UK Paediatric Glaucoma Society Meeting

26 January 2024 Cavendish Conference Centre London, UK



www.ukpgs.org.uk

UKPGS thanks all its exhibitors for their support at the 2024 meeting.

Representatives will be available in the Whittington Suite throughout the day.



Posters in the Whittington Suite	
1 Comparison of genetic resource utilisation	halmologists Deniz Goodman Miami, USA
2 Traumatic hyphaema	Aminah Iffah Jawaheer Taunton, UK
23 Aniridia and anterior segment dysgenesis:	on union Sirisha Senthil Hyderabad, India

Welcome to UK Paediatric Glaucoma Society 2024



Registration Desk: Open 08:00 - 18:30

Louise Richards will be available if you have any questions about proceedings.

WIFI access

Network: Cavendish WIFI Password: 12345cav

CPD

The Royal College of Ophthalmologists approves UKPGS to award **8** self-accredited points. An e-copy of your CPD Certificate will be emailed to you shortly after the meeting.

Meeting evaluation

Your feedback about the meeting is invaluable to UKPGS and the speakers presenting. Please complete your evaluation form throughout the day and hand it in at the Registration Desk before you leave.

Posters

Please take the opportunity to visit the posters located in the Whittington Suite during the meeting breaks.

Scan to view the day's programme online via the UKPGS website:



UKPGS 2024 Meeting Programme

09:00 - 09:10	Housekeeping announcements & President's welcome	Peng T Khaw, London, UK
09:10 - 10:02	SESSION 1: Research	

Co-Chairs: Sushmita Kaushik, Chandigarh, India and **Patrick Watts**, Cardiff, UK

1	Visual field indices in children with glaucoma compared to that in adult glaucoma patients with comparable retinal nerve fibre layer thickness	Ashok Kumar Singh Chandigarh, India
2	Utility of clinical exome sequencing for non-acquired childhood glaucoma	Sushmita Kaushik Chandigarh, India
3	Comparing the safety and efficacy of combined trabeculotomy and trabeculectomy (CTT) with and without Ologen in paediatric primary congenital and infantile glaucoma	Sirisha Senthil Hyderabad, India
4	Classification of the corneal changes in childhood glaucoma from ultrasound biomicroscopy (UBM) images, and determination of any prognostic value of specific changes	Ashok Kumar Singh Chandigarh, India
5	Distribution of port wine birthmarks and glaucoma outcomes in Sturge Weber syndrome	Daniel M Vu Boston, USA
6	Comparing the accuracy of ChatGPT-generated patient education materials about paediatric glaucoma and paediatric cataracts	Deniz Goodman Miami, USA
	09:47 - 10:02: Session Discussion Panel	

10:05 - 10:57 SESSION 2: Clinical

Co-Chairs: Ray Areaux, Minneapolis, USA and **Velota Sung**, Birmingham, UK

7	Genetic characterisation of ocular anomalies: <i>LTBP2</i> mutations and a unique ocular profile	Sirisha Senthil Hyderabad, India
8	Ab externo suture circumferential trabeculotomy versus gonioscopy-assisted transluminal trabeculotomy in primary congenital glaucoma	Serhat İmamoğlu Istanbul, Turkey
9	Efficacy of bent angle needle goniectomy as a primary and redo surgery for management of paediatric glaucoma (<i>Presented by Mohamed Awwad</i>)	Hend Helal Benha, Egypt
10	Phacomatosis pigmento vascularis: unravelling the clinical and demographic profile of a rare congenital malformation syndrome - a comprehensive series	Sirisha Senthil Hyderabad, India
11	Exploring the impact of retinopathy of prematurity and non-surgical interventions on intraocular pressure in preterm infants	Sushmita Kaushik Chandigarh, India
12	Longitudinal analysis of transition clinic follow-up in a tertiary glaucoma centre	Hussai Aluzri Birmingham, UK

10:42 - 10:57: Session Discussion Panel

11:22 - 12:08 SESSION 3: Tube Symposium

Co-Chairs: Sirisha Senthil, Hyderabad, India and Kenneth Yau, Manchester, UK

13	Ten-year outcome of tube shunt surgery at a tertiary centre in a paediatric population	Hussain Aluzri Birmingham, UK
14	Ologen augmentation of Ahmed glaucoma drainage devices: 1-5-year follow-up	Brenda Bohnsack Chicago, USA
15	A case of AADI (Aurolab aqueous drainage implant) with supramid stenting in a refractory secondary childhood glaucoma in congenital rubella syndrome post penetrating keratoplasty	Manju Anilkumar Madurai, India
16	Early outcomes of the PAUL glaucoma implant in intractable primary congenital glaucoma	Hamidu Hamisi Gobeka Afyonkarahisar, Turkey
17	Outcomes of Aurolab aqueous drainage implant in the management of refractory paediatric angle recession glaucoma, case series study	Mohamed Awwad Benha, Egypt
18	PAUL glaucoma implant effectiveness in an Irish paediatric cohort: a 24-month retrospective cohort study	Melissa Murphy Dublin, Ireland
	11:53 - 12:08: Session Discussion Panel	

12:10 - 13:00 SESSION 4: Tube Failure Symposium

Co-Chairs: Joe Abbott, Birmingham, UK & Hiranya Abeysekera, Colombo, Sri Lanka

19	Long-term outcomes of cyclophotocoagulation in refractory paediatric glaucoma: treatment response and success in 262 eyes	Maram Abdalla Elsayed Jeddah, Saudi Arabia
20	Management of failed Ahmed glaucoma valve in childhood glaucoma	Hiranya Abeysekera Colombo, Sri Lanka
21	Outcomes of revision surgery for aqueous shunt exposure in paediatric onset glaucoma	Alessandra Martins London, UK
22	Unrecognised intermittent glaucoma drainage tube blockage secondary to pharmacological pupil dilation	Ahmed Ali Newcastle Upon Tyne, UK
23	Outcomes following surgical decapsulation of glaucoma drainage device (GDD) for paediatric glaucoma	Samuel Simpson Newcastle Upon Tyne, UK
24	AGV valve slicing - a blessing in disguise	Sirisha Senthil Hyderabad, India

12:45 - 13:00: Session Discussion Panel

13:00 - 13:45

Lunch, Posters, Exhibition

13:20 - 13:45

UKPGS Annual General Meeting

13:45 - 14:05

GUEST LECTURE: Visual rehabilitation and glaucoma outcomes in severe congenital anterior segment developmental disorders

Susmito Biswas

Manchester, UK

14:05 - 14:38 SESSION 5: Teaching Glaucoma Symposium

Co-Chairs: Elena Bitrian, Miami, USA and Alessandra Martins, London, UK

25 Development of a skills transfer course	Deniz Goodman Miami, USA
26 Exam under anaesthesia for paediatric glaucoma: a simulation-based training	Vandana Persad Miami, USA
27 The synthetic eye model	Deniz Goodman Miami, USA
28 Training the next generation of paediatric glaucoma surgeons: an international effort	Elena Bitrian Miami, USA

14:31 - 14:38: Session Discussion Panel

12:38 - 15:20 SESSION 6: Rapid Fire - Research

Co-Chairs: Brenda Bohnsack, Chicago, USA and John Brookes, London, UK

29	Factors and outcomes associated with corneal oedema and Haab's striae in primary congenital glaucoma	Brenda Bohnsack Chicago, USA
30	Genetic changes associated with childhood glaucoma: a systematic review	Anika Kumar San Francisco, USA
31	Medium-term survival of developmental glaucoma treated with ab interno angles surgery: analysis of a large clinical database	Ta Chen Peter Chang Miami, USA
32	Early experience with micro-pulse transscleral diode cyclophotocoagulation for refractory paediatric glaucomas	Ray Areaux Minneapolis, USA
33	Exploring the potential of augmented reality and virtual reality as a diagnostic tool for glaucoma screening in ophthalmology	Krishnika Vetrivel London, UK
34	Value-cost comparison of standard automated perimetry and head-mounted perimetry	Deniz Goodman Miami, USA
35	Analysis of peripapillary vascular density with optical coherence angiography (OCT-A) in paediatric glaucoma. Comparative study with healthy subjects	Flora Xydaki Madrid, Spain
	15:07 15:20: Sossien Dissussien Penel	

15:07 - 15:20: Session Discussion Panel

15:20 - 15:30

Childhood Glaucoma Research Network (CGRN) update Alana Grajewski, Miami, USA

Refreshments, Posters, Exhibition

15:50 - 16:32 SESSION 7: Rapid Fire - Clinical

Co-Chairs: Ta Chen Peter Chang, Miami, USA and Christopher Lyons, Vancouver, Canada

36	Shedding light on ab interno canaloplasty: a novel surgical technique to manage paediatric glaucoma	Arjun Sharma Miami, USA
37	All aniridias are not same: the mysterious world of absent iris!!	Ashok Kumar Singh Chandigarh, India
38	Childhood glaucoma associated with Rubinstein-Taybi syndrome: Long-term outcomes of a case series	Cristina Ginés-Gallego Manchester, UK
39	Congenital glaucoma and Aicardi Goutières syndrome, a case report	Alfonso Miranda-Sánchez Madrid, Spain
40	Rare case of Adams Oliver syndrome with congenital glaucoma	Manju Anilkumar Madurai, India
41	Persistent tunica vasculosa lentis in glaucoma associated with neurofibromatosis - a case report (<i>Presented by Mohamed Awwad</i>)	Nader Bayoumi Alexandria, Egypt
42	Serous detachment of the macula in an Eye with glaucoma associated with port-wine birthmark after Initiating prostaglandin analogue therapy	Ta Chen Peter Chang Miami, USA

16:20 - 16:32: Session Discussion Panel

16:32 - 17:13 SESSION 8: Rapid Fire - Videos

Co-Chairs: Beth Edmunds, Portland, USA and Rizwan Malik, Abu Dhabi, United Arab Emirates

43	PRESERFLO MicroShunt-implantation surgery and a small case series of paediatric patients	Susana Duarte Lisbon, Portugal
44	Uveitis-glaucoma-hyphaema syndrome causing secondary paediatric glaucoma	Pooja Pendri Miami, USA
45	Juvenile xanthogranuloma with neovascular glaucoma masquerading as primary congenital glaucoma	Juan P López Santiago, Chile
46	Clinical evaluation protocol for childhood glaucoma	Manju Anilkumar Madurai, India
47	Surgical management of bilateral angle closure in a nanophthalmous patient during acute phacomorphic glaucoma attack in the right eye versus chronic angle closure	Rayan Abou Khzam Miami, USA

16:58 - 17:13: Session Discussion Panel

17:13 - 17:23

Prizes and acknowledgements

17:23 - 17:54

NOEL RICE LECTURE 2024: Standing on the shoulders of giants

Chris Lyons

Vancouver, Canada

GUEST LECTURE

Visual rehabilitation and glaucoma outcomes in severe congenital anterior segment developmental disorders

Mr Susmito Biswas

Consultant Paediatric Ophthalmologist. Manchester Royal Eye Hospital, Manchester, UK. Honorary Chair, MAHSC, University of Manchester.

Susmito Biswas is the Clinical Lead for the Paediatric Ophthalmology Service at the Manchester Royal Eye Hospital where he has worked as a Consultant for more than 20 years. He is currently Chair of the Royal College of Ophthalmologists Paediatric Subcommittee. He leads the regional Retinopathy of Prematurity Service in Greater Manchester.

He has an interest in paediatric anterior segment disorders, particularly corneal and external eye diseases.

He is one of the leading paediatric corneal transplant surgeons in the United Kingdom, receiving referrals from all over the North of England into his service. He also provides a wide remit of specialist paediatric anterior segment surgery including congenital and infantile cataracts and paediatric glaucoma.

He has published over 70 peer reviewed publications and written several book chapters covering a wide variety of topics in paediatric ophthalmology and anterior segment diseases.

NOEL RICE LECTURE

Standing on the shoulders of giants Professor **Christopher Lyons**

Investigator & Head, Department of Ophthalmology, British Columbia Children's Hospital, Vancouver, Canada. Professor, Department of Ophthalmology and Visual Sciences, Faculty of Medicine, University of British Columbia, Vancouver, Canada.

Born in France and educated in England from the age of 11, Christopher Lyons did his medical training at University College London and Westminster Medical School. After demonstrating anatomy and neuro-anatomy at Middlesex Medical School, he moved to Cambridge to start his ophthalmology training where his teachers included Mr John Cairns and Mr Peter Watson. He continued his Residency training at Moorfields Eye Hospital for five years, and subsequently did Fellowships in Paediatric Ophthalmology in Vancouver with Dr John Pratt Johnson, and Neuro-Ophthalmology in San Francisco with Drs Creig and Bill Hoyt. He finished his training with an Oculoplastic Fellowship at Moorfields with Mr Richard Collin.

He was appointed to the academic/clinical Staff at the University of British Columbia and Children's Hospital in Vancouver, Canada, where he has been based for 30 years, practicing as a Paediatric Ophthalmologist. On arrival, his colleague Dr Gordon Douglas immediately generously shared his Paediatric Glaucoma practice with him, nurturing his interest in paediatric glaucoma and assisting with appropriate surgical training in this sub-specialty. From 1994, Dr Lyons became responsible for the treatment of paediatric glaucoma for British Columbia. From the start, he has studied glaucoma techniques, including the indications and outcomes of goniotomy for a wide range of paediatric glaucoma types, and the use of mitomycin as a surgical adjunct to Ahmed valve implantation and revision.

Dr Lyons has published widely in paediatric ophthalmology including paediatric glaucoma. As a committed teacher, he has trained over 40 Paediatric Ophthalmology Fellows and many more Ophthalmology Residents. He was appointed to the rank of Professor in the faculty of Medicine in 2006, was UBC Residency Training Director for 12 years and, more recently, interim UBC Department of Ophthalmology Chairman from 2021 to 2023. He is co-editor of the 5th and 6th (current) editions of *Taylor and Hoyt's Pediatric Ophthalmology and Strabismus* and Associate Editor of the *Journal of the American Association for Pediatric Ophthalmology and Strabismus*. With his wife Philippa and three children Madeleine, Theo and Alex, he enjoys living a full life which includes many outdoor activities in beautiful British Columbia.

1 Visual field indices in children with glaucoma compared to that in adult glaucoma patients with comparable retinal nerve fibre layer thickness Ashok Kumar Singh

U Thakur, F Thattaruthody, SS Pandav, S Kaushik Advanced Eye Centre, Post Graduate Institute of Medical Education & Research, Chandigarh, India. Correspondence: ashoksingh1603@gmail.com

Purpose: To determine visual field defects detected by Humphrey's visual field analyzer (HFA) in children with glaucoma and compare them with visual fields (VF) in adults with comparable retinal nerve fibre layer thickness (RNFLT) on optical coherence tomography (OCT).

Methods: This cross-sectional study included 46 eyes of 35 glaucomatous children aged 6-18 years and RNFLT-matched adult eyes with glaucoma. The children underwent VF and RNFLT measurement, whereas the adults' data for VF and OCT were retrieved from the database. The main outcome measures were mean deviation (MD), reliability indices, time to do the test in each child and their comparison with RNFLT-matched glaucomatous eyes of adults. Children 6-11 years and 12-18 years were analysed separately.

Results: The mean age of the children was 11.37 ± 2.31 years. The average MD of children was significantly less than that of the normal adults despite having the same RNFL thickness (-7.86±3.96dB vs. -4.56±2.76dB, p=0.001). The difference between average MD in children aged 6-11 years and RNFLT-matched adults was significant (-6.91±3.36dB vs. -4.84±2.16dB, p=0.005). Similarly, the difference between children aged 12-18 years and RNFLT-matched adults was also significant (-9.20±2.73dB vs. -4.64±2.36dB; p=0.001). The average time taken in both subgroups of children was similar. The visual field indices were significantly worse in children aged 12-18 years.

Conclusions: Children appear to have lesser retinal sensitivity than adults. The VF appears worse in children aged 12-18 years than adults or younger children. It is worthwhile to consider developing a separate paediatric normative database for the VF assessment of children under 18 years.

2 Utility of clinical exome sequencing for nonacquired childhood glaucoma

Sushmita Kaushik

A Kumar Singh, S Choudhary, J Singh, F Thattaruthody, SS Pandav Advanced Eye Centre, Postgraduate Institute of Medical

Education and Research, Chandigarh, India. Correspondence: sushmita_kaushik@yahoo.com

Purpose: To study pathogenic variants detected by clinical exome sequencing (CES) in a cohort of non-acquired glaucoma (NAG) and to analyse their correlation with the phenotype and outcome.

Methods: In this prospective cohort study, children who presented with newly diagnosed non-acquired glaucoma between January 2021 and January 2023 underwent targeted gene capture sequenced on an Illumina sequencing platform (CES). Sequences were aligned to the human reference genome (GRCh38.p13). The pathogenicity of variants was determined using ACMG guidelines and targeted variant analysis was done by PCR and Sanger sequencing. The phenotype was studied during examination under anaesthesia (EUA). We correlated genetic variants to the phenotype and analysed the concordance.

Results: 171 children were analysed. One hundred and twenty-six children (73.7%) harboured genetic variants, of which 98 (77.8%) matched the phenotype; classified as pathogenic. Of these pathogenic variants, 54 (55.1%) were CYP1B1, 9 (9.2%) were PAX6, and six each (6.1%) were FOXC1 and LTBP2. Three children each (3.1%) had PITX2, FOXE3 and NF1 variants, while two had MYOC, COL2A and VCAN variants. Eight children had one miscellaneous variant each. CES was helpful in phenotype characterisation in eleven children in whom the actual diagnosis was missed on initial examination. Twenty-eight children (22.2%) harboured genetic variants of uncertain significance, which did not match the phenotype despite re-examination.

Conclusions: CES is a valuable strategy for identifying genetic variants in childhood glaucoma. One quarter of the children in our cohort had no variants on CES and may benefit from whole exome sequencing. Deep phenotyping and accurate genotyping may help open avenues for gene therapy.

3 Comparing the safety and efficacy of combined trabeculotomy and trabeculectomy (CTT) with and without Ologen in paediatric primary congenital and infantile glaucoma

Sirisha Senthil AK Mandal, R Krishnamurthy, V Prasad Sisodia *LV Prasad Eye Institute, Hyderabad, India.* Correspondence: sirishasenthil@lvpei.org

Aim: This single-centre randomised control study aimed to compare the safety and efficacy of two surgical approaches for the treatment of primary congenital and infantile glaucoma.

Methods: A total of 55 eyes from 55 children aged 1 month to 3 years were enrolled in the study, with patients randomised into two groups. Group A received combined trabeculotomy with trabeculotomy (CTT), while Group B underwent CTT with adjuvant Ologen Collagen Matrix. The minimum follow-up duration was 12 months during which intraocular pressure (IOP), corneal clarity, and the number of antiglaucoma medications (AGMs) were assessed at each visit.

Results: The median age at the time of surgery was 4 months for both groups. Baseline measures including median IOP, corneal diameter and corneal clarity were similar between the two groups (p=0.2). After 12 months the median IOP was 10 mmHg in Group A and 11 mmHg in Group B, with no significant difference (p=0.4). Both groups had a median number of AGMs of 0 and grade 1 corneal clarity. The complete success rate was 87% for Group A and 78% for Group B, while the qualified success rate was 93% for Group A and 94% for Group B. Six eyes in each group required topical AGMs with no sight-threatening complications observed and none of the eyes necessitated repeat surgery.

Conclusion: In conclusion, both surgical procedures, CTT alone and CTT combined with Ologen demonstrated similar success rates as primary interventions for primary congenital and infantile glaucoma at the one-year follow-up. The study found no additional benefits associated with the adjuvant use of Ologen when employed alongside CTT.

Classification of the corneal changes in childhood glaucoma from ultrasound biomicroscopy (UBM) images, and determination of any prognostic value of specific changes Ashok Kumar Singh F Thattaruthody, SS Pandav, S Kaushik

Advanced Eye Centre, Post Graduate Institute of Medical Education & Research, Chandigarh, India. Correspondence: ashoksingh1603@gmail.com

Purpose: To classify congenital corneal opacities in childhood glaucoma using ultrasound biomicroscopy (UBM) and to analyse the prognostic value of UBM.

Design: Retrospective cohort study. All childhood glaucoma patients who underwent UBM at the presentation from 2017 to 2023 were included. UBM images were compared with clinical photographs. Surgical outcome at 1 year after surgery were analysed as well. UBM images were classified according to new classifications which we propose:

Type 1 - Diffuse corneal thickening

Type 2 - Diffuse corneal thickening with intracorneal hyperreflectivity

Type 3 - Presence of Descemet's membrane discontinuity Type 4 - Presence of Descemet's and posterior stromal

defect

Type 5 - Presence of intracorneal clefts

Type 6 - Miscellaneous

9

Results: A total of 70 eyes of 43 children were included. Mean age of presentation was 5.9 months, and mean IOP was 22.78 mmHg. The majority of the children had corneal clarity of grade IV (47.14%) followed by III (24. 3%). Peters anomaly (27/70) was the most common diagnosis followed by PCG (17/70) and CEU (16/70). The majority of children having UBM types 3 - 6 had Peters anomaly whereas type 1 and 2 were majorly PCG or CEU. Children with UBM type 1 and type 2 had relatively good to fair outcome (59.5%), whereas types 3 - 6 had very poor outcome (53.13%). Children with type 1 and 2 performed well with CTT or angle surgery, whereas UBM types 3 - 6 required multiple surgeries and often required pupilloplasty.

Conclusions: We concluded that UBM types 3 - 6 were majorly Peters anomaly, had poor surgical outcome and required multiple surgeries. So, it is worth considering developing a uniform classification method of childhood glaucoma based on UBM.

5 Distribution of port wine birthmarks and glaucoma outcomes in Sturge Weber syndrome

Daniel M Vu

H Gjerde, AM Elhusseiny, I Oke, DK VanderVeen Boston Children's Hospital, Harvard Medical School, Boston, USA.

Correspondence: daniel_vu@meei.harvard.edu

Purpose: To identify which features of Sturge Weber Syndrome (SWS) were most associated with glaucoma onset, severity, and treatment failure at our tertiary care centre.

Methods: A retrospective chart review was performed for all children with SWS presenting between 2014-2020. Examination and imaging findings from dermatology, neurology, and ophthalmology were collected. Primary outcomes included factors associated with glaucoma development, progression to surgery, and treatment failure. Failure was defined as having a final intraocular pressure >21 mmHg, devastating complication, or ≤20/200 vision. Non-parametric tests and logistic regressions were used to detect statistical differences between groups.

Results: Twenty-three of 44 SWS patients (52.3%) developed glaucoma, and 6 (26.1%) had both eyes affected. Sixteen of 29 eyes (55.2%) required surgery, and 26.9% overall met our failure criteria (mean follow-up: 5.2±4.3 years). Glaucoma diagnosis was associated with bilateral port wine birthmarks (PWB; OR 5.9; 95%CI 1.3-43.2), PWB with any lower eyelid involvement (OR 9.7, 95%CI: 2.6-44.5), and choroidal haemangiomas (OR 3.8, 95%CI: 1.1-13.8), but was not associated with upper eyelid or leptomeningeal angiomas, seizures, prior hemispherectomy or pulsed-dye laser. Eyes that progressed to surgery were more likely to have PWB affecting the lower eyelid (OR 33.7, 95%CI: 4.5-728.0). No differences were found between eyes that did or did not fail therapy. In most cases, angle surgery failed but was a temporising measure before subconjunctival surgery. Discussion: Lower eyelid and choroidal angiomas were associated with greater glaucoma severity, hinting at nearby aqueous outflow impairment. However, upper eyelid and

leptomeningeal angiomas were not associated.

6 Comparing the accuracy of ChatGPT-generated patient education materials about paediatric glaucoma and paediatric cataracts Deniz Goodman

AL Grajewski, E Bitrian, EJ Savatovsky, TCP Chang Bascom Palmer Eye Institute, Miami, USA. Correspondence: dgm@bu.edu

Purpose: Evaluate the accuracy of ChatGPT responses to parents' frequently asked questions (FAQ) about paediatric glaucoma (PG) or cataracts (PC).

Methods: Ten FAQ and responses published on websites of national ophthalmology organisations and hospitals were compiled per diagnosis in this cross-sectional study. Questions were individually entered into separate ChatGPT-3.5 chats with and without the prompt: "Be specific and incorporate any applicable medical guidelines". Fourteen PC specialists graded responses to PC FAQ sentence by sentence as true or false, while 18 PG specialists, 7 glaucoma specialists, and 4 paediatric ophthalmologists similarly graded PG FAQ responses. The Mann-Whitney U test was used to compare the average accuracy between unprompted ChatGPT, prompted ChatGPT, and reference website responses.

Results: Among all PC questions, there was no significant difference between website responses (92.7%) and the accuracy of unprompted (93.4%, p=0.970) or prompted (91.4%, p=0.521) ChatGPT responses. Among all PG questions, the accuracy of both unprompted (96.1%, p=0.026) and prompted (94.6%, p=0.021) ChatGPT responses was significantly higher than the accuracy of website responses (88.5%). There was no significant difference between the accuracy of unprompted and prompted ChatGPT responses for PC (p=0.212) or PG (p=0.241) questions.

Conclusions: Unlike ChatGPT-generated PC information, ChatGPT-generated PG information is more accurate compared to information on reference websites. Given that ChatGPT is continuously updated, these results may vary over time and the public should be cautious when using online resources for medical information.

7 Genetic characterisation of ocular anomalies: *LTBP2* mutations and a unique ocular profile Sirisha Senthil

P Walvekar, A Varma, V Pochaboina, DC Prameshwarappa LV Prasad Eye Institute, Hyderabad, India. Correspondence: sirishasenthil@lvpei.org

Aim: This study aimed to uncover the genetic basis of a distinctive ocular condition featuring congenital megalocornea, persistent pupillary membrane, gross iridodonesis, ectopia lentis, and secondary glaucoma.

Methods: An 18-child cohort from 14 consanguineous families (age: 4 months to 12 years) underwent comprehensive ocular assessments and genetic analysis by whole exome sequencing validated by Sanger sequencing.

Results: The cohort displayed consistent ocular characteristics, including megalocornea, ectopia lentis, gross iridodonesis (100%), and high incidences of persistent pupillary membrane (78%), ectropion uveae (19%), and secondary glaucoma (72%). Lensectomy effectively controlled intraocular pressure when treated less than 2 years of age. 36% of eyes required glaucoma surgeries. Concurrent retinal issues affected 36% of eyes, with 20% developing retinal detachment after lensectomy or glaucoma surgery, and 5% experiencing suprachoroidal haemorrhage following primary glaucoma surgery.

Genetic analysis identified *LTBP2* gene variations in all 18 children, comprising 7 missense, 4 duplications, and 2 deletions characterised as pathogenic. These variations were uniform among affected siblings. None of these variations were found in population genome databases like the 1000 Genome Database or the ExAC database. These variations possibly affect functions related to microfibrils and elastin fibres, causing the typical phenotypic changes.

Conclusion: In conclusion, this study emphasises the need to distinguish this unique ocular phenotype linked to *LTBP2* genetic mutations from primary congenital glaucoma. Timely clinical diagnosis, targeted genetic testing, and lensectomy as the primary intervention are crucial. Comprehensive retinal assessment and prophylactic laser therapy can help prevent vision-threatening complications in affected eyes. This research contributes to our understanding of this distinct ocular condition and its genetic underpinnings.

8 Ab externo suture circumferential trabeculotomy versus gonioscopy-assisted transluminal trabeculotomy in primary congenital glaucoma Serhat İmamoğlu¹

A Olgun², HH Gobeka³, S Kuğu⁴

1 - Haydarpaşa Numune Teaching and Research Hospital, University of Health Sciences, Istanbul, Turkey. 2 - West Eye Hospital, Erbil, Turkey. 3 - Afyonkarahisar Health Sciences University, Afyonkarahisar, Turkey. 4 - Ophthalmology Clinic, Istanbul, Turkey.

Correspondence: ophserhat@hotmail.com

Background: To compare clinical outcomes of ab externo suture circumferential trabeculotomy (CT) versus gonioscopy-assisted transluminal trabeculotomy (GATT) in primary congenital glaucoma (PCG).

Methods: This retrospective study included 40 eyes from 34 patients receiving ab externo suture CT (Group 1, n=28) or GATT (Group 2, n=12) for PCG at Haydarpaşa Numune Teaching and Research Hospital, Istanbul, between September 2020 and September 2022. Success criteria was defined as complete success when an intraocular pressure (IOP) was ≤21 mmHg without anti-glaucoma medications (AGMs) and partial success when the IOP was ≤21 mmHg with AGMs. A need for further surgical intervention was regarded as failure.

Results: The median ages in Groups 1 and 2 were 6.5 (5-11.75) and 6 (1.5-21) months, respectively. Males (68.2%) predominated in Group 1, whereas the gender ratio in Group 2 was 1:1. Group 1 had a median follow-up duration of 14.5 (11-18) months, while Group 2 had 12.5 (1.25-14) months. Both groups achieved significantly decreased IOP [(Group 1: 27 (23.25-31.5) to 10.5 (8-15.5) mmHg, p<0.001) and (Group 2: 29.9 (28-31.5) to 13 (13-18.5) mmHg, p=0.013]. This also applied to AGM counts in Groups 1 and 2, which decreased from 3 (2-3) to 0 (0-1.5) and 3 (3-3) to 0 (0-2.75), respectively. Complete success was achieved by 83.3% in Group 1 and 82.1% in Group 2 (p=0.927).

Conclusion: Despite a comparable complete success ab externo suture CT had a greater qualified surgical success rate in treating PCG than GATT. Regardless, continuous monitoring is required for the long-term success of any PCG intervention.

9 Efficacy of bent angle needle goniectomy as a primary and redo surgery for management of paediatric glaucoma

Hend Helal M Awwad *Benha University, Benha, Egypt.* Correspondence: Hendhelal75@gmail.com

Purpose: To evaluate effectiveness of bent angle needle goniectomy (BANG) in treating congenital glaucoma, whether as primary or secondary treatment after previous failed surgeries.

Methods: This study focused on paediatric glaucoma patients who had BANG at the Benha Glaucoma Unit between 2020 and 2022. Patients were divided into two groups: Group A included patients who underwent BANG as their primary procedure, while Group B included patients who underwent BANG as a secondary procedure after previous failed glaucoma surgeries. Group A was further divided into neonatal (A1) and infantile (A2), while Group B was subdivided into post-trabeculotomy (B1) and post-trabeculectomy (B2). Success was determined by intraocular pressure (IOP) of 20 mmHg or less with or without medications.

Results: 48 eyes of 42 children were examined. Both groups experienced a significant reduction in IOP after BANG. At the end of one-year follow-up, groups A and B showed a 52.4% and 50.1% reduction in IOP, respectively. Success rates for primary and re-do surgery were 75% and 62.5%, respectively. Success rate was higher in group A1 (81.25%) than in group A2 (62.5%). On the other hand, post-trabeculectomy was more successful (64.28%) than post-trabeculectomy (60%). Failure rate was higher and occurred earlier in bilateral cases, eyes with higher baseline IOP, longer axial length, larger corneal diameter, positive consanguinity, and poor compliance to postoperative steroid treatment. There were no significant intra- or postoperative complications.

Conclusion: BANG is an effective surgery for managing paediatric glaucoma. It can also be used as a secondary surgery after failed previous surgeries.

10 Phacomatosis pigmento vascularis: unravelling the clinical and demographic profile of a rare congenital malformation syndrome a comprehensive series Sirisha Senthil

AK Mandal, P Kolipaka, D Molleti LV Prasad Eye Institute, Hyderabad, India. Correspondence: sirishasenthil@lvpei.org

Aim: Phacomatosis pigmento vascularis (PPV) is a rare neurocutaneous syndrome characterised by the coexistence of pigmentary nevi and capillary malformations, often accompanied by multisystemic involvement. Ocular manifestations in PPV are strongly associated with glaucoma. This retrospective case-study analysed the clinical profile and demographics of the largest cohort of PPV patients to date.

Methods: Over a 27-year period we examined 71 patients (137 eyes) with PPV at a tertiary care centre. Findings from structured case records revealed a patient population consisting of 22 females (30.9%) and 49 males (69.1%), with a median age of 1 year (range: 4 days to 40 years). All patients exhibited bilateral pigmentary and vascular malformations, with 70.4% presenting with bilateral glaucoma and 29.5% with unilateral glaucoma. Median visual acuity and intraocular pressure at presentation were documented.

Results: Systemic abnormalities, including epilepsy and MRI abnormalities were observed in a significant proportion (44%) of patients. Surgical intervention for intraocular pressure control was necessary in approximately two-thirds of the patients, 76% underwent combined trabeculotomy with trabeculectomy and 22% trabeculectomy. Post-surgical outcomes indicated a significant reduction in antiglaucoma medication use and intraocular pressure. However, surgical complications occurred in 13% eyes, some of which were sight-threatening.

Conclusion: In this PPV cohort, a majority of patients presented with bilateral glaucoma and systemic abnormalities. Approximately two-thirds of patients required glaucoma surgery, with a few sight-threatening complications. This study underscores the need for thorough ocular and systemic evaluations in all PPV patients and highlights the importance of close glaucoma follow-up.

11 Exploring the impact of retinopathy of prematurity and non-surgical interventions on intraocular pressure in preterm infants Sushmita Kaushik

A Kumar Singh, P Verma, D Katoch, P Kumar, F Thattaruthody, SS Pamdav Advanced Eye Centre, Postgraduate Institute of Medical Education and Research, Chandigarh, India. Correspondence: sushmita_kaushik@yahoo.com

Aim: To measure the intraocular pressure (IOP) in preterm infants with and without retinopathy of prematurity (ROP) and assess the effect of non-surgical treatment on the IOP.

Methods: This prospective study included infants with gestational age (GA) <34 weeks and birth weight (BW) <2000 grams presenting between 1st July 2021 and 30th June 2022. Those with ocular or systemic comorbidities and severe ROP requiring surgery were excluded. Eligible infants were divided into three groups according to the status of ROP. Group 1: No ROP. Group 2: ROP not requiring treatment. Group 3: ROP requiring non-surgical treatment. IOP was measured using a Perkins tonometer at presentation, one-month and three-month follow-up.

Results: 107 patients met the inclusion criteria; 40 (37.38%) with no ROP, 25 (23.36%) with ROP not requiring treatment, and 42 (39.25%) with ROP requiring laser photocoagulation or Anti-VEGF injection. The mean post-menstrual age (PMA) in the three groups was 37.89+2.74, 36.98+3.38, and 35.47+2.84 weeks respectively. In Group 1, the mean IOP at baseline, one-month, and three months follow-up was 15.0+2.43 mmHg, 14.29+2.25 mmHg (p=0.01), and 14.24+1.94 mmHg, respectively. In Group 2, the mean IOP at baseline, one-month, and three months follow-up was 14.95+2.89 mmHg, 14.61+3.15 mmHg, and 13.55+2.10 mmHg, respectively. In Group 3, the IOP rose transiently after intervention (13.46+2.54 mmHg to 15.40+2.76 mmHg (p<0.001)), which decreased to 14.31+1.87 mmHg at three months.

Conclusions: IOP in preterm infants declines significantly with maturity, likely due to the development of outflow channels. The IOP elevation after laser treatment must be anticipated and treated in time.

13

12 Longitudinal analysis of transition clinic follow-up in a tertiary glaucoma centre Hussai Aluzri

Jay Richardson, Velota Sung Birmingham Midlands Eye Centre, Birmingham, UK. Correspondence: hussain.aluzri2@nhs.net

Introduction: Due to risk of blindness in paediatric glaucoma, it is important to carefully manage the transition of patients from paediatric to adult care. During this period of transition, patients are often required to assume greater responsibility for their own conditions. We looked at a transitioning cohort over 5 years with the aim to identify any gaps in their management or progression in their glaucoma. **Methods:** We retrospectively reviewed electronic patient records for all patients who attended the transition clinic between 2017 to 2023. We analysed the following parameters: IOP, visual acuity, C:D ratio and the type or duration to further surgical procedures.

Results: We identified 27 transitioning patients (17 male, 9 female) with 54 eyes. The modal diagnosis (12 patients) was glaucoma secondary to: cataract surgery (8), uveitis (3) and silicone oil (1). Additional primary diagnoses included congenital glaucoma (5), anterior segment dysgenesis (2), juvenile open-angle glaucoma (2) and other rare ocular syndromes (2). Six patients were awaiting planned adult clinic follow-up; however two cases were lost to follow-up with one other seen at a local adult-clinic instead. The average time between transition and adult clinic follow-up was 86 days. At transition, average VA was 0.55 logMAR with an IOP of 17.25 mmHg, requiring 2.3 medications. Five patients (18%) required further surgical intervention with an 80% success rate.

Conclusion: This is the first study that investigates the efficacy of transition clinics, the risks of loss to follow-up, as well as quantifying the rate of further operations within this cohort.

13 Ten-year outcome of tube shunt surgery at a tertiary centre in a paediatric population

Hussain Aluzri¹

Shayan Samroo², Jay Richardson¹, Velota Sung¹

- 1 Birmingham Midlands Eye Centre, Birmingham, UK.
- 2 West Midlands Foundation School, Birmingham, UK.

Correspondence: hussain.aluzri2@nhs.net

Introduction: Glaucoma drainage devices are commonly employed in the management of paediatric glaucoma. We aim to evaluate the long-term (ten-year) outcomes in a paediatric cohort.

Methods: This is a single-centre retrospective analysis of (n=102 eyes) consecutive cases of glaucoma drainage device insertion (87 Baerveldt & 15 Molteno tubes) between 2003-2013. All cases were primary tube surgeries undertaken in children/adolescents (<18 years old) and included a minimum of 10-years follow-up.

Primary outcomes included: Complete success (IOP 5-21 mmHg AND >20% reduction WITHOUT medications), Qualified success (IOP 5-21 mmHg AND >20% reduction WITH medications) and Failure (IOP in excess of success criteria, further glaucoma procedures, NPL vision). Secondary outcomes included: Visual acuity, IOP, medications, cup-to-disc ratio and complications.

Results: 102 eyes of 82 patients were included with an average follow-up of 13.2 years. Preoperatively, the mean age was 9.5 years old. Causes of glaucoma were heterogeneous in this complex cohort (mean pre-op: VA 0.50 logMAR, IOP 29.4 mmHg, number of medications 2.8). At ~10-year follow-up, IOP and medications were significantly reduced (p<0.05) with mean IOP of 12.7 mmHg requiring 0.97 medications. The complete success rate was 34.2% with a qualified success of 65.7%. Of the 30 eyes which failed, rates of re-operation and progression to no perception of light were collated. 34 eyes required tube-shunt revision surgery.

Conclusions: This research represents the most extensive long-term evaluation of paediatric tube shunt surgery. Our results demonstrate the safety and efficacy of this procedure.

14

14 Ologen augmentation of Ahmed glaucoma drainage devices: 1-5-year follow-up

Brenda Bohnsack

Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, USA. Correspondence: bbohnsack@luriechildrens.org

Purpose: Ologen augmentation of Ahmed glaucoma devices (OAGD) has previously been shown to increase the short-term success of Ahmed glaucoma drainage devices (GDDs) in children. In this study, we present longer follow-up with a greater number of paediatric eyes.

Setting: Academic/University Children's Hospital.

Methods: Retrospective interventional case series of children (<18 years) who underwent OAGD between 2018 and 2022 with >1-year postoperative follow-up. Outcomes included intraocular pressure (IOP) and the number of IOP-lowering medications. Complete success was defined as IOP of 5-20 mmHg without glaucoma medications, visually devastating complication or additional IOP-lowering surgeries. Qualified success was defined as above except IOP control was maintained ± glaucoma medications.

Results: A total of 42 eyes of 28 patients underwent OAGD at median age of 2.4 years. Diagnoses included primary congenital glaucoma (12 eyes) and glaucoma secondary to non-acquired ocular anomalies (11 eyes), non-acquired systemic anomalies (12 eyes), following cataract surgery (3 eyes), and acquired conditions (4 eyes). Twenty-two eyes had previous glaucoma surgery (average 1.8 ± 0.7 surgeries per eye). Preoperative IOP was 29.3 ± 9.1 mmHg on an average of 2.8 ± 1.0 glaucoma medications. At final follow-up (2.0 ± 1.1 years, median 1.7), IOP (13.6 ± 4.8 mmHg) and glaucoma medications (0.5 ± 0.9 , median 0) were significantly decrease (p<0.0001).

Conclusions: OAGD shows continued IOP control in children with complete and qualified success rates of 67% and 90%, respectively. Survival with or without glaucoma medications was greater than 90% at 1 and 3 years of follow-up.

15 A case of AADI (Aurolab aqueous drainage implant) with supramid stenting in a refractory secondary childhood glaucoma in congenital rubella syndrome post penetrating keratoplasty Manju Anilkumar

M Udauy, K Naidu *Aravind Eye Hospital, Madurai, India.* Correspondence: manju@aravind.org

Case: A 1-year-old female child was referred for uncontrolled IOP in the left eye from elsewhere, had congenital rubella syndrome and had been operated on for PKP (penetrating keratoplasty) in both the eyes for congenital corneal opacity with sclerocornea, ultimately developing secondary glaucoma in the left eye. The baby was born out of a second-degree consanguineous marriage, full term caesarean delivery, with a birth weight of 2 kg. She was the second child by birth order. The baby had a NICU stay for 5 days with delayed developmental milestones. Her mother gave a history of fever during the fifth month of gestation. MRI Brain showed mild cortical atrophy with thinning of corpus callosum and was diagnosed with congenital heart disease with supravalvular pulmonary stenosis. The baby tested positive for IgM antibodies for both rubellas. The baby was able to reach out for nonilluminating objects well with the left eye and resisted occlusion in that eye. With glasses, the baby was able to fix and follow light in the left eye. On ocular examination, there was microcornea with clear graft in both eyes with central haze in the right eye. She was uncooperative for IOP measurement. The axial lengths recorded on the first visit were 18.02 mm in the right eye and 18.63 mm in the left eye. Both cornea and paediatric opinion was taken and the child was advised to continue steroid tapering in both PKP eyes along with glasses, with an aim to treat amblyopia. After the paediatrician and anaesthetist fitness, the baby underwent EUA in both the eyes and ST AADI with supramid stenting in the LE under GVP. The postoperative course was uneventful with the following findings recorded: the horizontal corneal diameter measured was 8.50 mm in the RE and 9 mm in the LE. The intraocular pressure was 14 mmHg in the RE and 26 mmHg in the LE. The fundus showed pale disc in the RE and hazy view in the LE. The baby was advised e/d dorzolamide with timolol in the LE along with the postoperative medications. The baby will be reviewed on a regular basis.

Conclusion: It is of utmost importance for a newborn with congenital rubella syndrome to undergo a thorough systemic examination. The decision of glaucoma surgery in such babies is very challenging with the refractory nature of the glaucoma. Despite good IOP control, several other factors like amblyopia needs to be addressed. This in turn leads to the disease demanding a multidisciplinary team approach.

15

16 Early outcomes of the PAUL glaucoma implant in intractable primary congenital glaucoma Hamidu Hamisi Gobeka¹

A Olgun², S İmamoğlu³

1 - Afyonkarahisar Health Sciences University,

Afyonkarahisar, Turkey. 2 - West Eye Hospital, Erbil, Iraq. 3 - Haydarpaşa Numune Teaching and Research Hospital,

University of Health Sciences, Istanbul, Turkey.

Correspondence: hgobeka@gmail.com

Background: To assess the efficacy and safety of the PAUL glaucoma implant (PGI), a novel glaucoma drainage device, in a paediatric cohort with intractable primary congenital glaucoma (PCG).

Methods: In this consecutive single-centre retrospective study, the medical records of seven intractable PCG patients having PGI surgery at West Eye Hospital between June 2022 and September 2023 were retrieved and reviewed retrospectively. A least three-month follow-up was necessary for the medical record analysis. A single surgeon performed all surgical procedures in the supertemporal quadrant, including the insertion of a 6-0 prolene ripcord intraluminal suture with the opposite end tucked under the temporal conjunctiva. The success criteria were defined as complete success when an intraocular pressure (IOP) was ≤21 mmHg without anti-glaucoma medications, and partial success when the IOP was ≤21 mmHg with medications. The need for further surgical intervention was regarded as failure.

Results: The median age at diagnosis was 11 years (9-13), with a male-to-female ratio of 3:4. There was a follow-up duration of 10 (9-13) months. Horizontal corneal diameter and axial length were 13 μ m (12.8-13.2) and 28.8 mm (24.5-25.1), respectively. There was a median of 2 (1-3) pre-PGI surgeries. All patients had their 6-0 prolene ripcord intraluminal sutures removed 2-6 weeks postoperatively. The IOP dropped considerably following PGI, from 30 mmHg (27-41) to 13 mmHg (11-14) (p=0.018), accompanied by a significant decrease in the number of anti-glaucoma medications, from 4 (3-4) to 0 (0-2) (p=0.017). There were neither hypotony nor post-PGI failure. Overall, five patients were completely successful, with two achieving partial success.

Conclusion: PGI could be a potent and reasonably safe surgical alternative in intractable PCG patients, with high qualifying success rates and significant decreases in IOP and the need for AGMs.

17 Outcomes of Aurolab aqueous drainage implant in the management of refractory paediatric angle recession glaucoma, case series study

Mohamed Awwad H Helal *Benha University, Benha, Egypt.* Correspondence: mohdzahir82.ma@gmail.com

Aim: To evaluate the effectiveness of Aurolab aqueous drainage implant (AADI) for treating refractory angle recession glaucoma in children.

Methods: This study was conducted at the Benha Glaucoma Unit from June 2018 to May 2022 and involved eight patients with uncontrolled angle recession glaucoma, each with one affected eye. All the patients underwent AADI. The study recorded the preoperative and postoperative intraocular pressure (IOP), the number of antiglaucoma medications, any complications, and the need for further surgeries.

Results: This study included eight children, with an average age of 10.4 ± 4.7 years at the time of surgery, and eight eyes. The mean preoperative intraocular pressure (IOP) was 38.62 ± 4.2 mmHg, and the mean number of medications was 3.62 ± 1.2 . All patients had a significant reduction in IOP, with a mean postoperative IOP of 15.6 ± 4.2 (p < 0.001) at the end of the one-year follow-up period. There were some postoperative complications, such as anterior chamber reaction in one eye treated with topical steroids, hyphaema in one eye, which was resolved spontaneously, and IOP >21 mmHg in three eyes. Two of the three eyes returned to normal after removing the intraluminal prolene filament, and one eye required topical antiglaucoma medication. No sight-threatening complications were recorded, and none of the patients required implant removal.

Conclusion: AADI is a safe, highly economical, and effective method for managing refractory angle recession glaucoma in children.

PAUL glaucoma implant effectiveness in an Irish paediatric cohort: a 24-month retrospective cohort study Melissa Murphy S Farrell Temple Street University Hospital, Children's Hospital,

Correspondence: murphm24@tcd.ie

Dublin, Ireland.

16

Purpose: The PAUL glaucoma implant (PGI) is a novel glaucoma drainage device used in the treatment of refractory glaucoma. This study discusses the effectiveness of the PGI in an Irish paediatric cohort.

Methods: A retrospective cohort study of all patients who underwent PGI surgery in Children's Hospital Ireland (CHI), Temple Street, Ireland over a 24-month period from September 2021 to September 2023. Primary outcome measures included failure (defined as intraocular pressure (IOP) >21 mmHg or <20% reduction), removal of the implant or further glaucoma surgery. Qualified success was defined as use of one or more topical medications. Secondary outcomes included mean IOP change, postoperative intraluminal suture removal, best corrected visual acuity (BCVA) and complications.

Results: A total of 13 cases (9 patients) were performed by a single consultant surgeon over the 24-month study period. Indications for surgery included congenital glaucoma, aphakic glaucoma, Sturge-Weber syndrome and increased IOP secondary to juvenile idiopathic arthritis with uveitis (JIA -U). 1 eye (8%) failed treatment, with a postoperative IOP of 25 mmHg 12 months post-surgery. Overall mean postoperative IOP at last clinic visit was 15.5 mmHg with a mean % change in IOP of 46%. 4 eyes (31%) required one or more topical medications. 7 eyes (54%) underwent postoperative intraluminal suture removal with one reported complication (8%) involving an incorrect suture material being used intraoperatively.

Conclusions: PGI is a useful and effective tool in lowering IOP in paediatric patients especially in those with complex or refractive glaucoma.

19 Long-term outcomes of cyclophotocoagulation in refractory paediatric glaucoma: treatment response and success in 262 eyes

Maram Abdalla Elsayed¹

B Alhazzaa^{1,2}, A Alshahrani^{1,3}, S Aljefri^{1,4}, I Al Obaida¹, AM Al Owaifeer^{1,5}, N Alotaibi¹, K Ahmad¹, R Malik^{1,6} 1 - King Khaled Eye Specialist Hospital, Riyadh, KSA. 2 - King Abdulaziz Medical City, Riyadh, KSA. 3 - Armed Forces Hospitals in Southern Region, Khamis Mushait, KSA. 4 - Imam Muhammad ibn Saud Islamic University, Riyadh, KSA. 5 - King Faisal University, Al-Ahsa, KSA. 6 - Sheikh Khalifa Medical City, Abu Dhabi, UAE.

Correspondence: maram.abdalla@gmail.com

Purpose: To report the surgical success and response to treatment for children undergoing cyclophotocoagulation (CPC) for refractory paediatric glaucoma in a large cohort.

Methods and design: Retrospective cohort study.

Participants: Children (aged <18 years) with a diagnosis of primary or secondary glaucoma, who underwent a first CPC at our institution between May 2010 and May 2019.

Data: Demographics, preoperative status, operative details and postoperative outcomes were collected. The cumulative probability of success was assessed.

Main outcome measures: For Definition 1, treatment success was defined as IOP \leq 21 mmHg at all the visits after the first 3 months without the need for additional glaucoma surgery or repeat CPC. For Definition 2, repeat CPC did not constitute failure. In addition, the response to CPC was classified as: (1) sustained (\geq 1 year) IOP lowering with a single or with; (2) multiple treatments; (3) temporary control of IOP for 6-12 months only; (4) no or transient (<6 months) effect only.

Results: We identified 262 eyes after assessing eligibility. The mean age at the time of first treatment was 5.33 ± 5.03 years, with a mean follow-up of 4.3 ± 4.2 years (31 eyes having 10 years or more follow-up). CPC was performed once in 149 eyes (56.9%), twice in 51 (19.5%) eyes, three times in 40 (15.3%) and ≥4 times in 22 eyes (8.4%). The success rates for Definitions 1 and 2 were 26.7% (95%CI: 21.7 to 32.4%) and 46.2% (95%CI: 40.2 to 52.3%), respectively. Cox regression showed that the older age was associated with a lower risk of failure after both single CPC (HR, 0.92; 95%CI: 0.88-0.96, p<0.001) and multiple CPCs (HR, 0.95; 95%CI: 0.90-1.00, p=0.073). One hundred and seven (41%) of eyes had sustained IOP-lowering with a single treatment; 56 (21%) with multiple treatments whilst 35 (13%) had a transient response and 64 (24%) had no response.

Conclusions: Glaucoma control in children with CPC often requires multiple treatments, with around a quarter of children responding sub optimally. Older children are more likely to exhibit successful IOP lowering.

17

20 Management of failed Ahmed glaucoma valve in childhood glaucoma

Hiranya Abeysekera

Lady Ridgeway Children's Hospital, Colombo, Sri Lanka. Correspondence: hiranyaabeysekera@gmail.com

Introduction: Implantation of a glaucoma drainage device (GDD) is the primary surgery in some forms of childhood glaucoma. This is also performed following failed angle surgery and trabeculectomy. Ahmed glaucoma valve (AGV) is the preferred GDD by the majority of experts. Attrition of the success rate of the AGV in long-term follow-up requires additional medical and surgical therapy to prevent progression of glaucoma.

Methods: All the children who underwent AGV implantation from 1/1/2016 to 31/12/2021 at the ophthalmology department of the Lady Ridgeway Hospital for Children, Colombo by a single surgeon were analysed retrospectively to determine the success rate of the procedure and the surgical procedures undertaken following the diagnosis of failed AGV.

Results: 68 eyes of 55 children were available for the analysis. After two years of implantation, 52% of the AGVs were diagnosed as failed. After five years of implantation, only 12% of the tubes were functioning. A second AGV implantation was done in 15% of failed AGVs. Cyclophotocoagulation was performed for 85% of the failed AGVs. Excision of the fibrous capsule was done in 44% of was AGV flushing performed patients. in 8%. Trabeculectomy was performed in another 12% of failed AGVs. More than one procedure was performed in 63% of the failed AGVs.

Conclusion: Failure rate of AGV in paediatric patients is very high. Most patients require additional surgeries to prevent the progression of glaucoma.

21 Outcomes of revision surgery for aqueous shunt exposure in paediatric onset glaucoma Alessandra Martins

Moorfields Eye Hospital, London, UK. Correspondence: alessandra.martins1@nhs.net

Objectives: To present outcomes of revision surgery undertaken over a five-year period in patients with paediatric onset glaucoma presenting with exposure of aqueous shunt. Surgical techniques to prevent re-exposure will be discussed.

Methods: Retrospective review of case notes for all consecutive aqueous shunt revision surgery for tube exposure undertaken by a single surgeon at Moorfields Eye Hospital 2015-2021. All patients underwent the same revision surgical technique which included a broad dissection and mobilisation of conjunctiva to avoid tension on limbal closure. The shunt was patched with a double layer of donor pericardium (Tutoplast) secured using fibrinogen glue (Tisseal) with conjunctival closure using 8.0 vicryl.

Results: Six patients with paediatric onset glaucoma underwent revision surgery for exposure during the ascribed time period. Three patients had a diagnosis of primary congenital glaucoma, with one each with a diagnosis of juvenile onset open angle glaucoma (JOAG), aniridia and anterior segment dysgenesis. Mean time from implantation to exposure was 53 months (range 1 week to 10 years). Over a mean follow-up of 70 months (range 24 - 96 months) there were no further episodes of aqueous shunt exposure.

Conclusion: The surgical technique described in study is a successful option for preventing further tube exposure and is a viable option for extending the lifespan of a functioning aqueous shunt.

22 Unrecognised intermittent glaucoma drainage tube blockage secondary to pharmacological pupil dilation Ahmed Ali

AJ Connor

18

Royal Victoria Infirmary, Newcastle Upon Tyne, UK. Correspondence: a.ali58@nhs.net

Case: We present an unusual glaucoma tube related complication in a 2-year-old female with bilateral congenital glaucoma secondary to *FOXC1* gene mutation.

After undergoing bilateral trabeculotomy at 4 weeks of age, persistently high intraocular pressure (IOP) and corneal clouding necessitated bilateral Baerveldt glaucoma drainage tube placement at 8 weeks of age. Following this procedure IOP control was achieved, and she remained under regular follow-up. At 25 months of age she was admitted to hospital with vomiting and drowsiness. CT scanning and lumbar puncture were normal and her illness resolved spontaneously after 2 days. Her parents reported a similar episode 6 months prior which had also resolved spontaneously.

She was admitted again at 32 months of age with a recurrence of vomiting and drowsiness. All 3 episodes had occurred 1-2 days following routine outpatient ophthalmology review. When an ophthalmology opinion was sought during the 3rd episode, IOP in the left eye was 66 mmHg with the internal opening of the glaucoma tube occluded by iris. The tube entry into the AC was close to an area of peripheral anterior synechiae which had tented up the pupil margin. Pupil dilation had further facilitated movement of the iris closer to the tube opening causing occlusion. Following treatment with miotics and systemic acetazolamide there was resolution tube blockage and normalisation of IOP.

Conclusion: This case highlights the importance of precise tube placement in the AC considering both depth and areas of adjacent iris abnormalities, with consideration of how this dynamic system may change with pharmacological dilation.

23 Glaucoma drainage Device (GDD) revision in paediatric glaucoma

Samuel Simpson

A Connor

Royal Victoria Infirmary, Newcastle Upon Tyne, UK. Correspondence: samuel.simpson4@nhs.net

Introduction: Glaucoma drainage devices (GDDs) play an important role in the management of paediatric glaucoma. The main cause of GDD failure is encapsulation and fibrosis around the GDD plate which restricts aqueous flow, leading to increased intraocular pressure (IOP). Surgical revision of the bleb involves decapsulation of the GDD plate and removal of the fibrotic tissue, with or without use of antimetabolites. This study aims to assess outcomes from GDD revision.

Methods: A retrospective search was performed to identify all cases of primary GDD revision surgery performed at a single tertiary centre within a 3-year period. Once cases were identified case note review was performed to gather relevant data for the 12-month period following revision.

Results: Seven eyes of six patients were identified with mean age of 11.7 years. Mean preoperative IOP was 33.1 mmHg on a mean of 4.1 medications. Eyes had undergone a mean of 2.3 previous procedures. At 3, 6 and 12 months postoperatively there was a mean percentage IOP decrease of 30%, 16% and 23%, respectively. Mean numbers of medications were 4.3, 2.8 and 3.3, respectively. There were no cases of significant visual loss. Two of the seven cases underwent further surgery within the follow-up period.

Conclusion: This case series would suggest that bleb revision surgery can achieve modest IOP reductions in the short term. However, this comes with a significant failure rate which would be expected to rise with longer term follow-up.

4 AGV valve slicing - a blessing in disguise

Sirisha Senthil

19

LV Prasad Eye Institute, Hyderabad, India. Correspondence: sirishasenthil@lvpei.org

Background: Primary congenital glaucoma is a rare and challenging condition in paediatric patients. We present a case of a 4-month-old infant with bilateral primary congenital glaucoma who underwent multiple surgical interventions, including trabeculectomy, trabeculotomy, transscleral cyclophotocoagulation (TSCPC), and Ahmed glaucoma valve (AGV) implantation, to manage intraocular pressure (IOP). Despite these efforts, IOP control was inadequate, necessitating a unique approach.

Case description: The patient exhibited typical signs of primary congenital glaucoma, including megalocornea, Haab's striae, and corneal haze. Initial management involved combined trabeculectomy with trabeculotomy for both eyes, which successfully maintained normal IOP for four years with medication. However, IOP control became a challenge in the left eye (LE), leading to a limited TSCPC. Subsequently, an AGV implant was placed in the LE, resulting in three years of IOP control.

Innovative intervention: Fibrous proliferation around the AGV bleb and its restrictive characteristics prompted the decision to perform AGV valve slicing along with bleb revision instead of considering a second implant or repeat TSCPC. This novel approach led to an optimally functioning AGV valve, effectively managing IOP and preventing further progression of the disease.

Conclusion: In refractory cases of primary congenital glaucoma, particularly in paediatric patients, the innovative technique of AGV valve slicing with bleb excision can serve as a successful alternative to additional implants or TSCPC. This case highlights the importance of adapting treatment strategies to the individual patient's needs and provides a promising solution for managing challenging cases of childhood glaucoma. Further research and follow-up studies may be warranted to assess the long-term efficacy of this approach in similar cases.

25a Development of a skills transfer course for surgical techniques in paediatric glaucoma Deniz Goodman

TCP Chang, AL Grajewski, E Bitrian Bascom Palmer Eye Institute, Miami, USA. Correspondence: dgm@bu.edu

Purpose: To effectively teach the surgical techniques required for management of paediatric glaucoma through a curriculum consisting of a didactic session followed by a targeted hands-on wet lab at the American Academy of Ophthalmology (AAO) conference.

Methods: The "Mastering Childhood Glaucoma Surgical Techniques" skills transfer course is the first and only paediatric glaucoma wet lab at the AAO conference. Hosted annually since 2021, this course has been perfected over the years to its current iteration, boasting 7 stations outfitted with QR codes of tailored step-by-step instructional videos, high-fidelity synthetic eye models, laminated exam under anaesthesia (EUA) checklists, cadaver eyes, and real surgical instruments. Using these amenities, attendees spend 75 minutes learning the basics of paediatric glaucoma surgical techniques, including goniotomy, trabeculotomy, shunts, cyclodestruction and EUA.

Results: Approximately 18 attendees participate in the course per year while 7 experts provide personalised instruction. This course began as a didactic session that has run continuously for over a decade. Initially, attendees practiced surgeries using a "dry-wet-lab" kit containing microscopes and synthetic eyes for practice in a lecture hall. Given the advantages of using human eyes, the course was expanded in 2021 to include a dedicated wet-lab session. Persistent challenges include obtaining sufficient instrumentation/materials and coordinating with industry to provide access to emerging surgical technology.

Conclusions: Given that few ophthalmologists specialise in paediatric glaucoma, our skills transfer course takes an innovative approach at providing a unique, risk-free, and supportive environment for learning the challenging techniques behind the most widely used paediatric glaucoma procedures.

20

25b The Ophthalmology Surgical Competency Assessment Rubric (OSCAR) for ab externo trabeculotomy in paediatric glaucoma Deniz Goodman

AL Grajewski, EJ Savatovsky, TCP Chang, E Bitrian Bascom Palmer Eye Institute, Miami, USA. Correspondence: dgm@bu.edu

Purpose: Develop and validate a surgical competency assessment tool for ab externo trabeculotomy in children.

Methods: A panel of four paediatric glaucoma experts from the Bascom Palmer Eye Institute (BPEI) developed a rubric for each step of ab externo trabeculotomy. Narrative descriptions of four competency levels (novice, beginner, advanced beginner, competent) were generated per surgical step based on a modified Dreyfus scale of skill acquisition. A second international panel of paediatric glaucoma experts was enlisted to review the tool and offer constructive feedback. The primary outcome was the final rubric after incorporating feedback that reached full consensus among the first panel of experts from BPEI.

Results: The Ophthalmology Surgical Competency Assessment Rubric for ab externo trabeculotomy (OSCAR-AET) consists of 12 steps and is the proposed rubric for evaluating surgical skill for ab externo-trabeculotomy procedures. Feedback from an international paediatric glaucoma expert panel was incorporated to establish face and content validity of this tool.

Conclusions: The OSCAR-AET may be used for evaluating surgical skill for ab externo-trabeculotomy procedures. Further investigations should determine the intra- and interobserver reliability of this tool.

26 Exam under anaesthesia for paediatric glaucoma: a simulation-based training

Vandana Persad

V Persad, TCP Chang, AL Grajewski, E Savatovsky, E Bitrian Bascom Palmer Eye Institute, Miami, USA. Correspondence: vxp533@miami.edu

Purpose: To introduce a simulation standardised model of exam under anaesthesia (EUA) in paediatric glaucoma for ophthalmology trainees. The aim of this model is to increase expertise, efficiency, standardisation of knowledge and optimise exposure to paediatric EUA.

Methods: Modified realistic size mannequins with 3-D printed eyes are used to simulate infants and the environment of an EUA in the operating room. The mannequins have 3-D printed eyes that allow for realistic examination of red reflex and fundus examination. A rubric for EUA is used and an EUA data collection sheet for systematic information collection is provided.

Results: Training begins with the infant masked to induce sedation and the trainee checks intraocular pressure with the mask on the infant's face. Attention is paid to positioning of the mask and ability to manoeuvre and check IOP while working in a tight space. Afterwards vertical and horizontal corneal diameters measurements, pachymetry, anterior segment slit lamp examination are performed, followed by gonioscopy and dilated eye exam. All steps are evaluated by a EUA rubric. The accuracy and efficiency of the trainee to collect pertinent information in the provided EUA sheet is evaluated. Ancillary testing such as the use of A and B scan, ultrasound biomicroscopy and intraoperative photography is trained and graded.

Discussion: Examination under anaesthesia of a paediatric patient with glaucoma is an important component of ophthalmology and glaucoma training. Exposure to both basic knowledge and complex examination techniques can be taught in a systematic and efficient manner.

Conclusion: A simulated model provides an accelerated development of expertise in a risk-free environment.

21

27a Utilisation of a synthetic eye model for teaching ab externo trabeculotomy with an ophthalmic simulated surgical competency assessment rubric (Sim-OSSCAR): a side-by-side comparison Deniz Goodman

AL Grajewski, EJ Savatovsky, TCP Chang, E Bitrian Bascom Palmer Eye Institute, Miami, USA. Correspondence: dgm@bu.edu

Purpose: Introduce a synthetic eye model (SE) as a tool for teaching ab externo trabeculotomy (AET) using a custom-made Sim-OSSCAR.

Methods: We established a structured session consisting of a didactic component with surgical videos and photos as well as a hands-on component between a paediatric glaucoma and instructor students, residents, or ophthalmologists with minimal to no experience with AET. The SE is equipped with sclera, Schlemm's canal, and trabecular meshwork that allows participants to perform AET. Using a previously created AET ophthalmic surgical competency assessment rubric (OSCAR) for in vivo human eyes, we developed a Sim-OSSCAR to specifically assess AET performance with the synthetic eye. A panel of expert paediatric glaucoma surgeons reviewed the tool and provided constructive feedback, which was incorporated after further discussion to establish face and content validity. Results: We present a side-by-side comparison video of AET, with each surgical step being performed on both an in vivo human eye and the synthetic eye. Expert commentary will accompany the video, and corresponding narrative descriptions for each step of the OSCAR and Sim-OSSCAR will be overlaid on respective sides to compare and contrast differences in anatomical fidelity and competency grading between the two types of eyes and rubrics.

Conclusions: The synthetic eye model is a reasonable approximation of in vivo surgery, and effectively allows for the simulation of AET in a step-by-step fashion among those without surgical experience. The Sim-OSSCAR is a validated analogue to the OSCAR that can be used to assess surgical skill with synthetic eyes.

27b Value-cost analysis of synthetic eye models for teaching paediatric glaucoma surgery Deniz Goodman

AL Grajewski, EJ Savatovsky, TCP Chang, E Bitrian Bascom Palmer Eye Institute, Miami, USA. Correspondence: dgm@bu.edu

Purpose: Compare the cost, required preparation, and anatomical fidelity between a synthetic eye model (SE), pig eyes (PE), and cadaver eyes (CE) for teaching paediatric glaucoma surgery.

Methods: Information from eye banks and scientific supply stores was used to determine the cost of eyes and materials. Surgical course instructors established representative wetlab scenarios to calculate cost estimates.

Results: The average cost of a SE, PE, and CE is \$70, \$2, and \$250, respectively. Relative to using CE, calculated savings are as follows: hosting a one-time wet lab for a typical residency class of 4 (PE: \$992, SE: \$720), a longitudinal (12 sessions/year) wet lab curriculum of the same class size (PE: \$11,904, SE: \$8,640), and a conference skills transfer session of 20 attendees (PE: \$4,960, SE: \$3,600). SE require significantly less intensive preparation, only necessitating lubrication and a 15-minute water soak, compared to PE and CE that must be stored at low temperatures to prevent degradation and soaked in formalin for several hours or injected with saline or viscoelastic agents to achieve physiological globe pressures. SE features a clear cornea and a human-like Schlemm's canal, thereby providing a more realistic model for teaching goniotomy and ab externo trabeculotomy compared to PE which may have a cloudy cornea, segmented canal, and tougher trabecular meshwork.

Conclusions: The synthetic eye model represents a costeffective and convenient alternative to cadaver eyes and pig eyes for teaching paediatric glaucoma surgery.

22

28 Training the next generation of paediatric glaucoma surgeons: an international effort Elena Bitrian TCP Chang, AL Grajewski

Bascom Palmer Eye Institute, Miami, USA. Correspondence: elenabitrian@miami.edu

Purpose: To present an international educational programme with a structured curriculum for teaching paediatric glaucoma surgery, that maximises physician learning for ophthalmologists, expanding their skills set into paediatric glaucoma surgery while enhancing patient safety. **Methods:** 17 glaucoma specialists from 4 different continents taking care of paediatric glaucoma patients were recruited to join a training programme for 3 weeks to 3 months (depending on the level of expertise). A structured curriculum consisting on clinical and surgical direct observation and didactic modules with slide sets, photos, surgery videos, followed by a wet-lab curriculum using both a 3D printed artificial eye and cadaver eyes were used to practice clinical and surgical skills. A rubric was used to assess how the trainee's performance improved.

Results: Trainees expanded their skillsets, learned new surgical techniques, improved their proficiency, surgical time and confidence performing different angle and filtering surgeries. Upon return to their home countries they practiced and teach the techniques learned.

Conclusions: An international training programme for paediatric glaucoma can highly improve the preparation and proficiency of the next generation of paediatric glaucoma surgeons.

29 Factors and outcomes associated with corneal oedema and Haab's striae in primary congenital glaucoma

Brenda Bohnsack

Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, USA.

Correspondence: bbohnsack@luriechildrens.org

Purpose: Investigate specific factors and outcomes associated with corneal oedema and Haabs striae in primary congenital glaucoma (PCG).

Methods: Retrospective review of patients with PCG (2011-2023) with pre and postoperative documentation of corneal findings. Preoperative details and final outcomes were compared between eyes with and without corneal findings. The right eye of bilateral cases and the affected eye in unilateral cases were included for analysis.

Results: Forty-two patients (77 eyes, 62% male) underwent initial angle surgery at 236±275 (median 132) days old. Corneal oedema and Haabs striae were present preoperatively in 56 (73%) eyes of 33 patients and 51 (66%) eyes of 30 patients. Analysis of one eye per patient showed that corneal oedema was associated with younger age (p<0.0001), higher IOP (p=0.0456), and shorter axial length (p=0.0102) at presentation and worse visual acuity at final follow-up (p=0.0024). Haabs striae were associated with older age at presentation (p=0.0155). After angle surgery, corneal oedema was present at 1-month in 70% with 95% CI [51,83], at 2-months in 27% with 95% CI[13,43], at 3-months in 15% with 95% CI[5,29] and at 1-year in 3% with 95% CI [0,13]. Corneal oedema did not resolve in 3 eyes of 2 patients after more than 4 years of follow-up.

Conclusions: Corneal oedema resolved in the majority of PCG cases within 2 months of initial angle surgery, but was associated with younger age, higher IOP, and shorter axial length at presentation and worse visual acuity at final follow-up.

30 Genetic changes associated with childhood glaucoma: a systematic review

Anika Kumar Y Han

23

University of California, San Francisco, USA. Correspondence: anika.kumar@ucsf.edu

Introduction: Many forms of childhood glaucoma have been associated with underlying genetic changes, and several genetic variants have been described. This systematic review aimed to summarise the literature describing genetic changes in childhood glaucoma.

Methods: This systematic review was conducted in accordance with the Preferred Reporting Items for Systematic review and Meta-Analyses (PRISMA) 2020 guidelines and registered with Prospero. A comprehensive review of Pubmed, Embase and Cochrane databases was performed using pre-defined search terms. Information was extracted regarding genetic variants including genotype-phenotype correlation. Risk of bias was assessed using the Newcastle-Ottawa Scale.

Results: Of 1,916 records screened, 196 studies met inclusion criteria and 53 unique genes were discussed. Among study populations, mean age±SD at glaucoma diagnosis was 8.94 ± 9.54 years and 50.4% were male. The most common gene discussed was *CYP1B1*, evaluated in 109 (55.6%) studies. *CYP1B1* variants were associated with region and population-specific prevalence ranging from 5% to 86% among those with primary congenital glaucoma. *MYOC* variants were discussed in 31 (15.8%) studies with prevalence up to 36% among patients with juvenile open angle glaucoma. *FOXC1* variants were discussed in 25 (12.8%) studies, which demonstrated phenotypic severity dependent on degree of gene expression and type of mutation. Overall risk of bias among studies was low; the most common domain of bias was selection.

Conclusion: Numerous genetic changes have been described in childhood glaucoma. Understanding the most common genes as well as potential genotype-phenotype correlations has the potential to improve diagnostic and prognostic outcomes for children with glaucoma.

31 Medium-term survival of developmental glaucoma treated with ab interno angles surgery: analysis of a large clinical database

Ta Chen Peter Chang

R O'Brien Bascom Palmer Eye Institute, Miami, USA. Correspondence: t.chang@med.miami.edu

Purpose: To evaluate the outcomes of ab interno angle surgery (AIAS, goniotomy and/or transluminal dilation of Schlemm's canal) in primary congenital glaucoma (PCG) and juvenile open angle glaucoma (JOAG) using the IRIS (Intelligent Research in Sight) Registry.

Methods: Survival analysis of IRIS Registry entries with AIAS as first surgery, cross-reference with International Classification of Diseases (10th Edition) codes for PCG/POAG. Failure definition based on insufficient intraocular pressure lowering, hypotony, loss of light perception or loss of eye.

Results: A total of 145 eyes of 112 patients with PCG (29.5% bilateral, 51.8% female) and 538 eyes of 383 patients with JOAG (40.5% bilateral, 49.6% female) had AIAS as first surgery, aged 7.9±6.2 years (median age 8.5, IQR 1-14), and 12.5±4.0 years (median age 14, IQR 10-16), respectively. At 3 years, there were 50 failures among PCG eyes and 211 failures among JOAG eyes, which yielded a survival of 42.6% for PCG eyes vs. 42.0% for JOAG eyes (sensitivity analysis: 42.4% for PCG and 43.9% for JOAG at 3 years). 54 eyes of 39 PCG patients had AIAS prior to the age of 3 years (38.5% bilateral, 51.3% female), with mean age of 0.6±0.7 SD years (median age 1, IQR 0-1). At 3 years, there were 14 failures among these eyes, which yielded a surgical success of 50%.

Conclusion: Three years after surgery, more than 50% of AIAS performed on PCG/JOAG failed. Modified failure criteria are needed to evaluate the outcomes of glaucoma surgery in children.

32 Early experience with micro-pulse transscleral diode cyclophotocoagulation for refractory paediatric glaucomas

Ray Areaux

24

University of Minnesota, Minneapolis, USA. Correspondence: areaux@umn.edu

Introduction: Compared to continuous-wave transscleral cyclophotocoagulation (CW-CPC) and endoscopic cyclophotocoagulation (ECP), data on micro-pulse transscleral cyclophotocoagulation (mpCPC) in children is limited.

Methods: Retrospective consecutive case series of eyes with refractory paediatric glaucomas at a tertiary centre that underwent mpCPC. Success was postoperative IOP less or equal to 24 mmHg with or without medications, without additional glaucoma surgery, and with at least 6 months follow-up.

Results: 10 eyes of 7 patients were included. 5 patients (8 eyes) were less than 2 years-old, 1 was 12 years-old, and 1 was 16 years-old. Types of glaucoma: primary congenital glaucoma (5), glaucoma following cataract surgery (2), glaucoma associated with non-acquired ocular anomalies (1), glaucoma in a non-acquired syndrome (2). Mean degrees of ciliary body treated was 300 (range 180 - 360). 8 eyes were treated once, 1 eye twice, and 1 eye thrice. Mean follow-up was 15.2 months (range 6 - 21). Mean preoperative IOP was 26 (range 21-36) mmHg. Mean postoperative IOP was 21 (19-23) for successes and 34 (25-42) for failures. The mean number of glaucoma medications (2.6) was unchanged from preoperatively to postoperatively. In eyes 26 months-old and younger, axial length progressed in all failures (5) and plateaued in all successes (3). Overall success was 40%. All failures required incisional glaucoma surgery. There were no significant complications.

Conclusions: mpCPC was performed safely but with limited success in this small cohort of heterogenous childhood glaucomas. mpCPC may be offered cautiously for refractory childhood glaucomas, emphasising that repeat treatments or surgical escalation may be necessary.

33 Exploring the potential of augmented reality and virtual reality as a diagnostic tool for glaucoma screening in ophthalmology

Krishnika Vetrivel

Kings College University Hospital, London, UK. Correspondence: krishnika.vetrivel@kcl.ac.uk

Introduction: Glaucoma is one of the leading causes of irreversible blindness globally. Early detection is crucial; yet, half of glaucoma cases remain undiagnosed. Whilst previous reviews have primarily focused on the application of VR/AR (virtual reality/augmented reality) in ophthalmology for education, surgical training and treatment, the potential of this technology as a diagnostic or screening tool is limited in current literature.

Aims: This review aims to provide an overview of the current state of research by exploring four key areas: limitations of existing screening tools, benefits and challenges of VR, the integration of VR with telemedicine and artificial intelligence (AI) for glaucoma diagnosis, and future directions.

Methods: A comprehensive search for original publications was conducted using the Medline and Embase databases. The initial search yielded 2878 studies, from which eight relevant studies were identified. The included studies had a varying risk of bias and applicability, according to the QUADAS-2 evaluation.

Results: VR offers advantages such as enhancing accessibility, improving disease management and reducing wait times. Integrating telemedicine and VR with AI offers the potential for remote collection of patient data, early disease detection, clinical forecasting, disease progression tracking, and optimised treatment planning. Nevertheless, potential limitations include possible myopia progression and marketing-driven heterogeneity.

Conclusion: Future studies should investigate the accuracy and applicability of AR/VR in diagnosing glaucoma across diverse patient groups, including paediatric cases, encompassing different types and severity levels. In conclusion, VR holds potential for glaucoma diagnosis but necessitates collaboration and further research before integration into clinical practice.

25

34 Value-cost comparison of standard automated perimetry and head-mounted perimetry Deniz Goodman

DE McLaughlin, HK Munshi, E Bitrian, EA Hodapp, AL Grajewski Bascom Palmer Eye Institute, Miami, USA. Correspondence: dgm@bu.edu

Purpose: Alternatives for resource-intensive standard automated perimetry (SAP) in visual field (VF) testing includes a head-mounted virtual reality visual field (VRVF) device. This study compares these devices by time, space, and equipment cost.

Methods: VF times, or time elapsed from the start of wait time for VF to the start of wait time for the next visit event, were collected retrospectively for SAP (February and May 2021) and the VRVF device (May 2021). Size specifications were quoted from the manufacturers. Device costs were provided by the hospital's purchasing department as well as by the device developer and webpage.

Results: Testing with a VRVF device shortened the mean VF time by 26% and 28% and the standard deviation by 50% and 52% compared to SAP times in February and May 2021, respectively (VRVF: 44.56±13.53 minutes vs. SAP [Feb]: 60.22 ± 27.27 minutes and SAP [May]: 61.94 ± 27.91 , p<0.05). The size specification of the SAP model was 46 L x 52 W x 58 H (cm) and the size specification of the VRVF model was 18 L x 13 W x 10 H (cm). SAP cost \$102,400 and VRVF cost \$36,000 over an estimated 20 years.

Conclusions: Incorporating a head-mounted virtual reality device for VF testing was time-, space-, and cost-effective. While VRVF testing is unlikely to replace SAP, the use of both devices within a clinic has the potential to improve efficiency and patient satisfaction for glaucoma evaluation.

35 Analysis of peripapillary vascular density with optical coherence angiography (OCT-A) in paediatric glaucoma. Comparative study with healthy subjects

Flora Xydaki

CD Méndez Hernández Hospital Clínico San Carlos, Madrid, Spain. Correspondence: fsxydaki@gmail.com

Summary: Glaucoma is a group of ocular pathologies that affect the optic nerve and leads to irreversible blindness. There are several subtypes of glaucoma; this study focuses on patients with infantile, primary congenital, juvenile and secondary glaucoma. Primary glaucoma affects children at birth or in the first years of life.

Childhood glaucoma is caused by an identifiable cause of increased intraocular pressure (IOP), such as secondary glaucoma, anterior chamber malformations associated or not with non-acquired syndromes, hereditary or not, or diseases acquired in the first years of life, secondary glaucoma because of cataract surgery, trauma or inflammatory processes, among others.

It has been shown that the density of retinal microcirculation is reduced in glaucoma, so the diagnosis of these patients the examination of vascularisation is important. Optical coherence tomography angiography (OCT-A) is a noninvasive technique, which does not require intravenous contrasts, and it is used to obtain images of the retinal microcirculation. 36 Shedding light on ab interno canaloplasty: a novel surgical technique to manage paediatric glaucoma Arjun Sharma

E Bitrian Bascom Palmer Eye Institute, Miami, USA. Correspondence: axs9291@miami.edu

Introduction: We present a novel surgical technique using a new minimally invasive gonioscopy-assisted transluminal trabeculotomy (GATT) device to manage glaucoma in a paediatric patient.

Methods: The operating procedure was recorded to illustrate the novel use of an all-in-one canaloplasty device, iTrack Advance. Surgical steps and pearls are described.

Results: A 7-year-old child with juvenile open angle glaucoma and an intraocular pressure of 32 mmHg on max medical therapy underwent GATT using the iTrack Advance device. A single 1 mm clear corneal incision was created, Miostat and viscoelastic to deepen and maintain the anterior chamber, followed by placement of a gonioprism for visualisation. Next, the illuminated microcatheter was inserted into Schlemm's canal with subsequent viscodilation. A 180-degree trabeculotomy was performed, followed by retraction of the microcatheter and removal from the eye. The catheter was repositioned and reinserted to address the remaining portion of the angle to achieve a 360-degree circumferential trabeculotomy. A single 10-0 vicryl suture was used to close the surgical wound. Using this technique, one fewer incision and suture decreased time under anaesthesia, and a decreased surgical time was observed.

Conclusions/Relevance: Angle surgery is the standard of care for glaucoma in the paediatric population. The use of an all-in-one canaloplasty device, iTrack Advance, offers a novel approach for surgeons to effectively treat paediatric patients with glaucoma by creating only one small surgical incision, allowing for fewer sutures, decreased complication risk, and decreased operating and recovery time as compared with other ab interno procedures.



37 All aniridias are not same: the mysterious world of absent iris!!

Ashok Kumar Singh TT Faisal, SS Pandav, S Kaushik Post Graduate Institute of Medical Education & Research, Chandigarh, India. Correspondence: ashoksingh1603@gmail.com

Aim: To discuss various conditions with absent/hypoplastic iris and the role of genetics in solving diagnostic dilemmas:

- *Case 1:* A 3-week-old boy presented with bilateral cloudy corneas, limbus stretching, and high IOP. There was presence of high iris insertion, multiple iris processes, peripheral iris tags and visible lens edge. The child also had telecanthus, a flat nasal bridge, an umbilical hernia, and micrognathia. Diagnosis of atypical aniridia with a possibility of Axenfeld-Reiger syndrome was made. The diagnosis was confirmed by clinical exome sequencing (CES), which showed a *FOXC1* mutation.
- *Case 2:* A 5-year-old boy presented with low vision and nystagmus. He had rudimentary iris stumps, stretched zonules, superiorly subluxated lens, pannus, limbal stem cell deficiency and absent foveal reflex. His sister and mother had identical features. CES showed deletion of the *PAX6* gene without *WT1* gene mutation. So, a final diagnosis of classical aniridia without systemic involvement was made.
- *Case 3:* A 2-month-old baby presented with bilateral cloudy cornea, intense photophobia, watering, high IOP and grade 4 cornea clarity. The child underwent CTT, and after corneal clearing, severe iris hypoplasia along with the presence of ectropion uveae and visible lens edge was seen. A clinical diagnosis of neonatal onset congenital ectropion uveae (NO-CEU) was made, which was confirmed by CES, which showed *CYP1B1* c1169G>A(p.Arg390His) variant.

Conclusion: Aniridia is not merely an absence of the iris but amalgamation of various ocular and systemic conditions. Genetics play a significant role in situations of diagnostic dilemma and helps in proper management.

38 Childhood glaucoma associated with Rubinstein-Taybi syndrome: long-term outcomes of a case series

Cristina Ginés-Gallego

C Fenerty, K Yau Manchester Royal Eye Hospital, Manchester, UK. Correspondence: c.ginesgallego@gmail.com

Introduction: Rubinstein-Taybi syndrome (RTS) is a rare autosomal dominant disorder characterised by intellectual disability, distinctive facial and skeletal malformations, broad terminal phalanges of hands and feet, bilateral cryptorchidism, as well as multiple ocular abnormalities, including congenital glaucoma. Since it was first reported in 1963, the systemic features and the genetics of this syndrome have been progressively better understood, leading to a prompt and more accurate diagnosis nowadays. However, the exact prevalence of ocular manifestations, in particular, glaucoma, is still uncertain, as most of the initial publications failed to report on intraocular pressure, or described clinical findings which could be mistaken for glaucoma.

Aim: To present long-term follow-up outcomes of three cases with childhood glaucoma secondary to RTS, and to illustrate the ocular findings of this infrequent condition.

Results: An eight-year-old male, as well as two twenty-three -year-old female twins, with more than twenty-year followup, were included. All of them had multiple bilateral surgeries: trabeculotomies, trabeculectomies, Baerveldt tube implants with repeat needling procedures, as well as cyclodiode. Significant differences were noted in the severity of the glaucoma and the need for surgery over time, even between the two sisters. Close follow-up and prompt surgical management when indicated were essential to maintain stable visual outcomes.

Conclusions: Glaucoma associated to RTS can lead to significant visual impairment at a very early age if untreated or misdiagnosed. A prompt, thorough ophthalmic assessment, including intraocular pressure measurement, is warranted to prevent the appalling consequences of irreversible vision loss in these young patients.

39 Congenital glaucoma and Aicardi-Goutières syndrome, a case report Alfonso Miranda-Sánchez

R Fernández-Narros, J García-Feijoo Hospital Clínico San Carlos. Madrid, Spain. Correspondence: alfonsomirandasanchez@yahoo.es

Introduction: Aicardi-Goutières syndrome (AGS) is a rare autosomal recessive inherited encephalopathy with a unknow prevalence. Various associated genes have been identified, including *TREX1*, *RNASEH2B*, *RNASEH2C*, *RNASEH2A*, *SAMHD1*, *ADAR*, and *IFIH1*. To date, only one case has been reported in which AGS, confirmed by genetic testing, was associated with congenital glaucoma.

Case: We present a case of AGS in a six-year-old child. A genetic study was conducted, which confirmed a mutation in the *RNASEH2B* gene characteristic of AGS. At the age of 6 years, the patient was referred to our hospital, a national reference centre for congenital glaucoma. A comprehensive evaluation under general anaesthesia was performed, revealing megalocornea in both eyes and a cup-to-disc ratio of 0.8 in OD and 1 in OS. IOPs was 25 and 30 mmHg in OD and OS, respectively. Goniotomy was performed in both eyes. Three weeks later, a follow-up evaluation was conducted. IOPs was 17 and 18 mmHg in the OD and OS, respectively, with no apparent changes in the optic nerves.

Review: In the comprehensive search of the updated literature, five cases were identified in which an association between AGS and congenital glaucoma had been established. However, in three of these cases, no confirmatory genetic diagnosis had been obtained. Additionally, in another case, the presence of congenital glaucoma could be explained by the coexistence of aniridia or the *CYP1B1* mutation in the patient. All cases were treated using trabeculectomy; however, we opted for goniotomy as a less invasive initial option with good results to date.

40 Rare case of Adams Oliver syndrome with congenital glaucoma Manju Anilkumar M Udaykumar Naidu *Aravind Eye Hospital, Madurai, India.* Correspondence: manju@aravind.org

Case: A 2-year-old male child was brought to clinic by the mother with complaints of white reflex in both the eyes since birth. The baby was born out of a non-consanguineous marriage, full-term normal vaginal delivery with a birth weight of 2.4 kg. The baby had a NICU stay for 3 days for oligohydramnios and congenital scalp defect (aplasia cutis). A short stature of 55.5 cm at 3 months was noted. On systemic screening, a small patent ductus arteriosus on ECHO and atelectasis of left lower lobe of the lung was noted on CT chest. USG cranium and abdomen was normal. At the first EUA, the child was found to have megalocornea with anterior polar cataract with hazy media with IOP of 33 mmHg in the right eye and 30 mmHg in the left eye, axial length of 23.56 mm in the right eye and 23.15 mm in the left eye and UBM was normal. He was started on e/d brinzolamide and latanoprost in both the eyes. Examination under anaesthesia with left eye trabeculectomy with MMC (mitomycin C) was done. As the IOP was refractory, subsequent AADI (Aurolab aqueous drainage implant) with supramid stenting was planned. When the IOP continued to remain refractory and with increasing axial length, a repeat superonasal AADI with supramid stenting was planned (left eye followed by the right eye). After the subsequent supramid suture removal, the IOP lowered to 7 mmHg in the right eye and 13 mmHg in the left eye.

Conclusion: Adams Oliver syndrome is a rare congenital syndrome. The ophthalmic manifestations constitute around only 10%. Systemic screening is indispensable. Early diagnosis, treatment and follow-up can prevent progression of glaucoma in such children.



41 Persistent tunica vasculosa lentis in glaucoma associated with neurofibromatosis - a case report Nader Bayoumi

N El Shakankiry

Alexandria University Faculty of Medicine, Alexandria, Egypt. Correspondence: naderbayoumi@yahoo.com

Summary: An 8-month-old female presented to the ophthalmology department of Alexandria Main University Hospital with an enlarged cloudy cornea of the left eye and a swollen left side of the face. The child was referred from the paediatrician with the diagnosis of neurofibromatosis type 1 (NF1). Examination revealed the right eye to be clinically free and the left eye to have an intraocular pressure (IOP) of 21 mmHg (under inhalational general anaesthesia), a corneal diameter of 16 mm, ectropion uvea, a cup/disc (C/D) ratio of 0.9, an S-Shaped upper lid deformity and an axial length (AL) of 28.06 mm. A diagnosis of secondary glaucoma was made and an uneventful combined trabeculotomy-trabeculectomy with mitomycin C was performed. The child presented on postoperative day 1 with a total fresh hyphaema that persisted for 2 weeks upon which an anterior chamber (AC) wash was performed. Intraoperatively, the source of bleeding was revealed to be a persistent tunica vasculosa lentis along the zonules of the lens. Bleeding did not recur following the AC wash for the following 6 months and the IOP was under control until the last follow-up despite a flat scarred filtering bleb. To the best of the authors' knowledge, the association of persistent tunica vasculosa lentis with glaucoma associated with NF1 has not previously been reported.

42 Serous detachment of the macula in an eye with glaucoma associated with port-wine birthmark after Initiating prostaglandin analogue therapy Ta Chen Peter Chang

A Mueller Bascom Palmer Eye Institute, Miami, USA. Correspondence: t.chang@med.miami.edu

Purpose: To describe a case of serous retinal detachment (SRD) in a child with glaucoma associated with port wine birthmark (PWB) after initiating prostaglandin (PGA) therapy.

Method: Single case report.

29

Case: An 11-year-old girl with PWB, ipsilateral diffuse choroidal haemangioma (DCH), and glaucoma experienced decreased vision upon starting treatment with bimatoprost. The patient was switched to latanoprostene bunod. Her vision remained reduced. Shortly after, she was diagnosed with SRD. Both SRD and vision improved following PGA cessation.

Conclusion: Patients with PWB are likely to have DCH and glaucoma. DCH itself poses a risk factor for SRD. Certain glaucoma management modalities may further increase this risk. Our report highlights the importance of regular surveillance for SRD in patients with DCH who are receiving PGA.

43 PRESERFLO MicroShunt-implantation surgery and a small case series of paediatric patients Susana Duarte

A Lima-Cabrita, R Barão, A Barata, C Brito, LAbegão Pinto, F Jorge Teixeira

Hospital de Santa Maria, Centro Hospital, Lisbon, Portugal. Correspondence: susanairduarte@gmail.com

Background: The PRESERFLO MicroShunt (Santen, Osaka, Japan) is a glaucoma drainage device intended for reduction of intraocular pressure (IOP). Although it is indicated for patients with primary open-angle glaucoma refractory to medical therapy, it may also be useful in the treatment of other types of glaucoma, such as childhood glaucoma.

Methods: To present a PRESERFLO implantation surgery along with a retrospective small case series in childhood glaucoma patients. Age, gender and diagnosis, as well as visual acuity, IOP, and treatment in the preoperative visit and all follow-up visits were collected. Outcome measures included IOP reduction from baseline, mean IOP change from baseline and medication use at 6 months.

Results: Eight patients were included, 5 (63%) females. The mean age was 11.9 ± 4.6 years. Childhood glaucoma diagnosis included primary congenital glaucoma (n=4), uveitic glaucoma (n=3) and one case of glaucoma secondary to neurofibromatosis. The average preoperative IOP was 24.6±3.6 mmHg (range 20 to 30) on 2.4±1.2 glaucoma medications, and average postoperative IOP was 19.6±9.6 mmHg (range 9 to 40) on 1.4±1 medications, at 6 months. Five patients (63%) had a ≥30% IOP reduction from baseline at 6 months. Three patients registered an increase in IOP at 6 months.

Conclusion: PRESERFLO can be used successfully to treat uncontrolled IOP in selected childhood glaucoma cases.

44 Uveitis-glaucoma-hyphaema syndrome causing secondary paediatric glaucoma Pooja Pendri

E Bitrian

30

Bascom Palmer Eye Institute, Miami, USA. Correspondence: pxp699@miami.edu

Introduction: Uveitis-glaucoma-hyphaema syndrome is a rare form of secondary glaucoma among paediatric patients. Its infrequency may lead to delayed diagnosis and management of this condition.

Case description: A patient with past ocular history of congenital cataracts status post cataract extraction with posterior chamber intraocular lens (IOL) insertion in both eyes and presumed glaucoma following cataract surgery presented 19 years after cataract surgery with an intraocular pressure (IOP) of 38 mmHg in the right eye on dorzolamidetimolol, travoprost, pilocarpine and oral methazolamide therapy. IOP was 19 mmHg in the left eye on no topical medications. Slit lamp examination of the right eye showed a single-piece acrylic IOL in the capsular bag, with a superotemporal haptic in the sulcus space. Gonioscopic evaluation revealed open angles with increased pigment in the right eye. Ultrasound biomicroscopy showed the IOL optic displaced superotemporally and tilted anteriorly with iris touch and the superotemporal haptic in the sulcus and contacting posterior iris. Fundus photos, optical coherence and Humphrey visual field testing confirmed progressive glaucomatous damage in the right eye. The patient underwent combined lens exchange with secondary sulcus lens placement and 180 degrees transluminal trabeculotomy (see video). Postoperatively, the patient had initial IOP reduction but then developed refractory glaucoma necessitating glaucoma drainage device implantation.

Discussion: This case illustrates the potential for progressive glaucomatous damage occurring years after paediatric cataract surgery with a malpositioned IOL and highlights the value of a careful examination. Recognition of this rare presentation is critical to the timely management and prevention of vision loss.

45 Juvenile xanthogranuloma with neovascular glaucoma masquerading as primary congenital glaucoma Juan P López¹

A Schweikart¹. P Montecinos².

1 - Clínica Alemana de Santiago, Santiago, Chile. 2 - Hospital Luis Calvo Mackenna, Santiago, Chile. Correspondence: jlopezg@alemana.cl

Introduction: Juvenile xanthogranuloma (JXG) is a primarily benign skin disorder mostly affecting infants. About 10% of the cases develop ocular involvement, being unilateral iris nodules with hyphaema and glaucoma the most common manifestation. Glaucoma may be secondary to a variety of mechanisms, including blockage of the trabecular meshwork with histocytes, red blood and inflammatory cells, peripheral anterior synechiae and neovascular glaucoma.

Clinical case: A four-month-old otherwise healthy baby girl presented with severe photophobia, epiphora and blepharospasm associated with bilateral corneal clouding suggesting primary congenital glaucoma (PCG). Examination under anaesthesia revealed Haab's striae OS and bilateral hyphaema and fibrin in the anterior chamber, which prompted a complete ophthalmic and systemic diagnostic workup to rule out intraocular and systemic malignancies, coagulopathies and child abuse. All of which were within normal limits. Iris neovascularisation was also observed OU without evident iris lesions or heterochromia iridis. Anterior chamber aspiration cytology revealed normal blood cells. Avastin was injected OU in order to treat the neovascularisation. The right eye responded well to topical glaucoma treatment but the left eye required an Ahmed valve implantation. During this procedure a peripheral iridectomy was performed and the biopsy specimen confirmed JXG. During the following months the patient was treated with periocular steroids achieving good glaucoma control in OD, but persistent severe corneal opacification in OS. No recurrence of the hyphaema, iris or skin lesions have been observed after 6 years of follow-up.

Conclusion: JXG can present early in infants with a clinical picture mimicking bilateral PCG.

31

46 Clinical evaluation protocol for childhood glaucoma

Manju Anilkumar

SR Krishnadas, C Pabloo, GV Puthuran Aravind Eye Hospital, Madurai, India. Correspondence: manju@aravind.org

Introduction: We need to have a structured clinical protocol for the evaluation of childhood glaucoma.

Methods: A clinical evaluation protocol was developed after examining 640 patients, with different diagnosis, in different age groups.

Discussion: We have found this structured protocol to be very useful for the evaluation of various types of glaucoma in children, and as the protocol for treatment and subsequent follow-up according to the severity of the disease.



47 Surgical management of bilateral angle closure in a nanophthalmous patient during acute phacomorphic glaucoma attack in the right eye versus chronic angle closure Rayan Abou Khzam

E Bitrian Bascom Palmer Eye Institute, Miami, USA. Correspondence: rxa961@miami.edu

Case: We present a 75-year-old female with nanophthalmia and bilateral appositional angle closure. We compare the surgical approach to the right eye during phacomorphic glaucoma attack (IOP of 50 mmHg "full IOP lowering agents", Hazy cornea with shallow AC, cell and flare), versus the left eye with chronic angle closure (IOP 19 mmHg, clear cornea, patent PI and shallow AC=1.39 mm with appositional angle closure).

- *Right eye:* A core 23G-vitrectomy was performed to create space posterior to the lens and aid in deepening of the anterior chamber, following that total lensectomy (capsule, cortex, and nucleus) was attempted using a fragmatome through a 20G-sclerotomy. An iridotomy was made using the vitrectomy cutter to prevent postoperative pupillary block. Vitrectomy ports and sclerectomy were sutured to prevent any leak/ hypotony and alleviate the risk of choroidal haemorrhage. Postoperative IOP improved to 18 mmHg and the anterior chamber demonstrated deepening. A secondary sutured IOL was placed later using the Yamane technique.
- *Left eye:* Cataract extraction, capsular tension ring placement to stabilise the bag and insertion of PCIOL was performed with no intra- or postoperative challenges. Postoperative IOP improved to 11 mmHg and the anterior chamber demonstrated deepening.

Conclusions: Identification of appositional angle closure before an acute attack saves the patient the risks of acute IOP rise, simplifies the surgical approach and promotes faster recovery. Phacomorphic glaucoma (PG) is a rare but clinically significant presentation requiring emergent cataract surgery. After cataract extraction, visual recovery is expected, intraocular pressure usually does not require additional surgeries.

32

P1 Comparison of genetic resource utilisation among ophthalmologists

Deniz Goodman¹

M Villalba¹, L Huertas¹, AV Dumitrescu , TCP Chang¹ 1 - Bascom Palmer Eye Institute, Miami, USA. 2 - University of Iowa, Iowa, USA. Correspondence: dqm@bu.edu

Purpose: Inherited diseases are frequently responsible for blindness. We compare the utilisation pattern of genetic resources between different types of ophthalmologists.

Methods: A 30-item survey containing true/false, multiplechoice, and Likert scale questions was sent to the American Association of Paediatric Ophthalmology and Strabismus (AAPOS). Respondents described their genetic practice patterns regarding referring to a provider, ordering tests, and interpreting the results. Nonparametric tests and descriptive statistics were used to compare practice patterns between respondents by role.

Results: There was a disparity in the proportion of respondents routinely referring to an in-house provider (paediatric ophthalmologist (PO, n=77): 59.7%; glaucoma specialist (GS, n=43): 20.9%; general ophthalmologist (GO, n=5): 0%. There was also a disparity in the proportion of respondents reporting that their lack of personal knowledge, practice guidelines, or training in ordering testing were the largest barriers to testing in their practice (GO: 60%, GS: 41.9%, PO: 29.3%). There was no significant trend in the frequency of ordering tests among PO (p=0.0544), GS (p=0.0813), or GO (p=0.2967). A decreasing frequency of test interpretation was significantly associated with PO (p=0.0308) and GS (p=0.0443), while all GO reported that they never interpret tests.

Conclusions: There exist discrepancies in genetic resource utilisation between types of ophthalmologists, which correspond to variations in provider knowledge. Genetic resource utilisation may be increased by publishing practice guidelines for ordering and interpreting tests as well as developing databases of local genetic providers.

POSTERS

P2 Traumatic hyphaema

Aminah Iffah Jawaheer

James Morris; Joseph Aslan, Kate Barnes *Musgrove Park Hospital, Taunton, UK.* Correspondence: aminah.jawaheer@somersetft.nhs.uk

Background: A 9-year-old girl was accidentally poked in the right eye with the sidearm of a pair of glasses.

Case: On presentation there was 70% right eye hyphaema obscuring the visual axis with vision down to HM and an IOP of 33 mmHg with no fundal view. Her left eye was unaffected with 6/6 vision, normal IOP and unremarkable ocular examination. She was commenced on g.maxidex, g.timolol and g.cyclopentolate. The hyphaema subsequently worsened to complete obscuration of the anterior chamber, with peak IOP of 52 mmHg and vision NPL, despite maximum medical management. The decision was then made to have an anterior chamber washout under general anaesthetic, which was done 6 days after initial review. There were no intraoperative complications. Vision steadily improved to 6/7.5 and IOP stabilised in the mid-teens. Clinically, there is residual corneal tattooing and pigmented fibrinous debris on the anterior lens capsule. Posterior examination is unremarkable and optic nerve function remains intact. The patient is on g.cosopt BD and a tapering course of g.maxidex. Other findings showed an abnormal clotting screen (low Factor XIII assay and Von Willebrand factor) which is being investigated by the paediatric and haematology team.

Conclusions: Where the IOP is not responsive to medical management, anterior chamber washout needs to be considered in traumatic hyphaema cases. It is also important to investigate any possible systemic risk factors for hyphaema in traumatic cases where the mechanism of injury is inconsistent with the severity of clinical findings.

P3 Aniridia and anterior segment dysgenesis: An uncommon union

Sirisha Senthil

K Naik, B Bhagga LV Prasad Eye Institute, Hyderabad, India. Correspondence: sirishasenthil@lvpei.org

Introduction: Congenital aniridia, typically characterised by partial or total iris absence, usually exhibits residual iris tissue when examined closely. However, we present a unique case of complete aniridia in a 1-month-old infant, revealing the absence of iris tissue with a detached Descemet's membrane, substantial corneal enlargement, severe haze, and elevated intraocular pressures. Genetic testing identified a *PITX2* gene mutation, associated with anterior segment dysgenesis 4 (ASGD4). This combination of complete aniridia and a *PITX2* mutation is exceedingly rare, signifying a severe form of anterior segment dysgenesis accompanied by aniridia and congenital glaucoma.

Case: The infant, born to non-consanguineous parents, displayed bilateral whitish eye discoloration since birth. Clinical examination unveiled megalocornea, grade 5 corneal haze and oedema in both eyes, along with a "golden arc sign" indicative of aniridia. A subsequent ultrasound biomicroscopy (UBM) depicted the total absence of iris tissue, a lens suspended by zonules, and a membrane connecting the cornea to the lens surface, likely a detached Descemet's membrane. Further evaluation under anaesthesia disclosed elevated intraocular pressures (IOP) measuring 26 mmHg and 22 mmHg in the right and left eyes, respectively, with a horizontal corneal diameter of 13 mm in both eyes. Genetic testing confirmed the diagnosis of aniridia.

Subsequently, the patient underwent combined trabeculectomy in both eyes after trabeculotomy was aborted due to Schlemm's canal non-visualisation. Peripheral iridectomy was omitted due to minimal peripheral iris remnants. Postoperatively, IOP was well controlled with partial improvement in corneal clarity. Persistent corneal haze prompted optical penetrating keratoplasty in both eyes, leading to clear grafts and controlled IOP. Clinical genome sequencing identified a likely pathogenic *PITX2* gene variant associated with ASGD4.

Summary: This case illustrates the rarity of complete aniridia linked with a *PITX2* mutation and underscores the importance of comprehensive management in addressing the challenges posed by such severe anterior segment dysgenesis conditions.

NOTES:	
	34

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35	35

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UKPGS is an affiliated society of Glaucoma UK. We thank them for their continued support.



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