Arch. Soc. amer. oftal. optom. (1960) 1 - III

EXANOPTIC AND SUPPRESSION AMBLYOPIA

WILLIAM SMITH, O. D.

California, U.S.A.

As a functional abnormality of visual perception, amblyopia of the exanoptic type, particularly, is a defect which is occupying the center of interest among all concerned with visual care. Exanoptic and suppression amblyopia is not a defect associated merely with strabismus. Indeed, it is know to be prevalent in many non-squinters and even in some instances in apparente orthophores.

The concept that every case of exanoptic amblyopia is the result of strabismus is being disproved by clinical evidence. So is the concept advanced by some that amblyopia is a contributing cause in all monolateral strabismus. While both contentions have some basis, clinical proof tends to disprove it as being a universal factor and one cause, unfailingly, producing the inevitable affect.

Serious clinicians and students of defects of the oculomotor and sensory apparatus are now convinced that amblyopia of the exanoptic and/or the suppression types are specific abnormalities affecting the capability of the visual apparatus to discern and/or crearly perceive and interpret visual impressions at the macular-foveal level. Perceptual inversion in which peripheral acuity, commonly lower than central in non-amblyopic eyes, is, in the exanoptic amblyopias, actually higher than is central acuity. While it is a recognized and accepted fact that strabismus and some types of heterophorias have amblyopia as associated defects, there are, similarly, instances in which subnormal visual acuity of the amblyopia type is present in eyes devoid of motor deviations. Ametropias, especially the anisometropias and antimetropias, particularly those with moderate-to-high astigmatism, are likely to have such defective acuity. The explanation for this may be lack of development of the foveal area because of purely optical incompatibility, in which the focus ing apparatus failed in the performance of its natural function. It is an accepted concept that any interference with the normal development of visual and sensory functions, even if such interference is only transitory, may

have a harmful effect on normal development of visual perception. It may serve either as a basic or, in mild cases, as a contributing factor for the incidence of exanoptic and/or suppression amblyopia.

From reports in ophthalmic literature, most writers seem to contend that amblyopia is an active inhibitory process in the cerebral cortex. However, after extensive work on amblyopia, Bietti [1] reported that he observed a reduction in the size of central and paracentral scotomas and in the size of enlarged blind spots after subconjuctival injection of oxygen. To Bietti this indicated that the process of suppression in squint is not purely cortical, but that in involves, also, the retina. With this contention Pugh [2] seems in some measure to agree. She bases her agreement on the observation that in many persons with strabismus or heterophoria, there is a reduced sensation of brightness in one oye (usually the eye with the active deviation(s.) even if vision in that eye is full and equal in measurement to that of the non-deviating eye. The studies by Pugh involved persons with and without ocular deviations and active amblyopia. The objective of these studies was to determine the adaptability to binocular light balance. The apparatus used consisted of stereoscopic slides. Persons with normal eyes saw the disparate elements on the slides centered correctly even if the intensity of the illumination on one side was reduced. It required a reduction of almost 99% of the full light to produce a shift of fixation toward the eye receiving the higher illumination. But in those subjects with motor deviations and especially those with amblyopia, the reduction in adaptability was manifested very early. The shift of the center of the targets toward the more brightly illuminated eye was quite pronounced. It was the necessary in some instances to reduce considerably the illumination of the better eye before balancing perception of the two stereograms. Using the electroencephelograph and tests, Keiner [3] arrived at the conclusion that amblyopia is a central nervous phenomenon and that the defect belongs to the cerebral cortex. Similar conclusions were reached by experiments conducted by electroencephelography by Parsons-Smith and Dyer and Bierman. [4] Parsons-Smith's studies led him to the conslusion (a) that amblyopia associated with strabismus is present in the nervous system. He localized the defect as being in the cerebral cortex, and (b) that it was probably due to delayed myelination.

---

(1) Bietti, G. C.: L'Action de l'oxygene sur les functions retiniennes: Son emploien clinique, Bull. et mem. Soc. Franc. Ophthal. 63:195-210, 1950 (Reported in A. M. A. Arch. Ophthal 48:95, July 1952).

(2) Pugh, M.: Brightness, Perception and Binocular Adaptation, Brit. J. Oph. 35:134-142, March 1951.

(3) Keiner, G. B. F.: New viewpoint on the Origin of Squint. A clinical statistical study on its nature, cause and therapy. The Hague, Martinus Nijnoff, 1951.

(4) Persons-Smith, G.: Activity of the Cerebral Cortex in Amblyopia, Brit. J. Ophth. 37:359-364, 1953.

Dyer and Bierman [5] reached similar conclusions by experimental studies of their own. These findings tend to support similar earlier conclusions by Chavasse. Work carried on by Burian and Watson further tend to support the findings of the earlier investigators mentioned.

Yet despite these findings and contentions, the exact site in the retina or the cerebral cortex where amblyopia of the exanoptic or suppression types is localized, is not yet determined. Nor is it definitely know exactly at what stage in the incidence of monocular squint or other etiological or contributing factors amblyopia develops.

According to Abraham, exanoptic or acquired amblyopia is due to one of two causes and is classified according to the type of suppression from which it is developed. It is Abraham's opinion that suppression is the cause of amblyopia rather than the result. This, however, is a contention with which not everyone agrees. Even in the face of much research on the subject, it is not definitely established at the presente time at what stage in the development of monocular squint amblyopia becomes established. Nor is it definitely known as to which comes first, the amblyopia or the suppression. What is currently accepted is that suppression is of two types, passive and active. The passive type results from lack of oportunity for the visual apparatus to function fully and properly. This may be due to uncorrected or inaquatelly corrected ametropias and to refractive abnormalities in which the conventional optical aids fail to raise visual acuity to normal because their application has been delayed too long. Such delay may have produced a type of refractive amblyopia. In this category may also be included hyperopias of high degree, myopias, and astigmias, especially the anisometropias in which the best corrected acuity is below 20/20. None of these defects need be associated with motor or sensory defects.

The active type suppression is usually associated with a motor and/or sensory defect. In these types, binocular central fixation (bifoveal fixation, that is) is either impossible or undesirable because of discomfort. In order to maintain comfortable perception even if not binocularly, images from one eye are suppressed. The inhibition is centered in the foveal area and is more intense than is the clinical fact that in ocular deviation, stimulation of the fovea of an affected eye is much more distracting than stimulation of the retinal periphery. With passage of time, the non-stimulated foveal area of a deviating eye tends to lose its sensitivity, and vision is lost from disuse. Suppression invariably is more intense and greater when binocular vision is attempted. Acutally, this is a form of selective

(5) Dyer, D., & Bierman, D. O.: Cortical Potential Changes in Suppression Amblyopia, Am. J. Ophth. 35:66-68, 1952.

cortical suppression of macular pattern vision to which, according to Burian, may be added other abnormal macular functions, such as increased frequency of flicker fusion and inversion of the pupilomotor index. In amblyopia, light perception is unaffected. Amblyopic eyes are known to fixate better in dim light and to behave like normal dark-adapted eyes. However, in exanoptic amblyopia, macular pattern vision is impaired not only in angular acuity, but also in separation acuity. In this type of acuity, insolated or individual letters or characters can be distinguished, but when groups of same-size characters are presented in a row, or all at the same time, some of them are indistinglishable. Weakness of fixation is a characteristic of amblyopia. Eccentric fixation, likewise, is a common manifestation. Some are of the opinion that eccentric fixation has a motor component, and not merely a sensory phenomenon. Muscle palsy is a factor to be looked for in strabismus cases in which eccentric fixation is manifested.

While the exact area in the cortex where amblyopia is localized is not definitely known, electroencephelography, nevertheless, has succeeded in determining that the cerebral cortex is the site of origin. The explanation for the inability to pinpoint the affected area however, lies in the fact that the amblyopic retinal, and, correspondingly, cortical areas, are not alike in all cases.

All nervous activities are reflex in nature and are of two types: the conditioned, or acquired reflex, and the unconditioned, or congenital. The conditioned reflexes originate in the cerebrum. The unconditioned are simple reflex activities of the cord and brain stem. The conditioned activities superimpose on the unconditioned reflexes, when the proper structures are ready to facilitate such response. With proper usage and development, conditioned reflexes can become firmly established and habitually ingrained to resemble unconditioned or inate reflexes.

According to Chavasse,[6] the reflex of inhibition differs from other reflexes in that it is of a negative nature. But, like its prototype, the positive reflex, it requires an adequate internal or external stimulus to produce it. The distinction between the two is merely one of degree, in which the higher associated reflex paths are involved. External inhibition occurs as a result of failure of motor or secretory activity due to some external or obvious cause. Internal inhibition is produced by some obscure cause, the stimulus of which arises in the higher cortical centers.

In the visual apparatus, either or both the motor and sensory components can be affected. When the inhibition is motor, as in strabismus or high heterophoria,

(6) Chavasse's Worth Squint: P. Blakiston's Son & Co. Philadelphia, Pa. ed. 7, 1939, p. 336.

a normally functioning sensory apparatus in the form of instrumentally induced fusional responses hasn't the power to induce response of certain beneficient types of motor action. When the inhibition is in the sensory apparatus (absence of foveal fusion; anomalous retinal correspondence) the result is absence of sensory stimulus-response which, in normal situations, helps to produce motor function.

Sensory inhibition is always reflex. It can be either exteroceptive, induced through the retina, or it can be proprioceptive. In the proprioceptive or postural type inhibition, a defect in the motor system helps to induce the inhibition. Both types of inhibition can be of either the congenital, or acquired types, and they can be either constant or intermittent. As in other types, the intermittent may turn into a constant type suppression, when contributing conditions arise to bring on such a change.

It is important here to give consideration to suppression of a secondary type. This type suppression has a variety of causes. Foremost among these are palsies, developmental abnormalities, traumas and ocular and cerebral lesions. These are indirect causes and bear no relationship to those which are responsible for the amblyopias and suppression in which the etiological factors are either sensory or motor.

Just as visual acuity over the retinal surface varies in degree according to the distance from the foveal rise toward the retinal periphery, so does retinal suppression vary also in the extent of intensity in the different parts of the retina. This applies to both the monocular as well as the alternating types of suppression. The variation in the intensity is in direct relation to the natural quality of the visual acuity in the respective regions of the various parts of the retina. This type of suppression is referred to as "contoured or regional inhibition". The macular-foveal area being the zone of keenest visual acuity, is, for some inexplicable reason, inhibited most intensely. Also, the retinal periphery, which under normal conditions, has a visual acuity of lower degree than the central retina, tends to manifest, in cases of suppression, higher visual perception. This inversion in acuity is one of the differential diagnostic signs between suppression amblyopia and a reduced acuity from a refractive defect. Deviation of the affected eye as little as one degree, often produces a most startling change in the measurable visual acuity. This may explain the reason why in amblyopia peripheral vision is oftentimes higher than is central vision.

Variability in the degree of retinal suppression over the retinal surface to which reference was made in the preceding paragraph, tends to produce a flattening of the retinal contours (or various retinal regions) sufficiently to make

it possible for the areas stimulated to achieve a requisite degree of dominance. In strabismus with eccentric retinal fixation, repeated stimulation of positive, repeatedly used and acquired fixation areas, may eventually lead to establishing these as secondary areas of retinal fixation, leading indirectly to the development of secondary retinal correspondence. In such cases the highest point of the flattened retinal areas or contours, may also be eccentric. It is well to remember that the degree of eccentricity of visual acuity differs in the normal from that of an eye with amblyopia. In an eye with amblyopia, the suppression, and hence the greatest visual loss, is at the fovea. In such cases, visual acuity may be comparatively higher in the extra-foveal area. In eyes with normally functioning foveas, the tapering of visual keeness is away from the foveal area and toward the retinal periphery.

A very important physiological law relating to vision is the Law of Use and Disuse.[7] This law postulates that functions of the conditioned or acquired type must be in a state of constant and repeated usage in order to become established as fixed and established physiologic functions. If not used, the performance of such functions tends to fade out or become obliterated. During the period that conditioned functions are being reflexely developed, ingrained and matured, they are both established easily and fade easily. In later life, they are increasingly harder to establish and are similarly more difficult to erase.

The Law of Immutability[7] is another very important law upon which development of the conditioned reflex is based. This law states that once they are established, conditioned reflexes are very nearly unchangeable. However, responses arising from such conditioning may be suspended or inhibited voluntarily. When a response is thus altered, it doesn't necessarily mean that the reflex, too, is changed. One can be altered without affecting the other. Also, it is possible to inhibit faulty reflexes, to change a reflex and to substitute voluntary control. And such voluntary control, if frequently and sufficiently repeated, can become established and instilled as habit, if not as a true reflex. Habits are defined as more elaborate patterns of response than are reflexes. The line of demarkation between the two, however, is very fine and is not clearly defined. Adjustment and change are readily possible.

In addition to foveal suppression and inhibition of retinal images, many exanoptic amblyopes also manifest abnormalities in their binocular sensorial relationships. Their space sense and their sense of localization is affected. This is manifested as anomalous retinal correspondence and abnormal spatial projection. It is well

(7) Smith, W.: Clinical Orthoptic Procedure. The C. V. Mosby Co. St. Louis, M. ed. 2, 1954, pp. 76-94.

to remember that the condition of abnormal retinal correspondence is entirely a state of anomalous binocular association in the brain and is of a compensatory nature. In this respect it differs from either "eccentric fixation" or from "false macula". Both these are uniocular abnormalities. Abnormal retinal correspondence is a binocular phenomenon.

Such abnormal functions are regarded as adaptive. They are presumed to be compensatory and of a sensory type. An existing strabismus is the underlying cause and the adapted, altered position of the eyes and resultant changed sensory performances, are the resultant clinical manifestations. In such cases, alternation from the normal tends to serve as a substitute and solution, or an escape from what might otherwise be an intolerable situation vision-wise. *Actually, it is a case of an impaired visual function adjusting itself to its environment.*

The basis for normal retinal correspondence is bifoveal performance under all conditions and situations, and by all tests. Abnormal, or anomalous retinal correspondence is widely regarded as an anomalous binocular association of a compensatory nature and is believed to be localized in the cerebral cortex. As such, it should not be confused with either "false macula" or "eccentric fixation". These are abnormalities of uniocular nature and do not have the same complications as anomalous retinal correspondence.

At this point it might be well to digress and consider the meaning of retinal correspondence in order that we might have a clearer understanding of this phenomenon both in the normal and abnormal states, and the implication of the abnormal state in the function of binocular perception.

Retinal correspondence is a sensory phenomenon in which the reflexes of foveal fixation and proprioception are basic and underlying factors. In the normal act of binocular perception, images of objects in the binocular field of vision are received and registered (without conscious awareness or effort) on corresponding receptors of the two eyes.

On paper this might appear to be a very simple performance. Physiologically, however, it is not so. In keeping with their common visual direction, in the normal state, the fovea of each eye is presumed to be such a receptor. Thus, in a pair of eyes whose visual axes are parallel, straight ahead fixation would place the images on the macula of each eye. When the cortical cycle is completed and the proper areas stimulated, the image is then, similarly, projected mentally, straight ahead into space, along corresponding pathways. Images of objects situated in the right visual field fall on the nasal side of the retina of the right eye and on the temporal side of the retina of the left eye. Mental projection of such images is to the right side.

Images of objects situated in the left visual field fall on the nasal side of the retina of the left eye and on the temporal side of the retina of the right eye. Here mental projection is to the left. When such a state of corresponding binocular impongement takes place in both visual fields, normal correspondence of retinal images is the result.

But what happens when there is squint and tests show retinal correspondence to be normal, nevertheless? An example of such a case would be one in which the esotropia is of the right eye. The image fixated is presumed to fall on the macula of the fixing left eye and on the nasal side of the retina of the squinting right eye. The nasal side of the retina of the squinting eye being the area stimulated, the image is projected to the right, or in the temporal half of the visual field of the right eye. If no suppression complicates, the manifestation is homonymous diplopia.

Supposing, however, that a similar muscle defect as cited in the preceding paragraph, is complicated with abnormal retinal correspondence, the fixated image falls, as before, on the macula of the fixing left eye, ands is projected straight ahead. It falls on the nasal side of the retina, not the macula, of the squinting right eye. But not being macularly adapted because the image is suppressed and prevented from reaching the macular and paramacular areas, it is projected, not to the right, as in the earlier example, but in the direction and position of the object.

In binocular single vision each macula projects directly to the object of fixation. Together, the two maculae thus serve in a corresponding capacity to bring the corresponding retinal images to the same visual direction. (Visual direction means direction of licalization and is not the same as visual line, line of fixation, or optic axis).

When a manifest deviation occurs, there is invariably a disruption in the alignment of the two eyes and with it, loss of binocular correspondence and visual direction. The images of the non-deviating eye continue to fall on the macula, but those of the deviating eye fall upon retinal areas, away from the macula. It is presumed that when this happens, diplopia results. The explanation given is that as a result, two images of the same object are present together in the visual field, simultaneously. The second image is perceived to a side of the object being fixated.

Occurring as it does in the formative years of a child's visual development, diplopia is an intolerable phenomenon which must be eliminated. Since the retinal area used by the deviating eye is away from the macular area, stimulation and use of that macula is supressed. Disuse follows because the images are not conveyed by the macular fibres to their respective termini in the higher cortical centers.

And so, in exanoptic amblypia we have pseudomacula elements whic lack the power of conveying images and a macular-foveal area which has become dulled from lack of conditioning and normal usage.

Without going further into the more intricate physiological, neurological and psychological aspects of retinal correspondence and the various ramifications which enter into the study of spatial perceptión, the meaning of normal retinal correspondence implies impingement by visual stimuli on normally corresponding retinal receptors, the foveae. When this happens, the response is bimacular and the perceptual response is that of normal correspondence. On the same premise, binocular fixation, in which the macula of the fixing eye and a peripheral area of the squint are simultaneously stimulated, will have anomalous retinal correspondence as the result.

Exanoptic amblyopia usually develops either very early in a child's life during infanthood, or later, during childhood, but before the sixth year. It is not occur in adulthood. If does, it is from a different cause. The extent of visual loss and the degree of the accompanying deteriorative processes depend largely on the age at which the motor devistion first becomes apparent, even if intermittently, or transitorily. It also depends on the type of the primary oculo-motor defect and on the associated complications, if there are any. These may be either systemic, emotional, traumatic, or functional. Since it is a conditioned-type anomaly, in some cases development of the exanoptic amblyopia may be a slow, gradual process. The visual loss may persistently remain of low degree, without manifesting any radical or sensational progressive changes as the child grows and develops. In other instances, especially the suppression type amblyopia, visual deterioration and loss may be rapid and quite intense. It may show severe progression in a relatively short period of time. It is a known clinical fact that the extent of visual deterioration in amblyopia can vary from the insignificant loss of a single line of letters to the extreme loss of performance of macular-foveal perception running to as low as mere recognition of finger vision at close range.

Defects of the motor system are not the only etiological or contributing factors in the onset of amblyopia exanopsia. Anomalies of refraction-hyperopia, myopia and astigmatism, especially of high degree, either simple or compound, are known to produce refractive amblyopia. Of the refractive errors, hyperopia and astigmatism, especially of the compound type, of high degree, and oblique axes, are the more culpable. Of the motor defects, presence of a vertical muscle defect as a component, is likely to produce a more intense type of amblyopia. This type is not as easy to correct as is the uncomplicated type. But they are amenable. A deep-rooted amblyopia should put the examiner on guard and should cause him to examine thoroughly the performance of the vertical motor muscles.

As a complicating defect in strabismus, exanoptic amblyopia may be secondary or even tertiary in origin. However, irrespective of this, prevention of incidence of this insidious defect or vigorous elimination of it when detected, is the most important part of the remedial procedure and one that should command utmost attention and concentration.

Suppression as a visual defect is known to exist in a variety of forms and degrees. It can be either central, affecting the macular-foveal area, or peripheral. Suppression can be intense and-deep-rooted (the type that has its origin in early infanthood), or it can be slight or mild. It can be alternating, or it can be confined to only one eye - monocular or uniocular suppression. And suppression confined to the central or macula-foveal area can have also a peripheral component, and vice-versa. Loss of physiologic rivalry is the first step in the development of suppression.

The younger the age at which strabismus occurs, the higher the eventual intensity of suppression if no immediate remedial steps are instituted. In the intermittent type squint, suppression is not as high or as intense as in the constant monolateral squints. Suppression of an alternating type invariably accompanies alternating strabismus. This is more prevalent in esotropias than in exotropias. Presence of a vertical or an oblique defect tends to increase the intenseness of the suppression.

In order for any treament method for eliminating amblyopia and suppression to be successful, it must encompass steps for thoroughly and permanently eliminating macular suppression and anomalous retinal correspondence. Visual perception, both quantitative and qualitative, at distance near, must be raised to as near normal levels as possible.

To be most effective, a well-rounded out treatment procedure for eliminating suppression and amblyopia must contain the following steps: (8).

1. Elimination of central suppression and establishment of a bifoveal path of conduction.

2. Elimination of anomalous retinal correspondence and abnormal fixation.

3. Development of spatial perception - normal proprioceptive sense by eliminating vicarious posture and torticollis.

4. Development of as high grade a sense of visual orientation as possible.

5. Development of visual acuity at all distance to as high a level as possible.

(8) Smith, W.: Clinical Orthoptic Procedure. The C. V. Mosby Co. St. Louis, M. Ed. 2, 1954, pp. 70-118.

SUMMARY:

Many practitioners are frequently confronted in their daily practices with simple, uncomplicated cases of amblyopia. In many instances, routine therapeutic measures such as occlusion of the non-amblyopia eye helps to achieve desired results. It is in the complicated cases, the cases in which suppression, together with amblyopia, anomalous retinal correspondence and a motor defect, help to intensify the abnormality, that clinical skill and experience are most essential.

The discussion presented touches but very lightly on this most complex subject. However, in writing it, the author tried to cover the most salient points of the subject matter. Likewise, the five steps in the corrective procedure presented are basic and rudimentary. They are steps which the author have been advocating and teaching in lectures and in clinical use. These steps have proven clinically to be consistently most effective.

6715 Sausalito Ave.
Canoga Park, California.