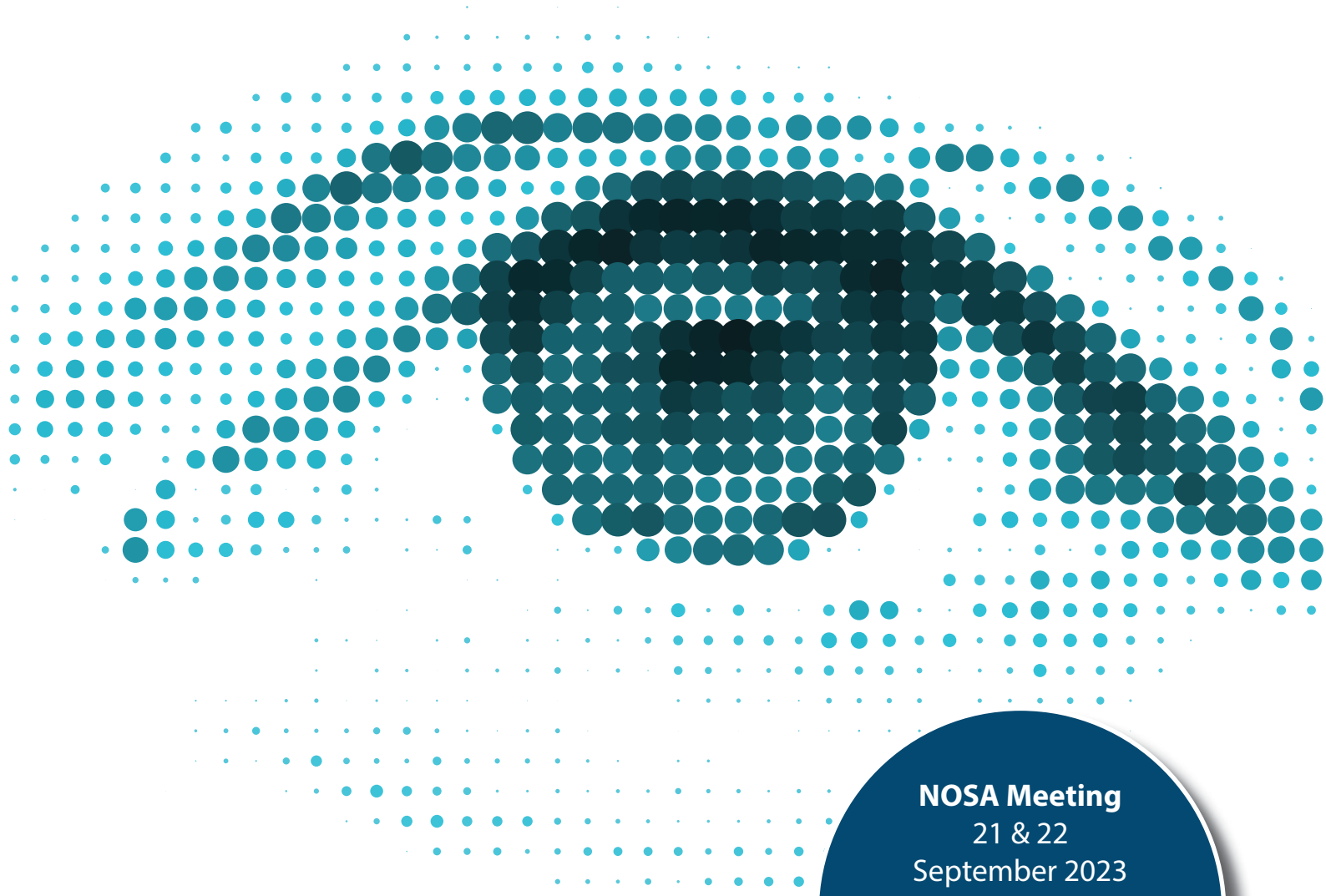
NOSA

Neuro-Ophthalmology Society of Australia

37th Annual Scientific Meeting

Crowne Plaza Hotel, South Australia



NOSA Meeting
21 & 22
September 2023

NeuroVision Training Weekend
23 & 24
September 2023

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Welcome

On behalf of the Council and the Scientific Committee of the Neuro-Ophthalmology Society of Australia (NOSA), I am excited to extend a warm welcome to all of you to the 37th NOSA Annual Scientific Meeting.

I would like to express my gratitude to everyone who contributed their time, especially the organizing committee, whose expertise, sound advice and meticulous planning have culminated in making this meeting happen. Their tireless efforts have ensured that this event is not only intellectually enriching, but also an enjoyable experience for all participants.

I am delighted to welcome our international guest speakers Professor Anthony Arnold from UCLA and Professor Susan Mollan from Birmingham UK. Our lineup of distinguished speakers and presenters is a testament to the quality and depth of the NOSA meeting that awaits us.

This meeting would not have been possible without the ongoing support of our sponsors. My sincere thanks to Abbvie/Allergan, Novartis, Roche, Encapsulate Pharma, Merck, Ipsen, Chiasi, Zeiss, Alexion and TEVA.

I extend my sincere appreciation to each and every one of you – the participants who have travelled from far and wide to be part of NOSA. Your presence and engagement are the lifeblood of this event. At NOSA, we pride ourselves on the collegiality of our society. It's lovely welcome back old friends and make new ones as that's what makes this a fun meeting. I encourage you to seize every opportunity to connect, learn and collaborate.

Thank you and welcome to NOSA 2023!

Professor Celia Chen
President, Neuro-Ophthalmology Society of Australia

NOSA
Neuro-Ophthalmology
Society of Australia

neurovision

General Information

Meeting Venue
Crowne Plaza Hotel
27 Frome Street
Adelaide SA 5000
Telephone: 08 7077 2222

Parking
There is convenient self-parking adjacent to the hotel at Auto Park on Frome Street for $25 for 24 hours.

Meeting Rooms
The conference will be held in the Visionary Ballroom located on the 2nd floor of the hotel.
The posters can be viewed in the Visionary Ballroom.

The reception desk, exhibition & catering will be held on the 2nd floor which includes the Elevate & Collaborate pre function area & Think Tank room.

Name Badges
Please ensure to wear your name badges throughout the meeting which will provided at the registration desk. This badge will be your official pass to sessions, catering, and social functions.

Registration desk
The registration desk will be in the foyer area on the 2nd floor & will be attended on the following days:

Thursday 21st September	7:30am – 4:00pm
Friday 22nd September	8:00am – 4:00pm
Saturday 23rd September	8:00am – 4:00pm
Sunday 24th September	8:00am – 12:00pm

Audio- Visual
Chris Taylor from Expert Audio Visual will be available throughout the conference to assist you with any technical assistance. Chris can be contacted on 0425 242 700.

NOSA Conference Dinner
Thursday 21st September
Time: 6:30pm
Venue: Ayres House
Address: 288 North Terrace, Adelaide
Dress Code: Smart

For any queries throughout the conference, please contact Kathleen Poon 0402 891 804

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References: 1. Dysport® Approved Product Information. **2.** Truong D et al. Parkinsonism Relat Disord. 2008;14:407–414.

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Date of preparation: August 2023. DYS258. DYS-AU-001488.

Prof. Susan Mollan

Susan Mollan is an Honorary Professor at the University of Birmingham and a Consultant Neuro-ophthalmologist at University Hospitals Birmingham (UHB) in the United Kingdom. She is an International Senior Examiner for the Royal College of Ophthalmologists. She is an active board member of the North American Neuro-Ophthalmology Society, on the research committee of the European Neuro-Ophthalmology Society, and the secretary to the British Isles Neuro-Ophthalmology Club and the United Kingdom Neuro-Ophthalmology Society.

She enjoys clinical research and is situated well within the large team in Birmingham to investigate rare conditions that affect the eye and brain. Her professional research specialties include Idiopathic Intracranial Hypertension (IIH), Giant Cell Arteritis (GCA), and the development of quantification of disease through ocular imaging. She is the director of ophthalmology research and the chief data officer for INSIGHT (a Health Data Research UK hub for eye health) at UHB.

Prof. Anthony Arnold

Dr. Anthony Arnold is Director, UCLA Optic Neuropathy Center, and has a research interest in ischemic and inflammatory optic neuropathies. He has authored several texts and more than 100 research publications in neuro-ophthalmology.

He has served as the UCLA ophthalmology residency program director from 1994-2017, and currently is Vice Chair for Education in the department. He has also served as the President of the national AUPO Ophthalmology Program Directors Council, Director of the American Board of Ophthalmology, Chair of the ACGME Residency Review Committee for Ophthalmology, member of the ACGME Board of Directors, Chair of the ACGME Ophthalmology Milestones Development Group, and President and Chairman of the Board of the North American NeuroOphthalmology Society

Pathologist
Associate Professor Sonja Klebe

Associate Professor Sonja Klebe is a surgical pathologist with a special interest in pulmonary pathology, ocular pathology and molecular aspects of diagnosis. Associate Professor Klebe undertook her basic medical training in Germany, obtaining her MD from the Free University of Berlin, graduating with the highest possible score, summa cum laude. She then undertook further studies and obtained her PhD in Immunology and Gene Therapy from Flinders University. She has been the recipient of grants, prizes and fellowships from the German Research Society, the National Society for Excellence in Study Germany, the Ophthalmic Research Institute, NHMRC, Glaxo Wellcome, RCPA and TSANZ. She is author or co-author of four book chapters, as well as approximately 140 peer-reviewed articles.

Associate Professor Klebe was Chief Examiner for the Royal College of Pathologists of Australasia in the basic pathological sciences, coordinating exams nationally and internationally. She is a regular speaker at conferences, both in Australia and overseas, on her chosen areas of interest, with more than 100 presentations to her name.

Neuro-Radiologist
Associate Professor Christen Barras

Associate Professor Christen Barras is a Diagnostic and Academic Neuroradiologist at Jones Radiology, Staff Specialist at Royal Adelaide Hospital, Research Fellow at South Australian Health and Medical Research Institute (SAHMRI) and Clinical Associate Professor of Radiology at The University of Adelaide.

After 5 years of surgical training including maxillofacial surgery, A/Prof. Barras completed a MMed and PhD in Stroke imaging, followed by Radiology training (FRANZCR) at Royal Melbourne Hospital. He completed the Pan-London Neuroradiology Fellowship in 2016, including training at Moorfields Eye Hospital, and is the only practicing Australian Radiologist to have become a Substantive Consultant Neuroradiologist at The National Hospital for Neurology and Neurosurgery, Queen's Square, London (2017).

A/Prof. Barras wrote the Neuroanatomy Chapter of Clinical Neuroradiology, The ESNR Textbook and Grainger and Allison's Diagnostic Imaging. He has several PhD students and broad research interests in stroke, clinical functional MRI, photoswitch molecules for visual reanimation, neuroanatomy, photon-counting CT, the glymphatic system, traumatic brain injury, and altered conscious states including hypnosis. He runs regular neurology and neurosurgery multidisciplinary meetings, registrar teaching and is a sought-after speaker in Neuroradiology.

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Abbreviations: HCP=healthcare professional; RMS=relapsing multiple sclerosis. **References 1.** MS Treatments. MS Australia www.msaustralia.org.au/treatments/ (Accessed 20 March 2023). **2.** KESIMPTA approved Product Information.

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NOVARTIS

9.00	Introduction and Welcome	Celia Chen
Neuroimmunology **Chair: John Crompton and Owen White**		
9.05	Plenary: MOGAD	Tony Arnold
9.35	Paediatric MOG-AD Optic neuritis. A 10 year retrospective review	Andrew Burbidge
9.50	Is MOG the new Syphilis	Chris Ovens
10.00	Neuromyelitis Optica Mimicking optic nerve glioma	Dianita Veulina Ginting
10.10	Exploring factors that prolong the diagnosis of Myasthenia Gravis	Elle Nguyen
10.25	Orbital Pseudotumor: The Role of Immunosuppressive Agents as Corticosteroid-Sparing Therapy	Yunita Mansyur
Morning Tea 10.40-11.10		

Neuro-Ophthalmology Investigations and AI **Chair: Sumu Simon and Neil Shuey**		
11.10	The rise of Neuro-ophthalmology big data and artificial intelligence research	Susie Mollan
11.40	Accessing the utility of Novel iPad-based entoptic perimetry in detection of neuro-ophthalmic conditions	Hong Jing Lee
12.05	Nerve fibre organisation of the human optic nerve and chiasm: what do we really know?	Christian Lueck
12.20	Multiscale modelling of the human optic chiasm: a possible explanation for bitemporal hemianopia?	Pratap Pawar
12.35	A Comparative of the Progression of Visual Fields After Conservative or Surgical Treatment of Pituitary Adenomas	Aine Kelly
Lunch 12.50-1.50		

IIH Symposium
Chair: Christian Lueck and Clare Fraser

1.50	Plenary: Idiopathic Intracranial Hypertension	Susie Mollan
2.20	Paediatric IIH series	Jude Fitzgerald
2.35	Why stenting relieves papilledema	Michael Halmagyi
2.50	Practical aspects of transverse sinus stenting	Hugh (Stephen) Winters
3.05	Stenting in IIH - patient selection & outcomes	Kate Reid
3.20	Visual outcomes of stenting after fenestration	Peter McCluskey

Afternoon Tea 3.35-4.00pm

Hereditary eye diseases symposium
Chair: Kate Reid and Susan Carden

4.00	Plenary: Leber's Hereditary Optic Neuropathy	Anthony Arnold
4.30	Inherited retinal diseases masquerade as papillodema and optic atrophy	Fred Chen
4.45	Beyond the disc elevation	Reema Madike
4.55	If it is not papillodema, then what could it be?	Chris Ovens
5.05	Feeling the pressure	Muhammad Khan
5.15	Assessment of Spontaneous Venous Pulsations in Idiopathic Intracranial Hypertension: A Pilot Study	Solmaz Bastani

AGM 5.30pm – 6.15pm

Conference Dinner 6:30pm

Ayres House
Address: 288 North Terrace, Adelaide
Dress Code: Smart

Walsh William Case sessions **Chair: Helen Danesh-Meyer**		
9.00	Plenary: OCT in Neuro-Ophthalmology	Susie Mollan
9.30	Cranial chaos	Ella Suo
9.45	Just another IIH patient	Randolph Dobson
10.00	To be determined	Eliot Smolyansky
10.15	More than meets the eye	Rachael Jeffrey
11.35	It's not all about the past	Jordan Ng
Morning Tea 10.45am-11.15am		
Walsh William Case sessions **Chair: Christian Lueck**		
11.15	Asian Pacific Plenary	Clare Fraser
11.35	Germ Warfare	Blake Colman
11.50	Blurred Vision? What a headache.	Garry Singh
12.05	How many ?	Jo Black
12.20	Metastatic vision loss	Sean Mullany
12.35	It's all in the eyes	Anna Tierney
Lunch 12.50pm- 1.50pm		

Vascular symposium
Chair: Ian Francis and Sara Booth-Mason

1.50	Plenary: Vascular supply of the eye	Tony Arnold
2.20	OCT angiography changes in acute NAION: A case series and protocol for a prospective longitudinal study	Garry Singh
2.35	Clinical Characteristics of Patients with Carotid-Cavernous Fistula (CCF) at a Tertiary Hospital	Sabrina Wardani
2.50	Preliminary Data from 6 year follow up of the impact of continuous positive airway pressure treatment on cardiovascular and cerebrovascular health outcomes in obstructive sleep apnea	Divya Rodrigues
3.05	Why the ipsilateral pupil was irregularly dilated five days following PERFECT pterygium surgery…!	Peter Tweedie

Afternoon Tea 3.20pm-3.45pm

GCA and tumours
Chairs: Mark Paine and Catherine Dunlop

3.45	Plenary: Giant Cell arteritis	Susie Mollan
4.15	The Temporal Arteritis Proforma Revisited- 2023	Aleeza Fatima
4.30	FESStival of Errors	John Crompton
4.40	Streptococcal Meningoencephalitis with WEBINO, bilateral blindness, pupil-involving third nerve palsies, right facial palsy, and upper motor neurone signs.	Amitouj Singh
4.50	A traumatised nerve	Andrew Coote
5.00	Evaluating immune-mediated contributors to acute uveitis	Serge Geara

Announcement of prizes
Thank you and Conclusion

MOGAD Optic Neuritis: Evolving Concepts

Author:
Prof. Anthony Arnold

Over the past two decades, our increased understanding of non-MS related autoimmune optic neuritis has created new paradigms for management. Myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) in particular has more recently been characterized, and recommendations for management are still evolving as new data become available. This session will review newly developed diagnostic criteria and critically evaluate the ongoing developments in treatment options.

Paediatric MOG-AD Optic neuritis. A 10 year retrospective review

Authors:
Dr Andrew M Burbidge, Dr Stephanie Tiew, Dr Eppie Yiu, Dr Shivanand Sheth

Presenter:
Dr Andrew Burbidge

Institution:
Royal Children's Hospital Melbourne

Email:
Dr.Andrew.Burbidge@gmail.com

Introduction: Myelin oligodendrocyte glycoprotein (MOG) antibody-associated disease (MOGAD), is a rare neuro ophthalmic disease entity with a paediatric predilection. The spectrum of presentations may include, any of: optic neuritis, acute disseminated encephalomyelitis(ADEM) and/or transverse myelitis. The aim of this study is to report the optic neuritis associated presentations at our institution of MOGAD positive cases.

Methods: In line with the tenant of the Declaration of Helsinki, we conducted a single center 10 year retrospective review of clinical case notes, serology and multimodal imaging result in a paediatric population. Mining the digital electronic medical records for all ICD coded H46 optic neuritis (including all subcodes) with data filtering for MOG positive serology (MOGAB and/ or MOGABO).

Results: A total of 80 cases of sero-positive MOGAD children were identified. Removing duplicate cases and those cases with ADEM and/or transverse myelitis. A subtotal of 33 cases of MOGAD (MOG-AB and/ or MOGABO) positive optic neuritis were admitted from the 1 st of January 2013 to 30 th June 2023. The Incidence of seropositive MOGAD optic neuritis at our institution was 4.351 cases per 100,000 admissions. Demographics: 64% (n=21/33) were female. Median age at presentation was 10.1yrs, [range 5-16 yrs.]. Bilateral cases were presents in 45.5% of presentations [n=15/33]. Clinically a RAPD was present at admission in 39.4% of cases.

All cases underwent MRI imaging brain and optic nerves and were treated with 5 days of Intravenous methylprednisolone as per a standard protocol. Median follow up was 655 days. Analysis of visual acuity and OCT Retinal Nerve fibre layer findings will be presented.

Conclusion: We present a large Australian cohort of paediatric MOGAD positive cases of optic neuritis. The Incidence of seropositive MOGAD optic neuritis at our institution was 4.351 cases per 100,000 admissions. Here-in we describe the demographic, visual acuity, and OCT findings of these cases to support the growing body of literature within the field.

Is MOG the new syphilis?

Authors:
Dr Chris Ovens, Dr John Leaney, A/Prof. Clare Fraser

Presenter:
Chris Ovens

Institution:
Save Sight Institute

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ovens.chris@gmail.com

Introduction: MOG-associated optic neuritis may masquerade as a variety of other optic neuropathies. We describe a case initially diagnosed as non-arteritic anterior ischaemic optic neuropathy in an 84yo man. We highlight the clinical features and considerations in diagnosis and treatment.

Clinical details: An 84yo man presented with 1 week of painless visual loss in the right eye. There were no other ocular or systemic symptoms. He had a history of non-insulin-dependent diabetes mellitus and hypertension. Visual acuity was RE HM and LE 6/9, with a right RAPD. Examination showed RE 360deg disc swelling with splinter haemorrhages, and LE inferior swelling, without intraocular inflammation. HVF showed inferior loss in LE. Inflammatory markers were normal. He received pulsed intravenous methylprednisone followed by oral prednisone taper, without significant improvement. Temporal artery biopsy was normal. MRI could not be performed due to positioning.

He presented 8 months later with new visual changes in the left eye. There was new 360deg disc swelling in the left eye, and his vision deteriorated from 6/12 to HM over the subsequent weeks despite a further course of oral prednisone. Additional bloods sent at this time returned a positive MOG-Ab on cell-based assay. A further trial of IV methylprednisone followed by IVIg was commenced, with no improvement. MRI showed asymmetrical nerve enhancement.

Discussion: This case highlights the difficulties in diagnosis of MOG-associated optic neuritis, which can present with a swollen optic nerve in older men, who may happen to have a disc-at-risk in the fellow eye. However, in one study, MOG antibody positive predictive value was 72% (even lower for low titre results), highlighting the importance of clinical assessment and the potential for false positive results. Ultimately this raises several questions – should more NAION patients have MOG testing done? Should more NAION patients receive steroids? And is MOG becoming another "great imitator"?

Neuromyelitis Optica Mimicking Optic Nerve Glioma in Children

Authors:
Dianita Veulina Ginting, Antonia Kartika Indriati, Rusti Hanindya Sari, Prettyla Yollamanda

Presenter:
Dianita Veulina Ginting

Institution:
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Introduction: Neuromyelitis optica spectrum disorder (NMOSD) is a central nervous system inflammatory demyelinating disorder characterized by recurrent optic neuritis and extensive longitudinal transverse myelitis. We present a case of NMOSD that was first suspected to be caused by a compressive lesion and attributed to an optic nerve glioma. Description: A 16-year-old girl presented to Neuro-ophthalmology outpatient services with a second episode of vision loss. The first episode was six months ago, when she developed visual loss in the left eye, which progressed to counting fingers over a period of one week. Brain Gadolinium-enhanced magnetic resonance imaging (Gd-MRI) suggested optic neuritis. She received intravenous methylprednisolone (1 g daily) for three days. Her vision gradually improved to 0.8 in two months. The second episode presented with vision loss in both eyes. Visual acuity was 0,1 and 0,5 in the right and left eyes, respectively. A visual field defect revealed bitemporal hemianopia, and brain Gd-MRI suggested an optic pathway glioma, Dodge stage 3. The vision loss progressed to no perception of light on the right eye and light perception on the left eye over the course of one week. In view of the recurrent episodes of acute visual loss, anti-aquaporin 4 antibody testing was done, which was positive. She received intravenous methylprednisolone (1 g daily) and Intravenous Immunoglobulin (IVIG) for five days, followed by oral tapering of methylprednisolone. In addition, she started on oral methotrexate (7,5mg/week). Her best corrected visual acuity on four months' follow-up was 0.8 and 0,63 in the right and left eyes, respectively.

Conclusion: This case underscores the importance of a detailed clinical history and a high index of suspicion for NMOSD in patients presenting with recurrent visual loss. Given the prognosis and the potential for the disease to recur, early diagnosis and prompt management with aggressive immunomodulation are indicated.

Exploring factors that prolong the diagnosis of Myasthenia Gravis

Authors:
Elle Nguyen, Meaghan Clough, Belinda Cruse, Anneke van der Walt, Owen White, Joanne Fielding

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Background and objectives: Myasthenia gravis (MG) is a condition with significant phenotypic variability, posing a diagnostic challenge to many clinicians world-wide. Prolonged diagnosis can lead to reduced remission rates and morbidity. This study aimed to identify factors leading to a longer time to diagnosis in MG that could be addressed in future to optimise diagnosis time.

Methods: Between 2010-2021, 110 newly diagnosed patients identified via electronic medical records across 3 institutions in Melbourne, Australia were included in this retrospective cohort study. Demographic and clinical data were collected for these patients over the first 5 years from diagnosis and at 10 years. Non parametric statistical analysis was used to identify factors contributing to a longer diagnosis time.

Results: The median time for MG diagnosis was 102(345) days. 90% of patients were diagnosed before one year. Females took longer than males to be diagnosed ($p= 0.013$). The time taken for first presentation after symptom onset contributed most to diagnosis time (median 17[141] days), with females and not-working as contributory factors. Neurology referral took longer if patients had diplopia ($p= 0.022$), respiratory ($p=0.026$) symptoms, or saw an ophthalmologist first ($p<0.001$). Outpatient management compared to inpatient was associated with a longer time to be seen by a Neurologist from referral ($p<0.001$), for the first diagnostic result to return ($p=0.001$), and for the result to be reviewed ($p<0.001$). Ocular MG had a median greater time to Neurologist review than generalised MG (median 5[25] days vs 1[13] days, $p=0.035$). Electrophysiology tests took longer for outpatients than inpatients (median 21[35] days vs 2[8] days, $p<0.001$). Outpatients were also started on treatment later than inpatients ($p <0.001$). There was no association of MG severity, ethnicity, age, medical and ocular comorbidities, public or private health service on diagnosis time. There was also no impact of time to diagnosis on Myasthenia Gravis Foundation of America (MGFA) outcomes, number of follow-ups or hospitalisations, or prevalence of treatments used. This study is limited by low patient numbers and its retrospective nature.

Conclusion: This study identified several factors that can contribute to a prolonged diagnosis time of MG. Patient and clinician education about MG and outpatient diagnostic efficiency needs emphasis. Further studies are also needed to explore the delayed presentation time of women and non-working patients in MG.

Orbital Pseudotumor: The Role of Immunosuppressive Agents as Corticosteroid-Sparing Therapy

Authors:
Cindy Hartono[1,3] , Yunita Mansyur[1,3,4] , Suriani Alimuddin[2,3] , Andi Pratiwi[1,3,4]

Presenter:
Yunita Mansyur

Institution:
[1] Department of Ophthalmology, Faculty of Medicine, Hasanuddin University
[2] Department of Internal Medicine, Faculty of Medicine, Hasanuddin University
[3] Wahidin Sudirohusodo Hospital, Makassar, Indonesia
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Background: Orbital pseudotumor is a noninfective inflammatory condition of the orbit with no identifiable local or systemic cause. Corticosteroids are the established primary treatment for orbital pseudotumor. Up to 80% of patients treated with steroids alone will eventually relapse during steroid taper and require the use of steroid-sparing agents.

Case illustration:
Case 1. A 31-year-old man presented with ptosis, swelling, pain, and ophthalmoplegia in the right eye. The ophthalmological examinations, laboratory, and radiological findings supported a myositis. The patient was treated using corticosteroid and cyclosporine A for three months and showed improvement for ptosis in the right eye with no more pain.

Case 2. A 23-year-old woman presented with ophthalmoplegia in both eyes, swollen eyelid and pain in right eye. The ophthalmological examinations, laboratory, and radiological findings supported dacryoadenitis and myositis. The patient was treated using corticosteroid and tacrolimus for two months and showed improvement for the dacryoadenitis with no more pain.

Discussion: Our case report highlighted the role of cyclosporine A and tacrolimus as steroid-sparing therapy with satisfactory outcome. Both cyclosporine A and tacrolimus are calcineurin inhibitors that inhibit T cell activities and inflammatory cytokines production.

Conclusion: Systemic corticosteroid remains the primary treatment for orbital pseudotumor cases. However, in such recurrence cases on steroid taper, the immunosuppressive drugs can be used as steroid-sparing agents. Cyclosporine A and tacrolimus had satisfactory outcome in our study but still need more time to conclude the therapy response.

Keywords: Orbital pseudotumor, cyclosporine A, tacrolimus

The rise of Neuro-ophthalmology big data and artificial intelligence research

Author:
Prof. Susie Mollan

Big data refers to large volume of data sets that are analysed in the health care environment to derive patterns of diseases and provide new information that may not have been discoverable by smaller cohort studies. Due to advances in storage, connectivity and artificial intelligence our ability to pool data and interrogate it have been made easier. Applying deep learning to retinal images for neuro-ophthalmology has been championed by the Brain and Optic Nerve Study with Artificial Intelligence (BONSAI) who have developed, trained and tested a deep learning system for the detection of papilloedema from fundus colour images. Detecting neurological diseases early with analysis of retinal and optic nerve head optical coherence tomography images represents a unique opportunity for Alzheimer's disease dementia which is set to exponentially increase in prevalence (AlzEye). Leveraging data from all sources, from electronic medical records to country wide databases, has found new patterns of prescribing and management in idiopathic intracranial hypertension that would have not been discovered without the directed use of large datasets. While there are limitations of these types of research, many barriers are being considered and overcome.

Learning objectives:

1. Understand the recent research findings for detection of papilloedema.

2. Recognise the opportunities and limitations of large datasets in rare diseases such as idiopathic intracranial hypertension.

3. Describe the challenges of directly employing artificial intelligence tools into clinical care.

4. Identify areas in your practice that could help large data studies in the future (i.e. maintaining clear records, engaging with coders, and deleting inadequate images).

5. 5. Develop an understanding safety in heath data research.

Assessing the utility of Novel iPad-based entopic perimetry in the detection of neuro-ophthalmic conditions

Authors:
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Presenter:
Hong Jing Lee[1]

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Background: To conduct a pilot study evaluating a novel tablet entopic perimeter, Visual Field Fast (VFF) in detecting neuro-ophthalmic scotomas compared to the standard Humphrey Visual Field (HVF) assessment. VFF's flickering screen allows for the rapid identification of scotomas.

Methods: A case-control study including 31 neuro-ophthalmic subjects (47 eyes) with 30 healthy controls (30 eyes) was conducted at a Singaporean tertiary eye centre. Comparisons were made between VFF and HVF: (1) Scotoma detection and localisation. (2) VFF scotoma area (%) agreement and correlation with HVF scotoma area, severity indices (Visual Field Index and Mean Deviation.) (3) Test times and qualitative descriptors of scotomas.

Results: With HVF pattern deviation, VFF's whole field scotoma detection sensitivity was 34/43 (79.1%) and specificity was 32/34 (93.9%) (kappa = 0.713). On quadrant localisation, sensitivity was 62/100 (62.0%) and specificity was 179/204 (87.7%) (kappa = 0.515). Scotoma area was underestimated (10.7% versus 38.0%, $p<0.01$) but correlated positively with HVF area ($r = 0.514$, $p <0.001$) and negatively with VFI ($r = –0.533$, $p<0.001$) and MD ($r = –0.446$, $p<0.01$). With HVF total deviation, VFF's scotoma detection was 81.0% sensitive and 94.1% specific (kappa=0.738) with a poorer quadrant localisation of 50.8% sensitivity and 87.5% specificity (kappa=0.400). Difference in scotoma area increased (10.7% versus 59.0%, $p<0.01$). Qualitatively, 35/39 (89.7%) subjects perceived scotomas as areas of reduced luminance and movement. VFF was faster than HVF SITA-Standard in neuro- ophthalmic (3.40±1.79 min versus 4.02±0.975 min, $p<0.02$) and control eyes (1.11±0.485 min versus 5.19±0.697 min, $p < 0.01$).

Conclusion: VFF accurately detected scotomas in neuro-ophthalmic patients for a broad range of pre-chiasmal, chiasmal and retro-chiasmal diseases. VFF demonstrated potential as an affordable and accessible screening tool for patients with a high clinical suspicion or in low resource settings.

Nerve fibre organisation of the human optic nerve and chiasm: what do we really know?

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Introduction: A recent anatomical study of the human optic chiasm cast doubt on the widespread assumption that nerve fibres in the human optic nerve and optic chiasm are arranged retinotopically: all nerve fibres passing through the chiasm, both ipsilateral and contralateral, appeared to cross each other multiple times and at many different angles. Preservation of retinotopy therefore seemed unlikely, suggesting the concept may have been based on historical conjecture.

Aim: To perform a scoping literature review of the microscopic anatomical arrangement of nerve fibres passing through the human optic nerve and chiasm.

Methodology: Initial systematic review of the literature supported by subsequent secondary referencing. Meta-analysis of quantitative information regarding nerve fibre numbers in the human optic nerve was performed with subsequent qualitative analysis of information relating to nerve fibre arrangement.

Results: The mean number of fibres in each optic nerve was 1.02±0.23 million with a large inter-individual range not accounted for by loss of nerve fibres with age. Very few published studies have addressed nerve fibre arrangement in the human optic nerve and chiasm. Available information suggests that nerve fibres in the anterior (orbital) portion of the optic nerve might be retinotopically arranged but that this arrangement is probably lost posteriorly with a more random organisation of nerve fibres, particularly when fibres pass through the chiasm. Multiple nerve fibre crossings are likely during the passage of individual nerve fibres through the chiasm for both ipsilateral and contralateral fibres. Of note, most nerve fibre crossings occur paracentrally, nerve fibres in the centre of the chiasm travelling coronally and largely in parallel.

Conclusion: The assumption regarding preservation of retinotopy in the human optic nerve and chiasm is probably not correct.

Significance: Further work will provide more precise anatomical information. In the meantime, information presented in anatomical and medical textbooks should be revised.

Multiscale modelling of the human optic chiasm: a possible explanation for bitemporal hemianopia?

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Introduction: The precise explanation for bitemporal hemianopia resulting from chiasmal compression remains unclear. Previous Finite Element Models (FEM) have supported the explanation offered by the 'crossing hypothesis', namely that nerve fibres which cross each other are more vulnerable to compression than those that run in parallel. However, previous FEMs did not incorporate the recent anatomical finding that all nerve fibres passing through the chiasm, both ipsilateral and contralateral, actually cross each other multiple times and at many different angles.

Aim: This study aimed to generate a more robust FEM, taking into account the number of crossings experienced by individual nerve fibres, the location of these crossings, and the angles subtended at each crossing.

Methodology: The locations, numbers, and angles of crossings were derived from an algorithm developed to simulate nerve fibre pathways traversing the chiasm. This information was then combined with a multi-scale FEM of chiasmal compression to model the strain experienced by individual axons as they passed through the chiasm: specifically, the cumulative normalised angle (CNA) and cumulative normalised (axonal, equivalent) strain (CNS) along the length of both ipsilateral and contralateral fibres were derived.

Results: A significant difference was observed between CNA and CNS values for contralateral and ipsilateral fibres, namely that 85% of contralateral fibres were potentially vulnerable to compression compared to only 25% of ipsilateral fibres. Of note, the model suggested that nerve fibre crossings were concentrated in the paracentral regions of the chiasm, consistent with recent anatomical findings.

Conclusion: Though based on updated underlying assumptions, the updated model continues to support the 'crossing hypothesis' as a potential explanation for the occurrence of bitemporal hemianopia.

Significance: The use of CNA and CNS to assess nerve fibre damage is novel and could potentially help explain the dysfunction resulting from nerve compression at other locations.

A Comparative of the Progression of Visual Fields After Conservative or Surgical Treatment of Pituitary Adenomas

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Pituitary adenomas comprise up to 16% of intracranial neoplasms. Although the majority of these are asymptomatic, primary ocular effects can include monocular and chiasmal visual field defects. Monitoring of visual fields is important to assess progression and response to treatment, which may be conservative or surgical. This study aimed to investigate the type of visual field defects seen in patients with pituitary adenomas and compare the progression of visual fields and size of tumours in patients who underwent surgical versus conservative management.

A retrospective review of 100 patients with pituitary adenomas who attended a pituitary clinic under an endocrine consultant between 1 April 2022 and 30 April 2023 was conducted. Data was analysed including types of pituitary growth, treatment and their outcomes. Tumour size was measured radiologically and the largest dimension was used to standardise results. Patients' eyes were investigated separately.

The cohort studied comprised 53 (53%) males and 47 (47%) females (mean age = 60 years). 52% were managed surgically. 71% of patients underwent Humphrey Visual Field 30-2 Testing. 83/142 (58%) eyes had no visual field defects. The most common visual defects were hemianopias (n=30, 35%), followed by superior temporal defects (n=17, 11%). Prior to treatment, larger visual field defects and tumour size were seen in patients treated surgically, with average mean deviation (MD) -4.9 and size 23.6mm, versus conservatively treated tumours, MD -4 and size 16.3mm. The average change in tumour size was 19.5mm after surgical intervention and 1.1mm after conservative management. Additionally, improvements in visual field defects for surgically managed patients were greater when compared to conservative management.

Our study supported the importance of visual field testing to highlight progression and response to treatment. We found patients who underwent surgical management had a greater change in both tumour size and visual field improvement than those managed conservatively.

Idiopathic intracranial hypertension

Author:
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Idiopathic intracranial hypertension is characterized by raised intracranial pressure that triggers disabling headaches and can cause permanent visual loss. There are no licensed treatments for the condition and the majority of approaches to managing the disease prioritize resolution of papilledema. There is a spectrum of the disease ranging from mild to severe, and for the majority of patients the visual prognosis is good. However, around 7% require escalation of treatment to save sight, with surgical choice being largely dictated by local expertise. Evidence is emerging that idiopathic intracranial hypertension is a systemic metabolic disease and there are a number of different factors that need to be considered. Addressing headache is of key importance to the patient and reducing long-term morbidity of the disease. Options for weight management should be sensitively discussed. In the future we should aim to reduce cardiovascular risk and improve maternal health by reducing the risk of preeclampsia, gestational diabetes and the need for caesarean-section at delivery.

Learning objectives:

1. Summarize the systemic disease associations of Idiopathic intracranial hypertension.
2. Differentiate between mild and sight threatening disease.
3. Describe the controversy in the management of visual failure in of Idiopathic intracranial hypertension
4. Discuss management of post- of Idiopathic intracranial hypertension persistent headache.
5. Consider the challenges of managing pregnancy in people with of Idiopathic intracranial hypertension.

Paediatric Idiopathic Intracranial Hypertension

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Audit was performed over 6 month period in 2022 and captured all patients undergoing review or new diagnosis for idiopathic intracranial hypertension. 21 patients met criteria, with 4 new diagnoses in the audit period. Retrospective analysis of casenotes from presentation was performed to determine cohort characteristics with regards to age, gender, BMI (where available), referral source, barriers to access, presenting symptoms, clinical findings, clinic imaging characteristics (with focus on OCT findings), radiological findings and lumbar puncture opening pressures. Average age of cohort was 11.7 +/-4.1 years with age range from 4 to 17 years of age at diagnosis.

Why stenting relieves papilledema

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Pseudotumor cerebri has many potential causes and can itself cause, in addition to papilledema and vision loss, headache, pulsatile tinnitus and skull base CSF leaks. Stenosis of the dominant transverse sinus is perhaps invariably present and can be due to intrinsic problems such as arachnoid granulations or to extrinsic compression from the intracranial hypertension itself. Stenting the stenosis will almost always relieve the papilledema but not always the headache. Here we present the physiological basis of the intracranial hypertension, supported by a mathematical model based on the Starling resistor – resistance increases with pressure. (Ahmed RM et al Transverse sinus stenting for idiopathic intracranial hypertension: a review of 52 patients and of model predictions. AJNR Am J Neuroradiol. 2011 Sep;32(8):1408-14.)

Practical aspects of transverse sinus stenting for IIH

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Management of Idiopathic Intracranial Hypertension (IIH) has been revolutionized by the advent of transverse sinus stenting (TSS). This review elucidates the practical aspects of TSS, outlining the patient's journey from diagnostic evaluations to postoperative management, and addressing the risk of recurrence.

Key to patient selection for TSS is preliminary cerebral venography and manometry, conducted several weeks prior. These tests discern sinus morphology and pressure gradient, dictating TSS suitability. The subsequent stent placement is less invasive compared to traditional surgical methods and requires an experienced neurointerventionalist.

TSS has demonstrated efficacy in crucially resolving papilledema, thus preventing vision loss and significantly enhancing the patient's quality of life. Mitigation of other symptoms is less comprehensive. However, potential periprocedural risks exist and must be transparently revealed to prospective treatment candidates.

Postoperative management, involving antiplatelet therapy, is paramount to prevent thrombosis, thus bolstering the safety and efficacy of TSS. Nevertheless, the risk of stenosis recurrence and symptom relapse necessitates continuous patient monitoring, with the help of a multidisciplinary team.

In conclusion, while TSS signifies a significant stride in IIH treatment, understanding its practicalities, from diagnosis to postoperative care, is vital. Further data is necessary to streamline patient selection, refine procedural techniques, and optimize postoperative management, thereby enhancing patient journey and outcomes.

Stenting in IIH - patient selection and outcomes

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Around 18% of idiopathic intracranial hypertension (IIH) is refractory to maximal medical therapy: some combination of acetazolamide, topiramate and weight loss. Innovations such as slow release exenatide may reduce the burden of medically refractory disease in the future, but currently at least, procedural intervention is the next step for these patients.

The ophthalmologist is ideally placed to identify those needing a procedure. Intervention is indicated when despite maximal medical therapy, there remains a threat to vision: progressive visual field loss, or persisting/ recurrent papilledema. Intolerable medication side effects, headache and patient sex also need to be considered.

90% of patients with medically refractory IIH will have a significant transverse sinus stenosis gradient of ≥ 8 mmHg at catheter venography. Endovascular stenting of the stenosis eliminates venous hypertension, and achieves truly impressive results: resolution of papilledema in 96%, and cessation of acetazolamide in 81%.

Other advantages of stenting include a safety profile that is favourable compared to CSF shunting: a lower rate of complication at < 1%, and a much lower rate of revision at 6%. The latter contrasts starkly with the 85% rate of shunt revision expected over a patient's lifetime. Finally, in the era of clot retrieval for stroke, stenting by a neurovascular interventionalist is more readily accessed than optic nerve sheath fenestration by an orbital ophthalmic surgeon in most Australian locations.

Visual outcomes of stenting after fenestration for IIH

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AT RPA Hospital Sydney, we have been shunting and fenestrating for over 30 years, and stenting for the last 20. What we recommend to patients depends both on the cause and the effect of their intracranial hypertension. Ideally, the management of each patient is individualized in a multidisciplinary Pseudotumour Cerebri Clinic.

In fulminant papilledema we recommend an immediate optic nerve sheath fenestration (ONSF) of the worse side. If there is no visual improvement within a week, then an ONSF of the better side is performed, followed in idiopathic intracranial hypertension (IIH) by a transverse sinus stent (stent). If there are concerns about postoperative surveillance after unilateral ONSF, in IIH fenestration is immediately followed by a stent.

In this presentation, we review our experience with ONSF in 70 eyes of 35 patients. There was a wide range of visual acuity and visual field outcomes following ONSF, with similar results from unilateral and bilateral surgery. Visual field mean deviation (MD) was more likely to improve than visual acuity (VA). The only patients failing to improve in either eye, and/or with continued deterioration after surgery, presented late with established optic disc atrophy.

Leber Hereditary Optic Neuropathy (LHON)

Author:
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While idebenone therapy for LHON has shown limited promise, the search for a more effective modality is ongoing. In recent years, several research groups have attempted gene therapy in this disorder, with controversial results. This session will review the basis and results of the major clinical trials, along with the controversies generated by them, focusing on the Gensight studies, RESCUE, REVERSE, and REFLECT.

Inherited retinal diseases masquerade as papilloedema and optic atrophy

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Background: Pseudo-papilloedema and pseudo-optic atrophy may rarely present as an inherited retinal disease. We report four cases that were initially referred with suspected neuro-ophthalmic pathology.

Case 1: A 43-year-old female presented with headache, moving rings in peripheral vision and "papilloedema" at age 31. MRV was normal. Opening pressure was 26 cmH 2 O. She returned with macular oedema at age 33 and received biologics for uveitis until age 40 when ffERG demonstrated a rod-cone dystrophy and a gene panel identified biallelic pathogenic USH2A variants.

Case 2: A 47-year-old female presented with headache, reduced peripheral vision and "papilloedema" at age 38. Opening pressure was 20 cmH 2 O and MRV was normal. Bilateral macular oedema at age 46 prompted further retinal investigations including ffERG which demonstrated a rod-cone dystrophy. A pathogenic PRPH2 variant was found.

Case 3: A 54-year-old Paralympic swimmer had bilateral visual loss from the age of 5 and was diagnosed with "Leber hereditary optic neuropathy". His brother had "optic atrophy". VA was 6/60 and 6/38. Discs were pale and global RNFL was 68-70 microns. Retinal exam and imaging were unremarkable. VEP showed delayed and reduced P100 on a background of an undetectable PERG and electronegative ffERG. A splice CACNA1F variant was found.

Case 4: A 30-year-old male with history of strabismus surgery, myopia and nystagmus presented with "early-onset optic atrophy". His brother was diagnosed with a "cone-rod dystrophy". VA was 6/19 R and 6/28 L. Discs were bilaterally pale and global RNFL was 64 microns. Foveal hypoplasia was noted and the ffERG was electronegative. A gene panel confirmed both brothers carry a frameshifting variant in CACNA1F leading to iCSNB. Conclusions: Neuro-ophthalmic symptoms should be assessed in the context of retinal structure and function. A ffERG may be informative to exclude retinal causes of pseudo- papilloedema and pseudo-optic atrophy when retinal structures appear normal.

Beyond the disc elevation

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A 16 year old male was referred by an optometrist for neuro-ophthalmology review with concerns of bilateral optic nerve elevation. The patient went for the optometry review to assess his visual driving eligibility and he denied any significant visual concern apart from 1 episode of suspected transient obscuration of vision. He denied history of tinnitus. He was not on any systemic medications and in particular no prednisolone, minocycline or Vitamin A derivatives. Past history include obesity with a body mass index of 37, delayed speech and hearing impairment. Family history include two sisters having eye and hearing issues.

On examination, his best corrected vision was 6/7.5 in both eyes. Pupillary reactions were normal and brisk with no relative afferent pupillary defect. Fundus examination was suggestive of small crowded optic disc bilaterally. Humphrey visual field showed generalised field constriction in the right eye and superior altitudinal visual field defect in the left eye.

The patient underwent MRI with MRV scan. MRI was normal but MRV showed bilateral venous sinus stenosis but no thrombosis. Lumbar puncture was performed by a paediatric neurologist and the initial lumbar puncture opening pressure was 21cm. A second neurology opinion and lumbar puncture showed an opening pressure of 26cm water. CSF analysis was within normal limits. The patient was treated as Idiopathic Intracranial Hypertension (IIH).

The disc appearance did not change with standard IIH treatment.

Subsequent work up showed peripheral fundus changes with pigmentary retinopathy change with the disc elevation. These changes are also seen his two sisters who have hearing and vision issues.

A diagnostic test was performed.

The patient and the family underwent genetic testing and the entire family was diagnosed with Usher Syndrome.

If it's not papilledema, then what could it be?

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Aim: To highlight ocular conditions that can present with disc oedema that could be mistaken as papilledema.

Methods: Patients with retinal dystrophies can present with disc oedema as one of their clinical signs. Here we present a cases series of such patients, where the disc swelling may be incorrectly diagnosed as due to raised intracranial pressure.

Results: Mer Tyrosine Kinase (MERTK) is a gene member of MER/AZL/Tyro3 receptor kinase family which has an essential role in phagocytosis at the level of the retinal pigment epithelium. A genetic variation in this gene can lead to an early onset, rapidly progressive retinitis pigmentosa (RP). An 8-year-old patient from a cohort of patients with MERTK-RP was first reviewed and worked up for disc swelling prior to any retinal signs or symptoms of nyctalopia. It wasn't until he was 16 years old that he reported nyctolopia and had electrophysiology studies diagnosing his retinal dystrophy. CRB1 (crumbs homolog 1) genetic sequence variations are associated with variable phenotypes of severe retinal dystrophies ranging from Leber Congenital Amaurosis (LCA) to retinitis pigmentosa. Specific retinal features such as preservation of the para-arteriolar retinal pigment epithelium, retinal telangiectasia with exudation, macular atrophy, nanophthalmos & optic disc drusen. CRB1-associated early onset severe retinal dystrophy has been seen to have an increase in retinal nerve fiber layer in follow up visits, indicative of disc odema.

Recently, independent discovery exome and genome sequencing in five unrelated families found a heterozygous missense variant in the alpha-kinase gene (ALPK1). All patient of the 5 families shared the ROSAH phenotype (retinal dystrophy, optic nerve edema, splenomegaly, anhidrosis and migraine headache) with low-grade ocular inflammation, pancytopenia, recurrent infections, and mild renal impairment in some.

Conclusion: We present three case reports of patients with retinal dystrophies of different genetic variability where optic disc swelling is one of their clinical signs.

Feeling the pressure in Alagille syndrome

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Alagille syndrome (arteriohepatic dysplasia) is a multisystem autosomal dominant disorder, dominated by the consequences of bile duct paucity and congenital heart disease. Variable penetrance means that presentations range from subclinical to life- threatening, with a mortality rate of up to 10%. The commonest ocular anomalies are posterior embryotoxon (95%), diffuse fundus hypopigmentation (57%) and mosaic iris stromal hypoplasia (45%). There is a characteristic facial appearance, and renal dysplasia may be seen.

Neuro-ophthalmic manifestations include optic disc changes (76%), cerebral arterial and venous abnormalities, and pseudotumour cerebri (PTC). Here we discuss the care of a lean young man with Alagille syndrome and extreme papilledema. He had challenging co-morbidities: an aortic valve replacement (AVR) after which he needed both warfarin and an iron transfusion; and chronic kidney disease (CKD) severe enough that transplantation work-up was underway.

Possible triggers for PTC in this patient included the syndrome itself and/or his low- iron state, as he was not taking any relevant medications. A cerebral venous catheter study did not demonstrate a stentable transverse sinus stenosis, or even significant venous hypertension. Hence stenting, the simplest option for a warfarinised patient, was not indicated.

Lumbar puncture (LP) was reluctantly sought, the CKD necessitating heparin infusion. LP did document elevated cerebrospinal fluid (CSF) pressure, confirmed normal CSF constituents, and provided temporising reduction in intracranial pressure. Shunting was deemed highly undesirable in a patient on warfarin, due to the revisions which nearly always follow. Bilateral optic nerve sheath fenestration under further heparin bridging was performed, and has succeeded in controlling papilledema for this very stoic patient.

The management team in this complex presentation included Neuro-ophthalmology, surgical Ophthalmology, Renal, Cardiology, Haematology and Neuro-radiologic Intervention. This highlights the dual role played by the multidisciplinary team – preserving vision, but also navigating safely through the minefield of the patient's co- morbidities.

Assessment of Spontaneous Venous Pulsations in Idiopathic Intracranial Hypertension: A Pilot Study

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Background: Spontaneous Venous Pulsations (SVPs) are rhythmic changes of the central retinal vein observed at the optic nerve head (ONH). Absence of SVPs have been suggested as a surrogate marker for assessment of elevated intracranial pressure (ICP). Using a handheld retinal imaging device, we assessed SVPs in a small cohort of Idiopathic Intracranial Hypertension (IIH) patients compared with ophthalmically normal controls.

Method: A total of 25 individuals were recruited (10 IIH (8 females) and 15 controls (12 females)). All participants had their pupils dilated and a 30 second recording of the ONH was captured using the Nun IR smartphone-based retinal imaging device (Odocs, New Zealand). Patient demographics including blood pressure, height, weight, and age was also documented. The equation developed and validated by Jonas et al for non-invasive ICP estimation was used to calculate ICP (ICP (mmHg) =(0.44×BMI)+(0.16×DBP) (0.18×age)–1.91).

Results: The mean age in the IIH and control group was 31±9 and 27±6 years. SVP was present in 11% and 100% of the IIH and controls, respectively. Estimated ICP was significantly higher in the IIH group compared with controls ($p<0.01$, 19.9±3.6 vs 14.3±3.1).

Conclusion: Using a handheld retinal imaging device we were able to visualise and screen SVPs for their absence or presence. The absence of SVPs could be a strong marker of elevated ICP, however, further studies in a larger cohort are required to develop an objective measure.

Optical Coherence Tomography Imaging Utility in Neuro-ophthalmology

Authors:
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There is a wide range of aetiologies that affect the optic nerves and anterior visual pathways including demyelinating, inflammatory, ischemic, traumatic, compressive, toxic, nutritional and hereditary causes. The optic nerve often shows structural changes alongside superficial and deeper retinal and choroidal vessels changes depending on the nature of the disease process. Optical Coherence Tomography Imaging has an expanding indication in neuro-ophthalmology playing an important role in the diagnosis and monitoring of many optic nerve-related diseases. There are a number of different devices that provide both quantitative and qualitative information, however not all the measures are interchangeable. As most proprietary platforms were designed for retinal diseases, there needs to be particular attention paid to segmentation and understanding of artefact is also important. As the technical ability of Optical Coherence Tomography continues to evolve, it is clear that the use for neuro-ophthalmology diseases will continue to grow.

Learning objectives

1. Develop a systematic and organised approach to the interpretation of different Optical Coherence Tomography imaging protocols

2. Identify common disease patterns using Optical Coherence Tomography.

3. Understand the importance identifying artefact and segmentation error, and correcting error.

4. Recognize the role of Optical Coherence Tomography in driving clinical decision making in neuro-ophthalmology.

9.30
Cranial Chaos

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A previously well 62-year-old truck driver was referred for an inpatient workup due to a six-month history of constitutional symptoms, dysphonia, and dysphagia. Six months prior, he experienced painless loss of vision in the right eye, progressing to perception of light. He was diagnosed with posterior segment ischemic optic neuropathy by a general ophthalmologist. He underwent treatment with gradually tapering prednisolone, resulting in limited visual recovery. About a month before his hospital presentation, he developed dysphagia for both solids and liquids, anorexia, hoarseness in his voice, weight loss, night sweats, and mild headaches.

9.45
Just Another IIH?

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A 38-year-old female was referred from her regional ophthalmologist for assessment of asymmetric disc swelling. She reports a history transient visual obscuration that began in her early teens whenever there was a postural change from lying to standing. This symptom completely dissipated but re-emerged over the past year in the right eye only. She reports a persistent mottled blur in her right inferior field that had been present for two years.

She reports a ten-year history of pulsatile tinnitus and migraines with visual aura three to four times per year. She gained 30kg over the preceding 10 years, however in the past 18 months was taking liraglutide with weight loss of 14 kilograms. Of note, her visual symptoms coincided with the history of 14 kg of weight loss in the past year. She reports no change in appetite, no night sweats or fevers and no exposure to tetracyclines.

She works as a sale assistant, is an ex-smoker with a 3-pack year history, no significant alcohol intake and lives with her husband. She has no significant family history.

10.00
"To be determined"

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A 34-year-old male originally from Somalia presented with 5 days of severe left retroorbital headache, left facial pain, reduced vision os, and horizontal binocular diplopia. Best corrected visual acuity was 6/6 od and 6/7.5 os. Colour plates were 14/14 od, 1/14 os, and he had a left RAPD. Fundoscopy was normal ou. Exophthalmometry measured 18mm od and 20mm os. He had a left 6th nerve palsy and reduced sensation over the left ophthalmic (V1) and maxillary (V2) trigeminal nerve sensory distributions.

10:15
More than meets the eye

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Purpose: To describe ocular imaging and electrophysiology in a patient with visual loss and gait disturbance.

Case: A 45-year-old female presents with a 1-year history of impaired distance vision and a 3-year history of impaired colour vision. Ten years prior, she was diagnosed with ovarian cancer during a routine laparoscopic investigation for polycystic ovarian syndrome. This was treated by intrauterine chemotherapy and hystero-oophorectomy. There were no regular medications and no known family history of eye disease. Her father passed away at 49 years of age. Systems review revealed poor balance for 5 years duration with increasing clumsiness.

She was unable to stand from a motorized wheelchair. Upper limb intention tremor and dysdiadochokinesis were present. She had limited up gaze, adduction deficit and hypometric saccades. Pupils were equal and reactive. Visual acuity was 6/30 R and 6/24 L. Intraocular pressures were 15 R and 11 L. Anterior segments were unremarkable. Fundoscopy revealed peripapillary atrophy around right optic disc. Fundus autofluorescence showed a hyper-autofluorescent ring nasal to the optic disc in the right eye. Optical coherence tomography (OCT) demonstrated foveal attenuation of the ellipsoid zone bilaterally as well as thinning of the inner retinal layers. There were no signs of retinal nerve fiber layer loss on OCT.

MRI showed olivopontocerebellar atrophy. Electrophysiology demonstrated generalized cone and macular dysfunction.

11.20
It's not all about the past

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Presenter:
Jordan Ng

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A 64-year-old woman who presented with 3-month onset of horizontal binocular diplopia on her left gaze with associated retro-orbital pain. Past medical history included 2 transcranial surgeries for a pituitary macroadenoma. There were no antecedent events apart from a recent recurrence of headache waking her from sleep.

The GP was concerned about recurrence of her pituitary macroadenoma. a plain CT scan that showed expanded pituitary fossa with empty sellar appearance but no tumour recurrence.

Her examination revealed best corrected visual acuity of 6/6 OU. There was also subtle left RAPD and visual field defect showed a left superior arcuate scotoma. The optic nerve examination showed temporal pallor, and this correlated with OCT findings of left inferior temporal nerve fibre layer thinning and ganglion cell dropout.

Motility examination showed an esotropia of 6 diopter prisms that increased to 10 diopter prisms on left gaze and orthotropic on right gaze which correlates with a lateral 6th nerve palsy.

An investigation is performed…..

11:35
'Germ warfare'.

Authors:
Blake D Colman[1,2], Tracie Tan[1,2], Robb Wesselingh[1,2], Subahari Raviskanthan[1], Catriona McLean[3], Anneke van der Walt[1,2].

Presenter:
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A 26-year-old previously well male presented with a two-month history of headaches, diplopia and sensory changes. He reported a gradual onset of left sided retro-orbital headaches and pressure associated with nausea prior to the development of horizontal binocular diplopia and facial parasthesias. Examination revealed a left cranial nerve VI palsy with left V1/2 distribution anaesthesia. Pulsed intravenous methylprednisolone was administered followed by an oral prednisolone taper, resulting in significant symptomatic improvement, however multiple attempts to wean prednisolone led to relapsing symptoms. He was referred to our institution 8 months after symptom onset where additional investigations were performed.

11:50
Blurred Vision? What a headache

Authors:
Dr Gurfarmaan (Garry) Singh, Dr Ayub Qassim, Dr Jude Fitzgerald, A/Prof. Sudha Cugati, Dr Devaraj Supramaniam

Presenter:
Dr Gurfarmaan Singh

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A 61-year-old male with a background of Non-Hodgkin Lymphoma in remission was referred to eye clinic for sudden onset blurred vision. This was associated with worsening headaches, tinnitus, dizziness and nausea. Four days following his initial presentation, he re-presented to clinic, complaining of bilateral eye pain.

12.05
How many?

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Pontomedullary cavernoma resection April 2021 resulted in Bilateral R>>L 6 th nerve palsies, R INO, ?L INO or ?R horizontal gaze palsy Plus R 7 th and 8 th nerve palsies

Sep 2021 – RMR Botox - no improvement

Jan 2022 – RMR recession 5.5mm and SR to LR transposition – no improvement

Initially wanted no further surgery due to lack of effect of previous surgeries. Now open to re-consider.

12.20
Metastatic vision loss

Authors:
Mullany, S., Berry, E.C., Slattery, J., Klebe, S., Siggs, O.M., Craig, J.E., Wechelekar, M.D., Chen, C.S.

Presenter:
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Case Details: An 82-year-old male of European ancestry undergoing immunotherapy with atezolizumab for metastatic non-small cell lung cancer presented with sudden onset unilateral vision loss, headache, and jaw claudication. He was presumptively diagnosed with central retinal artery occlusion and was commenced on high-dose intravenous methylprednisolone. A temporal artery biopsy was performed.

12:35
It is all in the eyes…

Authors:
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A 53-year-old female smoker presented with 5 week history of progressive vertigo, intractable nausea, vomiting, hiccups and 10kg weight loss. One week after onset of these symptoms, she developed horizontal binocular diplopia and ataxia. One week later she underwent an elective salpino-oopherectomy for ovarian cyst. Her symptoms persisted and she further developed oscillopsia and worsening of vomiting and hiccups. Examination revealed ocular flutter and an ataxic gait.

Vascular supply of the optic nerve head: implications for optic disc ischemia

Author:
Prof. Anthony Arnold

The vascular supply of the optic nerve head is complex and remains incompletely delineated. Over the past 50 years, investigators have attempted to clarify the relative contributions of the choroid, the short posterior ciliary arteries and the central retinal artery to the vascular beds of the inner retinal, prelaminar, laminar and retrolaminar segments of the nerve head. Various techniques have been utilized to study this microcirculation in normals and in various forms of optic disc edema, including ischemia. This session will review findings from most recent techniques, including swept source OCT angiography, and will suggest implications for the pathogenesis of ischemic optic neuropathy.

OCT angiography changes in acute NAION presentations: A case series and protocol to investigate

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Introduction: Non-arteritic Ischaemic Optic neuropathy (NAION) is a form of optic neuropathy that commonly affects individuals over the age of 50 and its annual incidence has been reported to be between 2.3-10.3 people per 100,000. The diagnosis of NAION is important to institute secondary prevention to prevent another end-organ ischemic attack. If untreated, approximately 15% of patients have contralateral eye involvement, resulting in bilateral visual loss. The pathogenesis of NAION is not fully understood and it is thought to be secondary to occlusive ischaemia of the short posterior ciliary arteries that supply the optic disc. OCT- Angiogram may help to determine the site of vaso-occlusive disease. Recent studies have demonstrated a reduction blood flow through the retinal peripapillary capillaries and peripapillary choriocapillaris in NAION.

Methods: Presentation of case series and proposal for a prospective longitudinal study.

Results: Described herein two patients with acute NAION within 7 days of symptom onset. The OCT-A showed demonstrated dilation of vessels at the disc. The OCT-A Retina Depth encoded layer showed associated superior hyperemic segments that correlated with the corresponding inferior altitudinal visual field changes.

Discussion: Based on these cases, evaluation of the peripapillary microvasculature may help to diagnose NAION in patients. Correlation of the structural changes and nerve function will help provide prognosticators for NAION. We present a protocol to assess the perfusion density of the superficial and deep capillary plexuses and measure the diameter of the central retinal artery using OCT-A to evaluate this.

Clinical Characteristics of Patients with Carotid-Cavernous Fistula (CCF) at a Tertiary Hospital

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Presenter:
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Introduction: A Carotid-cavernous fistula (CCF) is an abnormal arteriovenous communication between the cavernous sinus and the internal carotid artery (ICA) and or external carotid artery (ECA). Clinical manifestations are varied among CCF types. The diagnosis of CCF is made with clinical manifestation and neuroimaging. Clinical characteristics of patients affect the therapeutic decision.

Purpose: To describe the characteristics of patients with CCF at at a Tertiary Hospital.

Methods: This was a descriptive study conducted retrospectively. Data was gathered from medical records between January 2019 and December 2022 at National Eye Center Cicendo Eye Hospital. The subjects were patients who have been diagnosed with CCF. The patient's characteristics and clinical manifestations were recorded.

Results: A total of 79 patients (98 eyes) were included in this study. The subjects were mainly female (50.6%), The mean age was 41.12 ± 16.13 years old. History of trauma and systemic disease had been identified in 59.5% and 45.6% of patients respectively. Unilateral involvement occurred in 75.9% of eyes. Patients with onset of symptoms more than a month were 62.82%. 78.5% of eyes had best corrected visual acuity ≥6/60. The intraocular pressure was increased in 55.1% of CCF eyes. The types of CCF were Type A (33.7%), Type B (16.3%), Type C (45.9%), and Type D (4.1%) consecutively. The most common clinical manifestations were episcleral injection (100%), ophthalmoplegia (90.8%), proptosis (62.2%), and reduced visual acuity (53%).

Conclusion: Most CCF patients were dominated with female and patients in the age range of 20-59 years old category. A history of trauma was identified in most cases. CCF low flow type and Barrow's type C were the majority of CCF types. The three most common clinical manifestations were episcleral injection, ophthalmoplegia, and proptosis.

Keywords: Carotid-Cavernous Fistula, Barrow classification, Ophthalmoplegia, Episcleral Injection

Preliminary Data from 6 year follow up of the impact of continuous positive airway pressure treatment on cardiovascular and cerebrovascular health outcomes in obstructive sleep apnea

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Introduction: Obstructive sleep apnea (OSA) is linked to cardiovascular and cerebrovascular disease. Ocular manifestations of OSA include ischemic optic neuropathy, worsening of diabetic retinopathy and retinal vein occlusion. We have previously found that OSA was associated with quantifiable retinal vascular changes, with OSA disease severity at diagnosis independently associated with retinal arteriolar narrowing and reduced pulse amplitude. At the 2-year follow up, we demonstrated that there was progressive retinal arterial narrowing in severe untreated OSA whilst continuous positive airway pressure (CPAP) therapy appeared to attenuate retinal arterial narrowing. Here we present preliminary data from the 6-year follow-up.

Methods: 70 patients who previously participated in baseline and 2-year follow-up assessments will be invited to undergo a 6-year ophthalmic review including updated medical history, OCT, OCTA, static and dynamic vessel imaging. Central Retinal Arteriolar Equivalent, Central Vein Arteriolar Equivalent and Arteriovenous Ratio (AVR) were calculated using Visualis V 2.81 (Imedos).

Results: To date, 53 patients have been contacted with 27 patients completing review. Health outcomes are summarized in Table 1.

TABLE 1.

OSA severity/CPAP Usage	MI/ Angina	Hypertension	Hyperlipidemia	Diabetes	Peripheral Vascular Disease	Stroke/ TIA
Severe OSA+CPAP (n=5)	0	2	4	1	0	0
Severe OSA+No CPAP (n=7)	1	6	5	0	0	1
Mild/Moderate OSA + CPAP (n=2)	1	2	1	1	1	0
Mild/Moderate OSA + No CPAP (n=8)	2	6	4	1	2	1
Control (n=5)	0	2	2	2	2	0

No significant difference in mean CRAE, CRVE, or AVR was observed for individuals Mild/Moderate, Severe OSA with or without CPAP usage. Mean RNFL thickness in individuals with Severe OSA + CPAP (81.0µm ± 8.9) and Mild/Moderate OSA + No CPAP (73.9µm ± 13.5) was significantly thicker compared to the Control Nasally.

Conclusion: Based on the results so far it is unclear if CPAP provides a long-term protective benefit for cardiovascular and cerebrovascular events, however there is a trend for those not on CPAP to have higher rates of hypertension and stroke. The protective effect of CPAP on retinal vascular changes seen at the 2-year mark have not yet been seen in this 6-year review. Supported by RESMED

Why the ipsilateral pupil was irregularly dilated five days following PERFECT pterygium surgery…!

Authors:
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Presenter:
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An otherwise well 49-year-old man underwent uneventful right nasal PERFECT (Pterygium Extended Removal Followed by Extended Conjunctival Transplant) pterygium surgery. This surgical technique, advocated by Professor Lawrence W. Hirst, generally provides good surgical results, as well as good cosmesis and minimal pterygium recurrence. Five days postoperatively, the patient noticed an ipsilateral, dilated, irregular pupil. Considering that PERFECT pterygium surgery involves fairly posterior soft tissue orbital dissection, the patient was referred to the corresponding author for diagnosis and management.

Examination disclosed light-near dissociation of both pupils. On slit lamp examination, bilateral tonic re-dilatation was noted. The cranial nerves were intact. The surgical site appeared satisfactory.

Electronic Pupillometry (NPi-200 ®) disclosed pupil diameters of right 4.69mm and left 3.56mm. Following bilateral instillation of dilute 0.1% pilocarpine, the pupils measured right 4.09mm, left 2.48mm, a percentage reduction in pupil diameter of right and left 12.8% and 30.3% respectively, and a percentage reduction of pupil area of 13.1% and 51.5% respectively.

The patient's knee jerks and ankle jerks were examined and found to be absent bilaterally.

A diagnosis of coincidental bilateral Adie's Syndrome was made, readily explaining the patient's early postoperative dilated pupil.

The patient referred himself to another Neuro-ophthalmologist, who was unable to make a definitive diagnosis, but referred the patient for an MRI- brain, which was normal.

Initial management consisted of patient reassurance.

The appearance of this patient with his early-onset right and longstanding-left bilateral Adie's Syndrome pupils was confirmed not only by the Electronic Pupillometry, but by the typical light-near pupillary dissociation.

Even in surgery as relatively non-invasive as pterygium surgery, and therefore applicable to all ophthalmic surgery, each surgical patient deserves thorough preoperative examination. This examination includes corrected distance visual acuity, pupil reactions, and visual fields to confrontation, as well as detailed assessment of the patients' cranial nerves functions.

Giant Cell Arteritis

Author:
Prof. Susie Mollan

Giant Cell Arteritis is well known to be a critical ischaemic disease that requires immediate medical recognition to initiate treatment and where one in five people still suffer permanent visual loss. Recent national and international guidelines have supported the directed use of cranial ultrasound to reduce diagnostic delay and improve clinical outcomes. Immediate high dose glucocorticoids remain the standard emergency treatment for suspected Giant Cell Arteritis. As the immunopathophysiology has continued to be characterised, the opportunity for targeted treatment has become a reality for GCA. There are now a number of agents that have been shown in clinical trials to have superior clinical efficacy and steroid sparing effects. These advances have influenced a change to routine clinical practice.

Learning objectives
1. Recognise the clinical presentation of Giant Cell Arteritis.

2. Understand the importance fast-track patient referral pathway and the utility of cranial ultrasound in the diagnosis of Giant Cell Arteritis.

3. Be familiar with the interactions between the multi-disciplinary team members.

4. Discuss management options for new onset and relapsing Giant Cell Arteritis.

5. Consider the challenges in your clinical practice to change of routine care in light of international guidelines.

The Temporal Arteritis Proforma Revisited- 2023

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Temporal Arteritis (TA), aka Giant Cell Arteritis (GCA), is a potentially sinister illness. Failure to diagnose TA, and to treat the patient in a timely and appropriate fashion, may result in rapid and irreversible blindness, and death. TA has protean symptoms and signs, many of which are nonspecific, as they are seen in many illnesses. Clinicians must consider the diagnosis of TA especially when an elderly female presents on a Friday afternoon with new headache, amaurosis fugax or visual loss, jaw claudication and diplopia.

The Temporal Arteritis Proforma (TAP), first published in 2021, comprised 51 symptoms and signs. It has been distributed to and utilised in hospitals and clinics in Australia. The TAP has received favourable feedback from clinicians dealing with suspected TA. The TAP was distributed to NOSA delegates in 2021. The TAP was designed to be placed on the appropriate noticeboard or wall in Consulting rooms, Emergency departments and in hospital Outpatient clinics. Its aim was to make the numerous symptoms and signs of TA more easily remembered and utilised in formulating the diagnosis of TA. The TAP was therefore designed to improve the clinician's ability to diagnose TA by increasing his or her index of suspicion of its presence, especially as its presentation may vary considerably.

In earlier iterations of the TAP, the symptoms and signs were, as with the current TAP proforma, categorised into clinically appropriate sections. While the signs and symptoms of TA are not weighted for importance in the TAP, they are all relevant in assisting the diagnosis of TA.

In the most recent iteration of the TAP, numerous changes have been made, including the addition of Paracentral acute middle maculopathy as a sign of TA. The current TAP is available for delegates at the 2023 NOSA meeting.

FESStival of Errors

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In 2021, a 37 year old male presented with a superior field loss in his right eye noted just after having had FESS surgery in India a few weeks before. There was clinical evidence of a traumatic optic neuropathy. 3 further cases of FESS misadventures into the orbit but ruining extra-ocular muscles are presented briefly.

Streptococcal Meningoencephalitis with WEBINO, bilateral blindness, pupil-involving third nerve palsies, right facial palsy, and upper motor neurone signs.

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A 49-year-old Australian-Vietnamese male, previously healthy and an IT consultant, presented to a Tertiary Referral Teaching Hospital with a sudden severe headache, fevers, and neck stiffness. After lumbar puncture and neuroradiological assessment, he was diagnosed with Streptococcal meningoencephalitis. The patient was treated with high dose intravenous Benzylpenicillin. Gross cerebral oedema and purulent meningitis necessitated a decompressive craniectomy with an External Ventricular Drain (EVD). Four weeks later, he was later referred to Northern Beaches Hospital for Neuroophthalmological evaluation and systemic assessment.

The patient developed bilateral blindness as a result of Streptococcal meningoencephalitis, with vision ranging between no light perception, vague hand movements, and occasionally counting fingers. His visual fields were very limited. Wall-Eye Bilateral Internuclear Ophthalmoplegia (WEBINO) was observed during ocular rotations testing, with both eyes abducted in the primary position. Saccadic eye movement testing was challenging due to the patient's inability to fixate on targets, displaying slow adducting saccades. Bilateral abducting nystagmus of the contralateral abducting eye was present, consistent with WEBINO. Bilateral pupil- involving third nerve palsies were also noted, with fixed and widely dilated pupils. Doll's Eye testing demonstrated absent vertical gaze, assisting in evaluating slow adducting saccades and abducting nystagmus of the opposite eye. However, no papilloedema was detected, and a normal fundoscopy examination was noted.

Moderate bilateral deafness was present, but other Cranial Nerves, including corneal sensation, were normal. Upper motor neurone signs were observed, such as bilateral upper and lower limb hyperreflexia, and extensor plantar responses. A repeat MRI revealed widespread intracranial changes consistent with meningoencephalitis, with nonspecific changes in the midbrain and pons. Considering that the patient's brain was sterilized by penicillin, systemic intravenous steroids were introduced as a safe attempt to reverse the inflammatory response caused by the Streptococcal infection. Unfortunately, no significant improvement was noted. Despite being legally blind, rehabilitation was initiated.

A traumatised nerve

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A 30yo otherwise healthy male presented to the Emergency Department with pain and swelling in the left eye (OS) after being elbowed to the eye during sport 24 hours previously. On examination, visual acuity (VA) was 6/6 OD, 6/18 OS. He had bullous sub-conjunctival haemorrhage, mydriasis, and features of commotio retinae on fundoscopic examination.

Computerised tomography (CT) of the brain and orbits revealed a moderately displaced comminuted medial orbital wall fracture and a large lesion 2.2 x 2.3 x 2.4cm causing superomedial displacement of the left optic nerve and proptosis, consistent with intraconal haematoma. He underwent emergency canthotomy and cantholysis and was discharged with a plan for review in 1 week with repeat neuroimaging.

He represented 5 days later with recurrent chemosis, blurred vision and binocular horizontal diplopia. VA was 6/6 OD, 6/18 OS, with a RAPD OS, -1 abduction OS (presumed secondary to chemosis), 3-4mm lagophthalmos OS. Repeat CT demonstrated stable haematoma size. He was treated with 1g intravenous methylprednisolone, and discharged on topical steroids, lubricants, and antibiotics. Subsequent clinic visits revealed worsening VA to 6/60 pinholing to 6/18 OS and intraocular pressure up to 38mmHg, thought secondary to topical steroids, but subsequent neuroimaging was stable.

Repeat imaging 1 month post injury showed a persistent retrobulbar mass. Neuro- ophthalmology review noted VA 6/6 OD, 6/9 OS, RAPD OS. Humphrey visual fields suggested central scotoma. OCT RNFL thickness was 101 OD, 138 OS, with normal GCL thickness OU. suggestive of compressive neuropathy rather than traumatic aetiology.

MRI revealed a left intraconal T2 heterogeneous, partly contrast enhancing mass measuring 2.6 x 2.6 x 2.1cm with evidence of chronic bony remodeling displacing the optic nerve.

Tissue biopsy was S100 and SOX-10 positive, consistent with a schwannoma.

Evaluating immune-mediated contributors to acute uveitis

Authors:
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Background/Introduction: The aetiology of uveitis may be varied. Up to a third of all cases remain undifferentiated without a clear aetiology, which renders management challenging. We aim to identify clinical and immunophenotypic profiles which may be most consistent with an immune-mediated versus infectious aetiology, in order to improve characterisation of patients who are currently undifferentiated.

Methodology: We prospectively recruited all patients who presented to two tertiary uveitis clinics within 90 days of symptom onset. We performed clinical phenotyping, and collected serum and whole blood for immunophenotyping and genotyping.

Results: 65 patients are recruited (36 females,55%); median age 43y (range 16-79). 38/65 (59%) presented with their first episode of uveitis, and 27/65 (41%) as a relapse. 25/65 (38%) affected bilateral eyes versus 34/65 (52%) affecting one eye. Optic disc swelling was present in 13/65 (20%), and concurrent optic neuritis in 10/65 (15%). Patients were classified as having panuveitis in 16/65(28%); and isolated anterior (18/65, 32%), intermediate (4/65, 7%) or posterior (10/65,16%) uveitis. The ultimate diagnosis was infectious (16/65, 25%), immune (24/65, 37%) or undifferentiated (20/65, 31%). Median duration of follow-up was 12.3 months (range 3-24). Median visual acuity (of a total 88 eyes assessed) at disease nadir was 6/12(range 6/4-6/60) in the infectious group, 6/12(range 6/5-6/60) in the immune group, and 6/9(range 6/5-6/24) in the undifferentiated group (0.027 value; using ANCOVA). Median RNFL at latest follow up was 101 um (range 80-174) in the infectious group, 99 um (range 69-347) in the immune group, and 113um (range 46-296) in the undifferentiated group (p=0.33).

Conclusions/Future Directions: In this cohort of prospectively recruited patients, we are currently undertakingdetailed immunophenotyping including serological, antibody, and cytokine profiles to identify optimal differentiators of infectious vs immune-mediated causes of uveitis in a predictive model. We will utilise this model to characterise patients who are currently classified as 'undifferentiated' uveitis, in order to provide therapeutic and prognostic guidance.

Title	Presenter
Longitudinal Idiopathic Intracranial Hypertension visual outcomes in an Australian population.	Blake D Colman
Bilateral Atypical Optic Neuritis In An HIV Patient: A Case Report	Ni Luh Ayu Darmayanti
Neuro-Ophthalmology Manifestation in Young Patient with Primary Angiitis of Central Nervous System	Aulia Giffarinnisa
The effect of memantine administration as n-methyl d-aspartate receptor antagonist on retinal ganglion cellular atp level among mice models with methanol-induced toxic optic neuropathy	Antonia Kartika Indriati
Prognostic factors of unresponsive visual acuity after intravenous pulse methylprednisolone (IVMP) of first episode optic neuritis of Neuromyelitis optica spectrum disorders (NMOSD) patients	Pichaya Kulniwatcharoen
The WHY, The WHERE, The WHEN and The HOW of testing Visual Fields To Confrontation (FTC)…	Matthew K. Lee
Idiopathic Intracranial Hypertension presenting as Acute Acquired Comitant Esotropia	Lakshmi S. V. Mallapragada
Concurrent bilateral Optic Nerve Sheath Meningioma and Idiopathic Intracranial Hypertension: Occam's razor or Hickam's dictum?	Niamh Moloney
SD-OCT imaging of macular changes in Fabry Disease.	Stephanie S. Moon

Title	Presenter
"HaNDL" with positive Anti- MOG antibody: The Expanding Spectrum of anti-MOG associated disorders.	Verity Moynihan
The Efficacy, Adverse Effects and Economic Implications of Oral versus Intravenous Methylprednisolone for the Treatment of Optic Neuritis: A Systematic Review	James Pietris
Immune Checkpoint Inhibitors and Optic Neuropathy: A Systematic Review	James Pietris
Junctional Scotoma With Mass In Sellar - Para sellar Tursica: A Case Report	Pradistya Syifa Yudiasari
Journey through the Eye	Parthvi Ravat
Neuro ophthalmology education – what is the role for social media?	Elle Nguyen
Adult-onset Acute Necrotising Encephalitis with Bilateral Incongruent Homonymous Hemianopia	Jessica Redmond
Optic Nerve Swelling related to Infective Endocarditis	Jessica Redmond
Visual Function Recovery in Patient with Ethambutol-Induced Toxic Optic Neuropathy	Prettyla Yollamanda

Longitudinal Idiopathic Intracranial Hypertension visual outcomes in an Australian population.

Authors:
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Idiopathic intracranial hypertension (IIH) is a condition of rising prevalence and significant morbidity, typically affecting overweight women of reproductive age. There is a paucity of data regarding longitudinal clinical outcomes in Australia. We aimed to describe longitudinal clinical and paraclinical changes in a tertiary cohort of IIH patients.

A retrospective analysis was performed on adult patients diagnosed with definite IIH (Friedman criteria), prospectively enrolled in the neuro-ophthalmology database (NODE) at a single tertiary centre in Victoria. Demographic data were obtained at baseline with clinical evaluation, automated perimetry and optical coherence tomography at all sequential visits. Multivariate statistical analysis was performed in R.

116 patients were included; 93.1% were female. Mean (+/- standard deviation) time from first consultation to diagnosis was 6.72 (43.16) days, with an average follow-up duration of 352.7 days (range 0 – 1232) over 4.73 visits. Mean age at diagnosis was 28.8 (+/-6.8) years with a mean body mass index (kg/m2) of 39.1 (+/- 9.7). Papilloedema was found in 96.5%, mean Frisen grade of 1.96 (0.98). Mean CSF opening pressure was 31.29 (+/- 4.90) cmH20. No change in visual acuity was observed over time (mean LogMAR 0.02 in the right eye, 0.05 in the left). Time was associated with a reduction in retinal nerve fibre layer thickness ($p = 0.02$) and papilloedema grade ($p < 0.001$). BMI at time of diagnosis strongly correlated with mean perimetric mean deviation where a one-unit increase in BMI was associated with a 0.10 decrease in perimetric mean deviation ($p = 0.01$).

The demographic and clinical phenotype in our study are comparable with international cohorts. The main predictor of worse visual outcome was baseline BMI, providing a strong rationale for focused intervention on weight loss.

Bilateral Atypical Optic Neuritis In An HIV Patient: A Case Report

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Introduction: There are cases of bilateral Optic neuritis in HIV patients with caused by secondary infections including Syphilis, Toxoplasmosis, and CMV. This case also confirms that the awareness of HIV spread should be improved.

Case Presentation: A 26-year-old male who came with blurred vision in his left eye a month ago. Funduscopy examination showed that the margin of the optic disc on both eyes was blurred and hyperemic. 1 g of methylprednisolone was administered intravenously for three days before being tapered off. In the second week of follow-up, his visual acuity declined to 20/60 on the right and 20/400 on the left, and his fundus appearance was also worsening. We considered other causes for this case, so we conducted further examination. This patient was referred to the internal and dermatovenerology department. Serologic tests revealed a positive VDRL, TPHA, Anti-Toxo IgG, Anti-CMV IgG and human immunodeficiency virus antibody test. Visual acuity and color vision improved after one month of HIV treatment with Dolutegravir sodium, lamivudine, tenofovir, and cotrimoxazole. Syphilis treatment with Benzatin penicillin injection 2,4 unit IM single dose. He was given Pyrimetamine and Valgancyclovir for Toxoplasmosis and CMV management. Discussion: The diagnosis of the Atypical type of optic neuritis for this case is based on the unusual feature of the patient including male gender, bilateral involvement of the eyes and systemic association with HIV. Treatment for optic neuritis in HIV patients is given based on specific etiologies.

Conclusion: Patients with Atypical Optic Neuritis may be suspected to have HIV or other opportunistic infections. There was a considerable improvement in visual acuity with enhanced color vision after HIV medication. Bilateral atypical optic neuritis has a favorable visual prognosis, and it is vital to treat the underlying disease.

Keywords: Optic Neuritis, HIV, Syphilis, Toxoplasmosis, CMV

Neuro-Ophthalmology Manifestation in Young Patient with Primary Angiitis of Central Nervous System

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Introduction: Primary Angiitis of the Central Nervous System (PACNS) represents a rare inflammatory disease affecting the brain and spinal cord. PACNS associated ophthalmic findings, including vision loss is rare and may lead to irreversible blindness.

Case Description: An 18-year-old female was referred from neurology department with blurred vision in both eyes since a week before and worsened for the last three days. There was a history of right hemiparesis four months prior. Brain Magnetic Resonance Imaging (MRI) and Digital Subtraction Angiography (DSA) were performed, and the patient was diagnosed with PACNS by the neurologist. The hemiparesis improved after the second DSA, one month before the ocular manifestations occurred. The patient's visual acuity in the right eye was no light perception, and the left eye was 20/50. Relative afferent pupillary defect was present in the right eye. Funduscopy showed blurred margin of the optic nerve head with undetermined cup-disc ratio in the right eye and was diagnosed as an optic neuritis.

Conclusion: PACNS is a rare and severe disease whereas neuro-ophthalmic manifestation reports are limited to this date. Therefore, ophthalmologist and neurologist should have good collaboration to consider the importance of ophthalmologic examination of any ocular manifestation of patients suspected having cerebral vasculitis. To these days there still no proven treatment for atypical optic neuritis including due to PACNS.

Keywords: Primary angiitis of the central nervous system, ophthalmic involvement, visual loss.

The effect of memantine administration as n-methyl d-aspartate receptor antagonist on retinal ganglion cellular atp level among mice models with methanol-induced toxic optic neuropathy

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Background: Methanol Optic Toxic Neuropathy (MOTN) remains a health problem in developing countries with no current definitive therapy. Formic acid accumulation causes mitochondrial damage which disturbs the synthesis of adenosine triphosphate (ATP), axoplasmic flow disturbance, and an increase in reactive oxygen species (ROS), which ends in the death of retinal ganglion cells (RGCs). Decreasing RGCs density in MOTN has similar pattern as optic neuropathic in glaucoma, ischemia, and trauma, through nerve cell excitotoxicity process via the excessive activity of N-Methyl D-Aspartate (NMDA) glutamate amino acid receptor. Memantine is a receptor antagonist which blocks the NMDA receptor channels. The inhibition of NMDA receptor activity by memantine is expected to lower intracellular calcium levels, delay apoptosis, increase cellular ATP levels, and prevent further mitochondrial damage. This study aims to study the effect of memantine on ATP levels in mice on RGCs with MOTN.

Methods: An experimental study is conducted on 28 mice models with MOTN which were divided into 4 intervention groups: 1 (MOTN; enucleation on the third day), 2 (MOTN + Memantine; enucleation on the third day), 3 (MOTN; enucleation on the seventh day), and 4 (MOTN + Memantine; enucleation on the seventh day). All groups were exposed to N2O:O2 gas and given methanol orally. The level of RGCs ATP was assessed immediately after enucleation.

Results: The average level of ATP (nmol/µl): group 1 = 0.689 ± 0.079; group 2 = 1.022 ± 0.124; group 3 = 0.903 ± 0.275; group 4 = 0.794 ± 0.385. ATP level on group 2 was significantly higher compared to group 1 (p-value = 0.019).

Conclusion: Memantine administration on MOTN mice models until the third day may help alleviate mitochondrial functional disturbance, which can be observed from the significantly higher ATP level in the group receiving memantine.

Keywords: Memantine, Methanol Optic Toxic Neuropathy, ATP, Excitotoxicity

Prognostic factors of unresponsive visual acuity after intravenous pulse methylprednisolone (IVMP) of first episode optic neuritis of Neuromyelitis optica spectrum disorders (NMOSD) patients

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Objective: To explore factors of unresponsive visual acuity in patients with first episode optic neuritis of Neuromyelitis optica spectrum disorders (NMOSD) to intravenous pulse methylprednisolone (IVMP) treatment.

Methods: We retrospective chart reviewed patients who presented with first episode optic neuritis related NMOSD and were treated with a daily dose of 1 gm of IVMP at Chiang Mai university hospital between 2009 and 2022. We investigated baseline characteristics such as age, sex and preexisting autoimmune disease, together with neuro-ophthalmic features including baseline visual acuity, bilaterality, ophthalmalgia, presenting of optic nerve head edema, simultaneous transverse myelitis, and other cerebral symptoms. Laboratory investigation for NMO antibody and neutrophil to lymphocyte ratio, also duration of symptoms and treatment were investigated. All these potential factors were explored to prognose immediate visual acuity response to IVMP. Both univariable odds ratio and multivariable logistic regression were performed. Results: There were 54 NMOSD patients with 74 eyes in this study. 51 patients (94.4%) were female. Mean age was 43.87 years old (SD ± 16.05 years). NMO antibody was positive in 38 (83.61%) among 48 blood-tested patients. 31.48% of patients accompanied with transverse myelitis and 16.67% had other cerebral symptoms. Baseline visual acuity was 2.3 LogMAR (IQR, 1.48-2.3). More than half of patients (55.56%) received IVMP for 5 days. 46.30% of patients showed no improvement in visual acuity after IVMP. The median of post-IVMP visual acuity was 1.8 LogMAR (IQR, 0.89-2.3) and mean improvement was 0.45 LogMAR. Among all potential factors, the interval before treatment exceeded 30 days resulted in an increased odds ratio of 4.77 (p value 0.05, 95%CI 1.21-18.78) for immediate unresponsive visual acuity in univariable analysis. Although with adjusted multivariable analysis, it became statistically insignificant.

Conclusion: Late IVMP treatment in first optic neuritis of NMOSD might influence the visual acuity responsiveness, which suggested the additive treatments.

The WHY, The WHERE, The WHEN and The HOW of testing Visual Fields To Confrontation (FTC)…

Authors:
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The WHY:
a. FTC is the single most powerful test in all of Ophthalmology, Neurology and Neuro-ophthalmology, omitted at the clinician's peril.
b. FTC enables diagnosis of 26 possible different lesions.
VA (#1)

Lower nasal defect, one eye: Open Angle Glaucoma (#2), Retinal detachment (#3), AVCIC (Anterior Visual pathway Compression by Internal Carotid artery and others) (#4), Optic nerve drusen (#5)
Lower temporal defect, one eye: Retinal detachment (#6) or Vascular occlusion (#7)
Central visual loss: High refractive errors (#8), AMD (Age-related Macular Degeneration) (#9), Severe Optic Neuritis (#10), Central Serous Chorioretinopathy (#11)
Bitemporal Hemianopia (BTH): Chiasmal region lesion (#12); Optic nerve drusen (#13)
HHA (Homonymous Hemianopia) Stroke or Tumour: Temporal (#14 and #15), Parietal (#16 and #17), Occipital (#18 and #19)

Diabetic HHA due to Hyperosmolar Non-ketotic Hyperglycaemia (HNKH) (#20)
Dyscalculia for finger counting - dominant Parietal lobe lesion (part of Gerstmann's Syndrome) (#21)
Misinterpretation of object of regard due to Parietal lobe lesion (#22) [Professor Oliver Sacks: 'The Man who mistook his wife for a hat']
Diagnosis of Latent Nystagmus (#23)
Non-organic visual loss (#24)
Visual field desaturation: aka - Sensory Suppression (#25)
Assessment of Visual Fields for Driving (e.g. Estermann) (#26)

The Where: In ED, in the clinic or at the bedside.

The WHEN: Always !!

The HOW: For two eyes, requires 13.6 seconds of the clinician's examination time.
The clinician should:
Keep both of his/her eyes open.
Use only 1, 2 or 5 fingers.
Not wave his/her hands around.
Not use a red hat pin.
Test quadrants.
Demonstrate vertical and horizontal neurological meridia.
Move from NON-SEEING TO SEEING FIELD.
Assess desaturation (sensory suppression).

Idiopathic Intracranial Hypertension presenting as Acute Acquired Comitant Esotropia

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Introduction: Acute Acquired Comitant Esotropia (AACE) is a rare subtype of esotropia characterised by sudden onset of comitant esotropia with diplopia.

Case Description: A 33-year-old obese woman presented with headache & sudden onset, uncrossed, horizontal, binocular diplopia in primary gaze.

Uncorrected visual activity of both eyes was 6/6. Anterior segments were normal. Bilaterally, grade 2 papilloedema was seen on fundus examination. Bilateral extra-ocular movements were full and free. HCRT revealed 15 degree esodeviation of right eye for near (33cm) and distance (6m) fixation. Alternate cover test revealed fast redressal of both eyes. Both eyes were neutralised with 30 prism diopters base-out prism for near and distance fixation. Neurological examination was normal.

MRI Brain with contrast revealed partial empty sella and venogram showed partially hypoplastic left transverse and sigmoid sinus. The patient declined lumbar puncture, but given the clinical signs, a diagnosis of idiopathic intracranial hypertension (IIH) was made.

The patient was advised to lose weight and was started on acetazolamide 500 mg TDS. On follow up, headache, diplopia, esodeviation and papilloedema had improved.

Discussion: AACE secondary to neurological causes is rare, occurring in approximately 6% of cases. Neurological causes of AACE include intracranial tumours, Arnold-Chiari Malformation, IIH and ocular myasthenia graves.

Diplopia in the context of IIH usually results from sixth nerve palsy due to raised pressure causing compression of the abducens nerve. However, given preserved extra ocular movements, abducens palsy is unlikely to be the case for this patient. It has been hypothesised that in neurological causes of AACE, dysfunction of the supranuclear mesencephalic structures that control vergence eye movement are affected, leading to the clinical syndrome.

Conclusion: AACE can rarely be caused by neurological conditions. Therefore, neurological examination and imaging must be performed in all cases.

Concurrent bilateral Optic Nerve Sheath Meningioma and Idiopathic Intracranial Hypertension: Occam's razor or Hickam's dictum?

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Case description:
45-year-old female (BMI of 29) presented with headaches, tinnitus, blurring of vision and bilateral optic disc swelling. Initial non-contrast MRI brain revealed features of raised intracranial pressure (ICP). Two lumbar punctures showed normal opening pressures with normal constituents. Repeat MRI brain with contrast revealed bilateral optic nerve sheath meningiomas (ONSM).

The optic disc swelling gradually worsened. Radiotherapy for the ONSM was considered to carry significant risks with minimal potential benefit. As the MRI features of raised ICP were quite prominent, in the context of worsening optic disc swelling with ganglion cell loss, venogram was obtained which revealed a superior sagittal sinus (SSS) pressure of 25mmhg, stenotic gradient across SSS and left transverse venous sinus of 10mm HG.

She was unable to tolerate acetazolamide. To reduce any 'additional' pressure being transmitted to the optic nerve from presumed locally raised ICP, after multidisciplinary discussion, she underwent transverse venous sinus stenting. She had a transient complication of a jugular foramen syndrome with neuropraxia of the left cranial nerves IX, X and X1. She was treated with steroids and vocal cord botulinum toxin injections with complete resolution of symptoms.

Post stenting OCT revealed significant improvement in bilateral disc swelling when compared to her OCT prior to the stenting, suggesting that at least part of the disc swelling was from increased CSF pressure on optic nerves.

Discussion:
This case posed several diagnostic and therapeutic challenges, including inadequate initial MRI scan and delayed presentation to Neuro-ophthalmology.

It was debated if the prominent radiologic signs of raised ICP represented poor venous return via the cavernous sinus from the bilateral ONSM or she had IIH. We will further discuss the postulated mechanism between ONSM and signs of locally raised ICP.

The case also highlights the rare but debilitating complication of jugular foramen syndrome secondary to venous sinus stenting.

SD-OCT imaging of macular changes in Fabry Disease.

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Fabry disease (FD) is a rare, X-linked lysosomal storage disorder that can result in fatal end-stage renal disease, heart failure, and cerebro-occlusive events. Vague clinical symptoms and rarity often mean diagnosis and potential treatment is delayed. Ophthalmic findings in FD patients can be helpful in establishing an early diagnosis and timely treatment. SD-OCT imaging in FD patients shows hyper-reflective foci in characteristic patterns within the inner retinal layers. We found that the HRF was localised in linear distributions at the deep and superficial borders of the retinal inner nuclear layer, likely reflecting anatomic vascular plexuses and FD-related sphingolipid deposition within the vessel walls. These results support the use of SD-OCT to aid FD diagnosis in undifferentiated patients and highlight the potential for prognostication and disease monitoring.

"HaNDL" with positive Anti- MOG antibody: The Expanding Spectrum of anti-MOG associated disorders.

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Case Description: A 52-year-old male presented with global language dysfunction, headache and papilledema. History included migraine and testicular cancer with curative treatment 30 years prior. Acute stroke was excluded with normal CT brain/angiogram and MRI brain. Lumbar puncture revealed lymphocytic pleocytosis of 225 cells/µl, elevated protein 1.63 g/L, normal glucose and opening pressure of 32cmH2O. COVID-19 PCR was negative. He was empirically treated with antivirals and antibiotics for 24 hours until PCRs returned negative. EEG revealed diffuse encephalopathic pattern with focal left hemispheric slowing without epileptiform abnormality. Symptoms resolved within 24 hours. However, these episodes occurred recurrently every 2-3 days. Investigations for infective, autoimmune (including anti-NMDA, anti-VGKC antibodies), malignancy and paraneoplastic aetiologies were negative. Minimal left hemispheric leptomeningeal enhancement was seen on MRI.

In week three, he developed optic disc haemorrhages, bilateral vitreous haemorrhages and secondary maculopathy with enlarged blindspots on visual fields. Repeat CSF opening pressure was 20cmH2O, with persistent lymphocytosis and high protein. Intravenous methylprednisolone was commenced without clinical response.

Anti-myelin oligodendrocyte glycoprotein (anti-MOG) antibody via flow-based assay on fixed transfected cells subsequently returned strongly positive. At 6 weeks there was spontaneous clinical improvement with resolution of headaches and papilledema.

Discussion: Headache and neurological deficits with lymphocytosis (HaNDL) is a distinct neurological entity characterised by recurrent migraine and lymphocytic pleocytosis on cerebrospinal fluid without an identifiable aetiology. It is a diagnosis of exclusion. This case is unique as the neurologic phenotype is strikingly similar to that seen in HaNDL. Whilst leptomeningeal disease has been reported with anti-MOG antibodies, the association of HaNDL with anti-MOG antibody has not been reported. Given the lack of response to prednisolone, we cannot conclude that the antibody is pathogenic here; it may represent epiphenomenon. Further review of HaNDL and their association with anti-MOG antibody is necessary.

The Efficacy, Adverse Effects and Economic Implications of Oral versus Intravenous Methylprednisolone for the Treatment of Optic Neuritis: A Systematic Review

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Introduction: Optic neuritis may occur in a variety of conditions, including as a manifestation of multiple sclerosis. Despite significant research into the efficacy of corticosteroids as a first line treatment, the optimal route of administration has not been well defined. This review aims to explore the efficacy, adverse effects, and economic implications of using oral versus intravenous methylprednisolone to treat acute optic neuritis.

Methods: A systematic search of the databases PubMed/MEDLINE, Embase and CENTRAL was performed to July 2022, prior to data collection and risk of bias analysis in accordance with the PRISMA guidelines.

Results: Six articles fulfilled the inclusion criteria. The results showed that in the treatment of acute optic neuritis, oral methylprednisolone has a non-inferior efficacy and adverse effect profile in comparison to intravenous methylprednisolone. In a cost analysis, oral methylprednisolone to be more cost effective than intravenous methylprednisolone.

Conclusions: Oral methylprednisolone has comparable efficacy and adverse effect profiles to intravenous methylprednisolone for the treatment of optic neuritis. The analysis suggests oral administration is more cost effective than intravenous administration; however, further analyses of the formal cost-benefit ratio are required.

Immune Checkpoint Inhibitors and Optic Neuropathy: A Systematic Review

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Introduction: Immune checkpoint inhibitors are a class of monoclonal antibodies that are used as a mainstay of immunotherapy for multiple solid organ malignancies. With the recent increase in popularity of these agents, immune-related adverse events including optic neuropathy are becoming more frequently reported. This review aims to explore the association between immune checkpoint inhibitors and optic neuropathy through analysis of incidence, clinical features, investigations, treatment and patient outcomes.

Method: A systematic search of the databases PubMed/MEDLINE, Embase and CENTRAL was performed from inception to September 2022. Data collection and risk of bias analysis was subsequently conducted in accordance with the PRISMA guidelines.

Results: Eleven articles fulfilled the inclusion criteria. The results showed an increased incidence of optic neuropathy among patients receiving immune checkpoint inhibitor therapy compared to the general population. Presentation with painless reduced visual acuity and optic disc swelling was most common. Investigation findings were poorly documented. The only two patients who achieved full resolution of symptoms were treated with oral prednisolone.

Conclusion: There is a strong association between immune checkpoint inhibitor therapy and development of optic neuropathy. Although it remains uncommon, the incidence of optic neuropathy in this population exceeds that of the general population. Future research is needed to further characterise the risk profiles of patients who are most likely to develop ICI-associated optic neuropathy, and treatment pathways for these patients.

Junctional Scotoma With Mass In Sellar - Para sellar Tursica: A Case Report

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Introduction: Findings of visual field defects on examination are one of many things that may indicate visual pathway lesions. Visual field defects located on the vertical midline are pathognomonic for neurological defects, one of which is lesions in the chiasm. Lesions in the chiasm may be caused by congenital abnormalities, trauma, infarcts, aneurysms, or neoplasms. Pituitary adenoma is one of the most common tumours located in the chiasm. Early evaluation of pituitary adenoma can help improve treatment for pituitary adenoma patients.

Purpose: To present a case of junctional scotoma with mass in sellar - parasellar tursica

Case Report: A 69-year-old male came to Cicendo Eye Hospital with a chief complaint of blurred vision of the left eye (LE) for the past 5 years. He had no complaints regarding eye pain, diplopia ptosis, headache, hormonal imbalance, or signs of increased intracranial pressure. Visual field examination revealed superotemporal quadrant visual field defect on the RE and generalized visual field defect on the LE. Neuroimaging in this patient showed calcified solid mass in the sellar-parasellar ec suspected pituitary macroadenoma. The patient was diagnosed with Bilateral Optic Atrophy, Junctional scotoma ec Parasellar lesion, CN III palsy incomplete with pupillary sparing OS, CN VII palsy central sinistra, Mass a/r sellar-paraselar ec suspected pituitary macroadenoma, Immature Senile Cataract ODS, Hypertension Stage 2. The patient was consulted to the neurosurgery, neurology, and endocrinology department.

Conclusion: Loss of vision and visual field defect are one of the ocular manifestations that could indicate visual pathway lesions. Junctional scotoma is a visual field defect that may be caused by lesions in Wilbrand's knee such as pituitary adenoma. Early detection of pituitary adenoma patients may provide optimal and proper evaluations by several specialists.

Keywords: visual field defect, junctional scotoma, pituitary adenoma

Journey through the Eye

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The "Journey Through the Eye" approach is a metaphorical exploration of the intricate and interconnected parts of the eye, with each clinical sign serving as a significant landmark along the way. This article aims to provide a cohesive narrative of exploration, accompanied by visual representations to aid in recalling crucial CLINICAL SIGNS, benefiting clinicians in their practice even before investigations come into play.

The eye, a complex universe in itself, reflects numerous aspects of brain function. Our expedition commences with the iris, the gateway to perception, offering clinical insights into eye color and conditions such as coloboma, aniridia, iritis, krukenberg spindle, and melanoma. We continue our journey through the conjunctiva, uvea, and associated inflammatory disorders. In the cornea, we must not overlook the significance of "band keratopathy," indicative of hypercalcemia, akin to a frosted window obstructing the view. The lens, a pivotal landmark, holds key abnormalities like cataract and lenticonus, crucial for the neuro-ophthalmologist to focus and refract, like the lens itself. Moving onward, we encounter the star landmark, "the eyelid," featuring ptosis with lid lag, marcus gunn jaw winking, peep sign of myasthenia gravis, blepharospasm, apraxia of eyelid opening, collier's sign, Gifford sign, eyelid fasciculations, pseudoptosis, and more. We navigate through ocular muscle pathologies, both in the clinic and the ICU.

Finally, we delve into the retina, the light-sensitive tissue, and its connections to the brain. This part of the journey encompasses nystagmus, optic nerve, optic chiasma, optic tracts, optic radiations, medial longitudinal fasciculus, paramedian pontine reticular formations, brainstem, cortical stroke presentations, and beyond.

As we conclude this exploration of eye and brain localization in the clinic, we reflect on the significance of comprehending each clinical sign in neuro-ophthalmology. This journey serves as a valuable reference for clinicians, facilitating enhanced understanding, diagnosis, and management of complex eye-related neurological conditions.

Neuro ophthalmology education – what is the role for social media?

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Introduction: The spectrum of medical education is constantly changing. Medical students and junior doctors are now using picture mnemonics, songs, and stronger memory strategies to learning, as well as being more exposed to short snippets of information to process rather than longer didactic lectures and textbooks. Clinical based learning is being emphasised more.

Within neuro ophthalmology, similar adaptations have been made. "Neuro ophthalmology with Dr Andrew Lee" on Youtube (https://m.youtube.com/channel/UC5HcfsELV0W9AqtvJvpQQSg) has 65,000 subscribers (as at May 2023). Research into standard automated social media posts compared to visual abstracts showed a significant increase in viewership with visual abstracts, although no increase in full text clicks. Many medical colleges and societies are utilising social media to increase awareness of conditions, recent publications, and updates.

Methods: In January 2022, we (two Neuro-ophthalmologists) started an Instagram (@neuroptic) and Twitter (@neuropticEd) aimed at providing hand illustrated summaries of neuro-ophthalmology and related topics. The images were the same on both accounts, but captions and hashtags were different due to platform requirements. No paid advertising was organised for either of the accounts. However, social media algorithms commonly suggest accounts to contacts of subscribers. We also gained traction through word of mouth and mentions at NOSA 2022.

Results: In 18 months, there have been 100 posts, with 38,300 views on Instagram in total. There are 268 followers, most between 25-34 years of age. 38.8% of followers were from Melbourne, and 56.3% from Australia. 66.8% of followers were female. Early on, we also found that very simple illustrations gained the most views compared to illustrations explaining more complex concepts. This allowed us to tailor our illustrations towards simplicity, reflecting the nature of the social media platforms.

Conclusion: The preliminary data we have will continue to shape our educational posts to reach a wider audience in the future.

Adult-onset Acute Necrotising Encephalitis with Bilateral Incongruent Homonymous Hemianopia

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Introduction: Genetic acute necrotising encephalopathy (ANE1) is a rare encephalopathy following a viral febrile illness (commonly influenza) in patients with an autosomal dominant missense mutation in the RAN Binding Protein 2 (RANBP2) gene[1,2,3]. ANE1 presents with bilateral symmetrical thalamic lesions[2,3] and typically presents in young children[3].

Case: 45-year-old female developed acute onset binasal hemianopia and quadruple sectoranopia in relation to influenza A infection. The diagnosis was ANE1 with RANBP2 mutation. Her child had previously had acute disseminated encephalomyelitis (ADEM) in the context of influenza infection. In the adult case, MRI Brain demonstrated bilateral lateral geniculate nucleus lesions with T2 FLAIR enhancement. She was treated with intravenous methylprednisolone and plasma exchange, followed by oral prednisolone. After treatment, MRI changes improved and the patient's visual field deficit stabilised. There was subsequent improvement in the visual field testing with an ongoing binasal field deficit, and reduction in quadruple sectoranopia for several months following presentation and treatment.

Discussion: This case illustrates a rare disorder, with few presentations occurring in adults[3]. The visual field defects of binasal hemianopia and quadruple sectoranopia are uncommon, although consistent with bilateral lateral geniculate nuclei lesions. This diagnosis is an important differential to consider given the significant morbidity and mortality associated with ANE1[2].

References

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Optic Nerve Swelling related to Infective Endocarditis

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Introduction: Ocular manifestations of infective endocarditis (IE) include retinal artery occlusion, Roth spots and posterior uveitis. Optic disc swelling has rarely been reported.

Case: A 34-year-old female presented with penicillin-sensitive staphylococcus aureus IE with aortic, mitral and tricuspid valve vegetations. She had a right middle cerebral artery territory ischaemic stroke and pulmonary emboli. Visual acuity and colour vision were normal. Fundoscopy showed bilateral optic nerve swelling associated with bilateral peripapillary hemorrhages and a Roth spot in her left eye. The CSF opening pressure was 23cm of water. CT Venogram and MRI Brain did not demonstrate venous sinus thrombosis or any features suggestive of raised intracranial pressure. Three months later, following successful treatment of her IE, the ocular examination was normal with resolution of the optic disc swelling.

Discussion: Papilloedema is highly unusual in IE, and the small number of previously published cases come mostly from the pre-penicillin era. These suggest that optic nerve swelling may occur with minimal impairment of visual function, and that Roth spots are often a co-existent finding.[1,2]. Previous clinic-pathological studies may be consistent with a hypothesis that optic nerve swelling is an immunologic phenomena of IE, similar to Roth spots. It is possible that it is under-recognised, since visual symptoms are slight.

References

1. Dienst EC. Pathologic changes in the eye associated with subacute bacterial endocarditis. Archives of Ophthalmology. 1944;31(3):198. doi:10.1001/archopht.1944.00890030028002
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Visual Function Recovery in Patient with Ethambutol- Induced Toxic Optic Neuropathy

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Introduction: Tuberculosis (TB) remains an important health problem in Indonesia, with the use of ethambutol as an effective first-line therapy regimen. However, ethambutol use may cause permanent visual loss in a dose- and duration- dependent optic neuropathy.

Purpose: To report visual function recovery in a patient with ethambutol-induced toxic optic neuropathy (EON).

Case Report: A 23-year-old man came with painless gradual visual loss since three months prior to presentation. He had been diagnosed with pulmonary TB and consumed ethambutol 800 mg/day as one of the therapy regimens for 11 months. Visual acuity was 0.08 on both eyes. Ophthalmology examination showed reduced pupillary light reflexes on both eyes. He developed dyschromatopsia and decreased contrast sensitivity on both eyes. Humphrey visual field test showed bilateral cecocentral visual field defect, with increased retinal nerve fiber layer thickness on optical coherence tomography (OCT) test. Head computed tomography (CT) scan was within normal limit. The patient was diagnosed as EON. He was advised to stop ethambutol consumption immediately and given oral citicoline and zinc supplementation. After 1 month follow up, patient visual function was gradually recovered.

Conclusion: Ethambutol-induced toxic optic neuropathy is a sight-threathening condition. Regular screening of patients taking ethambutol by an ophthalmologist is recommended for early detection of the disease. If EON is suspected, Immediate discontinuation of the drug is advised to prevent potentially devastating visual loss.

Keyword: Ethambutol, Ethambutol-induced toxic optic neuropathy, Tuberculosis, Visual function recovery.

Saturday 23rd September		
08.55 – 09.00	Introduction and welcome	Clare Fraser and Celia Chen
Session 1: Introduction		
09.00 – 09.20	Practical clinical approach to afferent disease	Susie Mollan
09.20 – 09.40	MRI: visual pathway	Christen Barras
09.40 – 10.00	Interpretation of visual fields	Anthony Arnold
10.10 – 10.30	Imaging the optic disc – OCT in neuro-ophthalmology	Susie Mollan
10.30 – 11.00	Morning Tea	
Session 2: Swollen discs		
11.00 – 11.20	Approach to disc oedema	Celia Chen
11.20-11.50	Approach to Intracranial hypertension – IIH and more	Susie Mollan
11.50 – 12.30	Case challenge quiz (swollen discs) and investigations	Clare Fraser
12.30 – 13.30	Lunch	

Session 3: Optic neuropathies		
13.30 – 14.00	AION: Arteritic vs. non-arteritic	Anthony Arnold
14.00 -14.30	Inherited optic neuropathies	Kate Ahmed
14.30 -15.00	Optic neuropathy case quiz (10 mins each) - Nutritional - Toxic - Compressive	Megha Kaushik
15.00 – 15.30	Afternoon Tea	
Session 4: Miscellaneous		
15.30 – 16.00	Anisocoria: clinical evaluation and pharmacologic testing	John Leaney
16.00 – 16.20	Retinal mimics - cases	Sumu Simon
16.20 – 17.00	Help it's a child – case series and quiz - Strange discs - Paediatric optic neuritis - Paediatric anisocoria	James Smith Susan Carden
17.00-19.00	Welcome Drinks Luna 10 located on level 10 of the Crowne Plaza Hotel	

Sunday 24th September		
Session 5: Optic neuritis		
09.00 – 09.30	Typical optic neuritis:	Susie Mollan
09.30 – 10.00	When it's not MS-ON MOG, NMO	Anthony Arnold
10.00 – 10.15	Other inflammatory ON – SLE, ANCA, sarcoid	Anthony Arnold
10.15 – 10.30	Infectious optic neuritis	Anthony Fong
10.30 – 11.00	Morning Tea	
Session 6: The Posterior visual pathway		
11.00 – 11.20	Functional visual loss	Neil Shuey
11.20-11.40	Higher visual processing	Christian Lueck
11.40 – 12.00	Resources for learning	Elle Nguyen
12.00 – 12.30	Sunday quiz and cases - Optic neuritis - Higher processing	Celia Chen
12.30	Conclusion and thanks	Celia Chen and Clare Fraser
12.40 - 13.30	Lunch	

Prof. Anthony Arnold	Consultant Neuro-ophthalmologist Professor and Chief Neuro-ophthalmology Division	Stein Eye Institute UCLA Dept. of Ophthalmology Los Angeles, California USA
A/Prof. Christen Barras	Consultant Neuro-radiologist	Royal Adelaide Hospital University of Adelaide South Australian Health and Medical Research Institute
A/Prof. Susan Carden	Consultant Paediatric Ophthalmologist	Royal Children's Hospital and Royal Victorian Eye and Ear Hospital, Melbourne, Australia
Prof. Celia Chen	Consultant Neuro-Ophthalmologist	Professor of Ophthalmology, Flinders University and University of South Australia
Dr Anthony Fong	Consultant Neuro-Ophthalmologist	Department of Ophthalmology Royal Brisbane and Women's Hospital Princess Alexandra Hospital Gold Coast University Hospital
A/Prof. Clare Fraser	Consultant Neuro-Ophthalmologist	Sydney Eye Hospital Save Sight Institute, Sydney, Australia
Dr Megha Kaushik	Consultant Neuro-Ophthalmologist	Royal Prince Alfred Hospital Liverpool Hospital Sydney, Australia
Dr John Leaney	Consultant Neuro-Ophthalmologist	Liverpool Hospital Royal Prince Alfred Hospital Sydney, Australia
Prof. Christian Lueck	Emeritus Professor Consultant Neurologist	School of Medicine and psychology Australian National University Canberra, Australia
Prof. Susan Mollan	Consultant Neuro-ophthalmologist	Queen Elizabeth Hospital, University Hospitals Birmingham, University of Birmingham, UK
Dr Elle Nguyen	Consultant Neurologist and Neuro-Ophthalmologist	Alfred Health and Royal Victorian Eye and Ear Hospital Melbourne, Australia
Dr. Neil Shuey	Consultant Neuro-Ophthalmologist	St Vincent's Hospital and Royal Victorian Eye and Ear Hospital, Melbourne, Australia
Dr Sumu Simon	Consultant Neuro-Ophthalmologist	Neuro-ophthalmology Unit Department of Ophthalmology Royal Adelaide Hospital
Dr James Smith	Consultant Paediatric Ophthalmologist and Head of Department RNSH	Westmead Childrens Hospital Royal North Shore Hospital Sydney, Australia

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