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A Unilateral Myopic Shift Secondary to Sturge-Weber Syndrome Results in Refractive Amblyopia of the Fellow Eye in a Pediatric Patient

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Case Details

- 4-year-old female
- Hx secondary glaucoma OS. S/p trabeculotomy OS at 2 months of age. Full time glasses wear since 11 months of age
- Medical history: Sturge-Weber syndrome
- Current medications: Cosopt BID OS, Xalatan QHS OS, clonazepam, cetirizine

Pertinent Findings

- External: Port wine stain left side of face
- Cycloplegic retinoscopy
  - **+4.75 +3.00 x085 VA 20/70 OD**
  - **-0.25+3.00 x100 VA 20/25 OS**
- Alignment: Ortho
- IOP 21/23 iCare
- DFE: C/D 0.3 OD, 0.5 OS, ONH pink and distinct OU, macula WNL (+) FLR OU, periphery normal OU

Discussion

It is well documented that a glaucomatous high IOP causes an expansion of the globe due to excessive plasticity of collagen fibers in pediatric eyes[1,2]. The elongated axial length induces myopic shifts[1,2]. In this patient, the myopic shift in her glaucomatous eye resulted in a unilateral high hyperopic refractive error in the fellow eye with normal ocular health. This anisometropia is enough to cause amblyopia in the hyperopic eye.

In past appointments, the patient demonstrated equal visual fixation with the 10BD prism test. Since equal fixation was previously found, no amblyopia therapy was initiated. Now that the patient can verbalize optotype acuity, a moderate amount of amblyopia was found warranting additional treatment.

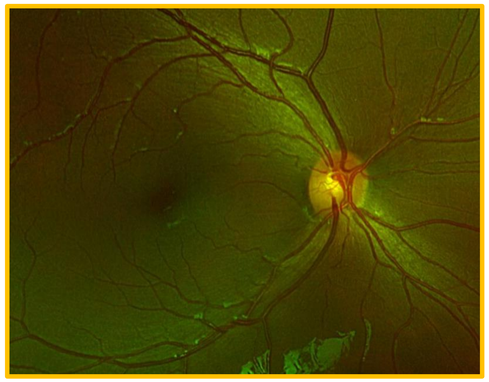

Figure 1: Optos image right eye

Figure 2: Optos image left eye

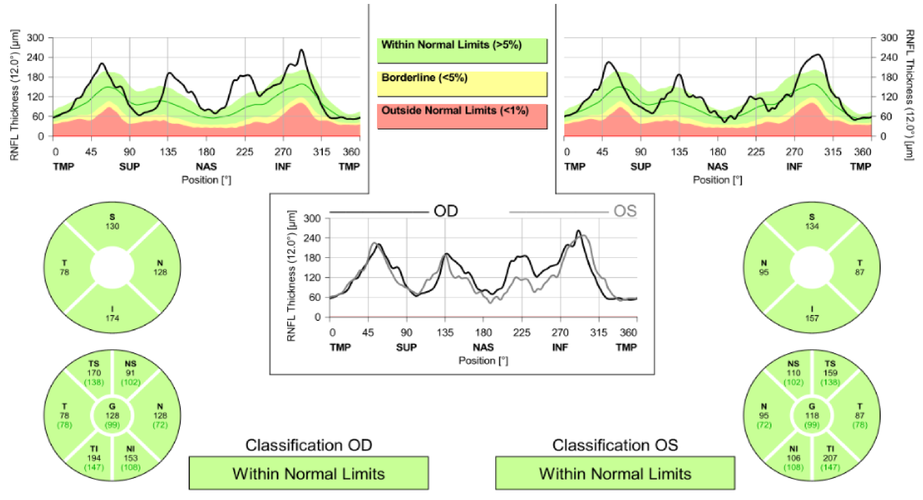

Figure 3: Spectralis OCT Imaging of both eyes. Mean RNFL: 128um OD, 118um OS

Treatment/Management

The mainstay treatment for moderate unilateral amblyopia is patching therapy or atropine penalization[3]. As the patient has already been in the correct optical correction since 11 months of age, it can be assumed she has reached her maximum vision improvement from spectacles alone[4]. The glasses prescription has been monitored closely since the initial visit and has remained stable. Based on the patient's refractive error, depth of amblyopia, and family's preferences, patching the left eye 2 hours a day was initiated.

The patient's glaucoma OS is well controlled with topical treatment s/p surgical intervention. The IOP and optic nerve health remain stable. Monitoring of her glaucoma will be done with serial IOP, OCT-nerve and dilated fundus exams. Once the patient is old enough, a baseline visual field will be attempted.

The current treatment plan is to monitor the patient in 2-3 months for VA and IOP check.

Take Home Message

In this case, the eye without the initial pathology developed an amblyogenic reduction in visual acuity. With the lifelong risk of glaucomatous vision loss, it is important to maximize visual potential in the healthy eye of our patient while they are still in their vision development period. In the setting of ocular pathology in pediatric patients, it is important to understand the secondary impacts that could occur during the vision development period.

References

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(4) Cotter S, Edwards A, Wallace D, et al. Treatment of Anisometropic Amblyopia in Children with Refractive Correction. Ophthalmology. 2006 June; 113(6): 895–903