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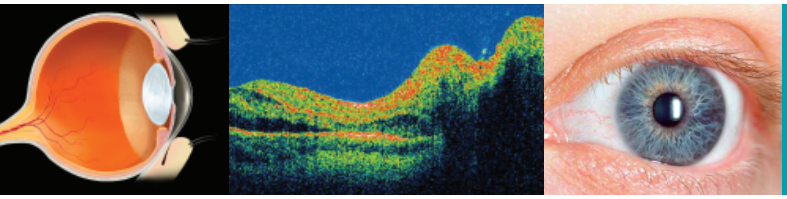
NEURO-OPHTHALMOLOGY



An International Journal of Neuro-Ophthalmology

Abstracts of the European Neuro-Ophthalmological Society (EUNOS) 15th Meeting

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ABSTRACTS

15th EUNOS Congress

BIRMINGHAM, UNITED KINGDOM

Editors' Introduction

Dear Delegates,

The first issue of the journal, *Neuro-Ophthalmology*, appeared in 1980. The founding Editor was A. Huber. The first issue of *Neuro-Ophthalmology* contained materials from the Third Meeting of the International Neuro-Ophthalmology Society (INOS) Meeting held in Switzerland. Sadly, INOS ceased to exist due to financial reasons. The last INOS meeting occurred in Singapore in 2012.

Fortunately, the European Neuro-Ophthalmology Society (EUNOS) is flourishing. The first meeting of EUNOS occurred in Zurich in 1993 and was held every other year up to 2019. Because of the Covid-19 pandemic, this present meeting in Birmingham, UK was delayed a year to 2022. Since the initial EUNOS meeting, the journal *Neuro-Ophthalmology* has been associated with EUNOS and we are delighted to be publishing the abstracts of this year's meeting.

The officers and organising committee have put together an outstanding programme for Birmingham. We congratulate them and look forward to a successful meeting.

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Editors-in-Chief
Neuro-Ophthalmology

EUNOS President's Introduction

Dear Colleagues,

Since the last meeting of EUNOS in 2019 we have all had to endure the profound effects, both to our personal and to our professional lives, of the coronavirus pandemic. As with so many other necessary adjustments to the way we work, we took the decision to defer our 15th biennial conference to this year. I am delighted that as a result of the success of the vaccine roll-out and infection control measures across many countries of the world we are now able to meet again in person. Tim Matthews and his colleagues in Birmingham have put together a fantastic programme which will showcase the scientific and clinical progress that continues to be made under challenging circumstances in our area of clinical practice. For many of us it will be the first major conference we are attending 'face-to-face' since the start of the pandemic, and I hope it will also be an opportunity to renew friendships and collaborations between colleagues from across Europe and worldwide.

Fion Bremner
President, EUNOS

Chair of the Organising Committee's Introduction: Welcome to Birmingham

Dear Colleagues,

It is with great pleasure (and some sense of relief) that I write to welcome you to the postponed 15th meeting of the European Neuro-ophthalmology Society (EUNOS) in Birmingham. As many of you will know, Birmingham is England's second city with a rich heritage including being at the heart of the industrial revolution. Later this summer, Birmingham will also host the XXII Commonwealth Games bringing together athletes from across the Commonwealth.

We are holding the conference at the centre of Birmingham University in their newly opened Elgar Concert Hall in the Bramall Music Building and the closely adjacent Great Hall. The University was founded by Joseph Chamberlain and granted a royal charter by Queen Victoria in 1900. At the time it was the fifth independent University in England (preceded by Oxford, Cambridge, London, and Durham). It is the original "red-brick" university. Those with an interest in art, should take time to visit the Barber institute of Fine Arts (on campus) which this summer will have several visiting masterpieces in addition to their own collection, causing it to be known as "one of the finest small art galleries in Europe".

The organising committee is very proud of the programme that we have assembled for you. Returning to the historic format of EUNOS, there will be a teaching day followed by 3 days of scientific symposia and clinical cases. As we are a forward-looking group, we have peered into the future in some of the sessions, to establish how we will be managing our patients in years to come.

COVID has played its role in the organisation of this meeting, and it will feature as a topic in at least two of the sessions. We have introduced a President's session this year and our President has chosen to discuss how COVID has impacted on our clinical practice. We will also be looking at how meaningful research during this time has impacted on the pandemic itself. We have arranged some exciting debates to stimulate and entertain you. We will also be exploring the role of Neuro-ophthalmology within the multidisciplinary team in various settings. As no trip to Birmingham would be complete without a round-up of the latest research in idiopathic intracranial hypertension, we will not let you slip away unsatisfied. Finally, we have had hundreds of abstracts submitted which the abstract committee scrutinised and have selected the best for platform and poster presentations.

While the committee all look forward to seeing as many of you in person as the current travel restrictions will allow, for those of you who are not able to travel, there will be full access to the scientific programme through our interactive app. For those who can attend, we have an exciting social programme to compliment the scientific programme and we are actively waiting to welcome you to Birmingham.

Tim Matthews
EUNOS 2022 Conference Chair

On behalf of EUNOS we would like to acknowledge the following people for taking time to assess the abstracts for the 15th EUNOS Congress:

Mr Ahmed Abel-Hay; Dr Luke Bennetto; Mr Fion Bremner; Mr Richard Blanch; Dr Sarah Cooper; Mr Michael Gilhooley; Dr Jenny Hepschke; Dr Simon Hickman; Dr Walter Jay; Miss Neringa Jurke; Dr Áine Ní Mhéalóid; Dr Hannah Lyons; Mr Ajay Patel; Dr Danilo Paulo; Dr Matidle Sassini; Dr Mark Thaller; Dr Francis Uchenko; Mr Jasvir Virdee; and Dr Andreas Yiangou.

Abstracts have been categorised under the following sections:

(1) Disorders of the Anterior Visual Pathway (Retina, Optic Nerve, and Chiasm)
(2) Disorders of the Posterior Visual Pathway and Visual Processing
(3) Idiopathic Intracranial Hypertension
(4) Neuro-Ophthalmological Disorders of Neurological and Systemic Diseases
(5) Neuro-Imaging
(6) Ocular-Imaging
(7) Ocular Motility Disorders and Nystagmus
(8) Orbital and Eyelid Disorders
(9) Pupils
(10) SARS-COV-2 Infection Related Neuro-ophthalmic Associations
(11) Miscellaneous

The committee were impressed with the quality of the abstracts and we would like to thank all the authors for their submissions.

Susan P Mollan
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EUNOS abstract coordinator

Monday 20 June 2022

09:00 Session 1: My Approach to		
Chairs: Timothy Matthews, Ahmed Abel-Hay		
09:00–09:30	Acute optic neuropathies	Ruchika Batra
09:30–10:00	Optic disc swelling	Grant Liu
10:00–10:30	Unequal pupils	Fion Bremner
10:30–10:45	Questions	
10:45	*Refreshments and posters*	

11:15 Session 2: My Approach to		
Chairs: Ruchika Batra, Mitch Strominger		
11:15–11:45	Nystagmus	James Benzimra
11:45–12:15	Double vision	Prem Subramanian
12:15–12:45	Unexplained visual loss	Ben Burton
12:45–13:00	Questions	
13:00	*Lunch and posters*	

14:00 Session 3: Investigations		
Chairs: Susan Mollan, Koni Webber		
14:00–14:30	MRI	Jonathan Micieli
14:30–15:00	OCT	Richard Blanch
15:00–15:30	EDTs	Aki Liasis
15:30–15:45	Questions	
15:45	*Refreshments and posters*	

16:00 Session 4: Cases		
Chairs: Michael Burdon, Hadas Stiebel-Kalish		
16:00–16:20	"A straightforward diagnosis at last"	Margaret Dayan
16:20–16:40	"The case of the black diamond"	Clare Fraser
16:40–17:00	"Hang in there"	Dan Milea
17:00–17:20	"Out back"	Aki Kawasaki
17:20–17:40	" . . . ."	Peter Quiros
17:40–18:00	Questions	

Tuesday 21 June 2022

09:00 President's Session

Chairs: Fion Bremner, Tim Matthews

09:00–09:05	Welcome to EUNOS 2022	Fion Bremner
09:05–09:20	Overview of the COVID-19 pandemic	Emma Wall
09:20–09:35	Ophthalmic manifestations of COVID-19/vaccination	Carlos Pavesio
09:35–09:50	Neurological manifestations of COVID-19/vaccination	Hadi Manji
09:50–10:05	Optic neuritis following COVID-19/vaccination	Axel Petzold
10:05–10:20	Haematological manifestations of COVID-19/vaccination	Marie Scully
10:20–10:45	Discussion	
10:45	*Refreshments and posters*	

11:15 Symposium: Idiopathic Intracranial Hypertension

Chairs: Steffen Hamann, Clare Fraser, Susan Mollan, Peter Quiros

11:15–11:40	Piecing the jigsaw together – new insights to IIH pathophysiology	Alex Sinclair
11:40–12:00	Techniques for non-invasive surrogate measurement of intracranial pressure	Steffen Hamann
12:00–12:20	How do you tell your patients to lose weight?	Sally Abbott
12:20–12:40	Why is it important to investigate and manage obstructive sleep apnoea in papilloedema?	Clare Fraser
12:40–13:00	Future treatment for IIH	Susan Mollan
13:00	*Lunch and posters*	

14:00 Free Papers Session 1

Chairs: Nancy Newman, Pinar Aydın O'Dwyer, Dan Milea

14:00–14:15	T01	The phase III REFLECT trial: Efficacy of bilateral gene therapy for Leber hereditary optic neuropathy (LHON) is maintained 2 years post administration	Patrick Yu Wai Man
14:15–14:30	T02	Neuro-ophthalmological characterisation of not-OPA1 dominant optic atrophy	Giulia Amore
14:30–14:45	T03	IIH Pressure: A randomised controlled trial of the GLP-1 receptor agonist exenatide in idiopathic intracranial hypertension	James Mitchell
14:45–15:00	T04	IIH Pressure Med: A randomised, sequential, trial of the effect on cognition of five drugs commonly used in idiopathic intracranial hypertension	Alex Sinclair
15:00–15:15	T05	Utilising optical coherence tomography measures as surrogates for raised intracranial pressure in idiopathic intracranial hypertension	Andreas Yiangou
15:15–15:30	T06	Segregation of decussating axons within the core of the primate optic chiasm shown by injection of a different tracer into each eye	Jonathan Horton
15:30	*Refreshments and posters*		

16:00 Symposium: Pituitary Multidisciplinary Team

Chairs: Ruchika Batra, Niki Karavitaki, Gordon Plant
MDT Panel: John Ayuk, Swarupsinh Chavda, Shazada Ahmed, Ute Pohl

16:00–16:20	Case 1	Naz Raoof
16:20–16:45	Hypophysitis; an overview	Gordon Plant
16:45–17:05	Case 2	Niki Karavitaki
17:05–17:15	Should pituitary apoplexy be managed surgically? For	Shazada Ahmed
17:15–17:25	Should pituitary apoplexy be managed surgically? Against	Tim Matthews
17:25–17:30	Summary	Ruchika Batra

Wednesday 22 June 2022

Free Papers Session 2

Chairs: Simon Hickman, Aki Kawasaki, Ajay Patel

09:00–09:15		Update on *Neuro-Ophthalmology* journal	Simon Hickman
09:15–09:30	T07	Deep learning can accurately distinguish between true papilloedema and optic disc drusen on retinal photographs	Dan Milea
09:30–09:45	T08	Opiate and headache prescribing patterns in women with idiopathic intracranial hypertension: A matched controlled retrospective cohort study	Susan Mollan
09:45–10:00	T09	Retinal fractal dimension in dementia: The ALZEYE study	Siegfried Wagner
10:00–10:15	T10	Association of retinal fractal dimension with incident cardiovascular events in a hospital-attending population in London, United Kingdom	Robert Struyven
10:15–10:30	T11	Increased risk of cerebrovascular disease among patients with non-arteritic anterior ischaemic optic neuropathy – a retrospective nationwide cohort study	Niv Levy
10:30–10:45	T12	Do retinal microvascular changes improve transient ischaemic attack (TIA) risk stratification scores? ABCD2 vs. ABCDEYE in the FOTO-TIA study	Valérie Biousse
10:45		*Refreshments and posters*	

Debates

Chairs: Andrew Jacks, Grant Liu, Nagini Sarvananthan

11:15	Can we treat nystagmus? – For	Steve Madill
	Can we treat nystagmus? – Against	Prem Subramanian
	Summary	
12:00	Can we treat NA-ION? For	Neil Miller
	Can we treat NA-ION? Against	François-Xavier Borruat
	Summary	Anthony Arnold
13:00	*Lunch and posters*	

14:00 Symposium: Meaningful Research in the COVID Era

Chairs: Richard Blanch, Alistair Denniston

Covid Research

14:00–14:02	Introduction	Alastair Denniston
14:02–14:22	COVID epidemiology and public health	Nagpal Hoysal
14:22–14:42	Research Informed Practice – How studies have changed the way we manage COVID-19	Liz Sapey
14:42 – 15:00	Development of COVID-19 vaccinations	Chris Green
15:00	Closing Remarks	Richard Blanch

Inherited Optic Neuropathies

15:00–15:15	Future treatment for Leber's hereditary optic neuropathy	Patrick Yu Wai Man
15:15–15:30	Future treatment for other inherited optic neuropathies	Nancy Newman
15:30	*Refreshments and posters*	

16:00 Symposium: Key Clinical Questions

Chairs: Lidia Alvarez, Deniz Atan

16:00–16:20	Should all multiple sclerosis patients have modern disease modifying treatments?	Martin Duddy
16:20–16:40	Is transient monocular visual loss a neuro-ophthalmic emergency?	Valérie Biousse
16:40–17:00	Should all NMO optic neuritis patients receive second line immunosuppression?	Saiju Jacob
17:00–17:20	Should we give corticosteroids for traumatic optic neuropathy?	Richard Blanch
17:20–17:30	Discussion	

Thursday 23 June 2022

09:00 Symposium: Visual Impairment in Neuro-ophthalmology and Approaches for Rehabilitation

Chairs: Anja Palmowski-Wolfe, Susanne Trauzettel-Klosinski, James Mitchell

09:00–09:15	Timing of vision screening and assessment in an acute population	Fiona J Rowe
09:15–09:30	Does reading training in vertical direction improve the hemianopic reading disorder? A randomised and controlled study	Susanne Trauzettel-Klosinski
09:30–09:45	Quantifying the Speedwheel – a nonverbal acuity test based on suppression of the optokinetic nystagmus	Anja Palmowski-Wolfe
09:45–10:00	Clinical applications of pupillography in visually impaired patients	Skorkovská Karolína
10:00–10:15	Visual perceptual learning in youth with infantile nystagmus: Feasibility and results of a home-based training App	Bianca Huurneman
10:15–10:30	Evolution and contribution of e-Health technologies in the clinical processes and rehabilitation of low vision patients related to neurological disorders	María B Coco Martín
10:30–10:45	Discussion	
10:45	*Refreshments and posters*	

11:15 Symposium: Radiation Treatment in Neuro-ophthalmology

Chair: Tim Matthews, Prem Subramanian, Ruth Huna-Baron

11:15–11:35	Radiotherapy for anterior skull base meningiomas	P Sanghera
11:35–11:55	Proton therapy for base of skull chordoma and chondrosarcoma – visual function considerations	G Whitfield
11:55–12:45	MDT discussion of cases	G Tsermoulas, S Chavda, P Sanghera, T Matthews
12:45–13:00	Discussion	

13:00–13:30	*Prizes and closing of meeting*	*Tim Matthews*

PLATFORM PRESENTATIONS

Platform T01

THE PHASE III REFLECT TRIAL: EFFICACY OF BILATERAL GENE THERAPY FOR LEBER HEREDITARY OPTIC NEUROPATHY (LHON) IS MAINTAINED 2 YEARS POST ADMINISTRATION

Patrick Yu-Wai-Man[1,2,3,4], Nancy J Newman[5], Prem Subramanian[6], Mark L Moster[7], An-Guor Wang[8], Sean Donahue[9,10,11], Bart Leroy[12,13,14], Valerio Carelli[15,16], Valerie Biousse[5], Catherine Vignal-Clermont[17,18], Alfredo A Sadun[19], Gema R Fernandez[20,21], Elizabeth Fortin[22], Rudrani Banik[23], Laure Blouin[24], Michel Roux[24], Magali Taiel[24], Jose A Sahel[25,26,27,28].

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Background and Aims: REFLECT is a Phase III randomised trial assessing the efficacy and safety of a bilateral intravitreal injection of lenadogene nolparvovec for the treatment of MT-ND4 Leber hereditary optic neuropathy (LHON).

Methods: Ninety-eight LHON subjects carrying the m.11778 G > A mutation and with vision loss ≤ 365 days received an intravitreal injection of lenadogene nolparvovec in the first-affected eye. The second-affected eye was randomly allocated to either gene therapy or placebo.

Results: Forty-eight subjects were treated bilaterally and 50 unilaterally (second-affected eye receiving placebo). At Year 2, a statistically significant improvement in best corrected visual acuity (BCVA) was reported from baseline in all eyes treated with lenadogene nolparvovec. A significant improvement from the nadir was observed in all eyes, reaching +20 and +17 ETDRS letters in the first- and second-affected eyes, respectively, for bilaterally treated patients ($p < .0001$), and +19 and +14 ETDRS letters in the first-treated and second-placebo eyes, respectively, for unilaterally treated patients ($p < .0001$). A better final BCVA was reported in subjects treated bilaterally compared with those treated unilaterally (+6 letters).

Conclusions: The statistically significant improvement of BCVA from baseline and the nadir reported at 1.5 years post administration of lenadogene nolparvovec was maintained at 2 years. The REFLECT results suggest a dose effect with bilateral injection of lenadogene nolparvovec.

Disclosures: None.

Platform T02

NEURO-OPHTHALMOLOGICAL CHARACTERISATION OF NOT-OPA1 DOMINANT OPTIC ATROPHY

Giulia Amore[1], Martina Romagnoli[2], Michele Carbonelli[1], Piero Barboni[3],Leonardo Caporali[2], Claudio Fiorini[1], Flavia Palombo[2], Valerio Carelli[1,2], Chiara La Morgia[2].

1. Department of Biomedical and Neuromotor Sciences, University of Bologna, Bologna, Italy. 2. IRCCS Institute of Neurological Sciences of Bologna, Bellaria Hospital, Bologna, Italy. 3. Studio Oculistico D'Azeglio, Bologna, Bologna, Italy.

Background and Aims: Heterozygous mutations in the AFG3L2 gene (encoding a protease involved in OPA1 cleavage) and the ACO2 gene (encoding the mitochondrial enzyme aconitase) are associated with isolated forms of dominant optic atrophy (DOA). We aimed to describe their neuro-ophthalmological phenotypes and to compare them with classic OPA1-related DOA.
Methods: Mutations in AFG3L2 and ACO2 genes were identified by a multigenic targeted panel. Neuro-ophthalmological evaluation included fundus examination, visual acuity (VA), colour vision, visual field (VF), optical coherence tomography of the retinal nerve fibre layer (RNFL) and ganglion cell layer (GCL). ACO2 and AFG3L2 patients were compared with a matched group of OPA1 patients.
Results: Forty-four eyes from 23 ACO2 and 26 eyes from 13 AFG3L2 patients were compared with 144 eyes from 72 OPA1 patients. All patients presented with bilateral temporally-predominant optic atrophy with various degree of visual impairment. Comparison among groups failed to reveal any significant difference in terms of VA, colour vision, VF parameters and RNFL thickness. Regarding GCL analysis, ACO2 patients had significantly higher thickness on average ($p = .01$) and in all sectors except supero-nasally compared with OPA1 patients, but only in the superior sector ($p = .02$) compared with AFG3L2 patients. The comparison between the AFG3L2 and OPA1 patients was unremarkable for the GCL analysis.
Conclusions: Despite a different genetic background, AFG3L2 and ACO2 patients showed a very similar neuro-ophthalmological phenotype. Overall, AFG3L2-related DOA is indistinguishable from OPA1-DOA, probably due to the effect of imbalanced OPA1 isoforms, while ACO2 patients only have a more preserved thickness of the retinal GCL compared with classic OPA1 patients.
Disclosures: None.

Platform T03

IIH PRESSURE: A RANDOMISED CONTROLLED TRIAL OF THE GLP-1 RECEPTOR AGONIST EXENATIDE IN IDIOPATHIC INTRACRANIAL HYPERTENSION

James L Mitchell[1,2], Susan P Mollan[1,3], Jessica Walker[1], Hannah Lyons[1], Andreas Yiangou[1,2], Zerin Alimajstorovic[1], Olivia Grech[1], Georgios Tsermoulas[4], Kristian Brock[1], Alex J Sinclair[1,2].

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Background and Aims: Raised intracranial pressure (ICP) causes significant morbidity and mortality in many conditions including idiopathic intracranial hypertension (IIH). The GLP-1 (Glucagon-like peptide 1) receptor agonist exenatide reduces ICP in pre-clinical data. The objectives of this study are to assess the biological effect of GLP-1 RA on ICP in participants with IIH.

Methods: This was a randomised, placebo controlled, double-blind physiology study of exenatide in women with active IIH (>25 cm cerebrospinal fluid [CSF] lumbar puncture opening pressure and papilloedema). Telemetric, intraparenchymal ICP monitors were implanted. Participants were randomised 1:1 to exenatide or placebo for 12 weeks. ICP was recorded at baseline, 2 weeks and 12 weeks.
Results: Sixteen participants were recruited; 15 were randomised and completed the study. Their parameters were: age 29.5 ± 9.5 years; body mass index 38.1 ± 6.2 kg/m^2; and ICP 30.6 ± 5.1 cmCSF. Significance was set at $p < .1$ as an early phase trial. ICP, the primary endpoint, fell significantly at 2.5 hours −5.7 (2.9) cmCSF ($p = .048$), at 24 hours −6.4 (2.9) cmCSF ($p = .030$) and at 12 weeks −5.6 (3.0) cmCSF ($p = .058$). Monthly headache days fell in the exenatide treated cohort (−7.7 [9.2], $p = .069$) and vision improved (logMAR acuity −.1 [0.04] $p = .004$).
Conclusions: We report the first human study assessing the biological effect of the GLP-1 agonist exenatide on ICP in IIH utilising telemetric ICP monitors. Exenatide reduced ICP at all timepoints, including acutely at 2.5 hrs and during chronic dosing. New therapies for ICP modulation are a significant unmet need.
Disclosures: Miss Grech reports scientific consultancy fees from Invex therapeutics during the conduct of the study. Professor Mollan reports other Invex Therapeutics, other Heidelberg engineering during the conduct of the study; other from Chugai-Roche Ltd, other from Janssen, other from Allergan, other from Santen, other from Roche, other from Neurodiem, outside the submitted work. Professor Sinclair reports consulting fees and stockholding with Invex therapeutics, during the conduct of the study. Other from Allergan, Amgen, Novartis and Cheisi. All other authors declare no competing interests.

Platform T04

IIH PRESSURE MED: A RANDOMISED, SEQUENTIAL, TRIAL OF THE EFFECT ON COGNITION OF FIVE DRUGS COMMONLY USED IN IDIOPATHIC INTRACRANIAL HYPERTENSION

James L Mitchell[1,2], Susan P Mollan[1,3], Jessica Walker[1], Hannah Lyons[1], Andreas Yiangou[1,2], Zerin Alimajstorovic[1], Olivia Grech[1], Georgios Tsermoulas[4], Kristian Brock[1], Alex J Sinclair[1,2].

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Background and Aims: Limited clinical data exist to guide treatment of idiopathic intracranial hypertension (IIH). Impaired cognition is increasingly appreciated in IIH. We examined the effects of five commonly used drugs on cognition in IIH.
Methods: We carried out a randomised, sequential, open label trial in women with active IIH. Participants were treated for 2 weeks, with the order randomised, with acetazolamide, amiloride, furosemide, spironolactone and topiramate. There was drug-washout between rounds. Cognition was assessed using the NIH Toolbox Cognitive Battery (NIHTB-CB) pre- and post-treatment. Headache frequency and severity was recorded with diaries. Analysis was by hierarchical regression, a priori significance was set at $p < .1$ as an early phase study.
Results: Fourteen participants were recruited. Mean body mass index was 38.1 (6.2) kg/m^2 and mean intracranial pressure was 30.6 (5.1) cmCSF at baseline. Baseline cognitive performance in the fluid domain was more than 1 standard deviation below expected, fully corrected T-Score (mean [standard error]) of 37.2 (2.55). Following drug treatment there was a further significant reduction in the fluid composite score associated with acetazolamide (mean T-score (SE), −5.00 (2.6), $p = .057$ and topiramate −4.14 (2.0), $p = .061$. Headache severity was also associated with worse cognitive performance $R^2 = .188$, $p = .0005$.

Conclusions: This study demonstrates that cognitive impairment is present in IIH and is quantifiable using a rapid standardised test. Headache severity impairs cognition. Cognition is further impaired by therapy with acetazolamide and topiramate. These results have important clinical implications when weighing up the pros and cons of prescribing in IIH.
Disclosures: Miss Grech reports scientific consultancy fees from Invex therapeutics during the conduct of the study. Professor Mollan reports other Invex Therapeutics, other Heidelberg engineering during the conduct of the study; other from Chugai-Roche Ltd, other from Janssen, other from Allergan, other from Santen, other from Roche, other from Neurodiem, outside the submitted work. Professor Sinclair reports consulting fees and stockholding with Invex therapeutics, during the conduct of the study. Other from Allergan, Amgen, Novartis and Cheisi All other authors declare no competing interests.

Platform T05

UTILISING OPTICAL COHERENCE TOMOGRAPHY MEASURES AS SURROGATES FOR RAISED INTRACRANIAL PRESSURE IN IDIOPATHIC INTRACRANIAL HYPERTENSION

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Background and Aims: There is an unmet need for non-invasive intracranial pressure (ICP) biomarkers. In idiopathic intracranial hypertension (IIH), ICP manifests as papilloedema, which can be quantified by optical coherence tomography (OCT). We sought to determine whether optic nerve head OCT in papilloedema could act as a surrogate measure of ICP.
Methods: This was a longitudinal cohort study with data from three randomised controlled trials. OCT and automated perimetry was followed immediately by ICP measurement. Cohort 1 utilised continuous telemetric ICP monitoring in one visit. Cohort 2 were evaluated at baseline, 3, 12 and 24 months and underwent lumbar puncture ICP assessment.
Results: One hundred and four patients were recruited. Amongst cohort 1 (n = 15) a range of OCT protocols were evaluated and optic nerve head central thickness (ONHCT) was found to be most closely associated with ICP (p = .03; r = .56) and superior to the current standard of care scan (retinal nerve fibre layer thickness). Cohort 2 (n = 89) confirmed the correlation between ONHCT and ICP longitudinally (12 and 24 months). Finally, bootstrap surrogacy analysis noted a positive association between ONHCT and ICP at all time points (e.g. at 12 months, an increase in ONHCT of 50 μm predicted an increase in ICP of 5 cmCSF).
Conclusions: OCT ONHCT reproducibly correlates with ICP and surrogacy analysis demonstrates its ability to predict ICP changes. OCT scanning is widely utilised internationally. Our data suggest that it has the utility to not only monitor papilloedema but also non-invasively predict ICP in IIH.

Disclosures: Professor Mollan reported personal fees from Heidelberg Engineering and advisory board fees from Invex Therapeutics during the conduct of the study and personal fees from Allergan, Santen, Sathera, Roche, Chugai, and Novartis outside the submitted work. Dr Mitchell reported grants from Ministry of Defence during the conduct of the study. Dr Brock reported personal fees from Invex Therapeutics during the conduct of the study; and equity ownership in AstraZeneca and GlaxoSmithKline, personal fees from Eli Lilly reimbursement of costs from Merck and Roche outside the submitted work. Professor Sinclair reported fees from Invex Therapeutics as a company director with salary and stock options during the conduct of this study; and personal fees from Novartis and Allergan outside the submitted work. No other disclosures were reported.

Platform T06

SEGREGATION OF DECUSSATING AXONS WITHIN THE CORE OF THE PRIMATE OPTIC CHIASM SHOWN BY INJECTION OF A DIFFERENT TRACER INTO EACH EYE

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Background and Aims: It is unclear why tumours cause more injury to decussating nasal retinal ganglion cell fibres, thereby giving rise to temporal visual field loss. To address this issue, the course of fibres through the monkey optic chiasm was examined following injection of a different fluorescent tracer into each eye.
Methods: Under general anaesthesia, cholera toxin subunit B Alexa Fluor 488 was injected into the right eye and cholera toxin subunit B Alexa Fluor 594 was injected into the left eye. After a week's survival for anterograde transport, serial coronal sections through the primary optic pathway were examined.
Results: Crossed fibres were confined mainly to a central core zone within the anterior and mid portions of the optic chiasm. This region of decussation was delineated by separate, interwoven sheets of green (right eye) and red (left eye) fibres. It expanded steadily to fill more of the optic chiasm as fibres coursed posteriorly. Eventually, crossed fibres became intermingled with uncrossed fibres, so that segregation was lost.
Conclusions: Crossed fibres are located primarily in a separate, central core zone of the optic chiasm. Sellar tumours concentrate their compressive force on this region within the optic chiasm, explaining why they so often produce bitemporal visual field loss.
Disclosures: None.

Platform T07

DEEP LEARNING CAN ACCURATELY DISTINGUISH BETWEEN TRUE PAPILLOEDEMA AND OPTIC DISC DRUSEN ON RETINAL PHOTOGRAPHS

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Background and Aims: Identification of true papilloedema due to raised intracranial pressure requires high neuro-ophthalmic expertise and expensive ancillary testing. Its ophthalmoscopic diagnosis can be easily mistaken with pseudo-papilloedema and, in particular, with optic disc drusen (ODD), especially if they are not ophthalmoscopically visible. We aimed to evaluate the performance of a deep learning system (DLS) to automatically distinguish between true papilloedema and ODD on standard retinal fundus images, in a large international multi-ethnic population.

Methods: We have developed, trained, and tested a DLS, using a total of 4,410 digital retinal images obtained in 2,108 patients, in 30 centres, from 20 countries. The reference ophthalmological and neurological diagnosis was established by the expert Neuro-ophthalmology providers, based on state-of-the-art diagnostic methods. The DLS performance was evaluated using its sensitivity, specificity, area under the receiver operating curve (AUC) and accuracy, following a binary classification of papilloedema versus ODD.
Results: The DLS was trained using 3,014 confirmed papilloedema images and 765 confirmed ODD images (476 visible, 289 buried). Subsequent external testing was performed in 323 patients from 3 independent centres (including 424 papilloedema images and 207 ODD images – 95 visible ODD and 112 buried 0DD). In the external dataset, the AUC to discriminate papilloedema from ODD was .98 (.97–.99), accuracy 91.9 (90.3–93.5), sensitivity 87.4 (84.1–91.1), and specificity 94.1 (92.5–95.9). After removing the visible ODD, the AUC to discriminate papilloedema from buried ODD was marginally reduced to .96 (.95–.98).
Conclusions: Deep learning-based image identification on conventional retinal fundus images has the ability to distinguish with high accuracy between true papilloedema and benign ODD. The implementation of such a DLS into a neurological environment could in the future improve diagnosis and management of patients with suspected papilloedema.
Disclosures: None.

Platform T08

OPIATE AND HEADACHE PRESCRIBING PATTERNS IN WOMEN WITH IDIOPATHIC INTRACRANIAL HYPERTENSION: A MATCHED CONTROLLED RETROSPECTIVE COHORT STUDY

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Background and Aims: Physician prescribing habits, opiate use and use of headache therapies have not been evaluated in a large matched cohort in idiopathic intracranial hypertension (IIH). The aim was to evaluate headache medication prescribing habits, opiate use and headache burden in women with IIH compared with matched women with migraine and population controls.
Aims and Methods: Data from IQVIA Medical Research Data, an anonymised, nationally representative primary care database in the UK between January 1995 and September 2019 was used. Women with IIH (exposure) were matched by age and body mass index with 10 control women without IIH but with migraine (migraine controls), and population controls.
Results: There were 3411 women with IIH, 30,879 migraine controls and 33,495 population controls included in the study. The adjusted hazard ratio (aHR) for new onset headache in IIH compared with population controls was 3.09 (95% confidence intervals [CI] 2.78–3.43). In the first year after diagnosis, 58% of women with IIH were prescribed acetazolamide and 20% topiramate. Twenty percent of women with IIH were prescribed opiates within the first year of their diagnosis, reducing to 17% after six years, compared with 8% and 11% of those with migraine, respectively. Women with IIH were prescribed more headache preventative medications compared with migraine controls.
Conclusions: Women with IIH were more likely to be prescribed opiate and simple analgesics compared with both migraine and population controls. Women with IIH trialled more preventative medications over their diseases course suggesting that headaches in IIH may be more refractory to treatment.

Disclosures: Professor Mollan reported personal fees from Heidelberg Engineering and advisory board fees from Invex Therapeutics during the conduct of the study and personal fees from Allergan, Santen, Sathera, Roche, Chugai, and Novartis outside the submitted work. Professor Sinclair reported fees from Invex Therapeutics as a company director with salary and stock options during the conduct of this study; and personal fees from Novartis and Allergan outside the submitted work. No other disclosures were reported.

Platform T09

RETINAL FRACTAL DIMENSION IN DEMENTIA: THE ALZEYE STUDY

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Background and Aims: Microvascular dysfunction is a hallmark of neurodegenerative diseases, such as Alzheimer's disease. We analysed the association between retinal fractal dimension (FD), a geometric index of complexity, and all-cause dementia in a large ethnically and socioeconomically diverse cohort in London, United Kingdom.
Methods: This was a retrospective cohort study linking ophthalmic imaging of patients attending Moorfields Eye Hospital aged ≥ 40 years with hospital admission data from 2008–2018. Multivariable linear regression modelled the association between FD, extracted from macular-centred colour photographs using the Vascular Assessment and Measurement Platform for Images of the Retina (VAMPIRE) software, with dementia.
Results: Among 96,995 patients, 4,353 had a diagnosis of dementia: of whom 1,222 had Alzheimer's disease, 885 vascular dementia, 229 mixed disease and 2017 unspecified dementia. Compared with those without dementia, patients with dementia were older (79.7 ± 8.6 versus 67.7 ± 12.4), more likely to be women (58.7% versus 50.6%) and more likely to have hypertension (83.3% versus 80.4%); all $p < .001$. Individuals suffering from dementia had reduced retinal FD after adjusting for age, sex, diabetes mellitus and hypertension (beta −.0047, 95% confidence intervals: −.005 to −.003, $p < .001$). There was no interaction between the covariates.
Conclusions: A strong inverse association was seen between retinal FD and prevalent all-cause dementia, after adjustment for known confounders of FD, confirming microvascular dysfunction as a pathophysiological correlate. Subsequent analyses will examine associations between other metrics of retinal morphology, derived from optical coherence tomography and incident disease.
Disclosures: Pearse Keane reports financial support from Allergan, Bayer, Heidelberg Engineering, Novartis, Roche and Topcon. He has acted as a consultant for Apellis and DeepMind.

Platform T10

ASSOCIATION OF RETINAL FRACTAL DIMENSION WITH INCIDENT CARDIOVASCULAR EVENTS IN A HOSPITAL-ATTENDING POPULATION IN LONDON, UNITED KINGDOM

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Background and Aims: Changes in retinal vascular morphology are related to cardiovascular morbidity. Here, we assessed the association between retinal fractal dimension (FD) and incident cardiovascular (CV) events in an ophthalmic hospital-attending population in London, United Kingdom.
Methods: This was a retrospective cohort study linking imaging of patients aged ≥ 40 years with hospital admissions with myocardial infarction and ischaemic stroke from 2008 to 2018. Retinal FD was calculated from macular-centred photographs using the Vascular Assessment and Measurement Platform for Images of the Retina (VAMPIRE) software. Odds ratios (OR) from multivariable logistic regression examined the association between incident cardiovascular (CV) events and retinal FD.
Results: Among a cohort of 62,143 patients, 674 had had a CV event after 1 year, 1,731 after 3 years, and 2330 after 5 years. Retinal FD was reduced in patients with an incident CV event (1.50) compared with controls (1.51, $p < .001$). After adjustment for age, sex, hypertension and diabetes mellitus, retinal FD was inversely associated with incident CV events with similar odd ratios for 1 year, 3 year and 5 year incidence (1 year: .91, .85–.97; 3 year: .91, .87–.95; and 5 year .90, .87–.93 per FD increase). Effect modification by sex was apparent. Stratification by sex showed a significant association of retinal FD with female sex only.
Conclusions: Retinal vasculature provides an easily accessible window into microvascular dysfunction. An inverse relationship was found between retinal FD and incident CV events but only among those of female sex. Subsequent research will assess other retinal vascular indices, the contribution of longitudinal oculomic biomarkers and prediction rather than aetiological objectives.
Disclosures: Pearse Keane reports financial support from Allergan, Bayer, Heidelberg Engineering, Novartis, Roche and Topcon. He has acted as a consultant for Apellis and DeepMind.

Platform T11

INCREASED RISK OF CEREBROVASCULAR DISEASE AMONG PATIENTS WITH NON-ARTERITIC ANTERIOR ISCHAEMIC OPTIC NEUROPATHY – A RETROSPECTIVE NATIONWIDE COHORT STUDY

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Background and Aims: Non-arteritic anterior ischaemic optic neuropathy (NAION) is the most common acute optic neuropathy among older adults. Data about the association between NAION and stroke is controversial and sparse. We aimed to investigate the occurrence of stroke in patients with NAION and the correlation between NAION and carotid artery stenosis (CAS) as a risk factor for stroke.
Methods: This was a retrospective nationwide, population-based-study of the "CLALIT" health insurance service sample cohort database between 2009 and 2019. Out of 1,200,000 insured individuals we included 605 patients with a new diagnosis of NAION without a preceding diagnosis of stroke. The control group was tenfold larger. The groups were matched by age, sex, and systemic cardiovascular risk factors.
Results: Stroke occurred in 18.7% and 14.7% ($p = .009$) of patients from the NAION and control groups respectively. Small vessel disease (SVD) occurred in 4.79% and 3.12% ($p = .028$) and CAS in 13.7% and 10.4% ($p = .012$), respectively. In multivariate analysis, the hazard ratios for stroke, CAS, and SVD in NAION patients were 1.32 ($p = .094$), 1.6 ($p = .002$), and 1.56 ($p = .031$), respectively.

Conclusions: According to our big data study results, there is a correlation between NAION and the incidence of stroke. Furthermore, NAION patients are at greater risk of having CAS compared with the general population. These data raise the question of whether ophthalmologists should assess NAION patients for stroke risk factors.
Disclosures: None.

Platform T12

DO RETINAL MICROVASCULAR CHANGES IMPR-OVE TRANSIENT ISCHAEMIC ATTACK (TIA) RISK STRATIFICATION SCORES? ABCD2 VS. ABCDEYE IN THE FOTO-TIA STUDY

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Background and Aims: The ABCD2 score is widely used for transient ischaemic attack (TIA) risk stratification in the emergency department (ED). We aimed to determine the effect of adding retinal microvascular findings and diffusion weighted imaging (DWI) positivity to the ABCD2 score for predicting the probability of a cerebrovascular event (CVE).
Methods: This was a prospective cohort of patients admitted to ED for suspected TIA/stroke. All had non-mydriatic fundus photographs looking for retinal microvascular findings (retinal haemorrhages, cotton wool spots, retinal emboli/occlusions, hard exudates, and microaneurysms). A stroke neurologist rated the probability each patient's presentation represented a CVE. Linear regression modelling was used for analysis.
Results: We included 395 patients (median age: 57 years (interquartile range 50–66); of whom 34 (9%) had retinal microvascular findings. Comparison of regression coefficients suggested that diabetes mellitus (DM) was replaced in the ABCD2 with microvascular findings, but when using the original ABCD2 point cut-offs (e.g., age ≥ 60 years: 1 point), both models had poor calibration (predicted versus neurologist-determined probability of TIA). By incorporating DWI positivity and either microvascular findings or DM and by using the full regression coefficients, excellent calibration was achieved (bootstrap bias corrected mean absolute error ~1%) and slightly favoured microvascular findings over DM.
Conclusions: Although we have reported that retinal microvascular findings among patients with suspected TIA are an independent factor differentiating TIA/stroke from mimics, incorporating them into ABCD2 does not markedly improve rule performance. However, the presence of diabetic ophthalmic microvascular disease better predicts CVE than simply the presence of DM alone.
Disclosures: None.

SYMPOSIUM

Visual impairment in Neuro-Ophthalmology and approaches for rehabilitation

Learning objectives: In this symposium we aim to increase awareness of the mutual importance of Neuro-Ophthalmology and Low Vision Specialists in patient care. Neuro-Ophthalmology concerns itself with diseases of the brain and the eye as a part of the brain. Despite best medical care these diseases frequently result in residual loss of vision involving acuity, contrast and/or fields and these patients benefit from low vision counselling. On the other hand, visual impairment is frequently associated with a neuro-ophthalmological disorder that may benefit from early diagnosis in order to, if possible, correct visual impairment or to prevent further impairment. For either area research aimed at improved diagnostics, visual training options and E health technologies can help speed up the diagnostic and rehabilitation process.

S01

TIMING OF VISION SCREENING AND ASSESSMENT IN AN ACUTE POPULATION

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Background and Aims: Stroke can have serious detrimental effects on the visual system and cortical processing, including eye movement disorders, visual field loss, low vision, perceptual and cognitive difficulties. There are considerable issues relating to the screening and, thus, identification of visual problems following stroke.
Methods: Ideally visual problems should be identified as early as possible after stroke onset. However, screening for visual impairment is neither standardised nor routine across many stroke units. In this presentation a review of the medical literature will be considered alongside data from two large UK-based cohort studies: the Vision In Stroke (VIS) study and Impact of Visual Impairment after Stroke (IVIS) study, which explore the visual consequences of stroke. Information will be presented on the timing at which specialist vision screening is feasible in an acute stroke population.
Results: A comprehensive visual assessment should include assessment of visual acuity, visual fields, ocular alignment, ocular motility, visual inattention and visual perception alongside documentation of past ocular history and current visual symptoms. Prospective epidemiology research shows that the majority of stroke survivors can be visually screened within 1 week of stroke onset with the median being assessed within 72 hours of stroke onset. Reasons for not being able to undergo vision screening or assessment are mostly related to severity of stroke where stroke survivors are unable to participate in with vision testing.
Conclusions: This presentation demonstrates that early visual assessment is feasible when undertaken by Orthoptists. The most common categories of visual impairment are impaired central vision and eye movement disorders followed by visual field loss and visual perceptual difficulties. Early visual assessment is important as a core stroke assessment in that information can be provided on visual status and its functional significance to the stroke team, patients and carers.

S02

DOES READING TRAINING IN VERTICAL DIRECTION IMPROVE THE HEMIANOPIC READING DISORDER? A RANDOMISED AND CONTROLLED STUDY

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Background and Aims: Reading training in vertical text orientation might improve reading performance in patients with homonymous hemianopia (HH), if they place the text along the vertical midline in the seeing hemifield.
Methods: Twenty-one patients with HH were randomly assigned to either vertical or horizontal reading training. They trained independently at home reading single lines of texts displayed on a computer screen for 2 × 30 min/day, 5 days/week, for 4 weeks. The main outcome variable was reading speed (RS) during reading printed standardised paragraphs of text (IReST) aloud. RS was measured before training (T1), directly after training (T2) and 4 weeks later (T3). Quality of life (QoL) was assessed by the Impact of Visual Impairment (IVI) questionnaire.
Results: Vertical training significantly improved RS in the vertical direction in patients with right HH. Horizontal training significantly improved RS in the horizontal direction, much more in patients with left HH. Both effects remained stable at T3. RS during training at the computer correlated strongly with RS of printed text. QoL showed a statistically significant improvement for vertical training.
Conclusions: The improvements of RS were specific for each training direction. The stable effect indicates the application of the newly learned strategies to everyday life. The side of the HH plays an important role: Left-HH patients benefitted from horizontal training; whereas right-HH patients benefitted from vertical training. However, the vertical RS did not reach the level of horizontal RS, but might be further improved by a longer training period.

S03

QUANTIFYING THE SPEEDWHEEL – A NONVERBAL ACUITY TEST BASED ON SUPPRESSION OF THE OPTOKINETIC NYSTAGMUS

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Background and Aims: In nonverbal children or adults, assessment of visual acuity with Snellen charts can be impossible especially, if in addition to low vision, developmental delay is present. We have thus developed an objective visual acuity test based on suppression of the optokinetic nystagmus (OKN), the "Speedwheel Test" (SW), and compared it with subjective visual acuity assessed with the Freiburg acuity test (FrACT; Bach M. Optom Vis Sci. 1996;73:49–53).
Methods: We obtained agreement limits for visual acuity assessed with the SW (SWA) compared with FrACT with Landolt-C and with tumbling E Symbols in 241 patients with various visual impairments, aged 4–88 years.
Results: Prediction intervals were not related to age. Subjective and objective visual acuity compared well, with a mean difference between Landolt-C symbols and logSWA of −.01 and between E symbols and SW of −.15. The results were influenced by the acuity range measurable with the respective tests, which was between −.2 and 1.43 logMAR for FrACT and between .05 and 1.0 logMAR for SWA. The distribution of visual acuity estimates of SW compared with FrACT C-test or FrACT-E in a similar manner as FrACT-C compared with FrACT-E.
Conclusions: The SW is an objective visual acuity test that does not require verbal feedback and shows a good correlation with subjective chart acuity. Thus it can be further explored in assessing visual acuity in a population otherwise unable to cooperate. Lower visual acuities may be tested by reducing the test distance, which here had been set at 1 m.

S04

CLINICAL APPLICATIONS OF PUPILLOGRAPHY IN VISUALLY IMPAIRED PATIENTS

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Background and Aims: With the discovery of intrinsically photosensitive retinal ganglion cells (ipRGCs), we have learned that the pupillomotor information delivered to the midbrain may originate not only in the outer retinal layer (activation of rods and cones) but also in the inner retinal layer (activation of ipRGCs), and pupillographic measurements in patients with various disorders of the visual pathway support the existence of two pupillomotor channels that drive the pupil light reaction – the subcortical (associated with the ipRGCs) and the supragenicular (driven by the rods and cones and receiving input from the visual cortex and extrastriate areas). The individual components of the pupil light reaction can be subtracted by the so-called chromatic pupillography.
Methods: By presenting light stimuli of varying intensity, duration, and wavelength, combined with specific backgrounds and adaptation conditions, it is possible to stimulate rods, cones, or ipRGCs.
Results: Clinical studies have proven the suitability of chromatic pupillography to detect changes in specific ophthalmologic disorders, ranging from impaired ipRGC function in glaucoma, age-related macular degeneration and diabetes mellitus, to reduced rod/cone function with enhanced ipRGC function in hereditary rod-cone dystrophies. In retinitis pigmentosa, pupillary responses are still recordable, even in patients with extinguished electroretinography recordings. Furthermore, protocols for chromatic pupillography have been developed for the objective assessment of efficacy in gene therapy in patients with CNGA3-linked achromatopsia. Chromatic pupillography may be combined with the presentation of focal light stimuli at different locations in the visual field, thus resembling classical perimetry and providing topographic information about the human retina.
Conclusions: Chromatic pupillography together with its possible modifications has great potential for examining the function of the retina and visual pathway in low-vision patients where other methods have reached their limits. Its clinical relevance is particularly evident since the development of new techniques to restore some vision in severely visually-impaired patients.

S05

VISUAL PERCEPTUAL LEARNING IN YOUTH WITH INFANTILE NYSTAGMUS: FEASIBILITY AND RESULTS OF A HOME-BASED TRAINING APP

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Background and Aims: In a previous project we demonstrated that a computerised visual perceptual learning paradigm improves visual performance in children with infantile nystagmus (IN) (Huurneman et al. IOVS 2016;57(10):4216–4246). Our training is a combination between a threshold acuity letter and visuomotor training. During training, a goal-directed saccade has to be initiated towards a threshold acuity letter under time constraints. This visuomotor component is included to train oculomotor and attentional control and invoke broader learning transfer. The previous version of our training, written in MATLAB programming language, could not be used at home without supervision.
Methods: The goal of the current project was to deliver a clinical training tool that could be used independently at home to induce substantial, long-term vision improvements in youths with IN.

Results: Thirty-six children (aged 7–18 years) with IN participated in this study. Children trained two times per week for a period of 20 weeks, resulting in a grand total of 40 training sessions (350 trials per training; total of 14,000 trials). Viewing distance was monitored with the use of a head target (which was visible 88 ± 8% of the time). The gain factor (real/desired distance) of the recorded viewing distance was .96 ± .02. Compliance with our training protocol was good; ~70% of the children completed 40 training sessions. Performance on the trained task improved single letter acuity (improvements: .084 ± .026 logMAR) and crowded letter acuity (improvements: .19 ± .05 logMAR). The larger improvements on crowded compared with single letter acuity were expected since the majority of training sessions consisted of a crowded letter configuration (30/40 sessions). Learning effects were not only visible on the trained task. They transferred to near and distance visual acuity as measured with vision charts, and improved stereopsis. Spatial and temporal aspects of reading performance and subjective functional vision (measured with the Functional Vision Questionnaire) improved too. Contrast sensitivity function did not change. Improvements were preserved 6 months after training.
Conclusions: Our novel training app is effective at improving a broad range of visual functions in children and adolescents with IN. Such home-based visual perceptual learning is a valuable rehabilitation tool for youths with IN. Average crowded distance vision improvements equal approximately 2 logMAR lines on the vision chart.

S06

EVOLUTION AND CONTRIBUTION OF E-HEALTH TECHNOLOGIES IN THE CLINICAL PROCESSES AND REHABILITATION OF LOW VISION PATIENTS RELATED TO NEUROLOGICAL DISORDERS

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Background and Aims: Over the last decade, the implementation of e-Health tools has increased significantly, as they promote earlier identification, better clinical outcomes and a greater patient interaction in the management of their diseases. In this context, vision rehabilitation is an area that has particularly benefited from technological advances, including larger screen sizes, higher screen resolutions, more processing power and lower hardware costs. These improvements allow not only a better quality of image testing but also new assessment and rehabilitation techniques using innovative algorithms, apps and devices. We aimed to quantitatively review and categorise the contributions of e-Health technologies in the comprehensive care of low vision patients due to neurological conditions, as well as to suggest future research needs in this emerging health-care field.
Methods: A systematic review in PubMed® was conducted using the terms "e-Health" or "m-Health" and terms such as low vision, visual impairment, visual assessment or visual rehabilitation. Reference lists of relevant articles, reviews and databases were also examined to find additional publications on the topic.
Results: Current evidence supports that e-Health technologies are a feasible option not only for improving visual and quality of life outcomes, but also for engaging patients in their own rehabilitation process. Besides, they promote better assessment when monitoring sudden symptoms or adverse events.
Conclusions: As many patients with disabling neurological diseases have mobility restrictions, technology has the potential to improve access to health-care services. Nevertheless, more research is needed in this field as most of the published papers need methodological improvements.

POSTER PRESENTATIONS

DISORDERS OF THE ANTERIOR VISUAL PATHWAY (RETINA, OPTIC NERVE, AND CHIASM)

P001

OCULAR MANIFESTATIONS AFTER ACUTE METHANOL POISONING

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Background and Aims: Acute methanol poisoning is often associated with a poor prognosis. The functional prognosis after survival is mainly based on the degree of ocular impairment, most frequently related to optic nerve involvement. The aim of this study was to describe the ocular manifestations after acute methanol poisoning during an outbreak in Tunisia.
Methods: The following tests were carried out: visual field; colour vision; and optical coherence tomography (OCT) with evaluation of the retinal nerve fibre layer (RNFL). Patients were classified in two groups. Group 1 included patients with a normal ophthalmological examination and group 2 included patients with abnormal ophthalmological findings.
Results: The data from 21 patients (41 eyes) were analysed. The mean age of the patients was 31 years. All patients were male. Patients were examined on average 4 days after acute methanol poisoning. Twelve patients (57%) were classified into group 1 and 9 patients (43%) into group 2. The differences were not statistically significant between the 2 groups in terms of age, serum methanol concentrations and RNFL thickness (p = .2, .3 and 1, respectively). The abnormal ophthalmic findings in group 2 included optic neuropathy in seven patients (12 eyes), bilateral central retinal artery occlusion in one patient and unilateral central serous chorioretinopathy in one patient.
Conclusions: Ocular manifestations after acute met-hanol poisoning were relatively frequent in our study. Optic neuropathy was the most frequent finding.
Disclosures: None.

P002

DISC OEDEMA IN BIRDSHOT CHORIORETINOPATHY

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Background and Aims: Birdshot chorioretinopathy (BSCR) is a rare form of autoimmune choroiditis. Disc oedema is reported in up to 38% and optic atrophy in 4% of patients, but is overall an understudied aspect of the condition.
Methods: This was a retrospective chart review of three treatment-naïve patients with BSCR and disc oedema.

Results: Two males and one female, with a mean age of 61 years (range: 52–67) had bilateral blurring of vision and floaters for 10–18 months. The initial mean visual acuity was 20/38 (range: 20/25 – 20/60). All had vitritis (range: trace – 2+) and typical bilateral birdshot lesions. Vascular/disc leakage and hypocyanescent lesions were present on fluorescein and indocyanine green angiography in all eyes. Disc oedema was bilateral in all patients (moderate in two and mild in one). Peripapillary retinal nerve fibre layer optical coherence tomography was available in four eyes at presentation with a mean thickness of 165.5 (range: 146–211) μm. Humphrey perimetry (two patients) demonstrated generalised depression (mean deviation −12.0 [range: −4.83 to −17.87] dB). Disc oedema resolved without significant macular ganglion cell-inner plexiform layer loss after corticosteroids. All had visual acuities ≥ 20/40 at latest follow up.
Conclusions: BSCR is a rare autoimmune choroiditis that primarily affects middle-aged white Caucasians. Pathology has demonstrated lymphocytic foci in the choroid, retinal vasculature and also pre-laminar optic nerve head, the last manifesting as disc oedema. A thorough examination of the vitreous and fundus in such cases should prevent unnecessary neuro-ophthalmological investigations.
Disclosures: None.

P003

RAXONE® TREATMENT FOR PATIENTS WITH DOMINANT OPTIC ATROPHY DUE TO OPA1 GENE MUTATION – A PILOT STUDY

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Background and Aims: Autosomal dominant optic atrophy (ADOA) is a disease of the retinal ganglion cells, in most cases caused by a mutation in the OPA1 gene. Currently, there is no therapy for ADOA. No prospective study about the effect of idebenone as a treatment for ADOA patients with OPA1 mutation exists.
Methods: In this trial, ADOA patients with genetically confirmed OPA1 mutation were treated with a standardised daily dose of 900 mg Raxone® (idebenone) for 12 months. The 12 months results are expected by April 2022 and will be presented at the meeting.
Results: The mean visual acuity at baseline (BL) and 6 months for the right eye (OD) were .52 and .55 logMAR, respectively and for the left eye (OS) were .54 and .52 logMAR, respectively. In both eyes stabilisation of colour vision (mean 4/21 Ishihara plates), contrast sensitivity measured with Pelli Robson charts (OD 1.11 to 1.16 log, OS 1.2 to 1.26 log) and the retinal nerve fibre layer was observed. Octopus visual field improved from 4.82 dB to 4.08 dB OD and was stable from 4.44 to 4.52 dB OS.
Conclusions: Our interim analysis shows a stabilisation of all parameters. Further examinations are necessary to evaluate the long-term therapeutic effect of idebenone in OPA1 ADOA patients.
Disclosures: Commercial compensation and support received in funds and research materials from Chiesi.

P004

CHALLENGES IN THE DIFFERENTIAL DIAGNOSIS OF ELEVATED EPISCLERAL VENOUS PRESSURE

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Background and Aims: The aim is to present the differential diagnostic pitfalls when the signs of elevated episcleral venous pressure show up and to discuss the procedures performed during the diagnostic process, considering their necessity compared with their invasive nature.
Methods: A 59-year-old female with presby-LASIK in her ophthalmic history, had persistent red eyes since her young adulthood. She presented at the refractive laser clinic with the complaint of gradually worsening of vision in her right eye (OD). On first examination her best corrected visual acuities were .6 OD and 1.0 in her left eye (OS).
Results: Besides the numerous tortuous and engorged episcleral vessels in both eyes (more prominent OD), the optic nerve head was widely excavated with a cup:disc ratio of .9 OD compared with a cup:disc ratio of .3 OS. Intraocular pressures were 14 mmHg OD and 10 mmHg OS. Based on the clinical presentation a diagnosis of low-flow dural arteriovenous fistula was made and the patient was referred to the neuro-ophthalmology department. Further diagnostic evaluation, which included cerebral angiography, did not confirm the initial diagnosis.
Conclusions: During the diagnostic process the following pathologies that are known to cause impairment of episcleral venous drainage need to be considered: carotid-cavernous-sinus fistula; thyroid related ophthalmopathy; Sturge-Weber syndrome; cavernous sinus thrombosis; superior vena cava syndrome; scleritis; head/orbital tumours; and idiopathic elevated episcleral venous pressure.
Disclosures: None.

P005

A CASE OF MONOCLONAL GAMMOPATHY OF UNDETERMINED SIGNIFICANCE (MGUS) – ASSOCIATED OPTIC NEURITIS

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Background and Aims: A 77-year-old white Caucasian female was referred to a tertiary neurosciences centre following progressive left painless vision loss over 2 months.
Methods: She denied any headaches, pain on eye movement or features of giant cell arteritis. The last had been excluded with a normal temporal artery biopsy. She had a history of a right unexplained optic neuropathy, which had occurred 20 years ago, for which she had not received treatment.
Results: Her medical history included hypertension, atrial fibrillation, and monoclonal gammopathy of uncertain significance (MGUS) with a raised paraprotein. On examination, her best corrected visual acuities (BCVAs) were 6/36 and 6/18 right and left, respectively, with a right relative afferent pupillary defect. The right optic disc was pale and the left disc was swollen with cotton wool spots. Her visual fields were globally restricted in the right and showed inferior hemifield loss in the left eye. Gadolinium-enhanced magnetic resonance imaging revealed an atrophic right optic nerve and no radiological abnormality of the left anterior visual pathway. She was admitted for 5 days of intravenous methylprednisolone treatment. Her BCVAs improved with treatment to 6/24 in her right eye and 6/9 in her left eye.
Conclusions: We hypothesise that the optic neuropathy was due to elevated paraprotein levels secondary to MGUS. Here we highlight and discuss the neuro-ophthalmological picture of optic neuropathy in the context of MGUS and paraprotein.
Disclosures: None.

P006

OPTIC NERVE HEAD ANATOMY IN PATIENTS WITH OPTIC DISC DRUSEN ASSOCIATED ANTERIOR ISCHAEMIC OPTIC NEUROPATHY

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Background and Aims: The presence of optic disc drusen (ODD) is a risk factor for non-arteritic anterior ischaemic optic neuropathy (NA-AION). The purpose was to investigate in depth, using optical coherence tomography (OCT), the anatomical optic nerve head characteristics of patients with ODD-associated NA-AION (ODD-AION) compared with patients with NA-AION without ODD (nODD-AION).
Methods: This was a retrospective case control study of 34 ODD-AION and 34 nODD-AION patients that had all been systematically OCT scanned using the ODD Studies Consortium ODD scanning protocol. Patients were compared in regards to demographics, clinical characteristics, and specific optic nerve head anatomical characteristics.
Results: ODD-AION patients were significantly younger than nODD-AION patients at the time of diagnosis ($p = .012$) and had fewer vascular risk factors ($p = .015$). ODD-AION patients had significantly more peripapillary hyperreflective ovoid mass-like structures (PHOMS) ($p < .001$) and prelaminar hyperreflective lines ($p < .001$) as well as smaller scleral canals ($p = .017$) compared with nODD-AION patients. No significant difference between ODD-AION and nODD-AION patients in regard to visual acuity, refraction, lamina cribrosa position, ganglion cell layer thickness or retinal nerve fibre layer thickness was found.
Conclusions: ODD-AION and nODD-AION patients present with distinctly different anatomical characteristics, establishing ODD and potentially also PHOMS as independent risk factors for developing NA-AION. The role in ODD-AION pathogenesis of having a small scleral canal, and the interaction between small scleral canals and ODD, remain to be elucidated.
Disclosures: None.

P007

OUTCOME OF TREATMENT WITH IDEBENONE OF PATIENTS WITH LEBER'S HEREDITARY OPTIC NEUROPATHY IN THE NETHERLANDS BETWEEN 2017 AND 2021

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Background and Aims: Treatment for Leber's hereditary optic neuropathy (LHON) with idebenone has been widely accepted. Nationwide reports on treatment results are scarce. Our aim is to present results of treatment of Dutch LHON patients with idebenone.
Methods: Patients had a confirmed mutation and reported loss of vision. Treatment was started as soon as possible in one of the three treatment centres in the Netherlands. Patients had 3 monthly follow up until 6 months after treatment.

Results: Data from 72 patients were analysed. Mean treatment duration was 23.8 (standard deviation [SD] 14.4) months. A positive response, i.e. a clinically relevant recovery (CRR) or a clinically relevant stabilisation (CRS), occurred in 53% and 11% of the patients, respectively. The magnitude of CRR was .41 SD 1.54 logMAR. CRR of visual acuity was associated with recovery of colour discrimination. The thicknesses of both the ganglion cell complex and the retinal nerve fibre layer were irreversibly reduced.
Conclusions: Our results confirm that idebenone may help to restore or maintain visual function in LHON patients and that they have a better outcome when compared with the natural course of untreated LHON patients as reported in the literature.
Disclosures: The funding party was not involved in the analysis and evaluation of the study results. The authors declare they have no conflict of interest.

P008

GA-DOTATATE PET-CT SCAN, NON-INVASIVE DIAGNOSIS OF OPTIC NERVE SHEATH MENINGIOMA

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Background and Aims: Optic nerve sheath meningiomas (ONSM) represent one third of all primary optic nerve tumours. Magnetic resonance imaging (MRI) findings are sensitive and specific but there are some cases of difficult diagnosis which can lead to delayed treatment. Histopathology remains the gold standard but is commonly avoided because it may lead visual morbidity.
Methods: We report a case of an orbital tumour diagnosed with a non-invasive somatostatin receptor (SSTR) positron emission tomography computed tomography (PET-CT) scan.
Results: A 61-year-old woman was referred for a painless progressive loss of vision in her right eye (OD). She had no significant medical history. Her best corrected visual acuity was 20/30 OD and 20/20 in her left eye. Pupillary examination showed a right relative afferent pupillary defect. The right fundus examination revealed marked optic disc oedema. Orbital MRI with contrast showed widening of the right optic nerve with normal signal on T2 and T1-weighted images. The imaging findings were not typical of meningioma and a glioma of the optic nerve could not be formally ruled out. An 8 Ga- DOTA0-[Tyr3]-octreotate (GA-DOTATATE) PET-CT scan showed an intense radionuclide in the orbital tumour indicating high expression of somatostatin receptors.
Conclusions: Meningiomas have a high SSTR subtype 2 density and can be visualised with a radio-labelled somatostatin analogues PET-CT scan. There are three gallium-68 conjugated somatostatin analogues, all of which bind with high affinity to SSTR. In this case, GA-DOTATATE was used, offering high resolution when using PET-CT technology. This case shows that GA-DOTATATE scan is a valuable tool for a non-invasive diagnosis of ONSM.
Disclosures: None.

P009

AUTOMATED DETECTION OF HYPERREFLECTIVE ELEMENTS IN THE OUTER NUCLEAR LAYER OF THE RETINA

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Background and Aims: Hyperreflective foci (HF) are described as small intraretinal, hyperreflective dot-shaped lesions when imaged using spectral domain optical coherence tomography (SD-OCT). The development of tools to effectively identify and quantify HF is of great importance and may assist in clinical diagnosis, progression, and treatment monitoring. We aimed to evaluate whether an automated image analysis pipeline may be used on SD-OCT scans to accurately identify, quantify and visualise HF in the outer nuclear layer (ONL) of the retina.
Methods: This is a longitudinal exploratory study investigating 14 eyes of seven patients presenting with various retinal disorders.
Results: In the manually labelled data set our image processing algorithm "blob detector" found 2555 candidates for HF, correctly detecting 350 out of 391 manually labelled HF, indicating that the detector missed around 11% of HF. The accuracy of the convolutional neural network classifier was assessed by splitting the 2555 hits in the manually labelled dataset into training data (2045 hits) and testing data (510 hits). On the testing data the classifier obtained an accuracy of 96.3%, a sensitivity of 84.4% and a specificity of 97.5%. Corresponding receiver operating characteristic curves on the testing data achieved an area under the curve of .989.
Conclusions: Automated image analysis and machine learning methods can be applied to successfully quantify and visualise the presence and flux of HF in the ONL of the retina. HF are likely a non disease-specific finding. Further prospective studies are necessary to explore the dynamic changes of HF in different retinal disorders.
Disclosures: Professor Ahmed Toosy has received speaker honoraria from Biomedia, Sereno Symposia International Foundation, Bayer and meeting expenses from Biogen Idec and Novartis He was the UK PI for two clinical trials sponsored by MEDDAY pharmaceutical company (MD1003 in optic neuropathy [MS-ON – NCT02220244] and progressive MS [MS-SPI2 – NCT02220244]).

P010

OPTIC NERVE AVULSION AND TOTAL RETINAL DETACHMENT AFTER BLUNT OCULAR TRAUMA: A CASE REPORT

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Background and Aims: Avulsion of the optic nerve is a rare and severe complication following blunt ocular trauma. The optic nerve is separated from the sclera by indirect forces because of the relatively weak structure of the lamina cribrosa area. It can lead to irreversible blindness.
Methods: This report presents a case of optic nerve avulsion and retinal detachment after blunt ocular trauma.
Results: A 10-year old girl presented with sudden loss of vision after she had blunt trauma after hitting a wall whilst riding her bicycle. Her visual acuity was light perception in the right eye. Dilated fundus examination of the right eye showed peripapillary haemorrhage and vitreous haemorrhage, prominently inferiorly. B scan ultrasonography showed a hyporeflective area posterior to the the optic nerve. Examination 1 week later of the right eye revealed total retinal detachment. Magnetic resonance imaging showed that the defect on the posterior aspect of ocular bulb was an avulsion of the optic nerve.
Conclusions: Optic nerve avulsion should be considered in cases of blunt ocular trauma with severe sudden vision loss.
Disclosures: None.

P011

PSEUDO-FOSTER KENNEDY SYNDROME SECON-DARY TO NONARTERITIC ANTERIOR ISCHAEMIC OPTIC NEUROPATHY WITH BILATERAL MACULAR ATROPHY IN HYPERTENSIVE AND HEAVY SMOKERS PATIENT: A CASE REPORT

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Background and Aims: Pseudo-Foster Kennedy syndrome is characterised by unilateral optic atrophy with contralateral optic disc oedema in the absence of an intracranial mass causing compression of the optic nerve. The most common aetiology is bilateral sequential anterior ischaemic optic neuropathy (AION). Non-arteritic AION (NAION) is the usual cause.
Methods: This case report describes pseudo-Foster Kennedy syndrome due to NAION with hypertension and tobacco smoking as the risk factors.
Results: A 59-year-old man presented with sudden loss of vision in the left eye (LE) 5 months previously and then sudden loss of vision on the right eye (RE) 1 week before admission. Visual acuity was hand movement perception in the RE and 20/150 in the LE. Dilated fundus examination revealed optic disc swelling, peripapillary haemorrhage, and macular atrophy in the RE and optic disc pallor with macular atrophy in the LE. Laboratory examination revealed dyslipidaemia. Optical coherence tomography (OCT) showed retinal nerve fibre layer (RNFL) thickening in the RE and RNFL thinning in the LE. Macular OCT showed thinning of the ganglion cell layers in both eyes.
Conclusions: Complete systemic evaluation and treatment of vasculopathic risk factors are required because NAION can occur in both eyes due to uncontrolled underlying disease.
Disclosures: None.

P012

LIFESTYLE AND ENVIRONMENTAL FACTORS IN AUTOSOMAL DOMINANT OPTIC ATROPHY

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Background and Aims: Autosomal dominant optic atrophy (ADOA) shows highly variable expression both within and between families. Both genetic and environmental factors may influence the expression of the disease. We explore the relationship between cigarette and alcohol consumption and sleep patterns and mini-mental state on the disease expression.
Methods: Smoking, alcohol consumption, dietary habits and past medical history relating to cardiovascular disease was assessed by questionnaire. A validated sleep questionnaire and week-long diary and a mini mental state examination were administered. All had mutations in the OPA1 gene. Thirty-five patients (14 ADOA pedigrees) participated.
Results: There was no statistically significant difference in visual acuity in 19 current/ex-smokers ($\chi 2 = 1.39$, df = 1, p = .23), and no significant correlation between visual acuity and lifetime number of cigarettes smoked (rs = −.09; p = .6). There was a significant correlation between units of alcohol consumed per week and visual acuity (rs = −.5, p = .003). No significant difference was found in 31 patients' perceived sleep patterns; sleep diaries (12 patients) showed that those registered severely visually impaired versus partially sighted napped more in the day (55% versus 18%) and awoke more in the night (91% versus 27%). Mini-mental state scores in 14 patients were normal.
Conclusions: This study suggests that there is no statistically significant correlation between smoking, diet and general health and severity of visual loss in ADOA. However, alcohol seems to affect the expression of the disease. Large studies of genetically homogenous and clinically characterised patients are needed to explore these issues further.
Disclosures: None.

P013

A CASE OF NON-ARTERITIC ANTERIOR ISCHAEMIC OPTIC NEUROPATHY IN A PATIENT WITH IDIOPATHIC THROMBOCYTOPAENIC PURPURA

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Background and Aims: Idiopathic thrombocytopaenic purpura (ITP) is a haematological disorder characterised by severely decreased platelet count of peripheral cause. Patients may present with cutaneous and/or mucous bleeding, possibly life-threatening organ haemorrhage and have a high thrombosis risk.
Methods: We describe a case of a 45-year-old woman with a medical history of ITP who was diagnosed with non-arteritic anterior ischaemic optic neuropathy.
Results: She had been treated with eltrombopag for two years ago after had been diagnosed with ITP, which was diagnosed due to an asymptomatic low platelet count. She consulted due to decreased visual acuity in her left eye, which had severe amblyopia. Her visual acuity was 1.0 in the right eye and .1 in the left eye, with subtle dyschromatopsia and a relative afferent pupillary defect in the left eye. The left optic disc showed superior oedema without any other fundus findings. Two months later she developed a severe mesenteric thrombosis with bowel ischaemia and she needed a partial resection of her small intestine.
Conclusions: People with ITP have an important risk of critical pathology so treatment is essential. An ischaemic optic neuropathy in a patient with a haematological disorder can be the first sign of an increased risk of another life-threatening complications.
Disclosures: None.

P014

AUTOSOMAL RECESSIVE "LHON-LIKE" OPTIC NEUROPATHY

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Background and Aims: This is a case report of the first patient with a genetically confirmed autosomal recessive (ar) form of Leber's hereditary optic neuropathy (LHON) caused by a mutation in the DNAJC30 gene in Croatia. With the development of molecular diagnostics, Stenton described a homozygous mutation in the nuclear gene DNAJC30 and identified arLHON.
Methods: A case of a 23-year-old male with subacute vision loss in both eyes with central scotomas is presented. Although the clinical picture was indicative of LHON, testing of the three typical genetic mutations was negative. Suspicion of an atypical form of optic neuropathy extended the work-up to sequencing the whole exome.
Results: Sequencing of the entire exome revealed the presence of a homozygous missense variant c.152A> G (Tyr51Cys): NM_032317.3 in the DNAJC3O gene, identified as the cause of arLHON.
Conclusions: Given the newly discovered pathogenic variants responsible for arLHON, in cases of unconfirmed diagnosis and with clinical suspicion for LHON, along with testing of the whole mitochondrial genome, nuclear genes should also be included, by sequencing the entire exome.
Disclosures: None

P015

ACUTE UNILATERAL VISION LOSS IN A DIABETIC: A SURPRISING DIAGNOSIS

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Background and Aims: Atypical acute optic neuropathy can be a diagnostic challenge. In those who present with a concurrent history of diabetes mellitus (DM) and sinus disease, the concern is for invasive fungal disease. These cases need a multidisciplinary approach to ensure that investigations and management are timely to save vision.
Methods: A 60-year-old Pakistani woman presented loss of vision in her left eye to 3/60 over 5 days. She denied pain, trauma, diplopia, headache, fever, or jaw claudication. She had weight loss, sinusitis and a dry cough. Her past medical history included Type 2 DM, non-specific inflammatory arthropathy and back pain.
Results: Three months earlier she had been diagnosed with a microvascular third nerve palsy, which resolved. Computed tomography (CT) and CT angiography excluded an aneurysm but showed mild sinus disease. At the current presentation contrast-enhanced magnetic resonance imaging was performed, which showed

thickening of the left retrobulbar and intra-canalicular optic nerve. CT of the head and sinuses confirmed a parasinusitis. She underwent emergency sinus debridement, followed by intravenous glucocorticoids and oral voriconazole. Her vision improved to 6/9. Microbiology for fungal elements and acid fast bacilli remained negative. A clinical picture following systemic survey and a positive anti-nuclear cytoplasmic antibody with PR3 confirmed granulomatosis with polyangiitis. She was commenced on rituximab.

Conclusions: In an atypical acute optic neuropathy in an immunocompromised patient appropriate emergency investigation must be performed to exclude potentially fatal conditions. Clinicians should be aware that invasive fungal sinusitis and vasculitis are important rare causes in the differential diagnosis of acute optic neuropathy.

Disclosures: Mollan reports other Invex Therapeutics, other Heidelberg engineering; other from Chugai-Roche Ltd., other from Janssen, other from Allergan, other from Santen, other from Roche, other from Neurodiem. All outside the submitted work. All other authors declare no competing interests.

P016

DNAJC30-LEBER'S HEREDITARY OPTIC NEUR-OPATHY: FURTHER EXPANDING THE CLINICAL PHENOTYPE

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Background and Aims: DNAJC30 recessive mutations have been associated with Leber's hereditary optic neuropathy (LHON) and Leigh's syndrome. The majority of patients carry the c.152A>G mutation and are from Eastern Europe, pointing to a founder effect. Incomplete penetrance, male prevalence and high percentage of clinical relevant recovery (CRR) have been described.

Methods: We here report the ophthalmological phenotype of five male Italian patients, all from East European ancestry, carrying the same homozygous c.152A>G mutation. The peculiar phenotype of one case who also developed an autoimmune/demyelinating disease is discussed.

Results: The age at onset was 19.2 ± 6.3 years and the interval from loss of vision between eyes was 3.25 ± .96 weeks. Visual acuity at nadir was .02 ± .02 in the right eyes (OD) and .05 ± .05 in the left eyes (OS), and at last follow-up was .32 ± .39 OD and .45 ± .43 OS. All patients were treated with idebenone and showed CRR. Mean retinal nerve fibre layer thickness at the last visit was 49.4 ± 12.5 OD and 49.2 ± 13.8 OS. One patient presented, 4 years after LHON onset, with vertigo and ataxia. Brain magnetic resonance imaging disclosed an area postrema lesion with gadolinium enhancement and cerebrospinal fluid evaluation found oligoclonal bands. Intravenous steroids were administered with complete recovery of the neurological symptoms, regression of the brain lesion and improvement of visual function.

Conclusions: We here report the ophthalmological and neurological features of a case series of five DNAJC30-LHON patients further expanding the clinical phenotype to the possible co-occurrence of an autoimmune disease, as already reported for classical mtDNA-related LHON co-occurring with multiple sclerosis.

Disclosures: Chiara La Morgia has performed consultancies for Chiesi Farmaceutici, Regulatory Pharma Net and Thenewway srl; received speaker honoraria from Santhera Pharmaceuticals, Chiesi Farmaceutici, Regulatory Pharma Net, Thenewway srl, First Class srl and Biologix; is PI/SI for clinical trials sponsored by GenSight Biologics and Santhera Pharmaceuticals.

P017

WHERE IS THE DISC SWELLING COMING FROM? A CASE REPORT

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Background and Aims: Whilst raised intracranial pressure (ICP) is the commonest cause for bilateral optic nerve head swelling, the differential diagnosis includes systemic conditions that cause uveitis. At examination it may be challenging to differentiate uveitis from raised ICP prior to invasive investigations.
Methods: A 20-year-old male presented with right-sided painful reduced vision, associated with photophobia and hyperaemia. Slit lamp examination showed anterior chamber activity of 1+ in both eyes. A diagnosis of anterior uveitis with cystoid macular oedema was made. He was treated with high dose oral glucocorticoids.
Results: Blood tests and a chest radiograph to exclude inflammatory causes were normal. T-spot was negative. HLA-B27 antibody tests were awaited. His bilateral disc swelling worsened on cessation of the glucocorticoids, and the differential of raised ICP was considered. Magnetic resonance imaging (MRI) was non-contributory to the diagnosis. The lumbar puncture opening pressure was 21 cm cerebrospinal fluid (CSF). The CSF protein was .64 mg/mL. A final diagnosis of HLA-B27 positive uveitis was made.
Conclusions: Physicians are mindful that withdrawal of glucocorticoids can precipitate raised ICP. Herein we review the evidence for this. Interestingly, the MRI showed that the bilateral disc swelling stopped in the mid-optic nerve, and may be suggesting in retrospect that it originated from the eye and not from raised ICP.
Disclosures: None.

P018

CLUSTERING ANALYSIS WITH OPTICAL COHERENCE TOMOGRAPHY DATA IN LEBER'S HEREDITARY OPTIC NEUROPATHY PATIENTS BY NON-NEGATIVE MATRIX FACTORISATION UNSUPERVISED LEARNING TECHNIQUE

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Background and Aims: One of the still unexplained features of Leber's hereditary optic neuropathy (LHON)

is the incomplete penetrance. Standardised predictive factors are able to establish the risk of conversion among asymptomatic LHON mutation carriers, but are not yet available for the medical community. However, there are well-described subclinical optical coherence tomography (OCT) signs that are able to distinguish asymptomatic and acutely symptomatic LHON patients.
Methods: This exploratory study included patients with genetically diagnosed LHON (49 asymptomatic, 18 acutely symptomatic) patients and controls (n = 24). We analysed retrospectively data from DRI-Triton-OCT. By using the non-negative-matrix-factorisation (NMF) unsupervised learning technique we aimed to divide eyes into groups with similar OCT profiles and to identify OCT features that mostly influence this NMF-clustering.

Results: The preliminarily obtained results showed that NMF-clustering confirmed the presence of OCT-data -structure by choosing to factorise the OCT dataset into three clusters (cluster 1, n = 51; cluster 2, n = 88; cluster 3, n = 34). One of these (cluster 3) was found to include only eyes previously and clinically classified as control (41%) or asymptomatic LHON carriers (59%). Regarding this cluster (cluster 3), the first three relevant OCT features were related to macular ganglion cell layer sectors. In fact, these are mostly affected by degeneration in symptomatic LHON, whereas their average thicknesses were significantly higher with respect to the other two identified clusters, compatible with the status of LHON carrier or control.
Conclusions: These findings underline how the NMF-clustering technique could be useful to extract meaningful information from huge OCT datasets in an unsupervised way and to contribute to understanding of key aspects of LHON natural history.
Disclosures: Martina Romagnoli reports consultancies from GenSight Biologics; honoraria from Santhera Pharmaceuticals; and she is Study Coordinator for clinical trials from GenSight Biologics and Santhera Pharmaceuticals. None of these activities are related to this study.

P019

A FUNCTIONAL SURPRISE

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Background and Aims: Functional disorders are common in Neuro-ophthalmology, but a high index of suspicion for an organic disorder in every patient is critical to effective diagnosis and management.
Methods: We describe a case of a 57-year-old white Caucasian woman who presented with fluctuating visual field defects and headache. She was diagnosed initially as having functional visual loss. She presented to eye casualty 4 weeks after symptom onset and was referred to our Neuro-ophthalmology clinic with extremely poor visual acuity.
Results: She was found to have bilateral optic neuropathy and neuromyelitis optica spectrum disorder was suspected. She was found to have myelin oligodendrocyte glycoprotein antibodies (MOG-Ab) and magnetic resonance imaging of her head and orbits with contrast showed bilateral optic nerve enhancement. She received intravenous methylprednisolone followed by plasma exchange. Her visual fields and vision improved rapidly over 1 month, recovering eventually to 6/6 in both eyes at 3 months after presentation.
Conclusions: Whilst functional visual loss is common, it is important that it is a diagnosis of exclusion, and documentation of normal visual function is recommended. Screening for MOG-Ab and aquaporin 4 antibodies is mandatory in patients with atypical presentations of optic neuritis
Disclosures: None.

P020

THE PROGNOSTIC VALUE OF OCT CHARACTERISTICS IN COMPRESSIVE OPTIC NEUROP-ATHY

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Background and Aims: Anterior visual pathway meningiomas (AVPM) may affect visual acuity (VA) and visual field (VF). Our purpose was to evaluate if the ganglion cell complex (GCC) thickness has a predictive value for visual outcomes in AVPM.
Methods: Data on ophthalmological examinations and optical coherence tomography (OCT) parameters of AVPM patients were retrieved from the pre-intervention, then the 6, 12 and 24 months post intervention evaluations. Correlation was measured between visual outcome and the pre-interventional OCT parameters. Patients were also divided into two groups: those with a thin and those with a normal GCC thickness.
Results: The medical records of 186 patients were analysed. Thirty-eight patients who met the inclusion criteria were included. There was a significant correlation between the pre-interventional GCC thickness and better VF 2 years post intervention ($r = .7$, $p \leq .001$) and better VA outcome at all time points ($r = -.5$, $p \leq .04$, .005, and .03, respectively). Furthermore, mean pre-interventional retinal nerve fibre layer (RNFL) thickness correlated with better VA outcome at 1 year and 2 years post intervention. Significant differences were noted between the thin GCC and the normal GCC group regarding VA improvement 1- and 2-years post intervention.
Conclusions: OCT parameters (RNFL and GCC thicknesses) have a predictive value in AVPM. There is strong correlation between pre-interventional GCC thickness and VA outcome. Although a thin GCC is generally considered a negative prognostic factor, improvement in clinical parameters was also evident in patients with thin GCCs.
Disclosures: None.

P021

OPA-1 HEREDITARY OPTIC NEUROPATHY TRIGGERED BY ETHAMBUTOL TREATMENT

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Background and Aims: Optic neuropathies that selectively affect papillomacular bundle ganglion retinal cells often induce mitochondrial dysfunction. They may be acquired or genetic. In some cases, the exposure to a mitochondrial toxic agent (ethambutol, disulfiram, or antiretrovirals) triggers an undisclosed hereditary optic neuropathy such as Leber's hereditary optic neuropathy (LHON) or dominant optic atrophy.
Methods: This is a case report of a 32-year-old male with presumed toxic optic neuropathy due to ethambutol. Visual loss and optic disc pallor in the temporal sectors progressed over 1 year after having discontinued the drug, so the suspicion of genetic optic neuropathy arose. Nutritional and toxic causes were ruled out.
Results: His visual acuity was .2 in the right eye and .15 in the left eye. His colour vision was normal, but there were caeco-central bilateral visual field defects. The macular ganglion cell thickness was below 55 μm on spectral domain optical coherence tomography. The peripapillary retinal nerve fibre layer thickness was reduced in both temporal sectors. Unexplained macular hyperfluorescence and P-wave amplitude reduction on multifocal electroretinogram were observed. Genetic testing revealed a heterozygous OPA-1 1822–1823 deletion in exon 19. LHON mutations were not detected.
Conclusions: Dominant optic atrophy is the most frequent amongst the hereditary causes, but less reported than LHON, to be triggered by ethambutol. Both LHON and OPA-1 mutations should be investigated in a presumed toxic optic neuropathy whenever visual function does not improve after stopping the drug or it keeps worsening.
Disclosures: None.

P022

A NOVEL OPA-1 MUTATION VARIANT IN THREE MEMBERS OF ONE FAMILY

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Background and Aims: Autosomal dominant optic neuropathy (ADON) is the most common hereditary optic neuropathy. OPA-1 mutations have been reported in 90% of cases. Several mutation variants have been reported and are associated with different clinical signs.
Methods: We report the genetic findings and the phenotypic clinical characteristics of a family with an OPA-1 novel mutation variant. Molecular genetic analysis and full relevant clinical examination have been performed and reported.
Results: Three family members had molecular genetic analysis confirming that they were heterozygous for the pathogenic OPA1 mutation c.2845del p.(Asp950fs). This variant has not been reported previously.
Conclusions: We detected a novel OPA1 mutation in a family of three which has not been reported previously in the GenomAD database.
Disclosures: None.

P023

LEBER'S HEREDITARY OPTIC NEUROPATHY (LHON) CAUSED BY ATYPICAL PATHOGENIC GENETIC VARIANTS

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Background and Aims: Apart from typical mitochondrial deoxyribonucleic acid (mtDNA) mutations, Leber's hereditary optic neuropathy (LHON) may also be caused by atypical pathogenic variants in the mitochondrial genome or by autosomal recessive mutations.
Methods: In eight bilateral optic neuropathy patients without common LHON pathogenic variants, next generation sequencing of mtDNA and the whole exome was performed. All participants also underwent a full ophthalmological and neurological workup. Cell respiration, as a mitochondrial function readout, was measured in peripheral blood mononuclear cells (PBMCs) using high-resolution respirometry (Oroboros).
Results: Genetic analysis revealed five novel variants of unknown significance in six patients: 2 MT-ND5: m.13042 G > T, homoplasmy; MT-NDI:m.4130A>T, homoplasmy; MT-CYB:m.15309 T > C, homoplasmy; MT-ND4:m.11443A>C, 65% heteroplasmy; and MT-ND6:m.14598 T > C, heteroplasmy 22%. Segregation analysis of the two cousins harbouring the 13042 G > T and 15309 T > C variants were typical for maternal transmission. A proband harbouring the 15309 T > C variant showed clinical improvement. The 11443A>C mutation was a de novo change previously unconfirmed in either of the proband's mother or brother. In two patients, an autosomal recessive DNAJC30 pathogenic variant was found; one of whom showed clinical

improvement. Mitochondrial function assessment revealed lower values of complex-I-linked respiration in PBMCs isolated from five patients with newly discovered mutations, compared with PBMCs from age- and sex-matched controls.
Conclusions: In the presented series of patients with the LHON phenotype, atypical genetic variants were found that may be causative of LHON and should be considered in further LHON genetic screening.
Disclosures: None.

P024

ISOLATION OF RETINAL GANGLION CELLS IN A MODEL OF DOMINANT OPTIC ATROPHY

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Background and Aims: Melanopsin-expressing intrinsically photosensitive (ip) retinal ganglion cells (RGC) survive late in dominant optic atrophy (DOA), long after other RGC sub-types have been lost; the mechanism of which is unclear. Our purpose was to optimise a pathway for the preparation and enrichment of single RGC suspensions from mouse retina to facilitate single cell ribonucleic acid sequencing (scRNAseq) approaches to investigate this resistance.
Methods: Retinas of mice (C57/BL6J,P21, both sexes) underwent mechanical disruption or papain enzyme treatment for 20, 30, 40 or 45 minutes (n = 6 retinas/group). Fluorescence assisted cell sorting (FACS) enriched suspensions for viable RGCs (CD90[thy1.2]+,CD48-,CD57-,CD15-,DAPI-). A FACS post sort purity check, synuclein gamma (SNCG) immunocytochemistry (ICC) and quantitative reverse transcription polymerase chain reaction (RT-qPCR) for RGC specific genes provided validation. RGCs from Opa1 +/TTAG_del mice (5 months old, n = 18 mice/group) were similarly isolated for scRNAseq.
Results: Viability following papain treatment was (mean ± standard error of the mean) 60.8 ± 2.4%, 54.4 ± 4.3%, 60.2 ± 5.9%, 59.3 ± 3.3% at 20, 30, 40, and 45 minutes, respectively with no difference between timepoints (one-way-ANOVA $F[1.28, 2.56] = 1.84$, $p = .30$). Mechanical dissociation gave lower viability (10.41 ± 2.90%; t-test $p < .0001$ versus papain [30 m]). Following FACS processing, papain (30 m) suspensions showed RGC-enrichment of 78.0 ± 6.6% on post sort purity check and viability of 96.5 ± 3.2% maintained (90.1 ± 1.8%) 5 hours later. ICC showed 75.2 ± 10.6% of cells in the RGC enriched sample stained for SNCG, compared with 13.1 ± 3.1% in the remaining suspension (t-test $p < .0001$); across the same comparison, expression of RGC-specific-genes demonstrated logfold increases (RT-qPCR) of: Thy125.3 ± .9; Brn3a61.4 ± .8; and Sncg7.5 ± .9.
Conclusions: This optimised protocol has allowed for enrichment of RGCs from wild-type mice and, for the first time, from a DOA model retina for use in scRNAseq. Such investigations will contribute to a greater understanding of the disease resistance of ipRGCs, potentially opening the way for novel neuroprotective therapies for DOA.
Disclosures: None.

P025

RETROGRADE MACULOPATHY CAUSED BY OPTIC CHIASMAL COMPRESSION

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Background and Aims: This case illustrates trans-synaptic retrograde axonal degeneration using optical coherence tomography (OCT) in an advanced presentation of optic chiasmal compression in a manner previously demonstrated on histopathology.
Methods: Case report.
Results: A 57-year-old man presented with a 2 month history of bilateral, gradually progressive painless vision loss. OCT showed bilateral extensive loss of the peripapillary nerve fibre layer and inner nuclear layer cystic changes nasal to the fovea. Confrontation visual field testing revealed bitemporal visual loss consistent with chiasmal compression. Head imaging confirmed a large pituitary macroadenoma causing severe compression of the optic nerves and chiasm. The binasal macular changes we identified correspond to the compression of decussating retinal ganglion cell fibres at the optic chiasm, and mirror the histological changes demonstrated in experimental chiasmal lesions over half a century ago.
Conclusions: Modern ophthalmological imaging demonstrates what was previously only possible histologically. The presence of retrograde maculopathy in the distribution of axonal damage highlights the primary role of ganglion cell loss in this phenomenon.
Disclosures: None.

P026

A CASE OF SUCCESSFUL TREATMENT OF NON-ARTERITIC ANTERIOR ISCHAEMIC OPTIC NEUROPATHY WITH INTRAVITREAL RANIBIZUMAB

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Background and Aims: Non-arteritic anterior ischaemic optic neuropathy (NAION) is the most common, yet untreatable acute optic neuropathy in the older population. Vascular epithelial growth factor (VEGF) inhibitors have been emerging as a possible treatment option. They might diminish the vasogenic component of optic disc oedema in NAION, although their reported efficacy varies. We would like to present a case of a 56-year-old female with a 4 day history of unilateral painless visual loss, who had an altitudinal visual field defect, optic disc oedema and flame shaped haemorrhages, consistent with NAION. She was successfully treated with an intravitreal VEGF inhibitor.
Methods: The patient's visual acuity progressively diminished, while the optic disc oedema and peripapillary haemorrhages intensified. Laboratory testing and head imaging findings were not significant, excluding compressive, infective, and inflammatory causes of optic neuropathy. Despite clinical signs of possible central retinal venous occlusion, electrophysiological findings were consistent with and optic neuropathy and no blood flow impediment was seen on fluorescein angiography either.
Results: She received a single intravitreal injection of ranibizumab. Her visual function started to improve in a few days and completely normalised within 3 weeks.
Conclusions: In selected cases, ranibizumab can be a viable treatment option for NAION. Further research is necessary to establish pathogenesis of the cases in which efficacy of such treatment is expected.
Disclosures: None.

P027

POSTERIOR ISCHAEMIC OPTIC NEUROPATHY SECONDARY TO GIANT CELL ARTERITIS MISDIAGNOSED AS NORMAL TENSION GLAUCOMA

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Background and Aims: Arteritic posterior ischaemic optic neuropathy (PION) is an acute ischaemic optic neuropathy characterised by the absence of optic nerve swelling. Discriminating glaucomatous from non-glaucomatous optic neuropathy can be difficult in our daily clinical practice.
Methods: We report the case of two patients with posterior ischaemic optic neuropathy (PION) secondary to giant cell arteritis misdiagnosed as normal tension glaucoma (NTG).
Results: Case 1: A 68-year-old female with a history of cerebral ischaemic stroke and bilateral carotid artery stenosis presented with bilateral visual loss. She was diagnosed with NTG. On questioning, she revealed the presence of asthenia, weight loss and jaw claudication. Case 2: A 75-year-old male presented with bilateral vision loss. He was diagnosed with NTG. Visual field features were not typical for glaucoma. Cerebral imaging, inflammatory biomarkers and temporal artery ultrasound were indicated. For both patients, the diagnosis of giant cell arteritis was suspected. Oral steroids were prescribed with improvement of VA in both eyes.
Conclusions: Better understanding and evaluation of pseudo-glaucomatous optic neuropathy is crucial because failure to diagnose promptly arteritic PION could worsen the visual prognosis
Disclosures: None.

P028

RØNNE'S WANDERING SCOTOMA: A DIAGNOSTIC CHALLENGE IN A CASE OF OPTIC NEURITIS

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Background and Aims: Painless, sudden onset monocular altitudinal visual field loss normally suggests a vascular pathology.
Methods: We present a case where the underlying cause was optic neuritis, and briefly review the literature and learning points.
Results: A 56-year-old man had 2 days of painless right superior field loss. He had visual acuities of 6/6 in each eye and Ishihara colour vision was reduced in the right eye at 11/17. He had normal pupillary reactions but had a dense right superior altitudinal visual field defect. Funduscopy and fluorescein angiography were normal. Inflammatory, infective and autoimmune screens were negative. Magnetic resonance imaging showed T2-weighted hyperintensity and swelling of the retrobulbar right optic nerve supporting the diagnosis of optic neuritis. Over 4 weeks the altitudinal defect transformed into a relative central scotoma. After 3 months the visual deficit resolved, with the sequela of relative right macular ganglion cell loss.
Conclusions: Altitudinal field defects are uncommon in optic neuritis. The transformation of a scotoma from one shape to another in optic neuritis was first highlighted by Rønne in 1927. Preservation of visual acuity allows clear demonstration of scotoma evolution, and provides a modern example of diagnostic difficulties in a mild case.
Disclosures: None.

P029

FAVOURABLE OUTCOME OF TREATMENT OF TWO DAY OLD SPONTANEOUS RETROBULBAR HAEMATOMA

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Background and Aims: Retrobulbar haematoma (RH) is a sight-threatening condition. Although it is usully the result of orbital trauma, it sometimes occurs spontaneously. RH rapidly cause compression or ischaemia of the optic nerve, and even cause central retinal arterial occlusion. Therefore, emergency surgical decompression is prerequisite for successful treatment. Conservative treatment usually gives poor results.
Methods: Case report of spontaneous RH and its treatment.
Results: A 78-year-old male developed spontaneous RH to his right eye, 2 days before hospitalisation. The visual acuity was light perception in his right eye, the eyeball was protruded and immobile, the conjunctiva haemorrhagic, and the pupil was dilated and not responding to light. A computed tomography scan confirmed spontaneous RH. He was on antihypertensive and warfarin therapy for cardiovascular disease. A lateral cantothomy brought subjective relief. For the next 3 days he was on mannitol, corticosteroid infusions and brimonidine, which improved his vision to finger counting at 2 m. Tapering therapy was given over the next 2 weeks. On follow-up at 3 weeks later, his right eye visual acuity was 20/25, with a regular eyeball position and normal ocular motility.
Conclusions: Medical treatment of RH with compressive optic neuropathy should be attempted after surgical decompression. In conclusion, the authors give possible explanations for the favourable treatment outcome, which is not expected in such cases.
Disclosures: None.

P030

UNDISEASED INTRACRANIAL ARTERIES COMPRESSING THE ANTERIOR OPTIC PATHWAY: A CASE REPORT

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Background and Aims: Optic nerve and chiasmal compression by anatomically normal intracranial vasculature has rarely been described as a cause of visual disturbance.
Methods: We present a case report of a 46-year-old female patient with a history of migraine, who complained of headache and rapidly progressive loss of vision in her right eye (OD), which had always been her worse seeing eye.
Results: Her best corrected visual acuity (BCVA) was 20/32 OD and 20/20 in her left eye (OS). There was no relative afferent pupillary defect but her colour vision was impaired OD. Visual field testing revealed generalised loss OD and peripheral loss of sensitivity OS. Biomicroscopy, funduscopy and optical coherence tomography of the macula and retinal nerve fibre layer were unremarkable. Magnetic resonance imaging (MRI) of the brain and orbits showed three small unspecific high signal areas in the subcortical frontal matter. Infectious and autoimmune markers were negative. Visual evoked potentials and full field

electroretinography were considered normal. At 6 months follow up her BCVA had improved to 20/25 OD and her visual fields were slightly improved. A second MRI revealed stability of the high signal areas, but also revealed contact between the supraclinoid portion of the right internal carotid artery and the optic chiasm. Computed tomography angiography confirmed remodelling of the right portion of the optic chiasm by the right internal carotid artery. Also, the left anterior communicating artery was tortuous and compressed the right optic nerve, which showed T2 high signal within it.
Conclusions: Although rare, vascular optic pathway compression should be considered in the setting of unexplained visual loss
Disclosures: None.

P031

ORBITAL MRI AND ULTRASONOGRAPHY FINDINGS IN PERIPAPILLARY HYPERREFLECTIVE OVOID MASS-LIKE STRUCTURES (PHOMS)

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Background and Aims: Peripapillary hyperreflective ovoid mass-like structures (PHOMS) are optical coherence tomography (OCT) findings that occur in several optic nerve disorders. Assuming that PHOMS are thought to correspond to herniating nerve fibres or to be secondary to axoplasmic stasis, the aim of this study was to investigate orbital magnetic resonance imaging (MRI) and ultrasonography findings in patients with PHOMS.
Methods: Fifteen patients (age range: 10–54 years; five males and 10 females) with PHOMS identified by special domain OCT (Heidelberg Engineering^{TM}) according to the multirater validation of PHOMS criteria (Petzold A. et al. Neuroophthalmology, 2020), with or without optic nerve head drusen, underwent neuro-ophthalmological examination, orbital ultrasonography (Aviso Absolu Quantel^{TM}) and orbital MRI.
Results: Orbital B-scan and A-scan ultrasonography showed optic disc elevation in 13/15 patients (86.6%) and the presence of a 30° sign in 2/15 (13.3%). Increased optic nerve sheath diameter was found on T2-weighted and T1-weighted fat saturated post-contrast MRI acquired in the axial and coronal planes in 6/15 patients (40%).
Conclusions: Orbital ultrasonography and orbital MRI may confirm the association of PHOMS with dysplastic anomalies of the optic nerve head or suggest impairment of perioptic cerebrospinal fluid dynamics
Disclosures: None.

P032

PRESSURE PERFECT, BUT MULTIPLE MISSED OPPORTUNITIES ALLOWED SLOWLY PROGRESSIVE VISUAL LOSS TO GO UNDETECTED

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Background and Aims: Compressive optic neuropathy can mimic normal tension glaucomatous optic neuropathy, therefore it is important to have a high index of suspicion in any young person with painless progressive bilateral or unilateral visual loss. Neuroimaging (magnetic resonance imaging [MRI] with intravenous contrast) of the head and orbits is the first line investigation.
Methods: A 45-year-old woman, with a background of acute lymphoid leukaemia treated with radio-chemotherapy during childhood, presented with bilateral gradual painless progressive vision loss. She did-not attend two clinic visits
Results: Her visual acuity was 6/9 in each eye at the first presentation. At this time her visual fields to confrontation were noted to be normal. However, she did not attend 2 clinic visits, hence did not have formal visual field testing. She was subsequently managed as normal tension glaucoma by her optician with topical latanoprost nocte. She re-presented after a further 2 years period with visual acuities of hand movement perception in her right eye and 3/60 in her left eye. There was no relative afferent pupillary defect. Dilated fundus examination revealed moderately pale optic discs bilaterally. Optical coherence tomography confirmed bilateral optic neuropathy. Emergency MRI of her brain and orbits with intravenous gadolinium contrast found a large tuberculum sella meningioma. The neurosurgical team performed a craniotomy for tumour management. She was registered as severely sight impaired.
Conclusions: Compressive optic neuropathy can mimic normal tension glaucoma, especially in young people. The importance of a formal visual field testing cannot be overstated. Where there is community shared care of patients, clinical oversight needs to be assured to ensure that missed opportunities to diagnose and intervene are not missed.
Disclosures: None.

P033

LEBER'S HEREDITARY OPTIC NEUROPATHY IN ASSOCIATION WITH THE VERY RARE MITOCHONDRIAL DNA MUTATION 14487 T > C

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Background and Aims: Leber's hereditary optic neuropathy (LHON) is usually characterised by visual loss in young males (15 to 35-years-old) carrying one of the three primary mutations of mitochondrial deoxyribonucleic acid (mtDNA) in over 90% of cases. Rare mtDNA mutations have been reported in association with LHON. We report the second family in the literature carrying a very rare mtDNA mutation MT.14487 T > C in the ND6 gene and manifesting as LHON with childhood and late onset.
Methods: Case report.
Results: A 65-year-old healthy woman complained of bilateral subacute painless loss of vision. Her son had experienced the same symptoms at 12-years-old, with complete visual recovery after many months. Clinical examination of the proband showed a visual acuity of hand movement perception bilaterally and temporal pallor of the optic discs. Visual field testing showed central scotomas. The three primary mtDNA mutations for LHON tested negative. LHON was strongly suspected and further genetic analysis were conducted with complete sequencing of mtDNA, which revealed a rare mtDNA MT.14487 T > C ND6 mutation (90% heteroplasmy in the urine). Idebenone treatment was started.

Conclusions: The current case highlights the importance of looking for rare mtDNA mutations when the clinical suspicion is high, even at a not classical age and in females. Moreover, this is the second case of LHON reported with the MT.14487 T > C ND6 mutation which has previously been associated mainly with LHON 'plus' phenotypes.
Disclosures: None.

P034

VERY DELAYED PRESENTATION OF A GIANT NON-FUNCTIONING PITUITARY MACROADENOMA

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Background and Aims: A 65-year-old woman presented with progressive bilateral visual loss, gradually deteriorating over several years, associated with frontal headache. She had lost weight (4 stone, from 17 stone) over a few months. Her past medical history included endometrial cancer, diagnosed 8 years previously, treated with hysterectomy and radiotherapy.
Methods: Her family had become increasingly concerned and, at their insistence, she attended an optician who referred her directly to the Emergency Department. She had no perception of light vision in either eye. Her pupils were amaurotic with grade 4 optic atrophy bilaterally. The remaining ocular and neurological examination was normal.
Results: Neuro-imaging showed a giant sellar/suprasellar tumour compressing and distorting the rostral aspect of the third ventricle, midbrain and frontal lobes. She was treated with hydrocortisone and urgent extended endoscopic transsphenoidal surgery, with second stage debulking 9 months later. There was no visual improvement and she was registered severely sight impaired. While an inpatient, she was reviewed by Neuropsychology, who identified significant executive dysfunction and a slowed information processing speed. Our patient explained she had not sought medical assistance earlier due to feelings of depression and being withdrawn. Community follow up with Clinical Psychology was recommended.
Conclusions: Mood disorders, including apathy and depression, are associated with pituitary tumours. This case illustrates the extremely poor visual outcome that can result.
Disclosures: None.

P035

HOW BLOOMS THE MORNING GLORY

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Background and Aims: Since its description as a distinct optic disc anomaly by Kindler in 1970, the morning glory disc anomaly, sometimes confused with peripapillary staphyloma, however affecting all three embryological layers, has baffled investigators regarding its origin. A clear pathophysiological mechanism is put forth to explain its development.

Methods: Patients with morning glory disc anomaly and/or peripapillary staphyloma in a neuro-ophthalmology and paediatric ophthalmology practices were examined over a 20 year period, and all published case reports were reviewed. Doppler ultrasonography as well as intravenous fluorescein angiography were also performed on four consecutive patients with morning glory disc anomaly.
Results: Morning glory and peripapillary disc anomalies are distinguished by the lack of any central retinal vasculature in the former, with exclusive compensatory cilioretinal disc vasculature developing to supply the retina with blood. Much as with Nettleship circumpapillary disc collateral vessels that develop in adults with permanent occlusive emboli of the central retinal artery (e.g., due to calcific emboli emerging from the heart), such retinal perfusion does not occur sufficiently rapidly to prevent neuronal loss. In the case of the morning glory anomaly, as central disc vasculature develops in utero, transitory retinal artery occlusion, such as with fibrin emboli, would suffice to cause it.
Conclusions: Twice as often left-sided, with a female predominance, the morning glory disc anomaly can result from occlusive central retinal vessel fibrin emboli emerging from the heart, also producing "moya-moya" like anomalies elsewhere. An increased incidence of prothrombotic factors can be noted in affected individuals or their mothers.
Disclosures: None.

P036

BILATERAL NEURORETINITIS AND RETINOCHORIOIDITIS IN A PATIENT WHO HAD NEVER VISITED EXOTIC PLACES: A CASE REPORT

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Background and Aims: A 42-year-old woman in good general health was referred to our clinic for electrophysiological examination in January 2018. She had noticed bilateral, sequential decrease in her vision in September 2017. At the time, her ophthalmologists found bilateral neuroretinitis with left eye predominance.
Methods: The exact cause could not be determined. She received antibacterial and antiviral therapy and the process stopped but it healed with marked sequelae. Immunological (autoimmune) diseases were suspected but not proven in the background. For that reason she feared forthcoming new manifestations.
Results: On examination we found best corrected visual acuities (BCVA) of 1.0 and .01 in the right and left eyes, respectively. A severe visual field defect was seen in her left eye, but the right visual field was also affected, although to a lesser extent. She had atrophic and pigmented changes all over her left retina (pseudoretinitis pigmentosa), while in the right retina, similar, but less pronounced changes were seen. All possible pathogens endemic in Hungary had already been excluded. It turned out that preceding her illness she had spent some weeks on a trip in the United States of America. Laboratory examination proved positive for Rickettsia rickettsii antibodies in serum, which is the causative pathogen for Rocky Mountain spotted fever.
Conclusions: In our presentation we also give an overview of the disease itself. By presenting this case we wanted to draw attention to the importance of taking a travel history.
Disclosures: None.

P037

USE OF FACTOR ANALYSIS TO DEVELOP A MODEL FOR IDENTIFYING CHANGES IN THE OPTIC NERVE REGION USING HAND-HELD OPTICAL COHERENCE TOMOGRAPHY IN CHILDREN WITH CHILDHOOD GLAUCOMA

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Background and Aims: Optical coherence tomography (OCT) is used widely diagnostically in adult glaucoma. However, despite hand-held OCT being available, its use in childhood glaucoma is limited. Our aim was to generate a model based on optic nerve and circumpapillary retinal nerve fibre layer (cpRNFL) parameters for clinical use in childhood glaucoma.
Methods: Hand-held OCT volumetric images were acquired from 28 children with primary glaucoma, 39 with secondary glaucoma, and 77 age-matched controls. Factor analysis was used to identify the contribution of parameters to glaucomatous changes or order to generate a binary logistic regression model.
Results: Factor analysis revealed two main clusters consisting of: (i) cpRNFL and rim parameters and (ii) cup and cup-to-disc ratio parameters. Using the two principal factors to generate a binary logistic regression model resulted in a receiver operating characteristic for detecting childhood glaucoma where the area under the curve (AUC) was .951 (lower and upper confidence intervals [CI]: .915, .987) for primary and .918 (CI: .861, .976) for secondary glaucoma. A similar result was obtained by using two representative parameters from each cluster (inferior quadrant of cpRNFL and cup area, where the AUC was .945 [CI: .903, .987] and .926 [CI: .883, .969], respectively).
Conclusions: The model suggests two distinct grouping of parameters, possibly because of different stages of disease progression. Combining the parameters derived from the optic nerve using hand-held OCT imaging into a simple model has important diagnostic potential for detection and monitoring of disease progression in childhood glaucoma.
Disclosures: Frank A. Proudlock has a commercial agreement with Leica Microsystems.

P038

UNILATERAL RADIATION PAPILLOPATHY FOLLOWING RADICAL TREATMENT FOR A HIGH-GRADE, UNDIFFERENTIATED INVASIVE PARANASAL SINUS SKULL BASE TUMOUR

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Background and Aims: A 43-year-old man, who was myopic, congenitally colour blind, previously well and a non-smoker, presented with a 4 week history of dizziness and headache, associated with an episode of vomiting. His wife reported that he had lost his sense of smell previously, without seeking medical attention.
Methods: Magnetic resonance imaging (MRI) showed a large nasofrontal enhancing skull-base mass, involving the nasal cavity, ethmoidal sinuses and bifrontal regions. He was commenced high-dose steroids. Whole body computed tomography was unremarkable. One month later, another MRI showed progression

into the left frontal sinus. Funduscopy showed haemorrhagic papilloedema. He underwent urgent skull-base neurosurgery. Histology showed a high-grade, malignant, undifferentiated tumour. The papilloedema resolved following surgery.
Results: Shortly after surgery, his visual acuity was 6/9 in his right eye (OD) and 6/6 in his left eye (OS). He had weekly cisplatin chemotherapy and 65 Gy radiotherapy (30 fractions). Neuroimaging showed that the meningeal enhancement had resolved but there were extensive post-treatment gliotic changes in the frontal poles. He developed diabetes mellitus secondary to the prolonged high-dose steroids. Two years after treatment was completed, he developed blurred vision: 6/9 OD and 6/24 OS. Funduscopy showed localised haemorrhagic disc swelling with exudates but no generalised retinopathy. His visual acuity worsened to 6/18 OD and 1/60 OS. Radiation papillopathy was diagnosed.
Conclusions: Blood tests (C-reactive protein, erythrocyte sedimentation rate, angiotensin converting enzyme, tuberculosis, Borrelia and syphilis) were normal or negative. MRI showed stable olfactory fossae cystic change extending to the left frontal lobe and ill-defined periventricular enhancement, suggestive of radiation necrosis. There was no enhancement of the anterior visual pathways. Intravitreal Avastin was trialled unsuccessfully.
Disclosures: None.

P039

LONG-TERM IDEBENONE TREATMENT CAN PROMOTE FAVOURABLE VISUAL ACUITY OUTCOMES IN PATIENTS WITH LEBER'S HEREDITARY OPTIC NEUROPATHY: RESULTS FROM THE PROSPECTIVE, NATURAL HISTORY-CONTROLLED LEROS STUDY

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Background and Aims: The LEROS study met its primary endpoint: long-term idebenone treatment resulted in clinically relevant benefit to patients with Leber's hereditary optic neuropathy (LHON). Here, we report on additional LEROS outcomes – clinically relevant worsening (CRW) of visual acuity (VA), and the transition between clinically relevant VA categories over time.
Methods: Patients with LHON treated for up to 24 months (n = 181) were stratified (subacute/dynamic [≤ 1 year from onset]; chronic [> 1 year]) and compared with an external natural history (NH) cohort (n = 372), matched by time since onset. CRW was defined as the loss of 2 logMAR chart lines and/or a worsening to off-chart.
Results: In treated subacute/dynamic and chronic patients, CRW was less frequent than in matched NH patients at 12 months (subacute/dynamic: 35.9% [23/64] vs 64.6% [53/82]; chronic: 6.3% [3/48] versus 18.6% [11/59]) and 24 months (subacute/dynamic: 33.3% [19/57] versus 57.6% [19/33]; chronic: 4.9% [2/41] versus 21.6% [8/37]). At baseline, 66.0% of treated subacute/dynamic patients had a best VA ≥ 1.0 logMAR. This increased at 6 months (76.7%) and decreased thereafter (57.1% at 24 months). At baseline, 70.1% of chronic patients had a best VA ≥ 1.0 logMAR (34.5% off chart) with a decrease thereafter (58.2% at 24 months; 16.4% off chart).
Conclusions: In LEROS, long-term idebenone treatment reduced worsening of VA. In addition, idebenone reduced the proportion of patients with a best VA that was ≥ 1.0 logMAR – the threshold of 'legal blindness' in many countries – or a best VA that was off chart.
Disclosures: LT and XL are employees of Chiesi Farmaceutici SpA. TK has received research support and personal fees from Santhera Pharmaceuticals, GenSight Biologics and Chiesi Farmaceutici.

P040

LONG-TERM EFFICACY AND SAFETY OF IDEBENONE IN PATIENTS WITH LHON IN THE SUBACUTE/DYNAMIC PHASE: RESULTS FROM THE PROSPECTIVE, NATURAL HISTORY-CONTROLLED LEROS STUDY

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Background and Aims: Idebenone is approved in Europe for the treatment of Leber's hereditary optic neuropathy (LHON) – a rare mitochondrial disorder resulting in severe, bilateral vision loss. Controlled data beyond a 6-month treatment duration are lacking. Herein, we report primary long-term results from the LEROS study.
Methods: Patients with LHON onset ≤ 5 years prior were enrolled and stratified by time since onset: subacute/dynamic (≤ 1 year) and chronic (> 1 year). Data from 181 patients treated up to 24 months were compared with an external natural history (NH) cohort (372 patients), matched by time since onset.
Results: The primary endpoint – the proportion of subacute/dynamic eyes with a clinically relevant benefit (CRB) from baseline following 12 months of treatment, versus matched NH eyes – was met (42.3% [60/142] versus 20.7% [40/193] [p = .002]). This difference was maintained after 24 months (52.9% [64/121] versus 36.0% [27/75] [p = .0297]). In treated subacute/dynamic patients, the median best visual acuity (VA) at baseline was 1.28 logMAR (n = 109). An initial worsening was observed at 6 months to 1.41 logMAR (n = 90), followed by a recovery to 1.30 (n = 81), 1.20 (n = 75) and 1.07 (n = 70) logMAR at 12, 18 and 24 months, respectively.
Conclusions: LEROS corroborates the outcomes of previous studies, demonstrating that long-term treatment with idebenone is safe and results in prolonged clinical benefit in patients with LHON in the subacute /dynamic phase.
Disclosures: TK has received research support and personal fees from Santhera Pharmaceuticals, GenSight Biologics and Chiesi Farmaceutici. XL and LT are employees of Chiesi Farmaceutici SpA.

P041

LONG-TERM EFFICACY AND SAFETY OF IDEBENONE IN PATIENTS WITH LHON IN THE CHRONIC PHASE: RESULTS FROM THE PROSPECTIVE, NATURAL HISTORY-CONTROLLED LEROS STUDY

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Background and Aims: Idebenone is approved in Europe for the treatment of Leber's hereditary optic neuropathy (LHON). Controlled data detailing treatment in chronic patients are sparse. Herein, we report long-term results from the LEROS study in patients with chronic LHON (1 to ≤ 5 years since onset).
Methods: Patients with LHON onset ≤ 5 years prior were enrolled and stratified by time since onset: subacute/dynamic (≤ 1 year) and chronic (> 1 year). Data from 181 patients treated up to 24 months were compared with an external natural history (NH) cohort (372 patients), matched by time since onset.
Results: As was observed in the subacute/dynamic phase, the frequency of a clinically relevant benefit (CRB) from baseline at 12 months was significantly higher in treated, chronic eyes than in matched NH eyes (50.3% [72/143] versus 38.6% [59/153] [p = .0087]). This difference was largely driven by a higher

proportion of eyes with a clinically relevant recovery of visual acuity (VA) (32.9% [47/143] versus 19.6% [30/153] [p = .0034]). The median best VA at baseline was 1.48 logMAR (n = 87) and improved to 1.32 (n = 81), 1.23 (n = 70), 1.26 (n = 66) and 1.16 (n = 55) logMAR at 6, 12, 18 and 24 months, respectively.
Conclusions: LEROS provides evidence of a significant therapeutic benefit of idebenone treatment in patients with LHON in the chronic stage, a group for whom viable treatment options are otherwise severely limited.
Disclosures: XL and LT are employees of Chiesi Farmaceutici SpA. TK has received research support and personal fees from Santhera Pharmaceuticals, GenSight Biologics and Chiesi Farmaceutici.

P042

UNILATERAL OPTIC NEURITIS IN A 5-YEAR-OLD GIRL WITH POSITIVE ANTI-MOG ANTIBODIES

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Background and Aims: Optic neuritis (ON) is a common presenting symptom in paediatric demyelinating disorders and may be associated with dramatic visual loss.
Methods: We describe the case of a healthy 5-year-old girl with a left eye (LE) ON with positive anti-myelin oligodendrocyte glycoprotein (anti-MOG) antibodies.
Results: A 5-year-old girl presented to the emergency room with left eye vision loss. At ophthalmological evaluation, the LE visual acuity was no light perception with a left afferent pupillary defect and papillitis on funduscopy. The examination of the right eye was unremarkable. No relevant signs were found on the cerebral and orbital computed tomography scan. A left ON was suspected so intravenous corticosteroid treatment was introduced, and the patient was hospitalised for aetiological study. Cerebrospinal fluid and magnetic resonance imaging were normal. Serum anti-MOG antibodies were positive. After 5 days of methylprednisolone, the left eye vision improved to counting fingers and the patient was discharged on oral prednisolone.
Conclusions: Paediatric ON is a rare disorder, which may occur as an isolated syndrome, or in association with diffuse neurological dysfunction, and is frequently associated with anti-MOG antibodies. Formal neuro-ophthalmological evaluation and follow-up are essential due to the possibility of relapsing disease and the development of other central nervous system lesions.
Disclosures: None.

P043

PSYCHOMETRIC VALIDITY OF THE VISUAL FUNCTION INDEX (VF-14) IN LEBER'S HEREDITARY OPTIC NEUROPATHY

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Background and Aims: The visual function index (VF-14) is a widely used patient-reported outcome measure for assessing vision-related activity limitation in individuals with a variety of ophthalmological conditions. The psychometric validity of the VF-14 for use by individuals affected by Leber's hereditary optic neuropathy (LHON) has not been reported previously. The purpose of this study was to determine the psychometric validity of the VF 14, when used by individuals affected by LHON, by Rasch analysis.
Methods: Rasch analysis was conducted using VF-14 data for 196 individuals carrying one of the three primary LHON mutations and affected by vision loss.
Results: Issues identified with the VF-14 included disordered response thresholds (12/14 items), local dependency (10/91 pairwise dependencies), and evidence of multi-dimensionality. However, the distribution of person estimates and item thresholds were fairly well matched; only one item (difficulty performing sports activities) showed misfit to the Rasch model, and there was good reliability (Person Separation Index .84). Rasch-informed VF-14 revisions included: removing both driving items and the misfitting sports item; rescoring response options across all items by merging the response categories of "a little" and "moderate amount"; and accounting for the dependency between two reading items.
Conclusions: Clinicians and researchers using the VF-14 with LHON patients should be aware of the limitations of the original version of the VF-14. Compared with the original version, the proposed Rasch-based structure of the revised VF-14 appears to offer improved psychometric performance and interpretation of vision-related activity limitation.
Disclosures: BSC is recipient of the Cambridge-Rutherford Memorial Scholarship awarded by the Royal Society Te Apārangi – Rutherford Foundation and the Cambridge Commonwealth, European & International Trust, and also the Aotearoa New Zealand Fellows Research Entry Scholarship awarded by the Royal Australasian College of Physicians (RACP). PYWM is supported by a Clinician Scientist Fellowship Award (G1002570) from the Medical Research Council (UK), and also receives funding from Fight for Sight (UK), the Isaac Newton Trust (UK), Moorfields Eye Charity, the Addenbrooke's Charitable Trust, the National Eye Research Centre (UK), the International Foundation for Optic Nerve Disease (IFOND), the UK National Institute of Health Research (NIHR) as part of the Rare Diseases Translational Research Collaboration, the NIHR Cambridge Biomedical Research Centre (BRC-1215-20014), and the NIHR Biomedical Research Centre based at Moorfields Eye Hospital NHS Foundation Trust and UCL Institute of Ophthalmology. The views expressed are those of the author(s) and not necessarily those of the NHS, the NIHR or the Department of Health.

P044

CAPTURING THE EXPERIENCES OF PATIENTS WITH INHERITED OPTIC NEUROPATHIES: A SYSTEMATIC REVIEW OF PATIENT-REPORTED OUTCOME MEASURES (PROMS) AND QUALITATIVE STUDIES

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Background and Aims: Patient-reported outcome measures (PROMs) are questionnaires that enable patients to report their experiences, and are frequently used in clinical trials to assess quality of life (QoL). It is unclear if the current PROMs used by individuals affected by inherited optic neuropathies (IONs) are suitable for use in this population. Our aim was to identify and comprehensively evaluate studies capturing the experience of individuals affected by an ION, focusing on PROMs and qualitative studies where the health status and QoL of these individuals have been explored.
Aims and Methods: We carried out a systematic review of MEDLINE, EMBASE, PsycINFO, CINAHL Plus, and Scopus databases.
Results: Out of 1326 unique articles identified, six studies were included. Five PROMs were identified: visual function index (VF-14); hospital anxiety and depression scale (HADS); a novel graphical online assessment tool (NGOAT) for reporting emotional response to vision loss; a new PROM informed by the DSM-V criteria for major depressive disorder; and an interpersonal and career 'impact rating' PROM. The psychometric performance of included PROMs were poorly described. Qualitative studies found that vision loss resulted in psychosocial losses, including loss of social and communication skills and loss of independence and freedom. Factors that modified the response to vision loss were also identified.
Conclusions: The current PROMs used by individuals with ION have poor content coverage, primarily measuring activity limitation and emotional well-being, and insufficient reporting of psychometric performance. There is a need to develop a PROM for individuals with IONs to report their experiences of living with their condition.
Disclosures: BSC is recipient of the Cambridge-Rutherford Memorial Scholarship awarded by the Royal Society Te Apārangi – Rutherford Foundation and the Cambridge Commonwealth, European & International Trust, and also the Aotearoa New Zealand Fellows Research Entry Scholarship awarded by the Royal Australasian College of Physicians (RACP). PYWM is supported by a Clinician Scientist Fellowship Award (G1002570) from the Medical Research Council (UK), and also receives funding from Fight for Sight (UK), the Isaac Newton Trust (UK), Moorfields Eye Charity, the Addenbrooke's Charitable Trust, the National Eye Research Centre (UK), the International Foundation for Optic Nerve Disease (IFOND), the UK National Institute of Health Research (NIHR) as part of the Rare Diseases Translational Research Collaboration, the NIHR Cambridge Biomedical Research Centre (BRC-1215-20014), and the NIHR Biomedical Research Centre based at Moorfields Eye Hospital NHS Foundation Trust and UCL Institute of Ophthalmology. The views expressed are those of the author(s) and not necessarily those of the NHS, the NIHR or the Department of Health.

P045

PRESUMED DUPILUMAB-INDUCED OPTIC NEURITIS: A CASE REPORT

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Background and Aims: Dupilumab is the first biological drug approved for moderate-to-severe atopic dermatitis treatment. It is a fully human monoclonal antibody against interleukin (IL)-4Rα that blocks IL-4/IL-13 signalling, inhibiting the janus tyrosine kinase – signal transducer and activator of transcription pathway. It has been considered a safe treatment for long-term use. Self-limited ocular surface pathology is the most frequent described adverse event.
Methods: We report a case of presumed dupilumab-induced optic neuritis.

Results: A 19-year-old female was evaluated for progressive vision loss, associated with intense photophobia and frontal headache. She had atopic dermatitis and had been treated with dupilumab for 6 weeks. Ophthalmological examination revealed a best corrected visual acuity of .5 in the right eye and .4 in the left eye. Ishihara colour vision was abnormal. Biomicroscopy and funduscopy were normal. Analytic and immunological studies were normal. Brain and orbital magnetic resonance imaging revealed focal enhancement of the right optic nerve sheath in the pre-canalicular segment. As the onset of visual complaints coincided with dupilumab prescription for atopic dermatitis, dupilumab withdrawal was decided, with gradual visual recovery.
Conclusions: We report a clinical case of a patient treated with dupilumab who developed bilateral optic neuritis.
Disclosures: None.

P046

SEPTO-OPTIC DYSPLASIA PRESENTING WITH READING DIFFICULTY

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Background and Aims: Septo-optic dysplasia (SOD) is classically described as presenting in childhood with a triad of optic nerve hypoplasia, midline brain abnormalities and pituitary dysfunction. More recently, it has been recognised that SOD may present in adults, usually with epilepsy, and other cerebral abnormalities.
Methods: We report SOD presenting in two adults with reading difficulty and asymptomatic severe visual field constriction.
Results: A 46-year-old man (visual acuities 6/6 in each eye) and a 32-year-old woman (visual acuities 6/7.5 in the right eye and 6/6 in the left eye) were independently referred with unexplained severe visual field constriction and evidence of 'optic atrophy' on optical coherence tomography imaging of their optic discs. Neither of them was aware of any problems with their peripheral vision, and both reported relief of their presenting symptom, reading difficulty, with the appropriate glasses. Brain magnetic resonance imaging revealed abnormalities consistent with SOD. In retrospect, their 'optic atrophy' was optic nerve hypoplasia, although the typical optic disc features of hypoplasia were not apparent.
Conclusions: SOD may present in adult life with apparent optic atrophy and severe visual field loss. The subjects' lack of awareness of their visual field constriction is consistent with a congenital abnormality. There are a lack of data on the range of visual field defects associated with SOD.
Disclosures: None.

P047

ACUTE AND CHRONIC INNER NUCLEAR MICROCYSTIC CHANGE IN THREE PATIENTS WITH NON-ARTERITIC ANTERIOR ISCHAEMIC OPTIC NEUROPATHY: A RETROSPECTIVE CASE SERIES.

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Background and Aims: The term microcystic macular oedema (MMO) was introduced to describe a change seen on optical coherence tomography (OCT) in multiple sclerosis. A similar change is associated with severe optic atrophy and the term "retrograde maculopathy" (RM) has been proposed. Similar cystic change has been earlier described in primary *retinal* pathology.
Methods: A retrospective observational case series was carried out at the University Hospital of Liège. Thirty-four cases (mean age: 60 ± 12.5 years) referred to the Neuro-ophthalmology with non-arteritic anterior ischaemic optic neuropathy (NA-AION) from 2014 to 2021 were reviewed. Standard ophthalmological tests had been carried out, including OCT.
Results: In 19 eyes an acute transient microcystic change in the peripapillary inner nuclear layer (INL) was observed on OCT associated with optic disc swelling, in some cases extending to the macula. This finding was associated with intra- and sub-retinal fluid originating from the optic disc. Subsequently, three of the 19 eyes developed 'classical' MMO/RM, which was restricted to the region of severe ganglion cell complex (GCC) thinning. The mean GCC thinning in the superior hemi-macula was −28.2 ± 5.2 μm (range: −22.3 to −30.3 μm; −33.3%) and in the inferior hemi-macula was −30.7 ± 5.6 μm (range: −24.3 to −34.8 μm; −31.0%). Peripapillary retinal nerve fibre layer (RNFL) thinning was also observed at 6 months (mean RNFL: 57.7 ± 2.5 μm).
Conclusions: We have observed two phenomena in which there is microcystic change in the INL in NA-AION, one reversible and the other long-standing, likely permanent. We propose a distinction between genuine tissue oedema of the INL and RM related to RNFL loss. The pathogenesis of RM, and why it is restricted to the macula will be discussed.
Disclosures: None.

DISORDERS OF THE POSTERIOR VISUAL PATHWAY AND VISUAL PROCESSING

P048

CEREBRAL MALARIA: INSIGHT INTO PATHOLOGY FROM OPTICAL COHERENCE TOMOGRAPHY

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Background and Aims: Clinical signs of malarial retinopathy (MR) help with diagnosing cerebral malaria (CM) and predicting outcome severity. This is the first study of hand-held optical coherence tomography (HH-OCT) in CM describing detailed in vivo retinal changes in MR to assess HH-OCT's diagnostic potential.
Methods: Children with MR (n = 43) underwent ophthalmoscopy, fluorescein angiography and HH-OCT during admission, then 1 month (n = 31) and 1 year (n = 8) post-discharge. Controls were comatose patients without malaria (n = 6) and age/sex-matched healthy children (n = 43). OCT changes and retinal layer thicknesses were compared.
Results: On HH-OCT, hyper-reflective areas (HRAs) were seen in the inner retina of 81% of the MR patients, corresponding to ischaemic retinal whitening on fundus photography. Cotton wool spots were present in 37% and abnormal hyper-reflective dots, co-localised to capillary plexuses, in 93%. Hyper-

reflective vessel walls were present in 84%, and intra-retinal cysts in 9% of cases. Vascular changes and cysts resolved within 48 h. HRAs developed into retinal thinning at 1 month (p = .027), which was more pronounced after 1 year (p = .009).
Conclusions: Vascular hyper-reflectivity may represent the sequestration of parasitised erythrocytes in vessels, a key CM feature. The mechanisms of post-ischaemic retinal atrophy and cerebral atrophy with cognitive impairment may be similar in CM survivors. HH-OCT has the potential for monitoring patients, treatment response and predicting neurological deficits.
Disclosures: None.

P049

THE UTILITY OF OPTICAL COHERENCE TOMOGRAPHY FOR DETECTING COMPRESSIVE CHIASMAL DISEASE

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Background and Aims: Sellar tumours may cause visual symptoms, including bitemporal hemianopia, but often their detection is incidental. Therefore, early diagnosis and treatment are crucial to preserve vision. Optical coherence tomography (OCT) is a potential tool to detect subclinical visual loss in this context. We aimed to evaluate the utility of OCT in detecting compressive chiasmal disease.
Methods: We retrospectively analysed patients with sellar tumours who underwent clinical, perimetric and OCT assessment. Chiasmal visual impairment was search using these three methods. Clinical and visual data were collected. Sixteen healthy individuals were used as controls.
Results: We included 41 patients (mean age, 57 ± 17 years; 63.4% females). Pituitary adenoma was the most frequent tumour (63.4%). Among patients with evidence of optic chiasmal compression on magnetic resonance imaging before surgery (n = 28), 18 (64.3%) showed pre- (7; 25%), per- (14; 50%), and/or post- (3; 10.7%) chiasmal visual involvement. Among these, OCT was abnormal in 16 (57.1%), being the most sensitive method to detect visual impairment (compared with perimetry [56%] and clinical evaluation [48%]). Using an adapted version of the normalised asymmetry score (aNAS, ratio between inferior temporal and nasal ganglion cell layer thickness) from the OCT data, we identified five additional patients with visual impairment (OCT sensitivity = 75%). Among other parameters, aNAS correlated with tumour size (R_2 = .598, p = .011).
Conclusions: OCT is a promising tool for detecting compressive chiasmal disease-related visual impairment and seems to supersede clinical and perimetric assessment in detecting early subclinical visual impairment in those patients. Customised analysis may further increase OCT sensitivity.
Disclosures: None.

P050

CONTRAST RELATED CORTICAL BLINDNESS FOLLOWING TRANSCATHETER AORTIC VALVE IMPLANTATION PROCEDURE: A CASE REPORT AND REVIEW OF THE LITERATURE

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Background and Aims: Transient cortical blindness (TCB) is an unusual, albeit fairly well recognised, complication that may arise following intravenous contrast agent use in angiography procedures. Amongst many hypotheses, the most popular pathophysiological process seems to be penetration of contrast through the blood brain barrier causing selective neurotoxicity in the occipital cortex.
Methods: In this report we describe an 84-year-old male patient diagnosed to have severe symptomatic aortic stenosis, who underwent a successful and uncomplicated transcatheter aortic valve implantation procedure. The contrast agent used was Iopamidol, which is a non-ionic and iodinated contrast agent.
Results: His relevant medical history also included hypertension on treatment and no history of any anaemia reported. No significant blood loss was noted during procedure, other medical observations remained normal. Six hours post-procedure, he developed gradual blurring of vision, progressing to perception of light only. He also complained of a vague headache and nausea. No other significant positive findings were noted intra-ocularly, and he had no other neurological deficits. Subjectively, his vision recovered back to normal over the next 4 days. The differential diagnosis at the time included cortical stroke (embolic/thrombotic phenomenon), posterior ischaemic optic neuropathy and contrast-related toxicity.
Conclusions: This was a distressing complication for the patient, whilst undergoing an unrelated and reported safe procedure. There is no established means of prevention or targeted treatment for this rare event. Hence, further data and research are required to better our understanding/management of TCB, in the event of contrast neurotoxicity.
Disclosures: None.

P051

RETINAL STRUCTURE AND FUNCTION STUDIED WITH ELECTRORETINOGRAM IN SPINOCEREBELLAR ATAXIA TYPE 3

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Background and Aims: Spinocerebellar ataxia type 3 (SCA3) is an autosomal dominant neurodegenerative disorder caused by expansion of a polyglutamine (polyQ)-encoding CAG repeat in the ATXN3 gene. The expanded ATXN3 (CAG)n size correlates directly with disease severity and progression, and indirectly with the age at onset. Because the ATXN3 protein regulates photoreceptor ciliogenesis and phagocytosis, we aimed to explore whether expanded polyQ ATXN3 impacts in retinal function and integrity in SCA3 patients.
Methods: We evaluated the retinal structure and function in five patients with SCA3 using electroretinogram (ERG). The neurological examination obeyed standard criteria for SCAs, and disease severity was classified according to the scale for the assessment and rating of ataxia (SARA) and the disease stage. Each participant underwent standard ophthalmological examination, spectral-domain optical coherence tomography (OCT, multifocal ERG and full-field flash ERG.
Results: Electrophysiological dysfunction of cones, rods and inner retinal cells was detected. Patients showed mild retinal thinning and decreased/delayed ERG photoreceptors and other retinal cells responses related to disease duration and severity. OCT analysis of these five patients revealed: i) reduced central macular thickness indirectly correlated with disease duration; ii) decreased thickness of the macula and the ganglion cell layer, and reduced macular volume inversely correlated with the SARA score.
Conclusions: Our results suggest that several structural and functional retinal deficiencies measured with ERG could represent a biological marker of disease timely evolution and severity.

Disclosures: Sanchez-Dalmau B: received compensation for consulting services and speaker honoraria from Chiesi and Sanofi-Genzyme and holds equity shares of Accure Therapeutics S.L. All other authors have nothing to disclose.

P052

PROGRESSION OF RETINAL GANGLION CELL COMPLEX AND RETINAL NERVE FIBRE LAYER IN SPINOCEREBELLAR ATAXIA-3 PATIENTS

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Background and Aims: The central nervous system affects of many diseases can be studied by imaging fundus structures with optical coherence tomography (OCT), a harmless technique that can easily report quantitative measures of eye tissue layers such as the retinal nerve fibre layer (RNFL) and ganglion cell complex (GCC). We present a retrospective cohort study of spinocerebellar ataxia-3 (SCA-3) cases to study quantitative RNFL and GCC changes with time.
Methods: We conducted a retrospective study in 17 patients with clinically and genetically confirmed SCA-3. Each patient's clinical history was retrospectively reviewed until the most recent ophthalmological evaluation (recruitment visit). A complete ophthalmological examination and spectral-domain OCT were performed in all patients to measure the peripapillary RNFL thickness, central macular thickness (CMT), macular volume and macular GCC thickness.
Results: Seventeen SCA-3 cases (34 eyes) were included. The mean age at disease onset was 40.2 years, with a mean disease duration of 9.9 years and an average follow-up of 44.9 months. Regarding OCT measures, mean CMT decreased as well as mean macular volume and average GCC thickness. By contrast, no statistically significant change was reported in overall RNFL thickness.
Conclusions: Advanced analysis of OCT images of the eye has become as a new tool to correlate clinical features of SCA-3 patients with objective anatomical measurements. Therefore, OCT-based quantification of retinal layers could be considered as disease biomarkers of great importance given the reported relationship with SCA-3 clinical features.
Disclosures: Sanchez-Dalmau B: received compensation for consulting services and speaker honoraria from Chiesi and Sanofi-Genzyme and holds equity shares of Accure Therapeutics S.L. All other authors have nothing to disclose.

P053

ORAL TERIFLUNOMIDE EFFECT ON VISUAL FUNCTION IN MULTIPLE SCLEROSIS: MULTI-FOCAL ELECTRORETINOGRAM RESPONSES AFTER 12 MONTHS OF TREATMENT

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Background and Aims: Development of predictive biomarkers of response to therapy is a priority for therapeutic optimisation in multiple sclerosis (MS). The retina represents an ideal model to investigate effects of therapy using non-invasive technologies such as multifocal electroretinography (mfERG) and

optical coherence tomography (OCT). In this study, we evaluated the change of first kernel mfERG responses in patients with relapsing-remitting MS (RRMS) after 12 months of starting teriflunomide treatment.

Methods: We conducted a prospective study in RRMS patients starting treatment with teriflunomide. Patients were assessed at baseline and after 12 months. Exploration consisted of 2.5% low contrast letter acuity, OCT, mfERG and expanded disability status scale. mfERG first kernel responses included N1 wave amplitude and peak time as well as P1 wave amplitude and peak time. Results were reported as mfERG sum response and sectorial responses by concentric rings (1–5).

Results: Twenty-four mildly disabled subjects were included by March 2018 and completed 1 year of follow-up. mfERG first kernel responses changed over 12 months showing a mean amplitude increase in N1 and P1 as well as a decreased peak time in N1 wave. Considering sectorial (ring) responses, significant increases were found in N1 amplitude (ring 5) and decreases in peak time (ring 1, 3 and 4) as well as increases in P1 amplitude (ring 3, 4 and 5).

Conclusions: In RRMS patients after 12 months of teriflunomide oral treatment we observed a significant improvement in retinal function, as revealed by increased wave amplitudes as well as decreased latencies.

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IDIOPATHIC INTRACRANIAL HYPERTENSION

P054

OBSTRUCTIVE SLEEP APNOEA IN WOMEN WITH IDIOPATHIC INTRACRANIAL HYPERTENSION: A SUB-STUDY OF THE IDIOPATHIC INTRACRANIAL HYPERTENSION WEIGHT RANDOMISED CONTROLLED TRIAL (IIH:WT)

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Background and Aims: Obesity is a risk factor for idiopathic intracranial hypertension (IIH) and obstructive sleep apnoea (OSA). We aimed to determine the prevalence of OSA in IIH and evaluate the diagnostic performance of OSA screening tools in IIH. Additionally, we evaluated the relationship between weight loss, OSA and IIH over 12 months.

Methods: This was a sub-study of a multi-centre, randomised controlled parallel group trial comparing the impact of bariatric surgery versus community weight management intervention (CWI) in IIH (IIH:WT). OSA was assessed using home-based polygraphy at baseline and 12 months. OSA was defined as an apnoea-hypopnoea index (AHI) ≥ 15 or ≥ 5 with excessive daytime sleepiness.

Results: Of the 66 women in the IIH:WT trial, 46 were included in the OSA sub-study. OSA prevalence was 47% (n = 19). The STOP-BANG had the highest sensitivity (84%) compared with the Epworth sleepiness scale (69%) and Berlin (68%) to detect OSA. Bariatric surgery resulted in greater reductions in AHI versus CWI (median [95% confidence intervals] AHI reduction of −2.8 [−11.9, .7], p = .017). Over 12 months there was a positive association between changes in papilloedema and AHI (r = .543, p = .045), despite adjustment for changes in body mass index (R^2 = .522, p = .017).
Conclusions: OSA is common in IIH and the STOP-BANG questionnaire appears to be the most sensitive screening tool. Bariatric surgery improved OSA in patients with IIH. The improvement in AHI was associated with improvement in papilloedema independent of weight loss. Whether OSA treatment has beneficial impact on papilloedema warrants further evaluation.
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P055

OBESITY IN FEMALE RATS DOES NOT CAUSE IDIOPATHIC INTRACRANIAL HYPERTENSION – ARE OBESITY-INDUCED SECONDARY FACTORS REQUIRED?

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Background and Aims: Idiopathic intracranial hypertension (IIH) is a condition characterised by increased intracranial pressure (ICP), impaired vision and headache, but with an unresolved aetiology. IIH occurs predominantly in obese (body mass index [BMI] ≥ 30 kg/m^2) women of childbearing age, although age, BMI and female sex do not encompass all aspects of IIH pathophysiology. Female IIH patients show a distinct hormonal profile, highlighted by androgen excess. To reveal the aetiology of IIH, we modelled female obesity in rats.
Methods: Rats genetically lacking the leptin receptor, Zucker rats, were grown to obesity (mimicking human BMI [hBMI] of 46 kg/m^2) and their ICP, brain water content, ventricular morphology, cerebrospinal fluid (CSF) production, and choroidal transcriptomic profile were assessed in comparison to their lean

counterparts (hBMI 23 kg/m^2). IIH-related androgen excess was mimicked by bi-weekly testosterone injections for 4 weeks in female Wistar rats followed by determination of their brain water content, CSF production, and choroidal transport activity.
Results: Obesity did not, on its own, create the elevated ICP characteristic of IIH, not did it alter various aspects of brain fluid dynamics or the choroidal transportome. Testosterone treatment, on the other hand, caused elevated CSF production due to increased activity of the NKCC1 transport protein.
Conclusions: These data indicate that while obesity and female sex are characteristic of IIH, these physiological aspects do not alone establish IIH pathophysiology, and that IIH-related androgen excess may be crucial in IIH aetiology. Future studies establishing an IIH animal model might require the combination of these, and possibly other, features.
Disclosures: None

P056

INTRACRANIAL HYPERTENSION SECONDARY TO SYSTEMIC LUPUS ERYTHEMATOSUS

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Background and Aims: Systemic lupus erythematosus (SLE) is an auto-immune disease that affects a variety of organs, mainly through inflammatory mechanisms. Intracranial hypertension (IH) is an uncommon presentation of SLE.
Methods: We report the case of a patient with SLE complicated by IH.
Results: A 36-year-old woman presented with a 1 month history of persistent headache. Her medical history was significant for a 3 year history of SLE that was not associated with antiphospholipid syndrome. She had diffuse arthritis and glomerulonephritis and was being treated with oral azathioprine and the antimalarial hydroxychloroquine. On examination, her visual acuity was 20/20 in each eye. Fundus examination revealed the presence of bilateral optic disc oedema. Magnetic resonance imaging was unremarkable. Lumbar puncture revealed an opening pressure of 28 cm cerebrospinal fluid (CSF) with normal CSF composition. She was started on oral acetazolamide. At follow-up, her visual acuity was 20/20 with complete resolution of the papilloedema in both eyes.
Conclusions: IH associated with SLE appears to be more frequent in patient with glomerulonephritis. This case warns us of the importance of fundus examination in all patients with SLE to raise suspicion of IH particularly if the patient has ocular symptoms.
Disclosures: None.

P057

IIH LIFE: A PROSPECTIVE LONGITUDINAL COHORT STUDY OF IDIOPATHIC INTRACRANIAL HYPERTENSION

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Background and Aims: There are limited longitudinal data evaluating visual and headache outcomes in idiopathic intracranial hypertension (IIH). Large prospective studies are required to help characterise key outcomes to help guide clinicians. We aimed to evaluate the disease course for non-surgical IIH patients, determined by their presentation to Neuro-ophthalmology with active IIH (papilloedema present) versus those in ocular remission (papilloedema resolved).
Methods: Our data source was the prospectively collected IIH Life database (2013–2021). Loess smoothers were used to trend measurements over time.
Results: Out of 490 patients with a confirmed diagnosis of IIH, 98% were female and the mean body mass index (BMI) was 38 kg/m^2. Visual outcomes in non-surgical patients showed little difference in Humphrey visual field mean deviation or visual acuity over time, despite higher optical coherence tomography (OCT) global peripapillary retinal nerve fibre layer (RNFL) thickness in the active group. For each unit increase in BMI RNFL thickness increased by 1.21 μm. High headache frequency was found to be determined by a personal migraine history and daily headache at diagnosis. Use of ICP medications did not affect visual or headache outcomes.
Conclusions: OCT appears to be a better measure for disease activity with visual field and acuity changing less. The headache burden was high in this cohort and they required complex headache management. In this cohort we were unable to establish the long-term benefit from the use of ICP lowering medications.
Disclosures: Professor Mollan reported personal fees from Heidelberg Engineering and advisory board fees from Invex Therapeutics during the conduct of the study and personal fees from Allergan, Santen, Sathera, Roche, Chugai, and Novartis outside the submitted work. Professor Sinclair reported fees from Invex Therapeutics as a company director with salary and stock options during the conduct of this study; and personal fees from Novartis and Allergan outside the submitted work. No other disclosures were reported.

P058

DOES THE TIMING OF IIH DIAGNOSIS IN RELATION TO PREGNANCY AFFECT OUTCOMES?

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Background and Aims: Idiopathic intracranial hypertension (IIH) is a metabolic neuro-ophthalmological disorder that is associated with body weight gain. One of the known commonest reasons why a woman of childbearing age would gain weight is pregnancy. The long-term impact of IIH diagnosed during pregnancy is not known.
Methods: We evaluated key outcomes such as vision (logMAR visual acuity, Humphrey visual field perimetric mean deviation [PMD] and optical coherence tomography [OCT] measures) and headache in a prospectively collected cohort within the IIH Life database (2013–2021). Comparison was made with those with a subsequent pregnancy, and those who never become pregnant.
Results: Women diagnosed with IIH in pregnancy had worse vision scores (Mean OCT retinal nerve fibre layer thickness and PMD) compared with the other two groups at baseline. None took intracranial pressure lowering medicines, few required a temporising lumbar puncture in the first trimester and fewer required sight-saving surgery. Overall, those who were not diagnosed in pregnancy had no sequalae, with the majority improving. All groups had comparable outcomes at 36 months.

Conclusions: IIH patient monitoring during pregnancy is important, not only for maternal health but physician communication. Medical intervention is limited due to the risk of teratogenicity. Those diagnosed with IIH in pregnancy, or those in whom IIH is exacerbated by pregnancy, are more challenging to manage and require individualised care plans.
Disclosures: Professor Mollan reported personal fees from Heidelberg Engineering and advisory board fees from Invex Therapeutics during the conduct of the study and personal fees from Allergan, Santen, Sathera, Roche, Chugai, and Novartis outside the submitted work. Professor Sinclair reported fees from Invex Therapeutics as a company director with salary and stock options during the conduct of this study; and personal fees from Novartis and Allergan outside the submitted work. No other disclosures were reported.

P059

IDIOPATHIC INTRACRANIAL HYPERTENSION: EVALUATION OF BIRTHS AND FERTILITY THROUGH THE HOSPITAL EPISODE STATISTIC DATASET BETWEEN 2002–2020

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Background and Aims: Idiopathic intracranial hypertension (IIH) predominantly affects women of reproductive age with obesity and who have a distinct profile of hyperandrogenism and insulin resistance. Polycystic ovary syndrome (PCOS) has an established adverse fertility phenotype that typically affects obese women. IIH may impact reproductive health.
Methods: We carried out a prospective cohort study of women with IIH aged 18–45 years from the English Hospital Episode Statistic dataset between 1 April 2002 and 31 March 2019. Comparison was made with PCOS and general population controls. The main outcome measures were pregnancies, gestational diabetes mellitus (DM) and pre-eclampsia, and method of delivery.
Results: Data were collected from 17587 IIH, 199633 PCOS, and 10947012 general population patients. The live birth rate, adjusted for age, was significantly lower amongst women with IIH (54.1%) compared with PCOS (67.9%), $p < .0001$ and the general population (57.7%), $p < .0001$. Pre-eclampsia and gestational DM risks were higher following a diagnosis of IIH (5.3 fold and 2.7 fold, respectively; $p < .0001$) compared with the general population controls. Following a diagnosis of IIH, elective caesarean section rates were more than twice that of the general population (odds ratio [OR] 2.4) and prior to a diagnosis of IIH (OR 2.2).
Conclusions: These data indicate that there are lower age-adjusted total pregnancy rates, increased risk of pre-eclampsia and gestational DM and a doubling of elective caesarean section rates in those with a diagnosis of IIH.
Disclosures: Professor Mollan reported personal fees from Heidelberg Engineering and advisory board fees from Invex Therapeutics during the conduct of the study and personal fees from Allergan, Santen, Sathera, Roche, Chugai, and Novartis outside the submitted work. Professor Sinclair reported fees from Invex Therapeutics as a company director with salary and stock options during the conduct of this study; and personal fees from Novartis and Allergan outside the submitted work. No other disclosures were reported.

P060

IDIOPATHIC INTRACRANIAL HYPERTENSION: EVALUATION OF ADMISSIONS AND EMERGENCY READMISSIONS THROUGH THE HOSPITAL EPISODE STATISTIC DATASET BETWEEN 2002–2020

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Background and Aims: With the increasing incidence and prevalence of idiopathic intracranial hypertension (IIH) in the United Kingdom, the aim of this study was to explore emerging themes in IIH using the Hospital Episode Statistics dataset and to quantify recent change in hospital admissions and surgeries performed within England.
Methods: Hospital Episode Statistics national data were extracted between 1 April 2002 and 31 March 2019, and followed up until 31 March 2020. All those within England with a diagnosis of IIH were included. Those with secondary causes of raised intracranial pressure were excluded.
Results: Between 1 January 2002 and 31 December 2019 28,794 new IIH cases were diagnosed. The incidence rose between 2002 and 2019 from 1.8 to 5.2 per 100,000 in the general population. Cerebrospinal fluid shunting was the commonest surgical procedure performed (6.4%), followed by dural venous sinus stenting (1%), bariatric surgery (.8%) and optic nerve sheath fenestration (ONSF) (.5%). Those requiring a shunt fell from 10.8% in 2002/2003 to 2.46% in 2018/2019. The mean 30 day emergency readmission rates for primary shunt, revision of shunt, bariatric surgery, stent, and ONSF were 23.1%, 23.7%, 10.6%, 10.0% and 9.74%, respectively
Conclusions: Increased awareness of the condition, specialist surgery and expert guidance may be changing admissions and surgical trends in IIH. The high 30 day readmission rate following primary shunt surgery for IIH requires further investigation.
Disclosures: Professor Mollan reported personal fees from Heidelberg Engineering and advisory board fees from Invex Therapeutics during the conduct of the study and personal fees from Allergan, Santen, Sathera, Roche, Chugai, and Novartis outside the submitted work. Professor Sinclair reported fees from Invex Therapeutics as a company director with salary and stock options during the conduct of this study; and personal fees from Novartis and Allergan outside the submitted work. No other disclosures were reported.

P061

EFFECTIVENESS OF BARIATRIC SURGERY VS COMMUNITY WEIGHT MANAGEMENT INTERVENTION FOR THE TREATMENT OF IDIOPATHIC INTRACRANIAL HYPERTENSION: A RANDOMISED CONTROL TRIAL

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Background and Aims: Idiopathic intracranial hypertension (IIH) causes headaches, vision loss, and reduced quality of life. Sustained weight loss in IIH is necessary to modify the disease. Is bariatric surgery superior to a community weight management intervention (CWMI) in sustaining the weight loss necessary to achieve sustained remission in IIH?

Methods: This 5-year randomised control trial (the IIH weight trial [IIH:WT]) enrolled women with active IIH and a body mass index of 35 kg/m^2 or higher in the United Kingdom (2014–2017). The primary outcome was change in intracranial pressure measured by lumbar puncture opening pressure at 12 months, as assessed in an intention-to-treat analysis.

Results: Sixty-six female participants (mean [standard deviation] age, 32.0 [7.8] years) were recruited. Intracranial pressure was significantly lower in the bariatric surgery arm at 12 months with an adjusted mean (standard error [SE]) difference of −6.0 (1.8) cm cerebrospinal fluid (CSF); 95% confidence intervals (CI), −9.5 to −2.4 cmCSF (p = .001) compared with the CWMI arm. Quality of life was significantly improved (adjusted mean [SE] difference, 7.3 [3.6]; 95% CI, .2 to 14.4; p = .04) in the surgery arm.

Conclusions: Bariatric surgery among patients with active IIH had favourable sustained outcomes with regard to reductions in ICP, disease remission, and superior quality of life outcomes at 1 months, and out to 2 years compared with a CMWI.

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P062

THE AMOUNT OF WEIGHT LOSS REQUIRED TO REDUCE INTRACRANIAL PRESSURE IN IDIOPATHIC INTRACRANIAL HYPERTENSION – RESULTS FROM THE IIH:WT RANDOMISED CONTROLLED TRIAL

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Background and Aims: The idiopathic intracranial hypertension (IIH) weight trial (IIH:WT) established that weight loss through bariatric surgery significantly reduced intracranial pressure (ICP) at 12 and 24 months as compared to a community weight management intervention (CWI). This study aimed to evaluate the amount of weight loss required to reduce ICP in IIH.
Methods: Adult women with active IIH and a body mass index ≥ 35 kg/m^2 were recruited. The relationship between ICP and weight loss was evaluated and categorised by the weight loss methods. A linear hierarchical regression model was used to fit the trial outcomes, adjusted for time, treatment arm and weight.
Results: Weight loss was significantly associated with reduction in ICP (R^2 = .4734, p ≤ .0001). Weight loss of 13.3 kg (standard deviation 1.76) was associated with disease remission (ICP <25 cmCSF). Normalisation of ICP occurred with 24% weight loss. At 12 months weight loss of between 5–10% caused a mean reduction in ICP of 4.4 cmCSF (standard error [SE] ± 2.15); whereas weight loss of 20–30% caused a greater mean reduction in ICP of −15.4 cmCSF (SE ± 2.89). Roux-en-Y gastric bypass achieved superior, more rapid and sustained ICP reduction in IIH.
Conclusions: The extent of weight loss can predict reduction in ICP in IIH. The amount of weight loss needed (24%) is unlikely to be achieved through diet alone, and early referral to a bariatric pathway should be considered. Roux-en-Y gastric bypass was the most effective surgical intervention to achieve remission in IIH.
Disclosures: Professor Mollan reported receiving personal fees from Allergan, Chiesi Farmaceutici, Heidelberg Engineering, Invex Therapeutics, Neurodiem, Novartis, Roche, Santen Pharmaceutical, and Santhera Pharmaceuticals outside the submitted work. Dr Mitchell reported receiving grants from the National Institute of Health Research during the conduct of the study and grants from the UK Ministry of Defence outside the submitted work. Dr Tahrani reported receiving grants from Novo Nordisk and Sanofi; personal fees from AstraZeneca, Boehringer Ingelheim, Bristol Myers Squibb, Eli Lilly and Company, Janssen

Pharmaceuticals, Merck Sharp & Dohme, Napp Pharmaceuticals, Novo Nordisk, and Sanofi; and nonfinancial support from Aptiva, AstraZeneca, Boehringer Ingelheim, Bristol Myers Squibb, Eli Lilly and Company, Impeto Medical, Merck Sharp & Dohme, Napp Pharmaceuticals, Novo Nordisk, and ResMed outside the submitted work. Dr Brock reported owning equity in AstraZeneca and GlaxoSmithKline, receiving personal fees from Eli Lilly and Company and Invex Therapeutics, and receiving reimbursement from Merck & Co and Roche outside the submitted work. Dr Frew reported receiving grants from the Birmingham City Council (United Kingdom) and the National Institute of Health Research outside the submitted work. Dr Sinclair reported receiving grants from the Medical Research Council of the United Kingdom and funding through a Sir Jules Thorn Award for Biomedical Science during the conduct of the study. No other disclosures were reported.

P063

INTRACRANIAL PRESSURE DETERMINES HEADACHE MORBIDITY IN IDIOPATHIC INTRACRANIAL HYPERTENSION: RESULTS FROM A RANDOMISED CONTROLLED TRIAL (IIH:WT)

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Background and Aims: Headache is the predominant disabler in idiopathic intracranial hypertension (IIH). The IIH weight trial (IIH:WT) was a randomised controlled trial in the United Kingdom investigating weight management methods in IIH. The aim of this sub-study was to characterise headache and investigate the association with intracranial pressure (ICP).

Methods: Participants with active IIH (evidenced by papilloedema) and a body mass index ≥ 35 kg/m^2 were recruited. At baseline, 12 months and 24 months headache characteristics and quality of life measures were collected. Lumbar puncture measurements of ICP were also performed.

Results: Sixty-six women with active IIH were included. The headache phenotype was migraine-like in 86%. Headache severity correlated with ICP at baseline ($r = .285$; $p = .024$). Change in headache severity ($r = .454$, $p = .001$) and monthly headache days ($r = .419$, $p = .002$) correlated with change in ICP at 12 months. Cutaneous allodynia was significantly correlated with ICP at 12 months ($r = .479$, $p < .001$). Bootstrap analysis noted a positive association between ICP at 12 and 24 months, enabling prediction of both change in headache severity and monthly headache days. ICP was associated with significant improvements in quality of life.

Conclusions: We have demonstrated a positive relationship between ICP and headache and cutaneous allodynia, which have not been previously reported in IIH. Those with the greatest reduction in ICP over 12 months had the greatest reduction in headache frequency and severity; this was associated with improvement of quality of life measures.

Disclosures: Professor Mollan reported receiving personal fees from Allergan, Chiesi Farmaceutici, Heidelberg Engineering, Invex Therapeutics, Neurodiem, Novartis, Roche, Santen Pharmaceutical, and Santhera Pharmaceuticals outside the submitted work. Dr Mitchell reported receiving grants

from the National Institute of Health Research during the conduct of the study and grants from the UK Ministry of Defence outside the submitted work. Ms Grech received personal fees from invex therapeutic outside the reported work. Dr Tahrani reported receiving grants from Novo Nordisk and Sanofi; personal fees from AstraZeneca, Boehringer Ingelheim, Bristol Myers Squibb, Eli Lilly and Company, Janssen Pharmaceuticals, Merck Sharp & Dohme, Napp Pharmaceuticals, Novo Nordisk, and Sanofi; and nonfinancial support from Aptiva, AstraZeneca, Boehringer Ingelheim, Bristol Myers Squibb, Eli Lilly and Company, Impeto Medical, Merck Sharp & Dohme, Napp Pharmaceuticals, Novo Nordisk, and ResMed outside the submitted work. Dr Brock reported owning equity in AstraZeneca and GlaxoSmithKline, receiving personal fees from Eli Lilly and Company and Invex Therapeutics, and receiving reimbursement from Merck & Co and Roche outside the submitted work. Dr Sinclair reported receiving grants from the Medical Research Council of the United Kingdom and funding through a Sir Jules Thorn Award for Biomedical Science during the conduct of the study. No other disclosures were reported.

P064

ANXIETY AND DEPRESSION PATTERNS IN WOMEN WITH IDIOPATHIC INTRACRANIAL HYPERTENSION: A MATCHED CONTROLLED RETROSPECTIVE COHORT STUDY

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Background and Aims: Incidence rates for depression and anxiety have not been previously evaluated in a large matched cohort study in idiopathic intracranial hypertension (IIH). Are anxiety and depression more common in women with IIH compared with age and body mass index (BMI) matched migraine controls and matched population controls?
Methods: Data from the IQVIA Medical Research Data, an anonymised, nationally representative database in the United Kingdom, between January 1995 and September 2019 were assessed. Incident rates for depression and anxiety were calculated in persons with IIH, migraine controls and population controls. Cox proportional hazards regression was used to calculate hazard ratios (HR) and 95% confidence intervals (CI).
Results: Anxiety crude incidence was 12.4, 12.6 and 8.2 per 1000 person-years in the women with IIH, migrainers and controls, respectively. Compared with controls, the adjusted HR for anxiety in IIH was 1.40 (95% CI 1.19–1.64); with migrainers, the adjusted HR was .98 (95% CI .83–1.14). Depression incidence was 20.1, 19.5 and 13.3 per 1000 person-years in the women with IIH, migrainers and controls, respectively. Compared with controls, depression-adjusted HR in IIH was 1.38 (95% CI 1.20–1.58); compared with migrainers, adjusted HR was .98 (95% CI .86–1.13).
Conclusions: In women with IIH, the hazard of anxiety and depression was approximately 40% higher compared with age and BMI-matched controls. Anxiety and depression are no more common in women with IIH as compared with a matched migraine population. Physician recognition and management of this co-morbidity in IIH is an unmet clinical need.

Disclosures: Professor Mollan reported personal fees from Heidelberg Engineering and advisory board fees from Invex Therapeutics during the conduct of the study and personal fees from Allergan, Santen, Sathera, Roche, Chugai, and Novartis outside the submitted work. Professor Sinclair reported fees from Invex Therapeutics as a company director with salary and stock options during the conduct of this study; and personal fees from Novartis and Allergan outside the submitted work. No other disclosures were reported.

P065

ALL-TRANS RETINOIC ACID (ATRA) – INDUCED INTRACRANIAL HYPERTENSION

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Background and Aims: A 21-year-old Polish male receiving treatment (second cycle) for pro-myelocytic leukaemia complained of intermittent horizontal diplopia. He also reported symptoms in keeping with pulsatile tinnitus. He harboured a divergence insufficiency esophoria but fundus biomicroscopy revealed significant, bilateral, severe optic nerve head swelling with normal optic nerve function.
Methods: His chemotherapy regimen consisted of all-trans retinoic acid (ATRA) and idarubicin. His medical history was otherwise unremarkable. His body mass index was 23 kg/m^2.
Results: Magnetic resonance (MR) imaging and MR venography revealed dilated optic nerve sheaths and distension of the globes only. A lumbar puncture revealed an opening pressure of 35 cmCSF with normal constituents. Blood analysis revealed neutropenia but no evidence of anaemia. There was no evidence of sepsis. He was diagnosed with ATRA-induced intracranial hypertension. Acetazolamide (500 mg twice per day) in combination with a reduced dose of ATRA for the remaining two cycles of his chemotherapy regime led to a rapid reduction in the signs and symptoms of raised intracranial pressure. This was sustained on completion of chemotherapy and acetazolamide was withdrawn.
Conclusions: ATRA-induced intracranial hypertension is a rare complication in adults receiving this treatment for promyelocytic leukaemia. Abolition of signs and symptoms can be achieved with either withdrawal of ATRA or the addition of acetazolamide in order to allow the patient to completed chemotherapy.
Disclosures: None.

P066

METABOLISM DIFFERS SYSTEMICALLY AND IN THE CENTRAL NERVOUS SYSTEM RELATED TO IIH DIAGNOSIS, RESPONSE TO INTERVENTION AND REMISSION

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Background and Aims: Idiopathic intracranial hypertension (IIH) is a disease characterised by raised intracranial pressure (ICP) and occurs predominantly in women with obesity; however the underlying molecular pathogenesis is not fully understood. Evidence is mounting to suggest that IIH is a systemic metabolic disease.
Methods: We applied untargeted metabolomic analysis using ultra high performance liquid chromatography-mass spectrometry to characterise the cerebrospinal fluid (CSF) and serum metabolite profiles in IIH patients at baseline and 12 months compared with gender and body mass index matched healthy controls with obesity to probe underlying disease mechanisms.
Results: We recruited 66 IIH patients and 20 control subjects. We identified two differential annotated metabolite features, formylpyruvate and maleylpyruvate/fumarylpyruvate, in CSF which were present at lower relative concentrations in IIH compared with control subjects and returned to control subjects' concentrations following intervention. These metabolites showed the opposite trend in serum. Twenty-one annotated metabolite features were present in serum, which differentiated IIH patients and control subjects. No previous studies have identified these metabolic pathways to play a direct pathophysiological role in IIH and therefore the reported study has identified areas of metabolism to target in future for IIH diagnosis and treatment.
Conclusions: These results support IIH being a systemic metabolic disease, not merely that causing pathology in the brain and optic nerves. The relative paucity of differential metabolites in the CSF and their role in CSF dynamics is of interest. The metabolite profiles require further mechanistic evaluation for potential biomarkers and therapeutic targets.
Disclosures: Professor Mollan reported personal fees from Heidelberg Engineering and advisory board fees from Invex Therapeutics during the conduct of the study and personal fees from Allergan, Santen, Sathera, Roche, Chugai, and Novartis outside the submitted work. Professor Sinclair reported fees from Invex Therapeutics as a company director with salary and stock options during the conduct of this study; and personal fees from Novartis and Allergan outside the submitted work. No other disclosures were reported.

P067

IDIOPATHIC INTRACRANIAL HYPERTENSION IN AN OBESE WOMAN: A CASE REPORT

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Background and Aims: Idiopathic intracranial hypertension (IIH) is a disorder characterised by raised intracranial pressure that predominantly affects young, obese women. The pathogenesis has not been fully elucidated, but several causal factors have been proposed. Symptoms can include headaches, visual loss, pulsatile tinnitus, back and neck pain, but the clinical presentation can be highly variable.
Methods: This case report describes a case of IIH in an obese woman with symptoms of headache and diplopia, and the presence of sixth cranial nerve paresis and papilloedema.
Results: A 16-year-old obese woman (body mass index > 30 kg/m^2) presented with headache and diplopia in both eyes for the previous month, but without visual loss. Her visual acuity was 6/6 in each eye, she had a sixth cranial nerve paresis, and funduscopy revealed papilloedema in both eyes. The results of head magnetic resonance (MR) imaging with contrast showed no intracranial space occupying lesion and MR venography

showed venous sinus stenosis. She was treated with acetazolamide 250 mg, once per day for 3 weeks, and instructed to diet to reduce weight. After 3 weeks of therapy the diplopia disappeared, the sixth cranial nerve paresis resolved and the papilloedema reduced.
Conclusions: IIH is a disease of women in the childbearing years, and its prevalence is increasing due to the worldwide obesity epidemic. Recognising and understanding the risk factors that truly contribute to intracranial hypertension is important in both diagnosing and understanding the pathophysiology of the disease.
Disclosures: None.

P068

I'VE SAVED MYSELF A SHUNT

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Background and Aims: Idiopathic intracranial hypertension (IIH) presenting with visually threatening features must be taken seriously. Interventions include cerebrospinal fluid (CSF) diversion procedures and optic nerve sheath fenestration, but interventions carry risks of complications and failure. Weight loss is disease modifying and what is unknown is whether rapid weight loss can impact acutely on IIH.
Methods: We describe a 22-year-old white Caucasian woman who presented with moderate-to-severe papilloedema and transient visual obscurations (TVOs) in both eyes. Neuroimaging (including venography) and lumbar puncture confirmed a diagnosis of IIH. No secondary causes were found. She reported approximately 19 kg weight gain over 12 months. Her body mass index (BMI) was 39 kg/m^2 and her weight was 111 kg.
Results: Management options were discussed as her visual fields showed increased blind spots. She opted for an emergency low calorie diet (500 kcalories/day) and oral acetazolamide (2 g daily in divided doses). Within 1 week, her papilloedema had improved. However, she was intolerant of acetazolamide, which was stopped. After 6 weeks she reported no TVOs. Her BMI reduced to 37.8 kg/m^2. Despite stopping acetazolamide within the first few days of treatment, the global retinal nerve fibre layer thickness reduced dramatically from 337 to 121 μm (right eye) and 304 to 107 μm (left eye).
Conclusions: The patient improved significantly and avoided a surgical procedure. She has remained in ocular remission at latest follow-up. Low calorie diets are effective, in motivated patients, to rapidly reduce weight and induce disease remission.
Disclosures: None.

P069

DIFFERENCES BETWEEN PRE-PUBERTAL AND POST-PUBERTAL PAEDIATRIC IDIOPATHIC INTRACRANIAL HYPERTENSION (IIH): A NARRATIVE REVIEW

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Background and Aims: The incidence of idiopathic intracranial hypertension (IIH) in adults is increasing in line with the rising incidence of obesity. In adults, the cause of IIH is not fully understood but there is increasing evidence suggesting that IIH is a disease of systemic metabolic dysregulation. In the paediatric population, the aetiology is even less clear and the contrasting presentations between the pre- and post-pubertal population suggest a different underlying pathogenesis.
Methods: We conducted a narrative review to evidence the differences between pre- and post-pubertal paediatric IIH. A detailed search of the scientific literature was performed using the PubMed database. This included English language papers and IIH-related keywords. The abstracts and full texts were reviewed by two independent assessors.
Results: Overall, the post-pubertal group had a much higher incidence and were more likely to be female and obese, compared with the pre-pubertal group. Presenting features in the post-pubertal group were more akin to adults, with headache as the dominant feature, whilst in the pre-pubertal group presentation was more variable and incidental. There are no evidence-based recommendations for management of paediatric IIH. Some studies had different definitions of pre- and post-puberty. Secondary causes of raised intracranial pressure may have confounded the accuracy of analysis as they have been included in some of the cohorts.
Conclusions: Post-pubertal children have a similar phenotype to adult cohorts, whereas pre-pubertal children do not display the same predilection towards female sex and obesity. This makes it harder to collate studies and compare when we are not using standard definitions or markers of puberty. In the future, defined criteria of 'paediatric' IIH would be helpful.
Disclosures: Professor Mollan reported personal fees from Heidelberg Engineering and advisory board fees from Invex Therapeutics during the conduct of the study and personal fees from Allergan, Santen, Sathera, Roche, Chugai, and Novartis outside the submitted work. Professor Sinclair reported fees from Invex Therapeutics as a company director with salary and stock options during the conduct of this study; and personal fees from Novartis and Allergan outside the submitted work. No other disclosures were reported.

P070

INVESTIGATION OF MITOCHONDRIAL FUNCTION AND THE ROLE OF GLUCOSE IN A BRAIN SLICE MODEL OF HEADACHE MECHANISMS

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Background and Aims: Cortical spreading depression (CSD), a propagating wave of depolarisation across the cerebral cortex, has been proposed to activate trigeminal nerve afferents and contribute to headache pathophysiology. Using a mouse brain slice model, we aimed to investigated the impact of hypoglycaemia and CSD on mitochondrial and metabolic function.
Methods: CSD was induced with 2 M Potassium chloride (KCl) to cortical regions of brain slices in the presence (10 mM) or absence of glucose, then mitochondrial oxidative respiration and metabolite profiles were assessed. Fluorescent calcium labelling demonstrated a wave of calcium movement throughout the slice following KCl application, characteristic of CSD activity.

Results: Mitochondrial integrity was not disrupted in brain slices following CSD and basal respiration remained unchanged. However, in the absence of glucose, CSD elevated mitochondrial oxidative capacity from complex I and II ($p = .002$) and increased maximal rates of uncoupled respiration ($p < .001$), which was rescued by 10 mM glucose. Metabolites altered following CSD included pyruvate ($p = .0315$) and lactate (p .003) in the presence of glucose, whilst decreases in lactate ($p < .0001$) and glutamate ($p = .0002$) occurred in the absence of glucose.

Conclusions: Energy deficits due to glucose deficiency may upregulate oxidative respiration and alternative metabolic pathways following CSD, to compensate. This begins to suggest that CSD induces disturbances in metabolic energy metabolism, which may contribute to headache. These changes could be amenable to nutritional intervention as an adjunctive therapy for those with idiopathic intracranial hypertension headaches.

Disclosures: O Grech reports scientific consultancy fees from Invex therapeutics during the conduct of the study. Professor Mollan reported personal fees from Heidelberg Engineering and advisory board fees from Invex Therapeutics during the conduct of the study and personal fees from Allergan, Santen, Sathera, Roche, Chugai, and Novartis outside the submitted work. Professor Sinclair reported fees from Invex Therapeutics as a company director with salary and stock options during the conduct of this study; and personal fees from Novartis and Allergan outside the submitted work. No other disclosures were reported.

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P071

DETAILED EVALUATION OF COGNITIVE PERFORMANCE IN IDIOPATHIC INTRACRANIAL HYPERTENSION AND RELEVANCE OF INTRACRANIAL PRESSURE

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Background and Aims: Idiopathic intracranial hypertension (IIH) is an increasingly prevalent disease, and although cognitive impairments have been reported in IIH, evidence supporting these deficits are scarce. Cognitive function is not commonly addressed during the routine evaluation of IIH and the contributing factors have not been defined.

Methods: The study design was a case-controlled study comparing cognitive domains between control and IIH participants at baseline. IIH participants were prospectively evaluated at 12 months as part of a sub-study of the IIH weight trial, a randomised, controlled, parallel-group, multi-centre trial comparing the

effects of a bariatric surgery pathway versus a community weight management intervention. Detailed evaluation of cognitive function was conducted using a bespoke battery of cognitive tests. These included the attention network test.

Results: We identified impaired attention networks (executive function) and sustained attention in IIH. These deficits were not permanent, as IIH participants exhibited improvement in executive function, sustained attention and verbal short-term memory over 12 months follow-up, which were associated with reduction in intracranial pressure (ICP). Cognition was evaluated before and after a lumbar puncture, with significant improvement in executive function. The impact of co-morbidities was explored, with headache severity, co-morbid depression and markers of obstructive sleep apnoea being adversely associated with cognitive performance. Cognitive deficits were associated with impaired reliability to perform visual field tests, the cornerstone of monitoring vision in IIH.

Conclusions: Our findings propose that cognitive impairment should be acknowledged as a clinical manifestation of IIH. We have demonstrated that cognitive deficits can improve over time and with reduction of ICP. Treating co-morbidities including depression, obstructive sleep apnoea and headache could improve cognitive performance.

Disclosures: O Grech consultancy work for Invex therapeutics (2020). Professor Mollan reported personal fees from Heidelberg Engineering and advisory board fees from Invex Therapeutics during the conduct of the study and personal fees from Allergan, Santen, Sathera, Roche, Chugai, and Novartis outside the submitted work. Professor Sinclair reported fees from Invex Therapeutics as a company director with salary and stock options during the conduct of this study; and personal fees from Novartis and Allergan outside the submitted work. No other disclosures were reported.

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P072

METABOLOMIC ANALYSIS REVEALS REMODELLING OF CENTRAL AND SYSTEMIC METABOLITE PATHWAYS IN IDIOPATHIC INTRACRANIAL HYPERTENSION LINKED TO DISEASE ACTIVITY AND HEADACHE GENERATION

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Background and Aims: The pathogenesis of idiopathic intracranial hypertension (IIH) remains poorly understood and this lack of knowledge hinders advances in IIH. Mounting evidence indicates that IIH is no longer considered exclusively a disease of the central nervous system, but instead involves systemic metabolic perturbation in excess of that driven by obesity. We sought to determine any metabolic disturbances in IIH and if they change in association with disease activity and treatment.

Methods: We conducted quantitative metabolomic profiling of cerebrospinal fluid (CSF), serum and urine in IIH patients (n = 84) compared with age, gender and body mass index matched controls (n = 20) and following 12 months of weight loss in IIH (n = 50).
Results: We identified a distinct metabolic profile in IIH featuring four predominant metabolites. Urea was lower in IIH (CSF $p < .001$, urine $p = .009$) and correlated with intracranial pressure ($p = .019$) and headache severity ($p = .031$). It significantly increased after 12 months of weight loss (CSF $p = .004$, urine $p = .043$). The lactate:pyruvate ratio was increased in IIH (CSF $p = .023$, serum $p = .004$) and decreased after 12 months of weight loss ($p < .001$). Acetate was higher in IIH ($p = .008$) and correlated with headache severity ($p = .03$) and disability ($p = .003$). It decreased after 12 months of weight loss ($p = .007$). The ketones 3-hydroxybutyrate and acetoacetate were altered in IIH CSF and normalised after 12 months of weight loss ($p = .019$ and $p = .015$, respectively).
Conclusions: This IIH metabolomics study has demonstrated systemic metabolic disturbances that are evident in the CSF, serum and urine of IIH patients, with the featured metabolites being urea, lactate: pyruvate ratio, acetate and ketone bodies. These alterations of metabolic pathways specific to IIH provide biological insights and warrant mechanistic evaluation.
Disclosures: O Grech consultancy work for Invex therapeutics (2020). Dr A Yiangou reports receiving speaker fees for educational talk from Teva, UK outside the submitted work Professor Mollan reported personal fees from Heidelberg Engineering and advisory board fees from Invex Therapeutics during the conduct of the study and personal fees from Allergan, Santen, Sathera, Roche, Chugai, and Novartis outside the submitted work. Professor Sinclair reported fees from Invex Therapeutics as a company director with salary and stock options during the conduct of this study; and personal fees from Novartis and Allergan outside the submitted work. No other disclosures were reported.

P073

DISCUSSING WEIGHT WITH PEOPLE WITH IDIOPATHIC INTRACRANIAL HYPERTENSION ASSESSING STIGMA, SUPPORT AND CHANGING THE DIALOGUE: RESULTS OF AN ONLINE SURVEY

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Background and Aims: Weight discussions are necessary in consultations between health professionals and people with idiopathic intracranial hypertension (pwIIH) because of the established links between obesity and outcomes in this condition. International guidance shows that the way health professionals engage with people living with obesity impacts on engagement, motivation and relationships.
Methods: This study was an anonymous online survey using Survey Monkey™, emailed to members of the national charity for pwIIH (IIH UK) and shared on social media. The survey aimed to gather information on the experience of weight-related discussions between pwIIH and health care professionals.
Results: The survey was completed by 625 pwIIH. Of these: 92% were not asked permission to discuss their weight; 79% did not find the person discussing weight empathetic or supportive; 86% reported being made to feel that IIH was their fault because of their weight; 67% felt that they had less favourable treatment because of this; and 78% were not offered support to lose weight. Of those who were offered support to lose weight, 84% did not find the support helpful or appropriate.
Conclusions: PwIIH expanded on questions qualitatively detailing negative impacts of language used and gave suggestions for improvements and support they would like to receive.
Disclosures: None.

P074

PHYSICAL ACTIVITY, QUALITY OF LIFE AND HEADACHE IN PEOPLE WITH IDIOPATHIC INTRACRANIAL HYPERTENSION: RESULTS OF AN ONLINE SURVEY

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Background and Aims: Idiopathic intracranial hypertension (IIH) is known to have a negative effect on health related quality of life (HRQoL). Physical activity levels are reduced and related to HRQoL in those with headache conditions, such as migraine. This has not been explored in people with IIH (pwIIH). This online questionnaire aimed to quantify physical activity levels and relationships between physical activity, quality of life, headache and clinical characteristics in pwIIH.
Methods: Primary measures were physical activity (PASIPD) and health related quality of life (SF-36®) with secondary measures of headache impact (HIT-6™), body mass index (BMI) and age.
Results: The questionnaire was completed by 164 pwIIH. Physical activity levels were low in pwIIH (median PASIPD score: 10.38 [interquartile range ± 17.6] metabolic equivalent [MET] h/day, PASIPD ceiling 199.5 MET h/day), similar to people with physical disabilities and other headache conditions. Engagement in exercise in pwIIH was low with 75%, 89% and 95% never engaging in light, moderate or strenuous exercise, respectively. Significant moderate correlations were found between the PASIPD total score, headache impact, and HRQoL (physical component score, physical functioning, physical role, general health, vitality and social role). Physical activity was not related to mental component score, age, current or at diagnosis BMI.
Conclusions: This survey showed that physical activity levels are affected in pwIIH. The results suggest improving physical activity levels in pwIIH could have positive impacts on HRQoL and headache impact. Future research should explore barriers, enablers and opportunities for pwIIH to increase physical activity levels and engage in exercise programmes.
Disclosures: None.

P075

A CASE OF PAPILLOEDEMA IN PARTIAL OPTIC ATROPHY – WHERE DOES THE FLUID GO?

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Background and Aims: Complications of idiopathic intracranial hypertension (IIH) in pregnancy are uncommon.
Methods: A retrospective case report is presented.
Results: A 37-year-old patient was referred with a 2 week history of bilateral blurred vision, associated with worsening headaches. She was 39 weeks pregnant with a history of IIH that had been managed with a lumbo-peritoneal shunt inserted 10 years previously. Recent review by the neurology team led to a diagnosis of migraine headaches as no significant disc swelling was seen, however she had a history of bilateral partial optic atrophy. On presentation, she had a visual acuity of 6/24 in her right eye (OD) and 6/12 in her left eye (OS). Her blood pressure and urine dipstick test were normal, indicating no signs of pre-eclampsia. Fundus examination showed pale discs bilaterally with profuse oedema at both maculae. Review of serial disc and macula optical coherence tomography (OCT) scans showed mild increase in retinal nerve fibre layer (RNFL) thickness and a definite increase in peripapillary intraretinal fluid, which extended to

both maculae over time, causing cystoid macular oedema and serous macular detachment. Referral to the neurology team confirmed raised intracranial pressure on lumbar puncture, secondary to a blocked shunt and exacerbated by pregnancy. Following a normal delivery, she underwent insertion of a ventriculo-peritoneal shunt. One week later, her vision had improved to 6/12 OD and 6/9 OS. Although both discs remained swollen, OCT showed a significant improvement in the macular oedema in both eyes.
Conclusions: In patients with partial optic atrophy, papilloedema can be difficult to detect as RNFL oedema may be subtle. Serial OCT imaging of the discs and maculae is especially useful in demonstrating early peripapillary intraretinal microcysts and subsequent tracking of fluid to the maculae, therefore enabling correct diagnosis and close monitoring of treatment response.
Disclosures: None.

P076

DESCRIBING THE IMPACT OF HEADACHE IN THE WORDS OF PEOPLE WITH IDIOPATHIC INTRACRANIAL HYPERTENSION: A QUALITATIVE SOCIAL MEDIA STUDY

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Background and Aims: Quantitative reports of headache in idiopathic intracranial hypertension (IIH) show that it affects quality of life, causes disability, is daily in most and has been identified as an unmet need in IIH. The qualitative lived experience of the impact of headache in people with IIH (pwIIH) is under reported. This study aimed to describe the lived experience of headache in the words of people with IIH.
Methods: This study was an anonymous online survey using Survey Monkey™. The survey was emailed to members of the national charity for pwIIH (IIH UK) and shared on social media sites.
Results: The survey was responded to by 512 people. Common words from responses were used to create a graphic using Wordle™. Thematic analysis of pwIIH qualitative responses identified themes in the data (changes in participation, changes in mood and well-being, wider IIH symptoms and the overwhelming, debilitating nature of headache). "It has completely changed my life … it effects my work, my social life, everything"; "It makes me feel worthless and like a failure and suicidal, I'm a shadow of the person I once was"; "IIH headaches changed my life … .I remember having more life, IIH has robbed me of my life".
Conclusions: This survey shows the breadth and depth of the impact of headache in IIH. Current measures do not capture all elements of resultant disability. Further qualitative research is needed to explore the lived experience of IIH to ensure that disability is not underestimated in this complex condition.
Disclosures: Professor Mollan reported personal fees from Heidelberg Engineering and advisory board fees from Invex Therapeutics during the conduct of the study and personal fees from Allergan, Santen, Sathera, Roche, Chugai, and Novartis outside the submitted work. Professor Sinclair reported fees from Invex Therapeutics as a company director with salary and stock options during the conduct of this study; and personal fees from Novartis and Allergan outside the submitted work. No other disclosures were reported.

P077

IDIOPATHIC INTRACRANIAL HYPERTENSION RECURRENCE IN THE ABSENCE OF HEADACHE: A CASE REPORT

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Background and Aims: Headache is a common presenting symptom of idiopathic intracranial hypertension (IIH). This presentation demonstrates a case in ocular remission who was medically managed for headache but who had recurrence of disease as evidenced by optical coherence tomography (OCT), and on questioning this was without symptoms of headaches or pulsatile tinnitus.
Methods: A 30-year-old white Caucasian woman was diagnosed with IIH (normal neuro-imaging and venography and a lumbar puncture opening pressure of 38 cmCSF, with a body mass index of 59 kg/m^2). During follow-up she had complete resolution of headaches with medical therapy (topiramate 100 mg nocte) and weight loss.
Results: On routine follow-up she had a flare up her papilloedema, as measured by OCT of the optic nerves. However, she reported her headaches as being 50% better than at the last visit (3 monthly migraine days; headache impact test score of 59; HAD A – 12; HAD D – 9). Her body weight had increased by 2 kg.
Conclusions: We report a phenomenon which is poorly documented in the literature with the majority of the case reports focussing on asymptomatic papilloedema at presentation rather than during follow-up. Where headache is controlled medically, routine screening may be prudent, particularly in the presence of weight gain.
Disclosures: None.

P078

HAEMORRHAGIC PAPILLOEDEMA OR TERSON'S SYNDROME: CAN WE TELL THEM APART?

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Background and Aims: Terson's syndrome is intraocular haemorrhage associated with intracranial bleeding, commonly subarachnoid haemorrhage. Papilloedema may exceptionally present with prominent retinal and preretinal haemorrhages obscuring optic disc swelling. Although both disorders have raised intracranial pressure in common, overlapping features in the fundi may be plausible.
Methods: We present retrospective serial case reports of two women with intracranial hypertension and haemorrhagic papilloedema. We also carried out a review of the literature of cases describing "unusual haemorrhages" in papilloedema or Terson's-like syndrome.
Results: A 64-year-old obese woman with hypercapnic respiratory failure caused by chronic obstructive disease and sleep apnoea, experienced headache, photopsias and bilateral visual loss. Bilateral optic disc swelling with prominent retinal peripapillary haemorrhages and preretinal haemorrhages in the posterior pole were observed. A 39-year-old woman experienced throbbing new onset headache, vomiting and paraesthesia caused by obstructive hydrocephalus. She had bilateral optic disc swelling with multiple retinal peripapillary haemorrhages in her left eye. Investigations disclosed neurosarcoidosis with skin and lung involvement.

Conclusions: Terson's-like papilloedema has been described in rapidly growing intracranial tumours and, less commonly, in pseudotumour cerebri cases. The abrupt onset of intracranial hypertension in subarachnoid haemorrhage or sinus vein thrombosis can lead to intraocular venous hypertension and rupture of retinal capillaries.
Disclosures: None.

P079

BILATERAL OPTIC NERVE SHEATH FENESTRATION PROCEDURE IN PATIENTS WITH FULMINANT IDIOPATHIC INTRACRANIAL HYPERTENSION

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Background and Aims: There has been an increase in incidence of idiopathic intracranial hypertension (IIH) co-existent with the increase in the incidence of obesity worldwide. IIH treatment usually consists of weight reduction and systemic acetazolamide. If these therapies lead to no improvement in visual symptoms one of the surgical procedures used is optic nerve sheath fenestration (ONSF).
Methods: We describe two cases of fulminant IIH, with significant improvements after bilateral ONSF after 2 months of follow-up.
Results: Two female IIH patients, 14 and 25-years-old, had severe papilloedema with severe visual loss / severe visual field constriction. The lumbar puncture opening pressures were 56 and 60 cmCSF, respectively. Both cases demonstrated no improvement in visual symptoms after 3 days of conventional therapy and the surgical method selected was bilateral ONSF in both. Rapid improvement in visual field in both patients was observed and the degree of papilloedema reduced. In addition, the 14-year-old patient showed improvement in visual acuity. Overall, most of the pathological initial symptoms disappeared.
Conclusions: In conclusion, bilateral ONSF had high efficacy and safety in the two IIH cases presented above and should be considered in cases of fulminant IIH. The decision to operate must be made rapidly to reduce the risk of permanent damage to the optic nerves.
Disclosures: None.

P080

A CASE OF A CHILD WITH BENIGN INTRACRANICAL HYPERTENSION AS A CONSEQUENCE OF A PROLONGED DIETARY MANAGEMENT

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Background and Aims: Pseudotumour cerebri occurs in cases of increased intracranial pressure with no obvious reasons. Benign intracranial hypertension rarely occurs in children, so it requires urgent diagnosis and exclusion of multiple neurosurgical disorders.
Methods: Although benign intracranial hypertension is uncommon in paediatric clinical practice, it can be a diagnostic, as well as therapeutic challenge.
Results: This paper presents a case of a 14-year-old girl who followed a rigorous diet, had hormonal imbalance, a cessation of a menstrual period, and clinical manifestations of pseudotumour cerebri. The discontinuation of the diet and oral application of carbonic anhydrase inhibitor for several months

contributed to the loss of subjective problems and recovery of clinically visible symptoms in the forms of diplopia and the fundus changes. The dominant symptom in our patient was headache and diplopia caused by abducens nerve palsy, while the presence of scotomas had not been registered, except for a slightly enlarged blind spot as a result of optic disc oedema.
Conclusions: The case of a 14-year-old girl who followed a rigorous diet indicates that benign intracranial hypertension is possible in this age, and management has to be by appropriate professionals and a paediatrician.
Disclosures: None.

P081

TERSON'S-LIKE SYNDROME SECONDARY TO IDIOPATHIC INTRACRANIAL HYPERTENSION (IIH)

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Background and Aims: A 30-year-old, obese female was referred by her optometrist with bilateral swollen optic nerve heads. Her vision had deteriorated quite suddenly over 6 days. Examination revealed severely swollen optic nerve heads with multi-layered haemorrhages within the peripapillary regions and posterior poles.
Methods: Her medical history included acne vulgaris and depression with psychotic episodes. She was being treated with olanzapine 10 mg and sertraline 50 mg, daily. She had gained 10 kg in weight over the preceding 8 months. She had no other neurological symptoms.
Results: Dedicated neuroimaging of her head revealed features suggestive of intracranial hypertension with no haemorrhages, aneurysms or venous sinus thrombosis. The results of extensive blood investigations including a thrombophilia screen were unremarkable. A lumbar puncture revealed an opening pressure of 40 cmCSF with normal constituents. She was treated with acetazolamide and, following consultation with her psychiatrist, her olanzapine dose was tapered until it was withdrawn altogether. After 2 months, her optic nerve swelling and haemorrhages had completely resolved. She had lost 10 kg in weight and her ocular fundi and optic nerve function remained normal after stopping acetazolamide.
Conclusions: This is a rare case of IIH causing a Terson's-like syndrome associated with severe papilloedema. Furthermore, this case is somewhat unusual due to the paucity of neurological symptoms. Patients with this condition can be managed conservatively without the need for vitrectomy.
Disclosures: None.

P082

CURRENT CONCEPTS COMPARING SPACEFLIGHT ASSOCIATED NEURO-OCULAR SYNDROME AND IDIOPATHIC INTRACRANIAL HYPERTENSION

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Background and Aims: Spaceflight associated neuro-ocular syndrome (SANS) has been described in astronauts following long duration (> 3 months) space flight. SANS describes a constellation of signs predominantly affecting vision. SANS is recognised as a significant health risk for astronauts, potentially limiting human space exploration. This syndrome shares similarities with idiopathic intracranial hypertension (IIH). The aim was to compare SANS and IIH.
Methods: Embase <1980 – 2021 Week-44> and Ovid MEDLINE(R) <1946 – 2021 Week-52> were searched for English articles. The search terms for articles in all year ranges with multiple combinations the key terms. A further search was performed between week-52, 2021 to 23, 2022.
Results: SANS occurs in the healthy male-dominated astronaut population, who experience chronic fluid shifts in microgravity, which may elevate intracranial pressure (ICP). With few subjects and limited diagnostic equipment available in space, experimental models are required to study this phenomenon. IIH is characterised by raised ICP of unclear aetiology, typically in young obese women with metabolic syndrome. Despite the different causes of SANS, IIH and idiopathic choroidal folds, they all manifest similarly with optic disc oedema, choroidal folds and reduced acuity. In this poster, we discuss how these entities likely share related pathophysiological mechanisms that can be targeted by similar treatments.
Conclusions: Recent clinical trials for IIH have identified novel therapeutic agents, which may also provide benefit to astronauts with SANS. Whilst the number of SANS cases remains limited, there is already wealth of clinical experience amongst neurologists with conditions such as IIH, which provides transferrable insights for space medicine.
Disclosures: Professor Mollan reported personal fees from Heidelberg Engineering and advisory board fees from Invex Therapeutics during the conduct of the study and personal fees from Allergan, Santen, Sathera, Roche, Chugai, and Novartis outside the submitted work. Professor Sinclair reported fees from Invex Therapeutics as a company director with salary and stock options during the conduct of this study; and personal fees from Novartis and Allergan outside the submitted work. No other disclosures were reported.

P083

OUTCOMES FOLLOWING VENTRICULO-PERITONEAL SHUNTS FOR SIGHT THREATENING IDIOPATHIC INTRACRANIAL HYPERTENSION: A PROSPECTIVE LONGITUDINAL COHORT STUDY

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Background and Aims: Idiopathic intracranial hypertension (IIH) is a rare neurological disorder characterised by raised intracranial pressure (ICP). Severe cases can manifest with papilloedema and rapidly deteriorating vision. Reduction of ICP can be achieved through surgical procedures, such as ventriculo-peritoneal shunt (VPS) insertion.
Methods: We established baseline measures of papilloedema and macular ganglion cell layer volume (GCLV) on optical coherence tomography (OCT), perimetric mean deviation (MD) and headache severity in people with IIH requiring VPS at a regional neuroscience centre. Data were collected prospectively. Loess smoothers were used to characterise outcomes post-operatively.

Results: Fifty-one patients underwent VPS insertion. The patients were 92% female (47/51), with a mean age of 28.1 years (standard deviation [SD] 8.4), mean body mass index of 37.4 kg/m^2 (SD 9.7), and a mean follow-up post-operatively of 330 days (SD 290). Characteristics of the worse eye at baseline included a MD of −11.4 (SD 9.5) dB, a mean retinal nerve fibre layer thickness of 376.2 [SD 121.4) μm and a mean Frisén papilloedema grade of 4.3 (SD .9). Post-operatively, papilloedema resolved completely by 4 months. Alarmingly, mean GCLV steadily declined from 1.05 μm^3 at baseline to .95μm^3 12 months post-operatively. Headache frequency fell from 12.7 days/month to 3.4 days/month at 3 months post-operatively, before increasing to 13.8 days/month by 12 months.
Conclusions: VPS insertion leads to a dramatic and sustained reduction in papilloedema by 4 months in IIH. However, macular ganglion cell layer loss continued at 12 months following surgery, which may predispose to future sight loss. Whilst headache severity often improves shortly following VPS, this is inconsistent and not sustained.
Disclosures: Professor Mollan reported personal fees from Heidelberg Engineering and advisory board fees from Invex Therapeutics during the conduct of the study and personal fees from Allergan, Santen, Sathera, Roche, Chugai, and Novartis outside the submitted work. Professor Sinclair reported fees from Invex Therapeutics as a company director with salary and stock options during the conduct of this study; and personal fees from Novartis and Allergan outside the submitted work. No other disclosures were reported.

P084

THE ACUTE AND CHRONIC EFFECTS OF ACETAZOLAMIDE AND TOPIRAMATE ON INTRACRANIAL PRESSURE

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Background and Aims: Lowering intracranial pressure (ICP) is the primary rationale of pharmacotherapy in idiopathic intracranial hypertension (IIH). There is limited evidence behind the use of the leading therapeutics, acetazolamide and topiramate, and how they lower ICP. We assessed the ICP lowering capacity of these drugs in a pre-clinical ICP model.
Methods: In a randomised, cross-over study, we assessed the capacity of acetazolamide and topiramate to modulate ICP in female Sprague Dawley rats (n = 10) using continuous telemetric ICP monitoring. We assessed single supra-clinical doses over 24 hours, and twice-daily clinically equivalent doses over 10 days. Drugs were delivered via oral gavage.
Results: Over 24 hours, acetazolamide (−.41 ± .08 mmHg, p = .016) and topiramate (−.44 ± .06 mmHg, p = .0016) lowered ICP compared with vehicle (−.01 ± .07 mmHg). Administering these drugs in combination had an additive effect at lowering ICP (−.76 ± .11 mmHg, p < .0001). ICP reached its lowest point 90 minutes after administration. Over 10 days, acetazolamide (−.35 ± .12 mmHg, p = .02) and topiramate (−.46 ± .14 mmHg, p = .006) lowered daily ICP compared with vehicle (.17 ± .08 mmHg) with no evidence of tachyphylaxis. Over the course of the average day, the effect of acetazolamide wore off overnight whereas the ICP lowering effect of topiramate was sustained overnight, relative to control.
Conclusions: We have demonstrated that acetazolamide and topiramate rapidly lower ICP following administration and their ICP lowering effects are additive. The ICP lowering effect of these drugs persists with no tachyphylaxis with long-term administration. These data provide the functional rationale for the clinical use of acetazolamide and topiramate in IIH.
Disclosures: None.

P085

MULTIPLE LUMBAR PUNCTURES AIMING TO RELIEVE HEADACHE RESULTS IN IATROGENIC SPINAL HAEMATOMA

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Background and Aims: Multiple lumbar punctures (LP) have historically been a strategy to relieve headaches associated with idiopathic intracranial hypertension (IIH) despite limited evidence of long-term efficacy. LP is typically a relatively straightforward procedure with minimal complications reported. Herein, we present a serious rare complication of an LP-related lumbosacral epidural haematoma which led to an incomplete cauda equina syndrome with urinary incontinence.
Methods: Multiple lumbar punctures had been part of this patient's management historically for her IIH.
Results: She presented with blurring of her vision, right-sided weakness and exacerbation of headache. Shunt dysfunction was questioned and she underwent magnetic resonance imaging (MRI) of her brain. An LP was then performed on the ward in the left lateral decubitus position using a pink Quincke needle (18 G with a cutting bevel). The LP was a traumatic tap, evidenced by blood-stained fluid. An opening pressure of 15 cmCSF was recorded. Her clotting and biochemical bloods were normal and she was discharged home. Five days after the LP she was re-admitted with new bilateral shooting leg pains and lower back pain. She was discharged with a plan to have urgent MRI of her spine as an outpatient. However, the following day she re-attended with new and unprovoked episodes of urinary incontinence, lower back pain and saddle paraesthesia. She underwent MRI of her spine, which revealed a 4 cm epidural lumbosacral haematoma at L1/S1. The haematoma was conservatively managed. She still reported saddle paraesthesia and urinary incontinence 7 months following the LP.
Conclusions: LP-related spinal haematomas remain a rare complication and this case demonstrates the need to appropriately consent patients prior to invasive tests. Multiple LPs used historically to treat IIH are not recommended by the consensus IIH guidelines.
Disclosures: Professor Mollan reported personal fees from Heidelberg Engineering and advisory board fees from Invex Therapeutics during the conduct of the study and personal fees from Allergan, Santen, Sathera, Roche, Chugai, and Novartis outside the submitted work. Professor Sinclair reported fees from Invex Therapeutics as a company director with salary and stock options during the conduct of this study; and personal fees from Novartis and Allergan outside the submitted work. No other disclosures were reported.

P086

MANAGEMENT AND OUTCOMES OF PERIPAPILLARY CHOROIDAL NEUROVASCULAR MEMBRANES IN IDIOPATHIC INTRACRANIAL HYPERTENSION: A CASE SERIES

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Background and Aims: Peripapillary choroidal neurovascular membrane (CNVM) secondary to papilloedema caused by idiopathic intracranial hypertension (IIH) is rare. Management options include observation since spontaneous resolution can occur, but permanent sight loss may occur, therefore intravitreal injection of anti-vascular endothelial growth factor (VEGF) agents is often considered on a case-by-case basis. IIH:Life is a prospective longitudinal evaluation of clinical outcomes in IIH (National Research Ethics Committee [14/LO/1208]). Out of 490 patients with a confirmed diagnosis of IIH, we identified four patients whose care was complicated by peripapillary CNVM.
Methods: Multimodal imaging with ocular coherence tomography (OCT) and angiography was critical for diagnosis and monitoring of these patients.
Results: We therefore found in .8% of people with IIH in a single neuroscience centre over a 9 year period had peripapillary CNVM. In each case the diagnosis of peripapillary CNVM was made clinically with supporting features observed on retinal imaging. Two patients underwent intravitreal anti-VEGF treatment with good outcomes. One patient spontaneously improved without treatment. One patient presented with extensive haemorrhage and underwent intravitreal anti-VEGF treatment, however they never recovered their vision.
Conclusions: There is a spectrum of this rare complication in terms of the clinical course. Clinical questions remain in particular as to how long we should observe people with IIH and a detected peripapillary CNVM without intervening with an intravitreal anti-VEGF agent. More evidence is required to inform safe clinical practice.
Disclosures: Professor Mollan reported personal fees from Heidelberg Engineering and advisory board fees from Invex Therapeutics and personal fees from Allergan, Santen, Sathera, Roche, Chugai, and Novartis outside the submitted work. Professor Sinclair reported fees from Invex Therapeutics as a company director with salary and stock options; and personal fees from Novartis and Allergan outside the submitted work. No other disclosures were reported.

P087

ACUTE NEURO-OPHTHALMIC PRESENTATION OF SIMULTANEOUS INTRACRANIAL HYPERTENSION AND MULTIPLE SCLEROSIS

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Background and Aims: The co-existence of multiple sclerosis (MS) and intracranial hypertension (IH) has been rarely reported. Whether this constitutes a coincidence or reflects a causal relationship remains to be settled. We report an acute neuro-ophthalmological presentation of simultaneous IH and MS with rapid, time-locked, initiation and progression of both entities.
Methods: A 23-year-old woman presented with a 2 day history of binocular vertical diplopia.
Results: On examination there was a left fourth nerve palsy. Additionally, there was bilateral papilloedema (Frisén grade 1). Perimetry showed blind spot enlargement of the right eye. Computed tomography venography showed bilateral transverse sinus stenosis with no thrombosis. Five days later the diplopia

worsened. Examination revealed a right internuclear ophthalmoplegia and a minor sixth nerve palsy. Whilst the fourth nerve palsy had resolved, the papilloedema had slightly worsened. Brain magnetic resonance imaging showed multiple demyelinating lesions, including one involving the right fourth nerve nucleus and the adjacent medial longitudinal fasciculus. The cerebrospinal fluid (CSF) opening pressure was 35 cmCSF and it showed oligoclonal bands. Acetazolamide and intravenous steroids were initiated.
Conclusions: In our case, the parallel worsening of signs attributed to MS (fourth nuclear palsy and internuclear ophthalmoplegia) and IH (bilateral papilloedema and sixth nerve palsy), suggests a causal relationship between the two clinical entities.
Disclosures: None.

P088

EVALUATING OUTCOMES OF WEIGHT LOSS AND DISEASE REMISSION IN A TERTIARY IDIOPATHIC INTRACRANIAL HYPERTENSION SERVICE

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Background and Aims Idiopathic intracranial hypertension (IIH) is a condition of raised intracranial pressure (ICP), associated with excess body weight. Research has shown that weight loss can reduce ICP symptoms and induce remission. We have set up a tertiary multidisciplinary service to support people with IIH, including a novel addition: online group consultations (GC). We set out to do a quality improvement project to assess the outcomes of weight loss and remission in our IIH service.
Methods: We carried out a retrospective case notes review of consecutive patients (n = 182) presenting to the IIH clinic from 2016–2022 under the care of a single physician (SHW).
Results: IIH was diagnosed in 182 patients, of whom 47.3% attended the traditional one-to-one appointments, and 52.7% transitioned to online GC. There was an overall IIH remission rate of 25.5% achieved, however 57% patients gained weight since diagnosis. Of the patients that attended the GC, the median maximum weight change before and after GC implementation were 2.6 kg and −.5 kg, respectively.
Conclusions: Weight loss is correlated with remission, as demonstrated in research trials. Some patients gain weight following diagnosis. Supporting weight loss is important to reduce the risk of visual loss due to weight gain. Our remission rate of 25.8% is one of the few reports on outcomes of an IIH tertiary service.
Disclosures: None.

P089

DEVELOPING A RODENT MODEL OF IDIOPATHIC INTRACRANIAL HYPERTENSION

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Background and Aims: Idiopathic intracranial hypertension (IIH) is a disease of raised intracranial pressure (ICP) leading to headache and papilloedema. However, we currently lack rodent models to explore the pathogenesis of raised IIH. We aimed to assess the effects of obesity, a feature of IIH, on ICP, and related symptoms in rats.

Methods: Female Sprague-Dawley rats received a high fat diet (60% fat, n = 14) or matched control diet (10% fat, n = 10) for 15 weeks. Following the diet, rats were implanted with telemetric ICP probes. Cutaneous allodynia, a headache feature, was assessed via von Frey filaments. Retinal anatomy was assessed by optical coherence tomography (OCT) and post-hoc histology.
Results: Compared with control rats: obese rats were 15% heavier (p = .002); had similar fasting glucose levels (p = .43); had raised ICP at baseline (2.77 ± .6 versus −.17 ± .7 mmHg, p = .0052), which correlated with abdominal adiposity (r = .54, p = .016); had cephalic cutaneous allodynia (163.1 ± 8.0 versus 213.8 ± 5.1 g, p < .0001); and had swollen retinal nerve fibre layers (RNFL) (28.8 ± .6 versus 24.8 ± 1.1 μm, p = .0026). RNFL thickness correlated with ICP (r = .639, p = .0058). RNFL bundles were thinner in obese rats (74.3 ± 3.6 versus 88.3 ± 3.9 μm, p = .0214). RNFL bundle thickness negatively correlated with RNFL thickness on OCT (r = −.62, p = .022).
Conclusions: Our data highlight that obesity increases ICP, accompanied by cephalic allodynia, papilloedema and retinal atrophy, mimicking IIH. Our unique model will facilitate deeper understanding of the molecular underpinnings of IIH and the development of novel therapeutics to treat IIH.
Disclosures: None.

P090

DETAILED INTRACRANIAL PRESSURE MONITORING IN IDIOPATHIC INTRACRANIAL HYPERTENSION PROBES DIURNAL AND POSTURAL VARIABILITY

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Background and Aims: There is currently limited understanding of the abnormal physiology of raised intracranial pressure (ICP) in those with idiopathic intracranial hypertension (IIH). This study aimed to characterise ICP measures over time and to evaluate ICP with predefined body positions and over a 24 hour period in the setting of active IIH.
Methods: Women with active IIH were recruited to 'IIH-Pressure' (a randomised controlled trial). A telemetric, intraparenchymal ICP monitor was implanted and ICP was recorded during a 24 hour visit and evaluated with changing position. Fifteen participants of mean age 29.5 ± 9.5 years, mean body mass index 38.1 ± 6.2 kg/m^2, and baseline ICP 23.5 ± 3.9 mmHg were included.
Results: At baseline, supine ICP rose over 5 mins and over 30 mins (n = 15). In a detailed analysis, it was observed to further rise out to 3 hours (n = 7). Mean (standard deviation) ICP while supine was 21.2 (4.8) mmHg; this increased in the left lateral position to 24.0 (3.8) mmHg, decreased on sitting to 10.1 (5.1) mmHg, and on standing to 10.3 (3.7) mmHg. Coughing and bending had a marked effect on ICP, both in terms of peak pressure and the amplitude of the waveform (p < .0001, all measures). There was a marked reduction in ICP late in the evening, with a rise in ICP through to 0700hrs (4.02 [1.77] mmHg, p = .02).
Conclusions: These data show novel findings of increasing ICP in supine postures with time. Following a normal day-night routine the night-time period in a recumbent position represents the longest period of exposure to the highest ICP. Overnight rising ICP may play a significant role in symptoms.

Disclosures: Miss Grech reports scientific consultancy fees from Invex therapeutics during the conduct of the study. Professor Mollan reports other Invex Therapeutics, other Heidelberg engineering during the conduct of the study; other from Chugai-Roche Ltd, other from Janssen, other from Allergan, other from Santen, other from Roche, other from Neurodiem, outside the submitted work. Professor Sinclair reports consulting fees and stockholding with Invex therapeutics, during the conduct of the study. Other from Allergan, Amgen, Novartis and Cheisi All other authors declare no competing interests.

P091

ASSOCIATION OF ADRENAL INSUFFICIENCY WITH PAEDIATRIC PSEUDOTUMOUR CEREBRI SYNDROME

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Background and Aims: Paediatric pseudotumour cerebri syndrome pathophysiology is complex and not well delineated. Therefore, it is important to identify potential contributors or targets underlying the primary pathogenesis for its development. To report cases highlighting the association of paediatric pseudotumour cerebri syndrome with adrenal insufficiency.
Methods: This noncontrolled, observational case series included paediatric patients diagnosed with pseudotumour cerebri syndrome and adrenal insufficiency at an urban academic children's hospital in Houston, Texas, from June 2015 to October 2019.
Results: Data were collected from five patients (age range, 5–10 years) diagnosed with pseudotumour cerebri syndrome and adrenal insufficiency. One was a girl; all were white and prepubertal. Three patients had unrecognised glucocorticoid exposure. All patients had bilateral optic nerve oedema that was initially treated with acetazolamide or topiramate, but cortisol functional testing by either 8 am cortisol or cosyntropin stimulation tests revealed a diagnosis of central adrenal insufficiency. Treatment with physiological doses of hydrocortisone resulted in resolution of optic nerve oedema and clinical symptoms of pseudotumour cerebri syndrome, as well as a shorter time receiving medical therapy.
Conclusions: In this case series, adrenal insufficiency was associated with both primary and secondary prepubertal paediatric pseudotumour cerebri syndrome. As a potential target specific to causative mechanism, physiological hydrocortisone therapy resolved the condition. Ophthalmologists and paediatric subspecialists should implement cortisol testing via either 8 am cortisol or cosyntropin stimulation tests at initial evaluation of all children with pseudotumour cerebri syndrome and risk factors for adrenal insufficiency, no predisposing causes, or non-response to conventional treatment. Further management and treatment should be in combination with Ophthalmology and Endocrine services.
Disclosures: None

P092

PREVALENCE OF ADRENAL INSUFFICIENCY AND GLUCOCORTICOID USE IN PAEDIATRIC PSEUDOTUMOUR CEREBRI SYNDROME

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Background and Aims: The pathophysiology underlying pseudotumour cerebri syndrome (PTCS) is complex and not well understood. There are clear differences between PTCS in adults and paediatrics. Few and isolated case reports have suggested that adrenal function may be involved, yet no large cohort study has examined this relationship.
Methods: We conducted a retrospective single-centre study of children who presented with a diagnosis of PTCS and had cortisol testing measured between January 2010 and September 2019. We included all subjects meeting the revised PTCS diagnostic criteria after the chart review. Based on morning, random or 1 μg cosyntropin stimulated cortisol levels, adrenal functioning was classified as: (1) insufficient (peak cortisol < 16 μg/dL and am cortisol < 5 μg/dL); (2) at risk (peak cortisol 16–20 μg/dL, am cortisol 5–13 μg/dL, or random cortisol < 13 μg/dL); or (3) sufficient (peak cortisol > 20 μg/dL and am or random cortisol >13 μg/dL).
Results: A total of 398 individuals were reviewed, and 64 were included for analysis. Of these, 40.6% were male, of mixed race and ethnicity with a mean age of 10.5 (standard deviation 4.7) years. Of these, 23% and 52% had insufficient or at-risk cortisol levels. The majority of those in the insufficient (70%) or at-risk (80%) groups were exposed to topical, nasal, or inhaled glucocorticoids but not systemic. Only 60% and 12% of those with PTCS with insufficient or at-risk cortisol testing, respectively, underwent definitive testing with a stimulation test.
Conclusions: Glucocorticoid use and hypocortisolism are prevalent in PTCS and need consideration as a potential underlying cause. Most children had insufficient or at-risk cortisol levels, and many did not undergo further testing/workup. Children who present with PTCS, particularly young males should be evaluated for adrenal insufficiency and its risk factors, including non-systemic steroids. Prospective studies are necessary to further evaluate the effect of cortisol in relation to paediatric PTCS.
Disclosures: None

NEURO-OPHTHALMOLOGICAL DISORDERS OF NEUROLOGICAL AND SYSTEMIC DISEASES

P093

PROPTOSIS AND OPHTHALMOPLEGIA BY CAROTID CAVERNOUS FISTULA: A CASE REPORT

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Background and Aims: Carotid cavernous fistula (CCF) is an abnormal communication between the carotid arteries and the cavernous sinus. Timely intervention is needed in these cases to prevent morbidity or mortality. We aim to report a case of indirect, spontaneous, and low-flow unilateral CCF.
Methods: We report the case of a 48-year-old man with a history of diabetes mellitus, who had a 3 month history of proptosis and ophthalmoplegia due to a CCF.
Results: We found decreased visual acuity, increased intraocular pressure, pain, chemosis, and ophthalmoparesis in the left eye. The blood glucose was 231 mg/dL. Results of neuro-imaging and digital subtraction angiography showed dilation of the left superior ophthalmic vein with left-to-anterior bulb proptosis, possibly due to an indirect type D carotid-cavernous fistula. After treatment, his vision improved but there was still ophthalmoparesis and mild proptosis. The patient's blood sugar decreased and it was planned they he would undergo coiling but he declined the procedure.

Conclusions: Indirect CCF often occurs spontaneously and is associated with systemic disease, but it generally recovers with conservative management. Other treatments for indirect CCF are radiosurgery and embolisation. It is recommended that clinical and radiological evaluation continue immediately after the procedure and after 6 weeks and 6 months.
Disclosures: None.

P094

FROM UNDETERMINED OPTIC NERVE ATROPHY TO MOHR-TRANEBJAERG SYNDROME: GENETIC TESTING AS THE CLUE WHEN AN UNCERTAIN CLINICAL PICTURE PROGRESSES

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Background and Aims: Mohr-Tranebjaerg syndrome is a rare X-linked genetic disease characterised by childhood hearing loss, progressive visual impairment, neuropsychological disorders such as paranoia, dystonia, and ataxia as a teenager, withevolution to dementia from the 4th decade onward. We report a case in which optic atrophy led to the diagnosis.
Methods: A male of 47-years-old experienced progressive visual loss. He had developed bilateral hearing loss within his first year of life, psychotic crisis and behavioural disturbance consistent with paranoid schizophrenia at 20-years-old, then cervical dystonia at 26-years-old. At 40-years-old he developed mild cognitive impairment and gait ataxia progressing to permanent wheelchair use.
Results: His visual acuity was 20/200 with bilateral pallor of the papillomacular bundle. Visual fields could not be obtained. Fifteen years before, his visual acuity was 20/40, no fundus abnormalities were found and his visual fields were normal. Dystonia had previously been attributed to neuroleptic used for schizophrenia. Presumed Wilson's disease had been treated with a chelating agent as the caeruloplasmin level was found low. Blood tests were normal and neuro-imaging showed no abnormalities. A genetic optic neuropathy panel was ordered with OPA-1 in mind. Nevertheless, a novel genetic variant (c.78_79del) mutation in the TIMM8A gene was detected. This genotype is responsible for Mohr-Tranebjaerg syndrome.
Conclusions: Mitochondrial disorders can be challenging to diagnose if patients are assessed by many specialists because of their multiple manifestations. Genetic analysis is not routinely performed so high clinical suspicion is needed. The detection of unexplained bilateral optic nerve atrophy is enough to warrant genetic investigations.
Disclosures: None.

P095

EVOLUTION AND CONTRIBUTION OF EHEALTH TECHNOLOGIES IN THE REHABILITATION OF PATIENTS WITH LOW VISION DUE TO NEUROLOGICAL DISORDERS

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Background and Aims: In recent years, the use of eHealth technologies has increased considerably worldwide. In this respect, vision can especially benefit from certain advantages offered by these tools, such as larger screen sizes and higher screen resolutions, for the rehabilitation of patients with acquired functional deficits due to different conditions. We aimed to review and categorise the contributions of eHealth technologies in the rehabilitation of patients with neurological vision impairment, as well as to summarise the gaps found in practice in this field of research.
Methods: We carried out a systematic review in the PubMed® and Embase electronic databases.
Results: Current evidence supports the use of eHealth services as feasible options to improve the visual and vision-related quality of life outcomes of patients with low vision and to facilitate the detection of low vision-related adverse events such as falls, which are associated with an increased risk of morbidity and mortality among the elderly. In addition, these tools have the potential to improve healthcare delivery to patients with reduced mobility and living in rural areas. However, more research with larger sample sizes is needed, as most of the studies published to date are not without methodological issues.
Conclusions: In view of the increasing age of the population and the contact and mobility restrictions caused by the COVID-19 pandemic, eHealth technologies are expected to play a key role in the provision of rehabilitation services for people with low vision due to neurological disorders in the near future.
Disclosures: None.

P096

TIMING OF VISION SCREENING AND ASSESSMENT IN AN ACUTE STROKE POPULATION

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Background and Aims: Stroke can cause serious impairments including eye movements disorders, visual field loss, low vision and perceptual difficulties. Issues in service provision relate to visual screening and, thus, identification of visual problems following stroke. Ideally, visual problems should be identified as early as possible after stroke onset. The aim is to present information on the timing and assessment choice of specialist vision screening that is feasible in an acute stroke population.
Methods: Data are presented from the Impact of Visual Impairment after Stroke (IVIS) study, which explored visual consequences of stroke.
Results: A comprehensive visual assessment should include assessment of visual acuity, visual fields, ocular alignment, ocular motility, visual inattention and visual perception alongside documentation of past ocular history and current visual symptoms. Prospective epidemiology research shows that the majority (79%) of stroke survivors can be visually screened within 1 week of stroke onset with the median being assessed within 72 hours of stroke onset. Reasons for not being able to undergo vision screening or assessment are mostly related to severity of stroke where stroke survivors are unable to participate with vision testing.
Conclusions: Early specialist visual assessment is feasible on acute stroke units. Central vision and eye movements are most commonly impaired after stroke. Early visual assessment is important within core stroke assessments, to inform stroke teams, patients and carers about visual status and its functional significance, and facilitate engagement in rehabilitation.
Disclosures: IVIS study funded by the NIHR, UK.

P097

BIOPSY-PROVEN GIANT CELL ARTERITIS ASSOCIATED VISION LOSS IN CHINESE: A CASE SERIES AND REVIEW OF THE LITERATURE

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Background and Aims: Only a few ophthalmological case reports of biopsy-proven giant cell arteritis (BpGCA)-associated vision loss in Chinese have been published in the English literature.
Methods: We describe three Chinese patients with BpGCA presenting with vision loss and a review of the English literature to provide an overview BpGCA-associated blindness in those of Chinese ethnicity.
Results: Three elderly Chinese patients were referred due to BpGCA-associated vision loss. In the English literature review, 11 cases (17 eyes) of BpGCA-associated vision loss in Chinese found and summarised. Of the 14 cases (including ours), the median age at diagnosis was 77 years, and nine (61.5%) were males. The most common extraocular manifestations were temporal artery abnormalities, headache, jaw claudication, and scalp tenderness. Two patients had occult GCA. All patients had glucocorticoids for treatment. Thirteen (56.5%) eyes had visual acuity of no perception of light at the initial visit and failed to respond to the treatment.
Conclusions: Although extremely rare in elderly Chinese patients with ocular ischaemic diseases, such as amaurosis fugax, anterior ischaemic optic neuropathy, posterior ischaemic optic neuropathy, central retinal artery occlusion, or ocular ischaemic syndrome, the diagnosis of GCA must be considered to avoid irreversible blindness.
Disclosures: None.

P098

OPTIC NEURITIS CAUSED BY ACUTE LYMPHOCYTIC LEUKAEMIA

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Background and Aims: Optic neuritis refers to inflammation of the optic nerve. It can cause vision loss due to swelling and destruction of the myelin sheath or direct axonal damage to optic nerve. Multiple sclerosis (MS) is the most common aetiology in adults, whereas in children it is often para-infectious. This report present an optic neuritis case due to leukaemia.
Methods: A 9-year-old boy presented with blurry vision in both eyes for the last 1 month, without any headache or ocular pain. His right eye (OD) visual acuity was 1/60 and his left eye (OS) had no light perception (NLP). Funduscopy revealed papilloedema in both eyes.
Results: There were no infection signs on laboratory results and no signs suggestive of MS, neuromyelitis optica spectrum disorder or acute disseminated encephalomyelitis on magnetic resonance imaging (MRI). In addition, no retrobulbar mass or intracranial space occupying lesion, infarction, haemorrhage, or increased intracranial pressure was found. He was treated with intravenous methylprednisolone 125 mg, four times per day for 3 days, followed by oral methylprednisolone 1 mg/kg, then a tapering dose. The visual acuity improved to 6/6 in each eye on the

sixth day. His visual acuity worsened again after 2 months to right eye 2/60 (OD) and NLP (OS). Bone marrow aspiration results showed acute lymphocytic leukaemia and spinal MRI showed metastases to the mediastinum. This recurrence may have indicated infiltrative optic neuritis.
Conclusions: This is a case of optic neuritis in which there was a significant visual acuity improvement after methylprednisolone, but with worsening after 2 months. Optic neuritis, which responds well to steroids but then recurs, should also be suspected as a result of an infiltrative malignancy, as demonstrated in this case.
Disclosures: None.

P099

A DIFFERENTIAL LOSS OF NERVE FIBRE LAYER THICKNESS AND RETINAL GANGLION CELL COMPLEX IN TOXIC AND NUTRITIONAL OPTIC NEUROPATHY

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Background and Aims: Toxic or nutritional optic neuropathies cause painless progressive vision loss, with central or caecocentral scotomas. Toxic aetiologies include antibiotics, chemotherapies, heavy metals, tobacco, etc. Nutritional causes are often due to deficiencies in B-complex vitamins or folic acid. These neuropathies result in papillomacular bundle damage and retinal nerve fibre layer (RNFL) loss. We present five cases of toxic and nutritional optic neuropathy.
Methods: All of the patients presented with reduced visual acuity and colour vision, had central or cecocentral scotomas, and had a history of alcohol abuse, tobacco use, or ethambutol exposure, with or without a nutritional deficiency.
Results: They all had diffuse, early loss of the retinal ganglion cell (RGC) layer, with relative preservation of the RNFL.
Conclusions: Prior studies have shown that these neuropathies can cause RGC loss, however our cases reveal that toxic substances or nutritional deficiencies may have a propensity to affect the RGC body first before the RNFL is affected. Mitochondrial dysfunction, caspase activity, and neurotrophin deficiency are implicated in RGC death. Toxins or nutritional deficiencies may affect these functions, or RGCs may have predisposing factors causing susceptibility to early death. Optical coherence tomography imaging of the RGC layer could identify toxic or nutritional optic neuropathies before optic nerve atrophy or RNFL loss are evident.
Disclosures: None.

P100

A REVIEW OF THE CLINICAL AND IMAGING FINDINGS IN MYELIN OLIGODENDROCYTE GLYCOPROTEIN ANTIBODY POSITIVE PATIENTS PRESENTING TO KINGS COLLEGE HOSPITAL, LONDON

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Background and Aims: The spectrum of myelin oligodendrocyte glycoprotein (MOG) antibody associated disease is emerging and associated clinical phenotypes include optic neuritis (ON), longitudinally extensive transverse myelitis (LETM), acute disseminated encephalomyelitis (ADEM) like presentation, and cortical encephalitis.
Methods: We identified all patients testing positive for MOG antibodies at King's College Hospital between January 2015 and August 2019 using Cogstack, our clinical analytics system, and reviewed their clinical phenotype, imaging and cerebrospinal fluid (CSF) findings.
Results: Twenty-two patients tested positive for MOG antibodies, of whom 11 were females and the mean age was 39 (standard deviation [SD] 18) years. The majority of the patients (15/22) presented with ON: 3 of whom had were bilateral ON and 2 had bilateral sequential ON. Other presentations included LETM (n = 2), ADEM (n = 2), brain stem syndrome (n = 1), lower cord/conus syndrome (n = 1), and seizures (n = 1). The mean logMAR visual acuity at nadir in patients presenting with ON was .69 (SD .58). Mean logMAR visual acuity at recovery was .1 (SD .54). Magnetic resonance imaging of the orbits was performed in 13 patients, of whom four had bilateral optic nerve involvement and five had longitudinally extensive optic nerve lesions.
Conclusions: The clinical and imaging presentation of MOG antibody positive patients presenting to King's College Hospital was similar to previously reported cohorts with the majority presenting with ON. Although the majority of patients made a good recovery five patients relapsed and two patients had poor visual outcomes.
Disclosures: None.

P101

CONCURRENT INTRAOCULAR AND ORBITAL LYMPHOMA OF CUTANEOUS T-CELL LYMPHOMA ORIGIN

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Background and Aims: Cases of primary and secondary intraocular lymphoma are usually of non-Hodgkin B-cell origin. Intraocular T-cell lymphoma is extremely rare. Orbital lymphomas are also mainly of non-Hodgkin B-cell origin, with only 3–11% being of T-cell origin. Furthermore, in cases of orbital lymphoma, under 13% are known to cause extraocular muscle enlargement.
Methods: Herein we describe a case of an elderly male with stage IV CD8+ cutaneous T-cell lymphoma, who developed simultaneous orbital and intraocular metastasis.
Results: A 64-year-old male with recently diagnosed metastatic cutaneous T-cell lymphoma, being treated with CHOP (cyclophosphamide, doxorubicin [Adriamycin], vincristine [Oncovin] and prednisolone), presented with horizontal binocular diplopia and blurry vision in both eyes that had been slowly progressive over the last few months. Examination revealed 20/100 vision in each eye, bilateral proptosis, a left-sided adduction deficit, and bilateral intermediate uveitis. Magnetic resonance imaging showed bilateral fat stranding and extraocular muscle enlargement. A lumbar puncture revealed lymphocytic predominance of the detected cells. A vitreous biopsy was performed and showed 52.4% atypical T cells, positive for CD2, CD3, CD8, and CD45, confirming the diagnosis of CD8 + T-cell lymphoma.
Conclusions: Our case is a unique example of concurrent intraocular and orbital lymphoma, which are rarely of T-cell origin. Additionally, secondary intraocular lymphomas usually metastasise to the uvea, while primary intraocular lymphomas frequently involve the vitreous. This case also illustrates the uncommon finding of extraocular muscle enlargement in orbital lymphoma.
Disclosures: None.

P102

IS IT THE GRAFT OR THE HOST?

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Background and Aims: Patients with complex systemic disease often present to Neuro-ophthalmology with diagnostic challenges.
Methods: We describe the case of a 60-year-old white Caucasian man, with a background of myelodyplastic syndrome, who underwent allogeneic stem cell transplantation in 2015 and had subsequently been diagnosed with chronic graft versus host disease (GVHD). He presented with high inflammatory markers, mild but progressive confusion, hallucinations, and features of bilateral optic neuropathy.
Results: His visual acuity was 6/36 in each eye, with no relative afferent pupillary defect and his optic nerve heads had mild temporal pallor. He had central visual blurring and poor colour vision. Neuroimaging was attempted but was limited by motion artefact. A lumbar puncture was performed, which showed raised protein (.79 g/L), but no other evidence of infection. Initially he was felt to have GVHD-related optic neuropathy. Intravenous aciclovir and meropenem were given, but his confusion worsened. An electroencephalogram showed global slowing. A differential diagnosis of progressive multifocal leukoencephalopathy was made. He underwent magnetic reasonnce imaging under sedation, which showed a differential of brain abscess/neoplasm. A biopsy was undertaken.
Conclusions: This case highlights the diagnostic dilemma and subsequent workup in patients with complex systemic disease, such as haematological malignancies and GVHD. Repeat investigations may be necessary when the diagnosis is unclear.
Disclosures: None.

P103

HORNER'S SYNDROME IN THE CONTEXT OF A SPONTANEOUS INTERNAL CAROTID ARTERY DISSECTION WITH 2 WEEKS OF EVOLUTION: A CASE REPORT

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Background and Aims: Horner's syndrome results from the interruption of the sympathetic innervation to the eye and can be a sign of multiple underlying disorders. Our purpose is to report the case of a Horner's syndrome in the context of a 2 week history of spontaneous internal carotid artery dissection.
Methods: This is a case report of a 53-year-old man observed in Centro Hospitalar Universitário do Porto. Horner's syndrome was confirmed with an aproclonidine test. Imaging studies were performed with cerebral computed tomography (CT) and a CT angiography of the cervical and supra-aortic vessels.
Results: The CT angiogram demonstrated an internal carotid artery dissection.
Conclusions: Horner's syndrome can be the presenting sign of a life-threatening condition. When associated with head or neck pain the hypothesis of a carotid dissection must always be excluded, even in the absence of known risk factors or in the presence of a long duration of symptoms, as in this patient's case.
Disclosures: None.

P104

GENOTYPE-PHENOTYPE CORRELATIONS IN AUTOSOMAL DOMINANT OPTIC ATROPHY

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Background and Aims: Autosomal dominant optic atrophy (DOA) is the commonest inherited optic neuropathy. Limited natural history data on DOA progression present a major challenge in establishing outcome measures for therapeutic studies. This study sought to establish the natural history of DOA in a large patient cohort with a confirmed molecular diagnosis.
Methods: This multicentre study included DOA patients carrying a pathogenic or candidate pathogenic OPA1 variant. Visual acuity (VA), visual fields, and optical coherence tomography (OCT) data were collected from clinical records. OPA1 variant data were retrieved from diagnostic reports.
Results: Of the 304 patients recruited into this study, 282 had isolated DOA and 22 had systemic features (DOA+). A missence variant was present in 45.5% of DOA+ patients compared with 20.6% of DOA patients (odds ration 3.22; $p < .01$). The mean (± standard deviation) rate of visual decline was .035 ± .162 logMAR/year. Final logMAR VA was significantly worse in the DOA+ group versus the DOA group (1.544 ± .817 versus .802 ± .441; $p < .001$), in patients who had a missense variant versus a loss of function variant (1.208 ± .512 versus .758 ± .470; $p < .001$), and in patients with a variant in the GTPase domain versus other functional domains (.985 ± .535 versus .751 ± .473; $p < .001$).
Conclusions: DOA is a genetically and phenotypically heterogenous disorder. We have identified several genotype-phenotype correlations that can be used for patient counselling and clinical trial design. The rate of visual decline in DOA is influenced by the underlying OPA1 variant and correlates with the presence of systemic "plus" features.
Disclosures: None.

P105

RARE CASE OF INTRACRANIAL HYPERTENSION SECONDARY TO ISOLATED CENTRAL NERVOUS SYSTEM B-CELL ACUTE LYMPHOBLASTIC LEUKAEMIA

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Background and Aims: Investigation of intracranial hypertension is important for appropriate management. Acute lymphoblastic leukaemia accounts for approximately 15–20% of all adult leukaemias but only 10% of these present with central nervous system (CNS) involvement. To our knowledge, this is the first reported case of papilloedema as sign of an undiagnosed isolated CNS lymphoblastic leukaemia. The aim of this report is to present the role of perimetry, diagnostic work-up and management of the case.
Methods: We present a case report of a previously healthy, 26-year-old male initially managed as a case of idiopathic intracranial hypertension with Frisén grade 4 papilloedema and enlarged blindspots on perimetry.
Results: Baseline perimetry was obtained. Cerebrospinal fluid (CSF) flow cytometry was performed and revealed B-cell lymphoblasts with a negative systemic leukaemia work-up. He was managed as intracranial hypertension secondary to a primary CNS leukaemia. His visual acuity and visual fields markedly

deteriorated, prompting urgent lumbar drain placement to temporarily address the recalcitrant IH. Serial perimetry was used to monitor for functional changes. Intrathecal and systemic chemotherapy with the hyperCVAD regime (hyperfractionated cyclophosphamide, vincristine, doxorubicin [also known as, adriamycin] and dexamethasone) successfully eliminated the CSF blast cells allowing for a permanent lumboperitoneal shunt placement, resulting in relief of the patient's symptoms and improvement of papilloedema and vision.
Conclusions: Detection of papilloedema in the absence of neuroimaging findings warrants workup with CSF cytology and flow cytometry to investigate for malignancy. We hypothesise that the infiltration of the leukaemic blast cells interfered with CSF outflow resulting in intracranial hypertension. Appropriate management was initiated and, in our case, lifesaving.
Disclosures: None.

P106

NEURO-OPHTHALMOLOGIC MANIFESTATIONS OF MULTIPLE MYELOMA: TWO CASES

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Background and Aims: Multiple myeloma (MM) is a plasma cell malignancy and the second most frequent haematological cancer. It is a progressive and incurable disease that affects the bone marrow and can affect any extramedullary tissue. Visual manifestations are rarely but the most frequent are secondary to orbital involvement. The aim of this report is to describe the neuro-ophthalmological complications of MM, which can be the first sign of a systemic disease, or it can appear in the course of the disease as an extramedullary extension of the malignancy.
Methods: We present two cases of MM who presented with visual symptoms.
Results: The first case was a 53-year-old man who consulted for proptosis with diplopia and blurry vision in his left eye (LE). He had decreased visual acuity, proptosis, ophthalmoplegia and optic disc oedema in the LE. Orbital magnetic resonance imaging (MRI) showed a left maxillary sinus neoplasm with orbital extension and a biopsy confirmed a plasma cell malignancy. The second case was a 55-year-old man with a history of treated MM who consulted due to diplopia and right proptosis. He had restriction of his right eye on horizontal movements. An MRI showed a plasmacytoma in the right lateral rectus muscle.
Conclusions: Ophthalmological manifestations of MM are infrequent. The most important causes of visual disturbances are plasma cell infiltration of the visual tissues or the complications secondary to haematological anomalies. Neuro-ophthalmological complications can be the first sign of a systemic disease or the extension of a known disease.
Disclosures: None.

P107

PROGRESSIVE THIRD NERVE PALSY SECONDARY TO CAVERNOUS CAROTID ANEURYSM, MISDIAGNOSED AS THYROID EYE DISEASE

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Background and Aims: Cavernous carotid aneurysms (CCAs) account for 2–9% of all intracranial aneurysms. Most patients with CCAs are asymptomatic, and the lesions are most often discovered incidentally on intracranial imaging acquired during the work-up for unrelated conditions. Symptomatic CCAs can occur due to mass effect, rupture and thrombosis. The most common symptoms of mass effect from an unruptured CCA are diplopia and pain. Diplopia can result from isolated oculomotor nerve palsies such as third and sixth cranial nerve paresis.
Methods: We present the case of an 88-year-old woman who had gradually worsening vertical diplopia
Results: She had previously been diagnosed several years ago with thyroid eye disease. She developed a partial pupil-sparing right third nerve palsy. Her thyroid status was stable but a noticeable right proptosis was noted so magnetic resonance imaging (MRI) was carried out. This scan showed a 14 mm right CCA which impinged on the third nerve. She was referred to the neurosurgeons and it was decided to manage her conservatively.
Conclusions: CCAs are uncommon and rarely cause symptoms. Her abnormal eye movements had been previously mistakenly diagnosed as thyroid eye disease despite no clinical evidence of thyroid pathology. MRI should be considered when assessing a new onset diplopia. Finally, clinicians should rely on eye movement assessment to distinguish between restrictive versus paralytic causes
Disclosures: None.

P108

OPHTHALMOPLEGIA AS A FEATURE OF ATYPICAL HSV TYPE 2 BRAINSTEM ENCEPHALITIS: A CASE REPORT

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Background and Aims: Herpetic encephalitis is mostly caused by Herpes simplex virus (HSV) type 1. Rarely, it can be caused by HSV type 2. Symptoms can include headache, fever, altered consciousness, neck stiffness, seizures, and changes in behaviour. Ophthalmoplegia is an uncommon manifestation of this condition.
Methods: A 78-year-old woman presented with a 6 day history of severe headache, generalised fatigue, photophobia, ocular pain and double vision. She had systemic hypertension and polymyalgia rheumatica and she was on long term treatment with 2 mg oral prednisolone per day.
Results: Clinically, there was a left pupil-sparing partial third nerve palsy and bilateral sixth nerve palsies. There were no signs of ptosis or fatiguability. Her Glasgow coma scale score was 15/15. Upper and lower limb examination was normal, with no areflexia or ataxia. Blood tests ruled out giant cell arteritis. A computed tomography scan of her head, magnetic resonance (MR) imaging of her brain MR angiography were normal. After performing a lumbar puncture, polymerase chain reaction (PCR) of cerebrospinal fluid (CSF) showed the presence of HSV type 2. She was diagnosed with HSV type 2 brainstem encephalitis and was treated with intravenous aciclovir, with significant improvement of her general medical and ophthalmological symptoms and resolution of the diplopia.
Conclusions: Ophthalmologists should include herpetic encephalitis in the list of differential diagnoses of acute ophthalmoplegia, especially if neuro-imaging is unremarkable . Moreover, the lack of cognitive impairment did not make clinicians think of encephalitis at first. This case emphasises the role of PCR of CSF to confirm the diagnosis of HSV type 2.
Disclosures: None.

P109

CHILDHOOD-ONSET LEBER'S HEREDITARY OPTIC NEUROPATHY – CLINICAL AND PROGNOSTIC INSIGHTS

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Background and Aims: Childhood Leber's hereditary optic neuropathy (LHON) is distinct from the adult form as it has a better visual prognosis and a more varied clinical presentation. It is often insidious, subclinical, slowly progressive and, in some cases, unilateral. The atypical age of onset and non-classical patterns of visual loss frequently results in significant diagnostic delays with initial misdiagnoses.

Methods: We carried to a cross-sectional retrospective study. The natural history of childhood-onset LHON in a large cohort of affected children was assessed and a classification system based on both the age and pattern of visual loss was proposed. Ophthalmological evaluation included best-corrected visual acuity, orthoptic evaluation, slit-lamp biomicroscopy, visual field and optical coherence tomography (OCT).

Results: Sixty-eight patients were stratified based on the age of onset of visual loss into: Group 1 at < 3 years, n = 14 (20.6%); Group 2 at ≥ 3 to < 9 years, n = 27 (39.7%); and Group 3 at ≥ 9 to ≤ 12 years, n = 27 (39.7%). Based on the pattern of visual loss, patients were categorised as: subacute bilateral, n = 54 (66.7%); insidious bilateral, n = 14 (17.3%); unilateral, n = 9 (11.1%); and subclinical bilateral, n = 4 (4.9%). Patients in Group 2 achieved a better visual outcome compared with those in Group 3. The mean ganglion cell layer thickness was highest in Group 2.

Conclusions: Children who lose vision from LHON before the age of 9-years-old have a better visual prognosis compared with those who become affected in later years, likely representing a 'form fruste' of the disease.

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P110

OPTIC DISC OEDEMA IN MCCUNE-ALBRIGHT SYNDROME

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Background and Aims: Craniofacial polyostotic fibrous dysplasia, as part of McCune-Albright syndrome, can have severe complications including vision loss. Also, patients with this syndrome are at greater risk of secondary intracranial pressure elevation due to medication side effects.
Methods: A 6-year-old girl with McCune-Albright syndrome and polyostotic craniofacial fibrous dysplasia and optic canal narrowing, developed signs of slowly progressive optic nerve compression on clinical examination including deteriorating visual acuity, positive relative afferent pupillary defect and bilateral optic disc swelling.
Results: Imaging using optical coherence tomography (OCT) revealed progressive retinal nerve fibre layer thickening. Prior to deterioration, the dose of triptorelin, a gonadotrophin-releasing hormone analogue, she was treated with for precocious puberty, had been increased. Medication cessation was followed by improvement in clinical and imaging findings.
Conclusions: McCune-Albright syndrome patients with craniofacial involvement and/or gonadotrophin-releasing hormone analogue treatment should be regularly monitored for clinical signs of optic neuropathy together with routine OCT imaging.
Disclosures: None.

P111

MOLECULAR INVESTIGATION ENDS THE DIAGNOSTIC ODYSSEY FOR FOUR FAMILIES WITH CHRONIC OPTIC NERVE SWELLING

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Background and Aims: Retinal dystrophy, optic nerve (ON) oedema, splenomegaly, anhydrosis and headaches (ROSAH) syndrome has been associated with a specific heterozygous variant, c.710C>T, p.(Thr237Met), in the ALPK1 gene. We report the clinical and molecular findings of four families, including a novel variant affecting the same codon, c.709A>G, p.(Thr237Ala).

Methods: We report the molecular cause of previously unsolved retinal disease patients as part of the NIHR BioResource Rare-Disease and United Kingdom's 100,000 Genomes Projects. Ultra-rare variants (minor allele frequency < .0001) in the ALPK1 gene were interrogated. An additional case was identified by exome sequencing.
Results: Four families with the mutations p.(Thr237Met), n = 3, and p.(Thr237Ala), n = 1, were identified. Pedigrees assessment led to the identification of 10 individuals (five females and five males) sharing the phenotype. The median age was 21 years (range 10–66, n = 9) at ascertainment and 14 years (range 3–32, n = 7) at ocular pathology presentation. Visual acuities ranged from 6/60 to 6/6 and were worse in older individuals. Optic nerve head swelling and outer retinal degeneration occurred in all patients with evidence of progression of the latter. Ocular inflammation, such as anterior uveitis (n = 1), intermediate uveitis (n = 2), pan-uveitis (n = 3) and vitritis (n = 1) was noted. Systemic findings included anhydrosis, splenomegaly, headaches, and the inability to breast-feed (in two adult females).
Conclusions: ALPK1 mutations should be considered in those presenting with unexplained chronic optic nerve head swelling and retinal degeneration, and systemic features should be sought. The progressive nature of the disorder and the involvement of the gene-product in the NF-kappaB signalling pathway encourages further investigation of the mitigating effect of immunomodifying therapies.
Disclosures: None.

P112

OPTIC NERVE HEAD FINDINGS AS THE KEY FEATURE PROMPTING GENETIC DIAGNOSIS IN A FAMILY WITH NOONAN SYNDROME

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Background and Aims: Noonan syndrome (NS) is a genetically heterogeneous, mostly dominantly inherited developmental disorder, characterised by short stature, facial dysmorphia, and congenital heart disease. Patients frequently show various ocular abnormalities. We describe a family featuring abnormal optic nerve head configuration as the key symptom leading to genetic diagnosis of NS.
Methods: We conducted full eye examinations including measurement of peripapillary retinal nerve fibre layer (pRNFL) thickness and macular ganglion cell layer (mGCL) thickness using optical coherence tomography (OCT). Genetic analysis was performed by exome sequencing in the index patients, and subsequent confirmation and segregation analysis in their parents by conventional deoxyribonucleic acid-sequencing.
Results: Two brothers, aged 11 and 14 years, were referred to our clinic because of suspected hereditary optic atrophy. Both siblings and their mother had congenital heart defects and mild facial dysmorphic features. Only the boys had short stature and one was highly hyperopic. Their anterior segments were normal, but all six eyes had abnormal optic discs with enlarged and deep excavations. OCT detected mildly thinned pRNFL (77–86 μm) and mGCL (31–33 μm). At the last visit, visual acuity was below 16/20 in all patients. Genetic analysis revealed the heterozygous missense mutation c.417 G > C p.(Glu139Asp) within the PTPN11 gene of both children and their mother.
Conclusions: Information in this pedigree is compatible with a diagnosis of autosomal dominant NS1 (OMIM #163950) caused by a mutation in PTPN11, which accounts for about 50% of NS cases. This is the first report on pRNFL and mGCL loss in NS using OCT.
Disclosures: None.

P113

MYLEOPROLIFERATIVE NEOPLASIA MASQUERADING AS IDIOPATHIC INTRACRANIAL HYPERTENSION: A CASE REPORT

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Background and Aims: We present a case report of essential thrombocythaemia presenting with idiopathic intracranial hypertension (IIH).
Methods: A 44-year-old woman with no significant past medical history except recently diagnosed hypertension was found to have bilateral optic disc swelling on routine optometry assessment. She reported no tinnitus, transient visual obscurations, focal neurology, or fever. She had a body mass index of 26.2 kg/m^2.
Results: Goldmann visual fields were normal with and she had normal visual acuity. Magnetic resonance (MR) imaging showed normal brain structure but signs of raised intracranial pressure. MR venography was unremarkable. Blood tests were normal except for a creatinine level 94 mg/dL and a platelet count of 490×10^9/L. Lumbar puncture opening pressure was 33 cmCSF and a diagnosis of IIH of unknown aetiology was made. She subsequently received intravitreal anti-vascular endothelial growth factor injection for a peripapillary choroidal neovascular membrane. A computed tomography scan of her abdomen found splenomegaly, small volume lymphadenopathy and skeletal mottling. She was referred to haematology who confirmed a diagnosis of essential thrombocythaemia on bone marrow trephine. She was commenced on aspirin and continues with regular monitoring of the papilloedema.
Conclusions: Myeloproliferative disorders are a cause of papilloedema, often secondary to venous sinus thrombosis predisposed by the associated hypercoagulable state. However, raised intracranial pressure could not be attributed to venous sinus thrombosis here and an alternative hypothesis had to be sought.
Disclosures: None.

P114

A RARE CAUSE OF PAPILLOEDEMA IN A RENAL PATIENT

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Background and Aims: A 30-year-old male, renal transplant patient presented with a generalised tonic clonic seizure. He had been commenced on erythropoietin (EPO) prior to the seizure. Magnetic resonance imaging (MRI) suggested posterior reversible encephalopathy syndrome (PRES). Due to ongoing headaches, an ophthalmological review was sought. This revealed bilateral optic disc swelling suggestive of papilloedema.

Methods: His optic nerve function was normal. The neuro-ophthalmology assessment prompted an urgent review of his neuroimaging, which confirmed enhancement of his leptomeninges (but not optic nerves), not typical for PRES. EPO and ciclopsorin were withdrawn by the renal team. He continued to be treated with mycophenolate and prednisolone.
Results: Investigations and results are summarised: MRI brain x 2 – progression of leptomeningeal enhancement over a 2 week period; computed tomography of thorax, abdomen pelvis – no evidence of malignancy; Epstein-Barr virus (EBV) IgG positive in serum; lumbar puncture (LP) (#1) opening pressure 27 cm cerebrospinal fluid (CSF) with CSF white cell count (WCC) of 39 (100% lymphocytes), raised protein, infection screen negative, cytology negative; LP (#2) opening pressure 28 cmCSF, with CSF WCC 45 (100% lymphocytes), raised protein, EBV positive and cytology negative; and a brain biopsy showed polymorphic post-transplant lymphoproliferative disorder (PTLD) and the latent membrane protein 1 was positive. Treatment included withdrawal of mycophenolate and 4 weekly rituximab infusions. His neurological signs and symptoms resolved.
Conclusions: PTLD is an uncommon, yet potentially serious complication of transplant medicine. The central nervous system is a rare site of proliferation and is difficult to diagnose. PTLD is a very rare cause of papilloedema but should be considered in patients with a transplant history suspected of harbouring this neuro-ophthalmological malady.
Disclosures: None.

P115

EYE KNOW WHY YOU HAVE LOST WEIGHT

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Background and Aims: A 75-year-old female was referred to the neuro-ophthalmology clinic with a 3 month history of deteriorating vision secondary to bilateral swollen optic nerve heads and a mild vitritis. She also complained of headaches and unintentional weight loss. Previous neuroimaging had revealed extensive white matter changes suggestive of severe ischaemia.
Methods: Her medical history included rheumatoid arthritis (not receiving immunomodulatory treatment) and hypertension. She had a 50 pack year smoking history.
Results: Results on admission included: computed tomography (CT) scan of her thorax, abdomen and pelvis with no evidence of malignancy; blood tests showed a raised beta-2 microglobulin level normal myeloma screen and the collapsin response-mediator protein-5 (CRMP-5) paraneoplastic antibody was pending; a vitreous biopsy showed no evidence of lymphoma; and the lumbar puncture opening pressure was 10 cm cerebrospinal fluid (CSF) with 7 white cells (100% lymphocytes) in the CSF and unmatched oligoclonal bands. She was prescribed oral prednisolone, which caused resolution of the optic nerve swelling and overall well-being. During outpatient follow up, results of the paraneoplastic antibody CRMP5 became available and was positive. A positron emission tomography (PET) scan showed mediastinal and hilar lymphadenopathy. A lung biopsy confirmed small cell lung cancer. She was treated with etoposide and carboplatin with adjuvant thoracic radiotherapy. There was no recurrence on follow-up.
Conclusions: The presence of the onconeural paraneoplastic antibody CRMP-5, led to the diagnosis of limited stage small cell lung cancer (Tx N2 M0) confirmed by PET scanning, that was not detected on CT initially. The treatment of which had led to complete resolution without recurrence.
Disclosures: None.

P116

SINUS ANOMALIES AND STENT-INDICATION FINDING: A "TYPICAL" CASE REPORT

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Background and Aims: Idiopathic intracranial hypertension (IIH) is a rare and difficult to diagnose and treat entity that sits between neurology, ophthalmology and neurosurgery. A retrospective case report is described.
Methods: I saw a 38-year-old woman with changing anisocoria left > right, flicker in the left eye, a feeling of retrobulbar tension, pulsatile tinnitus, holo-cephalic headaches, arterial hypertension, hypothyroidism and severe obesity since the age of 11-years-old.
Results: Examination showed good visual acuity, but severe papilloedema in both eyes. She had worsening of her visual field defects and hypometric saccades to the left following a lumbar puncture. Computed tomography and magnetic resonance (MR) imaging showed typical findings of IIH with an empty sella and slit ventricles. MR venography showed venous anomalies with rectus sinus stenosis proximally and transversal sinus/sigmoid sinus stenosis on both sides. After a lumbar puncture and intracranial pressure release, the headaches decreased.
Conclusions: The therapeutic options of IIH are all difficult and not very successful. However, in this case the venous sinus anomalies provide an optimal therapeutic option by stenting. This minimally invasive intervention had a good prognosis in this case.
Disclosures: None.

P117

REPORT OF BILATERAL SYMMETRIC VISUAL FIELD DEFECTS AND NERVE FIBRE LAYER THINNING ASSOCIATED WITH BRANCHIO-OTO-RENAL SYNDROME

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Background and Aims: Branchio-oto-renal (BOR) syndrome is a condition that affects the development of tissues in the neck and causes malformations of the ears and kidneys. This syndrome has variable presenting symptoms and signs. We report a new association with BOR syndrome.
Methods: We present the results of a full ophthalmological examination in addition to Humphrey visual fields and ocular coherence tomography of the nerve fibre layer in a patient with established BOR syndrome.
Results: Symmetrical visual field defects and retinal nerve layer thinning were detected.
Conclusions: We reported a novel association with BOR syndrome in the form of symmetrical visual field defects in an adult male patient. This sign has not been reported previously amongst BOR syndrome patients.
Disclosures: None.

P118

OPTIC NERVE NEUROSARCOID: ONE PATHOLOGY, TWO MECHANISMS FOR VISUAL LOSS IN THE SAME PATIENT

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Background and Aims: Optic neuropathy is the most common neuro-ophthalmological presentation of sarcoidosis. The neuropathy can occur in many forms including neuritis, perineuritis, granuloma formation, infiltration and ischaemic optic neuropathy. Papilloedema may be seen in cases of intracranial cerebrospinal fluid outflow obstruction.
Methods: We report a rare case of bilateral optic nerve involvement in a 53-year-old male with biopsy proven sarcoidosis who presented with bilateral sequential visual loss. Interestingly, the mechanism for loss of vision was different in both eyes; vascular and inflammatory and could be differentiated by the clinical presentation.
Results: In the right eye, the patient had acute painless visual loss with an inferior altitudinal defect obeying the horizontal midline, bearing the clinical hallmarks of a vascular presentation. The optic disc was 'lumpy' with visible granulomas and there was no improvement despite systemic steroid therapy. However, in the left eye, the visual loss was more gradual in onset with generalised visual field loss. The optic nerve was diffusely swollen with peripapillary nerve fibre layer haemorrhages and associated subfoveal fluid. The visual acuity and swelling improved significantly with steroid therapy, in keeping with an inflammatory presentation.
Conclusions: This case demonstrates two mechanisms for visual loss due to optic nerve head sarcoidosis in the same individual. The mechanisms can be differentiated by the clinical presentation, disc appearances and optical coherence tomography findings. An understanding of mechanism for the optic neuropathy may explain the visual prognosis and response to therapy.
Disclosures: None.

P119

DYNAMICS AND TREATMENT RESPONSE OF COMPARTMENTALISED SARCOIDOSIS USING LONGITUDINAL HIGH RESOLUTION RETINAL OPTICAL COHERENCE TOMOGRAPHY

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Background and Aims: Ocular sarcoidosis typically presents to the uveitis or neuro-ophthalmological service. Herein we describe the case of a 70-year-old female, with a recent history of appendiceal low-grade mucinous neoplasm, who developed profound right eye visual loss to hand movement perception over 2 weeks with unilateral optic disc swelling and optic neuropathy.

Methods: Optical coherence tomography (OCT) demonstrated the dynamics of vertically migrating intraretinal hyper-reflective foci, subretinal fluid, choroidal thickening and vitreous haze. Intracranial and orbital imaging, blood tests, lumbar puncture and whole-body positron emission tomography magnetic resonance imaging excluded neoplastic, infiltrative, inflammatory, infectious and known auto-immune causes.
Results: Given the hilar lymphadenopathy on a chest computed tomography scan, elevated serum angiotensin converting enzyme level, raised urinary calcium to creatinine ratio and eosinophilia, plus her ocular phenotype of right vitreous haze, optic disc swelling and neuropathy with thickened choroid, we made a clinical diagnosis of sarcoidosis. Treatment with corticosteroids led to rapid improvement of symptoms and the final visual acuity was .3 with pinhole, with 10/17 of the Ishihara plates correctly identified, although a central scotoma remained.
Conclusions: OCT permits dynamic monitoring of the inflammatory response on a cellular level between three adjacent compartments and a literature review suggests that intraretinal sarcoidosis with migrating hyper-reflective foci may be a novel, OCT-supported manifestation of this systemic disease.
Disclosures: None.

P120

VISUAL FUNCTION AND OPTICAL COHERENCE TOMOGRAPHY DATA IN THE MANAGEMENT OF OPTIC PATHWAY TUMOURS

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Background and Aims: The management of optic pathway tumours by the neuro-oncologists requires a multi-disciplinary approach with input from radiology and ophthalmology. The decision to treat or observe is based on deterioration of visual function and/or the progression of disease on magnetic resonance imaging (MRI). Assessment of visual function is of paramount importance, but this can be a challenging task in many children. This retrospective study analyses the use of optical coherence tomography (OCT) of the retinal nerve fibre layer (RNFL) and ganglion cell layer (GCL) in determining progression of disease.
Methods: We reviewed the case notes, MRI and OCT data of 30 children who were diagnosed with optic pathway tumours at Queens Medical Centre over the last 9 years (2013–2022).
Results: Gradual loss of RNFL and GCL thickness can occur without progression of the tumour or visual function. Acute thinning of RNFL and GCL suggest progression of the tumour, however there are patients who retain good vision despite 59% thinning of the global RNFL.
Conclusions: Our data show that gradual thinning (< 2 μm per year) of global RNFL thickness possibly suggests non-progression of the tumour. Rapid thinning of the RNFL suggests progression despite normal visual function. OCT findings help with the assessment, but cannot totally replace clinical assessment of visual function.
Disclosures: None.

P121

BRUNS SYNDROME – BROADENING THE PHENOTYPE

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Background and Aims: We present a 50-year-old woman with 6 month history of episodes of suddenly feeling weak with her legs giving way, triggered by sudden change of position or head-turning, but with no loss of consciousness. She had had left-sided tinnitus for years and 1 year worsening of migraines with fortification spectra, visual blurring, and vertigo.
Methods: Initially, she was diagnosed with having vestibular migraines. Her neurological examination was normal and attempted provocation of attacks with sudden changes in position and the Dix-Hallpike test were unsuccessful.
Results: Subsequent brain magnetic resonance imaging revealed a large lesion obstructing the foramina of Luschka and Magendie giving rise to obstructive hydrocephalus. Neuro-ophthalmology review showed bilateral papilloedema and no evidence of Parinaud's syndrome (no convergence retraction nystagmus or tectal pupils). Tumour biomarkers (bHCG-AFP-AlkPh-LDH) and a whole body computed tomography scan were normal. She underwent urgent cerebrospinal fluid diversion. She was given a radiological diagnosis of epidermoid cyst and is awaiting debulking.
Conclusions: This patient presented with non-specific episodes of lower limb weakness on the background of migraine with 'presumed' vestibular features. Assessing such symptoms in the context of established migraine can be challenging. Episodic headaches/vertigo with head-turning are described in Bruns syndrome, manifesting as abrupt headaches, vertigo, vomiting provoked by changes in head position due to intermittent obstructive hydrocephalous (mass lesion and ball-valve mechanism). We propose that this patient's acute episodes were similar phenomena highlighting importance of imaging in stereotypical attacks after triggers.
Disclosures: None.

P122

THE GREAT MIMIC: AN ELUSIVE DIAGNOSIS FOR A SIXTH AND SEVENTH CRANIAL NEUROPATHY

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Background and Aims: Sarcoidosis is an inflammatory multisystem disorder characterised by non-caseating epithelioid granulomas. It usually involves the lung, mediastinal lymph nodes, the skin and the eye. Ocular sarcoidosis may have several ophthalmological manifestations, most commonly uveitis, dry eye or conjunctival granulomas. Cranial neuropathy is the most frequent neurological manifestation.
Methods: A case report.
Results: A 53-year-old female presented to the ophthalmology department with right ocular pain, temporal headache and binocular horizontal diplopia. Findings included impaired right eye abduction, lagophthalmos, superficial punctate keratitis, hemifacial altered motility/sensitivity, compatible with right sixth and seventh peripheral cranial nerve palsies. Blood tests revealed increased erythrocyte sedimentation rate and negative angiotensin converting enzyme. Brain and neck vessel magnetic resonance imaging (MRI), cerebrospinal fluid (CSF) analysis and electromyography were normal. A thoracic computed tomography scan showed no mediastinal lymphadenopathy or lung involvement. Intravenous 1 g methylprednisolone for a 7 day trial led to no improvement. A whole body positron emission tomography scan revealed

extensive axillary lymph node activation. Excisional biopsy showed non-necrotising granulomatous adenitis, compatible with sarcoidosis. A diagnosis of cranial polyneuropathy secondary to sarcoidosis was made and immunosuppression was started.
Conclusions: The first recognised clinical manifestation of sarcoidosis often involves the eye. Neurological compromise is reported in 5–10% of cases. Diagnosis requires integrating MRI, CSF and histopathological findings. This case highlights the elusive nature of this disease and the necessity to thoroughly study cranial neuropathies.
Disclosures: None.

P123

BILATERAL TOXIC OPTIC NEUROPATHY INDUCED BY CHRONIC LINEZOLID TREATMENT

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Background and Aims: We report the case of bilateral toxic optic neuropathy from long-term treatment with linezolid in a patient undergoing systemic therapy for Mycobacterium asbcessus pneumonia.
Methods: A retrospective review of the case record was performed. Clinical evaluation, auxiliary testing, treatment, and follow-up data were retrieved.
Results: A 64-year-old woman presented with painless, bilateral vision loss. Her past medical history included Mycobacterium asbcessus pneumonia treated with amikacin and linezolid for the previous 7 months. Her best corrected visual acuity was 20/63 and 20/125 in the right (OD) and left (OS) eyes, respectively. Ishihara colour plates read were 3/16 OD and 2/16 OS. Funduscopy revealed bilateral temporal optic disc pallor. Automated perimetry showed bilateral blind spot enlargement, with an arcuate defect OD. Pattern visual-evoked potentials were extinguished to a 15 minutes of arc check size bilaterally. Bilateral toxic optic neuropathy induced by linezolid was suspected. Linezolid was discontinued and treatment with idebenone 500 mg daily was initiated. Three months later, her visual acuity, colour vision and visual field defects had improved significantly.
Conclusions: Toxic optic neuropathy may occur from prolonged linezolid treatment, requiring early diagnosis and prompt discontinuation of the drug. Linezolid-induced optic neuropathy is generally reversible after drug withdrawal. The benefit of the idebenone treatment requires further studies.
Disclosures: None.

P124

TB OR NOT TB – THAT IS THE QUESTION!

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Background and Aims: Chronic lymphocytic meningitis with marked hypoglycorrhachia (low cerebrospinal fluid [CSF] glucose level) is often considered synonymous with infectious meningitis. In endemic, high-burden areas of the world, tuberculosis is the first cause. In this report we highlight the diagnostic dilemmas arising in chronic lymphocytic meningeal infiltrative processes.

Methods: We review the clinical, radiological and CSF features of a patient who presented with worsening headaches, features of raised intracranial pressure and abnormal CSF, which had been considered pathognomonic of tuberculosis due to an extremely low CSF glucose level.
Results: A previously healthy young man of South Asian origin presented with an insidious onset of headaches and was found to have papilloedema. Magnetic resonance imaging of his brain and a whole body computed tomography scan were normal. The basic blood work-up was unremarkable. A lumbar puncture found a raised opening pressure of 32 cmCSF, a raised CSF protein level of 1.72 g/L, a low CSF glucose level of 1.0 mmol/L (compared with a plasma level of 5.0 mmol/L) and a CSF white cell count of 126 cells/μL (10% polymorphs and 90% lymphocytes) with a negative cryptococcal antigen test. On the basis of the suggestive CSF he was diagnosed with probable TB meningitis and anti-tubercular treatment was commenced. Worsening of symptoms with a new optic neuropathy 6 weeks later prompted a neuro-ophthalmology referral.
Conclusions: Herein we describe the journey leading to an unexpected diagnosis and, against all odds, a successful treatment outcome for this patient. We discuss the differential diagnosis with this type of CSF and a brief literature review of potential causes behind extreme CSF hypoglycorrachia in the context of lymphocytic meningitis
Disclosures: None.

P125

A CHILD WITH PAPILLOEDEMA – WHEN RESPONSIBILITY KNOCKS ON THE OPHTHALMOLOGIST'S DOOR

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Background and Aims: Papilloedema refers to optic disc oedema caused by increased intracranial pressure. Consultation requests for suspected papilloedema in children are frequent in Paediatric Ophthalmology. The distinction between papilloedema and pseudopapilloedema is crucial in the approach and prognosis of children, since failure to diagnose papilloedema can be life-threatening.
Methods: This case report describes a case of a 2-year-old child with a Chiari malformation and a history of ventriculo-cisternostomy due to hydrocephalus, who was sent to the Paediatric Ophthalmology Department due to bilateral papillary elevation. The study primarily involved retinography, ocular ultrasound (US), optical coherence tomography (OCT), and later fluorescein angiography.
Results: The results of retinography, ocular US and OCT were inconclusive. Fluorescein angiography identified some early capillary dilation and late papillary hyperfluorescence, with some papillary leakage. Therefore, a diagnosis of papilloedema was confirmed. Taking into account the results of the examinations performed, the Neurosurgery team decided to perform a ventriculo-peritoneal shunt with consequent progressive improvement in the clinical findings.
Conclusions: Studying papilloedema in clinical practice can be challenging. Paediatric age increases the difficulty in performing and interpreting the tests performed. We want to emphasise the importance of a complete study given the responsibility placed on the decision since it influences the treatment and prognosis of the patient.
Disclosures: None.

P126

THE PATTERN OF RETINAL GANGLION CELL LOSS IN WOLFRAM'S SYNDROME DIFFERS FROM OPA1 ASSOCIATED OPTIC NEUROPATHY

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Background and Aims: Wolfram's syndrome (WS) is a rare autosomal recessive neurodegenerative disease associated with WFS1 gene mutations. The mechanism and functional relevance to neurodegeneration of the mitochondrial involvement remains largely unsolved. We aim to describe the clinical phenotype of a cohort of WS patients, focusing on the pattern of optic atrophy, as compared with OPA-1 associated dominant optic atrophy (DOA).
Methods: Ophthalmological data from 25 WS patients were retrospectively retrieved and compared with 33 OPA1-related DOA patients.
Results: In our cohort, optic atrophy was present in 100% of WS patients. Visual acuity, mean deviation and retinal nerve fibre thickness loss were worse in WS patients with a faster decline from an early age compared with DOA patients, who displayed more stable visual function over the years. Conversely, ganglion cell layer (GCL) sectors were overall thinner in DOA patients from an early age compared with WS patients, in which GCL thickness started to decline later in life.
Conclusions: Our results show more severe degeneration of anterior visual pathways in WS, with fast deterioration from an early age. The retinal ganglion cells' axonal degeneration precedes by about a decade the cellular body atrophy. This differs from DOA, indirectly supporting the lack of primary mitochondrial dysfunction in WS.
Disclosures: Speaker honoraria from Santhera Pharmaceuticals, Chiesi Farmaceutici, Omikron Italia Farmaceutici, GenSight Biologics.

P127

WOLFRAM'S SYNDROME: ONLY A NEURODEGENERATIVE DISEASE OR ALSO A MACULOPATHY?

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Background: Wolfram's syndrome (WS) is a rare neurodegenerative disorder associated with multiple ophthalmic and systemic abnormalities. Optic atrophy is one of the earliest and most common ophthalmic findings of WS. Pigmentary maculopathy and diabetic retinopathy have been previously reported. We present a case report to describe a case of an adolescent patient affected by WS with subfoveal retinal changes on spectral domain optical coherence tomography (OCT).
Aims and Methods: A comprehensive ophthalmological evaluation, multimodal imaging (OCT, OCT angiography) and electrophysiological examinations were performed. Solar maculopathy and laser exposure were excluded.

Results: A 14-year-old male with WS and diabetes mellitus was evaluated. On OCT scans centred on the fovea, the ellipsoidal zone and Brüch's membrane – retinal pigmentated epithelium (RPE) complex appeared bilaterally interrupted. A backscattering effect under the interrupted area was reported. In contrast, the outer and inner retinal layers were preserved. Retinal thickness and foveal depression were within normal limits. No pigmentary or diabetic alterations were observed.
Conclusions: This case describes a macular alteration not previously reported, advancing the hypothesis that WS may affect not only the optic nerve, but also the outer retina. Further studies are needed to clarify this association.
Disclosures: None.

P128

PROGRESSIVE MULTIFOCAL LEUKOENCEPHALOPATHY: RAISING AWARENESS

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Background and Aims: Progressive multifocal leukoencephalopathy (PML) is a devastating demyelinating disease caused by JC virus reactivation in the setting of chronic immunosuppression due to different predisposing conditions. Neuro-ophthalmological symptoms are present in nearly 50% of cases emphasising the role of the neuro-ophthalmologist in timely diagnosis and potentially reducing neurological morbidity and mortality.
Methods: Retrospective case reports
Results: We report two cases of virologically confirmed PML with different clinical presentations and temporal courses. Case 1: Chronically progressive cortical visual loss occurred resulting in cortical blindness in an otherwise neurologically intact 75-year-old female with relapsing chronic lymphocytic leukaemia during treatment with rituximab and ibrutinib. Treatment was withheld with subsequent clinical stabilisation. Case 2: Rapidly progressive cognitive impairment with homonymous hemianopia, visual associative deficits, and general neurological decline occurred in a 67-year-old male during the treatment of a second relapse of mantle cell lymphoma with venetoclax, lenalidomide, and rituximab. Treatment was discontinued, but the patient deteriorated and passed away 4 months after the initial neurological symptoms.
Conclusions: Immunosuppressed patients with PML and underlying malignancy pose considerable diagnostic difficulty and a therapeutic dilemma. Early diagnosis of PML through heightened clinical awareness is the first crucial step in reducing disability and in prolonging survival.
Disclosures: None.

P129

A CASE REPORT OF LYME OPTIC NEURITIS AS THE ONLY MANIFESTATION OF NEUROBORRELIOSIS

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Background and Aims: Lyme disease is a multisystem zoonotic disease caused by the spirochaete Borrelia burgdorferi. The primary mechanism of transmission is through tick bites. Optic neuritis, described as inflammation of the optic nerve resulting in blurred vision and painful eye movements, is a rare complication of neuroborreliosis.
Methods: A case report.
Results: A 43-year-old female with a history of a tick bite presented to the UMC Ljubljana, due to impaired visual acuity in her right eye and painful eye movements. Ophthalmological examination revealed a relative afferent pupillary defect and optic disc oedema in her right eye. Magnetic resonance imaging showed right optic nerve swelling with contrast enhancement. The blood work-up showed positive titres of IgM and IgG Lyme-specific antibodies. A spinal tap confirmed intrathecal synthesis of IgG Lyme-specific antibodies. The patient received intravenous therapy with ceftriaxone and methylprednisolone and showed daily improvement. The optic disc oedema resolved, however partial atrophy of the right optic nerve was observed.
Conclusions: Isolated optic neuritis is a possible presentation of neuroborreliosis and it is important to include it in the differential diagnosis of optic neuritis in endemic areas. Neuroborreliosis can be confirmed by a spinal tap showing intrathecal synthesis of specific antibodies. Prognosis after specific antibiotic therapy is good.
Disclosures: None.

P130

TAKING CALCIUM EACH DAY KEEPS THE NEURO-OPHTHALMOLOGIST AWAY

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Background and Aims: A 28-year-old female was referred to the neuro-ophthalmology clinic with bilateral swollen optic nerve heads for 4 months and sudden onset of reduced vision in her left eye left vision. She had previously been diagnosed with autosomal dominant hypocalcaemia due to a calcium sensory receptor mutation. Her adherence to treatment was poor.
Methods: She had no neurological symptoms. She had experienced painless loss of her inferior visual field in her left eye on waking. She was normotensive and she weighed 60 kg (body mass index 22 kg/m^2). There were no vascular risk factors. She did not take any medications and denied illicit drug use.
Results: Her baseline investigations included the following: serum calcium 1.56 mmol/L, phosphate 1.31 mmol/L, parathyroid hormone 4.3 pmol/L, vitamin D 99 nmol/L; magnetic resonance imaging of head and spine with contrast were normal; and a lumbar puncture opening pressure of 16 cm cerebrospinal fluid (CSF) with normal CSF constituents. Treatment, directed by her endocrinologist, included calcitriol 500 ng daily and calcium carbonate 1.5 g/colecalciferol 10 μg (two tablets twice daily). Although her left visual field defect persisted, her right optic nerve function was preserved and her optic nerve head swelling started to improve 2 months later as her calcium levels increased. The swelling resolved following normalisation of her calcium levels 4 months later.
Conclusions: Rarely, hypocalcaemia may cause papilloedema due to secondary intracranial hypertension or an optic neuropathy secondary to disruption to axoplasmic transport resulting in a secondary ischaemic optic neuropathy as in this case. Treatment of hypocalcaemia may prevent progressive visual loss.
Disclosures: None.

P131

ICE DIAGNOSIS – WHEN IDEAS RUN OUT: A CASE REPORT

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Background Aims: Myasthenia gravis is an autoimmune disease that affects the neuromuscular junction, resulting in muscle weakness and fatigability. Ocular manifestations are often the presenting sign and, when isolated, define ocular myasthenia gravis (OMG). OMG diagnosis is often challenging.
Methods: We report a case of challenging diagnosis of OMG.
Results: A 75-year-old female was evaluated for asymmetrical bilateral ptosis with right eye predominance, associated with vertical diplopia for 1 month. She also referred bilateral severe ocular pain and progressive vision loss. Ophthalmological examination revealed visual acuities of .3 in her right eye and .1 in her left eye. On biomicroscopy, exuberant punctiform keratitis and cataracts in both eyes were observed. Fundus examination was normal. Macular and optic disc optical coherence tomography was unremarkable. Computed tomography (CT) scans of her head and orbit and CT angiography were normal, as was temporal artery ultrasonography. A therapeutic trial of oral corticosteroid was performed, without clinical response. An ice pack test reinforced the diagnostic suspicion of OMG. Pyridostigmine treatment led to clinic normalisation.
Conclusions: OMG is a challenging and rare diagnosis. Its presentation can simulate any extraocular muscle paresis. Perception of decreased visual acuity with pain from exposure keratitis are confounding symptoms that, in our case, led to diagnostic difficulties.
Disclosures: None.

NEURO-IMAGING

P132

PERILS OF OMITTING CONTRAST AGENTS FROM NEUROIMAGING: YET ANOTHER CASE!

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Background and Aims: To improve the diagnostic yield of intraorbital or intracranial tumours, magnetic resonance imaging (MRI) should be performed with gadolinium contrast. More importantly, correlating presenting clinical features with MRI features is vital in making the correct diagnosis.
Methods: We describe a case of a 75-year-old male with well-controlled type 2 diabetes mellitus, who presented with left complete pupil-sparing cranial nerve III palsy and a left optic neuropathy. An initial plain MRI of his brain was reported as normal but a subsequent MRI of his brain with contrast demonstrated a well-circumscribed left sphenoid wing tumour.

Results: His visual acuity was 6/6 in the right eye and hand movement perception in the left eye. He had a left relative afferent pupillary defect. There was a left complete ptosis, and near-complete ophthalmoplegia on the left except for abduction. Fundal examination revealed a moderately pale optic disc on the left. Repeat MRI with gadolinium contrast showed a rapidly progressive left orbital mass with a meningeal enhancement that had tripled in size over a 3 month period. On systemic screening he was found to have raised prostatic specific antigen level of 194 ng/mL. The biopsy results of the left orbital mass and the clinical outcome will be presented.
Conclusions: This case highlights the importance of engaging with radiology to ensure that MRIs ordered with contrast include it. The rapid increase in the size of the tumour over a 3 month period and the high PSA level makes a diagnosis of a metastatic orbital tumour highly likely.
Disclosures: None.

P133

PAEDIATRIC CIRCUMPAPILLARY RETINAL NERVE FIBRE LAYER DEVELOPMENT IN HEALTHY INFANTS AND CHILDREN USING HAND-HELD SPECTRAL-DOMAIN OPTICAL COHERENCE TOMOGRAPHY

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Background and Aims: We aimed to investigate the development of 3-dimensional full circumpapillary retinal nerve fibre layer (cpRNFL) in full-term infants and young children without sedation and dilation using handheld optical coherence tomography (HH-SDOCT) and also to establish normative age-adjusted values.
Aims and Methods: OCT images were collected from 231 children aged between 1 day and 14.5 years using HH-SDOCT and were then analysed semi-automatically using the GDx Nerve Fibre Analyser protocol. The developmental trajectories against log post-menstrual age for cpRNFL thickness in four quadrants were modelled using linear mixed models or fractional polynomials whenever appropriate.
Results: The thickness of the superior, inferior and nasal cpRNFL quadrants did not significantly change between birth and 14.5 years old. The 1st quartile (Q1), median (Q2) and 3rd quartile (Q3) were 100 μm, 110 μm and 120 μm, respectively for the superior cpRNFL quadrant; 99 μm, 110 μm and 120 μm, respectively for the inferior cpRNFL quadrant; and 60 μm, 68 μm and 75 μm, respectively for the nasal cpRNFL quadrant. In contrast, the temporal cpRNFL quadrant demonstrated a significant decrease between birth and approximately 18 months of age and then remained relatively constant up to 14.5 years (Q1 = 57 μm; Q2 = 62 μm; and Q3 = 70 μm > 18 months of age).
Conclusions: We describe the development of the full cpRNFL from birth to 14.5 years using 3-dimensional HH-SDOCT imaging. Interestingly, the temporal cpRNFL quadrant shows a different developmental trajectory to the superior and inferior cpRNFL quadrants despite all three quadrants comprising of fibres projecting from the macular region.
Disclosures: None.

P134

MAGNETIC RESONANCE IMAGING OF INFERIOR OBLIQUE MYECTOMY

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Background and Aims: Inferior oblique myectomy is the most commonly performed operation for the treatment of superior oblique palsy. After a segment of the muscle is excised, the proximal stump is cauterised and then allowed to retract into the medial orbit. What happens to this residual muscle tissue is uncertain.
Methods: A 35-year-old man presented with intermittent diplopia and findings consistent with a right superior oblique muscle paresis. He reported having undergone surgery at age 5 for strabismus, but could provide no further information. Orbital magnetic resonance (MR) imaging was obtained to evaluate the extraocular eye muscles.
Results: MR imaging showed a complete surgical detachment of the right inferior oblique, with the muscle remnant terminating 4 mm temporal to the lateral edge of the inferior rectus. The stump contacted posterior Tenon's capsule but did not insert into the globe.
Conclusions: MR imaging can be a useful tool to ascertain the status of the inferior oblique muscle when additional surgery is contemplated to address persistent head tilt and diplopia in patients with superior oblique palsy.
Disclosures: None.

OCULAR-IMAGING

P135

OPTICAL COHERENCE TOMOGRAPHY IMAGING OF A CASE OF PAPILLOEDEMA IN AN EYE WITH AN OPTIC DISC COLOBOMA

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Background and Aims: Optic disc coloboma (ODC) is a rare congenital defect that arises due to incomplete proximal optic fissure closure. We report the optical coherence tomography (OCT) findings when papilloedema occurs in an eye with an ODC.
Methods: Retrospective, descriptive case report.
Results: A 24-year-old woman with a history of a right ODC, presented with visual obscurations and headache. She was on lymecycline for acne vulgaris and reported recent weight gain. She had bilateral papilloedema, however the colobomatous area of the right inferior optic disc failed to show any swelling clinically or on OCT. Kinetic perimetry showed a coloboma-related superior visual field defect. A macular OCT showed nasal parafoveal schitic change. Computed tomography (CT) of her head and CT venography were normal. Her lumbar puncture opening pressure was 34 cm cerebrospinal fluid. She was diagnosed with pseudotumour cerebri secondary to the lymecycline and weight gain. She was advised to stop lymecycline and was started on acetazolamide. After 3 months, the colobomatous segment and schitic area remained unchanged despite resolution of the optic disc swelling, including the superior portion of the right optic disc.
Conclusions: The lack of swelling in the colobomatous part of the optic disc, i.e. an area without optic nerve fibres, is consistent with papilloedema being due to optic nerve fibre swelling secondary to axoplasmic stasis.
Disclosures: None.

P136

OSCAR-MS – A PROPOSAL FOR QUALITY ASSESSMENT OF RETINAL OPTICAL COHERENCE TOMOGRAPHY ANGIOGRAPHY

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Background and Aims: Optical coherence tomography angiography (OCT-A) allows high-resolution and non-invasive assessment of retinal vessels. During the last years, OCT-A has been increasingly used to study alterations of the retinal vasculature during different neuro-ophthalmological and neurological diseases. To date, standardised and widely-accepted criteria for quality control (QC) of OCT-A have been lacking. We aim to propose criteria for OCT-A quality assessment.
Methods: The following steps were undertaken: (1) Detailed literature review on OCT-A artefacts and quality; (2) Proposition of OCT-A QC criteria; (3) Application of criteria to evaluate a set of OCT-A scans; (4) Kappa-statistics for inter-rater agreement; and (5) Identification of reasons for inter-rater disagreement, revision of OCT-A QC criteria.
Results: We provide preliminary data from an ongoing study. After an extensive literature review, we identified seven major aspects that affect OCT-A quality: (O) obvious problems; (S) signal strength; (C) centration; (A) artefacts due to projection; (R) retinal pathology; (M) motion artefacts; and (S) segmentation error. The OSCAR-MS criteria were applied by four raters from two centres to evaluate a set of 40 OCT-A scans from patients with multiple sclerosis, Sjögren's syndrome, uveitis and healthy individuals.
Conclusions: We propose validated criteria (OSCAR-MS) for quality assessment of OCT-A, which might be considered in the context of clinical trials and studies.
Disclosures: Rebecca Wicklein, Christina Noll and Charmaine Yam declare no financial disclosures. Lilian Aly received a poster award sponsored by Novartis. Bernhard Hemmer has served on scientific advisory boards for Novartis; he has served as DMSC member for AllergyCare, Polpharma, Sandoz and TG therapeutics; he or his institution have received speaker honoraria from Desitin; his institution received research grants from Regeneron for multiple sclerosis research. He holds part of two patents; one for the detection of antibodies against KIR4.1 in a subpopulation of patients with multiple sclerosis and one for genetic determinants of neutralising antibodies to interferon. All conflicts are not relevant to the topic of the study. Ahmed Toosy has received speaker honoraria from Biomedia, Sereno Symposia International Foundation, Bayer and meeting expenses from Biogen Idec and Novartis. He was the UK PI for two clinical trials sponsored by MEDDAY pharmaceutical company (MD1003 in optic neuropathy [MS-ON – NCT02220244] and progressive MS [MS-SPI2 – NCT02220244]). Axel Petzold reports personal fees from Novartis, Heidelberg Engineering, Zeiss and grants from Novartis, outside the submitted work. He is part of the steering committee of the OCTiMS study which is sponsored by Novartis and is part of the steering committee of Angio-OCT which is sponsored by Zeiss. Benjamin Knier received a research award from Novartis (Oppenheim award 2020) and funding for preclinical studies that are not relevant to the topic of the current study.

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CLINICAL SIGNIFICANCE OF STRUCTURAL CHANGES IN THE PERIPAPILLARY RETINAL NERVE FIBRE LAYER AND OPTIC NERVE HEAD IN CHILDHOOD GLAUCOMA IMAGED USING HAND-HELD OPTICAL COHERENCE TOMOGRAPHY

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Background and Aims: To document three-dimensional changes in the peripapillary retinal nerve fibre layer (ppRNFL) thickness and optic nerve head parameters in paediatric glaucoma, using hand-held optical coherence tomography (HH-OCT) and to investigate how well these changes predict visual acuity.
Methods: All participants were imaged using HH-OCT and visual acuity was also assessed. Automated segmentation software measured ppRNFL thicknesses in four quadrants (at a 6° radius) as well as optic nerve head parameters. Linear mixed models were used to compare OCT parameters between groups and their relationship with visual acuity.
Results: From 103 paediatric glaucoma patients, we successfully imaged 31 children with primary glaucoma and 46 with secondary glaucoma (mean age: 6.66 ± 4.3 years). Seventy-seven age-matched controls were imaged for comparison. In glaucoma, ppRNFL thickness was significantly thinner for all quadrants compared with controls, with the largest changes occurring in the inferior (27–33%) and superior (29–35%) quadrants. The cup area was 102% and 158% larger and rim area 36% and 39% smaller, in primary and secondary glaucoma, respectively. Visual acuity correlated most strongly with inferior and superior ppRNFL quadrants, and cup depth, cup-to-disc ratio, cup area and volume, rim area and volume.
Conclusions: We demonstrate the feasibility of using hand-held OCT in a paediatric glaucoma clinic to document important indicators of glaucoma severity that are associated with reduced visual acuity.
Disclosures: Frank A. Proudlock has a commercial agreement with Leica Microsystems

P138

RETINAL NERVE FIBRE LAYER THICKNESS AND GANGLION CELL COMPLEX THICKNESS AMONG PATIENTS WITH MULTIPLE SCLEROSIS WITH AND WITHOUT A HISTORY OF OPTIC NEURITIS

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Background and Aims: Optic neuritis (ON) is the most common ocular manifestation of multiple sclerosis (MS), resulting in optic nerve atrophy and thinning of the peripapillary retinal nerve fibre layer (RNFL) observed optical coherence tomography (OCT). Nevertheless, a progressive loss of ganglion cells is also know to occur in these patients over time, which can also be observed on

OCT analysis. We aimed to evaluate both RNFL and the ganglion cell complex (GCC) thickness of eyes of MS patients with a previous history of ON and compare those with MS patients with no previous history of ON.
Methods: Data of all MS patients observed in our department since 2018 were collected (n = 55). Best-corrected visual acuity (BCVA), RNFL and GCC values were compared between patients who had had ON (ON group) against those who had no history of ON (non-ON group).
Results: Mean RNFL thickness was 79.3 ± 19.3 μm in ON group and 88.9 ± 9.9 μm in the non-ON group ($p < .001$). Mean GCC thickness was 65.8 ± 11.7 μm in the ON group and 75.8 ± 8.1 μm in the non-ON group ($p < .001$). When comparing eyes in the non-ON group with unaffected eyes from the ON group, GCC thickness was statistically lower in the latter group ($p < .048$).
Conclusions: ON history represents a risk factor for greater decrease in mean BCVA in MS patients, as well as for RNFL and GCC loss. However, a greater reduction of GCC thickness was also observed among eyes with a history of contralateral optic neuritis. Chiasmal involvement or possible subclinical episodes of ON may explain this difference.
Disclosures: None

OCULAR MOTILITY DISORDERS AND NYSTAGMUS

P139

ACQUIRED UNILATERAL PUPIL SPARING OCULOMOTOR AND BILATERAL ABDUCENS NERVE PARESIS IN NASOPHARYNGEAL CARCINOMA: A CASE REPORT

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Background and Aims: Acquired palsies of cranial nerves III, IV or VI are commonly initially referred to the ophthalmology emergency department and are often perceived as a sign of serious underlying pathology. Nasopharyngeal carcinoma is a malignancy that can cause paresis of cranial nerves III, IV, and VI.
Methods: This case report was aimed to describe III and VI cranial nerve paresis as the only initial manifestation of nasopharyngeal carcinoma and emphasise the need for a high index of suspicion.
Results: A 70-year-old woman could not glance to the right for 2 months. Two weeks later, a drooping right eye lid appeared. Right ocular motility testing showed motion reduction in the superotemporal, temporal, inferotemporal, inferior, nasal, and superonasal directions while left ocular motility testing showed reduction in the inferotemporal, temporal, and superotemporal directions with no pain on eye movement. She had right upper eyelid ptosis. Brain contrast-enhanced magnetic resonance imaging showed a nasopharyngeal mass extending to parapharyngeal space, sphenoid sinus, and medial fossa that compressed the right side of the optic chiasm, right internal carotid artery and right VI cranial nerve. She was referred to an ear nose and throat surgeon.
Conclusions: History and complete ocular evaluation along with appropriate an imaging examination is needed to find the aetiology of III and VI cranial nerve palsies. Management outcomes depend on the treatment of the underlying tumour.
Disclosures: None.

P140

AN UNUSUAL CASE OF IDIOPATHIC CAVERNOUS SINUS SYNDROME: A CASE REPORT

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Background and Aims: The cavernous sinus is a valveless dural-lined venous plexus located on either side of the sella turcica at the central skull base. Cavernous sinus syndrome is characterised by ophthalmoplegia and sensory deficits over the head due to combined deficits of the three contained cranial nerves (third, fourth, and sixth) that are involved with eye movements and the ophthalmic branch of the trigeminal nerve.
Methods: This report present a case of cavernous sinus syndrome in the right eye.
Results: A 33-year-old man had ptosis of the right eye followed by slow decrease in vision in that eye for 1 month. Upon examination, the best corrected visual acuity was 6/20. Pronounced limitation of ocular motility was present in his right eye. His pupil did not constrict normally and funduscopy revealed papilloedema in both eyes. Moreover, TORCH IgG antibodies (except type 2 herpes simplex virus IgG antibody) were elevated. Brain magnetic resonance (MR) imaging, MR angiography, and MR venography revealed an abscess in cerebellum of the right hemisphere, an inflammatory reaction in the right cavernous sinus and an arachnoid cyst in the right frontal lobe, providing suspicion of an immunocompromised aetiology
Conclusions: A case of cavernous sinus syndrome presenting as ptosis with poor vision is reported. Cavernous sinus syndrome should be included in the differential diagnosis of patients with ptosis.
Disclosures: None.

P141

A RARE CASE OF AN ISOLATED 3RD CRANIAL NERVE PALSY IN CAVERNOUS SINUS SYNDROME

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Background and Aims: A 77-year-old white Caucasian female presented to the emergency department with a 10 day history of a moderate occipital headache with binocular vertical double vision and right ptosis. She had a background of rheumatoid arthritis on tocilizumab, hypertension, and epilepsy. Her bedside observations were normal, she had significantly elevated erythrocyte sedimentation rate (60 mm/hour) and C-reactive protein level (87 mg/L).
Methods: A plain computed tomography (CT) scan if her head was reported to show intracranial pathology apart from a known longstanding left sphenoidal sinusitis. An Ophthalmology opinion was sought and a right isolated partial non-pupil involving 3rd nerve palsy was diagnosed with no other ocular abnormalities. A subsequent CT angiogram revealed no aneurysm. Rheumatology opinion was that of a low suspicion for temporal arteritis.
Results: She remained afebrile throughout her inpatient stay. Neurology agreed upon the finding of an isolated cranial nerve palsy. CT venography was requested and was initially reported as normal. However, the case was discussed at a skull base multi-disciplinary team meeting and here a neuroradiologist determined that the right superior ophthalmic vein was engorged suspected cavernous sinus thrombus. The patient was treated with antibiotics and low-molecular weight heparin.

Orthoptic review 2 weeks from presentation revealed full ocular motility, no ptosis, and an improved headache. Magnetic resonance venography 3 weeks from presentation revealed an improved superior ophthalmic vein filling defect and patent dural sinuses.
Conclusions: The possibility of a co-incidental ischaemic partial 3rd nerve palsy cannot be ruled out. The purpose of this case report is to emphasise the importance of specific neuro-radiology input and suspecting cavernous sinus pathology in an atypical presentation of an isolated cranial nerve palsy.
Disclosures: None.

P142

A PAINFUL CONUNDRUM

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Background and Aims: Recurrent painful ophthalmoplegic neuropathy (RPON) has an incidence of .7 per million and usually presents in children. The most commonly affected nerve is the oculomotor nerve. Fewer than 100 oculomotor nerve schwannomas (ONS) have been reported in the literature. A conservative management approach is advocated for small lesions. Larger lesions may require surgical resection.
Methods: We describe a case of RPON complicated by the presence of a third cranial nerve schwannoma noted on neuroimaging.
Results: A 23-year-old female presented with a 3 week history of a throbbing, worsening frontal headache, associated with new onset progressive diplopia and right-sided ptosis. She had a past medical history of migraine diagnosed at the age of 8-years-old with two previous episodes of RPON involving the third cranial nerve. On examination, she had a right partial third cranial nerve palsy with pupillary mydriasis and no other neurological defects. Magnetic resonance imaging of the head showed enlargement of a previously noted small nodular enhancing lesion in the proximal cisternal segment of the right oculomotor nerve, most likely representing a schwannoma.
Conclusions: ONS is a differential diagnosis for RPON. In this case, neuroimaging will need to be repeated to assess whether the ONS is causing the third cranial nerve palsy or whether it is secondary to RPON.
Disclosures: None.

P143

SEE-SAW NYSTAGMUS IN TRAUMATIC CHIASMAL DAMAGE TREATED SUCCESSFULLY WITH CLONAZEPAM

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Background and Aims: A traumatic chiasmal lesion is a rare cause of see-saw nystagmus.

Methods: A 31-year-old male, known to have chiasmal damage with bitemporal visual field loss from an accident 14 years earlier, complained of increasingly unstable vision. On examination his visual acuity was 1.0 and .6. His bitemporal hemianopia was stable. Assessment of his ocular motility showed see-saw nystagmus.
Results: On recording of his eye movements with an infrared eye tracking system the see-saw nystagmus was confirmed; the frequency was around 5–6 Hz. As the patient was limited in his daily activity by the nystagmus, he was treated with clonazepam. The starting dose of the clonazepam was .5 mg twice daily, later increased to three times daily. The amplitude of the nystagmus was halved on re-testing. The patient did not have major side-effects due to the medication and he could resume his daily activities. Recordings of the nystagmus pre- and post-treatment will be shown during the presentation.
Conclusions: See-saw nystagmus after traumatic chiasmal damage can be treated successfully with clonazepam.
Disclosures: None.

P144

DETECTING INTERNUCLEAR OPHTHALMOPLEGIA IN MULTIPLE SCLEROSIS: PREVALENCE IN A POPULATION-BASED COHORT

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Background and Aims: Internuclear ophthalmoplegia (INO) is a common eye movement disorder in multiple sclerosis (MS). INO can be detected more reliably by infrared oculography than by physical examination. Methods for diagnosing INO with infrared oculography in patients with MS and the association between INO and MS characteristics still need confirmation.
Methods: We investigated INO prevalence in a cross-sectional population-based cohort of MS patients with the same birth year (Project Y). Thresholds for the versional dysconjugacy index (VDI), assessed by infrared oculography, were used to detect INO. Clinical characteristics, visual functioning and visual complaints were compared between MS patients with and without INO.
Results: Two-hundred-twenty MS patients and 110 healthy controls were included. INO was detected in 53 (24%) of the MS patients. Nineteen (15%) healthy controls showed VDI values surpassing the threshold for INO. INO was associated with male sex (43% versus 22%, p = .002), greater disability (expanded disability status scale score 4.0 versus 3.5, p = .044), worse cognition (symbol digit modalities test score 49 versus 53, p = .046) and worse arm function (nine hole peg test time 22.20 versus 21.41 sec; p = .015) in MS patients. INO was not associated with disease duration, visual function or visual complaints.
Conclusions: INO is prevalent among MS patients and related to the clinical characteristics of MS. This highlights the relevance of assessing INO in MS using quantitative oculography. Validation of INO detection with oculography across MS patients with varying demographic characteristics is needed.
Disclosures: S.N. Hof, F.C. Loonstra, L.R.J. de Ruiter and L.J. van Rijn declare no conflict of interest. A. Petzold reports personal fees from Novartis, Heidelberg Engineering, Zeiss, grants from Novartis, outside the submitted work; and AP is part of the steering committee of the OCTiMS study which is sponsored by Novartis. AP is part of the steering committee of Angio-OCT which is sponsored by Zeiss. He

does not receive honorary as part of these activities. The NIHR BRC at Moorfields Eye Hospital supported AP. B.M.J. Uitdehaag has received consultancy fees from Biogen Idec, Genzyme, Merck Serono, Novartis, Roche and Teva. J.A. Nij Bijvank is supported by the Dutch MS Research Foundation, grant nr. 18–1027.

P145

A RARE CASE OF ISOLATED BILATERAL SUPERIOR GAZE PALSY WHO DEVELOPED FINDINGS CONSISTENT WITH DORSAL MIDBRAIN SYNDROME IN AN ELDERLY WOMAN WITH UNILATERAL THALAMIC STROKE WITHOUT MIDBRAIN INVOLVEMENT

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Background and Aims: Thalamic lesions are associated with possible neuro-ophthalmological deficits that are important to distinguish from those caused by lesions in the midbrain. Vertical gaze palsy, although a known manifestation of thalamic stroke, rarely presents as an isolated superior gaze palsy. The objective of this case report is to highlight the unusual occurrence of an isolated bilateral superior gaze palsy with elements of dorsal midbrain syndrome in a patient with a thalamic haemorrhagic stroke and no clear midbrain extension.
Methods: An elderly female presented in 2019 after a right sided thalamic stroke with left sided hemiparesis, normal pupil reactions and −5 limitation of superior gaze symmetrically.
Results: Her pupillary reactions on subsequent visits were sluggish with −4 limitation of superior gaze. Attempted vertical saccades produced some convergence without nystagmus. On subsequent scans the thalamic bleed completely resolved with a focal area of encephalomalacia without midbrain involvement. In 2021, her pupils were symmetrically large and sluggish with light-near dissociation, lid retraction and −4 limitation of elevation bilaterally. Attempted vertical saccades showed some convergence. Vertical optokinetic nystagmus responses on optokinetic drum produced convergence retraction nystagmus.
Conclusions: Vertical gaze palsies have been associated with midbrain lesions but rarely with isolated thalamic lesions. This report further supports the limited literature on thalamic lesions manifesting as isolated superior gaze palsies and highlights the importance of clinical examination with the optokinetic drum alongside brain imaging for diagnosis of this unusual presentation.
Disclosures: None.

P146

PATTERNS OF RESTRICTION: COGNITIVE BIASES AT PLAY IN THE OCULAR MOTILITY CLINIC

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Background and Aims: Up to 10% of myasthenia gravis patients (including ocular myasthenia) have no detectable antibodies to acetylcholine receptor or muscle-specific tyrosine kinase (MuSK). Staff in our specialist ocular motility clinic are specifically trained to consider myasthenia as a diagnosis, even if seronegative, enabling prompt identification and management. However, this can lead to over diagnosis.

Methods: We present two cases of patients initially diagnosed with presumed seronegative ocular myasthenia, who were shown, on revisiting the presenting history and clinical findings, to have different conditions causing their symptoms.
Results: Two females (A: 41-years-old; B: 69-years-old) presenting with diplopia were each diagnosed with seronegative myasthenia after initial subjective improvement with pyridostigmine treatment, but this was not sustained. Patient records and histories were revisited at subsequent reviews. Patient A had left high myopia (−19.0 D) with lateral rectus slippage on magnetic resonance imaging, and no variability of motility deficit between visits. She was re-diagnosed with heavy eye syndrome. Patient B described fleeting episodes of horizontal diplopia with a pulling sensation, but no ptosis or variability. A review of medical history revealed previous radiotherapy for a pituitary adenoma. She was re-diagnosed with ocular neuromyotonia.
Conclusions: In these cases, cognitive biases (anchoring, premature closure, and diagnosis momentum) led to patients continuing on inappropriate treatment and a delay in the correct diagnosis and management. Awareness of cognitive biases, engagement with metacognition techniques, and 'slowing down' can help reduce the effects of such biases on patient care.
Disclosures: None.

P147

SUPERIOR OBLIQUE MYOKYMIA: A VIDEO-DOCUMENTED CASE REPORT

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Background and Aims: Superior oblique myokymia causes intermittent episodes of monocular oscillopsia and cyclorotation due to paroxysmal contraction of the superior oblique muscle. It is a rare condition with characteristic symptoms and, if evoked in the office, the clinical appearance is distinctive and assists the diagnosis.
Methods: A 59-year-old woman, with a history of surgery for a cervical meningioma 8 years before, complained of daily episodes of oscillopsia lasting 5–10 seconds in her right eye, for the last 10 years.
Results: Although strongly suspected, superior oblique myokymia was not observed or elicited in the office for several years, and prior trials of topical beta-blockers or oral carbamazepine were unsuccessful. In an appointed consultation on one particularly stressful day for the patient, a high frequency, cyclotorsion movement of the right eye was observed and recorded. Superior oblique myokymia of the right eye diagnosis was confirmed. Th rest of the neuro-ophthalmological assessment including cerebral and cervical magnetic resonance imaging showed no other pathological findings. Carteolol eye drops for 3 months produced no response but the assessment of her condition helped in lowering her stress level and she eventually reported spontaneous improvement.
Conclusions: This video recording illustrates the characteristic clinical appearance of superior oblique myokymia. The distinctive eye movements can sometimes be evoked by asking the patient to look down whilst in adduction. We hope this video may help diagnose other patients with the same condition.
Disclosures: None.

P148

SUPERIOR OBLIQUE MYOKYMIA – RESULTS OF AN ONLINE SURVEY OF MEMBERS IN A FACEBOOK SUPPORT GROUP

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Background and Aims: Superior oblique myokymia (SOM) is a rare, monocular movement disorder involving contractions of the superior oblique muscle causing monocular oscillopsia and diplopia. Several medications have been reported to benefit some patients, but the efficacy of medical treatment has not been well established and little long-term follow-up data are available.
Methods: We created an online survey and invited members of an online SOM support group on Facebook to participate. Topics included subjective symptoms and disease course, therapy attempts and outcomes, and other aspects of SOM. The survey was performed online using limesurvey between June and December 2020.
Results: This cohort comprises 44 survey participants (age 46.5 ± 13.5 years, 77.3% female). Affected eyes were right, left, and both in 70.5%, 25%, and 4.5% of cases. Subjective symptoms were oscillations (86.4%), double vision (72.4%), and blurred vision (22.7%). The symptoms were permanent in 47.7% and episodic in 52.3%. Vascular trochlear nerve compression was found/excluded in 9.1% and 72.7% cases, respectively. Treatments tied included systemic beta blockers in 63.6%, anticonvulsants in 27.3% and topical betablockers in 97.7%, with little effect. Only 9.1% reported improvement and one patient reported remission. Neurosurgery (9%) was not effective and strabismus surgery (6.8%) improved symptoms in one case (2.3%).
Conclusions: Trochlear nerve compression was detected only in a small proportion of cases. Neurosurgery, strabismus surgery, systemic and topical pharmacological treatment were largely unsuccessful. Research on the pathophysiology of SOM needs to be continued to achieve more effective treatments in affected individuals.
Disclosures: None.

P149

THE GENOTYPIC AND PHENOTYPIC SPECTRUM OF FOVEAL HYPOPLASIA ASSOCIATED WITH NYSTAGMUS: A MULTI-CENTRE STUDY

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Background and Aims: Varying degrees of foveal hypoplasia (FH) represent different stages of arrested foveal development. The Leicester FH Grading System has been applied to albinism, and FRMD7, PAX6, SLC38A8 and AHR variants. We aimed to perform a comparative multi-centre study characterising the genotypic and phenotypic spectrum of FH in the aforementioned aetiologies.
Methods: Patients with known genetic associations of FH and nystagmus (n = 575) were identified from 10 centres from eight countries (75.6%), or extracted from publicly available datasets (24.4%). Genetic diagnosis was achieved using targeted panel-based sequencing or exome sequencing. Optical coherence tomography of the fovea was obtained in all subjects.
Results: The most common genetic aetiology for typical FH in this cohort was albinism (66.1%), followed by PAX6 (22.8%) and SLC38A8 variants (7.1%). All grades of FH were seen in albinism and PAX6 variants. All SLC38A8 cases demonstrated grade 3 or 4 FH. In AHR variants, only grade 3 FH was reported. In cases of FH and FRMD7 variants only grade 1 FH was observed.
Conclusions: Our data suggest arrested retinal development occurs earlier in SLC38A8 and AHR variants and later in FRMD7 variants. The defined time-period of foveal developmental arrest for albinism and PAX6 variants appears to demonstrate more variability. Our findings provide mechanistic insight into these disorders and have significant prognostic and diagnostic value.
Disclosures: HJK and MGT are consultants for Leica Microsystems.

P150

POSITIONAL MODULATION OF SIXTH NERVE PALSY IN PATIENTS WITH INTRACRANIAL HYPOTENSION SYNDROME

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Background and Aims: Sixth nerve palsy can occur with spontaneous intracranial hypotension (SIH). While headache and related symptoms (e.g., hearing loss) classically worsen when sitting-up, positional modulation of SIH-related sixth nerve palsy has not been thoroughly clarified. We aimed to investigate if acquired sixth nerve palsy in SIH is influenced by positional changes.
Methods: Alternate cover testing (ACT) in mid and lateral gaze (> 20 trials in each position) using a 1.5 m distance target was digitally recorded with video-oculography in the upright and supine positions. ACT eye movement tracings were manually measured using JRuler. Wilcoxon signed-rank test was used to compare measurements between positions.
Results: Two males, 29 and 50-years-old, presented with orthostatic headache and constant binocular horizontal diplopia, worse in left gaze, supposedly not affected by supine or upright position. On examination, in the upright position, there was a left sixth nerve palsy in both patients. Brain magnetic resonance imaging was remarkable for homogeneous pachymeningeal enhancement and no signs of spinal dural fistula in both, while lumbar puncture showed a low (< 8 cm cerebrospinal fluid [CSF]) opening pressure in both. After symptomatic treatment, one case resolved, while the other needed a blood patch for complete resolution. In both, ATC misalignment in the supine versus upright position was significantly greater (patient A, 12 versus 18 prism dioptres, and patient B, 17 versus 31 prism dioptres; $p < .001$).

Conclusions: We provide quantitative evidence for positional modulation of sixth nerve palsy in SIH. Upright position might lead to additional CSF leakage and/or cerebral vasodilation, possibly promoting further sixth nerve traction. Relying on patient's subjective notion of diplopia does not seem to be helpful to detect such modulation.
Disclosures: None.

P151

ALSTRÖM SYNDROME MIMICKING SPASMUS NUTANS: A CASE REPORT

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Background and Aims: Alström syndrome (ALMS) is a rare monogenic ciliopathy caused by a mutation in ALMS1 gene. Cone-rod dystrophy is its earliest and most consistent feature. Transient infantile cardiomyopathy, sensorineural hearing impairment, obesity, type 2 diabetes mellitus, hypertriglyceridaemia and progressive renal dysfunction are common and present through adolescence and adult life.
Methods: We report the case of an early-diagnosis of ALMS in a child presenting with similar features to spasmus nutans. We aim to highlight the diagnostic challenges of paediatric nystagmus and review the main features of this rare disease.
Results: A 5-year-old girl was referred to our department diagnosed with spasmus nutans. Her prior history included intense photophobia since birth, transient cardiomyopathy at the age of 5 months, poor vision, nystagmus and abnormal head movement starting at the age of 2. All developmental milestones were met and no other systemic conditions were present. Examination revealed a fine horizontal and vertical conjugate nystagmus, fast head bobbing and reduced visual acuity (.05 in each eye). Retinoscopy under cycloplegia showed compound hyperopic astigmatism. Ocular fundus examination was unremarkable. Eletcroretinography was consistent with cone dysfunction. Genetic testing revealed c.647–2A>G (r.spl) and c.1051C>T (p.[Arg351*]) variants in the ALMS1 gene.
Conclusions: Ophthalmologists are in a privileged position to early diagnose ALMS, ensuring further multidisciplinary care to achieve the best possible health outcomes.
Disclosures: None.

P152

THE VALUE OF OPHTHALMIC ASSESSMENT IN PATIENTS WITH UNEXPLAINED ATAXIA: BOUCHER-NEUHAUSER SYNDROME

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Background and Aims: A 30-year-old female presented with progressive ataxia. Her past history included amenorrhoea. She reported 'jumpy' vision and a head tremor, starting aged 17 years. Previous genetic testing for: spinocerebellar ataxias 1, 2, 3, 6, 7, and 12; Friedreich's ataxia; ataxia with oculomotor apraxia types 1 and 2; and ataxia telangiectasia was negative.

Methods: Ocular motor examination revealed saccadic intrusions and hypometric saccades. Visual acuities were 6/12 bilaterally. She correctly identified all of the Ishihara colour plates with the left eye but made five mistakes with her right eye. Automated perimetry showed extensive, patchy peripheral visual field loss bilaterally. Funduscopy revealed extensive, symmetric chorioretinopathy but her optic discs were normal.
Results: Magnetic resonance imaging of her brain showed cerebellar volume loss. Electromyography and nerve conduction studies revealed chronic neurogenic changes with mild, chronic denervation in distal muscles. Pattern electroretinography (ERG) s-waves were undetectable to a standard and large stimulus bilaterally. Dim flash rod ERGs and strong flash ERG a-waves were subnormal bilaterally. Single flash cone ERGs had an abnormal, broad a-wave shape and a markedly subnormal b:a ratio bilaterally. These findings are consistent with generalised retinal dysfunction affecting rods and cones, with severe macular involvement bilaterally.
Conclusions: From her clinical phenotype of ataxia, hypogonadotrophic hypogonadism and chorioretinal atrophy, Boucher-Neuhauser syndrome was suspected. Genetic testing confirmed our patient was compound heterozygous for a pathogenetic PNPLA6 splice-site variant (c2212-1 G > C) and a likely pathogenetic PNPLA6 missense variant (c3548G>A).
Disclosures: None.

P153

'WRONG-WAY' SKEW DEVIATION AS A REFLECTION OF CONCOMITANT FOURTH NERVE PALSY

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Background and Aims: Lesions of the fourth nucleus cause contralateral hypertropia and pathological excyclotorsion. Medial longitudinal fasciculus (MLF) lesions cause internuclear ophthalmoplegia (INO) and/or skew deviation (SD). SD is characterised by ipsilateral (midbrain/pons) or contralateral (medulla) hypertropia and pathological incyclotorsion. When SD and INO co-exit, the hypertropic/incyclopropic eye corresponds to the adduction deficit eye.
Methods: We report the case of simultaneous INO and SD due to a midbrain MLF lesion presenting with pathological incyclotorsion of the eye showing adduction deficit but hypertropia in the eye showing abducting nystagmus, as a reflection of concomitant involvement of the fourth nucleus.
Results: A 73-year-old male with atrial fibrillation presented with oblique binocular diplopia and imbalance. Examination revealed exotropia worse to the left, slow right adducting saccades, decreased left posterior head impulse test and right eye pathological incyclotorsion (0°), all suggestive of right INO and SD. However, a Hess test showed comitant left eye hypertropia. MLF ischaemia was presumed and imaging was ordered. At 1-month follow-up, the INO had resolved, the right eye incyclotorsion had normalised, while the Hess test now revealed left hypertropia, worse in right and downgaze, consistent with a left fourth nerve palsy. Magnetic resonance imaging showed a single ischaemic lesion affecting the right fourth nucleus and MLF.
Conclusions: 'Wrong-way' skew deviation (i.e. hypertropia of the eye showing abducting nystagmus) in a patient with INO, should raise the suspicion for fourth nucleus involvement ipsilateral to the INO. Herein, we have demonstrated that SD was initially counterbalancing the fourth nerve palsy-related hypertropia and causing pathological incyclotorsion.
Disclosures: None.

P154

SPLIT-TENDON MEDIAL TRANSPOSITION OF THE LATERAL RECTUS FOR PAEDIATRIC COMPLETE OCULOMOTOR PALSY

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Background and Aims: Split-tendon medial transposition of the lateral rectus (STMTLR) for complete oculomotor palsy can correct large angles of exotropia in adults, but outcomes are variable and complications are frequent. The aim of my study is to report the outcomes of this technique in paediatric cases of complete oculomotor palsy.
Methods: A retrospective review of outcomes was conducted on five consecutive patients with complete oculomotor palsy treated with STMTLR by a single surgeon between 2012–2011 at a tertiary paediatric referral centre. The primary outcome was the postoperative horizontal alignment, and the secondary outcome was demonstration of gain-of-function activity in the field of action of the paretic medial rectus muscle.
Results: Five cases of paediatric complete oculomotor palsy underwent surgical treatment with STMTLR. Subjects averaged 5.3 years of age (range 10 m – 16 y). Aetiologies were heterogeneous, and all presented with unilateral (n = 2) or bilateral complete oculomotor palsy with exodeviations ranging from 45 to >120 prism dioptres. Two subjects had bilateral disease secondary to miliary tuberculosis with central nervous system involvement. A third subject presented iatrogenically with complete bilateral third nerve palsies secondary to removal of a non-germinomatous germ cell tumour (NGGCT) of the pineal gland. The two remaining subjects had monocular involvement in their right eye: one from compressive neuropathy following a cavernoma midbrain haemorrhage; and one from a congenital right oculomotor palsy. Unilateral STMTLR was performed in all cases except the subject with NGGCT, in whom bilateral STMTLR was performed. Measurement of exodeviations post-op resulted in an average correction of 42 prism dioptres (range 37.5–47.5 prism dioptres) per operated eye. Four out of the five subjects regained limited but active adduction eye movements. The two unilateral cases demonstrated improved convergence.
Conclusions: STMTLR was a safe and effective approach for the surgical correction of paediatric oculomotor palsy in my case series. Paediatric patients may additionally benefit from STMTLR in with immediate gain-of-function activity in the transposed lateral rectus muscle, suggesting that adaptive neuroplasticity of visual target selection predominates over established agonist/antagonist neural signalling.
Disclosures: None

Orbital and Eyelid Disorders

P155

THE USE OF CONTACT LENSES AMONG KERATOCONUS PATIENTS IN SAUDI ARABIA: PREVALENCE, HABITS AND COMPLICATIONS

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Background and Aims: Keratoconus is a progressive, bilateral, asymmetrical condition characterised by corneal ecstasia and a thin, cone-shaped cornea. Keratoconus can affect visual acuity by inducing myopia and irregular astigmatism. Contact lenses (CL) play an important role in the correction of visual problems due to this condition, but pose certain difficulties.
Methods: This cross-sectional study included 112 keratoconus patients who were treated with CLs. The subjects were from different areas of Saudi Arabia. A voluntary self-administered questionnaire was used to collect data regarding prevalence, habits and outcomes of CL use among keratoconus patients.
Results: Of 112 respondents, 84.8% were treated with hard CLs, while 23.2% used soft CLs. Complications were reported among 57.1%, the most common being dry eyes. Regarding hygiene habits, 66.3% reported washing their hands before wearing their CLs, while 33.7% did not. Moreover, 69% of the participants made sure that there were no scratches or breaks in the edges of the CLs before wearing them. The majority of participants reported that they had never slept with CLs in (68.4%), while 13.7% kept their CLs on during naps.
Conclusions: This study shows good practice among keratoconus patients, health education on CL hygiene is recommended to improve patient behaviour and prevent severe complications. In addition, further research must be undertaken to evaluate the awareness of CL-related complications among keratoconus patients.
Disclosures: None.

P156

VRDN-002, A SECOND-GENERATION INSULIN LIKE GROWTH FACTOR-1 RECEPTOR (IGF-1 R) INHIBITORY ANTIBODY FOR THYROID EYE DISEASE: PRECLINICAL PHARMACOKINETICS AND CLINICAL PROMISE

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Background and Aims: VRDN-002 is a novel anti-insulin-like growth factor (IGF)-1 R monoclonal antibody intended for the treatment of thyroid eye disease (TED). It is engineered to incorporate Fc modifications for half-life extension to enable convenient dosing. We sought to compare the pharmacokinetics of VRDN-002 in cynomolgus monkeys to the marketed IGF-1 R antibody, teprotumumab, and to estimate potential human exposures.
Methods: VRDN-002 was administered to cynomolgus monkeys at various doses by intravenous (IV) infusion and subcutaneous injection. Teprotumumab at 10 mg/kg was administered by IV infusion. Antibody levels were measured using a human IgG enzyme-linked immunosorbent assay. Data were analysed with WinNonlin software. A semi-mechanistic model was constructed to estimate human pharmacokinetics.
Results: VRDN-002 pharmacokinetics by IV infusion was linear between 10 and 50 mg/kg. At 10 mg/kg, VRDN-002 half-life and AUCinf were 14 days and 2,300 μg.day/mL, respectively, versus 6.4 days and 779 μg.day/mL, respectively for teprotumumab. Subcutaneous bioavailability of VRDN-002 was 62%. Human exposure estimates were generated from the semi-mechanistic model.

Conclusions: VRDN-002 demonstrated a desirable pharmacokinetic profile in non-human primates, suggesting potential as a novel therapeutic monoclonal antibody for the treatment of TED. The prolonged half-life of VRDN-002 may enable lower frequency of IV infusion and/or low-volume subcutaneous injection; each possibility awaits clinical confirmation.
Disclosures: Viridian Therapeutics, Inc. and Cognigen Corporation.

P157

WHEN A RED EYE IS A RED-FLAG: A NON-TRAUMATIC CASE OF DIRECT CAROTID-CAVERNOUS FISTULA

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Background and Aims: Carotid-cavernous fistula (CCF) is a rare entity that occurs mostly due to trauma. The majority of CCF cases are direct between the internal carotid artery (ICA) and the cavernous sinus (CS).
Methods: We describe a case of a non-traumatic direct CCF secondary to spontaneous rupture of an ICA aneurysm followed by CS and superior ophthalmic vein thrombosis.
Results: A 67-year-old female with hypertension was referred to the ophthalmology emergency department due to a red right eye and eyelid swelling unresponsive to treatment. The right eye had complete external ophthalmoplegia, mild proptosis, a best-corrected visual acuity (BCVA) of 20/50, chemosis and the intraocular pressure was 48 mmHg. Tomographic and angiographic imaging confirmed the diagnosis. A flow-diversion stent and transvenous coil emboliaation were performed with the need for re-intervention. Ocular hypertension was managed with maximal treatment with the need for cyclophotocoagulation, which allowed optic nerve preservation while the reperfusion slowly occurred. Full recovery was achieved with a BCVA of 20/20 and no ocular hypotensive medication were needed.
Conclusions: The differential diagnosis of red eye should always consider sight-threatening and possibly life-threatening conditions as observed in this case.
Disclosures: None.

P158

ADAPTIVE DESIGN IN CLINICAL TRIALS: VRDN-001: A MONOCLONAL ANTIBODY FOR THYROID EYE DISEASE

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Background and Aims: Drug development understandably requires costly and time-consuming exploration of both effectiveness and safety. To increase efficiency and decrease participant burden, novel trial designs different than those historically conducted are needed.
Methods: Adaptive strategies are one such approach. They allow for prospectively planned modifications to aspects of the study based on accumulated data from subjects within the trial, as it is ongoing. Adaptations are prospectively specified before any comparative analyses of trial results are conducted.
Results: VRDN-001 is a humanised IgG1κ monoclonal antibody that binds to human insulin-like growth factor-1 receptor (IGF-1 R) and inhibits activation. A randomised, placebo-controlled multiple ascending dose (MAD) phase 1/2 trial evaluating safety and dose exploration is ongoing (NCT05176639). Driven by data from this trial, further exploration of VRDN-001 could include elements of adaptive design, including a masked interim analysis in which the least promising dose cohort(s) would be stopped with no further recruitment into those cohorts; the trial would continue with the placebo arm and the surviving dose cohort(s) thus protecting statistical validity and sparing thyroid eye disease patients ineffective treatment arms.
Conclusions: Adaptive design permits trial adjustment to information not available at initiation. Advantages include statistical efficiency, number of arms brought forward and reduced sample size requirements. Our exploration of VRDN-001 seeks to protect patient interests and burdens; incorporating principles of adaptive design is a methodology that may advance such interests.
Disclosures: Viridian Therapeutics, Inc and International Drug Development Institute.

P159

CHARACTERISATION OF VRDN-001, A HIGH AFFINITY AND POTENT ANTI-INSULIN-LIKE GROWTH FACTOR-1 RECEPTOR (IGF-1 R) INHIBITORY ANTIBODY FOR THE TREATMENT OF THYROID EYE DISEASE

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Background and Aims: VRDN-001 is an antagonist antibody to insulin-like growth factor-1 receptor (IGF-1 R) under development by Viridian Therapeutics for treatment of patients with thyroid eye disease (TED). Blockade of IGF-1 R has been shown to improve TED symptoms in randomised clinical trials. We evaluated the binding characteristics of VRDN-001 to IGF-1 R and its potency for inhibition of IGF-1 R phosphorylation.
Methods: Surface plasmon resonance was used to determine kinetic parameters and affinity. Binding to cell-surface IGF-1 R was assessed by flow cytometry. Inhibition of receptor autophosphorylation was determined using a commercial ELISA after stimulating primary human ocular choroid fibroblasts and A549 carcinoma cells with IGF-1 in the presence of inhibitory antibodies.
Results: VRDN-001 bound IGF-1 R extracellular domain with .57 nM affinity and exhibited slow dissociation. These results were consistent with cell binding studies. In IGF-1 stimulated human ocular choroid fibroblasts and A549 cells, VRDN-001 inhibited IGF-1 R phosphorylation with an IC50 of ~.1 nM and > 95% inhibition was observed at concentrations in the 1–10 nM range.
Conclusions: VRDN-001 bound IGF-1 R with high affinity and inhibited IGF-1 R signalling with high potency. VRDN-001 shut down IGF-1R signalling at clinically achievable concentrations. This in vitro profile suggests favourable characteristics for treatment of TED patients. A phase 1/2 trial for VRDN-001 in healthy volunteers and TED patients is underway (NCT050176639).
Disclosures: None.

P160

WHEN THE SMILE IS NOT THE MOST BEAUTIFUL CURVE – ODONTOGENIC OSTEOMYELITIS WITH ORBITAL INFILTRATION: A CLINICAL CASE

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Background and Aims: A wide variety of processes can produce space-occupying lesions in and around the orbit, requiring careful examination and history-taking. Causes range from benign lesions to malignant, life-threatening neoplasms that require urgent management.
Methods: We describe the case of a young healthy woman with new-onset diplopia caused by orbital infiltration of a 'mass'.
Results: A 22-year-old woman presented to the emergency room with new-onset binocular diplopia. At evaluation, left eye hypertropia was evident, with diplopia and pain in all directions of gaze. Orbital computed tomography and magnetic resonance imaging revealed sings of endodontic treatment of the 2.7 tooth, with apical rarefaction extending to the lateral wall of the maxillary sinus and densification of the contiguous muscle-adipose tissues with invasion of the left extraconal orbital compartment and distortion of the inferior and lateral rectus muscles. Since chronic osteomyelitis was suspected, the 2.7 tooth was extracted and the patient was treated with oral antibiotics and corticosteroids with full recovery.
Conclusions: Orbital pathology should always be considered in the differential diagnosis of new-onset diplopia. Osteomyelitis of the maxilla is a rare entity typically associated with odontogenic infections. This clinical picture illustrates how this pathological process can progress undetected and infiltrate the surrounding tissues, such as the orbit and its contents.
Disclosures: None.

PUPILS

P161

DIGITAL PUPILLOMETRY WITH STIMULATION OF THE MEDIAN NERVE FOR DIFFERENTIATION OF HORNER'S SYNDROME AND PHYSIOLOGICAL ANISOCORIA

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Background and Aims: Differentiation of Horner's syndrome from physiological anisocoria is an important task in Neuro-ophthalmological practice. We aimed to investigate if automated digital pupillometry combined with sympathetic stimulation of the median nerve ('buzzing') transiently enhances the anisocoria due to a sympathetic innervation deficit but not in physiological anisocoria.
Methods: In a prospective cohort study, we analysed a subgroup of 17 patients with anisocoria who underwent apraclonidine testing to determine the presence of Horner's syndrome. We used digital automated pupillometry under standardised light conditions combined with a 55 mA stimulation of the median nerve 2 seconds after light-off.

Results: Anisocoria at 3–4 seconds after light-off indicating the dilation lag was calculated. 'Buzzing' led to a significant increase of the dilation lag by .34 ± .23 mm in Horner's syndrome (p = .002). No significant change of the dilation lag was seen in physiological anisocoria (.1 ± .31 mm, p = .41). Compared with pupillometry alone, buzzing increased the sensitivity from 75% to 100% with an unchanged specificity of 100% to discriminate Horner's syndrome (dilation lag > .36 mm) from physiological anisocoria.
Conclusions: Stimulation of the median nerve causes a transient pupil enlargement ('bump') which is not recordable in pupils with a sympathetic denervation deficit. The addition of 'buzzing' can help enhance the diagnostic accuracy of pupillometry to distinguish Horner's syndrome from physiological anisocoria making pharmacological testing less important in the future.
Disclosures: None.

P162

TRANSIENT AND SUBCLINICAL HORNER'S SYNDROME REVEALING INTERNAL CAROTID ARTERY DISSECTION

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Background and Aims: Ocular symptoms or signs are frequently associated with internal carotid artery dissection (ICAD). The classic triad consists of pain in the ipsilateral neck, head and orbital regions, Horner's syndrome, and cerebral or retinal ischaemia. Patients presenting with painful Horner's syndrome should therefore require prompt investigations to exclude carotid artery dissection.
Methods: Report of two cases
Results: Case 1: A 61-year-old female presented with right temporal headache, an episode of transient visual loss and drooping of the right upper eyelid. Examination revealed anisocoria, with the smaller pupil in the right eye. Reversal of anisocoria was observed after instilling drops of apraclonidine. Neuro-imaging demonstrated ICAD. Her symptoms all resolved the next day. Apraclonidine testing a few weeks later led to no dilatation of the right pupil. Case 2: A 48-year-old male presented with drooping of the right upper eyelid and headache. Anisocoria was noticed, with cocaine pharmacological testing confirming a right Horner's syndrome. Neuro-imaging revealed ICAD. His symptoms all resolved a weeks later. However, cocaine drop testing still produced anisocoria, compatible with subclinical Horner's syndrome.
Conclusions: Transient or subclinical Horner's syndrome can be the presenting feature in ICAD; whilst straightforward when clinically evident, the characteristic eyelid ptosis and anisocoria may resolve in only a few days. If suspected by clinical history, pharmacological testing may be helpful in identifying subclinical cases.
Disclosures: None.

P163

ADIE ISSUES – A CLINICAL CASE OF ANISOCORIA PRESENTING TO THE EMERGENCY DEPARTMENT

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Background and Aims: Anisocoria is characterised by a difference in pupil sizes, and it occurs as result of disturbances in the efferent pathway dynamics. Aetiological diagnosis is mandatory due to the numerous associated causes, which range from benign to life-threatening.
Methods: We present a case of anisocoria in the emergency room and the consequent diagnostic work-up.
Results: A 41-year-old woman with no relevant medical history presented to the emergency room due to left periocular discomfort. Her visual acuity was 10/10 in each eye. Anisocoria (left > right) was evident and was worse in the light. Her ocular movements were normal and there was no evidence of ptosis. Anterior segment evaluation revealed vermiform movements of the left iris. No relevant signs were found on funduscopy. A diluted pilocarpine (.01%) test produced a miotic response in the left eye. Cerebral and orbital computed tomography and magnetic resonance imaging revealed no abnormalities. Syphilis was excluded. Adie's tonic pupil was diagnosed, and dilute pilocarpine was prescribed, with resolution of complaints.
Conclusions: Adie's syndrome is a relatively common neurological disorder of unknown aetiology comprising unilateral or bilateral tonically dilated pupils with light-near dissociation. When investigating a tonic pupil it is important to rule out other potentially more serious diagnoses such as ciliary ganglion lesions and neurosyphilis before assuming this benign condition.
Disclosures: None.

SARS-COV-2 INFECTION RELATED NEURO-OPHTHALMIC ASSOCIATIONS

P164

BILATERAL OPTIC DISC OEDEMA IN A PAEDIATRIC PATIENT AFTER PRESUMED CORONAVIRUS INFECTION (COVID-19): A CASE REPORT

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Background and Aims: Described first in December 2019, coronavirus disease 2019 (COVID-19) with its aetiological agent the new severe acute respiratory syndrome coronavirus-2 (SARS-CoV-2) has rapidly evolved into a global pandemic. Unlike adults, children are reported to have mild symptoms. Besides the respiratory symptoms, there are already reports of neurological involvement
Methods: We report a case of a young boy diagnosed with bilateral optic disc oedema following presumed infection with SARS-CoV-2.
Results: An 8-year-old patient was referred to our department due to a two week history of headaches and diplopia. His best corrected visual acuity was .9 in each eye but funduscopy revealed bilateral optic disc oedema. His IgM and IgG antibodies for SARS-CoV-2 and D-dimer test were positive. Other infectious and autoimmune laboratory tests were negative. Brain magnetic resonance imaging was normal and genetic testing for Leber's hereditary optic neuropathy was negative. We discounted multi-system inflammatory syndrome in children. Having his laboratory results, we diagnosed optic neuritis due to infection with SARS-CoV-19. He was successfully treated with corticosteroids.
Conclusions: Research into SARS-CoV-2 continues, therefore more data will be provided about the possible manifestations of infection with this novel virus. This case report is an example that shows that besides the involvement of ocular surface described in COVID-19, there can be more serious ocular involvement, such as optic neuritis or papillophlebitis.
Disclosures: None.

P165

BILATERAL NON-ARTERITIC ANTERIOR ISCHAEMIC OPTIC NEUROPATHY: OCULAR MANIFESTATION OF COVID-19 INFECTION

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Background and Aims: Non-arteritic anterior ischaemic optic neuropathy (NAAION) is a result of hypoperfusion of the optic nerve head. It is characterised by unilateral painless vision loss. Bilateral and simultaneous forms are uncommon.
Methods: We report the case of a patient with bilateral nearly simultaneous severe NAAION associated with COVID-19 infection and massive pulmonary embolism.
Results: A 51-year-old woman was admitted for erysipelas and was treated with penicillin G. She was referred for a sudden onset of vision loss of the left eye followed by the right eye 2 days later. On examination her visual acuity was limited to light perception in each eye. Anterior segment examination was unremarkable. Fundus examination revealed chalky white optic disc oedema in both eyes. Her blood pressure was 110/60 mmHg. The day after, she developed dyspnoea requiring her admission to the intensive care unit, her reverse transcriptase polymerase chain reaction test was positive for COVID-19 and a massive pulmonary embolism was noted on the chest scan. She passed away 3 days later.
Conclusions: COVID-19 infection induces a generalised hypercoagulable state associated with a higher risk of embolism. In this case bilateral NAAION was seen with a poor visual prognosis.
Disclosures: None.

P166

NEGATIVE IMPACT OF COVID-19 LOCKDOWN ON PAPILLOEDEMA AND IDIOPATHIC INTRACRANIAL HYPERTENSION

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Background and Aims: The enforced national lockdown due to COVID-19 limited access to medical services. We evaluated the impact on those presenting with papilloedema, and those with a pre-existing diagnosis of idiopathic intracranial hypertension (IIH).
Methods: We carried out a single United Kingdom centre prospective cohort study between 15 May 2020 (start of emergency clinics after the first 8 week national lockdown) and 31 July 2020. Demographics, diagnosis and outcomes were documented and compared with the same period in 2019.
Results: The study recorded 130 patients, 123 with a diagnosis of IIH and 92% were female. Mean ± standard deviation weight was 104.7 ± 25.9 kg. Mean logMAR visual acuity was .13 ± .22. Perimetric mean deviation was −5.69 ± 8.77 dB. Mean optical coherence tomography (OCT) retinal nerve fibre layer (RNFL) thickness was 128 ± 75 µm. Emergency cerebrospinal fluid diversion surgery was required in 13%, a 4.7-fold

(367%) increase compared with the same period in 2019. Weight increased in 58% (mean 6.2 ± 4.6 kg) and corresponded to a significant increase in papilloedema (mean OCT RNFL 15 ± 57.3 μm, p = .014). Elevated anxiety levels occurred in 64%.
Conclusions: There was a 367% increase in emergency shunting to save vision in IIH following national lockdown. Worsening of papilloedema, weight gain, and detrimental effects on mental health were recorded. Counter measures should be implemented to minimise harm in this rare disease during future service restrictions and lockdowns.
Disclosures: Professor Mollan reported personal fees from Heidelberg Engineering and advisory board fees from Invex Therapeutics during the conduct of the study and personal fees from Allergan, Santen, Sathera, Roche, Chugai, and Novartis outside the submitted work. Professor Sinclair reported fees from Invex Therapeutics as a company director with salary and stock options during the conduct of this study; and personal fees from Novartis and Allergan outside the submitted work. No other disclosures were reported.

P167

POST-COVID VACCINATION OPTIC PERINEURITIS WITH MULTIPLE CRANIAL NEUROPATHIES: A CASE REPORT

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Background and Aims: Autoimmune-mediated cranial neuropathy following vaccination is a rare entity. We aim to describe a case of unilateral optic perineuritis with multiple craniopathies in an elderly female post-COVID 19 vaccination.
Methods: Single case report
Results: An elderly Filipina was admitted for right eye pain, upper lid ptosis and diplopia which began after receiving second dose of a COVID vaccine. Examination showed visual acuity of 20/25 bilaterally, complete ptosis and exotropia with multiple motility deficits in the right eye. The rest of the Neuro-ophthalmological examination was unremarkable. Neuro-imaging revealed enhancement and T2-weighted fat saturated hyperintensity of the right optic nerve and nerve sheath. Apart from an elevated erythrocyte sedimentation rate, vasculitic, infectious, inflammatory, autoimmune screening and cerebrospinal fluid analysis were unremarkable. She was diagnosed with autoimmune syndrome induced by adjuvants (ASIA) syndrome as she had cranial nerves 2, 3, 4 and 7 dysfunction. She was treated with 3 day pulse of intravenous (IV) methylprednisolone followed by an oral steroid taper and an infusion of IV immunoglobulins.
Conclusions: Ophthalmologists should be aware that ASIA syndrome presenting as multiple cranial neuropathies, may develop following COVID-19 vaccination.
Disclosures: None.

P168

SEROPOSITIVE NEUROMYELITIS OPTICA FOLLOWING HERPES ZOSTER AND SARS-COV-2 INFECTIONS

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Background and Aims: Neuromyelitis optica spectrum disorder (NMOSD) is uncommon autoimmune central nervous system disease, with classic presentations of myelitis, optic neuritis, and/or area postrema syndrome. Approximately 75% of patients have aquaporin 4 (AQP4) antibodies. It is treated with glucocorticoids and often plasma exchange; and long-term immunosuppression is recommended. Different factors are involved in pathogenesis including infections
Methods: In this case report, a patient who had suffered from Herpes zoster (HZV) and SARS-COV-2 infections, developed later NMOSD which has been confirmed with the clinical and radiological findings along with positive serum AQP4.
Results: A 55-year-old African woman presented with an upper thoracic HZV rash and facial paresis, which quickly progressed into an area postrema syndrome, followed by encephalitis, bilateral visual loss and quadriplegia. Earlier presenting signs were masked by SARS-COV-2 and concomitant HZV infections. Initial magnetic resonance imaging (MRI) of her brain was inconclusive. However, clinical progression and a repeat head and spine MRI with contrast revealed features highly suggestive of NMOSD, supported with positive AQP4 serology. Despite aggressive treatment with high dose glucocorticoids, plasma exchange, intravenous immunoglobulin and cyclophosphamide, she retained profound neurological deficits with partial improvement only.
Conclusions: This is a unique case where both HZV and SARS-COV-2 infections immediately preceded a devastating NMOSD presentation. This case highlights the importance of early and aggressive treatment in NMOSD with additional diagnostic pitfalls when co-existent with an infectious trigger.
Disclosures: None.

P169

LATE ONSET SIXTH NERVE PALSY IN COVID INFECTION

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Background and Aims: Sixth nerve palsy is rare in young adults and is not well studied. Recently COVID-19 has been implicated as a cause of acute sixth nerve palsy.
Methods: Two young adults presented with diplopia of 1 week's duration. They had history of COVID-19 infection 2 months previously. On examination they both had esotropia with limitation of abduction in one eye.
Results: Systemic examination did not reveal any abnormalities. There was no evidence of any acute infections. Visual acuity was 20/20 in each each in both patients with unremarkable anterior and posterior segment examinations. They were positive for SARS-CoV-2 antibodies. Contrast-enhanced imaging of the head and orbits did not reveal any abnormalities. All blood investigations were normal. They were treated with oral steroids and after 2 weeks there was complete recovery in each patient.
Conclusions: The delayed damage appears to be mediated by the immune system as a post infective phenomenon. Our cases shows that delayed presentation can occur in COVID-19 and that it rapidly responds to steroids.
Disclosures: None.

P170

MYELIN OLIGODENDROCYTE GLYCOPROTEIN ANTIBODY–POSITIVE OPTIC NEURITIS AND TRANSVERSE MYELITIS AFTER mRNA COVID VACCINATION

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Background and Aims: The recent pandemic of coronavirus disease 2019 (COVID-19) has given rise to several new vaccines being produced to mitigate its impact. These new types of vaccines have been raising concerns about their side effects, which could involve (neuro)-ophthalmological complications.
Methods: We report the case of a 19-year-old male athlete presenting with headache, painful ocular movements, and blurred vision in one eye 2 weeks after the administration of the Pfizer-BioNTech vaccine.
Results: Visual acuity of the affected eye was not significantly reduced, but a profound central scotoma was present and the optic disc appeared swollen and hyperaemic. Magnetic resonance imaging results were consistent with optic neuriti, affecting the intraorbital and intracanalicular part of the optic nerve. Two weeks later he developed transverse myelitis, presenting with lumbar pain, urinary retention and lower limb paresthaesia. Serum myelin olygodendrocyte glycoprotein (MOG) antibodies were positive. He was treated with pulsed intravenous methylprednisolone and mycophenolate mofetil, which led to complete resolution of the symptoms.
Conclusions: Strong temporal correlation might suggest that vaccine components could potentially trigger specific immune mechanisms, leading to production of MOG antibodies associated with neuroinflammatory disease.
Disclosures: None.

P171

MYELIN OLIGODENDROCYTE GLYCOPROTEIN ANTIBODY-ASSOCIATED OPTIC NEURITIS FOLLOWING SARS-COV-2 VACCINATION

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Background and Aims: Myelin oligodendrocyte glycoprotein (MOG) antibody-associated disease (MOGAD) is characterised by serum or cerebrospinal fluid (CSF) anti-MOG antibodies. The most common phenotype of MOGAD includes optic neuritis. Five cases of acute MOG-ON possibly induced by COVID-19 have been described, and there are several reports after prodromal infections, such as herpes simplex and other viruses.
Methods: We carried out a retrospective review of the medical records including demographic, clinical and imaging data from a case.
Results: A 28-year-old female patient complained of right eye pain worsened by eye movements and blurred vision, starting 5 days after receiving the Moderna COVID-19 vaccine. The visual acuity in the right eye was 20/25 and in the left eye was 20/20. There was normal pupillary function. Fundus examination of the right eye showed 360° optic disc oedema, with flame-shaped haemorrhage. Blood analysis identified anti-MOG antibodies. Optical coherence tomography showed and increase in retinal nerve fibre layer (RNFL) thickness. The Farnsworth-Munsell 100 hue test showed yellow/blue changes and visual evoked potentials revealed prolonged latency and right retrochiasmal pathway delayed conduction. Magnetic resonance imaging (MRI) of the orbits revealed optic nerve asymmetry with thickening and enhancement

along with right optic nerve sheath. She had a normal MRI of her brain and normal CSF analysis. MOG antibody positive acute unilateral optic neuritis possibly induced by the Moderna vaccination was diagnosed and oral prednisolone was started. Full clinical and imaging recovery was achieved.
Conclusions: The association between immunisation and demyelinating manifestations of the central nervous system is well documented, but little is known regarding the complications of COVID-19 immunisation. Our case of optic neuritis following COVID-19 vaccination suggests that, along with infection, immunisation might trigger an anti-MOG autoimmune response. Recognising that will help us diagnose and promptly treat these patients.
Disclosures: None.

P172

VISUAL SNOW FOLLOWING COVID-19 INFECTION

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Background and Aims: Visual snow is a condition consisting of positive visual disturbance described as static and apart from an isolated case in the literature, it has not been previously linked to SARS-CoV-2 infection. We report a case of brain fog and visual snow in a young male following SARS-CoV-2 infection.
Methods: A 21-year old male presented with a few weeks history of visual snow, palinopsia, floaters and photophobia. He also complained of tinnitus and brain fog. Previously, he had been fit and healthy. His symptoms began shortly following diagnosis with SARS-CoV-2 infection. The patient had also received two doses of the Pfizer-BioNTech vaccine.
Results: On Ophthalmological examination there was anisocoria and partial right eyelid ptosis, which had been present from childhood as confirmed from photographs. The rest of the Ophthalmological examination was normal. Magnetic resonance imaging of his brain and orbits was unremarkable apart from the incidental finding of a pituitary microadenoma. Routine blood investigations and autoimmune screen were also within normal limits. At 4-months follow-up his concentration and most of his visual symptoms had started to improve but the visual snow persisted. He was unable to resume his studies due to his symptoms and therefore deferred for a year.
Conclusions: Emerging reports have shown that SARS-CoV-2 infection can cause alterations in brain functioning especially in regions associated with cognition, resulting in conditions like brain fog. Visual snow, just like brain fog, might be an emerging feature of post-COVID syndrome.
Disclosures: None.

P173

POST-COVID 19 VACCINATION NEUROMYELITIS OPTICA SPECTRUM DISORDER

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Background and Aims: There have been several cases of demyelinating disease reported in the literature linked to various vaccines. With the increasing availability of COVID-19 vaccines, there are rising concerns of possible neuroimmune complications. We report a case of neuromyelitis optica spectrum disorder (NMOSD) 3 weeks following receiving the Pfizer-BioNTech vaccine (PBV).
Methods: A 49-year-old female presented with a few days history of worsening right eye vision and pain on eye movement. Three weeks earlier she had received the second PBV dose. There was no history of fever, headache or malaise. He family history was negative for neurological disorders. She had been previously fit and healthy.
Results: On examination the visual acuity in her right eye was 6/12 and in her left eye was 6/6. There was a right relative afferent pupillary defect and right eye colour vision impairment. The anterior segments were healthy. Dilated funduscopy did not reveal any disc swelling or pallor. The rest of the neurological examination was unremarkable. Magnetic resonance imaging of the orbits showed intense contrast enhancement of the right optic nerve along the intra-orbital segment. There were no focal lesions or enhancement involving the brain parenchyma or spinal cord. An autoimmune and infectious screen were negative, including anti-myelin oligodendrocyte glycoprotein antibodies, however aquaporin 4 antibodies (AQP4) were positive.
Conclusions: She was diagnosed with AQP4-positive NMOSD and was pulsed with 1 g methylprednisolone for 5consecutive days with moderate improvement. As the reported cases of NMOSD following PBV remain few and the pathophysiology poorly understood, prospective research is necessary to confirm any link between COVID-19 vaccines and demyelinating disease.
Disclosures: None.

P174

IMPLICATIONS OF SARS-COV-2 INFECTION IN NEURO-OPHTHALMIC MANIFESTATIONS

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Background and Aim: We present a series of three clinical cases of patients with neuro-ophthalmic disorders in the context of the COVID-19 pandemic.
Methods: Three presented to the Ophthalmology clinic: a 45-year-old male patient with bilateral orbital cellulitis; a 52-year-old female patient with post-SARS-CoV-2 optic neuritis; and a 55-year-old male patient with ischaemic optic neuropathy prior to immunisation.
Results: The first case is a 45-year-old male patient who presented with a 4 day history of sudden onset binocular diplopia and painful red eyes. He tested positive for SARS-CoV-2. A head computed tomography scan showed bilateral orbital cellulitis with infiltration of the intra- and extraconal fat and infiltration of the rectus muscles (4.7 mm versus 5.4 mm). The second case is a 52-year-old female patient with a history of COVID-19 infection 1 month previously. She presented with decreased visual acuity in the right eye and a central scotoma. The third case is a 55-year-old male patient with hypertension. He presented with sudden painless decrease in visual acuity in right eye (uncertain light perception) approximately 3 weeks post-SARS-CoV-2 immunisation.
Conclusions: Although the cases were investigated quickly and effectively and the treatment was administered appropriately, in a multidisciplinary team (first and third cases), the evolution was not favourable in all three situations. Atypical neuro-ophthalmic manifestations may be present in the absence of systemic symptoms typical of SARS-CoV-2 infection.
Disclosures: None.

MISCELLANEOUS

P175

EEG AND RETINAL MORPHOMETRIC CONFIRMATION OF AMBLYOPIA TREATMENT EFFICACY WITH ADJUNCTIVE NEUROPROTECTIVE AGENT

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Background and Aims: It has been suggested that the results of traditional pleoptic and orthoptic therapy for amblyopia may be improved by the adjunctive use of neuroprotective agents. The degree of maturation of the cortical structures can be assessed by electroencephalography (EEG), whereas retinal morphometric parameters and be measured by optical coherence tomography (OCT). We aimed to assess amblyopia treatment efficacy with the adjunctive neuroprotective agents citicoline and vitamin B_{12} eye drops.
Methods: EEG rhythm indices and OCT parameters were evaluated in 79 amblyopic children. The treatment group (n = 57) was treated with conventional therapy plus eye drops with a neuroprotective agent for 2 months. The control group (n = 22) were treated with conventional therapy.
Results: More improvements in visual acuity and contrast sensitivity score occurred in the amblyopic eyes in the treatment group in comparison with the control group. Temporal retinal nerve fibre layer thickness in the treatment group increased significantly from 72.5 ± 14.6 μm to 78.5 ± 22.0 μm after treatment. Alpha indices increased to normal values in 73% of children with refractive and in 55% of children with strabismic amblyopia; delta and theta indices reduced.
Conclusions: These results suggest that the neuroprotective agents citicoline and vitamin B_{12} eye drops as adjuncts to conventional therapy of amblyopia improve visual function.
Disclosures: None.

P176

NANOS ILLUSTRATED CURRICULUM AND NANOS NEURO-OPHTHALMOLOGY TECHNIQUES OF EXAMINATION

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Background and Aims: Neuro-ophthalmology education is key for not only ophthalmologists, neurologists, but also other health specialities including emergency medicine, physical therapy among others.
Methods: The North American Neuro-ophthalmology Society (NANOS) has developed a curriculum for Neuro-ophthalmologists called the NANOS Illustrated Curriculum – edited by Dr Sachin Kedar and his team. NANOS has also developed a curriculum of Neuro-ophthalmology examination techniques (NOTE) edited by Dr Karl Golnik and his team.
Results: The NANOS illustrated curriculum is tiered for Neuro-ophthalmologists, Neurologists, and Ophthalmologists as well as learners at various stages of training (students, residents, fellows/practitioners). There are 433 images, 774 videos, 945 pdfs, and 22 audio files obtained through individual contributions from Neuro-ophthalmologists with links to textbooks such as Walsh and Hoyt, Leigh and Zee's Eye Movements and Shirley Wray's text. The NOTE curriculum has videos of all of the common techniques of examination and is

geared for non-Neuro-ophthalmologists and allied health specialists (Ophthalmic assistants, Physical therapists, and Rehabilitation specialists). After surveying 243 other physicians and allied health professionals, we developed 147 learning objects with backgrounds to examination techniques.
Conclusions: Increasing educational tools for all Neuro-ophthalmologists, Neurologists, Ophthalm-ologists and all allied health fields will strengthen Neuro-ophthalmology across the world. These resources are available by subscriptions through your library.
Disclosures: None.

P177

NEW, DIAGNOSTIC FLICKER TEST FOR OPTIC NEURITIS SHOWS SPECIFIC STAGES FOLLOWING DISEASE ONSET

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Background and Aims: The digital flicker test (DFT) examines the subjective brightness of a flickering field (0–60 Hz) and shows distinct patterns in acute optic neuritis (ON) (darkness enhancement at medial frequencies) and healthy eyes (brightness enhancement at these frequencies). We aimed to examine the diagnostic potential of the DFT in acute ON and to investigate the temporal development of the DFT response following ON while comparing with visual evoked potentials (VEP).
Methods: Darkness enhancement was expressed as a quantitative covariate (DFTDE). Results were compared with healthy controls.
Results: 112 patients were examined in the acute phase within < 31 days of onset (median 14.0 days [interquartile range: 8.75–21.0]). An abnormal DFT was present in 104 of the 112 patients (sensitivity 93%). DFT was abnormal in 2 of the 55 healthy controls and in 6 of the 15 differential diagnoses to ON (specificity 89%). Median DFTDE improved to 47.2% at 3 months, to 50.3% at 6 months and to 72.6% at > 8 months from onset. The DFT showed normalisation in 34% at 3 months, in 36.4% at 6 months and in 71.4% at final follow up > 8 months from ON onset compared with 13.3%, 22.4% and 28.6%, respectively for VEP.
Conclusions: We present a DFT that is an easy-to-use and is a sensitive diagnostic test for acute ON. The DFT shows a more pronounced temporal evolution following ON than VEP and may be of use in monitoring the course of ON.
Disclosures: None.

P178

THE TOP OPHTHALMOLOGICAL DIAGNOSES IN A LEVEL ONE NEUROLOGICAL REHABILITATION UNIT: COMPLICATIONS, BARRIERS TO REHABILITATION AND TREATMENT

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Background and Aims: Ophthalmological pathologies are very common following brain injury and can have a great impact on patient rehabilitation. In some cases, the resulting impairments are life changing. We present a case review of the most common complications affecting the eye following brain injury, the barriers to rehabilitation and treatments available.
Methods A case note review was performed of patients who had been admitted to a level one neurological rehabilitation unit in order to establish the frequency of ophthalmological complications. Data were compiled regarding the most common pathologies including treatments, barriers to rehabilitation, patient education and support.
Results: Thirty-five percent of current inpatients have an ophthalmological diagnosis secondary to their brain injury. Traumatic brain injury can be associated with direct trauma to the eye itself, as well as optic nerve damage. Non-traumatic injuries may lead to homonymous hemianopias, cortical blindness, oculomotor palsies, optic neuritis and Terson's syndrome. Common ophthalmological diagnoses are also prevalent in our patient group including exposure keratitis, conjunctivitis, chalazion, styes and blepharitis. Management focuses on specific treatments for individual conditions, patient education and support. Specialist advice from neuro-ophthalmology services is important for diagnosis and treatment. Multidisciplinary team working develops a bespoke rehabilitation plan for each patient.
Conclusions: Ophthalmological diagnoses are common following acquired brain injury and can cause significant barriers to recovery. Multidisciplinary neurological rehabilitation aims to maximise patient recovery through treatment, education and support. Working closely with neuro-ophthalmology services helps to identify problems early, optimise treatment plans and allow for timely follow-up.
Disclosures: None.

P179

SOME MECHANISMS OF TRANSFORMATIONS OF NEUROPLASTICITY AND CEREBRAL VISUAL IMPAIRMENT IN THE LIGHT OF MUTUAL CORRELATIONS OF INTRACRANIAL HAEMO- AND CEREBROSPINAL FLUID DYNAMICS AT THE RECOVERY STAGES OF PERINATAL ACUTE CEREBROVASCULAR STROKES

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Background and Aims: In recent years, research has been expanding in the direction of studying central haemodynamics, general and local intracerebral circulation, as well as orbital and intraocular haemodynamics when studying the dynamics of cerebral visual impairment (CVI) in children with pre- and perinatal encephalopathy.
Methods: I report the findings in 3500 children aged 3 months to 6 years. An acute stroke in the neonatal period occurred in 1500 of them. The following were collected: homoeostasis indicators-catecholamines in the blood; excretion in the urine; the sugar and cortisol in the blood; and electrolytes in plasma and erythrocytes. The reactivity of the blood pressure, the pulse and respiration to vegetotropic drugs and electroencephalogram recording sleep rhythm, emotional and motor activity were measured.
Results: It has been proven that the decisive mechanisms of recovery options depend on the orientation of the hypothalamic-pituitary relationship and the nature of homoeostatic aberrations. The overwhelming majority of these children had experienced catecholamine excite dumping syndrome over the years, and had a clinical picture of both cerebral pathology and cerebral visual impairment. By compiling histograms it was possible to establish that the effect of cholinergic mechanisms on general haemodynamics was less

pronounced than on intraorbital blood circulation. The assessment of haemodynamic responsiveness to the stimulus revealed a more significant fluctuation in the index of intracranial rather than general haemodynamics.

Conclusions: Analysis of variance proves that the stability of the mean dynamic blood pressure is one of the leading mechanisms for stabilisation of adaptive homoeostatic neuroplasty of the central nervous system and visual analyser. Disturbance of autoregulation in the the central retinal artery and superior and posterior cerebral artery play a key role in CVI.

Disclosures: None.