Response to Correspondence "In Pseudotumor cerebri, hormonal contraception is not associated, and the diagnosis remains as 'Idiopathic Intracranial Hypertension'"

REPLY

WE THANK DRS LEE AND FRANCIS FOR THEIR COMMENTS. WE agree that the diagnosis of idiopathic intracranial hypertension (IIH) should be reserved for conditions of intracranial hypertension (ICH) without a known etiology after sufficient investigation.

There are many conditions that can masquerade as IIH, which has been used synonymously with pseudotumor cerebri (PTC) in the literature. As chronicled by Wall and associates (1991), the terminology has evolved since Quincke described the condition in 1897.[1] The term IIH was proposed to convey the expectation that IIH is a diagnosis of exclusion.[2] This requirement is contained within the modified Dandy criteria, which specified that IIH patients must have signs and symptoms of ICH, elevated lumbar puncture opening pressure with normal cerebrospinal fluid constituents, no localizing neurologic finding except cranial nerve VI palsies, normal neuroimaging except for signs of increased intracranial pressure, and no other apparent cause.[3–5] The criteria assume the need for appropriate investigations into possible etiologies of ICH, including a neurologic examination, neuroimaging, a lumbar puncture with measurement of the opening pressure, and analysis of cerebrospinal fluid constituents, before making the diagnosis of IIH.

In the initial survey of IIH patients in our population-based study, the Rochester Epidemiology Project (REP) database was searched for patients with the diagnoses of IIH, intracranial hypertension, PTC, or papilledema.[6] This yielded 427 potential participants, only 63 of whom were confirmed to have IIH after ensuring that they met the modified Dandy criteria. Although we did not specifically screen for all coagulopathies, we excluded 13 patients with cerebral venous sinus thromboses because these patients do not meet the criteria of IIH/PTC. It is possible that some of the prior reported associations between birth control and IIH may have been unrecognized cases of cerebral venous sinus thrombosis because of the known prothrombotic risk of hormonal therapies, which can predispose patients to venous thrombosis.

While there is consensus that a diagnosis of IIH requires the exclusion of known etiologies of ICH, there is debate as to whether IIH should supplant the term "PTC," or represent a subset of PTC, which would also include secondary causes of ICH without an apparent tumor.[2,5] The use of PTC as a synonym of IIH has persisted in the common parlance, with one parenthetically equivalent to the other. Whether the term IIH or PTC is used, we think we can all agree that it is critical to exclude secondary causes of papilledema and ICH, such as tumor or venous sinus thrombosis, before these terms can be applied to patients.

KHIN P. KILGORE
Rochester, Minnesota, USA
MICHAEL S. LEE
Minneapolis, Minnesota, USA
JACQUELINE A. LEAVITT
RYAN D. FRANK
Rochester, Minnesota, USA
COLLIN M. MCCLELLAND
Minneapolis, Minnesota, USA
JOHN J. CHEN
Rochester, Minnesota, USA

CONFLICT OF INTEREST DISCLOSURES: SEE THE ORIGINAL article for any disclosures of the authors.

REFERENCES

1. Wall M, George D. Idiopathic intracranial hypertension. A prospective study of 50 patients. *Brain* 1991;114:155–180.
2. Friedman DI, Liu GT, Digre KB. Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. *Neurology* 2013;81(13):1159–1165.
3. Smith JL. Whence pseudotumor cerebri? *J Clin Neuro-ophthalmol* 1985;5:55–56.
4. Wall M. Idiopathic intracranial hypertension. *Neurol Clin* 2010;28:593–617.
5. Wall M, Friedman DI, Corbett JJ, et al. Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children: author response. *Neurology* 2014;83(2):198–200.
6. Kilgore KP, Lee MS, Leavitt JA, et al. Re-evaluating the incidence of idiopathic intracranial hypertension in an era of increasing obesity. *Ophthalmology* 2017;124(5):697–700.

Are Risk Factors for Growth of Choroidal Nevi Associated With Malignant Transformation? Assessment With a Validated Genomic Biomarker

EDITOR:

WE WOULD LIKE TO COMMENT ON HARBOUR AND ASSOCIates' American Ophthalmological Society thesis "Are risk factors for growth of choroidal nevi associated with malignant transformation? Assessment with a validated genomic biomarker."[1] The authors found that of the many clinical features associated with "malignant transformation" of a melanocytic tumor, only 2, tumor thickness greater than 2.25 mm and patient age (over 60), are significant. There are likely a few exceptions to this, such as