

## **Pediatric Ophthalmology**

### **E-00039**

Retinitis pigmentosa diagnosed in polyglandular autoimmune syndrome type-1

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#### **ABSTRACT (AS SUBMITTED)**

**Purpose:** To report retinitis pigmentosa as a rare ophthalmologic manifestation of the polyglandular autoimmune syndrome type-1.

**Methods:** We present a 9 year-old girl with complaints of nyctalopia seen by our pediatric ophthalmology service. Ophthalmologic evaluation, visual fields and electroretinogram (ERG) were performed.

**Results:** Dilated fundus showed alterations of retinal pigmentary epithelium and retinal arterioles of a diminished diameter. Goldmann perimetry revealed constricted visual fields. ERG had a non-recordable rod function while cone responses were diminished. These findings lead to the diagnosis of retinitis pigmentosa.

**Conclusions:** These results suggest that retinitis pigmentosa could be a manifestation of the polyglandular autoimmune syndrome type-1.

## **Pediatric Ophthalmology**

### **E-00040**

Ultrasound biomicroscopy in the management of retinoblastoma

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#### **ABSTRACT (AS SUBMITTED)**

**Purpose:** To define the characteristics and the role of ultrasound biomicroscopy (UBM) in the management of children affected with retinoblastoma.

**Methods:** A retrospective case series included patients for whom ultrasound biomicroscopy was used in retinoblastoma management at Hospital for Sick Children. The UBM characteristics of retinoblastoma and the clinical situations in which it was most useful were defined.

**Results:** Seventy eight patients (106 eyes) affected with retinoblastoma were studied using UBM to rule out anterior segment/ciliary region involvement. In thirty-nine patients, anterior involvement was observed (44 eyes). Of those, 25 eyes were IIRC Group E, 18 Group D and only one Group C. The types of anterior segment involvement included neovascular glaucoma in 23 patients and cataract due to tumour contact to the lens in 9 patients. In addition, nine other patients showed an obstructed view due to an extensive retinal detachment. In 11 additional eyes with extensive vitreal seeding, anterior segment/ciliary region involvement was excluded using UBM examination which allowed eye preservation.

**Conclusions:** The UBM provided a sensitive and reproducible visualization of the anterior retina and anterior segment allowing a better classification of eye pathology. The assessment of true disease severity led to optimized management of patients affected with severe retinoblastoma.

## **Pediatric Ophthalmology**

### **E-00058**

Management of stromal iris cysts in children

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#### **ABSTRACT (AS SUBMITTED)**

**Purpose:** Stromal iris cysts in children are very rare and can be complicated by endothelial touch, uveitis, glaucoma and decreased vision from obstruction of the visual axis and/or amblyopia. Many surgical techniques have been described in treating iris cysts with the aim of minimizing recurrence. We discuss our experience with stromal iris cysts, describing their features and suggested management.

**Methods:** We conducted a retrospective chart review of children with stromal iris cysts presenting to one surgeon (AA) at our hospital with a review of literature.

**Results:** Four children with stromal iris cysts were reviewed, age ranging from 25 days to 12 years. All children presented with a unilateral solitary stromal iris cyst. Three girls had primary iris cysts (two were congenital) and one boy had a secondary cyst following open globe repair. All four had decreased vision and photophobia on presentation and all had cyst-endothelial touch. Uveitis and cataract were found in the child with traumatic cyst. No child had glaucoma at presentation or in follow-up. Rapid cyst growth (over weeks) was seen in the congenital cases. Two children were treated with en-bloc resection of the iris cyst and two underwent aspiration and removal of the roof of the cyst. Follow-up ranged from two to seventeen months. Complications include one recurrence at two months which required adjuvant diode endolaser treatment and one visually insignificant cataract. The endothelial cell count measured postoperatively in the eldest child was decreased compared to the fellow eye but has stabilized at one year. One child continues to have subnormal vision related to deprivation amblyopia.

**Conclusions:** Photophobia associated with paediatric stromal iris cysts can indicate endothelial compromise and warrants early intervention. Endolaser photocoagulation of the cyst wall or en-bloc surgical excision with microsurgical instruments may reduce the possibility of recurrence. In children, we suggest early surgical intervention if an iris cyst is causing visual loss or corneal endothelial touch.