CONGENITAL OCULAR MOTOR APRAXIA A FORM OF HORIZONTAL GAZE PALSY*

BY

HOWARD REED AND SYDNEY ISRAELS

Children's Hospital, Winnipeg, Manitoba, Canada

COGAN (1952) has recently described an unusual congenital anomaly and suggested that it should be called "congenital ocular motor apraxia". In this disease all forms of vertical conjugate movements are normal, but lateral conjugate rotations of the eyes show abnormal features. Voluntary lateral gaze from the primary position to look at an object at one side is performed by a movement of head and eyes characteristic of the disorder. The head is jerked towards the object whilst the eyes rotate in the opposite direction. This contrary ocular rotation is presumably due to an uninhibited response to the vestibular reflex. When the head has turned beyond the object and fixation is possible, both the eyes and the head are straightened to look at it. Involuntary lateral movements are normal. Delayed walking, slight defects in coordination, and difficulty in reading also seem to be associated phenomena.

Cogan (1952) described four children with this anomaly, and Alfano (1955) has recorded another patient who may be an example of the same disease although there were a few differences in his case. We here describe the case of a child similar to those reported by Cogan.

Case Report

A Jewish boy aged 5 had shown abnormal eye movements since infancy. These had been recorded accidentally by his parents on an 8-mm. home ciné film (Fig. 1). The peculiar movements had gradually become less frequent but they still occurred particularly when he was frightened or ill at ease. His sense of balance had always been uncertain and, when walking through doorways or turning corners, he was liable to bump into the wall. This was especially apt to happen when his attention was diverted, so that his parents took care not to call him when he was about to enter a doorway or make a turn.

The child sat up at one year, walked at 19 months, and began to talk at about the age of 2½ years. All his "milestones" of development were later than those of his brother and somewhat later than the average. The parents were normal and not related to each other, and had one other elder son who was normal. There were no ocular abnormalities in the family. The pregnancy and birth of the patient were normal.

Examination.—He was a fit boy and no abnormality was detectable. Examination of his eyes revealed no anomaly apart from the unusual movements of head and eyes. His pupillary reactions and ocular fundi were normal. Refraction under cycloplegia showed him to have 2.5 dioptres hypermetropic astigmatism in each eye. The corrected acuity was 20/20 in each eye measured by the rotating E test.

The ocular movements on looking up and down were normal. He was able to follow

* Received for publication March 12, 1956.

FIG. 1.—Sequence of ciné pictures from a film taken by the parents at about the age of one year showing apraxic movements. The baby was looking to the left when his attention was attracted to the front.

- (a) Baby looking left.
- (b) Head rotating to right and eyes rotating to left.
- (c) Head facing front but eyes still looking left.
- (d) Head rotated to right and eyes looking left.
- (e) Head still facing right and eyes now almost looking front.
- (f) Head and eyes looking front.

the movement of a light to each side without difficulty, but when he was asked to look at an object to one side or when an object was suddenly moved into his peripheral field he appeared to toss his head to that side. On most occasions the rotation of the eyes in the direction opposite to the movement of the head was masked by closing the eyelids (Fig. 2), but occasionally the characteristic contrary conjugate ocular rotation was seen. When he was rotated in a swivel chair his eyes moved in a direction opposite to that of his head and body. Convergence was normal.

The skull x rays were negative.

FIG. 2.—Sequence of ciné pictures from authors' film.

- (a) Head and eyes looking left.
- (b) Head beginning to rotate to the front with the eyes rotating to the left.
- (c) Eyelids closed.
- (d) Head and eyes facing front.

Electro-encephalogram.—Dr. Michael G. Saunders of the E.E.G. Laboratory, Winnipeg General Hospital, reported as follows:

Two electro-encephalograms were taken. In both there was evidence of diminished α activity on the left side. Some slow spike forms were also present in the posterior areas on both sides. Photic stimulation produced bilateral, equal, and normal responses in the occipital areas. There

was no evidence to suggest an abnormality in the frontal areas, but some indications of involvement of subcortical systems associated with the occipital areas (Fig. 3).

FIG. 3.—Electro-encephalogram tracings.

Psychology.—Mr. K. G. Howard, of the Child Guidance Clinic of Greater Winnipeg, gave the following psychological report:

The boy was clean, neat, and well-dressed at the time of the interview. He accompanied the examiner to the office without hesitation and held his hand while in the hall. When walking, the boy tended to sway from side to side with each step he took. During the test the boy was responsive and cooperative, appeared reasonably at ease at all times, was able to sustain attention quite well, showed no marked distractibility, and appeared interested in the test items. The Revised Stanford-Binet Intelligence Scale, Form L (1937 revision) was administered.

RESULTS	Chronological Age:	5 years 0 months
	Mental Age:	5 years 4 months
	I.Q.	107

The boy successfully completed all items at the 4-year level and failed only “three commissions” at the 4 years 6 months level. In this test the child is required to do three simple tasks in sequence without confusion or error. At Year 5 all parts were correct except “definitions”. At Year 6 he gained credits on three items and failed in the vocabulary, bed chain, and mutilated pictures items. In the last the subject is shown pictures in which important parts are missing and he is required to name the missing part. When asked to give the examiner different numbers of blocks he was able to respond correctly when asked for three, nine, and seven blocks, but gave the wrong number when asked for five. He was able to discriminate between pictures when asked to pick out one which was different from the others in a group, and was able to complete an elementary maze satisfactorily. He was able to draw a square adequately, but his coordination appeared poor when he attempted to string beads, and he dropped beads on the floor twice when trying to place them on the string. All items were failed at the 7-year level. The I.Q. obtained falls in the average range.

Ciné Recording.—A hundred feet of 16-mm. film was used in an attempt to record the eye movements, but each time the pictures showed the eyes beginning to rotate in the opposite direction and then the closure of the lids (Fig. 2).

The early film (Fig. 1) demonstrated the rotation of the head beyond the desired direction with lagging of the eyes as if they were being dragged round. In the later film there was marked improvement in coordination and the head never overshot the direction of the object of gaze.

Discussion

Kestenbaum (1947) distinguished four types of gaze movement:

- (1) Schematic or command movement, which is entirely voluntary.
- (2) Optically elicited movement, which may be either voluntary or reflex in nature.
- (3) Follow movements, which also may be either reflex or voluntary in type.
- (4) Vestibular movement, which always occurs reflexly.

In our patient, in the cases of Cogan (1953), and possibly in the patient described by Alfano (1955), the first two types of movement were absent and the last two types were present. This corresponds to the acquired type of horizontal gaze palsy classified by Kestenbaum as the Oppenheim type.

In our case the pregnancy and labour were normal. Two of Cogan's cases were normal but the other two had features which may be significant: convulsions and signs of intracranial damage occurred in one infant after a prolonged labour; the mother of the other child was comatose for 12 hours during the second month of pregnancy as a result of carbon monoxide poisoning, and the child was born with cerebral palsy. None of the cases had a family history of any similar condition.

Alfano recorded the case of a boy with a condition which he has termed "spasm of fixation". In this case voluntary rotation to a lateral position of gaze was performed in the following manner:

The eyes were first half closed and then both the eyes and the head were rotated upwards and laterally to the desired position.

This was a first pregnancy and the mother had polyhydramnios. After a precipitate labour no cyanosis occurred but the child was found to have an occipital meningocele or encephalomeningocele, and this was removed at the age of one month. The presence of an occipital meningocele would suggest some atrophy of the underlying occipital cortex. Nevertheless, after a review of the literature and some discussion, Alfano concluded that the lesion was more likely to be due to "damage to the frontal oculogyric centres resulting from haemorrhage from the superior sagittal venous sinus incident to birth trauma".

All four of Cogan's cases were boys, as were Alfano's and the case described above. Whether this is of significance cannot be determined until reports of more cases appear.

Cogan and Adams (1953) reviewed the few reported cases of acquired ocular motor apraxia and described two adults who developed eye movements similar to those of the congenital type. In these acquired cases, the

jerky head movement characteristic of the congenital type was absent and both head and eyes were apraxic. One followed a post-partum thrombosis of the anterior part of the sagittal sinus, and the other was due to a tumour in the right fronto-parietal region. In all these cases, damage to the cortex and subcortical areas of the fronto-parietal region occurred.

Alfano suggested that since voluntary ocular movements, particularly lateral rotations, appear to be initiated in Brodman's Area 8 of the posterior end of the second frontal convolution, this particular defect of ocular movement may be related to a congenital defect of this area. Experimental and clinical findings suggest that in each hemisphere this centre is responsible for movements to the opposite side. Since the defect is bilateral, on theoretical grounds it would seem that there was defective development of Brodman's Area 8 or its immediate connexions on both sides.

In our case, however, the electro-encephalogram indicated a lesion in the posterior region of the brain and Alfano's cases certainly had some damage to the occipital cortex. Moreover, adjacent to the visual cortex is an optomotor area (Brodman 18) which is believed to be associated with the involuntary fixation reflex.

These cases, however, did not exhibit an isolated anomaly. In addition to the defect of lateral gaze, they all had defects of coordination and delayed development. It seems therefore that these children suffered from scattered foci of cortical damage.

Perlstein (1955), Breakey (1955), and other authors have reported a high incidence of gaze palsies in children with cerebral palsy. It is possible, therefore, that congenital ocular motor apraxia is related to certain types of cerebral palsy. Pathological and clinical studies of cerebral palsy cases may elucidate the site of the lesion in this condition during the next few years.

REFERENCES

- ALFANO, J. E. (1955). *Amer. J. Ophthalm.*, **40**, 724.
 BREAKEY, A. S. (1955). *A.M.A. Arch. Ophthalm.*, **53**, 852.
 COGAN, D. G. (1952). *Trans. Amer. Acad. Otolaryng.*, **56**, 853.
 ——— and ADAMS, R. D. (1953). *A.M.A. Arch. Ophthalm.*, **50**, 434.
 KESTENBAUM, A. (1947). "Clinical Methods of Neuro-ophthalmologic Examination", p. 208. Heinemann, London.
 PERLSTEIN, M. A. (1955). *Advanc. Pediat.*, **7**, 231.