ABSTRACTS

Neuro-Ophthalmic Literature Review

Carmen K. M. Chan, Evan Price, John H. Pula, Matthieu Robert, Michael Vaphiades, An-Guor Wang, and Sui Wong

ACUTE RETINAL ARTERY OCCLUSION IS FREQUENTLY ASSOCIATED WITH ACUTE CEREBRAL INFARCTION

Lee J, Kim SW, Lee SC, Kwon OW, Kim YD, Byeon SH. Co-occurrence of acute retinal artery occlusion and acute ischemic stroke: diffusion-weighted magnetic resonance imaging study. *Am J Ophthalmol* 2014;157:1231–1238.

Acute retinal artery occlusion (RAO) is essentially a stroke in the eye; therefore, it shares the same risk factors as acute cerebral infarctions. In fact, the American Heart Association and American Stroke Association recommend immediate brain imaging in all patients with acute retinal ischaemia. In this retrospective observational case series, the authors reviewed the results of diffusion-weighted (DW) magnetic resonance imaging (MRI) of the brain, done within 7 days of visual symptom onset, in 33 consecutive patients with acute RAO (18 central [CRAO], 15 branch [BRAO]). Acute cerebral infarctions were diagnosed on MRI in 8 patients (24.2%), including 5/18 (27.8%) of CRAO and 3/15 (20%) of BRAO patients. Three patients with acute cerebral infarctions had no systemic neurological symptoms. The cause of the RAO was identified in all patients with concomitant cerebral infarction (including atherosclerosis, cardiac embolism, etc.), but only in 36% of patients without cerebral infarction. Due to the high incidence of co-occurrence of cerebral infarction in patient with RAO, the authors recommended DW MRI in patients with RAO, and the patients should be worked up for stroke risk factors, particularly if the MRI revealed cerebral infarction.

Carmen Chan

THE MANAGEMENT OF PATIENTS WITH TEMPORARY MONOCULAR BLINDNESS

Naylor AR, Robinson TG, Eveson D, Burns J. An audit of management practices in patients with suspected temporary monocular blindness. *Br J Ophthalmol* 2014;98:730–733.

The highest risk of stroke after a transient ischaemic attack (TIA) is within the first 14 days, and temporary monocular blindness (amaurosis fugax) is a form of TIA. In 2008, the UK National Institute of Health and Care Excellence advised that patients with significant carotid stenosis should undergo carotid endarterectomy within 14 days of the index TIA. The Leicester Royal Infirmary, where the authors work, had set up a daily one-stop TIA clinic in 2008, where patients would see a stroke medicine consultant and receive brain imaging and carotid Doppler ultrasound imaging. In the EXPRESS study, a combination of being seen promptly in a TIA clinic and starting statin/antiplatelet agents resulted in a 80% reduction of the 90-day stroke risk. Their hospital protocol therefore stated all patients should be started on aspirin 300 mg daily and simvastatin 40 mg by the referring doctor, before referral. In this study, the authors audited the use of the TIA clinic by ophthalmologists in 2011 and 2012. It was found that although most ophthalmologists were aware of the TIA clinic, almost no one referred patients there. In addition, whilst most ophthalmologists would prescribe aspirin, almost no one prescribed statins according to protocol. This audit highlighted the fact that, despite the availability of a “one-stop” TIA

clinic, there was a general reluctance for ophthalmologists to refer patients there and a tendency not to follow the best medical practice guidelines. Further physician training is necessary!

Carmen Chan

NO DEFENESTRATION FOR FENESTRATION OR SHUNTING

Fonseca PL, Rigamonti D, Miller NR, Subramanian PS. Visual outcomes of surgical intervention for pseudotumour cerebri: optic nerve sheath fenestration versus cerebrospinal fluid diversion. *Br J Ophthalmol* 2014;98:1360–1363.

In this retrospective case series, the authors compare the visual outcomes of optic nerve sheath fenestration (ONSF) and cerebral spinal fluid (CSF) shunting for patients with papilloedema secondary to pseudotumor cerebri (PTC). This is the first report of such a comparison from a single centre. When the two surgical intervention groups were compared, they were found to be statistically equal in terms of preoperative best-corrected visual acuity (BCVA) and preoperative mean deviation (MD) on 24-2 SITA-standard testing using the Humphrey field analyser. The groups were also found to have equal postoperative BCVA. Postoperative MD was found to be worse in the ONSF group; however, it was also noted that this group had more severe median papilloedema on Frisén grading (grade 4) preoperatively as compared with the CSF shunting group (grade 2). Given the nature of this retrospective study, they were unable to analyse whether the difference in the amount of improvement in MD was significant between the groups, it is interesting to see that both procedures improved MD by an average of 6 dB. Complications and further visual loss did not vary significantly between the groups. The authors note that it is also important that while the differences in preoperative BCVA and MD between the groups were not statistically significant, such clinical factors may have influenced the surgeon's decision of procedure. Prominent PTC symptomatology (headache vs. papilloedema/visual loss) is also a known clinical factor that may have persuaded the team toward one procedure or the other. This study is helpful for the management of severe PTC and supports a prospective, randomised trial to further delineate the appropriate uses of ONSF and CSF shunting.

Evan Price

DAMAGE TO THE OPTIC RADIATION IN GLAUCOMA: A TOPOGRAPHICAL CORRELATION

Kaushik M, Graham SL, Wang C, Klistorner A. A topographical relationship between visual field defects and optic radiation changes in glaucoma. *Invest Ophthalmol Vis Sci* 2014;55:5770–5775.

Diffusion tensor imaging (DTI) has augmented the investigation of effects of various neuro-ophthalmic conditions on the visual pathways, particularly the optic radiation (OR). In this cross-sectional study, the authors aim to evaluate transsynaptic degeneration of retrogeniculate axons in glaucoma. Nine patients with primary open angle glaucoma and bilateral superior or bilateral inferior altitudinal visual field defects on Humphrey visual field testing were included in the study. Their unaffected hemifields were reportedly relatively well preserved. Nine healthy age- and sex-matched controls were also included in the study. Although prior studies have been conducted citing changes in the OR in glaucoma, this is the first study to note a topographic correlation between the damaged hemifield and the specific region of retrogeniculate damage. The affected hemifields in the glaucoma subjects were found to have lower average absolute thresholds on Humphrey visual field test than their unaffected hemifields, as well as corresponding retinal nerve fibre layer thinning on optical coherence tomography. The OR fibre bundles that correlate to these affected hemifields were found to be significantly decreased in number (3-fold to 4-fold) as compared with the unaffected hemifields in the same patients. In addition, the OR fibres in the glaucoma subjects that corresponded to both the affected and unaffected hemifields were found to have increased radial diffusivity (RD) and essentially unchanged axial diffusivity (AD) as compared with the controls. Mean diffusivity (MD) and fractional anisotropy (FA), which rely on RD and AD for their computation, were also significantly different as compared with the controls. It is argued that increased RD may reflect a decrease in the integrity of myelin or cell membranes, gliosis, or cytoskeletal changes. As the OR fibres that correlate to the unaffected hemifield were also noted to be different in quality from the control group, the authors speculate that DTI may be helpful in the evaluation of early glaucoma.

Evan Price

HOSPITALISATION RATES IN MULTIPLE SCLEROSIS OVER TWENTY-FIVE YEARS

Marrie RA, Elliott L, Marriott J, Cossoy M, Blanchard J, Tennakoon A, Yu N. Dramatically changing rates and reasons for hospitalization in multiple sclerosis. *Neurology* 2014;83:929–937.

Using health claims data from the Canadian province of Manitoba, the authors analysed the rates and causes of hospitalisation in 5797 patients with multiple sclerosis (MS) as compared against 28,769 age-, sex-, and postal code-matched controls. The data were composed of 25 years of claims spanning the period 1984–2011. It was found that the proportion of admissions for MS-related reasons dropped in the MS population significantly over this time (43.4% in 1984 to 7.8% in 2011). It was also discovered that the biggest declines in this proportion occurred before the advent of disease-modifying therapy. The authors speculate that changes in healthcare delivery may be at the root of such gains, such as the use of outpatient infusion therapy or increased utilisation of home care services, as well as better management of symptoms. Among the MS group, it was also noted that the all-cause hospitalisation rate decreased to a larger extent than that of the general population. However, admissions due to bacterial pneumonia, influenza, urinary tract infection, and pressure ulcer were more common than in controls. The authors assert that measures to prevent these specific conditions are needed to further reduce the hospitalisation rate in the MS population.

Evan Price

OCULAR NEUROMYOTONIA AND DEMYELINATING DISEASE

Menon D, Sreedharan SE, Gupta M, Nair M. A novel association of ocular neuromyotonia with brainstem demyelination: two case reports. *Mult Scler* 2014;20:1409–1412.

The authors describe two patients (both 15-year-old females) without previous neurologic symptoms who presented with episodic diplopia. Both patients were examined during symptomatic episodes and were found to have episodic deviation in abduction or abduction with supraduction. In both cases, brainstem demyelination was found. It was not reported if acetylcholine receptor antibodies were tested, but alternate cover test did rule out a latent phoria. The episodes were also not precipitated by sustained gaze in muscle direction, which is common in ocular neuromyotonia. Nevertheless, these case reports give support for imaging patients with episodic diplopia and also for considering ocular neuromyotonia as a cause of diplopia in demyelinating disease.

John H. Pula

NEW INSIGHTS IN BPES PLUS MICROCEPHALY

Dean SJ, Holden KR, Dwivedi A, Dupont BR, Lyons ML. Acquired microcephaly in blepharophimosis-ptosis-epicanthus inversus syndrome because of an interstitial 3q22.3q23 deletion. *Pediatr Neurol* 2014;50:636–639.

The authors report the case of a child with a blepharophimosis-ptosis-epicanthus inversus syndrome associated with microcephaly—a known contiguous gene deletion syndrome—and a deletion of chromosome 3q22.2q23, but which did not include the *ATR* gene, unlike previously reported cases with a similar phenotype. Instead, the authors hypothesise that the *PIK3CB* gene, which was included in the deletion, is responsible for the microcephaly in such cases.

Matthieu Robert

A NEW OCULAR MOTOR SIGN IN JOUBERT SYNDROME

Papanagnu E, Klaehn LD, Bang GM, Ghadban R, Mohny BG, Brodsky MC. Congenital ocular motor apraxia with wheel-rolling ocular torsion—a neurodiagnostic phenotype of Joubert syndrome. *J AAPOS* 2014;18:404–407.

The authors report two cases of children with Joubert syndrome and peculiar eye movements visible during ophthalmoscopic examination of the retina: a tonic, extreme, alternating cyclodeviation with a periodicity of 10–15

seconds in both eyes. In the first cases, these movements corresponded to a cyclic skew deviation. In the second case, they consisted in a periodic alternating skew deviation accompanied by a paroxysmal ocular tilt reaction (OTR) and they had been previously mistaken for a dissociated vertical divergence. As such movements share similarities with both seesaw nystagmus—classically seen in Joubert syndrome—and the paroxysmal OTR, the authors postulate that they must be modulated by the neuroanatomical pathways that normally subserve tilt rotation in the roll plane.

Matthieu Robert

A NEW ASSOCIATION INCLUDING CONGENITAL MYDRIASIS

Brodsky MC, Turan KE, Khanna CL, Patton A, Kirmani S. Congenital mydriasis and prune belly syndrome in a child with an *ACTA2* mutation. *J AAPOS* 2014;18:393–395.

Congenital mydriasis is a common misdiagnosis for aniridia in neonates when examined by non-specialists. Although much rarer, it does exist and is indeed—along with some other anterior segment dysgenesis—a differential of aniridia. It can be associated with generalised smooth muscle dysfunction, involving the bladder, bowel, and vessels. Brodsky et al. describe the association of a congenital mydriasis with iris hypoplasia and filiform radial iris strands, a prune belly syndrome (a rare congenital malformation with a wrinkled appearance of the abdomen, including loose abdominal musculature, urinary tract anomalies, cryptorchidism, limb deformities, and cardiac defect), intracranial arteries anomalies (dilation of the intracranial carotid arteries and marked tortuosity of the distal cerebral vasculature without moyamoya vessels), tortuosity, and dilatation of the retinal vasculature. An Arg179His mutation was found in the *ACTA2* gene, now known to be responsible for various phenotypes of generalised smooth muscle dysfunction.

Matthieu Robert

CONGENITAL PROSOPAGNOSIA, POOR FACE RECOGNITION, AND VISUOPERCEPTUAL DEFECTS IN ONE FAMILY

Johnen A, Schmukle SC, Hüttenbrink J, Kischka C, Kennerknecht I, Dobel C. A family at risk: congenital prosopagnosia, poor face recognition and visuoperceptual deficits within one family. *Neuropsychologia* 2014;58:52–63.

The authors administered standardised tests for face processing, a self-report questionnaire and general visual processing tests to an extended family of 28, in which many members reported difficulties with face recognition. They found evidence for a severe face processing (prosopagnosia), but intact nonface visuoperceptual skills in three family members—a father and his two sons—who fulfilled criteria for developmental prosopagnosia, supporting a familial transmission.

Michael Vaphiades

OPTIC NEUROPATHY CAUSED BY *PROPIONIBACTERIUM ACNES* PACHYMENINGITIS

Adesina OO, Stagg BC, Digre KB, Katz BJ, Quigley EP, Palmer CA, Warner JE. Optic neuropathy caused by *Propionibacterium acnes* pachymeningitis. *J Neuroophthalmol* 2014;34:264–267.

The authors report an 82-year-old man with optic neuropathy caused by *Propionibacterium acnes* pachymeningitis. The case and treatment is discussed.

Michael Vaphiades

ISSUES IN THE DIAGNOSIS AND MANAGEMENT OF THE PAPILLOEDEMA SHUNT

Chaudhary N, Griauzde J, Gemmete JJ, Pandey AS, Trobe JD. Issues in the diagnosis and management of the papilledema shunt. *J Neuroophthalmol* 2014;34:259–263.

In this case report and review of dural arteriovenous fistulas (DAVFs), the authors describe “the papilloedema shunt,” which shunts blood into the transverse or superior sagittal venous sinuses raising intracranial pressure and thus causing papilloedema. The imaging features of a DAVF that cause papilloedema may be subtle. If not detected on noninvasive neuroimaging, and if lumbar puncture is performed, there is a substantial risk of brain herniation. Digital subtraction catheter angiography is necessary to definitively diagnose and treat the disorder.

Michael Vaphiades

EYE MOVEMENTS IN VISUAL FIELD DEFECTS VERSUS PURE ALEXIA

Bormann T, Wolfer SA, Hachmann W, Lagrèze WA, Konieczny L. An eye movement study on the role of the visual field defect in pure alexia. *PLoS ONE* 2014;9:e100898.

Bormann et al. performed two experiments comparing the eye movements of four patients with pure alexia with controls with simulated visual field defects (sVFD) when reading single words. The analyses revealed quantitative and qualitative differences between pure alexic patients and unimpaired individuals reading with sVFD. The patients with pure alexia read words slower and exhibited more fixations. The serial, letter-by-letter fixation strategy was observed only in the patients but not in the controls with sVFD. The authors argued that sVFDs do not cause pure alexic reading.

Michael Vaphiades

TERSON'S SYNDROME IN PATIENTS WITH SUBARACHNOID HAEMORRHAGE

Skevas C, Czorlich P, Knospe V, Stemplewitz B, Richard G, Westphal M, Regelsberger J, Wagenfeld L. Terson's syndrome-rate and surgical approach in patients with subarachnoid hemorrhage: a prospective interdisciplinary study. *Ophthalmology* 2014;121:1628–1633.

In this prospective, uncontrolled, interdisciplinary study of 102 patients with subarachnoid haemorrhage (SAH), over a period of 24 months the authors analysed the need for surgical intervention in Terson's syndrome (TS) as well as the effect of pars plana vitrectomy (PPV) with or without internal limiting membrane (ILM) peeling. They concluded that pars plana vitrectomy and ILM peeling have beneficial effects on the visual rehabilitation of patients with non-clearing vitreous haemorrhage.

Michael Vaphiades

VISUOPERCEPTION TEST PREDICTS PATHOLOGIC DIAGNOSIS OF ALZHEIMER DISEASE IN CORTICOBASAL SYNDROME

Boyd CD, Tierney M, Wassermann EM, Spina S, Oblak AL, Ghetti B, Grafman J, Huey E. Visuoperception test predicts pathologic diagnosis of Alzheimer disease in corticobasal syndrome. *Neurology* 2014;83:510–519.

Frequently, patients with corticobasal syndrome (CBS) are found to have Alzheimer-type findings on autopsy. This Alzheimer disease (AD) pathology in CBS has implications in treatment and thus is useful information in these patients. The authors assessed 36 patients with the Visual Object and Space Perception Battery (VOSP) to distinguish AD from non-AD pathology in CBS. All patients were autopsied for definitive diagnosis. They concluded that in patients with CBS, failure on specific VOSP subtests is associated with greater odds of having underlying AD.

Michael Vaphiades

DIFFERENTIATING COMPRESSIVE FROM GLAUCOMATOUS OPTIC NEUROPATHY WITH SD-OCT

Danesh-Meyer HV, Yap J, Frampton C, Savino PJ. Differentiation of compressive from glaucomatous optic neuropathy with spectral-domain optical coherence tomography. *Ophthalmology* 2014;121:1516–1523.

The authors compared the optic disc topography in a total of 200 eyes with compressive optic neuropathy (CON) and open-angle glaucoma (OAG) using spectral-domain (SD) optical coherence tomography (OCT) and Heidelberg retinal tomograph (HRT). They discovered that compressive optic neuropathy is associated with significantly thinner nasal and temporal sectors compared with OAG, whereas OAG results in larger cups and cup volume with OCT measurements. The Heidelberg retinal tomograph was not able to differentiate CON from normal discs.

Michael Vaphiades

SELF-FACE HALLUCINATION EVOKED BY ELECTRICAL STIMULATION OF THE HUMAN BRAIN

Jonas J, Maillard L, Frismand S, Colnat-Coulbois S, Vespignani H, Rossion B, Vignal JP. Self-face hallucination evoked by electrical stimulation of the human brain. *Neurology* 2014;83:336–338.

Autoscopic hallucination (AH) is part of a larger class of such disorders called autoscopic phenomena, which are defined as illusory own-body perceptions, during which patients experience the visual illusory reduplication of their own body. Autoscopic hallucinations may be confined to the image of one's face (self-face hallucination) or head and shoulders, but they can also consist of more or less complete bodily images. The authors report the results of intracerebral electrical stimulations of the right medial occipitoparietal cortex (right precuneus and occipitoparietal sulcus) in two patients with epilepsy who underwent a stereo-EEG (electroencephalography). Immediately after the onset of the stimulation, the two patients reported seeing their current own face, facing themselves, in their left visual field. The authors concluded that the medial occipitoparietal junction has a key role in generating AH.

Michael Vaphiades

PREVENTING VISUAL FIELD DEFICITS AFTER NEUROSURGERY

Winston GP, Daga P, White MJ, Micallef C, Miserocchi A, Mancini L, Modat M, Stretton J, Sidhu MK, Symms MR, Lythgoe DJ, Thornton J, Yousry TA, Ourselin S, Duncan JS, McEvoy AW. Preventing visual field deficits from neurosurgery. *Neurology* 2014;83:604–611.

The authors assessed 21 patients who underwent anterior temporal lobe resection (ATLR) for refractory temporal lobe epilepsy (TLE) while employing optic radiation tractography in an attempt to reduce postoperative visual field deficits (VFD). They noted VFD in the contralateral superior quadrant were significantly less using intraoperative MRI (iMRI) guidance than without this technique. No patient in the iMRI cohort developed a VFD that precluded driving, whereas 13% of the non-iMRI cohort failed to meet regional driving standards. Seizure outcome and degree of hippocampal resection were unchanged.

Michael Vaphiades

VISUAL OUTCOMES IN PAEDIATRIC OPTIC NEURITIS

Wan MJ, Adebona O, Benson LA, Gorman MP, Heidary G. Visual outcomes in pediatric optic neuritis. *Am J Ophthalmol* 2014;158:503–507.

The authors retrospectively reviewed 59 paediatric patients with first-episode optic neuritis with the main outcome measures being visual acuity at 3 months and 1 year of follow-up. They determined

that the majority of patients regained normal visual acuity at 1 year, regardless of baseline clinical characteristics.

Michael Vaphiades

ACUTE VISION LOSS: JUST THE BEGINNING?

Colpak AI, Isikay I, Mut M, Soylemezoglu F, Kansu T, Foroozan R. Acute visual loss: just the beginning? *Surv Ophthalmol* 2014;59:548–552.

The authors present a 47-year-old man with sudden visual loss OD to no light perception (NLP), optic disk oedema, retinal ischaemia, and limited upgaze in the left eye. Cranial magnetic resonance imaging (MRI) revealed a thickened, enhancing left optic nerve. The patient's vision OD soon became affected, with worsening MRI necessitating a brain biopsy, which revealed glioblastoma. The paper discusses malignant optic nerve gliomas of adulthood.

Michael Vaphiades

THIRD NERVE PALSY AS THE INITIAL MANIFESTATION OF GCA

Thurtell MJ, Longmuir RA. Third nerve palsy as the initial manifestation of giant cell arteritis. *J Neuroophthalmol* 2014;34:243–245.

The authors present four patients with third nerve palsy as the presenting sign of giant cell arteritis (GCA). One patient had a complete third nerve palsy with pupil involvement, whereas the other three had third nerve palsies without pupil involvement. All patients had rapid improvement in symptoms and signs after high-dose oral prednisone was started, with all showing complete recovery within weeks. The authors concluded that GCA can rarely present with acute painful third nerve palsy, mimicking the presentation of a microvascular cause, and the presence of GCA symptoms or elevated inflammatory markers in a patient older than 50 years with an acute third nerve palsy should prompt initiation of high-dose steroid treatment and temporal artery biopsy.

Michael Vaphiades

VISUAL RECOVERY AFTER SURGICAL DECOMPRESSION OF AN OCCIPITAL INTRAVENTRICULAR CYST

Vedantam A, Yoshor D, Foroozan R. Visual recovery after surgical decompression of an occipital intraventricular cyst. *J Neuroophthalmol* 2014;34:255–256.

The authors present a 62-year-old woman with a chronic left homonymous visual field defect because of a right occipital cyst. Serial visual field examination documented stable visual fields for 12 months, after which there was worsening of visual fields associated with enlargement of the cyst. Surgical decompression of the occipital cyst resulted in marked improvement of the visual field defect over 9 months. This case demonstrates that surgical decompression of cystic lesions adjacent to posterior visual pathways can result in recovery of chronic visual field loss.

Michael Vaphiades

OPTOCHIASMATIC AND PERIPHERAL NEUROPATHY DUE TO ETHAMBUTOL OVERTREATMENT

Geyer HL, Herskovitz S, Slamovits TL, Schaumburg HH. Optochiasmatic and peripheral neuropathy due to ethambutol overtreatment. *J Neuroophthalmol* 2014;34:257–2578.

The authors present a 72-year-old man who developed visual loss and paresthesias after 11 weeks of exposure to a supratherapeutic dose of ethambutol. This case demonstrates the selective vulnerability of the anterior visual pathways and peripheral nerves to ethambutol toxicity.

Michael Vaphiades

INTRAVENTRICULAR TUMOUR PRESENTING AS PROGRESSIVE SUPRANUCLEAR PALSY

Morelli M, Fera F, Bono F, Fratto A, Arabia G, Quattrone A. Intraventricular tumor presenting as progressive supranuclear palsy-like phenotype. *Neurology* 2014;83:948.

The authors present a 70-year-old woman who presented like a progressive supranuclear palsy (PSP) patient but had a globular tumour compressing the midbrain. The authors conclude that neuro-imaging should be part of the work-up of PSP.

Michael Vaphiades

HICKAM'S DICTUM

Shaw HE. Tigers and snakes in neuro-ophthalmology. *J Neuroophthalmol* 2014;34:213–217.

The author reports three cases that ascribe to Hickam's dictum, which indicates that patients can have many illnesses and we cannot always ascribe the diagnosis to one disease. This is the counter-argument to Occam's razor, which implies that the simplest diagnosis is usually the best. The author cautions that the obvious diagnosis may not be the most important one. I personally am fond of a quote by Sherlock Holmes stating "when you have eliminated the impossible, whatever remains, however improbable, must be the truth".

Michael Vaphiades

EARLY OCT CHANGE IN DOMINANT OPTIC ATROPHY

Barboni P, Savini G, Cascavilla ML, Caporali L, Milesi J, Borrelli E, La Morgia C, Valentino ML, Triolo G, Lembo A, Carta A, De Negri A, Sadun F, Rizzo G, Parisi V, Pierro L, Bianchi Marzoli S, Zeviani M, Sadun AA, Bandello F, Carelli V. Early macular retinal ganglion cell loss in dominant optic atrophy: genotype-phenotype correlation. *Am J Ophthalmol* 2014;158:628–636.

Time-domain OCT has been used to examine patients with dominant optic atrophy (DOA), and a significant thinning of the peripapillary retinal nerve fibre layer (RNFL) was found, especially in the temporal and inferior quadrants. In this study, the authors examined 39 patients from 28 pedigrees with DOA using spectral domain OCT, which has the advantage of assessing macular retinal ganglion cells (RGCs) by measuring thickness of the ganglion cell inner plexiform layer (GC-IPL). They found that in DOA, loss of macular RGCs, as reflected by thinning of GC-IPL, occurs earlier than changes in the peripapillary RNFL thickness.

An-Guor Wang

VISUAL OUTCOME IN TRAUMATIC OPTIC NEUROPATHY FOLLOWING ERYTHROPOETIN

Entezari M, Esmaeili M, Yaseri M. A pilot study of the effect of intravenous erythropoietin on improvement of visual function in patients with recent indirect traumatic optic neuropathy. *Graefes Arch Clin Exp Ophthalmol* 2014;52:1309–1313.

Erythropoietin (EPO) is a glycoprotein hormone synthesised in the kidney. It may regulate red blood cell production in response to tissue hypoxia. Recently, it has been shown to exhibit a beneficial effect in the treatment

of patients with non-arteritic anterior ischaemic optic neuropathy. Here, the authors conducted a pilot study to evaluate the clinical efficacy of intravenous EPO in 18 eyes of 18 patients with recent indirect traumatic optic neuropathy. They found there is a significant improvement of visual acuity after EPO treatment compared with baseline.

An-Guor Wang

DON'T FORGET ABOUT OBSTRUCTIVE SLEEP APNOEA

Fraser CL. Obstructive sleep apnea and optic neuropathy: is there a link? *Curr Neurol Neurosci Rep* 2014;14:465.

Dr. Fraser comprehensively reviewed the current evidence of obstructive sleep apnoea (OSA) and its association with optic neuropathy, in particular, glaucoma, non-arteritic anterior ischaemic optic neuropathy (NAION), papilloedema, and raised intracranial hypertension. Evidence from studies of OSA in glaucoma is more convincing for normal tension glaucoma compared with primary open angle glaucoma. There is also a higher proportion of OSA in patients with NAION compared with controls. Possible mechanisms contributing to glaucoma or NAION in OSA include direct hypoxia or vascular dysregulation at the optic nerve head. Papilloedema with raised intracranial pressure is also associated with OSA, possibly due to increased cerebral perfusion pressure and cerebral venous dilation secondary to hypoxia and hypercapnia. It is not clear whether the association of these conditions with OSA is confounding or causative. Nevertheless, this review reminds us of the importance of looking for signs or symptoms of OSA in patients with glaucoma, NAION, and papilloedema.

Sui Wong

CHARLES BONNET SYNDROME LASTS LONGER THAN PREVIOUSLY THOUGHT, AND EDUCATION HELPS

Cox T, Ffytche D. Negative outcome Charles Bonnet syndrome. *Br J Ophthalmol* 2014;98:1236–1239.

This is the largest phenomenological survey of Charles Bonnet syndrome (CBS) to date. This questionnaire survey included 492 people affected by macular disease, identified through the Macular Society. Outcome was judged as negative based on the response to questionnaire on the effect of the hallucinations on their life being “fairly negative” or “very negative”. A key limitation of this study is that patients were identified based on “what they have been told, or judged themselves, to have had CBS”—i.e. potential errors of diagnosis (e.g. Lewy-body disease can cause hallucinations and affects a similar age group of patients). Therefore, some caution has to be applied with the interpretation of the results. Nevertheless, this study is of interest, showing that CBS lasts longer than previously thought, with 75% reported ongoing symptoms for 5 years or longer. At onset of CBS, 38% had a “fear-inducing” response and 46% reported CBS interfering with their daily activities. It is also highlighted that education early (CBS awareness at symptom onset) can help reduce the subjective assessment of negative outcome.

Sui Wong

VISUAL SNOW, OR “WAR OF THE ANTS”, PERHAPS

Schankin CJ, Maniyar FH, Digre KB, Goadsby PJ. “Visual Snow”—a disorder distinct from persistent migraine aura. *Brain* 2014;137:1419–1428.

Bessero AC, Plant GT. Should “visual snow” and persistence of after-images be recognized as a new visual syndrome? *J Neurol Neurosurg Psychiatry* 2014;85:1057–1058.

Schankin CJ, Maniyar FH, Sprenger T, Chou DE, Eller M, Goadsby PJ. The relation between migraine, typical migraine aura and “visual snow”. *Headache* 2014;54:957–966.

What coincidence that two papers on “visual snow” appeared simultaneously in two separate journals—and that both groups propose that “visual snow” should be considered a distinct visual disorder. “Visual snow” is the visual experience of continuous dots in the entire visual field, similar to static noise on an analogue television with no signal (or, according to Bessero and Plant, described as black bugs on white background, e.g. “war of the ants” in Scandinavia).

Bessero and Plant’s retrospective review of 27 patients showed that visual snow was associated with persistent after-images (persistence of an object when fixating and then looking away), or “trails” behind moving objects. These patients were young (2nd to 4th decade), healthy individuals, and predominantly female (2:1). Some patients also had tinnitus (15%) and migraine with aura (25%). Bessero and Plant also described two other groups (without “visual snow”), who had (a) isolated persistence of after-images and (b) solitary persistent phosphenes.

Schankin and colleagues report findings of a retrospective review of 22 patients, an Internet survey of 275 self-assessed data sets, and semi-structured telephone interview of 78 patients. One-quarter of patients had “visual snow” as long as they could remember (i.e. onset in childhood). For the others, onset was at age 21 ± 9 years, with equal female:male ratio. Other visual symptoms associated with “visual snow” included persistent after-images, trailing, floaters, blue field entoptic phenomenon, spontaneous photopsia, self-light of the eye, photophobia, and nyctalopia. Tinnitus, migraine with or without aura, anxiety, and depression were common comorbidities. Standard migraine treatments were unhelpful for the visual symptoms.

Next, Schankin and colleagues reported the results of [(18)F]-2-fluoro-2-deoxy-D-glucose positron emission tomography and statistical parametric mapping on 120 patients with “visual snow” (70 patients with comorbid migraine and 37 with comorbid typical migraine aura). This showed patients with “visual snow” had brain hypermetabolism in the right lingual gyrus and the left cerebellar anterior lobe adjacent to the left lingual gyrus.

Sui Wong