ABSTRACTS

Neuro-Ophthalmic Literature Review

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OCT MACULAR THICKNESS INVERSELY CORRELATES WITH PARKINSON'S DISEASE SEVERITY

Satue M, Seral M, Otin S, Alarcia R, Herrero R, Bambo MP, Fuertes MI, Pablo LE, Garcia-Martin E. Retinal thinning and correlation with functional disability in patients with Parkinson's disease. *Br J Ophthalmol* 2014;98:350–355.

Optical coherence tomography (OCT) is increasingly used in neurology as well as ophthalmology. Recent studies have shown that there is retinal nerve fibre layer (RNFL) thinning in patients with neurological disease (e.g., multiple sclerosis, Parkinson's disease [PD], and Friedreich's ataxia), even in the absence of any visual symptoms. In this study, the authors correlate OCT RNFL and macular thickness with disease progression in PD patients ($n = 153$) with no known eye diseases, and compared them with normal controls ($n = 242$). The peripapillary RNFL and macular thickness were measured with two spectral domain OCT instruments (Spectralis and Cirrus). PD severity was rated with the Hoehn and Yahr (HY) scale, Schwab-England Activities of Daily Living scale, and Unified PD Rating Scale. In PD patients, the average, superior, and inferior peripapillary RNFL were significantly thinned compared with normal, on both Cirrus and Spectralis. The macular thickness measurements (in all quadrants, except at the fovea), measured with Spectralis, were also significantly lower than in controls. There was a statistically significant (but low) inverse correlation between the thickness of most of the macular sectors (on Spectralis) and PD severity (HY scale). The authors speculate that the retinal thinning could be due to low dopamine levels producing axonal degeneration, leading to retrograde degeneration of inner retinal layers. The findings in this study suggest that OCT macular thickness may be used as an objective biomarker of PD disease progression and can help evaluate treatment effect.

Carmen Chan

AN IMPROVED NEUROLOGICAL HEMIFIELD TEST TO IDENTIFY VISUAL FIELD DEFECTS CAUSED BY CHIASMAL AND POSTCHIASMAL LESIONS

McCoy AN, Quigley HA, Wang J, Miller NR, Subramanian PS, Ramulu PY, Boland MV. Development and validation of an improved neurological hemifield test to identify chiasmal and postchiasmal lesions by automated perimetry. *Invest Ophthalmol Vis Sci* 2014;55:1017–1023.

Most ophthalmologists are familiar with the Glaucoma Hemifield Test (GHT), a built-in algorithm in the Humphrey Field Analyser (HFA) that analyses the probability of a certain visual field pattern is caused by glaucoma. The GHT compares pointwise the pattern deviation data from points above and below the horizontal midline. The authors of this paper previously described a Neurological Hemifield Test (NHT) that aimed to detect (subtle) visual field defects caused by chiasmal and post-chiasmal lesions, by pointwise comparison of pattern deviation data from points either side of the vertical midline. The original NHT used the higher NHT score from either eye, which means it was essentially a monocular test. In this paper, the authors describe an improved NHT

in which data from both eyes were used. In the improved NHT, the NHT laterality score is the difference between the scores of selected points to the left and right of the vertical midline, retaining the positive or negative sign. Therefore, patients with homonymous hemianopia should have NHT laterality scores of the same sign in both eyes, whereas patients with bitemporal hemianopia should have opposite signs in both eyes. The authors used the improved NHT in 633 patients (equal numbers of neurological, glaucoma, and glaucoma suspect patients), with 474 cases in the training set and 159 cases in validation set. Using the NHT laterality score from each eye combined with the sum of NHT scores, the area under the receiver operating characteristic (ROC) curve was 0.93, better than the original NHT test. The NHT scores also correctly identified the type of defect (bitemporal, right homonymous, left homonymous or binasal) in 96% (158/164) of neurological patients.

Carmen Chan

HANDHELD OCT PERFORMED UNDER SEDATION CAN BE USED TO PREDICT VISUAL LOSS IN YOUNG CHILDREN WITH OPTIC PATHWAY GLIOMAS

Avery RA, Hwang EI, Ishikawa H, Acosta MT, Hutcheson KA, Santos D, Zand DJ, Kilburn LB, Rosenbaum KN, Rood BR, Schuman JS, Packer RJ. Handheld optical coherence tomography during sedation in young children with optic pathway gliomas. *JAMA Ophthalmol* 2014;132:265–271.

Young children can be affected by optic pathway gliomas (OPGs), which occur sporadically or in association with neurofibromatosis type I (NF-1). OPGs do not always cause visual loss and chemotherapy is usually only indicated in OPG patients with new or progressive visual loss. Determining if a young child has visual loss may be difficult, due to poor cooperation with subjective tests (visual acuity, colour vision, visual field). Objective tests such as visual evoked potential (VEP) or neuro-imaging have little temporal relationship with visual loss. Peripapillary retinal nerve fibre layer (RNFL) thickness, as measured by optical coherence tomography (OCT), has been shown to correlate well with visual function. However, RNFL measurement requires considerable cooperation, even with a spectral-domain OCT instrument with eye-tracking function. In this study, the authors used a handheld OCT (HH-OCT; Biotigen) to measure the peripapillary RNFL in 33 children (64 eyes, median age 4.8 years) with OPGs (with and without visual loss) when they underwent magnetic resonance imaging (MRI) under sedation, and compared them with normal controls (20 patients, 31 eyes, median age 8.7). Only children who could cooperate with subjective visual acuity testing were included in this study. It was found that the OPG group with visual loss had thinner peripapillary RNFL. The authors suggested that RNFL measured under sedation by HH-OCT may be used as a surrogate marker for visual loss in young children who cannot cooperate with visual acuity testing; however, they do not recommend it for routine clinical use at this stage.

Carmen Chan

YES WE CAN

Umur Kayabasi A, Sergott RC. OCT and FAF in the early diagnosis of Alzheimer's disease. *Neurobiol Aging* 2014;35:715–724.

The objective of this study was to use retinal optical coherence tomography (OCT) and fundus autofluorescence (FAF) to test for signs of neurodegeneration in the retina of patients with Alzheimer's diseases. The rationale was that it has been proposed that beta-amyloid, which causes Alzheimer's disease (AD) in the brain, may be observed in different tissues of the eye (lens, retina, etc.) years before clinical onset. The authors examined 14 patients with a family history of AD, in 13 of whom there were only minor cognitive dysfunction. The results of the FAF examination of the retina revealed suspected regions. Interestingly, the OCT B-scans through these retinal lesions demonstrated plaque deposition within the retinal ganglion layer (GCL). Of note, many plaque deposits co-localised with drusen due to age-related macular degeneration. The authors conclude that these deposits may be signs of AD and may contain beta-amyloid. They emphasise that the deposits were predominantly in the GCL and did not necessarily co-localise with drusen. It is suggested that the detection of plaques in the retina may potentially help with the early diagnosis of AD. The reader does not know how many of these patients may have had subtle clinical signs of or genetic risk factors for glaucoma, a pathology that overlaps with AD.

Axel Petzold

NO WE CANNOT

Ho CY, Troncoso JC, Knox D, Stark W, Eberhart CG. Beta-amyloid, phospho-tau and alpha-synuclein deposits similar to those in the brain are not identified in the eyes of Alzheimer's and Parkinson's disease patients. *Brain Pathol* 2014;24:25–32.

The first suggestion that the retina may be a remote playground for Alzheimer's disease (AD) pathology was made by Hinton and colleagues (*N Engl J Med* 1986;315:485–487). Their postmortem data on 4 retinas and 10 optic nerves demonstrated reduced ganglion cell numbers and retinal nerve fibre (RNFL) loss in AD compared with controls. The present study tried to replicate these data by systematically investigating the eyes and brains from 11 patients with AD, 6 patients with Parkinson's disease (PD; with and without dementia), and 6 age-matched control subjects. Brain pathology was rated using the standard Braak score. Table 1 of the paper summarises the essentially negative data. For each case, the demographic data, clinicopathological diagnosis, and immunohistochemistry data for Congo red, beta-amyloid (clone 6F/3D), phospho-tau (clone AT8), and alpha-synuclein (clone 42) staining were shown. There were no differences between groups. The authors conclude that there is insufficient evidence to further support the hypothesis that abnormal retinal protein aggregation is a dominant feature in either AD or PD pathology. The reader does not know if patients with evidence for glaucoma were excluded from the study and whether or not this may be relevant for future studies.

Axel Petzold

31 CASES OF THE GREEN TUMOUR

Aggarwal E, Mulay K, Honavar SG. Orbital extra-medullary granulocytic sarcoma: clinicopathologic correlation with immunohistochemical features. *Surv Ophthalmol* 2014;59:232–235.

In one of the largest case series published, the authors report on 31 patients with biopsy-proven orbital granulocytic sarcoma (GS), describing the malignancy's clinicopathological and immunohistochemical characteristics. Most commonly found in the gums and skin, this rare orbital tumour is referred to by many names, including myeloblastoma and chloroma. This latter term shares its etymological roots with words like chlorine and chlorophyll, referring to the colour that myeloperoxidase (MPO) brings out in the tumour upon exposure to ultraviolet light (a quality that the authors note may not be present in one-third of GS). The median age at diagnosis of the patients in the study was 12, which is a few years older than the median ages cited in prior studies, and a slight female predilection was found (a finding that has been variable in the literature). All presented with proptosis, a palpable mass was found in 71%, and disc oedema was identified in 23%. Computerised tomography (CT) revealed a diffuse mass, most often in the superomedial orbit. As most of the above characteristics are not unique to GS, histology and especially immunohistochemistry have become integral to its diagnosis. The authors report positivity of MPO, CD34, CD117, and CD43 in 100% of their cases. After tissue diagnosis, 42% were found to have a concomitant haematologic malignancy and 72% of the remainder developed acute myeloid leukemia (AML) within the next 3–21 months. Additionally, the average survival of the patients with GS but without AML was 43 months, whereas previous reports have shown a median survival of about 36 months. This is notable in that the patients in this study were also diagnosed at an older median age.

Evan Price

TOWARD A BETTER UNDERSTANDING OF MMPs: A POTENTIAL OPTIC NEUROPATHY TREATMENT MODALITY

De Groef L, Van Hove I, Dekeyser E, Stalmans I, Moons L. MMPs in the neuroretina and optic nerve: modulators of glaucoma pathogenesis and repair? *Invest Ophthalmol Vis Sci* 2014;55:1953–1964.

In this detailed review of the expression and role of matrix metalloproteinases (MMPs) in the optic nerve and sensory retina, the authors report on the current understanding of their involvement in the pathophysiology of glaucoma. They describe the potential roles of MMP-1 and membrane type 1 MMP (MT1-MMP) in the optic nerve and MMP-3 and -9 in the retina, but note that the exact mechanisms of their involvement have yet to be clarified. The possibility of central nervous system (CNS) repair via MMP-based therapies is also discussed, with

gelatinases, an MMP subset, showing particular promise. Although only tangentially mentioned in the review, such interventions may also be applicable to other optic neuropathies of neuro-ophthalmic interest in the future.

Evan Price

TOURNAY'S ANISOCORIA

Robert MP, Plant GT. Tournay's description of anisocoria on lateral gaze: reaction, myth, or phenomenon? *Neurology* 2014;82:452–456.

This paper reviews the history of Tournay's anisocoria, which represents unequal pupil size occurring as a function of lateral gaze. This finding was well known in the early 1900s, but by mid-century was no longer discussed in the literature, and by the 1980s it was all but forgotten. Although some previous reports found that the anisocoria is present in most patients during testing, later reviews did not find it at all. The question remains: is this phenomena real or is it not true? The authors' response to this question requires us to understand the difference between reality and perception. In fact, what they determined is that the phenomenon is actually perceived (due to an optical illusion), but it is often not actually real. Whether or not the anisoria is perceived also depends on aperture size—it is not noted in miotic pupils. This is a fascinating review of a lesser known but intriguing neuro-ophthalmologic sign.

John H. Pula

A COMPUTERISED LANCASTER RED-GREEN TEST

Awadein A. A computerized version of the Lancaster red-green test. *J AAPOS* 2013;17:197–202.

Dr. Awadein, from Cairo, proposes here two computerised versions—one with a screen and another with a projector—of the Lancaster red-green test, and successfully compares it with the conventional test. This is not the first such attempt resulting in a publication. Such a device can indeed allow for easier performance from the patient, but above all, for more precise measurements of the results, in particular the torsion angles. This new device is not yet commercialised. Although the computerised versions of the visual field tests have spread to a considerable extent over the last decades, it is surprising that no computerised version of the screen tests for ocular motility disorders has been more successful so far.

Matthieu Robert

NEWS IN KCNV2 RETINOPATHY

Grigg JR, Holder GE, Billson FA, Korsakova M, Jamieson RV. The importance of electrophysiology in revealing a complete homozygous deletion of KCNV2. *J AAPOS* 2013;17:641–643.

KCNV2 retinopathy was first named “cone dystrophy with supernormal rod response” when described by Gouras in 1983. It is clinically a non-progressive or very slowly progressive retinal dysfunction (rather than a true “dystrophy”, from a clinical point of view). It does not only affect the cones, but also the rods. It exhibits a most specific electroretinographic (ERG) pattern, (although most often with normal rather than “supernormal” rod response), characterised by decreased cone responses, and a delayed rod response with very dim white flash, while with moderate increase in stimulus intensity the a-wave shape broadens and the b-wave amplitude dramatically increases, sometimes up to a supernormal extent. Such a phenotype is specific of homozygous mutations in the KCNV2 gene, which codes for a voltage-gated potassium channel. The authors report the first case of retinopathy associated with complete deletion of both KCNV2 alleles in a 6-year-old boy. The patient presented with poor vision, myopia, a manifest high-frequency, low-amplitude early-onset nystagmus (variably present in patients with KCNV2 retinopathy), structural macular changes, and a specific ERG pattern, though distinctive from usual cases by more severely affected—barely detectable—cone responses. The severe phenotype is thought to be linked with the genetic changes.

Matthieu Robert

NF1 AND MOYAMOYA

Duat-Rodriguez A, Carceller Lechon F, Lopez Pino MA, Rodriguez Fernandez C, Gonzalez-Gutierrez-Solana L. Neurofibromatosis type 1 associated with moyamoya syndrome in children. *Pediatr Neurol* 2014;50:96–98.

The presence of cerebral neurovascular abnormalities in neurofibromatosis type 1 (NF1) is well known; however, their characteristics and incidence are still a matter of study. Here the authors retrospectively reviewed 197 magnetic resonance imaging (MRI) scans of children diagnosed with NF1 and found four cases (2.3%) of moyamoya syndromes, three of which had been asymptomatic. The indication of systematic MRI scans in NF1 is debated, mainly regarding the question of detecting optic pathways gliomas. Neither the evolution of NF1-associated moyamoya syndromes nor the suitable treatment of such moyamoya, in case they would become symptomatic or evolve, is currently known. Among several conclusions and questions arising from this interesting article, one can deduce the current interest in further studying NF1 cerebral vasculopathies.

Matthieu Robert

THE HEIDENHAIN VARIANT OF CREUTZFELDT-JAKOB DISEASE

Parker SE, Gujrati M, Pula JH, Zallek SN, Kattah JC. The Heidenhain variant of Creutzfeldt-Jakob disease—a case series. *J Neuroophthalmol* 2014;34:4–9.

The authors retrospectively studied 3 patients with the Heidenhain variant of Creutzfeldt-Jakob disease (HvCJD), 2 patients confirmed by postmortem neuropathologic, immunohistochemical, and genetic studies. Rapid rate of visual and neurologic deterioration and abnormal diffusion-weighted imaging (DWI) were characteristic for HvCJD. The authors concluded that the combination of clinical neuroimaging and electroencephalographic (EEG) studies and 14:3:3 protein and other neuronal protein marker levels can lead to the diagnosis, as will immunohistochemical analysis and genetic testing at a specialised prion research centre.

Michael Vaphiades

AUTOIMMUNE RETINOPATHY

Grange L, Dalal M, Nussenblatt RB, Sen HN. Autoimmune retinopathy. *Am J Ophthalmol* 2014;157:266–272.

The authors provided a detailed review of the current clinical guidelines for the diagnosis, workup, and treatment of autoimmune retinopathy. Diagnostically, it is characterised by the presence of circulating anti-retinal antibodies along with electroretinographic and visual field abnormalities. They recognise, however, that little is known about the clinical course, prognosis, and treatment of this rare immunologic disease.

Michael Vaphiades

INFILTRATION OF THE VISUAL PATHWAY BY GLIOMATOSIS CEREBRI

Traynis I, Singer S, Winterkorn J, Rosenblum M, Dinkin M. Infiltration of the optic chiasm, nerve, and disc by gliomatosis cerebri. *J Neuroophthalmol* 2014;34:44–46.

The authors report an 18-year-old man with right frontal lobe gliomatosis cerebri and tumour infiltration of the optic chiasm and right optic nerve including the optic disc. This is a rare manifestation of this type of tumour.

Michael Vaphiades

SAGGING EYE SYNDROME

Chaudhuri Z, Demer JL. Sagging eye syndrome: connective tissue involution as a cause of horizontal and vertical strabismus in older patients. *JAMA Ophthalmol* 2013;131:619–625.

The authors used magnetic resonance imaging to evaluate rectus extraocular muscles (EOMs), pulleys, and the lateral rectus (LR)–superior rectus (SR) band ligament in patients with acquired diplopia of 11 men and 17 women (56 orbits) suspected of having sagging eye syndrome (SES) as the aetiology of their diplopia. They noted that patients with SES commonly exhibit blepharoptosis and superior sulcus defect. Significant inferolateral LR pulley displacement was confirmed in SES, but the spectrum of abnormalities was extended to peripheral displacement of all other rectus pulleys and lateral displacement of the inferior rectus pulley, with elongation of rectus EOMs. They concluded that small-angle esotropia or hypertropia may result from common involutional changes in EOMs and orbital connective tissue.

Michael Vaphiades

NEURO-OPHTHALMOLOGIC FEATURES OF CHORDOID GLIOMA

Al-Zubidi N, McGlynn MM, Chévez-Barrios P, Yalamanchili S, Lee AG. Neuro-ophthalmologic features of chordoid glioma. *J Neuroophthalmol* 2014;34:47–49.

In this photo essay, the authors describe the clinical, neuroimaging, and pathologic features of chordoid glioma in a patient presenting with a chiasmal syndrome.

Michael Vaphiades

FATAL ISCHAEMIC STROKE COMPLICATING ACUTE MULTIFOCAL PLACOID PIGMENT EPITHELIOPATHY

Tsang BK, Chauhan DS, Haward R, Whiteman I, Frayne J, McLean C. Fatal ischemic stroke complicating acute multifocal placoid pigment epitheliopathy: histopathological findings. *J Neuroophthalmol* 2014;34:10–15.

The authors report a 29-year-old man with acute left hemiparesis followed shortly by de novo convulsive status epilepticus. This was in the context of a 2-month history of flu-like symptoms, severe headaches, and retinopathy recently diagnosed as acute multifocal placoid pigment epitheliopathy (AMPPE). Neuroimaging demonstrated bilateral, multiple territory cerebral infarction. Despite intravenous methylprednisolone and craniotomy for the management of raised intracranial pressure, the patient deteriorated and died 14 days later. At autopsy, multiple infarcts of varying ages within a 10-day period were seen in association with a segmental giant cell vasculopathy of meningeal arteries. Stroke may develop in patients with AMPPE and usually develops within months, yet extensive and multiple territory strokes resulting in death, as in this patient, is rare.

Michael Vaphiades

BILATERAL OPTIC NERVE INVOLVEMENT IN IGG4-RELATED OPHTHALMIC DISEASE

Takahashi Y, Kitamura A, Kakizaki H. Bilateral optic nerve involvement in immunoglobulin G4-related ophthalmic disease. *J Neuroophthalmol* 2014;34:16–19.

The authors report a 62-year-old man with vision loss and a history of sinus surgery showing dense infiltration of immunoglobulin G4 (IgG4)-positive plasma cells. Computed tomography showed masses surrounding both optic nerves at the orbital apices and bilaterally enlarged infraorbital nerves. The patient underwent two cycles of intravenous pulse steroid therapy followed by a taper of oral steroids and his vision normalised; although the serum level of IgG was within normal limits, the IgG4 level remained elevated. The authors concluded that IgG4 may involve the optic nerves and steroid administration is the primary treatment. A slow taper however is essential to avoid relapse.

Michael Vaphiades

Neuro-Ophthalmology

RECOVERY OF OCULAR MOTOR CRANIAL NERVE PALSY AFTER HERPES ZOSTER OPHTHALMICUS

Chhabra MS, Golnik KC. Recovery of ocular motor cranial nerve palsy after herpes zoster ophthalmicus. *J Neuroophthalmol* 2014;34:20–22.

The authors performed a retrospective chart review of 21 patients with ocular motor cranial nerve palsy occurring at the time of herpes zoster ophthalmicus. Nine (50%) had complete recovery and 8 (44%) had partial recovery but no diplopia in primary gaze. One patient with complete ophthalmoplegia had persistent diplopia in primary position for recovery. The authors concluded that ophthalmoplegia secondary to herpes zoster ophthalmicus has good long-term prognosis for recovery.

Michael Vaphiades

SPONTANEOUS CEREBROSPINAL FLUID LEAK REPAIR

Chaaban MR, Illing E, Riley KO, Woodworth BA. Spontaneous cerebrospinal fluid leak repair: a five-year prospective evaluation. *Laryngoscope* 2014;124:70–75.

The authors prospectively evaluated 46 patients (average age, 51 years) with 56 spontaneous cerebrospinal fluid (CSF) leaks treated by a single otolaryngologist. Over 5 years, 21 subjects presented with recurrence of their CSF leak following previous endoscopic and/or open approaches by other physicians. Obesity was present in 78% of individuals. Fifty-two CSF leaks (93%) were successfully repaired at first attempt. With secondary repair, all CSF leaks were closed at last clinical follow-up averaging 93 weeks. Opening pressures via lumbar puncture averaged 24.3 ± 8.3 cm H₂O, which increased significantly to 32.3 ± 9.0 cm H₂O following closure of the skull base defect. Management of intracranial hypertension included acetazolamide or permanent CSF diversion, including five revisions of failed preexisting shunts.

Michael Vaphiades

RETINAL GANGLION CELL FUNCTION IN ASYMPTOMATIC LHON CARRIERS

Guy J, Feuer WJ, Porciatti V, Schiffman J, Abukhalil F, Vandenbroucke R, Rosa PR, Lam BL. Retinal ganglion cell dysfunction in asymptomatic G11778A: Leber hereditary optic neuropathy. *Invest Ophthalmol Vis Sci* 2014;55:841–848.

This is a 3-year follow-up study reporting serial visual function testing in asymptomatic G11778A Leber hereditary optic neuropathy (LHON) carriers. This follows the initial report of the LHON Gene Therapy Clinical Trial in 2010. The authors examined 45 LHON carriers (6 male and 39 female). They found that visual acuity, visual fields, and retinal nerve fibre layer thickness remained stable, but pattern ERG mean amplitudes dropped progressively by ~40% over 36 months. The authors conclude that there is a subclinical progressive dysfunction of retinal ganglion cells in asymptomatic G11778A carriers.

An-Guor Wang

90° AUTOMATED VISUAL FIELD AND SD-OCT IN ANTERIOR ISCHAEMIC OPTIC NEUROPATHY

Kernstock C, Beisse F, Wiethoff S, Mast A, Krapp E, Grund R, Dietzsch J, Lagrèze W, Fischer D, Schiefer U. Assessment of functional and morphometric endpoints in patients with non-arteritic anterior ischemic optic neuropathy (NAION). *Graefes Arch Clin Exp Ophthalmol* 2014;52:515–521.

The authors in this article report on the natural course of non-arteritic anterior ischaemic optic neuropathy (NA-AION) in a cohort of patients. They conducted a bi-centre study assessing functional and morphological features over 6 months from 16 patients with NA-AION. Ninety-degree automated perimetry (Octopus 101) revealed functional deficits immediately after onset of NAION, which did not change significantly over time. SD-OCT showed retinal nerve fibre layer (RNFL) swelling after onset, decreasing below normal limits at 2 months

in 80% of the patients. The authors conclude that functional defects were present from disease onset and remained unchanged irrespective of morphological changes.

An-Guor Wang

SIMULATING A NEUROLOGIC FIELD DEFECT

Ghate D, Bodnarchuk B, Sanders S, Deokule S, Kedar S. The ability of healthy volunteers to simulate a neurologic field defect on automated perimetry. *Ophthalmology* 2014;121:759–762.

Can a neurologic field defect be simulated by a healthy subject on automated perimetry testing with a short-duration stimuli? The authors conducted a study to assess for this possibility using a Humphrey SITA Fast 24-2 protocol. Thirty healthy volunteers new to automated perimetry were shown a diagram of a defect (hemianopia, quadrantanopia, central scotoma) to be simulated before the perimetry test. The authors found a high success rate and excellent reliability for these volunteers to simulate the neurologic field defects. The authors conclude that it is not difficult to simulate neurologic field defects on automated perimetry.

An-Guor Wang

A CASE OF BIOTINIDASE DEFICIENCY-RELATED OPTIC NEUROPATHY

Haines SR, Reid A, Longmuir RA. Optic neuropathy due to biotinidase deficiency in a 19-year-old man. *JAMA Ophthalmol* 2014;132:228–230.

Biotin, or vitamin B7, is an important water-soluble nutrient that aids in the metabolism of fats, carbohydrates, and proteins. Whereas most vitamins are loosely bound to proteins, biotin is covalently bound with protein, and cannot be easily released from food without biotinidase. Biotinidase deficiency is an autosomal recessive metabolic disorder. An infant with biotinidase deficiency may develop seizures, hypotonia, hearing loss, skin rashes, and optic atrophy. The authors report a case of biotin deficiency presenting as optic neuropathy in a 19-year-old.

An-Guor Wang