THE CONGENITAL STRABISMUS SYNDROME

Joseph Lang - Zurich
Switzerland

Children who squint from birth or from the first months of life often present a characteristic syndrome of dissociated vertical divergence, latent nystagmus, excyclorotation of the nonfixating eye and anomalous head position. This clinical entity may be termed 'congenital strabismus syndrome,' and has been analyzed by Crone (1), Anderson (2), Ciancia (3) and Lang (4). In 1967 Lang (4) presented the following data:

<table>
<thead>
<tr>
<th>Sign</th>
<th>Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dissociated vertical divergence</td>
<td>76</td>
</tr>
<tr>
<td>Latent nystagmus</td>
<td>47</td>
</tr>
<tr>
<td>Excyclorotation of the nonfixating eye</td>
<td>54</td>
</tr>
<tr>
<td>Abnormal head position</td>
<td>58</td>
</tr>
<tr>
<td>A-pattern 20%, V-pattern 17%, cerebral damage 15%</td>
<td></td>
</tr>
</tbody>
</table>

Although the percentage of these characteristics may vary, the symptomatology of the syndrome has been confirmed.

There seems to be some confusion regarding the terms 'infantile' and 'congenital.' Infancy refers to the first year of life, and the newborn period covers the first three weeks of life. In a strict sense congenital means present at birth. This designation may be possible when obvious anatomical malformations are involved, but defects of functional systems that are not fully developed at birth can be diagnosed only when the system is completely operative. However, pendular nystagmus, which is often not
apparent at birth, but appears at age three months, is termed 'congenital nystagmus' in the literature. Early onset strabismus is also usually not seen at birth; parents report that they noticed the squint 'from the beginning, when the child began to look.' Since the visual system continues to undergo development during the first four to six months of life, it would probably be simpler to use the broader definition of 'congenital,' as proposed by Costenbader (5,6) and replace the term 'infantile strabismus' with 'congenital strabismus.'

What is the pathogenesis of the congenital squint syndrome? Let us look at clinical findings and neuroanatomical facts and try to form a hypothesis regarding convergent squint and latent nystagmus (7,8).

Latent nystagmus is highly correlated to strabismus. Of 198 patients with latent nystagmus reported by Lang (9), 99% also had strabismus, whereas only 30% of 93 patients with nonlatent nystagmus had strabismus.

In infants with congenital strabismus, both eyes are in a convergent position at first. Upon ophthalmoscopical examination of fixation, the fixation star does not lie in the foveola, but on the nasal side of the fovea. One gets the clinical impression that only when fixation has moved to the foveola can latent nystagmus manifest itself. The fixation object in the nonoccluded eye slowly moves from the foveola to the nasal side and by a saccade returns to the foveola. Imbalance of the optokinetic nystagmus exists, indicating that adducting nystagmus cannot be elicited.
Finally one has to bear in mind that latent nystagmus is a typical strabological entity. There is no neurological disease producing latent nystagmus, not even multiple sclerosis, which otherwise can mimic everything.

Concerning the neuroanatomical facts, one must bear in mind that other visual pathways exist besides the retino-geniculo-striate system. The most important of these is the extrageniculo-striate pathway to the colliculus superior and from there to the pulvinar and the cortex, as shown in a sketch by Trevarthen and Sperry (10). This second visual pathway is claimed to be responsible for ambient vision.

Fig. 1: The two visual pathways. Black arrows: retino-geniculo-striate system. White arrows: extra-geniculo-striate system, after Trevarthen and Sperry (10), modified by Lang (4).

Furthermore, in the early stages of human foetal development, the primitive phylogenetic arrangement of complete decussation of the nerve fibres occurs in the chiasma. It is not until the 11th week that uncrossed fibres begin to appear. Bernheimer (11) has
demonstrated that 20-30% of the fibres of the optic tractus do not reach the corpus geniculatum laterale, but pass to the pre-tectal and tectal areas. According to Minkowsky (12) these are probably only crossed fibres that stem from the nasal halves of the retina.

Keeping in mind that congenital strabismus is often seen in cases of prematurity or cerebral damage, we may expect that in these cases the phylogenetically older extra-geniculo-striate system, subserving ambient vision, is dominant. This accounts for the convergent position of the eyes and for the tendency to fixate with a nasal part of the retina. As visual development proceeds in the first months of life, fixation slowly moves to the foveola to fulfil the task of discrimination of forms.

When both eyes are open, no nystagmus is seen, since opposite movements of the eyes are counterbalanced. When one eye is occluded, this balance is interrupted. Fixation then slowly glides from the foveola to the nasal part of the retina due to the tendency to ambient vision and then quickly returns to the foveola, bringing the object back to the area responsible for form discrimination.

We now have an explanation for latent nystagmus and congenital strabismus; however, dissociated vertical divergence is still an enigma. I would therefore like to present a new hypothesis, again based on clinical observations and on neuroanatomical facts.

Dissociated vertical divergence can be shown by occlusion of one eye: the occluded eye moves slowly upward and, when occlusion is abandoned, returns slowly downward. As shown by Bielschowsky
(13), when the fixating eye is darkened, the other eye slowly moves downward without reaching fixation. Dissociated vertical divergence may sometimes also be observed spontaneously when the patient is daydreaming or when he tries to read the optotypes. Impairment of binocular vision seems to be mandatory for dissociated vertical divergence, and Herings law is not valid.

All these movements are not saccadic, but very slow and tonic. They are influenced by difference in light in the two eyes. This phenomenon is of innervational origin and must not be confused with elevation in adduction as seen in V-patterns.

Dissociated vertical divergence can be found in a high percentage of congenital squint syndrome cases. All authors agree that it is not present in the first year of life, but appears in the second or third year. It is present in acquired unilateral blindness or aphakia, where binocularity has been lost.

Neurophysiology again brings us to the older extra-geniculo-striate system, in which light fibres subserving the pupillary mechanism do not reach the corpus geniculatum laterale but go immediately to the praetectum. It is also known that pupillary reactions are not consensual in lower vertebrates with total decussation. Light imbalance in both eyes then may induce an imbalance of tonus in the two oculomotor nuclei.

Warwick (14) has shown that in the oculomotor nucleus there is a crossed innervation to the superior rectus muscle and an uncrossed innervation to the medial and inferior rectus and the inferior oblique muscles. In cases of nuclear lesion the superior rectus muscle of the contralateral side and the medial and
inferior rectus and the inferior oblique muscles of the ipsilateral side are involved.

![Diagram of oculomotor nucleus organization](image)

**Fig. 2:** Oculomotor nucleus organisation (Warwick) and DVD.

When one oculomotor nucleus has more tonus than the other, slow sursumduction of the nonfixating eye is elicited. This movement is not controlled by the visual cortex or by brain stem centres, such as the medial longitudinal fasciculus or the mesencephalic reticular formation.

I am fully aware that both hypotheses are based on clinical and neuroanatomical findings which need further support. However, since there have been many recent reports on the geniculostriate system, I find it worthwhile to draw attention to the second visual system almost forgotten by strabologists.

The new hypotheses are unpretentious. They do not compel us to a special form of orthoptic or surgical treatment, but they may explain why even by most sophisticated technics we cannot reach a real cure in cases of congenital strabismus.
REFERENCES


