

DIFFERENTIAL DIAGNOSIS AND AN OUTLINE OF THE MANAGEMENT OF SENSORY ADAPTATIONS IN STRABISMUS

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Sensory adaptations occur very readily in the child under seven years who has a constant manifest strabismus. A better understanding of these adaptations aids the making of a prognosis and therefore will affect the management of the condition. Much has been written on this subject by ophthalmologists and orthoptists from all over the world. A few of the theories and the methods of management appear to conflict. However, when studying this subject in depth the greatest area of disagreement seems to be in terminology.

The aim of this paper is to collate the facts so that the student, who is embarking on the study of strabismus, can make a differential diagnosis, assess the prognosis and have some guide to the management of these cases. The inspiration to write such a paper came from the Symposium on Strabismus held in Cambridge, U.K. in September, 1973 where two authorities on sensory adaption in squint, von Noorden and Lang, agreed in their terminology and classification of microstrabismus.

The British definition of abnormal retinal correspondence is "a binocular condition in which there is a change in the visual direction of the retina, such that the fovea of the fixing eye has a common visual direction with an area other than the fovea of the deviating eye. The condition may occur which ever eye is used for fixation." This implies that each eye is capable of using the fovea for fixation. The fixing fovea would have therefore the principal visual direction, while the spatial values of the deviating eye have all shifted (nasally in convergent and temporally in divergent strabismus.)

Abnormal retinal correspondence can be classified as follows:

1. Microstrabismus without identity. Always unilateral - a small angled esotropia of less than 5° . Small amount of amblyopia present.
2. Convergent squint of 5° to 10° . Usually unilateral. Small amount of amblyopia present.
 - a. Harmonious A.R.C. with anomalous fusion and gross stereopsis. Usually claims four Worth's lights and a diagonal cross with the striate glasses. Cosmetically acceptable.
 - b. Unharmonious. Squint has an associated phoria. Often suppression on binocular tests.
3. Convergent squint of more than 10° , with early onset and often alternating, but can have varying amount of amblyopia. Vision in the deviating eye can be as little as 6/60, though this is rare as by definition each fovea is capable of fixing and this amount of amblyopia is usually associated with inability to fix centrally.
 - a. Harmonious - rare.
 - b. Unharmonious. Often has suppression on binocular tests.
 - c. Lack of N.R.C. but patient claims that slides cross at a less convergent angle compared with the objective measurement, that is, showing a tendency to A.R.C.
4. Small angled divergent squint. This is rare. Usually cosmetically acceptable.

Orthoptic Investigation of Arc

When investigating a patient with A.R.C. the case history, cover test and visual acuity provide some indication of the condition. A well established A.R.C. is often associated with a unilateral convergent strabismus of under 12° where the onset occurred around the age of $2\frac{1}{2}$ years. (The significance of this is that the patient has already been used to some binocular single vision.) The visual acuity of the deviating eye is normal 6/18 or better.

Using the visuscope there is central (foveal) fixation of either eye. When the star is superimposed objectively on the fovea of the one eye and with both eyes open, the patient will claim the star appears to the side of the fixation object (red light seen through mirror by the other eye).

On the synoptophore using simultaneous macular perception slides the objective and subject angles are compared. The subjective angle is always less than the objective angle. The difference between these measurements is known as the angle of anomaly. The angle of anomaly can be only 2 degrees or as much as 10 degrees. If the subjective angle is at 0° the angle of anomaly equals the angle of squint: this is known as Harmonious A.R.C. Usually these cases have a squint of 10° or less with more or less equal visual acuity. The smaller the angle of anomaly the more likelihood of the patient being able to do binocular tests. Therefore, in spite of not fixing the fusion slides bifoveally, the patient experiences a type of fusion with a range whilst seeing both controls. He experiences depth perception with the simple stereoscopic slides and also when viewing the fly on the stereotest. He will see 4 Worth's lights, a true cross with the striate glasses and he may claim physiological diplopia and be able to bar read.

In other words, complete adaptation has occurred to the squinting condition. The patient looks cosmetically satisfactory with good visual acuity and with binocular functions (though abnormal).

However, more frequently on the synoptophore, the angle of anomaly is less than the angle of squint. This is known as Unharmonious A.R.C. These cases sometimes have anomalous fusion but suppression can be a barrier. The patient may claim five Worth's lights and homonymous diplopia with red and green glasses and a spot light. However, when a correcting prism is used there is heteronymous diplopia (paradoxical diplopia). When asked to join the diplopia with prisms, a prism less strong than the correcting prism is chosen. (C.F. the synoptophore).

With the striate glasses usually two lights are seen with the streaks crossing above or below. The response in the after image test in cases of manifest squint with normal retinal correspondence is a true cross because both foveae are stimulated in turn. However, the response with A.R.C. is a T on its side, because the spatial value of one fovea has changed. (NOTE: The patient must be able to fix the centre of each bright light with the fovea for this test to be valid.)

Orthoptic Management of A.R.C.

Occlusion is given to all types if under the age of eight years in order to obtain the maximum visual acuity.

Type 1: Microstrabismus. No treatment. Kept under observation until 9 years, then discharged. Cosmetically excellent.

Type 2: Small convergent squint. No treatment. Might need intermittent occlusion. Kept under observation until 9 years then discharged. Cosmetically acceptable.

Type 3: Moderate to marked convergent squint. Probably not cosmetically acceptable, therefore, surgery advised.

- a. If harmonious A.R.C. present no pre-operative treatment is given. The position is assessed after surgery.
- b. If unharmonious A.R.C. present and the child is under seven and the visual acuity is equal, pre-operative treatment can be given to try to re-establish bifoveal fixation post-operatively.
- c. If only signs of attempting A.R.C. and the child is under seven and the visual acuity is equal, pre-operative treatment should be given to try to re-establish bifoveal post-operatively.

After surgery the situation is assessed. Several of these cases develop into a microstrabismus. The advantage of having a well developed A.R.C. is that consecutive divergence is unlikely to occur. Amblyopia is unlikely to recur if the patient has been supervised until 9 years old.

Some work is being carried out, particularly on the continent of Europe using prisms with these cases. One method is to over correct the angle so that the patient is artificially divergent. The theory is that the abnormal projection will not, therefore be used. Often very strong prisms are required.

Type 4: Small divergent squint. No treatment. Kept under observation until 9 years, (noting if there is any increase in divergence). Cosmetically acceptable.

Orthoptic Investigation of Eccentric Fixation

The British definition of eccentric fixation is "a unocular condition in which there is fixation of an object by a point other than the fovea. This point adopts the principal visual direction." That is to say the spatial values have changed in the amblyopic eye only. This change is present even if the non-amblyopic eye is occluded. In the investigation of patients with eccentric fixation, it will be found that the visual acuity is never equal, but can be as good as 6/9, i.e. where the fixation is parafoveal. Diagnosis is made using the visuscope. The fixation can be i) loss of central, that is wandering, ii) unsteady eccentric, iii) steady eccentric. The last has the poorest prognosis for the re-establishment of foveal fixation. Binocular visuscopy will show the degree of adaptation, that is to say the strength of abnormal binocular function.

Because the amblyopic eye cannot fix with the fovea the angle of squint is measured by the prism reflection test or by using corneal reflections on the synoptophore. Abnormal binocular function can also be assessed using the latter instrument. The nearer the fixation is to the fovea, the better the vision, which results in a stronger binocular link between the two eyes. If an instrument with Haidinger's brushes is available it can be used diagnostically to see if the patient can recognise the brushes with the amblyopic eye. Recognition is a good prognostic feature. There has been much discussion on the method of treatment of eccentric fixation especially the significance of occluding the amblyopic eye (inverse occlusion). The treatment generally accepted in Britain today is to occlude the fixing eye for one week and then to reassess the fixation of the amblyopic eye. If it is now foveal, or unsteady foveal, or wandering over the foveal area then this type of occlusion is continued until alternation occurs or there is no improvement in visual acuity over a period of six weeks. If however, there is not this type of fixation then inverse occlusion is advised for children under the age of seven. This is worn until the fixation becomes foveal. It is unusual to use inverse occlusion for longer than six months. If after this time, the fixation is not foveal, then occlusion of the fixing eye is given to eliminate as much of the amblyopia as possible.

Many orthoptists like using inverse occlusion as it helps the patient initially to cope with the psychological distress of wearing an occluder without also having to adjust to having poor vision.

To encourage foveal fixation red filter treatment can be given for twenty minutes two to three times a day. In the orthoptic department pleoptics can be given if the instruments are available and the child will cooperate. Quite young children can be taught to look for Haidinger's brushes even if they are unable to localize them. This helps to eliminate foveal suppression.

Orthoptic Investigation of Microstrabismus

The definition of microstrabismus is of a small angled esotropia less than 10^{Δ} (5°) usually with abnormal retinal correspondence and amblyopia of varying degrees in one eye. Bifoveal fixation is always absent.

Its etiology should be considered before the classification can be understood.

Primary

1. Idiopathic
2. Pathological lesion at fovea
3. Pathological lesion in media of eye
4. Pathological rejection of bifoveal fusion (Parks)

Secondary

1. Decompensated primary microstrabismus
2. After surgical correction of larger squint
3. After orthoptic treatment, particularly the accommodative squint of the convergence excess type
4. Optical - mainly anisometropia or premature reduction in hypermetropic correction or failure to prescribe full correction

The classification of both primary and secondary microstrabismus can be divided into three:

- a. With normal retinal correspondence and a small central suppression scotoma of less than $4A$
- b. With abnormal retinal correspondence, each eye fixing with the fovea under monocular conditions. Micro-strabismus without identity.
- c. With eccentric fixation and changed spatial values in the amblyopic eye where the "angle of anomaly" equals the angle of eccentricity. Microstrabismus with identity.

All types can have an associated heterophoria although it is more likely to occur with secondary microstrabismus.

In the investigation of microstrabismus the history will give an indication of the differential diagnosis between primary and secondary. The visual acuity will never be equal. The results of the visuscope examination will provide the differential diagnosis between a, b, and c above. In cases of N.R.C. and true A.R.C. the cover/uncover test will reveal a very small convergent squint.

In cases of eccentric fixation where the angle of eccentricity equals the angle of squint there will be no movement.

In doing the alternate cover test (on all three types) any underlying latent component will be revealed. Thus, in some cases the very small manifest squint increases on dissociation. But once the cover is removed and abnormal binocular function can be resumed the eyes revert back to the very small angle. This gives an indication of how well established the abnormal sensory condition is.

The four diopre prism test can aid the diagnosis. When bifoveal fixation is present (normal B.S.V.), if a 4^Δ prism base out is placed in front of either eye, that eye will move inwards very slightly. This B.S.V. will be regained.

When a microstrabismus is present, if the 4^Δ base out prism is placed in front of the eye with the better vision, this eye will move inwards. However, the other eye will move outwards (Hering's law); there is a shift of both eyes in the same direction. Bifoveal fixation is not taking place. When the prism is placed in front of the amblyopic eye there will be no movement of either eye as the image still falls within the small central suppression scotoma.

The synoptophore is used to aid the differentiation of cases of N.R.C. from those with A.R.C. This instrument is not helpful if eccentric fixation is present.

The patient may complain of separation difficulties when reading. These are similar to the crowding phenomenon which often occurs when the visual acuity of an amblyopic eye is tested. If the first part of a word appears to blur, run together or disappear, there is left microstrabismus. If the end of a word is affected then right microstrabismus is present (if the middle of a word is affected then foveal suppression is occurring).

With the striate glasses, the response in B.S.V. with bifoveal fixation is a diagonal cross, intersecting at the spotlight. In microstrabismus the diagonal line seen by the amblyopic eye is not complete. It has a central gap on each side of the spotlight. Unfortunately this defective cross is difficult for a child to recognise or to describe. The more accurate way of diagnosing this phenomenon is to allow him to keep looking through the striate glasses at the light whilst he draws exactly what he sees.

In spite of having subdivisions in microstrabismus, the management is the same for each type. As patients are cosmetically very satisfactory all the orthoptist can do is to try to obtain and maintain the best possible vision in both eyes. Total occlusion of the fixing eye in all cases under the age of 9 years is given. The occlusion is stopped once there has been no improvement whatsoever for four weeks. Care has to be taken that the patient does not get diplopia. This occasionally happens when the suppression associated with the amblyopia is reduced.

If the patient has symptoms due to an associated convergence insufficiency or decompensated heterophoria, orthoptic treatment can be given along orthodox lines. Similarly, if a convergent squint is noticeable on removal of the hypermetropic correction these patients can be taught to relax accommodation and to straighten the eyes.

Summary

The investigation and brief outline of the management of cases with A.R.C. or eccentric fixation have been considered, with particular reference to microstrabismus.

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