[1] GOOD SOCIAL FUNCTION IN A CHILD WITH SEVERE VISUAL IMPAIRMENT CAUSED BY CHORIORETINAL COLOBOMA

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At the Eye Department, we followed a child born with chorioretinal colobomas in both eyes. At the first consult it was assumed that he was blind. We followed him and he was helped by vision consultants. During the 7 years his vision improved to 0,08 measured using a snellen board.

At the last visit he appeared as a socially and intellectually well-functioning child. He went to normal school, and in his spare time he was able to ride his bike and swim.

This is an example illustrating how difficult it can be to predict the visual and social function of the new born with poor vision.

[2] OCULAR HYPERTENSION AND GLAUCOMA IN UVEITIS ASSOCIATED WITH JUVENILE IDIOPATHIC ARTHRITIS

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Background

Ocular hypertension and secondary glaucoma is a frequent complication of uveitis associated with Juvenile Idiopathic Arthritis (JIA). The aim of this study was to evaluate the prevalence, course and treatment of ocular hypertension (OHT) and secondary glaucoma in a Danish population of children with JIA-associated uveitis.

Methods

Medical records of consecutive children with JIA-associated uveitis under treatment at Aarhus University Hospital (AUH) in the period 2001 to 2014 were reviewed retrospectively.

Results

26 patients (50 eyes) were included in the study. Age at onset of uveitis was 1 to 13 years. Follow-up was 8.5 year (0.5-21). 58% of the children (15 patients, 25 eyes) developed increased IOP, which was steroid-induced in 80% (12 patients). In 11 patients (20 eyes) IOP was normalized after reduction or discontinuation of treatment with steroid, which in 6 patients involved starting systemic steroid sparing immunotherapy. 19% of the children
(5 patients, 6 eyes) developed glaucoma, which was steroid-induced in 2 eyes. Both eyes had severe glaucoma damage and one eye was buphthalmic and blind due to glaucoma. 4 patients (5 eyes) needed glaucoma surgery.

Conclusion

OHT and secondary glaucoma is a frequent complication in uveitis associated with Juvenile Idiopathic Arthritis. Treatment with steroid seems to be the main cause of IOP elevation and development of severe glaucoma and justify a low threshold for starting steroid sparing systemic immunotherapy in children with steroid-induced OHT.

[3] POTENTIAL EFFECT OF TUMOR NECROSIS FACTOR INHIBITORS ON TRABECULECTOMY WITH MITOMYCIN C FOR PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS -RELATED UVEITIC GLAUCOMA: A RETROSPECTIVE ANALYSIS

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Purpose

The majority of juvenile idiopathic arthritis -associated uveitic glaucoma patients require surgery to control intraocular pressure. Trabeculectomy with mitomycin C is a major treatment option. Chronic inflammation and young age are risk factors for failure. Factors that potentially protect from filtration failure are important to identify.

Methods

In this retrospective study were included 29 eyes of 29 consecutive patients (age, 3.1 to 20.4 years) who had uveitic glaucoma associated with juvenile idiopathic arthritis diagnosed when 16 years of age or younger and who underwent their mitomycin C-augmented primary trabeculectomy between April 1996 and January 2014. Fifteen patients were on systemic TNF inhibitors at the time of trabeculectomy to control their uveitis, arthritis or both. Fourteen patients did not receive TNF-inhibition since their uveitis and arthritis were quiet. No changes were made in their anti-rheumatic treatment preoperatively.

Results

The survival rate of trabeculectomies in patients with TNF-alpha inhibition was 73% at 1, 5 and 10 years postoperatively as compared to 57%, 16% and 0%, respectively, in those without TNF-alpha inhibition (*P*=0.004) by median 7.9 years of follow-up. The effect was observed especially in eyes without prior ocular surgeries. No other explanatory factors were found.

Conclusions

Our data suggest that juvenile idiopathic arthritis -uveitis patients may benefit from systemic TNF-inhibitor treatment at the time of MMC-augmented primary trabeculectomy.

[4] CHILDHOOD GLAUCOMA IN ASSOCIATION WITH MUTATIONS IN THE LTPB2 GENE

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Purpose

Homozygous mutations in the *LTBP2* gene associate with megalocornea, microspherophakia, lens dislocation, secondary glaucoma and rarely with primary congenital glaucoma. We present phenotype and management of glaucoma in 2 children carrying homozygous mutations in the *LTBP2* gene.

Methods

The diagnostic criteria for glaucoma were: intraocular pressure (IOP) above normal for age, enlargement of corneal diameter, axial elongation and progressive optic disc cupping. Both patients underwent lensectomy, vitrectomy and drainage tube implantation.

Results

Both patients presented with bilateral congenital megalocornea, microspherophakia and long axial lengths under normal IOP for age. In Patient 1, glaucoma of the left eye was diagnosed at the age of 3 years prior to surgery for lens dislocation and required multiple drainage tube surgeries. Glaucoma of the right eye was diagnosed at the age of 8 years and stabilized with medication. Patient 2 developed glaucoma at the age of 15 months prior to lens subluxations. He underwent a combined lens surgery and Molteno-3 tube implantation for both eyes at the age of 22 and 32 months. In all four eyes, anterior chamber angles were open without evident abnormalities. There was no sign of pupillary block or other lens-related mechanisms for glaucoma.

Conclusions

Diagnosis of glaucoma in association with the mutations in the *LTBP2* gene is challenging due to congenital megalocornea and axial elongation under normal IOP. Glaucoma is poorly controlled with medication and surgical treatment is complicated by lens dislocation.

NB

LTBP2-geenin mutaatio, josta käytetään merkintätapaa c.895C>T (p. Arg299Ter)

AC-OCT available, ES

Genetic test report ordered

[5] PULSATING ENOPHTHALMOS IN A CHILD WITH NEUROFIBROMATOSIS TYPE 1

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A 9-year-old boy with neurofibromatosis type 1/von Recklinghausen disease (NF1) visited the Department of Ophthalmology, Odense University Hospital, Denmark for a routine checkup.

The patient was predisposed to the disease as his mother is diagnosed, and also his sister is known to have NF1. During previous pediatric examinations multiple café-au-lait spots and a single plexiform neurofibroma have been identified. A child psychiatrist has found the patient to be slightly mentally retarded and to have a developmental disorder of speech and language. An MRI was performed 5 years earlier and showed no signs of abnormalities. Previous ophthalmological examinations were without signs of NF1.

However, this most recent routine ophthalmological examination revealed discrete left-sided pulsating enophthalmos, which became more evident when the patient was looking at something close up. The patient himself had not noticed this, and he wasn't bothered by diplopia. The motility of the eye was normal, but the eye slit a little smaller. CT of the head including orbits and facial bones was ordered and showed significant dysplasia of the sphenoid bone. The posterior part of the left orbit and parts of the orbital ceiling and lateral wall were lacking.

Pulsating enophthalmos in patients with NF1 is well described in the literature, but only a very few cases have ever been reported.

The case will be presented with video documentation of the pulsating enophthalmos.