Infantile strabismus: what is it? where is it?

Many ophthalmologists who have faced the challenging task of teaching medical students have been confronted by the apparent simplicity of the questions presented in this forum. In our own experience one of the most daunting questions that recurrently surfaces is what is really wrong in the patient with strabismus? The simple answer, of course, is it's not in the muscles, it's somewhere in the brain. Although the precise location of the 'somewhere' has yet to be defined, recent clinical and laboratory studies have begun to focus on several systems within the central nervous system that appear to be important in the genesis of congenital or infantile strabismus.

The provocative and crucial thesis of Keiner asserted that at birth illumination of the temporal retina drives the eyes nasal-wards. Keiner's thesis fundamentally states that esotropia develops as a postnatal consequence of light stimulus on the developing reflex pathways essential for binocularity. Indeed, Keiner went so far as to state that 'all children are born with the potentiality to squint and an almost total dissociation of the two eyes.' This thesis was later developed in more detail in the elegant and thought provoking work of Jampolsky. Jampolsky asserted that 'I offer the hypothesis that very early neonatal visual influences may be responsible for motor misalignment and anomalous motor development. Light stimulus in the premature insufficiency developed eyes (with yet incompletely resolved media diffusers in the vitreous and lens) fulfills the essential overall diffusion/stimulus criterion - the chain of exaggerated monocular and binocular dominances with altered muscle tonus.' Subsequently, a number of investigators have offered data to suggest that maldevelopment of the visual pathways mediating eye movement and motion processing may be the primary deficit responsible for the oculomotor deficits in naturally occurring infantile strabismus.

In both experimental and spontaneous strabismus in animals and strabismus in humans a permanent deficit in pursuit and optokinetic eye movements has been documented. This deficit is seen as a bias favouring stimuli that move in a nasal direction under monocular viewing conditions. This bias is, of course, similar to that observed in normal, healthy infants in the first months of life. These observations have led some authorities to look to the visual cortical areas known as MT and MST (areas involved in visual motion processing) as important in the genesis of infantile strabismus.

It is not to be inferred, however, that the evidence is overwhelmingly in favour of the cortex being the pathological site for congenital strabismus. Indeed, there may be multiple areas of the brain that are altered in the patient with childhood strabismus. For example, there is renewed interest in the vergence system and to what degree abnormality of vergence motor neurons might be important in some forms of strabismus. This is particularly true now that studies in the non-human primate have demonstrated both vergence tonic cells that increase their discharge directly in relation to the angle of convergence as well as a smaller group that increases the rate of discharge with divergence. These findings have renewed our interest in Duane's old proposal that esotropia might come about as the result of 'excessive subcortical convergence tonus opposed by cortical influences.'

In this issue of the journal, Sekiya and co-workers present a series of findings in patients with latent nystagmus that address this area of concern dealing with the genesis of infantile strabismus. Although it is true that latent nystagmus may occur without accompanying strabismus the impact of this current study is primarily in the area of understanding the pathophysiology of infantile strabismus. These authors suggest, based on their study of the pupillary light reflex in patients with latent nystagmus, that the visual sensitivity of the retina on both the nasal and temporal side is higher in these patients than in normal subjects. Moreover, these abnormalities were greatest in the patients with latent nystagmus and esotropia. From the results of this study the authors postulate that abnormal visual sensitivity determined by the pupillary light reaction in latent nystagmus appears to be a primary phenomenon and that the increase in visual sensitivity on the nasal and temporal sides is essential in understanding the pathophysiology of latent nystagmus. Whether or not it is correct that latent nystagmus occurs as a corrective nystagmus in response to esotropia caused by Keiner's reflex is, at the present time, uncertain. Indeed the lower prevalence rates of both latent nystagmus and dissociated vertical deviation in adult patients with long standing untreated infantile strabismus would directly challenge this thesis. Nevertheless, the study of Sekiya et al is important and fundamental to our understanding of both latent nystagmus and congenital strabismus. If their findings are duplicated by other investigators those of us who face the inquisitive medical student may be able soon to articulate a better answer to the question of - strabismus: what is it? where is it?

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What is the use of ocular blood flow measurement?

We may be impressed or otherwise by the nomenclature of methods of haemodynamic measurements of the eye such as ophthalmodynamography, oculo-oscillodynamography, pneumophotophysiography, bidirectional laser Doppler velocimetry (BLDV), or blue field entoptoscopy and be tempted to interpret the importance of the results as inversely proportional to the complexity of the name. The article by Fujio et al, in this issue of the journal, in which BLDV is employed to investigate the effect of panretinal photoagulation on retinal blood flow, provides an opportunity to reconsider the relevance or not of blood flow investigations to the understanding of disease processes and to the formulation of clinical practice.

When any method is employed to investigate physiological properties and particularly where technology is involved it is important to remember two possible areas of contention. Firstly, whether the assumptions have been correctly calculated? Without going into detail on a particular method, there are assumptions, often multiple, which have been made with each method that is used – for example, in this article Fujio et al rightly inform us that an assumption is made, during the calculation of blood flow, that the cross sectional area of a retinal vessel is circular. Such action is unavoidable in most blood flow measurements in clinical investigation because there are large gaps in our knowledge of in vivo ocular vessel dimensions and other circulatory properties. It is nevertheless important for the investigators (as in this article) to provide a means of accessing such information and for the reader to interpret the results accordingly.

This leads us to the second point for consideration – namely, interpretation. There is no technique available which provides direct measurement of blood flow in the eye. Doppler (ultrasound or laser) measures blood cell velocities, 1,2 tonography techniques measure ‘pulsatile blood flow’, 3,4 oculic compression techniques measure blood pressure, 5 videangiography records dye bolus velocities, 6 and blue field entoptoscopy provides white cell velocities. 7 Extrapolation of results to retinal blood flow requires vessel diametric measurements (impossible in other orbital vessels – for example, in the posterior ciliary circulation) which then involve more assumptions and can at best only allow estimation of the actual blood flow. It is, therefore, often difficult to know how to interpret the results of studies – for example, what is the relation of ‘pulsatile blood flow’ to total blood flow? An informed interpretation of the available methods is required, perhaps backed up by the knowledge obtained from studies of animal models (the methodologies of which should be more controllable).

The limitations in techniques of measurement of actual blood flow do not prevent their application in the clinical setting – for example, Fujio et al have suggested that BLDV could be used to predict the response to laser therapy and colour Doppler imaging has been used to detect retinal ischaemia in central retinal vein occlusion. 8 With this aim absolute blood flow measurement is not required, we need only look at the use of fluorescein angiography to see the application of an imperfect measure of blood flow to the clinical setting.

The complexity of the multiple influences on blood flow in the human means that it will be a long time before we fully understand the interaction of these influences in the normal individual, let alone in disease. The fact remains, however, that in a vascular organ such as the eye, where vascular mishap is commonplace, we cannot afford to ignore the information which these methods provide.

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