ORAL ABSTRACTS

NPOG SESSION I: GENETICS

MOLECULAR ANALYSIS IN DIAGNOSIS OF HEREDITARY DISEASES

Ingvild Aukrust, Norway

Abstract not available.

EXTENDED MOLECULAR GENETIC ANALYSIS IN PATIENTS WITH RETINAL DYSTROPHY

Karen Grønskov, Denmark

Abstract not available.

LUXTURNA:IMPLICATIONS FOR PATIENT TREATMENT

Ragnheidur Bragadottir, Norway

Abstract not available.

CONGENTIAL ANIRIDIA: A GENETIC DISEASE

Erlend Landsend, Norway

Abstract not available.

ALBINISM: CLINICAL CONSIDERATIONS

Lotte Welinder, Denmark

Abstract not available.

ALBINISM: GENETICAL CONSIDERATIONS

Karen Grønskov, Denmark

Abstract not available.

HEREDITARY FORMS OF CORNEAL VASCULARIZATION AND IMPLICATIONS FOR PATIENT TREATMENT

Cecilie Bredrup, Norway

Abstract not available.

KERATOENDOTHELIITIS FUGAX HEREDITARIA ASSOCIATED WITH NLRP3 MUTATIONS

Joni Turunen, Finland

Abstract not available.

ECTOPIA LENTIS ET PUPILLAE DUE TO MUTATIONS IN ADAMTSL4

Anne Christensen Mellgren, Norway

Abstract not available.

NPOG SESSION II: UVEITIS

NEW GUIDELINES FOR TREATMENT OF PAEDIATRIC UVEITIS

Stefan Berg, Sweden

Abstract not available.

TREATMENT OF SECONDARY CATARACT AND MACULAR OEDEMA

Sanna Leinonen, Finland

Abstract not available.

NPOG SESSION III: FREE PAPERS AND CASES TO LEARN FROM

VISUAL PERCEPTION IN CHILDREN WITH OPTIC DISC COLOBOMA

Athanasia Skriapa Manta[1,2], Evin Guven[3], Monica Olsson[1,2], Ulla Ek[4], Ronny Wickström[5] and Kristina Teär Fahnehjelm[1,2]

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Purpose: The aim of the study was to evaluate the visuoperceptual capability in children with Optic Disc Coloboma (ODC).

Background: ODC is one of the most common optic disc malformations in children. Visual perception (VP) is the ability to interpret visual information in order to comprehend our surroundings and can be divided into five different areas: recognition, orientation, depth perception, perception of movement and simultaneous perception. Deficiencies in VP may cause significant disability. Influencing factors include not only visual function but also cognition and other co-existing morbidities..

Methods: We performed a population-based, cross-sectional study of 23 ODC children (2-18 years). The children were part of a larger cohort of 31 children diagnosed with ODC. A systematic history taking from the parents aimed to assess the five areas of VP was performed. The age of the children, gender, other associated ocular diagnosis, behavioral- and developmental aspects reported previously will be considered during our analyses.

Results: Preliminary results indicate that there are problems in the area of perception of movements among ODC children for example in observing rapidly approaching objects. Detailed analyses are ongoing and will be presented.

Discussion: VP-problems in children with ODC seem limited but must be related to visual functions, cognition and other co-existing problems. The results of the current study may lead to new insights to how children with ODC and associated co-morbidity may have an increased incapability regarding visual perception. The study may also serve as reference for further research regarding VP in children with other optic disc malformations.

OCULAR CHARACTERISTICS IN CHILDREN AND ADULTS WITH FABRY DISEASE

Monica Olsson[1,2], Branka Samolov[1,2], Hani Tavakolikolagari[3], Karin Naess[4,5,6], Mikael Oscarson[7] and Kristina Teär Fahnehjelm[1,2]

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Purpose: The aim of this pilot study was to assess visual function and ocular characteristics in children and adults with Fabry disease (FD).

Background: FD is an X-linked recessively inherited lysosomal storage disease where decreased enzyme activity of alpha-Galactosidase leads to accumulation of Gb-3 (globotriaosylceramide) in organs and tissues throughout the body. Mainly males but also females risk premature death due to renal failure, cardiomyopathy and or cerebral stroke. Subepithelial corneal whorls, cornea verticillata, are detectable by slit lamp examination at a young age, long before irreversible organ damage occurs.

Methods: Clinical visual and ocular assessments including biomicroscopy, optical coherence tomography, keratometri and fundus photography were performed.

Results: 16 patients aged 11-58 years of age (10 men) were included. A vast majority of the patients had typical ocular pathology. Six patients had conjunctival tortuosity, 12 cornea verticillata, nine cataract and eight had torturous fundus vessels. In only one patient the initial ocular examination lead to diagnosis. Severe astigmatism and dry eyes were also found. Cardiomyopathy was present in seven patients, renal insuffiency in three, minor cerebral strokes in three and severe pain in extremities (acroparestesia) was present in 13 patients.

Discussion: FD disease is one of few diseases were ocular cardinal signs can signal systemic disease. Conjunctival beading/tortuosity, corneal verticillata and typical lens opacities may easily be overlooked. As early diagnosis and enzyme replacement treatment can slow progression of the disease and sometimes be life saving, pediatric ophthalmologists/ ophthalmologists must be updated on typical anamnestic and ocular signs of FD.

OPHTHALMOLOGICAL FINDINGS IN INFANTS WITH NON-SYNDROMIC CRANIOSYNOSTOSIS

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Aim: To report the ophthalmological findings in children with non-syndromic craniosynostosis preoperatively and 6-12 months after surgery.

Methods: Infants referred to Uppsala University Hospital for surgery of craniosynostosis were examined. Visual acuity was measured with Teller acuity cards or observation of fixation and follow. Strabismus and eye motility were noted. Refraction was measured in cycloplegia and fundoscopy was performed. Follow-up examination was performed at the children's local hospital.

Results: 112 infants with mean age 5.9 months were examined preoperatively. The main non-syndromic craniosynostosis was sagittal (68%). Preoperatively, all infants had visual behaviour normal for age. Mean refraction was +2.48 D in right and +2.51 D in left eyes. Strabismus was most common in children with unicoronar synostosis (55.5%). No infant had disc oedema or pale disc.

Postoperatively, 98 children were examined. Mean age was 16.2 months, mean refraction +1.67 D in both eyes. 4 children with unicoronar and 1 with metopic synostosis were prescribed eyeglasses. Strabismus disappeared in 3 children with sagittal synostosis and remained in 3 with unicoronar synostosis. 2 new cases with strabismus in operated unicoronar synostosis appeared. 1 infant with combined synostosis had disc oedema.

Conclusion: In our cohort, children with unicoronar synostosis had higher prevalence of strabismus and anisometropia, some of which developed postoperatively, which indicates the necessity of ophthalmological evaluation before as well as after reconstructive surgery.

OPHTHALMOLOGICAL FINDINGS IN FETAL ALCOHOL SPECTRUM DISORDERS – A FOLLOW-UP STUDY FROM CHILDHOOD TO YOUNG ADULTHOOD

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Purpose: To study if ophthalmological findings in fetal alcohol spectrum disorders (FASD) in childhood persist into young adulthood.

Methods: Thirty children (13 female) adopted from eastern Europe to Sweden in the 1990s and diagnosed with FASD by our multidisciplinary team at the median age of 7.9 years were followed up by the same team 13–18 years later, at a median age of 22 years. Visual acuity (VA), refraction, stereo acuity, strabismus, ocular fundus, and visuoperceptual problems (VPPs) were investigated.

Results: Median VA right/left eye (range) was 0.65/0.65 (0.1-1.0/0.1-1.0) in childhood; in adulthood 0.9/1.0 (0.01-1.0/0.1-1.25). Median refraction right/left eye (range) was 0.88/1.25 (-8.75 to +4.75/-9.38 to +5.25) spherical equivalent diopter (D) in childhood; in adulthood -0.25/-0.25 (-12 to +2.75/-13.25 to +2.63). Astigmatism (≥1D) was the most common refractive error; 40% and 47%, respectively. 43% had heterotropia and four individuals had optic nerve hypoplasia, equal at both examinations. Defect stereo acuity (>60") was noted in 67% in childhood compared with 73% in adulthood. VPPs were found in 8/27 (30%) in childhood and 18/27 (67%) in adulthood (p=0.01).

Conclusions: Ophthalmological findings, such as strabismus, fundus abnormalities and VPPs, are frequent in children with FASD and persist into early adulthood. Ophthalmological examination of children with FASD early in life is important for appropriate interventions. Given the high prevalence of FASD across the world, and a striking frequency of ocular abnormalities, ophthalmologists are not only the key in ameliorating ophthalmological impairments in FASD, but could also contribute in the diagnostic evaluation of the disorder.

References:

1. Andersson Grönlund M, Landgren M, Strömland K, et al. Relationships between ophthalmological and neuropaediatric findings in children adopted from Eastern Europe. Acta Ophthalmol 2010;88(2):227-234.
2. Brennan D, Giles S. Ocular involvement in fetal alcohol spectrum disorder: a review. Curr Pharm Des 2014;20(34):5377-5387.
3. Hoyme HE, May PA, Kalberg WO, et al. A practical clinical approach to diagnosis of fetal alcohol spectrum disorders: clarification of the 1996 Institute of Medicine criteria. Pediatrics 2005;115(1):39-47.
4. Landgren M, Svensson L, Strömland K, Andersson Grönlund M. Prenatal alcohol exposure and neurodevelopmental disorders in children adopted from eastern Europe. Pediatrics 2010;125(5):e1178-e1185.
5. Moore EM, Riley EP. What Happens When Children with Fetal Alcohol Spectrum Disorders Become Adults? Curr Dev Disord Rep. 2015 Sep;2(3):219–27.
6. Popova S, Lange S, Probst C, Gmel G, Rehm J. Estimation of national, regional, and global prevalence of alcohol use during pregnancy and fetal alcohol syndrome: a systematic review and meta-analysis. Lancet Glob Health. 2017 Mar 1;5(3):e290–9.
7. Strömland K. Visual impairment and ocular abnormalities in children with fetal alcohol syndrome. Addict Biol 2004;9(2):153-157; discussion 159-160.

PROFILE OF NORMAL GANGLION CELL- INNER PLEXIFORM LAYER THICKNESS IN HEALTHY 6.5 YEAR-OLD CHILDREN

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Purpose: To obtain macular ganglion-cell-inner plexiform layer (GC-IPL) thickness normative values in healthy eyes of full term born 6.5 year-old children.

Methods: The participants constituted the control group of the Stockholm cohort in the national follow-up Extremely Preterm Infants in Sweden Study (EXPRESS)1. They had been recruited from the Swedish medical register and were 6.5 years at examination. Cycloplegic refraction and visual acuity were evaluated and GCL-IPL was assessed with optical coherence tomography (OCT; Cirrus; Carl Zeiss Meditec, Dublin, CA). Neonatal clinical data, head circumference and intelligence quotient (IQ) at 6.5 years were collected for comparison.

Results: Hundred and eighty-four eyes from 92 children (53 boys/39 girls) were evaluated with OCT. The mean GC-IPL thickness measure was 86.0 (SD 5.4; range 70-100) um. The minimum GC-IPL thickness was 83.3 (SD 5.3; range 67-95) um. Mean GC-IPL thickness was positively correlated with spherical equivalent (P=0.013) but not with visual acuity, birth weight, gestational age, head circumference or IQ.

Conclusions: This study ensures a pediatric normative database of GC-IPL using spectral-domain OCT. This information may provide to diagnosis and monitoring of optic pathway diseases in children2.

References:

1. Hellgren K et al. Ophthalmologic outcome of extremely preterm infants at 6.5 years of age; Extremely Preterm Infants in Sweden Study (EXPRESS). JAMA Ophthalmol. 2016;134:555-562.
2. Muñoz-Gallego A, De la Cruz J, Rodríguez-Salgado M, et al. Assessment of macular ganglion cell complex using optical coherence tomography: Impact of a paediatric reference database in clinical practice. Clin Experiment Ophthalmol.2018:1-8.

NPOG SESSION IV: RETINOPATHY OF PREMATURITY

TEN YEARS OF ROP-SCREENING AND TREATMENT IN SWEDEN - RESULTS FROM THE SWEDROP REGISTER.

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A Wallin, St Erik Eye Hospital, Stockholm, Sweden

Purpose: Evaluation of the national screening program for ROP based on ten years of ROP-screening and treatment in Sweden.

Methods: Today all infants born at 30 weeks gestational age (GA) or less, as well as children born later but being extremely sick, are screened for ROP in Sweden during the neonatal period. SWEDROP, a national registry for ROP, has around 97% coverage.

Results: During the ten-year period between 2008 and 2017, 7249 infants with a GA of 30 weeks or less were registered in SWEDROP. Mean GA was 27.6 w (range 21-30 w) and mean BW was 1116 g (range 307-3245 g). During the study period ROP was found in 31.9% (range 26.8 -36.9%) and treatment for ROP was performed in 6.1% (range 4.1-7.8%) of the children. The incidence of ROP did not change during the study period, while there was a significant increase in the frequency of treatment (p=0.023). No infant with GA 30 w and only four infants with GA 29 w were treated for ROP. Eighty-two per cent (361/440) of the treated infants had laser only and 17.7% (78/440) were treated with Anti-VEGF, alone (17 infants) or in combination with other treatment.

During the ten-year study period altogether 46.038 examinations were performed, of which 5.232 (11.4%) in infants born with GA 30 w (30 + 0-6).

Conclusions: The frequency of ROP treatment was increased over the ten-year period, but no child born at 30 w GA was treated for ROP. Modification of guidelines is considered, with lowering the upper limit of screening with one week to less than 30 weeks GA. During the ten-year study period, this would have resulted in a reduction of 1681 infants (23.2%) screened for ROP and of 5.232 (11.4%) examinations.

LOW CONTRAST VISUAL ACUITY OUTCOME OF EXTREMELY PRETERM BORN 6.5 YEAR-OLD CHILDREN; EXTREMELY PRETERM INFANTS STUDY IN SWEDEN (EXPRESS)

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Purpose: To investigate low contrast visual acuity (LCVA) in relation to high contrast visual acuity (HCVA) in a national cohort of children born extremely preterm (EPT) in comparison to an age matched control group of full term children at 6.5 years of age.

Methods: Binocular distance linear VA with habitual correction was assessed at 3 meters with LEA SYMBOLS ® chart. The test was performed both with a high contrast chart (100%) and with a low contrast chart (2,5%). In order to evaluate the discrepancy between HCVA and LCVA for each child a contrast-ratio was calculated by dividing HCVA with LCVA. Cycloplegic refraction was measured.

Results: Threehundred and thirtythree of 483 EPT children (69%) were assessed together with 296 controls. The EPT group had significantly lower LCVA (mean 0.43 [SD 0.13]) than the control group (mean 0.51 [SD 0.11]; $p < 0.001$), and also a higher contrast-ratio (2.26 [SD 0.73] versus 2.12 [SD 0.54]; $p < 0.001$). Within the EPT group the contrast-ratio was significantly associated to high refractive errors and to severe intraventricular hemorrhage, but not to ROP or gestational week at birth.

Conclusions: The EPT group had significantly larger discrepancy between HCVA and LCVA than the controls and the discrepancy was associated to high refractive errors and intraventricular hemorrhage.

Reference: Hellgren, K., Tornqvist, K., Jakobsson, P., Lundgren, P., Carlsson, B., Källén, K., .Holmström, G. (n.d.). Ophthalmologic Outcome of Extremely Preterm Infants at 6.5 Years of Age: Extremely Preterm Infants in Sweden Study (EXPRESS). JAMA Ophthalmology., JAMA ophthalmology. , 2016.

PREDICTION, PREVENTION AND UP-TO DATE TREATMENT OF SEVERE ROP

Ann Hellström, Sweden

Abstract not available.

STABLE INCIDENCE OF RETINOPATHY OF PREMATURITY, BUT INSTITUTIONAL DIFFERENCES IN NORWAY

Beanca Grottenberg, Norway

Abstract not available.

OUTCOMES AFTER TREATMENT FOR RETINOPATHY OF PREMATURITY

Hajer Ahmad, Denmark

Abstract not available.

TWENTY-FIVE-YEAR-OLD OPHTHALMOLOGICAL FOLLOW-UP IN PREMATURELY BORN INDIVIDUALS

Dýrleif Pétursdóttir, Sweden

Abstract not available.

RETINOPATHY OF PREMATURITY IS A DIABETES-LIKE RETINOPATHY

Carina Slidsborg, Denmark

Abstract not available.

VISUOPATHY OF PREMATURITY - IS RETINOPATHY OF PREMATURITY JUST THE TIP OF THE ICEBERG?

Tora Sund Morken, Norway

Abstract not available.

NPOG SESSION V: FREE PAPERS AND CASES TO LEARN FROM

THE PEDIATRIC CATARACT REGISTER (PECARE): OUTCOME AT FIVE YEARS OF AGE FOLLOWING EARLY SURGERY FOR DENSE UNILATERAL CONGENITAL CATARACTS

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Purpose: To investigate the development of visual acuity (VA) and postoperative complications in a national register-based cohort study and compare results with earlier Swedish retrospective reports. Furthermore, to analyse screening for the detection of congenital unilateral cataract in Sweden.

Methods: Data were derived from the Pediatric Cataract Register (PECARE). All children operated on before 1 year of age between January 2007 and March 2014 with an available five- year follow-up record were included.

Results: The eligible number of children was 54. Visual acuity > 0.1 was found in 35 % (19/54) of the cohort more commonly among children who underwent surgery before 6 weeks of age compared to subjects operated on between the ages of 6 weeks and 1 year (42% vs. 25%). The proportion of eyes developing visual axis opacification (VAO) and glaucoma was higher among children who underwent surgery before 6 weeks of age compared to children operated later on, (57% vs. 32% representing VAO and 43% vs. 15% regarding glaucoma). Eight patients received a bag-in-the-lens IOL (BIL-IOL). Six of the eyes, 75% (6/8), achieved VA ≥ 0.3, and no VAO was detected. The majority, 65% (35/54), were operated on before 6 weeks of age and 86% (30/35) were referred from the maternity wards.

Conclusions: Visual acuity was higher compared to earlier Swedish reports but postoperative complications still remain a problem. Even though the number of subjects with BIL-IOL was limited, a trend towards improved results in the present study was shown and future studies are needed. Maternity ward screening is effective.

OUTCOMES OF BILATERAL CATARACTS REMOVED IN INFANTS 1 TO 7 MONTHS OF AGE CONCURRENT WITH THE INFANT APHAKIA TREATMENT STUDY

Bothun ED, Plager DA, Vanderveen DK, Freedman SF, Trivedi RH, Hodge DO, Lambert SR, Traboulsi EI, Anderson JS, Weil NC, Loh AR, Morrison D, Yen KG, Wilson ME (Toddler Aphakia and Pseudophakia Study)

Introduction: This study evaluates outcomes of bilateral cataract surgery in infants ages 1 to 7 months performed by Infant Aphakia Treatment Study (IATS) investigators during IATS recruitment and compares them to IATS outcomes of unilateral cases.

Methods: Retrospective clinical study at 10 IATS sites

Results: 178 eyes (89 children) were identified with median age of 1.8 months (range 1-7) at cataract surgery. 51 (29%) eyes of 26 patients received primary intraocular lens (IOL) implantation. Of the 60 children followed between 4-6 years of age with optotype visual acuity (VA) testing, corrected visual acuity was excellent (<20/40) in 45% of better seeing eyes and 20% of worse-seeing eyes. 2% had poor acuity (>20/200) in the better eye and 10% in the worse eye. Median best eye visual acuity was 20/50 (logMAR 0.40) (p=0.84)) in both aphakic and pseudophakic children. Unplanned reoperation occurred in 29% of right eyes (including glaucoma surgery in 9%).

Discussion: Good visual outcomes were obtained in both eyes following bilateral infantile cataract surgery. With or without the inclusion of children who tested poorly due to associated neurologic disease, the VA of the worse seeing eye in these bilateral cases is better than VA in unilateral cases included in the IATS. The rates of reoperation and glaucoma are consistent with the published IATS data. Aphakia management did not affect visual acuity outcomes.

Conclusions: Visual acuity after bilateral cataract surgery in infants younger than 7 months is better than VA following unilateral cataract surgery, but adverse events were similar.

References:

1. The Infant Aphakia Treatment Study Group. Comparison of Contact Lens and Intraocular Lens Correction of Monocular Aphakia During Infancy A Randomized Clinical Trial of HOTV Optotype Acuity. JAMA Ophthalmol. 2014;132(6):676-682.
2. Plager DA, Lynn MJ, Buckley EG, Wilson ME, Lambert SR; Infant Aphakia Treatment Study Group. Complications in the first 5 years following cataract surgery in infants with and without intraocular lens implantation in the Infant Aphakia Treatment Study. Am J Ophthalmol. 2014 Nov;158(5):892-8.

USE OF EXTENDED RANGE OF VISION INTRAOCULAR LENS IN PEDIATRIC CATARACT SURGERY

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Purpose: To evaluate safety and eligibility of extended range of vision (EDOF) intraocular lens (IOL) in pediatric cataract surgery to reduce spectacle dependency after cataract operation.

Methods: Visually disturbing cataract operated from seven children (10 - 18 yrs, median 14 yrs). For four patients operation was bilateral and for three unilateral. In the operation EDOF-IOL (Tecnis Symfony) was used (range +19.5 – 26.5 D), six spheric (ZXR) and five toric IOL (ZXT).

Results: Preoperative best corrected distance visual acuity (BCDVA, Snellen values) was 0.42+0.19 (median value 0.5, N=11 eyes) and increased after the operation to 0.91+0.16 (median 0.9, N=9) at 1 month, 1.0+0.13 (1.1, N=6) at 6 months, 0.90+0.21 (0.9, N=7) at 12 months and 0.93+0.32 (1.0, N=5) at 24 months. The four patients having bilateral implantations had uncorrected binocular near visual acuity (UCNVA) 0.65+0.19 (median 0.7) at one month after the operations. After bilateral implantation one child started to wear spectacles for far two years after the operation, none of them used spectacles for reading. One child having bilateral implantation reported of glare one month after the operation, none of the other patients reported any visual disturbances.

Conclusions: EDOF-IOLs implanted safely in 10-18 years old children. Both UCDVA and binocular UCNVA were good and gave high spectacle independency with good eligibility and low number of dysphotopsias.

A CASE OF ROP WITH DEVELOPMENT OF SEVERE CENTRAL EXUDATION FOLLOWING LASER PHOTOCOAGULATION OF PERIPHERAL RETINA

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Purpose: We present the case of a premature infant born at gestional age 24+0 weeks, birthweight 640g with a picture series showing an unusual development of bilateral macular exudation after laser treatment for ROP.

Methods: The infant was followed by routine screening from GA 31 weeks and fundus photography was taken with RetCam from the time ROP was seen.

Results: The patient had persistent ductus arteriosus and severe brochopulmonal dysplasia requiring oxygen treatment. From GA 31 weeks, she underwent weekly dilated fundus examination. At GA 33 weeks she developed stage 1 ROP in the nasal zone 2 regions, and from GA 34 weeks additionally stage 1 ROP developed in zone 2 temporally in both eyes. The patient was discharged from hospital at GA 37 weeks, needing oxygen treatment at home. Gradually, from week 37/38, central plus disease developed, and at 39 weeks she underwent laser photocoagulation of the periferal retina of both eyes. 6 days thereafter, a massive central serous exudation was seen, and severe plus disease was still present. Anti-VEGF (ranibizumab) was given intravitreally in addition to local corticosteroid treatment. The exudation regressed within the two following weeks, but central pigmentary changes in the foveal region remained permanently.

Conclusions: Despite laser treatment, reactivity in the blood vessels remained and was probably the cause of the massive central exudation. In this case, treatment with intravitreal anti-VEGF had a positive effect for the resorption of the exudation.

CASE REPORT OF LOW VISION IN A 14 YEAR OLD

Astrid Meistad, Norway

Abstract not available.

NSA SESSION I: ESODEVIATIONS: CLASSIFICATION AND EXAMINATION

PREVALENCE OF ESOTROPIA IN A NORDIC POPULATION

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Purpose: To determine the prevalence of esotropia (ET), strabismus in general, and strabismus surgery among Danish adults. Furthermore, to evaluate the frequency of strabismus-associated amblyopia among individuals with strabismus in relation to current national vision screening programme.

Methods: In total, 3785 adults in the Danish Rural Eye Study (DRES) underwent an interview regarding eye health and an objective examination including visual acuity (VA), Hirschberg test and retinal photography. If VA was <0.8 a full ophthalmological examination was performed. Participants were categorized into groups based on their birth date in relation to the introduction of the national vision screening programme.

Results: The prevalence of ET was 0.8% (29/3785; 95% CI: 0.5–1.1) and of exotropia (XT) 0.3% (12/3785; 95% CI: 0.2–0.6), resulting in an ET:XT ratio of 2.7:1. In total, the prevalence of strabismus was 1.1% (41/3785; 95% CI: 0.8–1.5); no differences were found in relation to gender or screening status. A history of strabismus was present in 4.6% (174/3785; 95% CI: 4.0–5.3), and a history of strabismus surgery was present in 0.8% (32/3785; 95% CI: 0.6–1.2) of the participants. Among participants with manifest strabismus, 24% had strabismus-associated amblyopia.

Conclusion: In this population-based study of strabismus prevalence among Danish adults, ET was the most common strabismus type. Our results are the first to display a predominance of ET among Caucasian Scandinavian adults. The prevalence was related neither to screening status nor to gender.

ACUTE COMITANT ESOTROPIA. A REVIEW ON TREATMENT OPTIONS

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Purpose: To reveal the etiologic factors for acute acquired comitant esotropia (AACE) in childhood and evaluate strabismus surgery for AACE.

Methods: A literature review and a retrospective file-based analysis of children referred with AACE from May 2000 to March 2013. All children underwent complete pre- and postoperative ophthalmological and orthoptic examinations.

Results: A total of 48 children were referred and included. Seven cause-specific types of AACE were identified. Intracranial disease was present in 6%, and 4 risk factors were identified to guide clinicians when to perform brain imaging. Strabismus surgery were indicated in 50% (24/48) of the children with AACE. All children were aligned within 8Δ of orthotropia postoperatively. A high success rate of regain of stereopsis postoperatively was achieved. Previous studies on smaller case series of children support our findings and report on the usefulness of strabismus surgery, but also botulinum toxin treatment and prisms in selected cases.

Conclusion: Findings of seven cause-specific types of AACE, including intracranial disease, suggest classification of AACE of childhood. The potential for regain of stereopsis was large. Strabismus surgery is effective in the treatment of AACE and provides a high success rate of regain of stereopsis among children.

ESOTROPIA: CLINICAL PRESENTATION AND CLASSIFICATION

Karin Sandvand, Norway

Abstract not available.

THE POTENTIAL FOR BINOCULAR FUNCTION IN ESOTROPIA

Aimee Sjaastad, Norway

Abstract not available.

NSA SESSION II: ESODEVIATIONS: TREATMENT

EARLY SURGERY IN INFANTILE ESOTROPIA – WHY AND WHEN?

Lisbeth Sandfeld, MD, PhD
Eye Dept., Zealand University Hospital, Denmark

A literature review of current knowledge about the outcomes of interventions in infantile esotropia.

Infantile esotropia develops before the age of 6 months. The treatment is surgical correction with or without additional Botox injections. The timing of surgery has been discussed during the past decades, – 'early' versus 'late surgery'? And how early is 'early'? What is achievable by doing interventions on infants?

Advantages and disadvantages in early versus late intervention, including sensory-motor outcomes, DVD development, and re-operation rates will be discussed with current recommendations from the literature.

STRABISMUS SURGERY IN TOPICAL ANESTHESIA FOR SMALL ESO-DEVIATIONS

Troels Vinding MD, DMSci, Lena Boulakh MD, Helena Buch Hesgaard MD, PhD.

Orthoptic examinations: Inger Holst DBOTroels Vinding, Denmark

The use of only topical anesthesia in strabismus surgery for small eso-deviations is a possibility in patients selected for local anesthesia.

The benefit of no tissue swelling with a perfect overview of the structures allows for gentle surgery.

Per-operatively the prism cover test can be performed as no paresis of the extraocular muscles or visual impairment are introduced. The previous planed surgery can be changed and the adjustment can be performed.

Sub-tenon anesthesia can be added if necessary.

16 consecutive operations, in only topical anesthesia with a single medial rectus muscle recessed and the possibility of intra-operative adjustment, were performed. The topical anesthesia used and the per-operative pain score results will be discussed as well as the post-operative alignment of the eye axis, the stereo acuity and the high satisfaction score found among the patients.

BIFOCAL GLASSES IN CONVERGENCE EXCESS ESOTROPIA: NORDIC EXPERIENCES

Gerd Holmström, Sweden

Abstract not available.

RETROEQUATORIAL FIXATION SUTURES IN ESODEVIATIONS: NORDIC EXPERIENCES

Cecilie Bredrup, Norway and Olav Haugen, Norway

Abstract not available.

NSA SESSION III: CASES TO LEARN FROM AND FREE PAPERS

CASE REPORT: RETROBULBAR BOTULINUM TOXIN A INJECTION FOR ACQUIRED NYSTAGMUS

Helena Buch Hesgaard MD, PhD, FEBO[1,2,3] and Troels Vinding, MD, DMSci

[1] Region Västra Götaland, Sahlgrenska University Hospital, Department of Ophthalmology, Mölndal, Sweden.

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[3] Copenhagen Eye & Strabismus Clinic, Capio-CFR Hospitals, Copenhagen, Denmark

Purpose: To describe treatment of rotatory nystagmus with functionally blinding oscillopsia.

Case: A 45 years old person, developed headache, vomiting, limb management problems, that progressed acutely. She got hospitalized. A CT scan of the brain revealed Pons hemorrhage with breakthrough to 4 ventricle and subarachnoid hemorrhage. There was no neuro surgical intervention indication. She was referred to rehabilitation as wheelchair bound patient with left hemiparesis and disabling ocular deficiencies compatible with pons lesion with affection of both 6th brain nerve cores as well as both facial nerves. Due to rotatory nystagmus and oscillopsia she was functionally blind. As treatment option medical treatment with Botulinum Toxin A as retrobulbar injection was offered and injected on the left eye.

Results: On 2 weeks follow-up after treatment full ophthalmoplegia was obtained on the left eye. The patient expressed great satisfaction with the result. For the first time after the injury, television could be viewed with right eye occlusion. Control consultation was planned for further treatment. Conclusion: Retrobulbar Botulinum toxin A injection is a treatment modality for rotatory nystagmus.

CASE REPORT: ESOTROPIA AND GRAVES' ORBITOPATHY

Laura Vanhatalo
Helsinki University Hospital

A 73-year-old man was diagnosed with Graves' orbitopathy secondary to hyperthyroidism late 2014. He received oral carbimazole, intravenous cortisone infusions, orbital radiation and finally a bimedial orbital decompression surgery in February 2016 due to a progressive visual field loss. He suffered from diplopia since the summer of 2015 and wore a monoculus on his left eye. The orbital MRI scan showed that all extraocular eye muscles were affected by Graves' orbitopathy, especially medial and inferior rectus muscles. He had strabismus related torticollis and restricted ductions both in abduction and elevation which was more noticeable on the left eye. Preoperatively he could fuse with a total prism of 50 PD (prism diopter) base out to near and far distance. He did not have any remarkable torsion.

In May 2018, we operated both of his medial rectus muscles with a donor sclera elongation.[1] With the free muscle technique, the total amount of recession was 12.5 mm on the right eye and 13.5 mm on the left eye. Postoperatively he was diplopia-free for the first two weeks but eventually developed a small esodiplopia to distance. He was symptomless and happy with a total prism correction of six PD base out in his spectacles. A small abduction deficiency persisted but the vertical balance remained good.

References:

1. Thorisdottir RL et al. Clinical evidence supporting the use of donor sclera as spacer material in complicated cases of strabismus surgery - retrospective evaluation of surgical results in 117 patients with thyroid-associated ophthalmopathy or congenital strabismus. Acta Ophthalmol. 2019 Feb;97(1):74-79.

INNOVATION IN OPHTHALMOLOGY – 10 STEPS WE LEARNED FROM DURING INVENTION OF SQUINTSCOPE®

Jon Peiter Saunte, MD
Head of Strabismus Team, Rigshospitalet Glostrup, Copenhagen, Denmark

Financial Disclosure: Holds European Patent for SquintScope® Patent pending for USA, Canada, India, China, South-Korea, Japan, Australia

Introduction and Purpose: Assisting personnel for ophthalmologists have become sparse, thus eye examination of small children may be difficult. The purpose of this study describes the innovation process of a device to allow for more successful eye examination in 2-6 year old children.

Methods: 10 steps in the innovation process:

1. Define problem
2. Create prototypes
3. What can and can't be shared with colleagues
4. Why team up with professionals from different specialities?
5. Patent application – pros and cons
6. Protecting domains
7. Lawyers: doctor's friend?
8. Funding: why and when?
9. Clinical testing: evaluate impact and utilization
10. Production, marketing and sales

Results:

1. Uncooperative children under 6 years of age may be difficult to examine for ophthalmologists.
2. 22 generations of 3D printed prototypes were designed with functional and aesthetic modifications performed
3. Contradiction: protection of intellectual properties versus valuable feedback from colleagues
4. A multidisciplinary team with combined competences provides synergy.
5. Patents protect intellectual property, and… may ruin you. Calculations to consider.
6. Registration®, TradeMarkTM, CE and FDA - important steps.
7. Lawyers: expensive friends you can't afford to miss.
8. Funding options for doctors, project stage is important for funding type

9. Clinical testing of SquintScope® revealed two new applications for use and saved children from eye examination under general anaesthesia.
10. 3D printing versus injection moulding. Lessons learnt from designers, engineers, and marketing companies.

Conclusions: Defining a clinical problem and working to find solutions may be challenging, surprising and hard work – but also fun! Utilization of the 3D printed SquintScope® demonstrated more successful eye examination in children aged 2-6 years.

References: Mads Faurholt og Lars Tvede: (Danish) Iværksætter – hvad vi lærte af at starte 30 virksomheder. 2017 Gyldendal Business, pp. 1---347

PROSPECTIVE OBSERVATIONAL STUDY OF DIVERGENCE INSUFFICIENCY TYPE ESOTROPIA IN ADULTS

Jonathan M. Holmes
Mayo Clinic, Rochester, MN, USA

Purpose: To report treatment outcomes for divergence insufficiency distance esotropia in adults.

Methods: In a prospective non-randomized multicenter observational study, adults with divergence insufficiency esotropia were enrolled when starting a new treatment (either prism, orthoptic exercises, or surgery). Divergence insufficiency esotropia was defined as distance esodeviation of 2-30 prism diopters, at least 1.25 times larger than near esodeviation, by prism and alternate cover test, and diplopia with frequency of "sometimes", "often" or "always" straight ahead in the distance by diplopia questionnaire. The primary outcome was "symptom success" at 12 months, defined as diplopia "rarely" or "never" straight ahead in the distance.

Results: One-hundred-fourteen participants were enrolled and initiated treatment: surgery (n=76, 67%), prism (n=34, 30%), or exercises (n=4, 4%). Prior treatment was reported mostly in the surgery group, using previous prism (n=61, 80%). Success criteria were met for 55 (89%, 95% CI=78% to 95%) with surgery and 17 (65%, 95% CI=44% to 83%) with prism. Success rates were high for both major types of surgery; bilateral medial rectus recession, 32 (91%) of 35 and bilateral lateral rectus resection 10 (91%) of 11.

Conclusions: Although success rates should not be compared directly in this non-randomized study, because there were important differences in baseline characteristics, success following surgery or prism was common. These data may be useful for counseling patients and for future randomized clinical trials.

CASE REPORT: RECURRENT ESOTROPIA

Aimée Sjaastad
Oslo University Hospital

Abstract not available.

CASE REPORT: NYSTAGMUS

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Abstract not available.

POSTER ABSTRACTS

THE PEDIATRIC CATARACT REGISTER (PECARE): OUTCOME AT FIVE YEARS OF AGE FOLLOWING EARLY SURGERY FOR DENSE UNILATERAL CONGENITAL CATARACTS

Gunilla Magnusson[1], Lovisa Borg[1], Birgitte Haargaard[2], Maria Kugelberg[3], Alf Nyström[1], Annika Rosensvärd[3] and Kristina Tornqvist[4]

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[4] Department of Clinical Sciences, Ophthalmology, Lund University, Lund, Sweden

Purpose: To investigate the development of visual acuity (VA) and postoperative complications in a national register-based cohort study and compare results with earlier Swedish retrospective reports. Furthermore, to analyse screening for the detection of congenital unilateral cataract in Sweden.

Methods: Data were derived from the Pediatric Cataract Register (PECARE). All children operated on before 1 year of age between January 2007 and March 2014 with an available five- year follow-up record were included.

Results: The eligible number of children was 54. Visual acuity > 0.1 was found in 35 % (19/54) of the cohort more commonly among children who underwent surgery before 6 weeks of age compared to subjects operated on between the ages of 6 weeks and 1 year (42% vs. 25%). The proportion of eyes developing visual axis opacification (VAO) and glaucoma was higher among children who underwent surgery before 6 weeks of age compared to children operated later on, (57% vs. 32% representing VAO and 43% vs. 15% regarding glaucoma). Eight patients received a bag-in-the-lens IOL (BIL-IOL). Six of the eyes, 75% (6/8), achieved VA ≥ 0.3, and no VAO was detected. The majority, 65% (35/54), were operated on before 6 weeks of age and 86% (30/35) were referred from the maternity wards.

Conclusions: Visual acuity was higher compared to earlier Swedish reports but postoperative complications still remain a problem. Even though the number of subjects with BIL-IOL was limited, a trend towards improved results in the present study was shown and future studies are needed. Maternity ward screening is effective.

STRABISMUS SURGERY ON A PATIENT WITH CUTIS LAXA: A CASE REPORT

S Lähdeoja[1], L Lindberg[1]

[1] Department of Ophthalmology, Helsinki University Hospital, Finland

Purpose: Cutis laxa is a rare disorder of connective tissue. Typically, the skin is loose, inelastic and wrinkled giving the appearance of prematurely aged person, and the joints are hypermobile. There is only little data on strabismus surgery on patients with cutis laxa1.

Methods: A 6-year old girl with ATP6V0A2-related cutis laxa, who had congenital esotropia, bilateral inferior oblique (IO) overaction and amblyopia, was referred for strabismus surgery. Preoperatively she presented with alternating esotropia of 75 prism diopters and +4 IO overaction together with a face turn due to cross-fixation. In addition, there was a small abduction defect and nystagmus. She was moderately hyperopic with symmetrical amblyopia of both eyes (visual acuity 0.3-0.4). Her face was narrow, the palpebral apertures were downslanting and she had a small lagophthalmos. On MRI scan both inferior recti were located more medially than normally, most likely due to excyclorotation of the bulbi.

Both tight medial recti were recessed 6mm. IOs were recessed 10mm and anteponated up to 12mm from the limbus. Care was taken not to overcorrect horizontal strabismus because of her narrow facial features. During the same general anesthesia, a pediatric ophthalmologist performed lateral tarsal strip operations to correct the downslanting lid apertures.

Results: Postoperative healing occurred normally. Two months postoperatively the cosmesis was significantly improved, although a small, variable esotropia and hypertropia remained. One year after the operation, good result had remained with moderate inferior oblique overaction at far-gaze and varying amount of esotropia and hypertropia at near. Visual acuity was 0.5 and a slight face-turn remained. The lid aperture had improved, although some problems with trichiasis continued.

Conclusions: Strabismus surgery may help improve the appearance and visual functioning of those patients with cutis laxa. It is important to attain preoperative MRI scans and take the typical facial features into account when planning the surgery. Oculoplastic interventions may also be needed.

References: Tas A, Gundogan FC, Kocaturk T, Altun S, Bayram Y. Oculoplastic approach to congenital cutis laxa syndrome. Aesthetic Plast Surg. 2013 Apr;37(2):417-20. Epub 2013 Feb 27.

OUTCOME OF THE PRIMARY HEALTH CARE OPHTHALMOLOGICAL SCREENING FOR CHILDREN AGED 18 MONTHS OLD OR YOUNGER

Linda Andersson
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Purpose: In Finland, preschool children undergo ophthalmological screening as part of the primary health care screening and counseling services. Purpose of this study is to evaluate outcome of the ophthalmological screening in the age-group of 18 months or younger.

Methods: We included all children aged ≤ 18 months referred during a 6 months period in 2012 from the primary health care to the Pediatric Ophthalmology Service of the Helsinki University Hospital. Ophthalmological data was retrospectively retrieved from patient records.

Results: Birth rate in our hospital district is 18 000 children in a year and all undergo the screen. There were 414 referrals in total. About half of the referrals were due to strabismus, only 5% of these had constant esotropia, 10 % had intermittent exotropia and over one third of these patients had findings consistent with age. 110 patients were referred due to blocked tear duct.

Only few vision threatening conditions were found in this cohort. One congenital cataract was found due to red reflex abnormality and one optic atrophy due to nystagmus. Two Horner syndromes were found due to anisocoria.

Conclusions: Based on currently analyzed results it seems that screening of strabismus at this age is difficult. The screen could be improved with additional education on this subject. The number of caught vision threatening conditions were low. It seems that in this age group there is no need for ophthalmological screen of current scale.

REAL-WORLD DATA ON RESPONSE TO THERAPY WITH IDEBENONE IN PEDIATRIC LEBER'S HEREDITARY OPTIC NEUROPATHY (LHON)

Marilena Cojocaru[1], Magda Silva[2], Xavier Llòria[2], Claudia Catarino[3], Bettina von Livonius[4], Claudia Priglinger[4], Thomas Klopstock[3]

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Purpose: LHON is a rare mitochondrial genetic disorder that results in severe, bilateral central vision loss. The Expanded Access Program (EAP), a named patient program , provides insights into the effectiveness of idebenone in pediatric (<12 years old) patients in a real-world setting.

Methods: A retrospective medical chart analysis of visual acuity (VA), expressed as logMAR was performed. VA efficacy was determined as a clinically relevant recovery (CRR), defined as an improvement from off-chart to reading one line on the ETDRS chart, or an on-chart improvement of two lines.

Results: Five patients were below 12 years of age at baseline (BL), with a time since LHON onset of 1.7 months to 5 years. The median best VA at BL was 0.94 logMAR (range 0.16 – 1.20). Following a median treatment duration of 33.6 months (range 6.8 – 40), median best VA at last visit (LV) was 0.08 logMAR (-0.18 – 1.36). Three patients achieved CRR from nadir (and BL) in both eyes, with a magnitude of recovery from 2 – 9 lines at first observation of CRR, which increased to 4 – 12 lines by LV. Idebenone was well tolerated, and there were no reports of serious adverse events related to the drug.

Conclusions: Idebenone had a favourable safety profile and led to clinically relevant recovery in LHON pediatric patients, consistent with results in adult population. Though limited by the low number of pediatric patients in this Expanded Access Program, the results suggest that further research is warranted to investigate the potential of idebenone in this population.

REAL-WORLD DATA ON RESPONSE TO THERAPY WITH IDEBENONE IN PEDIATRIC LEBER'S HEREDITARY OPTIC NEUROPATHY (LHON)

Marilena Cojocaru[1], Magda Silva[2], Xavier Llòria[2], Claudia Catarino[3], Bettina von Livonius[4], Claudia Priglinger[4], Thomas Klopstock[3]

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