COMMUNICATIONS

AMBLYOPIA *ex* ANOPSIA*

(SUPPRESSION AMBLYOPIA)

A PRELIMINARY REPORT OF THE MORE RECENT METHODS OF TREATMENT OF AMBLYOPIA, ESPECIALLY WHEN ASSOCIATED WITH ECCENTRIC FIXATION IN CASES OF STRABISMUS

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DURING the last two centuries the treatment of amblyopia *ex anopsia* in cases of strabismus has made considerable progress. One of the earliest forms of treatment was occlusion of the fixing eye. This was first carried out by Buffon (1743), who also showed the need for correction of the refractive error by means of spectacles. More than a century later, Javal (1896) thought that squint was primarily a fault of binocular function to be remedied by orthoptic exercises using a stereoscope. Amongst others, Chavasse (1939) intensified the use of occlusion in the treatment of amblyopia and stressed the importance of early corrective operation in squint, and Lyle has continued the use of these methods (Lyle and Walker, 1953). It was found, however, particularly by Chavasse, that in certain cases, occlusion did not result in any appreciable improvement in vision, and that in others the improvement was minimal. Although it was known that some of these unresponsive patients showed eccentric fixation, treatment other than occlusion was not carried out until 1940, when Bangerter began to use what he described as "pleoptic treatment" (Bangerter, 1955), in an attempt to improve uniocular vision by means of exercises involving the coordination of hand, eye, and brain. This form of therapy is now becoming recognized as a successful means of treating patients who do not respond to occlusion. In 1953, Bangerter of St. Gallen, Switzerland, developed a special method of restoring foveal fixation. In 1954, Cüppers of Giessen, Germany, developed a different method. He constructed the Visuscope for the diagnosis of eccentric fixation and the Euthyscope for the restoration of central fixation (Cüppers, 1956). His after-image method of treatment of amblyopia has recently been introduced into England and a series of patients has been treated by this method at the High Holborn Branch of Moorfields Eye Hospital.

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Clinical Material

The patients chosen for treatment were between the ages of 4 and 13 years and were selected from among those with amblyopia ex anopsia who had not responded to occlusion of the fixing eye. In some of these cases occlusion had been carried out for between 1 and 8 months with no improvement in visual acuity. Many of them had been treated surgically and all had been classified as being satisfactory cosmetically. In some cases the diagnosis of eccentric fixation, or loss of central fixation, had already been made. The age at onset of the squint varied from birth to 3 years.

Instruments*

Visuscope.—This is used for diagnosis, and is essentially a conventional ophthalmoscope with low illumination. A small star is inserted in the path of light, the image of which is seen projected on to the patient's fundus.

Euthyscope.—This is used for treatment; it is also a modified ophthalmoscope but with high illumination. A beam of light is projected to form a cone of 30° of arc on to the posterior pole of the retina. In the centre of this cone there is an unilluminated area of either 3° or 5° of arc, as shown in the Figure (opposite). There is also a green filter which enables the examiner to locate the macula exactly before turning on the white light.

Coordinator.—This is used for treatment, and its action is based on the entoptic appearances of Haidinger's "brushes"—an effect produced by polarized light only visible to the patient when he is using the fovea, because of the special anatomical construction of that part of the retina.

Methods

Diagnosis.—The unaffected eye is kept firmly closed. The examiner, using the Visuscope, focuses the small black star onto the fundus of the affected eye, and the patient is instructed to look carefully at the star. The examiner then notes the exact position of the star on the fundus in relationship to the fovea centralis. The following types of fixation have been encountered:

- (1) Central
 - (a) With nystagmus;
 - (b) Without nystagmus;
- (2) Eccentric
 - (a) Parafoveal and paramacular;
 - (b) Peripheral; this may be found between the fovea and the disc, or even on the nasal side of the disc.
- (3) Indefinite.

Treatment.—The first stage consists of total occlusion of the affected eye for a period of 3 to 4 weeks. When the fixation has become central, or is seen to be moving towards the fovea, the occlusion is changed to the unaffected eye, and pleoptic exercises are given. The after-image is produced with the Euthyscope, the pupil of the amblyopic eye being fully dilated. First the macula is found with the aid of the green filter, then the colour of the light is changed so that the macula is protected by the unilluminated area and a circular area of retina around the macula is stimulated; the light of the

* These instruments were all designed by Dr. C. Cüppers of Giessen, Germany.

Euthyscope, a 12- to 15-watt bulb, is projected on to the retina for 15 to 30 seconds; this results in the formation of an after-image corresponding to the area of retina stimulated. The after-image is either positive or negative depending on the intensity of suppression; it is usually positive at first and becomes negative later. The formation and observation of the after-image is facilitated by means of a flashing illumination on a white wall in the testing-room, the dark and light periods of the flashing being regulated independently.

The Coordinator is used for cases in which the fixation is near the macula and in which the after-image unavoidably covers the fovea as well as the eccentrically fixing point. It is also used in the later stages of the treatment of eccentric fixation after treatment has been begun with the Euthyscope. The patient is instructed to observe Haidinger's brushes and to bring and maintain them onto a fixation object controlled by his own hand. The Coordinator helps by showing the main projection of the macula outside the eye, and is therefore used to assist the restoration of foveal fixation, as well as to re-educate the coordination of eye, hand, and brain.

In-patients were treated daily and out-patients three times a week, each period of treatment lasting 30 minutes. Longer periods of treatment were found to be unsuitable because the patients became fatigued and no longer sufficiently cooperative.

Assessment of Results.—The patients under treatment for amblyopia were divided into two groups: those with central fixation, and those with eccentric fixation. After treatment had been completed, the improvement was assessed according to the visual acuity attained for distance :

- (a) None, less than 6/36
- (b) Slight, between 6/36 and 6/18
- (c) Satisfactory, 6/12 or better.

The results were judged on the "cortical" rather than the "angular" visual acuity*. Near vision was judged separately and the improvement assessed as follows:

- (a) None, less than N.36
- (b) Slight, between N.36 and N.12
- (c) Satisfactory, N.10 or better.

* The cortical visual acuity represents the ability to read a whole line of Snellen's test types accurately, whereas the angular visual acuity is the ability to read one letter per line. In suppression amblyopia, the angular visual acuity is usually better than the cortical.

FIGURE.—Diagram of Euthyscope projecting a beam of light onto the fundus. (Courtesy of Clement Clarke Ltd.)

The amount and duration of treatment depended on the degree of improvement obtained, and treatment was continued until it was clear that no further progress was likely to be made, or that further progress was likely to be so slow as to be uneconomic.

Results

(1) *Amblyopia with Central Fixation.*—There were twelve patients with amblyopia and central fixation (Table I, overleaf), eleven with convergent and one with divergent strabismus. All were between the ages of 5 and 13 years. When treatment was started the visual acuity for distance was between 6/18 and 6/36 (angular), and after treatment the results were as follows:

Improvement in Visual Acuity		Number of Cases
Distance	None	0
	Slight	2 (Visual acuity originally 6/36)
	Satisfactory	10
Near	None	0
	Slight	0
	Satisfactory	12

(2) *Amblyopia with Eccentric Fixation.*—There were 38 patients with amblyopia and eccentric fixation (Table II, overleaf), one with divergent, two with consecutive divergent*, and 35 with convergent strabismus. In the last group, two patients had a manifest vertical deviation, two a high degree of anisometropia, and thirteen a high degree of hypermetropia.

Of these 38 patients, 26 developed central fixation, and eight central fixation with nystagmus, and four remained with eccentric fixation. When treatment was started the visual acuity for distance varied between “the perception of hand movements” and 6/18 (angular), and after treatment the results were as follows:

Improvement in Visual Acuity		Number of Cases	
		Cortical	Angular
Distance	None	4	3
	Slight	16	8
	Satisfactory	18	27
Near	None	3	
	Slight	6	
	Satisfactory	29	

Before treatment by after-image stimulation was begun, occlusion of the unaffected eye had been carried out for an average period of about 8 months

* Consecutive divergent strabismus is a divergent squint resulting from an originally convergent squint, either due to over-liberal surgical correction, or to the mere passage of time.

for all but one of the cases with central fixation and all but seven of the cases with eccentric fixation, and no measurable improvement of visual acuity had resulted.

Discussion

The number of treatments necessary ranged from five to 53, the average being between twenty and thirty. On the whole less treatment was required for younger patients, particularly between the ages of 5 and 7 years, because they responded more quickly provided that their cooperation could be maintained. This is at variance with the opinion of Cüppers, who maintains that older children are more responsive to treatment. Patients with central fixation responded more quickly than those with eccentric fixation, and there was a more rapid response in those who were given daily treatment. The cases assessed as showing "no improvement" were mainly those in which there was a lack of cooperation on the part of the patient, or parent, in carrying out the treatment.

In those with central fixation, the after-image method of treatment rapidly overcame the suppression, although prolonged occlusion of the unaffected eye had previously failed to do so. In patients with eccentric fixation, the response to treatment appeared to depend upon the rapidity with which central fixation was regained; this in turn seemed to depend on the intensity of the macular suppression. As soon as the fixation became central, the angular visual acuity rapidly improved and total occlusion of the unaffected eye was carried out. This increased the rate of improvement of visual acuity, which showed a greater improvement in all cases if measured when the after-image was still present. This is said to indicate the degree to which the angular vision may be expected to improve, given sufficient time (Cüppers).

The cortical vision, especially for distance, was invariably slower to improve than the angular vision, and it was also observed that near vision improved to a greater extent than distance vision. This was thought to be due to the extensive use of Haidinger's brushes and the fact that the homework carried out was concerned with near vision, *e.g.* exercises such as crossing out all the letters "e" in a page of newsprint, as well as reading, writing, and drawing. It has been suggested that distance vision shows less improvement because children are not particularly handicapped with a visual acuity of 6/12 or 6/18, and therefore make less effort to see clearly in the distance.

Until recently it was believed that, in some cases, the visual acuity could not be improved beyond the point to which it had developed at the time of onset of the inhibition caused by the commencement of the squint. The amblyopia, represented by the visual acuity, which had developed at the time of onset of the squint, is known as the amblyopia of arrest (Chavasse).

This series included two cases in which the squint was known to have begun in the first few months of life (Cases 8 and 10 in Table II). It is

TABLE
RESULTS IN TWELVE CASES OF AMBLYOPIA
TREATED BY CUPPERES

Case No.	Age (yrs)	Diagnosis	Age at Onset of Strabismus (yrs)	Previous Treatment	No. of Treatments	Fixation of Affected Eye	
						Before Treatment	After Treatment
1	6	Right convergent strabismus Abnormal retinal correspondence	4/12	Yes	10	Central with nystagmus	Central
2	8	Right convergent strabismus High hypermetropia*	From birth	Yes	11	Central	Central
3	5	Left convergent strabismus Abnormal retinal correspondence Left hypertropia Myopia	15/12	Yes	8	Central with nystagmus	Central
4	9	Intermittent right divergent strabismus Myopia	5	Yes	12	Central with nystagmus	Central
5	6	Left convergent strabismus Abnormal retinal correspondence	4	Yes	19	Central with nystagmus	Central
6	6	Left convergent strabismus Abnormal retinal correspondence	9/12	Yes	5	Central with nystagmus	Central
7	9	Left convergent strabismus Abnormal retinal correspondence High hypermetropia	2	Yes	5	Central	Central
8	5	Right convergent strabismus Abnormal retinal correspondence	3	Yes	10	Central	Central
9	8	High hypermetropia	6	Yes	28	Central	Central
10	7	Left convergent strabismus	4	Yes	10	Central with nystagmus	Central
11	13	Esophoria Intermittent convergent strabismus Myopia	From birth	No	6	Central with nystagmus	Central
12	13	Left divergent strabismus Myopia	6	Yes	13	Central with nystagmus	Central

* High hypermetropia = more than +3D sph.

suggested that the visual acuity did not improve as a result of occlusion of the unaffected eye because there was an undiagnosed eccentric fixation, which the occlusion had only served to encourage. The vision, therefore, had only improved to the extent that the resolving power of the eccentrically fixing point would allow, which is usually not better than 6/24. In these cases diagnosis by the Visuscope revealed an eccentrically fixing point on the retina. After the restoration of foveal fixation, by the after-image treatment, the visual acuity subsequently improved, to 6/6 angular, and binocular single vision, with some range of fusion, was obtained. Cheiroscope drawing

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EX ANOPSIA WITH CENTRAL FIXATION
AFTER-IMAGE METHOD

Visual Acuity of Affected Eye						Binocular Function
Before Treatment		After Treatment				
Distance	Near	With After-Image	Angular	Cortical		
				Distance	Near	
6/36	N.36	6/6	6/9	6/12	N.5	Can alternate
6/18	N.18	6/4.5	6/7.5	6/9	N.5	Fusion and stereoscopic vision Straight on cover test
6/36		6/4.5	6/6	6/6	N.5	None
6/24	N.24	6/6	6/9	6/12	N.5	Binocular single vision for near and distance
6/24	N.14	6/4.5	6/6	6/12	N.5	None
6/36		6/6	6/12	6/18	N.6	Cannot alternate
6/36	N.24	6/4.5	6/6	6/6	N.5	Can alternate
6/24		6/4.5	6/6	6/12	N.5	Cannot alternate
6/36	N.10	6/12	6/12	6/12	N.5	Fusion and stereoscopic vision Straight on cover test
6/24		6/6	6/9	6/12	N.5	None
6/24	N.24	6/6	6/6	6/6	N.5	Binocular single vision
6/36	N.36	6/12	6/12	6/18	N.8	Binocular single vision for near

confirmed that this was true fusion.

Better and quicker results were obtained in those patients who had not had any previous occlusion, even when the fixation was sufficiently eccentric to be diagnosed by the cover test alone. Therefore, it is recommended that, in any case of amblyopia, a diagnosis of the type of fixation present should be made before occlusion, or any other form of treatment, is carried out.

The results obtained in this series are encouraging and it is hoped that the treatment will be continued. When this type of treatment is contemplated, it is desirable that it should be given as soon as the patient has developed the

TABLE I
RESULTS IN 38 CASES OF AMBLYOPIA
TREATED BY CÜPPERS*

Case No.	Age (yrs)	Diagnosis	Age at Onset of Strabismus (yrs)	Previous Treatment	No. of Treatments	Fixation of Affected Eye	
						Before Treatment	After Treatment
1	7	Left convergent strabismus Lack of normal retinal correspondence	8/12	Yes	21	Between disc and fovea	Central with nystagmus
2	10	Left convergent strabismus Abnormal retinal correspondence High hypermetropia*	6	Yes	36	Nasal border of macula	Central
3	7	Left convergent strabismus Abnormal retinal correspondence High hypermetropia	1½	Yes	36	Nasal border of macula	Central
4	6	Right convergent strabismus Abnormal retinal correspondence High hypermetropia	3	No	21	Between disc and macula	Central
5	7	Left convergent strabismus Normal retinal correspondence	4	Yes	8	Nasal border of macula	Central
6	8	Left convergent strabismus Abnormal retinal correspondence	1	Yes	14	Nasal border of macula	Central
7	6	Right convergent strabismus Anisometropia	1	No	12	Nasal to the fovea	Central
8	5	Right convergent strabismus Abnormal retinal correspondence	5/12	No	10	Between disc and fovea	Central
9	4	Left convergent strabismus Abnormal retinal correspondence	Uncertain	No	12	Between disc and fovea	Central with nystagmus
10	5	Left convergent strabismus Abnormal retinal correspondence High hypermetropia	3/12	Yes	16	Nasal to fovea	Central
11	5	Left convergent strabismus Abnormal retinal correspondence High hypermetropia	1	Yes	8	Nasal border of fovea	Central
12	7	Left convergent strabismus Abnormal retinal correspondence Left hypertropia	From birth	Yes	20	Nasal to fovea	Central with nystagmus
13	10	Right convergent strabismus Abnormal retinal correspondence High hypermetropia	From birth	Yes	35	Nasal border of disc	Parafoveal
14	5	Right divergent strabismus	8/12	Yes	16	Temporal to fovea	Central
15	6	Left convergent strabismus Lack of normal retinal correspondence High hypermetropia	From birth	No	28	Temporal to disc	Central with nystagmus

* High hypermetropia = more than +3D sph.

II
EX ANOPSIA WITH ECCENTRIC FIXATION
AFTER-IMAGE METHOD

Visual Acuity of Affected Eye						Binocular Function
Before Treatment		After Treatment				
Distance	Near	With After- Image	Angular	Cortical		
				Distance	Near	
5/60	None	6/18	6/24	6/60	N.24	None
6/36	N.36	6/4.5	6/9	6/12	N.5	Can alternate
6/24	N.10	6/6	6/9	6/12	N.5	Fusion and stereoscopic vision
Counting fingers at 30 cm.	None	6/18	6/36	6/36	N.6	None
6/24	N.24	6/9	6/9	6/12	N.5	Weak fusion No stereoscopic vision Seems straight on cover test
6/36	N.24	6/6	6/9	6/12	N.5	Can alternate
5/60	None	6/12	6/12	6/18	N.5	Can alternate for near
1/60	None	6/6	6/6	6/6	N.5	Fusion and stereoscopic vision
1/60	None	6/24	6/36	6/60	N.18	None Attendance insufficient
6/60	None	6/5	6/6	6/6	N.5	Fusion and stereoscopic vision Straight on cover test
6/36		6/6	6/12	6/18	N.5	Can alternate
6/36	N.10	6/4.5	6/18	6/18	N.5	Abnormal retinal correspondence
Counting fingers at 1 m.	None	6/36	4/60	4/60	None	None
6/36	None	6/6	6/9	6/9	N.5	Simultaneous macular perception
Hand move- ments	None	6/18	6/36	6/60	N.36	None Lack of normal retinal correspondence

Continued—

TABLE 1

Case No.	Age (yrs)	Diagnosis	Age at Onset of Strabismus (yrs)	Previous Treatment	No. of Treatments	Fixation of Affected Eye	
						Before Treatment	After Treatment
16	10	Left convergent strabismus Abnormal retinal correspondence High hypermetropia	24	Yes	41	Nasal to fovea	Central
17	10	Left convergent strabismus Abnormal retinal correspondence High hypermetropia	2	Yes	32	Parafoveal with nystagmus	Central
18	7	Left convergent strabismus Abnormal retinal correspondence	Uncertain	No	39	Nasal to fovea	Central
19	12	Right convergent strabismus	4	Yes	28	Nasal to fovea	Central
20	9	Right convergent strabismus Abnormal retinal correspondence High hypermetropia	1½	Yes	32	Temporal to fovea	Central with nystagmus
21	5	Right convergent strabismus Abnormal retinal correspondence High hypermetropia	1	No	27	Between disc and fovea	No real fixation
22	6	Right convergent strabismus Abnormal retinal correspondence Right hypertropia	16/12	Yes	25	Nasal to fovea	Central
23	5	Right convergent strabismus Abnormal retinal correspondence	23	Yes	7	Nasal to fovea Occasionally central	Central
24	7	Left convergent strabismus Abnormal retinal correspondence	?	Yes	33	Nasal border of fovea	Central
25	8	Left convergent strabismus Abnormal retinal correspondence	2	Yes	38	Nasal border of fovea	Central
26	8	Left convergent strabismus Abnormal retinal correspondence	1	Yes	34	Nasal border of fovea	Central
27	6	Right convergent strabismus Abnormal retinal correspondence	13	Yes	8	Nasal border of fovea	Central with nystagmus
28	9	Left convergent strabismus Abnormal retinal correspondence	?	Yes	15	Nasal border of fovea	Central
29	7	Left convergent strabismus Abnormal retinal correspondence	?	Yes	12	Nasal border of fovea	Central
30	10	Right convergent strabismus Abnormal retinal correspondence Anisometropia +5D astigmatism	2	Yes	53	Between disc and fovea	Central

I (continued)

Visual Acuity of Affected Eye						Binocular Function
Before Treatment		After Treatment				
Distance	Near	With After- Image	Angular	Cortical		
				Distance	Near	
6/24	N.36	6/4·5	6/9	6/12	N.5	Abnormal retinal correspondence
6/24	N.24	6/4·5	6/9	6/12	N.5	Superimposes slides First grade binocular vision only
5/60	None	6/6	6/12	6/24	N.8	Cannot alternate
6/18	N.18	6/4·5	6/6	6/9	N.5	Fusion poor
6/36	N.24	6/6	6/12	6/24	N.6	None
Counting fingers at 50 cm.	None	6/30	6/60	6/60	N.36	None
6/36	N.24	6/9	6/9	6/12	N.5	None
6/36	N.36	6/9	6/9	6/18	N.5	Can alternate
6/60		6/4·5	6/6	6/12	N.5	Diplopia caused by treatment for abnormal retinal correspondence
6/24	N.24	6/6	6/9	6/12	N.6	Binocular single vision for near
6/24	N.18	6/4·5	6/12	6/18	N.5	Can alternate
6/36		6/12	6/12	6/18	N.6	Can alternate
6/36	N.18	6/6	6/6	6/6	N.5	None
6/24		6/4·5	6/9	6/12	N.5	Fusion
1/60	None	6/9	6/24	6/36	N.24	Slight fusion

Continued—

TABLE

Case No.	Age (yrs)	Diagnosis	Age at Onset of Strabismus (yrs)	Previous Treatment	No. of Treatments	Fixation of Affected Eye	
						Treatment Before	After Treatment
31	12	Left divergent strabismus (consecutive) High hypermetropia	5/12	Yes	47	Temporal to fovea	Central binocular nystagmus
32	6	Left convergent strabismus Abnormal retinal correspondence	1	Yes	5	Nasal border of fovea	Central
33	9	Left convergent strabismus Abnormal retinal correspondence High hypermetropia	2	Yes	31	Nasal to fovea	Central
34	13	Right divergent strabismus (consecutive)	2	Yes	37	Temporal to macula	Central occasionally
35	10	Left convergent strabismus Lack of normal retinal correspondence	24	Yes	22	Nasal border of fovea	Central
36	6	Left convergent strabismus Abnormal retinal correspondence	6/12	Yes	6	Nasal border of fovea	Central
37	7	Right convergent strabismus Abnormal retinal correspondence	From birth	Yes	38	Nasal to disc	Central (unsteady)
38	8	Left convergent strabismus Abnormal retinal correspondence	1	Yes	18	No real fixation in fovea	Central binocular nystagmus

power to cooperate, in order to provide him with a high standard of visual acuity in each eye at the earliest possible age. Thus, he will have a reserve eye if he ever has the misfortune to lose the vision of one eye through trauma or some organic lesion. The probability of subsequent deterioration of vision in the originally amblyopic eye must, however, be borne in mind. Only in those cases in which restoration of binocular single vision, or some form of subnormal binocular vision, has occurred, or in those rendered freely alternating, can it be certain that there may not be some subsequent deterioration of the visual acuity gained in this way. However, if adequate treatment is given, it is unlikely that there will be a return to eccentric fixation once central fixation has been established, and it is unlikely that the visual acuity developed will later become grossly reduced. Follow-up examination of these cases will be needed in order to find this out.

Summary

Fifty cases of amblyopia were treated by Cüppers's after-image method; 38

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II (continued)

Visual Acuity of Affected Eye						Binocular Function
Before Treatment		After Treatment				
Distance	Near	With After-Image	Angular	Cortical		
				Distance	Near	
5/60		6/12	6/18	6/24	N.14	Fusion Binocular single vision at times
6/24	N.10	6/6	6/9	6/9	N.5	None
6/36	N.24	6/6	6/12	6/18	N.6	Can alternate
2/60		6/24	6/60	6/60	N.24	None
6/36	N.24	6/4.5	6/9	6/12	N.5	First grade binocular vision only
6/24	N.24	6/4.5	6/6	6/6	N.5	Can alternate
3/60	None	6/9	6/36	6/36	N.36	None
6/24		6/9	6/9	6/12	N.5	Can alternate

showed eccentric fixation in the amblyopic eye and the remaining twelve had central fixation. The vision in 38 cases improved to 6/12 and N.10 or better after treatment.

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