Editors' Note: The inability to visualize nigrosome 1 on a 7T brain MRI is suggestive of Parkinson disease, as described by Blazejewska et al. Mueller et al. report that similar sensitive and specific findings can be obtained with the more widely available 3T MRI. Replication of this finding on a larger cohort is warranted. In response to Liguori et al., Friedman et al. caution against diagnosing idiopathic intracranial hypertension syndrome on the basis of elevated opening pressure alone without other objective findings.

—Chafic Karam, MD, and Robert C. Griggs, MD

VISUALIZATION OF NIGROSOME 1 AND ITS LOSS IN PD: PATHOANATOMICAL CORRELATION AND IN VIVO 7T MRI

Christoph Mueller, Bernadette Pinter, Eva Reiter, Michael Schocke, Christoph Scherfler, Werner Poewe, Klaus Seppi, Innsbruck, Austria: Blazejewska et al.¹ identified a hyperintense ovoid area within the dorsolateral border of the otherwise hypointense pars compacta of the substantia nigra (SNc) in healthy controls (HC) and the absence of this MRI feature² in patients with Parkinson disease (PD) using T2*-weighted 7T MRI. Moreover, the postmortem 7T MRI with histopathologic correlations suggested that this MRI feature corresponded to nigrosome 1. We studied this MRI feature in consecutively recruited HC and patients with PD³ (16 HC, 53 PD; mean $\pm$ SD age, y: HC 54 $\pm$ 10, PD 67 $\pm$ 8; proportion female: HC 68%, PD 28%; mean $\pm$ SD disease duration, y: 8 $\pm$ 4) using susceptibility-weighted imaging (SWI) at 3T (Siemens Verio MRI scanner, Munich, Germany). Datasets of 2 HC and 18 patients with PD were not of diagnostic quality due to patient motion. In the remaining patients, presence of dorsolateral hyperintensity within the otherwise hypointense SNc was identified in 14/14 HC and 5/35 patients with PD. Absence of this MRI feature resulted in 85.7% sensitivity and 100% specificity to detect PD. We suggest that this MRI feature reported at 7T MRI using T2*-weighted sequences¹ can also be detected at 3T using SWI sequences and that absence of this MRI feature might have potential for diagnosing PD.

Author Response: Anna I. Blazejewska, Stefan T. Schwarz, Nin Bajaj, Dorothee P. Auer, Penny A. Gowland, Nottingham, England: Following our article on the application of 7T T2* MRI for PD diagnostics,¹ we were delighted to read about the successful use of 3T SWI for detection of nigrosome 1 absence in the substantia nigra in patients with PD by Mueller et al. We also studied the application of 3T SWI in PD cohorts as a fast and easily applicable diagnostic tool. Early findings indicated a sensitivity of 89%–95% and a specificity of 91% of nigrosome 1 detection using 3T MRI,⁴ which are similar to the results from the 7T MRI study.

Of interest, our rate of nondiagnostic scans was lower than that of Mueller et al. (29%). Their study in 14 controls and 35 patients with PD needs replication with a more robust protocol and in a larger cohort including atypical parkinsonism. Translation of the discovery made at 7T to 3T is particularly promising due to the general availability of 3T scanners. In addition, the lack of postprocessing required for nigrosome detection indicates a potential viable alternative to the more expensive nuclear medical techniques currently licensed.

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REVISED DIAGNOSTIC CRITERIA FOR THE PSEUDOTUMOR CEREBRI SYNDROME IN ADULTS AND CHILDREN

Claudio Liguori, Andrea Romigi, Maria Albanese, Maria G. Marciani, Fabio Placidi, Rome: Friedman et al.¹ updated the diagnostic criteria for

idiopathic intracranial hypertension (IIH). We would like to add the following case to reports of patients presenting chronic daily headache with elevated lumbar puncture (LP) opening pressure without papilledema.^{2,3}

We saw an obese woman with global headache associated with nausea and back pain that began 1 month earlier. Brain MRI/venography was normal, papilledema was not detected, and optic/ocular examinations were normal. LP revealed an opening pressure of 177 mm with normal CSF contents. Amitriptyline was initiated for chronic daily headache, the patient was advised to lose weight, and improvement was noted.

Six months later, the patient complained of severe, global, nonsteroidal anti-inflammatory drug-resistant headache taking place over the previous 4 weeks. She showed a 12-pound weight gain. The repeated brain MRI/venography was normal and we did not detect papilledema or optic/ocular abnormalities. LP documented an opening pressure of 313 mm with normal CSF contents. Treatment with acetazolamide was initiated and the symptoms resolved.

The revised criteria¹ did not support an IIH diagnosis; yet it is possible that our patient could have had IIH with a progressive evolution. Development of papilledema is possible. We suggest that patients presenting clinical conditions similar to our case could

be diagnosed with probable IIH requiring vigilant follow-up.

Author Response: Deborah Friedman, Dallas; Kathleen Digre, Salt Lake City; Grant Liu, Philadelphia: We thank Dr. Liguori et al. for their comments and appreciate that there will always be occasional exceptions to any rule. However, in our experience, patients who are diagnosed with pseudotumor cerebri syndrome (PTCS) on the basis of CSF pressure alone—without other objective findings—rarely have the disorder. We have seen devastating consequences in patients who were diagnosed incorrectly and subjected to unnecessary medications and surgical procedures that carry significant risk. The LP opening pressure is a snapshot in time and may be influenced by the presence of cephalgia and other technical factors. The therapeutic response to CSF removal and acetazolamide are not specific to PTCS.

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CORRECTION

***C9orf72* expansions are the most common genetic cause of Huntington disease phenocopies**

In the article “*C9orf72* expansions are the most common genetic cause of Huntington disease phenocopies” by D.J. Hensman Moss et al. (*Neurology*® 2014;82:292–299), there is an omission in the Methods section under the subheading “Case ascertainment.” The third paragraph should read “All *C9orf72*-positive cases were given a modified Goldman score,^{21,22} which was used to quantify the strength of the autosomal dominant family history (scoring was modified to give a score of 0 for no data, 4 for definitely no family history, and 4.5 for unknown or undescribed family history).” The full scoring system has been added to the supplemental data. The authors regret the omission.

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Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children

Claudio Liguori, Deborah Friedman, Andrea Romigi, et al.

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