

INTRODUCTION

Symmetrical bilateral decreased visual acuity in a pediatric patient is not always amblyopia. In the absence of amblyogenic factors, other differential diagnoses must be considered. Among the differentials are: ametropia, cortical visual impairment, infantile/juvenile cataracts, optic nerve hypoplasia, and retinal disorders such as Retinitis Pigmentosa or Stargardt. Only when all other factors have been ruled out, does the term malingering come into play.

CASE SUMMARY

An 11-year-old male with bilaterally reduced visual acuity presented for a vision therapy evaluation to ruleout amblyopia as the underlying cause. He has a history of being a poor responder and suspected malingerer at previous examinations. He reported increasing blurred vision since he was 6-7 years old. He also reported trouble seeing in the dark. According to his father, the family history was unremarkable for any vision loss. He was in the 5th grade but on a 1st grade reading level; he also reported that he did not like to read. His medical history is positive for asthma.

Table 1. Initial Examination Findings

		Findings
-	Best Corrected Visual Acuity	20/40+ OD 20/40+ OS
	Confrontation Visual Fields	Full to Finger Counting
	Manifest Refraction	OD: +1.75-1.75x178 OS: +2.00-2.75x170
	Cycloplegic Refraction	OD: +3.75-2.00x180 OS: +4.25-2.00x170
	Slit Lamp Exam	Syneresis inferiorly with brown pigmented flecks in the anterior vitreous OU
	Dilated Fundus Exam	Circumferential ring of flecks in the para-macular area OU, (-) foveal reflex OU
	Optos Testing with AF	Bilateral macular hyperfluorescence with a hypofluorescent para-macular ring, po scattered areas of hypofluorescnce 360 OU (Fig. 1)
	SD-OCT	Bilateral macular cysts (Fig. 2)

Upon follow-up examination in the retina clinic, the patients BCVA had decreased to 20/50 OD/OS. All other findings were stable. A diagnosis of Retinitis Pigmentosa was determined. The patient was immediately started on Neptazane 25mg PO BID to eliminate the macular cysts. The patient returned 3 weeks later for follow-up, at which time a multi-focal ERG was performed (Fig 3). The ERG demonstrated responses that are reduced by 60% under scotopic conditions and reduced by 50% under photopic conditions. These findings confirmed the diagnosis of RP. At his next follow-up, the patients BCVA was unchanged; he was not responsive to the Neptazane and we are awaiting pediatric approval to administer a sub-tenons injection of Kenalog.

Table 2. Causes of Bilateral Decreased BCVA in a Pediatric Patient

Differentials	Associated Clinical Findings		
Malingering ³	 Minimal refractive error on retinoscopy Tubular or ice cream cone defects on manual visual field te Cloverleaf pattern on automated testing 		
Ametropia	- Hyperopia, Myopia, Astigmatism		
Amblyopia	 Expected VA: 20/30-20/70 Refractive amblyogenic factor Deprivational amblyopia 		
Cortical Visual Impairment ⁵	 Variable BCVA to complete blindness, vision can vary from Peripheral vision more effective than central vision. Photophobia vs. compulsive light gazers Normal color vision Decreased depth perception Motion of target or of the patient improves vision 		
Infantile Cataracts ⁵	 Variable BCVA due to severity and type of cataract Leukocoria/Absent Red reflex Nystagmus Visual Inattentiveness Strabismus Photophobia 		
Optic Atrophy	 Pallor of optic nerve heads Nystagmus Underlying systemic condition Intracranial mass (+) RADP Abnormal color vision 		
Autosomal Recessive Retinitis Pigmentosa ¹	 Onset around 9yo, range from 3 to 18yo Decreased night vision Peripheral vision loss DFE: arteriolar attenuation, bone spicule pigmentation, pal Abnormal color vision (usually later in progression of diseased of the second strength o		

11-year-old Patient with Retinitis Pigmentosa Mistaken for a Malingerer: Lessons Learned

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osterior pole/mid-peripheral







Figure 1a/b. Red/green Optos photos Figure 1c/d. Optos photos with autofluorescence



Figure 2a/b. Horizontal SD-OCT 5-Line Raster of the macula Figure 2c/d. Vertical SD-OCT 5-Line Raster of the macula

Figure 3: ERG results. ERG scotopic response 47 uv OD; flat OS Rod/cone mixed OD 107 uv, OS 123 uv. Photopic response OD= 48uv at 40 msecdelayed; OS 64 uv at 43 msec. Flicker: OD only (OS electrode kept falling out)= 36.6 uv and delayed (37 msec)









DISCUSSION

Bilaterally decreased visual acuity is a finding with, more often than not, common and treatable underlying conditions. There are many differentials that must be ruled out. The most easily diagnosed is uncorrected ametropia which can be corrected with spectacle correction. In any case of reduced visual acuity, a careful differential diagnostic approach is necessary to determine if the cause of not organic.

Procedures	Potentia	
Distance and Near Visual Acuity	- Dista	
Pinhole Acuity	- Impr - Rules	
Whole Line vs. Single Letter Acuity	- Improv	
Amsler Grid	- Disto	
Pupil Evaluation	- Asym - Norn	
Dilated Fundus Examination	- Will re	
Color Vision	- Abno - Abno	

** If this testing does not yield reliable results, further testing with OCT, fundus photography with auto-fluorescence, VEP and ERG should be performed

If all of the diagnostic testing has been correctly performed and the best corrected vision is still decreased, then the diagnosis of malingering and amblyopia can be considered.

- desire for glasses.
 - responses:

 - better

 - alleviate the problem
- period of development ⁷
 - as:

	Isometropia	Anisometropia
Hyperopia	+8.00	+3.00
Myopia	-5.00	-1.00
Astigmatism	2.50	1.50

CLINICAL PEARLS

REFERENCES

- Grove Village, Illinois.
- . Erickson GB, Griffin, JR, Kurihara, JI. Streff syndrome: A literature review. J Opt Vis Dev 1994;25:64-69.

- 6. AOA Optometric Clinical Practice Guidelines: Care of the Patient with Amblyopia. 2014. 7. Lecture on Amblyopia by Jeffrey Cooper OD, at SUNY College of Optometry, Fall 2016

Table 3. Differential Diagnostic Testing

al Results

ance and near acuities would be inconsistent in cases of hysterical amblyopia and malingering

rovement suggests a diagnosis of uncorrected ametropia

s out underlying disease

ved acuity with single letters is common in patients with amblyopia

ortions on grid can indicate macular disease

nmetric pupil evaluation, such as an RAPD, can indicate an optic nerve dysfunction mal findings can help rule out a unilateral malingerer

eveal any organic causes involving the retina as well as an assessment of the optic nerve and macula

ormal in patients with rod-cone dystrophies

ormal in patients with unilateral optic nerve anomalies

• Malingering is defined as an intentional misleading of symptoms or subjective responses ⁴. Usually secondary gain is involved such as obtaining time away form work, financial gain or securing disability benefits. In the case of pediatric malingerers, the goal is not monetary, but usually associated with the

• Optometric tests and procedures can be performed to fool a malingerer into giving reliable

• Trial frame with +/-0.12 or 0.25 lens – indicate to the patient that this is the new refraction, ideally the child will not know any better and will most likely read the 20/20 line. • OKN Drum at 2-3 feet – patients eyes that follow the grating will have a VA of 20/200 or

• Conversing with the patient – getting to the root cause of the child's response can help

• Amblyopia is defined as a reduction in visual acuity due to abnormal visual experience during the critical

• Although amblyopia is more commonly a unilateral phenomenon, there are bilateral causes such

• Large refractive error – tend to have high hyperopic prescriptions, usually above 6Ds • Corneal and lenticular opacities

• Decreased vision can be attributed to blur caused by loss of foveal form vision.

• Most importantly, patients diagnosed with amblyopia must have had an amblyogenic factor present during the critical period.

Table 4. Refractive Amblyogenic Factors ⁶

• Ambylopia cannot be diagnosed with a patient that does not have an amblyogenic factor

• You must be sure that your patient is not malingering; children can sometimes be difficult, especially when he/she wants glasses more than anything and is willing to tell you just about anything to get them. Children can be misled with the use of low plus/minus lenses into thinking they are being corrected.

Be sure of a child's past medical and surgical history.

BEWARE of prior cataract surgery, hereditary conditions, metabolic/systemic diseases.

• ALWAYS dilate your patients if you cannot determine a non-pathological cause of the decreased vision

Alexander LJ (2002), Primary Care of the Posterior Segment: third edition. McGraw-Hill, New York. 2. Granet DB, Robbins SL, Baber LJ (2013), Challenging Cases in Pediatric Ophthalmology. American Academy of Pediatrics, Elk

4. Incesu AL. Tests for malingering in ophthalmology. Int J Ophthalmol 2013;6(5):708-717

5. The Wills Eye Manual: Office and Emergency Room Diagnosis and Treatment of Eye Disease. Philadelphia: Lippincott, 1994.