

AN OPHTHALMOLOGICAL SURVEY OF A SERIES OF CEREBRAL PALSY CASES

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Cerebral palsy (in this paper denominated CP) has been defined by Phelps¹ as 'a group of conditions which affect the control of the voluntary motor system and which have their origin in lesions of various parts of the brain.' The condition is classified according to the situation of the lesion. Three main groups into which 95% of the cases fall, are recognized:

1. *Spastic*. The lesion is in the cerebral cortex or pyramidal tract.

2. *Athetotic* (dyskinetic—Breakey¹⁰). The lesion is in the basal ganglia.

3. *Ataxic*. The lesion is in the cerebellum.

There are 3 other rare types, namely the rigid, the flaccid and the tremor types.

CP was first described by Little, in his communications of 1853 and 1862. His concept of a natal aetiology of the condition was challenged later by McNutt (1885), Osler (1889), Sigmund Freud (1897), and many investigators in the 20th century (see Anderson²).

Before 1939, treatment was directed mostly to operations performed on the most affected muscle-groups of the limbs but, as a result of the pioneer work of Phelps, Carlson and Perlstein, the line of treatment has swung to treating the child as a whole, with team-work by the parent, teacher, family doctor, medical specialists,

physiotherapist, occupational therapist, etc. Special clinics, hospitals and institutions have been set up, notably in the USA. In South Africa, too, there are several institutions which cater for these cases.

A ETIOLOGY

The condition is for the most part unavoidable and a decrease of its frequency in the near future is not to be expected. Evans^{3, 11} pointed out, however, that if our present knowledge was applied the incidence might be reduced by 1/3rd. The aetiological factors fall into 3 chronological periods (Phelps⁴):

1. *Prenatal*

(a) Congenital normal variations in the size, shape and functions of the brain.

(b) Congenital defective development (interference at any state of pregnancy, independent of heredity or specific prenatal environmental influences).

(c) Pathological prenatal conditions (e.g. vitamin or calcium deficiencies, disturbances of endocrine glands, liver or kidney, etc.)

2. *Natal*

Birth trauma, e.g. prematurity with rapid birth, forceps delivery, anoxaemia, Rh-negative mothers, etc.

(it is impossible to blame "poor obstetrics" for more than 3% of birth-injured CPs.⁴)

3. Postnatal

(a) Convulsions leading to cerebral haemorrhage during the first 3 months of life.

(b) Pertussis under 6 months.

(c) Encephalitis.

(d) Head trauma, e.g. falls.

(e) In adults, cerebral accidents.

INCIDENCE

Phelps⁴ has stated that in every 100,000 of the population 7 CP cases occur. Of these one dies under the age of 6; 2 are feeble-minded and require custodial care; and, of the remaining 4 who are mentally normal, one is severely handicapped, requiring custodial care and education, 2 are moderately affected and can be rehabilitated by treatment, and 1 is so mild that treatment is unnecessary. Phelps estimated therefore that in the USA there is a total of 200,000 cases up to the age of 20; i.e. CP is second to poliomyelitis as a child crippler. Levin, Brightman and Burt⁵ give much higher figures in their survey of Schenectady county.

Asher and Schonell⁶ considered the incidence of CP in the school-going population as 1 in every 1,000.

In South Africa, no estimate can reliably be made. This is partly due to the fact that such various grades of cerebral damage are possible, partly because of the relatively light concentration of doctors in the country areas, where the Native population through ignorance and superstition, economic or spacial reasons, seldom is able or willing to consult a qualified medical practitioner. Medalie⁷ has estimated an incidence of approximately 1,000 white children in the White population of 3,000,000.

Considering the population of South Africa as being over 12 millions, (12,646,375, the latest preliminary census figure) there should therefore be approximately 4,000 CP cases in the country.

OPHTHALMIC INVESTIGATIONS

Very little has been written in the ophthalmic literature on the subject of CP. The main contributors so far have been Guibor^{8,9} and Breakey¹⁰. As I have been fortunate enough to see a number of cases of this disability in my private practice, it was thought that a report might be of interest. These cases were drawn from a local school where school-going girls and boys from about the age of 5½ upwards are treated.

Material and Methods

A total of 73 unselected cases were examined. Some of these have been under my observation for 4 or 5 years. Table I shows them classified according to age, sex and type of case. It will be noted that 63 (or 86%) are

TABLE I. ANALYSIS OF TYPE OF CP ACCORDING TO SEX AND AGE

	5-9		10-14		15-20		Over 20		Total
	M	F	M	F	M	F	M	F	
Spastic	7	6	13	3	21	12	—	1	63
Athetoid	2	—	2	—	2	1	—	—	7
Ataxic	—	1	1	—	—	1	—	—	3
Total	9	7	16	3	23	14	—	1	73

spastic in nature. The sex relationship is approximately 2 males to 1 female (48 : 25); no valid conclusion can be drawn from this, as it may be merely due to the greater availability of accommodation for the male school children. The eye examination consisted of:

1. Visual acuity determination on the Snellan's chart or illiterate 'E' chart at a measured 6 metres; refraction with or without mydriatics (4% homatropine cocaine or atropine 1%) as indicated; subjective correction with lenses.

2. Muscle-balance studies with the cover test, the Maddox wing for near, and the Maddox rod with separate prisms or the Risley rotary prism for distance.

3. General eye examination.

RESULTS

The findings were considered under 3 headings: (1) Refraction, (2) muscle balance, and (3) other associated neural or neuromuscular defects.

1. The Status of the Refraction

As emmetropia, though the ideal optical condition is biologically unusual, it was arbitrarily decided to use the term 'normal' where the patient was ocularly symptom-free and the range of the refractive state lay between +1.00D.sph. and -0.50 D. sph. and between +0.50 D. cyl and -0.50 D. cyl. (axis immaterial). Table II

TABLE II. THE REFRACTIVE STATUS ANALYSED WITH REFERENCE TO THE TYPE OF CEREBRAL PALSY

Classification	Spastic	Athetotic	Ataxic	Total
'Normal'	47	5	2	54
More hypermetropic than +1.00 D. Sph.	1	1	—	2
More myopic than -0.50 D.Sph. ..	8	—	—	8
More than -0.50 D. Cyl. hypermetropic astigmatism	3	1	—	4

It was impossible for various reasons to test the refraction of 5 of the cases

reflects the findings in the various types of CP when classified under the headings of 'normal', more hypermetropic than +1.00 D. sph., more myopic than -0.50 D. sph., and more than 0.50 D. of hypermetropic astigmatism (compound myopic astigmatism was classed as myopia). It is to be noted that 54 (or 80%) can be considered 'normal'. There were 12 cases of amblyopia, mostly associated with strabismus or anisometropia. For various reasons it was impossible to test the refraction of 5 of the more spastic children.

2. Muscle Balance

The patient was considered to have a phoria (once more arbitrarily) if he had more than 4 prism-dioptres

TABLE III. THE PHORIAS AND TROPIAS ANALYSED ACCORDING TO TYPE OF CEREBRAL PALSY

	Spastic	Athetotic	Ataxic	Total
Heterophoria ..	9	3	—	12
Esotropia ..	7	—	1	8
Exotropia ..	3	—	—	3

of exo- or esophoria or 1 prism-dioptre of hyperphoria. The patient was considered to have strabismus if it was either constant or periodic at the time of examination. It was noted that in this series there were 12 cases of heterophoria, 8 cases of convergent strabismus and 3 cases.

TABLE IV. INCIDENCE OF OCULAR DEFECTS WITH CEREBRAL PALSY COMPARISON WITH RESULTS OF PREVIOUS INVESTIGATORS. PERCENTAGES.

	Guibor	Breakey	This Series
Amblyopia	25		16
Esotropia	51	40	11
Exotropia	9	8	4
Horizontal Conjugate Defects	33		
Ptosis	2	—	—
Spastic Lids		1	—
External Ophthalmoplegia ..			1
Nystagmus	9	2	4
Coloboma of Iris		1	—
Congenital Cataract		2	—
Papilloedema		1	—
Optic Atrophy	2	3	4
Previous Choroiditis			1

of divergent strabismus. Table III demonstrates these findings. It can be deduced from these results that there is a greater number of muscle defects in these CP cases than in the general population, a view held by previous investigators.

3. Other Associated Abnormalities

In this series, the following associated abnormalities were noted:

- Optic atrophy—3 cases (4.1%).
- Total external ophthalmoplegia—1 case.
- Nystagmus—3 cases (4.1%).
- Old healed choroiditis—1 case*.
- Anisocoria—1 case.
- Homonymous hemianopia—1 case.

* This case was fully investigated clinically and radiologically (including an examination for toxoplasmosis) with negative results.

Table IV is a comparison of the findings of this series with those of Guibor and Breakey.

DISCUSSION

It has been shown previously that the incidence, particularly of squints, is higher in the CP patients than in the general population. Guibor⁸ concluded that 75% of his CP cases had motor defects. He states that this percentage is probably higher than the statistics usually found. His patients appear to have been mostly seen below 2 years of age. The present results show a lower percentage of abnormalities possibly because (1) the cases belonged to an older age-group than Guibor's, and (2) severe CP cases were not seen. The cases seen could be considered as a selected group because, firstly, all cases were White (Europeans) and, secondly, a requisite for their admissibility into the local schools was that they were 'educable'. In other words they belonged to the rehabilitable 2/7ths of Phelps' classification. It is felt that severer signs would be demonstrated in cases with more severely damaged brains.

SUMMARY

1. A total of 73 cases of cerebral palsy were examined ophthalmologically.
2. A larger percentage of squints and other neuromuscular lesions were demonstrated than would be expected in a similar sample of the general population. This bears out the conclusions of previous investigators.

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