

UKNOS Joint Meeting with the Danish Neuro-Ophthalmology Society

27th September 2019

Downing College, Cambridge

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| 0845 | Welcome | Gordon Plant |
| **First session - Chair: Luke Bennetto** | | |
| 0900 | A survey on vintage steps in Danish Neuro-ophthalmology, from Rønne’s step and on | Hans Fledelius |
| 0930 | Five diagnoses you cannot afford to miss | Andy Lee |
| 1015 | Coffee |  |
| **Symposium on optic neuritis - Chair: Steffen Hamann** | | |
| 1045 | Optic neuritis: Diagnosis and ~~mimics~~ getting the diagnosis wrong | Simon Hickman |
| 1115 | New biomarkers in optic neuritis | Jette Frederiksen |
| 1145 | Update on CRION | Gordon Plant |
| 1215 | Paediatric optic neuritis | Ming Lim |
| 1245 | Lunch |  |

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| **Free papers – Chair: Gordon Plant** | | |
| 1345 | 1) Melanopsin: targeted ectopic expression for optogenetic visual restoration | Michael Gilhooley |
|  | 2) Progression over five years of prelaminar hyperreflective lines to optic disc drusen in the Copenhagen Child Cohort 2000 Eye Study | Steffen Hamann |
|  | 3) Idiopathic intracranial hypertension (IIH): From genomics to pathophysiology | Elisabeth Wibroe |
|  | 4) MOG antibodies: a unique biomarker in the evaluation of optic neuritis | Sudarshini Ramanathan |
|  | 5) Assessing neuro-imaging requests and reports submitted from the neuro-ophthalmology division within the eye hospital | Siddharth Sinha |
|  | 6) The nasal coastline of Traquair’s island.  On the influence of gaze direction. | Hans Fledelius |
|  | 7) The utility of optical coherence tomography (retinal nerve fibre layer and ganglion cell layer thickness), to measure axonal degeneration in multiple sclerosis subjects | Grace Levy-Clarke |
|  | 8) Functional-structural assessment of the optic pathways in patients with optic neuritis | Mathias Falck Schmidt |
| 1545 | Tea |  |
| **Interactive case presentations – Chair: Susie Mollan** | | |
| 1615 | 1) A case of central fusion disruption syndrome suggests the location of the motor fusion centre of the brain | Megan Jeffries |
|  | 2) Idiopathic orbital apex inflammation presenting with optic neuropathy | Ritu Chaturvedi |
|  | 3) Ophthalamological negligence? | Luke Bennetto |
|  | 4) Waves and canals | Joanna Jefferis |
| 1715 | Neuro-ophthalmology of outer space | Andy Lee |
| 1800 | Close of meeting |  |

**A survey on vintage steps in Danish neuro-ophthalmology, from Rønne’s step and on.**

Hans C Fledelius, Rigshospitalet, Copenhagen

Modern ophthalmology developed after von Helmholtz’ launching of the ophthalmoscope in the 1850’es. In Denmark the principle was introduced by first Copenhagen professor in ophthalmology, Hansen Grut, who - in an era dominated by continental tradition - was also open to Anglosaxon medical know-how. Hansen Grut’s first man was young Jannik Bjerrum (1851-1920), and *his* foreman again Henning Rønne (1878-1947), both later professors in Copenhagen. This also held for Marius Tscherning (1854-1939), who contributed to neuro-ophthalmology with important studies on accommodation, mainly performed under Javal at Sorbonne in Paris. In 1910 he succeeded Bjerrum on the chair in Copenhagen.

Bjerrum introduced the black curtain and precise small object campimetric visual field testing, thus to demonstrate and emphasize the *relative defects.* This markedly widened the concepts of glaucoma, and *Bjerrum’s* became eponym for *arcuate scotoma*. Added hereto, however, the full topic of neuro-ophthalmology was shared with his young assistant Henning Rønne, and a golden age was established for Copenhagen ophthalmic science. - In the century to follow, no Danish names have gained a similar fame, though currently with fiery souls to serve the clinical obligations within the discipline.

Rønne was an eager and continuously burning character. Based on personal clinical experience *and* pathology studies, in flourishing language he published predominantly in German ophthalmic journals. He shared quite many details with his contemporary reader - thus to expose a marked scientific transparency, for which inter al. Duke-Elder gave him much credit. Among Rønne’s topics were the visual field borders, the large pupil, the chiasmal decussation and the lateral geniculate body relay, arcuate hemiscotomas, and the peripheral crescent’s mono-representation in the temporal visual field.

A more humble line is drawn up to our present Danish era, where in particular courses given by Gordon Plant and Dan Milea have given a lift to both spirit and level of performance.

**Free papers – Chair: Gordon Plant**

**Melanopsin: targeted ectopic expression for optogenetic visual restoration**

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**Introduction**: Melanopsin is an optogenetic tool - it renders neural cells sensitive to light when exogenously expressed1 - and is emerging as a prime candidate for clinical optogenetic approaches to visual restoration2. The neural retina tissue surviving in the inherited retinal degenerations (IRD) after visual loss presents a model target for such approaches, however it is unclear if targeting specific neuronal populations within this tissue is advantageous over non-specific delivery. A fair comparison between these two methods is made difficult by the persistence of native photosensitive retinal ganglion cell (pRGC) responses in IRD models3. Here we describe a novel mouse model lacking all native retinal light responses to allow a more robust comparison of optogenetic targets.

**Purpose:** To test the hypothesis that retinae devoid of both canonical and pRGC photoreception will remove the “noise” of intrinsic responses and allow comparison of targeted expression of melanopsin in ON-bipolar cells with non-specific delivery.

**Materials and Methods:** Retina-degenerate mice lacking native melanopsin and expressing Cre recombinase in retinal ON-bipolar cells (L7.Cre,Opn4-/-,Pde6brd1/rd1) were used. At P45, intravitreal injections of adeno-associated virus containing the human Melanopsin gene (OPN4) driven by “floxed” EF1a (N=8,8), “non-floxed” CBA (N=8) promotors or saline (N=8) were administered. Eight weeks later, behavioural visual assays and ex-vivo multiple electrode array electrophysiological recordings of retinal light responses were performed.

**Results:** No significant difference was seen in behavioural responses, sensitivity of electrophysiological responses nor onset kinetics between the two treated groups. Expression in bipolar cells specifically however led to significantly shorter half-life (p<0.001) & duration (p<0.001) of responses.

**Conclusion:** This represents the first report in the literatureof restoration of light responses in a retina devoid of all native photoreception. While there was no apparent advantage in sensitivity between targeted and non-specific delivery, when Melanopsin was specifically expressed in bipolar cells, offset kinetics of responses were faster, presenting them as attractive targets for clinical optogenetic approaches to visual restoration.

***References***

1. Melyan, Z., Tarttelin, E.E., Bellingham, J., Lucas, R.J. and Hankins, M.W. (2005) Addition of human melanopsin renders mammalian cells photoresponsive. *Nature*, **433**, 741-745.
2. De Silva, S.R., Barnard, A.R., Hughes, S., Tam, S.K.E., Martin, C., Singh, M.S., Barnea-Cramer, A.O., McClements, M.E., During, M.J., Peirson, S.N. *et al.* (2017) Long-term restoration of visual function in end-stage retinal degeneration using subretinal human melanopsin gene therapy. *Proc Natl Acad Sci U S A*, **114**, 11211-11216.
3. Hattar, S., Lucas, R.J., Mrosovsky, N., Thompson, S., Douglas, R.H., Hankins, M.W., Lem, J., Biel, M., Hofmann, F., Foster, R.G. *et al.* (2003) Melanopsin and rod-cone photoreceptive systems account for all major accessory visual functions in mice. *Nature*, **424**, 76-81.

**Progression over five years of prelaminar hyperreflective lines to optic disc drusen in the Copenhagen Child Cohort 2000 Eye Study**

Lasse Malmqvist.1, Xiao Qiang Li1, Mathias Hvidtfelt Hansen1, Alexander Kai Thomsen1, Anne Mette Skovgaard3,4, Else Marie Olsen3,6, Michael Larsen1,2, Inger Christine Munch.2,5 and Steffen Hamann1,2

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**Introduction:** The purpose of the study was to examine five-year changes in eyes with optic disc drusen at baseline on optical coherence tomography scans and the relation of incident drusen to hyperreflective prelaminar lines.

**Methods:** The study included children who presented at baseline, when participants were aged 11-12 years, and again 5 years later. Grading for optic disc drusen was made in all. Grading for prelaminar lines was made in all children at follow-up and in eyes with optic disc drusen at baseline. Analyses included associations with scleral canal diameter at baseline in all children with optic disc drusen and a nested control group of 115 children without optic disc drusen. Data are reported as the number of children having at least one drusen or at least one hyperreflective line per person.

**Results:** The analysis included 724 children who attended both rounds of the study. Of these 11 (1.5 %) had optic disc drusen at baseline. Five additional children had developed optic disc drusen at follow-up, whereas optic disc drusen had disappeared in none, so that 16 (2.2 %) children had optic disc drusen in one or both eyes at follow-up. Children with optic disc drusen at the 5-year follow-up had had a mean scleral canal diameter of 1364 µm (IQR 81 µm), compared to 1457 µm (IQR 197) µm in 115 nested controls without optic disc drusen (P<0.001). Optic disc drusen at follow-up were associated with more hypermetropic refraction. All children who had optic disc drusen at follow-up also had prelaminar hyperreflective lines. In addition, such lines were found at follow-up in 24 of the remaining 708 children without optic disc drusen (P<0.001). Prelaminar hyperreflective lines with or without optic disc drusen were associated with a narrower scleral canal (diameter 1364 µm, IQR 119 µm) compared to absence of prelaminar lines (1486 µm, IQR 206 µm; P<0.0001).

**Conclusion:** This study provides the first evidence from a prospective study that small optic discs and prelaminar hyperreflective lines on optical coherence tomography are risk factors for the development of optic disc drusen. The association between prelaminar hyperreflective lines, hypermetropia and a narrow scleral canal supports that a crowded disc is an essential predisposing factor for the development of optic disc drusen.

**Idiopathic intracranial hypertension (IIH): From genomics to pathophysiology**

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**Relevant financial disclosures:** No relevant financial disclosure

**Introduction:** Currently an experienced neuro-ophthalmologist is essential in the correct diagnosis of idiopathic intracranial hypertension (IIH). Over 40% suffer from wrong diagnosis and thus wrong treatment in the acute stage and/or for years in the more chronic state with vision loss and poor quality of life to follow.

**Method:** We plan to include 70 cases and 70 sex, weight and age matched controls. All probable IIH patients from 2019-22 are invited to participate in the study. Of these, individuals diagnosed with IIH are enrolled as cases and the rest as controls. Patients are excluded if they suffer from other serious somatic disease or have insufficient Danish skills. At baseline and after 6 months all patients will go through a standard IIH workup according to hospital guidelines: Ophthalmological examination including autoperimetry, neurological examination, CT and/or MRI of the brain and orbits and lumbar puncture. In addition, patients will have an optical coherence tomography (OCT) performed and an OCT angiography. One saliva sample and one blood sample will be collected from each patient and eventually their parents and sent to Whole Exome Analysis in University College London.

**Outcome:** Our primary outcome is detailed insight into the structural and functional optic nerve changes in IIH by OCT and determining genetic disposition to the disease. Secondly, we aim to provide new diagnostic and prognostic biomarkers for IIH and thus form a more solid basis for further research.

**Perspective:** We hope to reveal a more efficient, non-invasive approach to IIH based on 1) already existing equipment widely used in the clinic (OCT and OCT angiography) and 2) a simple blood/saliva sample for genetic analyses. It is our hypothesis, that this approach will be efficient in the diagnosis of IIH, in monitoring the treatment (saving some patients from invasive procedures) and in the development of a better and more targeted treatment improving visual outcome. Secondly, the study will provide a basis for future studies of conditions with dysregulated brain water.

**MOG antibodies: a unique biomarker in the evaluation of optic neuritis**

Sudarshini Ramanathan 1,2, Fabienne Brilot 1,2,3, and Russell C Dale 1,2,4 on behalf of the Australian and New Zealand MOG Study Group

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Professor Russell Dale has received research funding from the Star Scientific Foundation, The Trish Multiple Sclerosis Research Foundation, Multiple Sclerosis Research Australia, the Petre Foundation, and the National Health Medical Research Council (Australia). Dr Dale has received honoraria from Biogen Idec as an invited speaker.

**Introduction/ Purpose:** Myelin oligodendrocyte glycoprotein (MOG) is a putative candidate antigen in antibody-associated demyelination1. We sought to identify the clinical phenotypes, radiological characterisation, treatment responses, and outcomes in MOG antibody-associated demyelination.

**Methods:** We performed a flow cytometry live cell based assay to detect MOG antibodies in adults with demyelination; undertook blinded neuroradiological assessment on 50 patients with first episode optic neuritis (ON) due to multiple sclerosis (MS), MOG, or aquaporin-4 (AQP4) antibodies; and evaluated treatment responses and outcomes in 33 children and 26 adults with relapsing MOG antibody-associated demyelination.

**Results:** MOG antibodies were strongly associated with recurrent and bilateral ON (BON) with optic disc swelling [9/23 adults with AQP4 antibody-negative neuromyelitis optica spectrum disorder v. 0/52 controls (p<0.001)]2. There were low rates of MOG antibody-positivity in Australian MS (1/76). Radiologically, bilateral longitudinally extensive optic nerve involvement was more common in MOG and AQP4-ON than MS-ON. MOG-ON exhibited anterior optic nerve involvement and optic disc swelling, while AQP4-ON more frequently exhibited posterior visual pathway involvement including the posterior segment of the optic nerve, chiasm, and optic tract3. ON was dominant at initial presentation [BON 32%, unilateral (UON) 22%] and throughout the clinical course (BON 19%, UON 34%) in relapsing MOG antibody-associated demyelination4. Patients were steroid responsive but 70% of episodes relapsed, especially at prednisone doses <10 mg daily or within 2 months of cessation. Immunotherapy, including maintenance prednisone (P=0.0004), intravenous immunoglobulin, rituximab, and mycophenolate, all reduced annualised relapse rates4. 59% of patients experienced residual disability, particularly with increasing relapses.

**Discussion**: MOG antibodies are strongly associated with ON. Relapsing disease is steroid responsive but vulnerable to relapse, responds to immunosuppression, and has the potential to result in sustained disability. MOG antibody-associated demyelination is a distinct clinical entity from MS and AQP4 antibody-associated demyelination, with pathogenic, therapeutic, and prognostic implications.

**References:**  
1. Ramanathan S, Dale RC, Brilot F. Anti-MOG antibody: The history, clinical phenotype, and pathogenicity of a serum biomarker for demyelination. Autoimmunity Reviews 2016; 15(4): 307-24.  
2. Ramanathan S et al. Antibodies to myelin oligodendrocyte glycoprotein in bilateral and recurrent optic neuritis. Neurology: Neuroimmunology and Neuroinflammation 2014;1(4):e40  
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4. Ramanathan S et al. The clinical course, therapeutic responses, and outcomes in relapsing MOG antibody-associated demyelination. Journal of Neurology Neurosurgery and Psychiatry 2018; 89(2):127-137.

**Assessing neuro-imaging requests and reports submitted from the neuro-ophthalmology division within the eye hospital**

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**Relevant financial disclosures:** No relevant financial disclosure

**Introduction:** Optic neuropathies refer to disorders within the optic nerve. Common conditions include optic neuritis, ischemic optic neuropathy and traumatic optic neuropathy (1). Similarly, isolated cranial nerve palsies are another common pathology due to dysfunction within the three eye motor nerves (2). These conditions can be idiopathic but can also be due to serious underlying pathologies including multiple sclerosis or tumours. Therefore, brain imaging is required to rule out these conditions (1,2).

**Methods:** 54 patients under the care of one consultant, imaged over 9 months at Manchester Royal Eye Hospital were analysed for this audit. With fixed timeframes set below: 

1. Most patients presenting with optic neuropathies should be scanned within 4-6 weeks (3).

2. Patients specifically presenting with typical optic neuritis should be scanned within 8 weeks (4).

3. Patients presenting with cranial nerve palsies except pupil involving III nerve palsy should be scanned within 6-12 weeks (5).

4. From scan request to report the set timeframe is <8-12 weeks.

**Results**: The results show that the overall compliance was 65.4% for time taken between requests and scans using the relevant guidelines. With 44.2% compliance from requests to reports using the timeframe set by the clinical supervisor.

**Conclusion:** Due to financial and radiology staffing shortages, the main solution would aim to effectively prioritise patients based on clinical need and streamlining patient bookings to reduce delays.

**The nasal coastline of Traquair’s island.  On the influence of gaze direction.**

Hans C Fledelius1

1. Copenhagen, Rigshospitalet, Denmark

Visual interpretation first depends on input from visual space and its refraction by the ocular media. Next, retinal photoreceptors transmit wavelength energy to propagating neural transmission further on. From posterior pole to ora serrata retinal photoreceptors are ready for the task, with decreasing resolution from foveal region to periphery.

Practically, the visual field is given by the reverse outward projection of the total retinal area actually involved. This seems to imply that in quite many visual situations a considerable amount of retinal receptors are not in use, anteriorly in particular. Theoretically, such functionally ‘blinded’ photoreceptors might be triggered into function If transmitted from hidden and dormant to sight-active conditions, for instance by a turn of the eye. In the horizontal plane, a lateral gaze direction thus would add a free sector to nasal visual space, by simply shortcutting the physical obstacle made up by the bridge of the nose.

Based on a 2015 observation, at present five normal emmetropes aged 11 to 59 years were further tested by kinetic supra-threshold Goldmann perimetry (IV,4e). On the specific question raised, a 25o lateral gaze direction did not increase the size of the recorded nasal visual field, but merely transposed it. Traquair just had to accept the size of his nasal beach. The indeed simple topic is discussed. How early is the individual’s visual field conditioned, by neural mechanisms, and apparently how fixed?

**The utility of optical coherence tomography (retinal nerve fibre layer and ganglion cell layer thickness), to measure axonal degeneration in multiple sclerosis subjects**

**Grace Levy-Clarke1, Levy Carrie2 and Mark Cascione3**

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**Relevant Financial Disclosure:** Investigator Initiated Study  
Grace Levy-Clarke, MD: Research support/consulting fees: Sanofi/Genzyme. Mark Cascione, MD: Research support/consulting fees: Sanofi Genzyme

**Purpose**: To Assess the Utility of OCT to measure Axonal Degeneration in MS subjects.

**Background**: The pathophysiology observed in other white matter structures in MS, appears to parallel the presence of inflammatory demyelination and associated axonal loss in optic nerve (ON). Additionally, the RNFL is a major site of axonal loss after ON. RNFL loss, subsequently leads to GCL loss. OCT allows for non-invasive retinal imaging, with accurate in vivo estimates of RNFL and GCL thickness

**Design:** Prospective cross-sectional study.

**Methods**: Baseline data for analysis was abstracted from clinical research source documents. Main outcome measures were OCT, patient determined disease steps (PDDS) scores, demographic and clinical characteristics. Descriptive statistics and simple linear regression analysis (to predict PDDS scores based on RNFL thickness} were calculated. (Statistical Package for the Social Sciences Version 24.

**Results:** 52 subjects of a planned cohort of 200 subjects: Mean RNFL thickness, stratified by age and ethnicity was thinner (mean 79.5μm +/- 12.7), OS and (mean 80.0 +/- 12.3μm), OD compared to published age matched normal patients (mean 100.1 microns +/- 11.6)1. Simple linear regression to predict PDDS scores based on RNFL thickness revealed: A significant regression equation, (F (1, 47) = 8.647, p < .005), with an *R2* of 0.16 (OS) and (F (1, 43) = 5.611, p < .022), with an *R2* of 0.12 (OD). Patients’ predicted RNFL-OS: 86.34 – 2.34μm for every 1- point increase in PDDS scores; patients' predicted RNFL-OD: 85.74 – 2.17μm for every 1-point increase in PDDS scores.

**Conclusions:** Preliminary results suggest an increase in PDDS may be predictive of a decrease in RNFL thickness. Subsequent analyses, stratification based on the type of disease modifying therapy and disease duration will further clarify the utility of OCT as an emerging surrogate endpoint in pharmacotherapy development for MS.

**Reference:**

1. Budenz DL, Anderson DR, Varma R, et al. Ophthalmology. 2007 Jun;114(6):1046-52 Determinants of normal retinal nerve fiber layer thickness measured by Stratus OCT.

**Functional-structural assessment of the optic pathways in patients with optic neuritis**

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**Relevant financial disclosures:** Dr. Schmidt declares that he has no conflict of interest. Dr. Pihl-Jensen has received support from Biogen Idec for a currently ongoing observational trial of VisionSearch 1 mfVEP measurements in optic neuritis patients. Dr. Frederiksen declares that she has no conflict of interest

**Introduction/Purpose:** Pattern-reversal visual evoked potential (pVEP) is widely used for the diagnosis of Optic Neuritis (ON), but this method has some limitations. The aim of this study was to examine the added value of multifocal visual evoked potentials (mfVEP) and spectral domain optical coherence tomography (SD-OCT) in the diagnosis of ON in patients that exhibit a normal pattern-reversal visual evoked potential (pVEP).

**Materials and Methods**: Thirty-three patients with the history of having ON and thirty gender and age matched healthy controls (HC) were investigated. We included patients who were suspected of having a first time ON and in whom pVEP showed normal results.Both eyes of the patients and HC were systematically investigated with SD-OCT, Visual Acuity, pVEP and mfVEP. The ON affected eyes of the patients were compared with only one randomly selected eye per person in the HC group. The fellow “non-affected” eye of patients was held as a separate group. Statistical analyses were performed (incl. t-test, Spearman's Rank-Order Correlation test) using SPSS Statistics, Version 24.0.

**Results**: In regard to OCT data, a significant difference was found in mean retinal nerve fiber layer thinning (RNFLt) between patients and HC (p=0.013) (i.e. 84.24 (±17.00) μm vs 93.48(±6.44) μm). An association was detected in patients between the inter-eye asymmetry of mean RNFLt and global (averaged) mfVEP amplitude (r = 0.565, p = 0.002).

When analysing mfVEP signals from sectors in the upper hemifield, a significant difference was found in mean mfVEP amplitude between patients and HC (p=0.005).

**Discussion**: In summary, this study provides evidence that pathologic changes are potentially measurable (via reduced RNFLt and focal analysis with mfVEP amplitude signals) in patients with clinical manifestations of ON although pVEP reports no abnormality. The mfVEP and SD-OCT may together be of value as supplementary tools in diagnosing patients on suspicion of a first episode of ON where pVEP reports no abnormality.

**Interactive case presentations – Chair: Susie Mollan**

**A case of central fusion disruption syndrome suggests the location of the motor fusion centre of the brain**

Megan Jeffries1 and Chris Hammond1

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**Relevant financial disclosures:** No relevant financial disclosure

**Introduction:** In 2012, a 30-year-old man, otherwise fit and well and with no medical history, developed sudden horizontal diplopia in all positions of gaze. There was no history of trauma, no history of unilateral poor vision, no history of flu like illness or intracranial pathology. He was found to have a small exophoria/exotropia in the primary position, measuring around 2 dioptres base-in, with stereopsis (60 seconds of arc on Frisby test), and described almost-constant diplopia with intermittent sensory fusion, and there was no motor fusion demonstrable. There was reduced adduction of the right eye with no abducting nystagmus of the left.

CT followed by MR imaging showed a small pontine cavernoma at the dorsal pons with associated intralesional haemorrhage, and associated developmental venous abnormalities. Over 7 years’ follow-up the diplopia has remained unchanged, and the MRI changes are unaltered. The patient was diagnosed with Central Fusion Disruption Syndrome. The loss of motor fusion caused by a small localized haemorrhage suggests that the dorsal pons may be the location of the horizontal motor fusion centre.

**Discussion:** In 1935 Alfred Bielschowsky1 described a case of ‘Horror Fusionis’ in which his patient was unable to fuse images under any conditions. This complicated case was eventually solved after discovering aniseikonia to be the cause of his fusional difficulty. However, since then there have been descriptions of cases of diplopia in which fusion simply cannot be obtained despite all efforts. This has been called Central Fusion Disruption Syndrome (CFDS).

In the largest series of patients with CFDS[[1]](#endnote-2), the causes included severe head trauma (42% of cases), intracranial pathology (15%), presumed post-viral (after a flu like illness, 5%), and, in 38% of cases, restoration of vision following prolonged unilateral traumatic cataract. In the vast majority of cases of CFDS, diplopia is intractable and the prognosis for treatment, other than occlusion, is poor.

**Reference**

1. Bielchowsky, A. Cngenital and acquired deficiencies of fusion. AmJOphtalmol 1935; 18:925-37.

**Idiopathic orbital apex inflammation presenting with optic neuropathy**

Ritu Chaturvedi1 and Mandagere Vishwanath1

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**Relevant financial disclosures:** No relevant financial disclosure

**Introduction:** Demyelinating optic neuritis (ON) is the most common cause of optic neuropathy. Progressive visual loss is noted over a course of few days to 2 weeks with spontaneous recovery to 0.3 or better in 80% of the cases. (1) A number of inflammatory, infective, tumours and ischemic conditions can cause optic neuropathy and need investigating if not fitting the typical pattern of a demyelinating aetiology.

**Materials and Methods:** We present retrospective consecutive case series of two patients with orbital apex inflammatory lesion, who presented with steroid responsive optic neuropathy.   
  
**Results:**  
Case 1: A 51yr old patient with a one day history of blurred vision in his right eye and a past history of amblyopia left eye. Examination confirmed right eye visual acuity of hand movement, relative afferent pupillary defect and absence of optic nerve swelling, leading to a provisional diagnosis of retrobulbar neuritis.

Due to the severity of visual loss in right eye and amblyopia in left eye he was treated urgently with intravenous methylprednisolone followed by oral prednisolone. Subsequent MRI scan showed changes at orbital apex which were deemed to be inflammatory. At 6 weeks, visual acuity improved to 0.0 (RE). Repeat MRI with contrast showed complete resolution of inflammatory apical lesion.

Case 2: A 55-year-old patient presented with progressive double vision and transient visual loss. He developed non-perception of light during attacks lasting approximately a minute four to five times a day. This was confirmed during such an episode in eye clinic. MRI scan showed an inflammatory orbital apex lesion. He was treated with intravenous methylprednisolone followed by oral prednisolone resulting in a marked improvement of his symptoms. Repeat MRI scan showed resolution of lesion. A complete body scan was undertaken to rule out lymphoma.   
  
**Discussion:** Optic nerve dysfunction needs to be appropriately investigated due to high risk of permanent visual impairment. Urgent imaging needs to be undertaken along with biochemical diagnosis. Histological diagnosis with biopsy at the orbital apex carries significant risk and was deferred as good clinical response to steroids was noted and maintained during the tapering stage.  
  
**Reference**  
1. Voss E, Raab P, Trebst C, Stangel M. Clinical approach to optic neuritis: pitfalls, red flags and differential diagnosis. Ther Adv Neurol Disord. 2011;4(2):123-34.

**Ophthalamological negligence?**

Luke Bennetto1

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Abstract to follow presentation

**Waves and canals**

Joanna M Jefferis,1 Nicholas T Skipper,2 Simon J Hickman3

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3. Department of Neurology, Royal Hallamshire Hospita, Sheffield, UK

Abstract to follow presentation



[www.uknos.com](http://www.uknos.com)

1. [↑](#endnote-ref-2)