Childhood Strabismus

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Introduction

Strabismus is a condition in which the two eyes are not parallel but one is deviated with respect to the other, horizontally and/or vertically. The cause of childhood strabismus is generally unknown, although weakness of one or several of the eye muscles may be present as will be shown in the case presentation below. The most common type of strabismus, without any obvious dysfunction of the eye muscles, is called concomitant strabismus, implying that the angle or size of the strabismus is more or less constant, irrespective of the direction of gaze. This is in distinction to strabismus due to muscle weakness, called incomitant strabismus where the angle varies with the direction of gaze.

Manifest concomitant strabismus is quite a common disease, affecting about 2% of the population. In this type of strabismus the eyes are constantly deviated in relation to each other. Latent concomitant strabismus is even more common and seen in more than half the population. In latent strabismus the eyes stay straight most of the time due to activation of the eye muscles over the visual system to keep the eye aligned in binocular single vision.

After the presentation of a case of complicated strabismus, the effects of strabismus on the visual functions will be described, and the connections between strabismus and eye muscle function and eye proprioception explored.

Case report

The following case report exemplifies recent advances in the management of a difficult case of incomitant strabismus. An infant boy presented with horizontal convergent strabismus of a
very large angle, as a part of a so called Möbius syndrome, a congenital disorder, involving weakness of both the outer (lateral) eye muscles and of many of the facial muscles (Fig. 1, left). Ordinary surgery for convergent strabismus failed due to the complete absence of function of both lateral eye muscles. In order to restore some ability to deviate the eyes to the sides and obtain an acceptable eye position, the tendons of the upper and lower vertical eye muscles were transposed to the insertion of the non-functioning lateral eye muscles. A weakening was also done of the inner (medial) horizontal eye muscles of both eyes by injection of Botulinum toxin into the medial muscles. This combination of surgical and pharmacological procedures resulted in fairly straight eyes (Fig. 1, right), a limited range of horizontal movements and almost normal vertical eye motility. Vision was normal in both eyes due to alternate fixation and well performed occlusion therapy during the first years of life (see Chapter I). Binocular vision was abnormal as a result of the early onset strabismus.

**Figure 1.** (Left) Boy 18 months old with Möbius’ syndrome, prominent convergent strabismus and slight vertical strabismus. (Right) Same boy at 9 years of age after strabismus surgery supplemented with Botulinum toxin. Much improved appearance, although some vertical strabismus remains.
Visual functions

In manifest strabismus where one of the eyes is constantly out of line, the child will experience double vision. In childhood the visual system is very flexible and double vision may be quite easily compensated and shifted into single vision by suppressing the image of one eye at a time. However, suppression may impair the visual development of the eye that is out of line most of the time, i.e. the image of the object fixated is falling outside the fovea centralis. In order to prevent reduced vision of the deviated eye treatment is instituted. It consists of occlusion of the better eye with a patch for a period each day. In most cases glasses are also prescribed, producing sharp images on the retina of the two eyes and supplementing the occlusion therapy. The therapy has to be started at an early age, and continued and supervised up to about age 10-12, during the so called plastic period of visual development (see also Chapter I). Suppression leads to disrupted binocular vision, the main part being loss of stereopsis, but this is a visual handicap of very limited consequences for the individual with strabismus. Reduced vision due to strabismus is uncommon in our country as a result of the extensive scheme for visual screening of young children and early treatment of children with impaired vision in one or both eyes (see also Chapter VI). Thus, we now know very much of the visual dysfunctions that are a result of manifest strabismus and we have reliable means to detect and treat them.

Motor functions

Some types of convergent strabismus are connected with refractive errors of the child’s eye, the most common being hyperopia (far-sightedness). In order to see clearly without glasses, the hyperopic eye has to accommodate and the motor activity of the ciliary (focusing) muscle inside the eye produces an impulse to converge the eyes, which may result in a manifest convergent strabismus. This type of strabismus may be cured by proper glasses to compensate the hyperopia.

In concomitant strabismus there exist only small abnormalities of eye muscle function. The eye muscles that move the eyes and control the gaze positions are known to be very fast and strong with respect to their size, and also very fatigue resistant. Fast movements are used in repositioning the eyes and delicate but long-lasting muscle activity is needed in steady fixation with both eyes directed to the same point in space. The muscle components for steady fixation have been shown to develop earlier than those for swift eye movements, showing that
steady control of the eye position is needed for proper development of visual acuity and binocular functions. Function of the muscle components can be adjusted in proportion to visual activity, and in strabismic eye muscles the components for steady control do not develop to a full extent when the need for steady control in binocular vision is reduced. In some of the divergent types of strabismus the eyes have to converge constantly in order to overcome an inherent outward deviation and subsequently the medial eye muscles become stronger in order to keep the eyes straight.

Control of eye position is mediated also by connective tissue structures outside the eye muscles proper. These structures can control the muscle tendon directions and therefore influence the actions of the muscle on position and movements the eye globe. The connective tissue structures are under influence of the eye muscles themselves, possibly independent of the main eye muscle activity. This would imply that eye position and eye movements are controlled by two muscle systems, one directly acting on the eye globe and the other indirectly by adjusting the sideway forces on the muscle tendon. The control of the connective components around the tendon could be of importance for high precision eye movement activities in e. g. prolonged fixations for near, but the clinical significance of this motor system and how it works in strabismus has to be further investigated.

**Eye muscle proprioception**

The disturbances that strabismus exerts on vision are treated with glasses and occlusion as described in Chapter I. There is as yet no effective treatment for binocular dysfunction. Even if the eyes seem properly aligned by surgery at an early age, and weak binocularity may be restored, the stereoscopic vision will always remain abnormal.

Eye muscle surgery is the common mode of treatment and it is generally performed at an age when the child can cooperate in mea in creating an acceptable eye position and facial appearance, so important for the self-esteem and social interaction of the child. Surgery also often creates conditions for some binocular vision although abnormal.

In addition to surgery, treatment with eye muscle injection of Botulinum toxin A has been recently introduced. Such injection can reduce small angle strabismus without surgery and has been used in childhood strabismus of both manifest and latent types. In may be useful also in the treatment incomitant strabismus as shown in the Case report.
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Conclusions

Concomitant childhood strabismus is a common eye disorder and about 2% of the population is affected by the manifest type and at least 50% by the latent type. The cause of strabismus is most likely a deficiency in the visual system leading to a misalignment of the eyes. Manifest strabismus is accompanied by reduced vision, sometimes manifested as monocularly reduced visual acuity and always as reduced binocular vision. Latent strabismus may cause visual discomfort and headache due to eye muscle fatigue in keeping the eye aligned. The ocular motor dysfunction is considered secondary to the primary visual deficits. In order to gain further understanding of the underlying basic mechanisms of strabismus more studies are needed of the visual and ocular motor development.

References
