CONCOMITANT ESOTROPIA OF LATE ONSET* PATHOLOGICAL REPORT OF FOUR CASES IN SIBLINGS

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CONCOMITANT convergent squint usually begins during childhood. Cosse (1899) states that in 90·2 per cent. the onset occurs between birth and the age of 5 years; Priestley Smith (1898) states that 300 out of 347 cases began during the first 5 years; Worth (1939) found that 70 per cent. of uniocular convergent squints began before the age of 4 years and that 88 per cent. of alternating squints began before the fifth year. Lagleyze (1913) believed that the average age at onset of convergent squint was 4 years and 3 months. Malbrán (1953) states that it is rare for a convergent squint to begin after 6 years of age; of 353 cases, 298 appeared between birth and the fifth year. Scobee (1951) found that 93 per cent. began before the fourth year; Keiner (1951) that 576 of 656 convergent squints had begun before the sixth year; Nordlöw (1953) that 92 per cent. of 518 cases had begun before the fourth year.

Thus, the percentage of concomitant convergent squints beginning after the fifth year is small, and we must subtract from them cases of esotropia of paralytic origin that may, in the course of time, have evolved towards concomitance; these belong to an already defined clinical entity and will not be considered here. We shall refer exclusively to non-paralytic concomitant esotropia of late onset, the pathogenesis of which has been variously interpreted by different authors. Three clinical forms have been described:

(1) A type described by Bielschowsky (1922) of late and acute onset in myopes accompanied by spontaneous diplopia. The affection begins as a rule in persons with neuropathic tendencies, as the result of a psychical or physical shock, and diplopia improves or disappears when the patients fix their sight at close range. Similar cases of this type were described by Weber (1947) and by Franceschetti and Moutinho (1951). The latter described two cases: one resulting from an injury to the orbital region, and the other followed a sea voyage made under adverse psychical conditions; both were cured by surgery. Dubois-Poulsen, Benmansour, and Attane (1955) have also presented two cases of this type.

(2) A second type is described by Franceschetti and Bischler (1948). This differs from Type 1 in that it appears in emmetropes or slight hypermetropes. The first case dealt with by these authors was that of a 15-year-old girl who, without apparent cause, suddenly developed concomitant esotropia in all fields of vision, for both near and distance, accompanied by diplopia. Franceschetti and Moutinho

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(1951) described three other similar cases of acute onset at 47, 64, and 9 years of age, which they proposed to name "acute concomitant convergent squint". Franceschetti (1952) presented another case of acute concomitant intermittent squint in a woman aged 40; in this case, however, the first episode had occurred when she was 20 years old, and although other neurological signs were absent, Franceschetti considered multiple sclerosis to be the most probable aetiology.

Meunier (1952) described the case of a boy aged 4 with diplopia due to concomitant convergent squint in all fields of vision. Although he identified this as belonging to the Franceschetti and Bischler group (Type 2) he proposed the name of "concomitant convergent squint with or without manifest diplopia"; the term "acute" does not satisfy him, as this type is not always of acute onset.

This type has rarely been reported in the literature except by Lagleyze (1913), Cords (1930), Terson (1935), Bailliart (1936) and Burian (1950).

(3) A third type of concomitant esotropia of late onset may appear as the result of ocular occlusion. Swan (1947) described four such cases and proposed the name "esotropia following occlusion". Two additional cases were described by Malbrán (1953) and ten by Norbis (1953). Paufigue (1931) and Bielschowsky (1940) also presented isolated cases of esotropia resulting from the occlusion of one eye.

Some authors have included these cases with Type 2, but we believe that, though pathogenically similar, they should be classed separately because of their specific cause.

Present Investigations

We present four cases of concomitant esotropia of late onset in four siblings (Figure):

Case 4.

Case 2.

Case 1.

Case 3.

FIGURE.—Four children of the same parents, aged 6, 10, 12, and 13 yrs, all with concomitant esotropia of late onset.

Case 1, a boy aged 12.—When he was riding on a merry-go-round at the age of 6, his left eye suddenly became convergent with spontaneous diplopia, and this condition has persisted unchanged.

Examination

Visual Acuity: without glasses: right eye 10/10, left eye 3/10; with glasses: right eye (+2.5 D sph., +1 D cyl., axis 10°) 10/10, left eye (-2 D cyl., axis 175°) 6/10.

Screen Test: with and without glasses, both eyes, esotropia 80△, concomitant in all fields of vision.

Normal correspondence on synoptophore and after-image test, but no range of fusion.

Operation.—5-mm. recession medial rectus both eyes. 7-mm. resection lateral rectus left eye.

Screen Test: Esotropia 40△ both eyes.

In spite of the persistence of marked residual esotropia, the patient has no diplopia. On Worth's 4-dot test he suppresses the left eye.

Case 2, a girl aged 10.—At the age of 6 she suddenly began to turn the left eye inwards with spontaneous diplopia, for no apparent reason. She was treated at another clinic with no improvement.

Examination

Visual Acuity: without glasses: right eye 10/10, left eye 1/10; with glasses: right eye (+3.25 D sph.) 10/10, left eye (+1.25 D sph., +2.5 D cyl., axis 180°) 1/10.

Screen Test: with and without glasses, both eyes, esotropia 80△, concomitant in all fields of vision.

Normal correspondence on synoptophore and after-image test, but no range of fusion.

Treatment.—Occlusion of the right eye was carried out for 6 weeks and the visual acuity in the left eye improved to 7/10. Esotropia unchanged.

Operation.—5-mm. recession medial rectus and 9-mm. resection lateral rectus left eye.

Screen Test: without glasses: esotropia 20△; with glasses: esotropia 10△.

Two years after operation, the esotropia was 25△ with no diplopia.

Case 3, a girl aged 13.—At the age of 9 she had begun to turn the right eye progressively inwards, with spontaneous diplopia. The strabismus was at first intermittent but gradually became constant.

Examination

Visual Acuity: without glasses: right eye 6/10, left eye 10/10; with glasses: right eye (-0.5 D sph., -0.75 D cyl., axis 130°) 9/10, left eye (-1 D sph., -0.5 D cyl., axis 75°) 10/10.

Screen Test: with and without glasses, both eyes, esotropia 60△, concomitant in all fields of vision.

Normal correspondence on synoptophore and after-image test, but with no range of fusion.

Operation.—5-mm. recession medial rectus both eyes.

Screen Test: esotropia 20△.

No diplopia.

Second Operation.—Marginal myotomy medial rectus and 8-mm. resection lateral rectus both eyes.

Screen Test: esotropia 6△.

Normal correspondence, fusion +9° to -5°.

Case 4, a girl aged 6.—After an attack of otitis media 3 months previously, she suddenly turned the left eye inwards, with diplopia.

Examination

Visual Acuity: without glasses: right eye 9/10, left eye 6/10; with glasses: right eye (+2 D sph., +1 D cyl., axis 180°) 9/10, left eye (+2 D sph., +1 D cyl., axis 180°) 9/10.

Screen Test: without glasses: esotropia 65△; with glasses: esotropia 50△, concomitant in all fields of vision.

Normal correspondence on synoptophore and after-image test, but no range of fusion.

Operation.—5-mm. recession medial rectus and 8-mm. resection lateral rectus left eye.

Screen Test: esotropia 16△ both eyes.

Normal correspondence, but no fusion.

A year later the esotropia increased to 35△, the state of binocular vision remaining the same.

Discussion

In these four siblings late concomitant convergent squint began suddenly, in three when 6 years old, and in the fourth more gradually when 9 years old. Esotropia began without apparent cause in Cases 2 and 3, while riding on a merry-go-round in Case 1, and after otitis media in Case 4. All showed spontaneous diplopia from the onset of the affection; esotropia was marked—over 50^Δ in all of them—and was characterized by variability of the angle of deviation. Visual acuity was good. In Case 2, visual acuity in the squinting eye was only 1/10, but this was rapidly improved by occlusion of the fixing eye. Cases 2 and 4 showed medium hypermetropia, and Cases 1 and 2 a moderate anisometropia. Retinal correspondence was normal in all four, but there was no range of fusion. All the patients had been operated on in other clinics. In three a residual esotropia persisted, which will require further surgery; and Case 3 was cured after two operations.

The age at onset is remarkable in these cases, since the late onset of a non-paralytic convergent squint is rare. Our cases should be included under Type 2 (described by Francéschetti and Bischler, 1948); they are not myopes as in Type 1 (Bielschowsky, 1922), nor was the onset caused by ocular occlusion as in Type 3 (Swan, 1947).

Heredity.—It should not be overlooked that the four patients are siblings, all with a similar strabismus, occurring at about the same age, with similar characteristics as regards vision, refraction, degree of esotropia, and retinal correspondence; this, together with the fact that the father shows an esotropia of 8^Δ with intermittent spontaneous diplopia, suggests that the condition is hereditary.

Many authors state that heredity plays an important role in the genesis of strabismus. Böhm (1845) drew attention to the frequent presence of squints in members of the same family. The coincidence of squints among siblings is too frequent to be disregarded: Pugh (1934, 1936) speaks of an “imitative squint”; Parinaud (1899) quotes a family of ten squinting children; Harms (1938) mentions four, and Lagleyze (1913) six siblings with divergent squint, a type in which the hereditary factor has been less frequently mentioned; Worth (1929) found that 51 per cent. of patients with convergent squints had family antecedents with a similar condition, Cords (1930) 10 per cent., and Darr (1945) 50 per cent. Keiner (1951) states that heredity plays an important role in 50 per cent. of squints appearing during the first year of life, and may also affect squints of later onset. Schlossman and Priestley (1952) reported that 47 per cent. of 158 cases came from families with a history of strabismus. Czellitzer (1922) found strabismus among siblings in 15 per cent. of 365 cases compared with 1 to 2 per cent. in the population as a whole. Heredity is thus undoubtedly important, but as Harms (1938) has said, the aetiological factors in strabismus are so numerous that frequency and importance of each of them needs to be determined.

Restricting ourselves to Malbrán's classification of the pathogenesis of squint, we find that there are both general and ocular aetiological factors which may be inherited:

(1) *General*.—These include neurological and psychiatric troubles. Javal (1896) insists on the frequency of neuropsychiatric affections within families, and other authors have stated that strabismus may co-exist with mental deficiency, idiocy, or other degenerative processes (Poulard, 1923). Pugh (1934, 1936), dealing with "imitative squints", found 28 per cent. of difficult children and 29 per cent. of psychopaths and neurological patients.

(2) *Ocular*.—First among the "motor alterations" are anomalies of the position of rest. Troubles of the "mechanism of coordination" comprise mainly weakness or absence of fusion. Other factors, in certain cases, include ametropia and amblyopia.

The anomalies of the position of rest are, according to Bielschowsky (1940) and Malbrán (1953), fundamental in establishing a permanent strabismus. Clausen (1922) and Bauer (1922) accept heterophoria, anomalies of refraction, fusion defects, and unilateral deficient visual acuity as inheritable factors; this opinion was also held by Lagleyze (1913) and von Graefe (1854, 1857). Harms (1938) considers that heterophoria and weakness of fusion (not excluding amblyopia) are apt to be inherited. Waardenburg (1954), studying heredity in strabismus, states that what is dominantly inherited is the anomaly of the position of rest (heterophoria), and that the true squint (heterotropia) is usually due solely to that anomaly, to which may be added certain predisposing secondary factors, such as hypermetropia and fusion defects.

Harms (1938) thinks that amblyopia is not hereditary, since it is not always constant in families or even in twins with strabismus. Waardenburg (1954) states that there is no evidence that amblyopia is inherited together with strabismus, and that amblyopia is not responsible for strabismus, since it may be prevented by the early treatment of the squint; he therefore concludes that sensorial alterations are always secondary to the squint. Finally, Weekers, Daenen, and Hacourt (1955) affirm that amblyopia is a complication of ametropia, or of strabismus, that it is only exceptionally genetic in origin, and that, though it may be acquired, it can be eliminated by re-education, even in squint of early onset. De Jaeger (1955) says that, although it has been proved that heredity plays an important part in the pathogenesis of squint, the same cannot be said of its cure, particularly in respect of amblyopia, since no difference is seen in the improvement of vision by re-education between inherited squints and those in which no factor of heredity is present.

On the other hand, Strazzi (1950), basing his conclusions on the study of two families, believes that amblyopia is often the cause and not the consequence of strabismus, through an inherited incapacity of the brain to receive impressions of images from one eye. Commenting upon this work, Burian (1950) states that Strazzi's idea should not be discarded, although it is not in accordance with the generally accepted theory.

Various authors also differ regarding the form in which strabismus may be inherited. Czellitzer (1922), tracing the genealogical trees of certain cases, proved the recessive character of the hereditary factors which influence the appearance of squint. Harms (1938) thought the origin of strabismus depended on an hereditary, polymorous, recessive complex. Schlossman and Priestley (1952) supported the recessive character of the heredity factor. Franceschetti (1954) accepted that strabismus was inherited preferentially in dominant form, but noted that divergent squints were inherited independently of refractive error, whereas in convergent squints hypermetropia was nearly always present. Waardenburg (1954) stated that the heredity factor was not regularly dominant because of a lack of penetrance of the genes; the motor factor was the main one in heredity, while other factors such as hypermetropia, although frequent and often acting as predisposing causes, were due to accessory genes.

We consider that heredity plays an important part in the genesis of strabismus, since our four siblings were all affected with a similar squint in similar conditions.

In considering the various aetiological factors which (in our cases at least) may have been inherited, the "imitative" factor should be remembered. This may correspond with an anomaly of the position of rest (since the existence of esophoria was proven in the father) and with weakness of fusion. Our cases all had normal correspondence, although their range of fusion was small or nil, just like their father who, with only slight esophoria, showed spontaneous diplopia in certain instances. Only one of the four showed amblyopia and this was soon overcome by occlusion. It appears to us that amblyopia is always an acquired factor, a consequence of strabismus, and that heredity plays no part in its development.

The small clinical differences between our four cases may be explained as individual variations within the main genotype.

Aetiology.—In the pathogenesis of non-paralytic concomitant squint of late onset, the main factors are an anomaly of the position of rest (pre-existing esophoria), with weakness or lack of fusion. To these fundamental factors must be added various secondary provocative factors.

Thus, in Type 1 (Bielschowsky), physical and psychical shock appear to act as the cause of onset. In Type 3 (Swan), the cause of onset is occlusion, since it interrupts fusion. In Swan's cases, hypermetropia was constant, but we believe with him that this is not an indispensable factor; hypermetropia was also absent in several of the cases described by Norbis (1953).

In Type 2 (Franceschetti and Bischler) the fundamental pathogenic factors are the same. Franceschetti (1952) explains the establishment of esotropia by the progressive reduction of fusion or by a shock to the divergence centre.

Meunier (1952) states that the pre-existing esophoria may be caused by a defect of the divergence centre, a reduction in fusion capacity, a transitory muscular paresis, or a spasm of convergence; he could not establish the cause of onset in the case he studied.

Pre-existing esophoria is accepted by the majority of authors; it remains to decide whether this is due to an anomaly in the mechanical position of rest (Bielschowsky, 1940; Malbrán, 1953); to an innervational object similar to the hyperkinesia of the convergence centre described by Adler (1945) and, therefore, of supranuclear origin; or to an anomaly of the harmonic reflex (anomaly of dissociated position). We must also include the hypothesis of Piper (1948), that it is due to liberation of the subcortical centres from the inhibition that governs separately the relative positions of the eyes.

Hypermetropia (although an accessory factor in certain cases, as in those described by Swan) generally has no part in the other clinical types, since the patients are either myopic, emmetropic, or slightly hypermetropic.

Another point on which the majority of authors agree, and one that we have been able to verify, is the existence of an escape reaction with its starting point in the fusion centre to avoid diplopia (the diplopia-phobia of Van der Hoeve, 1932).

Onset.—In considering the cause of onset, we may quote the words of Malbrán (1953):

The majority of squints result from a mixed mechanism: although fundamentally a motor mechanism, the sensorial troubles and those of the coordinating apparatus are important, though only exceptionally sufficient by themselves to carry the eye to a permanent squinting position.

Treatment.—Franceschetti and Moutinho (1951) and Meunier (1952) recommend orthoptic treatment, followed by anaesthesia of the medial rectus, and surgery. They advise proportionally less surgery in relation to the magnitude of the angle. Our four cases had already been operated on by other surgeons; Case 3 was cured by two further operations, and the others require further surgery.

Although this type of squint is rare, knowledge of it is necessary not only for practical purposes but also for the understanding of the pathogenesis of strabismus in general.

Summary

The age at onset of concomitant convergent squint is discussed; it rarely begins after the 5th or 6th year. The clinical types of convergent squints of late onset are listed, and four cases among siblings, in whom the squint appeared after the 5th year, are reported.

The importance of heredity in strabismus is examined. The factors most likely to be inherited are anomalies of the position of rest and defective fusion; these are the most frequently cited in the interpretation of the pathogenesis of concomitant esotropia of late onset, and seem to have been responsible for the defects in the cases described.

REFERENCES

- ADLER, F. H. (1945). *Arch. Ophthalm. (Chicago)*, **33**, 362.
 BAILLIART, P. (1936). *Bull. Soc. Ophtal. Paris*, **48**, 81.
 BAUER, J. (1922). *Abs. in Malbrán (1953)*.

- BIELSCHOWSKY, A. (1922). *Ber. ophthal. Ges. Heidelberg*, 43, 245.
- (1940). "Lectures on Motor Anomalies". Dartmouth College Publications, Hanover, New Hampshire.
- BÖHM, G. (1845). *Abs. in Malbrán* (1953).
- BURIAN, H. M. (1950). "Strabismus Ophthalmic Symposium", ed. J. H. Allen, p. 179. Mosby, St. Louis.
- CLAUSEN, W. (1922). *Münch. med. Wschr.*, 69, 292.
- CORDS, R. (1930). In F. Schieck and A. Brückner, "Kurzes Handbuch der Ophthalmologie", vol. 3, p. 540. Springer, Berlin.
- COSSE, F. (1899). "Le traitement du strabisme". Bordeaux. *Abs. in Malbrán* (1953).
- CZELLITZER, A. (1922). *Klin. Mbl. Augenheilk.*, 69, 519.
- DARR, L. P. (1945). *Amer. J. Ophthal.*, 28, 898.
- DE JAEGER, A. (1955). In *discussion of Weekers, Daenen, and Hacourt* (1955), p. 552.
- DUBOIS-POULSEN, A., BENMANSOUR, B., and MME ATTANE (1955). *Bull. Soc. belge Ophtal.*, No. 109, Fascicule II, p. 60.
- FRANCESCHETTI, A. (1952). *Ophthalmologica (Basel)*, 123, 219.
- (1954). In "Oftalmologia", ed. M. Amsler, A. Brückner, A. Franceschetti, H. Goldmann, and E. B. Streiff, p. 80. Salvat, Barcelona.
- and BISCHLER, V. (1948). *Confin. neurol.*, 8, 380.
- and MOUTINHO, H. (1951). *Bull. Soc. belge Ophtal.*, No. 98, p. 269.
- HARMS, H. (1938). "An Erbleiden des Auges", p. 63. Berlin. "Zeitfragen der Augen", p. 8. Stuttgart. *Abs. in Malbrán* (1953).
- JAVAL, E. (1896). "Manuel du strabisme". Masson, Paris.
- KEINER, G. B. J. (1951). "New Viewpoints on the Origin of Squint". Nijhoff, The Hague.
- LAGLEYZE, P. (1913). "Du strabisme". Rousset, Paris.
- MALBRÁN, J. (1953). "Strabismes et paralysies", trans. G. Sevrin. Editions Héraly, Charleroi.
- MEUNIER, A. (1952). *Bull. Soc. belge Ophtal.*, No. 101, p. 429.
- NORBIS, A. L. (1953). Unpublished observations.
- NORDLÖW, W. (1953). *British Journal of Ophthalmology*, 37, 593.
- PARINAUD, H. (1899). "Le strabisme et son traitement". Doin, Paris.
- PAUFIQUE, L. (1931). *Bull. Soc. Ophtal. Lyon*, 10, 11.
- and HUGONNIER, R. (1953). *Bull. Soc. Ophtal. Fr.*, p. 323.
- PIPER, H. F. (1948). *v. Graefes Arch. Ophthal.*, 148, 555.
- PUGH, M. A. (1934). *British Journal of Ophthalmology*, 18, 446.
- (1936). "Squint Training". Oxford University Press, London.
- SCOBEE, R. G. (1951). *Amer. J. Ophthal.*, 34, 817.
- SCHLOSSMAN, A., and PRIESTLEY, B. S. (1952). *A.M.A. Arch. Ophthal.*, 47, 1.
- SMITH, PRIESTLEY (1898). *Ophthal. Rev.*, 17, 159.
- STRAZZI, A. (1950). *Boll. Oculist.*, 29, 527.
- SWAN, K. C. (1947). *Arch. Ophthal. (Chicago)*, 37, 444.
- TERSON, A. (1935). *Bull. Soc. Ophtal. Paris*, 47, 22.
- VAN DER HOEVE, J. (1932). *Trans. ophthal. Soc. U.K.*, 52, 1.
- VON GRAEFE, A. (1854). *v. Graefes Arch. Ophthal.*, 1, abt. 1, p. 82.
- (1857). *Ibid.*, 3, abt. 1, p. 177.
- WAARDENBURG, P. J. (1954). *Docum. ophthal. (den Haag)*, 7-8, 422.
- WEBER, E. (1947). *Ophthalmologica (Basel)*, 114, 320.
- WEEKERS, R., DAENEN, P., and HACOURT, J. (1955). *Bull. Soc. belge Ophtal.*, No. 108, pp. 547, 552.
- WORTH, C. (1929). "Squint", 6th ed. Baillière, Tindall and Cox, London.