12 OVERVIEW OF SENSORY CHANGES IN STRABISMUS

Binocular sensory changes in strabismus

Diplopia and confusion

Diplopia occurs when a patient sees two images of one object. Figure 12.1A represents an adult with recent-onset left esotropia. The patient is viewing an isolated letter A with no other objects present in the field of view. The letter is imaged on the right fovea (f) but, because the left eye is convergent, it is imaged on a region of the left retina (p) that is not the fovea. In other words, the object is imaged on non-corresponding retinal points. Therefore, the object is perceived in two different visual directions, causing diplopia.

Everyday visual scenes are usually more complicated than the single object in Figure 12.1A. Figure 12.1B illustrates the situation, for the same patient, when there are two isolated objects in the visual field (of course, this is still an unrealistically simple example). The letter A is imaged on the fovea of the right eye and the letter B is imaged on the fovea of the left eye. Since the case is a recent-onset strabismus in an adult patient, the patient is likely to have normal retinal correspondence (NRC). This means that both foveae share the same visual direction, so the patient will see the two letters as being superimposed. The visual perception is described as *confusion*. Of course, the diplopia illustrated in Figure 12.1A would also be present in the situation illustrated in Figure 12.1B so, in normal everyday scenes, both diplopia and confusion will coexist. Depending on the visual scene and the magnitude of the separation of the images, diplopia may be more troublesome than confusion.

Suppression of the binocular field of the strabismic eye

Clearly, diplopia and confusion are undesirable and the visual system might be expected to develop sensory adaptations to avoid diplopia and confusion. In young patients, this is what happens. Hypothetically, one

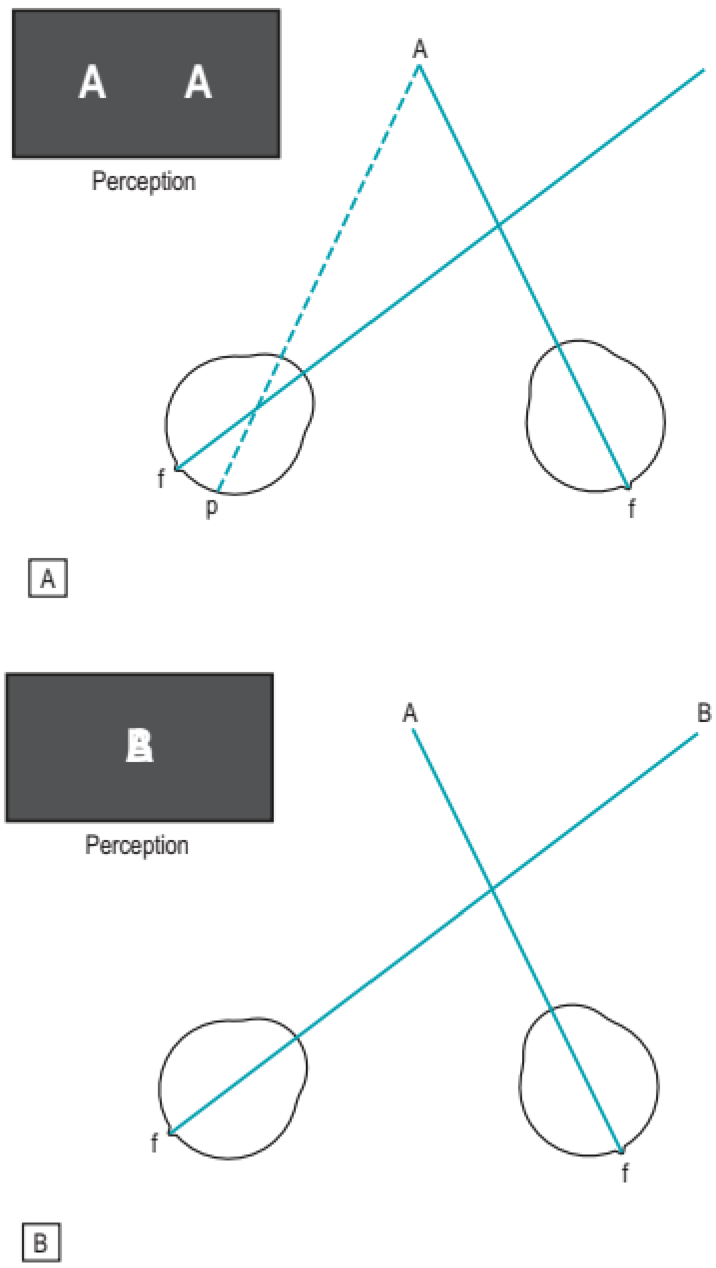

Figure 12.1 Illustration of (A) diplopia and (B) confusion in left convergent strabismus.

method of avoiding symptoms in strabismus might be to suppress the whole of the binocular field of the strabismic eye. This sometimes occurs (Joosse et al 2005), particularly in divergent strabismus (Ansons & Spencer 2001), and the investigation and treatment of suppression is detailed in Chapter 14. However, the visual system usually does not adopt such wasteful measures. Instead of having a large area of suppression, a strabismic patient who is young enough to have a reasonable degree of sensory plasticity usually will develop harmonious anomalous retinal correspondence (HARC). Suppression and HARC are fundamentally different and elicit different steady state visual evoked potentials (Bagolini et al 1994).

Anomalous (abnormal) retinal correspondence

The classical views on Panum's fusional areas and retinal correspondence have, as a result of research over the last 20 years, undergone much revision. The phrase 'corresponding retinal points' is something of a misnomer: a point image on one retina actually corresponds with point images falling in a Panum's area in the other eye. Several researchers have shown that Panum's area is not a fixated entity but that its size varies according to the parameters of the target. What remains unclear is whether, at a given retinal eccentricity, the size of Panum's area really changes or whether apparent changes are experimental artefacts.

Several studies have obtained data that have been used to argue that retinal correspondence can change in normal, non-strabismic observers (Fender & Julesz 1967, Hyson et al 1983, Erkelens & Collewijn 1985, Fogt & Jones 1998, Brautaset & Jennings 2006a) or that Panum's fusional areas are much larger than previously believed (Collewijn et al 1991). However, one very thorough paper has concluded from two experiments that retinal correspondence is fixed in non-strabismic observers (Hillis & Banks 2001).

There is certainly the need for some flexibility in the vergence system since, during everyday vision and head movements, small errors in vergence occur: one eye's visual axis may become misaligned with the object of fixation. This is particularly likely to happen after a large saccade and represents a small breakdown in Hering's law. This is probably why our visual system has evolved to have Panum's fusional areas rather than inflexible point-to-point correspondence.

Anomalous retinal correspondence in strabismus

In non-strabismic people, NRC can tolerate small vergence errors without losing fusion or stereopsis. This impressive feat of cortical processing is far surpassed by the ability of children, who are young enough to possess considerable neural plasticity, to exhibit large shifts in retinal correspondence to compensate for strabismus. The purpose of this abnormal retinal correspondence (ARC) is for a point on the retina of the good eye to correspond with a new point in the retina of the strabismic eye (not its natural, innately, corresponding retinal point). Clearly, the newly corresponding points should be set at the angle of strabismus. This is nearly always the case in ARC and there is said to be *harmonious anomalous retinal correspondence* (HARC). The angle through which the retinal correspondence has been shifted from the normal is called the *angle of anomaly*. The term anomalous *retinal* correspondence has been criticized because the abnormal correspondence occurs cortically, not on the retinae. Despite this semantic objection, it is often easier to conceptualize the effect of the HARC by considering retinae, so the convention will be followed here.

Research in non-strabismic subjects to investigate the largest amount of visual disparity that can still provide depth information may help to understand the basis for HARC (Dengler & Kommerell 1993). All the subjects who were tested could recognize disparities of up to 6°, and one up to 21°, without making compensatory vergence eye movements. It is possible that far-reaching interocular connections in normal subjects might also be utilized in cases of strabismus (Dengler & Kommerell 1993), although it should be noted that HARC is uncommon in vertical strabismus (von Noorden 1996).

The precise mechanism of HARC remains unclear. One view is that remapping of Panum's areas occurs (Lie et al 2000). Another view is that Panum's areas become enlarged. A third hypothesis is that in HARC the bifoveal assumption is abandoned and the position of each eye is registered separately, probably on the basis of muscle activity (Walls 1963, cited by Jennings 1985). This form of HARC would be most likely to facilitate the perception of direction, not depth and distance. It might account for HARC in large-angle strabismus, with the 'cortical remapping hypothesis' accounting for HARC in cases of small-angle strabismus (Jennings, personal communication).

To summarize, there are three types of binocular sensory status in strabismus. First, there may be no adaptation and diplopia and confusion result. Second, all the binocular field of the strabismic eye may be suppressed. Third, HARC may occur. The third option allows some rudimentary form of 'pseudobinocular vision' and is clearly the preferable outcome, so the question arises of why this does not always occur. This, and some limitations and consequences of anomalous retinal correspondence, will now be considered.

Factors influencing the development of HARC Although the precise neurophysiological basis of HARC is not known, the main theories all accept that this 'stunning feat of cortical processing' (Nelson 1988b) must inevitably have certain limitations. One of these limitations relates to the requirement for the visual system to be plastic for HARC to develop. It is therefore not surprising that a younger age of onset of strabismus is associated with a greater likelihood of HARC being present. Von Noorden (1996) states that HARC, albeit superficial (see below), can develop in the early teenage years. A survey of 195 patients by Stidwill (1998, p 41) found that although the condition was occasionally present in strabismus developing up to the age of 15, 97% of cases of HARC had developed in strabismus with an onset before the age of 6 years.

In cases of intermittent strabismus the visual axes will sometimes be straight and the patient will have NRC, yet at other times there will be a strabismus and the patient will have HARC. The change from NRC to HARC can be sudden or gradual. The term covariation describes the situation when the angle of anomaly covaries with the objective angle of strabismus. Covariation is likely to place additional neural demands on the visual system and hence constant strabismus will be more likely to develop HARC than intermittent or variable strabismus. For similar reasons, unilateral strabismus is more likely to develop HARC than alternating strabismus.

Von Noorden (1996, p 267) stated that the rate of occurrence of HARC 'is high in infantile esotropia, less common in exotropia, and uncommon in vertical strabismus'. Other authors have noted that suppression is more common in exotropia and in anisometropia.

Photoreceptor types, receptive field sizes and ganglion cell types vary across the retina. One degree of retina near the fovea has a much greater cortical representation than one degree in the periphery: this has been termed *cortical magnification*. The cortical processing task of remapping anomalously corresponding points must be easier if these points are at similar eccentricities from the fovea. Hence, small-angle strabismus is more likely to develop HARC than large-angle strabismus (Wong et al 2000).

Depth of HARC Patients who exhibit HARC can, under certain circumstances, be made to exhibit NRC. In other words, the neural substrate for innate NRC is still present. The difficulty in eliciting NRC is termed the 'depth of anomaly' (Nelson 1988b). The factors that make it easier for the visual system to develop HARC are also likely to make the HARC deeper. Therefore, it follows from the previous section that patients are more likely to have deeper HARC if there is younger onset (Kora et al 1997), a stable angle of strabismus, unilateral strabismus and a small angle.

The detection and treatment of HARC ARC can be thought of as 'pseudobinocular vision'. It was noted in Chapter 3 that a patient with weak normal binocular vision (e.g. a decompensating heterophoria) could, by using tests that tend to dissociate the eyes, be 'broken down' so that the heterophoria degenerates into a strabismus. An analogous phenomenon can occur with shallow HARC. If a patient with shallow HARC is tested with unnatural stimuli, such as after-images or the synoptophore, the pseudobinocular vision may be broken down into NRC, with resulting diplopia or compensatory suppression. If more natural, 'associating' tests are used, such as Bagolini lenses (Ch. 15), then HARC may be detected. This is why, if the practitioner is to discover whether HARC is truly present under normal everyday viewing conditions, naturalistic tests should be used. The factors that are particularly important in simulating normal visual conditions are listed in Chapter 14.

The likelihood of treatment succeeding is influenced by the depth of HARC and the age at which treatment is commenced. The shorter the interval between the age of onset of the HARC and age at the commencement of treatment then the better the prognosis. This raises the importance of regular professional eyecare for preschool children, especially if a strabismus is suspected.

Sensory function in HARC In HARC, a point in the peripheral retina of the strabismic eye is said to acquire, during everyday binocular viewing, the same visual direction as the fovea of the fixating eye; this point is directed towards the object of regard and is sometimes referred to as the *zero point*. The zero point has also been referred to as the pseudofovea but this can be confusing since 'pseudofovea' is also used to describe the eccentrically fixating area in eccentric fixation. When the good eye is occluded, the patient fixates with the eccentrically fixating point or, if there is no eccentric fixation, with the fovea and this is why the cover test works (for an exception, see Ch. 16).

The issue of the exceptionally small receptive field sizes at the fovea was mentioned above and this makes HARC difficult in two regions of the strabismic visual field. These areas are the fovea and the zero point. If HARC is not possible in these two areas then the alternative is suppression, and suppression at these two areas is a very common finding in the strabismic eye. These small suppression areas that occur in the presence of HARC are quite different from the complete suppression of the binocular field of the strabismic eye that occurs as an alternative to HARC. The central suppression areas in HARC are of the order of 1° (Mallett 1988a) and often cause, in the Bagolini lens test, the central part of the streak to be absent (Ch. 14). The central suppression areas are also why the modified (large) **OXO** test has to be used instead of the smaller unit to assess HARC (Ch. 14). The cortical task of 'remapping' will be increasingly difficult as the angle of the strabismus increases because larger peripheral receptive fields will have to be 'remapped' to anomalously correspond with smaller central receptive fields in the other eye. Therefore, if all other factors are constant, it seems likely that with larger angles of strabismus the suppression areas will be larger and pseudostereopsis and pseudomotor fusion will generally be worse.

The purpose of HARC is to compensate for the strabismus: to provide 'pseudobinocular vision'. The ultimate goal of binocularity is stereopsis, and some pseudostereoacuity is possible with HARC (Mallett 1977). This is more likely to be present and to be better with deeply ingrained HARC, particularly with small-angle strabismus (Henson & Williams 1980). Stereoacuity can be better than 100″ with the Howard–Dolman or Titmus circles tests (Jennings 1985), which measure contoured stereopsis, but it has been argued that random dot stereopsis cannot be demonstrated in a patient with strabismus (Cooper & Feldman 1978, Hatch & Laudon 1993), and Jennings (personal communication) has argued that stereoacuity would not be expected to be possible in large-angle strabismus, even in the presence of HARC. Rutstein & Eskridge (1984) argued that HARC may actually be detrimental to stereopsis and that some patients with small-angle strabismus have demonstrable stereopsis with random dot tests, which is indicative of normal correspondence. Yet another view is that stereopsis is not possible in any form of constant strabismus, even microtropia, and that findings to the contrary are attributable to monocular cues in stereotests (Cooper 1979).

The locus of the horopter and anomalous fusional space in HARC is much larger than in normal binocular vision (Jennings 1985).

Motor function in HARC The objective angle of strabismus is the angle of manifest deviation, measured objectively, for example by observing the eye movements during the cover test. The subjective angle is the angle of strabismus as perceived by the patient, from any diplopia they may have.

Table 12.1 Example of calculation of angle of anomaly in HARC and UARC

Angle	*HARC: habitual angle*	*HARC: total angle*	*UARC*
Objective angle	15 △ R SOT	20 △ R SOT	40 △ R SOT
Subjective angle	0	5	25
Angle of anomaly	15	15	15

In cases of NRC the objective angle will equal the subjective angle. In HARC patients will have single vision, so that their subjective angle is zero. The angle of anomaly is equal to the difference between the subjective and objective angles. So in HARC the angle of anomaly is equal to the objective angle: the HARC successfully corrects the full subjective angle of strabismus.

The objective angle normally obtained by the patient under undisturbed conditions is called the *habitual angle of strabismus* and the objective angle following prolonged or repeated dissociation is termed the *total angle of strabismus*. As the habitual angle changes to the total angle the angle of anomaly usually remains constant: the difference between the new total objective and subjective angles is the same as that between the habitual objective and subjective angles (Table 12.1, first three columns). The fact that the total angle is reduced to the habitual angle during everyday viewing implies that the HARC may induce some motor fusion to maintain the habitual angle. Indeed, vergence movements can occur in HARC and the patient can be seen to 'converge' to follow an approaching target, yet a cover test will reveal that the strabismus is present. Similarly, 'pseudo' fusional reserves can often be measured.

Unharmonious anomalous retinal correspondence The obvious alternative to HARC is NRC with diplopia or suppression of the binocular field of the strabismic eye. Another option, unharmonious anomalous retinal correspondence (UARC), is exceedingly rare and is best understood with an example. Imagine a young child who develops a small, stable strabismus and associated HARC. The purpose of the HARC is to prevent diplopia and confusion and to allow some rudimentary degree of binocular vision in the presence of strabismus. As mentioned above, the angle of anomaly will be equal to the objective angle of strabismus. Now, assume that after many years in his adapted state the patient suffers, for example, trauma and an extraocular muscle paresis resulting in a change in the angle of the strabismus, with consequent diplopia. If the HARC was shallow then the patient would revert to NRC. In this case the subjective angle (angle of diplopia) would be equal to the new objective angle and the angle of anomaly would be zero.

However, if the HARC associated with the old strabismus was very deep then the patient might continue with this HARC in the presence of the new strabismus. It is unlikely that a long-standing stable HARC could covary with a new change in the angle of the strabismus. Instead, the patient might develop 'a strabismus on top of a strabismus'. The objective angle would be the angle of the new strabismus, the subjective angle would be the difference between the angle of the old strabismus and the new strabismus, and the angle of anomaly would be neither zero nor equal to either of the subjective angles (Table 12.1).

It will be appreciated that this sequence of events is extremely unlikely (although UARC can also occur secondary to surgery), so why is UARC given such prominence in some textbooks? The reason is that many early methods of investigating retinal correspondence created very artificial conditions that tended to cause HARC to break down. It was sometimes concluded that these techniques were detecting UARC. Of course, if the patients really had UARC then they would complain of the symptom of constant diplopia. It would not make sense for the visual system to undergo extensive remapping only to leave constant diplopia.

The foregoing description of anomalous retinal correspondence can only be a very brief overview. There are many different theories on the aetiology of this condition and these have been thoroughly reviewed by Jennings (1985). Another detailed description of this condition was given by Nelson (1988b). Chapter 14 includes details of the investigation and treatment of HARC.

Monocular sensory changes in strabismus

There are two other sensory changes that may be present in strabismus, and these are monocular. They occur in the strabismic eye of a patient with unilateral strabismus and remain when the fellow eye is covered. Indeed, the dominant eye needs to be covered to detect and investigate them. They are amblyopia and eccentric fixation. These sensory changes, which occur in strabismus developing at an early age, are more fully described in Chapter 13 but will be introduced here.

Amblyopia

Amblyopia is an impairment of form vision with no obvious organic cause. In strabismus, amblyopia may assist in lessening the effects of confusion but there are other types of amblyopia that do not necessarily accompany strabismus (Ch. 13).

Eccentric fixation

This is a failure of an eye in monocular vision to take up fixation with the fovea. There are several theories, but little consensus, on its aetiology. These theories are discussed in Chapter 13. Usually, there are no accompanying changes to the localization system in the monocular vision of an eccentrically fixating eye (Ch. 13).

Clinical Key Points

- Diplopia, usually accompanied by confusion, is the obvious consequence of strabismus but can be avoided in young patients by suppression or HARC
- The precise mechanism for HARC is unclear, but it allows for 'pseudo-binocular vision', 'pseudobinocular' motor function, and possibly some 'pseudostereopsis'
- The factors favouring HARC are: esotropia, small angle, stable angle, and early onset. These factors also increase the likelihood of the HARC being deep
- UARC is very rare, and its prevalence is exaggerated by artificial test conditions
- HARC and suppression are binocular sensory adaptations: amblyopia and eccentric fixation are monocular sensory changes