CEREBRAL PALSY IN CHILDHOOD

(THE AETIOLOGY AND CLINICAL ASSESSMENT WITH PARTICULAR REFERENCE TO THE FINDINGS IN BRISTOL)

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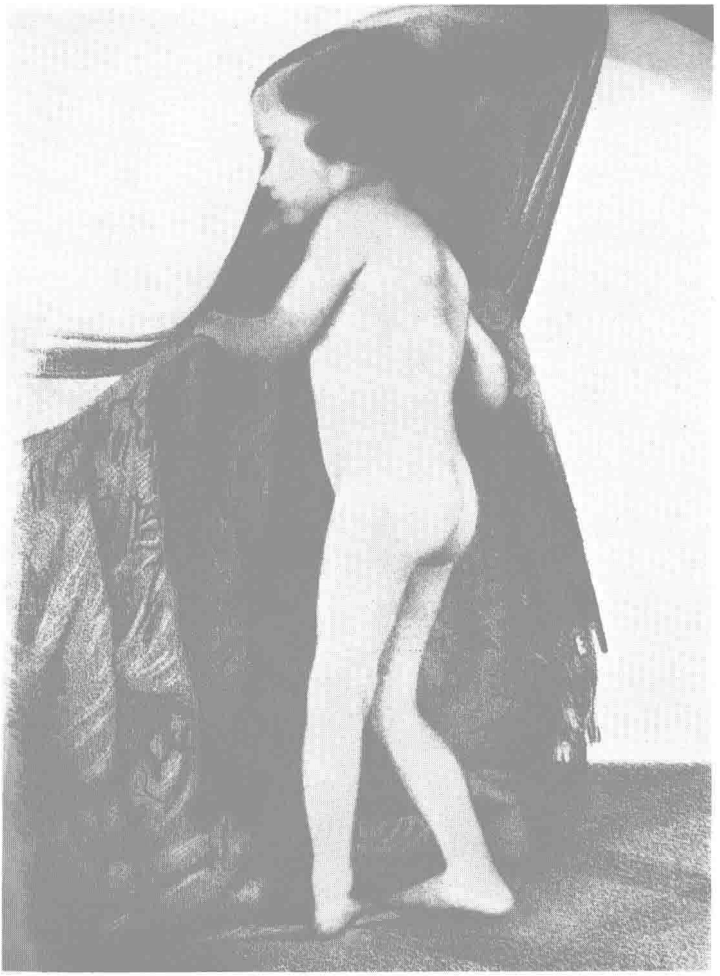

Drawing of a case of spasticity from Little's original article in the *Transactions of the Obstetrical Society, London*, 1862, **3**, 293.

PREFACE

DURING the past few years many articles have appeared in medical journals about various aspects of infantile cerebral palsy. From these articles, it is obvious that this disease is not a single entity, but a collection of clinical conditions which have in common two factors only. Firstly, there is a disease of the brain. Secondly, there is in consequence a defect of movement. This book is an attempt to analyse the aetiology of all the varying cerebral palsies in childhood, which may be antenatal, natal, or postnatal in origin, and to give at the same time a description of the child's other defects, such as deafness or sensory loss, and thus a clinical assessment of the whole child. It is an elaboration of an M.D. Thesis presented to the University of Bristol in March, 1956.

The survey on which these findings are based has been carried out during the last five years in Bristol, and has been assisted by research grants from the United Bristol Hospitals and the National Spastics Society. I would like to express my gratitude to these bodies.

I have also received invaluable assistance from many sources. I have to acknowledge the help and advice given me by Professor A. V. Neale, under whose direction the work has been carried out. I also wish to thank Dr. B. D. Corner and Dr. J. Apley for allowing me to see their cases. Dr. A. L. Smallwood has been of particular assistance in providing details of all the cerebral palsied children known to the Bristol Education Authority. Many cases have been referred by the Somerset Education Authority.

Dr. R. M. Norman has frequently given invaluable advice in discussing the likely pathology of these cases. Dr. R. Walker obtained figures of familial incidence of epilepsy. The electro-encephalographic records have been taken by the staff of the Burden Neurological Institute under the direction of Professor F. L. Golla. Many clinical problems have been discussed with Dr. P. Polani, and problems of deafness with Dr. L. Fisch.

Mr. R. V. Saunders has carried out all the intelligence testing, and has made observations of the child's specific learning difficulties. Miss E. H. L. Duncan has checked the numerical findings and given statistical advice.

Many interesting and exact observations on these children have been made by the various physiotherapists, speech therapists, and occupational therapists, and have been particularly useful. I would especially wish to thank Miss M. Ram and the staff of Claremont School for Spastics, Bristol, for their interest and co-operation.

The photographs were taken by Mr. F. A. Godman and the staff of the Medical Photography Department of the University of Bristol. Miss H. Perren has kindly carried out a large part of the secretarial work.

Finally, I must show my appreciation of the parents who are anxious to further the knowledge of their children's handicap, and have co-operated fully in obtaining records of the children.

Bristol, G. E. W.
November, 1957.

FOREWORD

By Peter Henderson, M.D., D.P.H.
Principal Medical Officer, Ministry of Education

One hundred years ago, Dr. William John Little, a physician at the London Hospital, gave, for the first time, a clear clinical description of cerebral palsy and noted its relationship to prematurity, difficult labour, and asphyxia neonatorum. In the years that have since passed, the infant death-rate has been substantially reduced, the control of some of the diseases that used to cripple many children has tightened, and medical and maternal care have greatly improved. Children born maimed and who once would have died in infancy now survive, and fewer of those born healthy are now deformed by disease in later childhood. The result has been that cerebral palsy now stands high, if not chief, among the conditions causing physical handicap in children. It was found in a survey in Bristol, which is the subject of this book, that the number of cerebral palsied children who reached the age of 5 years was 1·9 per 1000 live births. In recent years cerebral palsy in children has been studied by an increasing number of doctors and educationists and there have been many papers (some would say too many) and a number of books on the subject.

The particular merit of this book by Dr. Grace Woods is that it gives the findings of a team of workers at the Bristol Children's Hospital, under the combined direction of the Professor of Child Health at Bristol University and the Chief School Physician of the Bristol School Health Service, on a group of 301 children. The team included, in addition to Dr. Woods, a school doctor, the chief educational psychologist of the Bristol Child Guidance Clinic, and a health visitor ; the headmistress of the Bristol day special school for children with cerebral palsy was closely associated with it. The children attended the follow-up clinic frequently and those who were

at the special school were under close daily observation : some of the children were known to Dr. Woods for fourteen years.

The investigation, management, and education of children with cerebral palsy require the close co-operation of doctor, therapist, health visitor, educationist, and parent. Although this book is concerned mainly with the causation and the clinical description of the many manifestations of cerebral palsy, it sets an admirable example of co-operation between doctor, nurse, psychologist, and teacher.

This is a first-rate clinical study, undertaken with insight and patience. The different types of defect are described according to the upset of movement pattern. It is stressed that spasticity is not static and may be present in one group of muscles and then in another, depending on the posture and movement of the child : " The cortex knows nothing of muscles, it knows only movements," wrote Hughlings Jackson and this is aptly quoted by Dr. Woods.

It has long been known that deafness is more prevalent among children with cerebral palsy than among the normal child population ; 20 of the 301 children in this study were found to have defective hearing, and of the 32 athetoids 10 were affected. The hearing defect was considered to be of two distinct types : high-frequency deafness and ' auditory agnosia '. It is, however, often difficult to test the hearing of children who are also severely disabled by cerebral palsy and the results of hearing tests have to be interpreted with caution. It may well be that with more experience, and improved testing technique, a diagnosis of ' auditory agnosia ' will be made less often in future. The effect of marked hearing loss on the educational progress of a child who is also heavily handicapped by cerebral palsy has not yet been fully assessed. It can be investigated thoroughly only by persons skilled and experienced in the special methods used in the teaching of deaf children, and of children with severe cerebral palsy.

In addition to deafness, poor intelligence, and physical infirmity affecting adversely the education of cerebral palsied children, a number also appear to have specific perceptual difficulties that impede the process of learning : some, for example, may not be able to tell right from left, or may have

no idea of numbers, or are unable to distinguish between different shapes. There is evidence that these difficulties may arise from damage to the parietal lobes. Dr. Woods is convinced that it is as necessary to deal with these perceptual difficulties as it is to treat the defect of movement. She makes a strong plea for early diagnosis so that treatment can be begun early and the specific learning difficulties tackled in the pre-reading stage " before serious school work begins ".

This book does not discuss methods, or results, of treatment : that is not its purpose. It is, however, concerned with the causes of cerebral palsy. Since many severely affected children, after long years of expensive treatment and education, remain permanently incapacitated at the end of their school days it is of first importance that the causation of the condition should be investigated in the hope that measures can be devised to prevent its onset. Less than half the total number of children who were the subject of this study were born after a normal full-time birth following a normal pregnancy, yet there was little evidence of unskilled midwifery. It seems, therefore, that with present knowledge little further preventive action can be taken. The need for research is pressing since many children are involved and a substantial number, despite early diagnosis and skilled management, remain almost completely dependent on others for the rest of their days.

Much can happen to the developing baby during its nine months of intra-uterine life and there are likely to be many factors that may affect it adversely : their elucidation is one of the toughest tasks in medical research. The case histories of the 301 children investigated by Dr. Woods and her colleagues are recorded in detail and merit close study.

Dr. Woods has written a book that is notable for its wealth of clinical descriptions, revealing case histories, and for a wide review of the writings of others. It is a record of long and painstaking observation that challenges thought. No one, be he doctor, therapist, or teacher, who works with cerebral palsied children should miss reading it.

London,
October, 1957.

ABBREVIATIONS

A.E.G.	- -	Air-encephalogram
A.P.H.	- -	Antepartum hæmorrhage
E.E.G.	- -	Electro-encephalogram
E.S.N.	- -	Educationally subnormal
E.T.A.	- -	Elongation of tendo achillis
I.Q.	- -	Intelligence Quotient
O.S.	- -	Ordinary School
P.H.S.	- -	Physically Handicapped School
Oc.C.	- -	Occupational Centre
b.	- - -	Born
B.B.A.	- -	Born before arrival of doctor or midwife

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CEREBRAL PALSY IN CHILDHOOD

CHAPTER I

INTRODUCTORY

THE subject-matter of this book is based on the findings of cases referred to a Cerebral Palsy Assessment Clinic at the Bristol Royal Hospital for Sick Children. This clinic was run under the combined direction of Professor A. V. Neale, Professor of Child Health in the University of Bristol, and Dr. A. L. Smallwood, Chief School Physician, Bristol Health Services.

The prior purpose of the clinic was to assess the children for education and to place them in the most suitable school or occupation centre. It is likely that all cases between the ages of 5 and 17 years in the Administrative County of Bristol have been seen. Therefore the numbers are an accurate estimate of the proportion of cases of varying aetiology and movement defect in Bristol and possibly representative of the total population. The survey also includes some children under 5 years of age and many from outside the city boundary.

Cerebral palsy in childhood is the result of infantile cerebral palsy. It is, therefore, first necessary to define precisely what is meant by 'infantile cerebral palsy'. Courville (1950) defines it as "a clinical state resulting from some physical insult to the motor system of the developing or immature brain". Perlstein (1949 b) says: "Cerebral palsy is a condition characterized by paralysis, weakness, inco-ordination or any other aberration of motor function due to pathology in the motor control centres of the brain". Thus the term 'infantile cerebral palsy' can be widely used to include all cases where there is an upset of the normal pattern of movement due to a

cerebral cause. The cause may be an antenatal, natal, or postnatal damage or disease of the brain, and, using the term more widely than Courville, includes cases with a familial inheritance like hereditary spastic paraplegia. This wider definition will also include a number of cases which in the past have been crudely described as mentally defective. In many cases of mental deficiency there is a generalized rigidity, either mild or severe, which upsets normal movement. As will be shown later, some of these cases have a history of antenatal abnormality or obstetrical difficulty suggesting brain damage, and they can justifiably be included under the heading of cerebral palsy.

The term ' infantile cerebral palsy ' includes a wide variety of clinical conditions, and, as Phelps (1949) states, " Cerebral palsy differs from other handicapping conditions because it includes all functions of the brain. It does not represent only a motor handicap, but because of its origin in the brain may include mental and sensory deviations as well." Other cerebral functions such as sight, hearing, perception, understanding, feeling may also be defective to a different degree in each case, and because of the widespread or particular damage, no two cases are exactly alike.

It is customary when describing cerebral palsy cases to place them under definite headings such as (*a*) paraplegia ; (*b*) monoplegia ; (*c*) hemiplegia ; (*d*) quadriplegia ; (*e*) athetosis ; (*f*) ataxia ; (*g*) rigidity. These terms describe only the upset in movement pattern, and under each heading are cases of different clinical conditions. In fact a case in one group may more closely resemble a case in another group than in its own. It is not always easy to be certain into which group a case should be put ; for example, a severe tension athetoid may at a single examination appear to be a case of spastic quadriplegia. As these cases can only be assessed by a personal appreciation of the movement upset and not, as will be shown later, by set clinical signs, there may be human errors of judgement in placing a case in one group rather than the other. These clinical subdivisions are artificial, and have been formed because the emphasis in cerebral palsy is on the movement upset.

After giving figures of comparative incidence the cases are discussed in chapters under the seven headings given above.

As they represent widely differing clinical conditions, overall statistics of the whole number can be very misleading, i.e., one cannot get a true picture of the significance of obstetric factors if one includes cases which appeared to originate after birth. On the other hand, if cases are analysed in groups which appear to represent one clinical condition, the numbers may be too small for statistical appraisal. The method of statistical analysis has been used guardedly and in many cases only statistical ' hunches ' can be made.

In each chapter a bald numerical statement is made of the findings, and then an attempt is made to show how the cases may be grouped under definite aetiological or clinical headings. As very few cases similar to the ones we are seeing have ever come to post-mortem, much of this work must remain conjectural, and striking similarities may even be coincidences.

The numerical findings of each movement-defect group are then analysed to show the significance of varying factors in the birth process in the causation of cerebral palsy.

Following this, there are four chapters elucidating the other neurological signs and symptoms found in these cases. As already mentioned, the emphasis in cerebral palsy is on defects of movement, but because the condition is due to brain pathology there may be a wide variety of other defects, often of more importance to the child and his future welfare than the movement handicap alone. The varieties of visual defects, speech and hearing defects, loss of sensory discrimination, and epileptiform disturbances are discussed separately.

In a final chapter an attempt is made to show that because of the widespread brain damage there is a disorientation of the child's whole personality which may involve a general lowering of intelligence, behaviour problems, and particular difficulties in perception and conception which affect the child's ability to master the tasks of education.

CHAPTER II

HISTORY AND LITERATURE

No survey of the literature on cerebral palsy can be begun without mentioning the work of Little. His practice as an orthopædic surgeon brought him into contact with cases of severe deformity following untreated cerebral palsy and he gives an account of these cases in his book *On Deformities* published in 1853. Of 7533 patients treated at the Orthopædic Institute, Bloomsbury Square, London, he considered that the deformities in 138 were due to nervous affection or paralysis, and although he suggested various causes for the condition, such as diarrhœa and internal upsets, he noted that a large proportion followed a first pregnancy and many patients were premature (many weighed only 40 oz. at birth). He noticed the frequency of low intelligence, fits, and uncontrollable behaviour, and the value of massage, baths, gymnastics, and manipulations. He describes a case of marked improvement in an athetoid.

In 1862 Little wrote in an obstetric publication on the influence of abnormal parturition, difficult labour, premature birth, and asphyxia neonatorum on the mental and physical condition of the child, especially in relation to deformities. His description was in many ways in agreement with modern findings, and he mentions for instance that laryngismus stridulus may be present in infancy in these cases, a finding which has only recently been reaffirmed (Apley, 1953). Little states: "Treatment based upon physiology and rational therapeutics effects an amelioration surprising to those who have not watched these cases". A drawing from this book is shown in the frontispiece.

One or two earlier writers had described cases of cerebral palsy—Collier (1924) quotes Andey (1741), Delpech (1828), and Heine (1860). Little quotes Joerg (1828) as saying that

" too early and unripe born fœtuses may present a state of weakness and stiffness in the muscles persisting until puberty and later ".

Since Little's day there have been numerous articles dealing with specific sides of the subject. As cerebral palsy is the name given to a wide variety of clinical conditions, all the articles are of importance in understanding the complete nature of the condition and not all can be mentioned in any survey.

In 1895 Brissaud thought that prematurity at birth was the causal factor and considered that the essential pathology in symmetrical spastic cases was a lack of complete development of the pyramidal tract—an interesting theory which may contain some truth.

In 1885 Sarah McNutt wrote a paper on cerebral palsy giving clinical details and post-mortem findings in one case which showed meningeal hæmorrhage from pial vessels and venous sinuses, and suggested that this was the essential pathology.

In 1888 Gowers, in an article in *The Lancet*, was strongly of the opinion that most cases were due to cerebral damage at birth, and headed his article " Birth Palsies ". He states that " mental deficiency is not necessarily allied to the muscular disorder ; nor when it exists is there any necessary proportion between the two ". He says that the condition of birth palsy is not uncommon.

In 1889 Osler wrote on *The Cerebral Palsies of Children* from cases he had observed in hospital and in institutions for the feeble-minded. His group of 151 cases included 120 hemiplegics, and he considered a further 20 to be bilateral hemiplegics. Some of his cases were undoubtedly acquired cerebral palsies and he found a high incidence of feeble-mindedness.

Freud (1897) in a monograph discusses 53 cases of bilateral affection. He considered the aetiology of generalized spasticity to be one-third difficult birth and one-sixth premature birth.

Following these early papers, Collier, at the beginning of the century, had an immense influence on the thought given to problems of cerebral palsy and wrote papers in 1899 and 1924. His cases tended to be severe, and over a period of years