes used by payers, lawyers, quality and safety officers, auditors, and administrators to bludgeon thoughtful physicians who come to alternative conclusions. Our point is that guidelines are

frequently useful but were not brought down from the mountain on stone tablets; thus, blind acceptance should not be substituted for considered judgment in specific clinical situations.

Massachusetts General Hospital and Harvard Medical School (AJC), Boston; Mayo Clinic (GDC), Rochester, MN.

Disclosures: A.J. Cole serves on scientific advisory boards for BrainVital Corporation, Precisis AG, and Sage Therapeutics; serves as an associate editor for *Annals of Clinical and Translational Neurology*; receives publishing royalties from *UpToDate*; serves as a consultant to Sage Therapeutics, Clarus Ventures, and Precisis AG; receives research support from Neuropace and Sunovion; and has received stock/stock options from Precisis AG and Sage Pharmaceuticals. G.D. Cascino serves as an associate editor for *Neurology*; receives research support from the NIH; and receives royalties for Mayo Foundation- Mayo Clinic Ventures-High frequency nerve stimulation to treat lower back pain (Nevro, 2013).

1. Cole AJ, Cascino GD. First seizure management: I can see clearly now? *Neurol Clin Pract* 2015; 5:278–280.
2. Krumholz A, Wiebe S, Gronseth GS, et al. Evidence-based guideline: Management of an unprovoked first seizure in adults: Report of the Guideline Development Subcommittee of the American Academy of Neurology and the American Epilepsy Society. *Neurology* 2015;84:1705–1713.
3. Gronseth G, French J. Practice parameters and technology assessments: what they are, what they are not, and why we should care. *Neurology* 2008;71:1639–1643.
4. Ben-Menachem E, French JA. Guidelines—are they useful? *Epilepsia* 2006;47(suppl 1):62–64.
5. Krumholz A, Wiebe S, Gronseth G, et al. Practice parameter: evaluating an apparent unprovoked first seizure in adults (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology and the American Epilepsy Society. *Neurology* 2007;69:1996–2007.

Mal de débarquement syndrome

Bernard Cohen, MD, Mingjia Dai, PhD, Eric Smouha, MD, Catherine Cho, MD: The recent review of mal de débarquement syndrome (MdDS) by Saha and Fife¹ covered many aspects of the illness, but there is additional information that your readers should know. Using results from NASA experiments on humans² and experiments from our laboratory on subhuman primates,³ Dai devised a treatment for MdDS that has proven to be successful in a large proportion of patients with MdDS. We demonstrated that 70% of 24 people with MdDS were symptom free for an average of 1 year after treatment.⁴ Subsequently, we have been contacted by many patients with MdDS and currently have treated more than 100. Although we do not have adequate follow-up yet (treatment only began in this cohort 9 months ago), it appears that the “cure rate” is approximately the same as in the original report.⁴

Our initial report also indicated that there are physical findings associated with MdDS that can help make the diagnosis. All of the patients rocked, swayed, or bobbed at an average frequency of ~0.2 Hz (one cycle every 5 seconds). Many of the patients had lateral movement on the Fukuda Stepping Test, and some had vertical nystagmus, with quick phases of the nystagmus upward when the head was rolled to one side and downward when the head was rolled to the other side. They also had many associated physical complaints⁴ that are described in the Saha and Fife article.¹ These signs caused these people to be truly miserable. Many were unable to work and those who managed to soldier on found that their efficiency was very low. Depression was common. Several tried to commit suicide after failing to get relief from years of continuous oscillation.

Based on the physical findings, our laboratory studies in monkeys, and an early survey of the disorder,⁵ we postulated that MdDS was based on maladaptation of velocity storage in the vestibulo-ocular reflex. Treatment is essentially based on using low-velocity full-field stimulation while rolling the head at the frequency of the rocking, swaying, or bobbing. The physical findings and associated somatic signs disappear after successful treatment.

As treatment has progressed, it has become clear that there are 2 ways that MdDS is initiated. We have termed one form “classic MdDS,” which follows cruises or travel on the sea, extensive car trips, or turbulent flights. This has been the easiest form of the illness to treat, and the treatment is most successful in this group. We call the second form “spontaneous MdDS,”

which occurs when the rocking and associated signs develop without a preceding trip. In our recent study, these people were more difficult to treat and the cure rate was more than 50%.

Although vestibular testing can be of use, in general, MRIs, vestibular evoked myogenic potentials, auditory evoked potentials, and multiple blood tests really have nothing to offer diagnostically. The major diagnostic clues are a history of a previous cruise or flight or spontaneous onset and the rocking, swaying or bobbing at approximately 0.2 Hz (the frequency is generally higher after if the MdDS followed a flight or occurred spontaneously). A striking finding is that MdDS symptoms abate when riding in a car, plane, or boat but return promptly at the end of the ride.

Thus, MdDS is a treatable illness, and it is important that it be recognized as such by neurologists and otolaryngologists who see these patients in order to send them for potential reduction or cure of their symptoms and signs.

Icahn School of Medicine at Mount Sinai, New York, NY.

Disclosures: The authors report no disclosures.

1. Saha KC, Fife TD. *Mal de débarquement* syndrome: review and proposed diagnostic criteria. *Neurol Clin Pract* 2014;5:209–215.
2. Guedry FE Jr, Graybiel A. Compensatory nystagmus conditioned during adaptation to living in a rotating room. *J Appl Physiol* 1962;17:398–404.
3. Dai M, Raphan T, Cohen B. Adaptation of the angular vestibulo-ocular reflex to head movements in rotating frames of reference. *Exp Brain Res* 2009;195:553–567.
4. Dai M, Cohen B, Smouha E, Cho C. Readaptation of the vestibulo-ocular reflex relieves the mal de débarquement syndrome. *Front Neurol* 2014;5:124.
5. Brown JJ, Baloh RW. Persistent mal de débarquement syndrome: a motion-induced subjective disorder of balance. *Am J Otolaryngol* 1987;8:219–222.

Occasional resolution of multiple parenchymal brain calcifications in patients with neurocysticercosis

Nitin K. Sethi, MD, Prahlad K. Sethi, MD: We read with interest the case report on occasional resolution of calcified neurocysticercosis cysts on follow-up brain CT scans by Meneses Quiroz et al.¹ In 1985, one of us (PKS) reported 11 patients from India with appearing and disappearing CT lesions and seizures.² At that time, CT scan technology in India was still primitive and images were of low resolution. We reported both low-attenuation and mixed-attenuation lesions (high-attenuation lesion with perifocal edema) in brain parenchyma. In our patients, CT scan lesions disappeared without any specific therapy other than anticonvulsants, causing us to speculate about the underlying etiology of these lesions. We considered cysticercosis, tuberculosis, cryptic cerebral vascular malformations, a functional rather than a structural lesion (postictal edema), and a focal encephalitis peculiar to the Indian subcontinent as possible etiologies. In hindsight, what we reported in 1985 as appearing and disappearing CT scan abnormalities were calcified cysticercosis lesions.

New York–Presbyterian Hospital (NKS), New York; and Sir Gangaram Hospital (PKS), New Delhi, India.

Disclosures: N.K. Sethi serves as Associate Editor for *The Eastern Journal of Medicine*. P.K. Sethi reports no disclosures.

1. Meneses Quiroz LJP, Gonzales I, Pretell EJ, Saavedra H, Garcia HH; Cysticercosis Working Group in Perú. Occasional resolution of multiple parenchymal brain calcifications in patients with neurocysticercosis. *Neurol Clin Pract Epub* 2015 May 28. doi: 10.1212/CPJ.000000000000144.
2. Sethi PK, Kumar BR, Madan VS, Mohan V. Appearing and disappearing CT scan abnormalities and seizures. *J Neurol Neurosurg Psychiatry* 1985;48:866–869.

Neurology® Clinical Practice

Dystonia: Five new things

Stella Marousi, Brian D. Berman, Richard Grünewald, et al.

Neurol Clin Pract 2015;5:366-368

DOI 10.1212/01.CPJ.0000472923.37443.61

This information is current as of October 12, 2015

Updated Information & Services	including high resolution figures, can be found at: http://cp.neurology.org/content/5/5/366.2.full.html
References	This article cites 5 articles, 2 of which you can access for free at: http://cp.neurology.org/content/5/5/366.2.full.html##ref-list-1
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://cp.neurology.org/misc/about.xhtml#permissions
Reprints	Information about ordering reprints can be found online: http://cp.neurology.org/misc/addir.xhtml#reprintsus

Neurol Clin Pract is an official journal of the American Academy of Neurology. Published continuously since 2011, it is now a bimonthly with 6 issues per year. Copyright © 2015 American Academy of Neurology. All rights reserved. Print ISSN: 2163-0402. Online ISSN: 2163-0933.

