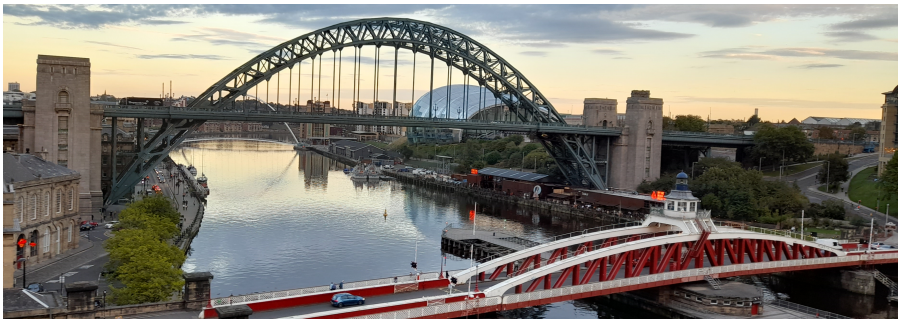
Annual Meeting
8th February 2024
Crowne Plaza Hotel

NEWCASTLE, UK



Uncertainties in Neuro-Ophthalmology

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08.00-09.15	**Neuro-ophthalmology Allied Professionals Breakfast Meeting**	
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2.45-3.00	**Elevated retinal ganglion cell sensitivity in paediatric optic disc oedema.**	Oliver Marmoy Great Ormond Street Hospital for Children NHS Foundation Trust
3.15-3.30	**Evaluation of static and kinetic perimetry practice in chiasmal compression (KINETIC study).**	Michaela Sherlock Faculty of Health and Life Sciences, University of Liverpool

3.30-3.45	**Pre-operative macular ganglion cell layer (GCL) and optic nerve retinal nerve fibre layer (RNFL) thicknesses as a prognostic marker for visual outcomes following pituitary tumour resection (PTR).**	Ben Amram University of Southampton
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Posters

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3	Evaluation of Optometrists' referrals to Ophthalmology for suspected optic disc swelling.	Ved Ramsewak, Chesterfield Royal Hospital
4	Group consultations in patients with idiopathic intracranial hypertension.	Prianka Gandesha, St Thomas' Hospital Eye Dept, London
5	Timing matters: Nocturnal hypotension as a cause of transient vision loss.	Matthew J. Hartley, Department of Ophthalmology, Royal Victoria Infirmary, Newcastle-Upon-Tyne
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7	Unmasking risks: A case study on subtle anterior ischaemic optic neuropathy from hyaluronic acid fillers.	H J Duncan, Department of Ophthalmology, Royal Victoria Infirmary, Newcastle-Upon-Tyne

8	**Visual electrophysiology: Clinical relevance in Neuro-ophthalmology.**	Kong Yien Chin, Evoked Potentials Service, Nottingham University Hospitals NHS Trust, Nottingham
9	**Sertraline induced raised intracranial pressure with bilateral optic nerve swelling.**	Hitesh Kumar Agrawal, Lancashire Teaching Hospitals NHS Trust
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11	**A curious case of downbeat nystagmus: A diagnostic dilemma.**	H Aglan, Ophthalmology Department, Bolton NHS Foundation Trust
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13	**A case report of bilateral optic neuropathy secondary to glial fibrillary acidic protein (GFAP) antibody-associated astrocytopathy.**	Shivaa Ramsewak**,** Medical Eye Unit, St Thomas' Hospital, London
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Speaker Biographies

Michael Burdon

Michael is a consultant ophthalmologist with a subspecialty interest in neuro-ophthalmology. After qualifying from St Thomas' Hospital medical school in 1985, he undertook postgraduate training in medicine at Oxford, ophthalmology at Birmingham, Brisbane and London, and neuro-ophthalmology at St Thomas' Hospital, the National Hospital for Neurology and Neurosurgery, and Moorfields Eye Hospital. In 1997 he was appointed as the first neuro-ophthalmology consultant in Birmingham with a remit to establish a supra-regional neuro-ophthalmology service. This department now has seven consultants, and has a world-wide reputation, training fellows from around the United Kingdom and overseas. In 2021 he moved to take up a part-time post at Bishop Auckland in the northeast of England.

Helena Lee

Helena specialises in Neuro-ophthalmology, Paediatrics and Strabismus and has an international research reputation in the area of infantile nystagmus, paediatric retinal development and optical

coherence tomography . She has researched the effects of idiopathic infantile nystagmus, achromatopsia and albinism on retinal development and was awarded the Fight for Sight Award in 2015 for her work on normal retinal development. She has recently been awarded a £1.4 million MRC Clinician Scientist fellowship to investigate the role of oral levodopa in improving visual development in infants and young children with albinism (the OLIVIA study).

Andrew Schaefer
Andy is consultant neurologist working at RVI since 2007. He also runs a neurogenetic clinic and is the clinical lead for the adult mitochondrial service in Newcastle - one of the 3 national highly specialised services for rare mitochondrial disorders.

Simon Hickman
Simon is a consultant neurologist with an interest in neuro-ophthalmology who works at the Royal Hallamshire Hospital, Sheffield, UK. He trained in neurology in Nottingham and London and carried out research looking into the recovery mechanisms following acute optic neuritis. He is the programme director of UKNOS.

Gemma Maxwell
Gemma is a consultant neurologist based in Sunderland Royal Hospital, having completed her training in the North East. She has an interest in both multiple sclerosis and neuro-ophthalmology and is involved in research in idiopathic intracranial hypertension. Another interest is medical education and she is a year lead for the new University of Sunderland Medical School, where she works her

hardest to reduce the prevalence of neurophobia in the next generation of doctors.

Alan Cunningham

Alan is a consultant neuro-ophthalmologist, ophthalmology IT lead, and head of department at the Newcastle Eye Centre, UK. Alan graduated from Newcastle University and trained in the Northern deanery before undertaking a neuro-ophthalmology fellowship at the Queen Elizabeth Hospital in Birmingham, and then at the Newcastle Eye Centre. He works closely with the pituitary and neurology services, also providing both a high flow cataract service and complex cataract service. Prior to ophthalmic training, Alan completed a Postgraduate Certificate in Medical Education with the University of Cardiff, and is a Fellow of the Higher Education Academy. Alan has a strong interest in technology and cannot leave a broken device alone! He developed programming skills whilst at school before working in a software house delivering election software. He enjoys all aspects of IT, from running a server, to building computers. His knowledge enables a rare link between the clinical and technical staff which has led him to overcome problems that have persisted for years.

Elizabeth Hill

Elizabeth, known by everyone as Liz, is a consultant neuro-ophthalmologist and ophthalmology urgent care lead at County Durham and Darlington NHS Foundation Trust. She completed her ophthalmology training in the North East followed by neuro-ophthalmology fellowships at the Newcastle Eye Centre and Moorfields. Prior to completing her medical training she qualified as an optometrist and worked in clinical trials at Manchester University. She has been involved in UKNOS, formerly UKNOSIG

since 2016 and is the abstract director. She is a keen runner and has completed twenty consecutive Great North Runs.

Gordon Dutton

Professor Gordon Dutton is a retired general and paediatric ophthalmologist who worked for over 20 years at the Royal Hospital for Sick Children in Glasgow. He's now Emeritus Professor of Visual Science at Glasgow Caledonian University. In retirement, in addition to his hobbies of walking and woodworking, Gordon has continued to pursue a special interest in vision and its disorders, and has contributed to a number of publications on this broad topic, and has been an invited speaker on these topics, both nationally and internationally. In his presentation he plans to focus on the visual manifestations of congenital and acquired pathology affecting the middle temporal and posterior parietal lobes, how common this is, how they can easily be missed, and the benefits of identifying them.

Platform presentations

A service evaluation and structure-function analysis of non-infectious paediatric optic neuritis treated at the Evelina Children's Hospital, London

Neil Clough, Paul Nderitu, Ailsa Ritchie
Ophthalmology Department, Evelina Children's Hospital, Guys and St Thomas' Foundation Trust, London

Introduction/Purpose:

To present a service evaluation of paediatric patients with non-infectious optic neuritis seen within the paediatric ophthalmology department at the Evelina Children's Hospital.

Methods:

Retrospective case-note review of all paediatric patients with a diagnosis of optic neuritis identified 19 patients over a 5 year period. Data was collected on demographics, investigations, diagnosis, treatment.

Results:

Age range at presentation from 6-17 years. Myelin oligodendrocyte glycoprotein antibody positive optic neuritis (MOG-ON) was the most common diagnosis (58%), followed by multiple sclerosis associated optic neuritis (MS-ON) (22%), neuromyelitis optica spectrum disorder (NMOSD) (10%), acute disseminated encephalomyelitis (ADEM) (5%) and clinically isolated syndrome (CIS) (5%). At presentation, all patients received intravenous methylprednisolone (IVMP) treatment, 21% received plasma exchange, and 47% were placed on long term immunosuppression. Mean final visual acuity (VA) of affected eyes was 0.13 logMAR (SD 0.39), mean final macular ganglion cell layer (mGCL) volume at 3.45 mm was 0.31 mm^3 (SD 0.08), and mean final peripapillary retinal nerve fibre layer thickness (pRNFL) was 75 µm (SD 17). 63% of affected eyes had normal (0.16 logMAR or better [1]) final VA, 16% had normal (>0.4 mm^3) final mGCL and 16% had normal pRNFL (>90 µm [2]) . Pearson correlation of VA to mGCL r = -0.452 (p = 0.079, R^2 = 0.205), and VA to pRNFL r = -0.150 (p = 0.565, R^2 = 0.023). 33% of patients had OCT abnormalities in the contralateral asymptomatic eye. 92% of patients were found to be vitamin D deficient.

Discussion:
Compared to published data (3), our cohort contains higher numbers of MOG-ON and MS-ON, lower numbers of CIS, similar numbers of NMOSD and ADEM, and lower numbers who achieve normal final VA. This is likely to be a reflection of the status of the Evelina as a paediatric neuro-inflammatory specialist centre. Our data show that VA may be falsely reassuring as a proxy for recovery or a 'good outcome', and that significant irreversible structural retinal changes in mGCL and pRNFL are present on OCT scans in many patients with normal VA. In our cohort, mGCL had a better correlation with VA than pRNFL. A significant number of patients with unilateral symptoms had bilateral OCT changes. We advocate for all paediatric patients with optic neuritis to have OCT scans done of both mGCL and pRNFL, and consider early treatment with IVMP at presentation while awaiting MOG and AQP4 antibody test results.

References:

1) Drover JR, Felius J, Cheng CS, Morale SE, Wyatt L, Birch EE. Normative pediatric visual acuity using single surrounded HOTV optotypes on the Electronic Visual Acuity Tester following the Amblyopia Treatment Study protocol. J AAPOS. 2008 Apr;12(2):145-9. doi: 10.1016/j.jaapos.2007.08.014. Epub 2007 Dec 26. PMID: 18155943; PMCID: PMC2497424.

2) Söhnel T, Meigen C, Hiemisch A, Wahl S, Ziemssen F, Truckenbrod C, Hübner K, Kiess W. Normative data for macular and retinal nerve fibre layer thickness in healthy German children and adolescents using optical coherence tomography. Ophthalmic Physiol Opt. 2023 Jul;43(4):922-934. doi: 10.1111/opo.13123. Epub 2023 Mar 17. PMID: 36930522.

3) Pineles SL, Henderson RJ, Repka MX, Heidary G, Liu GT, Waldman AT, Borchert MS, Khanna S, Graves JS, Collinge JE, Conley JA, Davis PL, Kraker RT, Cotter SA, Holmes JM; Pediatric Eye Disease Investigator Group. The Pediatric Optic Neuritis Prospective Outcomes Study: Two-year results. Ophthalmology. 2022 Aug;129(8):856-864. doi: 10.1016/j.ophtha.2022.03.021. Epub 2022 Mar 29. PMID: 35364222; PMCID: PMC10357378.

Elevated retinal ganglion cell sensitivity in paediatric optic disc oedema

Oliver R. Marmoy[1,2,3], Vasiliki Panteli[1], Richard Bowman[1], Nivedita Desai[1], Prab Prabhakar[1], Emma Hodson-Tole[3], Dorothy A. Thompson[1,2]
[1]Great Ormond Street Hospital for Children NHS Foundation Trust
[2]GOS-ICH University College London
[3]Manchester Metropolitan University, Manchester

Introduction:

Monitoring visual function in children with raised intracranial pressure (ICP) is challenging. We investigated whether the photopic negative response (PhNR) of the electroretinogram (ERG) provides useful information about retinal ganglion cell (RGC) function, that may improve the diagnosis and monitoring of children with, or at risk of, raised ICP.

Methods:

Forty-two patients were recruited prospectively. Patients were categorised according to ICP status (from lumbar puncture opening pressure or ICP bolt) at time of PhNR recording: (1) not raised; (2) equivocal; and (3) raised. The PhNR was recorded using skin electrodes and red-on-blue flash stimulation delivered using a handheld LED stimulator to a range of flash strengths. An ERG luminance-response series was recorded (ranging from -0.3 – 2.4 log cd.s/m^2). The B-PhNR amplitude was modelled using a gaussian curve to derive the coefficients Vmax (maximal amplitude), K (measure of sensitivity), σ (breadth of curve) and μ (position of the gaussian peak). All patients underwent optical coherence tomography (OCT) of the maculae and optic nerves to include global optic nerve head volume (gONHV), and central optic nerve head maximum thickness (cONH-MT).

Results:

Although no significant differences in any PhNR model parameters between ICP status groups ($p > 0.05$) were found, PhNR model coefficients showed significant negative correlations with OCT optic nerve head volume measures; particularly the gaussian curve position. PhNR model coefficients K, σ and μ had significant negative correlations with gONH-V of -0.314, -0.236 and -0.412 ($p = 0.003$, 0.029 and < 0.001, respectively). Significant negative correlations were also observed with cONH-MT of -

0.342, -0.275 and -0.459 ($p = 0.001$, 0.010 and < 0.001, respectively). This demonstrated that larger optic nerve head volumes associated with optic disc oedema relate to a 'leftward position' of the PhNR gaussian sensitivity curve.

Discussion:

This study is the first to explore the PhNR in paediatric conditions of raised ICP. We found a direct correlation between the PhNR luminance-response series and the degree of optic disc oedema. These findings demonstrate that the PhNR luminance-response series is activated by lower flash strengths and reaches Vmax to lower flash strengths in patients with more severe optic disc oedema, suggesting elevated proximal retinal sensitivity. The mechanisms responsible may relate to hyperexcitability of RGCs, or reduced inhibitory activity within retinal gap junctions as a consequence of disrupted axoplasmic flow consequent on the optic disc oedema. These findings suggest a PhNR luminance-response series is beneficial over standard single luminance PhNR recordings in disorders of raised ICP or when exploring RGC sensitivity.

Evaluation of static and kinetic perimetry practice in chiasmal compression (KINETIC study)

Michaela Sherlock, Fiona Rowe, Girvan Burnside, Lauren Hepworth
Faculty of Health and Life Sciences, University of Liverpool

Introduction:

There is clinical significance to the detection of visual field loss in chiasmal compression and capturing peripheral loss is important to the early diagnosis of chiasmal involvement. There is currently no specific perimetry programme recommended to accurately detect visual field loss in neurological conditions.

Aim:

To assess three visual field test programmes (Humphrey full field 120, 30-2 SITA fast and a kinetic template on the Octopus 900) and evaluate the diagnostic accuracy of these programmes in the detection of visual field loss.

Methods:

A prospective, cross-sectional, cohort study was conducted at two NHS trusts between 2021-2023. One hundred participants (163 eyes) with a diagnosis of chiasmal compression were recruited and three visual field programmes were completed on the same day. Results were independently graded for the presence/absence of a visual field defect plus type, location and severity of the defect.

Results:

Results of the three visual field programmes were classified using a binary response (yes/no) for the presence of visual field deficit/loss. Fifty-eight percent were classified as having a visual field defect on the SITA-fast 30-2 programme, 57% on the full-field 120 programme and 81% on the kinetic programme. The non-weighted kappa agreement coefficient for the presence of visual loss between the SITA-fast 30-2 static programme and the full field 120 was 0.42, (95% confidence interval [CI] 0.28, 0.56), between the SITA-fast 30-2 and the kinetic programme was 0.15 (CI 0.01, 0.29), and between the kinetic programme and the full field 120 was 0.26 (CI 0.12, 0.39). Using a visual field severity classification system (Wall and George, 1991) the weighted kappa coefficient for the severity of visual loss

between the SITA-fast 30-2 and the full field 120 was 0.61 (CI 0.52 0.70), between the SITA-fast 30-2 and the kinetic programme was 0.39 (CI 0.29, 0.49) and between the kinetic programme and the full field 120 was 0.41 (CI 0.316562 to 0.50676).

Discussion:

It is of clinical significance to test the periphery of the visual field in chiasmal compression. Kinetic perimetry was found to be most sensitive programme in detecting the presence of visual loss, with low agreement when compared to static programmes.

Pre-operative macular ganglion cell layer (GCL) and optic nerve retinal nerve fibre layer (RNFL) thicknesses as a prognostic marker for visual outcomes following pituitary tumour resection (PTR)

Ben Amram
University of Southampton

Introduction:

Pituitary adenomas often present with a reduction in visual acuity (VA) and visual fields (VF) due to mass effect on the optic chiasm. Treatment is normally surgical resection. After chiasmal decompression, recovery rates can vary. There is no established prognostic marker for visual outcomes following pituitary tumour resection (PTR). Optical coherence tomography (OCT) measuring optic nerve retinal nerve fibre layer (RNFL) and macular ganglion cell layer (mGCL) thicknesses have prognostic utility in other ophthalmic conditions. This study aimed to evaluate visual outcomes for patients who underwent PTR at University Hospital Southampton between 01/01/2018 and 31/12/2022 and to assess the use of pre-operative RNFL and mGCL thicknesses as prognostic markers for visual outcomes.

Methods:

A retrospective case review was performed. VA, VF mean deviation (MD) and pattern standard deviation (PSD) were recorded pre-operatively and at 0 - 6 months and 6 - 24 months post-operatively . Pre-operative optic nerve RNFL and mGCL thicknesses were recorded and correlated against post-operative visual outcomes.

Results:

At 0 - 6 months, VA (56.25%), VF MD (82.35%) and VF PSD (70.59%) had improved. At 6 - 24 months, VA (61.11%), VF MD (68.75%), VF PSD (75%) of all patients improved from their pre-operative measurement. Simple linear regression, adjusted for age and sex, was used to test if pre-operative OCT parameters were predictive of post-operative visual outcomes. The overall regression for nasal RNFL thickness in the right eyes (OD) was statistically significant ($R^2 = 0.836$, $F = 6.809$, $p = 0.047$). It was found that average (left eye [OS], $\beta = 0.392$, $p = 0.02$), (OD, $\beta = 0.481$, $p = 0.032$) and nasal (OS, $\beta = 0.217$, $p = 0.046$), (OD, $\beta = 0.537$, $p = 0.012$) RNFL thickness significantly predicted VF MD at 0 - 6 months. The overall

regressions for OS superonasal (R^2 = 0.911, F = 10.250, p = 0.044) and inferonasal (OS, R^2 = 0.913, F = 10.470, p = 0.043), (OD, R^2 = 0.908, F = 9.831, p = 0.046) mGCL thicknesses were statistically significant. Superonasal (OS, β = 0.335, p = 0.017) ,(OD, β = 0.921, p = 0.045) and inferonasal (OS, β = 0.844, p = 0.016), (OD, β = 0.0.992, p = 0.014) mGCL thicknesses significantly predicted VF MD recovery at 0-6 months. Nasal RNFL thickness predicted VA at 6 - 24 months. Superior and inferior RNFL thickness (0 - 6 months) and temporal and nasal RNFL thickness (6 - 24 months) were predictive of VF MD. Inferior RNFL thickness predicted VF PSD at 0 - 6 months. Superior, superonasal, inferonasal and superotemporal mGCL thicknesses predicted post-operative VA at 6 - 24 months. Superior and inferior (0 - 6 months) and superior, superonasal and inferonasal (6 - 24 months) predicted VF MD recovery.

Discussion:

Visual outcomes were in line with those in the literature. mGCL thickness has greater prognostic power than RNFL thickness for predicting visual outcomes following PTR.

References:

1) Muskens IS, Zamanipoor Najafabadi AH, Briceno V, Lamba N, Senders JT, van Furth WR, et al. Visual outcomes after endoscopic endonasal pituitary adenoma resection: A systematic review and meta-analysis. Pituitary. 2017;20(5):539-52.

2) Meyer J, Diouf I, King J, Drummond K, Stylli S, Kaye A, et al. A comparison of macular ganglion cell and retinal nerve fibre layer optical coherence tomographic parameters as predictors of visual outcomes of surgery for pituitary tumours. Pituitary. 2022;25(4):563-72.

3) Danesh-Meyer HV, Papchenko T, Savino PJ, Law A, Evans J, Gamble GD. In vivo retinal nerve fiber layer thickness measured by optical coherence tomography predicts visual recovery after surgery for parachiasmal tumors. Inv Ophthalmol Vis Sci. 2008;49(5):1879-85.

Poster Abstracts

1. Low risk disc clinic: A service evaluation of an Optometrist-led disc swelling clinic

Mousindha Arjunan, Anupa Patel, Oliver Backhouse
Leeds Teaching Hospital

Introduction:

An Optometrist-led low risk disc clinic was set up to address capacity issues for suspect papilloedema referrals. Bristol Eye Hospital reported a fivefold increase and rising trend in the number of disc swelling referrals from 2015 to 2020 (1). This could be due to widely available imaging modalities in the community, the Honey Rose case and overspecialisation of Ophthalmologists. This has impacted the burden on Neuro-ophthalmology clinics. As a result, Leeds Teaching Hospital designed a specialised Optometrist-led low risk disc clinic, set up in April 2022.

Methods:

A 6-month service evaluation was conducted. New suspect disc swelling referrals were triaged by a Neuro-ophthalmologist. Patients underwent a history, examination and disc imaging (optical coherence tomography retinal nerve fibre layer/disc volume/ganglion cell layer and Optos/autofluorescence). An alphabetic algorithm was created: A) definite disc swelling had same day Neurology referrals; B) presence of concerning symptoms but no definite diagnosis of disc swelling, patients would undergo an urgent outpatient magnetic resonance imaging (MRI); C) mild concerns but mostly normal discs, then patients would return to monitor for dynamic change; and finally D) patients without disc swelling or you had pseudopapilloedema were discharged.

Results:

Over 6 months a total of 49 patients were seen. The majority of referrals were from Optometrists (73%), Neurologists (11%), the emergency eye clinic (10%) and Ophthalmologists (6%). From those who attended clinic, 40% were reported as being symptomatic on their referral. According to our alphabetic grading system, out of the 49 patients, six patients met grade A and were referred to Neurology for further investigations (three

were diagnosed with idiopathic intracranial hypertension, two with dilated optic nerve sheaths and one with accelerated hypertension). Two patients met grade B criteria and were sent for outpatient MRIs which were unremarkable. Eight patients met grade C criteria and are being followed up to monitor for dynamic changes. 33 patients were discharged requiring no further investigation. However, what this service highlighted were three unusual cases. One patient was asymptomatic with significant disc swelling and the other two had a combination of dual optic disc pathology (tilted and crowded discs with swelling), which may have been missed without a face-to-face clinical examination.

Discussion:

This service was shown to reduce clinical burden, improve waiting times and cut down unnecessary referrals to Neurology. The value in having a face-to-face Optometrist-led clinic versus a virtual clinic is that atypical cases are less likely to be missed due to the subtleties involved in history taking and examination, therefore making an Optometrist-led clinic safer.

References:

1) McNicholl C, Gill A, Harrison R, Atan D. Impact of the COVID-19 pandemic and Honey Rose case on hospital attendances of patients suspected to have papilloedema. Eye (Lond). 2023 Jul;37(10):2157-2159. doi: 10.1038/s41433-022-02310-0. Epub 2022 Nov 28. PMID: 36443496; PMCID: PMC9702859.

2. Review of adult swollen optic disc pathway: A service evaluation and quality improvement study

Zhihang Cheng[1], Maryam Khan[2], Robert Cheeseman[1]
[1]St Paul's Eye Unit, Liverpool University Hospital
[2]University of Liverpool School of Medicine

Introduction:

Papilloedema, defined by optic disc swelling due to raised intracranial pressure, has seen a rising trend of referrals to hospital eye departments following a well-publicised legal case. This study aimed to evaluate the patient journey of adults presenting with suspected optic disc swelling and make recommendations for a more effective pathway for assessment.

Methods:
We performed a retrospective cohort study at a UK tertiary centre in patients aged 16 and above presenting with suspected optic disc swelling. The inclusion criteria were: patients presenting to the eye department with a clinical query of papilloedema or disc swelling suspicious of papilloedema. The exclusion criteria were patients with an established diagnosis of papilloedema or the referral stated an alternative cause for the disc appearances. The following clinical data were collected from the initial assessment: age; sex; symptoms of postural headache; blurred vision; diplopia; pulsatile tinnitus; and transient visual obscurations. The final diagnosis and aetiology of the optic disc swelling was collected at follow-up visits.

Results:
Over a 12-month period from March 2022 to March 2023, 125 referrals related to papilloedema or suspicious optic discs were received from optometrists. The average age was 32 (range 16 - 69 years). Seventy-six cases (61%) were referred to the ophthalmic emergency department directly, while 49 (39%) were referred by the GP e-Referral service as advised by optometrists, and triaged to our orthoptist-led suspicious disc clinic. Of these, 27 patients (22%) were further assessed for papilloedema by the medical team, with final diagnoses of idiopathic intracranial hypertension (22), venous sinus thrombosis (2), tumour (1), and normal opening pressure (2). Five cases exhibited optic disc swelling due to hypertensive retinopathy (2), optic neuritis (2), and giant cell arteritis (1). Thirty-nine patients (31%) were diagnosed with pseudopapilloedema, including peripapillary hyperreflective ovoid mass-like structures (20), disc drusen (10), tilted discs (6), and crowded discs (3). Notably, all cases requiring urgent intervention, such as those involving raised intracranial pressure, were directed to the ophthalmic emergency department. The study identified specific symptoms, including positional headache, pulsatile tinnitus, blurred vision, transient visual obscurations, and horizontal diplopia, with 92% specificity (confidence intervals 80-98%) for indicating raised intracranial pressure when two or more were present.

Discussion:

Our study showed that patients with suspected disc swelling, in our catchment area, are safely directed to the appropriate service by optometrists. There remains a significant false positive rate. The outcomes of our orthoptist-led suspicious disc clinic indicate its potential for managing low-risk patients through comprehensive assessments in a dedicated clinical setting.

References:

Mollan SP, Davies B, Silver NC, et al. Idiopathic intracranial hypertension: Consensus guidelines on management. J Neurol, Neurosurg Psychiatry 2018;89:1088-1100.

3. Evaluation of Optometrists' referrals to Ophthalmology for suspected optic disc swelling

Ramsewak VA[1], J M Jefferis JM[2], Hickman SJ[2], Pepper IM[2]
[1]Chesterfield Royal Hospital
[2]Sheffield Teaching Hospitals

Introduction/Purpose:

We see referrals for adults with suspected optic disc swelling from optometrists via written referrals in the out-patient clinic (OPC) and as emergencies in the emergency eyecare clinic (EEC). The OPC is run as a virtual clinic – a model we have shown as safe and effective (1). We compared patients referred by their optometrists to the OPC and EEC looking at the presence/absence of papilloedema in these patients.

Methods:

We retrospectively reviewed electronic notes and images of patients referred due to suspected optic disc swelling seen in either the OPC or EEC between 1st January and 31st December 2022. We excluded patients who were not referred via a community optometrist, who did not attend their appointment, who had optic disc swelling due to a cause other than raised intracranial pressure, and who had previously been seen in our department with optic nerve swelling or had a pre-existing diagnosis of idiopathic intracranial hypertension (IIH). All data were entered and analysed in Excel.

Results:
A total of 288 patient referrals were reviewed (128 OPC plus 160 EEC). Of the 128 OPC referrals, 90 (70.3%) met the inclusion criteria. The mean time between referral and review was 6.7 weeks, with 91.1% seen within 3 months. Following a single virtual review, 59 (65.6%) were found to have normal optic discs and 27 (30%) had pseudo-swollen optic discs. Another three (3.3%), after further review, were thought to have pseudo-swollen optic discs, leaving only one patient (1.1%) in the OPC group having true papilloedema. Of the 160 EEC referrals, 65 (40.6%) met the inclusion criteria. The mean time between referral and review was 1.1 days, with 48 (73.8%) seen on the same day and 60 (92.3%) within 5 working days. Following their EEC review, 16 (24.6%) were diagnosed as having normal optic discs, 24 (36.9%) had pseudo-swollen discs, and 25 patients (38.5%) were found to have papilloedema, of whom 21 had IIH. The likelihood of the patients having papilloedema was significantly higher when referred via the EEC pathway as compared to the OPC pathway ($p < 0.00001$).

Discussion:
According to our results, community optometrists were very successful in identifying patients with true papilloedema and referring them through the emergency pathway. There was a very low pick-up rate of papilloedema for patients seen via a written referral to the OPC. More work needs to be done to see if this latter group can be managed solely in the community.

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4. Group consultations in patients with idiopathic intracranial hypertension

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Background:
The increasing incidence of idiopathic intracranial hypertension (IIH) has led to increased service demands in Neuro-ophthalmology services throughout the country. Group consultations (GC) have been shown to be an effective way of delivering person-centred care, however this had not previously been investigated in IIH. To manage the service demand and improve quality of care, we set up group consultations (GC) at St Thomas' Hospital in 2019. The report from Phase 1 of our service delivery showed that patients preferred GC as opposed to the traditional face-to-face method of consultation, with median patient satisfaction scores of 9.5/10 and feeling listened to by clinician scores of 10/10. This, however coincided with the Covid-19 pandemic and we felt it was important to establish whether this was still patients preferred method of consultation.
Aim:
To re-audit the satisfaction rating of patients attending the IIH GC service from 2020-2023.
Methods:
Review of patient evaluation from 2020 to 2023. Patient evaluations were collected following each GC appointment.
Results:
We reviewed feedback for 241 patients. On re-evaluation, we found that 84% of patients found GC either extremely useful or very useful. Patients continue to feel listened to (median score 10/10) and felt they had enough opportunity to discuss their condition (median score 10/10). Only 2% preferred face to face consultations over GC.
Conclusion:
In summary, GC for patients with IIH are a success at St Thomas' Hospital. They continue to be a safe and effective way of reviewing patients with IIH and continue to be the preferred method of contact for our patient cohort. Our future aims include looking at remission rates between those seen in a 1:1 setting versus those seen in group consultation.

5. Timing matters: Nocturnal hypotension as a cause of transient vision loss

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Case report:

The patient, an 86-year-old female, with a significant cardiac history and polymyalgia rheumatica, sought evaluation for painless transient visual obscurations in both eyes exclusively at night for several months. The description of seeing 'a grey sheet across both eyes' prompted a thorough investigation. Her past ophthalmic history included bilateral cataract extraction. On examination, corrected visual acuity in the right eye was 6/12, and in the left eye was 6/9. Pupillary responses were brisk with no relative afferent pupillary defect. The anterior segment and posterior chamber examinations were stable. Fundal examination revealed healthy optic nerve appearances, and both maculae and retinae appeared unremarkable. Humphrey visual fields were grossly unreliable, while visual fields to confrontation were normal with no red desaturation. Ocular motility appeared full with no nystagmus. Optical coherence tomography results were within normal limits, and fundal auto-fluorescence was unremarkable. Additional investigations, including erythrocyte sedimentation rate, C-reactive protein, and ultrasound of the temporal arteries, did not support a diagnosis of giant cell arteritis. Despite an extensive ophthalmic evaluation, the initial diagnostic picture remained elusive. The patient's reported symptoms persisted even with a trial of increased oral steroids. Magnetic resonance imaging of the head and orbits with contrast revealed no intracranial or intra-orbital pathology, and electrodiagnostic testing demonstrated normal generalised retinal and optic nerve function. A fluorescein angiogram was not performed at this stage. On closer questioning, it was discovered that the patient had recently changed the timing of her antihypertensive medication to the evening. This crucial piece of information led to the diagnosis of nocturnal hypotension, causing inadequate optic nerve and retinal perfusion.

Notably, altering the timing of the antihypertensive treatment resulted in the resolution of the patient's visual symptoms.

Discussion:

This case underscores the importance of a meticulous patient history, including detailed diurnal timings of symptoms and treatments, in diagnosing conditions affecting optic nerve and retinal perfusion. The ultimate identification of nocturnal hypotension as the underlying cause highlights the need for a holistic and systematic approach in managing complex cases of transient vision loss.

6. Isolated third nerve palsy secondary to intra-cavernous internal carotid aneurysm: A report of two cases

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Case report:

We present two cases of transient, spontaneously-resolving, isolated, third nerve palsies resulting from internal carotid artery (ICA) aneurysms in the cavernous sinus.

Case 1:

A 55-year-old female presented with 3 days of diplopia and drooping of the left eyelid. There was no headache. On examination, her pupils were reactive but the left pupil was subtly larger. She had a left ptosis. There was an incomitant left hypertropia and exotropia, both worse on right gaze with limitation of the medial rectus. The rest of the examination was normal. A clinical diagnosis of left incomplete third nerve palsy was made. Computed tomography angiography (CTA) demonstrated a 5-6 mm aneurysm arising from the cavernous portion of the left ICA. On review the next day the motility findings had largely resolved. The neuro-surgical/neuroradiology multidisciplinary team decided to monitor closely without intervention.

Case 2:

A 62-year-old female presented with a 1 week history of right complete ptosis. The patient reported a similar episode of a ‘droopy eyelid’ a year

prior, which resolved itself. On examination a right-sided ptosis was observed. Orthoptic assessment showed a right exotropia and right hypertropia. A (-2 grade) limitation of the right eye in depression only was noted. All other findings were normal. A clinical diagnosis of incomplete right third nerve palsy was made. CTA revealed a right cavernous ICA aneurysm measuring 21 mm x 15 mm. The multi-disciplinary team decided that no further intervention was required currently. On review 15 weeks later the ptosis and ocular motility defects had completely resolved. Approximately a year later, she presented with recurrence of the right ptosis. CTA demonstrated that the aneurysm was unchanged but continued to exert substantial mass effect on the right cavernous sinus structures and temporal lobe. The interventional-radiology and neuro-surgical teams are now considering a flow diverting stent.

Discussion:

Ophthalmologists are well aware of the commonest aneurysmal cause of an acute third nerve palsy: a posterior communicating artery aneurysm. However, other aneurysmal sites can also affect the third nerve and can have more variable and subtle presentations. Cavernous sinus ICA aneurysms often involve multiple cranial nerves but, as in these cases, can present as an isolated intermittent third nerve palsy. Their management involves prompt neuro-imaging, multi-disciplinary team input and regular follow-up. Observation is often suitable for cavernous ICA aneurysms as their location means they do not tend to cause subarachnoid haemorrhage.

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7. Unmasking risks: A case study on subtle anterior ischaemic optic neuropathy from hyaluronic acid fillers

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Case report:

A 44-year-old woman presented to the eye casualty with painless visual loss superiorly in the left eye. She denied headache, double vision, weakness, recent trauma or pulsatile tinnitus. She is fit and well with no recent viral infections or recent vaccinations. There were no symptoms of giant cell arteritis. She did not take any regular medications. She did not smoke and drank around 10 units of alcohol per week. She worked as a school chef and was generally very active. The only recent hospital contact was with the maxillofacial team 8 months earlier when she was treated with ibuprofen, oral steroids, fexofenadine, aciclovir, and doxycycline for persistent upper lip swelling 7 months after having cosmetic lip fillers. Her visual acuity was 6/6 in each eye, but reliably performed Humphrey visual fields demonstrated left eye superior altitudinal visual field loss. Her intraocular pressures were normal. There was a left-sided relative afferent pupillary defect and her colour vision was reduced in the left eye. The anterior segments of both eyes were otherwise unremarkable. The right optic nerve looked crowded and had a ‘disc-at-risk’ configuration with a normal retina. The left optic nerve had 360 swelling with increased vessel tortuosity. The left retina was normal. Her blood pressure and random blood sugar were normal. Her weight was 54kg. A computed tomography (CT) sack of her head and CT venography were normal. Magnetic resonance imaging of her head and orbits was also normal. An extensive laboratory work-up was normal. Aquaporin 4 and myelin oligodendrocyte glycoprotein antibodies were negative. She was treated with 500mg of oral methylprednisolone to take daily for 5 days.

Discussion:
The lack of pain on eye movements, good visual acuity and the presence of optic nerve swelling did not fit a diagnosis of typical optic neuritis and to date her investigations into causes of atypical optic neuritis remain negative. We hypothesise that our patient may have received a smaller size of non-dissolvable hyaluronic acid particles during cosmetic lip filler injections, potentially leading to their persistence within the bloodstream and subsequent retrograde embolisation to the short posterior ciliary vessels. An acute association between hyaluronic acid fillers and central retinal artery occlusion has been reported, with a growing number of cases documenting various ophthalmic complications, including optic neuropathy, as the use of fillers becomes more prevalent. The practitioner's vigilance and awareness of the potentially devastating complications associated with intravascular injection during cosmetic procedures are paramount.

8. Visual electrophysiology: Clinical relevance in Neuro-ophthalmology
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Introduction/Overview:
Visual electrophysiology (VE) provides an objective measure of visual pathway function, including the retinal and post-retinal (e.g. retino-cortical) primary visual pathway. This presentation demonstrates the clinical relevance of VE techniques [1] in complementing clinical diagnosis or identifying the need for further investigation. VE uses a wide range of specialist tests including visual evoked potentials and electroretinography (VEP and ERG, respectively).
Four example clinical cases from neurology / ophthalmology origin are presented demonstrating how VE contributes to the diagnostic outcome. The examples we present include findings that were incidental or different from the initial clinical query. VE has also been useful in supporting clinical cases of possible functional neurological disorders and other congenital / retinal genetic disorders that may be previously undiagnosed.

Case 1: Neurological query of right optic neuropathy / quadrantanopia, however VE detected an abnormality of retinal origin (generalised and central retina) affecting both eyes.
Case 2: Query of left optic neuropathy for a patient with a history of mini stroke, VE detected left eye dysfunction of severe retinal origin with bipolar cell involvement; right eye function were normal.
Case 3: Ophthalmic query of acute zonal outer occult retinopathy with progressive visual field defect, VE did not detect evidence of retinal dysfunction however there was evidence supporting the likely presence of chiasmal lesion/compression, which were later confirmed on magnetic resonance imaging and visual fields.
Case 4: Query of right eye retinal or optic pathway dysfunction, VE showed normal retinal function but detected abnormal right eye optic nerve dysfunction with retinal ganglion cell involvement, multifocal-VEP test further showed the extent of field defect for the right eye.

Discussion/Conclusion:

The above example cases demonstrate the clinical relevance and importance of VE in supporting and complementing an accurate and more refined clinical diagnosis. In addition to these examples, VE also demonstrates its roles and relevance in detecting a wide ranging retinal / optic pathway dysfunction, including congenital / genetic disorders, that are not always apparent in eye clinic examinations alone.

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9. Sertraline induced raised intracranial pressure with bilateral optic nerve swelling

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Introduction:

Sertraline is commonly used for generalised anxiety and depressive disorders. It can cause ocular side effects such as optic neuropathy and maculopathy. We report an uncommon association of sertraline with bilateral disc oedema and features of raised intracranial pressure, which resolved when the medication was discontinued.

Case report:

A 32-year-old male referred to Neuro-ophthalmology with papilloedema by his optician. His only symptom was transient visual obscurations, especially on bending forward. There were no headaches, tinnitus or diplopia. There was no other medical history of note. He was on sertraline for anxiety, which had been started 2.5 years prior to this visit. On clinical examination there was no relative afferent pupillary defect and his extra-ocular movements were full. Both eyes had logMAR 0.0 vision. Fundus examination showed mild right optic disc oedema and lsevere left optic disc oedema. His blood pressure was normal. His visual field was normal in the right eye with a small inferonasal scotoma in the left eye with an associated enlarged blind spot. He underwent magnetic resonance imaging and venography which showed soft signs associated with raised intracranial pressure and transverse venous sinus stenosis. His lumbar puncture opening pressure was 30.4 cm of cerebrospinal fluid (CSF) with normal CSF constituents. He was started on oral acetazolamide and bendroflumethiazide. Despite increasing doses of acetazolamide over the course of a few months, there was no improvement in the optic disc swelling. The patient stopped his sertraline as he felt this was the cause of his problems after a discussion with his general practitioner, switching to citalopram. Within weeks of this, his optic disc swelling significantly reduced with resolution of the visual field defect. His acetazolamide and bendroflumethiazide were discontinued and at 3 years follow up, his optic disc swelling had not returned.

Discussion:

Sertraline is not a medication associated with raised intracranial pressure. It has been once reported in association with bilateral optic disc swelling, which resolved on discontinuation. In our knowledge our case is the first that links sertraline with raised intracranial pressure features. There is one

case report mentioning the association of sertraline with optic disc oedema (1). We are not sure of the mechanism related to raised intracranial pressure. This needs further study for similar cases in future. In this case report we want to highlight consideration of systemic medications in a case of optic disc oedema which are not commonly associated with it.

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10. Unilateral complete ophthalmoplegia and ptosis in an elderly female patient

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Case report:

A 79-year-old woman presented with a 1-week history of left ptosis and diplopia, with no associated cranial or systemic symptoms. Examination revealed unilateral complete ophthalmoplegia, ptosis and a minimally reactive pupil. The right eye was unaffected and visual acuity was normal bilaterally. Raised inflammatory markers and positive ultrasound Doppler of a temporal artery confirmed the diagnosis of giant cell arteritis (GCA). The patient responded well to oral corticosteroid therapy, showing near resolution of symptoms on 3-week follow-up. Due to the atypical presentation and significant electrolyte derangement, the patient was

further investigated for occult malignancy during her admission. Computed tomography scanning of her pelvis revealed bilateral multiloculated adnexal lesions of undetermined significance. The ovarian cancer-related tumour marker CA 125 was raised at 58 kU/L. Further magnetic resonance imaging (MRI) suggested these lesions may be benign, however, malignancy could not be definitively excluded. As such, she was referred for Gynaecology multi-disciplinary team (MDT) discussion.

Discussion:

GCA is a systemic autoimmune vasculitis which poses the risk of ophthalmic complications, of which sight-threatening presentations, such as ischaemic optic neuropathy is the most common and feared. Ophthalmoplegia secondary to cranial nerve (CN) palsies is much rarer, seen in only 2-15% of GCA patients. Our patient, who presented with complete ophthalmoplegia and ptosis secondary to multiple CN palsies with sparing of vision, is an especially rare presentation as per the literature. In most reported cases of CN palsies in GCA there is often some degree of accompanying visual involvement and rarely are multiple CNs affected. Such a presentation holds a better prognosis as visual loss in this setting is often permanent, whereas ocular CN palsies respond well to corticosteroid therapy. Ophthalmoplegia in GCA occurs secondary to microvascular ischaemia of the vasa nervorum of ocular CNs or extraocular muscles. The latter, however, would likely present with pain and visual loss as extraocular muscle ischaemia often forms part of orbital infarction syndrome in which all contents of the eye would be affected. Thus, this makes concomitant third, fourth and sixth nerve palsies the most likely aetiology in this case. The case gained complexity due to the atypical presentation, prompting exploration for underlying malignancy. The connection between GCA and malignancy is unclear; literature reports hint at a potential paraneoplastic link, more prevalent in haematological malignancies than solid tumours. While the MRI suggested likely benign leiomyomas, diagnostic uncertainty persists, thus our patient awaits further discussion in the Gynaecology MDT to assess lesion significance.

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11. A curious case of downbeat nystagmus: A diagnostic dilemma

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Case report:

A 59-year-old man presented with an 18 month history of blurred vision and dizziness. He complained of unsteadiness, using a broad gait and difficulty running. He had a history of chronic sinus disease and cough for over 20 years and had undergone sinus surgery. He was treated for hypertension and hypercholesterolaemia. He had previously consumed alcohol to excess, but had reduced consumption 2 years ago, and was emphatic that he was not drinking to excess. Vestibular function tests failed to identify a vestibular cause for his dizziness. He complained of blur, which changed with head posture. His visual acuities were 6/24 right and left. He favoured chin elevation and using this position had binocular acuity of 6/9. He had downbeat nystagmus, more marked on lateral gaze and elevation. Neurological examination demonstrated intention tremor and difficulty with heel-to-toe gait and a cerebellar aetiology of his nystagmus was suspected. Magnetic resonance imaging (MRI) of his brain was reported as normal. A neurological opinion was sought. He was noted to have reduced proprioception, loss of vibration sense in his legs, an unsteady tandem walk but no limb ataxia. Nerve conduction studies were consistent with a sensory neuronopathy or dorsal root ganglionopathy. Haematological investigations, including autoimmune screen, were

unremarkable, except for a low serum folate level, which was corrected without any change in his symptomatology. He tried clonazepam and gabapentin in an effort to reduce his nystagmus, which was felt to be responsible for his blur although he had no oscillopsia. He declined superior rectus recession. Repeat MRI performed a year after the first demonstrated disproportionate atrophy of the cerebellum and mamillary bodies, which was thought could be consistent with alcohol excess, although the patient denied this. Further investigations were undertaken to exclude paraneoplastic causes. A diagnosis of CANVAS ('cerebellar ataxia, neuropathy, vestibular areflexia') syndrome was then suspected due to the coexistent vestibular findings and neuropathy and the presence of chronic cough. This was confirmed with genetic testing.

Discussion:

CANVAS syndrome is a rare neurological autosomal recessive hereditary condition, which typically presents in middle age and is associated with progressive imbalance, sensory disturbance, dry cough and oscillopsia (1). Biallellic AAGGG expansions in the RFC1 gene are the genetic cause (2). Whilst rare, CANVAS syndrome should be suspected and genetic testing sought in patients presenting with ataxia and nystagmus of undetermined aetiology. Although there is no treatment which halts disease progression, vestibular rehabilitation may alleviate patient symptoms.

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12. Neuro-ophthalmology and immune checkpoint inhibitors - a diagnostic dilemma

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Case report:

A 56-year-old female patient presented to the Eye Emergency Department

on the advice of her oncologist. She reported a 6 day history of variable but worsening “blurred vision” which improved on covering either eye. She could observe the left eye drifting out of alignment, and reported impaired balance when walking. She had not noticed any diurnal variation or ptosis, and did not report any pain on eye movement. There was no significant ocular history. This patient had a diagnosis of transitional cell bladder cancer, which had been treated 9 months prior with surgical excision, and 4 cycles of neoadjuvant chemotherapy, followed by maintenance immunotherapy with Avelumab, a PD-L1 inhibitor. Ocular examination demonstrated bilateral limitation of adduction with bilateral abducting nystagmus and mild limitation of upgaze on the right. Her visual acuity was 6/6 bilaterally, colour vision and confrontation visual fields were full in both eyes, and ocular examination, including dilated fundoscopy, was otherwise unremarkable. Magnetic resonance imaging of her head with contrast was reported as normal, and anti-acetylcholine receptor and anti-neuronal antibody testing was negative. The patient was commenced on intravenous methylprednisolone under the care of Neurology and Oncology, with rapid improvement in symptoms. This was converted to oral therapy, and tapered over a short timeframe. Avelumab therapy was not continued by the oncology team. Her extraocular movements had returned to normal at orthoptic review 4 weeks following the initial presentation.

Happily, the patient remains disease-free without adjuvant therapy.

Discussion:

Immune checkpoint inhibitor (ICI) therapies are a key cancer treatment, and their use has increased exponentially over the past decade (1). The systemic and anterior segment side effects are more common (2), and Oncologists and Ophthalmologists may be more familiar with these. Neurological and neuro-ophthalmological side effects are rare, but include demyelination and myasthenia gravis, which are potentially fatal if missed (3). Additionally, this patient presented with a pseudo-internuclear ophthalmoplegia, in which the gaze paretic nystagmoid movements posed a diagnostic dilemma. The auto-inflammatory conditions precipitated by

ICI therapies are an additional factor to consider when assessing patients with a cancer diagnosis.

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13. A case report of bilateral optic neuropathy secondary to glial fibrillary acidic protein (GFAP) antibody-associated astrocytopathy

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Case report:

A 21-year-old male was referred by the Neurology team due to bilateral loss of vision whilst an inpatient. He had no past ocular or medical history of note prior to presentation. He initially presented to Accident & Emergency (A&E) 1 month prior to admission with headaches following a minor head injury, and had a computed tomography scan of his head, which was reported as normal. He re-presented to A&E with ongoing headaches and abdominal pain and was found to be in urinary retention. A urinary catheter was placed and he was discharged, but presented again the following day with headaches and fevers and was started on emipirical treatment for meningitis. Within 24 hours of admission he developed confusion and seizures, and was intubated for further management and investigation. Raised cerebrospinal fluid (CSF) lymphocytes and protein resulted in treatment for presumed central nervous system tuberculosis with adjunctive dexamethasone. Magnetic resonance imaging (MRI) of his

head showed punctate foci of diffusion restriction and perivascular enhancement, with extensive central cord involvement and enhancement of both optic nerves, indicative of glial fibrillary acidic protein (GFAP) antibody-associated astrocytopathy, which was later confirmed on CSF antibody testing. After the MRI, TB treatment was stopped and he received intravenous methylprednisolone, plasma exchange and intravenous immunoglobulins. Three weeks into his admission, he reported new bilateral loss of vision. Initial bedside ophthalmology examination on intensive care showed bilateral no perception of light vision, with minimal pupillary light responses, bilateral ptosis and ophthalmoplegia. There was no fundal abnormality seen on indirect ophthalmoscopy. Interval MRI brain, orbits and spinal cord showed extensive signal change throughout the midbrain and both optic nerves. Four weeks after initiation of treatment, repeat MRI brain and orbits showed reduction in the degree of optic nerve and midbrain enhancement. Six weeks after initiation of treatment, his visual acuity had improved to 6/6 unaided in both eyes, with full colour vision, normal pupillary light responses and resolution of the bilateral ptosis and ophthalmoplegia. Optical coherence tomography at that time showed bilateral optic atrophy, with moderate thinning of the retinal nerve fibre layer and macular ganglion cell layer volumes in both eyes.

Discussion:

GFAP astrocytopathy is a rare autoimmune inflammatory CNS disorder, with associated malignancy in 10 – 30% of cases (1). An estimated 25% of patients have visual involvement (2). We present a case of bilateral optic neuropathy due to GFAP antibody-associated astrocytopathy and documentation of OCT changes, which have not been previously reported (2,3).

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systematic literature review. Neurol Neuroimmunol Neuroinflamm. Sep 2023;10(5):e200146.
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14. A double hit on the optic nerve

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Case report:

A 59-year-old South Asian male was referred by his optician with right-sided optic disc swelling and was subsequently diagnosed with an optic nerve sheath meningioma, which was treated with fractionated CyberKnife radiosurgery. His optic disc swelling resolved, his visual function improved from 0.1 to 0.0 logMAR with normal colour vision and full 24/2 visual fields. He remained under routine follow up and underwent annual magnetic resonance imaging (MRI). Four years after the initial presentation he re-presented as an emergency with worsening vision, a constricted visual field and recurrence of the right eye optic disc swelling. The left eye remained normal. Initial thoughts were that the optic nerve sheath meningioma had reoccurred, however MRI showed that this was stable in size. Over the next few weeks the visual loss progressed to perception of light. The working hypothesis was optic neuropathy secondary to his prior radiation therapy, however the oncology team were surprised as treatment had been well within the usual treatment thresholds. At the same time he also presented to the emergency department with a cough and feeling unwell. Routine blood tests revealed the patient had advanced HIV infection with a CD4 count of 130. He was started on anti-retroviral treatment and after 4 weeks of treatment he noticed an improvement in vision in the right eye. The right optic disc appeared atrophic, however, his visual acuity improved to 4/60 in the right eye and he also noted a gradual improvement in his visual field over several months which markedly improved his quality of life, as he was now able to avoid bumping into things on his right-side.

Discussion:
In this case report we present a patient with unilateral progressive visual loss, initially thought to be secondary to prior radiotherapy treatment for an optic nerve sheath meningioma, who then had a moderate improvement in their vision once commenced on treatment for advanced HIV. We therefore hypothesise that this patient's right optic nerve, having already sustained a vulnerability due to the previous radiotherapy, was susceptible to further damage related to the immune changes resultant from advanced HIV. There are only a few case reports of HIV related optic neuropathy with improvement in visual function following treatment in the literature, and no cases where there is a history of prior radiotherapy (1,2). HIV infection should be considered in patients with worsening optic disc swelling, even when they already have another diagnosis.

References:
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15. Where does the fluid come from, where does it go?
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Introduction:
The case of a 37-year-old pregnant woman underscores the complexities in diagnosing and managing ocular complications amidst idiopathic intracranial hypertension (IIH).

Case report:
Initially presenting with bilateral blurred vision and escalating headaches over 2 weeks, her medical history unveiled a decade-long management of IIH through a lumboperitoneal shunt. Past episodes of papilloedema had resulted in bilateral partial optic atrophy, setting the stage for intricate diagnostic challenges. During her initial ophthalmology evaluation, the absence of clinical papilloedema led to a referral back to neurology with a

provisional diagnosis of migraine headache. However, the patient's symptoms continued to intensify, prompting her self-referral to eye casualty. On examination, her Snellen visual acuity was 6/24 in the right eye and 6/12 in the left eye. Diminished colour vision in the right eye (7/17 Ishihara plates) with normal colour vision in the left eye caught attention. Her ocular motility was full, and her pupils were equal and reactive to light with no relative afferent pupillary defect. Her anterior segments were normal in both eyes, however dilated fundus examination revealed bilateral pale discs with significant macular oedema. A review of serial optic coherence tomography (OCT) imaging unveiled subtle yet progressive increases in retinal nerve fibre layer (RNFL) thickness bilaterally, alongside the emergence of small peripapillary intraretinal cysts in the right eye during the initial ophthalmology visit. Subsequent imaging portrayed a marked escalation in RNFL thickness and the presence of intraretinal peripapillary cysts in both eyes. More strikingly, the right eye showed subretinal fluid tracking from the disc to the macula, while the left eye exhibited serous macular detachment and cystoid macular oedema. Despite prompt referral to neurology, diagnostic uncertainty persisted - a differential diagnosis of central serous maculopathy was raised, resulting in a referral to the medical retina team. The patient underwent spontaneous vaginal delivery 4 days later. Post-delivery, a lumbar puncture revealed elevated cerebrospinal fluid (CSF) pressure (35 cmCSF), coupled with redundant shunt tube findings on a computed tomography scan of her abdomen. Subsequently, the insertion of a ventriculoperitoneal shunt ensued, marking a turning point in the patient's management. Four months post-ventriculoperitoneal shunt insertion, there was a reduction in disc oedema and marked improvement in maculopathy bilaterally.

Discussion:

This case serves as a clarion call to the intricate nature of optic disc swelling, especially in patients with partial optic atrophy. Macular changes are under-recognised complications of papilloedema, with the potential to cause permanent central vision loss (2). Serial OCT has emerged as a critical tool in detecting subtle changes, cautioning against the

misinterpretation of macular complications in such cases as isolated issues.

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16. Advancing practice in Neuro-ophthalmology

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Advanced Clinical Practice is a rapidly expanding area across nursing and allied health professions and has continued to evolve in recent years supported by national frameworks and policy (1,2). The role of non medical professionals in Neuro-ophthalmology has also evolved in recent years in response to NHS policy and the drive for patient centred care, in addition to the workforce shortages identified in ophthalmology (3). This paper aims to to examine the range of different roles and models of care currently employed in clinical practice across the UK as well as the benefits in terms of cost, efficiency, patient safety, patient satisfaction and career motivation.

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2) NHS Longterm plan 2023. https://www.england.nhs.uk/wp-content/uploads/2023/06/nhs-long-term-workforce-plan-v1.2.pdf
3) RCOph Census 2022 https://www.rcophth.ac.uk/news-views/rcophth-census-2022-report/

17. Highs and lows: Optic nerve sheath fenestration in a refractory syndromic paediatric intracranial hypertension

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Case report:

A 3.5-year-old male presented with headaches and excessive blinking. Initial examination revealed hypermetropia, binocular vision of 0.275, equal and reactive pupils, normal colour vision, and swollen optic discs. The patient had a history of iron deficiency anaemia, asthma, pulmonary artery branch stenosis, left radio-ulnar fusion, high arched palate, testicular non-descent, wide-spaced nipples, and autism. Further investigations including ultrasonography of the optic discs, magnetic resonance (MR) imaging brain, MR venograpy, and a lumbar puncture, showed no drusen, normal neuro-imaging, and an opening pressure of 28 cm cerebrospinal fluid (CSF) with normal CSF constituents. A cardiology consultation and repeat iron studies were normal. Initial management with acetazolamide, furosemide, and topiramate failed. Ventriculoperitoneal shunt attempts were unsuccessful, prompting subsequent optic nerve sheath fenestration (ONSF). Post-procedure stabilisation and improvement in vision and optic disc swelling were noted. Five months post-ONSF, the disc swelling recurred, possibly in response to oral antibiotics for an asthma exacerbation. This responded to oral steroids, however weaning caused a recurrence of headache symptoms. Following interdisciplinary discussion a lumboperitoneal shunt was attempted, which failed and was removed due to infection. His symptoms and disc swelling responded to increased topiramate and prednisolone, however, Cushingoid side effects and increased right optic nerve swelling prompted repeat right ONSF with subsequent improvement in papilloedema and stabilised vision. Oral prednisolone was discontinued and he remained on a maintenance dose of topiramate. Further investigation confirmed normal visual evoked potentials. Genetic testing identified a de novo mutation in JAG1 associated with Alagille's syndrome (ALGS). The patient, presently symptom-free, maintains good visual function with mild-moderate optic disc swelling on topiramate. Further steroid treatment is undesirable due to a lumbar spine stress fracture, although DEXA scanning was satisfactory.

Discussion:

Papilloedema can be the first presentation of ALGS. When performed in a timely manner, ONSF can save vision and lead to improvement in visual function in patients with optic nerve swelling due to ALGS that is refractory to medical treatment. Ophthalmological abnormalities in ALGS patients include posterior embryotoxon (78-95%), optic disc abnormalities (21%), peripheral hypopigmented retinopathy (10%), lens opacity, and iris hypopigmentation. Embryotoxon is the most common, occurring in 78-95% of ALGS patients. Management necessitates a multidisciplinary team involving Paediatric Neurologists, Neurosurgeons, Ophthalmologists, Cardiologists, Respiratory Physicians, and Psychiatrists. This case underscores the challenges in managing a complex clinical presentation and the importance of a collaborative approach to diagnose and treat rare conditions like ALGS. The successful outcome following ONSF demonstrates the significance of tailored interventions in optimising patient outcomes in such intricate cases.

References:

1) Rock NM, Demaret T, Stéphenne X, et al. Intracranial hypertension and papilledema in a large cohort of pediatric patients with Alagille Syndrome. J Pediatr Gastroenterol Nutr. 2020;71:655-662.
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18. Another case of optic nerve swelling!

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Case report:

A 72-year-old limousine driver presented with a 1-week history of decreased vision in his left eye. He denied any eye pain, diplopia or photophobia. He had no headaches or systemic symptoms. His past ocular history included bilateral cataract surgeries. His past medical history was relevant for hypertension and hyperlipidaemia. His visual acuity was right 6/9 (pinhole no improvement) and left 6/18 (pinhole no improvement).

His intraocular pressures were right and left 9 mmHg. There was a grade 3 left relative afferent pupillary defect (RAPD). His extraocular motility showed mild abduction deficit bilaterally. Both eyes were proptotic with notable upper lid swelling. The left conjunctiva was mildly injected. The remainder of the anterior segment was unremarkable. Posterior eye examination showed bilateral swollen optic nerves. The left was worse than the right with circumferential disc hyperaemia. The remainder of the fundus examination was unremarkable. A computed tomography (CT) scan of the brain with CT venography revealed a symmetrical extracranial soft tissue process with thickened tissue proliferation into both orbit extraconal spaces and bilateral inferotemporal fossae. There was no sinus thrombosis. A vasculitis and infectious screen returned negative results for tuberculosis, sarcoidosis, syphilis, HIV, Hepatitis B and Hepatitis C. A blood smear and lymphocyte were negative for lymphoma. IgG subclass was added and showed markedly elevated IgG4 levels. The patient underwent an orbital biopsy of the left lacrimal gland, which confirmed IgG4 related disease. A positron emission tomography scan confirmed no other organ involvement. The patient was managed on systemic prednisolone with resolution of his orbital inflammation by 6 months. His visual acuity returned to 6/6 in both right and left eyes with resolution of the left RAPD. The patient developed steroid-induced hyperglycaemia and was subsequently commenced on rituximab as a steroid sparing agent.

Discussion:

IgG4-associated disease is characterised by sclerosing inflammation with infiltration of IgG4-positive plasma cells (1,2). It is associated with elevated serum IgG4 concentrations (2). Orbital presentations commonly involve the orbit and lacrimal gland resulting in lid swelling and proptosis (1-3). Conjunctival, sclera and extraocular muscle involvement has been reported (2). This case highlights a rare presentation of orbital IgG4-related orbital disease with bilateral optic nerve head swelling and an afferent pupillary defect with vision loss. The differential diagnosis includes idiopathic orbital myositis, marginal zone B-cell lymphoma, ANCA-mediated vasculitis and reactive lymphoid hyperplasia (2,3). Treatment for predominantly orbital IgG4-related disease includes systemic steroids,

radiotherapy or rituximab (1-3). Often orbital IgG4-related disease resolves after systemic steroid therapy, but relapse is often observed following therapy discontinuation (1-3).

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19. Not another RAPD!

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[2]Metro North, Queensland Health, Australia

Case report:

A 19-year-old male presented with a 2-day history of blurry vision in the right eye and a 1-week of right retroorbital pain. He denied any headaches or diplopia and had no past ocular history. His past medical history included previously treated pulmonary tuberculosis and a biopsied left frontal brain abscess, which showed fibro-inflammatory changes and was managed with systemic antibiotics. On presentation his visual acuity was right 6/24 (pinhole 6/9) and left 6/6. There was a grade 2 right relative afferent pupillary defect (RAPD). Subjective and objective optic nerve testing was reduced in the right eye. Formal perimetry revealed a monocular right inferior quadrantanopia whilst the left field was unremarkable. His extraocular motility was intact in all directions. The anterior and posterior eye examination was unremarkable bilaterally. Urgent magnetic resonance imaging (MRI) of his brain and orbits revealed a subdural empyema extending along the right anterior cranial fossa with thickening and enhancement involving the right orbital apex, right optic

canal and right canalicular segment of the right optic nerve. Atypical serologies, vasculitis screen, optic neuropathy including atypical antibodies, nutritional, autoimmune screens including IgG4 and infectious screen including syphilis, sarcoidosis, and mycobacterium culture all returned negative results. A lumbar puncture was negative for cultures and flow cytometry. After consulting with the Infectious Diseases team the patient was commenced on isoniazid, rifampicin, and pyroxidine with oral prednisolone for presumed cerebral tuberculosis. His right visual acuity deteriorated to 6/96. An urgent right pterional craniotomy and dural biopsy was performed. No compressive pathology was identified. Intra-operatively the dura was thickened and scarred with optic nerve swelling under the falciform ligament. The dural biopsy returned negative cultures and was re-evaluated and deemed not to show IgG4-related disease. The specimen however had crush artefact. His visual acuity returned to right 6/15 (pinhole 6/6) and his RAPD resolved on prednisolone. He was continued on empirical anti-tuberculosis medication throughout this time. He developed a right pupil-sparing third nerve palsy and his prednisolone was subsequently increased. Sequential MRI of his brain showed resolving dural thickening with the diagnosis of idiopathic hypertrophic cranial pachymeningitis (IHCP) most likely. The patient was weaned off steroids, commenced on rituximab and remains symptom free.

Discussion:

Hypertrophic pachymeningitis is a rare diffuse inflammatory process that causes thickening of the dura mater. It can be secondary from malignancy, infection and autoimmune processes (1-3). When evaluation fails to reveal a cause, it is termed IHCP (1-3). There are limited reports on the ocular manifestation of IHCP and its treatment. Clinical presentations include cranial nerve palsies, orbital pseudotumour, uveitis and cavernous sinus thrombosis (1-3) Diagnosis is based on a combination of radiological and histopathology findings. First line of treatment usually includes immunosuppressants such as corticosteroids (1-3). Azathioprine, cyclophosphamide, rituximab and methotrexate are other options (1-3). This case highlights IHCP manifesting as an orbital apex and a diagnostic challenge requiring multiple subspecialty input.

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20. When the ophthalmoscope is mightier than the stethoscope in chest disease

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Case report:

A 66-year-old woman was referred by the Eye Emergency Department for neurological investigation of suspected papilloedema. She complained of a 1-week history of persistent blurred vision affecting her left eye. This was associated with mild retro-orbital discomfort, which was independent of eye movement. There was no headache, nausea, vomiting, diplopia or transient visual obscurations. She was an active smoker with a history of well-controlled hypertension and dyslipidaemia. Her best-corrected visual acuities were 6/6 OD and 6/12 OS. There was a subtle left relative afferent pupillary defect. Her visual fields were normal on confrontation testing. Fundoscopy revealed optic disc oedema (Frisén grade - OD 2, OS 3). Recent routine fundus photography from her optician were reviewed and showed no evidence of optic disc oedema. Her ocular motility was normal. The remainder of her neurological examination was unremarkable. Magnetic resonance (MR) imaging of her brain and MR venography with contrast was unremarkable with no evidence of intracranial mass lesions, cerebral venous sinus thrombosis or leptomeningeal disease. A lumbar

puncture showed a normal opening pressure of 21 cm cerebrospinal fluid (CSF). However, CSF analysis unexpectedly showed lymphocytosis and a mildly raised protein level, raising the suspicion of atypical bilateral optic neuritis. Anti-myelin oligodendrocyte glycoprotein MOG and anti-aquaporin 4 antibodies were sent. MR imaging of her orbits showed left optic nerve hyperintensity with peri-neural enhancement. She was treated with intravenous steroids with subjective improvement of vision on telephone follow-up. Outpatient MR imaging of her spine was scheduled to assess sub-clinical cord involvement. While no cord lesions were demonstrated, a mass was identified in the upper lobe of the right lung. A serum anti-neuronal antibody panel was requested given the recognised association of anti-CV2 antibody-associated optic neuropathy and lung cancer. A positron emission tomography - computed tomography scan was suspicious for extensive stage small-cell lung cancer, which was confirmed on histopathology. The anti-CV2 antibody (CRMP-5) assay was strongly positive.

Discussion:

Bilateral optic disc oedema in an older adult smoker, as in this case is suggestive of raised intracranial pressure which in turn is commonly due to intracranial malignancy, often metastatic It was an unfortunate irony that although extensive investigation of optic disc oedema excluded intracranial or orbital neoplastic disease, the aetiology was nevertheless due to malignancy- small cell lung cancer with CV-2 antibody associated paraneoplastic optic neuropathy.

21. Is ocular giant cell arteritis one other thing to be worried about Covid 19 infection?

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Case report:

A 82 year old woman with a past history of multiple sclerosis and chronic kidney disease presented to the emergency department in August 2023 with sudden onset complete painless vision loss noted in the left eye for 1 day with associated blurriness of vision in the right eye. There were no

classical clinical symptoms of giant cell arteritis (GCA). She also had a history of breathlessness and feeling sick for 5 days preceding the eye symptoms. Examination showed no perception of light in the left eye with an associated relative afferent pupillary defect. Fundus examination showed chalky white disc oedema with features of central retinal artery occlusion. Although her right eye clinically looked normal, her visual acuity in that eye was reduced to 1/60. Blood tests revealed raised inflammatory markers. Based on the clinical features, a diagnosis of left-sided arteritic anterior ischaemic optic neuropathy (AAION) with associated central retinal arterial occlusion due to possible ocular GCA was made. She was admitted and started on intravenous methylprednisolone and referred for urgent rheumatology opinion. A temporal artery ultrasound showed the halo sign in the superficial temporal arteries and in a frontal branch on both sides, which were features consistent with clinical GCA. A COVID swab was sent due to her presenting symptoms, which turned out to be positive. She was treated with tocilizumab along with systemic steroids. The patient passed away 10 days after the onset of her ocular symptoms, unfortunately.

Discussion:

GCA is commonly seen in elderly females. Increased incidence of GCA following SARS-CoV-2 infection has been reported in literature. Various case reports have reported non-arteritic anterior ischaemic optic neuropathy associated with this infection. However, cases with AAION with GCA with concomitant COVID-19 infection has been sparsely reported. Our patient presented with classical clinical signs of ocular GCA in the left eye. Although her right eye was clinically normal looking, the vision in that eye was reduced and temporal artery ultrasonography showed changes on both sides signifying bilateral involvement. Even though it can be an accidental coincidence of AAION with the COVID-19 infection, the purpose of this case report is to argue that COVID-19 infection can be a possible risk factor for the onset of GCA and also to highlight potential ophthalmological complications of COVID-19. Further studies are required to determine the relationship between COVID-19 and GCA and whether this infection predisposes to early bilateral involvement.

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22. Bilateral optic atrophy: A presenting sign of classic WFS1 spectrum disorder (Wolfram syndrome type 1)

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Introduction:
Classic WFS1-spectrum disorder (Wolfram syndrome type 1) is a progressive neurodegenerative disorder, characterised by the onset of diabetes mellitus and optic atrophy before the age of 16 years. It is also known as DIDMOAD (diabetes insipidus, diabetes mellitus, optic atrophy, & deafness) due to the possible WFS1-related complications.

Case report:
We describe an 8-year-old boy who presented with early-onset exotropia aged 1 year, who subsequently developed reduced visual acuity and bilateral optic atrophy at the age of 5 years. Optical coherence tomography showed thinning of the retinal nerve fibre layer in both eyes. Magnetic resonance imaging of his brain showed thinning of the optic nerves and chiasm. Blood investigations, including blood glucose, were initially normal. Genetic testing (R41 Optic neuropathy gene panel) confirmed a likely genetic diagnosis of WFS1-related disease. Repeat blood testing showed raised blood glucose and HbA1c, confirming diabetes mellitus, and he was commenced on Insulin.

Dicsussion:

Optic atrophy in a child may be the presenting sign of WFS1 (Wolfram syndrome). It is important to investigate for associated childhood-onset diabetes mellitus. Biallelic pathogenic (or likely pathogenic) variants in WFS1 can be identified by molecular genetic testing.

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23. A typically atypical case

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Case report:

A 62-year-old heterosexual female, incidentally diagnosed with syphilis (initial rapid plasma reagin [RPR] [1:16]) and subsequently treated with procaine penicillin, presented 12 months later with sequential right, followed by left, optic neuropathy. At the time of assessment, the Snellen visual acuity (VA) was recorded to 6/9 in the right eye and 6/60 in the left eye. Bilateral disc swelling and hard exudates were observed. Serological examination revealed positive syphilis ELISA IgG/IgM and positive RPR (1:4). Notably, there was an absence of regional lymphadenopathy, and neither the patient nor her partner reported a history of injecting drug misuse or previous sexually transmitted infections. The patient was confirmed to be HIV-negative and remained so throughout the follow-up period. All other blood parameters were found to be within normal limits. T2 FLAIR magnetic resonance imaging displayed a swollen intra-orbital segment of the right optic nerve with high T2 signal, without evidence of intracranial involvement. Lumbar puncture results showed no significant abnormality. Four weeks into the re-treatment, the visual acuity in the left eye improved to 6/36, accompanied by reduction in the optic nerve head swelling.

Discussion:

Our presented case highlights a notable lack of response to the standard initial course of penicillin, necessitating a subsequent course of treatment with this antibiotic. Despite penicillin's longstanding recommendation as the primary therapeutic intervention for syphilis spanning over seven decades, treatment failure is documented in 10 - 20% of patients experiencing early syphilis. Recent investigations have identified diverse single-nucleotide polymorphisms (SNPs) in Treponema pallidum associated with penicillin resistance. In instances where patients manifest neurological symptoms or exhibit neuropsychiatric deterioration, coupled with a history of either treated or untreated neurosyphilis, a comprehensive evaluation is imperative. This evaluation should encompass brain imaging, cerebrospinal fluid (CSF) analysis, CSF RPR, and Venereal Disease Research Laboratory (VDRL) titres. Elevated suspicion of neurosyphilis relapse or refractory flares despite conventional penicillin therapy mandates a reassessment of treatment duration, appropriate antibiotic selection, and dosage adjustments. For cases of refractory syphilis, a judicious approach involves a reduced threshold for CSF examination to identify potential flares or resistance. Additionally, a 6-month clinical follow-up regimen with regular RPR and VDRL titres is recommended. In the event of a confirmed recurrence, alternative antibiotics such as doxycycline should be offered to the patient. These nuanced approaches aim to enhance the management of syphilis cases exhibiting resistance or recurrent neurological symptoms, thereby ensuring more effective and tailored therapeutic strategies.

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24. Botulinum toxin as a treatment and a diagnostic tool in adult strabismus

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Objectives:

Strabismus, or misalignment of the eyes, can arise from various aetiologies, including neurological conditions. Sixth nerve palsies and other neurological causes pose distinct challenges in strabismus management. This paper examines the efficacy of botulinum toxin as a therapeutic and diagnostic intervention in such cases. Beaumont hospital is unique, from an ophthalmic perspective in Ireland, given the fact that it is the leading centre for neurosurgery in Ireland. There are rare and diverse presentations of neurological and neurosurgical conditions that present with varying ocular pathology. Patients are affected by rare, and often complex, strabismic conditions affecting their quality of life. Setting up a new service for botulinum toxin injections for adult strabismus can be a valuable addition to Neuro-ophthalmological practice.

Methods:

All patients who had received botulinum toxin to an ocular muscle over the past 3 years were included. Data collection was via retrospective collection of data from healthcare records after identifying suitable patients from the theatre registry. No patients had previous strabismus surgery. The study was registered with the clinical audit committee in Beaumont hospital.

Results:

Thirty-two injections of botulinum toxin have been given to 18 patients since the introduction of the service in 2021 (average of 1.8 injections per patient). The average age of the patient cohort was 45.4 years. The

average angle of initial deviation was recorded (average 34 prism dioptre [PD)] range 14 – 50 PD) and then further measurements were taken at 4 weeks and 3 months for all patients (average angle at 3 months post treatment 17.9 PD; range 0 - 40 PD). The dosage of botulinum toxin given to each muscle was recorded and ranged from 2.5iU - 7.5iU. The presence of fusion pre – and post – treatment was recorded. Sixth nerve palsies accounted for 50% of the indications for treatment, none of which were attributed to a microvascular cause. Intracranial pathologies causing sixth nerve palsies included: pontine/basilar artery thrombosis; cavernoma; brainstem astrocytoma; acinic parotid cell tumour with cavernous sinus spread; carotid cavernous fistula; cerebellopontine angle meningioma; basilar artery rupture; and head trauma. A further subgroup analysis on outcomes was performed on the sixth nerve palsy patients. Other causes were further sub-classified into neurological or strabismic based on their aetiologies. The treatment had no effect in three patients overall, and strabismic conditions fully resolved in two patients post treatment. The average duration of effect in the remaining patients was 3.7 months. Five of these patients went on to have strabismus surgery with beneficial outcomes. The only complication recorded was transient ptosis, occurring seven times out of the 32 treatments (22%), the maximal duration of which was recorded as lasting 6 weeks.

Discussion:

Establishing a botulinum toxin injection service for adult strabismus is timely and aligns with modern Ophthalmological practice trends. Overall, we have shown that botulinum toxin for managing strabismus resulting from sixth nerve palsies and other neurological conditions has proven to be a promising and minimally invasive option with the correct patient selection. Early intervention with botulinum toxin can mitigate adaptations resulting from chronic strabismus such as contractures and head postures. Botulinum toxin not only serves as a treatment option but also as a diagnostic tool to evaluate the potential for fusion or binocular single vision in adults with strabismus. The presence or absence of fusion can influence the patient's decision and the ophthalmologist's approach to surgical intervention. As well as assisting in assessment of fusional

potential and trial alignment, the treatment can also serve to give a surgical response prediction and help provide clinicians with an insight into the risk of post-operative diplopia. Patients demonstrating binocular vision or diplopia after botulinum toxin treatment are more likely to sustain and benefit from binocular vision post-surgical correction. Their sensory system's response to temporary alignment acts as a surrogate marker for post-operative sensory adaptability.

25. A review of patients referred to Neuro-ophthalmology from ENT with tinnitus and radiological signs of intracranial hypertension: What proportion have papilloedema?

Jayne Tan, Jonathan Virgo
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Objective:

Neuro-ophthalmology is a small sub-specialty and in most centres demand for appointments is in excess of what is available. Urgent referrals from opticians for people with suspected papilloedema have steadily increased in recent years (PMID: 36443496). We have also observed a similar increase in referrals from hospital specialists for patients with radiological signs of possible intracranial hypertension. A large proportion of these referrals come from ENT, when brain imaging has been done to investigate tinnitus. In a study (PMID: 33871552) of 296 consecutive patients undergoing brain magnetic resonance imaging (MRI) for any indication, 49% of patients had at least one MRI finding suggestive of intracranial hypertension. However, the prevalence of papilloedema, detected by ocular fundus photography performed at the time of MRI, was only 2%. In this study we aimed to determine how common papilloedema is in people referred with tinnitus and radiological signs of possible intracranial hypertension. The findings will be of interest to Neuro- ophthalmologists who triage similar referrals.

Methods:

The electronic patient records were checked for all patients attending Neuro-ophthalmology clinic appointments between January 2022 and January 2023. Those referred by ENT for tinnitus and radiological signs of

possible intracranial hypertension were included in this study. Information was recorded for each patient including demographics, type of tinnitus (pulsatile vs non-pulsatile), type and number of MRI features suggestive of intracranial hypertension, body mass index, blood pressure, visual acuity, optical coherence tomography findings, presence/absence of headache and headache diagnosis, Neuro-ophthalmological diagnosis and management, including further investigations and treatment. Statistical analysis was carried out using Fisher's exact test or a T test.

Results:

As of 17/11/23 we have reviewed patients seen between Jan and April 2023. Of the 892 patients seen in Neuro-ophthalmology clinics, 33 patients (3.7%) had been referred from ENT due to tinnitus and radiological signs of possible intracranial hypertension. Five (15.2%) of the 33 had IIH with papilloedema, but in all cases the optic disc swelling was mild and vision was good. A further four patients (12.1%) were diagnosed with IIH without papilloedema. Statistical analysis will be carried out once data collection is complete.

26. What we know about the ocular manifestations of multiple sclerosis: An overview

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Imperial College Healthcare Trust, London

Introduction:

Multiple sclerosis (MS) is an autoimmune demyelinating disease of the central nervous system (CNS), in which there is dissemination of lesions in time (two or more clinical events) and space (multiple lesions seen on brain and spinal imaging). The pathophysiology of this disease is complex and involves a combination of genetic, environmental and immune factors. MS affects women to men in a 2:1 ratio, and it shows a geographical predilection for Europe and the Americas (1). The disease produces significant sensory, motor and autonomic effects, among which ocular symptoms are common. Indeed, in a large proportion of patients, the initial presenting symptom may be ocular in nature (2). This review aims to provide an overview of what is known so far about the ocular

manifestations of this demyelinating disease, and seeks to highlight those presentations that the ophthalmologist should be alert for when assessing patients with as yet undiagnosed MS.

Methods:

This review was conducted via a search of the existing literature in August 2022 in Medline via Ovid. A configurative approach to assimilation of existing research was taken, to construct a narrative review on what is known on the subject.

Results:

The review maps what is known or suspected about ophthalmological manifestations of MS into two broad, anatomically distinct categories: the afferent and the efferent pathway manifestations. Afferent manifestations discussed include optic neuritis, uveitis, and field defects resulting from afferent pathway lesions elsewhere. Discussion of the efferent pathway comprises internuclear ophthalmoplegia, nuclear and nerve palsies, skew deviation, nystagmus, saccadic intrusions and dysmetria.

Discussion:

MS can affect every portion of the visual system and thus produces a wide and varied range of clinical syndromes. It is important for the clinician to be aware of the hallmark features seen on clinical examination as, if detected promptly, early referral and initiation of treatment leads to improved disease outcomes (3).

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27. Peripapillary retinal nerve fibre layer thinning, perfusion changes and optic neuropathy in carriers of Leber's hereditary optic neuropathy-associated mitochondrial variants

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Introduction:

We investigated Leber's hereditary optic neuropathy (LHON) families for variation in peripapillary retinal nerve fibre layer thickness and perfusion, and associated optic nerve dysfunction.

Methods:

A group of LHON-affected patients (n = 12) and their asymptomatic maternal relatives (n = 16) underwent examination including visual acuity, visual-evoked-potentials, and optic nerve imaging including optical coherence tomography (OCT) and OCT angiography (OCTA) of the peripapillary retinal nerve fibre layer (RNFL). A control sample was examined also (n = 10). The software imageJ was used to measure perfusion by assessing vessel density (VD), and statistical software 'R' was used to analyse the data.

Results:

The LHON-affected group had significantly reduced peripapillary vessel density (VD) (median 7.9%, p = 0.046). Overall, the LHON asymptomatic relatives had no significant change in peripapillary VD (p = 0.166), though three eyes had VDs which fell below the derived normal range at 6% each, with variable visual acuity from normal to blindness; logMAR median 0, range 0 to 2.4. In contrast, RNFL thickness was significantly reduced in the LHON-affected group (median 51 µm, p = 0.003), and in asymptomatic relatives (median 90 µm, p = 0.01), compared with controls (median 101 µm). RNFL thinning had greater specificity compared with reduced perfusion for optic nerve dysfunction in asymptomatic carriers (92% versus 66%).

Discussion:

Overall, reduced peripapillary retinal nerve fibre layer perfusion was observed in those affected by LHON, but was not reduced in their asymptomatic relatives, unlike RNFL thinning, which was significantly

reduced in both groups versus controls. The presence of RNFL changes were associated with signs of optic neuropathy in asymptomatic relatives.

28. Optic Atrophy due to moyamoya in trisomy 13

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Introduction:

Moyamoya Disease (MMD) is a chronic, non-inflammatory, progressive, occlusive cerebrovascular disease characterised by stenosis at the terminal portion of the internal carotid artery and extensive collateralisation. Although the precise pathophysiology of MMD remains ill defined, a genetic association has been described (e.g., RNF213, BRCC3/MTCP1 and GUCY1A3).

Case report:

We describe a 38-year-old woman with trisomy 21 who presented with unilateral optic nerve atrophy due to MMD. She had many features of trisomy 21, including a short neck, protruding tongue, narrow palate, flat nasal bridge, upslanting palpebral fissures, epicanthal folds, and small ears. She had moderate intellectual and developmental delay and limited verbal communication. Her past ocular history however was normal and she had a previously documented 20/20 visual acuity in both eyes. The patient's mother noted a change in visual behaviour and an outside ophthalmologist noted optic atrophy in the right eye (OD).

On neuro-ophthalmological examination, the visual acuity was 20/80 OD and 20/25 in the left eye (OS). A relative afferent pupillary defect OD was present. The patient could not perform automated perimetry. Slit lamp biomicroscopy showed Brushfield spots on the iris in both eyes. Ophthalmoscopy showed diffuse optic atrophy OD and a normal optic disc OS. The remainder of the ocular exam was normal.

Investigations:

Laboratory testing for optic atrophy including serum vitamin B_{12} and B_9 levels, syphilis serology, complete blood count, tuberculosis interferon gamma release assay, and chest radiography were negative. Magnetic resonance imaging of the brain and orbit with contrast showed chronic lacunar infarcts in the periventricular white matter. There was absence of the left internal carotid artery (ICA) flow void, likely due to occlusion. The right optic nerve was atrophic with mildly increased T2 signal within the optic nerve. Computed tomography angiography of the head and neck with contrast showed chronic occlusion of the left ICA, right posterior cerebral artery, right middle cerebral artery and left anterior cerebral artery A2 segment with extensive collateralisation (moyamoya). Severe narrowing of the proximal left ICA beginning approximately 5 mm beyond the bifurcation was noted. A standard catheter cerebral angiogram confirmed the diagnosis of MMD. Vascular neurological and neurosurgical consultations were obtained, and aspirin 81 mg daily was started.

Discussion:

Moyamoya angiopathy is a rare condition, optic atrophy is an unconventional presentation, and they can be associated with trisomy 21. More studies are warranted to understand this disease to design randomised clinical trials assessing the best treatment modalities.

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The date and venue are to be announced