ANOMALOUS SENSORY RELATIONSHIP IN APPARENTLY CURED SQUINTS*

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DURING the last few years it has been found that many patients presumed to have normal binocular single vision after optical, orthoptic, and/or surgical treatment, have in fact a small angle of anomaly. From certain points of view this is not of great importance, as such cases are satisfactory cosmetically, and show good visual acuity in each eye with a limited degree of fusion detectable by certain tests. On the other hand, the amplitude of fusion is invariably restricted and true stereopsis is absent, and this is an unfortunate state of affairs in these days when normal binocular function is required in an increasing number of professions.

Even if (in common with some authors) we decry the value of stereopsis, there remain the risks associated with a small range of fusion—secondary divergence and the gradual establishment of amblyopia.

It is not unusual to encounter heterophoria of as much as twelve degrees, which, if associated with good fusion, causes no subjective symptoms. By contrast, quite low degrees of heterophoria, if associated with lack of fusion, may lead to intermittent or permanent decompensation.

This is not meant to imply that our standards are so high that our poorest results are those in which the residual angle is so small as to be hardly detectable by the ordinary methods of measurement; on the contrary, we classify such cases as "successful failures". An analysis of these results, however, suggests that perhaps these small residual angles might have been avoided, and perhaps also that, from the strictly sensory viewpoint (because it is harder to improve the sensory relationship in a case with a small angle of anomaly than in one with a large angle), we have made matters worse by treatment.

Methods of Investigation

Tests for Unioocular Fixation and Localization by Visuscopy

Before applying tests for binocular anomalous retinal correspondence (A.R.C.), it is necessary to investigate the unioocular fixation and localization, since a visual
SENSORY RELATIONSHIPS IN SQUINT

acuity of 6/12 or even 6/9 in the squinting eye (produced by occlusion of the sound eye) does not always exclude eccentric fixation. Cuppers (1958) has described two methods—active and passive—which may be used to detect eccentric fixation and localization by uniocular visuscopy.

Active.—The patient is asked to fix the star of the Visuscope. This allows the observer to determine, by the position of the black star on the fundus, whether the foveola or some other eccentric retinal point is being used for fixation.*

Passive.—The patient is asked to localize either the star of the Visuscope, or the centre of the light emitted by it, or the black spot of the Euthyscope, when any one of these is placed passively on the fovea.

The first stage in this test is to determine by active uniocular visuscopy which retinal point is used for fixation. The second stage is to ascertain whether this point corresponds to the principal visual direction as determined by active or passive uniocular visuscopy.

Disparity between these two tests is rare, but may sometimes be found in cases in which there is a scotoma of the foveola, fovea, or macula, usually extending as far as the optic disc (Fig. 1), where previous treatment by occlusion and/or surgery has been given.

The prognosis and treatment depend on whether the point used for fixation and the principal visual direction correspond exactly.

In determining the principal visual direction by active uniocular visuscopy, the patient is asked "to look straight ahead at the star even if he does not see it clearly"; it may then happen that he fixes the star with the foveola. This may be checked by passive uniocular visuscopy; the star is projected passively on to the foveola, and the patient may still have the subjective impression of seeing the star straight ahead. If the scotoma is so dense that the star is not seen, then one of the black spots in the Euthyscope or even the light of the Visuscope may be projected on to the fovea in the same way, and the patient is asked if he sees straight ahead or not. If, as in the previous instance, the patient sees the star of the Visuscope (or the black spot of the Euthyscope, or the light of the Visuscope)

* This point for fixation may also be determined by campimetry (Brockbank and Downey, 1959). It must not be confused with the real point of fixation which is the point that bears the principal visual direction. The former may be a retinal point on the border of a scotoma which, being outside it, has a higher resolving power.
straight ahead when the examiner sees it centred on the foveola then the fixation and the principal visual direction are considered to be foveolar. If the star is not seen straight ahead, the fixation and the principal visual direction are eccentric, and it is important to determine their position and direction and to know whether or not they correspond. For example, if a left eye has a temporal fixation on examination by active uniocular visuscopy but the patient subjectively localizes the star to the right, and if when the star is “placed” passively on the foveola the patient still localizes it to the right, then the principal visual direction is nasal. Thus, when uniocular diplopia is stimulated during the course of treatment, it is essential to know which image belongs to the fovea. In the case just mentioned, the left image would be the correct foveal image, but if the retinal point bearing the principal visual direction was either foveal or temporal (viz: the same point as that of fixation) then the correct foveal image would be to the right. With regard to prognosis, if an eccentric retinal point is used for fixation as well as being the “bearer” of the principal visual direction, then the treatment will be much more difficult than if the fovea still maintained its physiological principal visual direction.

The results of this uniocular test must be carefully compared with the results of the two following tests for binocular sensory correspondence (see below “Binocular Visuscopy”, and “Giessener Clinic Test”). It has been suggested that many cases of eccentric fixation have originated from abnormal retinal correspondence. Therefore, if the uniocular and binocular anomalies are identical, the treatment will be difficult because of their deep-seated and closely-linked development. This is particularly so in older patients; in those over 10 years of age, treatment devised to eradicate the sensory binocular anomaly should be undertaken only if circumstances are favourable, and in those over 14 years but rarely. On the other hand, if the uniocular and binocular anomalies are not identical, then the treatment may be undertaken with more hope of success, unless, of course, the uniocular eccentricity is very near to the fovea and/or the binocular angle of anomaly is small.

Detection of Small Angles of Anomaly

Small angles of anomaly, which are often the consequence of badly planned therapy, may be the result of an error committed in one of the three usual forms of treatment, optical, orthoptic, or surgical. The following four methods describe how to detect a small angle of anomaly. Possible optical, orthoptic, and surgical causes will then be discussed in general.

(1) Binocular Visuscopy (Cüppers) (Fig. 2, opposite).—The patient is placed at 2 m. from, and at right angles to, the centre of the Maddox tangent scale. Although it is not always necessary, both pupils should be dilated by the use of a mydriatic. The patient holds a small plane mirror in front of one eye at a convenient angle and is asked to fix the Maddox spotlight reflected in the mirror. The examiner then projects the Visuscope star on to the fovea of the non-fixing eye. If the patient says that the star overlies the light, the retinal correspondence is normal; if the star appears to one
side of the light, then the number on the scale on which the star appears to lie gives the angle of anomaly.

In the latter case, as a control, the star is moved across the fundus until the patient says it coincides with the light, and the point on the retina where the coincidence occurs is noted. These observations provide a check on each other.
This test should be used only where foveolar fixation exists,* and it should be carried out with both eyes fixing in turn.

(2) Giessener Clinic Test (Fig. 3)—The patient faces the scale and fixes the Maddox spotlight with the fixing eye through a dark red lens that allows only the light to be seen. An X-shaped after-image has been previously produced in the other eye (with an electronic flash) if the Visuscope has shown fixation to be foveolar; otherwise a "euthyscopic" after-image is created. This after-image is stimulated by alternately lighting and darkening the room, or by temporary intermittent occlusion of the non-fixing eye.

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* This test may be done in when the squinting eye has an eccentric fixation, but then only with the non-squinting eye fixing.
It is now impressed on the patient that he should concentrate on fixing the red light only, and should ignore the white light or the after-image.

The patient now sees a red spot over the figure on the scale which demonstrates the subjective angle of deviation, because, although perceived by the fixing eye, localization of the "red" light is made by the retinal point of the non-fixing eye corresponding to the foveola of the former, and the after-image created at, and seen by, the foveola of the squinting eye is seen over the figure corresponding to the objective angle of the squint.

In young children it is preferable to ask the patient to walk up to the scale and to point to the place where the objects appear to be situated.

Where the angle of anomaly is believed to be small, it is necessary to repeat the test at 5 or 6 metres from the tangent scale.

The test should also be repeated with each eye in turn, unless an eccentric fixation has been detected in the squinting eye.

(3) Foveolar Synoptophore Slides.—Small angles of anomaly may remain undetected when using conventional slides in the synoptophore. Those of Jampolsky, which have a very small central object, may detect these small angles of anomaly. We have verified this with home-made synoptophore slides with central pictures of 1·5 mm. subtending 45' (Fig. 4).

![Fig. 4.—Foveolar fusion slides. (Charlie Chaplin with walking-stick and flower as controls.)](image)

(4) After-Images, Haidinger's Brushes, and Synoptophore.—After-images and Haidinger's brushes in the synoptophore, as proposed by Küppers, will allow the detection of minute angles of anomaly, and are a helpful addition to the treatment of abnormal retinal correspondence. The use of a more powerful light source than usual* has made it possible to combine Haidinger's brushes with after-images and specially designed slides (Figs 5 and 6, overleaf).

* For simultaneous use of Haidinger's brushes and after-images, a double filament is employed— one filament is permanently incandescent, whilst the other is connected to a flashing device.
Where the fixation is known to be foveolar in both eyes, before measuring the abnormal retinal correspondence, the relation of the two after-images created in each eye consecutively is first determined*; and secondly, the relationship of the two after-images to each other by the introduction of a Haidinger brush. If the correspondence appears subjectively normal for these test objects, an objective control of their localization may be made by the decreasing field test.†

* Classically a vertical and a horizontal linear after-image are used, but this makes it very difficult for the patient to estimate small discrepancies. It is suggested that a cross and an X-shaped after-image be used.
† The part of the field which is known to correspond to the anomalous localization present before treatment is occluded for the non-fixing eye, and if Haidinger's brushes disappear then localization is not yet acquired for this object.
SENSORY RELATIONSHIPS IN SQUINT

499

As it is not possible to measure the angle of anomaly with Haidinger’s brushes or after-images, it is necessary to repeat the test with real objects. It is impossible for a child to determine with exactitude the distance between two such objects as after-images or Haidinger’s brushes without some point of reference such as a marked scale.

The objective angle having previously been measured, the tube of the synoptophore is placed at 0° before the fixing eye. A vertical Hering after-image is created in the non-fixing eye. A Maddox tangent-scale slide is placed before the fixing eye. The patient is then asked, whilst fixing the zero mark in the middle of the scale, where the after-image falls. A small foveal or macular triangle is then placed before the non-fixing eye and its position is noted. If the after-image, alone or in combination with the triangle, falls on the zero mark, then the correspondence is normal. If these two objects fail to coincide abnormal retinal correspondence is present*. The fixity of this can be determined if the after-image and the triangle coincide, or if there exists a different angle of anomaly for each object. If the angle of anomaly is the same for each object, the abnormality is more difficult to cure.

This test must be repeated, with the non-fixing eye fixing the zero of the Maddox tangent scale and another vertical after-image before the previously fixing eye.

Anomalies of Small Degree caused by Faulty Treatment

(1) Optical Correction.—Optical under-correction is uncommon in England where refraction is performed routinely after 5 days of atropization. In some Continental clinics, however, refraction is still performed under weak cycloplegia, or after a short period of instillation. This involves a serious risk of under-correction in cases of hypermetropia with accommodative esotropia, which may leave a small but constant deviation, which in turn causes a small angle of anomaly. The same thing may occur (in spite of a full hypermetropic correction) in patients who maintain a small deviation because the strabismus is not entirely accommodative. It is important, therefore, to measure the deviation at the outset with the patient wearing the full correction for any refractive error. If a deviation persists and the correspondence is anomalous, occlusion should be employed, but if the correspondence is normal, surgery (e.g. bilateral medial rectus recession) should be considered.

(2) Orthoptic Treatment.—Unless after-images or Haidinger’s brushes are employed to determine the nature of the retinal correspondence, there is always a danger of a small angle of anomaly developing. This danger is considerable where the treatment of abnormal retinal correspondence is undertaken by stimulation from the subjective angle in order to reach the

* It will be realized that the after-image may show a normal correspondence, but that an abnormal retinal correspondence may be present for real objects (i.e. Maddox scale and triangle).
objective angle, or from an angle that is intermediate between the two. In order to avoid this danger, it is advisable (as Cüppers has suggested) to use after-images and Haidinger's brushes for control and treatment of the abnormal retinal correspondence, suppression, and fusion, particularly where abnormal retinal correspondence has previously existed.

(3) **Surgical Treatment**.—An almost perfect surgical result is often worse, from the sensory point of view, than an obvious failure; a small angle of anomaly may result from an early operation which leaves a small deviation. By “early operation” we mean those performed before the age of 2 years.

Those who advocate early surgery (before 2 years, 18 months, or even at the age of only a few months) use a very simple, and at first sight very attractive, argument: “Put the eyes straight and binocular vision will develop under normal conditions as it does in non-squinters”. Such a policy could only be justified if, on the one hand, this statement was correct, and on the other if surgical results were a 100 per cent. perfect.

If we admit the proposition of Keiner (1951) that in concomitant strabismus there is a delayed “myelinization of the central nervous system”, or that of Burian (1958) that there is an “alteration in the sensory Anlage”, or if (with von Graefe, 1855, Hering, 1879, and Graefe, 1897, amongst many others), we consider foveal fixation and normal correspondence to be innate (a hypothesis also favoured by Cüppers), then early surgery would appear to lack a theoretical basis.

Even if we accept theories that advocate early surgery, a perfect surgical result would still be a *sine qua non*. This we believe is almost impossible for even the most skilled surgeon to guarantee.

Still more important is the fact that it is humanly impossible to investigate the motor anomalies in a child under 2 years of age. Motor anomalies are sometimes complex and need to be studied with great care. In cases of paralysis, which are non-congenital, where the sequelae have not had time to develop, it is impossible to estimate their importance or their evolution without a succession of precise measurements.

No surgeon interested in a functional or even a perfect cosmetic result, would care to operate unless he at least had precise measurements of the horizontal and vertical deviations in the cardinal positions, fixing with each eye separately for distance and near, with and without correction. In children under 3 years of age even an accurate measurement in the primary position may be difficult to obtain.

Likewise, one cannot measure the post-operative angle with any accuracy, or even know whether so-called “orthoptization” is taking place with the eyes really parallel or with a small angle of deviation. Obviously an angle of 5° can be detected by several methods, but to find really small angles of anomaly one needs careful cooperation on the part of the patient. In many cases an “orthoptic” result has been attained when a vertical and/or
horizontal prism has been prescribed, though this is only justified when one knows the exact angle of deviation.

We are offering no statistics, for it is only latterly that we have been able to apply the methods described to patients who had operations before the age of 2 years. We have, however, found that (except in a few very fortunate patients with perfect binocular vision) the great majority have a small angle of anomaly. In these cases, the more perfect the surgical result, the harder it was to resolve the anomaly by orthoptic treatment. At a first glance they appeared to have a normal correspondence in the synoptophore with conventional slides. A certain amplitude of fusion existed with stereopsis, and four Worth's lights were seen. Nevertheless, the methods described showed a small angle of anomaly in these cases. One cannot help feeling, therefore, that these patients would have been much better off if occlusion had been continued and surgical treatment postponed until the child had reached the age where detailed examination was possible before treatment.

Conclusions

It is suggested that the best method of treating squint in young children before 3 or 4 years of age (depending on the behaviour of the child), is either eccentric alternating or unocular occlusion of the fixing eye, or (in cases of eccentric fixation) of the amblyopic eye, until a correct diagnosis of the sensory and motor anomalies is possible. If feasible, a certain amplitude of fusion should be attained before surgery.

It appears to us that a sensory anomaly persisting after surgery is more harmful than the possible motor limitations. Early operation, therefore, would seem to be contraindicated by the subsequent danger of a small angle of anomaly. The slightly greater risks of general anaesthesia in children under 2 years of age should also be born in mind.

Early surgery may, however, be indicated in the following cases:

(a) When the cooperation of the parents is completely lacking. In this case it is perhaps preferable to risk leaving the child with a small angle of anomaly with a certain degree of peripheral fusion, rather than leaving a definite unilateral strabismus.

(b) When a very large angle of deviation is present.

(c) When obvious vertical anomalies associated with a definite torticollis may lead to eventual skeletal alterations.

(d) When the eccentric fixation is obviously linked to a paresis of a muscle (generally a lateral rectus).

Apart from these indications, until a guarantee of a good fusion has been assured, occlusion, with the full optical correction, should be continued in young children with squint, with careful testing by the methods outlined until the child is old enough to undertake exercises and/or is ready for operation.
SUMMARY

By the use of tests combining the Visuscope, Euthyscope, after-image, and Haidinger's brushes (which are described in detail) the authors have detected small angles of anomalous retinal correspondence in many apparently successful cases of cured squint.

These anomalies are so small as to be undetectable by conventional synoptophore slides. It is concluded that, as early surgery is particularly liable to lead to these small angles of anomaly (which are resistant to further treatment), its use should be limited to the small number of cases in which it is strongly indicated.

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REFERENCES