SESSION: Updates on Pediatric Vitreoretinal Surgery
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MODERATORS: Antonio Capone, Şengül Özdek

Wnt Retinopathies

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The Wnt signaling pathway plays a pivotal role in vascular morphogenesis in the eye, ear and central nervous system. Wnt ligands and receptors are key regulators of ocular angiogenesis during development. Mutations in Wnt signaling components cause rare genetic eye diseases in humans such as Norrie disease, osteoporosis-pseudoglioma syndrome (OPPG) and familial exudative vitreoretinopathy (FEVR) with defective ocular vasculature. This lecture will provide insights into the pathogenesis of Wnt retinopathies, their management, and avenues of pharmacotherapeutic research.
Keywords: Wnt signaling, Norrie disease, osteoporosis-pseudoglioma syndrome (OPPG), familial exudative vitreoretinopathy (FEVR)

Anti-VEGF drugs for pediatric retinal diseases? - ***Anna Ells***

The importance of fluorescein angiography in ROP

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Purpose: To present the findings of fluorescein angiography (FA) in ROP and the importance of this exam in ROP monitorization.

Introduction: FA is a relatively safe procedure in paediatric population. It enables us characterize the vascular anomalies associated with ROP that are not readily detected in indirect ophthalmoscopy. It can accurately characterize ROP stages, enhance vascular anomalies after anti-VEGF treatment, monitoring vascular interface in peripheral avascular retina and help in the detection of recurrence. Also has a role in the exclusion of the different diseases that are differential diagnosis of ROP.
Methods: A retrospective analyses of FA images obtained between January 2010 and June 2023 in a single institution. The authors present the fluorescein findings in the different ROP stages, vascular anomalies after anti-VEGF therapy, vascular interface in PAR and ROP recurrences.
Results: The authors present the fluorescein findings in: the different ROP stages, the vascular anomalies after anti-VEGF therapy, the characterization of the vascular interface in PAR and in ROP recurrences.

Conclusion: FA provides a body of information that help us in the characterization of ROP and in the monitorization of peripheral vascular changes after anti-VEGF treatment. It is a very promising exam that should be included in future studies in order to find reliable biomarkers for recurrence and progressive ROP.

Keywords: ROP, Fluorescein angiography, peripheral avascular retina

Surgical management of adult ROP complications

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In addition to potential manifestations in infancy, ROP is a life-long disease that can present with numerous late-onset complications. Some of these complications including peripheral retinoschisis, retinal detachment, and vitreomacular traction require surgical intervention. This presentation will review surgical approaches to adult ROP complications using a cased based approach.

Keywords: ROP, surgery

Laser Prophylaxis in Patients with Stickler Syndrome

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Purpose: To evaluate the association among laser prophylaxis treatment, retinal detachment (RD), and visual acuity (VA) in patients with Stickler syndrome (SS).
Design: Retrospective comparative case series.
Participants: Patients with SS.
Methods: Patients received extended vitreous base laser (EVBL), nonprotocol laser (NPL), or no laser prophylaxis treatment of any kind.
Main Outcome Measures: The 2 main outcome measures that were examined in these patients were rates of RD and VA.
Results: In this study, 230 eyes of 115 patients were included. Fifty-nine patients were women (51%). The median age at the time of laser prophylaxis treatment was 9.5 years (interquartile range [IQR], 6e13 years), and the median age of patients with RD was 11 years (IQR, 7e18 years). Of the 230 eyes, 92 did not undergo any laser treatment, 9 received NPL treatment, and 129 received EVBL treatment. Of the 129 eyes that underwent EVBL treatment, 4 (3%) had RD, compared with 74 eyes (73%) that had RD and did not receive laser or NPL treatment ($P < 0.001$). Eyes that received EVBL treatment had approximately 8 lines better vision, on average, compared with those that did not receive laser or NPL treatment (0.86 logarithm of the minimum angle of resolution; 95% confidence interval, 1.1 to 0.64; $P < 0.001$).
Conclusions: Treatment with EVBL seems to reduce the rate of subsequent RD and is associated with better VA in patients with SS.

Keywords: Stickler Syndrome, Laser Prophylaxis

The Long-term surgical outcomes of patients with posterior and combined PFV - ***Wei Chi Wu***

Severe anterior persistent fetal vasculature: the role of anterior retinal elongation on prognosis

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Purpose: To investigate surgical outcomes of eyes with severe anterior PFV and the role of associated anatomical anomalies on prognosis.
Methods: Consecutive series of 32 eyes of 31 patients with severe anterior PFV, defined as fibrovascular tissue totally covering the back of cataractous lens. Based on the degree of anterior retinal elongations, cases were classified as follows: Group 1, eyes with well-developed pars plana and minor/no abnormalities (n=11, 34%); Group 2, eyes with partially-developed pars plana and broader-based elongations (n=9, 28%). Group 3, eyes with no visible pars plana and fibrovascular membrane (FVM) having 360-degree continuity with peripheral retina (n=12, 38%). Complications, functional and anatomical outcomes were investigated.
Results: The median surgical age was 2 (1-12) months. The median follow-up was 26 (6-120) months. Seventy-three percent in Group 1 achieved finger counting or better vision with a single surgery and without any pupillary/retinal complication. Group 2 and 3 required 2.1±0.9 and 2.6±1.2 surgeries on average. Pupillary obliteration and RD occurred in 33% and 22% in Group 2, and 58% and 67% in Group 3. Retina remained attached after silicone oil removal in 89% of Group 2 and in 25% of Group 3. Phthisis developed in one eye (11%) in Group 2 and 6 eyes (50%) in Group 3. Incomplete removal of FVM was associated with a 4-fold increased risk of pupillary obliteration and a 3-fold increased risk of postoperative RD.
Conclusion: Peripheral retinal anomalies are common in severe anterior PFV and have a major impact on prognosis. Prognosis is favorable in cases with mild to moderate anomalies with complete removal of FVM and appropriate management of possible retinal tears. In eyes with 360-degree retinal elongations, severe fibrous proliferation and eventual eye loss are common.

Keywords: persistent fetal vasculature, anterior PFV, peripheral retinal elongation

Surgical Outcomes of Posterior Persistent Fetal Vasculature Syndrome: Cases with Tent-Shaped and Closed Funnel-Shaped Retinal Detachment

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Purpose: To determine the role of vitreoretinal surgery (VRS) for two different forms of posterior persistent fetal vasculature syndrome (PFVS); with tent-shaped tractional retinal detachment (TRD) and closed funnel-shaped TRD.
Methods: Retrospective, single-surgeon, consecutive series of 52 eyes of 44 patients with posterior PFVS who underwent VRS. Cases were divided into "tent-shaped RD" and "funnel-shaped RD" groups based on the preoperative TRD configuration. Functional and anatomical outcomes were evaluated.
Results: 30 eyes of 29 patients presented with tent-shaped TRD; 67% obtained counting fingers or better vision, 90% achieved complete or near-complete attachment of the retina. 22 eyes of 15 patients presented with leukocoric pupils associated with funnel-shaped TRD; 45% achieved LP vision, and cosmetically acceptable appearance in 86%. Of the patients with bilateral funnel-shaped RD, 70% had LP in at least one eye. The median length of follow-up was 12 (3-70) months.
Conclusion: VRS often provides functional vision and anatomy in posterior PFVS with tent-shaped TRD morphology. In funnel-shaped TRD morphology, where no treatment has historically been recommended, VRS may be considered with an aim of restoring LP and cosmesis, particularly in bilateral cases.

Keywords: Persistent fetal vasculature, posterior PFV, retinal detachment

Subfoveal Nodule in Coats' Disease: Stages, Prognosis, and Treatment

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Purpose: To investigate the prevalence, developmental stages, clinical and demographic factors affecting subfoveal nodule (SFN) development in Coats' disease.
Methods: The medical records and multimodal images of patients with Coats' disease were reviewed retrospectively in a tertiary university setting. Patients with Coats' disease at stages from 2A to 3A were included. SFN development was divided into five stages as follows: Stage 0, macular exudation without subfoveal hard exudate; stage 1, subfoveal exudation; stage 2, packaging of exudates; stage 3, vascularization of SFN; stage 4, subfoveal fibrotic scar. The factors which may have role in the formation and the tempo of SFN development were analyzed.
Results: Study included 43 eyes of 42 patients with Coats' disease with a mean age of 6.9±3.9 years and a mean of follow-up of 31.1±29.5 months. The prevalence of SFN was 48% at the beginning and 95% at final follow-up. There was no difference in baseline demographic and clinical characteristics among patients with different SFN stages. Patients developing SFN earlier than 9 months were significantly younger than those in longer period (>9 months) (5.6±3.2 vs 8.9±4.5, p=0.010), and the mean number of intravitreal (IV) anti-vascular endothelial growth factor (anti-VEGF) injection per year was lower in the latter group (3.6±1.2 vs 1.4±1.2, p=0.001). The risk of early SFN development was 2.4 times higher in patients younger than 7 years of age and 4 times higher in patients who received 3 or less IV anti-VEGF injections per year. Patients with considerably good final best-corrected visual acuity (BCVA) (<1.3 logMAR) had a higher mean number of IV anti-VEGF injections per year than those with poor final BCVA (>=1.3 LogMAR) (p=0.025).
Conclusion: SFN is commonly seen in Coats' disease and its prevalence was found 95% in our study. Anti-VEGF injection seems to decelerate SFN development and improve the functional result.

Keywords: Coats' disease, subfoveal nodule, intravitreal Anti-VEGF injection

Complex syndromic pediatric rhegmatogenous retinal detachment: Surgical technique & outcome

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110 eyes with syndromic pediatric complex rhegmatogenous retinal detachment (RRD), operated during the period from 2018 - 2022 were included in this study. It include eyes with stickler, Marfan, Konblock, congenital coloboma, Dawn and congenital retinoschisis. The RRD was associated with giant retinal tears, multiple retinal breaks, posterior retinal breaks, extensive lattice and/or proliferative vitreorerinopathy (PVR). The surgical technique included lensectomy with or without IOL implantation, 23 or 25 G. vitrectomy, unimanual or bimanual detachemnt & peeling of the posterior hyaloid (PH), extended internal limiting membrane (ILM) peeling, excision of the basal vitreous gel, retinal reattachment with PFCL, endolaser and direct PFCL/silicone oil 2000 Cs. exchange. Scleral buckle (SB) was used in the first 15 eyes. Retinotomy/retinectomy was only performed when mandatory
SiO was removed after achieving complete stable retinal attachment.
The number of operations needed to achieve retina reattachment, including SiO removal was 2 in almost 60%, 3 in 24%, 4 in 5%, 5 in 6.3%, and 6 operations in 5%. SiO could not be removed because of hypotony, persistent RD or PVR in 3%.
The challenges involved in such cases include: PH detachment & peeling, ILM peeling, recurrent proliferatio. Extended ILM peeling is a safeguard against epimacular recurrent proliferation.
No diifference was noted between eyes with or without SB.

Keywords: Syndromic pediatric RRD, extended ILM peeling

Development of Cataract following Lens-Sparing Vitrectomy in Pediatric Eyes

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Purpose:
We aimed to present the development of cataract and associated factors following lens-sparing vitrectomy (LSV) in the pediatric age group.
Method:
The research was a retrospective review of pediatric patients who underwent LSV from January 2011 to June 2022.The data was collected according to diagnosis, age at the time of surgery, presence of preoperative retina-lens apposition, presence of preoperative lens opacity, use of a tamponade, direct lens trauma during surgery, final anatomical success, as well as non-operative procedures like laser photocoagulation, cryotherapy, intravitreal anti-VEGF injections, and presence of any additional vitreoretinal surgery performed after LSV surgery.
Results:
294 eyes of 240 pediatric patients were included. Underlying diseases were ROP in 127 eyes, FEVR in 44,Coats in 16,posterior PFV in 19,trauma in 23,CXLR in 17 and miscellaneous (choroidal coloboma, terson syndrome, posterior uveitis, combined hamartoma of retina and retina pigment epithelium) in 48. The mean follow-up was 39 months (min 1, max 127, ± 33.2). Lens opacity developed in 67 of 294 eyes (22.7%). The mean time between primary surgery and the development of lens opacity was 27.2± 36.3 months (1-123 months), of which 32 eyes (10,8%) underwent lensectomy. In the remaining 35 eyes, surgery was not recommended.7 other eyes (29.1%) underwent lensectomy because of retinal problems.The mean time between primary surgery and lensectomy was 19.2 months (± 17.6, min:1, max:65 months).Direct lens trauma during surgery was observed in 6 (2%) eyes,and lensectomy was performed in 5 (83.3%) of these eyes.Retina-lens apposition (p: 0.00) and number of surgeries (p: 0.009) were found to be significantly associated with the lens opacity development.
Conclusions:
Development of lens opacity following LSV in pediatric ages usually takes much longer time than adults and develops in a lower rate.Lens opacity rate is higher in eyes with prior lens-retina apposition and with multiple vitreoretinal surgical procedures.

Keywords: lens sparing vitrectomy, cataract, pediatric eyes

Congenital X-Linked Retinoschisis: Surgical Needs And Outcomes In Long Term A Retrospective Multicenter International Study

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Purpose: To evaluate the need for vitreoretinal surgery (VRS), surgical success and factors affecting success in patients with congenital X-linked retinoschisis (CXLR).
Methods: This retrospective, multicenter study included the data of invited vitreoretinal centers after approval of the local ethics committee. A data set including demographic characteristics, phenotype, frequency of follow-up/surgery, surgical techniques and details, pre- and post-operative visual acuity and complications of CXLR patients followed-up were obtained and analyzed.
Results: The data of 635 eyes of 318 patients were analyzed. Mean follow-up time was 115±93 months. VRS was performed to 112 of 589 eyes (17.6%). VRS was needed most frequently in patients with complex phenotype ($p<0.0001$). The mean age at initial presentation was 9 (1-32) years and statistically signicantly lower in patients requiring VRS ($p=0.004$). The median baseline visual acuity (VA) of the patients underwent VRS was 1.6 LogMAR. The most common cause for VRS was rhegmatogenous retinal detachment (RRD) (61.6%) and retinal tears were most common in the lower quadrant (44.9%). The most frequent surgical procedure was vitrectomy alone in the whole group (42.9%), however, combined vitrectomy with scleral buckle was the most common procedure in eyes with RRD (53.8%) ($p<0.0001$). Inner-wall retinectomy was performed in 59.5% of eyes. Silicone oil tamponade was used in 53.8% of the eyes. Anatomical success was achieved in 68.5% with single surgery which was statistically higher in eyes with RRD and vitreous hemorrhage. There was no statistically signicant difference between the surgical method and maneuvers, and tamponade used in achieving success with a single surgery ($p>0.05$). Final anatomical success rate was 92.8%. The median final VA improved to 1.3 LogMAR ($p=0.006$).
Conclusions: VRS for CXLR related problems results in relatively lower single surgery anatomical success rates which increase to satisfactory levels with multiple surgeries. Functional results are also guarded in these eyes.

Keywords: congenital x-linked retinoschisis, retinal detachment, vitreoretinal surgery

Long-Term Surgical Outcomes of Pediatric Retinal Detachment Associated with Congenital Glaucoma

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Background: Congenital glaucoma is an extremely rare disease with an incidence of one in 10,000 live births. Increasing axial length due to congenital glaucoma predisposes to retinal thinning and retinal detachment (RD). The aim of the present study is to determine the long-term surgical outcomes of pediatric RD associated with congenital glaucoma.
Methods: This study was a retrospective, nonrandomized study including pediatric patients who underwent vitrectomy with silicone oil injection for RD associated with congenital glaucoma. Ocular examination reports and surgical findings of all patients were analyzed to assess for the anatomical and functional outcomes of the surgical intervention.
Results: The study included 20 eyes of 20 children. Mean age was 10.33 ± 3.94 years, range 5–16 years. Mean axial length of these eyes was 25.91 ± 2.15 mm (range, 20.31–28.91 mm). In 11 eyes, the IOP was <6 mmHg at the time of RD diagnosis. The mean LogMAR visual acuity improved from 2.13±0.45 to 2.02±0.61 (P =0.477) after surgery. Mean follow-up period was 24.05 ± 11.05 months, range 12–48 months. During the follow-up period, 6 patients (30%) had revision of surgery due to recurrent RD. Globe survival has been achieved in 19 out of 20 eyes (95%).
Conclusion: Ocular comorbidities like corneal scarring, amblyopia, or glaucomatous optic atrophy play a significant role in limiting final visual outcomes. Despite the poor visual prognosis of RD repair in eyes with congenital glaucoma, globe survival can achieve in such cases. Globe preservation has an important impact on patients' quality of life, psychological and social functionality. Moreover, ambulatory vision can be preserved in most eyes.

Keywords: Congenital glaucoma, Pars plana vitrectomy, Retinal detachment