SIGNIFICANCE OF CONGENITAL CYCLO-VERTICAL MOTOR DEFECTS OF THE EYES*

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THE fact that vertical disturbances are sometimes present in cases of horizontal strabismus has been early recognized and described. Strabismus surso-adductorius, a condition in which a marked upshoot of the eyes is seen in adduction, was first observed by von Graefe (1855) and accounted for in different ways by him and by others, who considered it as occasioned by an abnormal vestibular stimulation (Ohm, 1916, 1918), by anomalies of the scleral insertion of the internal rectus (Cords, 1922) or by an overaction of the inferior oblique (Graefe, 1875), either primitive (Bielschowsky, 1935) or secondary to an ipsilateral superior oblique or to a contralateral superior rectus insufficiency (Urrets-Zavalía, 1948a). Cases in which a downshoot of the adducted eye occurs, were also, although more rarely, noted by Ohm (1928), who attributed them also to labyrinthine disturbances.

These findings were contemplated as exceptional (de Lapersonne, 1905), or at least as relatively uncommon; Ohm (1928), for instance, detected elevation in adduction in only 10 per cent. and depression in adduction in 1 per cent. of all his cases of congenital strabismus. Vertical defects, however, are much commoner than was previously suspected; yet only recently has sufficient attention been given to the fact that they are the underlying cause of a great number of cases of exotropia or esotropia which were thought to be purely concomitant (Chavasse, 1939; White and Brown, 1939; Malbrán, 1940; Wagman, 1945; Anderson, 1947; Epstein, 1947; Urrets-Zavalía, 1948a, 1948b, 1948c, 1950a, 1950b, 1950c, 1950d, 1952; Spaeth, 1948).

More than 6 years ago, we pointed out the fact that when such is the case, the observation of the position of the eyes in direct elevation and depression is extremely important, since these may give rise to a relative divergence or convergence of the visual axes of a prominent diagnostic value. We stated at the time that although these cases are truly concomitant inasmuch as the lateral component of the deviation does not alter in direct dextroversion and laevoversion, they are utterly incommittant in that the magnitude of that component varies within wide limits in pure elevation and depression (Urrets-Zavalía, 1948c).

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Cases in which, owing to an overaction* of one or both inferior obliques, elevation is seen in adduction, ordinarily exhibit a relative divergence of the visual axes when looking straight up, and some degree of convergence when looking straight down, and this irrespective of whether a convergent or divergent horizontal deviation exists in addition to the vertical imbalance (Fig. 1A; Urrets-Zavalía, 1948a). The only previous superficial reference to this fact was made by Bielschowsky (1939) and recalled by Danis (1948).

Ringland Anderson (1947) noted divergence in elevation in two of his patients; his description, however, does not exactly fit the phenomenon—which he attributed to a paresis of both inferior obliques—as seen in typical cases.

Conversely, in cases in which, owing to a primitive or inhibitional paresis of one or both inferior obliques, depression in adduction prevails, a relative divergence of the visual axes is ordinarily found in depression, as well as a relative convergence in direct elevation (Fig. 1B; Urrets-Zavalía, 1948b).

FIG. 1.—(A) Divergence in elevation, elevation in adduction, and convergence in depression.

(B) Opposite phenomena: convergence in elevation, depression in adduction, and divergence in depression.

The convergence or divergence of the accompanying horizontal deviation depends on the action of secondary factors and is practically unrelated to the character of the vertical imbalance itself.

Although there still is no final answer to the question of the origin of these vertical troubles, on the basis of both embryological and physiological data and of the evidence furnished by clinical and surgical findings, we have

* Which in turn may be due either to a paralysis of the ipsilateral superior oblique, especially when unilateral and acquired, or to an insufficiency of the contralateral superior rectus, if bilateral and congenital.

ascribed the variations undergone by the lateral deviation in elevation and depression to the lack of balance which appears between the abducting and adducting components of the elevators, and between those of the depressors as well, as a result of the faulty contraction of one of these muscles (Urrets-Zavalía, 1948a, 1948b, 1948c, 1950c, 1952).

Let us first consider those cases in which elevation in adduction, divergence in elevation, and convergence in depression are present. In direct sursumversion, the overactive—or even normally contracting—inferior oblique and the primitively or secondarily defective superior rectus will cooperate in carrying the eyes upwards; yet the abducting component of the former

FIG. 2.—Action of individual ocular muscles of left eye (Tschermak-Seysenegg).

shall remain partially unopposed by the adducting component of the latter (as can be inferred from Fig. 2), and as a result the eye will move temporally. On the other hand, in direct deorsumversion, both the inferior rectus and the superior oblique will operate jointly to convey the visual axes downwards, but the abducting component of the latter being overpowered by the adducting component of the former, the eyes will move nasally in the course of their vertical displacement.

Mutatis mutandis the same holds true in cases in which the opposite phenomena of depression in adduction, convergence in elevation, and divergence in depression exist.

Krewson (1944, 1951), presenting statements and figures on the lateral component of the action of the inferior oblique, assigns to this muscle an adducting action whenever the visual axis is situated nasally with respect to a position of 6° of external rotation, and would seem at first glance to disagree with the interpretation proposed by us. However, this is not so, since Krewson's calculations refer to the modifications experienced by that component as the eye is moved on a strictly horizontal plane, but not to those which occur in elevation and depression. Accordingly, one must not deny the possibility of the inferior oblique having an external rotating action in elevation from the contention that an isolated contraction of this muscle

in the primary position brings about an internal, and not an external, displacement, since, as pointed out by van der Hoeve (1922, 1932), the position of the mobile insertion of the ocular muscles with respect to the centre of rotation of the globe, and accordingly, the muscle plane—and the muscular action itself—alters with every movement of the eye. This is why the affected eye becomes exophoric in elevation and esophoric in depression in cases of superior oblique palsy (Roelofs, 1949). On the other hand, if we admit that the inferior oblique possesses an adducting action even in sursumversion, the above explanation of the cause of divergence in elevation would still be valid, since the weakened adducting component of the insufficient superior rectus should allow the eye to reach that position of 6° of external rotation beyond which the inferior oblique acts as an abductor.

In this connection, a further fact deserves attention. Normally, in direct elevation and depression, not only are the obliques and the vertical recti called into play, but the external recti also contract slightly, their abducting pull being cancelled by the combined lateral components of the vertical muscles (Roelofs, 1949). If the result in a given case is insufficient or excessive, the visual axes will thereupon converge or diverge whenever they quit the horizontal plane.

To sum up, two fundamental groups of vertical—and more often than not bilateral and congenital—imbalances, as seen in cases of otherwise concomitant strabismus, may thus be established, according to:

- (i) the vertical changes suffered by the reciprocal position of the eyes as they rotate upon a vertical axis,
- (ii) the variations to which the horizontal relationship of both visual axes are subjected as they move around a transversal line.

As stated before, the presence and sign of the lateral deviation which eventually supervenes when dissociation takes place is immaterial to the intimate nature of the vertical disorders, which are primitive and may exist quite independently of such added disturbances. The fact that these vertical anomalies eventually become less obvious after surgical correction of the horizontal squint, far from proving them secondary to the latter, helps to demonstrate their primary quality, as does the possibility of their becoming more evident under the same circumstances (Urrets-Zavalía, 1952).

In previous publications we have elaborated upon the aetiology of the muscular defects which lead to the above-mentioned disturbances in the vertical field, and we have reached the conclusion that although their essential nature is still obscure—as to the neural or mesodermic character of the lesions—their bilateral quality and early appearance strongly suggest a developmental origin.

We feel that the presence of adhesions between the superior rectus and the superior oblique, on the one hand, and between the lateral rectus and the inferior oblique, on the other, as described recently by Johnson (1950) under the name of “adherence syndrome”, plays a certain part in the causation of quantitative and qualitative abnormalities

in the action of the vertical muscles. Not merely overactions and apparent pareses, but also some puzzling and hitherto unaccountable findings—such as the impressive role assumed by the horizontal action of these muscles, and the fact that the vertical imbalance is sometimes most obvious in extreme latero-version—might be adequately explained in this way. We have looked for and encountered the anomalies under consideration in a few cases; up to the time of writing, however, we are not able to state their frequency in percentages and properly to evaluate their importance.

Congenital vertical motor disorders of this kind have to be distinguished sharply from the so-called purely innervational, dissociated deviations (Bielschowsky's alternating *hyperphoria*), which must constitute an entirely different group in any classification of vertical motor defects (Bielschowsky, 1938; Malbrán, 1940; Urrets-Zavalía, 1948c). However, considerable confusion still exists, in that the former are often mistaken for the latter, especially when, owing to a more or less symmetrical insufficiency of one of the vertical muscles, an alternating *hypertropia* is seen in addition to a manifest lateral deviation; the fact that both types of imbalance may coexist sometimes also renders accurate interpretation difficult.

When the vertical defect is serious enough to preclude bimacular fixation and to prevent the development of normal binocular vision, or when, if early acquired, it represents an excessive burden for the vergence reflexes and causes a dissociation of the eyes, a secondary lateral deviation usually supervenes through the position of the eyes being determined no longer by the demands of bifoveal fixation, but by the action of other factors from which the divergent or convergent character and the degree of that secondary deviation depend. Among them, the type of the defect affecting the vertical muscles is less significant than the disturbing influence of an eventually coexisting ametropia or than the release of convergence from cortical control; this escape phenomenon, by virtue of which convergence becomes an unrestrained force, first mentioned by Chavasse (1939), has been extensively described (Urrets-Zavalía, 1950c, 1952; Adler, 1953).

When, on the contrary, the central fixation reflex is strong enough to overcome the mechanical hindrance and to achieve bifoveal fixation, normal binocular vision develops and the eyes are kept in alignment in the immediate vicinity of the primary position; fusion amplitude, however, is usually small and a tropia may be seen at times even in straightforward gaze. *Orthotropia* is thus attained, however precariously, in spite of the surprisingly large degree of coexisting *heterophoria*.

The normal reflex linkage between convergence and accommodation is in the former case loosened or, more commonly, even broken; a total disjunction of both functions occurs, extinction thus being the end-result of lack of reward. In the latter case this linkage is always preserved to a certain degree. The fact that the added lateral tropia exhibits at times some variation in near gaze depends entirely on the amount of dynamic convergence which is still in force, *i.e.*, on the age at which dissociation takes place, and has nothing to do with the type and magnitude of the vertical disturbance itself.

A superimposition of a cyclo-vertical imbalance and a horizontal deviation is then apt to occur in the cases under consideration. The cyclo-vertical imbalance may become apparent only in certain directions of gaze and is essentially inconstant; the lateral deviation is constant in so far as latero-version is concerned, but becomes extremely variable as soon as the eyes leave the horizontal plane. This partially concomitant character is related to the intervention of a supranuclear mechanism (Adler, 1953) only when the lateral component is convergent, for, if it is divergent, no such mechanism may be alleged to come into play, since the eyes then merely swivel back to their absolute position of rest.

Urist (1951, 1952) described the same abnormalities, which he erroneously considered to be secondary to the horizontal deviation (Urrets-Zavalía, 1952). Brown (1953) has also described these phenomena but without acknowledgement of our previously published investigations and conclusions.

As the collected evidence persistently suggests that most cases of lateral tropia with vertical imbalance are of a totally different nature from those of a purely horizontal character, a scheme of classification of strabismus was devised (Fig. 3), in which the former cases—designated *strabismus of an ophthalmoplegic type** so as to recall their main clinical feature—were

FIG. 3.—Urrets-Zavalía's schema for classifying so-called concomitant strabismus into three main categories, according to the dissociating influences:

- (I) Strabismus of an ophthalmoplegic type.
- (II) Strabismus of an anisaesthetic type.
- (III) Primarily accommodative strabismus.

Intermediate forms resulting from superimposed action of several agents are also represented.

S.F.: secondary factors, which determine the sign and the amount of horizontal deviation which almost invariably occurs as a result of dissociation (convergence vigour, purposive strabismus, etc.).

* The term *type* was chosen in order to avoid the aetiological connotations which the use of words such as *nature* or *character* would involve.

clearly distinguished from the latter, which may be divided into two different groups—*strabismus of an anisaesthetic type* and *primarily accommodative strabismus*—according to their resulting from a marked disparity of the retinal images or from an iso-ametropia. The classification just outlined includes the fundamental forms as well as the intermediate forms due to the superimposed action of several aetiological factors (Urrets-Zavalía, 1950c, 1950d, 1952).

As will be shown in a future publication (Urrets-Zavalía, 1954), certain slight facial malformations seem to be closely associated with both main types of congenital cyclo-vertical deviation. A more or less accentuated hypoplasia of the malar bones, a downward slanting appearance of the palpebral fissures, and an S-shaped contour of the lower lid margin, as seen in abortive forms of mandibulo-facial dysostosis (Franceschetti and Klein, 1949), occur frequently when an overaction of the inferior obliques is present (Fig. 4, and Fig. 5, overleaf).

FIG. 4.—Insufficiency of both superior recti, more noticeable on the right side.

In straightforward gaze (2) only a small degree of left hypertropia is present.

In elevation (1) and in depression (3) definite divergent and convergent deviations become manifest.

In dextroversion (4) marked elevation, and in laevoversion (5) slight elevation of the adducted eye is seen.

On the other hand, a mongoloid obliquity of the lid axes with a certain narrowing of the palpebral openings is seen in the much rarer cases in which an underaction of these muscles exists; the malar bones are then well developed and the lower lid border exhibits a somewhat straightened contour (Figs 6 and 7, overleaf).*

In order to ascertain whether these morphological anomalies are related to deeper architectural changes and whether they can be more properly evaluated on a quantitative basis, some craniometric measurements were made from lateral roentgen-grams of the skull. Only the radiologic sphenoidal angle defined by the nasion, ephippion, and basion,

* As parallax errors would influence strongly such rough estimates of palpebral obliquity as can be made as a routine procedure in clinical practice, the observer's eyes should be kept at a level with the eyes of the patient, and the latter's head maintained in an erect position.

FIG. 5.—(A) Bilateral overaction of inferior oblique.

In straightforward gaze (2), a slight right esotropia is seen, which becomes more obvious in depression (3) and disappears completely in elevation (1). Marked malar hypoplasia, anti-mongoloid obliquity of palpebral openings, S-shaped contour of lid margins.

(B) Marked insufficiency of right superior rectus.

In straightforward gaze (1) right esotropia and hypotropia are seen. In laevoversion (2) the vertical component disappears. In dextroversion (3) the vertical factor becomes still more evident.

No definite changes in the lateral component were recorded in elevation and depression. Some facial abnormalities were noted bilaterally, but were more obvious on the right side, *i.e.*, on the side of the defective superior rectus.

FIG. 6.—Bilateral underaction of inferior oblique muscle.

In straightforward gaze (1) only a small degree of right esotropia was present.

In depression (2) a definite divergence of the visual axes took place.

In elevation (not shown) no significant change in the horizontal deviation could be seen.

In laevoversion (3) and in dextroversion (4) depression of the adducted eye occurred (this was more evident clinically, as demonstrated by the cover test, than in the photographs).

Note normal development of malar bones, mongoloid obliquity of lid axes, and straight palpebral margins.

the nasal angle determined by acanthion, nasion, and basion, and the angle determined by acanthion, nasion, and ehippion (Delmas-Marsalet, 1942; Dyke, 1941) were taken into account.

According to the observations made by Bertolotti (1910, 1912, 1914) in cases of oxycephalia and Crouzon's cranio-facial dysostosis, a more or less marked reduction in these angles should be expected to occur whenever a certain degree of hypoplasia of the facial skeleton exists, as well as a certain increase of the same when the opposite situation

FIG. 7.—Bilateral insufficiency of inferior oblique muscle. In forward gaze (2) a pronounced alternating esotropia was seen, which became exaggerated when looking straight up (1) and diminished almost to nothing when looking straight down (3). Some facial peculiarities were seen, as in the preceding case.

prevails. Although our present experience is limited, we feel that in cases of cheekbone hypoplasia abnormally low values often correspond to the nasal angle and the acanthion-nasion-ephippion angle. However, as we are still unable to assign any statistical significance to the differences encountered, and as the goniometric changes which would result from the recession or the advancement of the acanthion may be exaggerated or counteracted by independent and purely accidental variations in the position of the ephippion and the basion, the importance of the changes encountered must be minimized.

Although the aetiology and pathogenesis of such abnormalities will not be discussed at present, we wish to emphasize that they obviously result from a developmental error and that their occurrence in cases of strabismus in which vertical disturbances can be demonstrated points to the latter as being also of a developmental nature.

In cases of strabismus in which neither divergence nor convergence in elevation or depression exist, and which do not exhibit the aforementioned facial abnormalities, the eventually coexisting vertical disturbances are for the most part secondary to the horizontal deviation.

Summary

In cases of strabismus, in which, owing to an overaction of the inferior oblique muscle, elevation in adduction is present, a conspicuous divergence of the visual axes is often met with in sursumversion, as well as some degree of convergence in direct deorsumversion. The opposite phenomena of convergence in elevation and divergence in depression are seen whenever a downshoot of the eye in internal rotation exists as a result of a defective contraction of the aforementioned muscle.

The incomitance exhibited by the horizontal deviation in pure elevation and depression, as well as the vertical imbalance to which it is due, are thought to be of developmental origin, because of their primitive character and frequent association with slight facial malformations.

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