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TREATMENT OF AMBLYOPIA WITH A PLUS LENS:
A LITERATURE REVIEW AND CASE REPORTS

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Biographical Sketches

Jerald T. Jolley graduated with an associate degree in Respiratory Therapy in 1988. He then finished his pre-optometry requirements at Weber State University in Ogden, Utah. He graduated with his bachelor's degree in Visual Sciences at Pacific University in 1992. He worked at LDS Hospital in Salt Lake City as a Respiratory Therapist before coming to Oregon. He has also worked at Oregon Health Sciences University Hospital in Portland while in optometry school. He graduated with his Doctorate of Optometry in May 1994.

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Treatment of Amblyopia with a Plus Lens: A Literature Review and Case Reports

Pathogenesis of Amblyopia

Amblyopia is a reduced visual acuity in one or both eyes that cannot be accounted for by uncorrected refractive error, ocular or neurological disease or structural abnormalities in the visual pathway.[1] Amblyopia can also be defined as 20/40 or poorer vision in one eye or a difference in visual acuity of at least one line when either eye is not correctable to 20/20.

Many theories about the pathogenesis of amblyopia have evolved over the years. The best known theory hypothesizes poor acuity as a result of a lack of neuronal development caused by light or form perception deprivation. The basis for this theory rests on the research for which Hubel and Wiesel won their Nobel prize for their studies on neurodevelopment. Using the animal model, they occluded an eye either by suturing the eyelid or by using an occluding contact lens during the period of susceptibility early in the animal's life. A decrease in visual acuity was noted in the deprived eye. Single unit recording of the striate cortex and histological examination of the lateral geniculate nucleus (LGN) showed marked neurophysiological abnormalities. Reduced cell area sections in the layers of the LGN subserved by the deprived eye were also noted. If occlusion was for a shorter period of time then visual acuity of the animal could be increased with prolonged periods of binocular vision. When occlusion was alternated daily they found that the number of cortical cells driven by each eye individually were approximately equal, and that the number of cells driven by both eyes—binocular cells—was decreased.[2] Unilateral deprivation is detrimental to the visual acuity of that eye, and alternating deprivation is detrimental to the binocular system.[3] Other studies on visual deprivation in animals have suggested that the longer the duration, especially at the height of the sensitive period, the deeper the amblyopia.[4]

Another theory indicates that amblyopia comes as a result of active neuronal inhibition of the amblyopic eye by the non-amblyopic eye. Bicuculline, a powerful convulsant which blocks the inhibitory effects of neuron cell firing, was used by Duffy on young cats with induced deprivation amblyopia. With the use of this drug he found that neuronal input was getting to visual cortex neurons from the deprived eye, where without the drug no stimulation from the deprived eye would reach the visual cortex. Spear used enucleation of the non-deprived eye to increase function of the deprived eye. By removing the non-deprived eye, tonic inhibitory effects were reduced and 30-40% of the cells of the visual cortex could now be activated by stimulation of the deprived eye.[5] Other inhibitory theories range from inhibition at the cortical level (central) to inhibition at the retinal (peripheral) level.[6]

The animal models that have been developed by the various researchers can help us understand the nature of the cause of amblyopia. Clinicians are divided on acceptance of the animal model, but information derived from animal research is helpful in the prevention and treatment of amblyopia in children.[7]

History of Amblyopia Treatment Methods

The treatment of amblyopia can be best described as an art and a science. Healthcare providers of all kinds, including both optometrists and ophthalmologists, have been searching for the perfect cure. Traditional occlusion therapy using a patch is considered by some as the gold standard, and is still today the most widely used method of amblyopia treatment.

The history of traditional patching goes back some time. Buffon first described occlusion in 1743.[8] Though he felt that strabismus was due to an inequality between the two eyes, he failed to fully grasp the significance of the refractive error. Donders was the first man to point out the neccessity of refractively correcting hyperopic convergence esotropia. Building upon the work of Buffon and Donders, Javal successfully used occlusion not only to improve acuity of amblyopes, but also to break down suppression with prolonged occlusion. Most ophthalmologists in the late 1800's rejected occlusion completely as a treatment for amblyopia because they felt that amblyopia was congenital in nature. Others felt that occlusion could cause psychological trauma to the patient. In the end, however, success would not be ignored, and practical experience eventually overruled dogma.[9] Worth in 1903 noted the recovery of visual acuity in amblyopic eyes after prolonged atropine administration to the sound eye.[10] In 1931 Baxter was the first to suggest using a strong high plus lens for partial occlusion. He used only enough plus fogging power to switch fixation from the sound eye to the amblyopic eye. Many other partial occlusion techniques were introduced in the early 1900's including smoked glass, opaque strips on dark lenses ("Javal Grid"), marled glass (a cloudy lens), a graduated set of clip-on occluders, and red filters.[11] Pouliquen, a European investigator, introduced the word "penalization" when he began instituting it as a treatment for amblyopia in 1958.[12] In some parts of Europe penalization has become the method of choice for the treatment of amblyopia, replacing conventional occlusion.[13] Penalization began to be reported in American literature in the late 1960's and 1970's.

Penalization

Penalization is defined as an optical reduction of form vision of the non-amblyopic eye at one or all distances of fixation.[14] The effect may be achieved by alteration of the spectacle correction and/or use of cycloplegic drugs.

Historically, two types of penalization have been described. (See Table 1.) The first is *penalization at near.* With this form of penalization the non-amblyopic eye is corrected for distance and is atropinized to prevent accommodation. (See Figure 1.) The amblyopic eye is given between +1.00 and +3.00 diopters sphere in addition to the distance correction. The object is to force the patient to use the amblyopic eye at near without any accommodative demand.[15]

The second form of penalization is *penalization at far.* With this form the non-amblyopic eye is fully atropinized and enough plus sphere is added to switch fixation from the non-amblyopic eye to the amblyopic eye. (See Figure 2.) The amblyopic eye is fully corrected at far without atropine. This method penalizes the non-amblyopic eye at far. The amblyopic eye is used in the distance and the non-amblyopic eye can be used at near. Penalization at far can also be utilized without using atropine in the non-amblyopic eye.

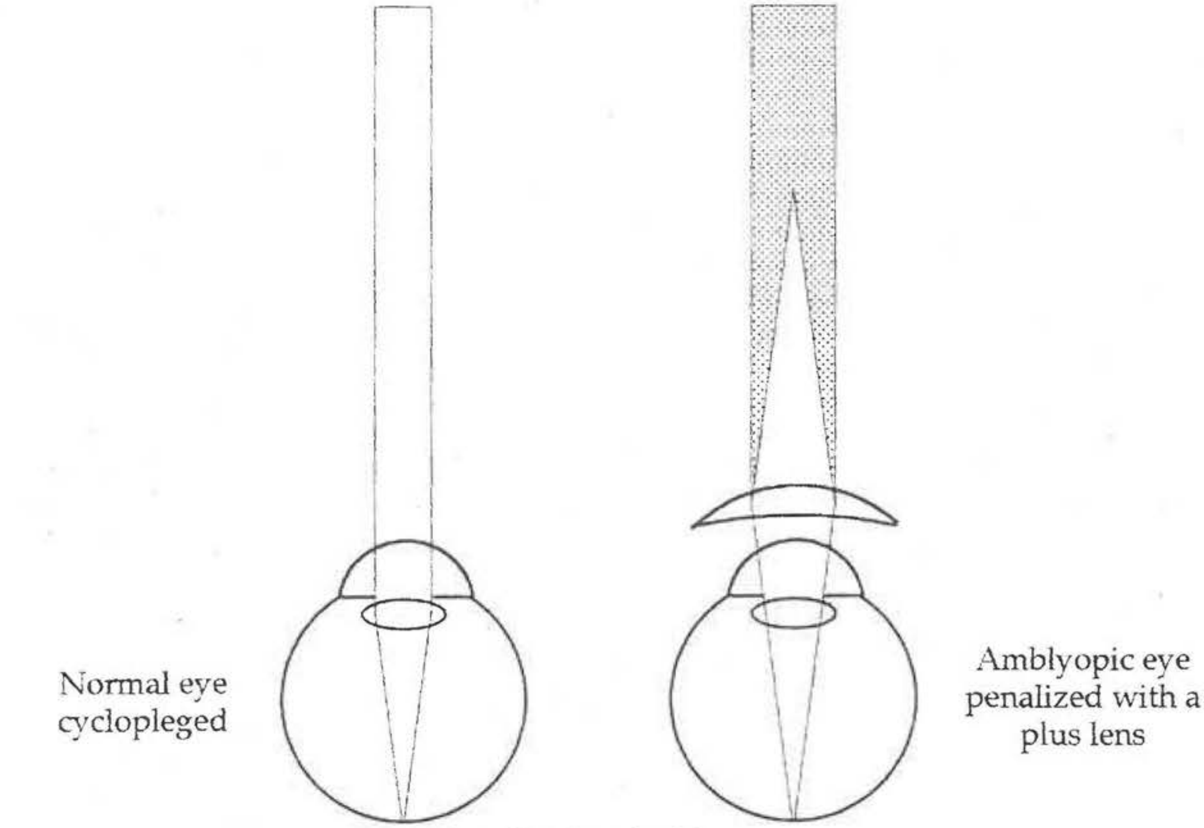
Some of the early theories advocated using penalization at near first and then as the patient has achieved maximum benefit, switching to penalization at far.[16]

Total penalization means to completely blur the image before the non-amblyopic eye. This may be done by atropinizing the non-amblyopic eye and then by placing a -4.00 diopter sphere lens before that eye. The amblyopic eye is corrected for the distance. This method may be used when the amblyopic eye can be used for both near and far distances.[17]

There are other techniques described in the literature. *Alternating penalization* is described by von Noorden and Attiah.[18] With this form of penalization two pairs of glasses are used. One pair has the plus sphere overcorrection in the right eye and the other pair has the overcorrection in the left eye. Alternating penalization can be used to preserve visual acuity in patients who have been successfully treated for amblyopia. With *selective penalization* the non-amblyopic eye is fully corrected and atropinized while the amblyopic eye is corrected with a +2.00 diopter sphere lens.

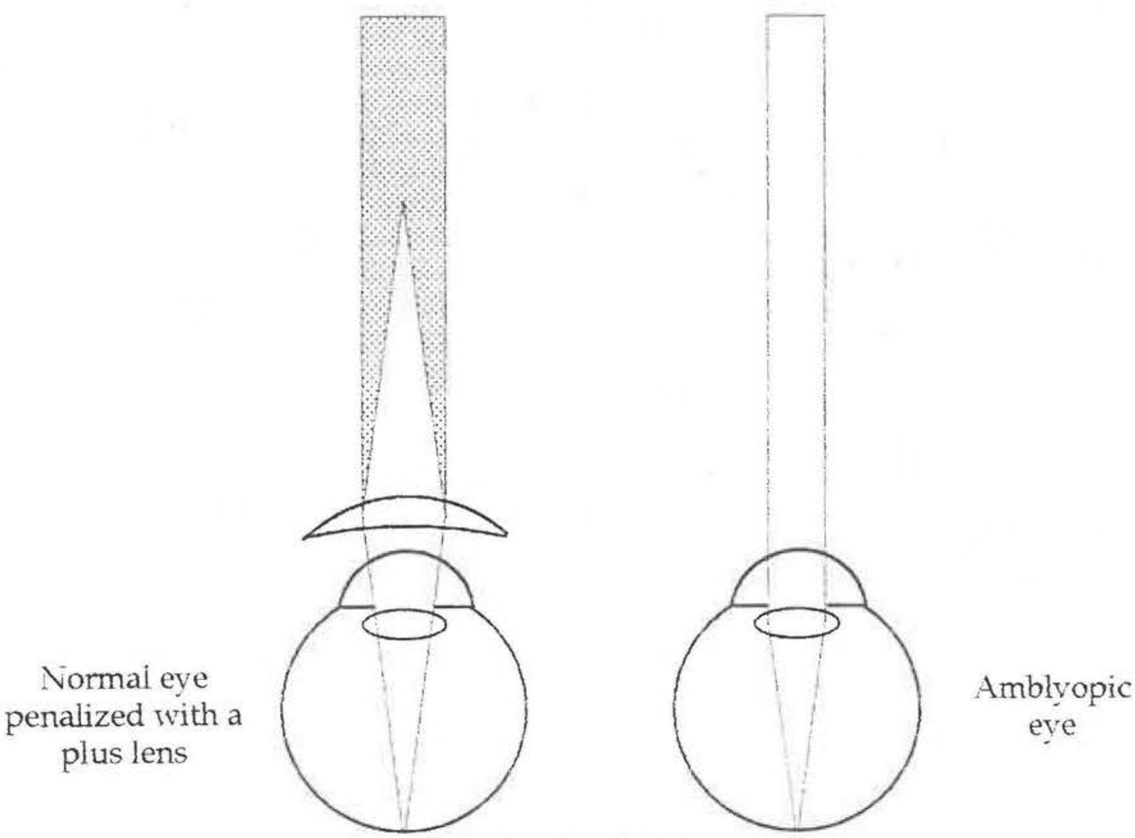
Table 1. Various Methods of Penalization

	Non-amblyopic Eye	Amblyopic Eye
Near	Full correction with cycloplegic and no add resulting in penalization at near.	Full correction with additional plus to allow for near vision.
Far	Full correction with additional plus resulting in blurred distance vision and clear near vision.	Full correction.
Complete or Total	Full correction with cycloplegic and -4.00 diopter sphere for blur at near and far.	Full correction.
Alternating	Two pairs of glasses: 1) Full correction with additional plus for penalization, resulting in blurred distance vision. 2) Full correction.	1) Full correction. 2) Full correction with additional plus for distance penalization.
Selective	Full correction with a cycloplegic for clear distance vision.	Bifocal with full correction for distance and +2.00 diopters in add for clear distance and near vision.
Slight or Mild	Full correction with +1.50 diopters for distance penalization, resulting in slight distance blur.	Full correction.



Penalization for Near

Figure 1.



Penalization for Distance

Figure 2.

Selective penalization is used to alternate or stabilize ocular dominance. Finally, with *slight or mild penalization* the non-amblyopic eye is overcorrected by +1.50 diopter sphere and is not atropinized while the amblyopic eye is fully corrected. This method is used as a maintenance therapy where no fusional result has been obtained at the end of strabismus therapy.[19]

What kind of patients can benefit from penalization? First, some authors have advocated the use of penalization over the use of traditional patching on the basis of cosmesis alone.[20] The stress of social situations in school age children prescribed a patch is enough to cause some patients and/or parents to reject patching as a form of treatment.[21, 22] The use of a high plus lens in a pair of spectacles or contact lenses gives a more natural appearance to the patient.[23, 24]

Second, a loss of acuity in patients who have previously been successfully treated with a patch is usually easily restored with alternating penalization.[25]

Third, another reason for penalization over patching is that patching an eye with nystagmus can increase the frequency of the nystagmus. The clinician must be very observant to prescribe the lens that causes the least amount of nystagmus.[26]

Fourth, neurological problems such as cerebral palsy can also be an indication for penalization, for the reason that it is generally tolerated better by the patient.

Fifth, the potential for binocularity is greater when optical penalization is used than when occlusion is used. The Optometric Extension Program recommends progressively cutting down the penalizing plus power as the patient's acuity improves in order for the patient to become progressively more binocular.[27]

Other reasons for using penalization over patching include skin allergies to adhesive tape and failure with traditional patching.

The literature is split when it comes to binocularity. As mentioned earlier, there are two different schools of thought as to the pathogenesis of amblyopia. On one hand penalization is praised because binocularity is maintained and binocular cells in both hemispheres of the cortex receive input. Penalization can also help to inhibit suppression of an eye by the patient and provide the possibility of normal retinal correspondence.[28] On the other hand, since with penalization both eyes are receiving stimulation the non-amblyopic eye still has an inhibitory effect on the amblyopic eye, thus reducing the possibility of binocular vision.[29] This paper will discuss penalization as a substitute, or at least as an adjunct to traditional occlusion with a patch.

Patient Selection for Penalization

Many studies on the subject of penalization utilize +1.00 diopter to +3.00 diopter sphere over best distance refraction as the means of plus penalization.[30, 31, 32, 33] Since patients vary in their responses to lenses, a single amount of overcorrecting plus sphere given to every amblyopic patient being treated with optical penalization is not the best choice. The goal of penalization is to switch fixation from the non-amblyopic eye to the amblyopic eye.[34, 35] Reduction of acuity of the non-amblyopic eye to levels below that of the amblyopic eye may not be enough to switch fixation. Fixation switch may occur and be determined by a number of different methods. In a controlled environment such as the major amblyoscope, fixation can be switched by flashing a light or by changing the amount of illumination. Snellen visual acuities, Bagolini striated lenses or the Worth 4-dot test are not adequate in determining fixation.[36]

Some objective tests have been advanced for the determination of fixation switch. The *Modified 4-prism diopter test* is a modification of the classical 4-prism diopter test. With this procedure the clinician starts with a strong plus sphere overcorrection of the non-amblyopic eye. Using the 4-prism diopter prism, which can be oriented in any direction over either eye to determine fixation, the clinician must determine the minimum amount of plus sphere over the non-amblyopic eye that the patient will take while still maintaining fixation with the amblyopic eye. The amount of plus is progressively reduced in the non-amblyopic eye until fixational movement is seen with the non-amblyopic eye.[37] The *10-diopter prism dissociation test* is helpful in determining fixation during the unilateral cover test. The procedure is performed the same way as the unilateral cover test, with the addition of ten prism diopters of vertical prism placed before an eye. The prism serves to amplify fixational movements by the patient.

One very good subjective test used to determine fixation switch is described by Repka et al., called the *Vectographic Fixation Test*. The 20/40 Snellen acuity line of a vectographic slide is presented to the patient. It serves as a good target because it has 6 letters: two are seen by the right eye, two by the left eye, and two by both eyes. Behind polaroid glasses, +0.25 diopter sphere is added to the non-amblyopic eye until the patient reports the switch in fixation. These researchers found that only an average of +1.25 diopter sphere was necessary to switch fixation, much less than the arbitrary +3.00 diopters prescribed by some clinicians.[38]

Since all the proposed methods of testing the amount of penalization mentioned here take a very short time, a combination of both objective and subjective methods should be used in determining the best amount of plus to provide adequate penalization.

To help predict prognosis a contrast sensitivity function (CSF) may be plotted before penalization treatment begins. CSF losses can be classified into two categories. Type I loss is specific to high spatial frequencies or smaller Snellen letters. Type II loss is a decrease in CSF across the whole spatial frequency range. Strabismic amblyopes generally fall into the category of a Type I loss. Type II loss generally describes the anisometropic amblyope[39]. Leguire et al. have expanded upon this concept. They theorized that for the amblyope in the 20/50 to 20/80 range the CSF can determine the amount of embeddedness of the disease process. For those patients showing a Type II loss, the amblyopia is expected to be more stubborn, requiring more lengthy and aggressive therapy.[40]

Amblyopia Treatment Success Rates

Several articles have reported success rates of various methods of amblyopia therapy. In 1977 Birnbaum did one such comprehensive survey. He reviewed 23 published amblyopia studies and concluded that between 32 and 40 percent of patients in age groups ranging from below 7 to above 16 were treated successfully when success was judged as 20/30 or better. When four lines improvement in visual acuity was used as the success criterion, better than 54 percent of patients under age 15, and nearly 42 percent of patients 16 and over were successfully treated. Oliver et al.[41] reported on success rates of amblyopia therapy in children more than 8 years old. In a prospective study of 350 children, they found that in children who were compliant, the improvement in visual acuity was almost equal between groups of subjects both above and below 8 years of age. Flynn and Cassady reported on their personal experience at Bascom Palmer Eye Institute. From 1966 to 1976, of 544 patients who were diagnosed with amblyopia, 439 were offered treatment. 30% of those patients were considered a success when success was defined as an improvement in visual acuity to 20/40 or better with foveal fixation.[42] Jenkins and Pickwell[43] treated 50 young amblyopic patients up to age 9 and found the success rate (two Snellen lines improvement in acuity) for conventional methods (occlusion with vision training) to be 50%.

For this paper 10 different studies were reviewed on the success rate of amblyopia therapy using some form of penalization. These results are summarized in Table 2. Without matching patients or success criteria from each of the studies we can say that a majority of these patient showed at least some success with penalization. Two of the studies, one by Repka et al.[44] and the other by Frank and

Table 2. Reported Success Rates

Study	Success Ratio	Definition	Inclusion Criteria	Type of Penalization	Duration
Gregerson et. al	17/23	Significant improvement 20/80 to 20/30	Mean refraction +3.00 Mean acuity 20/80 Ages 3-8	Optical + Drug at Near	6.5 mo.
McKinney and Boyers	5/10	Significant improvement	Refractive error > +2.00 in sound eye Mean acuity 20/100 Ages 4-7	Optical + Drug at Near, then switch to far	6.5 mo.
von Norden and Milam	10/17 7/8 for near	Improvement of 2 lines	20/100 Ages 2-12	8 Near 6 Total 1 Distance 1 Total-Alternating 1 Near-Total	8.4 mo.
Ron and Nawratzki	23/28	Improvement to 20/40-20/30 or 3 line difference	20/60 Presence of +1.50 Ages 6-12	Optic + Drug at near, then distance using filters	10 mo.
Repka et. al	22/34	Improvement in Snellen acuity	20/60 Ages 3-11	Distance only	3-31 mo. Mean 22.5 mo.
Timmerman	22/38 near 9/11 far	Acuity increase by .2	20/60 Ages 1-13	33 Near 11 Distance 3 mild 12 Alternating	5-15 mo.
Frank and France	15/24; 15/19 with good compliance	Improvement by 1 line Snellen	20/40 average Ages 5-11	Distance only	1-31 mo.
Ron and Nawratzki	26/46 <6yo 16/25 >6yo 13/21	Alternation in babies 20/30 or 20/24	20/40 10 mo.-11 yrs.	7 Near 4 Distance 21 Total 14 Near + Far	2-16 mo.
Willshaw and Johnson	6/10	Improvement 4/60-6/18 to 6/9 or better	2 lines or more acuity loss Ages 3-5	Optic + Drug at near, then distance	8 mo.-16 mo. mean 12 mo.
Haase	64/69	Improvement	Age 2-18	Optic + Drug at near	11 mo.

France[45] used penalization at distance exclusively. Their success rates were 22/34 (65%) and 15/24 (63%), respectively. The duration of the therapy they administered (1 to 31 months) appeared to be longer than other forms of penalization or combinations of penalization used by other researchers.

Case Studies

For approximately the last five years Ronald M. Pugh, O.D., of Spanish Fork, Utah, has used optical penalization at distance almost exclusively for the treatment of amblyopia. He has reported great success with this method. From his records we here present five case studies in which optical penalization has been used. No other treatment was used by these patients concurrently. Because this is a retrospective study, the cases reported here are anecdotal, given for the purpose of demonstrating that optical penalization can be effective in the treatment of amblyopia. For none of these cases was the amount of plus sphere overcorrection progressively reduced as the patient becomes more binocular, as the OEP Foundation suggests doing.

Case 1

M.L. was almost seven years old at his initial exam. Retinoscopy showed +0.12 sph OD, and +3.75 -1.00 x 180 OS. Uncorrected visual acuities at far were 20/20+ OD, and 20/70 OS. With this he was 5^{Δ} esophoric. Through a trial lens of +3.25 -0.50 x 180 over the left eye, this patient called off almost all of the 20/40 line. He was prescribed +3.50 sph OD as a distance penalization lens, and +3.50 -0.50 x 180 as a correction for the left eye. His mother reported two weeks after dispense that he was wearing the glasses a lot and was doing OK.

At a progress check six months later, M.L. had a corrected visual acuity of 20/20-2 in his left eye. No change in therapy was recommended.

$5\frac{1}{2}$ months later M.L. was correctable to 20/20+2 OS. On various visits over the next four years this improvement in acuity of the left eye remained stable and the right eye began to show between +1.50 and +2.00 diopters sphere of increased hyperopia. By the age of 12 penalization therapy was discontinued for M.L., who then began to wear his full correction in the form of disposable contact lenses.

Case 2

R.G. was almost six years old at his initial eye exam. His uncorrected visual acuities were 20/400 OD and 20/20 OS. With a subjective correction of +4.25 sph OD

R.G. was calling off the 20/100 letters, with +2.25 -0.50 x 180 OS he was 20/20. At this time he was prescribed the subjective.

Two months later R.G. was able to call off the 20/70 row with his right eye. At this time the left eye was prescribed a penalization lens of +4.50 and the patient was told to wear the glasses "all the time."

The following month R.G. was able to call off the 20/50 line with his right eye.

Nine months later R.G. was getting an aided acuity level of 20/25 OD.

One year later R.G. was getting an aided acuity level of 20/20+1 OD. This represents an acuity increase from 20/70 to 20/20+1 in 22 months by optical penalization alone. This acuity advance remained stable after another 16 months and the penalization therapy was stopped. This patient is now wearing contact lenses.

Case 3

J.J. was 7 years old when he was examined in the office of Dr. Pugh. A subjective refraction showed that he needed -7.00 sph OD, 20/60-2, and -5.25 -0.25 x 180 OS, 20/40-3. He had previously been wearing spectacles of -5.75 sph OD, and -5.00 sph OS. J.J. was prescribed the full -7.00 sph OD and was overplussed to -3.50 sphere OS. At a follow-up appointment three months later, the aided visual acuity of J.J.'s right eye had not changed from 20/60. It was reported that J.J. had hated his glasses, that they made him sick, and that he would not wear them at first, but that it was "OK now." Three months later at another follow-up appointment, the acuity of J.J.'s right eye had improved to 20/30-2. Over the next three years the acuity of that eye remained fairly stable: it was always between 20/40- and 20/30.

Case 4

C.D. was over $6\frac{1}{2}$ years old when her optical penalization therapy for amblyopia began. She was a right esotrope and had been prescribed a frosted lens OS since age three, which had brought her acuity OD from 20/100- to 20/30. At this visit she had an aided visual acuity of 20/50-2 OD, and 20/30+1 OS.
Retinoscopy showed +5.25 -0.25 x 180 OD, and +5.25 sph OS. She was prescribed +5.00 sph OD, and a penalization lens of +6.25 sph OS. $6\frac{1}{2}$ months later C.D. was 20/40+ OD. Another $4\frac{1}{2}$ months later she was still 20/40 OD with a stable refraction. With unchanged treatment she was 20/25-2 OD a year and a half later, and two months after that she refracted to 20/20-3 OD and 20/20 OS with +4.75 sph OD and +4.50 sph OS. At this time C.D. was over 9 years old, and in an effort to solidify the gains in acuity of her right eye, optical penalization was continued in the form of contact lenses with +1.50 diopters sphere overcorrection in the left eye.

Case 5

J.C. was a left esotrope and had previously undergone esotropia surgery. At the first exam in Dr. Pugh's office, J.C. was 11 years old and had an uncorrected visual acuity of 20/20-2 OD, and 20/100-1 OS. Two pairs of glasses were ordered for him: one with the full correction of +0.50 -0.62 x 112 OD, +0.75 -0.62 x 129 OS, and the other pair with a frosted lens OD, and the same lens OS as the first pair. J.C. was instructed to wear the frosted lenses $1\text{-}1\frac{1}{2}$ hours per day. Three years later his acuity was 20/70+1 OS. At this time optical penalization was initiated with a correction of +3.00 sph OD and +1.00 -0.25 x 090 OS. In four months his visual acuity OS had improved to 20/30-2. (The best acuity of his left eye at this time was 20/15-2.) J.C. did not come in for another exam for $3^1/_2$ years, at which time, because of non-compliance, his acuity OS had dropped to 20/60+1.

Conclusion

The treatment of amblyopia with penalization is challenging but rewarding. As with any treatment plan a very thorough case history must be taken. The amount of time the visual acuity has been decreased for the patient needs to be quantified. The age of onset, the presence of any disease process such as congenital cataracts, or other conditions which might have interfered with vision early in life during the sensitive period needs to be evaluated. The most severe effects of deprivation occur before the age of $2^1/_2$.[46] Visual acuity at far and near needs to be determined. Whole chart, isolated line and isolated letter acuities should be taken. An organic cause for the amblyopia should be ruled out. A full refraction including a cycloplegic refraction should be done. If present, the amount and direction of eccentric fixation as well as the presence of anomalous retinal correspondence (ARC) should be determined. If eccentric fixation or ARC are present then these need to be treated before penalization can be done.

When determining the amount of add to use for penalization, the maximum amount of plus that the patient will accept should be determined. After Reptka, et al.[47] had determined the amount of plus that would switch fixation, on follow-up visits they discovered that in some cases more plus (+0.87 for some patients) was needed to keep fixation switched, when the patient wasn't wearing his cycloplegic refraction. Haase found that the full manifestation of plus acceptance occurred after about 3 months of penalization.[48] Von Norden suggests using near penalization for deep amblyopia and then switching to penalization at far as acuity improves. The best results with penalization have been when the initial acuity has been better than 20/100.

One of the most frustrating issues facing clinicians in any treatment is compliance. Oliver et al. reported that the compliance of treatment of amblyopia by patching dropped from 72% of 2 to $5\frac{1}{2}$ year old patients to 47% of 8 to $11\frac{1}{2}$ year-olds.[49] A significant problem of compliance had been reported in numerous articles. Trick wrote "a patient's beliefs about his health condition and the efficacy of treatment are more important than the true state of his condition or therapy".[50] Press elaborates that the social consequences and the recognition of the child that he is suddenly "abnormal" can be very good reasons for noncompliance.[51] McKenny and coworkers summarized that cooperation of patients and parents was excellent with optical penalization therapy.[52] Gregerson, et al.[53] report that children spontaneously kept their spectacles on during penalization. School children can use their non-amblyopic eye at least part of the time and can participate more easily in normal school activities with optical penalization.[54] If optical penalization is a more acceptable therapy of occlusion to the patient, especially in older children, this should be the more preferred treatment.

From this review, we can see that optical penalization at far can be utilized in the treatment scheme of amblyopia. The efficacy of optical penalization is at least as good as traditional patching. Caution must be taken to avoid occlusion amblyopia, which can happen with any kind of occlusion or partial occlusion therapy. Follow-up visits must be done on a regular basis depending on the age of the patient. If clinicians don't at least try penalization in the treatment of amblyopia they are providing a disservice to their patients.[55] We feel that eyecare practitioners ought to seriously consider optical penalization as an alternative method of amblyopia treatment.

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