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Annual Meeting  
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ISH Venues  
Great Portland Street  
**LONDON, UK**



Anatomical Antagonists

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## Meeting Schedule

08.00-09.15	<b>Neuro-ophthalmology Allied Professionals Breakfast Meeting</b>	
09.30-09.40	<b>Welcome and Chair: Ailbhe Burke</b>	
09.40-10.10	<b>The optic nerves and oculomotor cranial nerves traverse the orbits.</b>	<a href="#">Sachin Salvi</a> Consultant Ophthalmologist, Royal Hallamshire Hospital, Sheffield
10.10-10.40	<b>The skull encases the brain and cranial nerves</b>	<a href="#">Richard Bowman</a> Consultant Ophthalmologist, Great Ormond Street Hospital, London
10.40-11.20	<b>The oculomotor cranial nerves have to negotiate the skull base and cavernous sinuses before reaching the orbit.</b>	<a href="#">Eleni Maratos</a> Consultant Neurosurgeon, King's College Hospital, London
11.20-11.50	<b>Coffee</b>	
	<b>Chair: Eoin O'Sullivan</b>	
11.50-12.30	<b>The optic chiasm is surrounded by potential enemies</b>	<a href="#">Hani Marcus</a> Consultant Neurosurgeon, The National Hospital for Neurology and Neurosurgery, London

12.30-1.00	<b>The sinuses lurk between the optic nerves</b>	<a href="#">Claire Hopkins</a> Consultant Rhinologist, St Thomas' Hospital, London
1.00-2.00	<b>Lunch</b>	
	<b>Chair: Margaret Dayan</b>	
2.00-2.30	<b>Service update: What do the stroke national guidelines mean for eye care services?</b>	<a href="#">Fiona Rowe</a> Professor in Orthoptics, University of Liverpool
	<b>Chair: Michael Gilhooley</b>	
	<b>Platform presentations from submitted abstracts</b>	
2.30-2.45	<b><a href="#">Optical coherence tomography findings following optic nerve sheath fenestrations in people with idiopathic intracranial hypertension</a></b>	Miss Joanna Jefferis
2.45-3.00	<b><a href="#">Supine Positioning in ICU: A Potential Model for Space-Associated Neuro-Ocular Syndrome (SANS)</a></b>	Dr Gagana Mallawaarachchi

3.15-3.30	<p><a href="#"><u>Can we trial interventions for homonymous hemianopia in children and young people? A pilot randomised control trial of peripheral prism glasses</u></a></p>	Ms Sian Handley
3.30-3.45	<p><a href="#"><u>Ocular Signs in Ocular Myasthenia Gravis</u></a></p>	Miss Abbie Ewart
3.45-4.15	<p><b>Poster viewing and tea</b> <b>Chair: Simon Hickman</b></p>	
4.15-5.15	<p><b>UKNOS Annual Lecture: Neuro-Ophthalmology – Quo tendimus?</b></p>	<p><a href="#"><u>Susan Mollan</u></a> Consultant Ophthalmologist and Honorary Professor, University Hospitals Birmingham</p>
5.15-5.30	<p><b>Prize giving</b></p>	
5.30	<p><b>Close of meeting</b></p>	

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## Speaker Biographies

### **Mr Sachin Salvi**

Mr Sachin Salvi is the clinical lead for the Sheffield Orbital & Oculoplastic Services as well as the Sheffield Ocular Oncology Service, one of three designated ocular oncology centres in England. He runs a regional orbital service managing patients with thyroid eye disease, orbital trauma, IIH requiring optic nerve sheath fenestration and other orbital complex problems such as orbital tumours and inflammation. He runs joint clinics with endocrinology and maxilla-facial surgery and is a member of the regional skull base MDT.

With his dual training and experience in ocular oncology and adnexal surgery (which includes oculoplastics, lacrimal and orbital surgery); he manages the full spectrum of eye cancers including eyelid, conjunctival, intraocular and orbital tumours.

He has trained extensively in India, UK and USA with Fellowship training at Royal Hallamshire Hospital, Sheffield; Moorfields Eye Hospital, London; and Cole Eye Institute, Cleveland Clinic, USA.

He co-authored the NICE approved UK National Uveal Melanoma Guidelines and has over 100 publications and book chapters to his credit. He has multiple awards and keynote lectures to his credit. He is chair of the international medical graduate sub-committee of the Royal College of Ophthalmologists, BRAAC committee member of British Oculoplastic Surgery Society, member of the Clinical advisory panel of Ocumel UK and advisory editorial board of Indian Journal of Ophthalmology.

He is the training director of the sought-after fellowship programme in ocular oncology and adnexal surgery at Sheffield. He is a Visiting Professor in Ophthalmology to Trinidad Eye Hospital and Honorary Senior Clinical Lecturer to the University of Sheffield.

He is regularly involved in voluntary ophthalmic work in Trinidad and Tobago as well as in India. His sporting passion is cricket and is the founding member of Sheffield Medics Cricket Club, currently positioned in the first division of the Yorkshire and Derbyshire league.

### **Mr Richard Bowman**

Mr Richard Bowman has been a consultant for over 20 years. He is a Consultant Paediatric Ophthalmologist at Great Ormond Street Hospital for Children in

London. He holds an honorary associate professorship at both Institute of Child Health and the London School of Hygiene and Tropical Medicine (LSHTM). He trained as a doctor at Cambridge University and Guy's Hospital medical school. His ophthalmology training was at Cambridge, the Tennent Institute of Ophthalmology, Glasgow, Moorfields Eye Hospital and Great Ormond Street. He is fellowship director at Great Ormond Street Hospital

Richard has published around 150 peer reviewed papers, several book chapters and has co-written a book for parents and carers of children with eye problems. His doctoral research won the annual award for the best medical doctorate at Cambridge University and the Moorfield Research Medal. He is academically active and has won around £1million in grant funding as principle investigator and has supervised/is supervising about 8 PhD students. He regularly lectures nationally and internationally.

He worked for several years in Tanzania performing high volume surgery and establishing a training programme in paediatric ophthalmology which served the whole of Africa and beyond.

He has a longstanding clinical and research interest in the diagnosis and management of childhood neuro-ophthalmology problems. This includes strabismus, nystagmus, cerebral visual impairment, optic nerve disease, craniofacial conditions, and CNS tumours. He is the lead for all these services at Great Ormond Street Hospital.

He has lectured nationally on nystagmus surgery and has recently been invited to write a book chapter on this subject. He has organized national paediatric ophthalmology teaching courses and lectures regularly at Moorfields, Great Ormond Street and LSHTM. He has been a member of various international examination boards for ophthalmology, including the International Council for Ophthalmology and, currently, the College of East and Southern Africa.

He is a member of 2 expert national and international subcommittees developing evidence based guidelines for assessing and managing children with cerebral visual impairment. He has previously sat on the paediatric sub-committee of the Royal College of Ophthalmologists. He has given 2 named lectures.

### **Miss Eleni Maratos**

Ms Eleni Maratos PhD FRCS(SN) is a consultant neurosurgeon who specialises in skull base pathology, pituitary disorders and degenerative spine disease. She is developing the orbital service at Kings College Hospital London in collaboration

with the ophthalmology, oculoplastic and maxillo-facial teams using both open and endoscopic techniques to tackle pathology of the orbit and skull base. She has a strong scientific background with a PhD in neuropharmacology and continues to publish and present her work at national and international academic meetings. She has a particular interest in collecting patient reported outcome measures and contributes to the national vestibular schwannoma audit as well as shaping the data set for the anterior cranial fossa audit.

Her enthusiasm for teaching has led to a collaboration with industry where she teaches on Skull Base dissection courses and presentation skills coaching. She runs “Hit the Ground Running on your Skull Base fellowship “, a post CCT course which prepares Skull Base Fellows from multiple disciplines for their year ahead.

### **Mr Hani Marcus**

Mr Hani Marcus is an experienced consultant neurosurgeon at the National Hospital for Neurology and Neurosurgery and Honorary Associate Professor at the UCL Queen Square Institute of Neurology. He specialises in treating patients with pituitary adenoma and related skull base tumours. His expertise lies in the use of “keyhole” endoscopic approaches, completing fellowships in Calgary, Zurich, and London before his appointment. In 2020, he co-founded the British Neuroendoscopy Society to drive the advancement of endoscopic techniques and led the largest prospective study of patients undergoing pituitary surgery worldwide, including 866 patients across 30 centres in the United Kingdom and Ireland. He has also authored international guidelines on pituitary surgery through the Pituitary Society, where he has served on several committees.

### **Professor Claire Hopkins**

Professor Claire Hopkins undertook her medical training at Oxford University and was awarded with a First Class Degree in Physiology and Distinctions in Medicine in 1993. She undertook specialist ENT training in London. She completed her Fellowship in Advanced Rhinology at Guy’s and St Thomas’, where she was appointed as an NHS Consultant. She is a Consultant ENT Surgeon and Professor of Rhinology at King’s College, London.

Her research focuses on improving outcomes from both medical and surgical treatments for nasal conditions. She has published over 400 peer-reviewed papers, 30 book chapters and is invited to lecture all around the world. She has written international guidelines on the management of sinusitis and nasal polyps,

and has been involved in the development and introduction of several advances in this field, including the use of biological therapies.

She is the Past-President of the British Rhinological Society, the Secretary General of the European Rhinologic Society, and the Rhinology Section of the Royal Society of Medicine. She is the Academic Chair of ENTUK's biannual conference and has represented ENT-UK on a patient safety panel at the Royal College of Surgeons and chaired the UK Commissioning Guidance Group for Sinusitis.

### **Professor Fiona Rowe**

Professor Fiona Rowe is Professor of Orthoptics at the University of Liverpool and Director of the VISION research group. She is an editorial board member for the British and Irish Orthoptic Journal, and lead orthoptic editor for the Cochrane Library Eyes and Vision group. She is the Past President of the Society for Research in Rehabilitation and a mentor for the National Institute of Health Research integrated clinical academic training pathway. She is the orthoptic representative on the Council for the International Strabismological Association and a member of the European Neuro-Ophthalmology Society Low Vision group. Her particular research interests include visual impairment due to acquired brain injury (leading a number of national and international stroke projects), visual field evaluation and control of ocular alignment. Professor Rowe is the author of two textbooks: 'Clinical Orthoptics' and 'Visual fields via the visual pathway', author for seven book chapters, and has presented and published her research extensively.

### **Professor Susan Mollan**

Professor Susan Mollan is a Consultant Neuro-Ophthalmologist at University Hospitals Birmingham and an Honorary Professor at the University of Birmingham. She is the first international member on the board at the North American Neuro-Ophthalmology Society (NANOS) and a research theme chair for the European Neuro-Ophthalmology Society. She has been instrumental to the stable running of the British Isles Neuro-Ophthalmology Club and is a past secretary of the UK Neuro-Ophthalmology Society. She actively works to provide pragmatic guidance for clinicians through research and collaboration. Her publications include the Idiopathic Intracranial Hypertension (IIH) consensus guidelines (2018); European League Against Rheumatism Giant Cell Arteritis (GCA) guidelines (2019); British Society of Rheumatology GCA guidelines (2020); the European Headache Federation GCA guidelines (2020); and the International Headache Society guidelines for randomised controlled trials in IIH. She and her colleagues in Birmingham reported the first randomised control trial into weight loss methods



in IIH (2021). She is an International Senior Examiner for the Fellowship of the Royal College of Ophthalmologists.



## Platform presentations

### Optical Coherence Tomography Findings Following Optic Nerve Sheath Fenestrations in People with Idiopathic Intracranial Hypertension

JM Jefferis, SL Hill, IM Pepper, SJ Hickman, SM Salvi  
Sheffield Teaching Hospitals NHS Foundation Trust

**Introduction:** Options for surgical intervention in vision-threatening idiopathic intracranial hypertension (IIH) include shunting, stenting and optic nerve sheath fenestration (ONSF). There is currently no evidence for the most effective intervention and choice is influenced by local expertise. In Sheffield, UK, we have experience in ONSF and have demonstrated its effectiveness at improving visual function in IIH. Optical coherence tomography (OCT) has become an essential tool in monitoring IIH. It can quantify and track optic nerve swelling whilst ensuring preservation of the macular ganglion cell layer (mGCL). Furthermore, OCT measures overcome some of the difficulties encountered measuring visual function in persons with severe headache. Changes in OCT parameters following CSF diversion surgery have been previously described. This study aims to describe changes following ONSF.

**Methods:** A retrospective review of OCT imaging from patients who underwent ONSF for sight threatening IIH between January 2020 and August 2023 in Sheffield. OCT imaging was performed with Spectralis OCT (Heidelberg Engineering). Parameters included: retinal nerve fibre layer thickness (RNFLT,  $\mu\text{m}$ ); total retinal thickness (TRT,  $\mu\text{m}$ ); and mGCL volume ( $\text{mm}^3$ ). Visual acuity and kinetic perimetry (Octopus) were also reviewed.

**Results:** Eleven patients who had bilateral ONSF were included. Mean (standard deviation) baseline RNFL in the worse eye (defined by a higher RNFLT at baseline) was  $410.2\mu\text{m}$  (113.5); whilst at 2 weeks post-operation it had dropped to  $193.9\mu\text{m}$  (54.7); and at nadir  $117.6\mu\text{m}$  (67.6); representing a relative reduction of 53% and 71%, respectively (one way repeated measures ANOVA  $p < 0.001$ ). RNFLT reduction was maintained at 12 months. Similar results were seen for TRT with relative reductions of 36% at 2 weeks and 52% at nadir ( $p < 0.001$ ) and maintenance of this at 12 months. mGCL volume, whilst trending slightly downwards from  $1.1\text{mm}^3$  (0.1) at baseline to  $0.9\text{mm}^3$  (0.11) at 2 weeks post-operation and  $0.93\text{mm}^3$  (0.16) at nadir was not statistically significantly reduced ( $p = 0.08$ ). Mean baseline logMAR visual acuity was 0.3 (0.34) and mean baseline Octopus I4e area was  $5729\text{deg}^2$  (4555); these improved to 0.26 (0.43) and  $6700\text{deg}^2$  (4686), respectively at latest review.

**Conclusions:** These results show rapid decline in optic disc swelling in the 2 weeks following surgery, which is maintained at 12 months. This is seen with maintenance of the mGCL volume, suggesting minimal/limited optic nerve damage at 12 months post-operation. The 12-month parameters are very similar to those described in shunting, but the changes may occur more rapidly after ONSF when compared to shunting.

## Supine Positioning in ICU: A Potential Model for Space-Associated Neuro-Ocular Syndrome (SANS)

Gagana Mallawaarachchi<sup>1,2</sup>, Ella Courtie<sup>1,3,4</sup>, Richard Blanch<sup>1,3,4,5</sup>

1. Department of Ophthalmology, Queen Elizabeth Hospital Birmingham, UK
2. Heartlands Hospital, University Hospitals Birmingham NHS Foundation Trust, UK
3. Institute of Inflammation and Ageing, University of Birmingham, UK
4. Surgical Reconstruction and Microbiology Research Centre, University Hospitals Birmingham NHS Foundation Trust, UK
5. Academic Department of Military Surgery and Trauma, Royal Centre for Defence Medicine

**Introduction** Space-associated neuro-ocular syndrome (SANS) is a significant health concern affecting approximately 15% of astronauts during extended space missions. It is characterised by structural ocular changes, including optic disc oedema, retinal nerve fibre layer (RNFL) thickening, and choroidal flattening. Supine or head-down tilt (HDT) protocols are widely used to simulate microgravity-associated ocular changes in terrestrial settings. Intensive care unit (ICU) patients are frequently maintained in prolonged supine positioning, providing a unique opportunity to evaluate whether this positioning could serve as a model for studying SANS-related retinal changes. We evaluated whether supine positioning during ICU admission affects retinal structure in general surgical patients and examined the utility of this protocol as a model for studying SANS.

**Methods:** 58 patients with planned post-operative ICU care after major upper gastrointestinal surgery were assessed prospectively. Optical coherence tomography was performed pre-operatively, during admission, and 3-6 months after hospital discharge to evaluate long-term effects.

**Results:** A total of 218 data points were included. Peripapillary global RNFL (RNFL G) thickness increased from (mean  $\pm$  SD,  $\mu\text{m}$ )  $97.76 \pm 12.97$  at baseline to  $100.4 \pm 12.38$  24-48 hours post-operatively in ICU, and returned to  $97.92 \pm 13.17$  at final follow-up. Similar trends were observed for all individual RNFL sectors. Linear modelling revealed that the increase in RNFL G from time point 1 to 2 was statistically significantly ( $2.01 \pm 0.496$  increase,  $p < 0.001$ ). Moreover, changes in RNFL thickness were independent of peripapillary total retinal thickness in all sectors, except I2, where significance was reached.

**Conclusion:** Supine positioning during ICU admission caused transient RNFL G thickening, similar to SANS, but which occurred within 24-48 hours of supine positioning. This may recapitulate some features of microgravity.

## Can we Trial Interventions for Homonymous Hemianopia in Children and Young People? A Pilot Randomised Control Trial of Peripheral Prism Glasses.

Sian E Handley<sup>1,2</sup>, Michael D Crossland<sup>3,4,5</sup>, Dorothy A Thompson<sup>1,2</sup>, Jessica Gowing<sup>1</sup>, Rosemary Wilson<sup>1</sup>, Vasiliki Panteli<sup>1</sup>, Mario Cortina-Borja<sup>2</sup>, Alki Liasis<sup>2,7</sup>, Richard Bowman<sup>1,2</sup>, Jugnoo S Rahj<sup>1,2,4,5,6</sup>.

1. Great Ormond Street Hospital for Children, London
2. UCL GOS Institute of Child Health, London
3. Moorfields Eye Hospital, London
4. UCL Institute of Ophthalmology, London
5. NIHR Biomedical Research Centre, Moorfields Eye Hospital, London
6. Ulverscroft Vision Research Group, UCL GOS Institute of Child Health
7. University of Nicosia Medical Centre, Nicosia, Cyprus

**Introduction:** Most intervention studies for homonymous hemianopia (HH) have been undertaken in adults with largely later onset field loss. Spectacle-mounted peripheral prisms are one option that can be used to increase awareness of objects in the non-seeing visual field of people with HH. Randomised controlled trial (RCT) evidence has shown their effectiveness in adults, but there is little evidence about their value in children and young people (CYP). We investigated whether it is possible to undertake trials of peripheral prisms in CYP with HH, comparable to those undertaken in adults which have demonstrated benefits.

**Methods:** We undertook a pilot of a double-masked crossover RCT of peripheral prisms. Participants wore both 40PD peripheral prisms and 4PD 'sham' prisms for approximately 4 weeks each. Minimisation allocated prism order balancing for side of HH. The primary outcome measure was parental and CYP intention (Yes/No) to continue wear. Secondary outcome measures were Functional Vision Questionnaire for CYP (FVQ\_CYP) and open-ended feedback.

**Results:** Twenty-two (47%) of eligible subjects participated: aged 5-17 years (mean 12) with 8 females. Ten had right HH. All completed the trial. One was excluded from analysis when new pathology was found. All CYP were able to complete the primary outcome i.e. indicating interest or not in continuing wear with parental support. A significantly higher proportion of CYP said "yes" to real (66.7%) compared with sham prisms (14.3%) (McNemar's  $p=0.016$ ). A similar proportion was reported by parents with 71.4% "yes" to real prisms compared to 19% to sham (McNemar's  $p=0.002$ ). The secondary outcome FVQ\_CYP was completed by 77% CYP at baseline and 50% at all visits. At trial end 15 (71.4%) chose to continue peripheral prism use and long-term follow-up is ongoing.

**Discussion:** Our findings show that interventional trials for HH are feasible in selected CYP, and families are willing to participate. Adaptions to HH trials designed for adults can be made to translate them to CYP with a range of neurodisability and so these factors should not be a barrier to participation. Both CYP and their parents showed a significant preference for peripheral prisms over sham. These findings will inform design of future studies including potential full-scale RCTs.

## Ocular Signs in Ocular Myasthenia Gravis

Abbie Ewart<sup>1</sup>, Sui H Wong<sup>2</sup>

1. Dept of Orthoptics, Moorfields Eye Hospital, London

2. Dept of Neuro-Ophthalmology, Moorfields Eye Hospital, London

**Introduction:** The pattern of ptosis and extraocular muscle (EOM) weakness in ocular myasthenia gravis (OMG), according to their serological subtypes, has not been well phenotyped. Understanding this is valuable in elucidating the pathophysiology of OMG. This study aims to define the pattern of ptosis and EOM weakness at the initial symptomatic episode in patients with OMG of different serological subtypes.

**Methods:** A prospective observational study of patients with OMG from 2013-2023. Data recorded included ptosis at presentation and following provocation, the presence of manifest deviations, diplopia in the primary position, the frequency of EOM involvement and EOM weakness rated according to the orthoptic convention on a 0-4 scale, where 0=normal and 4=maximum weakness.

**Results:** 248 subjects (148 [60%] males) with a median age of 61 years (range 25 to 93) were recruited in the prospective observational study. Data for 130 patients (86 [66%] men) were reviewed at the time of abstract submission. 7 were excluded as the diagnosis had not been confirmed by serology or neurophysiology (n=5), notes unavailable (n=1) or a subsequent alternative diagnosis was made on follow-up (n=1). 60/123 (48.8%) patients were seropositive with anti-acetylcholine receptor (AChR+), 5 (4.1%) with anti-low density lipoprotein receptor-related protein 4 (LRP4+), and 9 (7.3%) with anti-muscle specific (MuSK+) antibodies; 49 (39.8%) were seronegative (SN). Ptosis was present initially in 68/123 (55%) OMG patients: 40/60 (67%) AChR+, 3/5 (60%) LRP4+, 2/9 (22%) MuSK+, 23/49 (47%) SN. 16/123 (13%) had the presence of ptosis only on provocation; 6/16 (37.5%) AChR+, 2/16 (12.5%) MuSK+ and 8/16 (50%) SN. Manifest deviation was present in primary position in 71/123 (57.7%), of whom 34/60 (56.6%) were AChR+, 3/5 (60%) were LRP4+, 3/9 (33.3%) were MuSK+ and 31/49 (63.3%) were SN. Diplopia in the primary position was present in 66/130 (50.7%). The frequency of EOM involvement for the 246 eyes (1476 EOM) of 123 patients were: 63/1476 (4.3%) inferior oblique with mean severity 1.6; 94/1476 (6.4%) superior rectus with mean severity 1.7; 79/1476 (5.4%) lateral rectus with mean severity 1.4; 64/1476 (4.3%) medial rectus with mean severity 1.5; 36/1476 (2.4%) superior oblique with mean severity 1.6; and 84/1476 (5.7%) inferior rectus with mean severity 1.4. EOM weakness was seen in all serological subtypes, with 51/60 (85%) AChR+, 4/6 (80%) LRP4+, 8/8 (89%) MuSK+, and 43/49 (87%) SN, with no statistical difference on multivariable logistic regression (p=0.648).

**Discussion:** This large prospective OMG cohort shows that the distribution of ocular signs was similar across all serological subtypes.

## Poster Abstracts

### 1. Multiple Cranial Nerve Palsies due to Lemierre's Syndrome Associated with Atypical *Streptococcus pyogenes* Infection

Chathu De Silva, Toby Lindenbaum  
Royal Victoria Infirmary, Newcastle

**Case report:** We present a 38-year-old, normally fit and well man who presented to the eye casualty with red eyes and double vision. He also complained of a 2 week history of a sore throat, headaches, neck pain with stiffness and reported fever. His visual acuity was 6/9 in both eyes. He had bilateral periorbital oedema, chemosis, proptosis and general restriction of all extraocular muscles. His remaining cranial nerve exam revealed bilateral reduced corneal sensation and facial sensation in a V1 and V2 distribution. Optic nerve appearance and function was normal. He was pyrexial (39.1°C) and tachycardic (122bpm) with raised inflammatory markers, (WCC 12.57 and CRP 243). A CT head scan with venography showed a subtle blush of contrast within the central midbrain and a dilated right superior ophthalmic vein suggestive of a cavernous sinus thrombosis. He was admitted for IV antibiotics. His blood cultures were positive for Group A *Streptococcus pyogenes* and, given the recent history of sore throat, a CT scan of his neck and chest was undertaken. The scan showed a septic thrombosis of the right internal jugular vein, a right neck abscess, severe chest sepsis with multiple septic emboli and large necrotic consolidation in the right lower lobe, as well as a possible small splenic abscesses. These findings were consistent with Lemierre's syndrome. He was reviewed by multiple subspecialities and due to the early recognition and IV antibiotic treatment, made an excellent recovery. He completed a 4-week course of IV ceftriaxone. Serial CT scans showed resolution of his superior dilated ophthalmic vein and he had complete ocular and neurological symptom resolution.

**Discussion:** Lemierre's syndrome, often described as 'the forgotten disease' is a rare and life threatening condition. The syndrome is described as an oropharyngeal infection leading to thrombophlebitis of the internal jugular vein and septic embolization to other organs.

There is scarce evidence for the treatment of Lemierre cases, with the mainstay being prolonged systemic antibiotic therapy and surgical drainage of the infected site, if indicated. There is contested evidence surrounding the use of therapeutic anticoagulation.

Cavernous sinus thrombosis has been associated with Lemierre's syndrome amongst other ophthalmic complications. Whilst our colleagues in the ENT department may be more familiar with Lemierre's syndrome, within

Ophthalmology, it is a lesser-known cause of cavernous sinus thrombosis. Requiring a higher degree of clinical suspicion, the presence of ophthalmic manifestations indicate advanced, aggressive disease and is crucial that it is not missed on presentation.

## **2. The Crossroads of Leber’s Hereditary Optic Neuropathy and Autosomal Dominant Optic Atrophy: Unique Clinical Profiles in Patients with Coexisting Genetic Mutations**

Mohammed A. Halawani<sup>1</sup>, Nooran O. Badeeb<sup>2,3</sup>

1. Department of Ophthalmology, Ministry of National Guard-Health Affairs, Jeddah, Saudi Arabia
2. Department of Ophthalmology, University of Jeddah, Jeddah, Saudi Arabia
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**Background:** Hereditary optic neuropathies have highly variable genetic and clinical characteristics with a 1 in 10,000 prevalence. Both Leber’s hereditary optic neuropathy (LHON) and autosomal dominant optic atrophy (ADOA) are hereditary optic neuropathies characterized by mitochondrial dysfunction that induces destruction of the retinal ganglion cells and their axons, resulting in painless bilateral vision loss and symmetrical temporal pallor of the optic nerve heads.

**Case presentation:** Herein, we present six cases from one family with inherited ocular disorders. Three male cases had symptomatic bilateral vision loss, two were asymptomatic females while the other female had visual loss in her left eye accompanied by headache. She had a history of gazing at the sun with her left eye since childhood. All six patients had a normal anterior and posterior segment examination aside from the affected optic nerves except for the one female. The genetic examination, handled as a pedigree family diagram, showed that the first generation (grandparents) were unaffected and the second generation had an affected male (parents). In the third generation (grandchildren) an affected male (referred to as Case 3), diagnosed with ADOA - OPA1/heterozygous/nonsense/NM\_130873.2:c.2383C>T/p.(Gln795\*), married an affected woman (referred to as Case 4), diagnosed with LHON - MT-ND4/Missense/Homoplasmic/NC\_012920.1:m.11778G>A/p.ND4:(Arg340His). Their offspring (great-grandchildren) were one unaffected female, one affected female and two affected males diagnosed with two reported mutations inherited from their parents.

**Conclusion:** Mitochondrial optic neuropathies, which share mitochondrial malfunction selective injury and loss of retinal ganglion cells, are a substantial

cause of visual impairment with no cure. Herein, we reported two unique cases of combined LHON and ADOA mutations.

### **3. Metadiagnosis: Actuarial Estimation of Strength of Belief in Diagnosis With GCA as the Exemplar**

Louis Clearkin

Royal Free Hospital, London

**Purpose:** To present results of investigation into diagnostic error

**Methods:** Probabilistic evaluation of GCA in each patient referred to a single centre.

**Results:** 1) The correlation between informal estimation of diagnosis compared to the probability estimate that reference standard test was positive.

2) Influence of reference standard test result on final diagnosis.

**Discussion:** Viewing clinical findings dichotomously obscures diagnostic uncertainty. The limitations of clinical diagnosis can be offset by applying epistemological and quantified expressions of diagnostic belief, numeric probability, confidence in the probability estimate, and derived information. The practical relevance is illustrated by applying it to a common clinical problem. This process of “metadiagnosis” provides a new perspective whereby strength of belief in a diagnosis can be accurate and explicit.

### **4. Ocular Motor Cranial Neuropathy and Cardiovascular Risk Factors: A Systematic Review and Meta-analysis**

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**Aim:** This review examines the role of ocular motor cranial nerve palsy (OMCNP) in identifying uncontrolled vascular disease risk factors, investigates the link between major cardiovascular events and OMCNP, and looks into the recurrence rate of OMCNP to improve cardiovascular disease diagnosis and treatment.

**Methods:** A systematic review was conducted using PubMed, Web of Science, and EMBASE databases. The focus was on subjects aged 16 and older with OMCNP (3rd, 4th, and 6th cranial nerves) and one or more vascular disease risk factors, such as diabetes, hyperlipidaemia, smoking, high blood pressure, obesity, and obstructive sleep apnoea. Statistical analysis was conducted using SPSS, with a meta-analysis of proportions illustrated by forest plots.

**Results:** Among the study groups, hypertension had the highest prevalence at 67.71%, followed by diabetes at 47.89%, hyperlipidaemia at 36.10%, and smoking at 24.71%. Data on obesity and sleep apnoea in patients with OMCNP were

lacking. Cardiovascular events were more prevalent in patients with OMCNP, stroke at 13.65% and with heart failure at 7.63%. Evidence on myocardial infarction and the recurrence rate of OMCNP was limited.

**Conclusion:** This systematic review underscores the strong association between OMCNP and vascular risk factors like hypertension and diabetes. Despite prevalence data limitations in establishing causality, the findings underscore the necessity for standardised OMCNP management guidelines. Future research should prioritise prospective randomised controlled trials to clarify each risk factor's impact and intervention benefits in preventing neuropathy and associated cardiovascular events.

### **5. Newly Validated Eye-Tracking Test Quantifies Foveal Dark Adaptation Latency in Foveal Cones and Retinal Neurons: Implications for Assessing Macular and Neuro-Ophthalmological Disorders**

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3. Meddoc Research

**Purpose:** Accurate differentiation between retinal/macular disorders and neuro-ophthalmological conditions is crucial for effective diagnosis and management. This study aimed to validate a novel eye-tracking-based foveal and pupillary function test, the ACOLAPT (Acuity, Contrast, Light Adaptation, Pupillary Test), to establish previously unavailable biomarkers for retinal function. By evaluating foveal cone- and retinal neuron-related dark adaptation latency alongside detailed pupillary dynamics, we sought to provide clinicians with a comprehensive tool for differential diagnosis and monitoring. The test was validated on patients with age-related macular degeneration (AMD), diabetics without diabetic retinopathy (DR), diabetics with DR, and healthy controls (HC).

**Methods:** In a controlled clinical study, 17 patients with early to moderate dry AMD, 16 diabetic patients without DR, 16 diabetic patients with DR, and age-matched healthy controls were assessed using the ACOLAPT test. Foveal function metrics included detection acuity (DA), contrast sensitivity (CS), and foveal cone-mediated dark adaptation latency, assessing both photoreceptor and retinal neuronal function. Pupillary function was evaluated using eye-tracking-derived metrics such as pupil diameter, constriction and dilation velocities, latencies, accelerations, and jerk (rate of change of acceleration). The relative afferent pupillary defect log scale (RAPDlog) was also measured. Statistical analyses comprised receiver operating characteristic (ROC) curves for discriminative



capacity, intraclass correlation coefficients (ICC) for reliability, and Bland-Altman plots for repeatability. The Agreement Index (AI) and Stability Index (SI) were calculated to assess clinical utility.

**Results:** The ACOLAPT test effectively differentiated patients in the study from healthy controls, showing statistically significant impairments in foveal function variables. AMD patients exhibited increased cone-mediated dark adaptation latency, indicating dysfunction in foveal cones and retinal neurons. Pupillary function tests revealed significant alterations in pupil dynamics - including changes in diameters, velocities, latencies, accelerations, and jerk - in AMD and diabetic patients, reflecting retinal neuronal dysfunction. In contrast, neuro-ophthalmological disorders typically present with different pupillary response patterns, highlighting the test's potential in differential diagnosis. AI and SI indicated good repeatability and stability for both foveal and pupillary assessments.

**Conclusion:** The ACOLAPT test, combining eye-tracking-based assessments of foveal cone-mediated dark adaptation latency and detailed pupillary dynamics, offers valuable insights for distinguishing retinal/macular disorders from neuro-ophthalmological conditions. By quantifying impairments in retinal photoreceptor and neuronal function alongside specific pupillary response patterns, these metrics enhance the differential diagnosis of visual dysfunctions critical to neuro-ophthalmology. Validation in AMD and diabetic populations underscores its potential utility in clinical practice, and potential in Neuro-ophthalmological differential diagnosis and monitoring.

## **6. Microvascular Decompression of the Optic Nerve for Progressive Visual Field Loss**

Sammie Mak, Oliver Backhouse, Asim Sheikh  
Leeds Teaching Hospitals NHS Trust

**Case report:** A 51-year-old patient presented with a left supero-nasal visual field deficit. On examination bilateral fundoscopy was unremarkable, optical coherence tomography (OCT) of both nerve fibre and ganglion cell layers were within normal limits and there was no relative afferent pupillary defect (RAPD). Magnetic resonance imaging (MRI) demonstrated left pre-chiasmatic optic nerve compression between the internal carotid artery (ICA) and the falciiform ligament, causing mild distortion. Initial management was a conservative approach over 6 months. During this period there was both subjective and objective progressive loss of the left supero-nasal visual field. Repeat MRI revealed new focal T2 signal change within the optic nerve at the site of compression by the ICA. Subsequently,

a left supra-orbital craniotomy and microvascular decompression of the optic nerve was performed. Post-operatively, the patient had an immediate subjective improvement in the left visual field and Humphrey visual fields at 3 weeks post-operation showed dramatic resolution of the deficit.

**Discussion:** The intracranial optic nerve and supra-clinoid portion of the ICA are anatomically in close proximity. Whilst mass lesions often cause visual loss, the proximity of these two structures is typically physiological rather than pathological. However, it is well established that vascular compression of other cranial nerves (most commonly V & VII) can lead to their dysfunction and neurological symptoms. Microsurgical decompression is a standard treatment in these situations. Within the literature there is a debate as to whether carotico-falciform compression truly causes optic neuropathy or whether it is an incidental finding given the close physiological relationship between the structures. There are several case reports where microvascular decompression of the optic nerve has led to improvement in vision. On the contrary there have also been reports of deterioration in vision after one such operation, but which later improved with steroids. It has been argued that the mechanism of optic neuropathy from vascular compression may include both direct compression injury and chronic ischaemia from compromised regional perfusion. This case adds to the few previous case reports of optic neuropathy from vascular compression treated surgically. In conclusion, optic nerve compression from an ectatic ICA can lead to neuropathy via the same mechanism seen in other cranial nerve neuropathies. Furthermore, microvascular decompression in certain patients may lead to good clinical outcomes, but there are no clear evidence-based recommendations. However, due to the close relationship between the two structures, a compressive neuropathy should remain a careful diagnosis of exclusion.

## **7. Maximising OCT in the Differential Diagnosis of Papilloedema from Pseudopapilloedema**

Lorcan Butle

Newtownards, Northern Ireland

Differential diagnosis between the two conditions can be extremely difficult for any trainee ophthalmologist leading to unnecessary testing and possible harm to the patient. This presentation will discuss OCT findings that are found in papilloedema which will differentiate it from pseudopapilloedema. Discussing Bruch's membrane forward angulation, mGCL-IPL, wrinkles/folds, EDI OCT, PHOMs, HRB and information from the ODDS consortium to help guide the trainee in their assessment.

## 8. Occlusive yet Elusive: A Young Patient with an Unusual Deterioration in Vision Following Treatment for a Cerebellar Medulloblastoma

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2. University College London Hospitals

3. National Hospital for Neurology and Neurosurgery

**Case report:** A 19-year-old patient with type 1 diabetes mellitus and a cerebellar medulloblastoma (treated recently with surgery, chemotherapy & radiotherapy) presented with gradually worsening vision to 2/60 right eye (OD) and 3/60 left eye (OS) with a baseline of 6/6 OD & 6/9 OS. Being 17 at the time they were referred as possible diabetic retinopathy. Examination revealed bilateral disc swelling, cystoid macular oedema (confirmed on OCT) and intraretinal haemorrhages but no neovascularisation. Visual field defects were also noted (OD > OS). Fundus fluorescein angiography showed bilateral peripheral capillary dropout and macular ischaemia. This was diagnosed as bilateral central retinal vein occlusion (CRVO) though a trigger remained unclear with the differential diagnosis including radiation retinopathy following tumour treatment. Treatment with intravitreal therapy (IVT) and pan-retinal photocoagulation (PRP) laser was agreed upon following discussion with the oncologists and ophthalmologists. The patient received 4 x bevacizumab injections bilaterally (before turning 18) and 7 x Faricimab injections OS along with the PRP laser. The patient currently has no macular oedema or neovascularisation, and visual acuity is stable at 6/60 OD and 6/24 OS with pinhole improvement to 6/48 OD and 6/19 OS.

**Discussion:** Radiation retinopathy may cause retinal ischaemia and macular oedema - closely mimicking CRVO in many ways apart from the speed of disease progression.<sup>1</sup> Oncology input concluded that radiation retinopathy was unlikely as the levels of Gy used (2Gy/fraction) were within the tolerated range. However, CRVO is unusual in such a young patient as many classical risk factors for thrombosis were absent. Hypercoagulability secondary to malignancy is a possibility as brain tumours have a diverse effect on systemic coagulability.<sup>2,3</sup> However, studies are yet to indicate medulloblastomas having a procoagulant effect, with some subtypes displaying reduced tissue factor expression and thrombotic propensity.<sup>5</sup> Post-chemotherapy, the patient was in fact, frequently thrombocytopenic (as low as  $27 \times 10^9/L$ ) - posing a challenge for IVT as local guidelines advise a cut-off of  $100 \times 10^9/L$ . Chemotherapy itself may also elevate the risk of venous thromboembolism with reports suggesting certain agents possibly contributing to central retinal artery occlusion and CRVO.<sup>5-7</sup> Another concern was the initial use of IVT as bevacizumab is currently not licensed for use

in under 18s. As bevacizumab has not been proven to be inferior to other agents such as ranibizumab currently (LEAVO trial), it was used off-licence for the treatment of CRVO with macular oedema in our patient.<sup>8</sup>

#### References

[https://docs.google.com/document/d/1Z\\_2p4lO6dWL0UTpZ12eQGzeEeAt3Oau6lVQ8VZVlrLY/edit?usp=sharing](https://docs.google.com/document/d/1Z_2p4lO6dWL0UTpZ12eQGzeEeAt3Oau6lVQ8VZVlrLY/edit?usp=sharing)

### 9. Unmasking the Curious Case of Unilateral Flushing

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**Case report** A 54-year-old male noted striking asymmetrical facial flushing after cycling outdoors. The right side of his face was flushed from the exercise, but the left was pale, with a sharp vertical midline demarcation, which he photographed. On closer inspection, anhydrosis of the left side of his face was also present. Review in the Ophthalmology clinic further identified subtle left ptosis and miosis. Pupil size, measured with the Plusoptix Vision Screener, was 3.5 mm (right) and 3.1 mm (left). He subsequently underwent urgent investigations for Horner's syndrome. CT angiography revealed a dissecting aneurysm of the left internal carotid artery, measuring 8.6mm, at the level of the skull base. MRI head showed no evidence of acute infarction. A CT scan of his thorax revealed no apical lung or cervical rib lesions. We concluded the dissecting aneurysm of the left internal carotid artery was exerting intrinsic pressure on the cervical ganglion of the sympathetic trunk, causing Horner's syndrome with Harlequin's sign.

**Discussion:** Harlequin's sign is characterised by asymmetrical facial flushing and sweating with a sharp demarcation in the midline. It was first described by Lance et al. in 1988 and was named after its resemblance to the character "Harlequin" in Italian theatre. It is most evident after strenuous exercise, strong emotion or exposure to heat. The Harlequin sign is caused by unilateral dysfunction of sympathetic fibres carrying vasodilator and sudomotor nerves to the face. Normally, activation of these fibres causes vasodilation and sweating, resulting in heat loss to maintain thermoregulation. Unilateral dysfunction of these fibres causes ipsilateral vasoconstriction and anhydrosis, with compensatory hyperaemia and sweating on the contralateral side, resulting in the hemifacial discolouration characteristic of Harlequin sign. If the sympathetic deficit also encompasses the oculosympathetic fibres then concomitant Horner's syndrome is seen. It is extremely rare for internal carotid artery lesions to cause Horner's syndrome with Harlequin sign. This is because sudomotor and vasomotor

sympathetic fibres follow the course of the external carotid artery to innervate the blood vessels and sweat glands of the face, whereas oculosympathetic fibres ascend along the internal carotid artery. Therefore, internal carotid artery dissection usually only disrupts the oculosympathetic fibres, causing a partial Horner's syndrome with ipsilateral ptosis and miosis, without anhidrosis or vasoconstriction of the face. Therefore, this case illustrates the importance of keeping an open mind when investigating Horner's syndrome, as the underlying cause can be unexpected, and not readily explained by conventional anatomy teaching.

### **10. Sight Registrations via the Certificate of Visual Impairment in a Tertiary Neuro-ophthalmology Clinic: A Service Evaluation Spanning 6 years in Sheffield Teaching Hospitals NHS Foundation Trust**

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Sheffield Teaching Hospitals NHS Foundation Trust

**Introduction/Purpose:** Sight registration, via the certificate of visual impairment (CVI), is important in the care of patients with irreversible sight loss. Patients' quality of life is improved through access to social and financial support. Accurate CVI registration also helps provide epidemiological data. Some conditions causing sight loss in the Neuro-ophthalmology clinic are neither preventable nor treatable (such as hereditary or anterior ischaemic optic neuropathies), while some are potentially treatable causes, including optic neuritis and fulminant idiopathic intracranial hypertension (IIH). We also see patients with higher order visual processing difficulties (HOVPD), who, despite having good visual acuity, may have severe impairment of visual function, yet remain unaccounted for based on the current CVI criteria. There is also the dilemma as to whether patients with functional visual loss should be registered. This service evaluation aimed to understand the spectrum of pathologies that led patients to being registered as sight impaired (SI) or severely sight impaired (SSI) within our Neuro-ophthalmology service. We additionally sought to understand whether the registrations complied with CVI guidance, and which group of patients had potentially preventable/treatable causes of sight loss.

**Methods** The clinical notes of all patients registered as SI/SSI in the Neuro-ophthalmology clinic between June 2017 and June 2023 were reviewed.

**Results** We registered 80 patients as SI and 48 as SSI. The commonest aetiologies were stroke (n=21), non-arteritic anterior ischaemic optic neuropathy (n=19), and tumours (n=17). On review, 13 patients were registered as SI without strictly fulfilling the criteria, seven of whom were registered due to quadrantanopias. Two

patients were registered as SSI despite strictly fulfilling only the SI criteria. Five patients were registered due to HOVPD but none due to functional vision loss. Potentially preventable/treatable causes (n) included IIH (8), giant cell arteritis (GCA, 5), peri-operative complications (4), meningitis (2), multiple sclerosis (2), ethambutol toxicity (1) and radiation optic neuropathy (1).

**Discussion** Although the CVI guidance is necessarily ambiguous, it potentially contributes to variation in individual clinical practice for CVI registrations amongst ophthalmologists, particularly in relation to quadrantinopias, HOVPD and functional visual loss. Consensus within the Neuro-ophthalmic community could ensure equity of patients' access to CVI registration. Furthermore, if the classification of the aetiologies on the CVI form were refined in the Neuro-ophthalmology section, this could give greater insights into epidemiology of sight loss within our service, which is particularly important for potentially preventable causes of SI/SSI, such as IIH and GCA.

## 11. A Rare Ptosis

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**Case report:** A 64-year-old White Caucasian patient attended her GP with an unusual complaint - she had been unable to open her right eye spontaneously in the morning, or if she awoke overnight, for the past 9 months. It appeared to "reset" on mechanically elevating the right upper eyelid with a finger. Once the eyelid was lifted, there was no recurrence of ptosis during the day. She did not notice any variance in eyelid position, involuntary eyelid closure, diplopia or loss of vision. She did not report that the eyelids felt "stuck". There was a history of myopic LASIK with monovision and mild dry eyes, managed with topical lubricant drops. There was no history of neurological or autoimmune conditions. Her regular medication included HRT. Examination demonstrated normal vision, visual fields, pupils, and extraocular movements in both eyes. There was no apparent ptosis, and good levator palpebrae superioris function was present bilaterally. There was mild bilateral blepharitis and dry eyes, and the LASIK flap was clear in both eyes. Dilated funduscopy was unremarkable.

**Discussion:** This patient gives an excellent history of a rare phenomenon - awakening ptosis. While it was first described in the 1890s, there is no consensus on the correct nomenclature, with authors referring to awakening ptosis, benign unilateral apraxia of eyelid opening, sleep-induced apraxia of eyelid opening, and hypnopompic eyelid palsy all offered as descriptions. It is usually unilateral and resolves spontaneously within several minutes of waking, or on mechanical

elevation of the eyelid. It often resolves entirely within 18 months of onset. The case reports and series show a female preponderance, with the typical patient a White Caucasian female in their 60s. No association with neurological conditions has been reported, although coexisting autoimmune conditions are reported. There is discussion as to whether it represents a parasomnia with inappropriate persistence of REM sleep paralysis of the levator palpebrae superioris or is a variant of apraxia of eyelid opening. Reassuringly for our patient, neuroimaging and extensive investigation are not warranted in the absence of associated neurological findings.

## **12. Remission from Group Consultations for People 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. To manage the service demand and improve quality of care, we set up group consultations (GC) at St Thomas' Hospital in 2019. An mPhase 1 Quality Improvement (QI) report of GC showed that patients preferred GC as opposed to the traditional one-to-one (1:1) consultation, with median patient satisfaction scores of 9.5/10; and feeling listened to by clinician scores of 10/10. As Phase 1 was during the COVID-19 pandemic we repeated this QI evaluation in 2023 (Phase 2). The phase 2 QI report showed 98% of those asked in Phase 1 still preferred GC to 1:1. This was presented at the UKNOS 2024 meeting. Here, we report the Phase 3 QI evaluation on the effectiveness of GC in terms of IIH remission rate.

**Aim:** To audit the IIH remission rates of patients attending the IIH GC service from Nov-23 to Nov-24.

**Method:** Case note review of patients seen between Nov-23 and Nov-24.

Remission was considered for patients not on medication, having no papilloedema on examination and being free of IIH symptoms.

**Results:** 56 patients with IIH were seen in GC between Nov-23 and Nov-24. 14/56 (25%) of patients seen in GC during this period were declared to be in remission. Of these, 11/14 (79%) of patients in remission joined GC from the start of their diagnosis. Of those not in remission, 22/25 (88%) joined GC after a median of 41 months (range 9 - 175 months) following their diagnosis, Fisher's exact test  $p=0.06$ .

**Conclusion:** GC have successfully supported people with IIH safely, effectively, and continue to be the preferred method of contact at our unit. There was a trend to suggest that early entry into GC from time of diagnosis may support disease remission through peer support and lifestyle changes. Future study plans include comparing IIH remission rates in those seen in 1:1 setting compared to GC, and whether GC improves the likelihood of remission.

### **13. Blindsided by a Bad Bite**

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Mid & South Essex NHS Foundation Trust

**Case report:** A 43-year-old female with a history of type 1 diabetes mellitus presented with diabetic ketoacidosis (DKA) after a 10-day history of persistent vomiting. In the emergency department she was found to in severe metabolic acidosis, acute kidney injury and hypotensive requiring vasopressor support. She was managed with renal replacement therapy and the DKA protocol. After 4 days admission her consciousness improved and she reported bilateral loss of vision. Ophthalmological evaluation revealed visual acuity of perception of light, bilateral non-reactive dilated pupils and fundoscopy consistent with non-proliferative diabetic retinopathy. Neuroimaging, including magnetic resonance imaging and magnetic resonance venography were within normal limits. Optical coherence tomography showed no structural abnormalities of the macula or optic nerve in both eyes. A 3-day intravenous course of methylprednisolone led to no improvement in her vision. Follow up after 1 month showed bilateral disc pallor, which pointed towards the diagnosis of posterior ischaemic optic neuropathy (PION).

**Discussion:** Bilateral PION is a rare complication of DKA and has only been mentioned in a handful of case reports. The pathophysiology is thought to be associated with systemic hypotension and metabolic derangement. The retrobulbar optic nerve is particularly vulnerable to ischaemia due to its limited vascular supply in this compromised situation. Management involves addressing systemic factors, but the visual prognosis remains poor. This case highlights the need for prompt recognition and intervention in DKA to minimise systemic and ocular complications.

### **14. Risk Factor Analysis in a Transgender Individual on Hormone Therapy of Developing Idiopathic Intracranial Hypertension**

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## 2. Leeds

**Purpose:** To look into risk factors for idiopathic intracranial hypertension (IIH) in transgender individuals on hormone therapy.

**Methods:** All patients with IIH attending the Neuro ophthalmology services of Manchester Royal Eye Hospital between January 2023 to December 2023 and who had undergone gender transition were included in the study. Patients with obstructive hydrocephalus were excluded. Consent of disclosure was obtained.

**Results:** We identified five patients with IIH and a history of gender transition. One case with obstructive hydrocephalus secondary to type 2 Chiari malformation was excluded.

**Case 1:** 35-year-old female (F) to male (M) transition presented with headache and tinnitus. He had started hormone therapy 6 years ago with a recent rise of body mass index (BMI) by 6 points. The papilloedema resolved with oral acetazolamide and weight loss. He continued hormone therapy with intravenous (IV) testosterone.

**Case 2:** 30-year-old F to M transition was asymptomatic and incidentally noted to have papilloedema by his opticians. He had started transition 3 years ago and had recently shifted from a transdermal testosterone patch to IV testosterone. His BMI had recently increased by 7 points. The papilloedema resolved with oral acetazolamide and aggressive weight loss measures

**Case 3:** 20-year-old F to M transition presented with headache. Transdermal testosterone had been started 6 months previously. His BMI had risen by 2 points. After a further 6 months they stopped the testosterone as they did not wish to continue with gender transition. The papilloedema resolved without any treatment.

**Case 4:** 42-year-old M to F transition on oestrogen therapy presented with visual blurring and grade 4 optic disc oedema in both eyes with associated visual field and ganglion cell loss in both eyes. Her BMI had risen by 12 points. She was intolerant of acetazolamide due to renal co morbidities and underwent ventriculo-peritoneal shunting with complete resolution of the disc swelling.

**Conclusion:** Gender transition can significantly impact an individual's mental and social well-being. These patients have a dual risk of developing IIH secondary to hormone therapy and related change in BMI. Appropriate education and proper screening of individuals with high baseline BMI is recommended. Our review highlights that with proper weight management and a multi-disciplinary approach, patients can safely proceed with hormone therapy for gender reassignment.

## 15. Traumatic Optic Neuropathy: Belfast Urgent Eye Care Experience

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**Introduction:** Traumatic optic neuropathy (TON) may be caused by direct injury to the optic nerve or, more commonly, via indirect trauma transmitted by the skull base. Accurate initial diagnosis of TON may be challenging due to multiple confounding injuries (ocular, orbital and cranial). The aim of this study is to report the accuracy of initial diagnosis of TON, along with subsequent patient investigation, management and outcome.

**Methods:** Five years of data were reviewed from the Belfast eye emergency electronic data system (Symphony, EMIS Health) between 2019 and 2023, inclusive. Approval was obtained from the Belfast Health and Social Care Trust.

**Results:** Sixteen patients had an initial diagnosis of TON, of these 15 (93.8%) were male. Of the 16 patients with an initial diagnosis of TON, 13 (81.3%) attended follow-up, with eight (61.5%) of those attending follow-up having a final diagnosis of TON. At the initial presentation, seven (43.8%) had a relative afferent pupillary defect (RAPD) noted. Of the eight patients with a final diagnosis of TON, two (25%) were not reported to have an RAPD at the initial examination. Ten (58.8%) had computed tomography scanning of the brain and orbit, five (29.4%) had magnetic resonance imaging, and six (35.3%) were diagnosed with TON clinically. Two patients (11.7%) had fractures involving the optic canal. Two (11.7%) patients underwent electrophysiology. Thirteen (81.3%) were observed, two (12.5%) patients received glucocorticoids: one with oral prednisolone (prescribed by an outside centre); and one with a course of intravenous methylprednisolone. One patient underwent surgical decompression via fracture reduction. Five (38.5%) of the 13 patients who attended follow-up were reviewed in a Neuro-ophthalmology clinic. For patients with a final diagnosis of TON, the median presenting visual acuity was 1.0 logMAR (range: 0.3 to 3.0) and the final visual acuity was 0.6 logMAR (range: 0.0 to 1.8). The mean visual change was -0.52 logMAR (improvement). The presenting and final visual acuity correlation coefficient was 0.882 (strong positive correlation) with  $p = 0.038$ .

**Discussion:** The accuracy of initial diagnosis has been demonstrated to be limited (61.5% of those who attended follow-up) and this may be due to multiple confounding injuries. The use of glucocorticoids in the management of TON has decreased in clinical use, due to concerns of lack of efficacy and adverse event, and this is reflected in these data.

## 16. Eye Muscle Surgery in a Patient with Downbeat Nystagmus

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York Hospital

**Purpose:** To describe eye muscle surgery in a patient with acquired downbeat nystagmus (DBN) associated with autoimmune encephalitis.

**Methods:** Interventional case report. Analysis of clinical data before and after eye muscle surgery in a patient with acquired downbeat nystagmus who did not respond to prisms and medications.

**Results:** The patient had a null point in up-gaze with a compensatory chin down head posture. The aim of eye muscle surgery was to move the null position in up-gaze associated with DBN closer to primary position by moving both eyes down. She underwent bilateral superior rectus recessions of 5.0 mm combined with bilateral inferior oblique myectomy. There was a subjective improvement in oscillopsia, anomalous head posture (AHP) and visual acuity following surgery. There were no serious postoperative complications.

**Conclusion:** This study describes eye muscle surgery as a treatment option for patients with acquired downbeat nystagmus, which can result in an improvement in oscillopsia and AHP.

## 17. Optic Disc Swelling in Eye Casualty: Are We Doing it Right?

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**Introduction/Purpose:** Optic disc swelling is a common presentation to eye casualty that has vision and life-threatening implications. It is a manifestation of a broad range of pathologies affecting the optic nerve. Accurate assessment of any suspected optic disc swelling to recognise its cause is therefore crucial. However, accurate diagnosis of disc oedema is often challenging in busy eye casualty. We performed a clinical audit assessing the utility and safety of the Trust's 'Papilloedema Pathway' for patients presenting to eye casualty with suspected optic disc swelling due to raised intracranial pressure.

**Methods:** Electronic patient records were used for a retrospective data collection of adults and children presenting with suspected optic disc swelling who had investigations and referrals under the Trust's 'Papilloedema Pathway' from 1 March 2023 to 30 June 2023. Data collected included: basic demographics; presenting symptoms; examination findings; investigation results including head

imaging and lumbar puncture; diagnosis; treatment; and follow-up. Patients who did not follow the 'Papilloedema Pathway' and those who had inappropriate diagnosis or treatment were identified.

**Results:** 58 patients (8 male, 50 female) in eye casualty were managed using the 'Papilloedema Pathway' during the study period. The mean age was 29.6 years and there were 10 children. Headache was the commonest presenting complaint. Most patients (82.7%) presented with good vision ( $\geq 6/12$  Snellen visual acuity). Spontaneous venous pulsation was not documented in 78% of patients. Frisè grade was commonly used (72.4%) to grade the severity of disc swelling. 5 patients had severe papilloedema defined by Frisè grade 4-5. 13 patients (22.4%) were admitted and 38 patients (65.5%) had brain magnetic resonance imaging. 81% of our patients followed the 'Papilloedema Pathway', receiving timely investigation, diagnosis and management. The diagnosis was mostly made by non-consultant grades (73.2%) and only 58.6% of the diagnoses were correctly made on retrospective review. Two patients who were under the 'Papilloedema Pathway' had a final diagnosis of optic neuritis, one patient had ischaemic optic neuropathy and another had idiopathic intracranial hypertension, although was initially thought to have optic disc drusen. 22 patients were deemed to have a final diagnosis of pseudo-papilloedema and 10 patients had unnecessary lumbar punctures as a result. There was no patient safety incident reported from this audit.

**Conclusions:** The 'Papilloedema Pathway' was safely utilised in the eye casualty setting in managing patients with suspected papilloedema. Ongoing training of eye casualty staff is essential for accurate assessment and documentation of findings of optic disc swelling for appropriate diagnosis and treatment.

### **18. Interleukin-6 Receptor Signaling Inhibition With Satralizumab in Thyroid Eye Disease: Phase 3 SatraGO-1 and SatraGO-2 Trial Design**

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**Introduction/Purpose:** Thyroid eye disease (TED) is an inflammatory orbitopathy that can cause facial disfigurement and sight-threatening complications. There are two distinct phases of TED: an active phase characterized by ongoing inflammatory activity and orbital tissue changes; and a chronic inactive phase during which symptoms may improve but not fully resolve. There is a significant need for disease-modifying treatments for active TED as current therapies may be ineffective, cause relapses, and/or have considerable side effects, and for inactive TED, which is primarily treated surgically. Interleukin-6 (IL-6) plays a key upstream role in immune regulation and is elevated in people with TED. Blocking the IL-6 receptor (IL-6R) has the potential to reverse the manifestations of the disease, such as proptosis and diplopia, and restore normal immune function.

Satralizumab is a humanized monoclonal anti-IL-6R monoclonal antibody approved for the treatment of AQP4+ neuromyelitis optica spectrum disorder. Satralizumab utilizes innovative antibody recycling technology, enabling rapid and sustained suppression of inflammatory pathways. Here, we describe the SatraGO-1 and SatraGO-2 trials, which have been designed to evaluate the efficacy and safety of satralizumab as a disease-modifying treatment TED, with subcutaneous administration every 4 weeks (Q4W).

**Methods:** SatraGO-1 (NCT05987423) and SatraGO-2 (NCT06106828) are identical, global, phase 3, randomized, double-masked, placebo-controlled, 72-week multicentre studies that will recruit ~120 participants at ~70 sites across 20 countries. Participants  $\geq 18$  years with moderate-to-severe active TED or stable, chronic inactive TED are eligible provided the systemic disease is under control (euthyroid or mild hyper-/hypothyroidism). Participants will be randomized 1:1 to subcutaneous satralizumab or placebo at weeks (W) 0, 2, and 4 (loading doses) and then Q4W through W20 (maintenance doses). Based on the W24 proptosis response, non-responders will receive satralizumab Q4W and responders will be re-randomized 1:1 to satralizumab or placebo Q4W through W44.

**Results:** The primary endpoint is the proportion of active TED participants achieving a proptosis response ( $\geq 2$  mm proptosis improvement from baseline in study eye) at W24. Secondary endpoints include proptosis response in active/chronic TED participants and overall response ( $\geq 2$ -point improvement in clinical activity score from baseline and proptosis response in study eye) in active TED participants. Safety outcomes include incidence, seriousness, and severity of adverse events.

**Discussion:** SatraGO-1 and SatraGO-2 are designed to investigate IL-6R inhibition via satralizumab in TED. Satralizumab offers a potential disease-modifying

treatment in TED while minimizing safety risks associated with current treatments.

### **19. Internuclear Ophthalmoplegia and Pupil-sparing Third Nerve Palsy in a 6-Year-Old: A Case Report**

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Watford General Hospital, West Hertfordshire Teaching Hospitals NHS Trust, Watford

**Case report:** A 6-year-old girl presented with acute onset of diplopia and dizziness, peaking within 1 hour. Her history included 3 days of coryza, 2 days of fever responsive to paracetamol, and otitis media with effusion. There was no history of trauma and she was up to date with vaccinations. Her temperature was 38.1°C. Examination revealed right esotropia, intact direct, consensual and accommodation reflexes, equal and reactive pupils (3mm), a left internuclear ophthalmoplegia (INO), a right pupil-sparing 3rd nerve palsy, and bilateral restricted elevation with nystagmus. Neurological findings were otherwise unremarkable. Anterior segment examination showed quiet eyes with no adnexal, conjunctival, corneal, or lens abnormalities. Fundoscopy revealed bilateral elevated discs, with no macular changes and a flat retina. Magnetic resonance imaging of the brain and orbits showed high T2/FLAIR signal in the left midbrain, cerebral peduncle, and left basal ganglia, consistent with a recent ischaemic event. Cerebrospinal fluid (CSF) polymerase chain reaction revealed strongly positive Varicella zoster virus (VZV) IgG and negative IgM. VZV vasculopathy was diagnosed, and treatment included intravenous (IV) aciclovir, oral aspirin, and a tapering course of oral prednisolone. Bangerter occlusion foils were used to manage diplopia, alternating daily.

**Discussion:** The most common cause of acute diplopia is microvascular disease, however, this is unlikely in a child due to the lack of vascular risk factors. Another differential is stroke. While this has been reported in a child with acute diplopia caused by infarction secondary to a large patent foramen ovale, this is also unlikely. A multicentre Italian study found headache, strabismus, trauma, brain tumours, and demyelinating diseases to be the most common paediatric causes of acute diplopia. These were excluded based on history, examination, and imaging. Multiple sclerosis and brainstem infarction, the leading causes of INO, were also excluded. The patient's coryza and pyrexia suggested an infectious cause, confirmed as VZV cerebral vasculopathy via CSF analysis. VZV vasculopathy can result from primary infection or reactivation of latent VZV. IV aciclovir is the mainstay of treatment to resolve the infection and reduce complications, while

steroids improve recovery. Complications include stroke, vertebral or cervical artery dissection, and cerebral aneurysms. There is no clear guidance on antiplatelet therapy, but it may reduce stroke and aneurysm risk. Although aspirin is typically contraindicated in children due to Reye's syndrome, it was justified in this case to reduce aneurysm risk, similar to its use in Kawasaki's disease.

## **20. Understanding the Extent of Visual Snow Type Symptoms in Inherited Retinal Disease**

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**Introduction:** Visual snow syndrome (VSS) is a neurological condition of visual disturbance with a normal ocular examination. Due to the observation of referrals of patients with retinal diseases with VSS-like symptoms, this work aims to explore the extent and burden of such symptoms in patients with inherited retinal diseases (IRD).

**Methods:** Prospective, cross-sectional, single centre study performed in adult retinal genetics clinics at Moorfields Eye Hospital, London, from March 2024 to September 2024. All patients had a complete ophthalmological examination including best corrected visual acuity (BCVA) and had a confirmed or suspected diagnosis of IRD. To quantify patient-reported, VSS-like visual and non-visual symptoms, patients completed an 80-item questionnaire with the help of an examiner.

**Results:** Ninety-five patients (mean age  $46.0 \pm 17.8$  years, range 17-80; male:female ratio=54:41) with various IRDs were included. The main visual symptoms comprised photosensitivity (68.4% of patients), nyctalopia (62.1%) and floaters (49.5%). The main non-visual symptoms were tinnitus (24.2%), migraine headaches (23.2%) and anxiety (21.1%). Regarding the most bothersome symptoms on a scale from 0-10 with "0" as not bothersome, insomnia showed the highest values (mean  $6.8 \pm 2.8$ , 3-10), followed by migraine headaches (mean  $8.5 \pm 1.3$ , 5-10), and photosensitivity (mean  $7.2 \pm 2.6$ , 1-10). Symptoms of visual snow (flickering static across the whole visual field) were found in 12.6% (12/95) of patients (six with retinitis pigmentosa, four with Stargardt's disease, one with PRPH2 cone-rod dystrophy and one with CDH3 macular dystrophy). A greater proportion of this subgroup reported other VSS-like symptoms, such as palinopsia, photosensitivity, nyctalopia, diplopia, starbursts, halos, phonophobia, anxiety and depression, compared with the main group. The intensity of flickering vision did

not correlate with visual acuity ( $R=-0.15$ ,  $p=0.65$ ), nor with age at symptom onset ( $R=-0.08$ ,  $p=0.79$ ).

**Discussion:** The most commonly reported VSS-like symptoms in patients with IRD, as expected, included photosensitivity and nyctalopia, but floaters were also common. Interestingly, patients perceived associated non-visual symptoms as more disturbing. Flickering vision, similar to visual snow in VSS, was found in 12.6% of patients with IRDs, and a higher proportion of this subgroup reported additional VSS-like symptoms.

## **21. The Posterior Cranial Fossa as an Anatomical Antagonist: A Case Report**

William Wong

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**Case report:** A 25-year-old woman presented to the eye casualty at Moorfield Eye Hospital with a 1-week history of horizontal binocular diplopia and reportedly slightly blurred vision in the left eye. She had been having intermittent episodes of headache with associated nausea and vomiting over a 3-week period. She had no past medical or drug history of significance. Having seen a private GP a preliminary diagnosis of labyrinthitis was made and this had resolved at the time of presentation. She was referred to Watford eye casualty for further assessment and neuro-imaging. Review by the local ophthalmology team uncovered a left 6th nerve (CN VI) palsy and bilateral papilloedema on fundoscopy. Magnetic resonance imaging (MRI) of her brain and cervical cord revealed a large posterior fossa cystic lesion with a solid nodule and associated mass-effect at the pontomedullary junction and cerebellar hemispheres. There was also effacement of the fourth ventricle and lateral ventriculomegaly suggestive of early hydrocephalus. A referral was made to The National Hospital for Neurology and Neurosurgery (NHNN). A computed tomography (CT) scan of her thorax, abdomen and pelvis and MRI of her whole spine were both negative for von Hippel-Lindau disease. Subsequently, she was transferred to the NHNN and a posterior fossa craniotomy with midline tumour resection was performed. Histological examination of the resected enhancing nodule showed this lesion to be haemangioblastoma central nervous system World Health Organisation grade I. Surgery was performed successfully with no complications in the post operative period. A baseline post-surgical MRI was satisfactory demonstrating significant reduction in mass effect and decreased ventriculomegaly. She recovered well on the ward and was deemed medically fit for discharge. Three weeks post-surgery she represented to NHNN with fullness over the surgical wound. A CT head scan



revealed a pseudo-meningocele post craniotomy, which resolved upon insertion of a lumbar drain.

**Discussion:** This case report highlighted the unique challenges associated with definitive haemangioblastoma diagnosis and management between both medical and surgical specialties. As well as this, the posterior fossa itself adds to the complexity of management as an anatomical antagonist to the adjacent vital neovascular structures encased within, as seen in this case.

## **22. Enigmatic Proptosis and Palsy**

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The Leeds Teaching Hospitals NHS Trust

**Introduction:** This case aims to highlight the ophthalmic manifestations of IgG4-related disease in a postpartum patient

**Case report:** A 28-year-old, woman 10-days postpartum presented to the emergency eye clinic with sudden onset headache, reduction in visual acuity, and pain around the left eye. Initial examination demonstrated proptosis of the left eye, restriction of the lateral rectus muscle with diplopia on laevoversion and a reduction of visual acuity to 6/24. Due to her current hypercoagulable state following a caesarean section, urgent magnetic resonance imaging (MRI) was arranged. This excluded a cavernous sinus thrombosis but did not identify any particular pathology. As such the differential diagnosis remained broad including infectious, inflammatory and malignant causes. Investigations did not identify any clues except for a positive QuantiFERON test. Further MRI later revealed pachymeningeal thickening of the sphenoid wing, as well as enhancement in the superior orbital fissure, cavernous sinus, foramen rotundum and left pterygoplatatine fossa. Sequential imaging, lumbar puncture and discussion with the tuberculosis team, were not suggestive of an acute or latent tuberculosis infection. In collaboration with the wider team it was suggested that this may be IgG4-related disease. Once conclusively excluding any infectious causes she was started on high-dose corticosteroids. Serum diagnostic investigations for all inflammatory causes, including Immunoglobulins were all within the normal range. Over the next two months, steroid treatment led to an improvement in headache, visual acuity and pain but the proptosis and muscle paresis remained unchanged. The presumed diagnosis remains IgG4-related disease due to its clinical improvement on steroids and features upon presentation. Ongoing specialist input may shed further insight into the course of this complex presentation.

**Discussion:** IgG4-related diseases are rare, enigmatic and often a diagnosis of exclusion. IgG4-related diseases appear to have an association with pregnancy, but less frequently present in the postpartum period. With limited research on postpartum presentations of IgG4-related ophthalmic disease, the prognosis remains uncertain. This case highlights the need to consider systemic diseases in atypical presentations of diplopia, proptosis and nerve palsies.

### **23. Neuro-Ophthalmology in Eye Casualty – A Service Evaluation at Southampton General Hospital**

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2. Southampton General Hospital, Southampton

**Introduction:** Neuro-ophthalmological presentations span a variety of conditions in both paediatric and adult populations and account for a significant number of attendances at eye casualty. Given their complexity and diversity, they often require multiple attendances and follow-up in sub-specialist clinics. Here we aim to determine the burden of Neuro-ophthalmological conditions at Southampton General Hospital eye casualty and the appropriateness of referrals and follow-up.

**Methods:** We performed a retrospective review of all Neuro-ophthalmology cases presenting to eye casualty at Southampton General Hospital between August 2014 and August 2017. Attendances were categorised into 27 core Neuro-ophthalmological conditions and others were manually assigned a referral reason. Attendances were analysed by their demographics, triage time, appropriateness of investigations and follow ups.

**Results:** There were 3174 attendances for the core Neuro-ophthalmological conditions representing 2558 new presentations. Additionally, there were 828 attendances for other suspected Neuro-ophthalmological conditions with the most common referral reasons including new visual disturbance (54%), diplopia (18%) and field assessment (13%). Patients had an average age of 48 years. The majority (55.3%) attended eye casualty through referral (GP, optician, etc.), whilst the remaining were walk-in patients. 14% of total patients were current inpatients. The most common grouped presentations were headaches/migraines (49.8%), nerve palsies (13.1%) and optic neuropathies (11.8%). The mean number of follow ups was 1.3 per discrete presentation. Average triage time for Neuro-ophthalmological emergencies was 16 minutes. Average time spent in eye casualty across all conditions was 2 hours 19 minutes. Only 8.3% of patients had inappropriate investigations and 2.3% had inappropriate onwards referrals.

**Discussion:** There is a known national growing demand for eye casualty services with attendances increasing every year. Our study has confirmed the high presence of Neuro-ophthalmological presentations in eye casualty with almost 4000 attendances in a 3-year period. The majority of these were referred from colleagues. Patients were triaged and seen rapidly especially with Neuro-ophthalmological emergencies and turnaround from eye casualty was efficient at less than 2.5 hours. This is significantly less than the 4-hour target aimed by general A&E services. Additionally, the majority of patients were discharged from eye casualty either by onward referral or home within a single visit. Overall, there is evidently a large burden placed on eye casualty services at Southampton General Hospital by Neuro-ophthalmological presentations, which appears only to increase. However, local ophthalmologists are efficient in seeing and treating cases with few inappropriate investigations and referrals.

#### **24. Measuring Visual and Quality of Life Outcomes in Nystagmus: A Scoping Review**

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**Introduction:** Pathological nystagmus of all aetiologies can cause visual disability and reduced quality of life. Recent research has characterised the dynamic nature of the condition and questioned the validity of widely utilised outcome measures, such as visual acuity, as an accurate measure of the visual function in nystagmus. Visual acuity is simply the spatial resolving capacity of the visual system, whereas good visual function requires numerous other components including contrast sensitivity, colour vision, depth perception, and oculomotor functions. For these reasons, the literature on nystagmus, particularly interventional studies, has been inconsistent with regards to the outcome measures used. Therefore, we aimed to perform a scoping literature review to characterise all published measures and instruments for determining functional vision and quality of life in patients with nystagmus.

**Methods:** We developed a search query utilising relevant keywords for visual and quality of life outcomes after a preliminary data search. Included studies were either interventional studies or measurement studies that measured nystagmus outcomes under differing conditions. Our protocol was developed in line with the Joanna Briggs Institute's manual and was published on the INPLASY platform. We utilised PRISMA-ScR to report our study outcomes.

**Results:** 235 articles were included in the study, accounting for 5880 patients with nystagmus. Articles spanned six continents and eight decades of research.

Articles included 36 subtypes of infantile nystagmus and 31 subtypes of acquired nystagmus. 89 articles were measurement studies and 146 articles were interventional studies. We identified 10 grouped outcome measures including visual acuity (27 outcomes, 56 instruments), nystagmus features (29 outcomes, 3 instruments), visual function (12 outcomes, 34 instruments), ocular morphology (3 outcomes, 5 instruments), orthoptic features (17 outcomes, 46 instruments), reading assessment (7 outcomes, 5 instruments), visual search assessment (6 outcomes, 6 instruments), foveation/function models (8 outcomes), subjective outcomes (30 outcomes) and non-ocular features (6 outcomes).

**Discussion:** We have confirmed there is significant heterogeneity in outcome measures related to visual function and quality of life in the literature on nystagmus. It is evident that one or even a few measures are insufficient to measure all the relevant potential outcomes from interventional studies for people with nystagmus. Therefore, we propose a Delphi project utilising our data to determine a standardised core outcome set for interventional nystagmus studies.

## **25. Compliance with Ophthalmologic Monitoring Standards for Patients on S1PR Modulators in Secondary Progressive MS**

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**Background:** Sphingosine-1-phosphate receptor (S1PR) modulators, including siponimod and ponesimod, are increasingly used to treat secondary progressive multiple sclerosis (SPMS). However, these medications pose a risk of macular oedema, especially around 3-4 months post-treatment initiation. The European Medicines Agency (EMA) advises ophthalmologic evaluations within this timeframe and baseline assessments for patients with pre-existing ocular conditions for that reason.

**Aim:** This audit aimed to assess compliance with EMA guidelines for ophthalmologic monitoring in patients prescribed S1PR modulators for SPMS.

**Methods:** We conducted a retrospective review of patient records from 2021 to 2023, identifying patients commenced on a S1PR modulator. For each patient, we recorded pre-existing ocular conditions, baseline assessments, and timing of follow-up ophthalmologic evaluations.

**Results:** Out of 111 patients analysed, 15% received ophthalmologic assessments within the recommended 3-4 month timeframe, 17% were evaluated slightly outside this period, and 44% were assessed after an extended delay. Additionally, 24% had no documented assessments. Only two of the 12 patients with

significant past ocular histories received baseline evaluations. Following the establishment of a new clinic (ZKOTC) to streamline ophthalmological evaluation in September 2023, assessment times improved, averaging 3.3 months.

**Conclusion:** The audit revealed significant gaps in timely ophthalmologic monitoring for patients on S1PR modulators, particularly in documentation and pre-treatment screening. Initiatives to streamline data entry and improve clinic coordination are anticipated to enhance compliance. One initiative potentially includes a technician-led virtual ophthalmology clinic. Such a model could enhance coordination, address documentation challenges, and facilitate adherence to EMA standards. A re-audit will evaluate the impact of these interventions.

## **26. Bilateral Visual Loss in a Burns ITU Patient**

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**Case report:** A female in her fifties was admitted to Burns ITU following a significant flash burn in her garden due to an ethanol explosion. She was intubated with significant systemic affection. She also developed clonus and systemic infection. No ocular consultation was requested until day 45 post injury when she regained some consciousness and complained of not seeing anything. Ocular examination of the anterior segment was unremarkable. However, posterior segment assessment revealed significant bilateral symmetrical optic disc pallor and retinal haemorrhages involving the posterior pole and mid periphery. Neuroimaging was unremarkable apart from some ischaemic changes. Despite significant improvement of her general condition, her vision remained at no perception of light in both eyes. The differential diagnosis included ischaemic optic neuropathy, infective neuroretinitis and toxic optic neuropathy. Given the history, investigations and the clinical picture, the most probable diagnosis was bilateral ischaemic optic neuropathy (ION). After approval of her Burns ITU team, intravenous methylprednisolone was tried for 3 days with no response. The condition had significant effect on the patient and her family members and the Eye Care Liaison Officer was there for support.

**Discussion:** ION is a tragic condition that can present as postoperative visual loss in different surgical groups. It has also been described in critically ill patients with severe systemic inflammatory response syndrome, breathing support, haemodynamic compromise and hypoperfusion, requiring massive resuscitation. Few case reports describe the association between burns patients and vision loss due to ION. The blood supply of the optic nerve comes mainly from two routes:

the posterior ciliary artery that vascularizes the front part of the nerve, and the pial vessels, collateral of the ophthalmic artery, that vascularize its posterior part. ION is a result of an infarction at the dividing line between the two areas. ION is classified as anterior ION or posterior ION, depending on the location of the ischaemic lesion, in relation to the lamina cribrosa. According to its physiopathology it can be arteritic, or non-arteritic. Various factors may play a role in the development of ION in burns patients, such as aggressive fluid resuscitation, prolonged organ dysfunction, massive transfusion, hypoxia, ischaemia and prolonged prone position. Once developed, prognosis is very poor. Hence, prevention is the key if possible. Emotional and social support for the patient and the family is crucial.

### **27. Role of Pre-operative Retinal Nerve Fibre Layer Thickness Measurement with Optical Coherence Tomography in Predicting Visual Outcome in Untreated Compressive Sellar and Parasellar Lesions: Systematic Review and Meta-analysis**

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**Background:** This review evaluated the current literature detailing the use of preoperative retinal nerve fibre layer (RNFL) thickness, measured via optical coherence tomography (OCT), as a predictive tool for postoperative visual outcome in patients with untreated compressive sellar and parasellar lesions.

**Methods:** We searched Medline, Embase and CENTRAL on 21/07/2022 for observational studies reporting pre-operative RNFL thickness on OCT and post-operative visual outcomes (best corrected visual acuity [BCVA] and visual field mean deviation [VF-MD]) in adult patients (aged  $\geq 18$  years) with untreated compressive sellar or parasellar lesions. The Newcastle-Ottawa Scale informed our risk of bias and subsequent quality assessments. We used linear regression analysis with Pearson's correlation coefficients, including the coefficient of determination ( $R^2$ ), to investigate the relationship between preoperative RNFL thickness and postoperative visual function outcomes (BCVA & VF-MD). Studies with continuous pre- and post-operative data entered a random-effects meta-analysis.

**Results:** There were 27 eligible moderate to high-quality (NOS >6) observational studies identified from 155 records involving 1575 adult patients with compressive sellar or parasellar lesions. Statistically significant linear

relationships between pre-operative RNFL thickness, and post-operative visual outcomes: BCVA ( $p = 0.0030$ ) and VF-MD ( $p = 0.0019$ ) were identified. Less defined, statistically insignificant linear relationships between pre-operative RNFL thickness and the degree of change between pre- and post-operative BCVA ( $p = 0.1998$ ) and pre- and post-operative VF-MD ( $p = 0.8890$ ) were found.

**Discussion:** Pre-operative RNFL measurement on OCT appears to be a statistically significant predictor of post-operative visual outcome (BCVA; VF-MD) following resection of compressive sellar and parasellar lesions. Pre-operative RNFL does not, however appear to be a statistically significant predictor for post-operative visual recovery. There is a need for further studies to gain a better understanding of OCT and post-operative visual outcomes in patients with compressive sellar or parasellar lesions.

## **28. OCTOPIT (Optical Coherence Tomography in Optimising Timing of Intervention for Pituitary Tumours) Pilot Survey**

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**Introduction:** The OCTOPIT (Optical Coherence Tomography in Optimising Timing of Intervention for Pituitary Tumours) pilot survey aims to evaluate how optical coherence tomography (OCT) technology is currently used worldwide for managing asymptomatic non-functioning pituitary adenomas by gathering opinions from healthcare professionals. This survey is the first step towards launching a future, observational, multicentre study with the same name, the OCTOPIT study, which is being organised by the Institute of Neurosciences in Glasgow, in partnership with the University of Glasgow. The goal of the OCTOPIT study is to determine how OCT can be used in clinical settings for managing non-functioning asymptomatic pituitary adenomas and to investigate its potential role in timing interventions and enhancing patient outcomes. Additionally, this survey aims to find clinicians and healthcare trusts interested in joining collaborative research efforts in this field. Additionally, this survey aims to find clinicians and healthcare trusts interested in joining collaborative research efforts in this field.

**Methods/Results:** The survey was distributed to a diverse cohort of clinicians globally, capturing data on familiarity with OCT, its integration into practice and its perceived prognostic utility. A total of 45 respondents, predominantly neurosurgeons (56%), participated, with most holding senior positions as consultants/attending physicians (76%). The majority were affiliated with academic or university hospitals (71%) and practised across varied regions

including the UK/Republic of Ireland (53%), Western Europe, Australia, Sub-Saharan Africa and the Middle East. Key findings reveal a high level of familiarity with OCT (80%), but only 40% of respondents reported routine usage for asymptomatic patients. Amongst those using OCT, 53% indicated that findings influence clinical decision-making. Whilst 75% of participants advocated for routine OCT in preoperative evaluations, there was moderate agreement (average Likert score: 3.5/5) on its prognostic value for visual outcomes post-surgery. Access to OCT technology is available to 81% of respondents, though some rely on referrals to ophthalmology services. Discussion of OCT findings in multidisciplinary team meetings remains infrequent, with an average Likert score of 2.5/5, highlighting a gap in collaborative utilisation.

**Discussion:** This study underscores the need for standardised OCT protocols and enhanced interdisciplinary dialogue to optimise its clinical impact. A significant majority (87%) expressed strong interest in participating in prospective studies to further explore OCT's role in managing pituitary adenomas. These findings lay the groundwork for integrating OCT more comprehensively into routine assessment, informing future research and guideline development.

## **29. Persistent Papilloedema after Venous Sinus Thrombosis**

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**Introduction:** We present two patients with papilloedema secondary to cerebral venous sinus thrombosis (CVST). Despite anticoagulation treatment, resolution of thrombus and evidence of recanalisation, both patients had persistent papilloedema. Further investigations showed presence of a dural AV fistula (dVAF) in each patients as a target for intervention.

**Case 1:** A 34-year-old male presented to the eye clinic with bilateral disc swelling and headache after COVID-19. CVST was found over the right transverse and sigmoid sinus. He was treated with anticoagulants as well as acetazolamide. He continued to have persistent disc swelling and symptomatic headache after 9 months. A repeat venogram showed resolution of the thrombus and recanalisation. Suspicion of a dural AV fistula was raised on an early phase computed tomography (CT) venogram sequence and dedicated angiography demonstrated a dAVF over the right transverse sinus. He underwent radiological embolisation and subsequent follow-up showed significant improvement in papilloedema and headache symptoms.

**Case 2:** A 23-year-old female presented with bilateral disc swelling and headache after COVID-19. CVST was found involving the superior sagittal sinus, left



transverse and sigmoid sinus. She was treated with oral dabigatran and acetazolamide. After 1 year of treatment, repeat venography showed resolution of the thrombus and recanalisation, with improvement of optic disc swelling. However, she had worsening of the disc swelling after tapering her acetazolamide. Angiography was performed and a dAVF was identified at the superior sagittal sinus. She is awaiting intervention.

**Discussion:** In the literature, CVST and dAVF are known associations, but the causal relationship between the two entities is still a subject of debate. It is difficult to determine the initial pathology in our cases, however given the close anatomical relationship they are likely related. Our cases highlight the importance of considering the presence of dAVF in patients with persistent papilloedema after treatment for CVST. The identification of dAVF in both cases was pivotal in guiding further management, emphasizing that venous phase imaging alone may overlook such vascular abnormalities. The clinical improvement after intervention in our case indicate there is a role for embolisation when dAVF is found. Specifying an arterial phase imaging into CT venography allows better visualization of abnormal arteriovenous connections, which may otherwise be missed. Clinicians managing patients with CVST and persistent optic disc swelling should remain vigilant for dAVF and consider tailored imaging protocols, as timely diagnosis can significantly impact outcomes.

### **30. A Retrospective Audit of the Presentation and Initial Management of Anti-MOG Seropositive Optic Neuritis within University Hospitals Sussex**

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**Introduction:** An immune response to myelin oligodendrocyte glycoprotein (MOG) that triggered complement-dependent demyelinating activity was first identified in animal models in 1976. It was almost 40 years later that anti-MOG antibodies were confirmed to be associated with aquaporin-4 negative bilateral optic neuritis (ON). When presenting in adults, MOG antibody disease (MOGAD) often causes ON. Close scrutiny of clinical features and neuroimaging can often enable early distinction from more prevalent multiple sclerosis-related (or idiopathic demyelinating) ON. This distinction is important as high-dose methylprednisolone may be required to prevent long-term visual loss in MOGAD. Despite this, some cases go unrecognised at onset or treatment delayed. In this audit, we analysed sequential cases presenting to UH Sussex over an 11-year period to assess if diagnosis and treatment were provided in a timely manner.

**Methods:** This audit retrospectively reviewed 16 patients with MOGAD associated ON at UH Sussex between January 2012 and January 2023. Data collected included visual acuity, symptom laterality, presence and extent of optic disc swelling, pain, patient demographics (age, sex, ethnicity), autoimmune history, initial steroid administration, and whether MOGAD was considered as a diagnosis at presentation.

**Results:** Current observations from the initial data collected during this audit include: 62.5% patients were female, with mean age 42 years (range 18-71) at presentation; recorded visual acuities of affected eyes ranged from light perception to 6/9; 68.75% of patients had a unilateral ON initially, of whom two were bilateral-sequential; 31.25% had bilateral ON at onset; 62.5% had pain on ocular movements; and 68.75% of patients were given steroids at first presentation. Factors that may have influenced choice not to give steroids in the remaining patients included age and atypical features of presentation.

**Discussion:** Early findings suggest that the demographics of MOGAD patients at UH Sussex are broadly similar to the wider MOGAD population. Ongoing education is required to ensure that MOGAD cases are identified and treated early. Factors such as older age at presentation may be associated with delay in appropriate management.

### **31. A Rare Cause of Bilateral Optic Neuropathy in a Young Adult**

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1. St George's University Hospitals NHS Foundation Trust, London

2. Croydon University Hospital, Croydon

**Case report:** A 22-year-old woman presented with an 8-year history of bilateral worsening painless blurred vision and a sensation of her “eyes shaking” when trying to focus. She had a background of type 2 diabetes mellitus and anxiety. On examination she had reduced visual acuities of 6/36 in both eyes. Colour vision was absent bilaterally, gross bilateral optic disc pallor and bilateral pendular, presumed congenital nystagmus (present in primary position and mainly on horizontal gaze) were seen. Visual fields were abnormal with peripheral constriction on the right and reduced in the left temporal superior field. The remainder of the neurological examination was normal. Her body mass index was normal. Routine blood tests including B12, folate, aquaporin 4 antibody, MOG antibody, and thyroid function were all normal. Her HbA1C was 94. Non-contrast magnetic resonance imaging (MRI) of her brain and orbits was reported as normal. Visual-evoked potentials showed majorly symmetrically prolonged P100 latencies of 137ms on the right and 140ms on the left. Optical coherence

tomography reported bilateral significant optic pallor with reduction of ganglion cell layer. There was no evidence of diabetic retinopathy or macular oedema. A further MRI of her brain and orbits revealed small optic nerves and chiasm. A more detailed history revealed her mother also had diabetes mellitus and two of her maternal 1st cousins had severe congenital deafness. Genetic testing (R41 panel) was positive for autosomal recessive Wolfram syndrome 1 (compound heterozygous for a pathogenic variant and a likely pathogenic variant in WFS1). She is currently registered severely sight impaired and is being followed up by Neurology, Diabetology, Ophthalmology, and Clinical Genetics.

**Discussion:** Wolfram syndrome is also called DIDMOAD (diabetes insipidus, diabetes mellitus, optic Atrophy and deafness). It is an extremely rare neurodegenerative disorder with a prevalence of about 1/500,000. Wolfram syndrome 1 involves a mutation in the WFS1 gene on chromosome 4p16. Classical features include diabetes mellitus, central diabetes insipidus, optic atrophy and sensorineural hearing loss. Less commonly, patients can also develop urinary tract problems, psychiatric symptoms, neurological and autonomic disorders. Currently, management is focused on symptom control only with no definitive treatments. The prognosis is reported to be appalling with a median age of death of 30-years-old due to respiratory failure from brainstem atrophy.

### **32. “I’m Seeing Witches”: An Unusual Presentation of Epilepsy**

Akash Dharni, Joanna M Jefferis, Simon J Hickman  
Sheffield Teaching Hospitals NHS Foundation Trust

**Introduction:** We present the case of a 77-year-old woman with interesting octal visual and auditory hallucinations, secondary to temporal oedema from stereotactic radiosurgery (SRS) 2 years previously.

**Case report:** A 77-year-old woman was referred to the Neuro-Ophthalmology clinic due to hallucinations. She described seeing specific images of cartoon witches flying through the air, likening them to those seen in children's books, as well as letters on car licence plates moving around. She would see these images frequently during waking hours, particularly outside, and they were described as a nuisance. On occasion, she would tell the images to ‘go away’, knowing they were not real. The visual symptoms were accompanied by a rising abdominal discomfort, as well as auditory phenomena of hearing children’s nursery rhymes, but she experienced no change in conscious level during the episodes. She had attributed the hallucinations to starting ramipril and, as a result, her hypertensive medications had been changed 2 months prior to the Neuro-ophthalmological review, but with no change to her symptoms. She had history of a temporal lobe

meningioma, which had been treated with SRS 2 years previously. A few months prior to her presentation she had been admitted due to a headache. Magnetic resonance imaging showed temporal lobe oedema, thought to be secondary to the SRS. Her visual examination was normal, including normal visual fields in each eye. Temporal lobe epilepsy was suspected. A routine, awake, inter-ictal electroencephalogram (EEG) recording was performed, but was within normal limits. Following the EEG, a routine follow up was organised. In the interim period, she suffered a tonic clonic seizure requiring hospital admission and was treated with 50mg lamotrigine twice per day. After starting this she has had no further hallucination episodes and no further seizures.

**Discussion:** Complex visual hallucinations are rare in epilepsy. Epileptic visual hallucinations usually arise from the occipital lobes and are typically described as coloured blobs. When complex hallucinations have been reported the lesions have been temporal or temporo-occipital. Auditory hallucinations in epilepsy have been reported to arise from the temporal lobe. Our case is unusual in having the combination of both complex visual and auditory hallucinations.

### **33. It's All in the History**

Katie Parr, Katherine Smyth  
Royal Bolton Hospital, Bolton

**Case report:** We present a female, aged 59 years, referred with post-concussion syndrome following a minor head injury. She had identified struggles at work in her administrative role, making mistakes with invoices and payments. She underwent dementia testing, with no issues found. Previous eye examinations were unremarkable. She was insightful and able to describe the issues she experienced: problems reading and writing; as well as navigating kerbs. She struggled with contrast and experienced increasing visual difficulties in the dark. One of her biggest struggles was with shopping, particularly in the supermarket. Her husband, via telephone, identified further issues, including difficulties with object orientation and facial recognition (including her own). She attended clinic alone, navigating to the hospital independently and without issue. Her conversation and affect appeared normal. She was well kempt and interpersonally appropriate. Her Snellen visual acuities were 6/24 right and 6/36 left, but were 6/9 in each eye with uncrowded testing. Ocular examination, including confrontational visual fields appeared unremarkable. She had an intermittent near exotropia with diplopia, preferring the use of occlusion for near tasks. She was orientated in person and place, identified the day of the week and month correctly, but gave the wrong year, although identified this mistake

herself. She knew the prime minister and the monarch – notable after recent changes in these roles. She struggled to write her name, produce a drawing of a clock face and a simple line drawing. She had difficulty in locating a pen in a bag of equipment and could not describe the scene in the Boston Cookie Theft Picture. Neuroimaging was undertaken, which showed significant atrophic changes affecting the parietal lobes. Following referral for neuropsychological assessment, they found profound visual perceptual impairment, with notable impairment of visual construction. They did identify significant cognitive impairment, but felt that this was largely driven by her visual perceptual difficulties and they supported the diagnosis of posterior cortical atrophy as the underlying diagnosis.

**Discussion:** This patient demonstrates the need for careful assessment and thorough history when symptoms and signs do not correlate. Relative interview can be a valuable tool. With her normal affect and interaction, cognitive evaluation could easily have been over-looked as this appeared superficially unremarkable. We recommend that when visual problems are unexplained cognitive assessment can be useful in reaching a diagnosis and suggest a series of assessments that can be useful in the Ophthalmology clinic.

#### **34. High Pressure, High Stakes**

Nimesha Elizabeth Alex, Brinda Muthusamy, Aditi Vedi  
Cambridge University Hospital/University of Cambridge Department of  
Paediatrics, Cambridge

**Case report:** A 5-year-old boy with left adrenal high-risk neuroblastoma and liver metastases presented with bilateral optic nerve swelling during a routine baseline ophthalmic evaluation following 14 days of liquid isotretinoin pre-treatment as part of dinutuximab-based maintenance therapy. He had previously received intensive chemotherapy, surgery autologous stem cell transplant, and abdominal radiotherapy. He was asymptomatic of headaches or visual symptoms, but ophthalmic examination identified Frisè grade 2 papilloedema. Differential diagnoses included retinoic acid-induced idiopathic intracranial hypertension (RA-IIH), cerebral venous sinus thrombosis (CVST), relapsed neuroblastoma with central nervous system involvement, and optic disc drusen (ruled out with optic disc autofluorescence). Neuroimaging excluded relapsed neuroblastoma. A computed tomography scan confirmed bilateral optic nerve prominence, and magnetic resonance venography excluded CVST but revealed a small pituitary gland, indicative of elevated intracranial pressure. A lumbar puncture further confirmed IIH with an opening pressure of 35cm cerebrospinal fluid (CSF) and

normal CSF cytology. Recognising the essential role of isotretinoin in neuroblastoma treatment, the liquid formulation was switched to oral tablets, anecdotally associated with a reduced risk of intracranial pressure elevation. He was also started on acetazolamide to manage intracranial hypertension. He retains good optic nerve function and his papilloedema has responded to medical management. Regular monitoring of optic nerve function and disc swelling was scheduled monthly to ensure visual preservation. This case underscores the importance of balancing effective cancer therapy with the prevention of visual morbidity through proactive multidisciplinary management.

**Discussion:** Neuroblastoma is the most common extracranial solid tumour in children, 50% originating from the adrenal glands and the remaining anywhere along the sympathetic chain. Almost half of all patients with neuroblastoma have high-risk disease, carrying a poor overall prognosis, requiring multimodal treatment including chemotherapy, surgery, myeloablative therapy with autologous stem cell rescue, radiation, and immunotherapy. Treatment comprises of 14 days of oral isotretinoin followed by 10 days dinutuximab-beta for 5 cycles. Although overall well tolerated, isotretinoin carries rare but serious toxicity risks, including RA-IIIH. The challenge lies in balancing the benefits of isotretinoin with the risks of severe side effects, requiring careful, individualised management. In cases of visual deterioration, increasing oral acetazolamide or performing therapeutic lumbar punctures may be necessary to control intracranial pressure. In situations where life- and sight-threatening complications arise, prioritising life preservation is critical. This case emphasises the importance of routine and continuous monitoring and interdisciplinary collaboration to ensure effective oncologic treatment particularly in vulnerable paediatric patients.

### **35. Invasion Picking Off Nerve After Nerve**

Linda Lei, Johnson Ja, Farrah Jabeen

Neurology Department, Royal Free Hospital, London

**Case report:** The case is of a 74-year-old man who presented to A&E with a 1-week history of a central frontal headache and blurry vision. He had a background of ischaemic heart disease, end stage renal failure on thrice weekly dialysis due to IgA nephropathy (awaiting renal transplantation), hypertension, gout, glaucoma and type two diabetes mellitus. He had recently had a fall at a bus stop due to poor vision. When he initially presented in October 2024 he had an isolated left 6th nerve palsy with some confluent neck swelling on the right. This progressed rapidly over the next few weeks with the development of left 3rd and 4th nerve

palsies with ptosis, as well as a right vagal palsy with deviation of the soft palate and uvula. He went on to have magnetic resonance imaging (MRI) of the skull base which showed a neoplastic lesion, possibly arising from the clivus or the parapharyngeal soft tissues with invasion of the skull base and sphenoid sinus. The radiology highlighted the almost complete infiltration of the clivus. MRI of the neck revealed multiple large lymph nodes with suspicion of a mass encroaching into the inferior right carotid canal. He had an urgent biopsy under general anaesthetic of the postnasal space mass and ultrasound guided biopsy of the neck nodes under Ear Nose and Throat and is awaiting histological confirmation currently.

**Discussion:** The adage holds true in this case whereby ‘the brain sits on the nose’ and this case highlights the potential anatomical antagonists in the vicinity of the eye. The invasion of the clivus by a neoplastic process specifically highlights the course of the 6th cranial nerve. The progression of the neoplastic process into the sphenoid sinus and skull base explains the progressive involvement of the other cranial nerves. This case highlights the anatomical course of the vagus nerve passing through the carotid sheath with a vagal nerve palsy. The involvement and progression of further cranial nerves should prompt investigations for a neoplastic process that may well be a primary nasopharyngeal cancer.

### **36. Porous Walls and a 3rd Nerve Palsy**

Linda Lei, Johnson Ja, Farrah Jabeen

Neurology Department, Royal Free Hospital, London

**Case report:** An 82-year-old woman presented to hospital on the 23rd of May 2023 with a right parietal bleed in the context of rivaroxaban for known atrial fibrillation. On the 8th June she re-presented with a 3 day history of a headache, a fever of 38.9°C and raised inflammatory markers (C reactive protein [CRP] 34, white cell count 11). A head computed tomography (CT) scan showed fluid in the right sphenoid sinus with resolving appearances of the right parietal bleed. She was hypotensive and had a CRP the next day of 307. Her blood cultures grew *Streptococcus constellatus* and she was empirically started on co-amoxiclav for 2 days before being switched to ceftriaxone and then to metronidazole for 9 days. The headaches gradually worsened and became more localised to the right retro-orbital region. On the 13th June she developed blurred vision and she had a documented normal examination by Ophthalmology on the 15th June. On the 19th June she developed horizontal diplopia with a right sided ptosis. She was reviewed by the Neurology team the next day and was found to have a right-sided 3rd nerve palsy, which gradually worsened over the next few days on repeat

assessments. She had repeat CT head scanning on the 20th June, which revealed a completely opacified sphenoid sinus with partial opacification of the ethmoid sinus. Magnetic resonance imaging 2 days later revealed a further suggestion of inflammation involving the walls of the cavernous sinus. Her antibiotics were escalated to dual metronidazole and high dose ceftriaxone for 2 weeks. Her 3rd nerve palsy and headaches gradually completely resolved with antibiotics within 2 weeks.

**Discussion:** Even a localised sphenoid infection can affect the nearby cranial nerves as the wall is porous. Close team-working with microbiology, Ear Nose and Throat (ENT) and Ophthalmology is important to ensure the globe of the eye and optic nerve function is remaining intact whilst the sinusitis is treated with antibiotics. Visual acuity needs to be monitored regularly as the optic nerve function is an indication of progression of sinusitis. Early and appropriate antibiotic treatment is important as draining of any resultant collection is considered by ENT to be a last resort given the proximity to many other structures including other cranial nerves in the vicinity. *Streptococcus constellatus* can behave aggressively causing severe sinusitis. Of note, this bacterium requires early and aggressive treatment as there are reports of this pathogen causing progression and spread that leads to brain abscesses.

### **37. The Eyes as the Clue to the Diagnosis of a Systemic Disease**

Johnson Ja, Paul Watts

Royal Free Hospital, London

**Case report:** A 76-year-old woman presented with a 5-month history of unsteadiness of gait with paraesthesias in her hands and feet. She also had 12 kilograms of unexpected weight loss over the preceding 12 months. This occurred on a background of a 50 pack-year smoking history. Her past medical history was significant for hypertension, Grave's disease, thyroid eye disease and hypothyroidism. Her medications were ramipril and thyroxine. There was no significant family history. On examination she had exophthalmos with saccadic intrusions on smooth pursuit and hypometric saccades. There was downbeating nystagmus in primary position, which was more prominent on downward and lateral gaze. She had impaired suppression of her vestibular-ocular reflex. Her cranial nerves were otherwise normal. In her upper limbs she had clubbing of her fingers and there was pseudoathetosis of the outstretched fingers. Tone and power were normal. There were reduced reflexes in the upper limbs with finger-nose ataxia. Her joint-position and vibration sense were, surprisingly, normal. In the lower limbs reflexes were reduced with heel-shin ataxia. Her joint-position



and vibration were once again normal. She had an ataxic gait. She was investigated extensively. Her nerve conduction studies confirmed an axonal sensory neuropathy. Magnetic resonance imaging of her brain showed an incidental left frontal meningioma and moderate small vessel disease. Her lumbar puncture was inflammatory with a protein count of 2.53 and 3 white cells. Cerebrospinal fluid (CSF) cytology and flow cytometry were negative. A computed tomography scan of her chest/abdomen/pelvis revealed right-sided paratracheal and hilar lymphadenopathy, which was FDG-avid on positron emission tomography. She underwent an endobronchial ultrasound scan and biopsy and histology confirmed small cell lung cancer. Her serum and CSF antineuronal antibodies came back positive for anti-Hu. Her final diagnosis was small cell lung cancer with anti-Hu antibodies causing a paraneoplastic neurological syndrome manifesting as a sensory neuropathy and cerebellar eye signs. This is an interesting case of a patient with a paraneoplastic syndrome with cerebellar eye signs. A video will be provided and a discussion of paraneoplastic neurological syndromes and cerebellar eye signs will follow.

### **38. Evaluating the Effectiveness of a Joint Consultant-led Neuro-ophthalmology Clinic Service in a District General Hospital**

Hussein Al-Fazly<sup>1</sup>, Dr Zhaleh Khaleeli<sup>1,2</sup>, Dr Marcela Bohn de A Alves<sup>1,3</sup>

1. Watford General Hospital, West Hertfordshire NHS Trust, Watford
2. National Hospital for Neurology and Neurosurgery, London
3. Moorfields Eye Hospital NHS Foundation Trust, London

**Introduction:** The Neuro-ophthalmology clinic at West Hertfordshire NHS Trust was set up as a joint consultant-led clinic, with input from both an ophthalmology and a neurology consultant. It receives referrals from both the Ophthalmology and Neurology departments within the trust. We sought to evaluate the service as part of a quality improvement project.

**Methods:** We retrospectively reviewed all Neuro-ophthalmology appointments within a 1-year period from October 2023 to September 2024. Did not attend were excluded from the analysis. For each patient we looked at the dates when they were first seen by Ophthalmology/Neurology, the number of times they were seen in each department prior to their Neuro-ophthalmology appointment, their diagnosis pre and post Neuro-ophthalmology review, and Neuro-ophthalmology clinic outcome (discharge/follow up).

**Results:** 35 unique patients were identified (out of 39 appointments in total). Most referrals came from the Ophthalmology department, with eight patients seen by both Neurology and Ophthalmology, and three patients seen by

Neurology only. Patients were seen three times on average prior to their neuro-ophthalmology appointment (mean = 2.9, median = 3). Their Neuro-ophthalmology clinic appointment was on average 8 months after their initial review by Ophthalmology/Neurology.

**Diagnoses:** 14 patients had the same diagnosis as the pre-referral diagnosis. Three patients had a new diagnosis not mentioned prior to their Neuro-ophthalmology review, and three patients had a different diagnosis compared with their initial referral. For seven patients the queried diagnosis/differential in the referral was excluded by the Neuro-ophthalmology review. Four patients did not have a diagnosis after their first Neuro-ophthalmology clinic and were pending further investigations. Just over half of patients (n=18) were discharged after their first Neuro-ophthalmology appointment, one third (n=11) were booked for Neuro-ophthalmology follow up and a few (n=5) were booked for Ophthalmology follow up only.

**Discussion:** The Neuro-ophthalmology joint clinic is effective at providing a final diagnosis in cases of diagnostic uncertainty for patients who have been seen multiple times by Ophthalmology/Neurology.

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