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## The congenital strabismus syndrome

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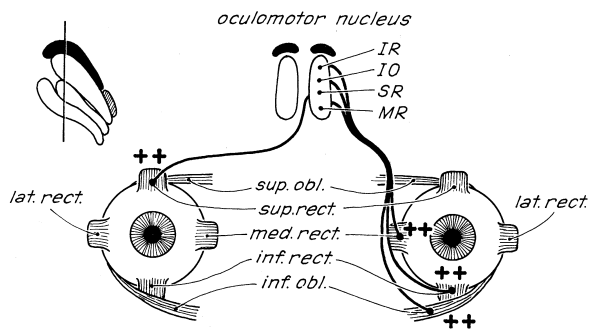
**Background** Years ago, I wanted to find out if strabismus at birth or starting in the first months of life presented a characteristic picture. At the first International Congress of Orthoptists in London in 1967, I reported on 82 patients squinting from birth or within the first four months of life.<sup>1</sup> In 92% of the cases there was a dissociated vertical deviation (DVD), in 57% a latent nystagmus (LN), in 65% an excyclo-torsion of the non-fixating eye and in 70% an abnormal head posture. There were 20% with an A-pattern, 17% with a V-pattern, 17% with cerebral damage, 95% were convergent. The four cases with primary divergence had cerebral damage. The percentage of the signs may change; later on, by ophthalmoscopy, the incidence of LN was much higher. I found this picture again and again and named it the *Congenital Strabismus Syndrome*, since it fulfilled the condition of a clinical entity with seemingly independent symptoms or signs.

This condition is also called “essential infantile strabismus”. Still, I prefer to use the term “congenital strabismus syndrome”, although it is often not present at birth, but may appear in the first four months of life or, as the parents say, ‘when the child began to look’. Birth in humans is actually a premature event in relation to the visual system. Birth does not take place when the visual system is mature, but before the head circumference of the neonate becomes too large to leave the womb. Nowadays, everybody agrees that binocular vision develops during the first four months of life. In a strict sense, ‘congenital’ means present at birth. This diagnosis may apply to obvious anatomical malformations, but in a functional system which is not fully developed at birth, the defect may show up only when the system is completely operative. Pendular nystagmus, which is often not present at birth but usually appears at the age of three months, is also termed ‘congenital nystagmus’ in the literature. I do not use the term ‘infantile’, which in my view is too vague. Infancy means the first year of life and so may include accommodative esotropia.

What is now the most important feature of this entity? Some, like Crone,<sup>2</sup> believe it is DVD. Thus, Keiner and Crone wrote that in DVD, the motor impulses from the lower retinal quadrant are deficient. Guyton<sup>3</sup> thinks that DVD is caused by cycloverversion, and Brodsky<sup>4</sup> refers to a righting reflex gone wrong.

**Dissociated vertical deviation** The slow elevation movements

Fig. 1. The oculomotor nucleus after Warwick and DVD, modification by Lang.



of DVD can be elicited by a difference of the incidence of light in the two eyes, as shown by the darkening test of Bielschowsky.<sup>5</sup> The eye that is occluded or darkened makes a slow upward movement. In case of amblyopia, when the fixating eye is darkened, the amblyopic eye makes a slow downward movement. These movements do not follow Hering's law. Bielschowsky believed that DVD was due to alternating or intermittent excitations of both cerebral centers for vertical divergence, but these centers have never been found. At the time of Bielschowsky, the 'Bernheimer Scheme' was valid for the oculomotor nucleus, where the nerves that innervated the eye muscles were uncrossed. It was not until 1953 that Warwick<sup>6</sup> was able to show that the paired oculomotor nucleus has crossed motoneurons for the superior rectus muscle and uncrossed motoneurons for the medial and inferior rectus and the inferior oblique. When one oculomotor nucleus, due to light imbalance, has a higher tonus than the other, a slow sursumduction of the contralateral eye and an infraduction of the ipsilateral eye is induced. This explains the enigma of DVD. Since binocularity is interrupted, this movement is not controlled by the visual cortex or the brain stem centers for vision. In 1985, I first made mention of this hypothesis for DVD based on the Warwick scheme of the oculomotor nucleus.<sup>7</sup>

As Bielschowsky<sup>8</sup> had already pointed out in 1913, DVD can also be acquired. When vision in one eye is impaired at a young age, the impaired eye makes spontaneous vertical movements. Discrete signs of DVD can also be seen in intermittent exotropia.<sup>9</sup> DVD can therefore not be accepted as the main characteristic of the congenital strabismus syndrome. In DVD there is often some cyclovertical instability. Regarding cyclotorsion, it may be useful to know that the eye, when rotating upward, makes an excyclotorsion, whereas when rotating downward and taking up fixation, the eye performs an incyclotorsion. This is in contradistinction to sea-saw nystagmus, where the eye makes an incyclotorsion when rotating upward and an excyclotorsion when rotating downward. This shows the difference between vestibular and visually induced eye movements. These torsion movements are more pronounced in A-patterns than in V-patterns.

The most important sign of the congenital strabismus syndrome seems

to be the LN. I avoid the oxymoron 'manifest/latent' and prefer to speak of it as LN or as nystagmus of the latent type. Kestenbaum called it manifested LN.<sup>10</sup> The term 'nystagmus in abduction' as used by Ciancia<sup>11</sup> may be confused with nystagmus in internuclear ophthalmoplegia.

In contrast to DVD, LN is never acquired. There is no neurological LN, not even in multiple sclerosis, which can otherwise mimic anything. I have found nystagmus of the latent type, be it more or less manifest, to be combined with strabismus in 99% of cases, while strabismus is found in only 30% of other forms of nystagmus.<sup>12</sup> LN is present in the congenital strabismus syndrome, but is not present in acquired forms of strabismus. Therefore, I believe it to be the fundamental sign of the syndrome.

Some authors believe that asymmetry in pursuit and OKN is the cause of LN. But how can you expect symmetry when LN is present and when one eye is covered? As I see it, LN and the congenital strabismus syndrome are due to a defect of fixation. The fixation mechanism is one of the most important visual mechanisms. Without fixation, the pursuit system, the saccadic system and the vergence system would not function correctly. We strabologists routinely check fixation by projecting the small star onto the fundus, be it by direct or indirect ophthalmoscopy. Normal children fixate the star of the ophthalmoscope with the fovea by an almost compulsory reflex at the age of 4-6 months.

In early onset strabismus, I have often seen that in the beginning not just one but both eyes were in a convergent position. The eccentric fixation can be documented by the first and fourth image of Purkinje. Also, I have often found fixation not to be foveal, but eccentric at the nasal side of the fovea in both eyes. This eccentric fixation changes into LN in the first six months of life. The fixation star then seems to drift from the foveola slowly to the nasal part of the retina and then back with a saccade to the foveola (in reality, the foveola drifts to the temporal side and returns with a saccade to the star). When both eyes are open, these movements are somewhat counterbalanced. When one eye is closed, LN becomes manifest.

**Hypothesis regarding latent nystagmus** What, then, does produce a binasal eccentric fixation in the congenital strabismus syndrome? My hypothesis for nasal fixation and a convergent position of the eyes relies on the preponderance of the nasal parts of the retina over the temporal parts, or of the crossed over the uncrossed optic fibers. Phylogenetically, it is known that there is a total crossing of the optic fibers in the optic chiasm in all vertebrates below the mammalia. In placental animals the crossing is incomplete, and the number of uncrossed fibers increases with the degree of frontality of the eyes. In cats and dogs, there are about 1/4 uncrossed fibers, in higher primates about 1/3, and 1/2 in humans only (Duke Elder<sup>13a</sup>).

From the ontogenetic point of view, "it is interesting that in the early stages of human development the primitive phylogenetic arrangement of a complete decussation of the nerve fibers occurs at the chiasm. It is not until the 11th week that uncrossed fibers begin to appear", according to Sakamoto as cited by Duke Elder.<sup>13b</sup>

Unfortunately, there exists no binasal hemianopia which could serve as a model for the congenital strabismus syndrome, but there are quite a few scientific findings and clinical hints supporting my hypothesis.

It is well known that phylogenetically and ontogenetically younger structures are predisposed to defects. Early onset strabismus affects a large number of premature infants or children with cerebral defects. I have seen identical twins where only the one with perinatal asphyxia showed a typical congenital esotropia syndrome, whereas the other never squinted in spite of an uncorrected hyperopia.

Mehdorn<sup>14</sup> and Herzau<sup>15</sup> have reported large nasal scotomata in amblyopia. Tychsen et al.<sup>16</sup> found that in monkeys squinting since birth, there is more metabolic activity in the cortical dominance columns which are driven by the nasal hemiretina. Also, the binocular horizontal connections between the ocular dominance columns were reduced by an average of 50-60% in strabismic as compared to normal monkeys.

In this syndrome, torticollis is mainly a turn of the face, but often a discrete head tilt toward the fixating eye can also be seen. In patients with the syndrome of congenital monophthalmus due to an organic lesion in one eye, there is a typical torticollis, namely a turn of the face toward the side of the fixating eye: the patient prefers to fixate with the adducted eye, since in this position nystagmus is diminished. To treat this torticollis one often has to operate on the better eye.

In contradistinction to this phenomenon, I found almost no LN in amaurotic cases with hypoplasia of one papilla and a deficient afferent pupillary reflex. As one optic nerve has no function, the cortical neurons belonging to the good eye have no competition from the amaurotic eye. Out of 14 cases with unilateral hypoplasia of the papilla, nine did not show LN.

In a boy with a cerebral hemispherectomy at age four after viral infection, this resulted in hemiplegia and hemianopia. A nystagmus with slow drift into the seeing hemiretina and a saccade towards the foveola was seen, like in LN.

Kommerell<sup>17</sup> has objected that if my hypothesis based on the preponderance of the nasal part of the retina were true, one would expect rapid eye movements into the nasal part of the retina rather than slow movements. All I can say is that saccades always bring the fixation to the fovea, or if the fovea is impaired in the direction of the fovea.

We all know that a convergent position of the eyes by itself may not be sufficient to cause a congenital squint syndrome. I have seen six cases of transient sixth nerve palsy after birth. After some months they recovered from their palsy and at the age of six months they reacted, without any doubt, positively to my stereotest.

There is also the example of Duane syndrome, where, in spite of a serious oculomotor defect, normal random dot stereopsis can be found. We should therefore be aware that an esotropia may not be the cause of sensorial changes due to misalignment, but that sensorial conditions may be the cause of congenital strabismus.

**Conclusion** Consequently, the phylogenetic and ontogenetic prevalence of crossed fibers of the nasal retina can explain all of the following: the bilateral eccentric nasal fixation, a position of both eyes in

adduction, latent nystagmus, the asymmetry in the pursuit system and optokinetic nystagmus, and the deficient horizontal connections in the visual cortex. DVD is due to light imbalance in the two parts of the oculomotor nucleus. These two new, simple hypotheses on DVD and LN may contribute to a better understanding and treatment of the congenital strabismus syndrome.

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