



Abstract Booklet

**Wednesday 4th – Friday
6th October 2023**

**Royal Society of Medicine
1 Wimpole Street
London W1G 0AE**



Free papers & rapid-fires



WEDNESDAY 04th OCTOBER 2023

10.00 Session II (P)

Paediatric Free Paper

Moderators: Miss Stephanie West, Southampton and Ms Elizabeth O'Flynn, Southampton

10.01 Retreatment for reactivation of ROP following initial anti-VEGF injections. A 5-year retrospective study - E Kollia (CF), H Razzouk (SAS), S Biswas (C), J Ashworth (P), Manchester Royal Eye Hospital

To present the retreatment rates and the characteristics of ROP reactivation, as well as the differences between bevacizumab and ranibizumab injections in premature babies treated in our department over the past 5 years.

A retrospective analysis of babies with treated ROP was performed. 89 babies who required treatment from 2017 to 2022 were examined. We studied the severity of their disease with regards to their gestational age, treatment time and type and the need of further treatment. We also focused on the comparison of anti-VEGF agents for ROP. 22 out of 89 babies (14 boys and 8 girls) with aggressive posterior retinopathy of prematurity (APROP) and mean gestational age of 25+3w received initially anti-VEGF injections. 16 of those (11 boys and 5 girls) required retreatment with diode laser. 9 out of these 16 babies were treated with ranibizumab (Lucentis) and 7 with bevacizumab (Avastin). It is also of note that only 2 out of 67 babies who initially received laser treatment needed a complementary laser session.

The majority of babies with aggressive ROP who receive anti-VEGF agents will most probably require further laser treatment. At an equal level of retinal damage, it seems that their response to ranibizumab and bevacizumab is similar.

10.10 Clinical impact of optic nerve defect referrals to paediatric ophthalmology department and evaluation of suitability for virtual service - N Gibbings (O), S Jain (C), R Jolly (C), Royal Free Hospital

Retrospective audit of paediatric patients referred to ophthalmology service with optic nerves suspicious of swelling. Using snapshot audit of 1109 clinical records (4 months), a total of 27 patients were identified as being referred for blurred disc margins (2.43% of clinical episodes). 5 of these were diagnosed with papilloedema (3 active, 1 late presenting whilst under monitoring, 1 resolving) with a true positive rate of 18.5%. Most common referral pathway was optician (74%). In Medisight diagnosis search of "optic nerve drusen" as diagnosis, 23 additional patients were located in addition to the snapshot cohort. 82.6% were referred via the optician for suspicious discs. Average wait time for an appointment for this cohort was 13 weeks (range 0 days – 22 weeks), 5 weeks for snapshot group (range 0 days – 38 weeks).

Long wait times for suspected papilloedema patients could impact clinical outcomes negatively, a wide spectrum of wait times reflects the referral pathway with short waits via A&E and urgent eye care. A significant proportion of referred patients do have disc swelling requiring intervention. Orthoptists and ophthalmic photographers are in a good place clinically to facilitate a virtual diagnostic pathway for these patients, with clear communication with paediatric ophthalmologists to ensure swift review.

10.19 The role of oFFA in characterising optic nerve pathologies in paediatric populations - B Muthusamy (C), R T Brady (T), H Kaza (T), N Jain (T), Cambridge University Hospitals NHS Foundation Trust

Optic nerve (ON) pathologies can present a diagnostic challenge in paediatric populations. Fundus fluorescein angiography (FFA) is a good diagnostic tool but can be difficult in children. Oral FFA (oFFF) is a less invasive alternative, and with the advent of superior imaging - ultrawide field (UWF) retinal camera, is a favourable alternative to intravenous FFA. This study aimed to demonstrate the safety profile of established oFFA protocol, characterise ON pathology, and compare the diagnostic accuracy of consultant ophthalmologists to specialist trainee ophthalmologists in interpreting oFFA studies following a focussed tutorial.

A retrospective review was conducted of paediatric patients undergoing optic nerve evaluation using oFFA with images taken at 15min, 30 min, and 45 minute timepoints between 2019 and 2022. Four cardinal oFFA abnormalities were identified: Normal nerve appearance, Papilloedema, ON head drusen and, inflammatory papillitis. Test sensitivity and specificity was determined by a survey of consultant and trainee ophthalmologists without oFFA experience, who completed a blinded interpretation of 15 oFFA images with one of four diagnoses.

oFFA series from 30 eyes (13 children) were identified, age ranging from 6-15 years. Our protocol produced high-quality imaging while generating 0% adverse side-effects. Interpretation analysis revealed a diagnostic accuracy of 81.7% for consultants with a sensitivity of 76.8% and specificity of 87.5%. Qualitative and quantitative feedback supported the task as a useful exercise.

oFFA with UWF imaging are a safe and effective imaging adjunct in diagnostically challenging ON pathology. Moreover, skills in test interpretation can be rapidly developed using a focussed learning tutorial.

10.28 Pathways to detection of non-infectious childhood uveitis in the UK: Findings from the UNICORN cohort study - S Kellett (RA), H Petrushkin (C), J Ashworth (P), A Connor (C), E McLoone (C), C Schmoll (C), S Sharma (C), E Agorogiannis (C), J Williams (C), J Choi (C), A Injarie (C), N Puvanachandra (C), P Watts (C), A Shafi (C), E Millar (C), V Long (C), A Kumar (C), E Hughes (C), A Ritchie (C), J Gonzalez-Martin (C), A Pradeep (C), S Anwar (C), K Warrior (C), B Muthusamy (C), R Pilling (C), J Benzimra (C), A Reddy (C), K Bush (C), D Pharoah (C), K Falzon (C), U O'Colmain (C), R Knowles (C), V Tadic (C), A Dick (C), J Rahi (C), AL Solebo (C), University College London, Institute of Child Health

Prompt detection of childhood uveitis is key to minimising negative impact. From an internationally unique inception cohort, we report pathways to disease detection. UNICORNS is a national childhood non-infectious uveitis study with longitudinal collection of a standardised clinical dataset and patient reported outcomes. Descriptive analysis of baseline characteristics are reported.

Amongst 150 recruited children (51% female, 31% non-white ethnicity) age at detection ranged from 2–18yrs (median 10). In 69%, uveitis was diagnosed following onset of symptoms: time from first symptoms to uveitis detection ranged from 0-739days (median 7days), with longer time to detection for those presenting initially to their general

practitioner. Non symptomatic children were detected through JIA / other disease surveillance (16%), routine optometry review (5%) or child visual health screening (1%). Commonest underlying diagnoses at uveitis detection were JIA (17%), TINU (9%, higher than pre-pandemic reported UK disease frequency) and sarcoid (1%). 60% had no known systemic disease at uveitis detection. At disease detection, in at least one eye: 34% had structural complications (associated with greater time to detection – 17 days versus 4 days for uncomplicated presentation).

The larger relative proportions of children with non-JIA uveitis reported here increase the importance of improving awareness of childhood uveitis amongst the wider clinical communities. There is scope for improvement of pathways to detection. Forthcoming analysis on the full cohort (251 recruited to date across 33 hospitals and 4 nations) will provide nationally representative data on management and the determinants of visual and broader developmental/well-being outcomes.

10.37 Anterior Segment Optical Coherence Tomography Detects Cells in Children without Eye Disease - A Bellchambers (T), S Kellett (RA), A Fraser (T), A Patel (Op), A D Dick (P), J S Rahi (P), A L Solebo (C), University College London

OCT quantification of anterior chamber inflammation is highly sensitive. We describe findings for children without eye disease.

Scans (13-line raster, Heidelberg Anterior™) were acquired from schoolchildren aged 5-17 years. Data collected included sex, ethnicity, and iris. Images underwent manual analysis by at least two independent readers. Statistical analysis comprised description of the maximum per line, and median and total cell count (maxcc, medcc, tcc) detected, and regression analysis of cell counts and collected variables.

Of the 120 children for whom data have been analysed, 59 were female (49.2%), and 25.6%, 8.6%, 20.5%, 34.2% and 11.1% of children had blue, blue-green, hazel, brown and dark brown eyes respectively. Mean age was 11.3 (range 5-14years). At least one cell was detected in 199/240 (82.1%) eyes. Medcc ranged from 0-2 per eye (median 0), maxcc 0-7 (median 2), and tcc per eye 0 – 28 (median 4).

Older age was associated with higher counts (regression coefficient for tcc 0.718565, $p < 0.001$, 95% CI 0.51- 0.93) with a median tcc for children aged 5yrs of 2 cells, versus a median tcc for children aged 14yrs of 6 cells. Iris colour, ethnicity and gender were not associated with cell count.

Healthy children without eye disease have findings on AS-OCT that would normally be considered representative of inflammatory cells. This finding will have consequences for disease control targets in childhood uveitis, and future AS-OCT based screening for children at risk of uveitis. The association of age with cell count is worthy of further exploration.

10.46 Flying baby anterior segment OCT in the diagnosis of Anterior Segment Dysgenesis - H Jasim (CF), O El-Haddad (C), S Amarakoon (C), Bristol Eye Hospital

A one day old baby was referred to Paediatric Ophthalmologists with unilateral right-sided dense corneal clouding, noted at the 24 hour postnatal review. This baby was born at 38 weeks gestation via forceps instrumentation and subsequent emergency caesarean Section. The baby sustained a right sided periocular haematoma, lateral canthal superficial skin abrasion and subconjunctival haemorrhage. The obstetricians were concerned this baby

had also sustained ocular trauma secondary to forceps use, such as a Descemet membrane tear (FIDMT).

We were able to perform anterior segment OCT with the Osiris MS39 AS-OCT (TM) at 2 days of age to assess the anterior segment anatomy and were able to establish an intact Descemet membrane, corneal stromal oedema and iridocorneal adhesions consistent with a diagnosis of anterior segment dysgenesis.

This is the first published report of using the flying baby technique for anterior segment OCT and allowed for rapid diagnosis of a developmental condition and exclusion of FIDMT. This avoided any delays in referral for further treatments, avoidance of potential medico-legal problems and most importantly, reassurance to the parents of the cause of corneal clouding. *All the acquired images will be shown in this presentation.*

10.54 Peripapillary Hyperreflective Ovoid Mass-like Structures (PHOMS) in Children: Optical Coherence Tomography Measurements and Refractive Status - L Pratt (T), S Rehan (T), J West (T), P Watts (C), University Hospital of Wales, Cardiff

Peripapillary hyperreflective ovoid mass-like structures (PHOMS) are a recently described entity. They are a common and non-specific cause of pseudopapilloedema. We aim to determine if there is a relationship between optical coherence tomography (OCT) measurements and refractive status on the presence of PHOMS.

Retrospective analysis of optic nerve head OCT scans from children seen in the suspected papilloedema virtual clinic between August 2016 and March 2021 at University Hospital of Wales, Cardiff. Three assessors graded each scan for the presence of PHOMS. Numerical data on the disc morphology (disc area (DA (mm²)) and scleral canal diameter (SCD (μm)) was obtained from the OCT scans. Refractive data was obtained from the initial optometric referral where available. Logistic regression analysis was performed to assess the effect of age, sex, spherical equivalent, DA and SCD on the likelihood of the presence of PHOMS. The SCD was significantly larger in eyes with PHOMS (mean diameter 1771 μm) vs no PHOMS (mean diameter 1621 μm). Odds ratio 1.0042 (1.0016 to 1.0069). The other variables were not significantly associated, but there was a tendency towards a younger age, larger disc area and the presence of a refractive error if PHOMS were present. Anatomical and developmental differences in the size of the scleral canal and optic nerve may explain the presence of PHOMS in children. In contrast to other recently published studies, we show that a wider scleral canal diameter was significantly associated with the presence of PHOMS.

11.03 Analysis of ganglion cell thinning in paediatric papilloedema- at what point does visual function become affected? - A Malem (C), A Reginald (C), M Wan (C), The Hospital for Sick Children (SickKid) Toronto

We studied the relationship between macula ganglion cell-IPL thickness (GCT) and visual loss in children with papilloedema.

Retrospective, single-centre study of patients (<18 years old) presenting with papilloedema between 2012-2022. Presenting age, sex, lumbar puncture opening pressure and aetiology were recorded in addition to initial and final BCVA, disc rNFL and macula GCT. Visual fields were performed when possible. We defined vision loss as a final BCVA of 20/40 or worse or an abnormal visual field (abnormal Goldmann VF or >-3.00DB mean deviation on Humphrey VF)

80 patients (160 eyes) were included. 53(66%) were female with mean age 11.5 years (3-18). Mean follow up was 21.3 months. Aetiologies were primary PTSC (66/80) and secondary PTSC in (14/80). 22 eyes (13.7%) from 14 patients experienced visual loss. Mean final GCT was 73um vs 83um ($p<0.001$ - AUC 0.86) and minimum GCT 68um vs 81um ($p<0.001$ - AUC 0.89) in those with vision loss vs those without. A mean GCT $<70\mu\text{m}$ and min GCT $<68\mu\text{m}$ were 100% predictive of visual loss. Those with visual loss had a significantly higher LP opening pressure (41vs49cmH₂O $p=0.006$) and higher average rNFL (242vs187 $p=0.04$). No difference was found for age, sex or aetiology. In our data, GCT was significantly reduced in paediatric papilloedema patients experiencing persistent visual loss. The highest risk occurred when min GCT was $<68\mu\text{m}$. This information is important for patient management and counselling.

15.15 Session V (P)

Paediatric Rapid-fire presentations

Moderators: Mr Alan Mulvihill, Abu Dhabi and Miss Bhavini Gohil, London

15.17 Tear proteins in premature babies at risk of retinopathy of prematurity -C Shipton (T), J Aitken, S Atkinson, R Burchmore, R Hamilton (o), H Mactier (C), S McGill (O), E Millar (C), A C Houtman (C), Greater Glasgow and Clyde, Glasgow

This feasibility study aimed to investigate the feasibility of collecting and analysing tear proteins from preterm infants at risk of retinopathy of prematurity (ROP). Additionally, we sought to identify any tear proteins which might be implicated in the pathophysiology of ROP.

Eligible infants were those undergoing ROP screening without other ocular pathology. Tear samples were obtained by Schirmer's test strips coincident with routine ROP screening. Mass spectrometry was used for proteomic analysis. All participants' parents gave written, informed consent.

Samples were collected from 12 infants, including two sets of twins. Gestation ranged from 25+6 to 31+1 weeks. Median postnatal age at sampling was 30.5 days (range 19 to 66). One infant developed self-limiting ROP. An adequate sample for protein analysis was obtained from each infant. 701 proteins were identified; 261 proteins identified in the majority of tear samples, including several common tear proteins, were used for analyses.

Increased risk of ROP as determined by G-ROP prediction criteria was associated with an increase in lactate dehydrogenase B (LDH-B) chain protein in tears. Older, more mature infants demonstrated increased concentration of immunoglobulin complexes within their tear samples and two sets of twins in the cohort showed exceptionally similar proteomes, supporting validity of the analysis.

Tear sampling by Schirmer test strips and subsequent proteomic analysis in preterm infants is feasible. A larger study is required to investigate the potential use of tear proteomics in early identification of ROP.

Conflicts declared: Recipient of Ophthalmology Trainee Research Award, funded by NIHR/Santen

15.22 Leukemic infiltration of Optic nerves Leeds experience - A Kumar (C), K Jan (T), Leeds teaching Hospitals Trust

Relapses of leukaemia typically manifest in the central nervous system (CNS) or bone marrow, however isolated ocular relapse is rare and has been quoted as 2.2%. Although optic nerve infiltration in leukaemia is rare, it raises concern of central nervous system involvement. Therefore, early detection and prompt treatment are crucial in preventing vision loss as well as systemic morbidity and mortality.

Literature review of leukaemic optic nerve infiltration and a case series of three paediatric patients were selected to demonstrate the presentation, treatment course and outcomes of intraocular relapse of childhood leukaemia.

We report 3 patients with previously successfully treated systemic disease who presented with optic nerve infiltration. All 3 patients had visual symptoms and intraocular involvement with normal systemic MRI and lumbar punctures at presentation. One patient had bilateral nerve infiltration, and was successfully treated with systemic chemotherapy, followed by whole body irradiation and stem cell transplant. One patient presented with unilateral infiltration treated with systemic chemotherapy, and is awaiting whole body irradiation then stem cell transplant. One presented with unilateral optic nerve infiltration and was treated with systemic chemotherapy but subsequently had systemic disease and was managed palliatively then passed away.

Leukaemic optic nerve infiltration is rare, challenging to diagnose and manage. Currently, there is no gold standard for its diagnosis and management so treatment relies on previous case reports. We demonstrate the importance multidisciplinary approach to managing the vision loss, as well as their systemic condition and taking into consideration their long-term prognosis.

15.27 Restricted diet causing irreversible visual impairment in children with autistic spectrum disorder: case series and review - F Ghazala (T), R Hamilton (o), D Mansfield (C), E Millar (C), Greater Glasgow and Clyde, Glasgow

Though micronutrient deficiency is recognized to cause visual system dysfunction, avoidant/restrictive food intake disorder (ARFID) has been poorly described in relation to this.

Review of 18 previously published similar cases highlights the importance of identifying other micronutrient deficiencies, even when vitamin A deficiency accounts for the presenting features. We present four patients with permanent visual loss as a result of highly restricted diets due to avoidant/restrictive food intake disorder (ARFID), and with autistic spectrum disorder (ASD).

The four cases reported here make a total of 22 reported cases of visual impairment due to ARFID-like restricted diets in boys with ASD. The severity of ASD varied widely across the 22 cases, but all had extremely restricted diets, in some cases tolerating only one or two food items. The most avoided food groups in children with ASD and food selectivity have been reported from the USA as vegetables, fruit, dairy and protein, with the most preferred food items being bread, chicken, cereal and yoghurt. In the 22 cases reviewed or reported here, tolerated foods tended to be predominately carbohydrate based, with dry or crunchy textures and beige or pale colouring, i.e. French fries, potato waffles, potato chips (crisps), rice, white bread, bagels, biscuits or cookies.

This case series and review highlights the need for heightened vigilance for visual problems in individuals with ASD-related ARFID and early and complete assessment of micronutrient deficiency.

15.32 When the optometrist suspects papilloedema in an asymptomatic child on a routine visit, there can be no papilloedema...- or can there? - AC Houtman (C), C Ambrose (SAS), K Graham (o), R Hamilton (o), The Royal Hospital for Children Glasgow

Paediatric ophthalmology and neuro-imaging referrals increased following the Honey-Rose trial. A dedicated 'blurry disc clinic' was established at the Royal Hospital for Children Glasgow. The accuracy of community optometry referrals for suspected papilloedema in asymptomatic children on a routine visit was investigated.

Consecutive case series of optometry referrals between May 2020 and Oct 2022. Referrals were retrospectively scrutinized for the reason of the optometry consultation, the presence/absence of symptoms suggestive of raised intracranial pressure (ICP), viz. headache, transient visual obscurations, tinnitus and nausea/vomiting. Referrals without a clear reason for attendance, symptoms of raised ICP, or clearly stating 'routine visit', were deemed routine visits. Optometry visits spurred by suggestive symptoms, and re-referrals of known (pseudo)papilloedema were excluded. (Pseudo-)papilloedema was investigated by an algorithm developed at RHCG which included OCT (pRNFL measurement, disc volumetry, radial disc scans for qualitative assessment of presence of drusen or PHOMS and posterior pole GCL measurement), supplemented with US B-scan for ONH-drusen and US B-scan ONSD, LP and MRI-brain in cases suspected of true papilloedema.

143 out of 295 referrals were included. Among these, 2 cases were diagnosed with IIH; all other cases had anomalous disc morphology or optic nerve head drusen. No cases of CNS tumours or hydrocephalus were diagnosed. No cases have presented back with true papilloedema within six months of the final inclusion date.

Referral thresholds for suspected papilloedema in asymptomatic children on routine visits appear very low; the detection rate of true papilloedema in this cohort is extremely low.

15.37 Scoping review of Homonymous Hemianopia in Childhood - S E Handley (CRF), D A Thompson (C), A Liasis (C), R J Bowman (C), J S Rahi (P), Great Ormond Street Hospital for Children, London

Cerebral visual impairment (CVI) encompasses a heterogeneous group of disorders and a spectrum of types of visual impairments. Research is needed to characterise the different forms of CVI and identify the specific needs of these groups to inform individualised patient care. Homonymous hemianopia (HH) is a definable visual field defect that affect some children with CVI. As part of a new research programme, we conducted a scoping review of the literature on HH in children and young people to map current knowledge and identify evidence gaps.

We used the PRISMA extension for Scoping Reviews methodology. Multiple online databases were searched using terms associated with "homonymous hemianopia" and "children". This yielded 1588 papers which were screened by two reviewers. Of these 1001 were excluded at abstract screen and a further 415 excluded after full text review, with full text unavailable for 15. Data were extracted and charted from 157 studies and additional grey literature.

Interim analysis shows reported studies are predominantly from high income countries with a paucity of higher-level evidence, and a preponderance of case reports. Most papers reported causative pathology and diagnosis of HH. There was minimal attention to or evidence relating to intervention. Child-specific grey literature on HH was limited. This review collates the current evidence-base for HH in children. It demonstrates the important evidence-gap relating to intervention in these cases that would help inform more individualised care. Similar scoping reviews may be prove useful in assessing the evidence relating to other definable groups within the CVI umbrella.

15.42 Optic Disc Swelling in children - a case series - S Parvizi (C), Surrey and Sussex Hospitals NHS Trust

Optic disc swelling in children is a common referral from the community. The concern of this finding is the likely association with raised intracranial pressure of which the causes can be numerous, with potentially devastating consequences.

In this case series of children presenting in a 2-year period between January 2021 and December 2022 to the eye clinic at East Surrey Hospital in Redhill, UK, red flags in the history, appearance of the optic disc on OCT and B-scan ultrasonography and any subsequent investigations including CT and/or MRI and lumbar puncture to determine the sensitivity of our tests in identifying children with true disc swelling who require intervention were assessed.

The analysed subset of 38 children had a referral to the eye clinic with either one or both optic discs appearing swollen, who after a full ophthalmological examination subsequently underwent OCT. Where these findings were equivocal, in addition to the aforementioned they underwent B-scan ultrasonography to determine any supporting diagnostic features. Under one-third of the patients (12/38) had findings of dilated optic nerve sheaths at the point of ultrasound, but of those half (6/38) were then found to have normal intracranial pressure as determined by lumbar puncture.

These findings illustrate the limitations of our investigative tests and importantly the diagnosis of true papilloedema being heavily reliant on a multidisciplinary assessment in order to reduce the risk of over investigation.

15.47 Unexpected retinopathy in a patient presenting with bilateral optic disc swelling - G Kiray (CF), V Panteli (C), N Enright (C), S Handley (o), O Marmoy (o), D A Thompson (C), R Henderson (C), Great Ormond Street Hospital for Children, London

A 12-year-old boy presented with 5 day history of blurry vision, “wobbly eyes”, tinnitus and difficulty seeing at night. Local ophthalmology noted bilateral optic disc swelling and referred him urgently for neurological investigations.

At presentation VA was RE 0.00 and LE 0.2 with normal Ishihara colour vision. His extraocular movements were full without manifest strabismus. Fundoscopy showed bilateral optic disc swelling. Electrophysiology unexpectedly revealed a functionally cone isolated retina with markedly abnormal rod function. Pattern VEPs indicated bilateral macular pathway dysfunction affecting left eye more than right eye. Wide field imaging showed bilateral diffusely scattered yellow-white flecks in the midperiphery of each eye. His kinetic visual fields were moderately restricted bilaterally. MRI showed a Chiari 1 malformation with cerebellar tonsil herniation, but LP opening pressure was normal.

Differential diagnosis included RDH5 retinopathy or vitamin A deficiency. On questioning he reported a diet restricted to only meat and biscuits. His vitamin A levels were subnormal at 0.14 umol/L (reference range 0.9-2.5umol/l) and he was started on high-dose Vitamin A supplements.

Four months after supplementation retinal appearances had normalised, the rod ERGs recovered, nyctalopia and visual field restriction resolved. PVEPs had improved but an element of LE macular pathway dysfunction remained. Optic disc swelling settled leaving mild temporal pallor, particularly of the LE with some RNFL loss.

It is important to recognise nutritional Vitamin A deficiency in children as prompt recognition and treatment can improve symptoms, reverse retinal pathology which we have demonstrated with electrophysiological findings.

15.52 x-linked retinoschisis: implications of visual impairment in childhood - S Balakrishnan (T), S Jain (C), Lancashire Teaching Hospitals NHS Trust

6 out of every 10,000 children in the UK are visually impaired or blind. 50% of these children have additional educational needs or learning disability (LD). Visual impairment (VI) impacts development of cognitive, motor and social skills and can mimic or exacerbate behavioural disorders or LD. This proves prompt recognition and diagnosis challenging. We report a 15-year-old male with reduced vision secondary to x-linked retinoschisis presenting with behavioural difficulty and LD. Prior to our clinic visit, he was undiagnosed and hence had not received any support for his VI despite regular visits to the opticians for several years.

His best corrected visual acuity was 0.44LogMAR and 0.76LogMAR in right and left eye respectively. Examination showed macula cystic changes and concentric ring of drusen-like deposits in the peripheral retina corresponding to fundus autofluorescence (FAF) findings. OCT showed macular schitic cavities. FAF showed irregular hypo-fluorescent central lesions with surrounding hyper-fluorescent concentric ring.

Following diagnosis, he accessed low vision clinic and was given aids which helped improve his academic performance and headaches. A supporting letter to school allowed implementation of significant educational support. Correction of refractive error by our optometrists provided some improvement of vision. He was also referred to genetics team for counselling. At 3-month follow-up, the patient and family report significant improvements in behaviour, concentration, and school performance.

Our case illustrates the importance of considering VI as a cause or exacerbating factor of behavioural disorders and LD. Prompt diagnosis and management can minimise adverse effects of VI on quality of life.

15.57 Trochlea, Trochlea, Trochlea - K Reed (T), A Natha (O), L Brocklesby (O), R Dahir (O), A Khaier (C), M Posner (C), Queen's Hospital, Romford

We present a case series of unusual trochlear pathology in young males.

Three patients presenting to Queens Hospital Eye department across 2022-2023 with trochlear pathology.

Firstly an acquired Brown's precipitated by prolonged up-gaze during a triathlon.

Resolution achieved with oral anti-inflammatories alone.

Secondly an acquired Brown's immediately following a heavy cocaine binge in a habitual user, recurrent sinusitis and rhinitis. Raised inflammatory and connective tissue markers found raising suspicion of SLE. No improvement with medical therapy.

Thirdly a case of trauma to the orbit resulting in unilateral superior oblique palsy and Brown's a few weeks prior to patient's wedding. A degree of spontaneous resolution, we discuss further management.

With excellent MRI imaging obtained we give pointers on imaging technique and modality. Notably with the case of prolonged up-gaze we report a treatment regime of oral anti-inflammatory which rapidly diminished symptoms within 4 weeks, of which discussion very little exists in peer review.

16.02 MOG associated encephalitis presenting as Idiopathic Intracranial Hypertension - V Panteli (C), A Alwis (C), N Desai (C), O Marmoy (o), D A Thompson (C), P Prabhakar (C), Great Ormond Street Hospital for Children, London

A young Caucasian male (7y) with normal BMI was atypical for his provisional diagnosis of Idiopathic Intracranial Hypertension (IIH), that resolved following a Lumbar Puncture (LP). At 8y he presented with a 2-week history of headaches and vomiting that started some weeks after flu vaccination and an upper respiratory infection.

Visual Acuity (VA) and colour vision were normal. Ocular motility was full. Fundoscopy and OCT showed recurrence of papilloedema, with enlarged blind spots on Kinetic perimetry. LP opening pressure was 30cm H₂O and CSF white cells were elevated (23). Repeat brain and spine imaging showed new white matter signal changes in keeping with neuroinflammation, as well as enhancement of the left optic nerve extending to the chiasm and optic tract. VA, colour vision and pupillary reactions remained normal.

Pattern VEP peak times were prolonged from the left eye compared to right eye to small check widths, consistent with relative macular-cortex pathway dysfunction. Hemifield PVEPs were slightly prolonged and reduced from the bitemporal fields indicating chiasmal dysfunction. Normal PERGs excluded PVEP delay associated with primary RGC disease. Further investigations showed oligoclonal band and serum-MOG antibody positivity.

Initial treatment with Acetazolamide 125mg bd for a week, following LP, was changed to IV methylprednisolone followed by oral prednisolone.

Symptoms improved significantly following LP and steroid treatment. He will be followed in a Demyelination Clinic.

MOG-associated disease has been reported with raised intracranial pressure and should be considered especially in children with atypical clinical phenotype for IIH.

16.07 Rapidly progressive chorioretinal atrophy following CAR-T cell treatment for Leukaemia - E De Smit (CF), S Burrige (N), S Ghorashian (C), R Bowman (C), H Petrushkin (C), R Henderson (C), Great Ormond Street Hospital for Children, London

We present the case of an 11 year old girl with acute lymphoblastic leukaemia (ALL). Despite treatment, she sustained multiple bone marrow relapses. Rescue therapy with Chimeric antigen receptor (CAR) T cells was performed. CAR-T therapy is a personalised immunotherapy where a patient's own T cells are genetically altered to recognise leukaemic antigen.

Six days after CAR-T infusion, she developed sudden bilateral visual loss and florid optic nerve swelling. She was treated with orbital floor steroid injections and her optic nerve appearance and vision improved. Within four weeks, she developed rapidly progressing chorioretinal atrophy in the right eye. A vitreous biopsy confirmed intraocular leukemia. Subsequent retinal biopsy demonstrated both ALL blast cells and CAR-T cells. She was treated with four 2-weekly intravitreal methotrexate injections. Systemic investigations showed no evidence of disease elsewhere. To date there has been no evidence of relapse.

This case demonstrates that CAR-T therapy can reveal intraocular ALL in patients who previously were asymptomatic. Despite being an immune privileged site, CAR-T cells can traffic to the eye. The literature on ocular complications post CAR-T treatment is limited. A handful of published case reports have described ocular ALL highlighted through CAR-T treatment but none with such extensive chorioretinal destruction.

We suggest that if a patient develops ocular symptoms or signs following CAR-T treatment, they be investigated urgently for local and systemic disease. In other reports, enucleation has been performed for isolated ocular ALL to prevent systemic spread. In this case, intraocular chemotherapy halted progression of ocular disease.

THURSDAY 05th OCTOBER 2023

12.00 Session III (P) (S)

Free Papers

Moderators: Una O'Colmain, Dundee and Clare Roberts, London

12.02 Medial Rectus Re-advancement: More bang for your buck? - K Naderi (T), N Quah (T), M Adamowicz (MS), S.Jain (C), Royal Free Hospital

Surgical management options of exotropia include medial rectus resection-lateral rectus recession surgery, as well as medial rectus re-advancement (MRR) in cases of consecutive exotropia. We compared the surgical outcomes of primary medial rectus resection-lateral recess surgery, to MRR in patients with consecutive exotropia.

Retrospective, electronic note review of consecutive patients undergoing primary recess-resect (RR) surgery for basic exotropia and divergence excess, and consecutive patients undergoing medial rectus re-advancement (MRR) +/- lateral rectus recession for consecutive exotropia in a teaching university hospital.

There were 88 patients in the RR group and 32 patients in the MRR group. The median age in the RR group was 29.50 years (range 7-83) and 42 years (10-87) in the MRR group ($p=0.077$). Median follow-up was 10 months (1-35) in the RR group and 1 month (1-11) in the MRR group. Post-operatively, there was a mean exotropia reduction of 24.93 Prism Dioptres (PD) (SD 14.67, $p<0.0001$) for near, and 22.72 PD (14.53, $p<0.0001$) for distance in the RR group. In the MRR group the mean exotropia reduction was 31.42 PD (19.43, $p<0.0001$) for near and 31.48 PD (14.53, $p<0.0001$) for distance. There was a greater reduction in the pre-operative exotropia in the MRR group compared to the RR group for distance ($p=0.036$), but did not meet statistical significance for near ($p=0.057$).

Based on our study cohort, MRR appears to give a greater effect for distance correction compared to RR surgery.

12.11 Prism Adaptation Testing in Children to improve Surgical Outcomes in Esotropia Surgery - T Liu (T), J Smedley (O), A Maudgil (C), Sheffield Children's Hospital NHS Foundation Trust

The aim of strabismus surgery is to improve alignment and ideally restore binocular vision. Prism adaptation (PAT) in esotropia is known to improve post-operative outcomes by allowing choice of target angle in line with pre-operative fusion potential. Reaching a successful end-point in PAT requires co-operation, which is not a given in the paediatric population. The purpose of this study is to describe the successful use of prism adaptation in children in planning target angles for esotropia surgery.

Retrospective review of 29 cases from a tertiary centre was conducted, including cases who underwent PAT prior to esotropia surgery over a 6-year period (Jan 2016- Jan 2023). Data was collected from the electronic patient record including patient age, diagnosis, measurements of presenting and prism adapted angles, surgery performed and surgical outcomes.

Successful prism adaptation was carried out in children of ages 3-16 . The average presenting angle of esodeviation was 20.9 PD in the distance and 27.3 PD at near. The average maximum prism adapted angle, which was the target angle used for surgery, was 35.3 PD (range 25-50PD). All patients underwent bimedial recessions except one, a re-do

who underwent LR re-advancement. 3 patients had one LR resection in addition (3/28). 89.7% of patients achieved binocular vision with stereopsis, 3.4% achieved binocular vision with simultaneous perception. 2 patients had no demonstrable BV, neither were overcorrected post-operatively.

Pre-operative PAT is an effective tool in pre-operative assessment of paediatric esotropia patients with binocular potential and can be used to optimise surgical planning and outcomes.

12.20 Convergence Insufficiency: Are we making a difference in patients' lives or is it a Waste of Time! - J L Z Jong (MS), Z Saleem (O), J Simmons (O), M Rhodes (O), J L Choi (C), Sheffield Teaching Hospital NHS Trust

Convergence insufficiency (CI) is a common condition that can impair visual performance and comfort during close visual work. This prospective study evaluated the effectiveness of interventions on clinical outcomes and quality-of-life using the adult strabismus quality-of-life questionnaire (AS20) in patients with CI.

Data was extracted from a database collected at first consultation from 2015 to 2022.

Demographics, interventions and outcomes of 84 patients with CI (mean age 47.0±24.9 years) were analysed.

Orthoptic exercises were prescribed to 56% of patients, 32% received prisms, 15% received no treatment, with 3 discharged on the same day. At latest follow-up review, 22.6% were recommended to continue exercises, 28.6% had prisms, 1 underwent bimedial resection and 2 had botox. The median follow-up was 5.5(5.0-55)months, 88.1% were discharged with 29.8% following failure to attend and 9.5% deceased. Near-point of convergence (NPC) improved from a median of 15(6-50)cm to 10(6-30)cm. The median AS20 score at presentation were 100(30-100) and 47.5(0-100), and post-intervention were 100(80-100) and 77.5(12.5-97.5) for psychosocial and functional components, respectively.

At the latest follow-up, the attendance failure rate was higher for exercises (36%) than for prisms (15%). Improvement was noted in NPC (33%) and mean AS20 scores was 9% higher psychosocially and 32.8% functionally, highlighting the benefits of intervention on patients' quality-of-life.

This cohort provides valuable insights into the clinical management of CI, as evidenced by improvements in NPC and AS20 scores. However, the study also found that long-term compliance with treatment is intrinsically challenging, emphasising the importance of disease education.

12.29 The Incidence, Clinical features, and Management of Essential Infantile Esotropia in the United Kingdom. A British Ophthalmology Surveillance Unit (BOSU) study – Final Report - P Watts (C), D Yeo (C), R Davis (C), W J Watkins (o), University Hospital of Wales

Studies from the UK have reported declining rates of surgery for childhood esotropia. It is not known if this equates to a reduced incidence of essential infantile esotropia (EIE). A national study was undertaken through the British ophthalmology surveillance unit (BOSU) to determine the incidence presenting features and management of EIE in the UK

Data from a prospective national observational cohort of newly diagnosed EIE presenting to clinicians in the United Kingdom over a 12-month period was collected. Cases with a

confirmed diagnosis by a clinician of a constant, non-accommodative esotropia ≥ 20 prism dioptres (PD), presenting at ≤ 12 months, with no neurological or ocular abnormalities were identified through BOSU. Follow up data was collected at 12 months. Data was collected on the age, gender, ethnicity, birth history, age at diagnosis, age at intervention, angle of esotropia, refraction, associated features of amblyopia, overelevation in adduction (OEIA), latent nystagmus and dissociated vertical deviation (DVD), method of management and outcomes.

During the period of observation between October 2017 to October 2018 a total of 57 cases were reported giving an incidence of EIE of 1 in 12,828 live births with a corrected incidence of 1 in 9027 live births allowing for estimated under reporting. The mean age of diagnosis and intervention were 7.05 ± 2.6 months (range 2 to 12 months) and 14.7 ± 4.9 (range 6.5-28.1 months) respectively. The majority were Caucasians 86.5% and 52.7% were female. Management was surgical in 59.6%, and botulinum toxin alone in 22.8%, 17.5% were observed. There was no significant difference in the age of presentation ($P=0.6$), gender ($P=0.8$), prematurity ($P=0.5$), deprivation indices ($P=0.68$), refraction ($P=0.7$), OEIA ($P=0.6$), DVD ($P=0.7$) or follow up ($P=0.3$) between the three groups. The preoperative angle of esotropia was smaller in the observation group ($P=0.04$). The post-operative angle of esotropia was not statistically significant between botulinum toxin or surgery ($P=0.3$) though the age of intervention was earlier in the botulinum group ($P=0.007$). Early intervention did not influence the motor post intervention outcomes between 0-10 prism dioptres of esotropia ($P=0.78$). Amblyopia ($P=0.02$) and latent nystagmus ($P=0.009$) was more common in the observation group.

The incidence of EIE in the UK is considerably lower than reported in other population-based studies. The preferred method of treatment was surgical with earlier intervention in those treated with botulinum toxin. An early age of intervention did not influence motor outcomes. Parental choice and amblyopia treatment were reasons cited for conservative management in the observational group.

12.38 The Global Retinoblastoma Outcome Study: Retinoblastoma Outcomes in Europe: a prospective, cluster-based analysis of 483 patients from 35 countries - R Bowman (C), D MacLeod (P), D Fabian (C) on behalf of global Rb study group, London School of Hygiene and Tropical Medicine, London

To describe presentation, treatment and outcomes for children presenting with retinoblastoma (Rb) throughout Europe during the year 2017.

Prospective analysis of patients diagnosed in Europe during 2017, then followed up for 3 years.

483 patients were recruited from centres in 35 countries within Europe (mean age 22 months, 0-128 months). 264/483 (55%) were male. 151 (31%) patients were from middle income countries and 332 (69%) from high income countries.

339/483 (70%) were unilateral and presented older (mean age 26 months) than the 144 (30%) bilateral cases (mean age 12 months $p < 0.0005$). Only 4/477 (0.8%) children had extra-ocular Rb at presentation (mean age 53 months vs 21 months for those without $p=0.002$). Middle income children did not present older but were more likely to present with cT3 than cT2 compared to high income children (RR 1.25 95%CI 1.08-1.44). Overall, 58% of children underwent enucleation over the three years, 36% of which as primary

treatment. Risk of enucleation was determined by disease stage and laterality but not socio-economic status.

12 (2.5%) children died from Rb. 3 of 4 (75%) of children with extra-ocular tumour at presentation died from Rb compared to 9/449 (2%) who had intra-ocular tumour (OR=146.7 13.9-1549.4, $p<0.0005$). 11/132 (8%) children from middle income countries died from Rb compared to 1/327 (0.3%) in high income countries (OR=29.8 3.8-232.0, $p<0.0005$).

Even within a wealthy continent such as Europe, socio-economic factors influence survival but not globe salvage rates. The majority of children still lose an eye.

16.00 Session VI (P) (S)

Rapid Fire presentations

Moderators: Fiona Rowe, Liverpool and Naomi Tan, London

16.02 Squint hook down: A tale of homeopathic strabismus surgery - E Cabourne (C), Kingston Hospital

A 6 year old boy was expected to undergo strabismus surgery for a symptomatic decompensating fully accommodative esotropia. He reported increasingly troublesome double vision.

With low hypermetropic correction, his visual acuity was 0.02 right eye and 0.04 left eye, his stereoacuity 85 seconds of arc, he had an 18 PD near esophoria and 8PD distance esophoria. Uncorrected, his visual acuity was 0.12 in both eyes with a 30 PD right esotropia at near and 20 PD at distance.

Whilst considering squint surgery, his mother, who is studying homeopathy, initiated treatment with an individualised homeopathic remedy of 30c nitric acid administered once a day on a sugar-coated dissolvable tablet. He completed an initial one-week course with success, although the effect was short lived with diplopia returning after one week. He proceeded with an additional two-week course which allegedly improved his symptoms and ocular alignment.

One month following this self-medicated treatment, our patient attended his scheduled outpatient review, to our surprise without diplopia and without spectacle correction. His unaided visual acuity was 0.04 in both eyes and he controlled a 20 PD esophoria for both near and distance fixation. Strabismus surgery was therefore postponed and active monitoring has resumed. At two months, Mum reports he remains asymptomatic. This is the first case described where individualised homeopathic treatment has demonstrated an apparent resolution of a fully accommodative esotropia. The longevity is yet to be determined but as homeopathy becomes ever more popular, Paediatric Ophthalmologists may require some basic awareness of such fascinating cases.

16.07 Surgical treatment of Heavy Eye Syndrome by Modified Loop Myopexy - A Agrawal (SAS), VSY Geh (C), Southend University Hospital

Heavy eye syndrome or convergent strabismus fixus is an acquired strabismus typically seen in eyes with high myopia. We present a case, discuss the aetiology and management, and include a short video illustrating the surgical procedure undertaken.

A 47-year-old highly myopic woman with h/o bilateral cataract surgery and B/L scleral buckling for retinal detachments, had left esotropia and hypotropia measuring more than

40 prism dioptres base-out and 12 prism dioptres base up. MRI orbits showed bilateral asymmetrical medial deviation of ocular bulbs, more on left side. Also, there was degeneration of lateral rectus-superior rectus band with displacement of lateral rectus downwards.

She underwent Botox to bi-medial recti after which she could demonstrate potential for binocular single vision. A left un-augmented loop Myopexy procedure and recession of the left medial rectus was thereafter performed under general anaesthesia.

After surgery, her eyes were binocularly aligned for near with minimal esotropia for distance.

This case suggests that patients with significant esotropia combined with high myopia should be suspected to have heavy eye syndrome. Orbital imaging should be undertaken to demonstrate the anatomical abnormality and muscle paths to confirm a definite diagnosis. Modified Loop Myopexy was found to be effective in this case of heavy eye syndrome.

16.12 Retinopathy in patients with mucopolysaccharidosis - M Noor (CRF), O McGrath (CRF), N Parry (P), T Aslam (P), J Ashworth (P), Manchester Royal Eye Hospital

The mucopolysaccharidoses are a group of inherited metabolic disorders resulting in abnormal degradation of glycosaminoglycans within lysosomes. Ophthalmic manifestations resulting in visual loss include corneal clouding, optic neuropathy and raised intraocular pressure, and retinopathy which occurs in MPS type I, II, III, IV. While corneal clouding may be stabilised with early treatment with HSCT or surgically treated with a corneal transplant, there is currently no known effective treatment for retinopathy.

We conducted a prospective observational study of patients with MPS who underwent fundus examination, OPTOS imaging, OCT, and electroretinography.

76 patients with MPS were studied, comprised of 45 MPSI, 9 MPSII, 13 MPSIV and 9 MPSVI patients. The age range was 3- 58 years of age. OPTOS imaging was performed in 65 individuals, OCT in 61, and electrodiagnostic assessments in 37 patients. Ten patients (7 MPSI, 3 MPS II) had fundoscopic signs of retinopathy, of which 5 had abnormal ERGs. Twenty one patients (17 MPSI, 2 MPSII, 2 MPSVI) had abnormalities on ERG, of which 5 had concurrent fundoscopic evidence of retinopathy. The onset of retinopathy in MPS patients was observed over a broad age range, with initial detection occurring between 2 and 53 years of age.

Retinopathy can be diagnosed on examination, imaging, or ERG in MPS patients as young as 2 years of age. The development of novel medicines, such as gene therapy, have potential to stabilise or improve retinopathy in the future. Therefore, phenotypic and natural history information pertaining to retinopathy in MPS is extremely valuable.

16.17 Reception age vision screening in Gloucestershire during the Covid Pandemic - R Hunn (O), Gloucestershire Hospitals NHS Trust

The UK NSC recommends that screening of children's vision should be offered to all children aged 4-5 years. Gloucestershire offers an Orthoptic-led reception age vision screening program to all children in mainstream local authority schools.

Screening is undertaken by Health and Wellbeing assistants and is an opt-out service. All children who record a vision of 0.225 logMAR or worse, or are unable to complete screening on the day, are referred to a diagnostic pathway. Children who record a visual acuity of 0.325 logMAR or less, or who are unable to complete screening on the day, are

referred to the HES. Children with VA of 0.225-0.3 are referred to an accredited High Street Optometrist. Children who are referred to the HES are seen within a one-stop combined Orthoptic and Optometry clinic within 6 weeks of referral.

In the academic year 2020-21, 6251 children were screened representing a coverage of 99.3%. This level of coverage is consistent with that achieved in all academic years since 2013/2014.

1026 children were referred following screening (16%) with 412 children fulfilling the criteria to be referred to the HES. Presenting visual acuities ranged from -0.1 to NPL. Refractive errors ranged from -8.50DS to +10.50DS with cylindrical errors up to +6DC. 77% of children were prescribed glasses at their first visit. The false positive rate for children with referring vision of less than 0.325 logMAR was 15%.

Children were managed by a combined Orthoptic and Optometry team with one children referred to the Paediatric Ophthalmologist.

16.22 Foveal hypoplasia in CRB1 associated retinal dystrophies - AC Rodriguez (F), B Higgins (o), V Tailor (T), S Malka (o), AM Collins (N), R Henderson (C), M Moosajee (P), Moorfields Eye Hospital

Foveal hypoplasia (FH) describes the underdevelopment of the fovea and can be classified based on a structural OCT grading scale (1 to 4). Although FH has been described in many ocular conditions and linked to visual acuity, it has not yet been reported in CRB1 retinopathies. The CRB1 gene plays a crucial role in retinal development and can result in a range of phenotypes such as early onset retinal dystrophy (EORD), retinitis pigmentosa (RP), cone-rod dystrophy and macular dystrophy (MD). Studies in CRB1 retinopathies have shown thickening and coarse lamination of retinal layers resembling an immature retina. Here, we report the grades of FH seen in a CRB1 cohort.

We identified 26 patients with molecularly confirmed biallelic CRB1 variants from Moorfields Eye Hospital and GOSH. Demographic and clinical data including best-corrected visual acuity (BCVA) and OCT (grading determined by two experienced ophthalmologists) was collected.

FH was identified in 11 (42%) patients (7 with EORD, 1 with cone-rod dystrophy and 3 with macular dystrophy), out of which 8 (72%) were classified as grade 1a and 3 (27%) as grade 1b. The most common variant found was c.2843G>A (p.Cys948Tyr), present in 6 (23%), but with no correlation with FH ($p=0.22$). Mean BCVA was 0.9 LogMar SD ± 0.13 in the FH group versus 0.9 LogMAR SD ± 0.17 in those without. No significant association between the presence of FH and BCVA ($p=1$) was found.

FH is seen in CRB1-retinopathies suggesting this gene is involved in foveal development, but mutations do not impact BCVA in this cohort.

16.27 Horner Syndrome: Can it be Familial? Case series in a family and review of literature - S Goyal (FD), S Verma (SAS), R Ranjan (C), R Goyal (C), Royal Glamorgan Hospital

Ophthalmic literature reveals vague and rare references to Horner syndrome on a hereditary basis. We present a case series of mother and son with Horner syndrome, which was confirmed pharmacologically. They noticed symptoms on the same side at a similar age and no serious pathology was found.

Retrospective case review of notes:

Case 1: An 11-year-old male presented with 6 week history of anisocoria, mild right ptosis, no heterochromia and no history of trauma or previous surgeries. The anisocoria was more noticeable in the dark, Horner syndrome was confirmed with apraclonidine test.

Case 2: Mother of case 1, 50-year-old female diagnosed with right Horner syndrome at the age of 14 in Austria. The presenting features were anisocoria, a lack of sweating on the right side of her face. Diagnosis was reconfirmed pharmacologically.

Case 1 was referred to paediatrics for a systemic examination which was normal. He was investigated with urinary catecholamines, MRI head and CT neck and thorax which were all normal. Case 2 was investigated in the past with a normal CT head.

Horner syndrome results in the interruption of the oculo-sympathetic pathway and can indicate serious pathology in the head, chest or neck. Our cases demonstrate that familial presentation could indicate an idiopathic aetiology as it is unlikely to have pathological Horner syndrome in two first degree relatives.

Our case series highlights the importance of eliciting a family history of Horner syndrome and examining the family members. Positive family history can reassure patients while awaiting results of investigations.

16.32 Impaired vision in children prenatally exposed to methadone: an observational cohort study - R Hamilton (o), A Mulvihill (C), L Butler (T), A Chow (RF), E Irving (P), D L McCulloch (P), A. McNeil (O), K. Michael (C), K M Spowart (C), J Waterson-Wilson (O), H Mactier (C), Royal Hospital for Children, Glasgow

Given previous observations of visual problems in children exposed to methadone in utero, we aimed to examine the prevalence of failed visual assessment at 8–10 years in children born to methadone-maintained opioid dependent (MMOD) mothers and relate this to known in utero substance exposure.

Follow up of observational cohort study of methadone-exposed and comparison children matched for birthweight, gestation and postcode of residence at birth. Participants were 144 children (98 exposed, 46 comparison). Prenatal drug exposure was previously established via comprehensive maternal and neonatal toxicology. Children were invited to attend for visual assessment and casenotes were reviewed. Presence of acuity poorer than 0.2 logMAR, squint, nystagmus and/or impaired stereovision constituted a 'fail'. Visual assessment pass/fail was compared between methadone-exposed and comparison children after adjusting for known confounding variables.

33 children attended in person: data were also derived from casenote review for all children. After controlling for maternal reported tobacco use, methadone-exposed children were more likely to have a visual 'fail' outcome, adjusted odds ratio 2.6, 95% CI 1.1–6.2; adjusted relative risk 1.8 (95% CI 1.1–3.4). Visual 'fail' outcome rates did not differ between methadone-exposed children who had (n=47) or had not (n=51) received pharmacological treatment for neonatal opioid withdrawal syndrome (NOWS); fail rate 62% vs 53% (95% CI of difference -11–27%).

Children born to MMOD mothers are almost twice as likely as unexposed peers to have significant visual abnormalities at primary school age.

Prenatal methadone exposure should be considered in the differential diagnosis of nystagmus. Findings support visual assessment prior to school entry for all children with a history of prenatal opioid exposure and may infer a teratogenic effect of methadone.

16.37 Development of a Questionnaire to study Fear and Anxiety Factors affecting Patients and their Families undergoing Strabismus Surgery - G Hogg (MS), S Joshi (MS), H Mason (MS), C O’Byrne (T), S Jain (C), Royal Free Hospital

The aim of this study was to develop a questionnaire to identify perioperative fear and anxiety factors affecting pediatric strabismus surgery patients.

First, we reviewed the literature to determine precipitants of fears and anxieties experienced by pediatric patients. Subsequently, we developed a questionnaire for pediatric patients undergoing strabismus surgery. This was a two part questionnaire, consisting of a 16-piece section for patients and a 22-piece section for parents. Finally, we piloted this questionnaire to validate its clinical use.

Common anxiety factors for children include pain, minor clinical procedures requiring needles, separation from parents and engaging with medical professionals. We used this information to develop a two part questionnaire for patients and parents. The questionnaire elicited positive and negative aspects of the patient journey, corroborated fears reported in the literature, and identified anxiety inducing factors specific to strabismus patients.

There is a lack of evidence regarding fear and anxiety specific to pediatric ophthalmology surgeries. Strabismus surgery carries unique fear inducing factors. Interventions which may alleviate the stress of pediatric surgery, therefore greatly benefit patient experience and surgical outcomes, and should be considered in the care of pediatric patients. Patient educational material is known to provide a sense of control to patients, helping to alleviate such fear.

Evidenced by the literature and the pilot questionnaire, there still exists anxiety inducing factors in pediatric surgery. Investigation into patient fears regarding pediatric strabismus surgery is needed to better understand how clinical staff can support patients perioperatively.

16.42 Engaging with young people to improve research, services and workforce development: eye-YPAG and “visually” workshops - V Taylor (RO), LM Brady (RF), J Miller (N), L Bays (o), J Zane (o), G Nagel (o), H Baker (o), R Crosby-Nwaobi (RF), A Dahlmann-Noor (C), Moorfields Eye Hospital

Involving children and young people (CYP) in service and research design improves quality and accessibility. Running events in schools to invite CYP to volunteer and explore careers in the NHS may contribute to uptake of training posts and developing the NHS workforce.

Here we evaluate two activities with CYP, our Young Person’s Advisory Group for research (eye-YPAG) and our workshop for secondary schools, “visually”.

We evaluated eye-YPAG in focus groups and online surveys with group members, parents/carers, researchers, facilitators and funders. We conducted thematic analysis and descriptive statistics. To evaluate “visually”, we monitored the numbers of workshops and young people applying for volunteering roles. We asked those who started working with us about their experience.

eye-YPAG members valued social and creative aspects as well as learning about research and developing skills and confidence. Researchers reported that CYP gave novel suggestions, modifying research plans, and that their different perspective was helpful in making research more relevant for children and families.

Over 6 months, we held 15 “visually” workshops in secondary schools. Ninety students applied for volunteering roles, and 20 have completed the Human Resources onboarding process. Young volunteers report that this work has increased their confidence and that they have gained insights into how a hospital works. One is considering training to become an orthoptist.

Both eye-YPAG and “visually” are available to all eye researchers and units in the UK and can facilitate outreach activities.

16.47 Unilateral lateral rectus resection as preferred procedure to treat patients with acquired distance esotropia - W Ghaffari (FD), N Mansoor-Ali (T), R Sadia (C), A Maino (C), Manchester Royal Eye Hospital

To report the results of a series of patients with acquired distance esotropia (ET) who underwent lateral rectus resection.

We retrospectively analysed data from 21 symptomatic patients with acquired esotropia who did not tolerate prisms. Twelve patients had myopic esotropia, 5 patients decompensated esophoria and 4 patients age-related distance esotropia. Patients were divided in two groups; myopic and non myopic. Two patients in each group had undergone surgery to medial recti muscles previously. Near and distance angles were measured over 2 visits. Data were analysed with t-test (paired and unpaired).

Average age was 47 (myopes), 77 (distance ET) and 26 years (esophoria). Mean spherical equivalent was -4.00 DS (myopes) and + 1 SD (non myopes). Lateral rectus resection ranged between 6 and 8 mm with an average of 7 mm in the myopic group and between 4 and 8 mm with an average of 6 mm in the non-myopic group.

The distance angle was reduced from 19 PD to 6 PD in myopes (mean difference 12 PD, $p < 0.0001$) and from 19 PD to 8 PD in non myopes (mean difference 12 PD, $p = 0.0011$).

There was no difference in reduction of distance angle between myopes and non-myopes ($p = 0.771$). All patients had complete resolution of diplopia after surgery without needing prisms. Near angle was also fully corrected in 19 patients. One patient with myopia and one patient with distance esotropia went on to have medial rectus recession.

Unilateral lateral rectus resection is an effective procedure for acquired distance esotropia in myopic and non-myopic patients.

16.52 Simultaneous Development of Acute Acquired Concomitant Esotropia in Two Siblings during the COVID-19 Pandemic: A Case Report - P Pujara (T), D Carter (O), K Bolton (C), R Nicholson (C), Portsmouth University Hospitals NHS Trust

A 5-year-old boy (sibling one) and his 11-year-old sister (sibling two) were presented to the hospital eye service in early 2021, having both developed acute-onset large angle esotropia within three months of each other. Neither had any significant past medical, ophthalmic, or family history. The siblings lived in the same household, and both experienced lifestyle changes as a result of the UK lockdown in response to COVID-19. Sibling one had a moderate right esotropia, initially maintained straight by corneal light reflex. He measured 45/50 prism dioptres (Δ) base out at near and 45 Δ base out at distance. Sibling two had esophoria which broke down into a right esotropia immediately on dissociation. The esotropia measured 30 Δ base out at near and 20 Δ base out at distance. At four month follow up, both siblings demonstrated a constant large angle esotropia

(sibling one: 54Δ base out at near and 45Δ base out at distance, sibling two: 45Δ base out at near and 40/45Δ base out at distance).

Each sibling was treated with right medial rectus recession (5.5 mm) and right lateral rectus resection (7 mm), and at a three-month follow-up, both were minimally esophoric with restored binocularity.

The unusual and abrupt changes in lifestyle imposed by the COVID-19 pandemic highlight the likelihood of an environmental aetiology for some forms of esotropia and raise the possibility that extended screen time may be a contributory factor.

16.57 Diagnostic performance of artificial intelligence for the detection of pathological myopia from colour fundus images: a systematic review and meta-analysis - N Tay (MS), J Prashar (MS), University College, London

Pathological myopia (PM) is one of the major causes of blindness worldwide and represents an imminent threat to eye health. Artificial intelligence (AI)-based methods are gaining traction in ophthalmology as highly sensitive and specific tools for screening and diagnosis of many eye diseases. However, there is currently a lack of high-quality evidence for their use in the diagnosis of PM.

We conducted a systematic review and meta-analysis of studies evaluating the use of AI-based tools to diagnose PM, according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidance. Five databases were searched and results assessed against inclusion criteria and quality assurance checklists. Summary receiver operating characteristic (SROC) curves were used to summarise model performance; sensitivity and specificity were calculated for each study and pooled.

11 studies were included in both the systematic review and meta-analysis testing approximately 165787 eyes. The area under the SROC was 0.9905. The pooled sensitivity, specificity and diagnostic odds ratio of AI-based detection of PM were: 95.9%, 96.5%, and 841.26 respectively.

This study provides evidence that AI-based diagnostic tools are highly specific and sensitive for the detection of PM using fundus images. There is potential for such tools to be incorporated into ophthalmic public health screening programmes, particularly in resource-poor areas with a substantial prevalence of high myopia.

17.02 Does the number of pre-operative assessments of strabismus patients influence the surgical outcome? - S McAuley (O), J Choi (C), I Zaman-Khan (MS), Royal Hallamshire Hospital, Sheffield

Surgical strabismus traditionally attended for several face-to-face (FTF) measurements before being listed for surgery. During the Covid-19 pandemic, a streamlined care-pathway was adapted to reduce FTF contact time to protect staff and manage floor-flow. Patients were listed for surgery after just one face-to-face orthoptic assessment, followed by a further measurement at pre-assessment.

This is a case cohort to look at surgical outcome of all consecutive exotropia patients who underwent re-do horizontal muscle surgery over the last 14 years, from a surgeon's prospective database.

65 patients had surgery before the pandemic (Group-A) and 28 since the pandemic (Group-B). The mean age was 34.9 and 39.1 years respectively. The mean pre-operative near-angle was 37.5 prism dioptres (PD) and distance-angle of 34.8PD for group-A. Group B was

39.3PD for near and 36.3PD for distance. 89.1% in group-A and 89.2% in group-B had suppression. Postoperative alignment within 15PD was achieved, after a combination of fixed and adjustable procedures, for 75.4% in group-A and 88.5% in group-B with predominant adjustable procedure. The mean 2-weeks post-operative alignment was -1PD for group-A and -1.3PD for group-B. Group-A had an average of 3.6 measurements, with 3.3 doctor consultant prior to surgery and group-B had 2 FTF orthoptics and 1 FTF doctor contact.

44.4% reduction in face-to-face orthoptics contact-time and 69.7% less consultant contact-time had not negatively affected the patient surgical outcome.

Strabismus surgery can be considered in selected cases with less FTF pre-operatively and improve cost-efficiency of our adult strabismus service with 'block-contract' status.

17.07 Diagnostic Monocular Occlusion in patients with binocular vision - W Ghaffari (FD), N Mansoor-Ali (T), A Budd (O), R Sadia (C), A Maino (C), Manchester Royal Eye Hospital

To determine if angle of deviation changes significantly after diagnostic monocular occlusion (DMO) in patients with BSV (binocular single vision).

We retrospectively analysed data from 136 patients with esotropia and 110 with exotropia, divided in BSV and non-BSV. Near and distance angles were measured over 3 visits and then after 2 hours monocular occlusion. Data were analysed with t-test and linear regression.

Esotropes with BSV had significantly larger increases in deviation after DMO than non-BSV patients for near (5 PD vs 3 PD, 95%CI 3-7 PD, paired t-test $p=0.0318$) and for distance (6 PD vs 3 PD, 95%CI 4-8 PD, $p=0.005$). There was a significant correlation between the amount of change after occlusion and the initial angle for near ($p=0.006$) and for distance ($p=0.010$) in BSV patients only.

In exotropes, there was no significant difference after DMO between BSV and non-BSV patients for near ($p=0.09$) and distance ($p=0.532$). There was a significant correlation between initial near angle of deviation and change in deviation after DMO in BSV patients ($p=0.0009$) but not in the non-BSV group ($p=0.07$). Distance angle of deviation before DMO was significantly correlated to the amount of change in deviation after DMO for both BSV ($p=0.022$) and non BSV patients ($p=0.015$).

Our study shows that diagnostic monocular occlusion should be performed in all exotropes with initial angles of deviation of less than 30 PD. In esotropes, DMO is more likely to show significant increases in deviation in BSV patients, especially for smaller initial angles (less than 25 PD before DMO). These changes should be taken into account when planning surgery.

17.12 Using visual data and teleophthalmology in Paediatric Ophthalmology with an app-free, browser-based, visual data platform: ISLACARE - A Elhence (F), L Christou (R), R Dodeja (C), A Raj (C), J Gonzalez-Martin (C), DCM Yeo (C), Alder Hey Children's Hospital NHS Foundation Trust, Liverpool

Visual data is underutilised in ophthalmology particularly within paediatrics. Following the pandemic, virtual and remote clinics in the form of telephone and video consultations have increased but there are limitations within paediatric ophthalmology as synchronous telephone or video calls are time-consuming.

Using a platform known as ISLACARE, we are able to run remote photo and video clinics that has the capability to support asynchronous or synchronous consultations. With this software, parents and clinicians do not need to create logins or download apps thus increasing compliance with the technology.

In an audit of 101 consecutive cases, the following was found. Mean age: 6.67 years (0-17years). The top 4 categories used in were anterior segment (36%), Strabismus (24%), Orbit/Trauma (17%), and Oculoplastics (12%).

On the use of photographs to support consultations , it was felt that 91% reduced time to treatment/supported clinical decision making and 75% improved clinician to clinician communication. We have found a 30% increase in capacity in our remote teleophthalmology clinics by utilising a pre-consultation proforma. A particular improvement has been in post-operative strabismus cases where 90% of all our first appointment checks are now done remotely.

We would like to demonstrate the clinical flow of how we use ISLACARE for asynchronous consultations, remote monitoring, and visual data archiving.

FRIDAY 06th OCTOBER 2023

09.50 Session II (S) (P)

Free papers

Moderators: Manoj Parulekar, Birmingham/Oxford and Rohit Jolly, London

09.51 Chiasmal Misrouting in Infantile Nystagmus Syndrome (INS): Phenotypes in Patients with Molecular Diagnoses – M J Gilhooley (T), M Moosajee (C), M M Neveu (SL), M Theodorou (C), Moorfields Eye Hospital

Chiasmal misrouting, once believed to be pathognomonic for albinism, has been reported in cases of INS, independent of melanin pathway disruption. The purpose of this study is to determine if there are clinical-electrophysiological parameters that correlate with particular genotypes in INS.

A retrospective chart review at Moorfields Eye Hospital identified 71 patients with a molecular diagnosis relating to INS. Visual acuity; presence of nystagmus, signs of albinism and OCT foveal hypoplasia grade were recorded alongside flash and pattern VEP (Visual Evoked Potential) amplitude and peak time. VEP asymmetry was assessed using the Pearson Correlation Coefficient (r).

Pathological variants in 8 genes (TYR, OCA2, HPS6, HPS3, HPS1, GPR143, FRMD7, SLC38A8, OCA1) were identified. Mean BCVA per group ranged from 0.38-0.74LogMAR

$F(0.72,3.5)=2.8$; $p=0.04$ one-way ANOVA. All genotypes demonstrated foveal hypoplasia (mode grade 4) except FRMD7 (all grade 1). In this cohort, positive flash and pattern VEP amplitude/peak time asymmetry correlated with clinical signs of albinism (flash VEP, $r=0.22(0-6yrs)$; pattern VEP, $r=0.17(6-65yrs)$). There was marked asymmetry in SLC38A8 patients ($r = -0.85$ to 0.93), a feature known to be associated with foveal hypoplasia 2.

This study provides a detailed genotype-phenotype correlation of VEP findings in a molecularly characterised INS cohort - useful in selecting clinically guided genetic testing and counselling patients.

10.00 Recognition of Intracranial Hypertension in Children using Handheld Optical Coherence Tomography: The RIO Study – S R Rufai (T), R Bowman (C), M G Thomas (CL), R Purohit (O), C Bunce (o), V Panteli (C), I Gottlob (P), C K Patel (C), D J Dunaway (P), D Johnson (C), F A Proudlock (o), T Lawrence (C), N U O Jeelani (C), Great Ormond Street Hospital, London and Oxford Craniofacial Unit, Oxford

Intracranial hypertension (IH) can damage the brain and optic nerve head (ONH) if unaddressed. Intracranial pressure (ICP) monitoring is invasive and carries risk. In this world-first diagnostic accuracy study, we assessed the role of handheld optical coherence tomography (OCT) to recognise IH in children.

We conducted a multi-centre, prospective diagnostic accuracy study at Great Ormond Street Hospital and Oxford. Children (<18 years) at risk of IH were recruited between September 2020 and August 2022. Handheld OCT was performed during ICP monitoring. The surgeons measuring/interpreting ICP were masked to OCT findings. We considered ICP raised if the median reading was above 20mmHg, or prolonged ICP spikes were present. Positive OCT parameters included anterior displacement of Bruch's membrane, raised rim and cup, cup obliteration and atrophy. Sensitivity/specificity analysis was

performed and Fisher's exact test was used to compare OCT parameters and ICP (raised/normal).

Handheld OCT was successful in 41 of 42 (98%) eligible children. Diagnoses included craniosynostosis (n=26), hydrocephalus (n=9), IIH (n=2) and other (n=4). Median age at examination was 6.5 years (range: 0.5-17, IQR: 4-13). For all OCT parameters, sensitivity was 90% and specificity 75% for IH. Fisher's exact test demonstrated a highly significant difference between OCT and ICP groups ($p < 0.001$).

Handheld OCT is feasible and valuable for recognising IH in children. The anterior displacement of Bruch's membrane alone delivered 100% specificity and can be considered a 'red flag' OCT sign for IH. Our findings could help improve the clinical management of children at risk of IH.

Conflicts declared: Frank Proudlock and Mervyn Thomas: paid consultants, Leica Microsystems. No other financial interests

10.09 Primary Outcomes of Management of convergence excess esotropia at Moorfields Eye Hospital - D Elfadaly (T), A-M Hinds (C), Moorfields Eye Hospital

Convergence excess ET (CXE) is an esotropia with binocular single vision (BSV) at distance fixation but esotropia on accommodation for near fixation with near distance disparity (1). In this retrospective chart review our 1st question: to evaluate the initial treatment to address CXE, whether this treatment successful or not, secondary question: to evaluate primary and final outcomes for control of near esotropia. Successful outcome defined as residual distance and near esotropia and near distance disparity of less than 10PD.

We included patients with CXE managed at Moorfields Eye Hospital from 2003 until 2022, defined as "esotropia with BSV at distance but esotropia on accommodation for near with near distance disparity over 8-10 PD while the eye is corrected with the full cycloplegic refraction". All age groups were included, amblyopic eyes were excluded.

668 patients were reviewed from 2005-2022, the mean age was 7.9 years (+/- 6), first line treatment was bifocal glasses in 60%, which was successful in 83%, bi-medial recession was offered to 12.5%, only 1/4 of which were improved, Botulinum toxin administered to 3%, bimedial posterior fixation sutures done in 1.5% which did not improve condition. Other lines included single vision glasses and Bangerder foil to relieve double vision. The final outcome was well controlled esophoria in 65% of cases. Binocular single vision achieved in 28%.

The management of convergence excess esotropia is still controversial, in our cohort most patients were managed with bifocals, the final motor and sensory outcomes were variable between patients. More than half of patients had satisfactory motor alignment. However, the sensory outcome was much less.

10.18 Virtual Strabismus Clinic: An Alternative Model of Care during the COVID-19 Pandemic - M Panahi (MS), D Mullinger (O), J Mistry (O), J Somner (C), A Vivian (C), Cambridge University Hospitals NHS Foundation Trust

Addenbrooke's Hospital introduced a virtual strabismus clinic in March 2021 to manage patient care during the COVID-19 pandemic. This study aims to explore the feasibility and utility of this care model by evaluating its effectiveness in delivering patient care.

Clinic data from April 2021 to April 2022 were retrospectively analysed, including patient demographics, referral information and outcomes. All patients underwent an initial

assessment by a specialist orthoptist, preceding virtual review by a consultant ophthalmologist.

The clinic saw 114 patients between the ages of 12 and 95 during this period, with an increasing number of patients seen per month. Within two months of the clinic's inception, wait times reduced by 59%: from 30.2 weeks to 12.5 weeks, remaining constant thereafter. Most referrals came from optometrists, with diplopia and identification of new or recurring strabismus being the most common complaint. Virtual review outcome varied significantly: 30.7% of patients were discharged, 16.7% listed for surgery, 34.2% received a repeat FTF review and a further 18.4% received a review virtually.

Following its inception, the virtual clinic was able to effectively accommodate patients despite capacity restraints. This was partly achieved through the effective utilisation of specialised orthoptists. Subsequent virtual review by a consultant ophthalmologist achieved positive patient outcomes.

Virtual clinics provide an opportunity to optimise patient care and maximise efficiency of clinical input. If applied appropriately, this model of patient care may reduce the NHS burden, improving wait times to facilitate faster intervention. Increasing consultant availability permits the treatment of a greater number of patients.

10.27 The efficacy of pre-operative multidisciplinary meetings for surgical management of strabismus - B Gohil (O), N Tan (C), R Jolly (C), N Yadav (CF), S Jain(C), Royal Free Hospital

Informal discussion regarding surgical management plans between strabismus surgeons is common but this limits potentially valuable multi-disciplinary input and learning opportunities. We evaluate the effectiveness of virtual multidisciplinary meetings to provide a platform for discussion of pre-operative strabismus surgical cases.

Weekly virtual MS Teams meetings are held on Monday mornings for one hour, attended by the three paediatric consultants, paediatric fellow, all trainees on the paediatric firm and orthoptists. The meetings are recorded and available for reference to the content and for those not present.

Presentations for upcoming surgery cases are prepared and presented by the fellow, with discussion from participants regarding examination findings and surgical options. Cases are anonymised to allow multicentre collaboration. The agreed management plan is documented in the patient's medical notes, and outcomes of challenging cases are discussed.

The management plan is formalised during the MDT. Where required, additional tests are arranged. There are opportunities for all participants to constructively challenge decisions. Trainees of all levels are actively engaged by presenting, listening to the rationale behind surgical plans, with the opportunity to ask and respond to questions.

Patients are informed that their case has been presented in the MDT to obtain multiple opinions, which gives them additional confidence. Orthoptists can see the impact of the measurements they provide, and how differing tests can change management plans.

This MDT has been a positive change to our surgical strabismus patient pathway.

Knowledge and teamwork have been strengthened using this innovative virtual discussion method.

10.36 Orthoptists in the Surgical First Assistant role - G Pullan (O), E Lynch (O), J Gonzalez-Martin (C), R Dodeja (C), A Raj (C), D Yeo (C), I Yip (C), Alder Hey Children's NHS Foundation Trust

At Alder Hey we have trained orthoptists who are surgical first assistants. We believe there are many benefits of this and would like to encourage other departments to utilise their orthoptists in improving their service and patient experience.

We are not training orthoptists to operate, we are training orthoptists to assist the surgeon and therefore the orthoptist is not 'stealing' training opportunities for registrars.

The orthoptist offers consistency in theatre over time. A registrar changes every 6 months and a fellow every 12 months, whereas orthoptists are a permanent main stay of the ophthalmology department.

On the day of the operation the orthoptist can offer strabismus assessments and repeat any orthoptic measurements. The orthoptist can assist with the measurement for adjustable suture operations. The orthoptist can also offer an expert second opinion on the type of procedure and the amount of surgery to be performed.

Orthoptists can assist in marking the correct eye for surgery, gaining consent, speaking to parents pre and post operatively, administering pre-operative drops, assisting with IT systems and being trained in all the ophthalmology software, performing video and photography recordings with the theatre cameras, bringing equipment from clinic to theatre.

An orthoptist trained to become surgical first assistant increases the surgeons surgical time to concentrate primarily on surgery, by reducing other theatre tasks this can lead to an increase in theatre slots. This will help the surgeon complete their job more efficiently and effectively, which improves the hospital service and patient experience.

Legend

(C) = Consultant, (O) = Orthoptist, (T) = Trainee, (CF) = Clinical Fellow, (CL) = Clinical Lecturer, (CRF) = Clinical Research Fellow, (F) = Fellow, (FD) = Foundation Doctor, (L) = Lecturer, (MS) = Medical Student, (N) = Nurse, (Op) = Optometrist, (P) = Professor, (RA) = Research Associate, (RF) = Research Fellow, (RO) = Research Orthoptist, (SAS) = Specialty Doctor, (SL) = Senior Lecturer, (o) = other



posters

Posters are located in the Atrium Well and Max Rayne Foyer.

**Authors have been allocated a time slot to be available by their poster for questions -
*this is not mandatory***

Atrium Well Boards 1-8

Time for questions: Wed 13.15 - 13.30

Secondary Idiopathic Intracranial Hypertension associated with Severe Aplastic Anaemia and Paroxysmal Nocturnal Haemoglobinuria

Dr T.Mannampat Shobh (CF), Ms S.Amarakoon (C), Bristol Eye Hospital

A 14 year old girl with severe aplastic anaemia and paroxysmal nocturnal haemoglobinuria requiring blood and platelet transfusion, presented with c/o severe headache and mild blurring of vision. She was on Ravilzumab, voriconazole, erythromycin, progesterone and was awaiting bone marrow transplantation.

Examination revealed bilateral haemorrhagic papilloedema and serous macular detachment. OCT disc confirmed the severe disc edema and OCT macula showed serous macular detachment. CT head and MR venogram confirmed papilloedema and excluded intracranial bleed, haemorrhage, hydrocephalus and cerebral venous sinus thrombosis. It showed features suggestive of raised intracranial pressure with dilated optic nerve sheaths and stenosis of lateral half of the right transverse sinus. Lumbar puncture was performed which showed opening pressure of 31cmH₂O.

She was treated with Acetazolamide and therapeutic lumbar puncture (LP) was done. She went on to have five further therapeutic LPs to control intracranial hypertension.

She was symptomatically better in 10 days. Papilloedema resolved and OCT showed a marked reduction in the RNFL thickness and serous macular detachment. Bone marrow transplantation (BMT) was performed from a fully matched sibling. She has remained asymptomatic post BMT.

Previous case reports have documented the association between anaemia and raised intracranial pressure. This case shows importance of prompt treatment and resolution of Intracranial Hypertension with treatment of aplastic anaemia.

Time for questions: Wed 13.30 - 13.45

Post-operative evaluation of orbital fracture patients with diplopia in down gaze S Poonoli (O), N Raof (C), The Royal London Hospital

Royal London Hospital is a major trauma centre for South east London. Due to this, high volumes of orbital fracture cases are seen by the ophthalmology.

Management of orbital fractures when patients are asymptomatic in primary position, but symptomatic with diplopia in down gaze is challenging. Surgeries in these cases have risk of post-operative diplopia in primary position. However, we often explore this option when the patient is troubled by diplopia in down gaze.

This study aims to review the effects of surgery on patients who are symptomatic only at down gaze.

We evaluated pre-operative and post-operative orthoptic findings of 7 patients aged between 24-79. These patients presented with diplopia in down gaze only, between January 2022- December 2022.

2/7 patients were asymptomatic in all positions post-surgery. 5/7 patients continued to have diplopia in down gaze. This study highlights the importance of managing patient expectations in these cases.

Time for questions: Wed 16.25 - 16.35

Functional neurology: 3 unusual ophthalmic presentations

J Hoole (O), Leeds Teaching Hospitals NHS Trust

Functional neurology is a complex poorly understood disorder where there is physical response and no pathology is identified e.g. non epileptic seizures. It is therefore termed a problem with the functioning of the nervous system not due to damage or structural disease of the nervous system. There is often a combination of symptoms. Underlying triggers may be emotional and psychological help may be needed particularly in the more severe cases. Patients are often resistant to this. There is still a tendency for some orthoptists and other clinicians to believe symptoms are deliberately made up or put on as in true malingering. Common presentations to ophthalmology include functional vision loss, spiralling visual fields and near reflex spasm.

Three unusual cases are presented:

- 1) Increasing inability to walk in a teenager whilst waiting for decision to treat diplopia associated with decompensating deviation due to superior oblique weakness
- 2) A 70 year old with near reflex spasm after withdrawal of long term painkillers and sleeping tablets both of which may have been given for a functional problem in the first place
- 3) Opsoclonus myoclonus signs / symptoms

Orthoptists / other clinicians need to be alert to functional neurology and consider it as a differential diagnosis with true malingering in atypical as well as common presentations understanding that symptoms are real with an underlying emotional or psychomatic aetiology. Treatment in ophthalmology is unlikely to help particularly in the more extreme cases. Treatment and help will depend on local neuro and psychology services.

Time for questions: Thu 13.20 - 13.35

Half and half syndrome: A rare cause of diplopia

P H Downes (O), F Kirtley (C), Shrewsbury and Telford Hospital NHS Trust

An 81 year old man presented to A&E with symptoms of numbness and tingling on the right side of his face, pins and needles extending down into the right hand and sudden loss of the use of his legs. He was admitted to the stroke ward where MRI imaging identified a left pontine infarct. The stroke rehabilitation team referred him to orthoptics querying a possible diagnosis of 3rd nerve palsy causing diplopia, but this assessment led to a diagnosis of a left internuclear ophthalmoplegia and a left 6th nerve palsy.

Orthoptic assessment, 2 months after the onset of the stroke, demonstrated vision of 6/6 bilaterally, a lateral rectus restriction, as well as adduction deficit of his left eye and on the right, abducting nystagmus but no movement deficit. Although convergence was normal, there was left hypotropia caused by slight under action of the left superior rectus. Skew deviation was investigated and excluded. There was no evidence of facial palsy.

His co-morbidities included hypertension, chronic kidney disease, hypercholesterolaemia. This patient is an example of half and half syndrome, an unusual presentation of diplopia and reported rarely. It is caused by a lesion involving the ipsilateral MLF, contributing half of the contralateral horizontal gaze palsy, and ipsilateral abducens nerve fasciculus, contributing half of the ipsilateral horizontal gaze palsy, whilst sparing the abducens nerve nucleus.

We present this case report to further expand the small evidence base of half and half syndrome and to help establish a wider knowledge of this rare INO plus condition.

Time for questions: Thu 15.40 - 15.55

MOG IgG optic neuritis in the paediatric population – an Irish case series

D Harford, D Townley, Galway University Hospital, Galway, Ireland

MOG IgG is a unique demyelinating cause with distinct features pertaining to treatment and prognosis. This is of importance as treatments are not universal for each cause of demyelinating disease. We describe two recent cases of MOG IgG associated optic neuritis in paediatric patients.

Case 1: A 10 year old female presented to eye casualty with a drop in vision in her right eye to CF from 6/5 accompanied by a swollen disc. MRI demonstrated a longitudinally extensive optic neuritis from globe to chiasm. Serum MOG IgG were found to be positive.

Case 2: A 6 year old female presented with a profound drop in visual acuity to 6/60 in her right eye accompanied by a swollen optic nerve head. Her MRI brain confirmed a long segment of T2 hyperintensity and swelling along the right optic nerve involving the intraorbital, intracanalicular and intracranial segments. Her blood work demonstrated anti-MOG antibodies.

Our series highlights a number of clinical findings which help identify MOG IgG optic neuritis in children. Firstly, the profound visual loss which often accompanies a presentation of MOG IgG optic neuritis in children was evident. Secondly, MOG IgG associated optic neuritis is reported to be associated with longitudinally extensive lesions along the optic nerve which we also describe. Thirdly, optic nerve oedema is frequently present which we also describe. MOG IgG associated optic neuritis is associated with an excellent response to steroids. Therefore a high clinical suspicion and a timely diagnosis are important.

Time for questions: Fri 13.15 - 13.30

Lessons Learned from COVID-19 (discharge delays in Paediatric Orthoptic clinic)

S Kaneshanesan (O), Barts Health

Paediatric Ophthalmology is experiencing increasing demand, especially with the growing East London population. A retrospective audit on discharge trends on all Paediatric patients on Orthoptic ASI (n=1340) due to the closing of outpatients in response to COVID-19.

Outpatients closed from March 2020 to July 2020 and January 2021 to March 2021 in response to COVID-19, and all patient appointments were cancelled and added to ASI (Allocated Slot Issues). In line with Public Health England guidelines, all patient notes were reviewed and triaged using the traffic light system (Moorfields). Electronic patient records (appointments (CRS) + clinical (Medisoft)) were used to identify discharged patients and type of interaction i.e. SMART triage, telephone consultation, number of face-to-face or WNB.

(Preliminary 725/1340) SMART n=114, TC n=>70, F2F n=>304 WNB n=>38. Preliminary data indicates at least 39% of Paediatric Orthoptic patients were discharged from March 2020 to April 2023.

Clinical and parental caution influenced some delays in discharge, which led to a retention of patients who were safe to discharge and transfer eye care to local Opticians. COVID-19

gave an opportunity to change attitudes towards follow-up duration and need, and increased efficacy and efficiency of the Paediatric Orthoptic clinics.

Conclusion: Reviewing notes identified numerous patients who were safe to discharge, and led to increase efficacy for high demand service.

Time for questions: Fri 13.30 - 13.45

Clinical Characteristics of Isolated Inferior Rectus Palsy

LM Roberts (O), A Agrawal (SAS), VSY Geh (C)

Southend University Hospital, Mid and South Essex NHS Foundation Trust, Southend-on-Sea, Essex

Isolated inferior rectus palsy is a well-recognized but uncommon clinical entity and is conventionally stated to be associated with Myasthenia Gravis. There is limited literature available on this condition. The aim of this report was to elucidate the aetiology and clinical characteristics of isolated inferior rectus palsy.

We undertook a 3-year retrospective study of patients with a diagnosis of isolated inferior rectus palsy. Records of 12 patients were available to be reviewed who attended between January 2021 and February 2023.

The mean age at diagnosis was 63.5 years (range 35 to 83 years). There were 5 men and 7 women. The main clinical presentations consisted of hypertropia of the affected eye, motility limitation in abduction and depression. Ten were tested for Myasthenia Gravis of which diagnosis was confirmed in 3 patients. Out of the remaining cases, the aetiology was unknown in one, four were microvascular and 2 had other causes like trauma and blood dyscrasia. Ptosis was found in 1 of the cases of Myasthenia Gravis.

The main aetiologies of isolated inferior rectus palsy involved myasthenia and microvascular events. Patients who present with isolated inferior rectus palsy should be investigated for Myasthenia gravis.

Time for questions: Fri 13.45 - 14.00

A novel phenotype associated with the GJA8 c.280G>A p.Gly(94Arg) variant

J Gilmour-White, A Churchill, M Tooley, Bristol Royal Infirmary

GJA8 is a gene that encodes for connexin 50, a transmembrane protein that plays a crucial role in lens development and homeostasis. A wide range of mutations in GJA8 have been identified as the underlying cause for a spectrum of ocular phenotypes. Notably, the GJA8 c.280G>A p.Gly(94Arg) variant has been established as a likely pathogenic mutation.

Previous studies have documented three cases in the literature that demonstrate a phenotypic spectrum consisting of aphakia, corneal opacity, microphthalmia, coloboma and glaucoma. In this report, we present a novel case of this variant with bilateral congenital aphakia, bilateral aniridia (iris tissue remnant visible in the periphery with transillumination), and corneal opacity. While coloboma of the iris and optic nerve have been reported in the past, we believe this to be the first documented case of aniridia associated with the GJA8 c.280G>A p.Gly(94Arg) variant. PAX-6 variants, which are a well-recognised cause of aniridia, were excluded. This case expands the current understanding of GJA8 variants and their associated phenotypes and underscores the importance of genetic GJA8 screening in individuals with developmental eye abnormalities.

**Atrium Well
Boards 9-16**

Time for questions: wed 13.15 - 13.30

Use of anti-Vascular Endothelial Growth Factor (antiVEGF) in the Treatment of Retinopathy of Prematurity (ROP), A Systematic Review and Meta-Analysis

S.J Udakumbura(SL), P Watts(C), University Hospital Of Wales, Cardiff

Retinopathy of prematurity (ROP) is one of the leading causes of blindness in children worldwide. The management of ROP has been revolutionised with the introduction of intravitreal anti-vascular endothelial growth factor(antiVEGF) agents. This review evaluates the safety and efficacy of intravitreal antiVEGF agents when used either as monotherapy, or in combination with laser/cryo in type 1 ROP.

A comprehensive literature search was conducted from 2000 to April 2022 using the following databases PubMed, EMBASE and CINHAL. Total of 12 randomised controlled studies (RCTs) and 7 comparative studies were selected after critical appraisal. RCTs were included in the meta-analysis.

There were 4628 eyes of 2571 infants with type 1 ROP were identified from the included studies. A meta analysis showed no statistically significant difference in retinal detachment, ROP recurrence, mortality and cerebral palsy between standard treatment (laser/cryo) and antiVEGF mono therapy. AntiVEGF mono therapy may reduce the risk of refractive errors in childhood compared to standard therapy. No significant risk of acute and long-term systemic complications noted with antiVEGF therapy.

AntiVEGF agents, as mono therapy is non inferior to standard therapy after evaluating for risk of retinal detachment and recurrence of ROP in infants with type 1 ROP. The data is not sufficient to make strong conclusions favouring routine use anti-VEGF agents as monotherapy in all types of type 1 ROP. Hence, further large scale RCTs with longer follow up period are necessary to evaluate the safety and efficacy of antiVEGF agents for type 1 ROP.

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Time for questions: wed 13.30 - 13.45

The Prevalence of Peripapillary Hyperreflective Ovoid Mass-like Structures (PHOMS) in Suspected Papilloedema in Children

S Rehan (T), L Pratt (T), J West (T), P Watts (C), University Hospital of Wales, Cardiff

Suspected papilloedema is a frequent cause of referral to paediatric ophthalmology clinics. To allow prompt assessment and manage demand, we run a virtual suspected disc swelling clinic. In these clinics, OCT disc images are taken and reviewed separately by an ophthalmologist. Recent publications have described a new finding called peripapillary hyperreflective ovoid mass-like structures (PHOMS) that may simulate papilloedema. We

aimed to retrospectively review the OCTs from these virtual clinics for the presence of PHOMs and report their frequency.

124 patients between 31st August 2016 and 17th March 2021 were appointed to the virtual clinic. 13 patients did not attend, and 1 had ungradeable images. In total 110 patient scans were reviewed by 3 assessors. The OCT appearance was compared with previously published images and descriptions of PHOMS. The agreement between assessors was also recorded.

The average age was 11.2 years (SD 3.4 years). PHOMS were identified in 74 patients (67.3%). 56.8% were bilateral. PHOMS were most commonly seen in association with other identified causes of pseudopapilloedema e.g. tilted disc and drusen (81.3%), but were also common in true disc swelling (66.7%) and otherwise healthy discs (55.4%). There was a high rate of agreement between assessors (Fleiss' kappa 0.97)

Misdiagnosis of true disc swelling can lead to unnecessary and invasive tests. PHOMS are found frequently within the paediatric population referred for suspected disc swelling. They may be an independent cause of pseudopapilloedema but are often seen in conjunction with other diagnoses e.g. true disc swelling or drusen.

Time for questions: Thu 13.20 - 13.35

Tales of the Unexpected! The Story of the Princess and the Satsuma—A rare presentation of Abducens Palsy

N V Corbin (O), B Cuckson (Op), M Shamir (SAS), North Cumbria Integrated Care NHS Foundation Trust, Carlisle

15 month old female, born at full term, presented to the orthoptist with a 6 day history of isolated vomiting, lethargy and 4 days of sudden onset left esotropia. Three consultations by GP had been undertaken without concerns raised.

Orthoptic examination found vision of 0.5 logMAR with Cardiff Cards. Approximately 20 prism dioptre esotropia for near and distance on cover test, with a marked restriction of abduction of the left eye. Dilated examination showed bilateral hypermetropia within normal limits for age. Fundoscopy showed left papilloedema with the fellow eye appearing normal. General observation showed ataxic gait, lethargy, prominent forehead, and reduced use of right arm.

Immediate admission to paediatrics with MRI under sedation. Radiological findings showed a mass in the lateral ventricles measuring 5.6cm x 6.1cm (equivalent to a satsuma) resulting in complete hydrocephalus, and a retro-cerebellar arachnoid cyst. Immediate management on dexamethasone and rapid deterioration resulting in specialist intubated transfer to tertiary centre in Newcastle. Immediate surgery for Extraventricular Drain (EVD), followed by surgical excision of mass by temporo-occipital craniotomy.

Spontaneous resolution of left abducens palsy following surgery. Histo-pathology showed Choroid Plexus Papilloma (CPP), WHO Grade 1. Normal developmental milestones resumed.

CPP is a very rare presenting aetiology of acquired Abducens Palsy, accounting for 0.4% of CNS tumours. Its slow growing nature often results in 6 months or more of symptoms before diagnosis. In a child with unexplained vomiting/nausea and headache and neurological signs, immediate imaging investigations should be sought.

Time for questions: Thu 15.40 - 15.55

Acquired Monocular Nystagmus in Pilomyxoid Astrocytoma

TKJ Chan (C), QX Lim (T), N Hall (T), A Stan (C), K Orr (C), Princess Alexandra Eye Pavilion

Acute onset monocular or asymmetric nystagmus is rare in the paediatric population. This tends to be seen in children in three scenarios: monocular visual loss; spasmus nutans; or as a presenting feature of an chiasmal/midbrain lesion. We aim to report a rare case of acute monocular nystagmus in a 4-month-old girl whose radiological and histopathological findings confirmed a diagnosis of pilomyxoid astrocytoma.

A 4-month old girl presented with right monocular nystagmus associated with poor feeding and weight loss. She was born full term via spontaneous vaginal delivery, weighing 7lbs 11oz following uncomplicated pregnancy. Examination showed monocular pendular nystagmus with horizontal and rotary component in the right eye only. There was no head bobbing or torticollis. Pupillary reflexes were normal with no relative afferent pupillary defect. She had symmetrical corneal reflexes and full eye movements. Dilated fundus examination showed healthy discs and normal fundi. Cycloplegic refraction showed +3.00D/+2.25D at 110° in right eye and +0.50D/+1.50D at 80° in left eye.

Subsequent investigations showed radiological findings of a highly aggressive brain tumour in the suprasellar region with widespread metastases in the posterior fossa, Circle of Willis, intracranial optic nerves and in C2, C6 and C7 spinal canal. Histo-immunopathology confirmed features of pilomyxoid astrocytoma. She underwent debulking surgery, ventriculoperitoneal shunt insertion and chemotherapy. Despite this, the disease progressed and she sadly passed away 5 months after initial presentation.

This case highlighted the importance and low threshold for neuroimaging in children presenting with acquired monocular nystagmus especially in those younger than 2 years old.

Time for questions: Fri 13.15 - 13.30

Novel inferior oblique muscle Y splitting procedure to minimize the anti-elevation syndrome

Amar Pujari (C), AIIMS, New Delhi

To describe novel Y splitting procedure of inferior oblique muscle to mitigate the anti-elevation syndrome.

A pilot, prospective interventional study was undertaken to assess the effect of inferior oblique muscle Y-splitting in patients with unilateral 3+ or more overaction. To correct primary gaze hypertropia and the excyclotorsion, a Y-splitting procedure was performed (along with routine horizontal muscle surgery as per the deviation) in 14 subjects. The effect of surgery was assessed at baseline and at 6 months post-intervention.

The mean age of 14 subjects was 25.14 +/- 7.70 years. The mean pre-operative hypertropia, excyclotorsion and inferior oblique muscle over-action was 18.42 +/- 3.50 PD, 14.14 +/- 2.65 degrees, and +3.21 +/- 0.42 respectively. Following surgery this was reduced to 1.57 +/- 1.74 PD of residual hypertropia (a net correction of 16.85 +/- 2.31 PD, p=0.005), 3.85 +/- 1.46 degrees of residual excyclotorsion (a net correction of 10.28 +/- 1.72 degrees, p<0.05), and +0.28 +/- 0.46 of residual inferior oblique over-action (a net correction ~+3) at the end of 6 months. Amongst fourteen patients, three patients still experienced residual/variable anti-elevation effect, and during the study period none of them experienced any adverse event and none of them required any additional surgeries.

While anteriorizing the inferior oblique muscle to correct primary gaze hypertropia and the excyclotorsion, a novel “Y splitting” procedure can be followed to achieve the desired results with mitigated anti-elevation effect.

Time for questions: Fri 13.30 - 13.45

Impact of red light from light-emitting diodes on subfoveal choroidal thickness and colour contrast sensitivity: proof-of-concept study

J Lam (CF/C), A Dahlmann-Noor (C), G Jeffery (P), Moorfields Eye Hospital NHS Trust

Daily viewing of low-level red laser light can slow progression of myopia in children, but effects on foveal photoreceptor health are unknown. An increase in subfoveal choroidal thickness (sfChT) may play a role in the mechanism of action of red light in modulating scleral remodelling and axial elongation. We conducted this proof-of-concept study to study the short-term effects of viewing 670nm light from red LEDs on sfChT. Six adult healthy volunteers viewed 670nm light from red LEDs for 3 minutes; three repeated the application once more later in the day. We measured sfChT on enhanced-depth imaging optical coherence tomography scans before and over 24 hours after the first viewing session.

Median sfChT increased from 257.5um (interquartile range IQR 165.8 to 331.6) at baseline to 280um (IQR 177 to 368.6) at 30 minutes, where it remained until 24 hours (n=6, p<0.05). Mean difference in sfChT from baseline to 30 minutes was 21.5(SD13.0)um, and to 24 hours, 18.1(SD23.6)um. Mean difference in sfChT from baseline to 24 hours in those participants who used the red-light treatment once (n=3) was 1.3 (SD1.8)um, and 34.9(SD23.3)um in those who had a second application later in the day, indicating that the second application may have prolonged the effect on sfChT increase (p=0.03).

Viewing red light from LEDs increases sfChT, an early biomarker of a therapeutic effect on scleral remodelling and axial elongation in progressive myopia in children, similar to that reported for low-level laser light. Red light from LEDs enhances photoreceptor function by increasing mitochondrial respiration, and possibly by improving oxygenation of retina and sclera downstream of an increase in choroidal perfusion.

Time for questions: Fri 13.45 - 14.00

Spectrum of visual dysfunction detected by a novel testing protocol within the Special School Eye Care Service.

RF Pilling (C), M Musleh (T), A Mankowska (Op), C Viner (Op), A Green (Op), University of Bradford

NHS England Special School Eye Care Service (SSECS) offers in-school visual assessments to children with special needs. The recommended testing strategy includes visual acuity, contrast sensitivity (CS), visual field (VF), stereoacuity, accommodation, eye movements (EM), refraction and parent-completed questionnaire (CVI5).

The aim of this evaluation is to report on the outcomes from the first cohort assessed by the locally commissioned service.

The service operates with opt-out consent. Data are routinely submitted to NHSEngland. The order of testing and tools used were determined by the optometrist to maximise each child's engagement. All children on whom testing was attempted were included.

78 case records were identified. The mean age of children tested was 9.6 years (range 7-12y). All six visual function tests were completed by 44% patients, with a mean of 5.

(range 2-6). Visual acuity was <6/15 in 31% cases. The most frequently omitted test was stereopsis (60% children completed), with >90% patients completing VF, CS, EM and accommodation. Only 20% children had a normal response to all tests. 44% parents returned the questionnaire; parents were more likely to complete this if their child had low visual acuity.

We found a higher proportion of special needs children with atypical visual function compared with previous studies. Children can engage well with the majority of tests with the possible exception of stereoacuity. The SSECS may have a role in identifying children who warrant further assessment for CVI related visual dysfunction.

Max Rayne Foyer Boards 1-8

Time for questions: wed 13.15 - 13.30

Usability of Delivery Aid for Vernal Keratoconjunctivitis Single-Dose Treatment K N Lebron (F), A D-Noor (C), Moorfields Eye Hospital NHS Foundation Trust

Usability of eye drop containers is particularly important for patient compliance when treating chronic eye diseases like vernal keratoconjunctivitis (VKC), glaucoma and ocular surface disease. Factors affecting treatment adherence include the convenience of use of the medication. The findings suggest that preferences may differ including the mechanical characteristics of the packaging or what the drop control is like. Santen has developed Dropaid VKC Single-Use, a delivery aid for use in combination with Verkazia in single use containers, that aims to help caregivers instill eye drops.

Eligible participants used Dropaid VKC Single-dose to administer CATIONORM PRO emulsion drops to a medical dummy, and then evaluated the usability characteristics of the delivery device by completing a questionnaire. Thirty adult participants were recruited and the usability was assessed through drop control, which was performed by an observer. Thirty patients completed the study. Most of the participants were females between 41 and 50 years old. 56% of the participants had experience using multi dose treatment and 43% using single dose drops, average of 3 to 5 years of overall experience. 76% of the participants found the general usability of the Dropaid VKC Single-dose container very easy, however 53% would recommend its use, 30% maybe and 17% would not recommend it. Most of the participants use the right hand to instil the drops, applying to the left eye first with approximately 2 drops per try.

Dropaid VKC Single-dose is a helpful tool to help caregivers instill eye drops from single use containers.

Time for questions: wed 13.30 - 13.45

Do my discs look big?

C McAtamney (O), P Anketell (O), A McCaw (o), S George (C), Belfast Health & Social Care Trust

Swollen discs can be a marker of a potentially sight or life threatening condition. In Belfast Health & Social Care Trust (BHSCT) an increase in referral rates has been noted (0.9/month pre-2016, 3.3/month in 2019, Stewart et al, 2017). The COVID pandemic reduced access to hospital eye services making assessment challenging therefore an

orthoptic delivered swollen discs clinic was developed which includes orthoptic assessment, imaging and virtual review by a consultant.

The database for all children attending the clinic was interrogated for this study. Visual measures and clinical outcomes were recorded for analyses.

This study reports on 217 children completed the clinic process; new=137, review= 80. Of the new patient were referred from an optometrist n=118, eye casualty n=4, paediatrician n=6, paediatric ophthalmologist n=5. Following review of the clinic reports 57% of all patients were discharged, this rises to 66% of all new patient referrals.

The development of this service has increased capacity within the paediatric ophthalmology service. Post COVID this service has continued for children requiring investigation of potential swollen discs. Extended scope training has been developed for orthoptists to with appropriate training to commence image review.

Time for questions: wed 16.25 - 16.35

TeleROP NI - A Service Improvement Initiative for Paediatric Ophthalmology in Northern Ireland

J Kearney (N), E McLoone (C), L Hamilton (o), Belfast Health & Social Care Trust

Every year in Northern Ireland, approximately 300 preterm infants require Retinopathy of Prematurity (ROP) screening. While fewer than 5-7% of the infants screened eventually require treatment, all eligible preterm infants must be screened at 1-2 weekly intervals from 31 weeks gestational age onwards. ROP is a time-sensitive eye condition in infants, which if undetected can result in lifelong blindness. This has life-changing implications for the infant and their family and significant adverse medico-legal implications for health care professionals and the National Health Service.

In September 2021, due to a paucity of Paediatric Ophthalmologists in Northern Ireland (NI), a novel three-tiered approach was introduced to manage the regional ROP service (TeleROP-NI). This consists of a regional ROP Clinical Coordinator, a weekly virtual ROP Multidisciplinary Team meeting and retinal imaging. As part of this initiative, non-medical ROP Imagers have undergone rigorous competency based consultant-led training in retinal imaging using handheld wide-field cameras. This poster summarises the development of a TeleROP-NI service improvement initiative and highlights a different approach for managing ROP screening and treatment.

Time for questions: Thu 13.20 - 13.35

Management of acutely acquired esotropia with botulinum toxin

N Patel (O), S Jain (C), R Jolly (C), Royal Free Hospital

We looked at the investigation and management of a child who presented with a new onset incomitant esotropia. We specifically look at the use of botulinum toxin (BT) as a diagnostic tool but also the possibility for its use as a therapeutic measure.

A 2-year-old male presented with an acute onset right esotropia with a significant abduction limitation. The orthoptic assessment showed a minus 4 limitation of abduction and a significant right face turn. Patching of the left eye could not prove an improvement in motility and binocularity was maintained with the face turn but diminishing with each visit. This led to a diagnostic dilemma with sixth nerve palsy being considered as a possible diagnosis. Considering the clinical features and acute onset deviation we did an MRI which was normal and then subsequently a decision made to inject 10 units of dysport BT into the

right medial rectus under GA. The needle was held in place for 90 seconds to avoid inducing ptosis or a vertical deviation.

The BT injection showed an excellent outcome as full abduction was proven in this child, with good binocularity in primary position and no residual deviation or compensatory head posture at three months following the procedure.

There is a limited consensus with regards to the best management of children presenting with acute onset esotropia especially those simulating a sixth nerve palsy.

The use of BT has showed an excellent outcome in this child and should be considered an option both as a diagnostic tool and a therapeutic measure in other children presenting with acute acquired incomitant esotropias.

Time for questions: Thu 15.40 - 15.55

Home visual acuity testing in young children using a smart phone App

Ahmed Ghoneim (CF), M Abbas (O), S Ludden (O), D ElFadaly (CF), A Dahlmann-Noor (C) (P), Moorfields Eye Hospital, London

VA is a core component of any eye-health consultation and management decisions.

OKKO-Health have developed a VA-app for home acuity-testing in young children by parents/carers, using pictorial Auckland Optotypes (TAO).

We approached children under the age of 16 years attending our clinics.

We recorded logMAR acuity at 3metres distance with Kay pictures or letters (Thomson-chart). Parents/carers were given an iPhone-XS with pre-installed OKKO App. We recorded age, gender, and worse-eye visual acuity.

We collected data from 46 children, mean age 6.5 standard deviation 3.3 years; 23 boys. 21 had amblyopia/refractive error/strabismus, 13 a lid/ocular surface condition, 5 congenital glaucoma, 5 healthy eyes/normal-for-age vision, and 1 each pseudophakia or anterior uveitis.

Median OKKO-TAO acuity was 0.19 interquartile range 0.00 to 0.40 and median VA was 0.13 logMAR (IQR 0.03-0.21). Mean difference between OKKO and reference standard was 0.08 logMAR, SD 0.24, 95 % CI (-0.15 to -0.04). Intraclass correlation coefficient was 0.7, 95% of measurements between +/- 0.48 logMAR

Results showed agreement between reference standard and TAO-element of the OKKO prototype. Limitations that near acuity (OKKO-app) may over-estimate distance acuity.

Comparing the vanishing-optotype, picture-only OKKO-TAO-prototype with a non-vanishing optotype reference standard affects our comparison, SD of 0.24 limits implementation into practice.

The updated version of the OKKO-Health app includes other targets, with a mechanism for brightness, grey scale and continuous measurement of distance between child and front-camera all to increase test performance. Evaluation is in progress.

Time for questions: Fri 13.15 - 13.30

Ocular Manifestations in Children with Autism Spectrum Disorder

U Karamchandani (Fd), N Oluonye (C), M Moosajee (C), Moorfields Eye Hospital

Evidence suggests abnormal visual and sensory processing contributes to developmental delays in children with autism spectrum disorder (ASD). However, current studies on ocular manifestations in these patients are limited by samples sizes, partly due to

behavioural challenges during assessments. We therefore aimed to evaluate this within the large Moorfields database.

Moorfields Eye Hospital records were searched for “ASD”, “Autism”, and “Autistic” to identify patients with this diagnosis. The ocular diagnoses and visual acuities were then recorded for patients aged under 20-years-old.

The study identified 1,245 patients with ASD, 870 demonstrated ocular manifestation(s). The mean visual acuity LogMAR (mVA) across the cohort was 0.40 (SEM: 0.17, range: -0.70-3.00). The most common conditions were strabismus (38%, mVA: 0.25, SEM 0.01, range: -0.70-1.50), then refractive errors (33%), amblyopia (7%), and retinal pathologies (6.9%, mVA: 1.12, SEM 0.10, range: -0.70-3.00). Sixty patients had confirmed genetic diagnoses with the majority being Lebers congenital amaurosis (23%, mVA: 2.58, SEM: 0.17, range: 0.80-3.00).

The prevalence of strabismus, refractive errors and amblyopia is much higher in this cohort than the UK paediatric population. Those with retinal pathologies, including dystrophies, retinopathy of prematurity, detachments, and genetic diagnoses, had low visual acuities or only perception of light. Given the importance of visual cues in childhood development, this may explain behavioural challenges.

Ocular manifestations occur more frequently in patients with ASD, and this may contribute to their behavioural and developmental delay. It is important to perform an ophthalmic evaluation and provide the necessary management if required.

Time for questions: Fri 13.30 - 13.45

Does ciclosporin A 1mg/ml cationic emulsion reduce the need for steroids in atopic, blepharo- and vernal keratoconjunctivitis?

A Dahlmann-Noor (C), M Hingorani (C), K Muthusamy (C), C Roberts (C), V Calder (P), NIHR Moorfields Biomedical Research Centre

Topical ciclosporin A 1mg/ml cationic emulsion (CsA) has recently received marketing authorisation in North America for vernal keratoconjunctivitis (VKC). It may reduce the need for topical corticosteroids and contribute to maintenance treatment of VKC, and also atopic and blepharokeratoconjunctivitis (AKC, BKC).

We reviewed the medical records of 485 children treated with CsA between 2015 and 2021, including 209 with VKC, 99 with AKC and 145 with BKC.

Median age at start of treatment was 9.7 years (IQR 7.2 to 12.8 years), 333 boys (62.5%).

Data were available for 12 months before/after starting CsA in 227 cases. The median number of inflammatory episodes requiring topical corticosteroids fell from 3 (IQR 2-4) to 1 (IQR 0-2), excluding steroid prescriptions concomitant with the first CsA prescription; Wilcoxon signed ranks, 2 tailed, $p < 0.01$. The number of clinic visits fell from a median of 4 (IQR 3 to 6) to 3 (IQR 2 to 5); Wilcoxon signed ranks, 2 tailed, $p < 0.01$. Stinging was a common adverse effect (4.7%); skin rash was unusual (0.6%). The most common reasons for discontinuation was a perception that the medication was no longer needed (32.8%) or the family not requesting, or the general practitioner not issuing, a repeat prescription (18.8%).

Csa 1mg/ml cationic emulsion reduces the need for topical corticosteroids and hospital visits and may improve the quality of life of children and families. Information and communication need to improve to raise awareness of VKC, AKC and BKC as chronic conditions requiring long-term treatment.

Time for questions: Fri 13.45 – 14.00

**Therapeutic botulinum toxin use in paediatric strabismus, the Sheffield experience
T Liu (T), M Naguib (FD), S Gosling (O), A Tandon (C), Sheffield Children's NHS
Foundation Trust**

Surgery is regarded as the mainstay treatment of paediatric strabismus. Botulinum injections, such as Botox, have emerged as a viable alternative. The procedure involves less trauma to the eye and is more tolerable in comparison to more lengthy and intense surgical options, especially in the paediatric population. This study aims to explore the efficacy of Botox in the management of paediatric strabismus.

A retrospective single centre study was conducted at a tertiary hospital ophthalmology unit. 41 patients were identified via electronic patient records between 2016 to 2021. Data collected included; age at time of procedure, primary and secondary diagnosis, muscles injected, Botox units, if patients underwent combined squint surgery, pre-operative and post-operative angle measurements, complications and whether patients underwent repeat Botox or further surgery.

Of the 41 patients, 80.5% underwent Botox injection to both medial recti. The mean difference in angle size by the third visit was a reduction of 23D. 14.6% required repeat botox after the third visit and 24.4% underwent further squint surgery later on. Of the 53.7% who experienced complications, 72.7% consisted of ptosis that self-resolved. Botox injections may be effective in treating strabismus but the effect may be temporary and patients may still require repeat injections or surgical treatment in the future. Overall, there may be a place for the use of Botox injections in the management of paediatric strabismus as an option prior to or after strabismus surgery for those who would like to opt for a less invasive procedure.

**Max Rayne Foyer
Boards 9-16**

Time for questions: wed 13.15 - 13.30

Have SARS-CoV-2 virus (COVID-19) lockdown restrictions impacted the prevalence and severity of retinopathy of prematurity in a London population?

K Reed (T), S Bazeer (T), I Hossain (T), D El Fadaly (CF), M Tsimpida (C), H Patel (C), A Das (C), A Khaier (C), M Posner (C), Queens Hospital, Romford

During the COVID-19 lockdowns in 2020 numerous global neonatal intensive care units (NICU) reported reduced incidence of pre-term births. Retinopathy of prematurity (ROP) is a vision-threatening disease that affects premature infants. We hypothesised that there may be a corresponding reduction in incidence and severity of ROP.

Comparison of birth weight (BW), gestational age (GA) and ROP stage from three East-London NICU during the UK lockdown period March to October 2020 matched with equivalent dates in 2019. Inclusion criteria; GA<32 weeks or BW<1501g.

264 ROP-screened pre-term babies included; 138 in 2019, 124 in 2020. 65 (46%) in 2019 and 46 (37%) in 2020 were diagnosed with maximal ROP grades 1-3. Babies were of similar GA (p=0.6) and BW (p=0.9) across the cohorts. There was, however, a statistically

significant difference in the severity of ROP with 13 (9.4%) 2019 babies vs 5 (4%) 2020 babies diagnosed with stage 3 ROP requiring treatment ($p < 0.05$).

Fewer pre-term babies were born in 2020 compared to the same time period in 2019. Of the pre-term babies that were born, fewer were diagnosed with ROP and fewer still had stage 3, treated ROP. The underlying reasons for this are uncertain and require further investigation but postulated suggestions include decreased incidence of maternal infections, decreased maternal activity and improved air quality.

This mirrors findings from a similar study in Japan indicating a reduction in sight-threatening ROP in babies born during Covid lockdown. Further investigation is required to determine a cause-effect relationship and to further inform expectant mothers.

Time for questions: wed 13.30 - 13.45

HSV2 induced Acute Retinal Necrosis in an 8-year-old child

C M Luxhoj (FD), R Jolly (C), H Petrushkin (C), D Hanumunthadu (C), Royal Free London NHS Foundation Trust

An 8-year-old male presented acutely with a red, itchy and painful right eye (RE). He had a past medical history of mild asthma, allergic rhinitis and a RE convergent squint but no other ocular history or relevant family history.

Visual acuity was RE 6/36 unaided, left eye (LE) 6/9 unaided. There was significant RE conjunctival hyperaemia and anterior chamber inflammation (cells 3+, flare 3+) with inferior keratic precipitates. Fundus examination showed vitritis in the RE with blurred disc margin, periphlebitis, temporal haemorrhages and retinitis. The LE was normal. Systemic assessment, autoimmune screen, syphilis serology and toxoplasma serology were normal.

Urgent aqueous biopsy was performed (which confirmed HSV2) with concurrent intravitreal foscarnet treatment.

Systemic antivirals were commenced: intravenous acyclovir followed by valaciclovir 20 mg/kg and subsequent addition of oral prednisolone.

Inflammation improved initially with treatment but progressed to secondary retinal detachment at two weeks post presentation. Prior to retinal detachment, the patient described worsening vision (to perception of light) but with stable ocular findings (no retinal detachment and improving ocular inflammation). Electrophysiology at this time was consistent with vision of 6/36 RE possibly indicating functional overlay.

HSV2 associated acute retinal necrosis (ARN) is a rare condition, particularly in children, and may present with no previous significant ocular or medical history. Functional overlay has the potential to further complicate management, especially in such complex cases.

Time for questions: wed 16.25 - 16.35

Development of a low-cost simulation model for neonatal speculum insertion

H St Ledger (O), S Jain (C), R Jolly (C), Royal Free London NHS Trust

Retinopathy of Prematurity (ROP) screening is a vital service for premature and low birth weight babies to identify early changes in the vasculature of the retina. One of the challenges we encountered while training orthoptists to participate in the service was the use of a neonatal speculum. We have devised a low-cost, high-fidelity model to help simulate this technique and up-skill clinicians.

A linear horizontal superficial incision was made in the skin of the grape using a no 15 blade to replicate the palpebral fissure. The opening created, allowed a neonatal speculum to be repeatedly placed and removed whilst observing for any underlying trauma to the grape.

The model was used by trainees, orthoptists, and non-medical individuals, and their pre and post procedure confidence was evaluated. We derived validity of the simulation by speaking to consultants and assessing their opinions with regards to how reliable the model was.

We observed an increased confidence in using the neonatal speculum following practice on the model eye.

This simple, readily available, and cost-effective simulation model was demonstrated to have a real-life clinical application, upskilling orthoptists and ophthalmic trainees, and to improve their confidence before utilising this skill in a clinical setting

We advocate the use of this model to help improve the confidence of clinicians and the wellbeing of the patient.

Time for questions: Thu 13.20 - 13.35

**The utility of Anti adalimumab antibodies (AAA) testing in paediatric uveitis patients
H.Razzouk (SAS), J.Ashworth (C), Manchester Royal Eye Hospital**

Adalimumab is well established as a biologic immunosuppressive treatment for children with non-infectious uveitis. It is increasingly recognised that treatment failure can be caused by the development of anti-adalimumab antibodies. We aimed to determine how often we test for AAA, how often AAA are positive, and the clinical utility of the test
Retrospective study of patients in the paediatric uveitis clinic who were on Adalimumab treatment for non-infectious uveitis. AAA was tested in the clinic when there was a concern about persistence or recurrent inflammation, or compliance issues

57 patients were on adalimumab as monotherapy (14%), Adalimumab with other DMARD treatment 85.8%, 92.98% of patient had JIA/idiopathic uveitis, 22.80% of patients were tested for AAA. 46% of those tested were positive for AAA (10.52% for total number of patients)

The time between starting the treatment and the presence of AAA varied between 3 months to 12 years. Higher levels of AAA were associated with lower levels of serum Adalimumab. However, some cases had negative AAA with low level of Adalimumab 50% of AAA positive patients were on both methotrexate and Adalimumab, 33.3% were on Adalimumab as monotherapy, 16% were on mycophenolate with Adalimumab
The development of Anti-Adalimumab Antibodies may be a factor when treatment failure occurs in paediatric uveitis. It may develop from 3 months after treatment until many years (12 years in our study). Further study is needed to assess the risk factors of developing AAA and whether a combination with DMARD treatment would prevent from this process

Time for questions: Thu 15.40 - 15.55

**Superior Oblique Tuck Surgery Outcomes: A 7-Year Data
D Gogi (C), R Lewis (O) J Hoole (O), I Simmons (C), St James's University Hospital,
Leeds**

To report the surgical outcomes of the superior oblique tuck procedure in the management of superior oblique palsies performed at a single centre over a 7-year period.

This is a retrospective study of superior oblique tuck performed over a 7-year period between 2016-2023 at a single centre. We evaluated patient demographics, the angle of deviation pre- and post-surgery in prism dioptres (PD), complications (iatrogenic Brown's Syndrome) and improvement of diplopia/ head tilt post-operatively.

18 eyes from 18 patients were identified. More than 60% of cases were congenital superior oblique palsy. 16/18 patients (96.91%) displayed a reduction in angle of deviation post-operatively and improvement of diplopia & head tilt. 4/18 patients (22.22%) experienced post-operative iatrogenic Brown's syndrome but none of these required further corrective surgery. Overall, 4/18 patients (22.22%) required additional extraocular muscle surgery to reduce diplopia further.

Our centre started Superior oblique tuck surgery in 2016. This data shows promising initial results of this surgery with excellent surgical outcomes with regards to reduction of the angle of deviation, diplopia, head tilt and limited complications. It is now the preferred surgical option in our centre, in Superior oblique palsy with incomitant vertical deviations in downgaze.

Time for questions: Fri 13.15 - 13.30

Primary Isolated Amyloidosis of Lateral Rectus: A Case Report

Devina Gogi (C), Janice Hoole (O), Ian Simmons (C), St James's University Hospital, Leeds

We report a case of external ophthalmoplegia in a woman due to an uncommon form of amyloidosis exclusively affecting the lateral rectus muscle.

A 46-year-old woman was treated as orbital myositis of lateral rectus of the right eye with oral steroids & NSAIDs as her orbital magnetic resonance imaging showed fusiform enlargement of the right lateral rectus muscle, without tendon involvement. She had right lateral rectus resection to correct her distance esotropia and did well initially but presented 3 years later with significant limitation of abduction of the right eye which increased exponentially with time. Routine laboratory examinations were unremarkable. A systemic work-up showed no evidence of systemic amyloidosis. However, extraocular muscle/ conjunctival biopsy confirmed the diagnosis of amyloidosis.

Primary isolated amyloidosis in the extraocular muscle is exceedingly rare; only 13 such cases without systemic involvement have been reported in literature, over the last four decades. This condition should be included in the differential diagnosis of extraocular muscle enlargement and tissue biopsy should be performed for diagnostic purposes.

Time for questions: Fri 13.30 - 13.45

They say my child can't see

P Anketell (O), J Jackson (Op), S George (C), on behalf of the NOVIC team, Belfast Health & Social Care Trust, Belfast

Cerebral visual impairment (CVI) is the leading cause of visual impairment in children in the developing world (Rahi et al 2003). Visual behaviours in CVI are impacted on by individual's current health and physical needs. In 2015, the neuro-ophthalmic vision impairment clinic (NOVIC) was developed in Belfast. NOVIC is a multidisciplinary clinic providing a collaborative investigation between ophthalmic and paediatric specialists shared through an outcome report.

Visual findings are recorded for all patients attending NOVIC. This study describes the visual outcomes of children with a diagnosis of CVI attending NOVIC between 2015 and 2019 inclusive. The database was interrogated to report ophthalmic findings, refractive error, presence of strabismus and visual field defects.

111 children (median age=4.71 years, range 0.29-17.09, female=46) attended the clinic. N=98 had a diagnosis of CVI. Visual ability ranged from 0.3logMAR to blindsight. The following findings were noted for those with CVI; 26.5% (n=26) had ocular anatomical changes, 76.5% of cases had a significant refractive error issued, 80.6% had strabismus, 63.3% had a visual field defect.

This study has provided an outline of the visual functions of patients with a diagnosis of CVI assessed in an MDT environment. The NOVIC MDT clinic facilitates collaboration between specialities and addresses common questions of 'what can my child see?' Outcomes from the clinic have influenced input for patients beyond health including education and housing. Further analyses of these subjective measures is underway which will provide a detailed picture of a large population of children with CVI.

Time for questions: Fri 13.45 - 14.00

Dominant Optic Atrophy: Diagnostic and Therapeutic Approach in the Paediatric Population

N Arruti (C), M Nieves-Moreno (C), P Rodriguez-Solana (RF), E Vallespin (C), S Noval (C), Hospital Universitario La Paz

To analyse the diagnostic and therapeutic approach in children diagnosed with Dominant Optic Atrophy (DOA).

Retrospective review of the paediatric patients' medical records with confirmed DOA seen in the Paediatric Ophthalmology Department in the Hospital Universitario La Paz from the 1st of January 2018 to the 1st January 2022.

A total of 11 children were identified in the study period. The main initial complaint was reduced visual acuity, present in eight patients. Mean visual acuity at baseline was 0.37 in the right eye and 0.35 in the left eye (decimal scale). Mean visual acuity at the end of the study period was 0.38 and 0.35 right and left respectively. Optic Coherence Tomography at the first visit showed a mean retinal nerve fibre layer thickness of 81.6 microns in the right eye and 80.5 microns in the left eye and a mean ganglion cell layer of 52.5 and 52.4 microns right and left respectively. Most common visual field defect found was a centrocecal scotoma and nine out of eleven patients showed bilateral temporal disc pallor at baseline. Six patients are currently being treated with off-label idebenone maintaining visual acuity without any side effects.

After the OPA1 gene was sequenced, seven different OPA1 mutations were identified. Two of those, c.267G>A and c.1406_1407del have not been previously reported.

Our study adds two novel variants to the mutation spectrum of the OPA1 gene. Early diagnosis of DOA is crucial, both for avoiding unnecessary consultations and for an appropriate genetic counselling.

Max Rayne Foyer
Board 17 &18

Time for questions: Fri 13.30 - 13.45

Setting up a modern nurse led retinopathy of prematurity screening service at Imperial College Healthcare NHS Trust

C Hennings (T), P Patel (O), S Wren (C), Imperial College Healthcare NHS Trust

Nationally last year, 7295 babies were eligible for retinopathy of prematurity screening. Increasing premature births, limited resources in screening staff and intensive schedules require alternative approaches. Models for safe and effective neonatal nurse led screening have been touted for near a decade

Within a level 2 neonatal unit a standard operating procedure for babies to be screened by neonatal nurses using a wide field digital retinal imaging (WFDRI) camera was deployed. This required: cost analysis, procurement, competency-based training, secure data management and audit of national guidelines. Babies were concurrently screened with Binocular Indirect Ophthalmoscopy (BIO) to ensure safety.

The Panocam by Visunex Systems was selected. During a 6-month period, 25 babies were screened. All babies received initial and termination examination with BIO and were possible WFDRI. Overall WFDRI was used 43 times for screening. All babies were screened on time and within appropriate intervals. No baby missed reviews and there is no delay in referral for treatment.

Numerous challenges exist but with-it rewards. Choice of WFDRI is a balance between cost, usability and effectiveness. A safe SOP working within established protocols needs scrutiny. Data management ensuring balance of governance and usability is testing. Empowering our workforce to take on this challenge has allowed skill development that was otherwise unavailable. We provide a base model for similar UK units to establish a ROP neonatal nurse lead digital screening program. We have shown it to be safe in its current deployment.

Time for questions: Fri 13.45 - 14.00

A rare case of bilateral optic nerve atrophy

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Optic atrophy is a leading cause of childhood visual impairment. It poses a diagnostic challenge and can be caused by ocular and systemic conditions. We present a case of Bosch-Boonstra-Schaaf Optic Atrophy Syndrome (BBSOAS), a rare neurological disorder characterized by a wide array of clinical features including optic atrophy, developmental delay and intellectual disability.

This 9-year-old girl was referred to us in 2021 for a second opinion regarding unexplained bilateral reduced vision (0.4 and 0.6). She had been first seen age 5months, with an intermittent exotropia and otherwise normal ocular exam. Aged 4years, she was re-referred by optician due to reduced vision (6/10 and 6/15) and found to have fine latent nystagmus and reduced vision bilaterally, with temporal optic disc pallor.

Electrodiagnostics and neuroimaging were reported as normal. Other than monitoring for poor weight gain, she was systemically well. We performed optical coherence tomography (OCT) that showed a global reduction in the retinal nerve fibre and ganglion cell layers, suggesting bilateral optic atrophy. Genetic testing detected a pathogenic deletion of the NR2F1 gene, confirming a diagnosis of autosomal dominant BBSOAS.

This rare case highlights the importance of performing objective tests, like OCT and the usefulness of genetic testing in appropriate cases. As genetic testing has been “democratised”, it also leaves the ophthalmologist as the initial contact for parents with the results of genetic testing- therefore we must be equipped to signpost them to regional counselling services and specialist clinical centres, as well as guiding the family towards appropriate supportive resources.



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