HYPERTELORISM: An Unilateral Case.

BY

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Ocular hypertelorism has been defined by D. M. Greig,¹ who was the first to differentiate this condition, as a congenital cranio-facial deformity produced by abnormal development of the portion of the sphenoid which is laid down in cartilage. This mal-development results in the formation of great wings which are under-sized, while the lesser wings are as big as, or bigger than, the great wings. The facial appearances are characteristic. They consist in wide separation of the orbits, a broad nasal bridge, and external strabismus. The head is brachycephalic with flattening of the occiput and bulging of the temporal regions. Mental deficiency is a feature of the cases, at any rate of the severe ones.

Greig was prepared to admit the possibility of ocular hypertelorism occurring as a unilateral deformity and in an addendum to his paper he writes as follows :—

Perhaps the following may not be irrelevant. In 1890, as a variety of plagiocephaly, Fridolin² described the skull of a boy aged 3 months. It was asymmetrical, high and short, and the right side flatter than the left. The frontal and parietal eminences were absent from the right side, and the upper two-thirds of the right half of the coronal suture were synostosed. In my opinion the partial synostosis and the absence of the right parietal and frontal eminences are related to each other, the synostosis having prevented their development, and, an unwonted strain being thrown on the anterior fontanelle, a large inter-sutural bone has been the result. This part of the deformity is not uncommon, and has no particular relation to hypertelorism, but the other part of the deformity present has; and the presence of two defects in the skull need excite surprise no more than the co-existence or multiplicity of congenital defects elsewhere.

The right orbital cavity is higher and narrower than the left, and the distance between the eyes is in consequence increased. The right orbital index is 155, the left is 84. In the right temporal region the great wing of the sphenoid is much narrower than in the left, the measurements being respectively 3 mm. and 11 mm. The right zygomatic bone is higher than the left, and the zygomatic arch is shorter. The right side of the root of the nose is 54 mm. from the acoustic meatus, the left is 69 mm. from the corresponding point on the left side . . .

It is obvious that synostosis of part of the right side of the coronal suture could not explain all these defects which are entirely unilateral. It is not difficult from what we have seen in hypertelorism to visualize the right side of the skull Fridolin so minutely describes. Further, if one can imagine the deformity as bilateral, the similarity to hypertelorism will be at once recognised and the fault ascribed to the sphenoid, especially that portion of its greater wings which is developed in cartilage. The explanation I have given of hypertelorism makes a unilateral affection not only possible but probable.

Enough has been quoted to show how completely D. M. Greig anticipated the existence of unilateral hypertelorism, and Fridolin's description can be compared with the features of the case of unilateral hypertelorism described in this paper.

DESCRIPTION OF CASE.

Winifred B., aged eight years, is the last child in a family of five. The other four children are of normal appearance. The first is a girl of twenty-three; the next is a boy of twenty-one, said to be the subject of renal trouble; the third (a girl) died of pneumonia; the fourth, a girl of fifteen, is of small stature but otherwise normal. Winifred was born when her mother was forty-two. Although labour was prolonged no instruments were employed. There have been no miscarriages. Both mother and father come of normal families.

Winifred was brought to the Children's Department at King's College Hospital because she held her head on one side, and it was found that she had a torticollis, probably of ocular origin.

Facial Appearance.

The general facial appearance is shown by the photograph (Fig. 1). The upper part of the face on the right side is flattened and the right supra-orbital margin is underdeveloped. The right eyebrow is somewhat higher than the left, and the right palpebral fissure is wider than its fellow. It appears on close examination that the right orbit is displaced outwards and a little upwards. In consequence, the nasal bridge is wide; the distance between the inner canthi is 34 mm. The inter-pupillary measurement could not be taken accurately on account of an alternating strabismus, but an approximate measurement of 60 mm. was made. The distance between the external canthi is 86 mm.

With the object of demonstrating more fully this facial asymmetry the photograph reproduced in Figure 1 was enlarged and two right halves made into one picture (Fig. 2) and two left halves into another (Fig. 3). Figure 2 gives the appearance of separation of the eyes and breadth of the nasal bridge characteristic of hypertelorism, while Figure 3 would pass for the photograph of a normal child.

Description and Measurements of Head.

The presence of hair made accurate head measurements difficult. The maximum horizontal circumference (measuring round the most prominent parts of the glabella and occiput) is 514 mm. ($20\frac{1}{4}$ "'). The greatest length (from the most prominent part of the glabella to the most prominent part of the occiput) is 197 mm. ($7\frac{3}{4}$ "'). The greatest parietal breadth is 168 mm. ($6\frac{5}{8}$ "') but a longer measurement was taken from a point above and in front of the left ear to a point above and behind the right ear ($6\frac{3}{4}$ "'). These measurements give a cephalic index of 85 and the skull is brachycephalic. There is very well marked flattening of the occipital region on the right side but none on the left.

Orbits and Eyes.

That the orbital measurements differ on the two sides is beyond doubt but such accuracy as is obtainable in the macerated skull, is not possible. On the right the orbital height is 34 mm. and the greatest width 33 mm., giving an orbital index of 103. On the left the orbital height is 31 mm., and the width 37 mm. giving an orbital index of 84.

Dr. Whittington, who has examined her eyes, reports, "Compound hypermetropic astigmatism for which she wears glasses. Has alternating concomitant external strabismus and does not fix a near object with both eyes. The right eye tends to diverge most with the screen test and turns a little upwards as well as outwards, but the right eye is the fixing eye of choice under ordinary circumstances. The vision of the left eye is the worse of the two, and the tendency to turn the face to the left and to tilt the head to the left is probably associated with faulty vision and defective muscle balance. Discs normal."

General Condition.

This little girl is a healthy and not unattractive child. Examination of the nervous system and visceral organs yields no abnormality. There is slight webbing of the second and third digits on each foot. There is the slightest suggestion of incurving of the little fingers but neither the hands nor feet are mongolian in character. There is no acrocyanosis.

Mental Condition.

Her mental condition is reported on by her teachers in the following words :

(Infants School) : "She was admitted at the age of five. During her first year her general progress was rather slow owing to timidity, but later she attained a good average degree of proficiency."

(Girls' School) : "She is below average in ability, but she has made great progress in reading and writing. Arithmetic she finds very difficult. She is a quiet little girl but is very interested in all school work."

On examination by the Binet-Simon tests Dr. N. H. M. Burke says of her : "This child is bright and intelligent in manner. She shows failure in tests requiring understanding of imagery and relationships. Observation, attention, and memory are good, and the general intelligence

Fig. 1.—Unilateral hypertelorism.

is well above the average. Her mental age is $9\frac{1}{2}$ years, giving an Intelligence Quotient of 117%." Thus the slight backwardness reported by the school teachers is not confirmed by the Binet-Simon tests.

Radiological Examination.

The radiological appearances are, in our opinion, inconclusive. The two sides of the skull are obviously not symmetrical, and there is definite bulging of the temporal region on the right side. The sella turcica is normal.

DISCUSSION.

In the case of Winifred B., we have an unusual ophthalmological condition to explain, namely, the occurrence of hypermetropia with external strabismus. It is well known that hypermetropia is usually associated with internal

strabismus. For an exception to this rule there are two theoretical explanations forthcoming. First, that a lesion of part of the third nerve nucleus may cause weakness of convergence. This, for example, is what occurs in the post-encephalitic state following encephalitis lethargica. Thus, Dr. T. H. Whittington found that among 100 post-encephalitic children there were eighteen cases of external strabismus and no cases of internal strabismus, although among unselected children internal strabismus is the commoner type of squint. It is reasonable to assume a partial lesion of the third nerve nucleus in these post-encephalitic children. Secondly, that a developmental defect of binocular

Fig. 2.

Two right (hypertelorhic) halves made into one picture. Note the widely separated eyes and width of nasal bridge.

Fig. 3.

Two left (non-hypertelorhic) halves made into one picture.

vision and the fusion centre* may occur. This is the explanation which seems more probable in the case under discussion. If hypertelorism is atavistic it would be expected that, together with the lateral displacement of the eyes, binocular vision would be absent. This, in fact, is the rule in hypertelorism and explains the ocular condition in the present case. Greig describes one of his cases in the following words: "Her eyes did not converge on near objects. Like a hare, she could not see objects directly in front of her so readily or so well as when they were placed laterally or when she turned her head away

* A fusion centre has been postulated by Worth but proof of its existence is wanting.

from them." Binocular vision is a recently acquired characteristic of man. Birds and many quadrupeds have laterally placed eyes. The horse is in an intermediate position; the ape with its arboreal agility is in need of binocular vision—a function which reaches its highest development in man.

Another point in the case here related is interesting. Her hypertelorism is right-sided. Although her right eye is the fixing eye of choice and the one which is more nearly emmetropic, yet it is the right eye which deviates more widely. The turning of her head to the left is an effort to bring objects into line with the right eye, and this explains her torticollis.

Ocular hypertelorism by derivation implies wide-apartness of the eyes (*ὑπέρ*, too much; *τήλε*, apart; *ὑεῖξω*, to separate). To describe a case as "unilateral ocular hypertelorism" is perhaps, somewhat contradictory, but the term "ocular hypertelorism" is established and has such merit that it should be retained.

CONCLUSION.

A case of right-sided (unilateral) ocular hypertelorism is described. External deviation of the fixing (right) eye is probably responsible for an associated torticollis. There is no mental defect.

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REFERENCES.

1. Greig, D. M., *Edin. Med. J.*, Edin., 1924, XXXI, 560.
2. Fridolin, J., *Virchow Archiv.*, Berlin, 1890, CXXII, 528.