

Article ▶ Successful Treatment of Severe Amblyopia

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ABSTRACT

Background: Amblyopia, with an estimated prevalence in the United States between 1% and 3%, is one of the leading causes of blindness in patients below the age of 70. Amblyopia is a reduction of best-corrected visual acuity that cannot be attributed to a structural abnormality of the eye or to visual pathways and that occurs as a result of refractive error, strabismus, or deprivation secondary to a visually significant ptosis, cataract, etc.

Case Report: This case report explores the use of full-time occlusion as a treatment for severe anisometropic amblyopia in a three-year-old male with high myopia secondary to Stickler Syndrome. Maximum occlusion led to quick and successful resolution of the patient's amblyopia.

Conclusion: The Pediatric Eye Disease Investigator Group (PEDIG) has performed many clinical studies that have shaped current amblyopia treatment. Full-time occlusion is sometimes viewed as an archaic treatment and is often not the first line of treatment for amblyopia for many practitioners. However, as in this case report, full-time occlusion can lead to quick and effective resolution of amblyopia. There are many advantages to full-time occlusion, including faster resolution of amblyopia, more successful treatment of amblyopia, and consistency in treatment.

Keywords: amblyopia, anisometropia, full-time occlusion, myopia, Stickler Syndrome

Introduction

Amblyopia is a reduction of best-corrected visual acuity that cannot be attributed to a structural abnormality of the eye or the visual pathways. With an estimated prevalence between 1% and 3%, amblyopia is one of the leading causes of blindness in the United States in patients below the age of 70.¹ Amblyopia may occur as a result of refractive error, strabismus, or deprivation secondary to a visually significant condition, such as ptosis or cataract.²

Visual maturation occurs predominantly during the first three years of life but continues until approximately the age of eight.¹ Any insult to the visual system during this time period may cause amblyopia. In addition, the treatment of amblyopia may be most successful in patients under the age of eight due to the neuroplasticity of the young visual system. Due to reports of improvement in amblyopia in older patients, it is presumed that some visual plasticity remains throughout life.³ However, the success rate of amblyopia treatment is much lower in patients nine years and older.⁴⁻⁶ This can be explained by the more vulnerable visual system (higher degree of neuroplasticity) seen in younger patients. For example, most patients nine and older will not develop amblyopia under the same conditions that will cause amblyopia in a two-year-old.

Current treatment modalities for amblyopia include patching therapy, Bangerter filters, atropine, and most recently iPad games with red/green glasses. Although there is a continuous pursuit for newer treatment modalities, these modalities have not been found to be effective for severe amblyopia when compared to patching therapy. Based on prospective randomized studies performed by the Pediatric Eye Disease Investigator Group, when prescribing patching for severe amblyopia, practitioners have a presumed choice of six

hours or full-time occlusion. Many choose six hours due to the perceived lesser burden on the family and patient. However, the perceived burden has been shown to be equal with full-time and six hours of occlusion.⁷ In addition, many families, when educated of their choice between six hours and full-time occlusion, will choose full-time occlusion.⁸ This case report explores the successful treatment of severe anisometropic amblyopia with full-time occlusion.

Case Report

Patient JH, a 3-year-, 0-month-old male, reported to clinic due to parental concern for vision because he held objects very close to his face. JH's medical history was significant for Stickler Syndrome Type I, diagnosed at one month of age. JH's presenting signs of Stickler Syndrome included cleft palate, failed newborn hearing screening, and family history of Stickler Syndrome. Family ocular history was significant for keratoconus, retinal detachment, and congenital cataracts.

At JH's initial examination, visual acuity was recorded as central, steady, and maintained at near OD and OS. The patient would not fixate on distance charts and was uncooperative for visual acuity testing such as matching Allen pictures. Pupils were equal, round, and reactive with no afferent pupillary defect. His extraocular muscle movements were full and comitant, and no strabismus was present upon cover test with both distance and near fixation. JH was unable to perform stereoscopic testing due to comprehension. Anterior segment and posterior segment examinations were unremarkable. Cycloplegic retinoscopy revealed OD: -12.00 DS and OS: -9.50+1.50x180. The patient was given a prescription for the full cycloplegic refractive error, and full-time spectacle wear was encouraged. The patient was

instructed to return to clinic in two to three months for an acuity, alignment, and compliance check.

The patient returned for follow-up three months later. His parents reported good patient compliance with spectacle wear. Visual acuity was OD: 20/400 (6/120) and OS: 20/60 (6/18) with Allen pictures. The patient exhibited extreme resistance to cover of the left eye. Visual acuity testing was of questionable accuracy due to patient comprehension and fatigue. The patient's vision was also evaluated for fixation preference and was assessed as OD: central, steady, non-maintained and OS: central, steady, maintained while fixating a distance target. The patient was unable to perform stereoscopic testing due to comprehension. Retinoscopy performed over the spectacles while the patient fixated on distance objects was plano OD and OS. The patient's extraocular movements were full and comitant OU. This examination revealed that despite full-time spectacle wear for three months, severe amblyopia of the right eye remained. Due to the severity of the anisometric amblyopia, full-time occlusion of the left eye was prescribed. The patient was instructed to return to clinic in one month for assessing efficacy of the prescribed occlusion therapy.

At the patient's follow-up visit one month later, the parents reported excellent compliance with full-time occlusion and spectacle wear. Visual acuity had improved to OD: 20/50 (6/15) and OS: 20/30 (6/9) with Allen pictures. Retinoscopy performed over the spectacles remained plano OD and OS. JH was orthophoric with cover test at distance and near fixation. Due to the success of treatment and excellent compliance, the patient was recommended to continue full-time occlusion and return for a follow-up examination in one month.

The patient reported for follow-up three months later. His parents again reported excellent compliance with full-time occlusion. Visual acuity had improved to OD: 20/30 (6/9) and was stable OS: 20/30 (6/9) with Allen pictures. Retinoscopy performed over the spectacles remained plano OD and OS. JH continued to be orthophoric with cover test at distance and near. By the fourth month of full-time occlusion, the patient's amblyopia had resolved. Because JH had been treated with full-time occlusion, tapering treatment was necessary to decrease the likelihood of amblyopia recurrence. Therefore, the parents were instructed to decrease JH's patching to two hours per day for one month, then discontinue all patching for one month, and return for visual acuity assessment and cycloplegic exam in two months. Unfortunately, the patient was lost to follow-up.

Discussion

Treatment Options for Severe Amblyopia

The Pediatric Eye Disease Investigator Group (PEDIG) has performed many clinical studies that have shaped current amblyopia treatment. PEDIG's investigation comparing six hours to full-time occlusion in severe strabismic and anisometric amblyopic children revealed that "six hours of prescribed daily patching produces an improvement in visual acuity that is of similar magnitude to the improvement

produced by prescribed full-time patching."⁹ Although the final outcome in visual acuity was similar in the two groups (4.8 lines of improvement in the six-hour group and 4.7 lines of improvement in the full-time group), the full-time occlusion treatment group appeared to reach that final outcome more quickly than the six-hour treatment group. In the six-hour treatment group, only one patient was able to reduce patching due to equal visual acuity, while eight patients in the full-time occlusion group were able to reduce patching for the same reason within the first four months of treatment. In addition, PEDIG found that patients younger than five years old with baseline visual acuity between 20/200 and 20/400 showed greater improvement (6.9 lines) compared to those older than five years old (4.4 lines).¹⁰ However, the full-time occlusion group had fewer patients younger than five, which could cause the data to favor the six-hour treatment group.

Other treatment options for amblyopia include atropine, Bangerter filters, and newer innovations such as iPad games with red/green lenses. PEDIG has explored the use of atropine as treatment for severe amblyopia, finding that although there have been reported improvements in visual acuity, residual amblyopia is present in the majority of cases of severe amblyopia after several months of atropine therapy.¹⁰ For many patients with severe amblyopia, atropine alone is not a severe enough penalization to result in favoring fixation with the amblyopic eye, therefore the patient continues to suppress the amblyopic eye. The use of Bangerter filters for the treatment of moderate amblyopia has also been explored.¹¹ However, Bangerter filters alone may not penalize vision to a high enough degree to be successful treatment for severe amblyopia.¹² Newer treatment options such as iPad games with red/green lenses have been explored in moderate amblyopes.^{13,14} However, like Bangerter filters and atropine, the penalization of the non-amblyopic eye may not be severe enough to improve the vision in the amblyopic eye at as quick a rate or as successfully as occlusion. In addition, there have been no studies to investigate the effectiveness of Bangerter filters or iPad games in severe amblyopes. Therefore, in these patients, occlusion is the most efficient and effective treatment.

Full-Time Occlusion

Due to JH's young age and the severity of his amblyopia, the goal of full-time occlusion was to resolve his amblyopia as quickly as possible. There were many factors that led to the decision to prescribe full-time occlusion as the treatment for JH's amblyopia. First, because JH was three years old at the beginning of treatment, his visual system was especially vulnerable to the effects of patching, leading to an even faster recovery of vision. As was shown in the PEDIG study comparing six hours and full-time occlusion, patients under the age of five with severe amblyopia treated with full-time occlusion had the greatest improvement in vision.¹⁰ Second, it has been found that higher amounts of patching have the most influence on visual outcome in unilateral amblyopia,¹⁵

and maximum therapy (full-time occlusion) leads to faster and more complete resolution of amblyopia.¹⁶⁻¹⁹ After only one month of full-time occlusion, JH's amblyopia had improved from 20/400 to 20/50, and after only four months of full-time occlusion, JH's amblyopia had resolved.

Many practitioners have reservations with prescribing full-time occlusion due to perceived patient intolerance and poor compliance. However, PEDIG's study comparing full-time occlusion to six hours of occlusion found that the treatments were equally tolerated.¹⁰ In addition, when parents are educated about treatment options and efficacy, they are more likely to prefer full-time occlusion compared to other amblyopia treatment options.²⁰ An advantage to full-time occlusion is consistency in treatment, which can lead to better compliance. It has been shown that more consistency in treatment has led to better patient compliance, especially in younger patients.¹⁹ It has been theorized that because the patching regimen does not change, the child more readily adapts to patching as part of their normal routine. JH's parents had great compliance and satisfaction with the treatment selection of full-time occlusion. When questioned about compliance, his parents expressed that it was easier to leave the patch on from "breakfast to bedtime" than to monitor the hours of occlusion. In addition, full-time occlusion has a definite endpoint to treatment: equal visual acuity or visual plateau. Because the initial treatment is the most aggressive treatment, there is no inconsistency or change in treatment. In contrast, if part-time occlusion or atropine is prescribed and amblyopia remains, increasing the amount of occlusion time is necessary until amblyopia resolves or the maximum treatment has been explored.⁸ As was shown in this case report, it is important to note that tapering treatment is necessary for patients who have reached maximum therapy (full-time occlusion). It has been shown that if treatment is stopped abruptly, there is greater chance of recurrence of amblyopia.⁹ Anecdotally, parents find it easier to taper patching hours than to increase patching hours.

There are many factors that can influence success of amblyopia treatment, including age at onset of insult resulting in amblyopia, age at initiation of treatment, severity of amblyopia, and compliance with treatment. As in this case, despite the severity of the patient's amblyopia, due to his young age, aggressive treatment, and good compliance, this patient's amblyopia successfully resolved, showing that full-time occlusion leads to effective and efficient resolution of severe amblyopia.

Conclusion

Full-time occlusion has many advantages, including increased compliance and reduced treatment time. Practitioners should not be deterred by perceived patient intolerance or presumed poor compliance. As in this case report, the patient and parents were compliant and happy with the success of full-time occlusion, which led to quick and successful resolution of amblyopia.

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